Invisible but exposed

An innovative research lens on congenital bowel disorders

By

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TABLE OF CONTENTS

LIST OF FIGURES

LIST OF TABLES

ABSTRACT

Hirschsprung's disease (HSCR) and/or anorectal malformations (ARM) are two of the most common causes of neonatal bowel obstruction. While improvements in surgical approaches have significantly reduced the mortality rate for these babies over the past fifty years, surgery does not always result in the desired outcome of faecal continence for the child. Since the diagnosis of HSCR/ARM is only possible after the baby is born, the news usually comes as a shock to parents who had no indication their baby had a congenital abnormality, often requiring major surgery within days of their birth. Parents feel isolated, and this is exacerbated if the child continues to have varying levels of faecal incontinence, a state which is often interpreted through the lens of poor parenting or childhood anxiety in schools and among other parents. This doctoral research focuses on the experiences of those families who have a child born with HSCR/ARM-related faecal incontinence, from diagnosis as a neonate to their experiences at school, and some older adults who have contacted the support group I co-founded, the Bowel Group for Kids Incorporated. The thesis is configured through a multi-lens approach to capture critical stakeholders through a child's life, including paediatric surgeons, parents, other children, teachers, and allied health professions.

My significant, original contribution to knowledge is to demonstrate that the medical model of Hirschsprung's disease or anorectal malformation is not sufficient to enable not only the treatment, but the life of these citizens. I draw on my personal experiences as a nurse who cares for these families; as a parent of a child born with Hirschsprung's disease, an educator of future clinicians and

as cofounder of the first Australian parents' peer-to-peer support group for children born with a congenital bowel disorder, founded in 1994. Surveys were distributed to paediatric surgeons, organisers of support groups globally and parents of affected children. Interviews were conducted with parents and teachers, and a review of school policies was conducted to understand how these children were accepted and supported within the school environment. To consolidate this research, it was important to include the stories from the people who have the lived experience caring for these children once they are discharged from hospital following corrective surgery. My analysis offers a revaluation and reconfiguration of Goffman's theory of stigma (1963) as relational, demonstrating how faecal incontinence is frequently positioned by others as a 'spoilage' of the child's identity. I highlight how parents struggle to become 'wise,' in Goffman's sense: normal individuals who are accepted by the stigmatised as people who do not, and cannot, shame them. Through peer-to-peer support groups, and through active advocacy for their children, parents carve out the space of being 'wise' from their children's earliest years, anticipating the stigma that will befall their children. My significant original contribution to knowledge in this doctoral thesis is the ability to draw on the position of a nurse caring for children born with congenital abnormalities, as a mother of a child born with significant disabilities, an educator of future nurses and doctors and as the co-founder of a peer to peer support group to support those affected by congenital bowel disorders. This thesis demonstrates the need for support in all domains of children's lives who have a taboo condition like faecal incontinence, resting on rigorous and compassionate multidisciplinary clinical care, the value of peer-to-

peer support for parents and families from diagnosis and throughout their child's life as needed, and enlightened, non-judgemental educational policy.

DECLARATION

I certify that this thesis does not incorporate without acknowledgment any material previously submitted for a degree or diploma in any university; and that to the best of my knowledge and belief it does not contain any material previously published or written by another person except where due reference is made in the text.

Signed.....Eunice GRIBBIN.

Date.....20 June 2023

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My sincere appreciation goes to Professor Tara Brabazon for your willingness to support me when all else failed. Your innate ability to turn your mind to any subject as though it has been your life's work is exceptional. You have taken me on a journey I could never imagined possible, one that allowed me to write from the heart to bring this taboo subject into focus. I thank Professor Jamie Quinton sincerely for coming on board to support my work with such enthusiasm. Thank you both, your kindness and mentorship, your guidance and belief in me will remain with me always.

I acknowledge the many doctors and nurses who have supported my work and cared for my son Adam through very difficult times, always having his best interests at heart and never giving up. I am grateful to the hundreds of families I have had the great privilege to meet over the past thirty three years, without your shared experiences, this research would not have been possible. Thank you to the colleagues and friends who helped set up the support group which has enabled our collective experiences to support new families and grow awareness to this hidden, but exposed disorder.

To my dearest friends who have stood by me, encouraging and supporting me and keeping me on track through this seemingly endless journey, even when I had serious doubts. Special thanks goes to my gorgeous family, who have supported my quest for education and greater awareness for families affected by congenital abnormalities. My dear husband Michael of over fifty years, you have always encouraged me, even after I drew a smiley face on the calendar marking the end of my final degree, a Masters; only to announce a few weeks later I wanted to enrol in a PhD! You've shrugged it off with disbelief and a smile. You care so beautifully for our son Adam with whom you share a special bond, you're his best mate. Together, we are immensely proud of our three daughters who have exceeded our expectations and grown into the most compassionate, inspiring young women despite of, or perhaps because of their challenging upbringing with a little brother who required more care than we could have ever imagined. Thank you to our wonderful son-in-law who instantly stepped up to become the big brother Adam never had, showering him with unconditional love and acceptance. Our three gorgeous grandchildren adore their Uncle Adam and bring endless joy into our lives.

I am immensely grateful for the opportunities I have been afforded from the hundreds of people who have generously shared their experiences as parents, grandparents, carers or the children and adults born with anorectal malformations themselves. The insight gained has broadened my understanding of the challenges people face in all walks of life and across the world. Supporting new families through to helping older people understand a condition they were born with but had neither comprehended nor discussed the condition with anyone has been particularly rewarding. Historically, information and discussions between families and clinicians was limited and health support over the affected child's lifetime was minimal. It is through the culmination or shared experiences of those who have come before us; the emergence of bowel clinics specific to the

needs of families affected by congenital bowel disorders; the growth of a diverse range of support services online and traditional peer to peer support organisations and through education and open discussion that we have any hope of changing the future to improve the quality of life and inclusiveness for these children and their families.

This has been a journey like no other and would not have been possible without the special people in our lives, and the knowledge and experience gained through the lens of Adam's difficult journey over the past 34 years.

Molte grazie.

You can only know something when you have lived it.

Theresa Plane, aged 89 years, Senior Australian of the Year 2023

GLOSSARY OF TERMS

Aganglionosis	Absence of nerve cells.
Anastomosis	Surgical connection between two structures. It usually means a connection that is created between tubular structures, such as blood vessels or loops of intestine.
Anorectal	Pertaining to the anus and rectum.
Antegrade colonic enemas (ACE)	Surgically created opening into caecum to administer enemas that flow in the natural direction of the colon from the right colon to the left.
Anus	The opening at the lower end of the alimentary canal, through which the faeces are discharged. It opens out from the anal canal and is guarded by two sphincters.
ARM	Anorectal Malformation
Barium enema	Insertion a chalky substance into the rectum which is then viewed by x-ray. It is used to assess gastrointestinal conditions.
Bile	Also called gall, greenish yellow secretion that is produced in the liver and passed to the gallbladder for concentration, storage, or transport into the first region of the small intestine, the duodenum. Its function is to aid in the digestion of fats in the duodenum.
Bilious vomiting	Green vomit. Assumed to be caused by a mechanical obstruction until proven otherwise.
Bowel	Part of the digestive system, which is also called the gastrointestinal (GI) or digestive tract or gut. The digestive system starts at the mouth and ends at the anus.
Bowel Motility	Spontaneous movement (of the bowel).
Caecostomy	Opening into the Caecum (first part of large bowel) usually to place a device or tube to enable washouts to be performed as in antegrade colonic enema treatment.
Chait	Colon cleansing by administration of antegrade enemas can be useful in severely constipated patients with spina bifida not responding sufficiently to retrograde bowel management.
Colon	Large intestine, which is about 1.5 meters (5 feet) long.
Colostomy	Opening into the colon through the abdominal wall for the passage of faeces.
Congenital	Present at birth.
CVS	Chorionic villus sampling (CVS) is a pregnancy test that checks the baby for genetic or biochemical abnormalities
Defaecation	The opening of bowels.
Diagnosis	The process of determining the nature of a disorder.
Dilatation	Expansion of an organ or vessel.
Disease prestige	The collective perception of a disease's "worthiness;" the degree to which the sufferer "deserves" care and support. As a social construct, it changes over time and between

	communities. A disease's prestige reflects the community's commitment to care, support and economic investment.
Distal	Farthest from a point of reference.
Distension	An uncomfortable swelling in the intestines usually caused by excessive amounts of gas and fluid.
Duhamel procedure	The Duhamel procedure was first described in 1956 as a modification to the Swenson procedure. A retrorectal approach is used, and a significant segment of aganglionic rectum is retained. The aganglionic bowel is resected down to the rectum, and the rectum is oversewn.
Functional Enterocolitis	Voluntary or involuntary passage of formed, semi- formed, or liquid stool encopresis into a place other than the toilet for more than one time per month in a child over four years of age for at least three months.
ENT	Doctor who specialises in conditions of the Ear Nose and Throat
Enterocolitis	Inflammation of the intestines.
Excoriation	Burning or blistering of skin from contact with faeces/urine.
Faecal continence	To have control over the natural impulse to defaecate.
Faeces	Body waste discharged from the intestines.
Fistula	An abnormal tube-like connection between two hollow organs such as between bowel and bladder.
HAEC	Hirschsprung's associated enterocolitis.
HSCR	Internationally recognised abbreviation for Hirschsprung's disease.
Gastroenterologist	A doctor who specialises in conditions of the digestive system, including liver, biliary tract and pancreas.
Gastrocolic reflex	A physiologic response in which the simple act of eating stimulates movement in the gastrointestinal tract.
Genetic condition	A condition that is inherited.
Gastroscopy	A test to examine the oesophagus, stomach and duodenum using an endoscope.
Gastrostomy	Opening into the stomach to enable feeding directly into the stomach via a button device or tube.
Girth measurements	Girth measurements, although considered inaccurate, are often used as a method of determining abdominal distension
Ileostomy	Surgical creation of an opening into the ileum (last part of the small bowel) for the purpose of evacuating the bowel contents.
Ileovesicostomy	Also known as "cutaneous ileocystostomy", "ileal chimney", or "bladder chimney") is an uncommon urologic diversion in which a loop of small bowel is anastomosed/augmented to the dome of the bladder. This loop of bowel then exits through a urostomy.
Jejunostomy	Tube placed either directly into the jejunum (second part of the small bowel) or fed through a gastrostomy tube to enable feeding

Laxative	A drug used to stimulate or increase the frequency of bowel motions.
Manometry	A test using a special tube/balloon to measure of pressure within various parts of the gastrointestinal tract.
Meconium	The first stool passed by a newborn infant normally occurring in the first 24-48 hours after birth. It is characterised by its dark green sticky appearance.
Megacolon	Excessive dilatation of the colon.
Migration	The movement of cells etc. from one position to another.
Mother Guilt	A feeling of guilt, doubt, anxiousness or uncertainty experienced by mothers when they worry they are failing or falling short of expectations in some way.
Mucosa	Mucous membrane, lines the entire gut from mouth to anus.
Mucous fistula	Non-functioning stoma, usually the second stoma of a double barrel colostomy which secretes mucous but does not contain any faeces.
Non-functioning Encopresis	Faecal soiling, or the involuntary passage of fluid or semisolid stools into the clothing. Non-functional includes anorectal malformations, Hirschsprung's disease and spinal cord anomalies.
Paediatrician	A doctor who specialises in children's medicine.
Paediatric surgeon	A surgeon who performs operations on children.
Peristalsis	The wave like movement of the bowel which pushes food along the intestines towards the rectum.
Perforation	Creation of a hole in an organ (in this context the bowel).
Polydactyly	A baby born with one or more additional digits; the name comes from the Greek poly (many) and dactylos (finger).
Premature	An infant born before completion of 37 weeks gestation.
Preterm infant	Infant usually weighing less than 2500 grams (5 ½ lbs).
Primary pull through procedure	Primary pull through procedure involves removal of the entire aganglionic colon, with an end-to-end anastomosis (re-joining) of the normal colon to the low rectum.
Rectosigmoidectomy	Rectosigmoidectomy (rĕk″tō-sĭg″moy-dĕk′tō-mē): Surgical removal of the rectum and sigmoid colon.
Reflux	A backflow of liquid against its normal direction.
Retrograde	Going backwards or moving in the opposite direction.
Resection	The surgical removal of a portion of an organ such as the intestines.
Sphincter	Ring of muscle that opens and closes the anus.
Sphincterotomy	To cut or stretch the anal sphincter muscle.
Stoma prolapse	Extended protrusion of the bowel through the stoma opening.
VATER/VATCERL	Acronym for Vertebral defects, Anal atresia, Cardiovascular defects, Trachea defects , Oesophageal fistula/atresia, Renal defects, Limb anomalies

Vomit	The act of disgorging stomach contents through the mouth, also refers to the disgorged matter itself.

PROLOGUE

My experience of Hirschsprung's disease (HSCR) and anorectal malformations (ARM).

I began my nursing training in 1975 at Camden District Hospital, an hour's drive and 70km southwest of Sydney. There was no Campbelltown hospital, and Liverpool hospital was just another small district hospital. The local coal mines were working at full capacity and there were a lot of young families in the area, so Camden Hospital ran very busy wards including maternity, children's, the emergency department, operating theatres, ICU and the general male and female wards. The area had attracted a new influx of specialists from Sydney to support the growing need and offered a semi-rural lifestyle to raise their families.

I loved nursing. As a child, I used to walk to the sanitorium where my mother worked as a nurse caring for sufferers of tuberculosis. I would be allowed to help in some small way and knew this was what I wanted to do. Mum and I used to watch 'Your Life in Their Hands,' an English documentary showing operations being performed for the first time on television (Essex-Lopresti, 2006). My training was hospital-based in a joint training school with Liverpool and Fairfield where we would come together for six-week blocks of lectures, mostly given by doctors and occasionally by a nurse educator. This was an apprenticeship model, moving through three years in a systematic way, acquiring competence towards the end of third year to run a ward. There was a definite hierarchal system within hospitals at that time, if a more senior person entered a room, you stood up, hands behind your back, white starched uniforms meticulously maintained, makeup, jewellery or anything below the elbows was not permitted, including a wristwatch.

I gravitated to the critical care areas of nursing and worked in maternity, emergency, children's ward and the operating theatres and I thrived on emergency situations. In the last few months of my three-year course, I was asked to take over the running of the children's ward at Camden Hospital as the registered nurse who was in charge there had been asked to leave, I had no idea why. I accepted and thrived on the responsibility but found it frustrating that the rules were rigid. Parents were not permitted to stay with their children, the teaching at the time was children were more distressed when their parents were around, and we had a job to do. There was a huge diversity on the ward from babies brought in overnight with croup to be set up in oxygen tents; terrifying for them and alarming for the parents; to adolescents in traction from fractures sustained in their many adventurous or sporting activities. We also had two isolation beds for infectious diseases. At the end of general nurse training, we were known as Sisters, a title carried over from the Sisters of Charity who arrived from Ireland in 1838 and established a hospital (with patronage from a non-Catholic) in 1857.

I took a position in the operating theatres and whilst there were a lot of routine theatre cases, we had our fair share of complicated and/or emergency cases. We would treat and transport the urgent or complicated cases on to the large tertiary hospitals, and I had a burning desire to know what happened when we transferred these patients to the city hospitals. Once I reached a level of competence, I always wanted to learn more.

Working in tertiary hospitals

I was fortunate and grateful for the support of surgeons I worked with who encouraged me to apply to do the post basic operating theatre course. There were only five city hospitals that ran such a course, and they were renowned for only accepting their own graduates. Those of us who trained outside of the city were not normally considered. I first applied to Royal North Shore Hospital, but quickly realised what I was up against when the first question asked at interview was 'What did your father do?' I then applied - somewhat daunted by that experience - to St Vincent's Hospital in Darlinghurst. Fortune was on my side as this was the first year St Vincent's had offered the course to 'outsiders,' and they had never accepted a student who was married. I was accepted with strict conditions after multiple interviews which I am sure were designed to reveal if I had the determination to stay the course. I was very grateful for the opportunity and knew I had a responsibility to future students not to abuse the trust bestowed on me.

St Vincent's did not cater for paediatric patients, so we were seconded out to what was then known as the Royal Alexandra Hospital for Children at Camperdown. Not having children of my own at that stage I found it upsetting seeing babies and young children undergo what seemed like such torture. I felt for the parents who were not permitted into the theatres or even anaesthetic bays in those days and had to say goodbye to their children at the theatre doors. This was most distressing for all concerned and very frightening for the children. I came home upset every day and vowed I could never work in a paediatric operating theatre.

St Vincent's was an education like no other, we cared for sex workers from Kings Cross when they came through emergency for routine antibiotics, through to tragic trauma cases such as a young woman who had suffered a vicious vaginal attack with a broken bottle. We operated on her for hours as she lost her uterus, spleen and a large segment of her bowel, but was deemed 'lucky to be alive' at the end of it. I often wonder what happened to her. As students, we rotated through all specialties, and I will be forever grateful I had the opportunity to work with outstanding surgeons such as the late Dr Victor Chang. I did not realise at the time that this was the forefront of specialisation in Australia. Once the course was over, I worked in a variety of public and private operating theatres in and around Sydney with newfound confidence.

Our own four children were born between 1982 and 1989 and I became a 'stay at home mum' for many years. Our three daughters were born first without incident and then in 1989 our son was born. Having been in the profession, I was fortunate to be able to choose an obstetrician I knew and trusted. I recall him explaining at the first consultation when I was pregnant with our son Adam, that the Government were now offering Chorionic Villus Sampling (CVS) to any woman over 35 years of age. I saw no point as our third daughter had just turned one, so why would situations change from one pregnancy to another in such a short time, even though in those days I was considered an 'older mother'. The pregnancy was uneventful, and Adam was born late one night after a long protracted induced labour. It was not until the following day that I learnt that there were concerns Adam may have Down syndrome. Within a few days we were in an ambulance being transferred to the then Prince of Wales Children's Hospital. These early days in Adam's life are enfolded in the research arc of this

thesis, interrupting and activating the research literature and aims of this doctoral thesis.

This was the start of many admissions, surgeries and procedures, all in an attempt for Adam to gain faecal continence. Each time Adam was admitted to the children's hospital, the staff would say, "Oh it's Adam, not Amy." Curiosity got the better of me and I wanted to know who Amy was. Eventually I was fortunate to meet Doreen, Amy's mother who was also a registered nurse. Although we were strangers, Doreen and I – Amy and Adam - shared the same surname. Amy was five years older than Adam and had been born with Down syndrome and imperforate anus, one of the types of congenital anorectal malformations.

Doreen and I met with Jenny, a skilled paediatric nurse at Prince of Wales Children's Hospital who was integral to the care of these children on the surgical ward and had a passion for understanding congenital bowel anomalies. It became clear, as we tried to understand the complexity of these rare conditions, that neither we nor the paediatric surgeons had all the answers we required. We realised that if we had trouble navigating our way through these rare conditions, with the benefit of working in the system, then how difficult must it be for families who were not familiar with the hospital system and had been handed a shock diagnosis after the birth of their baby that was difficult to spell let alone comprehend. We decided the best solution was join forces to offer what little bit of collective experience and knowledge we had with the aim of bringing families together to support each other. In 1994, with the support and encouragement of our team of paediatric surgeons, the Bowel Group for Kids was formed.

Even though I was not working outside the home at the time, I enjoyed the return to the health system albeit in a unique way. Life was busy but sharing what we had learnt with other parents and carers we met along the way was our way of activating diverse knowledge, skills and education with others. The unexpected bonus was we met other parents in a similar situation and were able to support each other simply by being able to discuss the issues we were facing that were unique and unable to be shared with family and friends as they were not in line with the accepted stages of child developmental.

The issues were not the usual toilet training process for toddlers. These children and their parents were trying to follow a normal pathway of bowel and bladder training, but their start to life was vastly different to most babies. Hirschsprung's disease and anorectal malformations require major surgical intervention, some require diversionary stomas for bowel and sometimes bladder as well. Many children need ongoing management for life: something that was not clear when Adam was born. At that time, the condition was usually treated more as a surgical problem. Once the operations were over, the advice was generally to call the surgeon if there was a problem. The trouble was, when the baby was born with such complex problems and underwent major corrective surgery from birth, it was difficult to isolate the cause of what our children were experiencing; was it normal childhood experiences such as teething, fussy eating, an upset tummy, or was something to do with the anorectal malformation.

The surgeons at both the children's hospitals in Sydney were very supportive of our efforts to brings parents together and told us they were also learning along the way. This was more than we could have hoped for and gave us all a feeling

of working together as a team for the best outcome for the children, rather than the hierarchical system of the past.

I wrote a letter to the Down syndrome Association asking to be connected with other parents who had a baby born with Down syndrome and either Hirschsprung's disease or an anorectal malformation. The association itself did not include associated conditions on their database and to my knowledge, this remains the case even though it would be invaluable for parents whose children have other conditions known to be associated with Down syndrome. My letter was uncovered recently, and I was surprised to realise that my interest in this area of research began so long ago. This request printed in the Down syndrome newsletter, was the start of my journey into developing and growing with a network of parents whose loved children had an invisible and stigmatising condition:

> Parents of a five-year-old child who has both Down syndrome and Hirschsprung's disease has sought ADSA's assistance in contacting other parents with a view to sharing valuable information about their children.
>
> Their own child is doing well, but his social development is being impaired by the ongoing problems associated with faecal incontinence following the definitive surgery. They have exhausted all other avenues and are now working closely with a team of specialists at the Prince of Wales Children's Hospital in Sydney to come to a satisfactory means of managing their child's physical situation.
>
> It appears that there is very little knowledge at this stage of this combination of disabilities; the Prince of Wales team is therefore keen to conduct a survey Australia-wide to determine how many children are involved, and their various health situations, in the hopes of improving their

> understanding of this condition. The type of information they will be seeking is:
>
> Parent/s' name/s, address, telephone number; child's name, date of birth.
>
> At what age the child was diagnosed with Hirschsprung's disease; if/when surgery was performed, and at which hospital, paediatric surgeon; age at closure of colostomy; Are they continent following closure? If not, what problems have been experienced? Any current medications - that is, laxatives or enemas.
>
> Any parents interested in contacting the Gribbin family to share their experiences; and/or permitting their information to be passed on to the Prince of Wales's medical team to assist their survey, please contact Eunice. (Gribbin, 1994)

What followed was the life-changing connection with twenty-seven other parents who formed the basis of the support group members. The response of parents to the support group was always the same: they thought they were the only ones who had a child with this rare condition along with Down syndrome. Through the support group, we have met some extraordinary people around the world. I was fortunate to be invited to meet with support group organisers we had corresponded with from Germany, Italy and France when I presented at paediatric surgeons' conferences in in Frankfurt 2013, Berlin in 2015 and Milan and Ljubljana in 2017.

Whilst the medical care in Australia is effective and professional, it is the day to day lived experience that doctors cannot possibly be expected to know or understand, which is where parent-to-parent support groups can fill that void. Some of the babies of families who joined the inaugural Bowel Group for Kids, are now in their thirties, and some are giving back to the community, sharing

their own firsthand experiences with families who are just starting their journey but with much more information and resources than we had access to, the internet has certainly changed the way we can access information instantly.

Working at the Children's Hospital Westmead

In 1995 the Royal Alexandra Hospital for Children moved from Camperdown where it was when I specialised in theatre nursing, to Westmead, about twenty-six kilometres, from Sydney which is part of the Greater Western Sydney Region. By then, I had been back at work for just over a year working casually in various hospitals. Having been around the Children's Hospital for the past five years as a parent, I had a new appreciation for paediatric nursing, so I decided to apply to the new Children's Hospital at Westmead for a position as a perioperative registered nurse. I was fortunate to be accepted and enjoyed eight years working there. This gave me the opportunity to not only use my skills as a perioperative nurse, but as the mother of a child who had been through multiple surgeries, so I knew firsthand what families and their children were experiencing. I was able to visit the parents of children born with an anorectal malformation pre and postoperatively and offer my support as a parent. The clinicians I worked with were supportive and keen to understand more about the lived experiences and needs of the families.

I found this rewarding - not to give any form of medical advice as this was the domain of the treating team, but just to be a supportive person who understood from the unique perspective of being a mother of four young children, and a nurse. I also began running in-service sessions for the nursing staff to expand their understanding of the many surgeries that were performed in the theatre

suite. This helped develop my love of teaching which grew over time to teaching nursing and medical students both on campus at university and in the clinical setting. Years later when I was working in general theatres, an obstetrician I was assisting, asked me to help him understand what life was like raising a child with Down syndrome. This surprised me as I had worked with him for many years and knew he had delivered hundreds of babies, some of whom were diagnosed with Down syndrome. He explained that despite his vast experience in obstetrics, once a mother leaves hospital after the birth, he had very little further contact, other than a short postnatal visit six weeks later. After the baby's birth, their care becomes the domain of the paediatrician and the obstetrician really wanted to understand what we had experienced. This encounter has stayed with me to this day, and when appropriate, I include my personal experiences when teaching nursing and medical students.

Teaching at University

In 2001, I accepted an education role at university facilitating nursing students during the clinical component of their training, as well as lecturing and running tutorials on campus. I thrived in this new aspect of nursing and found I had a deep love of teaching in the various settings. I mainly supported students in their final placements before becoming registered nurses due to my background being mostly critical care and paediatrics.

School Nurse

My experience in nursing has been far more varied than I ever imagined it would be and I doubt it would have extended beyond my original specialisation of operating theatres had it not been for the unique experiences gained after my

son was born. My love of paediatric theatre nursing and general paediatrics came as somewhat of a surprise given it was something I was sure I would never be strong enough to endure after my earlier experience as a novice. That was before I became a mother to three amazing daughters who showed such love and resilience after their brother was born. In 2006, the special school my son attended, advertised for a registered nurse to set up a formal nurse's station. As Adam was 16 years of age, he had two years left at the school. I saw this as an opportunity to extend my skills in understanding the needs of children with disabilities from a different perspective, while supporting the staff and children who cared for him. Up until this time any form of nursing care at the school had been provided by the teachers aids who had first aid experience. The school accepted children with intellectual disabilities, so the main focus was not on their physical disabilities as such. I saw my role as adding to the wonderful work the aids were already doing, I established policies, provided in-services for staff, set each classroom up with hand sanitiser for the children to use, over a decade before this became standard procedure following COVID-19, but I saw it as essential to reduce the spread of infection. Action plans were streamlined for children with specific conditions such as a young girl in Adam's class who had a moderate intellectual disability and Type I diabetes. She had trouble eating so she used to spend her lunchtime in my room so I could supervise her food intake, administer her insulin and monitor her blood sugar levels. Another child I felt was capable of performing her own self catheterisation but had resisted any involvement. I worked with her, her mother and the continence nurse I knew from the Children's hospital, and collectively, the young girl developed independence and this also provided a level of protection as she got older rather

than having others attend such a personal procedure when she left school. Another child suffered from uncontrolled seizures and would stop breathing, requiring me to call paramedics, but I did manage to implement strategies to reduce the number of times the ambulance service was called to the school. At the end of Adam's schooling, he was scheduled for more major surgery, and after two years there, I felt I had reached the limit of what I could achieve and accomplish in this location and yearned to be back in clinical nursing. So, when Adam went into hospital, I finished up at the school which turned out to be the best option as we came close to losing him in ICU, just days after the surgery so I needed to be with him constantly.

In 2008, after leaving the special school and Adam had recovered from surgery, I approached the new medical school that had opened at the University of Western Sydney[1] where I was working in nursing. The medical school was calling for experienced educators to teach undergraduate medical students. I decided to apply and transferred from nursing to medicine. This brought even greater opportunities to share my personal experiences with the young students when appropriate. Whatever the setting, be it education, the peer to peer support group or presenting at a conference, personal accounts are generally well received by the audience and this was certainly the case for my students who enjoyed the lived experience when it was in context of the subject we were exploring.

I taught across most subjects; problem based learning, law and ethics, communication involving actors delivering bad news or being an angry patient,

[1] The name has since changed to Western Sydney University.

this gave the students the skills to manage these situations in a safe environment. During fourth year, students were engaged in clinical practice during rotations of paediatrics, mental health or obstetrics and gynaecology, they were required to write a weekly online journal using reflective practice strategies they had been taught. This involved writing about an encounter during one of these rotations to explore how they felt about a given situation and how it could have been handled better. One unexpected revelation was how many of the students reflected on their own mental health when discussing encounters with patients diagnosed with mental health conditions. As the online journal was only accessible to the student and myself as a facilitator, this provided a safe platform to reveal aspects of themselves they had been too afraid to expose. They explained this enhanced their understanding of the patient experience and gave them an opportunity to compare their own lived experience with that of their patients, not unlike (Allen & Lavender-Stott, 2020) found in their students' experience exploring the taboo topic of abortion which opened their minds to the fact that their lived experience had previously shaped their mindset.

One aspect of the course was for students to be focused on patients in the community. I was asked to allow students to interview Adam as part of this program. This was a powerful opportunity and expanded to include Adam, my husband and I attending tutorials to share our experiences as parents of a child with multiple disabilities. In time, a colleague and friend who also had a son the same age as Adam with Down syndrome, joined us. The students were able to ask us any questions and gain an understanding of the complexity of anorectal malformations. Having the two boys attend was invaluable, whilst they shared a diagnosis of Down syndrome, that is where the similarities ended. Adam was

non-verbal, Brad (name changed for privacy) was very vocal and did not have any associated medical issues, this was a perfect opportunity for students to understand that a primary diagnosis in one person does not mean all people with that diagnosis are affiliated the same way. I often come across former students who are now qualified doctors who tell me they have fond memories of those sessions; the experiences have remained with them as they were able to learn so much about caring for children which they felt was missing from textbooks and formal lectures. A surprising benefit of this program, was the students' ability to teach me, the educator, gaining insight into the concerns of students when it came to caring for people with disabilities, was something I had not anticipated. These experiences have changed the way I teach students and has brought a deeper understanding to the program and institution where I work (Allen, 2022). What I have been able to share in all these roles is the perspective of a parent and the child to our young clinicians in the hopes they will have a better understanding of the needs of families.

Neonatal Bowel Clinic

As part of this research, I have been fortunate to attend the Neonatal Bowel Clinics at both Randwick and Westmead campuses of the Sydney Children's Hospital Network (SCHN). There, I have met families, some from the support group, others who had no idea support was available. These multidisciplinary clinics offer access to a surgeon, stomal therapy nurse, psychologist, dietician and social worker as deemed appropriate. Having this opportunity for families in one setting rather than having to attend multiple appointments is wonderful. These clinics also provide ongoing routine follow-up appointments which it is hoped, will develop data on the long-term outcome to guide future practice.

Over time, I realised that my perspectives as a mother and co-founder of a support group could be combined with my nursing experience to delve deeper from an academic viewpoint to question the validity and types of support systems parents could access when a child is born with a congenital abnormality. Initially my doctoral research was directed towards quantitative data, with data being collected from children's hospitals in Sydney, Newcastle and Canberra. It quickly became clear that it would be very difficult to investigate through these methodologies whether or not being a part of a support group improved the quality of life for these families and their affected children. I then started to shift the focus to the lived experience, whilst the data collected was analysing demographically, the main focus was now on interviews and questionnaires from the perspective of families, clinicians and support groups. Theoretically, I aligned my research through a re-engagement with Goffman's Stigma. This 'old theory' offered new insights and an innovative frame for these complex epistemological, methodological and ontological issues.

The level of medical care has never been in question throughout this research. Australia has a strong medical care service for children. What is missing though is the understanding that each family is different and as clinicians, we cannot possibly understand what life is like for those caring for a child who has faecal incontinence. My doctoral research aims to fill that gap by showing what kinds of support systems are most beneficial to support families when their baby is diagnosed with Hirschsprung's disease or an anorectal malformation. This is especially important in both conditions as even now in 2023, neither can be diagnosed antenatally and come as a terrible shock to parents shortly after birth. The research will show that having information about support services at the time

of diagnosis is important. The ability to talk to someone who has been through this same situation as they find themselves in, can be helpful. It is hoped the inclusion of support services as part of the healthcare team when a baby is born with Hirschsprung's disease or an anorectal malformation, will help guide families through the whirlwind of care required in hospital, at home and throughout their child's education.

This prologue offers the first lens on this research. It is experiential, but demonstrates the early alignments between universities and public health, learning and living. From this position and positioning, my research works between these spaces to create new insights, methodologies and approaches to understand Hirschsprung's disease as a community of care and in care, rather than an individual 'problem' to be named, labelled, judged and diagnosed.

INTRODUCTION

Patient experience

As parents, the excitement of a much-wanted pregnancy looms large. So much to consider. Perhaps, like us, this one was a 'surprise package' evident on a flight home from the UK via America. Either way, if the news of a pregnancy is welcomed, women move into an intricate social space. Heavily pregnant woman are greeted with a knowing smile, no need for words. We see new mothers with babes in prams, babies cooing at their dads and older siblings, an unsettled baby we yearn to comfort, these small glimpses into our future is all consuming.

Planning attends the arrival of a newborn; furniture to buy, equipment seemingly essential yet sometimes never used, the house to organise, children's rooms to rearrange to accommodate the new baby. The day our baby was born, we were thrust into a typical whirlwind of joy and excitement. Over the next few hours or days, sadly it became clear something was not quite right. He was kept in the special care nursery longer than I felt necessary. At each nappy change the absence of meconium was noted, this is the first dark green tarry stool a baby passes within the first twenty four to forty eight hours, it consists of material ingested in utero such as amniotic fluid, swallowed cells, bile and water. I became frustrated thinking, 'why is there such a focus over a seemingly irrelevant milestone, we just need to get home.' Midwives reassured us it was just routine, but unbeknown to me, they had started measuring my baby's abdomen for signs of distension (swelling). In an acute bowel obstruction of the newborn, once the baby starts to feed, the baby's abdomen will often be distended as waste products cannot be passed out of the body due to the blockage. Some babies

may vomit bile (bilious vomiting). Concerns were raised, and we soon found ourselves surrounded by eager medical students as a consultant fired questions at them. The situation seemed surreal as we realised, they were talking about our baby who was just perfect in our eyes and I just wanted to go home, our girls needed me and I had promised them I would only be away a few days.

The question of Hirschsprung's disease was never raised with us in hospital, although I later learnt his paediatrician had suspected it and ordered an abdominal X-ray. The suspicion that Adam may have been born with Trisomy 21 loomed large and could not be confirmed until genetic test results were available, which could take up to six weeks. Our paediatrician was very astute. He knew that I wanted to distance myself from doctors and the hospital. I convinced myself they were all mistaken in their diagnosis. He looked like his sisters. He did not look like babies I had cared for that were born with Down syndrome. It was only after I insisted on taking him home after a weeklong stay and no answers, that reality hit. By the next morning, I knew only too well, he had an acute bowel obstruction, the rest is history.

The grim reality that something was not quite right weighed heavily, but what? I had had all of my antenatal check-ups, ultrasounds confirmed all was as it should be, I did not drink or smoke, and had taken as much care of myself as is humanly possible with three children under the age of six and another on the way. I avoided the ever-growing list of foods to be eliminated during pregnancy and nothing seemed out of the ordinary compared to my other pregnancies, so how could anything be wrong?

Then the doctors broke the news they had concerns our baby may have a bowel obstruction. Babies are routinely checked within the first 24 hours to confirm the presence of an anus, and that it is correctly positioned. If not, this is known as imperforate anus, also termed an anorectal malformation. The condition has varying degrees of severity from a thin covering of tissue over the anus, to entire sections of the lower bowel missing with no visible anal opening. Another cause of a congenital bowel obstruction is Hirschsprung's disease where there is an absence of ganglion (nerve) cells in a section of bowel from the anus back to varying points in the gastrointestinal tract, preventing the passage of stool. The affected segments range from ultra-short at the anus to the most severe form which encompasses the entire gastrointestinal tract, including the oesophagus. This is very rare and is largely inconsistent with life. I recalled learning about congenital bowel disorders during my theatre training, but had never encountered any patients myself, probably because up until Adam was born, I had worked in adult critical care areas, not paediatric.

We entered the unfamiliar world of the healthcare system from a parent's perspective, which felt very strange to be on the wrong side of the bedside with a whirlwind of surgeons, and specialist nurses. As our precious baby underwent multiple tests and procedures, then major surgery to correct the anomaly at just thirteen days of age. This was not the new beginning we anticipated, why could this abnormality not have been detected during pregnancy? As a mother, I immediately wanted answers, and my mind went into overdrive…

Mother guilt emerged. Was it something I did wrong? Did I eat something I should not have? Was it the chest infection I had during early pregnancy? Was it

because we were travelling to distant parts of the world? Did I inadvertently take some 'over the counter' medication in the early weeks before I realised I was pregnant?

So many unanswered questions. We were reassured by the healthcare team that these abnormalities were present from the time of conception. They do not occur at any stage throughout the pregnancy, and some may be genetic. We started to ask family members if they knew of anyone who had a similar condition. Some families we have met have expressed when questioning their families, an elderly relative may recall a sibling or aunt who had a baby who died in early infancy with a huge tummy, but these vague recollections do little to dispel the fear of the unknown. Whilst Danish surgeon Dr Harald Hirschsprung named the rare condition in 1988, corrective surgery was not devised until the 1940's when Swenson and colleagues recognised the puzzle of symptoms for the condition known as Hirschsprung's disease. Historical cases are often difficult to confirm, and the mortality rate was high during experimental surgical techniques (Stephens 1950). As parents of three young children and a newborn with major complications, we were exhausted as we came to terms with the reality of the situation and tried to explain this strange phenomenon to family and friends. How I often wished I were not a nurse, I was always analysing his symptoms using the mechanism of injury I taught my students, rather than being able to just be his mum, knowing too much from a nurses perspective was not always helpful in this situation, or throughout the many hospitalisations to come.

After surgery, we navigated years of bowel management in an attempt to normalise bowel function, learnt ways to fend off unwelcome advice from well-

meaning friends and strangers, then entered the complex world of the education system that was ill prepared to support our child due to his faecal incontinence. We became advocates for our child in an attempt to ensure this did not impact upon his right to access a good education, strengths we never realised were simmering under the surface.

All being well, we would all come through this terrible situation relatively unscathed, no doubt more resilient as we learnt to manage any residual incontinence our child may have been left with. But what about us as parents, how has it impacted our relationship with each other and our other children, we worry about them too. There was always a feeling we never had enough time to share around as our child born with a bowel disorder often had to take priority. Despite our constant concerns, we are fortunate that our three daughters, who are now in their thirties, have learnt invaluable life lessons we could never have formally taught them; acceptance, compassion, living life in the moment; they are wise beyond their years, and we admire their ability to pick themselves up, dust themselves off and carry on, no matter what life throws at them. We sometimes wonder, did growing up with Adam have a detrimental effect, or has it taught them far more than we realised.

When Adam was born, the shock of multiple diagnoses coming at what seemed like lightning speed sent our heads spinning, how quickly life changes. The focus during the day spent in labour was more about getting the job done, having this baby and getting home as soon as possible. After all, I was a very organised person, I had prepped the girls, stocked up with groceries and as I had to be induced, all was going to plan, very neat and organised. Then the labour went

much longer than expected, Michael had to go and collect our little girls who were being minded at their aunt's along with her four year old triplets and seven year old child. We were anticipating the baby would be born before dark so they could all celebrate and go home; but alas this was not to be. Once Michael and our girls were home, there was no way to contact them as our telephone had been cut off due to a fault with the provider. A neighbour was able to convey the news of Adam's birth to them so at least they knew he had been born before they came in the next day. By the time they arrived to see Adam, I had been delivered the devastating news that he may have Down syndrome. How could everything have gone so wrong in such a short time? We all needed time to comprehend what this meant, all the while trying to shield our daughters from our pain and concern. We had no idea the enormity of what was about to unfold over the following few days. We felt like we had been hit by a train, how would we ever get back to the normality we took for granted? Solace is found in the most unexpected places and in our case, a colleague of Michael's sent us a poem which I feel describes the complex, unpredictable but enriched life that we never thought was possible in those early days, but luckily we have experienced much more of as a family since Adam was born. The poem 'Welcome to Holland' was written by Emily Perl Kingsley in 1975 when her son was born with Down syndrome. She was working on the television production *Sesame Street* as a writer at the time and continued to do so until she retired in 2015.

Welcome to Holland:

I am often asked to describe the experience of raising a child with a disability - to try to help people who have not shared that unique experience to understand it, to imagine how it would feel. It's like this……

When you're going to have a baby, it's like planning a fabulous vacation trip - to Italy. You buy a bunch of guide books and make your wonderful plans. The Colosseum. The Michelangelo David. The gondolas in Venice. You may learn some handy phrases in Italian. It's all very exciting.

After months of eager anticipation, the day finally arrives. You pack your bags and off you go. Several hours later, the plane lands. The flight attendant comes in and says, Welcome to Holland."

"Holland?!?" you say. "What do you mean Holland?? I signed up for Italy! I'm supposed to be in Italy. All my life I've dreamed of going to Italy."

But there's been a change in the flight plan. They've landed in Holland and there you must stay.

The important thing is that they haven't taken you to a horrible, disgusting, filthy place, full of pestilence, famine and disease. It's just a different place.

So you must go out and buy new guide books. And you must learn a whole new language. And you will meet a whole new group of people you would never have met.

It's just a different place. It's slower-paced than Italy, less flashy than Italy. But after you've been there for a while and you catch your breath, you look around.... and you begin to notice that Holland has windmills....and Holland has tulips. Holland even has Rembrandts.

> But everyone you know is busy coming and going from Italy... and they're all bragging about what a wonderful time they had there. And for the rest of your life, you will say "Yes, that's where I was supposed to go. That's what I had planned."
>
> And the pain of that will never, ever, ever, ever go away... because the loss of that dream is a very very significant loss.
>
> But... if you spend your life mourning the fact that you didn't get to Italy, you may never be free to enjoy the very special, the very lovely things ... about Holland (Kingsley, E.P. 1987).

Background

This thesis explores the difficulties families face throughout a child's life when their baby is born with Hirschsprung's disease or an anorectal malformation. Hirschsprung's disease and anorectal malformations are rare, complex congenital anomalies which, despite advances in medical knowledge and technology, remain largely undetected until after the baby is born. This introduction sets the background, rationale and aims of the research conducted and presented in this doctorate. The medical background, treatment and complications of Hirschsprung's disease and anorectal malformations will be explored. The impact on families can be devastating when they are thrust into a complex medical environment of a neonatal intensive care unit, operating theatres and recovery suits, often long distances from home. The priority when a baby is diagnosed with Hirschsprung's disease or an anorectal malformation is accurate diagnosis then the extent of the condition, avoidance of life-threatening infections and surgical intervention to relieve a bowel obstruction and correct the anomaly. Once the family returns home from hospital, the parents often face a challenging time managing a new regime of bowel management and conflicting advice from well-

meaning friends and relatives. Due to the rare nature of the conditions, baby health clinics, general practitioners and even paediatricians often have limited experience. The baby's surgeon will often manage follow up to a certain period, but in the absence of a mechanical issue, or the likelihood of a surgical intervention being useful, there is little the surgeon can offer if continence is not achieved. Parents can find themselves marooned with no one to turn to for advice.

My significant, original contribution to knowledge is to demonstrate that the medical model of Hirschsprung's disease or anorectal malformation is not sufficient to enable not only the treatment, but the life of these citizens. Acknowledging my own experience as a mother, nurse, educator of future clinicians and co-founder of a peer to peer support group for families who have a child born with Hirschsprung's disease or an anorectal malformation, this thesis draws on parents' stories to gain insight directly from those living through this traumatic experience. Surveys were conducted of members of peer to peer support groups, the group organisers and paediatric surgeons internationally. The goal of this research is to ensure that the lives of citizens and their families are understood with context, texture and meaning, rather than a diagnosis. A diagnosis is not an ending, but the beginning of new nodes and modes of community.

Hirschsprung's disease and imperforate anus

Hirschsprung's disease and anorectal malformations are often used together, with the latter denoting conditions like imperforate anus. Hirschsprung's disease is a congenital malformation which is characterised by a lack of ganglion cells in

the intestine stretching continuously from the anus to an indeterminant region higher in the rectum, preventing normal peristaltic activity through the bowel, and therefore resulting in a non-functional colon.

Hirschsprung's disease is the most common cause of large bowel obstruction in the neonatal period (Levene 2008; Barr 2013; Lotfollahzadeh et al., 2022). The disease does have a genetic dimension, and one-fifth of cases are familial; however, half the cases of Hirschsprung's disease have no known genetic connection (Tam and Garcia-Barceló 2009)

The Bowel Group for Kids was fortunate to have the inspirational Olympian Duncan Armstrong join the organisation as its Patron in 2006. Duncan was the perfect person to support the organisation and encourage children affected by these conditions not to allow their conditions to hold them back from their aspirations. Duncan himself was born with Hirschsprung's disease and tells his story:

> My 'faulty plumbing' was operated on at an early age and I learned to adjust my lifestyle around my new relationship with the toilet. For me I didn't really feel any different from anyone else I simply had special rules to learn about foods, eating times, and number of trips to the toilet.
>
> From the age of two or three up until I was around 24 I had to be very disciplined about what I ate, when I ate it and how far from a toilet I was at all times. This sounds very difficult but really it was just the way I was, I can't really remember being sad about it or missing anything.
>
> I just had to stay on top of it. Sure, there was the all-day toilet workouts, and the unbelievable painful gas days, and the

embarrassing 'bum-trumpet' moments. But that was just part of who I was, I was special and sometimes being special means, you work a bit harder than everyone else. This also has the effect that you seem to enjoy the simple things a little more than if you didn't have such a specialty in your life.

I've never felt that Hirschsprung's ever really held me back from parts of my life in which I really wanted to succeed. I set my heart on competing for Australia at around the age of 15, so my training in the swimming pool exploded with intensity. This also brought on plenty of explosions of another kind! Intense training really upset my bowel and for the first year or so of hard swimming I almost had to re-educate my body to cope. Many times, I'd be halfway through a great training session only to end up spending the next two hours in the pool's toilet while all my friends and training squad were finished and long gone home. My body took a great deal of time to adjust to intense training, I'd fall sick and lose important weight at crucial times because my bowel couldn't take the training regime. But slowly over time and with great patience I found the foods that work best, a supplement routine that supported me, and training times that helped me cope. I never gave up trying to find the rhythm for which my bowel could work at its best. Sometimes I would make some BIG mistakes and pay for it, but slowly over time I worked through the misfires and found what best suited me. This allowed me to chase and achieve my dream of wearing the Green and Gold at the Olympics.

These days I'm blessed with 3 beautiful children and wonderful wife. My youngest son was also born with Hirschsprung's disease which brought a new perspective on the condition from my own situation to understanding what it was like from a parent perspective, until I had this experience myself, I did not understand what my own parents went through. From about the age of 24 when I retired from swimming my health has been mostly normal. I go to the toilet twice a day every

> day and will probably enjoy this routine for the rest of my life, everything else in my life is incredibly normal.
>
> I know there are families whose child or family member is doing it a great deal tougher than I had it with my Hirschsprung's. I'd like to encourage all of you to keep searching for the right 'life rhythm' that will empower your lives. I believe there's an individual lifestyle out there for all of us to cope with our bodies and to achieve what we want in this life. Don't be afraid to get connected with doctors, hospitals, other parents, online, and through BGK. The more information you can obtain the better the decisions you make for your family.
>
> I'm looking forward to working with everyone at BGK and responding to the challenge of raising the public's awareness of The Bowel Group for Kids.

One woman researched her family history uncovering four generations and nine family members who were born with Hirschsprung's disease. Daphne herself was born with the condition as were two of her sons and her grandson, the youngest family member is her grandson who is seventeen years old and the oldest living member being her mother who is eighty seven and was only diagnosed at age twenty nine after years of bowel complications and difficulties that went unexplained.

The symptoms of Hirschsprung's disease in neonates include abdominal distension, vomiting and failure to pass meconium within 24-48 hours of birth (Ambartsumyan et al., 2020; Comes et al., 2021; Gorbatyuk, 2022; Thapar and Borrelli, 2018).

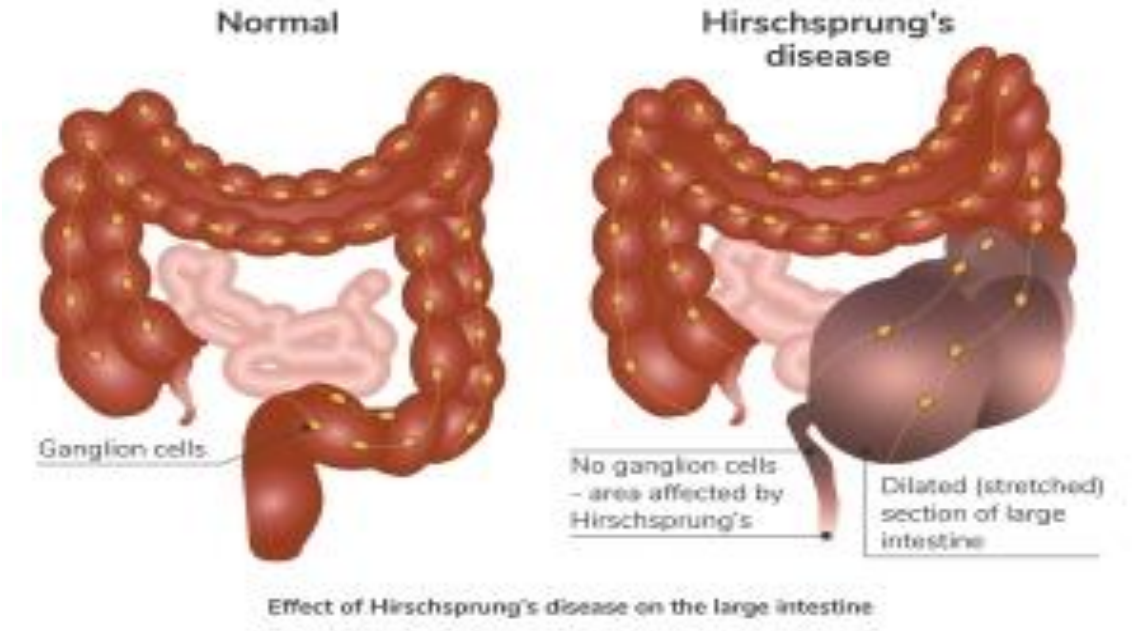


Figure 1: Normal colon verses colon showing Hirschsprung's disease

The illustration on the left depicts a normal large bowel (colon). The illustration on the right shows a narrowed section of colon at the end which is devoid of ganglion (nerve) cells. Without ganglion cells, peristalsis, the wave-like action that pushes stool through the bowel does not occur; resulting in a grossly dilated (stretched) section of colon above which occurs when a baby feeds and stool is prevented from passing through to the anus. (Hirschsprung's Disease | CSOR | NPEU, n.d.)

Imperforate anus affects the distal anus and ranges from minor and easily treated conditions of a membrane covering the anal opening, to more complex anomalies involving the rectum, urinary and genital tracts which may involve connecting fistulae. The latter has a poorer functional prognosis (Wood, Levitt. 2018).

Figure 2: Imperforate anus before and after corrective surgery

The illustration on the left depicts imperforate anus, closed at birth. The illustration on the right, shows the anal opening, corrected with surgery. (Imperforate Anus Information | Mount Sinai - New York, 2021)

The incidence of both HSCR and ARM is 1 in every 5,000 live births. In New South Wales in Australia, there are about twenty new cases diagnosed annually of Hirschsprung's disease and imperforate anus, so it is not unusual to find even the paediatric surgeons themselves may experience only a few cases each year. When one considers these twenty new cases may be spread across the entire state and between all the general practitioners, baby health clinics and paediatricians, it is easy to see the lack of experience is due to the rarity of the condition. There are three dedicated children's hospitals in New South Wales where all major paediatric surgery is performed. One of the paediatric hospitals is situated twenty-six kilometres from Sydney at Westmead, one in Randwick 6 kilometres southeast of Sydney and the third in Newcastle 170 kilometres from Sydney. Between these three hospitals there are about twenty paediatric surgeons servicing all cases in NSW. For families living in rural or remote areas of New South Wales, having a baby born with one of these conditions is especially difficult. Often families are separated for long periods of time whilst one parent remains in Sydney to care for the newborn undergoing major surgery, whilst the other parent manages the remaining family in their hometown, often hundreds of kilometres away. This additional stress on the family adds to the complexity of the situation. Once the baby recovers sufficiently from major surgery, they may be transferred to a rural hospital closer to the family home, but this also brings challenges if the staff are unfamiliar with these conditions. In our own situation,

I chose to remain in the tertiary hospital rather than transfer closer to home as I felt more confident in the care he was receiving with staff who knew him well.

These conditions are widely considered to be medical and technical challenges. The primary, life-saving focus in a newly diagnosed baby is to relieve a bowel obstruction or correct the anomaly to allow the passage of stool. What is not realised is that for some of these children, a normal functioning bowel can be very difficult to achieve, leading to the little-known social side of this issue following surgical correction of faecal incontinence or soiling, referred to as non-functional encopresis (Yilanli and Gokarakonda, 2022). This is especially important for a condition that is often lifelong, where the focus is placed heavily on the parents and the child to instil toilet training, toilet timing and bowel management, (Lotfollahzadeh et al., 2022) and the condition is especially stigmatised in the wider community.

The stigma associated with the condition is paradoxically increased because of the expectation of a cure. Upon learning that a friend or family member's baby has been diagnosed with one of these anomalies and is about to undergo major surgery, it is natural to assume that the baby will make a full recovery and then begin life like any other newborn. Whilst the medical care for these conditions in Australia is not in question in this thesis, it is societal expectations and understanding for children and their families living with faecal incontinence that lags behind. Faecal incontinence in children can pose great difficulties for parents and their affected children. If all avenues have been exhausted for surgical or medical intervention, then parents are often left to manage the problem alone, excluded from society, but the problem remains with devastating effects for the

child's development. Due to the rare nature of the condition, there is limited understanding of the surgical intervention or long-term management by clinicians outside of the major paediatric hospitals. The family general practitioner, baby health clinic nurse or clinicians at a local hospital are often in the difficult position of rarely, if at all seeing children born with anorectal malformations. Occasionally clinicians will recommend a support group to parents, but often this is an afterthought when all else has failed.

This thesis explores the impacts that support services can provide from a collective of shared experiences from people sharing the same experience and how an introduction to a support group and/or one of the newer hospital run paediatric bowel management clinics, ought to be integrated into clinical care at the time of diagnosis, forming a means of support when and if the parents feel they need it. This may be short term or longer until the child achieves continence or develops a system of self-management. Some families remain in a support group long after they need the support themselves to give back to the community and help support new families coming along.

My fourth child and only son Adam was born with Down syndrome and Hirschsprung's disease in 1989. We were told if he had been born five years earlier, it was unlikely surgery would have been offered, the only option being palliative support until he died, effectively solving the problem by allowing him to die when surgical intervention was possible. Surgery for Hirschsprung's disease had been successfully performed for many years but when combined with Down syndrome, it was largely uncharted waters. I describe Adam's history with the condition in the following chapter. As described in the prologue, I have been

involved for over 29 years with a peer support group for the families of children with Hirschsprung's disease or an anorectal malformation, which has brought together many formerly isolated parents to share their experiences and support each other with the goal of raising community awareness, advocating to improve the lives of affected children.

The importance of parent support when Hirschsprung's disease or an anorectal malformation is identified is of great significance due to the inability to screen for these conditions antenatally. Imperforate anus is often associated with other conditions that may be detected during the twenty week morphology scan. Once some of these other conditions are identified the clinician is alerted to the possibility the baby may have a cluster of anomalies known as VATER or VACTERL. No such opportunity exists for Hirschsprung's disease (Jakobson-Setton et al., 2015). Even in the neonate, it can be difficult to detect and is usually suspected when there is a delay in the passage of meconium within 24-48 hours after the baby is born (Buonpane, C., Lautz, T. B., and Hu, 2019). If the mother is discharged from hospital within that period, neonatal diagnosis can be further compromised.

