

THE FUNCTIONAL ABILITY OF CHILDREN WITH ARTHROGRYPOSIS IN THE EXECUTION OF ACTIVITIES OF PERSONAL MANAGEMENT

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A research report submitted to the Faculty of Health Sciences, University of the Witwatersrand, Johannesburg, in partial fulfilment of the requirements for the degree of Master of Science in Occupational Therapy.

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DECLARATION

I, Frances Geyser, declare that this research report is my own work. It is being submitted for the degree of Master of Science in Occupational Therapy in the University of the Witwatersrand, Johannesburg. It has not been submitted before for any degree or examination at this or any other university.

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ABSTRACT

This study investigated the functional ability of 19 children with arthrogryposis aged six months to seven and a half years in the execution of age-appropriate personal management tasks. The level of independence, amount of caregiver assistance and modifications used during each task were ascertained using the Pediatric Evaluation of Disability Inventory (PEDI). Participants were divided into functional age groups namely infant, toddler, preschool and school aged. Results showed that the infant and toddler group performed above expected levels in the self-care domain, whereas the preschool and school aged group performed significantly below the expected level. A significant correlation between the amount of caregiver assistance needed and the participants' level of independence was found especially in the preschool and school aged group. There was little use of modifications and participants relied on compensatory methods and assistance to execute personal management tasks. Guidelines were established to encourage therapists to make more use of modifications in furthering age appropriate independence of this population.

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All the children and their families that agreed to participate in the study.

My family and friends who always supported me.

DEDICATION

I dedicate this study to every child with arthrogryposis in South Africa, and every individual that works with them to help them reach their full potential. And to my father, who always encouraged me to further my education.

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OPERATIONAL DEFINITIONS

Activities of daily living – serves as an umbrella term including all those activities related to the care of one's own body referred to as personal management in this study e.g. bathing and showering, toileting and toilet hygiene, dressing, swallowing/eating, feeding, functional mobility etc. It further encompasses more complex instrumental activities of daily living enabling the individual to function within a community including occupations such as the care of others and pets, child rearing, communication management, driving and community mobility, financial management, health management and maintenance and home establishment and management (1).

Arthrogryposis multiplex congenita or arthrogryposis - a non-progressive condition characterized by multiple congenital contractures in the limbs (2).

Amyoplasia – the most common subtype of arthrogryposis, mostly characterised by symmetrical involvement of all four limbs (2, 3).

Distal arthrogryposis – an autosomal dominant subtype of arthrogryposis where only the distal parts of the limbs are affected (2, 3).

Caregiver assistance – the amount of assistance a child needs from a caregiver during the execution of self-care tasks(4).

Modification – equipment and environmental modification used by the child during the execution of a self-care task. The type of modification is described as being child-orientated equipment, rehabilitation equipment or extensive modifications(4).

Child orientated modification - a modification or assistive device that is non-specialised and not only limited to the disabled population e.g. baby spoon or straw(4).

Rehabilitation equipment – equipment or assistive device usually prescribed by a health care professional and specific to the disabled population e.g. long-handled, angled spoon, one handed cutting board(4).

Extensive modification – alteration to the living environment or specialised equipment e.g. bath lift, wheelchair, ramp(4).

Personal Management – self care: all activities related to the care of your own body and which are fundamental to enable basic survival and well-being(5).

Splinting – treatment modality used by occupational therapists to address a hand injury or congenital deformity (6).

Passive stretching – treatment modality usually used in conjunction to splinting to improve a joint's range of motion and usually executed by the caregiver on recommendation of a health care professional (7).

Remedial therapy approach – therapy aimed at restoring an impaired skill to the previous functional level, or to establish a skill not yet developed(5).

Rehabilitative (compensation, adaptation) approach – using compensatory methods and assistive devices to enable the execution of a task, usually when the skill can not be remediated (5).

ABBREVIATIONS

AAMR - American Association on Mental Retardation

AMC - arthrogryposis multiplex congenita

AMPS - Assessment of Motor and Process Skills

CMJAH – Charlotte Maxeke Johannesburg Academic Hospital

DASH – Disabilities of the arm, shoulder and hand measure

FIM - Functional Independence Measure

HPCSA – Health Professionals Council of South Africa

ICF – International Classification of Functioning, Disability and Health

IFSSH - Federation of Societies for Surgery of the Hand

LSEN- Learners with special educational needs

NORD - National Organization for Rare Disorders

OTPF – Occupational Therapy Practice Framework

OTT – Occupational Therapy Technician

PEDI- Paediatric Evaluation of Disability Inventory

POP – plaster of paris

SD – Standard Deviation

CHAPTER 1

INTRODUCTION

Congenital deformities of the upper extremity have been reported to occur in 0.16 to 0.18 percent of live birth with approximately 10% of the above-mentioned infants having partial to complete absence of a limb(8). In 1983 the International Federation of Societies for Surgery of the Hand (IFSSH) classified congenital deformities into seven categories(9) according to the morphologic and structural characteristics(10) for clarity as so many different deformities exist.

Although the classification is widely used in clinical settings up to the present day, it has however been criticised as it does not have significant practical value when prescribing occupational therapy, which considers the functional performance of each individual child(9). Especially upper limb function is necessary for the execution of most personal management tasks like feeding, dressing, toileting and grooming and most of these tasks are of a bilateral nature. A lack of function in one, or both upper limbs will thus compromise an individual's independence in the execution of these tasks. The development of independence in personal management tasks is not only important for maintaining bodily health and hygiene, but also culturally significant as the mastery thereof is expected by the broader community in which a child functions (11). Most schools expect a level of independence in personal management tasks before they accept a child into the school (12, 13), and it serves thus as a prerequisite for the engagement in other occupations. Mastery of self-care tasks also leads to a sense of mastery and positively contributes to the child's self-esteem and self-reliance. As it provides a sense of autonomy it sets the foundation for the mastery of other more complex skills, as the child develops (11).

One of the conditions with clinically significant severe functional limitations referred to occupational therapy in hospitals and schools for Learners with Special Educational Needs (LSEN) are children with arthrogryposis multiplex congenita (AMC) as upper limb function and therefore independence in personal management tasks is significantly compromised. Arthrogryposis multiplex congenita or arthrogryposis is a non-progressive condition characterized by multiple congenital contractures that cause significant functional limitations in the limbs and occurs in only 0.03% of live births.



Figure 1.1 Typical presentation for upper limb deformity in arthrogryposis

The typical presentation of arthrogryposis is a loss of muscle mass surrounding the shoulder girdle and elbow with a general inability to actively flex and elevate the shoulder(2).(Figure 1.1)

The elbow is usually fixed in extension with limited flexion, as a result of weakness or absence of the biceps muscle, which hinders the execution of functional activities of daily living (14).The wrists are flexed and there is a loss of opposition of the thumb.

This disorder presents in approximately 150 different forms and is treated by a variety of professionals, including orthopaedists, geneticists, and neurologists and rehabilitation therapists, all of which tend to focus on a different aspect(15). Although classification of congenital hand deformities and understanding the aetiology and medical intervention for arthrogryposis are important, it does not do much in terms of guiding rehabilitation as each case should be assessed and managed individually(9). Therefore occupational therapy is recommended as early as possible after birth and the outcome of treatment in occupational therapy irrespective of the severity of the condition, is geared towards increasing the functional use of the upper limbs and optimizing independence in occupational performance, especially in tasks of personal management(16).

1.1 Problem statement:

The incidence of arthrogryposis multiplex congenita in South Africa is unknown, but a number of these cases are encountered by the researcher at Charlotte Maxeke Johannesburg Academic Hospital (CMJAH) and by occupational therapists in public hospitals and LSEN schools. Evidence for the occupational therapy management of these children in this context is minimal. Although surgical intervention can alter a congenital deformity, it cannot eliminate it and thus hand function in terms of everyday activities in this population remains compromised.

Little research has been published on the ability of children with arthrogryposis to execute activities related to personal management. Previous research has concentrated on the components of hand function and poor fine motor ability which has been shown to have a poor correlation with their ability to participate in activities of daily life(17, 18). All the research published on the treatment and functional ability of children with arthrogryposis was conducted in first world countries where they had access to adequate health services. There is no research published on the outcomes for these children in the South African setting. Health and education services may be less accessible to children especially those that reside in rural settings with only basic health care facilities. Those that have access to specialised hospitals may find access to therapeutic and surgical services challenging due to financial restraints and limited resources. The question must be asked about what the functional outcome of these children is and how will it compare to those in first world countries? It is thus important to establish the functional ability in the occupational performance areas of this population for occupational therapy treatment to be effectively prioritised and to choose between remedial and adaptive therapy.

1.2 Research question:

What is the functional ability of children with arthrogryposis aged six months to seven and a half years in the execution of age appropriate activities of personal management?

1.3 Purpose of the Study

The purpose of the study is to establish the ability of children with arthrogryposis in the Witwatersrand and Tshwane areas under the care of an occupational therapist for at least three months to carry out age appropriate activities in the occupational performance area of personal management, using a standardised disability inventory.

The assessment of the child's ability to complete these self care activities will include the use of environmental modifications that support functional performance as well as the amount of assistance required from a caregiver. This will assist an occupational therapist to determine whether a remediative or an adaptive therapeutic approach will be suitable in treating these children, and to determine how much caregiver assistance and modifications are needed.

1.4 Aim of the Study

To determine the functional ability of children with arthrogryposis aged six months to seven and half years under the care of an occupational therapist for at least three months, in the execution of age appropriate activities of personal management.

1.4.1 Research objectives:

- To compare the ability of children between six months and seven and a half years with arthrogryposis, to execute functional activities related to personal management.
- To establish the type of environmental modifications that support functional performance used by children between six months and seven and a half years with arthrogryposis in the execution of functional activities related to personal management.
- To establish the type and amount of assistance provided by caregivers to children between six months and seven and a half years with arthrogryposis in the execution of functional activities related to personal management.
- To compare the results obtained regarding the ability, modifications and caregiver assistance to the PEDI norms for four developmental age groups of children with arthrogryposis.

1.5 Justification of the study

A thorough literature search indicated that there is a lack of research into the functional performance of children with congenital hand deformities, especially arthrogryposis in personal management tasks. Without research it is difficult to prioritise treatment aims effectively, as well as to provide intervention to accommodate for the functional loss and provide compensation for future personal management, play and school performance.

Therapists encounter children with arthrogryposis in the clinical setting and evidence for treatment is very limited. It is hoped that by determining the functional independence in self care of children with arthrogryposis, evidence for therapy will provide guidance in treatment prioritisation and planning. This is necessary as therapists needs to establish which self care activities can be remediated and which needs to be addressed using adaptive therapy. It would also give an indication of how much caregiver assistance and modifications are needed in different activities of personal management.

Arthrogryposis is recognized by the National Organization for Rare Disorders (NORD), and although it is more difficult to do research on a rare disease, they believe that because of the rarity of these conditions, the families and patients are more driven to find effective treatment methods and encourage research of these disorders(19). Multiple associations and support groups in Britain, Ireland, Australia, America and other countries testify to this. Successful research done on this disorder will encourage further research

of other rare diseases. The NORD reports that of the 7000 rare diseases only a few hundred have treatments, and research should thus be encouraged(19).

CHAPTER 2

LITERATURE REVIEW

This literature review will consider congenital deformities of the upper limb with specific reference to arthrogryposis. The classification and medical intervention for arthrogryposis will also be considered. The need for appropriate identification of dysfunction in everyday activities and functional outcomes for children with arthrogryposis on which evidence based practice in occupational therapy can be measured will also be presented.

2.1 Congenital deformities

The development of the upper limb is dependent on multiple genetic and environmental factors. An abnormality in any of these factors can lead to the formation of a congenital deformity like arthrogryposis(20). The prevalence of congenital deformities has led to a need for a universal classification system. Congenital deformities are grouped together according to their morphologic and structural features. The International Federation of Societies for Surgery of the Hand (IFSSH) established seven categories of classification namely(9):

- Failure of formation
- Failure of differentiation
- Duplication
- Overgrowth/ gigantism
- Undergrowth/ hypoplasia
- Congenital constriction band syndrome
- Generalised skeletal abnormalities

The Japanese Society of Surgery for the Hand changed the classification to include abnormal induction of rays (syndactyly, triphalangeal thumb etc.) and unclassifiable cases(21). There are not a lot of research studies on the epidemiology of congenital abnormalities of the upper limb available in literature. One study showed that the incidence of failure of formation and duplication is higher in males, whilst the incidence of undergrowth and constriction band syndrome is higher in females(22).

Congenital hand deformities are rare with an estimation of 22.91 cases per 10 000 births according to the IFSSH. Out of these 22.91 cases arthrogryposis was 0.2 cases (9).The incidence of arthrogryposis is said to be between 1 in 3000 and 1 in 5100 births(23).

2.2 Arthrogryposis

2.2.1 Classification of arthrogryposis

Arthrogryposis is classified under failure of differentiation on the IFSSH classification system(24). The first description of a case of arthrogryposis was published in 1841 by Otto(25). Thereafter, all other cases described considered the aetiology as being primary contractures resulting in the joint deformities. The word arthrogryposis is derived from two Greek words meaning hooked or curved joints(3).

Later research showed that the primary cause of the joint contracture is actually due to a lack of functional muscle and the term was changed to arthromyodysplasia to indicate muscular and skeletal dysfunction(25). Although the term arthrogryposis multiplex congenita (AMC) is still used, it is important to remember that the term arthrogryposis does not refer to one specific condition but describes a group of 105 different disorders where more than two joint contractures are prevalent in children(23, 25).

As the disorder has varying clinical features a classification system was developed to describe the aetiology of the disorder more accurately. Bamshad, Hall et al(26, 27) divided the presentation of arthrogryposis into three groups: the first describing a group with only limb involvement, the second being a group with limb and other body part involvement, and the third being a group with limb involvement as well as central nervous system dysfunction. Survival in the third group is uncommon as half of this population dies soon after birth(23).

The greatest number of children with arthrogryposis fall into the first group described by Bamshad, Hall et al, otherwise known as amyoplasia, or distal arthrogryposis which is characterised by a prenatal lack of functional muscle in the limbs. The number of limbs affected varies and only one joint to multiple joints in all four limbs can be affected. Ten percent of children have additional abdominal structural abnormalities and more than half have genital abnormalities(2).

The typical clinical presentation in the upper limbs of amyoplasia is that of restricted and internally rotated shoulders, extended elbows with limited flexion and limited pronation of the forearms. Flexion deformities of the wrists and fingers are also common. In the lower limbs hips are either flexed and internally rotated or extended and dislocated, knees are either flexed or extended and clubfeet are common(14, 23).

Distal arthrogryposis is a subclinical group describing less severe hand or foot deformities like ulnar deviation of the hand and finger flexion deformities, clubfeet or other foot deformities. Proximal large joints are mostly spared but may have mild flexion deformities

that are more easily correctable than those seen in amyoplasia(23). Although the elbow joint is not restricted there may be weakness of the biceps muscle also making hand to mouth movements difficult(14). Distal arthrogryposis is subdivided into ten different types(2).

2.2.2 Aetiology of arthrogryposis

Arthrogryposis is non progressive and present at birth(8, 24). There is no single cause of arthrogryposis and more than one factor seems to be involved including genetic and environmental factors which leads to a decrease in fetal movement in utero. A joint may be developing normally but due to the lack of movement more connective tissue forms around the joint, tendons are not lengthened and the bones surrounding the joint becomes more flattened(3). These changes result in limited range of joint movement after birth.

A lack of movement in utero can be ascribed to a number of reasons. Potential aetiologies include a neuropathic cause which affects the brain, spine or peripheral nerves. It is thought that central nervous system dysfunction especially affecting the motor cortex, leads to a decrease activation of the cortico-spinal tract and spinal cord motor neurons which leads to a decrease of foetal movement. This is especially true for amyoplasia or classic arthrogryposis. Injury to the spinal cord can also limit foetal movement resulting in comorbid arthrogryposis(2, 25,28).

Abnormality of muscular structure and function like muscular dystrophies and mitochondrial abnormalities although rare, as well as connective tissue abnormalities, limited space in utero as seen in twin pregnancies, maternal diseases such as Multiple Sclerosis and Myasthenia Gravis have been implicated in the aetiology of this condition. Trauma affecting the fetus's neuromuscular junction, and lastly impaired vascularity of either the uterus or the fetus are also thought to play a role(2, 23).The lack of fetal movement in utero resulting from the conditions described above has been associated with the limb muscles becoming fibrotic, collagen proliferation and joint capsule thickening(26).

Although there has been little research on the aetiology of arthrogryposis most researchers have tried to understand what causes the deformities. In a study done by Hall and Fahy in 1990 when they reviewed 828 arthrogryposis cases it was found that the main birth factors related to arthrogryposis were twin pregnancy, too little or too much amniotic fluid, and severe nausea during pregnancy(29). Sodergard, Hakamies-Blomqvist et al in 1995 found similar results in their smaller sample of 52 and identified twin pregnancies and too much amniotic fluid as the main factors associated with the condition(30). It has however been proposed that arthrogryposis is caused by multiple

genetic factors along with the more than other 35 associated muscular disorders which result from genetic aetiologies(23). Distal arthrogryposis is linked to an autosomal dominant genetic disorder with the genes implicated being those responsible for the production of acetylcholine or associated receptor proteins(2).

Like most congenital contractures though the aetiology for most cases remains unknown(2). Arthrogryposis is generally only diagnosed after birth, but ultrasound is now able to detect it in utero. A lack of fetal movement as perceived by the mother can also suggest the presence of arthrogryposis(23). Since it is a non progressive condition the treatment starts at birth and generally is surgical in nature in an attempt to minimise the deformities.

2.2.3 Upper limb deficits in arthrogryposis

The incidence of upper limb deformities vary, with symmetrical involvement of more than 90% of this population presenting with elbow joint deformities in either flexion or extension. More than 80% of patients have wrist and shoulder deformities in various degrees. In general all four limbs are mostly affected, with only lower limb involvement being more common than only upper limb involvement. The severity of the deformities is worse distally, with proximal joints being less affected(23, 25).

Children with arthrogryposis usually have very limited shoulder movement due to muscle weakness of the deltoid and external rotator muscles, resulting in the shoulder being internally rotated and adducted. Individuals with arthrogryposis have limited shoulder abduction and external rotation, making reach, which is a crucial component in most self care activities, difficult. Range of motion in the elbow joint varies but the joint is mostly in extension with limited active flexion. The forearm is mostly pronated(2, 7). The triceps muscle is more developed than the biceps and brachialis muscle making hand to mouth movements without assistance impossible(7, 14). Flexion and ulnar deviation of the wrist is common in children with arthrogryposis and intra-articular and intercarpal adhesions are present, limiting wrist extension(2, 7). Hand deformities vary and may include flexed and overlapping fingers, ulnar deviation of the metacarpal phalangeal joints, thumb-in palm deformities or webbing of interdigital spaces. Thumb opposition is limited and children with arthrogryposis mostly make use of finger adduction (interdigital) to grasp objects(2, 7).

Especially upper limb function is necessary for the execution of most personal management tasks like feeding, dressing, toileting and grooming and most of these tasks are of a bilateral nature. A lack of function in one, or both upper limbs will thus compromise an individual's independence in the execution of these tasks. The

development of independence in personal management tasks is not only important for maintaining bodily health and hygiene, but also culturally significant as the mastery thereof is expected by the broader community in which a child functions (11). Most schools expect a level of independence in personal management tasks before they accept a child into the school (12, 13), and it serves thus as a prerequisite for the engagement in other occupations. Mastery of self-care tasks also leads to a sense of mastery and positively contributes to the child's self-esteem and self-reliance. As it provides a sense of autonomy it sets the foundation for the mastery of other more complex skills, as the child develops (11).

2.3 Intervention for the upper limb in arthrogryposis

2.3.1 Surgical intervention

Families often rely on surgery to improve the appearance of the child's limbs, but at the same time this may compromise function(31). Surgery is most often indicated for children with amyoplasia but even surgery which aims to increase function tends to be unsuccessful(9) and even with surgery these children can require assistance into their adult life(2).

There is some debate about exactly what the most appropriate age to start with surgical intervention is, but it is mostly advocated to take place between six and 18 months. Earlier corrective surgery should be completed between three and 12 months of age, because this is the time in which important grasps and fine motor coordination develops and the child's active immunity has matured. If surgical intervention takes place after certain grasp patterns have been established it may not be able to alter those patterns(8, 32).

The advantages of early corrective surgery is the child's ability to recover better as skin is more elastic, nerve recovery is better and blood vessels are more visible(9). Some literature shows that delaying surgery until after one year of age makes it more problematic as contractures become more severe(8). Further surgical procedures then take place as the child grows and it becomes necessary(9, 32).

The procedures most commonly advised in surgery are related to improvement of the deformities that form as a result of arthrogryposis. Procedures to improve joint positioning of the shoulder include an external humeral derotation osteotomy at the shoulder joint(2). Other surgery to increase the range of motion includes a carpectomy at the wrist with soft tissue release and remodelling of the carpal bones. Muscle transfers also allow

replacement of missing movements like wrist extension and prevent the recurrence of the flexion deformity(14, 23).

