

Congenital Hemiplegia and the Neglected Upper Limb

A Thesis for the Degree of Doctor of Philosophy in Medicine

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DEDICATION

This thesis is dedicated to the memory of my father, Eduardo Domenico Russo, who died after a brave battle with lymphoma in 1991. He recognized the value of pursuing your dreams and goals, and working hard to bring them into reality. He instilled in me a real passion for working towards a goal, and for knowing the truth about how things in life really work – two vital skills for anyone wanting to complete a PhD. His motto was “*Find your passion, aim to be the best, and never give up!*”

My wish is that he could know how helpful he was to me during this difficult and challenging pursuit, and how grateful I am for his influence in my life.

Eduardo Domenico Russo



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DECLARATION STATEMENT

I certify that this thesis does not incorporate, without acknowledgement, any material previously submitted for a degree or diploma in any university and 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.

Signature _____

Date ____/____/____

Dr Remo Nunzio Russo

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SUMMARY OF ABBREVIATIONS

1. ACTR – Australian Clinical Trials Registry
2. ADL – Activities of daily living
3. AMOS – Analysis of a moment structure
4. AMPS – Assessment of motor and process skills test
5. AT – Assistive technologies
6. BoNT-A – Botulinum toxin A
7. CFA – Confirmatory Factor Analysis
8. CFI – Comparative fit index
9. CP – Cerebral Palsy
10. CPRHand – Cerebral Palsy Register measure of Hand Function
11. DIAT – Dynamic Interference of Tone - Upper Limb
12. DTRs – Deep tendon reflexes
13. GAS – Goal Attainment Scale
14. GLM – General Linear Modelling
15. GMFCS – Gross Motor Function Classification System
16. HCP – Hemiplegic Cerebral Palsy
17. ICF – International Classification of Functioning
18. IQ – Intelligence Quotient
19. MACS – Manual Ability Classification System
20. ManHands – Manipulative Hand Skills
21. MAS – Modified Ashworth Scale
22. OT – Occupational Therapist

- 23. PEDI – Pediatric Evaluation of Disability Inventory
- 24. PedsQL – Pediatric Quality of Life Inventory
- 25. PRP – Paediatric Rehabilitation Physician
- 26. QUEST – Quality of Upper Extremity Skills Test
- 27. R1 – Range of motion at “first catch”
- 28. R2 – Full passive range of motion
- 29. RMSEA – Root Mean Square Error of Approximation
- 30. SACPR – South Australian Cerebral Palsy Register
- 31. SAE – Serious Adverse Event
- 32. SPSS – Statistical Package for the Social Sciences
- 33. UPLIFn – SACPR Upper Limb Functional Assessment Scale

THESIS SUMMARY

Hemiplegic cerebral palsy (HCP) is a static neurological condition that primarily affects one side of the body. There are associated cognitive and functional problems impacting on the life of affected children. Lower limb effects have been extensively studied with clinical/laboratory based tools, to the point of being very reliable to assist families in decision making to achieve better functional outcomes. However, there is a paucity of evidence for the effects on upper limb dysfunction. Some reasons for this lack of evidence is that upper limb function, not being reliant on repetitive, stereotypic motor and sensory function like the lower limb, is impacted upon by many related factors including cognition, environment (including adaptive devices), self-concept, pain, quality of life and other factors interacting in a complex manner.

The primary focus of this work is on functioning and independence, with an exploration of the clinically relevant factors that could be measured that impact on these functional outcomes. Associations in the areas of self-esteem and self-concept, quality of life and the experience of pain are explored. The secondary aim is to explore the effects of botulinum toxin injection to improve functioning.

These aims are achieved through conducting two related studies. The first (Part 1) was a cross sectional analysis of children with HCP recruited from a population based cerebral palsy register, with an analysis of the children's functional level as defined by the Assessment of Motor and Process Skills, clinical neurological examination, as well as measures of quality of life, self-esteem/self-concept, pain,

caregiver burden, and use of orthoses and assistive devices. The second (Part 2) was a single blind randomized control trial recruiting children from Part 1, with a focus on functional improvement and the attainment of individualised predefined goals. All participants undertook regular occupational therapy and the intervention group had upper limb botulinum toxin injection.

Children with hemiplegic cerebral palsy are resilient, with levels of self-esteem equivalent to sex and age matched typically developing peers. They report significantly lower levels in some self-concept domains (such as physical and scholastic competencies), and children recognise their limitations from a young age. They self-report lower levels of quality of life, and higher levels of pain. The impairments most strongly associated with functional level and independence are muscle power and sensation, indicating prediction of and improvement in functional independence requires a focus on sensory testing and strengthening. Tone is less strongly associated, however the degree of upper limb tone is related to the need for intervention. Knowing the degree of upper limb muscle tone from a young age is helpful in assisting families with children with a new diagnosis of HCP. Children who had an acute reduction in tone with the use of botulinum toxin injection, however, achieved their stated functional goals more quickly, with an associated boost in self-esteem, unlike the control group, who had lower levels of self-esteem during the study period possibly related to a focus on functional gain with a slower rate of improvement with therapy alone.