While an imperforate anus may be suspected antenatally when any of the associated anomalies are detected during routine ultrasound, the presence or absence of an anus is not able to be confirmed until the baby is born and close inspection takes place. This contrasts with conditions such as Down syndrome where there is an array of prenatal testing routinely offered to women in Australia (*A Guide for Expectant Parents*, 2018). As early as ten weeks gestation, a non-invasive prenatal blood test (NIPT) can analyse both maternal and foetal cell-free

DNA circulating in maternal blood (Benn et al., 2013). If first line screening suspects an anomaly, then counselling and diagnostic testing such as chorionic villus sampling (CVS) or amniocentesis is recommended. If a positive antenatal diagnosis is made, this allows parents/carers time to absorb the information, have discussions with their team of clinicians and speak to their local Down Syndrome Association to gain insight into how it may affect their family and even speak to other parents if they choose to do so. This multidisciplinary approach to a prenatal diagnosis of Down syndrome offers families choices allowing them to decide if they wish to proceed with the pregnancy or terminate at an early stage. Prenatal diagnosis of Down syndrome is considered progress, allowing parents the choice of whether they wish to raise a child with a disability. Whilst many aspects of raising a child with Down syndrome can be discussed prenatally such as the likelihood the child with require additional educational services, including early intervention and further educational supports in schools, information relating to the need for neonatal intensive care admission or associated medical and surgical conditions are less clear. Some cardiac anomalies can be detected on ultrasound which allows greater opportunities to prepare for surgical intervention at birth, no such opportunity exists to prepare for the surgical intervention or long term consequences of Hirschsprung's disease or an anorectal malformation (Seither et al., 2021).

Despite attempts to recognise subtle signs of anorectal malformations during routine antenatal ultrasound, the only advances to date have been disappointing and merely alert the obstetric team to be aware of signs in the neonate such as monitoring for Hirschsprung's disease by the passage of meconium in the neonate or close inspection of the anal position or patency in imperforate anus.

The focus on diagnosis remains a purely surgical one. Although the incidence of Hirschsprung's disease is greater in babies born with Down syndrome, a prenatal diagnosis of Down syndrome is far more likely to elevate clinicians' concerns about cardiac anomalies, which – unlike Hirschsprung's disease can be detected on prenatal ultrasound (Stoll, Dott, Alembik, and Roth, 2015). In the vast majority of babies who are diagnosed with Hirschsprung's disease there was no abnormality of the foetal bowel detected during (Jakobson-Setton et al., 2015).

Most mothers in Australia undergo routine antenatal screening and are afforded excellent antenatal care in various forms such as through a GP obstetrician, midwifery care or private obstetricians, using shared care models or single care models for any of these clinicians (Government Department of Health, 2020; RANZCOG Committee, 2016). Being told their baby has been born with a bowel disorder despite all of this prenatal testing can be emotionally draining and come as a terrible shock. There has been little change in this over the last twenty years (Hinton, Locock, Anna-May, and Knight, 2018).

Rather than welcoming their new baby and proudly sharing their joy with family and friends, parents find themselves in a confusing whirlwind of unfamiliar hospitals, clinicians, the complex environments of neonatal intensive care units, and medical jargon used to describe their baby's condition that sound like a foreign language. Most babies, if they are in general good health and not considered an anaesthetic risk, will undergo major surgery within days or weeks of life. Babies considered stable enough following surgery are discharged either to a hospital closer to the family home to complete their recovery, or if they are well enough to be discharged home into the care of the family, most parents

leave the major paediatric hospital believing the worst is behind them. For many babies, this is the case, but for some, it marks the beginning of a long relationship with the paediatric team and many hospital admissions. Without appropriate support, parents are often left questioning what has gone wrong and if they or their child are the cause of their child's incontinence.

Appropriate support during the early days of diagnosis and prior to discharge in the neonatal period, from people who have experienced the same situations, can offer families support at this crucial time. Bringing a newborn baby home from hospital often presents a steep learning curve for families whose baby has a normal start to life. The support from family and friends is often difficult to understand when the baby has been born with the additional complexities of an anorectal malformation. Therefore, research is required to understand the services that are rarely studied or recognised, and the 'invisible' support that can transform the lives of children and families.

My significant original contribution to knowledge is activated by the privilege of being a part of so many families lives as they navigate the unknown world of paediatric surgery, postoperative care and the effects of the condition not only for their child, but siblings, grandparents and extended family. These rich associations from my experiences as a paediatric theatre nurse, an educator of future clinicians, being a mother of an affected child and cofounding the peer to peer support group have given me a unique perspective, beyond the medical model. Providing support to families in hospitals as they nurse their newborns following major bowel surgery, with stomas normally associated with bowel cancer in older adults; the many telephone conversations, hospital visits, family

picnics, parent and teacher conferences, letters received, questions raised on social media sites and the submission of parent stories for the support group newsletters 'Segments' have developed into a wealth of insight into problems and solutions I may have been unfamiliar with. I have watched babies thrive and grow into young adults who refuse to define themselves by the conditions they were born with and go on to live fulfilling lives.

Interestingly, when sharing their stories at Bowel Group for Kids annual conferences, some mothers relay a worrying memory of the early years in their child's life, concerned about the long-term psychological effects of their child being subjected to many doctors' consultations, examinations and invasive procedures. In contrast, the affected person, usually in their late teens to mid-thirties, often recollect on their early years in a very matter-of-fact way. They did not see themselves as different from their peers, and they just got on with life. Nevertheless, some children and young adults have damaging memories – or continue to have faecal incontinence and have suffered due to the stigmatising attitudes held around faecal incontinence in the general community – it can be seen as poor behaviour, attention seeking, or inadequate parental or self-care (Akin Sari 2014, Hamid et al., 2007).

Several older men aged in their fifth and sixth decade who were born with an anorectal malformation have contacted the Bowel Group for Kids, having lived in an era where such issues were never discussed until now. Most of these men had never spoken to anyone previously and through support and research, we have been able to uncover some of their old medical records, reigniting long-suppressed emotions. One gentleman, Greg Ryan, has taken the approach of

bringing his situation to light through the publication of an autobiography and speaking at conferences both in Australia and overseas in the hopes of educating the community about the long-term effects some people experience (Ryan, 2019). In his first book "A Secret Life: Surviving a Rare Congenital Condition" he describes the physical impact and shame of having imperforate anus.

> It is so personal and taboo in all cultures that our bowel functions are the last topic anyone wants to discuss. From my earliest memory I have chosen to keep my condition as my secret. Only my parents and my doctors were aware of my trauma. The thought that anyone else would know was too embarrassing and humiliating for me to bear (Ryan 2019, p 5).

In his book, Ryan outlines the personal impact of the social stigma associated around defecation, particularly faecal incontinence, which I explore in this thesis more generally through the experience of families of school-age children. Overcoming the stigma which appears to contaminate families of affected children, can seem at times to be an impossible task. However, peer support groups such as Bowel Group for Kids and lonely acts of witness such as those of Greg Ryan offer a glimpse of a future in which this may be possible. As a result of Greg Ryan's book and advocacy, Senator Anne Urquhart advocated in the Australian Federal Parliament for more support for people with imperforate anus and anorectal malformations, the first time this condition has ever been mentioned in parliament anywhere in the world. Senator Urquhart posted on Facebook with a link to her speech "We hear you; we are with you" (Senator Anne Urquhart, 2018). Currently, with the advent of greater awareness and

understanding of the often hidden disorders, support is provided for families much earlier with psychologists and clinicians all on board, and we hope never to see a repeat of this level of suffering for our children going forward.

When children born with anorectal malformations are adults, it can be difficult for parents who have been so closely involved with their child's life to step back and reflect, to enable and improve the experiences of those who follow. Recently I was contacted by a mother of six, seeking help for her 50-year-old son who was born with Hirschsprung's disease and has been struggling with difficulties evacuating his bowel for some time. Neither her or her son had found appropriate support or advice and so turned to a support group initially set up for new families. Speaking to this mother was so enlightening as she explained her son's early start to life. She lamented the lack of support but was relieved to speak to someone who understood the difficulties they experienced. Every new encounter adds to the richness of my understanding of the difficulties people face and how important it is to be able to communicate with someone who has lived through similar experiences. It is difficult to fully appreciate each person's perspective unless you have lived through similar experiences. This thesis draws on my original contribution to knowledge from the unique perspective as a nurse caring for these children, an educator of future clinicians, the mother of a child born with Hirschsprung's disease and co-founder of a support group for these families.

Hirschsprung's disease or an anorectal malformation all carry appreciable risks, both during and after surgery, and in their own right. I have witnessed the immense sadness when despite excellent care, babies and young children die from complications of Hirschsprung's disease or an anorectal malformation.

Fortunately, a death from complications of anorectal malformations are exceptions today.

From a professional point of view, I have been involved as part of the theatre team operating on these babies and young children and have had the opportunity to talk to parents before and after surgery to offer some small comfort from a parent's perspective in understanding the worry as they await a visit from the surgeons. In recent years, many clinicians have established bowel clinics for these children in the major paediatric hospitals, providing the opportunity for families to see surgeons, specialist nurses and allied health professionals during one visit. I have been invited to attend these clinics as a researcher and provide support for parents/carers. This is a vast improvement to the fragmented care that was the norm when my son was born 34 years ago when Individual appointments with each specialist were time consuming and lacked continuity amongst the team of clinicians.

Whilst there is a plethora of literature on the short-term outcomes of children born with Hirschsprung's disease or an anorectal malformation, thus literature mainly focuses on the biological features of Hirschsprung's disease and surgical procedures and treatment techniques for constipation. The literature on how the children and their families manage once the corrective surgery is over, is comparatively limited. What evidence there is suggests that there is a need for more consideration of how complex (and prolonged) after-care is for children who have undergone surgery for Hirschsprung's disease. In a study of the psychosocial functioning of nineteen adolescents who had neonatal surgery for Hirschsprung's disease and their families, Diseth et al (1997), found that the

parents were upset about performing procedures at home which were distressing for themselves and their child, and the young adolescents had little or no social support to guide them through incontinence, soiling and ridicule at school. A study of adults who had been born with a high anorectal atresia reported that adults with the condition were left to work out their own form of bowel management, often with poor results (Hassink et al 1996). The expectation of both surgeons and parents have evolved over time from child survival to the achievement of a good quality of life preferably continent of faeces, without the need for repeat or redo surgeries which are often unpredictable. A study of seventy survivors of surgery for Hirschsprung's disease in Helsinki, found that a high proportion of parents with children with bowel and/or bladder dysfunction suffered from anxiety. In the Helsinki study, it was the parent support groups rather than the professional psychological services that decreased their anxiety (Neuvonen I., Kyrklund J., Rintala P., and Pakarinen P., 2017). Additional stressors were poor organisation of transfers from paediatric services to adult services when the child entered adulthood. (Diseth, Bjørnland, Nøvik, and Emblem, 1997). Unfortunately, transition to adult services remains a difficult and. unresolved process to this day (Bitencourt et al., 2021; Nardone et al., 2020; Samarasinghe et al., 2020; Toulany et al., 2022).

Relationships have developed over time with like-minded parent support organisations around the world. This has provided insight into how they operate, what services they provide, how they are funded and if their services are integrated into clinical care. I have also met the organisers at professional conferences and experienced how they are supported by health professionals which seems to be the norm, particularly in Europe. A recent connection in

Columbia reminded me of how fortunate we are in Australia as they lacked funds for basic supportive care. The Bowel Group for Kids supported their endeavour to set up a peer to peer support group for families by sharing the resources we had developed over many years (Figueroa G Luis Mauricio and Soto Ch Mercy, 2022).

Clinicians have been donating their time and expertise for many years to support the hospitals in developing nations such as Nepal. Professor David Croaker a highly experienced Australian paediatric surgeon with a particular interest in Hirschsprung's disease and anorectal malformations has been visiting Nepal since 1997, first as a student, then a general practitioner and since 1988 as a paediatric surgeon. He has seen many changes In the country over the years, but it remains a challenge to ensure longevity in babies born with these rare conditions. Many countries still do not have the luxury of stoma appliances that we enjoy in Australia which are heavily subsidised by the government.

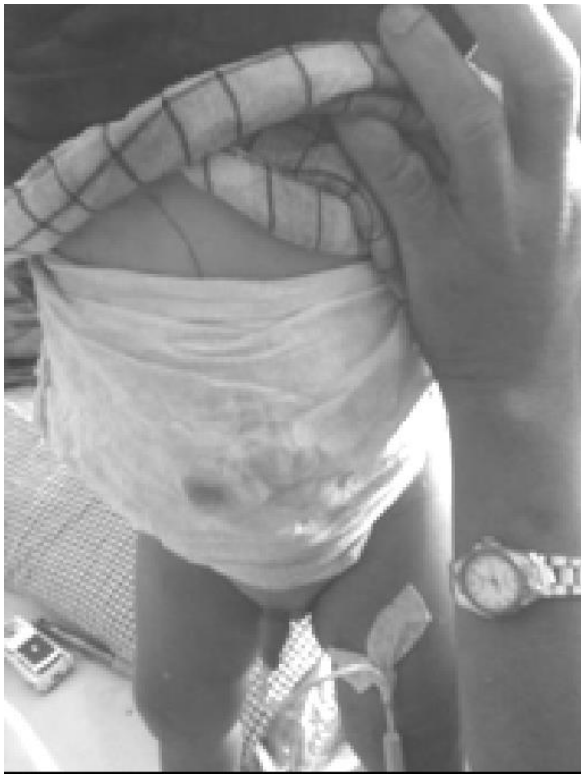


Figure 3: Toddler in Nepal wearing a cummerbund

The picture above shows a toddler in Nepal who had a stoma fashioned as a result of being born with Hirschsprung's disease. As Nepal does not have the benefit of stoma supplies, the parents use cotton binding in the form of a cummerbund to wrap around the child's abdomen to absorb faecal matter. These are then washed and reused. (Paediatric Surgery in Nepal, Professor Croaker David, 2011)

Most families find it cathartic writing their child's story. Several parents say they wished they had kept a diary of their baby's journey from birth to the present as they often forget the details as they move to the next stage. I often wished I had taken photographs of my son during his many hospital visits, not only for us, but for our now adult daughters to have a better understanding of that time. This may well reflect the disparity in memories of children and their parents, as our daughters have little memory of the times I was away from home with their brother in hospital. Our daughters tell me they have very little recall of what Adam went through and they also relay that they do not feel they missed out on our attention, although we have vivid memories of feeling torn between the needs of Adam and the needs of our girls. This was reiterated as a mother and adult daughter spoke at one of the Bowel Group for Kids annual conferences. The recollections of the early years with countless hours spent at doctors' appointments or hospital visits, was confined to the mother. The adult daughter was unaware her childhood had been any different to any of her peers (Beyond the Horizon, the Bowel Group for Kids Annual Conference, 2007)

Parents' concerns for their baby can be a rush of emotions on hearing the diagnosis shortly after birth, as they fast-forward through their child's life in their mind wondering how this condition will affect them, will they need further surgery, have ongoing continence issues, will they have friends, be accepted, be able to go to mainstream school, how will they broach the subject to a prospective

partner, will it affect their relationship, will they be able to have sex, have babies of their own, is it genetic? The questions can be all consuming and almost impossible to answer with certainty. Once they are assured of their baby's survival, the parents focus tends to move towards their child's quality of life. Surgery for these babies was pioneered around the middle of last century. During that time parental and clinician focuses were aligned to the baby's survival as mortality rates were higher. Mortality rates for babies born with Hirschsprung's disease or an anorectal malformation have improved considerably since the 1950s. Clinicians focus today is on diagnosis, extent of the condition, the general health of the baby, fitness for major surgery, appropriate surgical approach, timing, concerns over adverse risks such as enterocolitis or overwhelming sepsis and the urgency of the surgery. Parents on the other hand, expect that their baby will survive, so quality of life and knowing what the future holds has become paramount.

This is where support tailored to the family's needs can be invaluable. In the situation where the affected child is now an adult and announces they are pregnant; their parents are immediately taken back to their own experience and the joy of a new grandchild is mixed with concern that this could happen again. Many parents born with an anorectal malformation have relayed their mix of joy and trepidation when their baby is born as they await the early signs that the bowel is functioning normally. The first few bowel actions are met with much celebration, bringing strange looks from staff as this is something only their family would understand. Sadly, this is not always the case and some families have more than one child born with the very same condition they were, resurfacing

memories for grandparents and yet providing unique support for their child and grandchild.

The thesis explores the situation that develops when parents and the multidisciplinary team of clinicians come together through the birth of a baby born with Hirschsprung's disease or an anorectal malformation. The primary focus for clinicians at the time of a suspected anomaly is on accurate diagnosis, creating a plan of care and managing possible surgical/medical complications. Support for the parents is varied and dependent on several factors; for example, in Australia, major paediatric surgery is only performed in tertiary paediatric hospitals based in major cities. Specialist paediatric hospitals may be far from the family home making family support difficult. Parents and carers may have other children which can be a source of stress deciding if staying with the affected baby in hospital is plausible. English may not be the first language of parents or carers, and there is very little written material aimed at parents in languages other than English.[2]

Parents and carers may be offered a visit from a social worker to help them connect with services to manage other carer responsibilities with small children or other family members. Family and friends often find it difficult to support the parents due to the limited knowledge and/or understanding of Hirschsprung's disease or an anorectal malformation which are often seen as a birth defect that will be fixed with surgery in the early weeks of life. There is little comprehension of the impact removing often large segments of bowel can have on faecal

[2] An exception is the Spanish translation on the US KidsHealth.org site *Enfermade de Hirschsprung* which includes detailed and sensitive translations into Spanish, including audio Accessed 28 March 2021 from (https://kidshealth.org/es/parents/hirschsprung-esp.html). This should be the model for multilingual information about HD in other languages.

continence. This can add to the parents' stress as they continually explain the condition and the need for ongoing management, often dealing with unwelcome advice about toilet training.

Aims and Contributions

This doctoral research has three aims:

a) identify the impact for parents or carers of a child born with Hirschsprung's disease or an anorectal malformation.
b) identify the impact of peer support groups for families of children born with Hirschsprung's disease or an anorectal malformation.
c) identify the challenges children, young adults and their families face when dealing with faecal incontinence beyond societal expectations.

My significant original contribution to knowledge in this research is to contribute to a greater understanding of the support needs of families when a baby is born with Hirschsprung's disease or an anorectal malformation and further, when the child does not meet societal expectations for faecal continence, how that may impact on the school aged child. There is great potential for support groups and clinicians to work together to improve long-term outcomes for young people and adults born with Hirschsprung's disease or an anorectal malformation. Being told their baby must undergo major surgery is often overwhelming. In the standard surgical textbook on anorectal malformation, the authors of a chapter on family support noted that good relationships with support groups are critical to care and to knowledge generation (Jenetzky and Schwarzer 2006).

Outline

This thesis is divided into eight chapters.

- Chapter One provides insight into my own son Adam's journey from birth to thirty four years of age. This chapter describes the various surgical procedures such as the pull-through procedures, which will be referred to in passing throughout this thesis and the challenges we have faced.
- Chapter Two explores the stigma associated with faecal incontinence and reviews Goffman's theory in comparison to how stigma is used today.
- Chapter Three reviews the planning and processes necessary when travelling with a child who suffers from faecal incontinence.
- Chapter Four reviews the literature on support groups in general and focuses on the limited number of support groups globally that have developed specifically for Hirschsprung's disease or an anorectal malformation, setting this in the context of the data on long-term outcomes of the various surgical procedures.
- Chapter Five sets out the methodology for this thesis.
- Chapter Six explores parents' narratives of their personal experiences and the difficulties they have faced in a challenging system that has not caught up with the child's needs.
- Chapter Seven focuses on the Australian education system and how children with Hirschsprung's disease or an anorectal malformation are supported, included or excluded.
- Chapter Eight presents the results of studies into the understanding paediatric surgeons have about support groups, and some descriptive data on participants in the nascent self-help group the Bowel Group for Kids Incorporated. Finally the conclusion of the study points to further research

and the areas for improvement to help drive change for acceptance and inclusion for these children and their families.

The imperative of this doctoral research is to render the invisible visible. The goal is to speak the unspeakable, and care through the carelessness of medicalizing disability and impairment. Most importantly, this PhD offers research strategies – through stories, storying, and innovative writing modalities – to demonstrate how the marginal and marginalized can be rendered present, active, engaging and community-building.

CHAPTER 1: 'TAKE A WALK IN MY SHOES'.

Understanding the personal challenges faecal incontinence brings.

This chapter reveals the personal story behind this thesis: the experience of one family and a young boy with Hirschsprung's disease, who grew into a man. Elliott Mishler, (1984) talks of all narrative encounters between clinical experts and patients as a struggle between the "voice of the lifeworld" and the "voice of medicine". My work in this thesis will bring the voice of a life/world to transpose the voice of medicine. Adam is my son, but his experience is that of many children, and the experience I discuss of my family is that shared by many parents of children with this condition. In the prologue, I spoke of my professional work as a nurse caring for children born with Hirschsprung's disease or an anorectal malformation. My experience as a parent of a child with Hirschsprung's disease has shown me a completely new world – a familiar world was made unfamiliar and presented a unique opportunity to cofound a support group for these families. This thesis draws on all aspects of 34 years of experience as a nurse, an educator, the mother and cofounder of a support group to understand the complexities of raising a child born with a congenital bowel disorder, that despite corrective surgery, some of these children are unable to gain the continence required to fit into the world as we know it. In the early years sanctuary is found in the safety of their homes, but there comes a time when the child has to venture out into the world, a world that cannot understand the difficulties they face. It is the culmination of my unique experiences as a nurse, educator, mother and

supporter compared to the medical model, that forms my significant, original contribution to knowledge.

I set out the personal story of the need for, and the development of, a peer to peer support group against the background of dealing with a stigmatised disorder, one that has a complex and uncertain surgical treatment. Hirschsprung's disease and anorectal malformations are complex diseases which are difficult for the parents to explain to family and friends. These conditions are rare, most people in the community have never heard of them and they are often difficult to comprehend. Stone, (2018) explains how diseases have certain levels of prestige for example, breast cancer, childhood cancer and heart disease have a high disease prestige. These conditions are easily recognised in society and attract significant philanthropic support. Unfortunately, conditions such as Hirschsprung's disease or an anorectal malformation have a low level of disease prestige. This low level of disease prestige continues once the definitive surgery is over in the early years of life as it is considered the problem is now solved. Despite the best surgical advances and expertise, some children born with Hirschsprung's disease or an anorectal malformation fail to gain faecal continence by the accepted norm of around four years of age. For these children there is a greater need for clinicians and educators to help fill the void in validation and support. This research addresses an illness which is of low prestige, which causes a great deal of social suffering and is often considered a hidden disorder until the child attends preschool or school, and then the problem is exposed leading to a turbulent time during the child's formative years.

There are very few narrative studies on Hirschsprung's disease or anorectal malformations. This may be because the stories are challenging to tell, and the key symptoms of faecal incontinence are stigmatising and unpleasant. Nicole Murphy, a science teacher researched the condition after her son was born and wrote an informational resource guide for parents and doctors (Murphy, 2011). Greg Ryan's personal account of his own experience of being born with imperforate anus in the 1960s reveals his experience through deformity and deep depression and the need to keep his condition a secret for most of his life (Ryan, 2019).

1.1 The pre-history of Adam

I had been working as a registered nurse for fifteen years and was used to occupying a leadership role. I had managed a paediatric ward in the days when parents were not allowed to stay and had been in control as a perioperative nurse. When I became pregnant in 1988 at the age of 36 years with my son Adam - an unexpected surprise after a holiday to my parents on the Isle of Man – I expected to manage the pregnancy and raising of our infant with the same degree of organisational capability I enjoyed both in a professional capacity and in raising our three daughters, then aged just one, three, and five.

We live on a property that is located about 100 km from Sydney. Being a theatre nurse, I ran our home like clockwork. I was organised and was convinced preparation was the key to success during the birth of the new baby. The pregnancy was uneventful, which was strange given I had previously had multiple threatened and complete miscarriages and an ectopic pregnancy. At my first antenatal visit my obstetrician commented that the government was now offering

free chorionic villi sampling (CVS) to mothers over 35 years of age, and that this had to be performed no later than 11 weeks gestation. He asked if I wanted to do this. I knew CVS carried a risk of miscarriage and given my history of miscarriages, I dismissed it, convinced it was unlikely and satisfied we would manage whatever life threw at us, never truly believing anything untoward would happen. How wrong I was.

The prenatal testing would have most likely shown the baby I was carrying did in fact have Down syndrome. However, the biggest issue in his life has not been his intellectual disability. There are plenty of educational and support services for children and adults with Down syndrome. Managing Hirschsprung's disease on the other hand, has been a much tougher road to tread. And to date, there is no routine prenatal screening or testing that can detect Hirschsprung's disease. It is only able to be diagnosed after the baby starts feeding and fails to pass stool adequately that suspicion may arise that this could be an acute bowel obstruction (Amandullaevich and Danabaevich, 2022; Katz et al., 2017; Pan et al., 2022a).

1.2 "Some features that are worrying me".

After a long and difficult birth, Adam was finally born late on a Friday night. He was placed in the Special Care nursery as he had a little difficulty maintaining his body temperature. I was tired and I vaguely remembered a nurse waking me during the night and asking if anyone had spoken to me, I was so tired, I thought nothing of it at the time and slumped into a deep sleep like no other. In the morning, I was basking in the joy that it had taken us 13 years to have our first successful pregnancy, and now we were blessed with four beautiful children,

three girls and now a son, life could not get any better. I felt our lives were perfect and nothing could touch us.

I asked to see Adam in the Special Care nursery or for him to be brought to me. The response was always the same, full of reasons why this was not possible, none revealing the actual truth, all with good intentions to allay my fears but it simply deepened my concerns. 'He was tired; he was resting; the staff still had trouble maintaining his temperature, so he was being kept warm; I should just enjoy the rest while I could.' I was frustrated but it was not unusual in the late 1980s for babies to be placed in the Special Care nursery to allow both mother and baby to rest and recover.

As it was a Saturday, the paediatrician rostered for the weekend – a stranger to me – came to see me with a nurse. He was wearing jeans with his shirt only half tucked in, discomforting details for someone who was from an era of nursing where we identified and aligned professional pride with our neat, pristine uniforms. He said,

> I've been going over your baby for the last four hours and wanted to talk to you about some features that are worrying me.

Those are words no mother wants to hear. I was suddenly very focused and eager for an explanation, and any pain I was suffering disappeared in that one short sentence. I thought, who is this person coming here saying things like this about my baby? I sat up and demanded to know, "What features?"

The paediatrician proceeded to enumerate those features starting from the top of Adam's head and working his way down. By the time he got to the ears and mouth I stopped him, as my mind raced to identify the unmistakable features, I

said..."Are you trying to tell me you think my baby has Down syndrome?" He said…

> Sometimes you can look at a baby and say yes without hesitation. In your son's case I really can't be sure, but if you ask me what's in the bottom of my heart, (he put his hand on his heart) … yes, I think he does have Down syndrome.

His words and demeanour dispelled any fears I had of his ability to provide the very best of professional and compassionate care for my baby, I knew then he was a keeper.

Deathly silence filled the room. Strangely, my thoughts went to the other three mothers in the room who must have overheard what was going on from behind the curtains around my bed. I think this was the beginning of the confusion I grappled with for many years. Which hat was I wearing? Mother or nurse? Physically I was in my comfort zone, in a hospital where I worked and knew the staff and cared for patients. But I was now the one in the curtained bed, receiving bad news. I was the mother of a baby born with a disability and no road map to follow. I felt vulnerable, confused and in disbelief that my best laid plans were not being played out.

What really hit me was how vulnerable our patients and parents must feel. Knowing the system and the staff it was very hard to separate the two, especially now there was work to be done. So many thoughts went through my mind, I knew the common associated conditions and wanted answers and a definitive diagnosis.

In 1989, chromosome testing had to be sent to a specialist service in Sydney and often took six weeks to come back. How was I going to explain this to my husband Michael, our girls, our family, especially given there was no definitive diagnosis yet? I did not want other people to know what was happening until we knew for sure and were ready to discuss the situation. We had a lot to deal with and the last thing I wanted was lots of visitors with inevitable questions.

There were no mobile phones, no internet, our home phone had just been disconnected without warning by the provider due to an administrative error. I could not call Michael to warn him of the devastating news. I knew he would be coming in with our daughters, eager to see their new baby brother. Our youngest daughter shared her birthday with Adam. She was just two years old the day he was born, and the others were four and almost six. I could never have imagined how our lives were about to be turned upside down.

Thirty years later, I met Adam's paediatrician in the corridors of the Children's Hospital where we worked. We were chatting and he revealed that he felt deep regret over the way he told me his suspicions about Adam's diagnosis on that first day I met him. I was apparently the first mother he had delivered the news to on her own and he vowed never to do that again without support for the mother. His sense that he had clumsily left me alone with the diagnosis had plagued him his entire career, I had no idea he felt that way. He had in fact tried to delay telling me the diagnosis alone. He had tried to call Michael but because of the disconnected telephone, he had not been able to contact him to come in to be with me when he discussed his concerns about our baby. The paediatrician need not have spent all those years worrying about the way he had broken the

news to me. What had seemed like an egregious professional misstep to him was not something we had ever given thought to. We were focusing on the crisis at hand and understood it was just the way it was, in a way, to me it was better as there was no one to mind our three girls and they would have all been at the bedside when the news was delivered which could have made it so much worse. I was saddened to think he had carried that burden for so long and yet to our family, we referred to him as the 'conductor of the orchestra', the one person I could go to for help when things inevitably went wrong with Adam's care and he would ferry me out to whichever specialist Adam needed. Sadly our beloved paediatrician and close friend, died too young just a year ago, I am so grateful I had the opportunity to ease his mind at least giving him comfort for his final few years.

Michael arrived with the girls to see their baby brother. The curtains remained around me, while I tried to quietly tell Michael what was happening, the girls were excited and wanted to see Adam, but he remained isolated in the Special Care nursery. Somehow, we got through the first day, Michael took charge of the situation and organised the girls. I had been the organiser in the family, and now I was a distraught mother, yet struggling with the nurse inside me that wanted to take charge and control of this train wreck. When the paediatrician came in the next day and asked if I had any questions, I bombarded him with an extensive list I had made. Would Adam be able to walk? Talk? Go to school? Kick a ball? When would the results of the testing be back? How was his heart? Were there any other associated conditions? My obstetrician was clearly upset over the outcome, but I feel he must have been rather relieved his role did not extend to

explaining about Adam, once a baby is born they become the patient of the paediatric team.

The next week was a blur of waiting for the chromosome test to come back, and all the while I kept thinking the medical and nursing staff had it all wrong. I convinced myself Adam looked just like our three daughters. Our eldest daughter just about to turn seven years of age, brought me a card she had made with a Band-Aid stuck to it. She wrote,

"I know you are upset mummy, so this will make it better."

Adam remained in the Special Care Nursery for the next six days. He was tube fed and did not wake for feeds and I wanted so much to just go home and put this all behind us. By day six I'd had enough, I told the nursery staff I was taking him to my room and going to wait for him to wake for a feed. He weighed 3.8kg at birth so I knew he was not going to starve to death. Eventually he did wake; I fed him and announced that we were going home, confident this was all behind us. So off we went, our phone still not connected. The paediatrician had said to call him the next day to see if the chromosome results were back. I drove to a neighbour's place about 10 minutes away and asked to use their phone. The paediatrician told me the results were back and he was sorry to say, that, yes, Adam did in fact have Trisomy 21, the most common form of Down syndrome. I drove home at a loss for words and just broke down and cried when I saw Michael. Our poor girls must have been so confused.

Adam screamed all night. I did all the wrong things and offered a feed every time he woke. By morning, he had the most distended abdomen I have ever seen,

hard as a rock. He would not wake even when we bathed him. Oddly, there seemed to be no activity from his bowel, not even a single noise through the night. His extremities were turning blue, and his breathing was laboured. I knew we were in trouble.

As a mother, having medical knowledge can be a burden, occupying antithetical position to that of a mother. As a nurse, I immediately began to think of the mechanism of injury and was able to connect the dots very quickly and started eliminating the most serious issues until I narrowed his symptoms down to some of the most likely conclusions, none of which I wanted to be true. Stone, (2019) describes the blurring of roles and confusion of identity, especially when your child requires long term admissions to hospital. She expands on the difficulties experienced as a health professional such as fitting in, if you attend a mother's group with your baby and disclose you are a health professional, then you open the flood gates to members wanting medical advice. At the same time, you are disconnected from your own profession as you are now on 'the other side' struggling between being a parent to your child, but not being treated like a mother as everyone assumes you understand the situation. Often no explanations are offered as you are seen as 'able to cope because you are a nurse or doctor,' when all you want is to be a mother and be looked after the way you look after families in these situations. Well-meaning comments from people generally do not help, they simply retraumatise you. Comments such as…'good job you are a nurse, you know what to do', or 'God only gives you what you can handle', I found very difficult to deal with, but over time realised most people are uncomfortable and have no idea what to say, the most helpful are those who are simply there for you, no need for words, or they bring food, wash up or mind the

children so you can have a much needed shower, a place for solitude to shed a tear in private.

Realising I had to act, I rang the Special Care Nursery where Adam had been admitted and they suggested he may have a meconium plug and as I was a nurse, they advised me to insert a cotton bud into his anus and see if it released the obstruction. I was not comfortable with that and felt we were looking at something far more serious than a meconium plug, so we drove to the emergency department of the hospital where he was born 45 minutes from our home. On the way Adam vomited bright green bile and I knew then there was no mistaking it, this was an acute bowel obstruction. I started running through my mind the possible options. High on the list was a vague recollection of Hirschsprung's disease although I could never remember having nursed anyone with it. I knew we would not be coming home right away, we dropped two of our three daughters off on the way to hospital and took our two year old with us. Knowing our girls were well cared for helped us to focus on Adam, they stayed with my dearest friend who is more like a sister to me, having met during our nurses training in 1975, we remain forever close.

1.3 "An interesting case that you may never see again".

At the hospital, the paediatric registrar had overheard the conversation in the nursery and was expecting us, he took us straight in. Abdominal X-rays were taken, the registrar did a rectal exam which caused an explosion of faecal matter and confirmed the suspicions of Hirschsprung's, Unbeknownst to me, our paediatrician had his concerns when we were still in hospital and had taken abdominal X-rays at a few days old and had been monitoring Adam's girth

measurements. The comparison was dramatic. Adam's colon was so distended it filled his entire little abdomen which was pressing on the diaphragm causing difficulty breathing known as dyspnoea and his extremities to have a bluish hue due to lack of oxygen known as cyanosis.

The registrar left the examination room and spoke to a group of young doctors waiting just outside our room. I overheard him telling them:

> You should come and have a look at this baby, he's such an interesting case and you may never see this again.

I wanted to pick Adam up and run out of there. From a nurse educator perspective, I knew that his comments were not intended to cause harm, but the effect is very different from a parent's perspective though. It is so enlightening when you are a patient or parent of a sick baby, nothing in clinical training prepares you to understand how people can be affected by our comments like living the experience from the 'other side' (O'Keeffe, 2014).

Adam and I went by ambulance to the Children's Hospital in Sydney about an hour away. It was to be the start of a lifelong association with the hospital which moved when Adam turned 18 years of age to the adult section of the hospital. It was pouring with rain. The ambulance ride from Camden was long and cold. A nurse sat with me in the back of the ambulance, and we made small talk, more like two nurses escorting a baby, rather than a mother and a nurse. I wished I could switch off and just be a mother, but I was afraid if I became 'just a mother' I would have no say, I would be too emotional and lose control, this was my way of keeping my emotions in check, but how I longed to be comforted as a mother.

In the whirlwind of the emergency department, the most distressing issues I faced were the seemingly small aspects of emergency care, the necessary evils like accessing a vein. Adam was very small and trying to cannulate a neonate is far from easy. In the end the only success they had after multiple attempts was in his foot. He was screaming and I found it intensely distressing to be so helpless and so confused as to why this upset me so much. After all I was often the nurse doing these things and I did them calmly and professionally. Now I was a mess. My mind kept wandering to the babies I had cared for. Had I recognised how distressing the parents found these necessary procedures? Was I compassionate enough? I always prided myself on never seeing a patient as 'just another case' or 'the appendix in bed six,' but I really had no idea what it felt like to be a parent handing over your neonate to a stranger. One thing was for sure, this experience would ensure I paid more attention to the parents in future, that was, if I was ever going to return to nursing.

We were admitted to the Paediatric Intensive Care Unit (PICU). Michael had picked up the girls and driven down to be with us. The poor girls were confused; I recall their little faces looking up at me saying…

> but you said you would be coming home with the baby after four days, remember you drew it on the calendar, why are we here, why isn't he coming home, what is wrong with him?

I had a lot to learn about being more flexible. Strangely, the area of theatre nursing I loved most was emergencies. I loved the unpredictable, patient's lives hanging in the balance, caesarean sections where the baby was in distress, the rush of snapping into action, quick thinking and changing situations. My actions and directions to staff had to be precise and informed. I could not make a mistake

or delay, or we could lose the patient or baby on the table. And yet here I was feeling very vulnerable in this unpredictable predicament. Being a parent of a child in the intensive care unit or recovery unit after major surgery is an experience most parents who are clinicians will never forget and one that shapes future practice (Janvier et al., 2016).

Adam seemed so big in the PICU amongst the premature babies I had been used to. Then came the heartbreaking decision to leave him. No matter how much I longed to remain with my baby, I also needed to go home with our girls and comfort them. I never realised how difficult it can be as a mother to prioritise your young children who are frightened and bewildered, when all you want to do is fall in a heap yourself, but you just have to pick yourself up and soldier on.

Although we lived over an hour away, I could at least drive in every day to be with Adam after dropping the girls at pre-school and school and with the support of friends helping to mind our two-year-old. But what of people who lived in rural or remote areas, their decision to stay or go home was made so much more difficult. Adam went through a barrage of tests over the following week: abdominal and cardiothoracic scans, X-rays, endless blood tests. My head was spinning from all the different clinicians who came and went. Fortunately, he did not have any cardiac problems, but a rectal biopsy confirmed earlier suspicions of Hirschsprung's disease. Surgery was planned for 27 April 1989, the day after our eldest daughter had her seventh birthday. Michael understood how torn I was between being at home with our three girls or going to the hospital to be with Adam, the reality was, he was at work, there was no parental or compassionate leave available for men in those days, and there was little I could

do for Adam. I was unable to feed him due to the bowel obstruction, he had a rectal tube in place to deflate his abdomen and he was receiving nutrients via an intravenous infusion. Michael explained how helpful our eldest had been and it would be unfair to her to be away on her birthday, so naturally I stayed home and I must have bought everything she thought of for her birthday in some distorted way of making up for the trauma they had all endured.

1.4 The surgical years

The following day, when Adam was 13 days old, he had a rectosigmoidectomy and a colostomy was formed. I asked the surgeon about primary pull-through to avoid a stoma (Teitelbaum and Coran, 2003). A primary pull-through procedure was routine in adults so why should it not be for a neonate? He explained that sufficiently small sized instrumentation was not available in Australia at that time and the stoma would allow the bowel to recover before re-joining it. The routine surgical procedure in the 1980s in Australia for this condition in a neonate was removal of the affected colon and formation of a colostomy This was an improvement from an earlier three stage procedure of not closing the stoma until it was sure the re-joining of the bowel had healed.

I was discovering more about the complex identity shifts between being a nurse and being a mother of a child with a chronic condition. There was always this frustrating opinion from the hospital staff and people who knew us, 'oh well, mum's a registered nurse so she will be OK, or she can manage.' I somehow remained very focused and in nurse mode right up until the time they brought my little baby out into recovery following his first major surgery at just 13 days of age. Although I knew what to expect, seeing my baby bearing a colostomy

which took up his entire abdomen with tubes everywhere was a shock for which I was unprepared. The mother in me broke down. I called Michael who was at work. These were the days before paid parental leave, and so taking time off to be with your baby in hospital was not an option for men. As this was familiar territory for me, I had assured Michael that morning I would be fine. Fortunately, he was able to leave work and be with me in the hospital for a while before returning to work.

Figure 4: Child's abdomen wearing a colostomy bag and anatomical placement.

The illustration on the left depicts a child's abdomen with a colostomy on the left with a stoma bag in place to collect faecal matter. The illustration on the right shows the stoma placement in the colon.

Down syndrome was the least of our worries. Down syndrome for us meant that it took Adam a bit longer to reach each stage in life, and once he did, he stayed in each stage much longer. Despite his slow progress, he was an absolute delight. Adam fed, slept, and seemed to have awareness of life around him and the girls delighted in their new baby brother, oblivious to his disabilities. When he was around fourteen months of age he would reach the milestone to undergo the pull through procedure and close the colostomy. I remember saying to friends at

preschool as we said our farewells for the Christmas break, "we just have to get through this next operation and then he will be fixed."

How naïve I was. I have since learnt not to mention the 'F' (fixed) word. We now talk in terms of manageability. The thought had crossed my mind, this first year has been so trouble free with a stoma, I actually found changing a colostomy bag much easier than changing nappies, perhaps we should leave well alone, but the aim was always to move towards 'normality' a normal functioning bowel like most people had, after all one had to conform to societal norms. On reflection, how far removed was this from the attempts to normalise victims of the Thalidomide disaster in the 1960s and 1970s with artificial limbs that failed to live up to their expectations (Blue, 2021)? The pursuit of normality began. We were introduced to a new paediatric surgeon who focused on Hirschsprung's disease to perform the corrective surgery, there were only three paediatric surgeons at the Children's Hospital at the time so each one operated on one rare condition to maximise experience, one did Hirschsprung's, another imperforate anus and the other did any chest operations such as trachea-oesophageal fistulas. I met with Stomal Therapy Nurses and have enjoyed a long and wonderful relationship with them to this day. They have been a lifeline on the end of the phone, email or each time Adam was in hospital. I have presented talks for many years to nurses considering stoma nursing as a speciality to help them understand the needs of children with stomas.

At the age of 14 months Adam had a Duhamel's procedure performed and the colostomy was closed. Little did we know our life was about to detour yet again. What followed were years of obstructions, bowel washouts, evacuation

procedures all in an attempt to get his bowel working normally. It all proved to no avail. When he was 5 years old the decision was made to try a new procedure, the antegrade colonic enema (ACE) which consisted of inserting a Bard gastrostomy button into the caecum to perform antegrade washouts.

Figure 5: Caecostomy with tubing and anatomical position
This illustration shows a Caecostomy with tubing connected to a fluid filled bag for enema administration.

'This image has been removed due to copyright restriction. Available online from: https://uihc.org/childrens/educational-resources/cecostomy-tube-care-pediatrics

As there is no natural opening in the right side of the colon, a surgical opening is created to allow the antegrade colonic enema (ACE) to be administered. there are various methods using different connections to insert tubing which is connected to a fluid filled bag. The depiction above shows one type of ACE procedure.

There were only two children who had had this procedure performed at the main Children's Hospital in Camperdown where I had spent part of my theatre training. Both these children were born with Hirschsprung's disease but neither of them had Down syndrome and both were much older than Adam. We were embarking on unknown territory. This was a first for the Children's Hospital at Randwick and for his surgeon.

A roundtable meeting was organised with me, Adam's gastroenterologist and his team of paediatric surgeons, to determine the best course of action. The options were to go back to a stoma or try this new ACE procedure. The decision was

made to do a barium enema first so we had a baseline image of Adam's bowel before proceeding to either option. The barium enema showed a hugely distended colon with no hope of evacuating faeces. The decision was made to try the ACE procedure, I still had niggling thoughts of whether this was the right decision, or should we just go back to a stoma, but it was thought to be a backward step and the gastroenterologist felt he would not cope with a stoma as he was a very active child. The surgery was uneventful, and we started the washout regime after a few weeks when everything had healed. The idea was to pour saline solution in through the Bard button while Adam sat on the toilet with the hope that it would clean out the colon within half an hour, leaving him clean until the next washout. It sounded great in theory. I remember a Stomal Therapy Nurse coming to assist me at home when we started the first washout, he had never seen one before so we were learning together. We poured a litre of normal saline in through Adam's Bard button and waited and waited and waited, the nurse had to leave as it was taking too long. Twenty-four hours later, I was still waiting as Adam who was only 11kg waddled around with all this fluid sloshing around in his bowel. Eventually it started to trickle out, but it took days for him to evacuate it all. Over the next year we tried everything: laxative drops which caused intense pain; osmotic bowel washout preparations; doing it in the bath; at night; in the morning, Unfortunately, despite our best efforts, the procedure never functioned for Adam in the way in which it was intended.

Another meeting and a second barium enema were organised for comparison. The radiologist compared the results of the pre and post ACE procedure films and said:

> If there was no date on the films, I wouldn't be able to distinguish between the two, nothing had changed.

I felt so defeated when I saw the barium enema results, and knew it meant we were headed for a permanent colostomy. We had fears about how Adam would respond. He was such a lively little fellow; he had started walking at three and a half and once he started, we could not keep up with him. How was a stoma going to fit with such an active child? This had been a concern voiced by the doctors in our earlier meetings, but we had no choice. Our lives had become impossible, we were trying to lead a normal life in a very abnormal, socially unacceptable situation.

Importantly today, the patient experience is being gradually introduced into medical school curriculum. I was fortunate to be involved in one such program where I teach medical students. Rather than them listening to lectures, the students were given a list of consenting patients to visit in their homes over several sessions to gain insights into their lived experiences. This inclusion of the patient experience to assess the goals and the quality of patient care is invaluable to understanding complex care (Gaille, M. 2019).

1.5 Living with Hirschsprung's disease

Faecal soiling is the most distressing complication of Hirschsprung's disease for families and for children (Athanasakos et al., 2006; Doolin and Richards, 2021; Monica Carissa et al., 2022; Mahmud et al., 2022; Xie et al., 2022; Zhang et al., 2022). Yet there has been very scant coverage on this in the medical and surgical literature that focuses on functional outcomes (Ivana et al., 2021; Kandil Ibrahim

et al., 2021; Liu et al., 2020; Mudasir A et al., 2021; ShR, n.d.; Teitelbaum et al., 2000; Wesson and Lopez, 2020; Xie et al., 2022). The narrative turn in medicine has not yet turned to capturing the narratives of parents of a young child with faecal soiling from Hirschsprung's disease. Accounts online tend to focus on inspiring stories of survivorship[3].

The family impact of Adam's faecal incontinence as a young child around the period of the surgery discussed in the previous section was of being constantly on the edge of not-coping. We could not go out as a family, one of us had to stay home with Adam as he was incontinent of faeces all the time. We would get ready to go out and he would need bathing just as we were about to leave. I remember I always bought long pants with elastic at the ankles for Adam in an attempt to contain the liquid stools he would pass that would go straight through disposable nappies. One morning, I was rushing to get the girls to school and just as we were getting in the car Adam vomited and was incontinent all over one of the girls. I just sat on the floor tears welling up for the dreadful predicament we were in. It was devastating trying to manage and so unfair on the girls who never complained, but 'mother guilt' was forever present.

One day we came to visit a friend in the Children's Hospital and took the children up to McDonalds in Randwick for a rare treat. Adam did not eat anything from there, but the girls enjoyed it. When we were about to leave to go back to the hospital, Michael picked Adam up and he was incontinent of faeces all over him. It cascaded everywhere. I took Adam to the disabled toilet to clean him up, only

[3] For example, those on the Children's Hospital of Pennsylvania "Patient Stories" website, accessed 7 July 2015 from https://doi.org/10.1007/s00383-006-1651-6

to find that I had left the basket with all his supplies in the car. After using all the paper in the bathroom, I faced the problem of a child with no usable clothing who could not leave McDonalds without some form of protection against another episode of faecal incontinence. I had a moment of inspiration: in operating theatres we always put spare bin liners under the bin being used for quick changes. I looked in the disabled toilet's garbage bin, sure enough they had done the same. So, Adam came out of the bathroom wearing a new style of pants – I had punched two holes in the bottom of the garbage bag for his feet to go through and pulled the garbage bag up to his waist. At least we would be able to get to the hospital with some form of protection where I could bathe him as the staff knew him well.

In March 1995, Adam had another major operation to remove the left side of his colon (a Hartmann's procedure) and a new colostomy formed. We thought that would be it: we viewed going back to a stoma as a last resort. How wrong we were. What followed were more years of stoma prolapses, attempted prolapse repairs and obstructions. Every time we drove to the Children's Hospital for another admission, as we drove past the airport, he knew where he was going and would start crying and pleading, 'no hospital, no tube' meaning the nasogastric tube which he hated. I had to keep watch through the rear vision mirror as the child booster seats in the 1990s did not include a crotch strap so Adam used to wriggle down in an attempt to escape the impending doom he must have felt and ended up on the floor of the back seat many times.

1.6 Adam's reaction to stoma surgery, aged 5 years.

As for how Adam reacted to all this invasive surgery, I will never forget the day I bathed him at home after the colostomy was formed when he was 5 years old. He looked down at his little tummy, pointed to the stoma and cried. What had we done? And yet, what choice did we have? Gradually he came to accept it and now, at 34 years of age, he has learnt to change the bag and base plate himself. He needs supervision to ensure he has it on properly, but he is fiercely independent and insists, "I do it myself."

Childcare or afterschool care was always challenging, there were limits on what was considered within the realms of what they were willing to accept and adapt to allow a child to attend. I ran into these issues many time throughout Adam's early life, Adam had started attending an after school care program where our three daughters attended when I was working at the Children's Hospital, my shift was 10am to 6pm and the hospital was over an hour away. These hours allowed me to take everyone to school in the morning, but I could not be home in time for them in the afternoon and we had no family to assist at that time. One day I received a call from the owner of the day care centre who informed me Adam could no longer attend, their reasoning was that other parents did not like the risk their children might see his stoma bag or what if it leaked. I sensed it was more likely the staff who had a problem with the situation than parents whose children attended as it was unlikely they knew about his condition other than he had Down syndrome. Amazingly, one of the nurses I worked with heard about the situation and offered to mind Adam after school. I was cautious and explained he had a stoma, and it may leak, she brushed it off and said there were several of her family members who were born with Hirschsprung's and a stoma did not faze her at all. They also had an adult cousin who was born with Down syndrome

but they never found out if he had Hirschsprung's, I was so relieved, not only by her offer, but her ability to make his situation inconsequential, where others saw it as a major problem. I was intrigued about her story and as Hirschsprung's disease is more prevalent in Down syndrome, I found it hard to believe her cousin escaped the condition. As I was working at the Children's hospital at the time and involved with research with my first supervisor, I asked the staff in Medical Records if they could search for this man's records. As the hospital had relocated many years earlier, any old records were archived offsite. The staff called me and I could sense the excitement in their voices, they found the records, a small tattered brown file. When I collected them, I read the small file and sure enough, it showed he had a rectal biopsy as a baby which confirmed Hirschsprung's, but no surgery was offered as he was not expected to live! It gave me great pleasure to hand the file over to the family.

So began a 'new normal' in our lives again. I must admit we had not realised how abnormal it had become. We could now plan and take the entire family out together. For unknown reasons, around the age of eleven, Adam deteriorated neurologically. He underwent many outpatient tests and consultations, some with psychiatrists who we found were less than helpful. Often the consultation would involve a couple of simple questions like "does he flap his hands?" when I answered no, I was told then no, he is not autistic, and that was that. No discussion, no deeper analysis of his signs and symptoms, it was simply a case of, if you cannot tick that box then you do not fit the profile, so that was the end of the consultation. Often these cost between $250 to $400 and gave us no clue as to what was causing this rapid decline. He was later diagnosed with autism

after weeks of observation by a clinician at home, in school and in the clinical environment.

Following a three-day hospital stay at the children's hospital, his neurologist performed a barrage of tests, lumbar puncture, CT scans and blood tests. The underlying cause was eventually revealed as severe Vitamin B12 deficiency. This was thought to be due partly to self-imposed dietary restrictions and partly to ongoing inflammation of the first part of the small bowel, the ileum where Vitamin B12 is absorbed, using intrinsic factor secreted by gastric parietal cells. B12 deficiency has a range of symptoms, including peripheral neurological impairment, neurocognitive impairment, and megaloblastic anaemia (Green, 2017). The consequences of late diagnosis can be dire. Adam began a trial at the hospital of Vitamin B12 via a nasal spray, but the results were not successful. He has been having three monthly injections of Vitamin B12 and blood tests to monitor levels ever since. Unfortunately, due to the length of time it took to obtain an accurate and correct diagnosis, Adam suffered some neurological impairment that is now permanent.

1.7 Transition to adulthood

At age 18 Adam ran into trouble again and needed further surgery, the stoma was prolapsing constantly and his peristomal skin was grossly excoriated. The decision was made to relocate the stoma which entailed further major abdominal surgery. NSW Health was supposed to have a transition program to guide families

from paediatric to adult hospitals in place, but this never eventuated in our case. It was seven years later that the Agency for Clinical Innovation released guidelines on principles of transition of young people to adult care, but in practice this remains very disjointed (Brodie, 2014).

At eighteen years of age, Adam was considered too old for the Children's Hospital. Despite a long association with the hospital both as a nurse, a parent and running the support group, letters of support from his surgeons, we were discharged from paediatric care. Moving to the hospital's adult service was a very different experience. We thought we had everything in hand. His adult surgeon worked with our paediatric surgeon as they performed the operation together.

The missing person in all this planning was Adam. We could not prepare him for the operation or explain why he would experience so much pain. His surgeons had worked out a plan that Adam would remain in ICU postoperatively as he had no concept of what was happening, I now know we ought to have discussed these plans in detail with the anaesthetist before admission. I knew as soon as the anaesthetist did his flying preoperative visit in the anaesthetic bay, we were headed for trouble. I did not know him and he did not wait to listen to the plans and dismissed any suggestion of Adam going to ICU postoperatively. Here I was faced with a perpetual problem, as a health professional you know the system, you try to be the 'good parent' otherwise you run the risk of being labelled as 'difficult, that demanding mother… who does she think she is? We make the decisions.' We waited for hours for the surgeons to emerge, all had gone well but the remainder of his large bowel was redundant and had to be removed, so he had effectively lost all his large bowel over multiple operations between the ages

of five and eighteen years and now had a permanent ileostomy with no way back, it could not be reversed. How I wished I had been strong enough when he was a baby and said 'enough, no more, let's leave well alone'. Whether this would have avoided further surgery we will never know. In recovery I again asked if he was going to ICU, tried to explain the plans but was told no, he was being returned to the ward.

Postoperatively, Adam struggled as I had feared. It was heart wrenching to see him suffer, unable to comprehend why we were doing this to him. It felt like a battle all the way. I had to plead my point that Adam could not stay in hospital alone. He was non-verbal and I had to stay with him, as was always the case in paediatrics. But this was adult care. It was different. No parents or carers ever stayed with the patient. Eventually they found a chair for me to sleep on in his room. It was exhausting both physically and emotionally.

The next couple of days were the most distressing. Adam suffered in pain and despite pain relief, nothing settled him. He pulled out his intravenous line which was the only means of administering pain relief and fluids. The anaesthetist on the pain team came around and said to me, you know we have to really question when you see a young person suffering like this, 'Just because we can, should we?' I knew only too well what he meant, but at what point do you say enough is enough. It is hard enough to make those kinds of decisions in the neonatal period as to whether a child should live or die, but we had come this far, there was no turning back and if we could have had a crystal ball when he was born, would it have changed anything? I really doubt it. I knew the pain team anaesthetist was struggling with the services available, as opposed to the need

for Adam to be cared for in a paediatric sense. When Adam was in the Children's hospital, anything invasive or distressing such as central lines, tubes or MRIs would be performed under sedation or anaesthetic. No such provisions were made for people who chronologically were deemed adults, yet developmentally remained childlike. Just because we have the ability to perform surgeries and procedures which have no doubt prolonged the lives of children like Adam into adulthood, the support services particularly within adult hospitals have not kept up with the medical and surgical advances resulting in a failure to meet their functional capacity. Without the supportive environment acknowledging the inclusion of parents to accompany their adult child and the transition of supports carried through from paediatrics to adult hospitals, we are failing these families, and the young adults are suffering as a result. This has to change.

By the third postoperative day, I knew Adam was in trouble. He was not able to take deep breaths because of the intense abdominal pain. His oxygen saturations (SpO2%) which should have been above 95%, read around 75%. The nurse shrugged it off as probably a faulty probe, but his symptoms told me otherwise and I insisted he call for help. Adam was scheduled to return to theatre to have a central line put in to replace the one he pulled out. As we approached the theatre flap doors, he screamed and pulled back in the bed. I then encountered a nurse who tried to make me leave Adam despite his evident terror, she announced, mothers were not permitted in theatre. I was furious, as a theatre nurse I would never do this to any patient who was vulnerable, but as a mother, I held no court in this arena. This again shows the stark contrast between paediatric and adult hospitals. A person may be an adult chronologically, but not developmentally, and in those cases, they will need the support of a parent or

carer as they cannot possibly understand what is happening and are often terrified. Yet another battle to overcome, I was exhausted from constantly advocating, but thankfully the anaesthetist overheard the commotion and came out and let me stay throughout the entire procedure.