Limited elbow flexion can be corrected with an elbow flexorplasty and a posterior elbow capsulotomy, thus releasing the joint and lengthening the triceps muscle(2, 23). A transfer of the triceps or other muscle tendons can also be done to improve active elbow flexion. Usually elbow flexion in one arm is corrected making hand to mouth movements possible, whilst the other arm is left in extension for the execution of other activities of daily life(2, 14). A thumb release and opponensplasty is done to improve pincer function, with other contractures being released and corrected with skin grafts(2, 23).

Although surgery is considered one of the main options for children with arthrogryposis to improve their functional range, they are at an increased risk of developing pre and postoperative complications. Their venous access is difficult and as their jaws may be affected it makes endotracheal intubation difficult. They are at risk of developing hyperthermia during surgery and their varied muscle mass makes the distribution of pharmaceutical agents challenging. Postoperatively they have a greater risk of aspiration and they present with vertebral instability(23). A study on 87 patients with arthrogryposis noted the following pre and postoperative complications: gastrointestinal difficulties including constipation, decrease weight gain leading to failure to thrive as well as chronic upper respiratory infections(23). Since this population has been shown to be more at risk for surgery this option needs to be adopted with caution and non invasive procedures should be considered first in the treatment of these deformities.

2.3.2 Non invasive intervention

Usually, in order to limit the need for future surgery, a multidisciplinary team provides non invasive treatment with the goal of decreasing deformities. This intervention is started in the first four months postnatally and is seen as a crucial period to prevent fixed deformity.

Passive stretches of the elbow, wrist and finger joints are advocated to maintain joint integrity and serial splinting often during nighttime is advocated to improve functional range(2, 14). This is usually done by a combined team of a physio and occupational therapists and carried out as part of a home programme where the caregivers are educated on how to apply the treatment protocol in their home environment(23).

A study done on 63 patients who began treatment at younger than 18 months, showed that serial splinting and daily passive stretches improved both wrist and elbow mobility, thus making this type of intervention successful with an improvement in elbow mobility of 38% and wrist mobility of 43%(14). An older study which also described the outcome of

rigorous passive stretches and serial splinting showed a definite improvement with the need for later surgery decreasing from 5.6 to 2.7 procedures per patient after implementation of the programme. They also acknowledged that the younger the infant when treatment starts, the better the results(33).

The multidisciplinary team including the surgeon should work together to decide on the best treatment protocol for each specific patient(6). Most tasks can be executed unilaterally if needs be, but at times the only way to maximise hand function is through surgery especially for activities like toileting, dressing, playing and writing(9). If there is no improvement with splinting within the first six months, surgery is advocated(8). If surgical procedures are carried out, the occupational therapist continues to play an active role in post operative scar management and hand therapy(9). As children are more prone to injure themselves initial splinting or casting after surgery is advocated(6).

2.3.3 Occupational therapy intervention

Therefore one of the roles of occupational therapy in children with arthrogryposis is the use of splinting. Splinting is not only used after surgery but also initially in the form of serial splinting to position joints, improve functional range of motion and enhance muscle function(9, 23). Splinting should be used with caution, taking the child's age and intellectual ability as well as the family's need into consideration as this will affect the level of compliance(6). The younger infant under six month's of age more readily accepts a thermoplastic splint but as the child's skin integrity is not as good skin breakdown can occur. Therefore splints should be closely monitored during this time.

Splints should be difficult for the child to remove and should initially only be used for short periods of time. The older the child becomes the more difficult is it to apply splints as a child between six to 18 months is aware of strangers and splinting is often accompanied by crying and squirming. Toddlers do not like to be restricted and it becomes difficult to keep splints on for the necessary length of time. Children between four and seven years are more task orientated and become more compliant as they begin to understand and follow rules(6).

Different types of splint are prescribed, depending on the joints that need to be addressed and the age of the child. A full hand splint provides wrist, finger and thumb support and improves extension. Materials like elastomer putty can be inserted to decrease ulnar finger deviation. A functional wrist splint is usually applied, either dorsally or volarly, depending on the amount of support that is required. It still allows for functional finger movement whilst maintaining wrist extension and is good for day time wear. Dynamic wrist extension splints can be used to oppose the deformity by giving resistance during

active flexion and promoting extension but should be supervised and only used for short periods of time. Elbow splints are used to promote either flexion or extension depending on the type of deformity. When placed posteriorly it promotes elbow extension and when placed anteriorly pulls the elbow into flexion(34).

Splinting is an adjunctive modality used by occupational therapists in aiming to improve function in activities of daily life. They therefore also need to assess and treat all aspects of hand function including strength, sensation, bilateral coordination, grasps and the functional use of the upper limb(6).

Occupational therapists may be involved in facilitating normal development and assisting in the development of neck and head control, the ability to roll, independent sitting, crawling, standing and walking. Because the physical limitations of these children will hinder normal movement patterns, compensatory movement patterns are encouraged and promoted as long as it contributes to a functional outcome. Less used muscle groups are still targeted and strengthened through various therapeutic techniques(34). Intrinsic hand muscles are strengthened using various activities incorporating different mass and precision grasps against graded amounts of resistance. As thumb opposition is necessary for most functional grasps the strengthening of the opponens and thenar muscles will be a priority. The use of an inter digital grasp, common in arthrogryposis, will be discouraged as more functional grasp patterns are facilitated in therapy(34).

Literature warns that it doesn't help to only assess the physical aspects of hand function, as the assessment of how the hand is used in functional activities is necessary for prioritising treatment and guiding families. As these children have musculo- skeletal abnormalities, normal prehension patterns may never be obtained, but they may be functional by using compensatory movement patterns or assistive devices(31).

Therefore essential outcomes that should be addressed and facilitated in occupational therapy are those related to independence in all the occupational performance areas. Personal management or selfcare are addressed first with feeding initially the most important developmental activity. Hand to mouth movements are encouraged in therapy and during play to prepare the infant to self feed(6). During early infancy the caregiver will also be taught how to provide the child with adequate postural support to make feeding easier. If the child finds grasping the utensils difficult it may benefit from built up foam handles or the use of an universal cuff. Modifications such as long handled, bent or rotated utensils can make hand to mouth movements easier and decreases the need for pro and supination. Scoop dishes with elevated sides can help children who struggle to scoop food onto a utensil. Non slip mats can assist plates from moving if the child cannot

stabilise with the non preferred hand. A child who does not have adequate elbow flexion for hand to mouth movements can be taught to flex the trunk forward to reach the hand or be provided with an elbow block to position the hand closer to the mouth.

As children with arthrogryposis struggle to pick up cups for drinking, straws can be used or cups with handles depending on their degree of hand function. As a child enters adolescence food preparation becomes part of its functional daily skills. Positioning of food and kitchen utensils is important so that they can reach and grasp with success. Assistive devices like special knives, openers and chopping boards are designed to promote independence in the kitchen and compensate for insufficient grasp strength(34). Dressing independence is another therapy aim and the dressing activity is broken up into steps and practiced and redirected until success is achieved. The young child needs immediate gratification and relies on external encouragement to remain motivated to perform a dressing activity(6). Pull-on loose fitting shirts and pants with elastic waist bands make dressing easier. Clothes can be modified to compensate for inadequate hand function by using velcro tabs behind buttons, zipper pulls and velcro shoes. Dressing aids like sock aids and shoehorns can be used if a child cannot reach its feet or does not have adequate hand strength to put on socks and shoes. For those who can afford it a dressing frame can assist a child with dressing the lower and upper extremities(34).

A child with arthrogryposis must be encouraged to be independent in personal hygiene tasks. If an infant cannot maintain an upright position in a bath a bath support can be fitted promoting alignment. If a child cannot transfer into and out of the bathtub independently grab bars, bath or shower benches, swivel chairs or hand-held showers can be recommended.

A soap dispenser may be accessed more easily than a normal bar of soap and a long handled sponge can be used for washing purposes. Hair care can be made easier by providing the child with long handled bent hairbrush. Levers on taps are easier to use than normal taps. Even toothbrush handles can be built up to promote easier grasp. Electric toothbrushes are easier to use as it decreases the need for rigorous up and down and side to side arm movements(34).

Toileting independence is crucial as schools are reluctant to accept children who require assistance with toileting tasks. Toilet transfers can be made easier by fitting a step and handrails or grab rails next to the toilet. The management of clothing is a big challenge and clothes should be adapted with zipper-pulls, elastics, Velcro tabs or material loops to enable the child to pull down his pants and underwear and to pull it back up again. Girls can be encouraged to wear skirts or dresses which make toileting easier. Because of

limited joint movement the child should be taught to wipe from front to back and the toilet paper should be placed within the child's reach. School and public restrooms frequently used by a child with arthrogryposis should be adapted to make toileting independence possible(34).

As the child gets older they have to become more independent in personal management tasks to make school attendance possible. Functional school related fine motor skills like cutting and drawing is then addressed to make the child ready for more formal schooling(6). To assist with writing the child can be provided with an adaptive writing aid. Children with severe restrictions in hand function can be given adapted writing and drawing aids that can be accessed using the mouth. Computers can be readily accessed by children with arthrogryposis whether using a mouthwand, a keyguard, an on screen keyboard or a trackball or Headmaster Plus. Speech recognition can be used if a child cannot physically access a computer(34).

Other therapy aims will include independence in communication and mobility. Often assistive devices are called for depending on the need of the child and these are utilised and adapted to enhance independent function as these patients have normal cognitive abilities and are able to access these devices(2, 6, 23). Occupational therapists also make use of support groups where families with children with similar conditions can support and learn from one another(6).

2.4 Outcomes of Arthrogryposis

2.4.1 Overall outcomes

Surgical outcomes vary, depending on the type and severity of the deformity. A long-term study of fifty-two patients showed that flexion contractures of the hips were more successfully treated than extension contractures and only six of the fifty-two patients were wheelchair bound, the other being able to walk independently or with walking aids like crutches. Upper limb surgery was aimed at elbow function. Out of 13 surgeries done, 5 were successful but the rest failed. Only two patients received wrist surgeries, and mostly the degree of function was achieved by compensation, and not as a result of normal range of motion or movement patterns(30).

Research has shown that there is no link between the number or type of contractures and the intellectual outcome of these children. The same study mentioned above used the Wechsler Intelligence Scale for children and adults and the average intellectual score of this population was slightly above average. This study further showed that 35 of the fifty-two patients attended a normal school and six attended a school for learners with special

educational needs, the rest not being at school going age at the time of the research (30). Another review of 38 patients with arthrogryposis showed that out of the 15 school aged children, 13 could read and write, the other two being undocumented(29).

However, literature shows that children with any disability are at a greater risk of developing psychological difficulties, although no formal research has been done on arthrogryposis to predict the psychological outcome(35). Family involvement and acceptance is key as most children continue to depend and live with their families into adulthood(29, 35).

In general the prognosis of these individuals is good, with 94 percent reaching adulthood and, with no additional complications, a life expectancy to middle and late adulthood is possible(23). Retrospective research studies have shown that children with arthrogryposis can graduate from mainstream schools and go on to get a tertiary education. Some patients marry and have children and are employed, all depending on the degree of their disabilities and the success of their intervention(30, 36). The literature also advises that the functional outcome in adulthood is not as dependent on the number of physical limitations, but rather on personality, coping skills and education. This article emphasises the importance of caregiver education and assistance in the management of these children(36).

2.4.2 Functional performance outcomes

Even if surgical or other types of intervention is not successful, the child with arthrogryposis may be able to compensate for his limitations and in using alternative methods to execute tasks may still achieve a good functional outcome(14). According to a study done by Carlson et al, and Hahn(37, 38) with intervention children with amyoplasia can have no limitations in the execution of activities of daily life, achieving reasonable independence. They used a sample size of 34 patients with an age average of 27 years, and 70% of their sample achieved reasonable independence in activities of daily life. The sample was however made up out of different subgroups of arthrogryposis due to limited numbers which makes the interpretation of the results more complicated (23, 29).

Another study done by Sells et al. reviewed the records of 38 children and found that patients with amyoplasia still may require assistance from carers in adulthood as while they are mostly independent in feeding, they are less independent in grooming, dressing and bathing activities. All the children from their sample still resided with their parents. They however, did not use a standardised assessment but coded function according to levels of independence and warned that the results may vary as children of different age groups were included in the study(23, 29).

Their results were similar to those in a study where the functional outcome of a sample of 28 children with arthrogryposis using the modified Stanford Health Assessment Questionnaire for Children indicated that 58% achieved functional independence, 25% had major difficulties and 17% required total assistance(28). A long term retrospective study of 52 patients by Carlson et al found even fewer, only 46% of older children with arthrogryposis reached full independence in activities of daily life, 10 % had some limitations in tasks required fine motor dexterity and 8% required full assistance from carers(36).

It must be emphasized that all the patients in the above-mentioned studies had received intensive occupational or physiotherapy intervention, most starting soon after birth. They also had rigorous surgical intervention although in all of the participants in this research most surgeries were done on the lower limbs to improve ambulatory function(28, 29). Eighty percent of patients in one of the above mentioned studies continued receiving therapy as teenagers and with ongoing orthopaedic and therapeutic intervention 85 percent of this population were independent in mobility and the execution of activities of daily life(2, 28).

A child with functional limitations may require adult assistance to execute some functional tasks of everyday life, but can also make use of assistive devices to become more independent. A review of 96 children found that these children used functional adaptation patterns when executing tasks and only relied on simple assistive devices like anchor mats, scoop dishes, rocker knives, grip handles and inverted cutlery for feeding; reaching sticks, button hooks, shoehorns, dressing frames and elastic shoe laces for dressing; utility brushes and comb sticks, long handled bath-brushes, suction cup nail clippers and toilet paper holders for personal hygiene tasks. More complex assistive devices were not used and soon discarded(37). Five out of 27 patients in the above mentioned study could live independently in an adapted house, thus relying on assistive devices and structural adaptations(30).

Literature on managing children with arthrogryposis advises health care professionals to design and modify mechanical aids, adjust chair and table heights, make use of assistive devices for feeding, dressing and personal hygiene, and adapt utensils and clothing to encourage independence(39).

To assess the functional outcome of these children both hand function and actual task execution should be taken into account. Research has indicated that that which a child can do in a clinical setting considered capacity by the International Classification of Functioning (ICF) may differ from what that child actually does in the home environment

or in what the ICF considers their actual performance as a multitude of factors affect this. Functional assessments should thus be paired with caregiver questionnaires as done in a study on children with congenital transverse reduction deficiencies(40). No published previous studies could be found that use standardised assessments to ascertain the level of participation of children with congenital hand abnormalities, as most rely on interviews and questionnaires of the child's functioning in various contexts(31).

2.5 Assessment of Function – Personal Management

When assessing a child's independence in various functional areas, gaining information from carers, teachers and observing the child's ability in various settings is important. Independence is not only achieved when the child can execute the specific task, but he or she should also be able to collect what is needed before executing the task. As parents can be biased or have a skewed perception of their child's ability these results may not be accurate and a more standardised assessment process is called for, hence the use of criterion or norm -referenced assessment tools(31).

The disc-o-gram was developed by Mennen to document the functional ability of children with arthrogryposis as well as any improvements through surgical and non-surgical intervention methods. On the disc-o-gram, all joints movements of the body is documented in terms of range of motion and functional ability using the DASH (disability of the arm, shoulder and hand) measure, which is a self-report questionnaire and not designed specifically for use in the paediatric population, or targeting all aspects of self-care skills(41, 42).

There are a few more comprehensive assessments available to assess a child's level of function in self-care tasks such as the American Association on Mental Retardation (AAMR) Adaptive Behaviour Scale that includes an independent functioning domain, the Assessment of Motor and Process Skills (AMPS) that looks at the underlying skills necessary to perform a self-care task, the Battelle Developmental Inventory that looks at grooming, toileting, dressing and eating independence and the Functional Independence Measure (FIM) that was specifically developed for children with physical disabilities similar to the Paediatric Evaluation of Disability Inventory used in this study(11).

2.5.1 The Paediatric Evaluation of Disability Inventory

The Paediatric Evaluation of Disability Inventory (PEDI) consists out of a function test and a questionnaire and is a standardised assessment that measures a person's capacity to perform an activity as well as the actual performance of the activity up to age seven and a half years(43). Personal management is assessed with Functional Self-Care Scales which

include self-care tasks like feeding, the use of utensils, personal hygiene, dressing and toileting tasks(44).

The PEDI also has a Caregiver Assistance Scale which measures the amount of assistance provided by the caregiver during a given task as well as a Modifications Scale which describes the types of modifications used by the child or the caregiver to execute the task(43). The inventory has been designed to be used in a mildly, moderately and severely disabled population and as it is helpful in identifying developmental delays it was standardised on a normative sample(43).

The psychometric characteristics of the PEDI include excellent internal consistency using Cronbachs' alpha within three Functional Skill Scales: self-care ($\alpha=0.99$), mobility ($\alpha=0.97$), social function ($\alpha=0.98$) and three Caregiver Assistance Scales: self-care ($\alpha=0.97$), mobility ($\alpha=0.95$), social function ($\alpha=0.95$) and inter-rater/intra-rater reliability with intraclass correlation coefficients of 0.95-0.99. Inter respondent reliability scores are not as high which is why only the researcher administered the test during the study. Strong correlations were found between mean scale scores and the child's age for both functional skills scale and caregiver assistance scales indicating good construct validity. Good concurrent validity has been shown with the Battelle Developmental Inventory Screening Tests (BDIST) ($r=0.73$, disabled – $r=0.70$, non-disabled – $r=0.81$. PEDI caregiver assistance)(45).

The PEDI was designed for use with young children with a physical disability, and is not limited to one specific condition, but can be used to assess any limitations resulting from a physical disability(46). The PEDI was the assessment of choice in a study assessing hand function in various samples of children with musculoskeletal dysfunction including those with congenital deformities in the form of radius deficiencies(18). The PEDI was also used in a study by Jane Case-Smith(47) to assess hand function including tactile discrimination, defensiveness, grasp and strength in a population of 40 preschool children (31). Children with Osteogenesis Imperfecta were assessed using the PEDI measuring the effect of spondylolysis on their functional ability(48). Although successful in assessing functional independence in the above mentioned populations it has not yet been used in children with arthrogryposis.

There is no published data on the validity of the PEDI in the South African context but it has been used in one South African study, on a population of children with Cerebral Palsy (49).

CHAPTER 3

RESEARCH METHODOLOGY

This chapter describes the research design used, the selection of the participants and data collection process as well as all ethical considerations taken into account during this process. The data analysis used is also explained.

3.1 Research design

For this study a quantitative cross-sectional design (immediate ex post facto comparisons) was used. Cross-sectional studies involve observation of all of a population, or a representative subset, at one specific point in time. The purpose of this design was to measure the study population in terms of some immediately measurable dependent variable(50). In this study, children with arthrogryposis were assessed once and compared to a normative population in terms of functional independence in self-care using a standardised test.

The outcome variable in this study was the independent execution of personal management tasks. The independent variables were the age groups. All these variables were measured at one time. The researcher compared these variables to draw conclusions regarding the relationship between the independent functioning of children with arthrogryposis in different age groups. This enabled the researcher to compare functional independence between age groups, as well as against a normative sample in various functional activities related to personal management.



Figure 3.1 Diagrammatic representation of the research procedure:

3.2 Selection of participants

3.2.1 The study population

Previous studies done on congenital hand deformities noted that a small number of participants are common in studies with uncommon congenital deformities(51). Due to the rarity of this condition the methodology described in the original research proposal underwent slight changes to make the study more viable. It was decided in order to complete statistical analysis that an attempt would be made to obtain a sample of 30 children under the care of an occupational therapist in the Witwatersrand. As the entire population of children with arthrogryposis in the Witwatersrand area was half of the sample size required the Tshwane area was also included to increase the number of participants. Some of the inclusion and exclusion criteria were found to be too limiting so these were altered during the data collection procedure to increase the sample size.

The total population of children with arthrogryposis in the required age groups available in the Witwatersrand and Tshwane areas under the care of an occupational therapist was sought. Total population purposive sampling technique was used and the entire population of these children were recruited into the study. After contacting all public hospitals and public LSEN schools in the Witwatersrand area, a population of 16 children was identified, with the assistance of the heads of the various Occupational Therapy Departments of the schools and hospitals detailed in Table 3.1.

Hospitals that specialised in specific diseases like tuberculosis and tropic diseases as well as psychiatric institutions were not included in the study as they do not treat congenital physical or orthopaedic conditions. After all the hospitals were contacted it was evident that all patients with arthrogryposis were either referred to a secondary or tertiary hospital where more specialised health care could be obtained, rather than being seen at a primary health care level, thus following the guidelines set out by the Department of Health(52).