Postoperatively in the recovery room Adam deteriorated further and the decision was made to intubate him, put him on life support and transfer him to ICU. The trauma of the past few days had taken its toll and I was shattered, but the one shining star was that Adam was finally where he should have been immediately postoperatively, in ICU. If he had gone there as planned, it is most likely he would have avoided the trauma of the past few days. That was the first time I relaxed, once he was ventilated in ICU, I knew he was no longer suffering and would receive the one-to-one care he needed in those initial days.

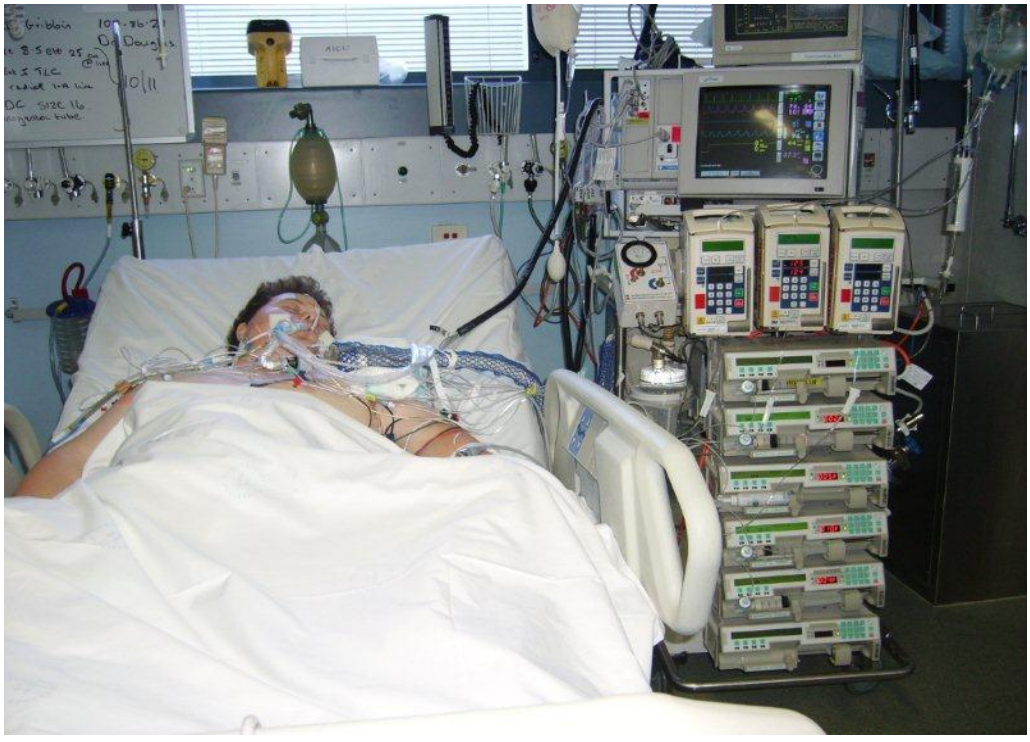


Figure 6: my son Adam in ICU aged 18 years.

Adam in ICU on life support aged 18 years after more major abdominal surgery to remove the remainder of his colon.

A few days later, as I was walking out of ICU, I overheard one of the female intensivists call out 'we're extubating the Downs boy today.' I stopped dead in my tracks and for a moment contemplated whether this was a battle worth fighting. To a parent whose child had been through so much it was yet one more battle to add to the list. I took a deep breath and walked out of the ICU, reminding myself what I often said to our girls, 'pick your battles'.

Ingstad and Christie (2001) point out the transformative effect on clinicians of experiencing illness in hospital. All members of the healthcare team need to be parents, patients or have someone they care for in hospital at some stage. We can teach ethics and instil compassion to our students, but there is nothing like being on the receiving end to realise how our passing remarks can cut so deep when people are at their most vulnerable.

A few days later I spoke to the next round of intensivists as they discussed their plans for Adam. He would remain on a ventilator for one more day, before reviewing if he were ready to be extubated. I questioned that the alternative – to use a full-faced mask for non-invasive positive pressure ventilation – would have been frightening and intolerable for him. Although there is no research comparing the psychological impact of invasive and non-invasive mechanical ventilation, it seemed likely to me that the full-face mask would be traumatising for Adam, especially given he could not cope with a normal Hudson face mask. They assured me they would do their rounds at 8am so I made sure I was there the next morning to hear their plans. Unfortunately, the intensivist who had made the insensitive remark was rostered on that next morning. When I arrived, she had already extubated Adam and he was terrified as she tried to put the full-face mask on his face. I spoke to her about the team's plan, and she dismissed my

concerns and simply said…'well if he won't breathe up well, we will have to intubate him again,' as though he was deliberately being difficult. The ICU nurse and I encourage Adam to breathe well without the mask and somehow, we got through the day.

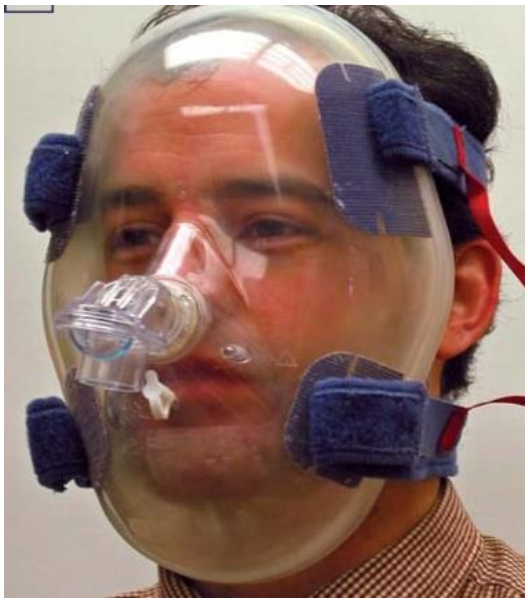


Figure 7: Example of a full-face mask

Example of a non-invasive positive-pressure ventilation (NPPV) full-face mask on a patient. Retrieved from https://www.degruyter.com/document/doi/10.7556/jaoa.2007.107.4.148/html

Adam was able to return to the ward after a few days and we eventually made it home, exhausted and battle weary from the experience.

1.8 Adam today

Knowing Adam had a life-threatening bowel disorder that required major surgery, put a new perspective on his Down syndrome. It has never been a major issue since birth. We just see him as 'Adam.' Obviously as he gets older the impact of his intellectual impairment is more worrying as we face the prospect of what will happen to him when we are no longer able to care for him, but we take the view that we must take each day as it comes, it is difficult to plan for the unknown. Adam is known as a gentle giant and loves being with his family especially now,

he has two little nieces and a nephew to play with. They are so refreshing as they just accept him for who he is, and they always include him in their games calling out 'come on Uncle!'.

Occasionally I wonder: what if he were born now? Would things be different? Would he have been saved the years of hospitalisations, operations and procedures; would he have a stoma? The advances, research and support for Hirschsprung's disease and anorectal malformations has grown globally and I have been fortunate to have been a small part of the research through one of my past supervisors and colleague. Despite this, there are still cases where the children born with these rare conditions have complications or comorbidities and things do not go as planned. We know so much more now, but making decisions in the flurry of the moment, and without access to a knowledgeable peer, is always difficult. Had we known then what we know now, we probably would never have taken that first step of closing Adam's initial colostomy at 14 months of age. But hindsight is an impossible guide. Like all parents in that situation, we chose what we thought was for the best, and we made the decisions with the best information available at the time. As I discuss through this thesis, in retrospect the knowledge we had was limited, especially about the lived experience of children and families with Hirschsprung's disease or anorectal malformations, we were picking it up as we went along. As there was then, there remains, a need for support for new families from those who have walked this lonely path before, to assist them by providing insight, support and understanding for vulnerable children and their families.

1.9 Socialising when your child is incontinent of faeces, facing the reality of an invisible disorder.

Having an invisible disability in one sense protects the affected person from constant gazes or questioning, and yet the invisibility of a disorder fails to provide any form of support of understanding when needed. There has been growth in the area of raising awareness that not all disabilities are visible. Whilst this is a welcome change to the long held perception that unless one can see a person's disability, then just how disabled can they possibly be? What is clear in the literature reviewed, is the term 'invisible disability' is largely restricted to mental health, Autism, sensory impairment such as hearing or vision loss, or certain conditions that are intermittent such as arthritis or chronic pain. (Aspland. C, 2021; Carlisle, 2022; Evan, 2020; Folkenroth-Hess, 2021). What is missing in the literature in relation to this thesis is the inclusion of faecal incontinence when hidden or invisible disabilities are discussed. It is unclear if this is simply an area poorly understood particularly where children and young adults are concerned, or is it destined to always be the poor relation, hidden away, never to be revealed due to its vile and repugnant nature.

When accessing suitable facilities, the disabled toilet is the obvious choice for those who live with incontinence or have a stoma, but in accessing these facilities if one's mobility is not affected, it can bring condemnation from onlookers for using facilities when you do not appear disabled. This was evident when a male model approached a conference centre in urgent need of toilet facilities due to his stoma bag malfunction that was leaking faecal matter, his request for access was flatly refused (BBC News, 2018).

I recall instances that seamed insignificant at the time they occurred, but over time wear you down. Being in the baby changeroom of David Jones changing

Adam's nappy, another mother asked how old he was, I found myself telling a white lie, saying he was 18 months old when in fact he was over three years of age. Why? Well, it was a form of protection, I had soon learnt giving people the socially acceptable answer saved a lengthy explanation about Hirschsprung's disease. Few knew about this strange sounding condition, and it would likely make them uncomfortable so why go there? It felt good at the time, I had fended off further interrogation and questioning of my parenting abilities. On reflection, I was ashamed to realise, I had contributed to a failure to change societal attitudes.

Our neighbour who had three older boys, felt she had a duty to inform me with great confidence, the reason Adam was still in nappies at this age was because I had girls and clearly had no idea how to toilet train a boy, is there even a difference? Another time I took him for an appointment to the Cochlear Implant Centre, there was only one bathroom, Adam had soiled again, I had learnt to dress him in long pants with elastic around the bottom in an attempt to capture the leakage. Cleaning him up and changing him was no quick task and I was quickly running out of toilet paper! it was not long before there were several knocks on the door wanting to know how long I would be as other children needed to use the toilet. The comment from our accountant one year who feared Michael and I may have separated as someone had told him we were never seen out together anymore. This was simply a way of coping because taking Adam anywhere with faecal incontinence was no easy task, so it was often easier for one of us to stay home. The experience we had as a family of being invited to be on board one of the Sydney ferries for the annual Ferrython on Australia Day one year. Adam was only about four years old; the girls were so excited and off we

went. Fortunately, they remember it fondly, racing the ferries across the harbour at great speed, meeting the cast of the soap opera, Home and Away were highlights for them. I do not have many memories as I spent most of the time below deck in the bathroom cleaning Adam. The gastroenterologist we were seeing at the Children's Hospital whose parting comments one day in full view of waiting parents was, 'I think you just need to be more diligent with the toilet training.' I was enflamed. I turned and blurted out, 'if you'd like to take him home for a week and see how you go trying to toilet training him, be my guest.' I was fuming. We switched specialists. The dietician who came to assess Adam at home when he was around two and a half years of age. I was struggling to stop breastfeeding, but this was all he would tolerate, fortunately, typical of Down syndrome, he was small for his age, so it did not look out of place providing I hid the truth. The dietician provided her opinion, Adam's problems were simply caused by my inability to move on from his traumatic start to life, I was dwelling on the past and had to get over the fact that he was born with Hirschsprung's disease, he had the surgery, it was fixed, and I was clearly the problem. Her solution, she wanted to take Adam there and then and admit him to hospital where she was going to force feed him, I would not be permitted to visit, and that would fix him. She was promptly shown the door. No doubt to lodge her long report on this terrible mother. I called his wonderful paediatrician who organised abdominal X-rays and scans which quickly showed his bowel was totally blocked and that is why he could only tolerate breastfeeding. We were admitted back into the children's hospital where he underwent an evacuation under anaesthetic and daily washouts which allowed him to slowly start tolerating a suitable diet.

One of the leading surgeons who I worked with at the Children's Hospital, accepted some children failed to gain continence, but had the attitude that eventually these children start to manage their own bodies around age nine or ten. By then he said they were more aware when they had soiled and smelled, so would take themselves off to the bathroom to clean themselves up. He honestly thought this was an acceptable progression. The lack of understanding of the impact faecal incontinence has on a child's formative years, especially when they start preschool or school was staggering, these are the very years children develop friendships and bonds that carry them through school, it was not something a child could just start establishing prior to high school when the patterns of isolation and stigma had been firmly embedded.

These stories are relentless and horrific. But the reality was that all these seemingly minor little instances, built up, reinforcing the impending doom I felt as Adam was unable to meet the normal milestones of toilet training at a socially acceptable age. The future looked bleak, the older he got, the harder it became to fend comments and condemnation off. Whilst there was a lot of support for Down syndrome, faecal incontinence was a whole different ball game, it was isolating, intimidating, demoralising, embarrassing for the girls and the effects severely limited what we could do as a family.

One of the most difficult aspects of all this was his treating team really had no manual to follow, these were uncharted waters, after all, it had only been a few years before Adam was born that children with Down syndrome who had complex medical conditions were offered surgery as the outcomes were unlikely to be favourable. We had meetings with his treating team, tests and samples of his

bowel biopsies were sent to clever people researching unusual cases around the world, from Melbourne to London, sadly no enlightening answers were forthcoming.

The invisible disorder of faecal incontinence was difficult to cope with in society. It was socially unacceptable, was not a welcome topic of dinner conversation, did not attract community support and the congenital abnormalities were largely unknown. Most people associated a stoma or 'bag' with the elderly and bowel cancer, people were shocked if they found out your baby had bowel surgery or a stoma at birth or as a young child. I always found it curious that everyone seemed to have an opinion on how to solve the problem. Well-meaning people you met in passing would have a solution or gently try and point out the error of your ways. I often felt that I could cope with a fair amount of criticism but how did families manage who did not have the strength or support behind them to prevent them falling into the depths of despair?

Fortunately - today - there is much more support and services available to assist families navigate these difficult and isolating experiences with online social media, specialists providing online media education, parent support groups and hospital bowel clinics, none of which were available when Adam was born and have only started to grow in recent years. Despite this, societal attitudes to faecal incontinence remains a taboo subject.

1.10 Is coping the best mechanism?

Most nurses are problem solvers. We see a problem or behaviour, research it, find a solution and 'fix it,' seems straight forward to me. Adam had undergone the 'definitive' surgery for Hirschsprung's disease and yet he was not 'fixed.' I look back on those terms – definitive and fixed, how could it be definitive or fixed when it failed to achieve the desired result. According to the Cambridge English Dictionary the word definitive means; firm, final, and complete; not to be questioned or changed: the word 'fixed' refers to something that is stable, secure, set. Suffering from faecal incontinence was far from any of those definitions. The trouble was no one had any answers in the early 1990's.

My way of managing the problem of Adam's incontinence was to 'cope.' In hindsight, it was probably the worst thing I could have done. I came from an era where women did not complain, common clichés of that time were 'pick yourself up, dust yourself off and get on with life,' 'don't look back you're not going that way". That was the way to 'cope.' Being a nurse reinforced this, after all that is what we did, coped with all sorts of disasters, I flourished in those environments, but this was different, I was used to fixing problems, having control, but this was one problem I just could not fix. I felt defeated and moved to the only mode I knew, coping, which in effect is similar to protective mode described by J. A. Cummings, (2018) where she describes parents 'stepping up' and 'doing what needs to be done; in any given situation. For all intent and purposes, the outward appearances were that everything was fine; after all, that was easier than launching into extensive explanations about the condition and why surgery had not restored his little body to the level of accepted societal norms. The truth was exhausting and demoralising, no one knows what goes on beneath the facade of the perfect family, where you struggle to fit in with a society that has yet to catch

up. I have no answer for the well intentioned questions about how we cope, the truth is, we merely try to exist in a world our son does not fit neatly into. The proverb that time heals all wounds is simply not true, time just moves you through the various stages of grief in the hopes you find acceptance of your new situation that you cannot change. I realise I was a perpetrator of my own problem, coping meant hiding the difficulties of managing Adam's incontinence which did not solve them, it simply removed them from view and allowed them to perpetuate, it saved other people from feeling uncomfortable in our presence. I regret not getting angry, not being the mum who demanded more, not being the squeaky wheel that got attention, but that is hard to do when you are in the difficult position of being a nurse professionally but seeking help as a mother around colleagues you work with.

I did not realise it at the time, but when a dear friend, one of those treasures you just gel with, and have for life, offered to mind Adam for a few hours while Michael and I went out for lunch, was the turning point I needed. Offering to mind Adam was so foreign to us, why would anyone do that? I went into organisation mode, there was a lot to prepare before I could hand him over. I dropped him off a little concerned about how she would cope, I should not have worried, we were similar vintage, so had grown up as copers. I left her with a new pack of disposable nappies, wipes, nappy disposal sacks, barrier creams and what seemed like a wardrobe of spare clothes, all for a matter of a few hours. I am sure it looked like overkill to her. When we arrived at the school carpark to pick him and our other children up, my friend was not there, I was worried, this was not like her, she was a teacher and always punctual. I feared the worst, soiling.

She arrived later looking uncharacteristically hassled. Adam was clad only in a nappy. She had used all the clothes I gave her after multiple episodes of faecal incontinence and said after she had put him in the car seat to drive to the school pick-up, he had 'done it again!' Luckily, it was summer, and she took him out of the car, hosed him off and put the last nappy on. Seeing things from the perspective of my friend was a wakeup call that I had lost all perspective of how abnormal things had become and utterly unfair to our three daughters.

Why had I remained silent? Why had I put up with this for so long, surely something could be done? I guess for me, it was just what I had to do, my days were spent between the bathroom and laundry, apart from caring for our girls. Michael was great, he worked long days in the city and did what he could when he came home, but being the nurse, anything medical he naturally felt I was better to cope with and I took on that role as a matter of course. I think I had completely lost track of what 'normal' was, and our new normal was far from it. But it gradually crept in and took over our lives and I prided myself, dealing with each disaster as it occurred. Amazing what you can conceal behind closed doors, once you step outside, no one would know.

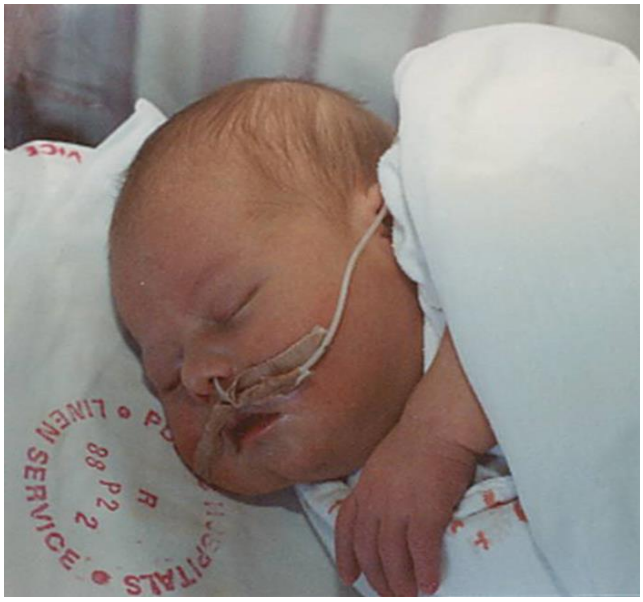
I recalled mothers I had observed during our many trips to the Children's Hospital, being very vocal and demanding action be taken to help their child with whatever ailed them, I was always in a room towards the end of the ward near the parents' room, which was just a large room full of army cots for mothers who stayed overnight to sleep on. But I could hear these mums and knew I could never make such a fuss, I had the perspective of the other side of the nurses' station, how difficult it was to manage a ward full of sick children and their

parents. Interestingly, it was those mothers who got the medical reviews, the social workers came to see them, nurses spent more time with them, maybe their way of dealing with things was superior? I just could not bring myself to behave that way. This was reinforced by Adam's surgeon who could often be heard to say…'oh it's OK mum's an RN, she knows what to do, or yes mum can be discharged early as she's more than capable of implementing these treatment regimen at home.' In reality, I just wanted to be a mother, which is what I was to Adam, not his nurse, hard to do in your place of work.

The impact on our lives was not something that could be easily explained when I sought help from his treating team, how could they possibly understand the impact this was having, but then in the early 1990's no one had an answer. Our surgeon explained the transition clinicians were slowly going through, a sign of success for these children since the condition was identified in the 1950's was survival; it was not unusual for many children to die or not be offered surgery at all. Times and surgical techniques had improved the earlier mortality rate and now it was the norm for children to survive, but at what cost, what if the surgery could not restore continence? Many children eventually gained control of their bowels as they aged, but that often traversed their formative years, meaning they were excluded from friend groups and sleep overs, they often missed school. For those who did not gain continence, it was a long and painful road of trial and error.

A phrase comes to mind that I was used to hearing in nursing and would hear many times through Adam's life…'just because we can, should we'? I realise I probably made things worse by not being more vocal and not demanding they

'do something.' Was I afraid of being seen as a failure, not coping, not trying hard enough, being a complainer? It was my friend's recall of how disastrous those few hours had been that day, and the culmination of many difficult days we had endured at home, which made me take stock and realise we could not go on like this any longer. In no way was she being judgmental, rather, she was in disbelief at what a day with Adam must entail. It was a blessing in many ways, the difficulties we had been experiencing were exposed, I could no longer pretend everything was fine. The arduous journey to seek help began.



"Although the world is full of suffering...it is also full of overcoming it"

Helen Keller

Figure 8: Adam as a newborn and at his sister's wedding enjoying time with dad.

CHAPTER 2: THE STIGMA OF AN INVISIBLE DISORDER.

Sameness breads more sameness, until you make a thoughtful effort to counteract it.

(Obama, 2019).

Goffman (1963) introduced researchers and activists to the attributes of stigma as an "attribute that is deeply discrediting (p.11)". The concept of stigma is easily understood when used in a derogatory way that isolates people due to their differences such as race, religion, colour, or visible disability. Stigma is less easily understood when considering a hidden disorder or difference; one that if revealed risks rendering the person vulnerable to guilt and shame. This thesis aims to expand on the theory of stigma by providing a unique lens on the lives families when a baby is born with a congenital bowel disorder such as Hirschsprung's disease or imperforate anus. My significant original contribution to knowledge comes from the unique perspective of being a nurse caring for these babies and their families; an educator teaching our future nursing and medical students, a mother of an adult son born with Hirschsprung's disease and lastly, cofounder of a support group in Australia born out of recognition of the desperate need for education and support for families and their children born with a congenital bowel disorder, throughout their journey to adulthood.

Children who have faecal incontinence, and their families are frequently alienated from friends and family who have misguided feelings of revulsion in their inability to 'control' their bodily functions. Professional caregivers such as nurses and doctors may share those feelings, inadvertently shaming the parents as not trying hard enough (Butcher, 2020) These children are often the victims of bullying at school, suffering name calling and exclusion (Garcia J. et al., 2005). Faecal incontinence in children is distressing for families due to the associated stigma, which can have devastating consequences for the child that can last throughout their school years (Dobson, 2016; Rajindrajith et al., 2013).

2.1 Unpacking the lived experience:

This thesis unpacks the lived experience providing insight and education to future carers, families, and clinicians, in the hopes the conditions these children are born with, are seen in a similar light as any other congenital abnormality such as cardiac anomalies which foster empathy, and community support rather than social isolation as they suffer in silence.

Patients can experience feelings of guilt and shame and a sense of 'incompetence,' which can be connected to childhood experiences. Children learn the etiquette of accepted behaviour around the excretion of bodily waste, but children associate faecal incontinence as 'being bad' or punishable and the cause of humiliation (Garcia J. et al., 2005). Similarly, nurses and caregivers can encounter feelings of disgust and revulsion. Dealing with incontinence is a fundamental nursing skill taught in the early stages of training. Caring for people with faecal incontinence may not hold the status of a highly accomplished

clinician, but to the affected children and their families, if managed with care, competence and discretion, is one of the most highly valued skills (Norton, 2004).

Some children who are born with congenital bowel disorders do not survive. Fortunately with the advances in technology, operative techniques and medications, this is a rare event today, but it can have a strong effect on nurses caring for these children and their families (Nugent et al., 2022). The sense of loss and sadness can happen even if the encounter was brief but can have a profound effect on their ability to care for children with disabilities or serious illnesses in the future. I recall my first encounter with seriously ill children affected me greatly, I was undergoing a one-year specialisation in operating theatre nursing which involved spending time at the main children's hospital in Sydney. This was before I had my own children and the experience had such a profound effect on me, I was convinced I lacked the strength of character to pursue a career in paediatric theatres. Fortunately, an experienced children's nurse advised me to go and see the children in the ward that we had operated on that day, I was so surprised to see the children happily playing or at least no longer distressed and came to realise children may not always harbour memories of distressing situations long term, they are more interested in living in the moment and playing. Little did I realise at the time, I would have my own child undergoing multiple lifesaving surgeries or that it would change the trajectory of my working life to become a paediatric theatre nurse myself. This change in focus evolved into realising the need for support for these children and their families and led to the development of a peer to peer support group.

What is it like for a child who is born with a congenital bowel disorder? How does it affect their developing years and their family? This thesis unravels the implications of being born with a disorder that is largely a taboo subject, a disorder that must be hidden and yet impacts a child's very social existence. Do the words; low profile, silent disorder, hidden disorder, signify low clinical significance or low priority for research? How can we possibly expect society to have any understanding of the implications of these rare conditions if they are neither discussed nor fully understood.

Understanding how faecal incontinence disconnects children and their families from society is so important if healthcare workers and educators have any chance of supporting them through these difficult times. These children and their families are thrown into a world where faecal incontinence is shunned by society and yet they become the pseudo social anthropologists living a different life, quite alien to them and not one of their choosing. Families often find themselves distanced from their usual social supports as their priorities change to tend to their children's increased care needs, doctors' appointments and school meetings. (Bliss et al., 2010; Devendorf et al., 2021; Else-Quest et al., 2022; Golics et al., 2013; Hendry, 1999; Judd-Glossy et al., 2022; Leigh, 1982; Lister, 2017; Murphy. et al., 2022; Stackhouse, 2018; Svetanoff et al., 2022; Thompson et al., 2021).

2.2 Goffman's 'stigma' and understanding its impact on daily life.

Goffman, (1963) explained the term stigma originated from the Greeks who used the term to describe bodily signs that were cut or burnt into the bodies of slaves, criminals or traitors to display them to the world as unusual or bad. He categorised stigma into three broad types, physical which he referred to as

'*abominations of the body*,' character being perceived as '*weak will, domineering or unnatural passion*; If the person had been imprisoned or was known to have a mental illness, held very strong beliefs, or was known to be dishonest, then the stigmatising characteristic was inferred. The last form of stigma Goffman referred to was tribal, meaning people who were born into a particular race, nation or religious group would be rendered as stigmatised simply from their origins (Goffman, 1963).

The[EG1] reason exploring stigma was so expansive is that my interpretation of Goffman and Gofman's stigma, is that stigma is more widely defined of which disability is just a part of the wider culture of stigma. If I was to remove other elements of stigma, it would be removing the more robust interpretation of stigma.

Do we as people dress and act differently depending on the situation? We dress differently depending on the venue and we present ourselves in such a way so as not to stand out or draw attention to ourselves. How we dress to go to lunch with a close friend or family would most likely be quite different to how we would prepare ourselves to present at a conference of peers or those of higher ranking than ourselves. To Goffman, stigma is not derived from the person or their difference. Rather, it is summoned by those without that characteristic. For example, a person who is of the Christian faith would not be stigmatised amongst their cohort but could be stigmatised if they attended a Muslim Mosque. Likewise, a person who is an amputee, may feel stigmatised amongst non-amputees, but not if he was within a group of people who had also lost their limbs. The same can be said about skin colour, a light skinned person may feel stigmatised if they

live in an area where most people have dark skin. In understanding Goffman's definition of stigma, we know he was not referring to specific identifiable features of a person, but the circumstances in which they found themselves. Goffman's reasoning helps us understand why people who have a specific disability, may seek out support from people who have suffered the same fate, we feel comfortable discussing feelings with people who are more likely to understand and accept us for who we are.

Jacobsen and Smith, (2022) described Goffman as a pair of eyes, seeing what we do and how we do it. Goffman enabled us to discuss the dark secrets of society, those normally repressed for fear they would expose the truth of much human life (Scott and Schlenker, 1981). Goffman's understanding of stigma has changed over time to depict bodily signs of physical disorder. Whilst the concept of a visible disability is easy to appreciate, what of those that are invisible?

Expanding on the various types of stigma related to disability, (Goffman, 1963) explored physical disabilities and how they may affect people, whilst also capturing the beauty people who were marginalised, saw in everyday life, often missed by those considered 'the normals'. The concept of supports between people with similar deformities was considered a welcome discovery, one who had walked the journey before could offer insights to those who possessed the same form of stigma.

Another focus of this Goffman-inspired literature is human trafficking. Niumai and Rajesh, (2022) found children under the age of 18 years who were trafficked from Assam under the false promise of education and money, remained stigmatised as traffic survivors long after their release when police rescued them.

Reintroduction to their families was difficult and without the right kinds of support, they risked being re-trafficked. To their friends and families, they were often seen as trafficked survivors, despite their efforts to shed that stigmatising title.

The use of the term stigma has shifted over the past decades to focus primarily on disability issues, in Howerter, (2022) her thesis examined the implications for racially stigmatised groups considering the recent riots in the United States where non white people were marginalised as less than whites in society. The movement grew with the deaths of George Floyd and Breonna Taylor; Black people who were killed by white police officers, this fuelled the discrimination perceived to exist in all areas of their lives such as education, housing, and employment opportunities.

The language people use can be derogatory, demeaning, dismissing, and decentring, especially if the person using the terms is influential in society. For example, former President of the United States, Donald Trump, described the coronavirus using terms that were derogatory to the Chinese people such as "Chinese virus," "Wuhan Virus," and "Kung Flu" (Al-Hindawi et al., 2022). This only inflamed relationships and marginalised the Chinese people living in America. A difference was connected to an illness and stigmatized.

Via a conference paper (Minton et al., 2021) explored early pregnancy loss as a concept almost accepted as inevitable in many cases. The notion that it is best to keep the news of a pregnancy quiet until after twelve weeks has passed when the rates of miscarriage decline considerably, this simply stigmatises the loss as something not to be discussed. Pregnancy loss is a difficult concept to reconcile,

particularly if it is not a topic widely discussed. Supports following a miscarriage can be difficult when the pregnancy was unknown to family and friends. Yet to the mother, it can be devastating whilst fathers are often at a loss to know how to support their loved ones whilst feeling the loss of their unborn child themselves.

Abortion is another topic rarely discussed. It is a stigmatized, and often illegal or criminalised behaviour, which confirms a woman has control (or lacks control) over her reproductive rights. The misogyny emerges in a patriarchal political and judicial system that denies these rights and stigmatises actions and behaviours. In her thesis, Allan, (2021) compared the standpoint of women using an online social networking site Twitter to engage with the #ShoutYourAbortion hashtag which was used to challenge the stigma of abortion against the pro-life discourses of abortion. The site gave women a platform to share their firsthand experiences relating to the reasons for their choice and control over their bodies, such as for health and reproductive rights. Following the 1986 explosion of the Chernobyl Nuclear Reactor, women were stigmatised as it was thought their future babies would be born disabled. Many women who were pregnant at the time, or became pregnant afterwards, were forced to have abortions (Damaj, n.d.). This often left doubt in our minds as we were on the Isle of Man when Adam was likely conceived and like other areas of the British Isles, received some radioactive fallout from the 1986 Chernobyl disaster which affected the higher country, sheep and necessitated legal controls on sheep farming (Mckenna and Longworth, 1995).

Of relevance to this thesis is 'courtesy stigma' which is experienced by many parents of children born with Down syndrome. It activates empathy, knowing looks and stereotyping. These were the significant findings in a Japanese study of twenty-three families who had a child born with Down syndrome. Hearing terms such as '*they're always so happy*,' or '*they're very musical aren't they*.' Parents expressed coping strategies to deflect or ignore this form of stigma, often experienced in public or by health professionals (Watanabe et al., 2022).

In an online post, parents of children born with Down syndrome expressed their frustration at the stereotypes afforded their children. Comments such as:

> 'If I had a child with Down syndrome, I'd have to have it adopted. I couldn't cope.' Well, don't have children then.
>
> (Stacey B.)

> I don't know how you do it every day!' 'Do what? Love my son? Be a parent to him? Take him out in public? Teach him? Delight in him? Because you wouldn't?
>
> (Stacey B.)

> I can't believe you had one of those. You were so clever in school.
>
> (Paula H.)

> They all look the same. Bless them.
>
> (Mary C.)

> Maybe she'll grow out of it.
>
> (Gianna A.)

> It'll go away after his heart surgery.

(Holly H.)

> When my son was in ICU after a traumatic, preventable postoperative experience, I overheard the intensivist, in a loud voice announcing 'we're going to extubate the Down's boy today' I braced myself and stored the comment away in my mind to include in a letter of complaint to the Chief Medical Officer when my son recovered. It was a case of pick your battles.

(Eunice G.)

> After I had my son, a friend who was pregnant contacted a mutual friend to see if Down syndrome was contagious. She was afraid her son would 'get it' from mine.

(Peggy W.)

Slogans such as the following, do not help, they only retraumatise us:

> Everything happens for a reason,
> God never gives you anything you can't handle (Stone, 2019).

Courtesy stigma can have a profound effect on families, particularly if the child's disability is invisible, it can often be seen as a behaviour problem, parents find themselves the target of unsolicited advice. In a study focusing on fathers of autistic children, the research found fathers experiences were quite different to mothers and yet the most research into autism focusses on the experiences of mothers (Alareeki et al., 2022). Fathers expressed frustration over the misinformation or ignorance they encountered in society, comments such as:

> They'll say something like, that is not very healthy what you're feeding your kid… Like, this is all he'll eat!

(Patrick, father of 8 and 10 year old children)

> Friends and strangers in our community used offensive terms like idiot, retarded, stupid. It's very offensive. I don't call them again. It happens.
>
> (Celio, father to David aged 9 years)

> We went out and made him a hat and, on the top, it says please respect my space I have autism. Just to try to help with it [judgement from strangers]
>
> (Graham, father to 2 year old Aaron)

> When people hear the term autism, they think of the film Rain Man, which was described as detrimental to the understanding of autism. They think of autism as screaming and not wanting to get on an airplane and things. Don't get me wrong, great movie, but you are really stereotyped.
>
> (Celio, father to David aged 9 years)

> Stereotyped assumptions were also experienced as people often thought children with autism all have special gifts.
>
> (Patrick, father to 10 year old Hope and 8 year old George)

In a study exploring the lived experiences of fathers of children diagnosed with autism, it was clear, fathers were concerned about their child's future and went to great lengths in order to support them. However support for fathers was not as easily found as it was for mothers and often required a different approach. The way fathers were told about the diagnosis from professionals was remembered as unsympathetic and left them in a state of shock, they expressed the power differential was evident, they had hoped the diagnosis had been delivered in a more supportive and encouraging way (Camilleri, 2022)

Associative stigma may be experienced by family members, caregivers and friends of people who suffer a stigmatizing illness or condition such as mental health or infectious diseases such as HIV, Ebola, Zika virus, Covid-19 and Monkeypox. Goffman, (1963) explained that because of the close relationship these people have with the affected people, they may share the same stigmatising experience. Whereas people working in areas to service and support the needs of the stigmatised person such as doctors, nurses, or police, may have close contact with the stigmatised person, but are not personally affected by stigma due to their association.

Stigma is seen in gendered ageism. This applies particularly to older women who are marginalised in society and the workforce. Women find themselves discriminated against when training opportunities and promotions arise (Chiu et al., 2001). Older women are often labelled as having poor memory, lower productivity, being inflexible and reluctant to change. There is a perception that older women are resistant to taking orders from younger people in more senior roles (Íñiguez-Berrozpe et al., 2022).

Sorella (2022) identified three barriers to mature aged women returning to education. Situational barriers refer to the multiple roles mature aged women often must manage to enable them to embrace furthering their education such as, paid work, childcare and the caring responsibilities of disabled family members or older parents. These women are often referred to as the Sandwich Generation (Alburez-Gutierrez et al., 2021; Atab et al., 2013; Estioko et al., 2022; Kartseva and Anatoly Peresetsky, 2022; Patrick et al., 2022; Rajahonka, n.d.; Turgeman-Lupo et al., 2020). Parenting skills are often put under scrutiny as

women feel the stigma of their perceived selfishness when they choose to further their education (Grant, 2011; Kirkman and Fisher, 2021; Lundberg et al., 2022).

Older Russian women face sexism and ageism once they reach post-pension age. No longer able to work, they form a protective barrier around the stigma of their situation by choosing to adopt the role of a babushka, an unpaid carer for their adult children and grandchildren, giving them a new purpose, one that is recognised in their society as meaningful (Shadrina, 2022).

Within a patriarchal system, the ideology is that the feminine is secondary, static, and marginalized. Such a system configures a perspective of women who are often stigmatised when it comes to developing new skills. Australian universities are heavily attuned to a stereotypical student, the young, white student often portrayed in university advertising and on their websites (Mallman and Lee, 2016). Rarely if at all, are older women promoted as examples of learning opportunities in our universities and workplaces. Blattman. C, (2015) conducted a blog post and found the consensus was anyone starting on the PhD journey over the age of 35 years, would struggle to be accepted into major universities as they are likely to be questioned over how much can be contributed post-graduation. Questions arose around the stigma of being in a class with much younger students or even finding the supervisor was younger than the student. Institutional barriers overlap with situational barriers in that the challenges facing older women returning to further their education, often find the institutions are inflexible to their needs, favouring the school leaver who can conform to the timeframes of the academic program. Added to these two barriers is the fear of

failure; older women face additional challenges and often question their decision and ability to succeed.

Universities have well-established policies for staff and students with regard to all forms of discrimination, harassment, vilification, and victimisation; programs for students with disabilities, codes of conduct, bullying prevention, gender equality, policies for Indigenous students and sexual harassment to name a few. A review of one university revealed 272 policies listed for staff and 131 policies for students (Western Sydney University, 2022), yet the barriers remain for older women and universities continue to portray the young, fresh faced students as aspiring applicants encouraged to enrol.

2.3 My personal experience of stigma

I understand and grasp the impact of stigma in both my personal and professional life. Growing up, I had no concept of the word stigma, but I came to understand later in life that I had certainly experienced it throughout my childhood. I grew up on a small island having been adopted out as a baby, it was clear my features singled me out as different to my peers and those differences were the catalyst for endless questions and torment. Once my background became known, peers and even teachers disregarded my feelings in pursuit of their own curiosity (Friedlander, 1999). Typical of the era of the 1950s and 1960s, the subject of adoption was taboo and never allowed to be discussed at home. The past was the past. It would not be until I was in my mid-thirties, pregnant with our son, that I was able to discover my true identity which made everything clear; I was the daughter of an Italian woman who had fled the Mussolini regime, joining her brother who had been working for the Italian Embassy in London, when World

War II began. He was interned on the Isle of Man and made a new life there once the war was over. Attempts to find my birth mother were in vain as I later learnt she had died long before I had discovered my origins.

At 31 years of age, in a foreign country and unable to be with the man she loved, my birth mother was faced with making the most difficult of decisions at a time when there was no support or tolerance for unmarried mothers. I cannot imagine the stigma imposed on her during that time. Documents uncovered show she was disowned by the Catholic Church, the very institution that ought to have supported her. The only way for a mother to avoid the disgrace and have a successful economic life was to relinquish her child or be sanctioned by marriage (Crawford, 1997; Luddy, 2011; Wegar, 1997).

I was given the greatest gift of all. My life. The best way I could honour my birth mother was to live my best life and forge my own path, which is what I chose to do. I always wanted to be a nurse; I grew up watching my adoptive mother care so beautifully for her patients so it seemed a natural progression, it was what I knew. I was fortunate to experience excellent training as a registered nurse during the era of hospital training after my husband and I migrated to Australia as newlyweds. This led me to further my training specialising in operating theatre nursing and I gained confidence in the skills I attained as the senior nurse responsible for the theatre teams I was fortunate to work with. I thrived as a nurse and later as a mother to our three little girls, priding myself with my organisational skills, this confidence was put to the test when our son was born, things were not as I had planned.

My personal experience of stigma as an adult, came with the birth of our fourth child. Our son, now an adult, who was born with multiple anomalies including Down syndrome, Hirschsprung's disease, profound hearing loss which he has bilateral implanted bone conductor hearing aids for; he is non-verbal and has a degree of autism spectrum disorder. I have experience stigma many times over his 34 years, starting with the stigma of having a child with Down syndrome. I was often questioned as to why I failed to have a 'test' during pregnancy, to the enquirer that would have simply eliminated the perceived problem. Testing back in 1988 relied on amniocentesis which carried a risk of termination and could not be performed until around 20 weeks gestation. The newer chorionic villi sampling (CVS) test had only just been introduced to women over 35 years of age and had to be performed prior to 11 weeks gestation. As we had just returned from overseas, I was nearing that landmark when I saw my obstetrician, so it was not an option. Nor did we wish to pursue it; our three daughters were one, three and five, so we saw no reason to be concerned and were committed to whatever came our way, after all, as a nurse I had experienced the birth of babies born with Down syndrome, how hard could it be?

Stigma was even evident throughout the pregnancy as people felt compelled to assume we were embarking on a fourth pregnancy purely to fulfil some mystical goal of having a son. This was not true. We delighted with our three girls after years of infertility. This pregnancy was a surprise, the gender was inconsequential. When Adam was born, a young registrar came to talk to me, picked up my file and without even looking at me asked, so how old are you? It was the last straw of many negative interactions, I snapped and said…'well you have the file in front of you, and I'm sure you can read.' The ignorance of such

comments is astounding as women as young as 18 years have birthed babies with Down syndrome, certainly the risk increases with age, but at 36 when Adam was born would be considered the norm by today's standards.

Many aspects of stigma and stereotypes followed us throughout his life. The occupational therapist who condoned his lack of crawling progress as my inability to try hard enough, the fact was Adam had been in hospital since birth having undergone countless major surgeries. His abdomen was heavily scarred, he had a stoma, and he clearly found it uncomfortable to lay on his stomach. I was warned of the peril I was facing that if he did not crawl, he would never progress to walking and I would be to blame. This is not a new phenomenon, mothers and often both parents are blamed for negative outcomes and experiences of their children (Toews et al., 2019) Adam finally took his first steps to everyone's delight at three and a half years of age, even friends said it was better than winning the lottery. He rapidly progressed to running everywhere shortly afterwards and was a livewire as a young boy, how I wanted to contact that therapist to let her know how damaging and wrong her remarks and predictions had been.

The paediatrician who specialised in Down syndrome we saw shortly after Adam was released from hospital following surgery gave a bleak picture of his future. We were advised he would be unable to perform simple tasks we took for granted. For example, to learn how to turn on a tap, we would need to take his hand, place it over the tap and turn it for him, this would need to be performed multiple times as his brain would never remember the steps. I would have loved to know where he got his information from. One day, we found Adam as a toddler

merrily hosing our car - with the doors open, clearly turning on a tap was the least of his inadequacies.

As a baby, and up until Adam was around three years of age, it was relatively easy to avoid the stigma of faecal incontinence, which was seen as the norm, and even accepted in Down syndrome as being a bit slow to accomplish. As he got older though, countless questions arose around our ability to control his faecal incontinence. Often perfect strangers felt compelled to offer unsolicited advice with an air of superiority. It was easy to appreciate that to some people, he had a problem at birth which had been fixed with surgery, so why was there still a problem, the easiest solution was to blame the parents. Suggestions went from the sublime to the ridiculous, naturally this took its toll, and I stopped trying to explain, often making light of a situation (Lash, 2022), or even reducing his age thereby avoiding the unnecessary explanations that were never understood.

One institution; public education, flooded our lives with ideologies, labels, and blame. School was a major source of stigma, there is no avenue to support a child who has faecal incontinence, even preschools require children to be toilet trained prior to enrolment. We were constantly made to feel inadequate, or that he had a behaviour problem, again dating back to the bowel problem being surgically corrected so the aftermath was hard for anyone to comprehend. Surgeons found it difficult to fathom as without a mechanical problem there was no option for surgical intervention, gastroenterologists eventually put it down to parental failure. Social outings became impossible, faecal incontinence is not something you can time or predict so the easiest solution was for one of us to take the girls and one of us stay behind with Adam at home. Stigma was

everywhere, as I tried to normalise family life for the girls who were keen to pursue physical culture, ballet lessons, swimming at the local pool, outings with friends, birthday parties, special trips such as New Year's Eve celebrations, the Easter Show, seeing Santa at David Jones in the city. Sadly, so many of these were tainted with long interruptions as they were too young to be left alone while I took Adam to bathrooms to clean him up, so they had to tag along, fortunately they seem to have little recollection of these instances, but I wear the memories of these stigmatising situations deeply embedded in my mind. Could things have been different? Mother guilt is ever present in any mother's mind but felt more acutely when one of the children has a disability, it is not uncommon for the mother to feel they are failing their other children as the one with a disability ultimately takes priority (Kerr et al., 2021; Smith and Blamires, 2022; Yoosefi Lebni et al., 2021).

On reflection, Adam had been born at a pivot in medical history. If he had been born a few years prior, he would not have been offered surgery but simply put on supportive fluids until he died. Surgical techniques and medical interventions were advancing at a rapid rate, and this was a time of change where parents were questioning more, researching, and demanding more be done to save their babies. Research overseas was often presented to doctors, something I am guilty of as I often turned up at appointments with yet more promising research printed out, hoping this was the answer we were looking for. As a mother you search for anything that will help your child and alleviate their suffering. It was now possible for a baby born with Down syndrome and a life-threatening bowel disorder to be saved. As such we were caught up in an era of trial and error. Yes, surgical intervention could remove affected bowel and bypass the anomaly which

would in effect, save his life, but the prospect of the quality of that life had not been tested or considered, we were in fact the guinea pigs of that era with no tests cases to refer to. What followed were years of frustration, disappointment, constant hospitalisations, the pain and suffering of multiple surgical interventions, all with the elusive goal of faecal continence. Advance thirty years and things are very different now, whilst Adam was one of the many test cases for multiple interventions, many to no avail, some babies succumbing to their fate. Today, the options are more streamlined, the delay in returning to a stoma or primary stoma formation is shortened, if continence is not achieved in the normal timeframe or soon after pull through surgery, we now understand the importance of preventing long term damage to the bowel wall from chronic distension. Medications and interventions have improved and are introduced early, preventing the trauma Adam and many others suffered at that time. Medical professionals have joined with parents to learn from each other; social media platforms have enhanced the ability for better delivery of information and shared experiences, it is hoped, these advances will change the way care is provided for future families when their baby is born with one of these rare conditions (Ryan et al., 2020).

Experiencing the difficulties our son encountered, led me to further my understanding of the complexity of congenital bowel disorders. Firstly, it seemed a natural progression to combine my personal and professional experience and set up a support group for families to share their experience and knowledge of these rare conditions. The timing was optimal as there was no support system for these children or their families. A paediatric nurse who had a special interest in these conditions, along with another nurse who was also the mother of a little

girl who had Down syndrome and a congenital bowel disorder, agreed to join forces and the support group was formed. Many parental support groups begin when parents experiencing similar situations, and form a community to share experiences and support each other. Groups such as the 'The Parents Village' began when two friends who were professional women, found themselves overwhelmed and isolated when their first child was born. Together, they formed the group in 2016 due to the need for a 'village' to learn from, lean on and laugh with. https://www.theparentsvillage.com.au/

One of the Men's Sheds in Queensland created a special interest group to support men whose families have children diagnosed with autism. This developed when one of the fathers recognised that his wife was more in tune with their son's challenges because she did most of the caring. But it left him feeling alone with no one who understood his experience. The group runs with the support of the Department of Education and Training Autism Hub bringing men together to share experiences (FACT, 2023)

2.4 My significant original contribution to knowledge

This doctoral research is a culmination of my unique experience as a nurse, an educator of future health professionals, a mother, and cofounder of the support group to write this thesis. This unique lens of combined experiences form my significant original contribution to knowledge and led me to pursuing a PhD in my late fifties. It proved challenging in many ways. I was not the archetypical student who had completed an honours or master's in research following onto a higher degree. I was starting from a foundation of hospital nursing training in the 1970s to a bachelor, Graduate Diploma in Adult Education, and a Master's in

Health Law, none of which was seen as the standardised progression into a PhD. I was on my own in this journey. I enquired around all the NSW universities about a potential supervisor for my research into the supports that are needed when babies are born with a congenital bowel disorder, Hirschsprung's disease and anorectal malformations. I was unable to secure a supervisor who had any interest in the topic so contacted a long-time colleague I had worked with for many years in the operating theatres of the Children's Hospital and through his research into Hirschsprung's disease. He had also been instrumental in helping set up the support group. My intention was to ask whom he would recommend. To my surprise he enthusiastically offered to support me by being my supervisor. It seemed the perfect choice, we knew each other well and worked and researched together, what could go wrong? On reflection, I was both naïve and lacked the skills to assess the situation. He supported my application which seemed to progress relatively easily which I now realise lacked academic rigor. I had no idea what to expect from a supervisor and quickly realised I was largely on my own as he ran an extremely busy paediatric surgical practice, was often on call, ran clinics and had little time to devote to supervision. Sessions were ad hoc, infrequent and progress was slow. I am grateful for the opportunities afforded me during my time under his tutelage as I was fortunate to speak at numerous conferences both locally and in Europe, all the while feeling inadequate as most of the presenters were accomplished surgeons or surgical registrars. I felt stigmatised as I was the only nurse or mother in attendance and felt the imbalance of power acutely. On rare occasions, there was enthusiasm from surgeons, albeit the younger cohort. One young registrar approached me after my presentation in Milan in 2015 entitled 'A Collaborative Approach to Holistic

Care in Paediatric Anorectal Malformations' and was very supportive as he explained no one ever taught him what I had shared, and that he felt compelled to understand more. I did feel as though he was in the minority although he gave hope that attitudes were gradually changing.

Questions arose as to why I was doing a PhD at my age. What was the point? The implication was that completing a PhD was an expected pathway to progress one's career through university, which was a rather ignorant and narrow view on lifelong learning. I had been employed at various universities for over twenty years on a casual or contract basis, I was not pursuing a career as a full time academic, nor was a position likely to avail itself as tenured positions were fast becoming obsolete. My reasons were quite simplistic; as an educator I understood the importance of sharing knowledge, I wanted to culminate my unique perspective of congenital bowel disorders as a nurse, an educator, a mother and founder of a support group. Surely the years of experience across all aspects of these children's lives was worth sharing, otherwise education is incomplete if kept to oneself.

Unfortunately, my experience throughout the PhD was poor. Two of the three supervisors were never actively engaged. One I briefly met as I caught up with him in a corridor of the hospital requesting he sign a document, his comment was, *'doing something important are we'?* He had no idea who I was and clearly no interest in finding out, my only relevance to him was a silent addition to his resume. I emailed an article to the second appointed supervisor for review which failed to elicit a response. I called him to ask if he had received it, the reply was, 'oh I don't do email', so I had to print the article out, drive over an hour to deliver

it and wait; feedback never eventuated. Over time it was obvious I had one, very busy supervisor who later admitted he had his own agenda which was purely statistical based on data collection and he neither understand nor supported qualitative research. The thesis was not progressing the way I intended, I wanted to give these families a voice, to bring their lived experiences into focus. Whilst existing data was important to review numbers of children affected; percentages of how many did well post-surgery, they failed to elicit what goes on behind closed doors. None focused on the difficulties these children and their families faced navigating systems designed to only accommodate children who were socially continent. Storying was the basis of my thesis, not pure data collection and comparative studies, without storying, the crucial question of why faecal incontinence matters cannot be understood. Therefore, my supervisory pathway through this doctoral program continued the stigma, the neglect, and the marginalization.

It took a long time to realise I had to speak up and although it felt uncomfortable raising the issue with the university regarding my long-time colleague, it had to be done. I was assured a new supervisor would be found. Months went by, eventually a new supervisor was appointed, but he was on sabbatical for the next year, so I experienced more delays. Once I commenced with this supervisor things seemed to progress in the right direction, and he actively encouraged using 'What's app' as a platform for our weekly meetings as opposed to the previous routine of driving three and a half hours from Sydney to Canberra to meet with my previous supervisor who could often only spare 30 minutes to discuss progress. Research was going well until he abruptly left the university just six months into our sessions, I was never given an explanation. A further six months

went by before I was appointed another supervisor. Therefore, I understand invisibility, marginalization and not fitting into the expectations of systems and structures.

The burning desire to never give up was a strong as ever, if I gave up now, who would speak up for the many children and young adults like my son? Yes I was frustrated and disenchanted but speaking up for those who through no fault of their own, spend their lives masking the pain and isolation that stems from faecal incontinence had to be uncovered. The risk and harm of not exposing the effects faecal incontinence has on a child's social and cognitive development, would be a disservice to the very people who have entrusted their stories to me. This thesis is a testament to all the incredibly brave young people and their families who have endured years of humiliation, exclusion and frustration as they navigate the difficult path through their formative years towards adulthood, who are wise beyond their years. Stigma has transformed in its role and place in the social sciences since I first learnt of the concept in 1963. We have a greater understanding of many different types of stigma such as courtesy, public, affiliate, experienced or perceived (Čolić, 2021).

2.5 Educating future clinicians:

In healthcare, before nursing education moved into universities, students spent their entire training caring for patients which was complimented with theoretical learning, usually from doctors and later, nurse educators. Since the advent of the nursing degree in 1982, most of the learning is theoretical, taking place on campus with skills practiced on manikins in laboratories such as bed making, catheterisation and wound care. It is not until the undergraduate nursing

students attend clinical placements, that they experience the complexity of nursing. Many students hold stigmatising attitudes when faced with upcoming mental health clinical placements (Lim et al., 2020). With education and understanding, students felt less anxious and more prepared and appreciated the importance of nursing care not only in mental health facilities, but in many aspect of general nursing.

An important aspect of clinical placement for students is modelling, observing the experienced clinician interact with clients provides a catalyst for students to practice in a safe, supervised environment and allows the opportunity for formative feedback. Cummings et al., (2015) in their study of clinical supervision, found clinical supervision was an important aspect of training when it was a purposeful and proactive endeavour. I was fortunate to be involved in the inaugural non-governmental organisation (NGO) program at one university whereby I would supervise nursing students during a two-week placement as part of their mental health component. There, they had the wonderful opportunity to sit and talk to clients who came together in a safe space. Many of whom were homeless men, who had the most amazing stories to tell if only someone would give them the good grace of time to listen. Some, so traumatised by their service during the Vietnam war, they found it impossible to slip back into the society they once enjoyed. These placements were a stark contrast to most Mental Health placements where students attended an inpatient unit to observe patients admitted during to an acute crisis, usually behind the security of a safety glass screen. The NGO placement gave them an understanding of how people who suffered from a stigmatising mental health condition actually lived in society, rather than the narrow perspective of the acute hospital setting.

Understanding disease mechanism is vital to dispelling myths and reducing the stigma the patient feels. For example, there are many forms of diabetes mellitus which are largely misunderstood in the community, often the broad term diabetes is used leaving the patient feeling stigmatised as it is assumed they have made poor lifestyle choices.

Although type 1 diabetes mellitus (T1DM) and type 2 diabetes mellitus (T2DM) do not have the same causal link, they both carry the burden of stigma, as to the observer, it is often assumed both are caused by eating to excess or consuming a high intake of sugar. This can be particularly difficult for children who are diagnosed with T1DM (Brunton, 2022). The onset is often rapid, without warning and changes their quality of life permanently as they navigate a complex health system and must adopt a new approach to food monitoring, carbohydrate intake offset by insulin delivery by multiple subcutaneous injections every day. All the while managing stigmatising comments from family, friends and the general public. This is particularly challenging if the diagnosis appears in adolescence when there is a perceived carefree lifestyle of the young, finding their way, which all ends abruptly as their life is at risk if the strict regimen of diet and insulin delivery is not followed.

The stigma associated with T2DM is felt not only from the public, but from healthcare providers entrusted with managing their care (Himmelstein and Puhl, 2021). People with Type 2 diabetes suffered a sense of double stigma from being overweight and being perceived to have caused their own diagnosis (Puhl et al., 2022). A study conducted in Ghana (Botchway et al., 2021) found people who suffered from self-stigma, were ashamed or worried about what people thought

which often led to them keeping their diagnosis a secret to avoid the perceived shame which in turn led to poorer health outcomes. In low-income countries, the use of peer support groups was considered valuable to help inform people when healthcare was limited. In contrast non-disclosure was found to be quite common in high income areas due to perceived stigma about societal attitudes (Browne et al., 2013). Enacted stigma occurs when healthcare workers blame the person for their condition rather than offering supportive advice, this can lead to self-stigma and lower self-esteem and can result in the person looking for alternate avenues of support which can be detrimental to their health outcome. Poor disease management also occurs when people fear seeking professional advice due to the perceived stigma (Browne et al., 2016).