Table 3.1 Identification of possible participants at public hospitals and public LSEN schools in the Witwatersrand area

Government Hospitals	Number of possible participants	Government LSEN schools	Number of possible participants
Chris Hani Baragwanath	2	Hope School	1
Coronation Hospital	0	Forest Town	0
Dr Yusuf Dadoo	0	West Rand CP school	2
Edenvale Hospital	0	Ezibeleni School	0
Far East Rand Hospital	0	Ithembalihle	0
Germiston Hospital	0	Muriel Brand	0
Heidelberg Hospital	0	Frances Vorweg	0
Helen Joseph Hospital	0		
Charlotte Maxeke Academic Hospital	11		
Kopanong Hospital	0		
Leratong Hospital	0		
Polosong Hospital	0		
Sebokeng Hospital	0		
South Rand Hospital	0		
Tambo Memorial Hospital	0		
Tembisa Hospital	0		
Total number of possible participants	13	Total number of possible participants	3

Schools that catered for learners with physical and intellectual disabilities were not contacted as cognitive delays were one of the exclusion criteria of the study.

As only 16 possible participants could be identified in the Witwatersrand area the study was broadened to include the total population in the Tshwane area and a further 11 possible participants were identified from the following hospitals and schools (Table 3.2).

Table 3.2 Identification of possible participants at public hospitals and public LSEN schools in the Tshwane area

Government Hospitals	Number of possible participants	Government LSEN schools	Number of possible participants
Dr. George Mukhari	6	Pretoria CP school	2
Kalafong Hospital	1	Nuwe Hoop school	0
Jubilee Hospital	0		
Steve Biko Academic Hospital	2		
Mamelodi Hospital	0		
Pretoria West Hospital	0		
Tswane District Hospital	0		
Total number of possible participants	9	Total number of possible participants	2

One parent who heard about the study through word of mouth contacted the researcher asking whether her daughter could be included in the study. As she met the inclusion criteria she was included as a possible participant, which resulted in a total of 28 possible participants.

3.2.2 Inclusion and exclusion criteria:

Initially inclusion and exclusion criteria were set that limited the sample size. These criteria were changed and participants who had undergone previous surgery were included, since most of the participants had lower limb surgical intervention for clubfoot correction. Initially the inclusion criteria required that the child should have had six months of occupational therapy but this time was shortened to three months to make the inclusion of younger participants possible. The age for inclusion was changed from 12 months to include participants from six months of age. Most of these participants only started receiving occupational therapy intervention at the age of three months which would put all of them under the care of an occupational therapist for three months only. The final criteria used for the study were:-

Inclusion criteria

Children

- under the care of an occupational therapist for a minimum period of three months, in the Witwatersrand or Tshwane area
- between the ages of six months and seven and a half years,

- with a diagnosis of arthrogryposis, made by a medical practitioner,
- whose parents/legal guardians gave consent for them to be included in the study.

Exclusion criteria

- children with a neurological cause to the condition impacting on cognitive function as diagnosed by a medical practitioner.

Even though the inclusion criteria stated that children between the ages of 6 months and seven and a half years can be recruited for the study, there were no participants between 5 years 9 months and 7 years 6 months available to participate in the study. All other demographic and medical differences between the subjects were documented and taken into account during data analysis.

3.3 Measurement techniques

3.3.1 Demographic questionnaire (Appendix A)

The questionnaire, developed for the purpose of the study, had questions regarding the personal information of both the participant and the caregiver, so that participants could be contacted at the parent's request. A code was allocated to each participant and marked on the questionnaire to ensure confidentiality. The demographic questionnaire aimed at taking all contextual factors into account as it would impact on the functional outcome of the participant. Questions regarding birth, health and development of the child were included as this would impact on functional performance outcomes. Questions regarding schooling and whether the participant had access to private or public health services were included to determine whether there was a difference in the functional outcomes of different demographic groups. Specific questions related to Occupational Intervention received and surgical intervention done or planned were also included as this would impact on the functional outcome of the participant, and this study was aimed at specifically guiding Occupational Therapy intervention. For this reason Occupational Therapy was divided into splinting, group sessions or individual sessions. Exactly what therapy aims were addressed under group and individual sessions was outside the scope of this study as this study involved the parents and not the treating therapists.

3.3.2 Paediatric Evaluation of Disability Inventory (PEDI)(Appendix B)

The outcome measure used in this study was the Paediatric Evaluation of Disability Inventory. This test was developed to measure development and the functional skills of

children with disabilities. The test is divided into three categories: self-care, mobility and social function. The test was designed for children between 6 months and 7.5 years of age and can be used for any diagnosis resulting in functional problems(45).

The PEDI is made up of three different measurement scales.

Part I of the inventory assesses discrete functional tasks through 73 self-care items. Skills were rated as 0 (unable, or limited in capability) or 1 (able to perform the skill, or beyond the level). The mobility and social function sections are scored in the same way.

Part II, Caregiver Assistance, assesses 20 complex functional activities and was rated on a scale between 0 (total assistance) and 5 (independent)(45). This was the parent interview section of the inventory. The criteria for every score from 0 to 5 are set out under each item in the PEDI manual to eliminate subjectivity.

Part III or Modifications, assesses the child on 20 complex functional activities and was rated as N (no modification), C (child-oriented modifications), R (rehabilitation equipment), or E (extensive modifications). A description of what is meant by no modification, child-oriented modifications and extensive modifications under each item was given to simplify scoring and eliminate subjectivity. This was the therapist and parent interview section of the inventory.

For the purpose of this study only the self-care domain was used. This domain was made up of 73 self-care tasks and rated the child's ability as either unable or capable. Parts II and III of the self-care domain were also administered to ascertain the amount of caregiver assistance and modification needed during self-care task execution.

There are several methods to administer the PEDI, dependent on what specific domains are relevant to the particular child. Methods include administration by parent report, administration by professional judgment, and administration by a combination of methods(45). For the purpose of this study the following items were administered by the researcher and was observed and scored using professional judgement:

- use of utensils (B);
- use of drinking containers (C),
- handwashing (G),
- pullover/ front-opening garments (I);
- fasteners (J);
- pants (K);
- shoes/socks (L)

The researcher set up the following toolkit to ensure that all the items were administered in the same manner using the same materials to eliminate bias. Where the participants' age group was of such that no independence in any of these tasks could be expected, the researcher observed the assistance the caregiver gave to the participant and the child's response when they were very young as per the standardised assessment in the execution of these tasks (i.e. lifts arm to be dressed at 1 year).

Use of utensils (B)

A medium sized fork, spoon and child-safe blunt knife were used to assess use of utensils. The child was given a variety of food items including chips, apple quarters, cheese cubes, bread, butter and jam and yogurt. The researcher observed the child whilst finger feeding the chips and apple quarters, using a spoon to eat the yogurt, using the fork to eat the cheese cubes and a knife to butter the bread and cut it.

Use of drinking containers (C)

The child was provided with a medium sized juice bottle and a medium sized plastic cup from which to drink. The child was asked to pour from the juice bottle into the cup. Infants where observed drinking from their bottles which the caregiver was requested to bring with to the assessment.

Handwashing (G)

The child was provided with a bar of soap and a washbasin with a tap. A towel was available to dry their hands after hand washing. If the child was unable to reach the wash basin a step was provided to stand on.

Pull over/front opening garments (I)

The clothing that the child was dressed in was used for the assessment of this item. A back up T shirt and front opening shirt was available if clothing was not appropriate for this item.

Fasteners (J)

A special doll whose clothing was adapted to include a zip, snap buttons and medium sized buttons were provided for the assessment of this item. If a child had clothing on with any of the above mentioned fasteners, the fastening thereof was also included in the assessment.

Pants (K)

The pants that the child had on were used for the assessment of this item. A back up pair of pants with an elastic waist was available if the child's clothing was not appropriate for this item.

Shoes and socks (L)

Caregivers were asked to bring children dressed with socks and shoes to the assessment. A back up pair of Velcro shoes and shoes with shoelaces were available to assess the fastening of these if the child's own shoes were not appropriate.

The questions were only answered as capable or unable, thus eliminating any room for subjectivity.

The rest of the items were administered by parent report namely

- food textures (A);
- toothbrushing (D);
- hairbrushing (E);
- nose care (F);
- washing body and face (H);
- toileting tasks (M);
- management of bladder (N);
- management of bowel (O)

The researcher completed the assessment by questioning the caregiver on the items administered by parent report. The time required for administration of the PEDI can be from anywhere between 20-30 minutes when administered by an experienced therapist, and took the researcher approximately 30 minutes(45).

Scoring

In Part I: Functional Skills all the items scored as 1 (capable) were added up to get a self-care domain sum. In Part II: Caregiver assistance, the scores ranging from 0 to 5 for each item was added up to get a self-care sum. In Part III: Modification scale the number of N's (no modification), C's (child-oriented modifications), R's (rehabilitation equipment), or E's (extensive modifications) were added to get the self-care modification frequencies. The Normative Standard Scores for Part I and II were derived using the tables for the specific age group as set out in Appendix IV and V, and converted to Scaled Score using the tables in Appendix VI and VII (45).

The PEDI provides the age ranges at which 90 percent of children in their normative sample mastered these items (45).

3.4 Research Procedure

3.4.1 Recruitment of government hospitals and LSEN schools.

The researcher contacted all schools and hospitals within the Witwatersrand and Tshwane districts to ascertain which institutions might have possible participants. A meeting was arranged with the Heads of Occupational Therapy of all the hospitals and schools with possible participants. At the meeting the researcher explained the purpose of the study, the inclusion and exclusion criteria, the data collection procedure as well as the benefits and risks involved. The Heads of Occupational Therapy Departments at the various institutions were provided with information letters with a consent slip and asked to distribute these to participants that fit the inclusion criteria. Those interested in participating in the research would then consent and make their contact details available to the researcher.

Out of the possible 28 participants the following where not able to take part in the study.(Table 3.3)

Table 3.3 Reasons for exclusion of possible participants

Number of participants	Reason for non participation
4	Identified as possible participants at the beginning of the study but could not be located at time of first meeting as they relocated.
4	Comorbid medical condition resulting in exclusion
1	Age resulting in exclusion

3.4.2 Recruitment of participants.

After the contact details of the 19 participants fitting the inclusion criteria were made available to the researcher, they were phoned and a meeting arranged at a time and place that was suitable for them. This was either at the hospital during their normal occupational therapy appointment or at the school at a time where the parents would normally drop off or fetch the child. If the meeting incurred travelling costs they were reimbursed. Two appointments were made at the participant's home as travelling was difficult.

3.4.3 Data collection procedure.

Data was collected over a seven month period. The assessment consisted of a single meeting set up with the parent of caregiver at a time that was convenient for them and took about an hour to complete.

Demographic information for each participant was collected using the questionnaire and coded to ensure confidentiality. The researcher read the questions out to the caregiver to make sure that all questions were understood and captured the information on the form. At George Mukhari Academic Hospital some parents were not English literate and an occupational therapy technician (OTT) was asked to translate the questions after permission was given by the caregiver. In these instances the OTT remained in the room to assist with the translation of the instructions when the items in the PEDI were administered.

The first section of the PEDI was then administered with each child participating in the study. This was done at the various hospitals or schools, in a room made available by them for the researcher to use. The rooms differed in size but the researcher requested that certain uniform items were present in all rooms as far as possible namely a small table and chair, basin and step, small bench and carpeted area. All the children were assessed using the same sequence and the same materials.

For hygienic purposes the first item that was assessed was item G (handwashing). The participant was taken to a basin, with a step so that he/she could easily access it. All the basins had the same twist knob taps and looked similar. For younger participants and participants that could not access the basin themselves, the carers were asked to wash their hands as they would normally do so. All participants were provided with a washcloth and soap which they could use. They were provided with a hand towel to dry their hands after washing them.

For the next few items the child was seated at a small table if independent sitting was possible, alternatively they were seated on their carer's laps. Two of the participants were strapped into their seats as they were unable to maintain a seated position without external support. Item B (use of utensils) and item C (use of drinking containers) was administered in this position.

Items I (pull over/front opening garments), J (fasteners), K (pants) and L (shoes and socks) were administered either on a carpet or a small bench depending on the child's preference. The participant could assume the position they normally do during dressing tasks either, sitting on the bench, lying or sitting on the carpet, or standing up. The

participant was asked to take off or put on their shirts and pants, shoes and socks if they were appropriate, or they used the clothing items provided by the researcher. They were given a doll with different fasteners and asked to unfasten and fasten again, and if appropriate the fasteners on their clothes were used. If the participant was unable to do it independently their carers were asked to assist them as they normally would.

All observations were documented on the Functional Skills, Self-care domain form of the PEDI. Items were rated as unable or capable and observations were documented next to each item on the form.

3.5 Ethical Considerations

After ethical approval for the study was obtained from the Human Research Ethics Committee (HREC) at the University of Witwatersrand, an application for ethical approval was submitted to the Department of Education and granted (Appendix C). Permission to conduct the study was requested from the CEO of all the hospitals in the Witwatersrand and Tshwane area with the assistance of the Heads of the Occupational Therapy Departments of these institutions (Appendix D). Participants were supplied with an information letter explaining and describing the purpose and the nature of the study (Appendix E) by the heads of the Occupational Therapy Department and asked whether they would like to participate. The information sheet for both the parents and the child, explained the reason for the study in understandable terms. It also explained the nature of the research procedure. All participants were informed that participation in the study is not compulsory and that they could withdraw from it at any given time with no consequences. The parents of the participants will be able to have access to the research findings at the end of the study.

Caregivers willing to participate in the study then made their contact details available to the researcher to arrange a meeting. At the meeting the purpose and the nature of the study was verbally explained to them again and they were asked to sign an informed consent form (Appendix F) and where possible the children were also asked to give verbal assent or written assent to participate in the study (Appendix G).

Confidentiality was maintained by assigning a code to every participant instead of using their personal information. The original questionnaires with personal contact details were locked away and only accessible to the researcher. Personal information about the parent/legal guardian was recorded so that should the standardised assessment reveal delays in certain areas of self-care activities, those participants' parents could be provided with the names and contact details of appropriate services for treatment.

Data will be stored according to the Health Professions Council of South Africa (HPCSA) regulations for six years if the study is unpublished and for two years if it is published.

No parent experienced psychological distress when completing the demographic questionnaire but the treating occupational therapist that was known to them was available to support them if need be. They could then be referred to an appropriate health professional for further counselling and support. No participant experienced emotional or psychological distress, but the parents were present throughout the assessment to help alleviate any feelings of anxiety should the need arise.

3.6 Data analysis

The background information of each participant was coded and collected as nominal data e.g. age group, birth history, type of occupational therapy intervention and entered into an excel data spreadsheet. Descriptive statistics in the form of means and percentages were used to analyse this data.

Normative standard scores from the self-care domain were calculated and converted to Z scores to determine the level of functioning on the entire sample. Due to the small sample size and the heterogeneous nature of the sample for the purposes of data analysis the participants were grouped according to their ages into four respective developmental groups according to the guidelines set by the National Centre for Education Statistics(53)(see Table.3.4).

Table 3.4 Grouping of participants by developmental age groups for data analysis

Age group	Age ranges
Infants	6-12 months
Toddlers	12-36 months
Preschoolers	36-60 months
School aged	>60 months

This was necessary as the developmental milestones of personal management differ in these age groups. Self-care tasks were grouped into four categories namely eating and feeding, personal hygiene and grooming, dressing and toilet hygiene according to the Occupational Therapy Practice Framework (5). The normative scores for each age group's performance in the four categories of self-care tasks were determined using the PEDI.

Standard scores were calculated for each participant and as the tables in the PEDI only lists standard scores between 10 and 90 (54), a standard score of >90 or <10 was achieved if a participants performance was either below or above the extreme score for the child's age. This range >90 or <10 was entered as 90 or 10 into the excel worksheet to enable calculation. This procedure was used for the self-care items and the Caregiver-Assistance Scales.

The standard scores were analysed using descriptive statistics to determine an average or mean percentage for the level of function on the PEDI, in each self-care category for each age group. These percentages were compared to the expected percentage within each age group at which these self-care tasks should be mastered according to the PEDI. Children in the preschool group are expected to achieve a score indicating they have mastered 80% of the items for feeding for example, while those in the school going group should achieve a score of 100%.

Normative standard scores from the self-care domain were converted to Z scores for comparison across the age groups. A Chi squared test was used to determine if there was a significant difference between the expected level of independence of each age group as set by the PEDI and the level of independence observed on the PEDI.

The Modification Scale was analysed by giving a numerical value to each section (see Table 3.5) so that the mean for the modification count for each age group could be established.

Table 3.5 Score allocation for the Modification Scale

Modification scale:description	Numerical value
None (N)	4
Child (C)	3
Rehab (R)	2
Extensive (E)	1

Chapter 4

RESULTS

4.1 Introduction

This chapter presents the results obtained from the study. A total of 19 participants aged six months to five years nine months were recruited from Government Hospitals, LSEN schools and word of mouth were assessed using the Self-care Domain as well as the Caregiver Assistance Scale and Modification Scales of the PEDI.

4.2 Demographics

4.2.1 Gender and Age:

There were more males than females in the sample with a total of 11 male and eight female participants. When the participants were divided into the developmental age groups (Figure 4.1) a greater percentage of males were found in the infant, preschool and school going age groups.

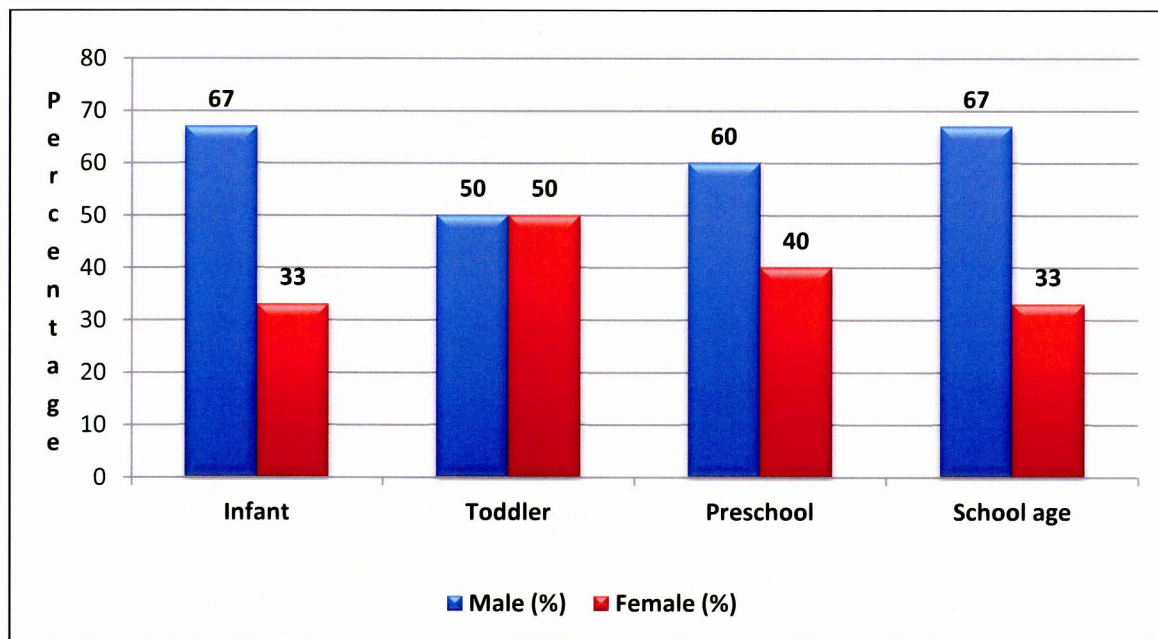


Figure 4.1 Gender of participants according to developmental age groups (n=19)

The participants' ages ranged from seven months to 5 years 11 months. Participants were not evenly divided amongst the developmental age groups with the majority of the participants in the toddler and pre-school groups. (Table 4.1)

Table 4.1 Number of participants in the developmental age groups (n=19)

Age group	Number of participants
Infants (6-12 months)	3
Toddlers (12-36 months)	8
Preschoolers (36-60)	5
School aged (>60 months)	3

4.2.2 Birth and medical history

4.2.2.1 Maternal age

Eight of the participants were born as the result of a planned pregnancy and none of the mothers reported to having smoked or used alcohol during their pregnancies. Five mothers were taking medication during their pregnancies including Anti-Retro Viral treatment, antibiotics or blood pressure medication.