Understanding human conditions, be they visible or not, can help dispel long held beliefs and attitudes which are stigmatising to those affected. The challenge comes when a newly discovered condition arises in the community that we do not understand, this raises fear and can be stigmatising to the affected person.

2.6 The stigma of diseases

Once trades between countries was possible, and more so when people were able to travel across nations, the risk was imminent that diseases would spread. There is a long history, be it folklore or fact. Hoppe (2018) explored the origins of disease names, specifically those relating to geographical locations and found these were stigmatizing towards innocent people of those locations, even though the disease may not have had anything to do with them. The Gobi desert was supposedly the source of the Black Death, reaching Dorset in 1348, the cholera pandemic in England in 1832 thought to have entered on a ship from Hamburg,

the Spanish Flu was in fact the influenza pandemic just over one hundred years ago and yet had no correlation with Spain other than an early news report suggesting Madrid being the originating source (Choi, 2021). The connection of diseases to foreign populations can spread irrational fear against people from those regions, stigmatising them as though they themselves are responsible. Fortunately, the advent of the social media has meant governments can correct misinformation early, but once it has begun, xenophobia is hard to contain.

Since the Spanish Flu, we have seen similar fearful reactions to the 2009 influenza A (H1N1) virus, Ebola virus, and the Zika virus. Whilst the Zika virus (ZIKV) became common knowledge globally around 2015 when it spread rapidly, it was first discovered in 1947 in the Zika forest in Uganda. Zika spread fear among pregnant women as the effects on the developing foetal brain could be devastating. In a study conducted by (Marbán-Castro et al., 2020), women expressed fear of mosquitos, the disease itself and stigma associated with being identified as having Zika and the risk posed to others and their unborn baby.

Stigma was clearly evident when HIV first appeared in Australia forty years ago, when a woman gave birth to a 28-week premature baby girl in Australia she called Eve. Little Eve was given several blood transfusions in her fight for life, tragically, one of those transfusions came from a donor who had a virus that attacks the body's immune system called human immunodeficiency virus (HIV), untreated this can lead to acquired immunodeficiency syndrome (AIDS). In 1982, it was unknown that HIV could be transmitted via blood products. The family were ostracised due to ignorance around transmission. Eve and her family suffered the unimaginable stigma of being considered contaminated, people

avoided contact for fear they would be infected, with further education and understanding, it became clear, that was never the case. That knowledge was unfortunately, too late for Eve and her family who were eventually forced out of the country and sought sanctuary in New Zealand (Harris. K, 2022). It was around this time that homosexual men were blamed for transmitting HIV via blood donations. Sexual intercourse between consenting adult males was illegal in NSW until 1984 when antidiscrimination legislation was passed. Stigmatisation of homosexuals was rife during this time, with degrading notices and signs posted such as the 'No Gay Blood' posted at the Ford Motor Company plant blood collection unit. Heterosexual men lined up to give blood to prove to workmates they were not gay (Sendziuk, 2005).

Over the past three years, the world has experienced the effects of stigma during the Coronavirus disease (COVID-19) pandemic, this has caused discriminatory behaviour in a way that has been difficult to contain. Families holding opposing views have become estranged (Lattanner and Richman, 2017), staff with opposing views or decisions regarding public health orders and their perceived human rights in workplaces, hospitals and schools risked the disease spreading rapidly (Martin, 2021). The endless association of China as the perpetrator of COVID-19 has been difficult for people of Chinese origin who felt stigmatised no matter how they behaved during the pandemic (Choi, 2021). Comments were identified such as:

> I don't want to wear a mask in grocery stores because people look at me all weird. They might be thinking I'm already sick, walking around in the public and spreading the virus, you know?

> It is OK for White people to wear a mask, or to not wear it. . .. But as Asians, we must be careful.
>
> People look at me like I AM the virus. [By wearing a mask] I am being protective [of] my own health and everyone else's.

Nurses and midwives during the COVID-19 pandemic found they experienced stigma in ways they could not have imagined when the virus began. Nurses reported neighbours feeling threatened as though they were carriers of the virus. Nurses felt overwhelmed by the increased workload and unsupported by management. During lockdowns, the increasing pressure of home-schooling children, being torn between the sense of duty as a nurse to work but wanting to be home to care for their children was stressful. Burnout was common during the height of the pandemic as staff were off on sick leave due to contracting covid themselves or having someone at home with the virus, meaning they had to isolate (Marsden et al., 2022; Shiu et al., 2022).

As I held certification as a nurse immuniser, I accepted a role as a vaccination nurse at the local respiratory clinic during the Coronavirus (COVID-19) pandemic administering vaccines. It was an experience like no other, we were administering hundreds of vaccines a day to adults and children at the time. One day, an 8 year old girl who had Down syndrome came bounding into my room for her vaccine with her father, she instinctively ran up to me to give me a hug and I reciprocated, her father started to apologise for her 'behaviour' and I said to him please do not ever apologise for her beautiful nature and spontaneity, I told him about Adam and how I understood their lack of inhibitions, which he was grateful for. It goes against societal norms to accept close contact between a heath professional and a child but in this case, the father was in attendance. The encounter made me

wonder, how sad that her father felt the need to apologise for his daughter simply because she did not fit in with societal norms.

Parents and caregivers have been advocating for greater understanding of the needs of the people they care for, be it children born with congenital abnormalities, adults with acquired mental illness or conditions that set them apart from mainstream populations because of a stigmatising appearance, condition, or physical impairment. Scholz et al., (2018) found whilst the delivery of services has changed over the last 30 to 40 years, there remains a significant power imbalance with providers retaining superiority over consumers, who are often seen as a resource to be used rather than the partnership that was hoped for. The stigma of their child's condition felt like a constant battle when dealing with authorities. Whilst on the one hand parents were often rejoicing in the achievements of their children born with a disability, when it came to accessing services, the only way to secure support for their child was to focus on negativity and what they could not do. This was certainly our experience when Adam was transitioning from high school to what was then called post school options, to ensure he received the level of supports he required, meant we had to focus on every aspect of life he could not do himself or without supervision, this felt very uncomfortable as we always focused on his capabilities.

Comments from the very people who are employed to assist parents were often seen as uneducated and unhelpful in their quest for families to access services for their child. Comments such as *'when did the Down syndrome start? when will they grow out of it' 'so is he better now'* added to their stress (Thomas, 2021).

Kašperová (2021) found the reaction of customers to people with visible disabilities varied, some lost contracts once the customer met with them, this led in some circumstances to choosing to run a faceless business. This of course was not always possible, and contrary to this experience, running a business that was based on the lived experience, such as a man who was wheelchair bound, set up a business selling clothing for men in a similar situation (De Clercq and Voronov 2009). This was of great benefit to the customers who felt their needs were understood thereby reducing the stigma often felt.

2.7 Conclusion

It is clear stigma punctuates many aspects of our lives, the implications for change within clinical practice is huge. This thesis highlights the need for support networks, collaborative healthcare relationships, clear and accurate information and adequate government and financial support. By understanding the experiences of people who are stigmatised more deeply, there is an opportunity to guide the changes needed for improvement and provide a more meaningful picture into what this experience looks like.

CHAPTER 3: TRAVELLING WITH A CHILD WHO HAS A STOMA OR FAECAL INCONTINENCE

It is important throughout this thesis that the stories of families who have so generously shared their experiences with me be told. Without first-hand knowledge of the difficulties and challenges families face when their child does not fit with societal norms of toilet training, we cannot begin to understand the problem. Through ignorance, the child remains isolated, shunned and their problem hidden from society. It is through storying that we begin to learn the extraordinary depths parents have gone to in order to help their child, often to no avail leaving them to figure out solutions alone. Without understanding the lived experiences of children who have residual faecal incontinence following corrective surgery for Hirschsprung's disease or anorectal malformations, we cannot begin to build the supportive interventions these children need once they leave the security of home. The natural progression of preschool, school, socialisation with peers, sleep overs, camps, travel, employment; all present challenges some find too difficult to overcome and so they retreat to the safety of home, missing out on the greatest adventure of all, a normal life. Faecal incontinence is regarded as a taboo topic, a dirty word, certainly not one to enliven dinner conversations, and yet not presenting these stories through the lens of families would be a failure to grasp the opportunity entrusted to me through my own experience as a nurse, mother and co-founder of a support group. My significant original contribution to knowledge is framed by – and built on - the comparative research between the learned experience as a clinician and that of families who come together to support each other due to their shared experiences. My life experiences as a mother of a child born with significant

disabilities, a nurse, an educator and cofounder of a support group aims to assist clinicians, educators and the general public understand the difficulties some of these children and their families face. Change cannot occur if we do not expose the problem, research the options, and create change. We may not be able to correct the underlying problem, but we can certainly address the stigma associated with it and provide the services to support these children rather than concealing and perpetuating the problem.

Travelling with a child who has a stoma or faecal incontinence is an education and certainly never dull. Packing for a holiday is often filled with excitement. Travel arrangements have been made, accommodation booked and itinerary constructed. Most of us pack far more than needed, and sometimes the weather is not what we anticipated, and we need to buy more items overseas, or donate items we only needed for this trip to charity shops. But what do you pack when your child is no longer a baby but is incontinent of faeces? Planning takes on a distinctive and unique perspective. Most hotels require an additional room if there are three adults whereas a couple with a child can be accommodated in one room, so the cost doubles. Basic hotel rooms are no longer suitable, accommodation needs to have facilities to wash and dry clothes quickly and easily, not through an expensive laundry service offered by the hotel. Packing, preparing documentation to confirm the need for additional baggage all adds to the plan.

Our experiences of overseas travel have been varied and each time we have learnt new things and are convinced next time will be easier, it rarely is. We are fortunate to have family scattered worldwide but If we do stay with family, it is

only ever short term as we find it easier to have our own space and there is less stress about leakage happening overnight soiling bedding and when washing soiled clothing, I would feel extremely uncomfortable using someone else's washing machine in their home.

3.1 Preparation before our journey.

We never leave Australia without comprehensive travel insurance. This is never a simple task, as congenital bowel disorders are a pre-existing condition, whether they have a stoma or not, and most policies will not cover this without a review by the insurers medical team and an additional fee. One year we were denied cover for Adam without reasons to substantiate that decision. I asked for peer reviewed research evidence to support their decision, within minutes they came back with an authorisation. If Adam were to require hospitalisation whilst we were overseas, we may need to change or cancel our accommodation or flights, so we make sure the insurance covers this as cancellations can be costly. We choose not to suspend or cancel our Australian private health insurance whilst we are overseas. If we ever needed to return early and Adam needed to be hospitalised, we prefer to know his cover is active as soon as we touch down. Some countries have reciprocal arrangements with Australia for health care needs, so we always check before leaving and check the smart traveller website for current information[4]

3.2 The dreaded packing

I start a few weeks before we are due to leave by making a list of items starting with the morning routine throughout the day up until Adam gets up the next

[4] Travel information accessed 6 October 2022 at https://www.smartraveller.gov.au/

morning. We have learnt that it is easier to be over prepared than underprepared, we now pack bed protectors with us, see picture below, and a set of sheets for Adam's bed in case there are accidents.

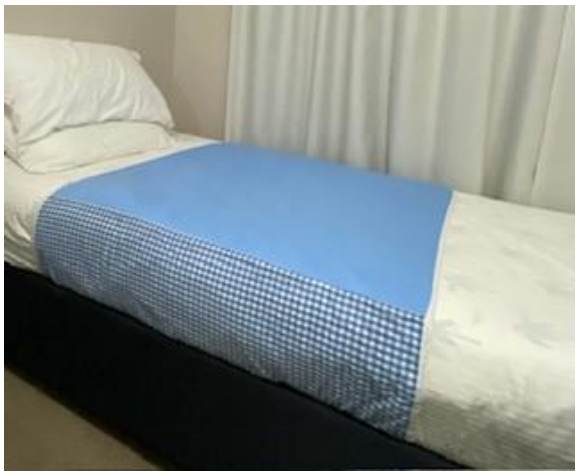


Figure 9: An example of a bed protector on my son Adam's bed, easy to change when accidents occur.
https://www.brollysheets.com.au/products/brolly-sheet-blue

We have been caught out in the past, running out of stoma bags. This is not something that can be bought at any pharmacy. One solution was to empty the contents of the used bag into the toilet, fill it with water a few times to rinse it out and reuse it. This was far from ideal. We have also approached the local hospitals for some emergency supplies when needed and they have usually been able to offer some assistance. We now pack more than anticipated, which means counting the days we will be away, multiplying by five changes a day and adding more, just in case. We have also packed and mailed stoma supplies to a destination to save weight restrictions, particularly when going from an international flight to a small aircraft that has much less baggage allowance. This often brings some strange responses, but better than being caught without them.

One suitcase alone is never enough for Adam's supplies which include a sleuth of incontinence aids such as bed linen, pads, nappies, nappy bags, creams and

stoma supplies along with all his medications. Then there are the clothes, so many sets are required to ensure we do not run out between destinations. This is even more problematic if we travel through Europe, then head over to our birthplace, the Isle of Man, which is notoriously windy and much colder than Europe.

As for accommodation, we try to find self-catering apartments which have a laundry and kitchen equipped enough to manage. Occasionally a hotel room will have a microwave included, but it is rare. One year in Rome, we spent most of the time trudging around the streets, midsummer with bags full of dirty laundry trying to find a laundromat. Whilst Rome has so many amazing attractions, sadly, laundromats are not one of them.

I type up letters listing every single condition Adam has, every current medication and appliances he needs for travelling. I make appointments with his general practitioner and take the letters along and ask them to sign them. That way when we are inevitably stopped at security, we have as much documentation as possible to support our need for carrying so many medical supplies and special foods or drinks. I also take a current list of his medical history which I have been adding to since he was born, once I created the original document, editing and adding procedures and operations was easy, it also lists all his current doctors with telephone numbers in case we run into difficulty, and he needs medical assistance overseas.

This became a lifesaver one year when I was presenting at two conferences in Europe, and we had organised a holiday to follow. The travel was gruelling, moving every two days. The first talk was in Milan, then a few days later,

Ljubljana, Slovenia. Although not related to his bowel disorder, on the Friday evening before leaving Slovenia to head to Rome, we were settling Adam to bed and removed the external component of his implanted hearing aid and the internal component snapped off. Without it, he was profoundly deaf so could not hear anything. I emailed his Ear Nose and Throat surgeon in Australia; he emailed back after contacting a colleague in Rome and sent us the details of his clinic where an appointment had been set up for the very next day. We could not believe it, getting into a specialist on a Saturday at short notice would be highly unlikely in Australia. Going to an emergency department for something like that or related to his bowel would be complicated as these conditions are so rare, they need the expertise of specialists.

The surgeon in Rome assessed Adam and we were relieved to learn surgery could wait until we returned home. What was remarkable was he organised a temporary device that would give Adam some level of hearing until he underwent surgery back in Sydney. The device was not available in Rome, but the surgeon organised one to be sent to Sorrento where we were headed the next day. When we arrived at the hotel, the package was waiting for us containing his emergency device and you could not wipe the smile off Adam's face; he could hear again.

3.3 Events we did not anticipate.

The extra time required at security check-ins due to the additional items we needed to carry onboard, we cannot risk all his medications and stoma supplies going in the aircraft hold in case bags are lost, so we spread things across all bags, including carry-on bags. The restriction of carrying specific foods and liquids on board the aircraft is problematic as Adam will not eat regular airline

foods and has a small selection of food he will or can tolerate. If he were a baby, additional liquids in the form of baby foods or formula would be permitted, but not a person who chronologically is expected to adhere to adult rules.

Despite meticulous planning, at each security check, there are questions, items scrutinised, and supervisors called to inspect our documentation is in order. This of course is particularly difficult on long haul international flights as opposed to domestic flights. One year we were fortunate to have the contact details of the head of security who advised us to ask for him on arrival, the difference was unbelievable, when we arrived, he came down to security and simply waved us through, sadly he was no longer there the next time we flew, and his successor did not afford us the same privilege.

One year when Adam entered the whole-body imaging machine at airport security, alarms bells were set off as his ileostomy bag showed up on screen. The security guards immediately jumped into action wanting to know what he had hidden under his clothing. If we had been in an English-speaking country this would not have been too difficult to explain, but in Singapore, it took quite a bit of explaining to convince them it was not a concealed weapon or drugs. Apparently using disabled people or children to conceal illegal drugs or weapons is not unusual.

3.4 Seating arrangements:

We have been fortunate to amass enough frequent flyer points to fly business class. We realised after a disastrous British Airways flight just how important

seating configuration is. These seats were two to a row, but in opposing directions to each other which meant we had to climb over the persons legs behind if their seat was reclined. One poor man found this intolerable as we had to take Adam to the toilet frequently as the air pressure in the cabin meant his stoma bag filled with air very quickly. Some business class seating arrangements are single rows which would be unsuitable for Adam as one of us always needs to sit with him. We generally choose two seat rows and take the third seat on the adjacent row. We also take turns having a sleep and ask the airline stewards to wake us if they see Adam getting up.

3.5 Toilets:

We all know how ridiculously tiny aircraft toilets are. Try fitting two adults in without drawing unwanted attention. Privacy is non-existent, one of us takes Adam to the toilet and we have to leave the door open, providing as much privacy as we can, once he has passed urine, he shuffles around to face us while we change his stoma bag. Each time we hope this will not draw unwanted attention, but it inevitably does with strange looks from passengers and the stewards inevitably checking nothing untoward is taking place.

We have travelled extensively with Adam except in the past few years of course with the restrictions due to the pandemic. We prefer to travel to countries where we can find serviced apartments and suitable foods without much trouble. We try to travel outside of peak holiday times as there are less crowds, and it is not so hot in Europe which puts Adam at risk of dehydration and sodium loss due to his condition.

We often have well-meaning friends suggesting we leave him with our girls or put him into some form of respite so we can travel alone. Whilst this may seem of benefit to some people, it would not suit us, we could no longer go off enjoying ourselves leaving him behind, he loves to travel and always jumps onto the window seat on the plane. Yes, there is a lot more planning involved but Adam has gone through more in his short life than most of us would experience in a lifetime, but that does not mean he should not experience living life to the full which every parent wants for their child.

The following chapter explores a review of relevant literature depicting research already reported on, this will identify and characterise where gaps in the knowledge exist, leading to this research that frames the investigation into understanding ways in which clinicians, support organisations and institutions can work together with families to contribute to a greater understanding of the needs of children born with a rare, congenital bowel disorder from birth to adulthood, and support their families throughout this journey.

CHAPTER 4: LITERATURE REVIEW

The first section of this thesis offered a discussion of positionality when approaching research into Hirschsprung disease. There was attention to the relationship between experience and expertise, past and present, opinion and research, subjectivity and truth. From this frame, this chapter presents a review of the existing literature on Hirschsprung's disease and anorectal malformations, and patient and family support groups. In doing so, this chapter identifies a gap in the existing literature: the lack of literature focusing on patient and family support services for children born with Hirschsprung's disease and anorectal malformations. The literature review has been presented at this point of the thesis, rather than as chapter one or two, because the experiences of families, parents, children and adults must be granted priority. From acknowledging the diversity of these experiences, the academic lens is presented and probed. There is a substantial medical literature on Hirschsprung's disease and anorectal malformations. This literature focuses on the anatomical and biological aspects of the conditions, how and when they are diagnosed, the various surgical techniques used to treat the conditions - such as primary pull through, laparoscopic assisted pull through, single or two stage procedure and whether the procedure should be performed under the protection of a resting colostomy which can be closed a few months later when the surgical repair has sufficiently healed. An emerging literature is now documenting the long-term outcomes of surgery for Hirschsprung's disease and anorectal malformations. Whist there seems to be an improvement happening over time, there was certainly a significant number of years where these children faced the difficulties associated with faecal incontinence which can persist into adulthood (Aliev et al., 2020;

Bogusz et al., 2021; Pan et al., 2022b; Puri. P, 2019; Wesson and Lopez, 2020). According to the World Health Organisation (WHO) Declaration of the Alma Ata (WHO 1978) "just as government must provide adequate health and social measures" the people of any country "have the right and duty to participate individually or collectively in the planning and implementation of their health care". The literature on patient and family support services in medical contexts is also expansive, addressing the patient's experience of participating in a support group, feelings of belonging and shared experiences with people who understood their situation. Historically, peer support groups often began due to a lack of knowledge or understanding of the complexities faced by people with emerging or rare conditions. Another major driver for support organisations was a need to connect with others in a similar situation due to the isolation they felt living a life their friends and family found difficult to relate to (Ansell and Sarah Insley, 2013; Basset et al., 2010.; Castellano, 2012; Cyr et al., 2016.; Davidson et al., 1999; Faulkner and Kalathil, 2012.; Rogers and Patterson, 2021.; Borthwick et al., 2022; Tang, 2021; Wang et al., 2022). The growth of peer support organisations has been expansive with the availability of the internet to connect people. The benefits of online access to supports are increased access from anywhere in the world providing there is internet access. The limitations can be the limited feedback when participants ask questions, the quality of those moderating the site, difficulty in authenticity of members particularly when sensitive issues are being discussed or those relating to childhood conditions (Gary and Remolino. L, 2001). As I discuss in this chapter, the literature on support groups for families with children with Hirschsprung's disease and anorectal malformations is very scant (Gupta et al., 2021; Kula Sahin and Karakas, 2022).

4.1 Hirschsprung's disease and anorectal malformations

Hirschsprung's disease and anorectal malformations are frequently associated with genetic disorders. The most common genetic disorder associated with Hirschsprung's disease is Down syndrome. The first report of Down syndrome in association with Hirschsprung's disease was in 1956 (Vacher, Garcia and Palacio 1956). It was seven years later that an increased occurrence in the associated condition was recognised (Bodian and Carter 1963). We now know that up to 10% of children with Hirschsprung's disease have Down syndrome, and 1% to 2% of children with Down syndrome have Hirschsprung's disease (Heuckeroth, 2015) while (Bradnock et al., 2017) found infants born with Down syndrome were forty times more likely to have Hirschsprung's disease.

Moore, (2018) concluded children born with Hirschsprung's disease and Down Syndrome may achieve an acceptable degree of bowel continence following definitive surgery, but there was a less confident prognosis for these children. In a comparison of children born with Hirschsprung's disease, with and without associated Down syndrome, Friedmacher and Puri, (2013) concluded that children with co-morbid Hirschsprung's disease and Down syndrome continued to have persistent bowel incontinence and a higher risk of the serious life-threatening complication Hirschsprung's associated enterocolitis.

Approximately 45% of children born with Mowat-Wilson syndrome, a disorder caused by mutation of the ZEBR2 gene, also have Hirschsprung's disease (Coyle and Puri 2015). They also found because children with Mowat-Wilson syndrome have an increased prevalence of long segment Hirschsprung's disease, they are more likely to have ongoing bowel problems following definitive surgery. Thus,

children with Mowat-Wilson syndrome have an increased surgical prevalence of stoma formation compared to those who did not have the syndrome (Coyle and Puri 2015).

Children with congenital central hypoventilation syndrome (CCHS) – a rare, life-threatening condition caused by a mutation in the PHOX2BG gene may suffer from multiple disorders reflecting prenatal neurological development, including Hirschsprung's disease. 1.5% of all cases of Hirschsprung's disease are in children with CCHS. These children are also more likely to suffer a severe form of Hirschsprung's disease, with 10% of them suffering total colonic Hirschsprung's disease (Croaker et al 1998).

There are a set of common features associated with anorectal malformations which are arranged into two groups, acronymically denoted as VATER and VACTERL. VATER is a condition associated with Vertebral defects, Anal atresia, Tracheo-esophageal fistula and Renal anomalies. VACTERL has the additional anomalies of Cardiac defects and Limb abnormalities (Ratan et al., 2005; Solomon, 2011).

4.2 Complications of Hirschsprung's disease.

A serious complication of Hirschsprung's disease is Hirschsprung's associated enterocolitis (HAEC), a potentially fatal bowel infection. The diagnosis of HAEC is often slow or incorrect due to complex symptoms that may be misdiagnosed as viral gastroenteritis. Recent data shows HAEC ought to be suspected in children presenting to the emergency department with symptoms of lethargy and abdominal distension, with investigation using plain X-rays of the abdomen

(Gosain et al., 2017; Nakamura et al., 2018; Sellers et al., 2018). Sadly I have experienced two children, one a baby, one a toddler who have died from HEAC.

4.3 Long-term outcomes of surgical interventions

Studies of long-term outcomes of surgery for Hirschsprung's disease or anorectal malformation are critical, but technically challenging. To undertake them, hospitals must keep adequate registers and records of surgery, and up to date details of young people as they grow up; and the surgical community must be interested enough in these outcomes to invest in such studies. Most countries do not have national linked data systems for health care. It is not surprising that the most detailed study to date (Meinds et al 2019), has emerged from the Netherlands, which is a pioneer in data linkage across health services.

This body of literature is beginning to demonstrate that faecal continence can be a delayed outcome for children after surgery, sometimes not until late adolescence, with a small group not achieving it at all. For example, in a single institute study in Japan over a period of 47 years followed up 153 children who had reached the age of at least 17 years (Ieiri et al., 2010). They determined that two-thirds of children born with Hirschsprung's disease and treated with definitive surgery in childhood, had achieved "excellent" bowel continence by adulthood. However, 19% of the population still experienced intermittent soiling, and 17% experienced faecal incontinence.

In a follow-up of a retrospective cohort of ninety-eight children using self-report questionnaires of children's long-term experiences with continence, Catto-Smith et al., 2007) found 26% of the overall population diagnosed with Hirschsprung's disease between 1974 and 2002 that faecal incontinence was common after

surgery for Hirschsprung's disease and had a significant impact on social activities. For the subset with Down syndrome and Hirschsprung's disease, an unexpected outcome was adverse reactions to certain foods (Catto-Smith et al., 2006).

In a 22.5-year follow-up of children treated at the Eastern Ontario Children's Hospital, bowel continence was relatively poor in children under the age of 15 years, though it improved by late adolescence (Yanchar and Soucy, 1999). The authors pointed out that suffering incontinence into mid adolescence had an impact upon the quality of life and social well-being of children which may not have been appreciated by surgeons.

The most rigorous long-term study was conducted in the Netherlands, (Meinds et al 2019) in a multicentre study of children treated in all six paediatric surgical clinics. Researchers were able to enrol 346 participants (55.5% of the total population) over the age of 8 years, of whom half were still under the age of 17 years, and half were adults. Compared to healthy controls in the Netherland population, adults who had been surgically treated for Hirschsprung's disease were more likely to have incomplete evacuation (47% vs 27%), and faecal incontinence (16% vs 6%). Adults who had been treated with Hirschsprung's disease had lower rates of faecal incontinence than children and adolescents (16% vs 38%).

All these studies have affirmed that quality of life indicators are affected by faecal incontinence, and that faecal incontinence in childhood and adolescence is particularly deleterious for quality of life.

4.4 Patient and Family Support Groups

This section addresses the literature on patient and family support groups in general. In some cases, a support group is formed and run by people who join together on the basis of common experiences to help one another, often the common experience being similar medical conditions or life situations. These people may eventually come together in a group to provide emotional support, share information, education and advice on various aspects of their condition or situation. These groups aspire to offer a better understanding of the condition; management options and how to navigate the service systems available; these aspirations are yet to be proven.

Generally, groups are formalised out of a need for a contact point for new members and professionals along with the drive and passion to meet again and compare notes, with the aim of improving the quality of life of its members and offer support to those who find themselves in a similar situation. Often simply the relief of being able to discuss their situation with someone who has shared similar life experiences and who truly understands what the issues are encourages people to remain connected. PhD students reported feeling understood and supported by other group members, which helped them feel less isolated and anxious (Panayidou and Priest, 2020). Apart from providing emotional and practical support, support groups can be a much-needed resource of shared information (Lee and Meadan, 2021; Mikal et al., 2021; Nielsen et al., n.d.; Panayidou and Priest, 2020b; Roesch-McNally et al., 2021; Tonkin-Hill et al., 2022)

Support groups generally do not operate for profit and rely heavily on donations and small membership fees to provide their services to members. Enquiries through a health care provider may offer links to organisations already formed or with consent of both parties by being put in touch with others in a similar situation if no formal group exists (Ball et al., 2021; Parsons et al., 2021; Pilon et al., 2021). In some instances, simply the commonality of a group of people in each situation can develop informal forms of a support network. (Scannell-Desch, 1999) studied female nurses who served in Vietnam and remarked on the depth of bonding these nurses experienced. Whilst they may not have formed an official 'support group' as such, many have continued a very close unique bond for over 35 years because of their joint experience during the war.

Some groups do provide an active therapeutic space. The power of self-help groups is famously demonstrated by the very successful program; Alcoholic Anonymous (AA) which had its origins on 10 June 1935 (Oka and Borkman, 2000); this day is well known as the Day of Dr Bob's last drinks and marked the foundation day for the organisation. AA began following the meeting of two men referred to as Dr Bob, an Akron surgeon, and Bill W, a New York stockbroker who were both hopeless alcoholics. They supported each other towards sustained sobriety and were eager to support other alcoholics to recover from this addiction and lead normal lives. It took another three years before the Alcoholics Foundation was formally established. By 1939 over one hundred Alcoholics had become sober following the principles developed by AA, now known as the 'Twelve Steps of Recovery Alcoholics Anonymous 2010'.

In contrast to AA, early forms of self-help and support groups for mental illness around the 1960s were focused around supporting the relatives of people who were diagnosed with a mental illness rather than the patient themselves

4.5 Support groups in Australia

One of the earliest support groups formed in Australia was the Benevolent Society which began following a 'Meeting of Friends' in 1813 initiated by Edward Smith Hall. The group began by providing outdoor relief to the poor, blind, aged and infirm within the struggling colony of New South Wales. Initially it was known as the Society for the Promotion of Christian Knowledge and Benevolence and five years later changed to the Benevolent Society (Evans and Curthoys, 2013). In 1920 the society cared for women during labour which led to the formation of the District Nursing in Australia and social welfare programs (Gaudron. M, 2008).

The very early groups catered more for the general community who were unable to care for themselves due to poverty or disability. In 1860, a Scottish migrant Thomas Pattison who was deaf formed the first support services for a specified disability, the Deaf and Dumb Institution in Liverpool Street Sydney. The name and location have changed over the years to reflect the change of focus to that of education of deaf and blind children and the greater links to the community. It is currently known as the Royal Institute for Deaf and Blind Children (*Royal Institute for Deaf and Blind Children*, n.d.).

In 1924 while convalescing following gall bladder surgery, Reverend Stanley Drummond established a charitable organisation called The Far West Children's Home in Manly, to allow children from remote areas, respite with a holiday by the sea. With the help of Dr Montcrieff Barron, who provided honorary medical

services, the organisation became a health care provider. In 1931 Drummond began the first baby health clinics to remote areas The organisation – now called Royal Far West - still stands today and provides services for children from outback New South Wales. (Winks and Pike, 2006).

Community based organisations then evolved into self-help organisations such as The New South Wales Society for Crippled Children which was formed in 1929 and changed its name in 1995 to The Northcott Society in honour of Sir John Northcott who was Patron of the Society from 1946 to 1957. The Society owes its origins to the combined efforts of Sir Robert Wade, a prominent member of the British Medical Association and Mr B Gelling, a member of the Rotary Club. Gelling's advocacy for crippled children in New South Wales resulted in the Rotary Club of Sydney making "*The Education and Vocational Training of Crippled Children*" their community project for 1928-29.

The Northcott Society's long-term objective was to "*prepare the children with disabilities, emotionally and socially, to take their place as regular members of the community*". The Northcott Society's success has been attributed to their ability to remain committed to their objectives and a willingness to adapt and change with the times by establishing new projects as the needs of its members arise (Fletcher, 1998). This is evident in the Society's history of developing and subsequently closing services such as Orthopaedic Hospitals, Special Schools and Sheltered Workshops. The closures were due to financial decisions and services were becoming more community focused rather than their original ideas of educating children with special needs in isolation. Initially, the Society believed this isolated form of education to be the best way for providing additional support

whilst not hindering the progress of children not affected. The Northcott Society continues to grow and develop with the introduction of early intervention programs; respite services; open employment and post school option programs (Fletcher, 1998).

The Diabetic Association of Australia was founded in 1938 by Sir Kempson Maddox to provide benefits and services and to be a powerful voice for those living with diabetes. The organisation continues to provide educational programs and advocacy services, and to fund vital research into better treatments so those in the diabetes community can live their best lives (*We Have a Long and Proud History*, n.d.)

In 1952 a mother set up the Epileptic Welfare Association in response to the appalling treatment of her daughter who suffered from severe epilepsy, and at the age of nine years was placed in a psychiatric hospital as that was the only place that would take her (Trusler 1952). In her words, her daughter came out of the hospital 'a broken child.' Determined that no other child would have these experiences she started the support group currently known as Epilepsy Action Australia.

4.6 Support groups for patients with anorectal malformations.

Hirschsprung's disease and imperforate anus are considered rare diseases. To be termed a rare disease, a condition must affect a small number of people compared to the general population and specific issues are raised in relation to their rarity (*Rare Voices Australia*, 2022). According to (Rode, 2005) a rare disease is a disease that occurs infrequently or rarely in the general population".

In order to be considered as rare, each specific disease cannot affect more than a limited number of people out of the whole population, defined in Europe as less than 1 in 2,000 citizens.

Whilst there are a plethora of support groups for any number of conditions such as for mental health, cancer and alcoholism, the supports for families who have a baby born with an anorectal malformation are limited globally to around thirteen groups. Most of these small organisation's rely on donations and small membership fees as they rarely attract government support.

Parents often feel alone and isolated when their child has a rare condition, recognising the difficulty in finding like-minded families who understood their experiences, eased the journey and provided a means of education, understanding and the opportunity to gain experience from others who had walked their path before (Anderson et al., 2013; Gioldasi et al, n.d.; Baumbusch et al., 2019; Oyania et al., 2021). For some older members, mentorship was an important aspect of their membership long after they had gained knowledge and experience themselves. Often members remained active in the groups to provide support as a role model to new emerging members and their families, who were comforted to see their progression and the hope of a life beyond the diagnosis (Doyle, 2015; Heisler, 2007).

Another emerging role of support organisations is the linking with research facilities and clinicians, often the health professionals will approach support organisations to support their research by distributing research information to their members. This provides the researchers with a select group of people, specific to their research (Griggs et al., 2009; Ingelfinger and Drazen, 2011).

The emergence of the "power-to-the-people" social movement in the 1960s was a catalyst to the self-help group development as people wanted to have more control and self-advocacy in their health management (Baldacchino and Rassool, 2006). Schipper (2008), in an address to an International Workshop on anorectal malformation in Nijmegen, Netherlands gave a first-hand account of living with this chronic, congenital condition. Schipper was born with an imperforate anus and is a Board member of the Dutch support organisation *Vereniging Anusatresie*, a support group for people born with anorectal malformations. Schipper explained how in the past, treatment and research has been mainly focused on cure and care. Whilst this was of great importance to save lives, develop surgical techniques and study the genetic causes of the condition, it did little to address the long-term complications such as incontinence. Of increasing concern have been the challenges faced by adults born with this condition. Transition to adult care is rarely a smooth one following the all-inclusive multidisciplinary approach of paediatric care. In contrast, adult care tends to be highly specialised around specific disorders with little interaction between specialists. Many adults born with anorectal malformations or Hirschsprung's disease remain under the care of paediatric surgeons simply because they are unsure of which specialist to be referred to. It is only in recent years that adult surgeons have been required to care for these people as the mortality rate was so high previously. Over the years it has become apparent that the needs of people born with a congenital bowel disorder may well increase as they get older both physically and psychosocially which has seen the change of focus of support groups in this area to one of paediatric to whole-of-life centred care.

In 1995 the Bowel Group for Kids (BGK), formally known as the Australian Pseudo-obstruction Support Association (APSA) was formed by a group of mothers and interested health professionals who sought support from each other due to a lack of information and understanding of living with the once fatal congenital bowel disorders; Hirschsprung's disease and/or imperforate anus. As discussed in the prologue, I am a founding member of this group. We aim particularly to respond to the lack of understanding of management once the surgery is over and how families may help their children to avoid the problems associated with the potentially fatal condition, Hirschsprung's Associated Encopresis.

4.7 Types of support groups

There are many types of groups offering support that have developed out of the needs of members from all walks of life in the community. Some examples of support are, church gatherings, charitable organisations, welfare groups, volunteer organisations, support groups; either professionally or peer led, self-help groups; either individual or combined, mutual-help groups, telephone support services, personal group meetings, and social media groups such as Facebook groups, or group chat networks on Twitter.

With the advent of the internet one of the first things parents do when their baby is diagnosed with a congenital bowel disorder, is search for the condition and find one of the many groups online. The various modes of social media such as blogs, vlogs, webinars, Twitter or Facebook, provide access to online communities for rare conditions from anywhere around the world. Online groups are often established by parents and offer a means of accessing information from

supporting families in similar situations. Almost all are purely dedicated online formats with no personal contact, and most do not provide evidenced based guidance (Wittmeier et al., 2014). Traditional support groups conduct meetings, offer conferences, workshops and printed educational material and often have their own form of social media for members. A newly emerging trend is for specialist hospitals to set up parent support networks specific to the conditions they manage, for example, the Colorectal and Pelvic Reconstruction (CCPR) at Nationwide Children's Hospital in Columbus, Ohio, has set up a closed Facebook group for carers and patients. Whilst groups such as the CCPR are a great resource for parents to share stories, ideas, encourage others and ask questions, they are moderated by the CCPR staff, rather than by parents.

Self-help groups are usually formed by the end user of medical systems, such as a sufferer of a medical condition and increasingly are run by volunteers who are representatives of professional groups such as nurses, social workers or psychologists (Adamsen, 2002). As in the case of Nationwide Children's Hospital's CCPR group, some sites for support of parents are run by hospitals with minimal input from parents. An interesting absence in the field of self-help groups for patients with anorectal malformations is the absence of "astroturfing" groups – that is, the development of an apparent grass-roots support or lobby group which is funded by industry, usually the pharmaceutical industry. Such groups often act to advance the interests of industries, and treatments, while ostensibly being a peer-support group. This probably reflects the absence of a significant interest by pharmaceutical or medical device industries in conditions associated with faecal incontinence, which are untreatable by either industry at present. It is likely that the self-help landscape may change, with the introduction of more

industry players, should precision medicine yield marketable interventions aimed at the genomes of people with Hirschsprung's disease or anorectal malformation.

Although it might seem as though support groups are mainly just for women and children, there are a large number that are set up for an equally wide range of situations specifically for men. Examples include 'Dads in Distress,' which began by offering peer support to men going through separation from their partner and/or children. The organisation has grown to become Parents Beyond Breakup which has three frontline services; Dads in Distress, Mums in Distress and Grandparents in distress programs catering for those who are separated from children or grandchildren due to separation or divorce of the parents.

Two other organisations which have seen rapid growth in similar areas of support recently are Mensheds Australia, established in 2002, and the Australian Men's Shed Association which began in 2005 (Golding, 2008). The two organisations appear to have similar aims and objectives, that of supporting men who are traditionally reluctant to discuss problems and may find themselves isolated and lacking the ability to seek help and support for a variety of reasons such as the loss of employment, financial loss, or death of a long-time partner. The idea is an updated version of the shed in the backyard which has traditionally been seen as an environment where most men feel comfortable. By providing a community shed, men can connect and work collaboratively towards living productive and meaningful lives which in turn, is hoped will provide support for the health and wellbeing of men across Australia (Golding, 2015). In 2010 the Australian Men's Sheds Association was recognised by the Federal Government for the role it plays in addressing social isolation, health and well-being. This was followed by the

inclusion in the National Male Health Policy and funding for the direct financial assistance to Men's Sheds. The organisation has now expanded to include men who are living with long term disabilities or fathers of children with autism which gives them an opportunity to be less isolated in society (Hansji et al., 2015).

As people live longer, there is a greater likelihood they will suffer depression because of isolation through losing loved ones. Mental health in Australia is a growing concern with little resources to support the needs of people suffering with a mental illness. Fortunately, there has been a rapid growth of support; self-help and mutual support groups for family members whose loved one suffers from severe psychiatric disabilities. In America, the National Alliance for the Mentally Ill (NAMI) reported in 1979 just one hundred family representatives came together to form NAMI, by 1997 there were 1100 Chapters and over 1400 members (NAMI 2019).

The lack of traditional support mechanisms such as churches, family and community and isolation of families; working and/or single parents, find people looking for alternatives to this societal inadequacy. The emergence of self-help groups or support groups goes some way to filling those gaps not only for medical conditions but for the myriad of life situations such as divorce, abuse, gambling etcetera (Baldacchino and Rassool, 2006). Support groups vary in the way they are established, funded and run. Some are run by peers who become experts in the day to day lived experience of a medical condition whilst other groups may be facilitated by health professionals who could support patients and their families in making the best use of online information to better manage their condition (Bartlett and Coulson, 2011; Mo and Coulson, 2010).

Support groups generally provide information and education to members; a resource for health professionals and school educators; produce newsletters and offer group meetings, get-togethers and conferences. More active and financial organisations may also advocate for individuals and lobby Governments to implement change that would benefit people with the condition. Dadich, (2006) explored what self-help support groups could offer young people who experience mental health issues. A sample of fifty-three participants undertook open-ended interviews which revealed that most came to the organisation looking for change, while those with poor well-being sought recovery from their condition. Participants reported a sense of emotional support through empathy and friendship and importantly, by way of advice that was of great practical value. They also found being in the support group offered them opportunities to gain experience about their condition and more about the kinds of services and health professionals and treatment options. There was reportedly a sense of connectedness and the ability to expand social networks. One of the great benefits of information that comes out of support group discussions is the wealth of lived experiences rather than being purely biomedical. Self-help support groups can be of benefit to young people offering them a degree of respite (Dadich, 2008).

Under the Commonwealth Disability Discrimination Act 1992, a Standard has been constructed for the right to participate in education and training without discrimination for students with disabilities. Under the Act, it is unlawful for any person (including a child) to be discriminated against by an educational authority based on a disability (Children's Rights Australia 2019). The Disability Standards for Education states…the education provider must take reasonable steps to

ensure that the student is able to use support services used by the students of the institution in general on the same basis as a student without a disability, and without experiencing discrimination (Department of Education and Training, 2005).

Some support groups are well recognised within the NSW Department of Education and their information sheets are used to provide information, education and forms for schools to issue to parents to complete when their child enrols, or the condition is diagnosed. The organisations in reciprocal agreements with schools are the Asthma Foundation, Diabetes NSW, Epilepsy Action Australia, Cancer Council and Hepatitis NSW (NSW Department of Education, 2014, 2016a, 2016b, 2016c, 2017).

An interesting benefit of support groups not immediately recognised is that of the helper-therapy principle as described by Riessman, (1997) which he summarises as "those who help are helped most". As early as 1965, Reissman realised that although the people receiving help may not always benefit; he felt certain that the people giving help profited from their roles. Lynn Wolfson, a person with Hirschsprung's disease who advocates through the US support organisation G-PACT (Gastroparesis Patient Association for Cures and Treatment) provides testament to the benefits of giving back to the community:

> Once we can master our personal challenges, there is no greater joy than giving back to others.
>
> (Wolfson, 2019)

The 'Lindsay Leg Club' model offers another example (Lyndsay. 2004). This is a peer support model for managing wound care and leg ulcers in the elderly, in a

community-type setting with peer support, social interactions and assistance with goal setting. A comparison with this model and a control group managed in the traditional community nurse setting in their own homes showed that the participants in the Lindsay Leg Club group had significant improvement in outcomes in quality of life; pain levels; morale; self-esteem; independence in activities of daily living and ulcer healing. This study shows the potential to apply this model of care to other community type settings (Edwards et al 2009).

Stang and Mittelmark, (2010) explore the empowerment process emerging among participants in a support group of women undergoing treatment for breast cancer. Hospitalised patients often report a feeling of lack of control and vulnerability during an illness, hearing from others who were optimistic helped those who were naturally pessimistic in their outlook. Participation in a self-help group for people being treated for breast cancer helped to empower patients and provide a valuable contribution to recovery. In a study of support groups for Australian Indigenous women, one woman expressed how her experience of hospital treatment for breast cancer made her feel useful to those who were afraid of the pending treatment (Finn et al., 2008)

SWSLHD (1980) in self-help groups and its impacts upon people with a life-threatening disease such as cancer or HIV. Her findings were encouraging in that participants reported the following benefits on order of importance, coming to an understanding of one's problem, spending time with others in the same or similar situation, a strengthening of relationship and family relations, receiving understanding and support, learning from other people's experiences, forming

new friendships in the group, an improved sense of being normal, knowledge of the body and the disease.

Self-help groups are also useful for siblings within families. Nolbris et al (2010) studied children whose siblings had been diagnosed with cancer and were undergoing treatment or had died as a result. They found that siblings had a greater sense of togetherness and shared experiences with others in the group and were proactive in helping each other with advice and encouragement.

Support groups offer a welcome adjunct to the existing services by providing support and hope from those who have lived through the experience and can offer insight and experience to newly diagnosed people. Frese and co-authors (2001) identified a problem with the focus in recent years on evidence-based practice for mental illness, which is conventionally treated by many health care services as being of little relevance to the everyday struggles experienced by patients dealing with mental illness. Policy makers have realised the importance of combining evidence-based practice with consumer led support services to provide optimal recovery potential. Self-help groups are credited with providing "decreased symptoms, increased coping skills, and increased life satisfaction among long-term versus short-term members" (Salzer, 2002). The evidence-based practice movement is clarifying the areas where self-help groups are most useful in improving health outcomes; for example, there is good evidence self-help groups are not suitable for people in an acute psychological crisis (Adamsen 2002). In these situations, trained mental health professionals were required.

Although there may be many organisations for similar conditions or the same organisation in different states, they may not perform the same functions or

support activities. This is mainly due to the individuals involved and the needs of the geographical area. Mannion et al (1996) compared the characteristics of members of self-help support groups for families in relation to mental illness with those of people with non-members. They found that members of support groups tend to have smaller social networks and therefore, that the lack of social support could be an incentive for people to join support groups. Support group members were more likely to have a high-functioning relative, and to utilise more developed adaptive strategies. Family members who are struggling with a chaotic relative may find it difficult to join support groups – indicating that outreach and partnering models may be needed for such family members to access support groups.

Mandell and Salzer (2010) found parents of children with autism were more likely to join support groups if their children were self-injurious; had sleep problems or severe language deficits. Another interesting finding from their survey was that parents joined support groups if their diagnosing clinician referred them to a group. Clinician referral is poor in congenital bowel disorders partly because the clinician may not see the need as the condition is considered largely a surgical problem and long-term issues are not always anticipated. Often when parents do find a support group for Hirschsprung's disease or anorectal malformation themselves, they will say they wish they had been referred to the support group earlier.

Face-to-face support groups are not preferred by all sectors of the community. Adolescents found the anonymity of an online suicide prevention community was their only solace in times of distress (Greidanus and Everall, 2010). Being able to

express their true reasons for seeking help without having to see someone allowed them to feel comfortable to reach out for help. Trained crisis intervention moderators were online to assist with professional social support and refer people to offline services. Those who had received help soon became custodian of the helping role and therefore the helped became the helper (Greidanus and Everall, 2010).

A common theme reported throughout the studies was the sense of belonging and comfort being part of a group of people who shared the same experience engendered for participants. Benefits included helping each other with advice and encouragement, leading both participants - adviser and recipient – to having a sense of control and an improvement in coping with their situation. This is consonant with Reissman's conceptualisation of his helper therapy (1997). This unexpected benefit was also noted by Heisler (2007) who found that peer support models also potentially benefit both those receiving and those providing support. The unifying feature of these programs is that they seek to build on the strengths, knowledge, and experience that peers can offer. Peer support interventions build on the recognition that people living with chronic illnesses have a great deal to offer each other; they share knowledge and experience that others, including many that health care professionals cannot always understand. If carefully designed and implemented, peer support interventions can be a powerful way to help patients with chronic diseases live more successfully with their conditions.

The evidence base for the expert patient model of peer-led health facilitation was developed at Stanford School of Medicine Chronic Disease Self-Management Program (1997), using Bandura's social cognitive theory of behaviour (Bandura,

1978). This model pointed to the key predictors of successful behaviour change as being confidence in the ability to conduct the action and an expectation that it is achievable. Patient-led self-management programs for chronic disease received a considerable boost with the NHS Expert Patient Programme, launched in England in 2001 (Griffiths et al., 2007). This followed the Stanford model, of a six-week programme aimed at improving self-management for different chronic diseases facilitated by lay volunteers. Implementation was tricky, in part because the model could be co-opted inadvertently into a medical dominance framework (Fox et al., 2005). However, the model has proven with many conditions to be a cost-effective strategy to reduce the severity of symptoms, decrease pain, improve life control and activities, and enhance the satisfaction of the patients in the UK (Lorig et al., 1993; 2008). Critics have warned that the model may be oversold as a way of shifting costs and responsibilities for their recovery onto patients themselves, under a rhetoric of respect (Rogers et al.,, 2009) and that the model may have limited applicability to poor countries where access to adequate health care delivery is still insufficient (Xiao et al., 2015).

4.8 Knowledge Gaps in the literature

One of the major gaps in the literature is recognition of, and programs to meet the needs of children born with bowel disorders, who often suffer in silence and are ostracised during the early years at school due to faecal incontinence. Having a bowel disorder is not something children wish to share openly and fails to attract community support or funding for better services.

Schipper (2008) explains the role of patient organisations as threefold: education and provision of information, patient to patient support, and advocacy. The

importance of early referral to support groups by hospital staff is imperative for families who have a baby born with a congenital bowel disorder such as Hirschsprung's disease or an anorectal malformation. This is a time when they are confused, frightened and vulnerable as their newborn baby requires lifesaving surgery for a condition most have never heard of. (Schipper, 2008) points to the fact that this early referral to a support group should be standard practice but is underestimated by the medical community and is rarely practiced. A common misconception is that Hirschsprung's disease or anorectal malformation are surgical problems and once the surgery is over the problem has been solved. We now know this is not always the case. Long term management from babies requiring anal dilatations cause distress to the parents who feel they are hurting their child and worry the children may have long term psychological problems from the procedures. The adults with Hirschsprung's disease or anorectal malformations may no longer need these treatments but may still need further surgical intervention to enable devices for antegrade enema treatment in an attempt to have some level of control of their continence during the day.

Whilst there have been some attempts at determining the effectiveness of support groups, few papers have addressed the benefits to clinicians from being able to provide a means of support to their patients on the lived experience with chronic conditions. (Emerick, 2014) noted that support groups tended to fall into three main categories; those that were "supportive" groups in which partnerships with professionals could occur but were problematic were considered moderate. Then there were radical "separatist" groups, he found these to be a minority of those represented and they were unlikely to succeed in partnership with professionals. The groups most likely to succeed were called "partnership" groups

and were found to be more conservative and worked with health professionals to provide support for clinicians and patients.

Whilst most papers analysed were able to list the benefits of support groups on general wellbeing and a sense of connectedness with others in a similar situation; there was an obvious lack of evidence to prove that belonging to a support group actually led to a better physical outcome for patients or their families. A model called 'Peer Coaches' was described as a way of providing peer support for patients with diabetes and other chronic conditions (Heisler, 2007). Volunteers undergo eight to thirty-two hours of training with a focus on communication skills, empathic listening and supporting patients rather than attempting to take over the role of health care provider. This model has been used in providing support for patients with a wide range of conditions, such as breast cancer; prostate cancer; postpartum depression, heart failure patients and HIV/AIDS. Her work identified the need for further studies into the best method of integrating peer support services into existing clinical and outreach services. This is a different model to the expert patient model described in the previous section, where expert patients could be devolved into a quasi-professional role with the health sector. The peer coach is a side-by-side model in which the focus is primarily supportive.

In a study looking at patients who had long-term experience of using the antegrade continence enema procedure (ACE), Yardley et al., (2009) found that patients transitioning from paediatric to adult services were vulnerable and often felt isolated and abandoned. The reasons related to going from the "wrap-around" model of care in the paediatric setting to the insular adult care where

surgeons were unfamiliar with the conditions or the specific continence management of the ACE procedure.

Gartner and Riessman, (1982) found that the inclusion of peer support services into traditional mental health services enabled a greater numeric outreach to people with mental illness in the community and assisted people to become more independent and interdependent. A study exploring the quality of life of patients born with Hirschsprung's disease or an anorectal malformation found that those who had a severe form of the disease or had an additional congenital abnormality would benefit greatly from inclusion in a support group (Hartman. et al., 2008). This was particularly the case with girls. The rationale was that these girls with severe disease were particularly vulnerable to loss of self-confidence and self-imposed social isolation, for which support groups may mitigate some of social distress.

4.9 Conclusion

In this chapter, I have reviewed the literature discussing Hirschsprung's disease or anorectal malformations, associated conditions and support groups. Whilst there is a large volume of biomedical literature relating to these congenital bowel disorders, there is a relatively new literature on the long-term follow-up of persons who have Hirschsprung's disease and anorectal malformation. These painstaking, long-term studies have indicated that faecal soiling and incontinence is not necessarily cured by the definitive surgical treatment that aims to corrects distorted anatomy. This is not because the surgery itself causes the poor outcome, but rather points to the persistent neurological dysfunction associated with the condition.

Improvements in continence occur into adulthood, indicating that the highest rate of post-treatment incontinence occurs during the most vulnerable developmental years of childhood and early adolescence. If this knowledge is only now emerging in the surgical literature, then the completeness of information given to parents making complex decisions about their children's health appears to be inadequate. Thus, there is a critical need for ancillary support for parents and families from the point of diagnosis to adolescence.

This chapter has indicated that there is a critical knowledge gap in the literature on the importance of parent support for the caregivers of these children. It is this support from families who understand the complexities of the day to day caring for a baby or child who has undergone major bowel surgery in the first few weeks of life that is vital.

Charities, self-help groups and community-based support groups have been around for centuries in various forms; from support networks of communities brought together by circumstances; life experiences; illness or conditions. They have evolved from the Victorian model represented by Alcoholics Anonymous (AA), to the current disease specific organisations represented by the Bowel Group for Kids (BGK) and others. Little objective evidence is available to inform clinicians or educational institutions for children of the role and effectiveness of organisations specific to faecal incontinence, or the effectiveness on health outcomes for patients who are members of a support group. In the following chapters the thesis will provide insight from surveys responded to by parents, paediatric surgeons and parent support groups on the benefits and challenges of

being a member of a support group aiming to improve the day-to-day lived experience for children born with a congenital bowel disorder.

The focus of support groups today has changed considerably from face-to-face meetings and attendance at annual conferences to online support groups on social media sites, virtual conferences, webinars and vlogs (Bridson et al., 2021; Brooks et al., 2021; Carter et al., 2012; Eysenbach, 2005; Gothberg et al., 2013; Mbah, 2022; Mitchell et al., 2005; Van Herck et al., 2021).

Whilst there is a plethora of research into support groups, how they are formed, who runs them, who the participants are and how they are supported or support others in a similar situation, the gaps in the literature are the lack of ability to see the needs of peer support from different perspectives. My original contribution to knowledge in this thesis, is the ability to understand and share the needs of patients, health professionals, parents and most importantly, the participants of a support group. This thesis brings together a unique collaborative insight gained from the lens of a nurse who cares babies born with a congenital bowel disorder; an educator of future nurses and doctors, as a mother of a child born with Hirschsprung's disease at a time when there was little knowledge of the long term consequences of the conditions and as co-founder of the first support group in Australia to offer knowledge, education, and support for these rare conditions.

CHAPTER 5: METHODOLOGY

5.1 Introduction

This chapter outlines the qualitative and quantitative methodologies used in this thesis. The aims of this research were to identify the impact of Hirschsprung's disease or imperforate anus on the lived experience of children and their families, identify the impacts of peer support groups of these families, and the challenges they face when dealing with faecal incontinence. To explore this research area, I have adopted a framing institutions approach (Watkins-Hayes et al., 2012), arguing that children and parents of Hirschsprung's disease navigate several framing institutions which construct their own experiences of the illness, and impact on how positive and negative these trajectories may be. The framing institutions are the hospital, an arena which includes the staff of the hospital; the profession of surgery and its technical interventions; schools; and parent groups. Additional frames come from the experiences of other children and other parents. In this chapter, I first describe framing institutions as a theoretical approach. I then describe the qualitative and quantitative approaches used to explore the impact of soiling on young people with Hirschsprung's or imperforate anus from different analytical perspectives.