The mothers' ages ranged between 19 and 44 years and the mean maternal age at the time of delivery was highest for the preschool developmental age group (Figure 4.2)

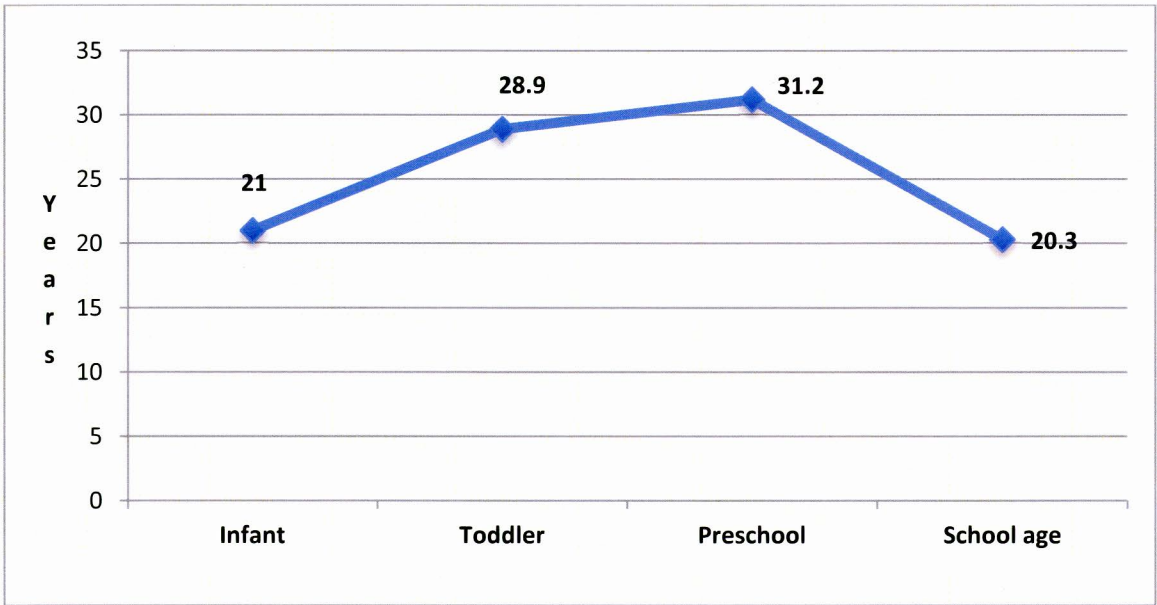


Figure 4.2 Mean maternal age according to developmental age groups (n=19):

4.2.2.2 Birth History

The mean gestational age of the participants was 39.7 weeks with only two participants being born prematurely at 31 and 32 weeks. Half of the participants were delivered by Caesarean Section and this was the most common form of delivery in all the developmental groups except the toddler group.

4.2.2.3 Previous and planned surgical intervention:

While five participants had not yet had surgery, the other 14 participants underwent some surgical procedure to correct a joint deformity. No surgeries had been conducted on any of the infant participants although most of them had non surgical orthopaedic intervention, mostly plaster of paris (POP) casting, to correct clubfeet.

The mean percentage of surgeries conducted in upper limbs and lower limbs indicate that lower limb surgical intervention had occurred most frequently in the younger age groups with upper limb intervention increasing between the preschool and school aged groups. (Figure 4.3)

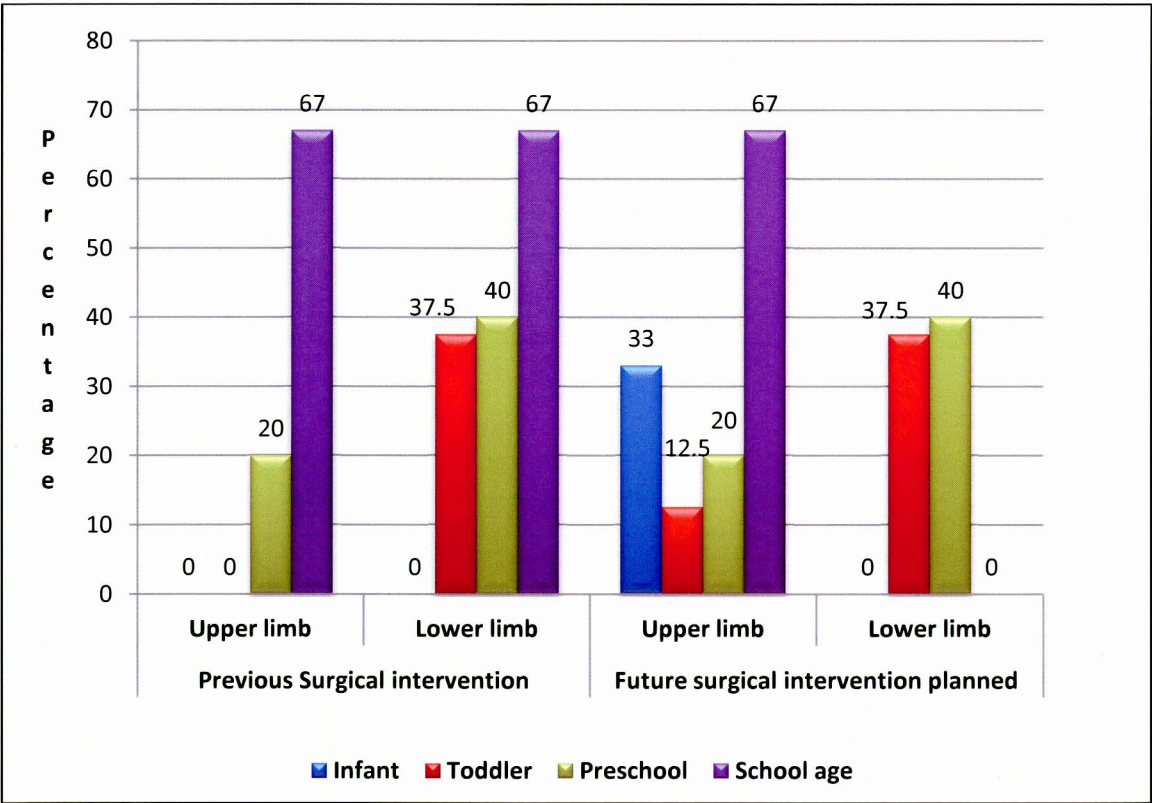


Figure 4.3 Mean percentage of previous and planned surgeries to the upper and lower limbs of participants according to developmental age groups (n=19)

Future surgery for the upper limb was planned for 5 of the participants particularly for the infants and school age children but the caregivers did not know exactly when surgery would be conducted. All lower limb surgery appears to have been completed before the children are of school going age.

4.2.3Occupational Therapy Intervention

All of the participants had had access to occupational therapy services and underwent splinting during some stage of their treatment.

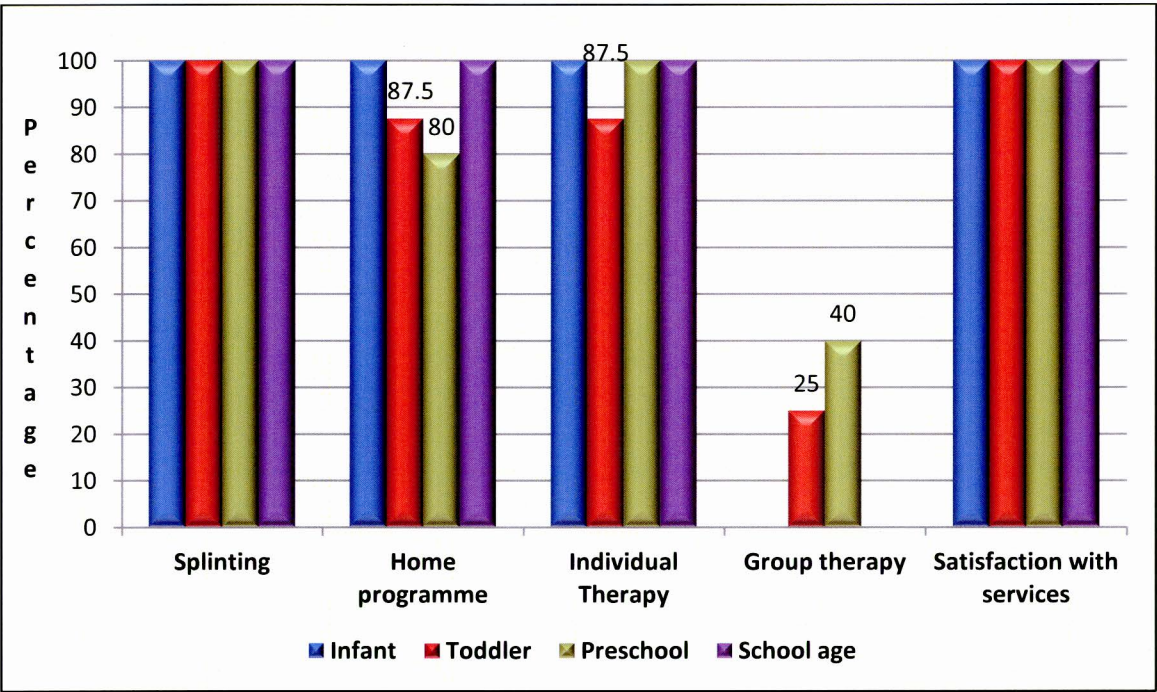


Figure 4.4 Percentage of participants involved in aspects of occupational therapy and their satisfaction with the service according to developmental age groups (n=19)

More than 80% of the participants had received an additional home programme to follow and had individual therapy with less than 40% of participants receiving group therapy. All participants reported that they were satisfied with the occupational therapy services they had received. Most satisfaction was expressed about improvements in hand function, splinting and the ability of the child to carry out activities independently.

Seventeen of the participants started attending occupational therapy in the first year of their lives and the other three before their second birthday indicating referral to occupational therapy occurs in the early developmental phase. All the participants received at least three months of occupational therapy. When asked about present dysfunction in the participants, parents expressed concerns about independence, hand

function, walking and schooling. Only parents of two participants did not express any concerns.

4.3 Level of Function on the Paediatric Evaluation of Disability Inventory (PEDI)

The data for the independence in the various self care items and how much assistance and modifications the participants needed was grouped into areas of basic activities of daily living and personal activities of daily living described in the Occupational Therapy Practice Framework (OTPF) (5) for discussion and comparison namely:

- Eating and Feeding
- Personal hygiene and grooming
- Dressing
- Toilet hygiene

4.3.1 Self-Care Domain

The functioning for the self care domain for the total group and the four developmental age groups identified for data analysis, was determined using z scores.

As reflected in the Z score progression, the infant group’s functioning is still within the norm, whereas the toddler group falls within the borderline range. The infant and toddler still rely a lot on caregiver assistance for the execution of functional self care tasks which is age appropriate. The preschoolers are more than two standard deviations and the school aged group more than 3 standard deviations from the norm.

Table 4.2 Z scores of total group of participants in the self care domain and in the developmental age groups (n=19)

	Z scores
Total group	-1.65
Developmental Age groups	
Infants (6-12 months)	-0.48
Toddlers (12-36 months)	-0.9
Preschoolers (36-60)	-2.03
School aged (>60 months)	-3.17

This indicates that as the expectancy for independence in the execution of self care tasks increases this population's ability to meet the demands decreases and their scores deviates progressively further from the norm.(Table 4.2)

The analysis of the different parts of the self care domain was then completed and the individual tasks will be discussed under the areas of basic activities of daily living and personal activities of daily living. The raw score of each participant in every item was added up and converted into a percentage using the total score that could be achieved in the item. The percentages of each participant under each age group were added together and the mean percentage calculated which was depicted in graphs. The PEDI provides the expected age of mastery of the self-care items(55), which could be compared with the actual scores of the participants.

A chi-square test was used to compare the expected percentage function of the participants with their actual mean percentage function for their total function in all aspects of personal management on the PEDI.

Table 4.3 Expected mean percentage function of the participants with their actual mean percentage function for their total function in personal management

Age group	Expected mean percentage Function (SD)	Observed mean percentage function (SD)	Chi Squared	p value
Infants (6-12 months)	4.37 (9.09)	13.04(19.21)	51.08	0 .000**
Toddlers (12-36 months)	35.48(22.45)	44.35(24.43)	48.69	0 .000**
Preschoolers (36-60)	81.46(13.37)	54.03(22.36)	221.34	0 .000**
School aged (>60 months)	100(0)	65.77(20.35)	241.82	0 .000**

Significance set at $p= 0.05^*$, 0.001^{**}

There was a statistically significant difference between the expected mean percentage function and the observed mean percentage function for all four age groups. For the infants and toddlers the observed mean percentage function was significantly higher than expected. For the preschool and school aged children it was significantly lower.

4.3.1.1. Eating and Feeding

For the purpose of this study food textures are included under eating and the use of drinking and feeding utensils has been clustered together under feeding.

Table 4.4 Mean percentages of total group of participants in the self care items of food textures, use of utensils and use of drinking containers in the developmental age groups (n=19)

Age group	Food textures Mean (SD)	Use of utensils Mean (SD)	Use of drinking containers Mean (SD)
Infants (6-12 months)	58.3(1.52)	20 (1)	13.3(0.57)
Toddlers (12-36 months)	93.75 (0.70)	52.5 (1.99)	65 (1.83)
Preschoolers (36-60)	100 (0)	48 (2.24)	44 (2.03)
School aged (>60 months)	100 (0)	66.7 (2.88)	60 (1.73)

Food textures describe the child's ability to orally take in different types of food, produce a bolus and swallow. Use of utensils describes the child's ability to use different methods to get the food and liquid to their mouths according to their age group.

Eating

The mean expected age of mastery for all textures of food is between 3.5 – 4 years of age. The participant's scores in these tasks were all age appropriate and no mother reported any adverse reaction to any type or texture of food (Table 4.4).

Feeding

Use of utensils and drinking containers were however more challenging with scores indicative of a below mean performance as all school aged children are expected to have achieved 100% in this aspect as they should be able to manage a cup, spoon, fork and knife and pour liquids into a container. Less than 70% of this sample achieved this for eating utensils and drinking containers (Table 4.4)

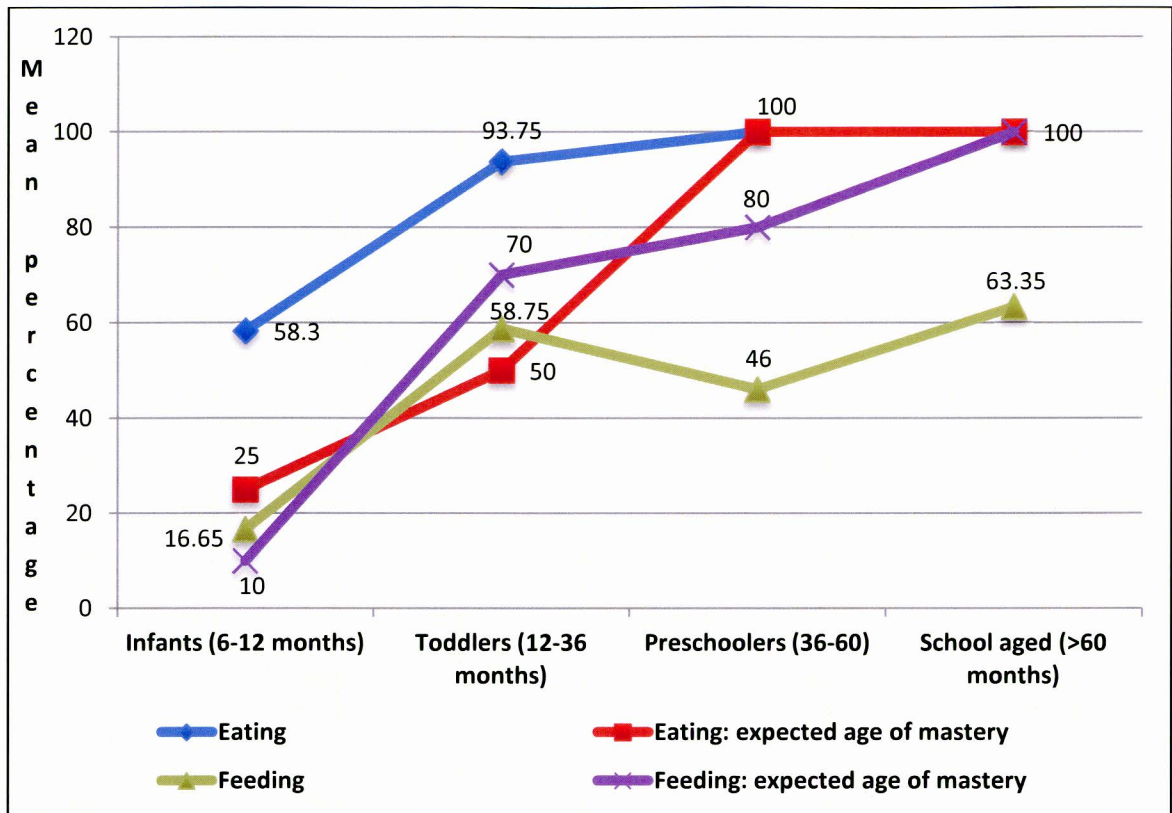


Figure 4.5 Mean percentage function of participants for eating and feeding according to developmental age groups (n=19)

Whereas there was a steady increase with age in the ability to eat food of all textures, the mean percentage scores for the use of utensils and drinking containers was lowest for infants who are expected to finger feed. While it increased for toddlers a decrease was seen when more dexterous and unilateral function was required from preschool children (Figure 4.5). The mean percentage function for school age children was just above 65%.

4.3.1.2 Personal hygiene and grooming

Personal hygiene includes all self-care items related to toothbrushing, hairbrushing, nose care, handwashing and washing of body and face.

Grooming

Grooming includes the items on tooth brushing, hair brushing and nose care.

Table 4.5 Mean percentages of total group of participants in the self care items of toothbrushing, hairbrushing and nose care in the developmental age groups (n=19)

	Toothbrushing Mean (SD)	Hairbrushing Mean (SD)	Nosecare Mean (SD)
Infants (6-12 months)	13.3 (1.15)	8.3 (0.57)	0 (0)
Toddlers (12-36 months)	57.5 (1.45)	28.1 (1.12)	70 (1.69)
Preschoolers (36-60)	60 (1.41)	45 (0.97)	88 (1.2)
School aged (>60 months)	86.7 (0.57)	41.7 (1.15)	80 (1.73)

The expected age of mastery for toothbrushing and nose care is 72-78 months and as the mean age of the children in the school aged group is only at 66 months these activity milestones could not be compared to the normative age group (Table 4.5).

The activity milestone of nosecare was however at and above 80% for the school going and preschool group respectively indicating a reasonable level of independence for these older participants.

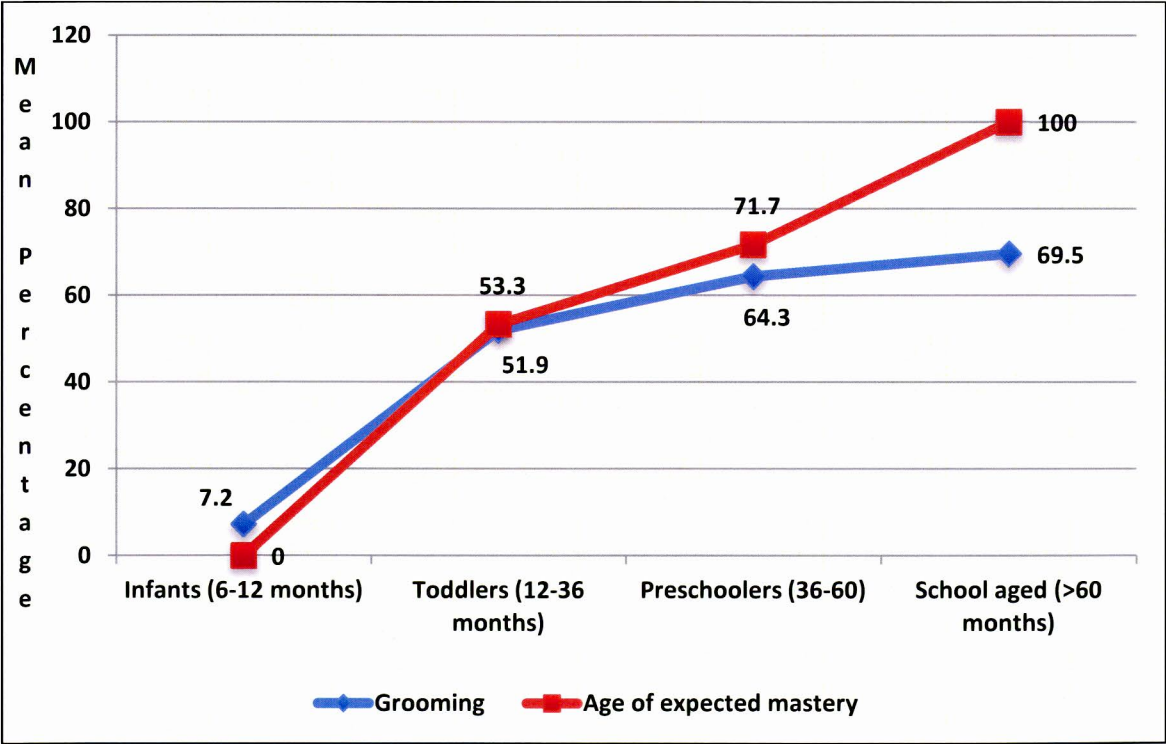


Figure 4.6 Mean percentage function of participants for grooming according to developmental age groups (n=19)

The preschool group along with the toddlers only had around a 60% level of function for tooth brushing. However this improved to nearly a 90% level in school children indicating a marked improvement with age for this activity milestone. (Figure 4.6)

Hair brushing should be mastered by 42-48 months, thus the preschool group should have achieved this milestone. The mean percentage of this group was however 45%, so they are performing well below the level expected when compared to the normative sample. A similar finding for the school age groups indicated there was no improvement in this aspect as the children got older and this aspect of grooming remained the most difficult for all participants.