5.2 Theoretical approach

In Western medicine, the biomedical approach tends to present the patient as defined through their diagnosis which addresses their physiological or anatomical disorder. This disorder is treated, or reordered, by the physician. While it is useful, and sometimes essential, for the patient to receive medical or surgical intervention, what is missing is the opportunity to understand how the condition

impacts on the person's day to day existence. The biomedical approach focuses on the condition the person is suffering to the exclusion of being able to recognise the psychosocial effect; the focus is on the disease not the person (Priya, 2012). Patients suffer illnesses, doctors diagnose and treat diseases (Eisenberg, 1977). The role of the doctor in the neonate with an apparent bowel obstruction is to investigate, diagnose and treat the disease. The biomedical focus on disease has the benefit for neonatal surgeons of making their job technical (dealing with the non-functional segments of bowel, aligning functional segments of bowel). However, the biomedical model does not help children and families address the areas that are "bracketed out" of their conceptualisation of the problem of Hirschsprung's disease or anorectal malformation – the fact that conditions that address the distal rectum and anus will accrue to the sufferer a socially isolating burden should the distal rectum and anus not become fully functional.

In this thesis, I juxtapose the biomedical view of Hirschsprung's disease or anorectal malformation with the concepts of stigma and framing institutions that may extend or mitigate the stigma experienced by children with incontinence and their families.

This thesis fills the gaps revealed in the literature review. Whilst the literature focuses primarily on the various approaches and new developments in surgical techniques, it is the lived experiences of the people affected by congenital anorectal malformations that are essential to understand the complexity of the outcomes following surgical correction. The innovative strategies to which parents' resort to help their children engage in normal everyday activities are discussed and the importance of sharing coping strategies through peer-to-peer

support groups are explored. Whilst the format of support groups has changed from likeminded people meeting in person, to sharing their stories on social media; the underlying concept of experienced parents supporting new families to navigate the complex needs of children who deal with faecal incontinence daily remains the focal point of any support organisation.

5.3 Framing institutions approach

The importance of framing institutions cannot be underestimated in the lives of children born with Hirschsprung's disease or imperforate anus. Whilst the hospitals and clinicians play a vital role in saving these children's lives through surgical intervention, it is the ongoing support and acceptance of the community, schools and support organisations that can help facilitate the child's coping strategies (Watkins-Hayes et al., 2012).

Stress is an emerging problem in many people living with a chronic illness (Compas et al., 2012), and this is particularly so for children born with Hirschsprung's disease or anorectal malformation. Hirschsprung's disease has a long history of confusion around the aetiology of the condition, which was first described in 1691, but it took until 1886 before the condition was recognised as a congenital bowel obstruction and then until the 1950's before surgical approaches were successful in removing the affected segment of bowel and survival became a possibility. As such, the focus since that time has been on perfecting surgical techniques. Faecal incontinence is seen as a problem, a failure, a burden, an impairment and something to be endured in secrecy, when in fact it is a chronic illness that with a shift in focus from a supportive

environment in society, schools and parent support groups could drive a change to the stigma associated with it. Interpretation is important in helping children see their outlook as a challenge to be overcome with strategies and support rather than a problem to be endured in secret (Burry, 1991; Thomas, 2012). In this thesis, I explore the ways that different institutions – from biomedicine to the family, to school – all contribute to the framing of Hirschsprung's disease or anorectal malformation as a chronic disease, rather than an acute surgical problem. These institutions do not necessarily work in concert; in fact, as was discussed in Chapter four the literature indicates that the biomedical framing of Hirschsprung's disease or anorectal malformation did not move towards a sharing the institutional frames of the family until the longitudinal follow-up studies of Hirschsprung's disease or anorectal malformation surgery had indicated that failure of full correction of the neurological abnormality (in terms of achieving full function) might be the norm. This move to shared institutional framing by family and biomedicine is now influencing the framing of the condition by schools, whereas will be seen in Chapter 7, the framing of Hirschsprung's disease or anorectal malformation (and faecal incontinence, which the major symptom noted at school) has borrowed from behavioural psychology and moral notions of good parenting.

5.4 Storying as a method to understand

Participants within this research generously shared their stories during workshops, at conferences, through social media, over the telephone and during interviews. It is through story telling that we have the privilege of gaining insight into the day to day lived experience of the people affected by varying degrees of

faecal incontinence and how this invisible disorder affects lives of the children and their families. Without storying, we cannot begin to understand the affects a silent disorder has on a child's life and the challenges parents and carers face every day in an effort to protect their child from isolation and damaging comments. Storying sits well in this research which is qualitative based (Gravett. 2019).

5.5 Navigating stigma

The second theoretical approach used in this thesis is the notion of stigma, and its impact upon children. this section looks at stigma and its impact on children. When a child suffers faecal incontinence, the effects for the child and the family are far reaching. Faecal incontinence/soiling is particularly stigmatising and makes navigating this journey very hard. Goffman (1963) defines a stigmatised person as one who possesses "an attribute that makes him different from others...and of a less desirable kind" (p.3). Goffman defines a set of criteria for depth of stigma, noting that this is directly related to its visibility, obtrusiveness and whether it occurs in a range of situations. All these criteria apply to faecal incontinence; it is highly stigmatising as it is something the child is unable to hide due to the unpredictable nature of which the child has no control.

In Goffman's terms, stigma is a relational state, in that it is determined by another person's reaction, leading them to deploy stigmatising actions or words; this is then internalised by the person and results in what Goffman called "a spoiling of normal identity." The condition which attracts stigma is not in itself foul or fair; stigma is not determined by attributes. This notion of stigma is very important for this thesis, because it assumes that stigma can be ameliorated, modified or

expunged through attention to relations, rather than to the attributes of the condition itself. In other words, stigma can be minimised not by the kind of deep internalising of shame which Greg Ryan (2019) describes in his account of his life with imperforate anus, but by increasing the openness and support around the person who is being stigmatised. The sense of spoiled identity, again, is not a developmental, internal state, but rather one that reflects the perspective of the stigmatising other. Goffman's notion of stigma also enables us to understand why this condition is often kept secret by people who have it; stigma cannot be created for a condition that is not known.

In his classic book, *A Sociological History of Excretory Experience*, David Inglis provides a rare study of changing attitudes to excretion through history (Inglis, 2001). He notes that this necessary physiological activity is the subject of taboos, disgust and shame, arguing that has escalated since the advent of the bourgeoisie and the invention of reliable plumbing and waste management systems. These have removed the practice of excretion from the street, or the forest, or the plantation – if not in plain view, at least undertaken with the certain knowledge of the community – to small, enclosed spaces. The fact that domestic sanitation became known by the architecture surrounding it (a small "closet" for water) rather than the much more impressive technical achievement of piped water or flushing technology tells us about the desire for these actions to be made private, removed from the public eye. Inglis terms this notion of how we should manage defecation "faecal habitus" – the cultural organisation and interpretation of faecal events.

Inglis, (2001) sees faecal habitus as a combination of two factors: the cultural symbolic classification borrowing from dirt and impurity, and applied to faeces, which leads it to be viewed as dangerous and contaminating; and a set of excretory practices that carry symbolic meaning. These excretory practices are (a) private, (b) involve regulation of sensory experience of sight, sound and particularly smell, which is profoundly connected to emotional readings; and (c) are constructed and deflected through words that euphemise or hide the practice. In general, excretion is not a polite topic of conversation; we talk about it through joke words or through words that society regards as harsh.

Faecal habitus enforces and rationalises the application of stigma to children and adolescents affected by faecal incontinence or soiling due to Hirschsprung's disease or anorectal malformation. As can be seen in the story of my son Adam, the excretory practices which are regarded as acceptable faecal habitus may all be violated in a young person with faecal incontinence or soiling due to Hirschsprung's disease or anorectal malformation. It may not be private, as in the case described in the experience of my child's explosive faecal incontinence which was a regular occurrence. It is difficult to regulate sight and sound and particularly smell. Incontinence is often noted by smell first, which sets in motion an immediate disgust reaction among others – a reaction which might have been resisted if the first sensory warning was through sound or vision (Krusemark et al., 2013). It is clear however, that with time this affective reaction can be managed since families and carers are able to disconnect the affective impact of the smell of defecation. Finally, we do not have a language that enables easy conversation about defecation. A medical condition of serious weight is often reduced to using non-technical, euphemistic language. To bring about change for

these families we first must understand what is known about the condition and how it is managed by gaining insight from the key stakeholders. To enable this, the methods used in this thesis were to explore how families manage this difficult situation; the parents understanding of the condition, how they were given the diagnosis, how much they wanted to know about the long term effects of the condition, what services were available and what they found helpful to support their child. Support groups relevant to congenital bowel disorders were surveyed to understand how they were set up, what methods were used to support parents, their degree of interaction with health professionals and hospitals. Paediatric surgeons were surveyed to gain insight into their understanding, and use of support groups and finally the paediatric hospitals were surveyed to understand how support group information was displayed and distributed to families. This broad collating of information was then combined with my own lived experience as a nurse caring for these children, as the mother of a now adult son born with Hirschsprung's and cofounder of a support group formed to help parents and the wonderful stories from parents and older children collected over many years that provided a rich collaboration of information which is my original contribution to knowledge on the subject of faecal incontinence.

5.6 Conceptual approach

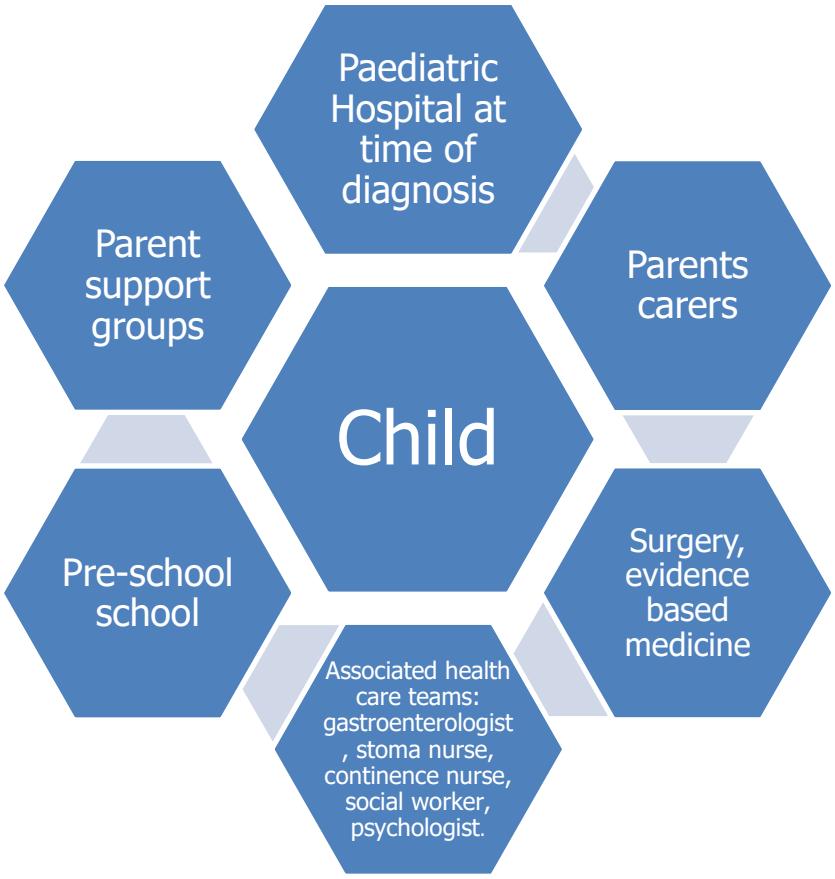


Figure 10: Conceptual approach used in this thesis of different frames.

The conceptual approach uses the concept of institutional frames in different domains of the life of the child with Hirschsprung's disease or anorectal malformation being the parents, the school, hospitals, health practitioners and peer to peer support services. These institutional frames surround and determine the way in which the child experiences (or does not experience) stigma. The frames are: biomedicine (as typified in surgery and evidence-based medicine); the support teams of allied health workers and psychologists, who use a social model of health and biomedicine together to frame the condition as primarily biomedical with some social underpinnings; the hospital institution, which as an organisation has its own norms and regulates when and how the child is reviewed, and how they will be transferred to other organisations, especially as they become adults; parents and carers (the institutional frame of the family);

school (the institutional frame of behavioural psychology and education); and parent support groups (a political institutional frame about equality of opportunity and democratisation of knowledge sharing).

The methodological approach deployed in this thesis, which involves several different data collection methods and analyses, is located across all of these domains, in order to describe the different institutional frames. Whilst the data collection provides an analysis, it is my original contribution to knowledge in this subject that collates the depth of understanding from all institutional frames. As a nurse who cared for these children in theatre, I could relate to the situation not only on a professional level but a personal one as the mother of a child who had endured the same fate, I also provided support to the parents before and after surgery as the cofounder of a support group set up to help parents navigate this new and daunting arena they were thrust into when their baby was diagnosed with a rare congenital bowel disorder. Whilst teaching future nurses and doctors, I incorporate the importance of listening to parents and research available support systems that the family may find helpful.

5.7 Methodological approach

A summary of the research methods is presented in Table 6.1. Much of this research is qualitative. Through qualitative research I was able to ensure that parents were able to express their own feelings about how having a baby born with a congenital bowel disorder has affected them and their family's life personally. This allowed a window into the complex, sensitive issues they have encountered, the strategies they have put in place for their unique situation and where the gaps are in supporting families (Berg, 2001; Finlay, 2011). The

sensitive nature of faecal incontinence also required a qualitative approach to unpack the complexities which are unique in children. Qualitative research was used to explore this sensitive area where there was a lack of existing literature in the lived experience of these families, has been utilised (Liamputting 2013).

The study required a sensitive approach given the nature of faecal incontinence which is rare and can be stigmatising for the child and the family. Given most of the literature for these congenital bowel disorders is biomedical and focuses on the surgical intervention, the lack of published literature on the lived experience of the child and their families, a qualitative approach was appropriate. Each participant's contribution formed a valuable insight into the lived experience of families who have a baby born with either Hirschsprung's disease or an imperforate anus. This allowed a unique view of the participants' experience which it is hoped will help gain a greater understanding of the needs of children and their families who, despite having survived many years of surgery, interventions and bowel management programs, are unable to attain complete control of their bowels prior to starting school. It is the richness of parent and older children's stories that brings meaning and understanding to what these children and their families endure, without a story, how can decisions be made to improve their situation (Kahneman, 2011)

A literature review search strategy utilised combination of the following terms: support groups, support organisations, anorectal malformations, Hirschsprung's disease, imperforate anus, and long-term follow-up. More than one hundred individual articles, reports and several theses have been analysed for the key themes that will be explored in the research.

Table 5.1: Summary of research questions, methods, participants and sites included in this thesis

Research question	Participants	Site	Method	N
Determine long term follow up of patients and their use and opinions of support groups	Paediatric surgeons	Australia, New Zealand; England; France; Germany; Italy; Colombia; Austria; Iran; Iraq; Netherlands; Poland; Russia; Sweden; USA	Survey	45
Understand the way care of families during their long careers. Has there been a change in the involvement of families.	A nurse unit manager (NUM) who has over thirty years of experience in charge of a major paediatric hospital ward. A paediatric ward nurse who has over twelve years' experience caring for these children.	Canada Australia NSW	Key informant interviews and informal discussions	2
Understand the difficulties experienced by parents at critical stages of their child's lives. Understand what parents found helpful throughout these experiences.	Parents	Nation-wide; New Zealand; USA; England; France; Philippines.	Participant observation Informal discussions	600
Families experience of a support group for HSCR/ARM.	Parents	Nation-wide	Survey	95
Understand the process and supports available to a child when they have ongoing faecal incontinence following definitive surgery for HSCR/ARM	Educators	Nation-wide	Participant observation Informal discussions Policy review	13
Data collected from hospital patient records for children born with HSCR	Children	Paediatric Hospitals NSW and ACT.	Case series analysis	430
The role and efficacy of support groups in paediatric medical conditions.	Parent support groups	Nation-wide; New Zealand; England; France; Germany; Italy; Netherlands; USA	Survey	28

Do hospitals have a system in place to disseminate information about support groups to families when their child is diagnosed with HSCR/ARM	Hospitals	Nation-wide	Telephone survey	10

5.8 The domain of the parent and carer

The aim of this subcomponent of the doctoral research was to understand the parents' experience from diagnosis to post-surgical experiences. with that of nurses, paediatric surgeons and paediatric hospitals. Privacy was of the utmost concern, the delicate nature of the conditions being researched and the ages of the children whose conditions were being discussed, required all identifying information to be made completely private to avoid any possibility of their personal information being linked back to them in later years through online searches.

Method and instrument: An anonymous online survey, with twenty-one items, comprising six demographic questions, two on the length of post-operative follow-up; one about family involvement, the remaining twelve on parent experiences of support groups, the referral system, and what they found helpful or unhelpful.

Recruitment: Parents were selected from the database of a support group in Australia, along with parents interviewed at a paediatric hospital clinic specific for babies and young children born with Hirschsprung's disease or imperforate anus. Participants' children varied from those who had had a postoperative pull-through surgical procedure to some remaining under the care of paediatric surgeons when they became adult patients. Consent forms were collected, and parents were asked to respond to a Survey Monkey questionnaire which they were able to access online anonymously.

The data in this online survey were supplemented with informal discussions with family members of people with Hirschsprung's disease or imperforate anus,

conducted over 25 years, over the telephone, email, social media and in person at annual conferences with permission. In this I acted as an observant participant (Abu-Lughod 1988), in that the data were also part of my everyday work as a peer-support provider through the support group. Parents would contact me to discuss the birth and unexpected diagnosis of their baby with someone who had experienced the same situation. Parents wanted education to understand the condition, terminology, and some would want to know more about the long-term outlook. There were a minority of parents whose child had a late diagnosis. Discussions on management topics were conducted over social media sites. These ranged from coping with the diagnosis, products used, managing ongoing incontinence after surgery, particularly at school, visiting friends or sleepovers, to camps and the future from the perspective of adolescents.

Analysis: The data in the online survey (n=95) were analysed through descriptive statistical methods. The findings were then considered in relation to the emergent data provided through parent discussions and social media posts.

5.9 The domain of the surgeon and evidence-based medicine

The aim of this component was to describe practices of long term follow up of patients and paediatric surgeons' use and opinions of support groups.

Method and instrument: Anonymous survey, delivered online. The survey consisted of asked seven items relating to Demographics; three items related to non-surgical bowel continence postoperative follow-up; and eight items related to their understanding of and use of support groups.

Recruitment: Surgeons were approached through the Colorectal Club Meeting held annually in different locations in Europe and through social media, email and in person at annual conferences.

These data were supplemented by a critical review of the surgical literature on outcomes of Hirschsprung's disease and imperforate anus, noting the different ways in which "acceptable outcome" was framed.

Analysis: The data in the online survey (n=45) were analysed through descriptive statistical methods. The findings were also analysed in relation to the framing of outcomes research for surgery for Hirschsprung's disease and imperforate anus in the scientific literature.

5.10 The domain of the allied health workers, nurses and other members of the support team.

Aim: This section aimed to understand the way care for families had changed over the years, and especially to review the changing involvement of families.

Method: In-depth interviews with two key informants. Informal discussions were conducted in person in Australia and via private social media between Canada and Australia. The interview in Australia was recorded with permission and of 90-minute duration. The topic was understanding any change in focus of caring for babies, children and their families over a 30-year period. A short discussion was conducted with a nurse with over 12 years' experience as a senior nurse caring for these children who has a special interest in neonatal surgical bowel conditions. The intention was to understand from her perspective of caring for these children over time, whether the approach and involvement of parents had changed in that time.

Recruitment and participants: The key informants were a nurse unit manager (NUM) who had over thirty years of experience in charge of a major paediatric hospital ward, and a paediatric ward nurse who had over twelve years' experience caring for these children.

Analysis: These data were analysed inductively, generating thematic chapters, and these themes were then cross-referenced to other components across different domains.

5.11 The domain of school

Aim: This component aimed to understand the process and supports available to a child when they have ongoing faecal incontinence following definitive surgery for Hirschsprung's disease and imperforate anus.

Method: Informal interviews with key informants from education; policy review; discussion with state department officers.

Participants: Eight department of education websites were analysed to assess the types of medical conditions recognised and care plans implemented.

Two assistant principals were informally interviewed to gain perspective of the process when a child is enrolled in the school who has ongoing faecal incontinence.

Telephone discussions were held with three Learning and Wellbeing Officers within the Department of Education NSW to understand the process when a child enrols in a school who has faecal incontinence due to being born with Hirschsprung's disease and imperforate anus.

Analysis: The policy documents were categorised according to a pre-developed coding tree and were analysed using descriptive statistics. The interviews were analysed inductively and applied across different domains.

5.12 Parent support groups

Aim: To assess the role and efficacy of support groups in paediatric medical conditions

Method: Online anonymous surveys. There were ten items addressing organisational features of the support group (governing bodies, years of service, nature and structure of the support group. Paid staff and volunteers; five items related to funding and conditions covered by the organisation, twenty-three items related to services and method of delivery to families and clinicians.

Recruitment: Questionnaires using Survey Monkey were sent to two hundred support groups to determine their structure and effectiveness. Of those, 172 were rejected as their focus was mainly adult based. There were. twenty-eight respondents whose support organisations were specific to children, from these, nine were designed to support families of children born with bowel conditions, of those, only six were specific to anorectal anomalies such as Hirschsprung's disease or imperforate anus.

Participants: twenty-eight members of parent support groups – located in Australia and internationally

Analysis: Simple descriptive statistics, reinforced by work with parents through the Bowel Group for Kids group.

5.13 Paediatric surgical units

Aims: The aim of this component was to determine if hospitals have a system in place to disseminate information about support groups to families when their child is diagnosed with HD or ARM

Methods: Survey with many open-ended questions, exploring existing hospital-based mechanisms for disseminating information about support services to parents via their website or a display of pamphlets publicly available.

Participants: Staff in paediatric hospitals (n=28) were contacted directly

Analysis - Simple descriptive statistics.

This was supplemented by data on 430 children born with an anorectal malformation has been collected from the medical records at Sydney Children's Hospital Randwick and Westmead campuses, John Hunter Hospital in Newcastle and the Canberra Hospital from 1999 to 2015. This has provided a rich data set of demographics, survival, severity of the conditions, associated anomalies and the need for ongoing care.

Statistics on the incidence of Hirschsprung's disease and its various associated conditions at New South Wales and Canberra children's hospitals were collected from 1960 onwards. These were reviewed for the years 1960 to 2014. It should be noted that these figures do not include children who presented to non-specialist hospitals, and who were then NOT referred to specialist units at Sydney Children's Hospital, Randwick, the Royal Alexandria Hospital for Children Camperdown, Children's Hospital Westmead, Newcastle or Canberra. Information is likely to be incomplete before 1975, as small numbers from Newcastle and

Canberra will be missed, and not all Sydney records from that time were retrieved. The annual incidence of Hirschsprung's disease and Hirschsprung's disease with Down syndrome was calculated, and the percentage of Hirschsprung's disease with Down syndrome was then also calculated.

5.14 Ethics approval

Ethics Approval HREC reference: LNR.13.SCHN.435 was granted to conduct a long term follow up of patients with Hirschsprung's disease and anorectal malformations. Governance Site Authorisation was granted for the Children's Hospital at Westmead; SSA reference: LNRSSA/14/SCHN/241 and for Sydney Children's Hospital Randwick; SSA reference: LNRSSA/14/SCHN/242. Ethics approval was granted from Australian National University High Risk HREC committee. Ethics approval was granted whilst I was collecting data during my early years at the Australian National University. No data relating to this ethics clearance was collected after 2015.

A secondary data set was gathered through open access conference materials gathered from public conferences and workshops. Information from the workshops was shared with attendees. Permission was granted to use their information in newsletters and online as stories, all names were changed, and any identifying material from the online stories was removed to protect the children in the future from anyone accessing their information through online searches. Even though permission was granted to use their details in those forums, due to the sensitivity of the information I have anonymised the names, ages, and any identifying material to ensure no possible trace back to the affected children.

At the start of public conferences, all delegates signed off the right to testimonies being used in a public fora, allowing their material to be freely used, whilst protecting the identity of the children. Public dissemination and qualitative community research using open access material, via conferences was utilised, with clear sign off by all delegates. The Bowel Group for kids Facebook site (https://www.facebook.com/groups/BGKAustralia) and the Bowel Group for Kids (https://www.bgk.org.au/) website can be accessed, and the online community visited.

It is integral to this doctoral research that attention be placed on data collection, noting the sensitive nature of the conditions of participants. Most of the participants were parents of children born with a congenital bowel disorder. The children were too young to be interviewed themselves, so permission was granted from parents on the condition that all possible means of tracing back to the child were deidentified or removed. This was accomplished by changing names, ages or position in the family to remove any connection to the child, or their geographical area. Protecting children's identities became a major focus of the support group I co-founded. The widespread household use of the internet was not apparent until the late 1990s and most people did not have easy access or affordability until the early 2000s. This brought about change in the way the support group ran as what started as a member based, hard copy newsletter where members happily shared their child's story and pictures, needed to change to protect identities. The risk of a teenager searching the world wide web for a friend's name was now a reality and could be shared online. A detailed search had the potential to locate old newsletters or articles that contained sensitive information and pictures about their early years being born with a congenital

bowel disorder, their surgeries or difficulties experienced with incontinence. Whilst parents in general happily discuss their child's bowel habits and toilet training escapades, no teenager wants that information found on the internet linked back to their name. Digitisation had an impact on what and how much data could be collected, I was unable to use any data that would be identifiable, this made tracing back to participants difficult, but did not interfere with the overall thesis.

5.15 Limitations

The limitations of this research are typical of research in this area and relate to the low numbers of Hirschsprung's disease and imperforate anus which are rare conditions and the small number of parent support groups globally that have a specific focus on supporting these families. It was imperative that the identities of participants in the research were protected, any form of information that could be traced back to the children or their families was changed, names, ages, position in the family, and if appropriate, the sex of the child so there was nothing that could expose these families or the child. The sensitive and private information had to be protected to ensure their information could not be traced. When requesting permission to publish stories in the Bowel Group for Kids newsletter, I advised parents not to use their child's name, age or any identifying information such as the school they attended due to the risk of exposure in later life through online searches and social media. The names of the families throughout this research were changed to avoid any possible link to the child in later life.

The richness of this study was guided by my own experience over the past thirty years giving me a first-hand understanding of the difficulties we had experienced as a family, which in turn meant I have gained the trust of the parents I had met and interviewed over the twenty-nine years of running a parent support group, and through my professional role as a paediatric theatre nurse caring for these children.

5.16 Conclusion

This chapter has outlined the methodological framework of this thesis, using institutional framing and stigma to explore how children with Hirschsprung's disease and imperforate anus can be socially isolated and under supported. This theoretical approach highlights how socially challenging conditions which are engaged with disordered faecal excretion are, and the impacts that they may have on social identity.

This theoretical framework is conceptualised into six domains in which institutional framing can occur, all of which rotate around the child. These domains are the paediatric hospital (organisational institutional framing), surgery (biomedical framing); parents and carers (the family); nurses and allied health teams (biomedicine and social support); schools (behavioural psychology and educational institutional framing); and peer support groups (institutional framing of social equity and democratisation of knowledge).

A complex set of methods have been used to explore all these domains, comprising interviews, surveys, case series analyses of children with Hirschsprung's, and my own work as an observant participant over 29 years of work with the peer support group Bowel Group for Kids.

The importance of this research is to understand the complex nature of faecal incontinence, not just data on how many children are diagnosed each year, what surgery is performed, the survival and long term serious medical conditions, but the lived experience. Whilst the advance in medical imaging means a clearer, accurate diagnosis, a prenatal diagnosis still eludes us to this day. Improvements in surgical techniques has shortened hospitalisations, improved the physical appearance for children who no longer bear the huge scars of open abdominal surgery and postoperative care and understanding of bowel function has improved over time. The goals of diagnosis and early surgical intervention are survival and faecal continence. Today it is a rare occurrence that a child may die as a consequence of being born with a congenital bowel disorder (Rosen and Holder, 2022; Short et al., 2022),

One aspect that remains problematic and a constant source of difficulties for children and their parents, is when there is not a favourable outcome from the surgery. Any form of faecal soiling or incontinence is often seen as a failure of the surgery leading to further consults with the treating surgical team often to no or little avail. If there is no mechanical problem to fix, then further surgery is unlikely to improve the situation. Unfortunately, despite the progressive steps in imaging, surgical technique and postoperative care, there is no way of knowing the outcome until the child reaches a developmental stage when it is expected the child will gain faecal continence. Some children may show early signs of difficulties passing stool and various interventions can be utilised to alleviate this such as rectal dilatations, stool softeners bowel washouts and enemas; continence, however, is difficult to predict.

At the birth of any new baby, parents are understandably shocked when given a diagnosis their baby who appears perfectly healthy, has been diagnosed with a congenital bowel disorder. The number of clinicians involved, the assessments, imaging and consultations are overwhelming at a time when the parents were anticipating taking their baby home. Even as an experienced theatre nurse, I found myself exhausted at the level of activity when it became clear Adam had an acute bowel obstruction, blood tests, further consults, intravenous fluids as breast feeding had to cease and milk expressed as nothing could be taken orally that would exacerbate the bowel distension. I found this incredibly difficult to manage and distressing to see my baby go through so much. It is this insider knowledge and unique experience as a nurse, educator, mother and support group co-founder that strengthens this research to a level of understanding not experienced by clinicians alone.

This research fills the gap left between the shock of the medical diagnosis which has to take priority when the condition is first recognised, and when the baby is discharged home into the care of parents who are often exhausted and traumatised. The importance of this research is to ensure parent stories are told and new parents are given the opportunity to talk to another parent who has gone through the same situation as no one can possibly understand unless they have lived through it. Typically, the baby is diagnosed shortly after birth when it becomes apparent they are not passing the first tarry stool known as meconium with 24 - 48 hours. To the extended family and friends, the baby has some obscure problem they have never heard of, that is being dealt with before they come home and even if they visit, there is nothing to indicate the seriousness or long term outcomes of the problem. Whilst social workers and counsellors a.re

an important adjunct to support parents navigate the system, it is understanding from other parents who have walked this path before, that new parents find most supportive.

CHAPTER 6: "SURGERY DONE – PROBLEM SOLVED!" PARENT'S ACCOUNTS OF THEIR LIVES WITH A CHILD WITH A CONGENITAL BOWEL DISORDER

This chapter addresses the parental perspectives of having a baby with Hirschsprung's disease or an anorectal malformation. In this chapter, I uncover what parents were told about their child's condition at the stage of diagnosis, in particular their understanding of the outcomes of surgical intervention. The material in this chapter is shaped from discussions with parents seeking peer-to-peer support in person, over the phone, and in hospitals. Material has also been collected from discussions at conference and workshop sessions held for parents. Some material has also been gathered from posts on social media by parents and family of children with Hirschsprung's disease and anorectal malformations. Ethics and privacy have been respected throughout the process.

These[EG2] data sources were gained from discussions over the past thirty years with families of children born with anorectal malformations in a multitude of settings and situations. Whilst working as a registered nurse in the operating theatres at the Children's Hospital, with the surgeon's permissions, I would visit families on the ward before or after their child underwent surgery, to offer comfort and an opportunity for them to discuss their experiences raising their child born with an anorectal malformation. As a mother myself of one of these children, I was able to show I understood from a personal perspective, yet being in my role as a theatre nurse[EG3], I was able to assure them, that they were in the very best of hands. I would visit the family after surgery to offer comfort and support if it was so desired.

This[EG4] was also an opportunity to provide information to the families on the value of Allied Health Teams consisting of the stomal therapy and continence nurses, where a Gastroenterologist may be of assistance and if the parents, siblings or the affected child needed support, I would explain the role of the hospital Social Worker and a Psychologist to help them understand these health professionals are very experienced and an excellent source of information and professional support if need be. Often parents expressed a fear they would be seen as not coping if they reached out for help from allied health professionals, I was able to dispel those fears and offer contact information for the future.

As[EG5] a mother in hospital caring for my own son, I occasionally met other parents whose child also had an anorectal malformation so we were able to comfort each other. As anorectal malformations are rare diseases, there can be a sense of loneliness when your child is in hospital as there is unlikely to be another child in at the same time.

Other[EG6] data sources were gathered through the Neonatal Clinic sessions where I was invited to attend and speak with families.

Each of these data sources formed a basis to the conceptual frames depicted at point 5.6 of this thesis.

The chapter begins with narratives from adults with Hirschsprung's disease of their diagnosis and treatment in the mid-twentieth century, when it was common for diagnosis to be delayed. This is important because surgical outcomes are better if the procedures are conducted when the baby is young. Today there are fewer cases of late diagnosis of both conditions (Amata et al., 2018; Stensrud et

al., 2012). I then present people's narratives of managing faecal incontinence, and their sense that they were under or mis-informed by overly optimist surgeons that anatomical correct equates with complete cure. Finally, I discuss the roles and place of dedicated peer-to-peer support groups for parents of children with Hirschsprung's disease or anorectal malformation.

6.1 Who are the parents?

Table 6.1 summarises details about the informants in this chapter. Accounts by sixteen parents, fifteen of which were women, are incorporated. The accounts refer to twenty affected individuals, of whom five had anorectal malformations, and fifteen had Hirschsprung's disease. Four of the children with Hirschsprung's disease also had Down Syndrome. The corpus of narratives presented here enables an in-depth view of the lived experience of people with Hirschsprung's disease or anorectal malformation and how it has impacted upon their parents. The importance of their stories cannot be ignored, every life matters and by telling their stories, we begin to understand the complexity of problems encountered when a child is incontinent of faeces following surgical correction for a congenital bowel disorder, it is through their stories we see why it is important to understand their situation and the impact it has on their lives, as it is only then we can enact the changes needed to support these children through to adulthood (Monarth, 2014).

Table 6.1: Details of people with HSCR or ARM and parents. (All identifying information has been completely eradicated to protect the children who are the focus of this research).

Child	Age	Gender	Condition[1]	Parent	Communication mode with informant
Leila	3	F	ARM/cloaca	Jenny	Interview
Leah	10	F	ARM	Mary	Telephone
Lillian		F	HSCR	N/A	Social media post
Sandra		F	ARM	N/A	Interview
Chloe	8	F	HSCR	Fiona	Interview
Lucia	6	F	HSCR/DS	Nicole	Telephone contact
Saxon	4	M	HSCR/DS/CHD	Amber	Interview
Rachel	4	F	HSCR/DS	Cathy	Telephone contact
Damien	9	M	HSCR	David	Interview
Sarah	7	F	ARM	Avril	Conference workshop
Samantha	5/12	F	HSCR	Chris	Interview
Liam	7	M	HSCR	Olivia	Interview
		M	HSCR	Rose	Telephone contact
James	3	M	HSCR	Vera	Social media post
Adam	30	M	HSCR/DS	Self	Personal recall
Isabelle	8	F	HSCR	Daphne	Telephone contact
Eliza	7	F	HSCR	Veronica	Interview
Jacob	6	M	ARM	Sandra	Interview
Bethany	5	F	HSCR	Mary	Conference workshop
Mary	35	F	HSCR	N/A	Telephone interview
June	23	M	HSCR	Michael	Email response

[1]DS, Down Syndrome; CHD, Chronic Heart Disease, HSCR Hirschsprung's, ARM Anorectal malformation.

6.2 Missing the diagnosis

Lillian was born in 1936. Her childhood was marked by severe constipation, severe enough to warrant serious surgical intervention.

> [M]y mum gave me an enema each day for the first 5 years of my life. I went to a Sydney hospital and had a bilateral lumbar sympathectomy as described in (Rankin and Learmouth 1939) but results were disappointing. No investigations were conducted. (Lillian, personal communication, 10 August 2011).

Her Hirschsprung's disease was diagnosed at the age of 29 years, after she had married, had four children, all the time suffering severe constipation "sometimes up to three months at a time". Further family research has revealed nine of her family members over four generations were born with Hirschsprung's disease, three of whom died shortly after birth in the 1930s and 1940s.

Sandra, who was born in 1965, was also undiagnosed at birth.

> I was my parent's first child and was sent home without anyone noticing I had an imperforate anus. I suffered terrible nappy rash, still the fact I did not have an anus was missed. My mother noticed I had faeces leaking from my vagina. She took me straight back to the doctor who confirmed I had no back passage, and we were sent straight to the children's hospital. My bowels exit high into my vagina and no attempt was made to correct this. (Sandra, personal communication, 12 May 2015).

Because Sandra had an exit point for her colon into her vagina, the connection, a fistula was enlarged, rather than a stoma being created, as would have occurred if she were male.

> Mum had to insert dilators into the bowel through the vagina at every nappy change. …I suffered from severe constipation at times and one doctor was

> going to charge my poor mother with neglect. My mother was amazing, she just managed it on her own… Finally finding the support group has not only helped me, but it also helped my dear mother who was finally able to stop feeling guilty that it was something she did.

Surprisingly even today, there are reports of babies being sent home without a diagnosis. This is plausible if the mother is part of the early discharge program which offers discharge within 24 hours if the mother and child appear well. Hirschsprung's disease is considered if the baby has not passed meconium in the first 24-48 hours or fails to feed as expected or vomits bile, so it could be missed if the baby and mother were discharged early. For an anorectal malformation, there are checks in place where the midwife follows a checklist which includes parting the baby's buttocks and checking that there is an anus. Despite this, up to one in five neonates with anorectal malformations have a delayed diagnosis after 48 hours of life (Turowski et al., 2010).

> When my baby was born the nurses ticked the checklist that she had an anus, then the paediatrician checked it, problem was she did not have one. It was not until I noticed something at 4 months of age that we got some action. After the operation I was told it was a low anorectal malformation so toileting should be ok other than she may take longer to toilet than other kids and may have constipation. That was ten years ago, and boy was it an understatement.

(Mary, mother of Leah aged 10, personal communication, March 11, 2016).

Parents were often told they were overreacting:

> Shortly after the delivery of our daughter, my husband noticed her anus was really tiny and seemed close to her vagina, we were told all babies are different. We asked if this had any connection to her additional digits on both hands and no thumbs, this too was dismissed. We had a geneticist

> appointment to discuss her hand abnormalities called polydactyly. The doctor organised an X-ray of her spine, soon after we got home, they called us back as they discovered she had a collection of symptoms known as VATER. We later saw a surgeon who took one look at where her anus should be and said it was not an actual anus it was a fistula and she needed surgery straight away. How could so many doctors and nurses see her and not notice this.
>
> (Jenny, mother of Leila aged 3, personal communication, 21 March, 2016).

The literature suggests Jenny is not alone as a parent in feeling ignored by doctors and nurses. (Maxwell and Barr, 2003) studied how parents were not included in communications relating to their child, as though their concerns were not important. Listening to parents provides an opportunity for clinicians to learn from their experiences, gain an insight into what parents find helpful when their baby is undergoing major surgery or is in the neonatal intensive care unit (Hinton et al., 2018).

6.3 Trust in the surgical cure

While everyone will have their own comfort level regarding how much they want or need to understand, the results show there is no consistency in how these conditions are being presented to families. The congenital abnormality is seen as surgically correctable, or in the words of a European colleague, 'surgery done - problem solved' (Schipper, 2008). Historically the emphasis has been on correcting the defect and saving the baby's life. For many children, today we know this is often just the beginning of learning to live with the long-term effects of Hirschsprung's disease or anorectal malformation. But those babies have grown up and are now seeking a better quality of life (Hartman E. et al., 2015; Hartman et al., 2006; IeiriI et al., 2010).

From our own experience with our son Adam born with Hirschsprung's disease and Down syndrome, our hopes that once the stoma was closed and the bowel re-joined, then life would return to normal: we now know this was unachievable. The representation of an anatomical cure being the same as a functional cure was presented to many families and remains so today.

Moore, (2016) regarded Hirschsprung's disease as surgically correctable with most patients achieving socially acceptable anorectal function. The functional outcome depended on several factors, length of bowel affected, surgery, complications and the presence of, family support, and associated anomalies of the patient.

> We were told that our daughter no longer had Hirschsprung's disease after the pull through at 4 months! My poor gorgeous girl nearly eight now and struggles daily.
>
> (Daphne mother, of Isabelle, personal communication, 10 August 2011).
>
> We were also told after the surgery she no longer had Hirschsprung's disease and was "fixed" and would just be the type of child that may feel the cold so take an extra jumper. We were warned she might take a little longer to be continent, but everything was now "fixed". Result - permanent ileostomy, ileovesicostomy and an ileal conduit Chait· No control of bowel or bladder.
>
> (Veronica, mother of Eliza aged 7, personal communication, 24 March 2016).

The inconsistency of information presented to parents of babies born with a congenital bowel disorder creates confusion, anger, guilt that they must be doing something wrong, and invites criticism and aversion from society. Parents, mainly mothers as primary carers, and their affected children often bear the brunt of

cruel comments and taunts when the children fail to reach societal expectations of faecal continence. Children suffer taunts at school, parents are often made to feel they are either not coping or not trying hard enough, which often results in withdrawal from society at large for the entire family.

6.4 Differentiating normal childhood issues from those related to the condition

Even in children without any significant issues at birth, parents can struggle to decide if the child needs to go to the doctors or hospital. There are a plethora of websites and social media sites parents can access to try and determine what constitutes a problem, but when the child was born with Hirschsprung's disease or an anorectal malformation, how do you decide if it relates to the underlying condition or just normal childhood illness? (Fields, n.d.)

The year was 1990, following the final pull through surgery at 14 months of age. The surgeon inspected Adam's wound and abdomen, asked a few questions about his feeding and bowel function and said everything looked good and to come back if there is any problem. No instruction was given on what to look for, so we were always second guessing, was this a problem or normal childhood development, was it related to Down syndrome?

Vera relayed the same concerns for her son James aged 3 years:

> We were told he needed surgery and that would fix the problem. It was never explained to us that there may be some long-term continence issues. I would have preferred to be prepared as I felt I was in the dark, you do not know if it is related or something normal like teething, reaction to foods etc. being told the problem was fixed made me reluctant to ask if it could

be related to Hirschsprung's disease. (Vera, mother of James, personal communication, 3 November 2017).

6.5 Consequences of trust in the surgical cure

Almost all parents recalled faecal incontinence being radically understated as a potential ongoing problem. This is an example of the patient's perspective being under-recognised

> We were told that surgery would "fix our daughters problem". It was only when she developed complications, they explained that there could be long term complications.

(Avril mother of Sarah aged 7, personal communication, 3 November 2017).

Olivia, mother of Liam now aged seven, demanded to know the worst possible outcome

> I insisted on knowing the worst-case scenario, I cope better with that level of information, but it is not for everyone. Our surgeon was pretty realistic in her expectations. (Olivia, mother of Liam, personal communication, 3 November 2017).

Many parents reported that they had consented to keep having their child operated on in the hopes of achieving continence for their child and did not trust their own instincts. When considering the ethics of each decision, a doctor should respect parents' opinions, even if they are sometimes unorthodox, unless they entail harm to the child (McDougall, R., Delany, Clare, and Gillam, 2016).

A procedure that surgeons view as corrective can co-exist with ongoing faecal incontinence. This struck many parents as a failure of surgical imagination, or

worse, an abrogation of recognition of the child's experience. Some parents noted that surgeons may focus on the anatomy, rather than the actual experience.

> The doctors said I would not have any problems now because they had cured my Hirschsprung's. As a young adult I did my own research and finally feel like I understand a bit more now about the issues that come with it.
>
> (Mary, age 35, personal communication, 1 February 2018).

Some families were assured the shorter the segment of bowel affected equated to a good outcome; this was not always the case.

> Our son's surgery went really well, he only had short segment Hirschsprung's disease, so they were confident that he would be fine. Well, he has had enterocolitis twice, Botox, had further biopsies to try and work out why and ended up with a colostomy. The whole time his surgeons were stumped as to why he kept having issues.
>
> (David, father of Damien, personal communication, 9 April 2014).

A parent who described herself as someone who actively wanted to be told potential negative outcomes (*I work on preparing for the worst but hope for the best*) was presented with what in retrospective was an overly rosy presentation of outcomes.

> We were told the surgery would fix him; it would just take him longer to potty train because of nerves to the brain to get the message across that he needs to empty. We were not told anything about possible complications like incontinence.
>
> (Sandra, parent of Jacob aged 6, personal communication, 23 January 2014).

In the following account, a parent describes the value of a surgeon giving a direct and unadorned projection of the hope for the future. The parent was told that there was significant variation in outcomes.

> [S]he would consider it a great success if the child was able to go to school and remain clean all day without accidents. The hope was that we would achieve success, but we would monitor for signs of incomplete emptying, constipation, smearing, incontinence etc. This straightforward approach helped us a great deal as we felt we had a realistic approach and were well prepared on what to look out for right from the start

(Mary, parent of Bethany aged five, born with Hirschsprung's disease, personal communication, 24 April 2014).

This kind of surgery is challenging, and the course is unclear. Parents ought to be given realistic outcomes, without being overly negative. Parents should be honestly informed that likely outcomes that include the facts that children may have residual chronic constipation or faecal incontinence due to disruption of the normal neuronal pathways or manipulation of the anorectum during surgery. (Bragagnini Rodríguez et al., 2017; Loening-baucke, 2002).

6.6 Need for support from other parents.

Many parents find relief in talking to other parents who are or have experienced similar situations as the ones they find themselves in. The importance of hearing from others who have walked the same path before, builds trust and validity and an understanding of what they are experiencing (Haven, 2007).

Cathy relayed isolating moments in the NICU with baby Rachel who was born with Hirschsprung's disease and Down syndrome, these worrying times were

relieved by telephone calls with a member of the support group. Fiona's daughter was eight years of age before Fiona, overwhelmed and isolated, was told about a support group, and her perspective of her own competence and capability changed when talking to others who understood. (Cathy, personal communication, 20 August 2015).

When Nicole's daughter Lucia was 6 years of age, Nicole found herself feeling at the end of her tether with the constant management of incontinence. She asked the surgeon if there was precedent for returning to the child having a stoma. His reaction was one of horror that anyone could suggest this. Meeting other parents helped her reconcile her decision as reasonable. (Nicole, personal communication, 16 December 2015).

In a similar situation, Amber considered going back to a stoma for her son Saxon aged four who was born with Hirschsprung's disease and Down syndrome. In this case, while the surgeon was supportive, her family and friends were in disbelief. (Amber, personal communication, 12 November 2015).

Some parents find their experience of parenting a child born with Hirschsprung's disease or an anorectal malformation is so different from their friends, that they just stop talking about the condition to them, but of course this can isolate them from normal parenting conversations. The usual conversations for parents from birth to school age revolve around stages of development, childhood illnesses, preschool or school selection, their child's social development, where they buy clothing, to name a few.

For parents of children born with Hirschsprung's disease or anorectal malformation the kinds of conversations range from toileting issues, foods that constipate or cause a loose stool, multiple doctors' appointments, tests, medications and soiling. Worries about acceptance into their local pre-school or school, and once there, managing their disorder discreetly to avoid teasing or bullying. The risk of soiling once they reach an age where toilet training is a societal expectation, the endless washing of bed linen and clothing, the list goes on. All these conversations are foreign to anyone who has no understanding of what parents are going through when the child has ongoing faecal incontinence.

6.7 The hidden disorder

A concern raised by many parents is not being taken seriously. They say they just knew something was not right, but were often dismissed by midwives, doctors or baby health clinic nurses as the perceived problem was '*nothing to worry about*.' The conditions of Hirschsprung's disease or anorectal malformation are rarely known to the parents of a newly diagnosed baby. The medical terminology is no different to other professions, they all have their own 'language' so to speak. What is missing though, is the terms are not explained to parents who are left with a complex list jumbled up in their minds, not only of the condition but the terms and acronyms used to deliver the plan of care for their newborn. Whilst there is an urgency about 'getting on with the task at hand,' it is important parents are not left to turn to searching online to decipher this new language as that can be fraught with danger as not all websites are dependable. Parents report feeling overwhelmed by the conversations, firstly the shock of being told their newborn who appears perfectly normal and healthy, has a condition they have never heard of.

Some comments I hear often are:

> …Hirsch what? What does that mean,
>
> …I had no idea, did they catch a disease, and if so where from?
>
> …What do you mean they don't have an anus? What does VATER or VACTERL stand for, when will we know if my baby has any of these?
>
> …Nurses and doctors tried to explain it to me, but I was at a loss to understand anything except that my tiny baby had had an operation and now had a colostomy. Again, what did that mean?

Most mothers feel immediate guilt that they have done something wrong during the pregnancy. Mothers will report they strictly adhered to the pregnancy dietary recommendations which is increasingly complex in the twenty-first century. There is a need for clinicians to be aware of the emotional rollercoaster parents often feel after giving birth and then being told there is something wrong with their baby, all rational thought is lost. (Landsman, 1998; Skotko and Bedia, 2005; Wigander, Öjmyr-Joelsson, Frenckner, Wester, and Nisell, 2018)

Going home from hospital can be a daunting task after the relative safety of being surrounded by teams of health professionals who appear completely at ease and competent in their management of tiny babies with stoma's, tubes, wounds and managing procedures. Some terms parents find particularly difficult especially after the shock of an unexpected diagnosis. Stomas, for example, are often seen as things that old people who have bowel cancer have. The terminology for different segments of the bowel can be confusing. Parents often worry that they will hurt their baby through washouts, enemas and anal dilatations. These intimate practices require new quasi-nursing skills, outside the expectations of

normal parenting of a baby. The dilators look like instruments of torture and parents report feeling very uncomfortable, even deviant, and certainly not something they ought to be doing to their baby.

6.8 Siblings

Sandra recalls the positive and negative experiences that have resulted from her younger brother being born with Hirschsprung's disease and how the experience has changed her life entirely. She recalls many times doing her homework at her brother's bedside in hospital and having a set of clothes ready on her desk at home just in case the hospital called in the middle of the night.

> Friends never really understood, it was a lonely time and I feel I grew up before my time but that made me very independent. I think a lot of people who haven't had a sick child, forget life goes on for others at home – and that is hard. Having no one to talk to about it, but this experience – long and painful as it was, made me architect my career today. (Sandra, personal communication 16 February 2017).

Jessica recalls Adam who was born with Hirschsprung's disease and Down syndrome being terrified and how we had to pretend we were not going to hospital, if he knew he would cry and say 'no tube, no needles. We didn't really go on family holidays; it was too hard, and a lot of time was spent in doctors' surgeries or at the hospital.' Jessica and Adam are just two years apart so are very close and she probably recalls the most because our other two boys were at school. Fortunately, none of our children have much recollection of the terrible procedures that were necessary to perform at home and they adore their brother. The concern now is more for the future, (Mary, personal communication, 12 July 2018).

A particularly challenging time for parents is when their affected adult child is pregnant. This can bring back haunting memories and they worry that their grandchild may have the same condition.

> i worried the entire pregnancy wondering if my son's baby would also be born with Hirschsprung's disease. This time brought back all the memories of what living with a child with Hirschsprung's disease was like. I tried not to worry the new parents but made my own enquiries so that should there be any signs of this disease I would be able to support my son and his partner. I arrived at the hospital just after the birth and found that I was the grandmother of a perfectly healthy child. It is a great feeling and I just love every minute of being a grandmother

(Rose, mother of son with Hirschsprung's disease, personal communication, 4 September 2018).

I recall being present during the pregnancy scans of our grandchildren; our daughter and her husband were keen to see the baby in general and the sex, I couldn't help but focus on the abdomen to see if it seemed large. I know this would not have shown any signs of Hirschsprung's disease or anorectal malformation, but still, I just had to check, fortunately they are all very healthy.

6.9 Transition to adult care

Transition to adult care services is a difficult time for families who often describe feeling like the hospital had become an extension of their family, a safe haven when their child needed admission. They were known to the staff which made explaining the issues easier. The adult services are far from ideal for children who have grown up in paediatric care where clinicians consult as a team including the parents. Parents find themselves in a completely different situation moving over

to adult care. There needs to be much more attention to this important area. The transition process is often not considered until the child is a teenager, but this process needs to start long before then (Lemacks et al., 2013). This is reiterated by a set of key principles designed to guide clinicians in how to best prepare and execute the transition process for long-term paediatric patients and their families (Brodie, 2014)

6.10 Conclusion

This review of parent experiences has demonstrated the need for support groups to be an integral part of patient care, based on the parent's wishes. Many parents were dismayed to discover they could have had support from when the baby was first diagnosed. Parents should be provided with basic support group information such as what the group offers and how to contact them. Not all parents have the same need, but if given information by way of a pamphlet, they can contact the support group when and if they choose, at their own pace. Many support organisations will come and visit the parents in hospital if they so wish, while others may prefer to just communicate through email or talk on the telephone. This chapter has also addressed the quondam failure of surgeons to recognise the social and parental impact of continued faecal incompetence after corrective surgery. The fact that anatomical correction can co-exist with serious ongoing functional impairment is often glossed over by surgeons. Many parents assert that they had been give insufficient advice about the true likely outcome of surgery for their child. There is a clear need for better communication and therapeutic alliances between clinicians and parents to clarify the real impact of long-term faecal incontinence, and whether surgery can prevent this.

CHAPTER 7: EDUCATIONAL EXPERIENCES OF HIRSCHSPRUNG'S AND ARM STUDENTS

This chapter addresses the legal and social issues around school readiness, social norms and expectations, and the experiences of students with Hirschsprung's disease or anorectal malformation and their parents and teachers. This chapter begins with an overview of the provision of support for children with disabilities in Australia. I then review Hirschsprung's disease and anorectal malformation as an invisible and stigmatising disability. Using data gathered from focus groups and workshop sessions at annual conferences for the Bowel Group for Kids and interviews with teachers, I demonstrate that in schools, planning and responding to faecal incontinence is complicated by pre-conceptions about continence. Conflating these two types of incontinence results in further stigmatising of children with Hirschsprung's disease or anorectal malformation and delegitimating parental concerns about a condition that is often reviewed as being surgically "cured.". It is assumed that all children of school age will be continent of faeces, the odd accident around urinary leakage is an accepted norm for children of four or five, full of excitement and some anxiety at the start of the school year, but ongoing faecal incontinence beyond the social norm is a completely different concept and totally intolerable.

The[EG7] data used in this chapter was gathered from publicly available open sources. I looked at freely available public information then gathered up unobtrusively already existing data sets already available around Australia. In this thesis, I interpreted the data collected from parents and educational institutions to form the basis for this chapter.

7.1 The rocky road of transition to the ultimate goal - mainstream school.

When Adam was born in 1989, the focus of education for children with Down syndrome was very much towards integration into mainstream schools, so that was what we aimed for. After all, our three daughters went to our local mainstream school before him; the teachers, most of the families and students all knew Adam, and that he had Down syndrome and seemed so supportive and encouraging about his future. In the 1980s, children with Down syndrome were more likely to live with their families rather than sent off to institutions, hidden from society. How hard could it be? But something niggled at me, were people just being kind, sympathetic or was it bordering on patronising?

I was advised to start him in early intervention classes as soon as possible after birth, the emphasis was on 'the earlier the better.' I began searching for potential places, in 1989 of course there was no internet to search, you were really on your own trying to find services. I recall discussing with his paediatrician, why wasn't there a central agency that could provide a list of facilities these children may need with contact details? I quickly learned the only way to find out where suitable services existed, was word of mouth. I had called the local council, preschool, schools, early childhood nurses; all to no avail. I joined a group of mums for children with Down syndrome whom I found to be the best source of knowledge, some had children a bit older than Adam so had already sourced the best services. The only drawback was, Adam did not just have Down syndrome, he had spent the first six weeks of his life at Prince of Wales Children's Hospital at Randwick, (now known as Sydney Children's Hospital). A community nurse had called me one day in between my dashing between trying to be home for the

girls and driving down to see Adam at Randwick and trying to establish breastfeeding. She was quite firm in her tone that it was critical for Adam to access early intervention services such as speech and occupational therapies, delays could be detrimental to his development. Yet another blow to mother guilt.

I tried to explain, our goal at the moment was for him to recover from major surgery at 13 days of age and getting him home from hospital. When he eventually came home, a colleague who was a lovely community nurse came to do a home visit for Adam, I recall opening the front door just enough to warn her, as one of the girls, Adam and I had all come down with a gastroenteritis. The house was a disaster zone, one of the girls had emptied the lower shelves of the linen cupboard to play hide and seek. My friend was a welcome friendly face, but I was wracked with embarrassment at the state of the place. We had no family in Australia and of course I never asked friends for help, why would I? I could run a children's ward with children ranging from babies to teenagers with a raft of conditions with orderly precision, or an operating theatre with a calm ordered approach, surely, I could run one household with only four children. From outward appearances, we were like any other family with four young children, despite the rough start, the baby's operation was over, we were home from hospital, and I was a nurse so of course I had everything in hand.

Early intervention classes had just started up in Camden only 30 minutes from home, so it seemed perfect, sadly the reality was quite different. They did not have regular therapists and the program was yet to be properly developed, it was very much a trial, and I was haunted by the earlier call that delays in early intervention would delay his progress.

The group of mothers from the Down syndrome Association told me about a brilliant facility at Parramatta. I felt pressured to pursue this or he would never learn to walk or talk. Whilst this was a brilliant, established service with trained therapists, again the stumbling block for Adam was his disrupted start to his early life which all stalled his progress. The occupational therapist's comments left me feeling inadequate and a failure. Adam would often be asleep from the long drive from home, they insisted he had to experience tummy time, or he would never learn to crawl. This would have been fine except he hated going on his tummy as it pressed on where he had major surgery and squashed his stoma. I felt I was constantly being reminded of what children with Down syndrome needed, but this was always being challenged due to his hidden disability of having Hirschsprung's disease. The comments were becoming all too familiar… '*yes but he had the surgery for that so that's been fixed now*.' It was as though to outward appearances, he was like another baby with Down syndrome, but the reality was, he was not, and this was becoming more problematic as time went by. As he got a little older, the issue of our failure to establish tummy time loomed again, instead of crawling, Adam had discovered he could scoot around on his bottom at breakneck speed. Something the girls ended up in fits of laughter as they tried in vain to copy him. The occupational therapist insisted he had to learn to crawl, or he would never walk. I felt the burden of responsibility weighing heavily on my shoulders. Why was it so hard for people to understand his difficult start to life?

Hearing tests when Adam was born were not available until babies were around six months of age. The closest Australian Hearing Centre was at Fairfield, so off I went with all children in tow as it was school holidays. The news was

devastating, Adam was profoundly deaf and needed hearing aids to hear anything at all. Adam also wore glasses from about six weeks of age as he was short sighted and had a turned eye. So, hearing aids were just one more device to add to the list. Today hearing tests are conducted within the first few days of birth. We returned to the carpark to drive home only to discover our car, fully equipped with prams, baby car seats and Christmas presents had been stollen, the worst blow was my camera with early photographs of the family when Adam was born, gone forever, no doubt the film was discarded as unimportant to the thieves. We walked to the police station to report the stollen car (*no mobile phones back then*), the officers were most unsympathetic, and said, well what did you expect when you parked in the council carpark. I called Michael and somehow, we got home, all six of us in a sedan without a single child car seat, I cannot remember how we did it. We were six weeks without a car as the insurance company refused to pay out until enough time had gone by for it to be found, we also lost the no claim bonus and had to pay more for a similar vehicle. It felt as though everything was against us. Life went on, I kept travelling to Parramatta for Adam's therapies on a weekly basis, dropping the girls at school and preschool along the way, our youngest came with us.

I cannot recall how, or who the recommendation came from, but I was put in touch with the Royal Institute for the Deaf and Blind (RIDBC) at North Rocks. It had never occurred to me that he would be eligible for this service. If only we had known about it earlier, it would have saved so much angst. We attended an assessment with the most wonderful, supportive and encouraging staff, after which a comprehensive report was received and an offer of one of their therapist to come to our home on a regular basis. I could not believe our good fortune.

Adam was now 3 years of age, and we were introduced to Helen who was a retired nurse and now therapist. She was a godsend to our family, she was encouraging, supportive and never once made me feel inadequate. She was always thrilled at his progress rather than focussing on what he could not do.

Adam attended a 'pre-preschool' from the young age of two years and nine months. This was a Government run facility aimed at preparing children with intellectual disabilities for life in mainstream preschool and school. The facility was located in one of the poorest socioeconomic area of Campbelltown called Claymore. A relatively new suburb that quickly became home to disadvantaged families, often single parents where generational unemployment was the norm. Driving through the suburb was such a steep learning curve, kids on the streets looking unkempt, adults sitting around smoking or drinking alcohol. Driving into the school was like a different world, the buildings were quite new, the school had been opened in 1981 and surprisingly had more facilities than our local schools in the Camden area. I was escorted to the 'special school' building, a demountable on the perimeter of the grounds as far away from the main building as possible. This was a small, one room building, no bathroom facilities, not even running water. Police cars often frequented the school grounds. I was terrified at the thought of our precious baby attending here. I should not have worried, the staff were amazing, they were very welcoming, friendly, educated and accommodated Adam who was still shuffling along on his bottom to get around as he did not walk until he was three and a half. Adam attended there two days a week and due to his special needs, we managed to successfully gain funding through the Department of Education for a full bathroom with hot and cold water to be installed. His early years at Claymore went by smoothly, the staff adored

him, they were caring and taught him early learning skills. The facility felt somewhat protected and far removed from the mainstream school which was plagued by violence, poor attendance and a constant police presence. Fortunately, there was never any reason to step into the mainstream school, I recall only going inside once to meet with the Principle to discuss Adam's enrolment and felt sadness realising the fate the children in the area had ahead of them.

Our first confrontation came as a shock when we applied for him to attend the local Government run preschool our girls had attended. The committee was made up of parents that we knew well, many of whom were avid churchgoers, and considered themselves model citizens. In contrast, we were not regular churchgoers, we came from the Isle of Man which had a similar in demographic to Ireland which was steeped in great divisions between Catholic and Protestant faiths, neither of which appealed to us. Michael was brought up Catholic and I was raised Protestant, so our union was frowned upon, but we considered we lived a good Christian life by example. I was utterly devastated and shocked when the preschool advised me, they were unable to accept Adam as they did not have the facilities to cater for his 'needs.' This really was the first of what was to become a recurring theme, the unspoken words, the inability to elaborate on what exactly those 'needs' were. I was so confused, what 'needs,' were not considered suitable for integration, the main buzz word around inclusiveness? I felt like everyone knew the truth behind the decision, but no one was willing to voice it.

My imagination was in overdrive. Were people tiptoeing around me? I felt excluded, shunned as though everyone was whispering about the situation. I no longer felt a part of the very community we had embraced. The stark reality of life with a child who has faecal incontinence, beyond what is considered the norm of three or so years began to weigh heavily on my shoulders. Goldberg et al., (2020) explored the lives of families who did not meet the expectations of societal norms such as parents of colour, those with lesser means, or whose child had a disability or were from same sex parents, and how they often felt marginalised and were made to feel they were not welcomed in the school community or by other parents, this had negative flow-on effects as parents felt ostracised by their peers and became less engaged in school activities. The only other option for us was a long day care centre as they took younger children that were yet to be toilet trained and were willing to accommodate him, it certainly was not my first choice as it was more child minding than educationally based preparing children for school.

The progression to mainstream school where our daughters attended was short and disastrous. Adam was the first child with special needs accepted into the local school, the area was predominantly populated with Caucasian, middle-class families. Despite all the tremendous work that had been done in early intervention programs to prepare for this moment, the school chose to go it alone and refused any advice from experienced teachers and clinicians who had been a major part of Adam's life. Where Adam had thrived on structure in the past, the school's intention was to allow him to explore what he wanted to do and let him decide, this quickly became problematic as he simply wanted to play outside and find his sisters. This was totally unfair to Adam and other children in

kindergarten who started to question why they couldn't go out and play like Adam. The school's solution to his 'bad behaviour' was to engage a behaviour support person who attempted to discipline Adam, of course he was quickly deemed non-compliant. I was informed they could only accommodate him two hours a day from 9 to 11am, so I had to take him and the girls to school, come home, go back again to pick him up, then back again to collect the girls at 3pm. Adam had been used to a full day at pre-preschool or day-care so he found the unstructured days difficult, and he was six years of age by then, I was back at work and so needed him to go to school. We realised this was never going to work in its current form and so withdrew him and enrolled in the only school accessible mid-year, which was a school for special purpose, the students had a higher level of need than he required, but he was welcomed and of course they found what they saw as his minor transgressions, easy to manage. The following year he was accepted into a support class in a mainstream school, again some distance from our home but there was no other option. He flourished there as they had a gated section for infants with play equipment which he loved, and the entire school were being taught sign language, so his hearing loss was not an issue. Apart from when he constantly threw his hearing aids away, which occasionally required the services of the fire department to come and rescue them from a drain where they could be heard whistling.

This went well until he had to move up to the primary stage of the school away from the safe haven of the gated infant's stage. He kept wanting to go back and play on the equipment. His time there lasted until he reached year 5, then we were politely told they could no longer cope with his needs, when I asked for them to elaborate, it all came down to the dreaded bowel issue. Unless I could

'do something' about it, they could no longer offer him a place. Integration clearly had its limitations which did not extend to continence issues.

Mainstream high school was never going to work as Adam would require a much greater level of independence than he could manage. So again, I felt defeated as we headed to another special school which was for children with intellectual disabilities. They had excellent teachers aids who were able to manage his stoma without issue. Whilst the classes were small compared to mainstream, the diversity of students was vast, Adam was the quiet one happy to sit in a corner playing alone, whilst teachers tried to manage other more vocal or aggressive children. This was far from the opportunity we hoped for, but the only option open to him.

Adam attained a modified Higher School Certificate at age 18 years, this was more tokenism really, but without continuing education to year 12, children with disabilities were unable to access any of the post school option programs in those days. These programs had two streams, transition to work or community access, these were a gradual transition away from the workshops of the past. If children with disabilities were unable to secure gainful employment, these programs were the only option for them when they left school. The National Disability Insurance Scheme (NDIS) was not introduced until many years later.

The sad reality was, we were caught in an era of trial and error, as though Adam was always at the forefront of what was to come, a guinea pig, the subject of scientific experimentation. Whilst we spent the first six years of his life in an out of hospital trying varying methods to establish normal bowel motility, medicines, bowel washouts, implanted devices, daily irrigations all to no avail, today most

children undergo one operation providing they are well at birth, to remove the affected segment and anastomose the bowel to establish continuity. If successful bowel function is not restored within a short time postoperatively, other options are implemented early or a resting stoma may be implemented to prevent the bowel overstretching. It is most unusual now to put the child through years of suffering before action is taken. If Adam had been born today, it is more likely than not, his path would have been a very different one which may well have changed the course of his educational years which were fraught with problems more due to faecal incontinence than Down syndrome.

7.2 Schooling in Australia

Preparations for the first day of school begins in earnest during the year before most children enter kindergarten. In Australia, if a child attends pre-school or a long day care centre, this can be anywhere from birth to 5 years and 11 months of age. Preschool children participate in a School Readiness Program during the second half of the year prior to starting school. These programs attempt to determine school readiness based on social and emotional skills, the child's approach to learning and their level of language, cognition and general knowledge, but place a significant emphasis on the age of the child (Lewit and Schuurmann, 2000). Education in Australia is compulsory, and children start school between five and six years of age, with minor variations between the states and territories. The school year in Australia starts in January and finishes in December.

Australia is a signatory to the United Nations Convention on the Rights of the Child (United Nations, 1989), and therefore every child under the age of eighteen

has the right to access free school education appropriate to his or her needs, which allows the child to reach his or her full potential. The Australian Education Act of 2013 (Parliament of Australia, 2013) states that:

> All students in all schools are entitled to an excellent education, allowing each student to reach his or her full potential so that he or she can succeed, achieve his or her aspirations, and contribute fully to his or her community, now and in the future…and should not be limited due to their personal circumstances.

For children who are diagnosed with a recognised disability, there are processes to follow to ensure their health and safety at school. In Australia state education departments retain primary responsibility for the education of a student with a health condition, even if the student's health status precludes them from attending their regular school or another learning institution (Hopkins, 2015).

Children who have a diagnosed chronic medical condition that requires ongoing management or may entail periods of absence from school can and do wish to remain engaged in education. A child recovering from surgery, such as the children born with Hirschsprung's disease or anorectal malformations, or those experiencing bouts of infection or periods of unmanageable incontinence should have education provided to them no matter where they may be, be it at hospital or at home (Closs, 2000).

Due to advances through digitisation, and the ability to manage more health care needs in the home, the traditional model of a hospital school may well become a thing of the past. Children who were hospitalised for long periods would attend

the hospital school when they were well enough, during their long hospital stay, or the hospital school educators would attend their bedside for short lessons based on how long the children were able to manage. Today children can interact online with their schools; they can participate in programs in live time and remain engaged in the school community remotely whilst they recuperate in hospital or at home (Wilkie, 2011). Due to the recent COVID-19 pandemic students all over the world have experienced lockdowns and school closures, which has led to an explosion of online learning for children across the curriculum (Dhawan, 2020)

A recognised disability as defined in the Commonwealth Disability Discrimination Act 1992 as:

> total or partial loss of the person's bodily functions; or
>
> total or partial loss of a part of the body; or
>
> the presence in the body of organisms causing disease or illness; or
>
> the presence in the body of organisms capable of causing disease or illness; or
>
> the malfunction, malformation or disfigurement of a part of the person's body; or
>
> a disorder or malfunction that results in the person learning differently from a person without the disorder or malfunction; or
>
> a disorder, illness or disease that affects a person's thought processes, perception of reality, emotions or judgement or that results in disturbed behaviour; and

includes a disability that: presently exists; or previously existed but no longer exists; or may exist in the future (including because of a genetic predisposition to that disability) Disability Discrimination Act 1992 No. 135, 1992 accessed 5 September 2019 at https://www.legislation.gov.au/Details/C2016C00763

Therefore, children who have ongoing faecal incontinence due to Hirschsprung's or imperforate anus, do meet the criteria of total or partial loss of the person's bodily functions.

A national review of hospital-based educational support recognised that at a federal level, students with many chronic health conditions could be considered to meet the criteria of disability, but that there continues to be inconsistency within each state's education system as to what constitutes a disability (Hopkins, 2015).

Care Plans for Departments of Education around Australia were assessed to determine which conditions were listed as recognised conditions requiring support. The following table lists the health care conditions and practices recognised by each state and territory in Australia.

Table 7.1: Health care conditions and practices recognised in state and territory educational policies

Recognized Condition	NSW	ACT	VIC	SA	WA	NT	QLD	TAS
Allergies	✓	✓	✓	✓	✓	✓	✓	✓
Anaphylaxis	✓	✓	✓	✓	✓	✓	✓	✓
Asthma	✓	✓	✓	✓		✓	✓	✓
Childhood cancer	✓			✓				
Cystic fibrosis				✓				✓
Diabetes, hyper and hypoglycaemia	✓	✓		✓				✓
Epilepsy	✓	✓						✓
HIV and AIDS	✓							✓
Bleeding disorders								✓
Osteogenesis imperfect				✓				

Spina bifida and hydrocephalus				✓				
Practices								
Clean intermittent self-catheterization				✓				
Continence care needs				✓				
toileting and hygiene		✓						✓
Ileostomy/colostomy care								✓
Gastrostomy								✓
Medications	✓	✓		✓		✓	✓	
Spinal health	✓							
Student injuries						✓		
Infection control								
Head lice	✓	✓	✓			✓	✓	
Hepatitis	✓	✓						
Infectious/contagious diseases		✓				✓	✓	
Insect viruses and allergies							✓	
Living with bats							✓	
Meningococcal	✓							

My analysis of the plans for schools in the six states and two territories in Australia confirms that recognition of a child's disability differs widely across the country. While all states and territories have care plans for the more common conditions such as asthma, allergy and anaphylaxis, some have care plans for very specific conditions such as spina bifida, cystic fibrosis or insect viruses. Whether these specific care plans developed from having a child with one of these conditions enrolled in the school or it was considered important in their geographical area is unknown. What is clear is there is no consistency nationally except for the common conditions. There are also national policies in place, further adding to the inconsistency across the country. South Australia and Tasmania were the only states that included continence, ileostomy or colostomy care (Heays and Debra, 2019c, 2019a, 2019b) and the Australian Capital Territory and Tasmania included toileting in their list of recognised care plans.

More complex healthcare needs than those listed in existing policies require an application directly to the school. The child's eligibility requirements for medical intervention support and funding are then assessed based on the child's current needs. The criteria vary across the country but in general terms the condition must require that the student needs specialised assistance at school and the staff may need to undergo specialised training and/or instructions by a recognised healthcare professional to perform certain procedures. This training must be updated annually to ensure the staff member remains current and is assessed as competent.

For students who attend schools in New South Wales, there is an assessment process for the care needs of the student so they can participate with dignity in the school setting and develop independence, as indicated through the following eligibility criteria:

> To be eligible, students must have essential educational needs that are directly related to their identified disability and that cannot be met from within the full range of resources available from the school and region (NSW Department of Education 2019).

In education as in healthcare, funding is based on a deficit model where the focus is on what the child cannot do. In healthcare this equates to the disability being seen as a problem that needs to be fixed or managed. This is a difficult concept for most parents to reconcile as they spend their lives focusing on what the child can do and what they are capable of doing. As memorably articulated by Hotchkiss, (2016) the deficit model of teaching implies that the teacher "injects" the learning to make good a deficit. This stands in direct contrast to the belief

that the teacher's role is to draw out learners' tacit knowledge and understanding through questioning and facilitation.

The ACT has developed a '*Schools for all program*' to work towards meeting the needs of all students including those with a disability. Their measure of success was defined thus:

> The issues posed by students with complex needs and challenging behaviour should not be problematised, but instead seen as an opportunity, invitation and challenge to further exemplify the inclusive vision of the ACT and its schools (Shaddock, Packer and Roy, 2015 p14).

The Nationally Consistent Collection of Data on School Students with Disability (NCCD) collects data annually from both government and non-government schools about Australian school students with disability. This collection of information is then used to understand the needs of students with disabilities and the best levels of support they require. Schools only include the data of students with recognised disabilities. The school must have evidence that the school has provided an adjustment(s) for a minimum period of 10 weeks of school education, excluding school holidays unless the student engages in school-based activity such as a camp during these times.

In 2000, the UK Government called for a public consultation exercise to understand the existing practice and provision for children with medical needs in schools. As a result, the UK Department for Education and Skills (DfES) jointly published with the Department of Health (DoH) statutory guidance "Access to education for children and young people with medical needs 2008". In a study

conducted to determine best practice in the provision of effective services for children and young people with medical needs, the University of Manchester researchers (Farrell and Harris, 2003) recommended a range of strategies for children who are absent from school due to their medical needs to ensure continuation and inclusion of their educational needs in collaboration with their school and class. Recommendations for future practice included having a school policy for children's education when they are unable to attend with a specific staff member responsible for their educational needs. This would include protocols to guide school staff to maintain contact with the students, liaise with the care providers such as hospitals, and to ensure work is provided in a timely manner. When the students were returning to school either part time or full time, there should also be provision for flexible reintegration and reduced timetables to support them.

7.3 Needs of children with faecal soiling

For children born with a congenital bowel disorder such as Hirschsprung's disease or anorectal malformation, and who have ongoing faecal incontinence or soiling, the process and preparation for school readiness is much more complex. As discussed in earlier chapters, children born with Hirschsprung's disease or anorectal malformation who have undergone life-saving surgery in the early days or weeks of life are often left with long term faecal incontinence or soiling, requiring life-long bowel management programs and support from those involved in their daily care. Menezes et al., (2006) interviewed 259 patients to determine the long-term outcomes of people who had received corrective surgery for Hirschsprung's disease in childhood. Although interviewees had experienced many different surgical procedures for the condition, the entire population

reported a high prevalence of continuing faecal incontinence. What was important in determining better outcomes was the quality of the system of long-term follow-up and support for these children. Several studies have demonstrated a clear association between faecal incontinence and poorer psychosocial functioning and parental criticism (Catto-Smith et al., 1995); (Damon et al., 2004). Although there have been great advances over the past thirty years in surgical management of Hirschsprung's disease or anorectal malformation, the psychological impact remains poorly recognised (King, 2017).

7.4 The invisible disability

An "invisible disability" is something that is "non-visible," "hidden," "non-apparent," or "unseen", and is generally defined as a physical, mental or neurological condition that limits a person's movements, senses, or activities that is invisible to the onlooker. Faecal incontinence is one such invisible disability. Social expectations today demand any issues to do with toileting and defecation are kept private and never spoken about in public. According to Inglis, (2001), defecation is considered one of society's most unmentionable topics and one that is associated with disgust, secrecy, shame and guilt. Despite general lack of discussion of the topic, faecal incontinence has a huge impact on a child's life, especially when they are away from the protected environment of home. The fact that faecal incontinence is so socially abhorrent, along with Hirschsprung's disease or anorectal malformation being seen as surgically correctable at birth, can lead to a lack of understanding, disbelief and judgments. That the child had undergone corrective surgery and still has faecal soiling is often assumed to mean that the problem could no longer be medical but must instead be in the realm of parenting or childhood behaviour.

Anna, the parent of Andrew, recalled a social worker encouraging her to "think positive thoughts" about her son's surgery, suggesting that the soiling occurred because of her excessive focus on the gut. Andrew at the time was 6 years of age and soiled several times a day.

Alice, now aged 9 years, was born with cloaca, a severe form of anorectal malformation. Her mother Joan recalled:

> The school does not understand no matter how much I have tried to explain my daughter was born without an anus and most of her rectum, and yes, they corrected it with surgery, but it has left her with soiling she cannot help.

The difficulties with conditions such as faecal incontinence is the assumption that all children will develop at what is considered a normal rate and by the time they are of school age, they will be able to manage their own continence. Whilst this is true for most children and is the case for many children born with a congenital bowel disorder, but for the children whom continence eludes them despite concentrated efforts from the child and their parents, the prospect of starting school can be terrifying. Starting school is a time when children without a disability often feel anxious, this is exacerbated for both the child and the parents, through no fault of their own, the child is unable to gain continence.

At a teaching session, I asked a group of medical students what they considered could be classified under the heading of 'invisible disability.' The answers were as expected: blindness, deafness, some mental illnesses, autism spectrum disorder, multiple sclerosis, chronic pain. When prompted about faecal or urinary

incontinence, they acknowledged they had not considered it. Invisible, stigmatising conditions like incontinence need much more focus in preschools, schools and included in the curriculum of all health professionals. This doctoral research provides the insights and lens into such consciousness and – and in – curriculum.

7.5 Faecal Incontinence explained.

Faecal incontinence is frequently seen as a behavioural problem, and this has coloured the way teachers understand the problem. Ongoing incontinence issues for children born with Hirschsprung's disease or anorectal malformation is a different issue to those who were not born with one of these rare conditions. The term 'encopresis' was introduced in 1926 by Weissenberg as the faecal equivalent of enuresis which is the involuntary passing of urine, or urinary incontinence (Burns. C., 1941).

Encopresis is divided into functional and non-functional forms. It is important to clarify the difference between functional and non-functional encopresis, as the terms can seem counter intuitive. Non-functional encopresis poses problems for the child's function in daily life. Functional encopresis, on the other hand, has no implications for the child's overall function. Functional encopresis does not have an organic cause. It is defined as repeated involuntary faecal soiling in the child's underwear and can involve passage of anything from a smear to (rarely) a normal-size bowel movement. The most common reason for functional encopresis is retentive constipation with overflow incontinence (Har, A; Croffie, 2010). In their research into toilet design in early childhood settings (preschools or long day care centres), Cliff and Millei (2011) questioned the "panoptic" design

of bathrooms in these facilities. Children's bathrooms have wide vantage points which enable supervision of several children at a time by limited numbers of staff. No doubt this is partly an economic consideration, but these facilities bear no resemblance to the child's experience of a safe and private bathroom at home. A teacher asked the children why they were lining up when there were other spare toilets in the facility, the children responded by saying:

> Rebecca: I don't want to use them.
>
> Teacher 1: Why? Is there a problem?
>
> Tilly: Yeah, people can see us.
>
> ZM: Do you think it's ok to watch one another on the toilet?
>
> Zoey and Talia in chorus: No, it's not ok. (Cliff & Millei, 2011)

One boy flatly refused to use the toilets at preschool and could be seen crouching, holding his legs together and clutching his abdomen. He was considered a 'problem' and recent advice from a counsellor suggested teachers "had to be hard on him" in order to correct his behaviour. Other children were reported to restrict drinking at preschool and would wait to use the toilet at home, often 'busting' by the time they were in the sanctuary of their own home. This raises the issue of 'holding on' and whether, even at this young age this may be a precursor to functional encopresis. Constipation is one of the most common reasons for consultation with a paediatrician Hiscock et al., (2017), and there are now constipation clinics in some children's hospitals in Australia. The problems often continue when the child starts school.

Functional encopresis is considered correctable with combined parent and child education, behavioural intervention and long-term compliance with the medical treatment regimen. Recovery rates are slow, but if the regimes are followed and adhered to, most children see a dramatic improvement. Along with initial medication, simple measures which are often successful include recognising the gastrocolic reflex, toileting routines after meals and correct toilet sitting techniques (Loening-baucke, 2002).

Figure 11: Getting into the right squatting position

Correct toilet posture Elbows to knees, a straight back and a footstool. Retrieved from https://www.coloplastcare.com/en-CA/bowel/routines/build-the-basics/r1.3-toilet-positions/

Non-functional encopresis on the other hand, does have an organic cause, Hirschsprung's disease or anorectal malformation being two of them. While these conditions are rare and the child has usually gone through extensive surgery in the early years, there remain a proportion of children who, despite the expertise of paediatric surgeons, specialist continence nurses and the tireless efforts by parents, remain incontinent of faeces or have faecal soiling (Sanders and Bray, 2009). Various forms of interventional management are available (Table 8.2).

Table 7.2: Interventional management of non-functional encopresis

Rectal interventions	**Method**	**Comments**
Suppositories	Inserted rectally	Difficult once the child reaches 2 years +
Enema	Inserted rectally	As above
Rectal colonic irrigations	Placement of a rectal catheter or cone into the lower rectum or past the anal sphincter. A solution is then attached to the device and flows in.	Intended to wash out the colon into the toilet
Peristeen	Bowel washout system that can be used to manage faecal incontinence and chronic constipation for children over 3 years of age.	Allows the child to participate in their own management. Child sits on toilet as solution flows in, bowel is emptied into toilet. See diagram below.
Surgical interventions	Method	Comments
Antegrade colonic enema	ACE, MACE, Caecostomy, The Chait Trapdoor Catheter. An opening is made into the caecum or first part of the large bowel on the right lower abdomen and a tube or device is inserted, or the caecum is pulled up to form a fistula through the umbilicus, this is less obvious on the child's abdomen but requires daily catheterisation or the stoma will close.	Various methods exist, all with the same aim, to washout the bowel in the direction of the flow. See diagram below.
Stoma; colostomy, ileostomy.	Surgically created opening of the bowel into the abdomen	Requires the wearing of an external appliance as the stoma is incontinent. Examples are depicted in chapter 3 Adam's story

Jing, the mother of Liam, aged 10 years explained the difficulty teachers had in recognising Liam's problem as medical.

> The biggest issue is the school doesn't see his soiling as a medical issue, it is viewed more as an issue with him or my parenting. Like he cannot be bothered to go to the toilet. They have even asked what kind of diet I am feeding him. I wish it were that simple. Despite explaining the condition, the response is often the same: Didn't they [the surgeons] fix

> the problem? A social worker told me to stop dwelling on the past and realise that while he was born with an imperforate anus, it was fixed so it is time to move on.

Sarah voiced the same concerns for her son John aged 9 years:

> Despite giving information to the school, each teacher had no idea about the condition, and I felt, really preferred not to know. They made me feel like his soiling was obviously a case of poor parenting again.

7.6 Disability symbols: making the invisible visible.

Because Hirschsprung's disease or anorectal malformation are invisible, problems arise when the child is out in public with or without their family. They need facilities to clean themselves properly and a more private space than a small cubicle in a toilet block. There are several disability symbols used to denote spaces that are accessible for people with disabilities, as in the wheelchair symbol (Figure 11). The wheelchair symbol was developed in 1968 and indicates access for people with limited mobility. This sign is universally used for access to bathrooms or a telephone that is lowered for wheelchair users. It can also be used to indicate that the person themselves is disabled (for example, for disability parking permits).

Figure 12: Blue square overlaid with a white image of a person in a wheelchair indicates accessibility for people with limited mobility. The symbol is known as the International Symbol of Access (ISA)

Retrieved from
https://commons.wikimedia.org/wiki/File:International_Symbol_of_Access.svg

Not all people who require accessible facilities, have mobility issues. There is a push for a new symbol that more clearly represents people who require accessible facilities, but do not have mobility issues. The Accessible Icon Project in the US and the Forward Movement in Canada feel the original sign is more about the chair rather than the people it is supposed to represent. A new sign was designed to suggest a person in a wheelchair may well have some ability to move. Not everyone is comfortable with the new Dynamic Symbol of Access sign (figure 12) as it implies a level of independence and ability, not everyone who uses a wheelchair has.

Figure 13: Dynamic Symbol of Access created by the Forward Movement

Retrieved from https://torontoguardian.com/2017/05/forward-movement/

There is an array of other symbols which are explained in further details in appendix D., but no such symbol exists for faecal incontinence. Without a visual cue, people in the community cannot recognise when a person with a hidden disability may need additional support. Using a disabled toilet when the child or adult does not have a visible disability, singles them out from their peers and often brings derogatory comments about abusing facilities intended for people in wheelchairs.

Internationally, some changes are occurring in this space. In Alaska, the symbol of a lower care 'i' can be added to a person's driving licence to depict an invisible disability.

Figure 14: Alaskan documentation for persons with invisible disabilities

Retrieved from https://invisibledisabilities.org/ida-getting-the-word-out-about-invisible-disabilities/announcing-idas-national-disability-id-initiative/

Figure 15: Invisible Disabilities International Disability ID Card
Retrieved from https://invisibledisabilities.org/national-disability-id/idid-card/

In 2016, an idea grew from a comment at London's Gatwick Airport when a person was asked "How can we recognise that one of our passengers may have a non-obvious disability?" Discussions were held with several organisations such as the Alzheimer's Society, the National Autistic Society and Visualise Training and Consultancy which resulted in the sunflower being chosen as a symbol to identify passengers with a hidden disability who may need additional help while travelling. The Hidden Disabilities Sunflower program has expanded globally and in 2020, the program partnered with Bayley House, a disability support provider in Melbourne who distributes lanyards and badges for people to wear to identify they may need assistance of some kind. A discussion on a social media site, raised concerns that the Sunflower symbol is not widely recognised by the public or staff at key locations such as airports. This raises the issue that with so many symbols and National Awareness Days, has the impact of these forms of recognition diminished due to overuse or simply the inability for the public to understand what a symbol stands for? Without a national campaign to launch awareness, it is easy to understand who people would not find it easily recognisable.

Figure 16: Hidden disabilities sunflower badge

Retrieved from https://hiddendisabilitiesshop.com.au/hidden-disabilities-id-card.html

Figure 17: Hidden disabilities sunflower poster

Retrieved from https://hiddendisabilitiesshop.com.au/a4-sunflower-poster.html

The campaign for recognition of invisible disabilities was started in Australia by Marni Walkerden in 2018. The slogan "Think outside the chair" is the inspiration for the campaign's inclusive disability symbol.

Figure 18: Invisible disability icon, Australia

Retrieved from https://thinkoutsidethechair.com.au/

Walkerden's campaign was inspired by a campaign two years earlier instituted by Crohn's and Colitis UK: 'Not Every Disability is Visible.' They drew on some ideas already proposed on the website of Enuresis Resource and Information Centre (ERIC). ERIC was originally founded to support families of children who had urinary incontinence, but this later grew to include faecal incontinence. One of ERIC's members wrote on their website about her daughter's experience.

> My eight-year-old daughter has both Asperger's and constipation with overflow soiling. On first sight our daughter does not 'look disabled,' and we've had many a tut and a shake of the head when we exit disabled loos out and about. The issue is that my daughter does not want everyone knowing her business. She is extremely private and found having her diagnosis and medical issues discussed repeatedly with strangers to be very embarrassing. Perhaps a card (or for children a badge) would be a good idea? (ERIC, ND.)

Figure 19: Not every disability is visible icon.

Retrieved from https://crohnsandcolitisuk.s3.amazonaws.com/imager/NEDIV-Logo.png

Figure 20: Not every disability is visible, accessible toilet sign
Retrieved 2019 from https://crohnsandcolitis.org.uk/our-work/campaigns/not-every-disability-is-visible/access-to-toilets

Whilst the array of disability signage is welcome, I have demonstrated some signs are confusing leading to uncomfortable experiences where people with a disability, particularly those with an invisible disability find themselves having to explain in great detail to people in authority such as airports or places of work, why they require additional support or access to disabled facilities. Entering a disabled toilet or accessing a designated disabled carparking spot can often bring disdain and verbal abuse from onlookers who fail to understand their need and are not afraid to voice their disgust that a person who is not wheelchair bound is using such facilities.

7.7 Growing up with non-functional encopresis

Hirschsprung's disease or anorectal malformation are invisible and stigmatising conditions. The subject matter does not attract community support nor does it evoke comfortable dinner conversation (Cavet 2000). Often the stigma is not only directed towards the child, but is also felt by the parent, particularly mothers who are seen as the primary carer in many situations. Mothers often feel a sense of responsibility for their child not being able to achieve continence, despite there being a known, complex medical issue that required major surgery. This often

leads to an imbalance of power between the schools and the mother (Manago, Davis, and Goar, 2017).

Jody, parent of John aged 6 years born with imperforate anus commented:

> I have tried to educate the teachers, who I appreciate have not heard of it before, but I feel they think its poor parenting and I am just not trying hard enough because he should be toilet trained by now. (Jody, parent of John, personal communication, 13 July 2016)

Children who have ongoing soiling or incontinence need careful support in schools, enlisting the help from all involved in the child's medical care. This includes liaising between the parents/carers, teachers, support staff, medical and nursing practitioners and the child themselves to put plans in place to support them without directing unwanted attention towards them. Shared experiences with a focus on parenteral self-efficacy (Bandura, 1982) help improve the outlook for these children (Grano et al 2008).

In a ground-breaking study of its time, Ludman and Spitz (1996) explored the psychosocial impact of ongoing faecal incontinence in children born with ARM by conducting separate interviews with 160 children aged between the ages of 6 and 17 and their parents. There were clear differences in attitudes at different ages, and between the genders. Boys aged 6-7 years seemed oblivious to the social nature of their condition whilst girls were withdrawn. In the 8-11 years age group, boys tended to deny the condition, and girls were secretive. Through the age of adolescence all genders were initially in denial but eventually came to accept their situation. The relationship with peers and parents was often difficult.

Parents often expressed frustration that their child was oblivious to the smell of soiling while the children found themselves excluded from social events such as parties, sporting events and friend bases. Some teachers in high schools attempted to help the children by having a special card system that they had to show to be permitted to leave the room. Although this was well intentioned, in some cases the children found it drew more attention to the fact they were 'different.' Avoidance behaviours developed in some children who would feign illness, or leave school, to avoid sporting events that may involve using communal showers for fear of being singled out.

One sixteen-year-old boy epitomised the difficulties of growing up with such a disorder;

> If one looks okay, then there is no need for anyone to know you have a problem - if they know, they probably will not want to know you, or sit next to you.
>
> (Ludman and Spitz, 1996, p. 563)

During a focus group held during a Bowel Group for Kids Conference, Flavia described her despair for her 11-year-old daughter Elena:

> The other day we were at the park and a group of boys from her class ran past shouting, "She poos her pants." I was devastated for her (Flavia, mother of Elena, personal communication, 4 March, 2017).

Debbie, aged seventeen, who was born with total colonic Hirschsprung's disease, described her need for privacy and to not be singled out:

> If other kids notice my scars and ask about them, they keep asking even if I don't want to talk about it. I don't feel I should have to explain all the time. The girls are talking about their holidays and the new bikinis they have bought. I feel I can't wear one because of all my scars (Debbie, personal communication, 4 March, 2017).

7.8 The difficulty of finding the right educational facility

My own son Adam attended three different preschools and four different schools, during which times we faced never-ending questions about his condition. His first preschool experience at two years and nine months was a special preschool for children with developmental delay which was within the grounds of a public school in a low socioeconomic area. This demountable building was situated on the boundary of the school grounds without running water. Due to Adam's faecal incontinence, we applied for and with the school's help, were successful in having full bathroom facilities incorporated which proved beneficial to students and teachers alike.

The normal transition from this early intervention preschool was a mainstream preschool, so we approached the local community preschool that our three daughters had attended. Feeling confident they had known Adam since birth and the committee was run by local mothers whom we encountered every day at preschool or the local school, I was shocked when I was told that they did not have the facilities to cope with a child such as Adam. I was never asked to attend a meeting; it had been conducted without notice and behind closed doors. I felt let down by our local community. This was a government run preschool and as we had successfully applied and achieved full bathroom facilities for the early

intervention preschool, I would have done the same for them which would have benefited many children in the community, not just Adam. But the door was closed to him. The only option was a long day care centre where they catered for younger children so were used to toddlers in nappies, even though Adam was 4 or 5 by this stage.

The bespoke option that worked best for us was private care. An enrolled nurse of my acquaintance offered unexpectedly to care for Adam while I was at work. I raised the concern about his soiling. By this stage he was booked for further surgery where the likelihood of returning to a stoma seemed to be the only option. I recall asking her about how she felt about managing a stoma and was surprised when she told me she had several family members who had stomas due to Hirschsprung's disease. I was grateful beyond belief and so relieved that Adam finally was accepted somewhere, except he would not experience the social interaction of peers.

When the time came time for him to be enrolled in school, I approached our local primary school that our three girls attended at the time. Adam was the first child they had accepted with special needs. Unfortunately, it did not go well. The school refused to accept any advice from his previous placements, the hospital, or specialists, or me, which I found curious as this would normally be the case for any child that required any form of additional support. The school engaged the local GP's wife as a teachers aid. Everyone, from teachers, to the aid, to the other parents had their own ideas on how to 'manage' Adam. All solutions became increasingly unworkable, culminating in a final condition that he could only to attend for two hours a day, a process which resulted in me undertaking

nearly two hours of commuting back and forth to pick up Adam and his sisters at different hours. Our three daughters found the treatment of Adam unsettling, reporting that he was repeatedly to be found sitting outside the classroom, even in the two hours he was allowed there. We began our search for a second school.

The only option was a high-level support school that he was really not suited on a long-term basis. This turned out to be a pleasant break for him, as he was the pupil most capable of complying within the structured environment. However, he was no longer with his sisters at school, and the burden of taxi-ing him back and forth for long distances was considerable.

Adam started at his third school the following year aged six turning seven and thrived in the kindergarten to year 3 area. The school had play equipment and devoted teachers. Things became challenging when he moved from the supportive environment of the K-3 area to a support class in Year 4-6 area. The school had over 1200 children and countless teachers, many who were unfamiliar with Adam. Adam was a lively child, who had no understanding of rules such as out of bounds. He was constantly being given detention cards which had little meaning to him, so they would be cast aside, resulting in more serious consequences. By year 5 it had become obvious the time had come to look for a suitable place for year 6. The only options we were eventually faced with were to find another school, or for me to be on call to change him every time he soiled. cWe embarked on a new form of management using the anterograde enema treatment, but after a year this had not changed his situation at all. Returning to a stoma was the only option.

Adam's fourth school was a special school for children with intellectual disabilities in Camden. This was not the ideal option as he was quite capable of achieving much more than what the school could offer, but there are few options between a mainstream school or a special school for students with disabilities. This was where Adam spent his remaining school years. The school was able to support his medical needs of a having a stoma, rather than his intellectual needs. I was asked to apply for the new role they were establishing of a school nurse. This had the added benefit of enabling me to spend time with Adam at school for the remaining two years he was there.

The issues we faced with Adam's primary schools were always presented as arising from *other parents*, who were reportedly disturbed by Adam's medical conditions and soiling. That may have been the case, but it seemed in practice that the issues were more the teachers' difficulties in responding to his faecal incontinence and lack of acknowledgement from the Department of Education of the need for support in this area. It became obvious from the persistent comments from the schools, either communicated by telephone or in face-to-face meetings to discuss his future. We were told that that the schools did not have a problem with Adam attending their school, so long as he was continent. Discussions were always geared towards finding a more 'suitable' place for him: one with better facilities, one where there were more support staff. In reality, no such school existed apart from one for high needs students which we felt denied Adam the opportunity for an education where he could reach his potential.

It came down to a need for greater understanding of these rare conditions, engaging the right support and staff who are willing to learn how to best help

the child and be supportive rather than dismissive. As a mother I found it very frustrating to have questions addressing my parenting skills: was I feeding him properly? was I even *trying* to toilet train him. Then questions would centre around the hospital and his doctors such as they thought the last operation was supposed to 'fix him' and couldn't *they* (the doctors) *do something*?

My experience is by no means unusual. In a focus group, parents were asked to describe their own interactions with schools. Glenda, the mother of Adriana aged 9 years who had been born with an anorectal malformation reported:

> I am so tired of negotiating with school. We have had meeting after meeting. The continence nurse was supposed to attend as well but has cancelled three times, I don't blame her. I know she is so busy between the hospital which covers a huge area, not just our school. The school doesn't understand no matter how much I have tried to explain that my daughter was born without an anus and most of her rectum, and yes, they corrected it with surgery, but it has left her with soiling she cannot help (Adriana, personal communication, 1 February 2019),

Sally described her frustration at repeatedly having to brief the staff at the school of her daughter aged 10 years:

> Why do we have to go through this every year or every time my child has a new teacher? We have the plans in place and a care plan in her file and in the staff room, and yet it is like reinventing the wheel every time. The conditions are hard to explain. This year we gave them a pamphlet produced by a support group I belong to, it has certainly made a huge

> difference instead of me having to explain the condition, fingers crossed, but I worry, will it be read by those who need to? (Sally, personal communication, 14 June 2018),

Non-functional encopresis is essentially a hidden disorder, but the problems cannot be underestimated. Faecal incontinence poses problems many schools have never encountered before. If the child does not have an associated developmental delay such as Down syndrome, then it can be difficult to reconcile that a child who looks and functions as though they are perfectly healthy, has this condition that is invisible and also is taboo. Nor is it a recognised condition such as asthma or diabetes. For children in high school where it is commonplace for the adolescent to move around to different rooms with different teachers all day, the child has to adapt to a new environment of not having one dedicated teacher who can be confided in and support the child throughout the year. Some families opt for a change of schools or boarding school as chance for a fresh start if things have become too difficult.

7.9 Inclusive education

In a review of inclusive education (Forlin, 2006) discussed how inclusive practices are 'founded on the need to provide for all children regardless of ability, disability or cultural or socio-economic difference'. This poses great challenges for the teachers to accommodate these differences before even considering the learning and support needs of students with a vast array of disabilities. An interesting issue is that while schools are trying their best to incorporate inclusivity for all, what happens when the adolescents who have been in a relatively protected school community enter the workforce? Can inclusive education prepare young

people for life beyond that supportive environment if their disability remains. Surely inclusivity needs to be a whole community project, not just relegated to the domain of teachers.

For children who are blind or deaf, there are specialist schools, support classrooms within mainstream schools and we often see the entire school being taught sign language to bring a sense of understanding and inclusiveness to the school. We have schools, or support systems within mainstream schools, which cater exclusively for children with an intellectual disability, such as Down syndrome or autism. Exceptions and allowances are made for children born with a congenital heart defect that may require portable oxygen or modifications during physical activities. There is an understanding for children who miss school due to chronic congenital abnormalities such as cystic fibrosis. We have specialist schools or support teachers within mainstream schools for children with a recognised physical disability and there is support and empathy from both the community and the school for children born with cerebral palsy, paraplegia, quadriplegia, and if they also require continence support, it is seen as part of the package they were born with, almost understandable that they may lack control of those bodily functions.

These schools or classrooms have dedicated teachers who provide the support these children need to allow them to receive education in a supported environment. This raises the question then as to why this is accepted and supported. Is it because they have an overriding disability that is visible, and therefore incontinence is seen as just part of the package? For my own son Adam, this has long been my thought. Following the pull through surgery which is

termed as 'definitive surgery' his faecal incontinence was seen as a 'problem.' He had Down syndrome, but he was mobile. He had language skills when he was younger, he could do a lot of things for himself, he was capable of dressing, eating, following directions, he was continent of urine. Why couldn't he control his bowels, particularly when the problem had been 'fixed' long before he started school?

Once the decision was made to return to a stoma when he was about six years of age, the perception change. This was now seen as just another one of his medical conditions, not unlike his two implanted hearing aids for profound deafness. Understandably for the schools a stoma was much more manageable. I taught the relevant staff how to change his appliances and apart from the odd accident, the years of turmoil abruptly ended. We saw a completely different approach, with empathy and understanding. The school staff may not have understood the condition, but continence was no longer an issue they had to deal with.

This highlights the diversity required of teachers to comply with inclusive education which stipulates that Adam be included in school and entitled to an education. Having Down syndrome was the easy part. Down syndrome is recognised as an acceptable form of disability, and he could well have been educated in a mainstream school if he had no other condition. The fact that he also had faecal incontinence changed things dramatically. There are no standard provisions, protocols, or policies to accommodate a child who suffers from faecal incontinence, Down syndrome is seen as an acceptable and manageable disability, unlike the aberrance to faecal incontinence. As much as most parents

of children with Down syndrome will attest, the outdated stereotypes (Keighley, 2017) - "they're so happy!" "they're so friendly" and "they love music," for example - to project an image of a child who will be relatively easy to manage and teach. Having in one's classroom a child who may or may not have an intellectual disability, and faecal incontinence, demands a real commitment to inclusive education that the teachers may not have been prepared for.

There is now a realisation than understanding the diversity of inclusive education needs to start long before the teachers are in front of a classroom (Campbell et al., 2003). Forlin, (2006) argued that if trained teachers have the will, then they will seek out the competence. The thought was that if teachers were exposed to families and had to understand the range of disabilities before they were teaching a class themselves, this may well change their own beliefs about inclusive education in a positive way. This faith in experiential education is also applied to the medical students I teach; many have had the opportunity to spend a few weeks in special schools and the program Adam was involved in for a few years when he finished school as a young adult. There is no doubt that for some students, this was their first encounter with a person with a disability and it certainly makes an impact. Whether this is a lasting impact is yet to be seen.

Where do children born with a congenital bowel disorder who do not have any visible disability 'fit' into any of our existing education systems? Children who were born with Hirschsprung's disease or anorectal malformation who have ongoing faecal incontinence are poorly understood. Faecal incontinence does not attract community support, no-one puts up their hand to volunteer to be the staff member who will go and help this child clean themselves or shower them and

most schools lack those facilities. This is not a criticism of the education system or teachers and support staff, rather, it represents a failure to comprehend why the child is incontinent. One can understand why the comparison may be made to other congenital conditions that are surgically corrected in the early years of life, so by the time the child reaches school age, there is little evidence that a problem existed at all, or it is managed outside of school hours so does not pose a problem at school. Hirschsprung's disease or anorectal malformations are rare, congenital abnormalities that most people have never heard of, and despite bowel function being something every human being relies on as a mostly daily necessity, in our society today, it is managed behind closed doors (Inglis, 2001).

Parents were asked in one of the Bowel group for Kids workshops about the resources in schools

> My child comes home with soiled pants nearly every day. I am at my wit's end. I have had meetings galore, there are care plans in place, but they say there is not enough staff to clean him up each time. They just don't understand, I hate this disease.
>
> (Patricia, personal communication, 4 March, 2017).

> Just because our kids have a rare condition, doesn't make it any less important. If my child needed an EpiPen they would give it without question, but just because soiling is not life threatening doesn't mean it is any less important to my child.

(David, personal communication, 4 March, 2017).

Without constant and open communication, many parents would not have survived the ordeal of schooling. Most of the problems parents encountered were simply due to ignorance and due to a condition that a family members, friends, and community members were coming to terms with. Often the team of specialists, the school, the support staff, and the whole family are learning as they go along. This can be exhausting as parents feel they have to question every step of the way, or they fear their child would be inappropriately placed had they not challenged every quick fix that was suggested.

As parents of children born with Hirschsprung's disease or anorectal malformation know, there is a gradual transition from advocating for our children, to slowly handing over the reins to the many people we entrust with our child's care. Every parent hopes they have done enough groundwork, engaged the right people and supported their child enough to navigate their way towards independence. Clearly much more remains to be done to help educate school staff about the long-term effects for children born with Hirschsprung's disease or anorectal malformation.

7.10 Secondary school

Once children reach adolescence and begin to move away from home, they are unlikely to respond to parents badgering them with question about toileting and volumes of water drunk and reminding them to pack spare underwear when they go out with friends.

Mary, mother of Sarah aged 15 years who was born with imperforate anus became guarded about asking her if she had remembered to take enough water. The response, Mary said, was usually just 'the look".

It is widely expected that all children who are around four years of age will be continent of both bowel and bladder and have 'good toilet habits' (Reeves et al., 2012). Toilets are a crucial space for all school students and yet can be the source of ongoing physical and psychosocial development. In a survey of school students aged 4-11 years, children were asked for their perceptions of school toilets (Barnes and Maddocks, 2002). The majority reported they would resist using the school toilets for defecation and only do so if desperate, preferring to wait until they got home. Some comments were concerning to the researchers. In some schools, toilet paper was only accessible from the teacher if they wanted to have their bowels open; students were only allowed to leave the classroom to go to the toilet in break times; the toilets were dirty with urine all over the floor and the locks had been kicked off. The adverse conditions in school toilets which results in children resisting to defecate at school may result in chronic constipation (Barnes and Maddocks, 2002; Dimitri, Davidson, and Wright, 2006; Vernon, Lundblad, and Hellstrom, 2003).

Sarah, a fourteen-year-old who participated in a workshop for younger persons with faecal incontinence echoed these points. Sarah was born with an imperforate anus:

> It is hard at school because going to the toilet can be noisy and smelly, it's embarrassing as the toilet doors have big gaps, so I have to try and go when no one is in the toilet block. When my bowel hurts, I feel I can't tell

anyone because they won't understand (Sarah, personal communication, 4 March, 2017).

While teachers have gained a vast knowledge base on school readiness, this preparation rarely includes any education on the elimination needs of children starting school (Boyt, 2005). (Tatlow-Golden, O'Farrelly, Booth, and Doyle, 2017) recognised the lasting effects and health consequences of delaying toilet use in young children and highlighted the importance developing school policies incorporating the knowledge and experience of both teachers and nurses. The ERIC Nurse Early Interventions Project was launched in 2013 to raise awareness and education about childhood continence and was aimed at all frontline health professionals, importantly, the project identified a gap in teacher training and support (ERIC Nurse Project, 2016). This gap was later rectified with the development of the "Right to Go" campaign and charter for schools along with education sessions for school staff (ERIC Nurse Project, 2012).

For students with a recognised but invisible disability such as faecal incontinence there is a disconnect between using the 'normal' school toilets as opposed to using disabled toilets, if they exist in the school space, or using the teacher's toilet. While using an alternative toilet is seen as a good compromise, it separates the student and puts them 'in their place' (Kitchin, 1998). As discussed above, school toilets are notorious for being inadequate, unclean, and seen as a place of risk and anxiety for some students, offering very little in the way of privacy or clean facilities (Slater et al., 2019). Teachers are in a difficult situation because they are at risk of being accused of inappropriate behaviour if the child needs

assistance. Likewise, the child is faced with possible ridicule if they are unable to meet societies expectations.

As the child reaches maturity, their perception of the conditions may change, indicating a level of acceptance. For Stephen aged 17 years, Hirschsprung's was not stigmatising, just a marker of individual difference – as different as others with more obvious differences.

> Well, Patrick wears glasses and hearing aids, that's just part of him. Darren has two front teeth like a rabbit but that's him, he has always had them, and he always will. And so, the way I see it, I have Hirschsprung's…no big deal. I had it all my life, always will. That's me. I'm as normal as they are (Stephen, personal communication, 4 March, 2017).

Hannah aged twenty-nine born with imperforate anus, echoed this account. She was now a mother of twins who are not affected by the condition:

> I was born with an imperforate anus; my mum and I gave our own account of our experiences at one of the Bowel Group for Kids conferences. When we compared our talks, I realised that mum had a different view of what I had been through. She worried I wasn't enjoying a normal childhood, always at the doctors or in hospital, she was concerned I would be psychologically damaged having had lots of bowel management and rectal examinations, but to me, I didn't know any different and just saw it as that's the way it was (Hannah, personal communication, 30 June 2018).

Schools are instruments of inclusion that could have important impacts upon how students experience their body in society. Children spend at least six hours a day

at school in close association with each other, the teachers and support staff. There are multiple opportunities to discuss the importance of inclusion, our differences and acceptance of these, particularly in personal care needs which may arise in school toilets, change rooms and swimming venues. While inclusive education for all can be considered a global commitment, this research identified different approaches across each state and territory in Australia, so a more uniform approach to the care needs of children at school is needed and a recognition of the impact incontinence may have on a child and their siblings who attend the same school and their parents.

In 2014, the Australian Medical Association released a position statement which recognised the importance of support within schools for children and young people who have chronic medical conditions and special needs. Their position was that these children and young people should not be prevented from engaging in education, and that this was particularly important for children who spend significant periods of time in hospital due to their medical conditions or those who are absent from school for health reasons. The AMA also recognised the importance of shared learning through education and support for teachers regarding essential and emergency medical care, right through to medical students and treating doctors to engage with the student and their school to enhance their learning experience (Strait, 2014).

Many parents of children born with Hirschsprung's disease or anorectal malformation lament the difficulties they face if they approach their general practitioner (GP) to write a supporting letter to the school on their child's behalf simply because the GP has little understanding of the complexities of the

conditions. Long-term management is often exclusively the domain of the treating specialist teams of paediatric surgeons or gastroenterologists who are not as accessible as the local family physician.

Due to the spectrum and complexity of soiling and/or incontinence, it is important to include the child's clinicians in determining the kinds of supports required based on medical need rather than a formal diagnosis. No two children who began life with a congenital bowel disorder have the same needs, unlike some conditions such as Type 1 diabetes where each child must be administered insulin to survive (Strait, 2014).

7.11 Conclusion

The provision of supportive and integrated care is required for many students who present with a chronic medical condition. Incontinence and/or soiling is a complex condition that requires sensitive, supportive interagency cooperation to ensure they have the same learning opportunities as other children to learn. Changes are needed to our education system, the training of teachers and support staff from preschool to secondary school and inclusion of a greater awareness of encopresis should be part of medical and nursing training.

CHAPTER 8: EXPLORING PATIENT SUPPORT GROUPS: ORGANISATIONAL STRUCTURES, ACTIVITIES, AND WORKERS

8.1 Introduction

An important aspect of this research was to avoid bias by reporting on support groups globally, similar to the Australian Bowel Group for Kids that I was heavily involved in. By understanding how other support groups operated, I was able to compare similarities or differences between them. This was only possible due my in depth understanding of how support groups operate, what was important from a participant's perspective and how support groups can assist clinicians in understanding the needs of families when a child is born with a congenital bowel disorder. It is through the unique knowledge and experience gained over 40 years as a registered nurse; the vast learning experience enhanced over 33 years managing my own son's complex needs, consolidated with 26 years of running the support group where I was fortunate to further my knowledge of other parent's experiences that culminates into my contribution to knowledge that has a unique perspective on congenital bowel disorders. This chapter sets out the quantitative analysis of data on twenty-six patient support groups, seeking to describe the organisational structure of these groups, and key elements that respondents for the groups thought led to their success. The survey included questions on the organisation's structure, its funding sources, how members contacted support groups and continuity of membership. The services offered by the groups were explored, whether campaigns were currently running to change government policy related to the specific conditions and if the organisations had a method to evaluating the effectiveness of their services. It was important to

understand how other support groups were formatted for comparison and to gain insight into what additional means of support may help parents through the difficult years supporting their child who was born with a rare congenital bowel disorder. It was not feasible to conduct a study only relevant to Hirschsprung's disease or anorectal malformation as the numbers of professionally based support groups for Hirschsprung's disease or anorectal malformation are very small worldwide. Other support groups for rare or comparatively uncommon conditions were included for comparison. Most of these related to gastrointestinal conditions and included support group for families who had a child born with short gut syndrome, paediatric gastro-oesophageal reflux, coeliac disease, gastroparesis, chronic intestinal pseudo-obstruction, and colonic inertia. There were a few non gastrointestinal surgical conditions of childhood: trachea-oesophageal fistula, spina bifida, hydrocephalus, cleft lip and palate[EG8].