Washing

More than 90% of the normative group could thoroughly wash and dry their hands between 48 and 54 months.

The preschool group in this study scored 72% in this task and is thus below the normative group. There was only a 1.3% improvement in performance between the preschool and school aged child that might suggest a plateau in performance (Table 4.6).

Table 4.6. Mean percentages of total group of participants in the self care items of hand washing and washing of body and face in the developmental age groups (n=19)

	Handwashing Mean (SD)	Washing body and face Mean (SD)
Infants (6-12 months)	0 (0)	0 (0)
Toddlers (12-36 months)	57.5 (1.80)	22.5 (0.99)
Preschoolers (36-60)	72 (1.01)	44 (1.6)
School aged (>60 months)	73.3 (1.52)	53.3 (2.08)

Children between 6 and 6.5 years (72 – 78 months) are expected to obtain soap, thoroughly wash their body and face and dry themselves thoroughly (Figure 4.7). The school aged group in this study (mean age 66 months) only scored 53.3 % on this task, indicative of a delay in independent washing. Again the percentage independence improved with age for this aspect.

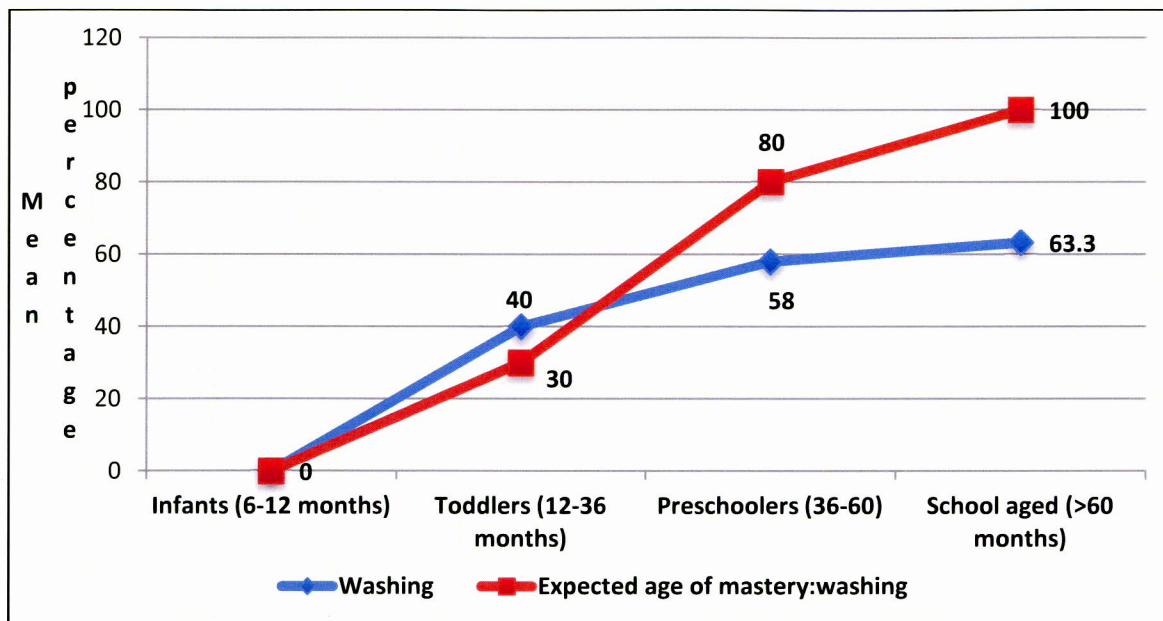


Figure 4.7 Mean percentage function of participants for washing according to developmental age groups (n=19)

4.3.1.3 Dressing:

Dressing includes all tasks related to putting on different items of clothing or garments as well as aspects requiring more dexterity like shoes and socks, and managing fasteners.

Garments

When comparing the groups' performance on front opening garments compared to pants it can be seen that their ability to manage front opening garments plateaued at a 40% level of functioning from the toddler to the school aged group with no improvement in performance (Table 4.7)

Table 4.7 Mean percentages of total group of participants in the self care items of pullover/front opening garments and pants in the developmental age groups (n=19)

	Pullover/front opening garments Mean (SD)	Pants Mean (SD)
Infants (6-12 months)	13.3 (0.57)	0 (0)
Toddlers (12-36 months)	40 (1.41)	27.5 (0.91)
Preschoolers (36-60)	40 (1.78)	32 (1.85)
School aged (>60 months)	40 (1.73)	60 (2)

The normative group could independently manage a front opening shirt at age 48-54 months and could manage the fasteners on the front opening garment at age 66-72 months, so the preschool and school aged participants had marked reduced independence for this activity milestone (Figure 4.8).

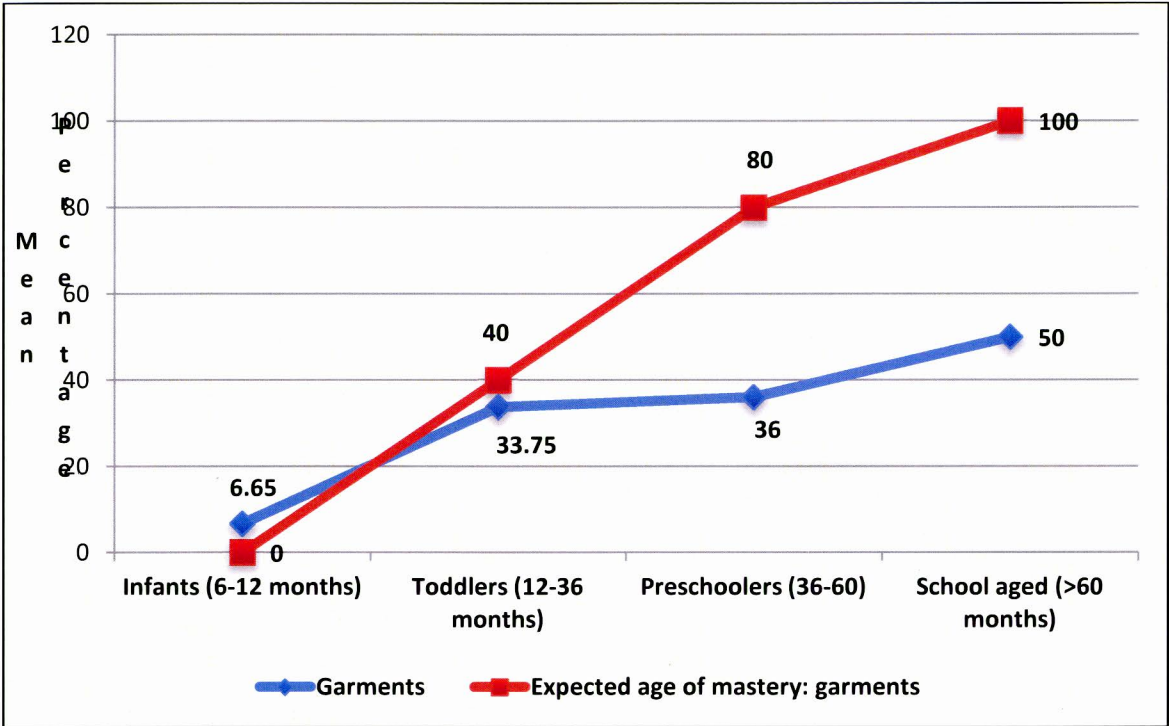


Figure 4.8 Percentage function of participants for dressing in garments according to developmental age groups (n=19)

The ability to manage pants increased with age and the participants performed better in this task reaching nearly 60% in the school aged children. According to the normative group school aged participants should be able to put on and fasten pants at 66 – 72 months. This group only scored 60% for this activity milestone and although they performed better in this task than in the front opening garment task, their performance was still well below that of the normative group.

Shoes and socks and Fasteners

By 48 months of age the normative group could put shoes and socks on, and they knew the correct foot by 6 years (72 months) of age. Although there is a steady increase in performance between all the age groups, the school aged child still performed at 50% for this activity milestone, well below the 100% achieved by the normative group. (Table 4.8)

Table 4.8 Mean percentages of total group of participants in the self care items of fasteners, shoes/socks in the developmental age groups (n=19)

	Fasteners Mean (SD)	Shoes/Socks Mean (SD)
Infants (6-12 months)	0 (0)	6.7 (0.57)
Toddlers (12-36 months)	35 (1.58)	12.5 (1.06)
Preschoolers (36-60)	44 (1.32)	36 (1.83)
School aged (>60 months)	40 (2)	46.7 (2.08)

A 72-78 month old child is able to manage all zips and buttons independently. The school aged group could not be compared to this but as they score below 50% a delay in independence for this activity milestone can be expected (Figure 4.9). There was a decrease in performance between the preschool and school aged group due to the expected competence of fine motor bilateral dexterous hand function as the child gets older (Table 4.8).

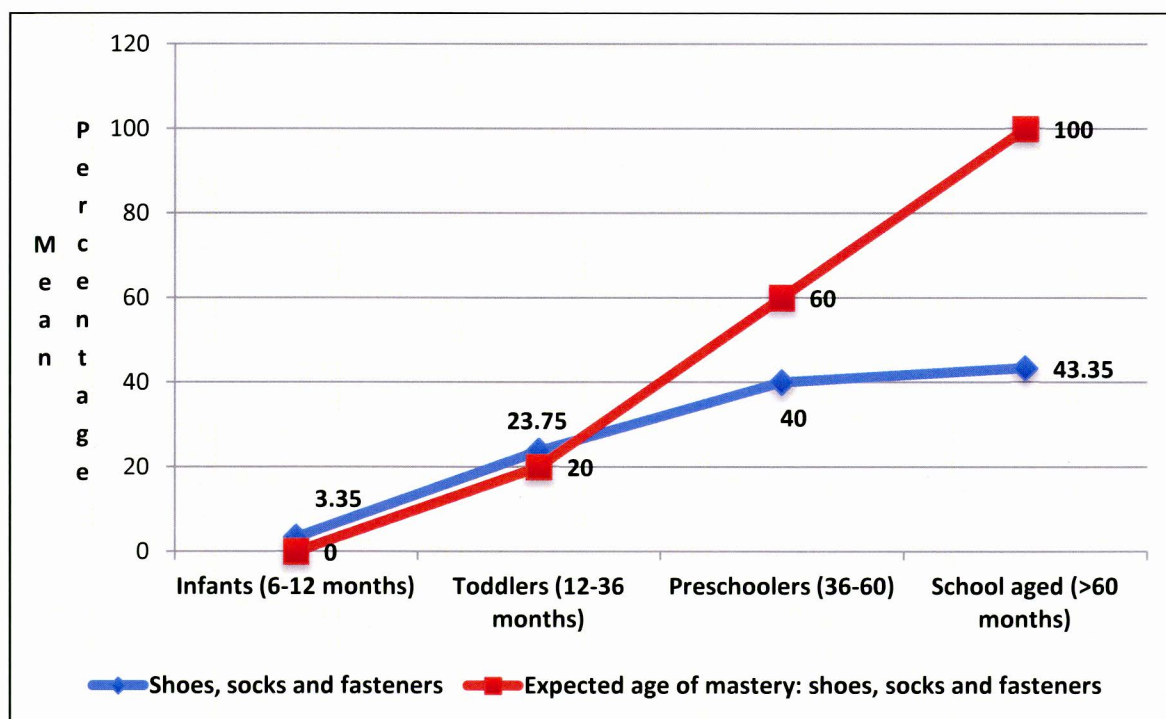


Figure 4.9 Mean percentage function of participants for dressing shoes and socks and doing fasteners according to developmental age groups (n=19)

4.3.1.4 Toilet hygiene

Toilet hygiene includes the physical task of going to the toilet, managing clothes, wiping, and using the toilet appropriately as well as the ability to manage the bladder and bowel, knowing when there is a need for toileting and staying dry.

Table 4.9 Mean percentages of total group of participants in the self care items of bowel and bladder management and toileting in the developmental age groups (n=19)

	Management of bowel Mean (SD)	Management of bladder Mean (SD)	Toileting Mean (SD)
Infants (6-12 months)	13.3 (0.57)	13.3 (0.57)	0 (0)
Toddlers (12-36 months)	32.5 (1.18)	32.5 (1.18)	17.5 (0.99)
Preschoolers (36-60)	56 (1.72)	64 (1.16)	28 (1.95)
School aged (>60 months)	86.7 (0.57)	93.3 (0.57)	46.7 (2.51)

The normative group was able to toilet independently by age 72-78 months and although there was a steady increase in the participants with age for this aspect the school aged group had achieved less than 50% in this activity milestone so a future delay in this activity could be expected (Table 4.9).

A child of 54 – 60 months should have no wetting incidents and a child of 42-48 months should have no soiling incidents. In the preschool group only 64% and 56% scored at a functional level for bladder and bowel management respectively and most of them were still wearing nappies (Table 4.9).

The functional level for these milestones has improved markedly for school aged participants with over 90% achieving independence in bladder management and 87% achieving bowel management (Figure 4.10).

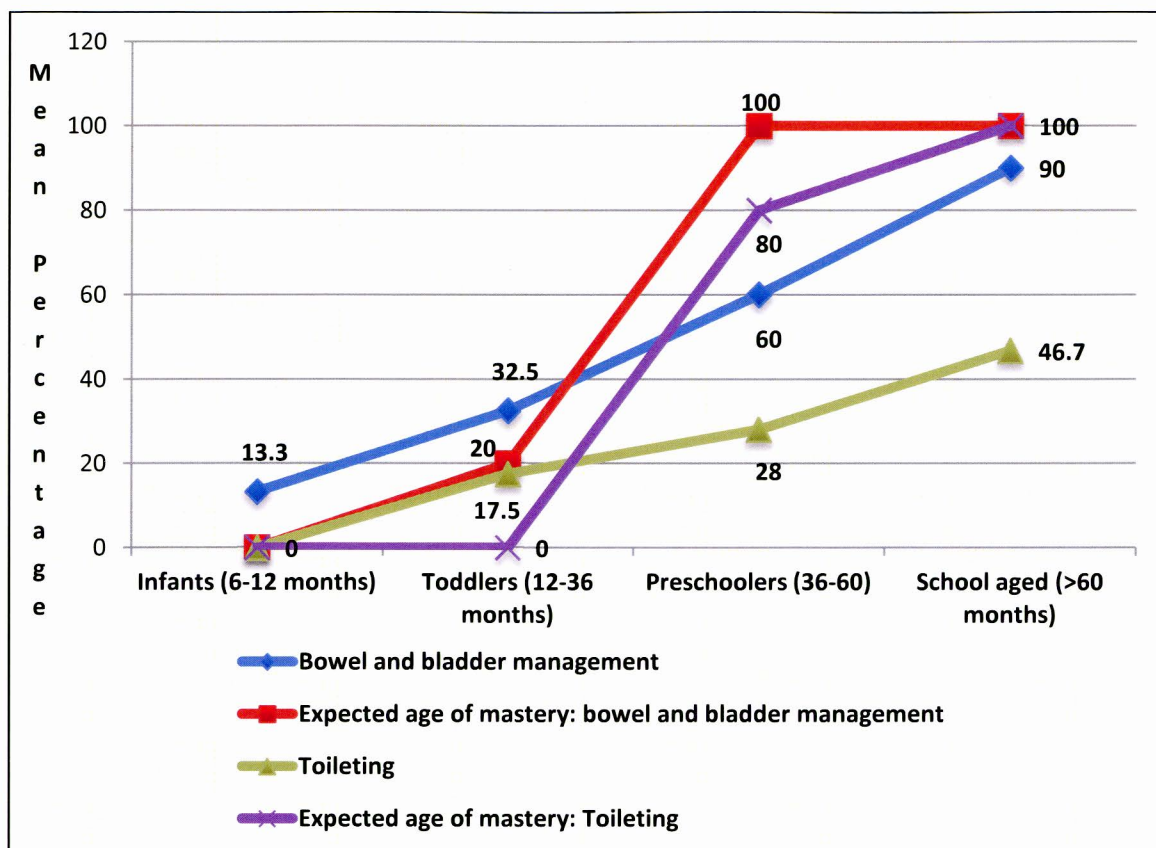


Figure 4.10 Mean percentage function of participants for bowel and bladder management as well as toileting according to developmental age groups (n=19)

4.3.2 Caregiver Assistance

The caregiver assistance scale was designed to indicate how much assistance a caregiver gives during a functional task. The less assistance given, the greater the score. High percentages thus indicate greater independence than low percentages.

Table 4.10 Z scores of total group of participants in the caregiver assistance scale self care domain and in the developmental age groups (n=19)

	Z Scores
Total group	-1.15
Developmental Age groups	
Infants (6-12 months)	0.4
Toddlers (12-36 months)	-0.28
Preschoolers (36-60)	-2.95
School aged (>60 months)	-1.78

4.3.2.1 Eating and Feeding:

While all the children were independent in eating, no need for assistance with feeding was only achieved by more than 90% of the normative group between 78-84 months of age. Thus since the school aged participants in this study had a mean age of 66 months it was not expected that they would no longer need some assistance with feeding.

As expected there was a marked increase in independence in feeding between the infant and toddler group. No change was seen between the performance of the toddlers and preschoolers but a further increase in independence in feeding was evident in the school aged child (Figure 4.11).

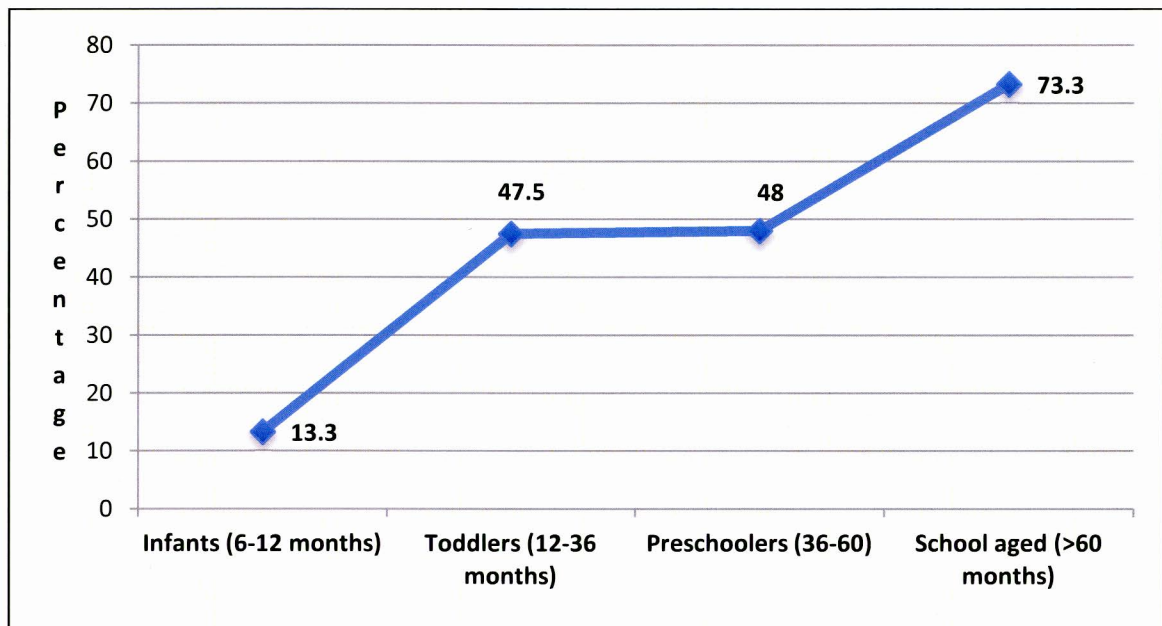


Figure 4.11 Mean percentage independence from caregiver assistance of participants for feeding according to developmental age groups (n=19)

4.3.2.2 Personal hygiene and grooming

The development of increased independence in both grooming and bathing tasks followed a similar pattern to that seen with feeding as there was an increase between the infant and toddler group, a similar performance between the toddler and preschool group, and a rapid increase again in the school aged group. (Figure 4.12)

Since both grooming and bathing tasks require no caregiver assistance above 7 years (84 months) of age, assistance can still be expected even in the school aged group in this study. The amount of caregiver assistance that is age appropriate for these participants can therefore not be determined.