The data used throughout this chapter was sourced from publicly available sites such as Facebook and the support groups data available online. I developed the data sets with my own leadership and the sources were all open access. Support groups were sent the surveys I developed and were asked to respond.

8.2 Results

Most groups had similar origins, usually an educated parent who had a child born with the condition and a zest for further knowledge and understanding and a desire to share this with others in a similar situation. The length of time groups had been operating ranged from over fifty years to seven years. Few had paid staff and half had good relations with the local medical and para- medical fraternity. The majority had a team of long-term enthusiastic volunteers who

remained with the group to support others long after their own child was no longer a concern.

A desire to connect with and learn from others in a similar situation was reported as the main reason people joined the organisation (Table 8.1). Half of the organisations reported members had a more severe form of the condition than non-members .

Table 8.1: Reasons for joining the support group

Reason for joining the support group	Percentage of groups (%)
Education and information	74
Lack of family support	17
Connect with others in a similar situation	83
Online forums/information board	48
Learn from more experienced members	70
Attend conferences and informal meetings	26
Access to equipment and supplies	4

8.2.1 Organisational structure

The majority of the groups were formally constituted with Boards of Directors, or other governing structures, and 77% were incorporated organisations (Table 9.2). Incorporation protects individuals from being held individually responsible for their actions, and this is particularly important since 92% of the organisations use volunteers, with nearly half of the organisations having ten or more volunteers. More than half the services provided training for volunteers, but only 20% of organisations had a code of ethics for volunteers to follow. Twenty-three percent did not have a medical advisory board.

Table 8.2: Types of support group organisational structures

Management of groups	Percentage of groups (%)
Board of Directors	8
Incorporated	77
AGM	84
Paid staff	58
Volunteers	92
Medical advisory board	77

One of the biggest concerns for the organisations was funding. Sixteen percent had joining fees and 65% had annual membership fees, but the groups were too small for this to constitute an appreciable source of funding. Many groups engaging in high-effort, low-return activities like sausage sizzles, fundraising evenings (Table 9.3). Only one group was in receipt of a philanthropic trust, which would provide some security.

Table 8.3: Funding sources for support groups

Type of funding source	Percentage of groups (%)
State or federal government	45
Corporate sponsorship - regular	23
Corporate sponsorship - occasional	42
Annual fundraising	20
Crowdfunding	24
Raffles and lotteries	20
Trivia nights and dinners	7
Sausage sizzles	7
Merchandise	4
"Attention events" e.g., cycling for a cause	4
Philanthropic trusts	4

8.2.2 Services offered by the sector.

Nine organisations surveyed provide printed newsletters, while eight produced online versions. A small proportion organised sibling support, camps, and formal dinners, whilst twelve organisations provided local meetings, thirteen ran conferences. eighteen organisations provided education for health professionals, whilst eight provided information brochures about the conditions covered. Only eight were able to provide in-services to pre-schools or schools, whilst six organisations visited families in hospital or their homes. Seventeen respondents provided training for volunteers. Increasingly social media such as Facebook, forums, chatrooms, email groups and twitter are being used as a means of gaining quick responses to queries from members who have previous experience or just to share thoughts with someone who understands. Physical meetings were

less frequent, either due to distance, time restraints or the embarrassment of attending a group with a personal issue such as incontinence.

Half of respondents conducted their own research, whereas feedback about their own organisation and its programs was gained at the conclusion of events. Most of the feedback was generalised relating to the suitability of a venue, what families would like to read in future newsletters or the kinds of speakers they would like to hear from at future session. By far the emphasis from participants at conferences in most organisations and certainly the Bowel Group for Kids, was hearing from other parents and older children who had walked their path before. Presentations from older children who were able to give their own accounts of their recollections of growing up with a bowel disorder and how they managed through challenging times was very encouraging. For parents of younger children, hearing success stories, even after difficult earlier years, were reported as highlights of the day. Next was the workshops that brought families together in their child's age groups where specific questions were asked and insights were gained, then shared among the entire audience. Insight from professionals who had significant experience managing the conditions was welcome, but the real life stories from families and older children were by far the most well received and requested for inclusion in future conferences.

The Bowel Group for Kids developed pamphlets for families to assist them, their families, carers and teachers to understand the condition and complexities their children are dealing with. These have been updated regularly and include pamphlets on the support group in general, the conditions covered, toilet training, general information around trips to hospital and many more in collaboration with

stomal therapy nurses, surgeons and anaesthetists. These ought to be available nationally to all families as soon as their child is born with Hirschsprung's disease or an anorectal malformation.

8.2.3 Engagement with the health sector

Eighty percent of those surveyed reported an affiliation with hospitals and doctors. Whilst the majority of the groups surveyed had a good rapport with healthcare teams, most found promoting the group one of the biggest hurdles as it really needed a champion in the hospital system to maintain the connection or regular attendance by support group organisers. Most people found out about the support group through word of mouth (71%), or through direct referral from health professionals (83%). The organisations that reported the most difficulty in this area were those relating to the bowel as they do not attract community support nor are they considered newsworthy by the media. Families may also prefer not to be associated with an organisation that is for a condition they prefer to maintain privacy about such as a bowel condition. The collaboration between clinicians, the hospitals and parent support groups are seen as important to provide a well-rounded view, respecting each other's knowledge and experience.

There is a role for clinicians within the hospital system to connect parents with support organisations if they so wish. Not every parent wants or needs that connection with others who are brought together due to some congenital anomaly or illness, but the information should be available at the time of diagnosis, so they have a point of contact if they choose to follow it. The benefits of shared knowledge and experience between new and more experienced parents, cannot be understated. Parents say they no longer feel alone and

isolated dealing with conditions that are poorly understood by the public and they find hard to discuss with family and friends.

8.3 Conclusion

Despite differences in specific circumstances, there is a wealth of experience to be gained from sharing the lived experience of a particular illness (Dadich, 2008). The relationship between support organisations and clinicians is of vital importance to provide a well- rounded balanced support for patients and their families. Respect for each other's knowledge is an important factor in the relationship; support groups can provide insight to parents from the unique perspective of the day-to-day lived experience. Healthcare providers on the other hand have learnt about the condition through training and experience and can provide diagnosis and management of the condition. It is this shared wealth of knowledge that can be of great support to the formal health system and benefit all parties (Adamsen 2002; Boyle et al., 2003).

The non-profit organisations provide a valuable resource of accumulated knowledge and education on dealing with the day-to-day issues encountered with specific conditions. Transition periods experienced by children were peak times for parents to seek help from others whose children were older. Typical times of need were identified as when the primary caregiver returned to work requiring the child to be placed in some form of childcare; transition from pre-school to school, infants to primary and onto high school. The transition from paediatric to adult care services is recognized as a difficult time for families.

The groups relied heavily on the founding members and volunteers who gave their time, wealth of knowledge and expertise freely There are significant

concerns about the sustainability of these organisations as many do not have strong sources of funding. Volunteers – while critical resources for support groups – also need frameworks that clarify their roles, including codes of ethics. There is a need for a more collaborative approach to include optimal support services to families who have a baby born with Hirschsprung's disease of an anorectal malformation.

CONCLUSION

TIME FOR CHANGE IN THE WAY CARE IS PROVIDED THROUGHOUT THE LIFESPAN OF PEOPLE BORN WITH HIRSCHSPRUNG'S OR IMPERFORATE ANUS.

This doctoral research explores the organisational cultures encircling the child with Hirschsprung's disease or anorectal malformation through the way this condition is institutionally framed within the hospital, the surgery, the family, the school, peer support groups and allied health clinicians. Hirschsprung's disease and anorectal malformations are rare congenital conditions affecting 1 in 5000 live births and are sometimes associated with other conditions, the most common being Down syndrome. My significant original contribution to knowledge is shaped by my extensive experiences as a nurse, and educator of future clinicians, as the mother of a child with complex disabilities and draws on 29 years of work with Australia's first peer to peer support group for children and families affected by Hirschsprung's disease or anorectal malformation. Interviews and workshops were conducted with parents and children, surveys of paediatric surgeons and peer support groups, and an analysis of educational support policies and procedures for children with congenital bowel disorders formed a collective understanding of these complex conditions which can have lifelong consequences.

Whilst the surgical approaches to these conditions have advanced over the years, this thesis demonstrates that the recognition and support for children in society, schools and the workplace, who suffer ongoing faecal incontinence has remained hidden and the problems this creates has not been addressed. This thesis

demonstrates the areas of urgent need for these children who through no fault of their own are unable to achieve faecal continence according to societal expectations.

The standard surgical approach to Hirschsprung's disease and to anorectal malformations traditionally involved the creation of a stoma to enable the anatomically compromised bowel to empty. This was followed sometime later by a complex procedure in which the healthy bowel was anastomosed in order to allow faecal evacuation via the anus and the stoma was closed. More recently, improved surgical techniques have enabled a single stage procedure avoiding the formation of a stoma in otherwise healthy children. Although there has been a great deal of surgical literature exploring various surgical techniques and ensuring that immediate poor outcomes such as Hirschsprung's associated enterocolitis (HAEC) are minimised, there have been some gaps in the literature on the long-term follow-up of these children. Surgery, by its very nature, tends to view success or failure in terms of immediate outcomes. Surgery on a neonate with Hirschsprung's disease or an anorectal malformation is lifesaving; a paediatric surgeon was therefore likely to gauge their success or failure on whether the child lived, without substantial complications such as life threatening infections postoperatively. Subsequent procedures conducted on infants and young children seek to protect a child from the experience of having a stoma for life, and paediatric surgeons again are likely to view the outcome here as whether the child is able to develop functional use of their bowels.

The large long-term studies now being conducted in centres with decades of surgical experience for these young children with Hirschsprung's disease or an

anorectal malformation, demonstrate that whilst most people do gain faecal continence, a substantial number of surgery recipients have ongoing faecal incontinence, ranging from soiling to explosive loss of control. This is most marked in childhood and early adolescence and generally improves somewhat in later adolescence and adulthood as the person learns to manage the condition. Repeatedly, the follow-up literature states that paediatric surgeons may have underestimated in their representations of outcomes to parents, and in their own conceptualising of success, the likelihood of ongoing faecal incontinence, and its impact upon children and their families.

That they have underestimated the prevalence of faecal incontinence reflects failure of evidence generation in paediatric hospitals addressing long-term outcomes for surgical procedures on neonates and young children. Such studies are technically challenging, as they require the generation of registers of children with these conditions, and the development of formal follow-up. We are familiar now with the requirements of such follow-up for procedures – for example, in the outcomes of breast cancer surgery, comparing total mastectomies and lumpectomy and node excision. However, surgical procedures using devices are often implemented with insufficient monitoring and rigor, as in the current example of transvaginal mesh being used for primary management of uterine prolapse, which has been banned in many countries and become the focus of medical litigation (Izett et al., 2018).

Children who receive life-saving paediatric surgery have historically not been followed into adulthood. Part of the problem is the complex transition of children with medically challenging conditions requiring surgery transition into adult

specialist care. These transitions are often haphazard, and many of these children will transition from being viewed as primarily a surgical object of care to becoming viewed as belonging within chronic medical care services, managed by internal medicine specialists. In fact, the preliminary long-term data of children – now adults – with Hirschsprung's disease has prompted calls for long-term surgical follow-up and monitoring for adults with complex medical needs, with particular attention on the transitions in adulthood and adult care (Cairo et al., 2018). In the absence of expansive longitudinal data sets, it has been possible to be overly sanguine about the experiences of children, adolescents, and adults with Hirschsprung's disease.

The long-term data emerging over the last decade confirms what parents have reported through my research: that some children continue to experience major difficulties with continence, and that this can continue to impact upon them through their adolescence. Although the surgical follow-up data suggests that adults often develop better control, the psychosocial impact of faecal incontinence on children growing up is considerable, as is the management burden upon parents of the affected child.

If the failure to appreciate the prevalence of long-term faecal soiling or incontinence reveals shortcomings in surveillance and monitoring of surgical procedures, lack of recognition of the impact of this on a child's identity reveals a failure of imagination. This failure of imagination is reiterated throughout this thesis – from teachers who view children with faecal incontinence as a sign of poor parenting or behavioural stubbornness, to the surgical establishment which interprets as a success a finding that only one fifth of respondents regularly

experience incontinence, to the hospital institutions which do not regard these children as in need of support into adult institutions.

Stigma and Hirschsprung's disease or anorectal malformation

This thesis has addressed an uncomfortable topic. Faecal incontinence is a condition which is readily stigmatised, reflecting the cultural faecal habitus described by (Inglis, 2001). The affective dimensions of guilt and shame associated with faecal habitus – triggered in part by the sensory dimensions of excretion – mean that adults and adolescents with anorectal malformation or Hirschsprung's disease are very vulnerable and should be viewed within the hospital setting as needing heightened support in the community.

In his personal memoir of growing up with anorectal malformation, Greg Inglis describes the strategies used to try to mitigate stigma:

> I must deal with the uncertainty of what may occur with my bowels every single day of my life. I learnt to cope with being known as the smelly kid at school; on the football field with my best-mates by using the strategy of rubbing dirt onto the back of my shorts, so no-one would notice the inevitable soiling patch; at high school I wore my school jumper tightly around my waist, no matter the weather, to camouflage the stain on my school pants.
>
> (Inglis 2018, p 6)

It is no coincidence that Inglis in this excerpt focuses particularly on school. This is a time when our psychosocial identity is being formed, and when (in Goffman's

terms) stigma can "spoil" that identity. The research described in this thesis indicates that schools are unprepared for children who have faecal incontinence associated with Hirschsprung's disease or anorectal malformation. This is expected, given its rarity. *Functional* faecal incontinence (that is faecal incontinence not associated with a medical condition) is however not particularly uncommon in the early school years, associated as it is with constipation. Most teachers will have at least some experience with a kindergarten child with functional constipation with some overflow. It is therefore not surprising that they bring the framework of behavioural psychology (sometimes also, dietetics) to give meaning to the experience of the child with non-functional faecal incontinence.

This downplaying of the significance of medical need for these children is also related to the popular notion that Hirschsprung's disease or anorectal malformation are surgically curable conditions, corrected soon after birth. This faith in the notion of surgical cure can persist independent of the evidence presented to teachers, clinicians, and family friends that the child still has incontinence, leading the person to the inevitable conclusion that the incontinence is really a matter of behavioural control or poor parenting.

The role of support groups for Hirschsprung's disease and anorectal malformations

Since the setting of stigma is a relational process, re-figuring social relations may go some way to ameliorating stigma. Our cultural faecal habitus is likely to be resistant to change. Public health /campaigns around toileting and sanitation attract little international attention, again reflecting the cultural disinclination to

prioritise this issue over other more "saleable" campaigns. The Sustainable Development Goal related to defecation (SDG 6: to ensure availability and sustainable management of sanitation and water for all by 2030) now appears unlikely to be met (United Nations, 2019), and the statistic that more people in the world have access to phones than have access to effective toilets has been cited since 2010.

Support groups for conditions that may be socially stigmatised are particularly important. This thesis has recounted parents' narratives of the importance of peer to peer support in connecting previously isolated parents and carers. While connections and information-sharing would be valuable for any carers for children with any rare condition, parents in support groups such as the Bowel Group for Kids are able to share experiences with one another that they may not be able to talk about in other fora. Management of persistent faecal incontinence requires some advanced technical skills, sharing of tips and tricks, and sharing of parenting advice. Thus, peer to peer support groups seem to act to help destigmatise the condition to a degree, by providing a forum for parents to share experiences and normalise what would to most people appear to be very abnormal. It is likely that those people who have had an excellent outcome from surgery are unlikely to seek support or persist with a support group. The peer to peer support group therefore acts as an avenue for knowledge exchange and peer support for those who have the most complex symptoms to manage.

The future of support groups

Support groups have moved on from the early days of parents seeking help through telephone calls or letter writing to a support group who in turn would

publish the letter and relevant articles in newsletters three or four times a year, mailed to the homes of subscribers. Most support groups now have a presence of social media, and some of them are solely present of social media. Whilst closed Facebook groups and microblog sites like Tumblr or Reddit offer ways for participants to readily communicate online with any question, they may suffer from not offering tailored communication and real person supports. The progenitor of the self-help movement, Alcoholics Anonymous, continues to maintain a person-to-person presence. Online AA chatrooms are available, but the organisation is careful to state that these groups do not replace attendances at meetings. Their argument is that peer to peer mentoring and support works best when humans are in each other's presence or are talking to each other through real-time synchronous communication (such as on the telephone).

The movement to online support groups can result in professionalising of the support group, as in the case of the support groups moderated by the staff of paediatric hospitals. Using staff members to moderate a support group brings in some expertise, but it runs the risk of ablating the voice of the patient and running a particular argument about medical treatments. Pharmaceutical and medical device industries have a history also of setting up or co-opting support groups to become lobby groups for treatments, or to encourage uptake of these treatments. These industries currently have little interest in Hirschsprung's disease or anorectal malformation support groups presumably due to the lack of financial incentive as a result of the small numbers. It is possible however, that hospital based support groups may face conflicts of interest in supporting or advising parents who wish to pursue a complaint or contest treatment plans. Due to the COVID-19 pandemic of the past three years, support groups have been

left little option but to conduct their business online, and like other aspect of business; as a community we have become accustomed to working from home, doing business online, shopping online and children have had to adapt to learning online at times. This rapid change of circumstances thrust upon the community due to the pandemic, has certainly accelerated how we use online systems. Support organisations are no different, the use of social media has increased to a point that for many people seeking help, this is their first port of call in the hopes of finding a group of people who are experiencing the same set of symptoms or congenital abnormality as they or their children are. Connecting with people online can be less confronting than entering a room full of people and being expected to participate in some way.

I have demonstrated in this doctoral research that peer to peer support groups for families, carers, and children with Hirschsprung's disease or anorectal malformation warrant heightened support given the stigmatising nature of the condition. There is a case that they should receive dedicated state support to provide an independent support group, perhaps analogous to the Multiple Sclerosis Support groups associated with MS Australia. These are integral to support patients after diagnosis and through their journey with MS including making decisions around treatment.

An effective support group ought to be part of the paediatric surgeon's referral network for every child after diagnosis – again analogous to the MS Australia support groups. This will then provide the parents with some form of contact if they wish to pursue it, once their baby is discharged from hospital, this is when they are most vulnerable and would most likely benefit from peer to peer support.

The narratives of parents have indicated that there are some critical times when access to peer to peer support groups may have the most value. These are firstly, at the time of diagnosis, usually shortly after the birth of a baby. Parents reported celebrating the joy of giving birth to their baby who appeared perfectly normal and healthy, only to discover within days that they have a serious congenital abnormality which requires urgent life-saving major surgery.

The second critical period for support is after the shock of surgery. Most parents were understandably upset seeing their tiny baby recovering from major surgery, many trying to juggle other children at home, sometimes travelling great distances as these operations are only performed at major paediatric hospitals in the cities. In New South Wales there are three such paediatric hospitals which are hundreds of kilometres from many rural and remote towns.

The third critical period is when learning to manage "the new normal." Parents reported having to learn how to manage stoma appliances, feeding tubes, severe excoriation of the buttocks, bowel washouts, anal dilatations and long hospital stays and follow up appointments. Often their children suffered from constipation, incontinence or stool retention. There is a huge need here for skills transfer. Meeting an everyday parent from a peer to peer support group who has managed to master these skills for their child can be empowering for the parent navigating "the new normal." Toilet training for children who have endured invasive bowel surgery is also a related period with skills and knowledge sharing in this highly complex area between parents. The shared knowledge is invaluable and not something other parents can understand or relate to.

Parents can lose heart when their child is battling to manage at school, and they have not gained faecal continence according to societal expectations. The realisation that despite successful surgical intervention, there were some children that were unable to achieve continence was a severe blow to families, the affected child and often a source of confusion and frustration for clinicians. It soon becomes clear non-functional encopresis or faecal incontinence because of being born with Hirschsprung's disease or an anorectal malformation was not an isolated situation. This discovery steered the research into a much greater depth of analysis of the information conveyed to parents following the definitive surgery, teacher understanding of these conditions and the availability of appropriate school support staff and facilities, such as school toilet blocks. The child's well-being and resilience up until this period in a child's life has largely been the responsibility of the family, who often manage this hidden disorder without support, hoping it will resolve by the time the child reaches school age which is often not the case.

Education system

The research explored the lack of awareness, education, support systems and facilities available in the Australian public school system for children born with Hirschsprung's disease or anorectal malformations. While there is a clear recognition of well-known childhood conditions, such as asthma, allergies, anaphylaxis, and diabetes evidenced by the provision of care plans across all states and territories, no such uniformity exists for children who have faecal incontinence. Health practices were consistent for the administration of medications, but support material for staff to understand and help children with incontinence, toileting or stoma care, were rarely addressed.

Parents can approach the school for adjustments for their child which are actions taken to enable the students with a disability to access and participate in education on the same basis as other students. Whilst this is a welcome recent addition to the provision of care in schools, this requires the recognition of Hirschsprung's disease and anorectal malformations to be classed as a disability. There is also a process for parents to make the application for adjustment which involves the provision of letters of support from all treating clinicians and meeting certain criteria. In Australia the National Disability Insurance Scheme does not recognise any disability that is classified as a medical condition, this is due to the notion that there are other governmental supports for anything medically related. Whilst this may be the case for many medical conditions, faecal incontinence following a diagnosis of Hirschsprung's disease or imperforate anus, fails to attract support in any arena.

Building a culture that supports parents and peer to peer support groups.

Support groups for rare conditions need to move beyond relying on the key individuals who often generate the foundational energy for these groups. Supporting one's own child with high medical needs can be challenging enough, let alone being available to respond to the needs of other parents, no matter how worthwhile that endeavour is. Peer to peer support groups that are founded on the hard work of individuals in the community may not survive the departure of that individual.

There is a need to bring peer to peer support groups into the heart of the medical and educational institutions, while maintaining their independence. One option may be to have an umbrella organisation oversighted by government, which

resources peer to peer support groups for agreed chronic or high needs conditions. Care would need to be taken that this did not amount to a "popularity contest" with the most socially acceptable or prominent conditions being foregrounded much like occurs currently in donations from wealthy organisations at the end of the financial year, the conditions that are publicly appealing gain the support whilst those conditions that are stigmatised, fall short. Conditions that run the risk of being stigmatised should be prioritised in such a model. This might include conditions that result in faecal incontinence, or Tourette's syndrome, or some skin conditions such as ichythosis.

On a practical note, paediatric surgery as a discipline can summon partnering opportunities with parents and other clinicians, especially paediatric gastroenterologists who are unlikely to be called upon despite one of their roles being bowel management. This should include regular sessions including parent experiences in conferences relevant to Hirschsprung's disease and anorectal malformations, and more regular engagement with parents and adults with these conditions in surgical journals, as currently happens in the British Medical Journal and Lancet. All paediatric surgeon consultation rooms and bowel/continence clinics should have material on peer to peer support groups, which should be given to all families in hospital at the time of the baby's diagnosis, with contact details for parents.

With regard to schools, there is a need for a national set of guidelines for managing Hirschsprung's disease or anorectal malformation in schools, which should be co-produced with representatives from education, paediatric surgery, paediatrics and parents involved in peer to peer support groups. These could

then be used to frame schools' responses to individual children's needs. Schools which have managed to provide good support for children with Hirschsprung's disease or anorectal malformation should be recognised and supported to mentor other schools in a school support network for complex and stigmatising conditions. This is analogous to the national process used to develop the Asthma Friendly Schools policy.

Future research

This research has been exploratory and has demonstrated areas for further research. Hirschsprung's disease and anorectal malformations are distributed throughout the world. There is a need to conduct further research on peer to peer support groups for culturally and linguistically diverse Australians, and whether there are needs of or learnings from these populations. There is a huge gap in peer to peer support groups for Hirschsprung's disease or anorectal malformation in developing countries, and support groups in richer nations could consider seeking funding to partner with and mentor support groups in lower and middle income countries.

Future researchers may wish to explore more definitively the outcomes in terms of satisfaction and self-efficacy of models of care for families with newly diagnosed Hirschsprung's disease or anorectal malformation which are integrated into care, or which have a more distant advisory role. The surgical fraternity should partner with parents to identify the patient-centred outcomes of care that should be prioritised in their evaluations of surgical outcomes. There is an urgent need for greater research to bring about change in schools and transition of care to adult care for these children who require ongoing support and medical and

surgical intervention but have outgrown the accepted age to remain within the supportive paediatric space they have known since birth.

This thesis has shown the lives of these children matter and we as citizens and researchers need to do better in the way we support them once the surgical intervention is over. Whilst the medical fraternity often rejoice in a new surgical technique that improves recovery time and reduces scaring for children, the dilemma often comes later as there is no guarantee that a new technique will improve the long term outcome or quality of life for these children if faecal incontinence persists. This is when parents may hear the feared comment *'just because we can, should we?'* meaning, just because we are able to save this child's life by performing surgery, have we done them any favours if they now face the developing years with faecal incontinence and multiple hospitalisations in search of meeting societal expectations. The answer ought to be, yes we most certainly should, and we need to look at what is needed to support these children and their families who suffer as a consequence, otherwise we have not progressed from Nazi ideologies of Eugenics when people with disabilities were hidden from society in institutions or worse, by euthanising them. My significant original contribution to knowledge throughout this thesis is the ability to understand the complex and different perspectives of the parent, nurse, educator and the organiser of a peer to peer support group bringing people together whose children share the socially isolating condition of faecal incontinence past the age of societal expectations.

Children with Hirschsprung's disease or anorectal malformation are remarkably resilient in their commitment to living openly and well with a difficult condition.

The support of surgeons, gastroenterologists, physicians, nurses and educators is critical to them being able to live as full a life as possible. This thesis has argued that peer to peer support groups for parents and carers will extend that value to patients and their families across all the domains of the child's life.

This research lays the groundwork for an entire research field currently not explored, including public health and sociology. The medical model alone is insufficient to support these children and their families through to adulthood. Faecal incontinence associated with Hirschsprung's disease and imperforate anus are hidden disabilities heavily stigmatised in society. This research has explored and uncovered the difficulties children and their families face when born with a congenital bowel disorder, but this is just the beginning, this research demonstrates the vast gaps in the literature and understanding of clinicians and educators in how important it is to understand and address the difficulties these children face every day through no fault of their own. Children and their families find themselves ostracised, excluded and shunned in society. There is an urgent need to understand what can be done to support these children so they have the opportunity to develop friendships, enjoy the things children ought to such as play dates, swimming, school camps, parties and friend bases without fear of being shunned because they have uncontrolled soiling. The investigation and analysis throughout this thesis clearly demonstrates the urgent need for collaboration between clinicians, families, the children themselves and the institutions they frequent such as schools and hospitals. The children – their lives, their stories and their experiences - are key in understanding the difficulties they face when venturing away from home in the normal progression of life. A poster developed by children affected by these conditions during one of the BGK

conferences is appended to this thesis and clearly demonstrates even as parents, we cannot possibly completely understand how they are feeling and what we as parents can do to help them. Above all, the children and their families must be given a voice to have input in how we as a society, as clinicians, as educators help navigate these invisible disabilities to best support them to enjoy life in a community, the very foundation of our lives.

We cannot all live an ideal existence. Life is not a fairy tale. To summon Emily Perl Kingsley's poem once more: whilst 'Holland has windmills it also has Tulips.' What are we – as researchers – to do to ensure that children and adults with invisible disabilities can live with decency, respect and understanding? The first stage must be to understand them, and create contexts where they can live, thrive and diversify beyond stigmas and beyond labels. Faecal incontinence requires care and consciousness, but also contexts to enable support and respect.

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APPENDICES

1 Long-term follow-up of paediatric anorectal anomalies and the role and efficacy of parent support groups for such conditions.

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ORIGINAL ARTICLE

Long-term follow-up of paediatric anorectal anomalies and the role and efficacy of parent support groups for such conditions

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Abstract
Background/Purpose There are a multitude of parent support groups for most life situations and medical conditions. The aim of this study was to discover defining characteristics and to evaluate the structure and effectiveness of parent support groups in paediatric anorectal anomalies.
Methods Over 200 non-profit organisations and parent support groups were sent questionnaires to determine their effectiveness. Many of these were excluded as they were deemed not relevant to the final study. A final 20 groups were included for review, questions were based around origins, activities, education and evaluation, organisational structure and their affiliations with the health care team.
Results Most groups had similar origins, usually an educated parent who had a child born with the condition and a zest for further knowledge and understanding and a desire to share this with others in a similar situation. Only 20 % received government funding; few had paid staff and half had good relations with the local medical and paramedical fraternity. The majority had a team of long-term enthusiastic volunteers who remained with the group to support others long after their own child was no longer a concern.
Conclusions Some support groups offer a valuable service to families alongside the medical and paramedical fraternity. These groups provide families with the day-to-day lived experiences through social media, networking and meetings. Some provide more formal educational conferences designed to bring families, the medical and paramedical fraternity together to learn valuable lessons from each other.

Keywords Anorectal malformations · Hirschsprung's disease · Support groups · Parent support groups

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Introduction

Historically, families lived closer together than today and provided support for each other through difficult times. Young families had the benefit of their parents and grandparents to learn from and to support them through various transitions in their lives. The ability to travel brought the opportunity to move away from home and experience life elsewhere; or to move for better employment opportunities or better educational facilities for their children. However, in recent years there seems to be a trend to remain or move closer to family for support and child-minding as both parents are more likely to work [1].

Support groups have a long history. In Australia, the Benevolent Society was founded 200 years ago, by citizens concerned for their fellows. By the early 20th century, the charitable concept had broadened to include the self-help of Alcoholics Anonymous, a peer support group (admittedly closely supported by the Salvation Army), and then in the mid-20th century to disease specific peer support groups like the Coeliac Society. Alongside those newer efforts, the Benevolent Society has flourished to support a variety of social welfare programs, including the Institution of District Nursing in Australia [2, 3].

Today, these various kinds of non-profit organisations coexist in many different forms: some professionally led, others not; some relying heavily on the internet, and others not [4–6]. Often groups include many of these systems within their own model to provide a variety of support allowing members to choose their preferred method of contact.

Materials and methods

Qualitative data were collected through SurveyMonkey questionnaires sent via email to a wide mix of over 200 non-profit organisations and parent support groups. Many of these were excluded as they were deemed not relevant to the final study. A final 26 groups were included for review.

Questions were related to how the organisation was structured, when it began, who formed the group, whether the organisation was staffed by volunteers or paid workers, what training was offered to volunteers and if they followed a Code of Ethics. We also looked at how the organisation was funded, for example, Government funding, membership fees, donations, fundraising activities or corporate sponsorships. Questions surrounding the demographics of the organisation were asked to determine the geographical area serviced, age of members and conditions covered. We wanted to find out how long members remained in the organisation, was it long-term for their own benefit or short-term whilst they gained information and understanding of the condition then moved on. Importantly, we wanted to know how members found support groups when they were in need: was it through hospital or health professional referrals, word of mouth or online searching via a search engine? We asked questions around what services the organisations offered to members, health professionals and schools or pre-schools. We were interested to know if the organisations ran any campaigns to change Government policy related to the conditions covered by their organisation or if they conducted any research or had any way of evaluating the effectiveness of their services (Table 1).

Results

Organisational structure

The majority had Governing bodies such as a Board of Directors, Board of Trustees and Executive Committee or were self-governed. 77 % of respondents were incorporated organisations, which protects the individuals from prosecution. 84 % of respondents held AGMs, the vast majority of organisations were run purely by volunteers.

Table 1 Organisational structure

How groups are managed	Percentage of groups
Board of directors	8
Incorporated	77
Deductible gift recipient	85
AGM	84
Paid staff	58
Volunteers	92
Medical Advisory Board	77
Joining fees	16
Membership fees	65

Table 2 Funding

Groups are funded by	Percentage of groups funded
Government funding	45
Corporate sponsorship	23-regular; 42-sometimes; 35-never
Donations	76-members
Third party such as Everyday Hero	24
Annual fundraising events	20
Raffles/lotteries	20
Selling merchandise	4
Dinner/trivia nights	7
Sausage sizzles	7
Events cycling	4
Philanthropic trusts	4

Only one organisation did not have deductible gift recipient status (DGR) which allows them to receive tax deductible gifts (Table 2).

Funding

35 % never received any form of sponsorship with 65 % receiving this form of funding, regularly or occasionally. 45 % of organisations received government funding, while the remaining organisations received none. 17 organisations charged membership fees and only 4 charged joining fees. 76 % of organisations were heavily reliant on donations from members; an increasing trend is through a third party such as Everyday Hero; some via the general public and one through Philanthropic trust grants (Table 3).

Formation of the organisations

The length of time the groups had been in operation ranged from 1967 to 2005. The vast majority of founding members were parents, some groups had the support of interested clinicians, doctors, nurses and allied health professionals.

Table 3 Formation of the organisations

Foundation of the groups	Percentage of groups
Year groups were formed	1967–2005
Founded by parents	85
Founded member health professional	Nurse-36; doctors-57; allied-14
Professional members	0–4,500
Members affected by the condition or their carers	27–12,000
Age demographic paediatric	4
Age demographic adults	4
Age demographic both paediatric and adult	92

Table 4 Geographical area covered

Geographical area covered	State	National	Australasia	Worldwide
	20	44	8	28

Most provided support for paediatrics through adult services. Health professional members ranged from none to 4,500, those members affected by the condition ranged from 27 to 7,500 (Table 4).

Geographical area covered

44 % provided support nationally around Australia, while 28 % offered worldwide coverage (Table 5).

Services offered by the organisations

Nine organisations surveyed provide printed newsletters, while eight produced online versions. A small proportion organised sibling support, camps and formal dinners, while 12 organisations provided local meetings, 13 ran conferences. 18 organisations provided education for health professionals, whilst 8 provided information brochures about the conditions covered. Only eight were able to provide in-services to pre-schools or schools, whilst six organisations visited families in hospital or their homes. 17 respondents provided training for volunteers, whilst 20 organisations had a Code of Ethics for volunteers to follow. Increasingly social media such as Facebook, forums, chat rooms, email groups and twitter are being utilised as a means of gaining quick responses to queries from members who have previous experience or just to share thoughts with someone who understands. Physical meetings were less frequent, either due to distance, time restraints or the embarrassment of attending a group with a personal issue such as incontinence (Table 6).

Table 5 Services offered by the organisations

Groups offer the following services	Percentage of services
Volunteer training	60
Code of ethics for staff to follow	84
Conference	54
Local meetings	50
Christmas party	38
Picnics	42
Formal dinners	8
Organised camps	21
Respite for families	4
Printed newsletters	38
Online newsletters	33
Sibling support	13
Educational material	35
Email and/or phone support	27
Chat rooms/discussion groups	17
Forum	57
Facebook page	78
Education for health professionals	65
Education for pre-school/schools	30
Visit families at home or in hospital	26-yes; 35-on request only

Table 6 How members hear about the organisation

Members hear about the organisation by	Percentage of groups gain members by
Via an online search engine for the condition	79
Referrals from health professionals	83
Advertising in hospitals with brochures or posters	12
Advertising in doctors rooms with brochures or posters	8
Word of mouth	71
Advertising in baby health clinics	4
Health professionals stock brochures	4
Phone and membership forms	11
Information service such as a 1800 phone number	4

How members hear about the organisation

The majority of requests about the organisations come via an online search by the parents or carers or interested family member or from the treating clinician. 74 % came via Word of Mouth, a small proportion came from advertising in hospitals, doctor's waiting rooms or baby health clinics (Table 7).

Table 7 Membership use of the organisation

How long do members remain in the organisation	Percentage who remain
Short-term whilst they need information and support	14
Long-term for their own needs	55
Long-term to support new members	5
A mix of both short- and long-term	26

Table 8 What factors determine who joins a support group?

People join a support group because	Percentage
Members have a more severe form of the condition than non-members	52
Education and information	74
Lack of family support	17
Connect with others in a similar situation	83
Online forums/discussion boards	48
To learn from more experienced members	70
Attendance at conferences and informal meetings	26
Financial support	0
Access to equipment or supplies	4

Membership use of the organisation

Most members of the support organisations reviewed remain long-term for their own needs, a few to support others whilst a few only remain short-term, the rest had a mix of short- and long-term member (Table 8).

What factors determine who joins a support group?

The main factor that may determine who joins a support group was overwhelmingly cited as a desire to connect with others in a similar situation. Half reported members had a more severe form of the condition than non-members, a high proportion sought education and information or to learn from others who are more experienced at living with the condition and only six felt members joined to attend conferences and informal meetings. Only four organisations cited a lack of family support as a reason for people who joined their group.

Involvement in campaigns

Six organisations were actively working on or had been successful in running a campaign to change services offered through government agencies (Table 9).

Table 9 Research functions

Groups involved in research	Percentage
Conducts its own research	47
Considers requests for contribution to research projects	70
Has not contributed to any research projects to date	12
Recruit people for clinical trials	4
Evaluation of own programs	100

Table 10 Professional affiliations of support groups

Affiliations of patient groups with	Percentage of groups with affiliation
Doctors	80
Hospitals	80
Clinics	35
Nurses	50

Research functions

The majority of respondents contributed to research requests, whilst almost half conducted their own research. Organisations were asked how they evaluate the effectiveness of their own programs, 77 % had feedback evaluations after events, 55 % surveyed their members directly, 45 % evaluated at committee meetings and 23 % gained feedback through various forms of communication with members such as email, forums, social media and phone support. Only one organisation had a method of comparing members and non-members to determine if being a member of a support organisation improved their quality of life (Table 10).

Professional affiliations of support groups

Affiliations with hospitals, doctors, nurses and clinics were encouraging. Some additional comments offered by the organisations have been summarised as follows: support groups are left open for people to participate without obligation. Different experiences are represented to ensure that all aspects of the disease are represented. Some groups try and match people with similar circumstances together to share experiences.

Discussion

We wished to survey the composition and characteristics of patient peer support organisations, with a view to analysing how well they matched both patient and practitioner expectations. The study includes organisations whose

primary aim is to support families who have a child born with some form of congenital malformation. We included mostly gastrointestinal anomalies such as Hirschsprung's disease, anorectal malformations, cloacal extrophy, short gut syndrome, paediatric gastro-oesophageal reflux, coeliac disease, gastroparesis, chronic intestinal pseudo-obstruction, and colonic inertia, along with a few non gastrointestinal surgical conditions of childhood: (tracheaoesophageal fistula, spina bifida, hydrocephalus, cleft lip and palate).

Groups surveyed were founded between 1967 and 2005 by parents of affected children and most of those had the support and collaboration of an interested clinician who specialised in the particular condition. The number of members was broken down into professional members whose numbers ranged from none to 4,500 and members ranged from 27 to 12,000; (members represented those who were either affected by the condition the organisation supported, or were the affected person's parents/carers interested family members). All but one organisation had DGR, an important encouragement to donors. The majority of organisations included in this study were formally set up with governing bodies and had structured committees consisting of a president, secretary, treasurer and various committee members. State and territory legislation for incorporation provide a simple and more affordable means of creating a separate legal entity for small, community-based groups with limited resources. An incorporated association is also a legal entity separate from its individual members and can hold property, sue and be sued [7].

A major issue for most organisations is the lack of funding from government or sponsorships. Most organisations gained funding through modest membership fees or donations, though some organised fundraising activities within the group. A growing trend is using third party fundraisers such as Everyday Hero, to increase the funding options. We are unable to say if it has increased the net amount of funding actually received. Only one organisation received funds through a philanthropic trust.

Most organisations provided training for their volunteers and followed a Code of Ethics. A small number of organisations were actively working on campaigns to bring about change at a governmental level, while 70 % of organisations contributed to research requests from outside sources. Half of respondents conducted their own research, whereas feedback about their own organisation and its programs was gained at the conclusion of events. It was clear that the groups relied heavily on the founding members and volunteers who gave their time, wealth of knowledge and expertise freely.

Members had a tendency to remain in the organisations long-term to continue gaining support and a small number remained to support others coming along with newly diagnosed children. The overwhelming reason cited for joining support groups was to connect with others in a similar situation and to learn ways of managing the condition day to day, consistent with the findings of Law et al. [8]. New members are able to gain insight from existing members, even though each person's situation might be slightly different, there is a wealth of experience to be gained from sharing the lived experience of a particular illness [9]. Organisations generally accumulate educational information which is both relevant to the condition and provides valuable advice for others who may be involved in a child's care such as family members, pre-school and schools. One-third of organisers felt members had a more severe form of the condition than non-members. Interestingly, only one organisation felt members joined due to a lack of family support.

Services offered by the support groups ranged from printed or online newsletters, local meetings and conferences whilst over half provided education for health professionals, one-third provided information brochures for schools and pre-schools to assist the staff to better understand the child's condition. To a smaller degree, some organisations were able to offer in-services to schools or visit parents in hospital or the home.

Although good affiliation with healthcare teams in hospitals, private practises and clinics was claimed by most respondents, in reality most groups found promotion of their organisation very difficult. This was particularly so for conditions relating to the bowel as they do not attract community support nor are they considered newsworthy by the media. The relationship between support organisations and clinicians is of vital importance to provide a well-rounded balanced support for patients and their families. Respect for each other's knowledge is an important factor in the relationship; support groups can provide insight to parents from the unique perspective of the day-to-day lived experience. Healthcare providers on the other hand have learnt about the condition through training and experience and can provide diagnosis and management of the condition. It is this shared wealth of knowledge that can be of great support to the formal health system and benefit all parties [10]. Time is another important factor as clinicians who are time-poor can focus on the medical management of the condition, whilst parents can gain information about reputable support groups who can provide practical information [11].

The internet has alleviated some of the difficulties parents have in finding appropriate support when they most need it, but information on the web can be bewildering and come from a variety of sources [12]. Most support groups have websites with information for new parents that at least comes from a known and (hopefully) trusted provider. The ability to join the group online and find information and

support as soon as a diagnosis is made is a useful and very modern role for the peer support group. It has enabled parents to connect with others, diminishing isolation for conditions that may be poorly understood by relatives and friends and which do not attract community support or media attention. Increasingly popular is the advent of social media sites which have enabled parents and older children to ask questions and gain responses almost immediately to a current issue and be able to discuss issues with someone who understands the situation with a degree of anonymity [13–15].

The non-profit organisations provide a valuable resource of accumulated knowledge and education on dealing with the day-to-day issues encountered with specific conditions. Transition periods experienced by children were peak times for parents to seek help from others whose children were older. Typical times of need were identified as when the primary caregiver returned to work requiring the child to be placed in some form of child care; transition from pre-school to school, infants to primary and onto high school. Shipper reported in a presentation at the International Workshop ARM in Nijmegen that the transition from the multidisciplinary approach in paediatric care to the individualised specialist adult system was a major problem for children born with an anorectal malformation. This same issue has been reported in other areas where paediatrics patients experience difficulty transitioning to adult care services [16, 17].

It is at these times that we see the peer support organisations having a particularly important role, the children and young adults themselves and the parents who care for them on a daily basis have great insight into their everyday needs and can help determine what aspects of care they require. Their wealth of experience is also valuable in preventing a reinvention of the wheel and it also gives people a sense of giving back to the community by sharing their experiences with parents of newly diagnosed children [18].

Acknowledgments We thank the organisations involved in this research for their insight into the services they provide.

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2 Aligning services between clinicians, paediatric hospitals and support groups for children born with anorectal anomalies

Australian National University

Aligning services between clinicians, paediatric hospitals and support groups for children born with anorectal anomalies.

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September 2014

Introduction

Clinicians provide excellent medical and surgical care for children born with anorectal anomalies such as Hirschsprung's disease (HSCR) and anorectal malformations (ARM), but parents struggle with the day-to-day lived experiences that may be more appropriately served by parent support groups.

The aim of this study was to determine if a more collaborated approach between paediatric hospitals, clinicians and support groups would provide a better service to families.

Materials and Methods

Questionnaires were sent to 45 Paediatric surgeons in Australia, New Zealand and Europe to determine long term follow up of patients and their use and opinions of support groups.

Six support groups specific to anorectal anomalies in the same regions were surveyed to determine their affiliations with health care teams.

10 paediatric hospitals in Australia were polled to determine the inclusion of support groups as an adjunct to existing services.

Results of all three were compared.

Results

Paediatric Surgeons:

The majority follow-up patients until they transition to adult services.

Most were in favour of support groups, but noted the following barriers:

- Lack of contact with support groups
- Preferred a hospital based service
- Time constraints
- Unaware of how to find groups
- Hard to evaluate their services.

Support Groups:

- Generally formed by parents
- Some supported by clinicians
- Services varied
- Funding was problematic
- Disseminating information to parents, clinicians and hospitals proved difficult.

Paediatric Hospitals:

- Majority did not have a framework for the inclusion of support groups.
- Two provided links via website
- One had endorsed a support group through meetings and a MoU.
- One hosptial undergoing redevelopment has included a formal process for assessment and inclusion of support groups.

Discussion

Whilst the majority of clinicians saw the value in support groups, most found it difficult to determine the quality of such groups or lacked the time to research them.

Whilst providing a valuable service to families, support groups lacked the resources and/or knowledge on how to connect with clinicians and hospitals.

There was an inconsistent approach in hospitals to the inclusion of support group information; some provided links on their website to the support groups and others had information pamphlets displayed.

Conclusion

The authors feel the time is right for a more formal approach towards the inclusion of support groups as an adjunct to the services currently provided by clinicians and paediatric hospitals.

The aim of all parties is to improve the quality of life for children born with anorectal anomalies.

By bringing these services together, we can work towards achieving that aim by providing a full range of services within our paediatric hospitals.

PatCH

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3 The prevalence of Hirschsprung disease (HSCR) associated with Down syndrome (DS)

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The prevalence of Hirschsprung disease (HSCR) associated with Down syndrome (DS)

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Introduction

The introduction and widespread adoption of antenatal ultrasound screening in the last decades of the 20th century are rightly seen as a great boon for obstetric care. One of the consequences of that screening was that many congenital problems were able to be diagnosed in utero. The changes to abortion law in the preceding decades then made abortion of children with antenatal problems seem like a viable option for many. The actual effect these changes in attitude and technology is having is unclear. Hirschsprung disease is a condition that is not to our knowledge ever diagnosed antenatally on ultrasound. Down syndrome on the other hand can be picked up by a variety of methods. We asked whether there would be a noticeable drop in the proportion of Down Syndrome / Hirschsprung to total Hirschsprung cases during the last 50 years which encompasses those social and technical changes.

Materials and Methods

Statistics on the incidence of Hirschsprung's disease and its various associated conditions at New South Wales and ACT children's hospitals were collected from 1960 onwards. These were reviewed for the years 1960 to 2014. It should be noted that these figures do not include children who presented to non-specialist hospitals, and who were then NOT referred to specialist units at Sydney Children's Hospital, Randwick, RAHC, Camperdown, Children's Hospital Westmead, Newcastle or Canberra. Information is likely to be incomplete before 1975, as small numbers from Newcastle and Canberra will be missed, and not all Sydney records from that time were retrieved.

The annual incidence of Hirschsprung's disease and Hirschsprung's disease with Down syndrome was calculated, and the percentage of Hirschsprung's disease with Down syndrome was then also calculated.

Data collection for this study was approved by the relevant institutional ethics committees, 1995 and 2015.

Results

There were altogether 921 Hirschsprung disease children born in NSW and the ACT from 1960 to the end of 2014. Out of this group, 72 had Down syndrome. 365 children were born with Hirschsprung's disease in the first 20 years (20 with Down syndrome), and 468 in the last 20 years of the time period (40 with Down syndrome). There is no significant difference in the proportions of Down syndrome in the first 20 years compared with the last 20 years ($p = 0.1050$ Fisher exact test).

During the 55 years covered, the population of New South Wales went from approximately 3.9 million to 7.5 million, accounting for the increasing total numbers.
Down syndrome proportional birth rates are given in the table.

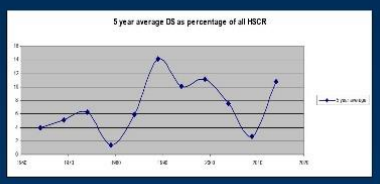

Figure 1. No evidence for a decrease in DS/HSCR proportion over 55 years, and maybe even a slight increase.

Discussion

There is no indication in this data that significant numbers of Down syndrome children are being aborted. It may be that our annual incidence of Down syndrome/Hirschsprung disease is simply too small a sample to be indicative, although with a total sample size of 72 this seems unlikely. 40 to 50% of Down syndrome children are born with congenital heart disease, and some of these also have Hirschsprung disease. Even without accompanying cyanotic heart disease, a newborn Down syndrome child with a congenital bowel obstruction in 1960 would have been a challenging management problem. It is possible that not all Down syndrome children in the first decades of the study were referred for active treatment to the specialist centres. If this is true, then postnatal loss early on compensates for antenatal loss in the second half of the study. It may also be true that increasing numbers of older mothers more recently are compensating with increased total numbers of Down syndrome conceptuses. Demographic studies considering the genetic contribution of trisomy 21 to the aetiology of Down syndrome need to consider such confounding factors. Long-term follow-up studies of children with major disabilities need also to take into account changing attitudes to conception and the value of the life of the child.

Conclusion

This study found no indication that the relative incidence of Down syndrome with Hirschsprung disease was declining over the decades. If anything the incidence is slightly higher now than 50 years ago. Antenatal diagnosis seems to have had no effect on the incidence of this particular significant combined condition.

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4 Role of community peer support groups in management of patients with anorectal anomalies from the paediatric surgeon's viewpoint

Australian National University

Role of community peer support groups in management of patients with anorectal anomalies from the paediatric surgeon's viewpoint

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October 2013

Introduction

Community support groups for patients are increasingly popular.

The aim of this study was to discover paediatric surgeons' opinions on parent support groups and their approach to managing anorectal anomalies such as Hirschsprung's disease (HSCR) and anorectal malformations (ARM).

Materials and Methods

Paediatric surgeons were surveyed through a questionnaire in Australia, New Zealand and Europe.

45 respondents with slightly more in Australia than elsewhere.

We asked about:

- Demographics
- Management of anorectal anomalies
- Did they engage others in patient care
- Opinions of support groups
- Their concept of an ideal support group.

Results

Length of specialist practice ranged from 5 to more than 20 years.

The majority saw up to 5 new patents annually of both HSCR and ARM and followed up patients until transition to adult services.

50% engaged the services of other health professionals.

40 out of 45 were in favour of support groups.

Barriers to referring people to support groups were reported as:

- No contact with support groups
- Parents did not ask
- Prefer a hospital based service
- Concern groups had the potential to raise unnecessary fears.
- Time constraints
- Unaware of how to find a good support group.
- How they are run, who runs them
- Do they train volunteers/staff and follow a code of ethics

Discussion

Most were generally in favour of support groups, however, we are mindful this may indicate that only those positive about support groups responded.

Support groups can provide the day to day lived experiences and support for parents.

Support groups could be particularly helpful in anorectal anomalies due to the low incidence and therefore small experience of individual surgeons who have limited time and resources.

Conclusion

We found many surgeons lacked information or contact details on support groups and did not have independent coordinators of paramedical services for colorectal conditions.

Some surgeons engage well with support groups providing opportunities to learn from each other.

Aligning services between clinicians, families and support groups may be a first step in support groups becoming a more inclusive part of health care.

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5 Do parent support groups have a role in anorectal anomalies?

Australian National University

Do parent support groups have a role in anorectal anomalies?

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October 2013

Introduction

The aim of this study was to analyse groups for paediatric anorectal anomalies.

With significantly improved mortality and morbidity for children born with anorectal anomalies, there has been a shift in focus to functional outcome with the aim of improving the quality of life for these children,

Materials and Methods

Questionnaires were sent to support groups to determine their structure and effectiveness. Of 28 respondents, we selected 9 pertaining to bowel related conditions of which six were specific to anorectal anomalies.

We asked about:

- The groups origins
- Organizational structure
- If they had paid staff or volunteers.
- What services they offered members
- Methods of delivery and evaluation.
- Importantly, were they affiliated with the relevant healthcare teams.

Results were compared to our previous survey of paediatric surgeons to assess both viewpoints.

Results

Most support groups had similar origins, usually an educated parent who had a child born with the condition, some with the support of a clinician.

Funding was an issue and most had a team of long-term enthusiastic volunteers who remained with the group to support others long after their own child was no longer a concern.

Disseminating the group's information to those who need it most was difficult.

Services such as social media sites and online forums provide quick access to daily living experiences.

More formal support was provided through newsletters, educational material and conferences which had the potential to bring families, the medical and paramedical fraternity together.

Discussion

The process of finding a reputable support group was difficult with no central point of contact. This lack of coordinated services was considered laborious and far from optimal.

The improvement in morbidity and mortality rates for babies born with anorectal anomalies today has paved the way for parents and clinicians to work more closely together to improve the quality of life for these children.

Whilst there are very few support groups whose primary focus is anorectal anomalies, those surveyed are working hard to support families and clinicians to this desirable outcome.

Conclusion

The majority of support groups offer a valuable service to families alongside the medical and paramedical fraternity.

The authors feel the time is right for a more collaborated approach to include optional support services to families who have a baby born with an anorectal anomaly.

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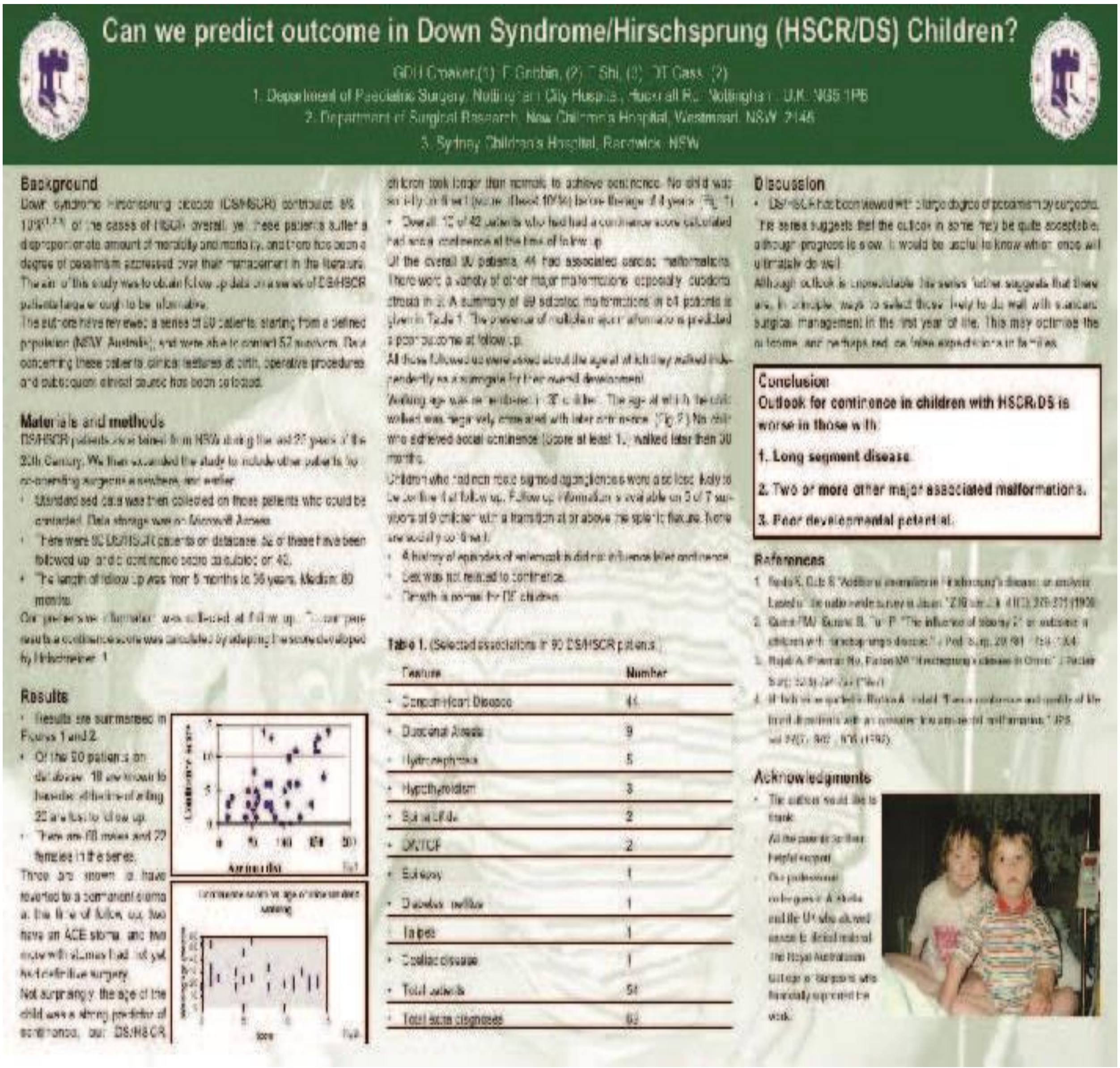
6 Can we predict outcome in Down Syndrome/Hirschsprung (HSCR/DS) Children?



7 Bowel Group for Kids Support Group Information

HERE'S WHAT MEMBERS SAY ABOUT BOWEL GROUP FOR KIDS:

"When our daughter was born, we were devastated and overwhelmed! There was so much to take in, a condition we'd never heard of, and being in the NICU was scary and isolating. Having the BGK there to talk to was so calming."

"My husband was born with Hirschsprung's disease and when our baby was diagnosed, we thought we were the only ones. It was such a relief to talk to someone who also had a parent and child born with the condition."

"I found the pamphlets so helpful. It saved me repeating myself to each of my son's carers and now his school. The information is clear, simple to understand and helps those caring for him understand his needs better."

"Thank you for being there from the beginning"

MEMBERSHIP

Membership is open to families, health practitioners and others with an interest in HSCR/ARM. We provide:

- Extensive educational resources specific to the needs of children born with HSCR or ARM
- Access to National, short educational seminars, video linked to other states and NZ, annual events and social activities
- Access to a network of families experienced at caring for children with HSCR/ARM
- Information packs for schools

ONLINE SUPPORT

Members also have access to our online support:

- Practical articles to support children born with these conditions.
- Our private Facebook group for parents/carers and clinicians
- Our private Facebook site for young people with these conditions
- Living with Hirschsprung's Disease handbook
- Latest medical research
- Newsletters

JOIN US NOW

Annual Membership $30.

Contact us or take a look around the website

www.bgk.org.au
enquiries@bgk.org.au

Bowel Group for Kids is a registered charity. Donations over $2 are tax deductible.

Bowel Group for Kids

ABOUT BOWEL GROUP FOR KIDS

Caring for children born with Hirschsprung's disease (HSCR), imperforate anus or anorectal malformation (ARM) since 1994

www.bgk.org.au
enquiries@bgk.org.au

Bowel Group for Kids Support Group Information (Cont)

EDUCATION, PRACTICAL INFORMATION AND SUPPORT

Bowel Group for Kids is the only national organisation dedicated to providing education, practical information and support to families of children born with Hirschsprung's disease (HSCR), anorectal malformation (ARM) and associated conditions.

These rare congenital conditions affect one in every 5000 babies born and children are often diagnosed in confronting and distressing circumstances. Families must come to grips with a condition they may never have heard of before at the same time as dealing with complicated surgery, hospital stays and uncertainty about the future.

Bowel Group for Kids exists to support your family through this challenging time.

We provide a space to connect with others going through the same thing and offer practical advice on the day-to-day challenges of living with these conditions at every stage of life.

Whilst your doctors provide the medical advice, Bowel Group for Kids provides advice on the day to day, practical living experience.

NB: All BGK information and resources are neither intended not implied to be a substitute for professional medical advice

Connect with us online anytime!

OUR PUBLICATIONS

Our publications provide practical information to assist with the practical care for children with HSCR/ARM. Titles include:

- Hirschsprung's disease: A Guide for Schools
- Anorectal Malformation: A Guide for Schools
- Stomas: A Guide for Schools
- Basic Stoma Care
- Antegrade Colonic Enema Information
- Toilet training after pull-through surgery
- How to help your child in hospital
- Coping with a hidden disability in a mainstream school
- Anaesthesia and your child

And many more available to members.

Bowel Group for Kids

JOIN BOWEL GROUP FOR KIDS

Annual Membership $30

OUR WEBSITE

Visit the Bowel Group for Kids website for access to educational resources for people living with HSCR and ARM as well as up-to-date research. Members also have access to educational seminars and information packs for schools.

www.bgk.org.au
enquiries@bgk.org.au

A MESSAGE FROM OUR PATRON DUNCAN ARMSTRONG

I have never felt that my Hirschsprung's disease has ever held me back - I was even able to achieve my dream of wearing the green and gold at the Seoul Olympics in 1988 and break a swimming world record!

Now as parents of a child with Hirschsprung's disease, my wife Rebecca and I share in that tremendous feeling of strength and support from the community at Bowel Group for Kids.

I'd like to encourage all of you to keep searching for the right 'life rhythm' that will empower your lives. Don't be afraid to connect with doctors, hospitals, other parents and through the BGK.

The more information you can obtain, the better the decisions you will make for your family.

Success is a process – you learn how to achieve and then make it happen,

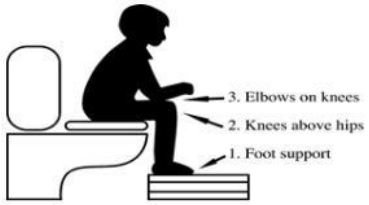
Warmest regards,
Duncan Armstrong

8 Anorectal Malformation: A Guide for Schools

CHILDREN WITH ARM NEED THE FOLLOWING SUPPORT

- **Fluids**: One of the functions of the large bowel is water absorption. Children who have had parts of their bowel removed need to maintain better than average fluid intake. It is critical to allow them to have a water bottle on their desk and encourage drinking throughout the day.
- **Smearing and soiling**: Incontinence and constipation are common in children with HSCR and they may not be aware when soiling happens. This may happen because of the condition itself or the effects of surgery. It may also be because their bowel is never completely empty and is constantly sending the message that they need to go. As a result, the children have become immune to the feeling of needing to go.
- **Smell**: Children who continually soil may become immune to the smell. They need support and understanding and ways to discreetly communicate the problem.

'Constant exposure to the same message tunes down the message'



Correct posture for using the toilet

TAKE HOME MESSAGES FOR THE SCHOOL

- Understand the condition
- Be prepared and patient
- Establish a routine to meet the child's needs
- Never deny access to toilets or drinking facilities
- Work with the family to establish strategies

OTHER PUBLICATIONS FROM BOWEL GROUP FOR KIDS

- About Bowel Group for Kids
- Hirschsprung's Disease: A Guide for Schools
- Stomas: A Guide for Schools
- Toilet Training After Pull-Through Surgery
- Antegrade Colonic Enema Information

GLOSSARY OF TERMS

Bowel: consists of small and large intestines, sometimes referred to as 'colon' 'gut' or 'gastrointestinal tract'
Congenital: present at birth
Defaecation: the opening of bowels (poo; motions; No. 2; big jobs etc)
Faeces: poo, stools, motions (and any other terms that may be used at home)
Ostomy: a surgically created 'stoma' in the abdominal wall for the removal of faeces
Stoma: Greek word meaning mouth or opening
Stool Retention: the retention of a soft stool due to poor muscle tone

The Bowel Group for Kids are here to help!

www.bgk.org.au
enquiries@bgk.org.au

Bowel Group for Kids

A GUIDE FOR SCHOOLS IN UNDERSTANDING ANORECTAL MALFORMATION

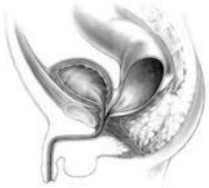
Information for schools and preschools about caring for children with an anorectal malformation (ARM)

www.bgk.org.au
enquiries@bgk.org.au

Anorectal Malformation: a Guide for Schools (cont)

WHAT IS ANORECTAL MALFORMATION?

Anorectal malformation (ARM) is a congenital abnormality where a baby is born with no anus meaning there is no exit for faeces. This is also known as imperforate anus. It affects 1 in every 5000 children born. There are many variants in boys and girls, from a membrane covering the anal opening, misplaced anal opening, to more serious types where a fistula develops between the urinary or reproductive system. There are a wide range of medical issues with varying complexities associated with imperforate anus known as VATER or VACTERL. Two examples of ARM:



Male with fistula

Female Cloaca

The effects will be different for each child. Babies born with imperforate anus require surgeries at birth. In some cases, they may also have a temporary stoma - an artificial opening that has been surgically created as an exit for waste that is fitted with a pouch to collect the waste matter. In severe cases, is still in place when the child starts school.

Imperforate anus may be also be accompanied by a collection of other anomalies known as **VACTERL/VATER.** These include:
Vertebral defects, **A**nal atresia, **C**ardiovascular defects, **T**rachea defects, **O**esophageal fistula/atresia, **R**enal defects, **L**imb anomalies.

WHAT DOES THIS MEAN FOR MY CHILD AT SCHOOL?

Due to the condition itself and the delicate corrective surgery they have had, children with this condition often experience delays in achieving faecal continence, whereby the child may have limited control of their bowel by school age. Children may have little or no sensation in their anal sphincter. They may experience soiling due to constipation, stool retention or loss/damage of the anal sphincter.

Coupled with this, children born with ARM have varying amounts of large bowel missing at birth. The greater degree of bowel that is lost, the more liquid the stools will be. It is important to understand a liquid stool does not indicate infectious diarrhoea, it may be a normal consequence of the degree of bowel lost.

WHAT DOES THE SCHOOL NEED TO DO TO PREPARE?

- Understand that families have been through many treatments, surgeries and management plans to establish their routine.
- Discussion should be around what the condition is and what it means for the child.
- Arrange a meeting with principal, class teacher and parents/carers to discuss what the condition means for the child and what toileting needs the child has. Include the child if appropriate.
- Further meetings need to be arranged just prior to the child starting school to clarify any changes to routine and reiterate the plans.
- Develop a management plan in collaboration with health practitioners and parents/carers
- Set up a communication book, bowel chart or other routine to ensure consistency between home and school.

WHAT CAN TEACHERS DO TO SUPPORT A CHILD WITH ARM?

- Support the child discreetly & confidentially.
- Always allow immediate access to toilet and drinking facilities - even if the child needs multiple attempts in a short period.
- Parents will provide a changing bag containing clothing, wipes and disposable bags.
- The child may not have the sensation to use their bowel de to the problems associated with the anal sphincter and may need to be on a toilet schedule.
- Use the same language around toileting as at home.
- Look out for gestures that indicate a child needs to toilet and establish a 'secret signal' to alert the child that they may need to go.
- Children with this condition may need several attempts to pass a bowel motion.
- Always allow access to facilities even if they need to go several times over a short period.
- Aim for the child to clean him or herself but understand they may need assistance to begin with to ensure good hygiene.
- Reinforce the importance of hand washing.
- Give praise when the child responds to prompts
- Ensure regular dialogue with parents and carers to evaluate progress.

Bowel Group for Kids

Caring for children born with Hirschsprung's disease (HSCR), imperforate anus or anorectal malformation (ARM) since 1994.

www.bgk.org.au

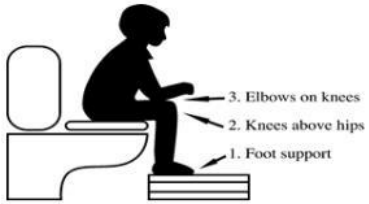
enquiries@bgk.org.au

9 Hirschsprung's disease: A Guide for Schools

CHILDREN WITH HSCR NEED THE FOLLOWING SUPPORT

- **Fluids**: One of the functions of the large bowel is water absorption. Children who have had parts of their bowel removed need to maintain better than average fluid intake. It is critical to allow them to have a water bottle on their desk and encourage drinking throughout the day.
- **Smearing and soiling**: Incontinence and constipation are common in children with HSCR and they may not be aware when soiling happens. This may happen because of the condition itself or the effects of surgery. It may also be because their bowel is never completely empty and is constantly sending the message that they need to go. As a result, the children have become immune to the feeling of needing to go.
- **Smell**: Children who continually soil may become immune to the smell. They need support and understanding and way to discreetly communicate the problem.

'Constant exposure to the same message tunes down the message'



Correct posture for using the toilet

TAKE HOME MESSAGES FOR THE SCHOOL

- Understand the condition
- Be patient
- Establish a routine to meet the child's needs.
- Never deny access to toilets or drinking facilities
- Work with the family to establish strategies

GLOSSARY OF TERMS

Bowel: consists of small and large intestines, sometimes referred to as 'colon' 'gut' or gastrointestinal tract'.
Congenital: present at birth.
Defaecation: the opening of bowels (poo; motions; No. 2; big jobs etc)
Faeces: poo, stools, motions (and any other terms that may be used at home).
Ganglion cells: These nerve cells are normally found in the bowel and are very important for the process of peristalsis.
Peristalsis: The wave like movement of the bowel which pushes food along the intestines towards the rectum to allow emptying.
Ostomy: a surgically created 'stoma' in the abdominal wall for the removal of faeces.
Stoma: Greek word meaning mouth or opening

OTHER PUBLICATIONS

- Bowel Group for Kids General Information
- Anorectal Malformation: A Guide for Schools
- Stomas: A Guide for Schools
- Toilet Training After Pull-Through Surgery
- Antegrade Colonic Enema Information

www.bgk.org.au
enquiries@bgk.org.au

Bowel Group for Kids

A GUIDE FOR SCHOOLS IN UNDERSTANDING HIRSCHSPRUNG'S DISEASE

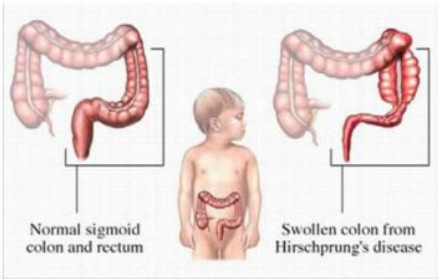
Information for schools and preschools about caring for children with Hirschsprung's disease

www.bgk.org.au
enquiries@bgk.org.au

Hirschsprung's disease: A Guide for Schools (Cont)

WHAT IS HIRSCHSPRUNG'S DISEASE?

Hirschsprung's disease (pronounced HURSH-sprungz) affects 1 in every 5000 children born.



It is a condition in which the ganglion cells (nerve supply) to the colon is absent or incomplete. These cells are essential for the bowel to move stool through to the rectum. Babies with the condition are often born with an intestinal obstruction requiring surgery. This usually involves removing the parts of the colon that do not work. The effects of Hirschsprung's disease (HSCR for short) will be different for each child.

WHAT DOES THIS MEAN FOR MY CHILD AT SCHOOL?

Children with Hirschsprung's disease may have delay achieving faecal continence, even at primary school age. In some cases, they may also have a temporary stoma – an artificial opening that has been surgically created as an exit for waste that is fitted with a pouch to collect the waste matter.

Most cases of childhood faecal soiling are due to constipation or stool retention. Liquid stool can leak around the more formed stool in the colon, so stools can range from hard to liquid, but it is important to understand this is not an infectious diarrhoea, rather a normal consequence of the condition.

WHAT DOES THE SCHOOL NEED TO DO TO PREPARE?

- Understand that families have been through many treatments, surgeries and management plans to establish their routine.
- Discussion should be around what the condition is and what it means for the child.
- Arrange a meeting with principal, class teacher and parents/carers to discuss what HSCR means for the child.
- Further meetings need to be arranged just prior to the child starting school to clarify any changes to routine and to reiterate plans.
- Develop a management plan in collaboration with health practitioners and parents/carers.
- Set up a communication book, bowel chart or other routine to ensure consistency between home and school.

WHAT CAN TEACHERS DO TO SUPPORT A CHILD WITH HSCR?

- Support the child discreetly & confidentially.
- Always allow immediate access to toilet and drinking facilities - even if the child needs multiple attempts in a short period.
- Use the same language around toileting as at home.
- Look out for gestures that indicate a child needs to toilet and establish a 'secret signal' to alert the child that they may need to go.
- Aim for the child to clean him or herself, but understand they may need assistance to begin with to ensure good hygiene.
- Reinforce the importance of hand washing.
- Give praise when the child responds to prompts.
- Ensure regular dialogue with parents and carers to evaluate progress.

Bowel Group for Kids

Caring for children born with Hirschsprung's disease (HSCR), imperforate anus or anorectal malformation (ARM) since 1994.

Contact us today!

www.bgk.org.au

enquiries@bgk.org.au

10 Stomas A Guide for Schools

CHANGING AN APPLIANCE

Modern stoma products are designed for ease of use and discretion. Most have gas filters to allow for odour-free passage of 'wind'. This also prevents the pouch from ballooning with air or detaching unexpectedly. There are two main types:

- A one-piece system where base plate (or wafer) and bag are in one piece and can be drained.
- A two-piece system has a base-plate that attaches to the abdomen and a pouch that attaches to this. The wafers can be left in place for a few days.
- A belt may be attached for additional bag security.

EQUIPMENT REQUIRED

Pouch changing is not a sterile procedure.

- Gloves
- A new ostomy pouch
- Odour neutralising nappy sacks
- Moist wipes
- Appropriate disposal bin
- Hand washing facilities
- Air freshener (optional)

DISPOSAL

The contents of the pouch can be emptied into the toilet before disposing of the pouch in a deodorised nappy sack. Never flush a used pouch down the toilet. Your healthcare provider can advise you regarding local policies for special disposal bins

POSSIBLE PROBLEMS

Skin excoriation: Redness, rashes or broken skin around the stoma or any pain around the stoma needs to be discussed with a parent/carer.

Bleeding from the stoma: Slight bleeding may occur from the surface of the stoma if it is rubbed during cleansing. This is not a problem and a light dusting of stoma powder will stop the bleeding. If the bleeding appears to be coming from inside the stoma, **seek immediate medical advice.**

TAKE HOME MESSAGES FOR THE SCHOOL

- Understand the condition
- Be patient and prepared
- Establish good strategies that meet the needs of the child
- Stomas can get wet and swimming is okay
- Sport is great but be careful with contact sports
- Stomas have no nerve endings so they can't cause pain
- Bleeding from inside the stoma requires urgent medical attention

OTHER PUBLICATIONS

- Bowel Group for Kids General Information
- Hirschsprung's Disease: A Guide For Schools
- Anorectal Malformation: A Guide for Schools
- Toilet Training After Pull-Through Surgery
- Antegrade Colonic Enema Information

GLOSSARY OF TERMS

Bowel: consists of small and large intestines, sometimes referred to as 'colon' 'gut' or gastrointestinal tract'.
Congenital: present at birth.
Continence nurse: Nurse who advises on bowel and bladder issues.
Ostomy: a surgically created 'stoma' in the abdominal wall for the removal of faeces.
Stoma: Greek word meaning mouth or opening

Bowel Group for Kids

A GUIDE FOR SCHOOLS IN UNDERSTANDING CHILDREN WITH STOMAS

Information for schools and preschools about caring for children with a stoma

www.bgk.org.au
enquiries@bgk.org.au

Stomas A Guide for Schools (Cont)

WHAT IS A STOMA?

A stoma is an artificial opening in the abdomen that looks like a red spout. The name comes from the Greek word for "mouth".

A stoma is surgically created to provide an exit for faeces or urine. A person with a stoma has no control over the passing of this waste and so stomas have a pouch fitted to collect the waste matter. They are often a temporary solution to help until further surgery can be carried out.

WHY ARE STOMAS CREATED IN CHILDREN?

There a several types of stoma. The prefix describes their site. The suffix 'ostomy' is another word for stoma:

- A **colostomy** (**col**-ostomy) is an opening in the large bowel brought through the skin and onto the abdomen to provide an exit for solid faeces.
- An **Ileostomy** (**ile**-ostomy) is an opening in the last part of the small intestine (ileum) brought through the skin and onto the abdomen to provide an exit for liquid or semi-solid faeces.
- A **ureterostomy**, (**ureter**-ostomy) **vesicostomy** or **Ileal conduit** are forms of stoma that provide an exit for urine.
- Children born with Hirschsprung's disease or imperforate anus, may at some stage have a stoma.

Example of stoma appliances

WHAT DOES THE SCHOOL NEED TO DO TO PREPARE?

- Understand the child and their family may have been living with a stoma for some time and will know how they need to be supported.
- Arrange a meeting with principal, class teacher and parents/carers to discuss what having a stoma means for that child.
- Discuss with the family what having a stoma means for the child.
- Develop an individual management plan in collaboration with health practitioners and parents/carers that meets the child's needs and support the staff to care for the child.
- Further meetings need to be arranged just prior to the child starting school to clarify any changes to routine and reiterate the plans.

WHAT CAN TEACHERS DO ONCE A CHILD WITH A STOMA IS ENROLLED?

- Ensure discretion and confidentiality.
- Ask the family about special requirements or restrictions around sports/activities.
- Ensure good access to toilet and drinking facilities.
- Families should provide the school with a discreet bag containing clothing, moist wipes, ostomy appliances and disposal bags.
- Aim for the child to clean him or herself but understand they may need assistance to begin with to ensure good hygiene.
- Reinforce the importance of hand washing.
- A communication book or pouch change chart can be very helpful in ensuring consistency between home and school.
- Be consistent with language used at home and at school.
- Allow for adequate time for the student to change stoma bags.

SPORTS

Children with stomas *might* have restrictions around contact sports, depending on the age of the child and type of contact sport. Change the pouch prior to sporting activities to reduce the risk of accidents from leakage. Special stoma guards are also available.

SWIMMING

Swimming is not restricted, but a change of pouch prior to swimming is essential. A stoma guard or smaller pouch may be used. A rash vest is a good camouflage to prevent embarrassment

FLUIDS

One of the functions of the large bowel is water absorption and any loss of the large bowel leads to increased risk of dehydration. Children who have had parts of their bowel removed need to maintain better than average fluid intake. Children who have an ileostomy have no large bowel and are more at risk. Allow them to have a water bottle on their desk and encourage drinking throughout the day.

STOMAL THERAPY AND CONTINENCE NURSES

These are registered nurses based in major hospitals who specialise in stomas, wound care, skin care and continence issues. They are essential in the support of children born with congenital bowel disorders.

Contact Bowel Group for Kids:

www.bgk.org.au

enquiries@bgk.org.au

11 Email response granting authorship permission to publish a poem in this thesis written by Emily Perl Kingsley, 'Welcome to Holland'.

From: Emily Kingsley <epkingsley@gmail.com>

Sent: Tuesday, November 29, 2022, 5:00 AM

To: Eunice Gribbin <grib0021@flinders.edu.au>

Subject: Re: Form Submission - WTH form

Name: Eunice Gribbin

Email Address: grib0021@flinders.edu.au

Purpose of reprint:: I am the mother of a wonderful young man Adam, aged 33 years. Adam was born with Down syndrome and diagnosed with Hirschsprung's disease at one week of age. Hirschsprung's causes an acute bowel obstruction at birth requiring major surgery and some children are left with faecal incontinence, it is also more prominent in DS.. We found Hirschsprung's quickly took over from any concerns we had of Down syndrome and has been the major focus of his difficulties in life, multiple surgeries, losing all his large bowel and the formation of a permanent bag. He has endured so much more than most people would in their lifetime. I have made my life's work as a nurse to provide education, understanding and support to parents who have a child born with a congenital bowel disorder. We started a support group in 1994 with other colleagues and mothers we met in hospital www.bgk.org.au Now I am putting the finishing touches on my PhD and when Adam was born I was sent your beautiful poem and it still brings tears to my eyes today! I have read the background story of your gorgeous boy, I had not known the history behind it and would love

permission to incorporate the poem and maybe some part of your story to my thesis. The focus of the thesis is the 'Unique perspective of congenital bowel disorders from the lens of a nurse, mother and support group co-founder'.

Congratulations on your and your son's brilliant work and thank you for your poem that moves me so much.

With every best wish

Eunice xx

Dear Eunice:

How nice to hear from you.

I was moved by your story. Many years ago I fostered a tiny baby who was born with Down syndrome and rejected by his parents. Within 24 hours of my taking him in, we discovered that he has Hirschsprung's disease and required surgery immediately. The experience of dealing with changing colostomy bags on a child less than a week old was daunting but also strangely rewarding. We really had this baby's life in our hands. He grew up to be a delightful spirited little boy and was adopted by a marvelous loving family. He continues to do well.

I am delighted to give permission for you to use "Welcome to Holland" in the thesis you are writing. You can find more background on me and my work at my website: www.emilyperlkiingsley@gmail.com.

I have a couple of requests -

I am enclosing herewith a complete and accurate copy of Welcome to Holland so you can be sure you are using an "authorized version." It's been used in many places and in many versions. I would appreciate your reprinting it unchanged and uncut, nothing added, nothing deleted, using this official version.

Please credit me as the author, using my full (three-part) professional name: Emily Perl Kingsley and the copyright notice:

Reprinted by permission of the author.

Please let me know that these terms are acceptable to you.

I am not charging you any copyright fees for this usage.

I send all warmest wishes and best of luck with this important project!! All warmest holiday greetings to you and your beloved family.

Sincerely,

Emily

EMILY PERL KINGSLEY

Welcome To Holland

by Emily Perl Kingsley

I am often asked to describe the experience of raising a child with a disability - to try to help people who have not shared that unique experience to understand it, to imagine how it would feel. It's like this……

When you're going to have a baby, it's like planning a fabulous vacation trip - to Italy. You buy a bunch of guide books and make your wonderful plans. The Colosseum. The Michelangelo David. The gondolas in Venice. You may learn some handy phrases in Italian. It's all very exciting.

After months of eager anticipation, the day finally arrives. You pack your bags and off you go. Several hours later, the plane lands. The flight attendant comes in and says, "Welcome to Holland."

"Holland?!?" you say. "What do you mean Holland?? I signed up for Italy! I'm supposed to be in Italy. All my life I've dreamed of going to Italy."

But there's been a change in the flight plan. They've landed in Holland and there you must stay.

The important thing is that they haven't taken you to a horrible, disgusting, filthy place, full of pestilence, famine and disease. It's just a different place.

So you must go out and buy new guide books. And you must learn a whole new language. And you will meet a whole new group of people you would never have met.

It’s just a different place. It's slower-paced than Italy, less flashy than Italy. But after you've been there for a while and you catch your breath, you look around.... and you begin to notice that Holland has windmills....and Holland has tulips. Holland even has Rembrandts.

But everyone you know is busy coming and going from Italy... and they're all bragging about what a wonderful time they had there. And for the rest of your life, you will say "Yes, that's where I was supposed to go. That's what I had planned."

And the pain of that will never, ever, ever, ever go away... because the loss of that dream is a very very significant loss.

But... if you spend your life mourning the fact that you didn't get to Italy, you may never be free to enjoy the very special, the very lovely things ... about Holland.

* * *

On Wed, Nov 23, 2022 at 11:56 PM Squarespace <form-submission@squarespace.info> wrote:

Sent via form submission from Emily Perl Kingsley

Name: Eunice Gribbin

Email Address: grib0021@flinders.edu.au

Purpose of reprint:: I am the mother of a wonderful young man Adam, aged 33 years. Adam was born with Down syndrome and diagnosed with Hirschsprung's disease at one week of age. Hirschsprung's causes an acute bowel obstruction at birth requiring major surgery and some children are left with faecal incontinence, it is also more prominent in DS.. We found Hirschsprung's quickly took over from any concerns we had of Down syndrome and has been the major focus of his difficulties in life, multiple surgeries, losing all his large bowel and the formation of a permanent bag. He has endured so much more than most people would in their lifetime. I have made my life's work as a nurse to provide education, understanding and support to parents who have a child born with a congenital bowel disorder. We started a support group in 1994 with other colleagues and mothers we met in hospital www.bgk.org.au Now I am putting the finishing touches on my PhD and when Adam was born I was sent your beautiful poem and it still brings tears to my eyes today! I have read the background story of your gorgeous boy; I had not known the history behind it and would love permission to incorporate the poem and maybe some part of your story to my thesis. The focus of the thesis is the 'Unique perspective of congenital bowel disorders from the lens of a nurse, mother and support group co-founder'.

Congratulations on your and your son's brilliant work and thank you for your poem that moves me so much.

With every best wish, Eunice xx

12 LNR.13.SCHN.435 - Ethics Approval SCHN - 15 June 2014

The Sydney children's Hospitals Network
care, advocacy, research, education

Contact for this correspondence:
Research and Development
Ethics & Governance Administration Assistant
Phone: (02) 9845 1253
Facsimile: (02) 9845 1317
Email: ethics.schn@health.nsw.gov.au

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Tel +61 2 9845 0000
Fax +61 2 9845 3489
http://www.schn.health.nsw.gov.au/
ABN 53 188 579 090

15 June 2014

Ms Eunice Gribbon
School of Medicine
Australian National University

Dear Ms Gribbon,

HREC Reference: **LNR/13/SCHN/435**

Project title: ***Long term follow up of patients born with Hirschsprung's Disease***

Sites Listed: **The Children's Hospital at Westmead**
Sydney Children's Hospital, Randwick

Thank you for submitting the above project for single ethical and scientific review. This project was first considered by the Executive of the Sydney Children's Hospitals Network Human Research Ethics Committee (HREC) at its meeting held on 20 December 2013. This HREC has been accredited by the NSW Department of Health as a lead HREC under the model for single ethical and scientific review.

This lead HREC is constituted and operates in accordance with the National Health and Medical Research Council's *National Statement on Ethical Conduct in Human Research* and *CPMP/ICH Note for Guidance on Good Clinical Practice*.

I am pleased to advise that following the receipt of further information, the HREC Executive granted ethical approval of this research project. Your approval is valid from the date of this letter.

Document Reviewed	*Version*	*Date*
LNR, Submission Code AU/6/92B715		
Questionnaire		
Emaster Information Sheet	1	14 August 2012
Emaster Consent Form	1	14 August 2012
Audit Survey, Long Term Follow Up HSCR		
Response to Further Information		undated

Please note the following conditions of approval:

1. The co-ordinating investigator will immediately report anything which might warrant review of ethical approval of the project in the specified format, including:
   -Unforeseen events that might affect continued ethical acceptability of the project.

2. Proposed changes to the research protocol, conduct of the research, or length of HREC approval, will be provided to the HREC for review in the specified format.

3. The HREC will be notified, giving reasons, if the project is discontinued at a site before the expected date of completion.

4. The co-ordinating investigator will provide an annual report to the HREC and at completion of the study. The annual report form is available on the Hospital's intranet and internet or from the Secretary.

5. Your approval is valid for 5 years from the date of the final approval letter. If your project extends beyond five years then at the 5 year anniversary you are required to resubmit your protocol, according to the latest guidelines, seeking the renewal of your previous approval. In the event of a project **not having commenced** within 12 months of its approval, the approval will lapse and reapplication to the HREC will be required.

Should you have any queries about the HREC's consideration of your project please contact the Research Ethics Administration Assistant on (02) 9845 1253.

You are reminded that this letter constitutes ethical approval only. You must not commence this research project at a site until separate authorisation from the Chief Executive or delegate of that site has been obtained. A copy of this letter must be forwarded to all site investigators for submission to the relevant Research Governance Officer.

The HREC wishes you every success in your research.

Yours faithfully

Ms Jillian Shute
Executive Officer
Sydney Children's Hospitals Network Human Research Ethics Committee

13 Governance Approval Letter CHW 14 January 2015

The Sydney children's Hospitals Network
care, advocacy, research, education

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Date: 14 January 2015

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ABN 53 188 579 090

Mrs Eunice Gribbin
ANU School of Medicine
PO Box 40 Oakdale NSW 2570

Site Authorisation Letter

Dear Mrs Gribbin,

HREC reference number: LNR/13/SCHN/435

SSA reference number: LNRSSA/14/SCHN/241

Project title: **Long term follow up of patients born with Hirschsprung's disease and their involvement in support groups**

Site: **Children's Hospital at Westmead**

Thank you for submitting an application for authorisation of this project. I am pleased to inform you that authorisation has been granted for this study to take place at the above site.

The following conditions apply to this research project. These are additional to those conditions imposed by the Human Research Ethics Committee that granted ethical approval:

1. Please advise us of the date when the project starts at this site.
2. Proposed amendments to the research protocol or conduct of the research which may affect the ethical acceptability of the project, and which are submitted to the lead HREC for review, are copied to the research governance officer.
3. Proposed amendments to the research protocol or conduct of the research which may affect the ongoing site acceptability of the project are to be submitted to the research governance officer.

Yours sincerely,

Asatina Viviani
Research Governance Officer

14 Governance Approval Letter SCH 16 March 2015

The Sydney children's Hospitals Network
care, advocacy, research, education

Contact for this correspondence:
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Yours sincerely,

Asatina Viviani
Research Governance Officer

15 Families' experiences of Support Groups

Families experiences of support groups

***1. Please enter the year of your child's birth**

Child's year of birth

***2. What sex is your child**

- ◯ Male
- ◯ Female

***3. Is there a family history of Hirschsprung's disease or imperforate anus?**

- ◯ Yes
- ◯ No
- ◯ Unknown

If you answered yes, please give details

4. At which paediatric hospital was your child cared for?

- ☐ Sydney Children's Hospital, Randwick, NSW
- ☐ Westmead Children's Hospital, NSW
- ☐ John Hunter Children's Hospital, Newcastle, NSW
- ☐ Camperdown Children's Hospital, (moved to Westmead 1994)
- ☐ Mater Children's Hospital, QLD
- ☐ Royal Children's Hospital, Herston, QLD
- ☐ Women's and Children's Hospital, Adelaide, SA
- ☐ Royal Children's Hospital, Parkville, VIC
- ☐ Monash Children's Hospital, Clayton, VIC
- ☐ Princess Margaret Hospital for Children, Perth, WA
- ☐ Royal Hobart Hospital, Tasmania
- ☐ Starship Children's Health in Auckland City, NZ

Other (please specify)

Families experiences of support groups

5. Which clinicians are involved in your child's care?

- [ ] General Practitioner
- [ ] Paediatrician
- [ ] Paediatric Surgeon
- [ ] Paediatric gastroenterologist
- [ ] Stomal therapy nurse
- [ ] Continence nurse
- [ ] Continence Physiotherapist
- [ ] Physiotherapist
- [ ] Psychologist
- [ ] Dietician

Other (please specify)

***6. For how long was your child followed up with their Paediatric surgeon**

- ◯ After pull through surgery only
- ◯ up to 5 years
- ◯ up to 10 years
- ◯ up to 15 years
- ◯ up to transition to adult care
- ◯ Our Paediatric surgeon still sees our adult child
- ◯ Not applicable

Other (please specify)

7. Do you find you can discuss your child's condition with family or friends?

- [ ] Family
- [ ] Friends
- [ ] Work colleagues
- [ ] None of the above

Families experiences of support groups

***8. How did you hear about a support group for your child's condition?**

- [ ] Did not know there was a support group.
- [ ] Online search
- [ ] The hospital
- [ ] My child's surgeon
- [ ] Stomal therapy nurse
- [ ] Gastroenterologist
- [ ] Baby health clinic
- [ ] Word of mouth from friend or family

Other (please specify)

***9. When and how should families be informed about support groups for their child's condition?**

- [ ] Online search themselves
- [ ] Families given a pamphlet so they can follow up when they feel ready.
- [ ] As soon as baby is diagnosed
- [ ] Treating surgeon at post op follow-up
- [ ] Stomal therapy nurse in hospital
- [ ] A hospital visit from a support group representative.
- [ ] Baby health clinic

Other (please specify)

10. What age was your child when you heard about a support group?

11. Did you join a support group?

- ◯ Yes
- ◯ No

Families experiences of support groups

12. If you joined a support group, what were the reasons for joining?

- [ ] Information and advice
- [ ] To learn more about the condition
- [ ] To meet others in similar situation
- [ ] To discover what the future held
- [ ] So my child could meet others with the same condition
- [ ] My child was soiling
- [ ] My child was ready for pre-school/school and not yet continent

Other (please specify)

13. If you joined a support group, how long did you remain a member?

- ◯ 1 - 3 years
- ◯ 3-5 years
- ◯ more than 5 years

Families experiences of support groups

16. If yes to previous question, please describe how you feel your child's or families quality of life improved being in a support group.

17. If you have other children in the family, have they been affected by having a sibling with Hirschsprung's disease or imperforate anus?

- ◯ Yes
- ◯ No
- ◯ Not to our knowledge

18. If you answered 'Yes', in what way are your other children affected by having a sibling born with Hirschsprung's disease or imperforate anus?

19. What other services could a support group offer to assist your child and/or your family

16 Paediatric Surgeon Survey

Paediatric Surgeon Survey

1. What is the main area of your practice?

- ◯ Burns
- ◯ Colorectal
- ◯ Chest
- ◯ General
- ◯ Urology

Other (please specify)

2. How many years have you been in specialist practice?

- ◯ 0 - 10 years
- ◯ 10 - 20 years
- ◯ 20 years +

3. Are you Male or Female?

- ◯ Male
- ◯ Female

4. How many new patients do you see annually with Hirschsprung's disease?

- ◯ 0
- ◯ 1 - 5
- ◯ 5 - 10
- ◯ 10 +

5. How many new patients do you see annually with an anorectal malformation?

- ◯ 0
- ◯ 1 - 5
- ◯ 5 - 10
- ◯ 10 +

Page 1

Paediatric Surgeon Survey

6. How long do you think patients born with HSCR or ARM should be followed up?

- ◯ 6 weeks post pull-through surgery
- ◯ 1 year post-surgery
- ◯ 2 years post-surgery
- ◯ 5 years post-surgery
- ◯ Transition to adult care

7. Do you follow up on your own or in joint practice with other medical or paramedical specialists?

- ☐ Own
- ☐ Paediatrician
- ☐ Stomal therapy nurse
- ☐ Continence nurse
- ☐ Continence physiotherapist

8. Do you have an estimation of patients non-attendance at clinics?

- ◯ None
- ◯ 1-5%
- ◯ 5-10%
- ◯ 10%+

9. Are you aware of support groups for the conditions you manage?

- ◯ Yes
- ◯ No

10. Do you think support groups have a place in the care of paediatric patients?

- ◯ Yes
- ◯ No

11. Do you suggest support groups to your parents/patients?

- ◯ Yes
- ◯ No
- ◯ Sometimes; if I feel they are seeking further information, or wish to talk to others in a similar situation.

Paediatric Surgeon Survey

12. Are any of the following barriers to you referring people to support groups?

- [ ] I have not had a Support Groups contact me
- [ ] Parents do not ask about support groups for their child's condition
- [ ] I only refer if the parent/patient asks
- [ ] I do not have time
- [ ] I prefer to refer to hospital based support or ancillary systems such as specialist nurses, psychologist, social workers etc
- [ ] I do not get feedback from patients as to whether a support group was beneficial
- [ ] I do not think support groups are professionally managed
- [ ] Concern about the quality of information support groups will offer
- [ ] Concern over support groups interfering with my management plan
- [ ] Concern parents/patients will meet others with a more severe degree of the condition which may raise unnecessary fears.
- [ ] None of the above

Other (please specify)

13. Have you met with any of the organising committees of support groups to determine relevance to your group of patients?

- ◯ Yes
- ◯ No

14. Would you be more inclined to suggest support groups to patients/parents if you had a better understanding of:

- [ ] How they are set up
- [ ] Who runs them
- [ ] If they provide training for their volunteers & follow a Code of Ethics
- [ ] What services they provide

Other (please specify)

15. Would you consider meeting with support group organisers to work out a way of working together without encroaching on each other's territory?

- [ ] Yes
- [ ] No

Paediatric Surgeon Survey

16. Are you concerned that support groups encourage dependence rather than a healthy attitude to returning to normality once the situation is resolved?

- ◯ Yes
- ◯ No

Other (please specify)

17. Would you consider having information pamphlets about support groups in your reception area for parents

- ◯ Yes
- ◯ No

18. What would your concept of ideal support group be

17 The role and efficacy of Not for Profit groups in paediatrics

The role and efficacy of NFP organisations/support groups in paediatric

1. Who are the Governing bodies of your organisation?

2. Is your organisation Incorporated?

- ◯ Yes
- ◯ No

3. Is your organisation registered as a deductible gift recipient (DGR)?

- ◯ Yes
- ◯ No

4. Do you hold an annual AGM:

- ◯ Yes
- ◯ No

5. How many paid staff does your organisation have?

- ◯ None
- ◯ 1-5
- ◯ 5-10
- ◯ 10+

6. How many volunteers does your organisation have?

- ◯ None
- ◯ 1-5
- ◯ 5-10
- ◯ 10+

The role and efficacy of NFP organisations/support groups in paediatric

7. What positions are held within the organisation?

- [ ] President
- [ ] Vice-president
- [ ] Treasurer
- [ ] Secretary
- [ ] Membership Secretary
- [ ] Corporate Sponsorship Coordinator
- [ ] Conference Coordinator
- [ ] Committee members
- [ ] Other (please specify)

8. Does your organisation have a medical advisory board?

- ◯ Yes
- ◯ No

9. Please list any fundraising activities your organisation carries out.

10. Does your organisation receive corporate sponsorship?

- [ ] Regular Sponsorship
- [ ] Sometimes
- [ ] Never
- [ ] Other (please specify)

11. Does your organisation receive Government funding

- ◯ Yes
- ◯ No
- ◯ Occasionally

12. Do you charge membership fees

- ◯ Yes
- ◯ No

13. Do you charge joining fees?

- ◯ Yes
- ◯ No

14. Do you receive donations?

- ☐ from members
- ☐ Via Everyday hero or other online third party
- ☐ Other (please specify)

15. Does your organisation have a means for members to contact each other?

- ☐ Facebook
- ☐ Forum
- ☐ Chat room
- ☐ Twitter
- ☐ Other (please specify)

16. What year was the organisation formed?

17. Who were the founding members?

- ☐ Parents
- ☐ Nurses
- ☐ Doctors
- ☐ Allied Health Professionals
- ☐ Business People

Other (please specify)

The role and efficacy of NFP organisations/support groups in paediatric

18. What geographical area does your organisation cover?

- [ ] Australian Capital Territory
- [ ] New South Wales
- [ ] Northern Territory
- [ ] Queensland
- [ ] South Australia
- [ ] Tasmania
- [ ] Victoria
- [ ] Western Australia
- [ ] Australia wide
- [ ] Australasia
- [ ] Worldwide

19. What conditions does your organisation support?

20. What age demographic does your organisation support?

- [ ] paediatric
- [ ] Adult
- [ ] Both

21. How many professional members does your organisation have?

22. How many members affected by the condition/s does your organisation have?

23. How long do your members maintain membership?

- ◯ Short term whilst they need information and support
- ◯ Long term for their own needs
- ◯ Long term to support new members
- ◯ Other (please specify)

The role and efficacy of NFP organisations/support groups in paediatric

24. What do you think are factors that may determine who joins a support group?

- [ ] Members have a more severe form of the condition than non-members
- [ ] Education and information
- [ ] A lack of family support
- [ ] Connect with others in a similar situation
- [ ] Online forums/discussion boards.
- [ ] To learn from others who are more experienced at living with the condition
- [ ] Attendance at conferences and/or informal meetings
- [ ] Financial support
- [ ] Access to subsidised equipment or supplies
- [ ] Access to medical advisory board
- [ ] Other (please specify)

25. What services does your organisation provide for members?

- [ ] Conference
- [ ] Local meetings
- [ ] Christmas party
- [ ] Picnics
- [ ] Formal dinners
- [ ] Organised Camps
- [ ] Respite for families
- [ ] Printed newsletters
- [ ] Online newsletters
- [ ] Sibling Support
- [ ] Other (please specify)

26. Does your organisation provide education for health professionals?

- [ ] Yes
- [ ] No
- [ ] Information brochures
- [ ] Only on request
- [ ] Other (please specify)

The role and efficacy of NFP organisations/support groups in paediatric

27. Does your organisation provide in-service sessions for Pre-schools/schools?

- [ ] Yes
- [ ] No
- [ ] Information pamphlets only
- [ ] Other (please specify)

28. Does your organisation provide training for volunteers and/or staff?

- ◯ Yes
- ◯ No
- ◯ Other (please specify)

29. Do your volunteers or staff follow a Code of Ethics?

- ◯ Yes
- ◯ No
- ◯ Other (please specify)

30. Does your organisation run any campaigns to change Government policy on health care issues, if so please describe.

31. Does your organisation conduct any research

- [ ] Our organisation conducts its own research
- [ ] Our organisation considers requests for contribution to research projects
- [ ] Our organisation has not contributed to any research projects to date

Other (please specify)

The role and efficacy of NFP organisations/support groups in paediatric

32. How do members hear about your organisation?

- [ ] Online search
- [ ] Referrals from healthcare providers
- [ ] Advertising in hospitals
- [ ] Advertising in doctors waiting rooms
- [ ] Word of mouth
- [ ] Advertising in baby health clinics
- [ ] Other (please specify)

33. Does your organisation have an affiliation with healthcare providers?

- [ ] hospitals
- [ ] doctors
- [ ] nurses
- [ ] clinics
- [ ] Other (please specify)

34. Do representatives visit families at home or in hospital

- ◯ Yes
- ◯ No
- ◯ Sometimes if requested

Other (please specify)

35. How does your organisation evaluate the effectiveness of programs?

- [ ] Feedback evaluations after an event
- [ ] Surveys to members
- [ ] Surveys to healthcare providers
- [ ] Evaluation at committee meetings
- [ ] Other (please specify)

The role and efficacy of NFP organisations/support groups in paediatric

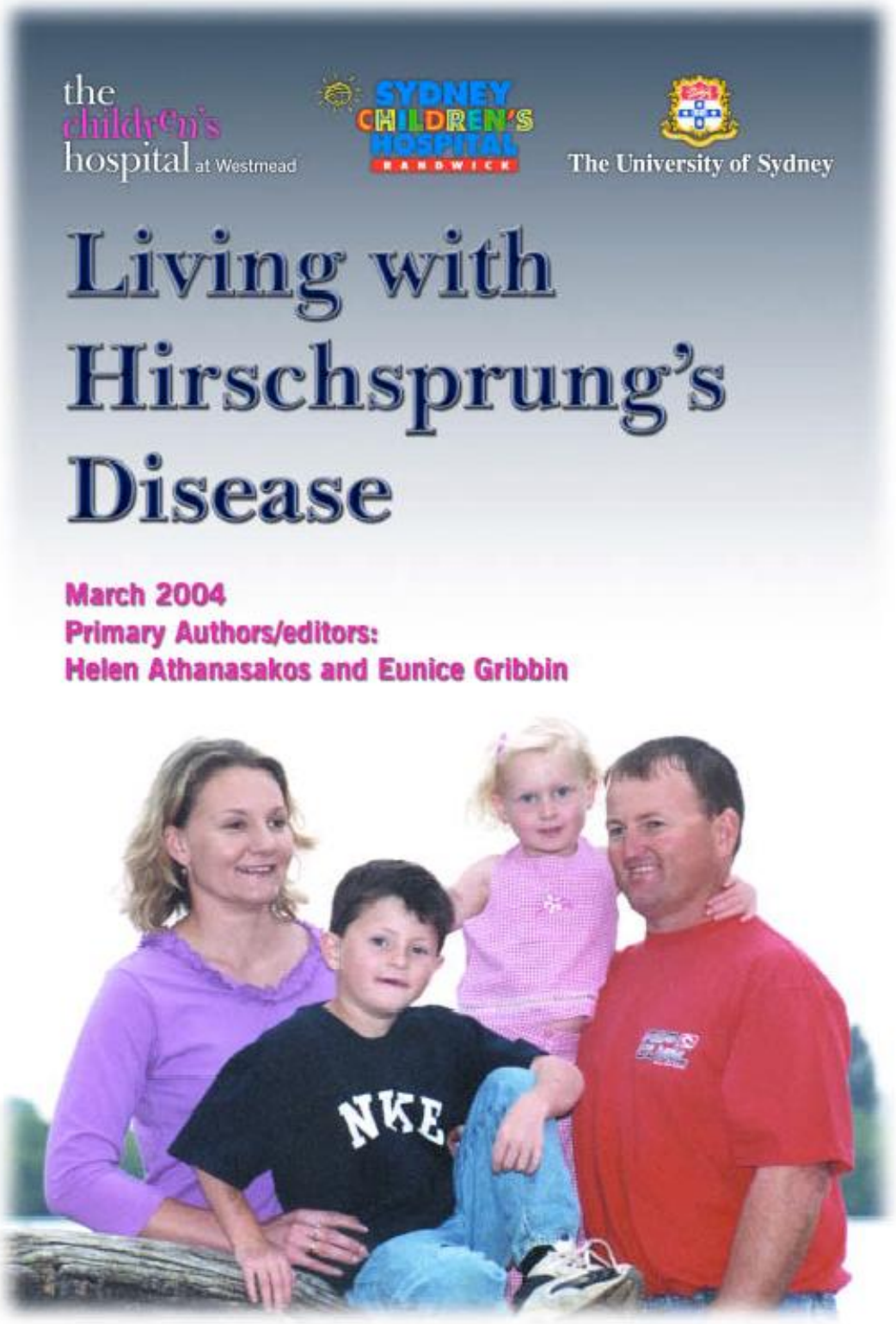
36. Does your organisation have a way of comparing members and non-members to see whether they have:

- ◯ an equal or better quality of life
- ◯ an equal or better medical outcome
- ◯ Our organisation does not have a way of comparing members and none members

37. If yes to previous question, what methods were used?

38. Any other comments you would like to add regarding how your support group runs?

18 'Living with Hirschsprung's disease' Handbook:



CONTENTS

Cover: Living with Hirschsprung's disease.
Tim and Lana Robertson with Lachlan (6) and Haylee (3)

ISBN 0957795157

the children's hospital at Westmead

FOREWORD

Dr Ian Kern

In 1888 Hirschsprung described the disease that carries his name but it was not until the late 1940s, that Swenson and Co-workers associated the physiology and pathology of HSCR to devise the first curative operation. Since that time there has been an increasing knowledge about the disease itself, its genetic background, its association with other diseases such as Down syndrome and the somewhat mysterious entity known as Intestinal Neuronal Dysplasia (IND), and its management. Initially, the treatment was divided into three stages but now we are seeing one-stage operations and the use of the laparoscope to enable patients to recover more quickly with less surgical complications, better bowel functioning and overall quality of life.

Although Hirschsprung's disease is not a common disease, probably affecting 1 in 5000 newborn babies, it is a complicated problem and parents/carers have to put in an enormous amount of effort. Any problem is best treated by understanding its cause, knowing what to expect and how to deal with it in everyday life. The sharing of knowledge is a major factor in allowing parents/carers to deal with the problem. I have always been impressed by the devotion of parents/carers of children with Hirschsprung's disease and the importance of their input. I always described to them the nature of Hirschsprung's disease and its management but have always found they will get more out of another parent/carer.

This handbook has been prepared to enable parents/carers to have an understanding of Hirschsprung's disease, how to manage children with the disease and what to expect for the future. Both the primary authors and contributing authors are eminently qualified to prepare this handbook. The handbook has been developed from both The Children's Hospital at Westmead and Sydney Children's Hospital Randwick.

I have known Eunice Gribbin for fifteen years when her own son was afflicted with this condition. She is a registered nurse and has spent some time working with many workers who are trying to solve the problem of Hirschsprung's disease both academically and practically. She is co-founder/secretary for the Bowel Group for Kids (*formally Australian Pseudo-obstruction Support Association (APSA)*, which conducts seminars and produces a newsletter called Segments. I have found Eunice to be a woman of great empathy and her help in supporting other parents has been of the utmost importance.

Helen Athanasakos has had her interest in the disease stimulated by the fact that her brother was diagnosed with Hirschsprung's disease in 1988 and through her medical studies. She has just completed her Master's degree in this subject at the University of Sydney in conjunction with the Department of Academic Surgery and Psychological Medicine at The Children's Hospital at Westmead. Her Masters thesis investigated the long-term outcomes of the condition for the child with HSCR and the family/carer involved. She has conducted many seminars for families with HSCR, contributed articles to "Segments", published works and has formed a partnership with Eunice to produce the handbook.

As I have said above, information is vital in helping the parents/carers coping with children who are suffering who have Hirschsprung's disease. I am confident that this handbook will fill the gap and help parents/carers to cope on an everyday basis and remind them that they can receive help from such people as Eunice and Helen.

think. kids.

Published by
the children's hospital at Westmead

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www.chw.edu.au

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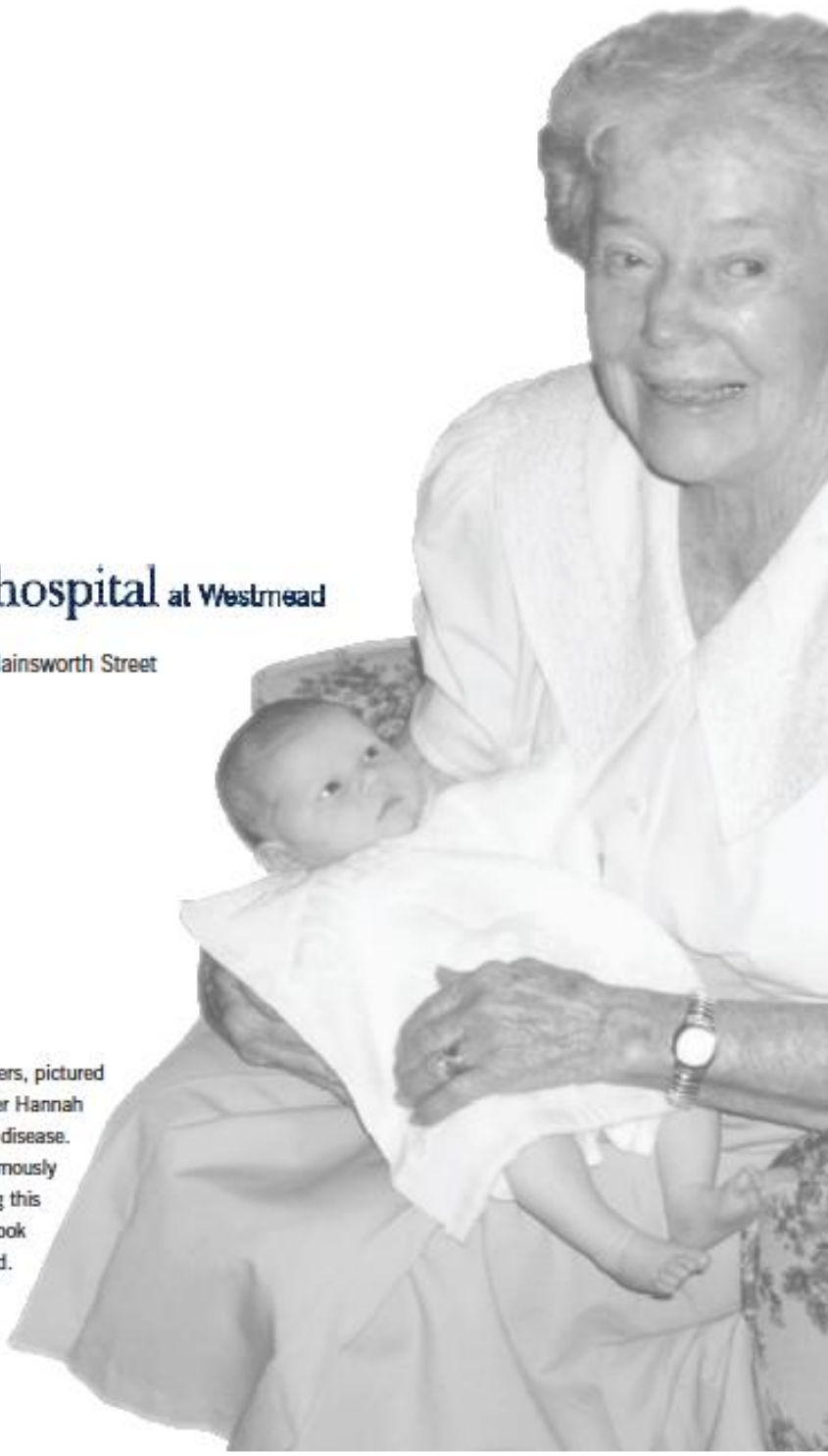

Special thanks to the late Dot Peters, pictured here with her beloved granddaughter Hannah who was born with Hirschsprung's disease. Dot lived for her family and posthumously donated generous funds enabling this second edition of the HSCR handbook to be printed.

19 Unedited and anonymous comments made by the children who attended the Bowel Group for Kids conference December 2007, self-named the 'scar club'.

The Scar Club:

- Parents and teachers don't really understand.
- It's hard to express pain.
- The scar site gets itchy as well as painful.
- Please don't ask all the time about pain.
- Don't massage my stomach – it doesn't help or if you do it PLEASE be gentle.
- Don't ask the child to do something that won't help e.g. Panadol makes it worse.
- Just let them do what they know will help – Don't fuss or overreact.
- Need to eat slowly – can't hurry up as it causes vomiting and pain.
- Let them make choices on how they can manage pain – they will let you know if it's not resolving or if we need more intervention.
- Don't ignore complaints of pain e.g. when it happens a lot don't just say "go & sit down".
- Leave them alone when in the toilet; don't hassle them or keep checking "are you alright in there?" it makes them lose concentration.
- If they say they are full, let them stop eating, don't force them.
- If they say "this food gives them a sore tummy" don't make them eat it.