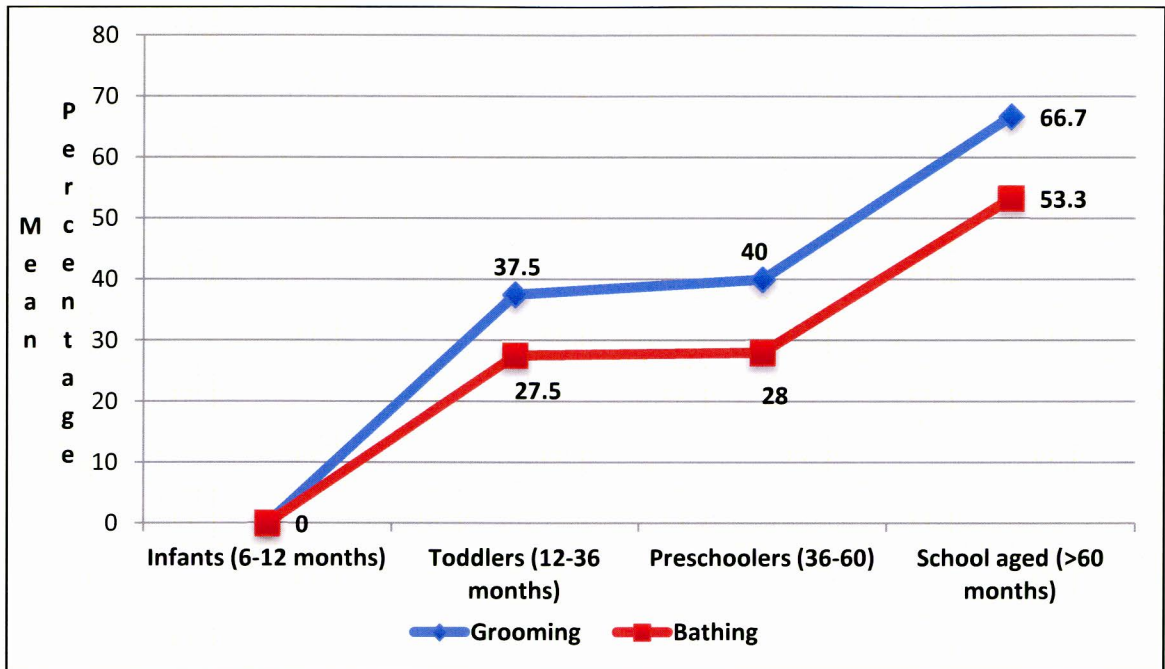


Figure 4.12 Mean percentage independence from care giver assistance of participants for personal hygiene and grooming according to developmental age groups (n=19)

4.3.2.3 Dressing

The amount of caregiver assistance needed for the dressing of upper and lower body are exactly the same in the infant and preschool group(Figure 4.13)

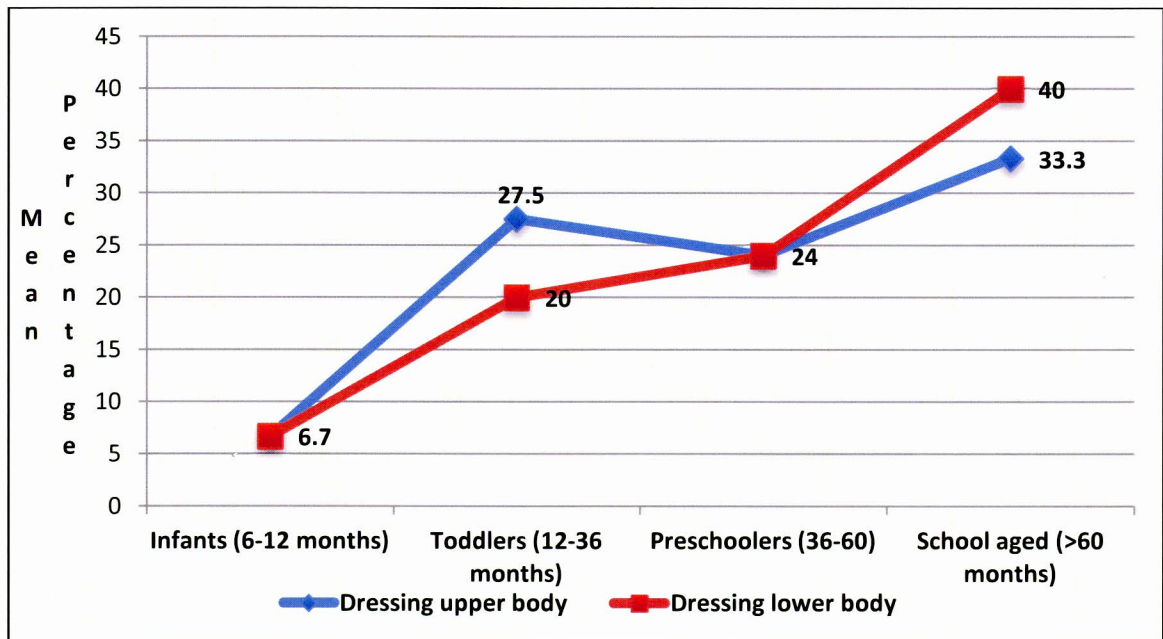


Figure 4.13 Mean percentage independence from care giver assistance of participants for dressing according to developmental age groups (n=19)

The toddler group needed more assistance with dressing their lower body, whereas the school aged group needed more assistance in dressing their upper body. This was related to the type of clothes worn in different age groups.

Independent dressing of the upper body is expected between 66-72 months and the lower body between 78-84 months. It can be seen that the school aged group still achieved a mean percentage score far below the normative group in upper body dressing indicating they required assistance from their caregivers over 76% of the time as they only had 33.3% independence in this aspect. They needed caregiver assistance 60% of the time for lower body dressing as they achieved 40% independence for this activity milestone.

4.3.2.4 Toilet hygiene

Toileting includes how much assistance is needed in managing clothing, using the toilet, cleaning up accidents and monitoring schedules.

Table 4.11 Mean percentages of total group of participants in the caregiver assistance items of toileting, bladder management and bowel management in the developmental age groups (n=19)

	Toileting Mean (SD)	Bladder management Mean (SD)	Bowel management Mean (SD)
Infants (6-12 months)	0 (0)	0 (0)	0 (0)
Toddlers (12-36 months)	17.5 (0.35)	22.5 (0.35)	22.5 (0.35)
Preschoolers (36-60)	24 (2.16)	60 (1.87)	52 (2.30)
School aged (>60 months)	46.7 (2.30)	100 (0)	100 (0)

The assistance needed for bowel and bladder management is much less than the assistance needed for toileting as toileting includes tasks like managing clothing, wiping and managing the toilet and the previous section outlining the results of the self-care items shows that the participants had difficulty in both the garments, fasteners and hand washing items. Although toileting should be independently executed by age 66-72 months, the school aged participants in this study still need a lot of assistance in this regard.

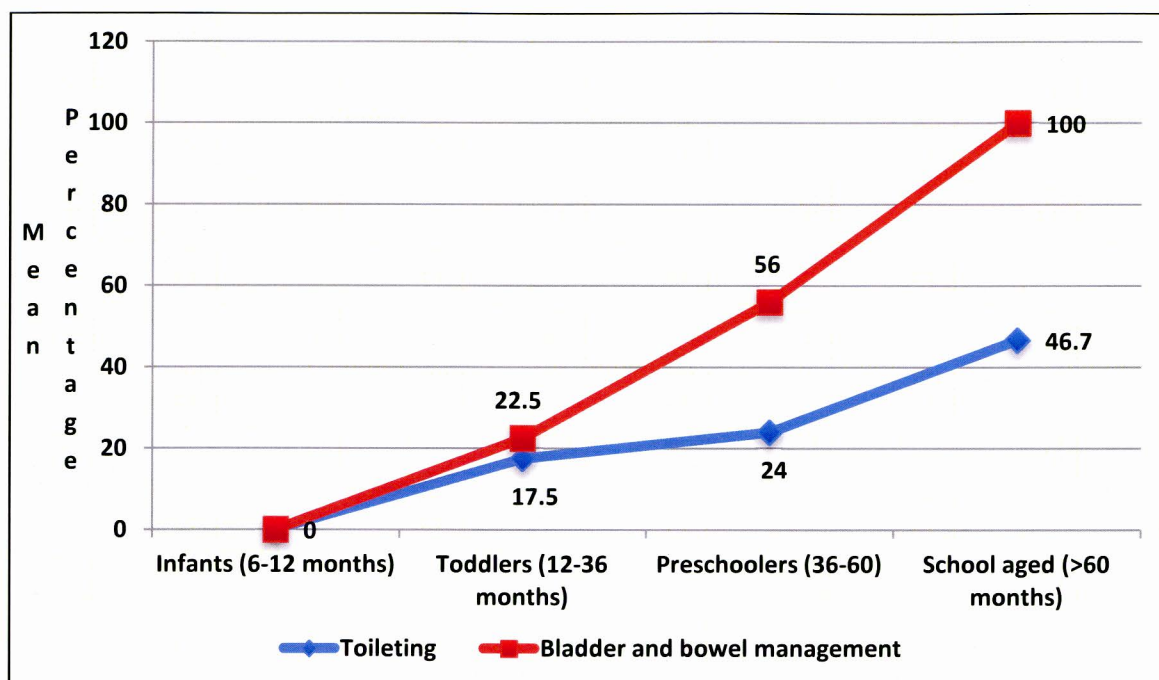


Figure 4.14 Mean percentage independence from caregiver assistance of participants for toilet hygiene according to developmental age groups (n=19)

4.3.3Modification scale

The modification scale was given a numerical value using the same format as the caregiver assistance scale and changed into a percentage. The higher the value, the less modifications are used.

Table 4.12 Mean percentage of total group of participants in the modification scale self care domain and in the developmental age groups (n=19)

	Mean (percentage) (SD)
Total group	97.74 (0.87)
Developmental Age groups	
Infants (6-12 months)	98.95 (2.94)
Toddlers (12-36 months)	97.65 (2.64)
Preschoolers (36-60)	97.5 (3.77)
School aged (>60 months)	96.87 (8.83)

4.3.3.1 Eating and Feeding:

Most modifications were child orientated and school aged children were most dependent on modifications out of all the age groups. Modifications used included allowing the child

to drink through a straw or only filling the cup half way to make it lighter so that the child can pick it up.

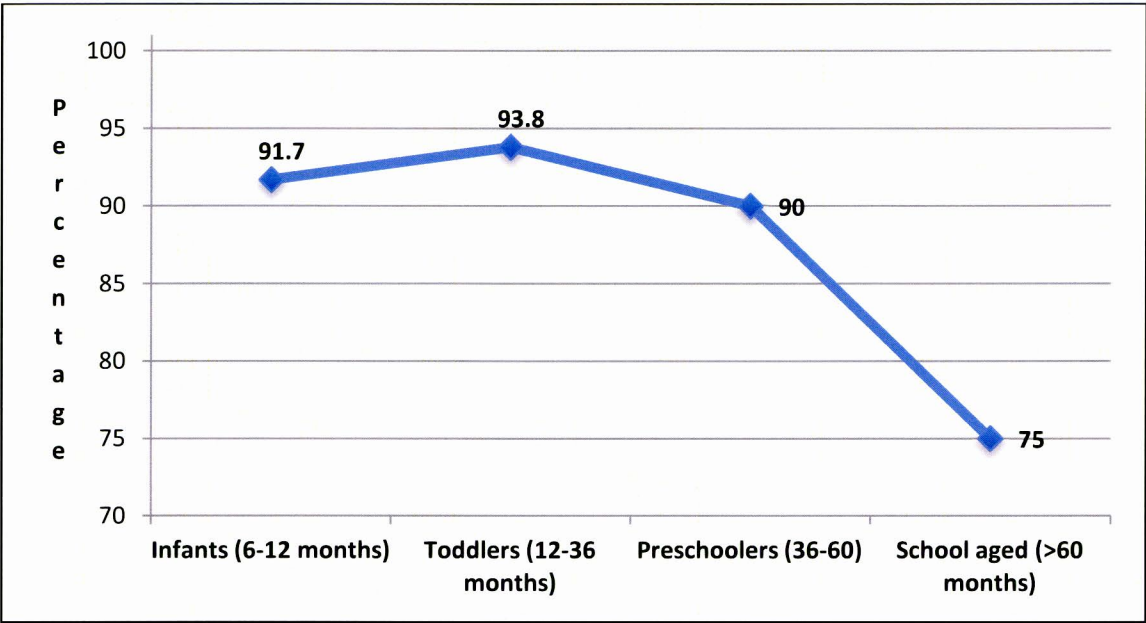


Figure 4.15 Modification score for participants for feeding according to developmental age groups (n=19)

Many of the participants did not rely on modifications but rather used compensatory methods including using mouth to hand rather than hand to mouth movements, picking up food with their mouths instead of using a utensil, picking up a cup bilaterally with back of hands, or picking up cup by biting it and tilting head backward to allow for drinking.

4.3.3.2 Personal hygiene and grooming

Preschoolers used most modifications for bathing purposes whereas the rest of the age groups did not use any. Toddlers used some modifications for grooming whereas the rest of the age groups did not use any. Modifications included filling the bathtub with a small amount of water so that the child can lie down, or not closing the taps tightly so that the child will be able to open them. Instead of relying on modifications participants used compensatory methods instead.

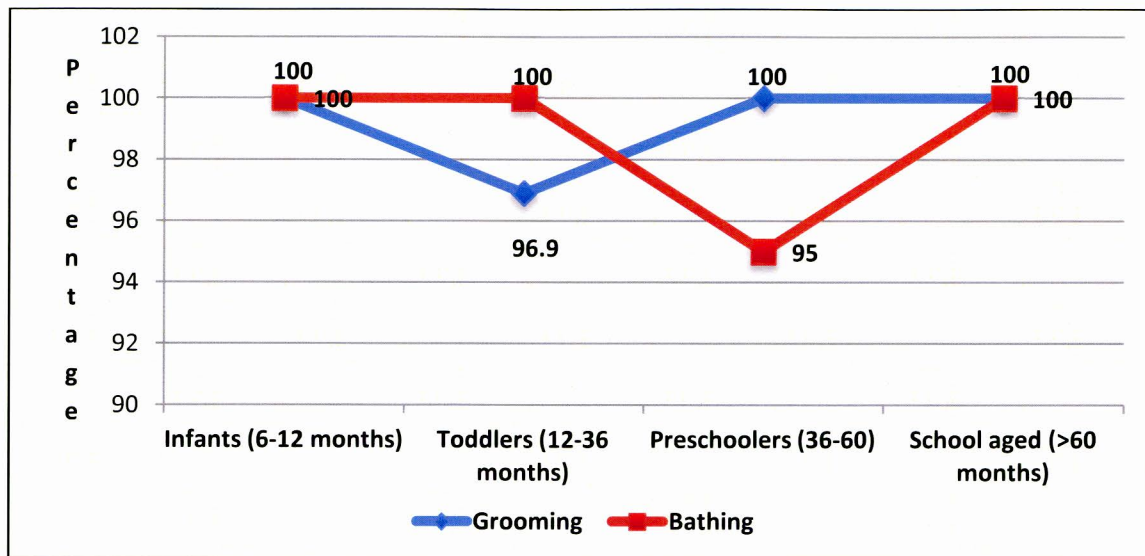


Figure 4.16 Modification score for participants for personal hygiene and grooming according to developmental age groups (n=19)

These include bending down to reach toothbrush in hand and moving head side to side to enable brushing, wiping nose on sleeve rather than using a tissue, washing hands by rubbing the back of hands together, bending down to reach washcloth to enable washing of face and holding a cloth with their mouths and trying to wash their bodies.

4.3.3.3 Dressing:

Toddlers used some child-orientated modifications for dressing tasks related to the upper body and preschoolers for dressing tasks related to the lower body whereas the other age groups did not use any modifications.

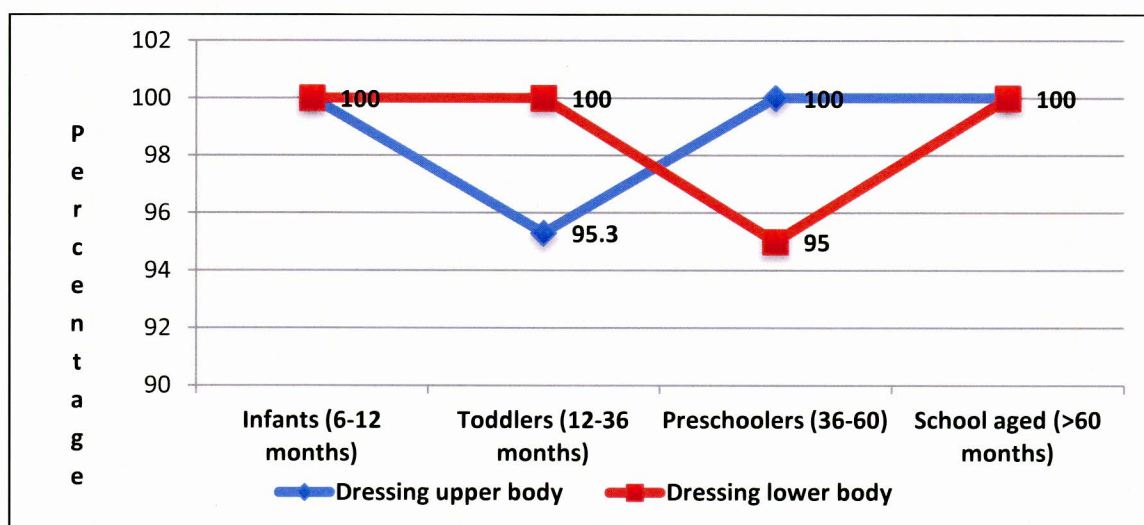


Figure 4.17 Modification score for participants for dressing according to developmental age groups (n=19)

The modifications were in terms of buying clothes with more zips and Velcro fasteners than buttons, and shoes with Velcro fasteners rather than shoe laces. The participants did not use any compensatory methods for dressing and was mostly reliant on caregiver assistance as seen in the Caregiver assistance section.

4.3.3.4 Toilet hygiene:

Only the toddler group used some modifications for toileting but no other modifications were used by any of the other age groups. They were very dependent on caregiver assistance and the majority of the participants were on nappies. (Table 4.11)

Table 4.13 Mean percentages of total group of participants in the modification items of toileting, bladder management and bowel management in the developmental age groups (n=19)

	Toileting Mean (SD)	Bladder management Mean (SD)	Bowel management Mean (SD)
Infants (6-12 months)	100 (0)	100 (0)	100 (0)
Toddlers (12-36 months)	95.3(0.37)	100 (0)	100 (0)
Preschoolers (36-60)	100 (0)	100 (0)	100 (0)
School aged (>60 months)	100 (0)	100 (0)	100 (0)

4.4 Correlation between Self-Care and Caregiver Assistance

The Spearman's rank-order correlation test was used to establish the strength of association between the variables outlined in tables. All correlations were significant at a value of $r=0.73$ and a p value of $p<0.05$ indicating that the more independent the child was the less caregiver assistance he/she needed. Independence thus lessened the burden of care on the caregiver.

4.5 Summary

The descriptive statistical analysis on the 19 participants showed that males were more prevalent than females in the study, and the distribution of the participants were uneven, with the majority being in the toddler and preschool group. Out of the 19 participants, 14 had undergone previous surgery and most of the participants received some non-surgical

intervention like casting or splinting. All of the participants received occupational therapy at some stage and all were satisfied with the intervention that they received.

The preschool and school age groups had negative Z scores at two and three standard deviations below the mean for the self-care domain indicating that as the participants became older, their ability to carry out personal management independently at an age appropriate level became more compromised. The infant and toddler group performed significantly better than the preschool and school aged group. A chi-square test confirmed this and showed that the infant and toddler group performed at a significantly higher level when compared to the expected level of mastery on the PEDI, where-as the preschool and school aged group performed at a significantly lower level than expected when compared to the PEDI.

The preschool group scored the lowest in the caregiver assistance domain, with a negative Z score of nearly three standard deviations from the norm. The school aged group had a negative Z score of nearly two standard deviations from the norm. There was a significant correlation between the amount of caregiver assistance needed and the level of independence of the child, indicating that the less independent the child, the more caregiver assistance is needed.

The modification scores showed that the number of modifications were limited as most participants relied on compensatory methods.

CHAPTER 5

DISCUSSION

5.1 Introduction:

In this chapter the deficits in function of the participants aged six months to five years nine months are discussed. The discussion considers the participants in the age groups used to analyse the data with function in the specific aspects of personal management reviewed. Guidelines for therapists to prioritise treatment and use when treating a child with arthrogryposis to enable their independence in self care skills have also been developed.

5.2 Demographics of the participants:

The wide range of the participants' ages from six months to five years nine months was due to the difficulty of finding a sample of children with this rare condition. The greater percentage of male participants may have been due to the small sample size and lack of participants, as the literature indicates that males and females are equally affected.(3) From the 19 participants, 3 were Caucasian, and 16 were African.

The mothers' demographic data showed no major discrepancies between maternal age(Table 4.2) so this factor, as with other research, cannot be seen as a contributing to the development of the condition. Some studies indicate that there are maternal illnesses that can contribute to the development of arthrogryposis, mostly diabetes myelitis, myasthenia gravis and multiple sclerosis(3) but none of the mothers in this study reported having any of these conditions. Five mothers were on medication during their pregnancy but there is no research investigating the effect of these medications, which included Anti-Retro Viral treatment, antibiotics and blood pressure medication, on the development of arthrogryposis. None of the mothers reported abusing substances during their pregnancies as drugs like cocaine and alcohol if taken at certain critical times have shown to result in congenital contractures(56).

5.3 Birth, social and medical history:

Although there are many known subgroups of Arthrogryposis Multiplex Congenita with different signs, symptoms, and causes(57) for inclusion into this study a diagnosis of arthrogryposis without defining different types was accepted. None of the mothers knew the cause of their child's arthrogryposis which is supported by the literature which states that often the cause of the condition remains unknown (2). None of the participants had

any neurological defects, as those who did were excluded from the study. Most of the participants had amyoplasia of varying severity, with two participants having distal arthrogryposis. Amyoplasia is described as being sporadic with no genetic cause, whereas distal arthrogryposis has been linked to a specific chromosome and is autosomal dominant (3).

The mean gestational age of the participants was 39.7 weeks which is towards the upper end of normal. Many mothers reported that they did not feel the infant move during their pregnancy and the main reason for delivery via caesarean section was that the foetus was in a breech position. This can be a result of decreased foetal movement (3) which is one of the characteristics of arthrogryposis, and which can explain why the foetus did not turn in preparation for birth. Most cases of arthrogryposis can be diagnosed during the second or third trimester in utero by routine ultrasound scans, and is usually triggered by the mother's concern of decreased foetal movement(57). Only one mother who made use of the private health care sector was aware that her child had arthrogryposis before delivery, where as the other 18 participants were only diagnosed after birth. Early diagnosis gives the opportunity for family counselling concerning the condition and planning for the delivery and neonatal medical care, and can thus be improved in the South African setting, especially in the public health care sector which was used by 16 of the 19 mothers.

Many of the mothers that made use of public health care services did not understand their child's condition and the long term consequences as well as the future treatment planned. They did not know why the joint range of their children was restricted but only knew that the limb was 'not working properly'. When asked what they understood under the term arthrogryposis many said that they did not know and that no one ever explained to them what it meant. Many of the mother's did not understand that arthrogryposis it a condition and thought that after surgery their children would have normal limb functioning. The caregivers in the public health care sector often did not have access to information regarding surgical intervention options and could not say what surgery was planned for their children in the future. They were reliant on the advice of the health care providers, mostly plastic surgeons that they consulted. The level of their education was undocumented but most of them were either employed as domestic workers, or were unemployed, staying home to look after their children relying on social grants as a source of income. This may have affected the scores of the PEDI as during the administration of the assessment it was seen that many mothers did not know that they have to encourage independence, readily accepting the role of assisting their children maximally in the

execution of self-care tasks, hence the lack of independence and modifications in the execution of these tasks.

The participants that had access to private health care services had more frequent surgical intervention as their caregivers were often the driving force behind surgical and other medical intervention, researching possible treatment options themselves. Although the level of these mothers' education was not documented, they were all employed in the formal work sector having furthered their education. All of these mothers kept medical records of previous surgeries done and knew which future surgeries were planned.

Out of the 19 participants, 13 had undergone surgery to correct a joint deformity. No surgeries had been conducted on any of the infant participants although most of them had non surgical orthopaedic intervention, mostly plaster of paris (POP) casting, to correct clubfeet.

Of the toddler group, 37.5% received lower limb surgery, with future lower limb surgery planned for a further 37.5%. Most of the participants received surgery for clubfoot corrections, with more club foot corrections and knee surgery planned. No previous hand surgery was done on this group, and one participant (12.5%) was going to undergo hand surgery in future. The caregivers was informed that hand surgery will commence once the lower limbs have been corrected. This participant made use of the public health care sector.

In the preschool group 40% had received lower limb surgeries for clubfeet and knee correction, whilst future surgery was planned for another 40% for further hip and foot correction. One participant in this group received hand surgery in the private health care sector and another participant was going to receive future hand surgery, also in the private health care sector.

Sixty seven percent of participants in the school age group had received lower limb intervention, including clubfeet, ankle and knee surgery and no future lower limb surgery was planned. All lower limb surgery appears to have been completed before the children are of school going age. Of this group two participants (67%) have also received hand surgery to correct wrist, finger and elbow deformities and both of them were going to receive further hand surgery in future. One of these participants made use of the government health care sector and one of the private health care sector.

The mean percentage of surgeries conducted in upper limbs and lower limbs indicate that lower limb surgical intervention had occurred most frequently in the younger age groups with upper limb intervention increasing between the preschool and school aged groups.

(Figure 4.3) Both the public and private health care sectors performed lower limb surgeries. One of the hand surgeries were conducted in the public health care sector, and two in the private health care sector. Three hand surgeries were still planned in the public health care sector and two in the private health care sector. There is thus not a considerable difference in the number of surgeries done when comparing the public and private health care sectors but the participants that made use of the private health care sector were operated on more frequently, especially in the lower limbs.

Only three of the participants in this study received surgical intervention under 12 months of age, one in the public health care sector and two in the private health care sector. The other participants only received surgery after 12 months of age and the surgery in their younger years were aimed at lower limb intervention with upper limb surgical procedures only taking place in the preschool and school aged groups.

The procedures most commonly advised in surgery are related to improvement of the deformities that form as a result of arthrogryposis. Lower limb deformities need to be corrected before the walking age of 18 months, and clubfoot surgery can be done as early as 4 weeks of age(58). Most of the participants were treated with serial casting from early infancy to correct club feet. Club feet surgery only commenced from late infancy and stretched throughout the toddler and preschool age groups. Mobility of the participants was thus limited and most of them reverted to crawling or rolling being delayed in walking due to their deformities. The toddler participants who received previous lower limb corrections were to undergo further surgical procedures for foot correction but the caregivers did not know the exact nature of the surgery. Lower limb surgery in South Africa is thus done at a later stage than literature advocates, as it seems that more emphasis is placed on non surgical methods like casting and splinting. The reason for this was outside the scope of the study, but may be a result of limited resources making non surgical intervention a more viable treatment option.

The literature and the IFSSH report on the management of the upper limb in arthrogryposis completed in 2005 advocate early surgical intervention for the upper limb(59). Upper limb surgery is expected to take place between three and 12 months of age so that the child can still develop the different grasps and good fine motor coordination. It warns against late surgical intervention as altered grasp patterns would already have been established which would then be difficult to change(7, 8,32). A better recovery is also expected from a younger child and a delay of surgical intervention after one year of age causes contractures to become more severe which makes intervention difficult(9)(8). More upper limb surgeries is done in cases of amyoplasia(7) and 17 of the 19 participants in this study was diagnosed with this condition. Of these participants, three

received hand surgery, one a wrist carpectomy, and the rest hand and elbow surgery not specified. Five more upper limb surgeries were planned in future, and it seems that surgical intervention of the upper limb contrary to what is recommended only takes place at a later stage in both the South African public and private health care sectors. Since surgical intervention is delayed in South Africa, it is even more important to start with non invasive procedures like splinting from an early age to prevent contractures from getting worse.

In this sample, three participants could walk after receiving lower limb surgery, and one could weight bear on the lower limbs with support. The other 6 participants still reverted to rolling, crawling or using a wheelchair to move around even after multiple lower limb surgeries. Literature shows that with the necessary early surgical intervention the majority of individuals with arthrogryposis should be ambulatory, with only a small number of individuals still having to rely on a mobility device to move around (29, 30) an outcome which is not consistent with the participants in this study. This could be due to a lack of early surgical intervention.

Nine of the participants could achieve mouth to hand, with the rest using compensatory methods or required assistance. Even the most functional participants could not pick up heavy objects as hand function remained compromised, with limited wrist extension, supination and elbow flexion. Two of the participants had no hand function, one after having had a wrist carpectomy, and used their mouths to execute some tasks. Therefore surgical intervention can also not be totally relied upon to improve function and greater emphasis should be placed on modifications and compensatory methods as these children will physically not be able to execute all tasks in a normal way. A study of 52 participants with a mean age of 16.2 indicated that only 2% of the participants required assistance as a result of poor hand function, with the rest having minimal or moderate restrictions that did not impact on their independence. Compensatory methods were used by those having moderate restrictions (30). Because the participants in the above mentioned study were much older, the results could not be generalised to this study, but it can be noted that the outcome for hand function with early intervention is promising, which again is not consistent with the outcome of the participants in this study.

It was commendable that all of the participants had been referred to and had access to occupational therapy services, with 17 of them starting therapy in the first year of their lives and the other three before their second birthday. Referral to occupational therapy is therefore occurring in the early developmental phase, but can be done earlier as splinting should be started only a few days after birth to promote a better outcome(7). As mentioned in the literature review previous studies have shown an improvement in wrist

and elbow mobility with serial splinting and daily passive stretches and will thus promote the outcome in the absence of early surgical intervention(14, 33). In fact surgery is only recommended after six months if there is a failure to progress with these conservative measures(60).

5.4 Level of Function on the Paediatric Evaluation of Disability Inventory (PEDI)

Overall, the Z scores of each age group indicated that as the participants got older, their independence decreased. The infant group scored within the normal limits as they are mostly dependent on caregiver assistance. The toddler group had a borderline score as they were expected to start becoming more independent in personal management tasks, especially those related to feeding and dressing. The preschool group had a Z score two standard deviations below the norm, and they still struggled with dressing and feeding tasks, but also with tasks related to toileting, washing and grooming. The school aged group's score was three standard deviations from the norm and the level of independence in these tasks did thus not improve.

The results showed that the caregiver assistance increased accordingly. The infant and toddler group's caregiver assistance Z score fell within the norm, whereas, from all the age groups, the preschoolers were most reliant on caregiver assistance, with a Z score of -2.95. The more independent the participant, the less reliant they were on caregiver assistance, decreasing the burden of care. Most of the modifications were used by the school-aged group, which decreased the level of caregiver assistance slightly ($Z = -1.78$), but modifications were limited to child-orientated modifications with no rehabilitative or extensive modifications, even though there was a need for them.

These results showed that not enough emphasis is placed on independence in personal management tasks, which falls within the scope of Occupational Therapy. Modifications and assistive devices were not being used to promote independence and lessen the burden of care on the caregiver.

Because the results indicated that the age of the participants affected the significance of their functional deficits and therefore the level of function for each age group used for the analysis will be discussed separately.

5.4.1 The infant group (6-12 months)

The infant group had an overall Z score of -0.48 in the Functional Skills, Self-Care domain section which indicated their level of function falls within normal limits. At this age group

the infant is mostly reliant on caregiver assistance for functional tasks like dressing, feeding, toileting and the physical restriction due to loss of limb function will thus not have an impact on the execution of such tasks. The infant participants in this study managed all food textures very well and as they were still bottle/breast fed they scored within the norm for feeding items.

Grooming and personal hygiene, dressing and toileting tasks all fell within the normal range as it is appropriate for the infant to be totally dependent on the caregiver(11). The infant group had a Z score of +0.4 in the Caregiver Assistance section which indicates the appropriate amount of caregiver assistance for this age group. The modification percentage was 98.95% and the highest of all the age groups indicating the least number of modifications used, as the infants relied mostly on caregiver assistance and thus did not need modifications. The only modification use was a child-orientated one being a baby feeding cup. The infant participants' scores were significantly higher than those expected on the PEDI, specifically in the eating item as they could manage more food textures and scored 33.3% more than the normative population (Figure 4.5). This is possible as the infant participants had no oral motor limitations that could limit their feeding abilities and the item selection of the PEDI was skewed towards the lower end of the functional skill continuum(46).

5.4.2 The toddler group (12-36 months)

The toddler group had a Z score of -0.9 in the Self-care domain, functional skills, which indicated greater deficits with their level of function. As it was a borderline score close to -1 SD below the norm it already gave an indication of possible future delays.

The toddler participants scored slightly above the norm in grooming, washing and toileting tasks as well as using fasteners and managing shoes and socks. Developmentally it is seen that children in this age group still require a moderate amount of caregiver assistance in all the above mentioned tasks and are only starting to assist in these tasks (opening mouth during toothbrushing, holding hair in position when combed, holding hands out to be washed, assisting with clothing management) which will explain their high scores as the need for selective movement and hand dexterity is limited. The toddler participants scored slightly below the normative group in feeding and dressing tasks as during feeding the toddler is expected to learn how to finger feed, manage a spoon and fork and this increases the level of difficulty of this task. Two of the participants in this group were unable to finger feed, one was unable to hold a bottle or cup, one could hold a bottle but was not able to drink from it, three could use a spoon but not a fork, three could use a knife to butter bread, three could drink from a cup using one hand only and three

used both hands when drinking or pouring liquids. These participants should have learnt to lift the cup to one hand which requires sufficient hand strength as well as a coordinated hand to mouth movement. Dressing required that the participants be able to remove a shirt and pants independently, increasing the complexity of these tasks. Four of the participants in the toddler group could remove their shirts, and three could remove their pants, the rest were unable to.

The toddler group has a Z score of -0.28 in the Caregiver Assistance section of the PEDI which still falls within the normal range. They required the most assistance during toileting and a smaller amount of caregiver assistance in feeding tasks which is appropriate as feeding independence should be developing in this age group. Their modification percentage was 97.65% with more child-orientated modifications than in the infant group, including rubber spoons, soft children's hairbrush, sippy cups, shirts with buttons to make dressing easier and a children's toilet seat. There were no rehabilitative modifications even though all the children attended occupational therapy and feeding modifications should have been introduced as these children need to have some level of independence when attending preschool.

5.4.3 Preschooler (36-60 months)

In general the preschool group of three to six years is a very important age for the development of independence in self care tasks. The preschool participants scored more than 2 SD below the norm during the Functional Skills Self care items, which was significantly below what is expected from their age group ($p=0.00$).

Preschool is the age group where total independence in bowel and bladder management and toileting is developed and potty training plays a very important role(11). By the age of four, there should be no wetting or soiling incidents and the preschooler should be fully independent in bowel and bladder management(55). The preschool participants in the study achieved the lowest scores in the toileting task and required the greatest amount of caregiver assistance in this aspect. Toileting is a very complex task and requires more than control over emptying the bladder or bowel, with the greatest deficits occurring in managing clothing, accessing the toilet and wiping. Three of the participants in the preschool group could indicate when they needed to empty their bladder or bowel but did not have the mobility to access the toilet independently. They could also not manage their clothes fasteners or pants and underclothes when going to the toilet. They did not have the upper limb range of motion to wipe themselves and needed assistance with accessing taps when washing their hands. Two of the participants in this group were still

permanently on nappies as a result of their joint restrictions. Only one of the participants, diagnosed with distal arthrogryposis in this group was independent in toileting.

Dressing independence starts to develop before the age of three years and the preschool participants should have been able to manage 80% of dressing tasks(55). The dressing tasks expected of this age group require more skill than just removing certain types of clothing as children are expected to put on pants and a shirt, even though they may still need some assistance with fasteners. Even putting on shoes is expected, although they need assistance in determining the correct foot. Using zips and snap buttons should be mastered by the age of six years(55).

Putting on and fastening clothes require a lot of bilateral hand use as well as fine motor manipulation which was not possible for most of the participants in the study as they had limited joint ranges and hand function. Thus they scored far below the expected level of function in these tasks and there was no significant improvement in the dressing score from the toddler to the preschool age group, although a great improvement is normally expected(11). The preschool group relied on the greatest amount of caregiver assistance for toileting and dressing tasks, both of which is expected to be executed with minimal caregiver assistance.

The preschool participants were expected to manage 80% of feeding tasks independently. This group was able to eat all types and textures of food and was on par with the normative population. However, they struggled with feeding independence scoring below the expected level of independence. As the nature of feeding tasks become more bilateral with the expectations of being able to use utensils to feed themselves and prepare food such as buttering a piece of bread, or cutting soft foods with a knife and fork, the lower the participants scores were. Two of the preschool participants ate by bending forward and picking up food with their mouths. The other participants positioned the spoon in their hands by wedging it between their fingers and then bent down to put it in their mouths, but were unable to use a knife to cut or butter bread as they could not supinate their forearms or hold the knife using thumb opposition. Preschoolers should also be able to pour liquids from one container into another and drinking from a cup should occur with one hand only(55). The participants still continued to grasp their cups between their hands and a lack of rotation in their forearms meant the level of functioning decreased from the toddler to the preschool group as they could not pour liquids.

Personal hygiene including washing and grooming also showed deficits in level of function. The preschool participants scored only slightly below the expected level of independence for this aspect, but the largest difference between scores was for executing

the washing tasks. It is still age appropriate for a child to rely on caregiver assistance for grooming tasks but 80% of washing tasks should be independently executed in this age group. This includes obtaining the soap and washcloth and washing the body thoroughly, and washing and drying hands thoroughly and managing taps(55).

Using soap and a washcloth requires bilateral hand use and dexterity which made the task difficult for these participants, even though they could execute washing unilaterally. They had trouble washing their entire body as managing to reach all body parts require good range of motion in all planes. Managing taps also requires good hand function and lateral wrist movement which most participants lacked. These limitations in range and hand function seen in arthrogryposis and especially in the participants in this study made the execution of these tasks very difficult. In general the preschool group had very low scores when compared to the normative group with significantly lower levels of independence ($p=0.00$) in dressing, washing, feeding and toileting tasks.

A certain level of independence especially in terms of toileting is expected from a school aged child when attending school and if these tasks are not addressed in this age group it will have a negative effect on attending preschool as being toilet trained is a prerequisite for being admitted into some preschools (12, 13). Finding the appropriate school was a great concern for most of the parents of the preschool group as a delay in independence in self-care tasks may thus have an impact on the type of school a child can go to.

Many parents of the participants in this age group were not aware that they should encourage independence and just continued to assist the participants as they have been doing previously. Therefore the Caregiver Assistance Score was -2.95 SD below the norm, which indicates that this group required far more caregiver assistance than the normative group. The score indicates that the participants relied on the same level of assistance as the toddler group, and there was thus no increase in the level of independence between the two age groups.

The participants required a lot of caregiver assistance and had a score of independence from caregivers of between 24% and 28% in the Caregiver assistance scale for most items except feeding and grooming. Except for dressing this indicated a large dependence on the caregivers.

No assistive devices were used during dressing tasks and the only child-orientated modification was the use of loose fitting clothes. Surprisingly only one child made use of a child-orientated modification namely only filling the tub to a certain level so that he/she can lie down flat in it, although there are a lot of assistive devices designed to increase bathing independence available. There was also a lack of knowledge of the use of

assistive devices although many of these will make a positive contribution towards the development of independence. There are many assistive devices for toileting on the market but not one of the participants knew of or used any, which is of concern as all the participants had access to an occupational therapist who is trained in the use of assistive devices.

5.4.4 School aged group (>60 months)

It is important to note that only one of the participants in the school aged group attended a mainstream school, the rest attended schooling for Learners with Special Educational needs (LSEN). A school aged child should be independent in all self-care items, including fasteners. Managing shoes, socks and fasteners were most problematic for the school aged participant, as it requires a lot of precision grasps and hand muscle strength, both of which was lacking in this population. Their score showed no significant increase from the preschool group and there was thus no improvement in function in this area between preschool and school going age even though they had had upper limb surgery and more surgery was planned. The two participants who received upper limb surgery had better range of motion in wrist extension and supination but function was not much improved as they still could not pick up a glass and lowered their mouths to drink and eat.

Many teaching assistants in the schools who cater for learners with special educational needs took on the role of the caregiver during school hours. This included assisting the participants with toileting as although the participants achieved a 100% level of independence in bowel and bladder management most caregivers reported that the child relies on them for managing toileting. The child tells them when he/she needs to be taken to the toilet but cannot manage their clothes or wiping. The Caregiver Assistance toileting score was thus very low (46.7%) indicative of a high level of caregiver assistance. No modifications were used during toileting tasks.

The group achieved a similar low score (50%) in the dressing task with the greatest amount of caregiver assistance required. Again no dressing modifications were used and the children relied mostly on assistance. There was a 14% improvement from the preschool group, as most of the preschool participants' lower limbs were in plaster of paris making independent dressing of the lower limbs and shoes and socks difficult. Dressing the upper body remained unchanged from the preschool to school aged group.

Feeding and washing both scored under 65% and washing did not show a significant increase from the level achieved by the preschool group. Although washing required a lot of caregiver assistance, feeding did not and feeding had the highest level of independence from caregiver assistance of all the self care items indicating the least

amount of caregiver assistance out of all tasks was required. Feeding had the greatest number of child orientated modifications, including straws and sports bottles for drinking and using bowls instead of plates for easier scooping up of food making feeding easier. Even though it did not assist the child to be totally independent it did lessen the amount of caregiver assistance. No rehabilitative or extensive modifications were used for feeding or washing to reduce the amount of caregiver assistance needed, though the participants were all receiving occupational therapy. The participants who received upper limb surgery had better positioned joints but it did not make a tremendous difference in their functional ability in feeding or washing tasks when compared to the preschool group as they still lacked fine motor dexterity and hand muscle strength.

There were similar results found for grooming which also did not show a significant increase in scores between the preschool and school aged group and the school aged participants. Most of the school aged participants were not able to brush their hair due to limited shoulder and elbow range of motion, and one of the participants also struggled with nose care and tooth brushing. In the whole group the use of modifications was limited to some child-orientated ones like an electric toothbrush and no rehabilitative assistive devices were used. The caregiver assistance total score was more than one standard deviation from the norm (-1,78) and in general indicated that the participants relied on more caregiver assistance than the normative group.

Through the functional independence Z scores of the self-care domain deviating further from the norm it can be seen that the functional ability of the participants in this study decreased according to the age groups. They became less able to participate at the same level as the normative group as the activity demands and need for independence increased. Thus the children need to attend and receive assistance from occupational therapy as more challenges to improve independence are faced as they reach preschool and school age.

5.5. Implications for occupational therapy: guidelines for intervention of self care skills

Since occupational therapy is aimed at treating a child holistically and ensuring independence in all areas of life, guidelines discussed below are aimed at improving functional independence in self care tasks which was the focus of this study. As discussed in previous sections the development of independence in personal management tasks is not only important for maintaining bodily health and hygiene, but also culturally significant as the mastery thereof is expected by the broader community in which a child functions (11). Most schools expect a level of independence in personal

management tasks before they accept a child into the school (12, 13), and it serves thus as a prerequisite for the engagement in other occupations. Mastery of self-care tasks also leads to a sense of mastery and positively contributes to the child's self-esteem and self-reliance. As it provides a sense of autonomy it sets the foundation for the mastery of other more complex skills, as the child develops (11). It also decreases the burden of care on the caregiver, which gets more important as the child gets older.

Although personal management tasks are the focus of this study the therapist should also continue to address all other aspects of development including play, social and emotional well being, scholastic tasks and especially technology which limits the need for physical skill and still makes learning possible. Every child should be individually assessed and their and their family's needs addressed. Because these participants in this study did not suffer any neurological impairments they have normal intelligence, sensory and perceptual functioning and thus have great potential to be very functional with therapeutic intervention(61).

Throughout the analysis of the research results it was seen that there was a lack of modifications used by the participants although their function could greatly be improved by these. As all of the participants had access to occupational therapy services it is of concern as occupational therapists are trained in the use of assistive devices and should be able to provide the participants with the appropriate devices. Unfortunately, locally there is limited access to assistive devices, as most assistive devices are produced and imported from other countries. The public health care system does not always have the funds to obtain imported assistive devices for their patients, and private medical aids do not always cover these devices. Alternative methods or low cost materials should thus be utilised to substitute bought assistive devices as much as possible. Normal sports bottles or straws can be used to make drinking easier, plastic place mats can be placed under plates to provide a non slip surface and utensils can be adapted with built up handles made out of splinting material cut-offs or tubi grip. Button hooks can be made out of a dowel stick and a piece of wire, and velcro cuffs can be used to compensate for poor grasp strength. Clothing can also be adapted by attaching small pieces of velcro at the back of buttons and a wash mitt can be made by sewing a wash cloth together. By using these methods assistive devices can be produced and modifications can be made without requiring a lot of funds or a high level of skill(62).

5. 5.1 The infant group (6-12 months)

The infant group scored within the norm for all self-care items and this age group will thus provide the ground work for developing functional independence during later age groups.

Therapy will thus be more aimed at facilitating normal development and assisting in the development of gross motor function including neck and head control, the ability to roll, independent sitting, crawling, standing and walking as far as the child's restrictions will allow(34). Compensatory movement patterns are encouraged and promoted as long as it contributes to a functional outcome and less used muscle groups are still targeted and strengthened through various therapeutic techniques(34). Another important aspect during this age group that needs to be prioritised are client factors including maintaining range of motion especially in the hands, wrists, elbows and shoulders, strengthening the necessary muscles as preparation for the independent execution of self-care tasks and positioning the hand for promoting hand function. Therefore splinting should be used in this age group, and early surgical intervention advocated if it can contribute to a positive functional outcome(34).

During the study it was evident that most of the caregivers did not understand the condition or the effect thereof and were under the impression that later surgical intervention will remediate all restrictions. Caregiver education thus also needs to be a priority during this age so that the parent can be prepared to assist in the training of independence starting in the next age group.

5. 5.2 The toddler group (12-36 months)

While the emphasis on client factors should be continued therapy in this group should also target the developing independence in dressing and feeding tasks as both of these tasks scored slightly below the normative group in this study. Feeding is initially the most important developmental activity and hand to mouth movements are encouraged in therapy and during play to prepare the infant to self feed(6). Utensils can be modified to include built up foam handles or the use of an universal cuff or long handled, bent or rotated utensils. Scoop dishes and no slip mats can assist the child in getting the food onto the utensil. Many children naturally compensate for a lack of hand to mouth movements by bending forward and this should be encouraged. Nearly all of the participants used this movement during feeding tasks.

A wooden block can be placed on the table on which the child can place their elbow and get the hand closer to the mouth(34). When sitting on the ground, some of the participants in the study used their knees as a platform on which to rest their elbows. In the study modifications used for drinking included straws and light cups for lifting but cups can further be adapted to have handles for easier management.

Dressing independence should be encouraged and parents educated in the type of clothing that the child can manage with more ease. Front opening shirts and pants with elastic waists are easier to remove which will promote age appropriate independence(34).

5.6.3 Preschooler (36-60 months)

The results of this study show that preschoolers struggled with all the functional skill items in the self care domain and a large emphasis should be placed on independence when treating this specific population. As there is a general lack of knowledge on how to improve independence, caregivers should be educated to encourage the child to execute tasks themselves and not to just rely on caregiver assistance. Emphasis should also include toileting, grooming and washing with more independence.

Although many of the tasks that need to be executed during this age group are bilateral in nature and require good range of motion and hand function, there are a lot of modifications and assistive devices that can be brought in to assist the child in executing each task as independently as possible. A home visit should be done to evaluate the environment in which the child is expected to execute these tasks(34).

The child should thus be assessed whilst executing the task and compensatory methods and assistive devices according to the child's limitations should be investigated and implemented. Feeding tasks requiring bilateral hand use can be made possible by providing assistive devices like special knives, or one handed chopping boards which are often used by adult stroke patients(34). Soap dispensers will make washing easier if a child cannot manage a bar of soap and long handled bent hairbrushes can be provided for brushing hair as most children with arthrogryposis do not have the range of motion to bring their hand to their heads. Adapting normal combs or brushes with Velcro cuffs can enable brushing if the child bends forward(61).

Having a bath mitt that the child can pull over his hand eliminates the need for grasping a washcloth. Bathing equipment can be expensive like bath benches and swivel chairs which allow a child with lower limb flexion deformities to transfer into the bath but grabrails or hand held showers can also be used to assist in bathing tasks. When the child has access to a shower a shower bench can be used to compensate for insufficient standing endurance(34).

Electric toothbrushes eliminate the need for up and down and side to side movements and the child can move his head side to side instead, as was seen by one participant in the study.

Clothing management during toileting tasks can be made easier by attaching a zipper pull to the zip, using elastic waisted pants, replacing buttons and hooks with velcro tabs and attaching material loops to underwear and pants to enable the child to pull his pants down and up by hooking their forearms into the loop(62). Another adaption to assist with the raising and lowering of pants is by attaching hooks to the wall. A child can stand with its back against the hooks facing upwards at thigh level and then lower its torso for pants to be pulled upwards. For pulling pants down the child can stand with its back against down facing hooks and then by moving upwards the hooks will catch onto the pants and pull them down(34).

During toileting wiping from front to back should be encouraged(34). Thus simple modifications and assistive devices can make independence in these tasks possible. Of course, as the degree of physical limitations vary between individuals, an assessment of the child's hand function and mobility should be taken into consideration before prescribing a specific assistive device.

5. 6.4 School aged group

In the school aged group independence in all self-care items should already be developing, with only the more difficult tasks like fasteners being mastered. If appropriate intervention occurred in the preschool group, the child should have a reasonable degree of independence (with compensation and assistive devices) in the home environment. A school visit should then be conducted to make the same degree of independence possible in the school environment. As the child progresses in school therapeutic aims will change to improve independence in scholastic tasks, as self-care independence should already be in place. As a child reaches teenage years specific self care items like fastening and unfastening a bra, using a shaver, or wearing make-up will again need to be addressed(34).

5. 6 Limitations and strengths of the study

5.6.1 The use of the PEDI

The PEDI was the assessment tool of choice due to its design for determining the functional abilities of children with disabilities (46). The PEDI was easily administered and easy to understand. The different scales gave a good indication of how the child's functioning relates to the amount of caregiver assistance and modifications, and as children with arthrogryposis rely on assistance and modifications it gave a good indication of where intervention should be aimed at. Because it provides different age groups, the performance of each group could be established and be compared to the other age

groups to see the trajectory of how a specific skill develops over time. As it aims to separate out functional meaningful units, the Self Care domain gave specific information of different self-care skills(46). Strengths and weaknesses in different tasks could thus easily be identified and compared.

One limitation of the study was that only the self-care domain was used. Although the self-care domain could be used in isolation (46) it would be better to get an indication of the participants mobility and social function domain as it would give a more holistic idea of the participant's function.

The only item on the PEDI that was not always culturally appropriate was the hair brushing item as some of the black participants had hair weaves and braids that do not require any brushing.

The item selection of the PEDI was skewed towards the lower end of the functional continuum as it is aimed to assess children with disabilities(36). The participants' scores in other assessments that are not skewed should thus be lower than the scores they achieved in the PEDI.

5.6.2 Sample size

Although a small sample size is usually a limitation, a study on this condition would require years in order to get a greater sample size, which is not always possible. A small sample size thus provides the researcher with the opportunity to conduct a study when only a short time period is available. If a study is researching a new hypothesis where the outcome is questionable it is better to do a study with the participants at hand, even if it is a small sample size, to get an idea of the conclusions that can be drawn as a basis for larger, more time consuming confirmatory studies(63).

As arthrogryposis is such a rare conditions, only a small number of children participated in this study. As there is no research done on arthrogryposis in South Africa, and especially on occupational therapy Intervention in arthrogryposis in South Africa there is a need for research to guide therapeutic intervention. The result from the study should be generalised with caution and other confirmatory studies are required with this population to get a better reflection of their functional skill performance as a smaller sample size will lead to larger confidence intervals which might not produce statistically significant results (63).

CHAPTER 6

CONCLUSION

6.1 Introduction

This study was conducted using a quantitative cross-sectional design to determine the level of function of children with arthrogryposis aged six months to seven and half years under the care of an occupational therapist for at least three months, in the execution of age appropriate activities of personal management. The assessment tool used was the Paediatric Evaluation of Disability Inventory (PEDI) specifically the self-care domain. An additional demographic questionnaire was used for medical and other personal information that was relevant to the study. Nineteen participants were recruited in the Witwatersrand and Tshwane areas. The various results were grouped according to four developmental age groups and data from each age group was analysed using descriptive statistics.

Although the sample size was small the study gave a good indication of which self care items the different age groups found difficult, as well as how much they relied on caregiver assistance and how many modifications they used which gives valuable information in terms of guiding therapists in the South African context when treating a child with arthrogryposis.

6.2 Main findings and implications for Occupational therapy practice

The results indicated that there is a decline in functional independence from the infant to the school age groups, with the older populations becoming more dysfunctional during self-care tasks as the activity demands increase. There was a positive correlation between the self-care skills and caregiver assistance indicating the amount of caregiver assistance increases as the child's level of independence decreases which makes the burden of care on the caregivers greater.

The infant participants scored within the norm as they mostly rely on caregiver assistance at this age. The toddler group showed a slight delay in feeding and dressing independence and still relied mostly on caregiver assistance. The preschool participants on the other hand showed the greatest delay in functional independence in all areas of

self-care including feeding, grooming, washing, dressing, managing shoes/socks and fasteners and toileting and still relied heavily on caregiver assistance.

The school aged participant's performance did not significantly improve when compared to the preschool group and they still had delays in all areas of self-care except eating. Throughout all the age groups very little modifications were used and parents were not aware of various available assistive devices that could be used. As all the participants received occupational therapy (specifically splinting, individual therapy and home programmes) it seems as if therapy was more aimed at the physical client factors of the child and not aimed at improving functional independence through compensatory methods or the use of assistive devices. A remediative approach was thus used with less focus on a rehabilitative treatment approach.

Occupational therapists should advocate early surgical intervention (before one year of age) as the literature suggests as all of the participants only received intervention from late infancy and their toddler years onward. Therapy should be aimed at preparing the infant for functional independence by addressing range of motion, hand function, muscle strength and bilateral hand use. Dressing and feeding independence should become a priority during the toddler years, with further intervention to promote washing, grooming, toileting, and managing shoes, socks and fasteners during the preschool years. The child should be trained to use compensatory methods to achieve a functional outcome and the use of modifications and assistive devices should be implemented where necessary. During the school aged years the school environment should be adapted to promote the same level of independence that the child has in its home environment before the focus of therapy shifts to school related tasks.

6.3 Future Research

There is a great need for more research into arthrogryposis, especially in the South African context, as most of the research done have been done in other countries. Especially Britain, Ireland, America and Australia are active in supporting and educating families that care for a family member with arthrogryposis by various associations and support groups. As it is such a rare condition, literature and previous studies are limited, and do not aim at guiding occupational therapy intervention, but therapists still encounter patients with the condition and need to be effective in the treatment thereof. Research into all aspects of development including gross motor, fine motor, play, social, emotional and scholastic performance would assist in guiding holistic treatment to ensure an optimal outcome.

The NORD report that although more challenging, research into rare conditions should still be encouraged as it still affects the lives of many individuals and families (19).

Overcoming the obstacles related to the research of rare conditions is possible, and will benefit, not only individuals with arthrogryposis, but also individuals with other conditions that limit function and requires intervention.

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APPENDIX A: DEMOGRAPHIC QUESTIONNAIRE

CODE: _____

Personal Information

(to be kept separate)

Parent/ Caregiver's information

Mother/Caregiver's Name: _____

Residential address: _____

Code: _____

Postal address: _____

Code: _____

Telephone number (Home): _____ (Work): _____

(Cell phone): _____

Child's information

Name : _____ Surname: _____

Date of birth : _____

Background questionnaire

The information obtained from the following questions will give the researcher a complete understanding of your child. All information is confidential and only the researcher will have access to it.

Code: _____

Date: _____

Child

Age: _____

Language: _____

Male/ female : Male ☐ Female ☐

School/ _____ crèche

: _____

—

Who looks after the child during the day? _____

Medical background

How old was the mother when the child was born?

Was the pregnancy planned? Yes ☐ No ☐

Did the mother smoke during the pregnancy? Yes ☐ No ☐

Did the mother consume any of the following during pregnancy? Please tick if yes:

Alcohol ☐ Medication ☐ Caffeine ☐

At how many weeks was the baby born?

Type of delivery? _____

Were forceps used? Yes ☐ No ☐

Was the baby's cry immediate? Yes ☐ No ☐

Were there any problems at birth? Yes ☐ No ☐

Please explain

At which clinic or hospital was the baby managed after birth?

Where was the child diagnosed with arthrogryposis?

Are the doctors planning to do surgery on your child in future? Yes ☐ No ☐

Please explain

Where has your child received any Occupational Therapy intervention up to date?

At what age did occupational therapy treatment start? _____

How long has your child been receiving occupational therapy? _____

Which of the following treatment modalities were included in therapy? Please tick

Splinting ☐

Home programme ☐

Individual therapy sessions ☐

Group therapy session ☐

Are you satisfied with your child's progress up to date? Yes ☐ No ☐

Explain

What are your main concerns at this stage?

Thank you!

APPENDIX B: PEDI ASSESSMENT SHEET

Pediatric Evaluation of Disability Inventory

VERSION 1.0

Stephen M. Haley, Ph.D., P.T., Wendy J. Coster, Ph.D., OTR/L, Larry H. Ludlow, Ph.D.,
Jane T. Halfwanger, M.A., Ed.M., Peter J. Andrellos, Ph.D.

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SCORE FORM

ABOUT THE CHILD

Code: _____

Age Year Month Day

Interview Date _____

Chronological age _____

Diagnosis (if any) _____

GENERAL DIRECTIONS

Below are the general guidelines for scoring. All the items have specific descriptions. Consult the Manual for individual item scoring criteria.

PART I Functional Skills:

197 discrete items of functional skills

Self-care, Mobility, Social Function

0 = unable, or limited in capability, to perform item in most situations

1 = capable of performing item in most situations, or item has been previously mastered and functional skills have progressed beyond this level

PART II Caregiver Assistance:

20 complex functional activities

Self-care, Mobility, Social Function

5 = Independent

4 = Supervise/Prompt/Monitor

3 = Minimal Assistance

2 = Moderate Assistance

1 = Maximal Assistance

0 = Total Assistance

PART III Modifications:

20 complex functional activities.

Self-care, Mobility, Social Function

N = No Modifications

C = Child-oriented (non-specialized) Modifications

R = Rehabilitation Equipment

E = Extensive Modifications

PLEASE BE SURE YOU HAVE ANSWERED ALL ITEMS.



PEDI Research Group, Center for Rehabilitation Effectiveness, Sargent College of Health and Rehabilitation Sciences, Boston University,
635 Commonwealth Ave., Boston, MA 02215-1605 • Phone (617) 358-0175, Fax (617) 353-7500

Part I: Functional Skills

SELF-CARE DOMAIN

Place a check corresponding to each item:
Item scores: 0 = unable; 1 = capable

	UNABLE 0	CAPABLE 1
A. Food Textures		
1. Eats pureed/blended/strained foods	☐	☐
2. Eats ground/lumpy foods	☐	☐
3. Eats cut up/chunky/diced foods	☐	☐
4. Eats all textures of table food	☐	☐
B. Use of Utensils		
5. Finger feeds	☐	☐
6. Scoops with a spoon and brings to mouth	☐	☐
7. Uses a spoon well	☐	☐
8. Uses a fork well	☐	☐
9. Uses a knife to butter bread, cut soft foods	☐	☐
C. Use of Drinking Containers		
10. Holds bottle or spout cup	☐	☐
11. Lifts cup to drink, but cup may tip	☐	☐
12. Lifts open cup securely with two hands	☐	☐
13. Lifts open cup securely with one hand	☐	☐
14. Pours liquid from carton or pitcher	☐	☐
D. Toothbrushing		
15. Opens mouth for teeth to be brushed	☐	☐
16. Holds toothbrush	☐	☐
17. Brushes teeth; but not a thorough job	☐	☐
18. Thoroughly brushes teeth	☐	☐
19. Prepares toothbrush with toothpaste	☐	☐
E. Hairbrushing		
20. Holds head in position while hair is combed	☐	☐
21. Brings brush or comb to hair	☐	☐
22. Brushes or combs hair	☐	☐
23. Manages tangles and parts hair	☐	☐
F. Nose Care		
24. Allows nose to be wiped	☐	☐
25. Blows nose into held tissue	☐	☐
26. Wipes nose using tissue on request	☐	☐
27. Wipes nose using tissue without request	☐	☐
28. Blows and wipes nose without request	☐	☐
G. Handwashing		
29. Holds hands out to be washed	☐	☐
30. Rubs hands together to clean	☐	☐
31. Turns water on and off, obtains soap	☐	☐
32. Washes hands thoroughly	☐	☐
33. Dries hands thoroughly	☐	☐
H. Washing Body & Face		
34. Tries to wash parts of body	☐	☐
35. Washes body thoroughly, not including face	☐	☐
36. Obtains soap (and soaps washcloth, if used)	☐	☐
37. Dries body thoroughly	☐	☐
38. Washes and dries face thoroughly	☐	☐
I. Pullover/Front-Opening Garments		
39. Assists, such as pushing arms through shirt	☐	☐
40. Removes T-shirt, dress or sweater (pullover garment without fasteners)	☐	☐
41. Puts on T-shirt, dress or sweater	☐	☐
42. Puts on and removes front-opening shirt, not including fasteners	☐	☐
43. Puts on and removes front-opening shirt, including fasteners	☐	☐

J. Fasteners

UNABLE
0
CAPABLE
1

44. Tries to assist with fasteners
45. Zips and unzips, doesn't separate or hook zipper
46. Snaps and unsnaps
47. Buttons and unbuttons
48. Zips and unzips, separates and hooks zipper

K. Pants

0 1

49. Assists, such as pushing legs through pants
50. Removes pants with elastic waist
51. Puts on pants with elastic waist
52. Removes pants, including unfastening
53. Puts on pants, including fastening

L. Shoes/Socks

0 1

54. Removes socks and unfastened shoes
55. Puts on unfastened shoes
56. Puts on socks
57. Puts shoes on correct feet; manages velcro fasteners
58. Ties shoelaces

M. Toileting Tasks (clothes, toilet management, and wiping only)

0 1

59. Assists with clothing management
60. Tries to wipe self after toileting
61. Manages toilet seat, gets toilet paper and flushes toilet
62. Manages clothes before and after toileting
63. Wipes self thoroughly after bowel movements

N. Management of Bladder (Score = 1 if child has previously mastered skill)

0 1

64. Indicates when wet in diapers or training pants
65. Occasionally indicates need to urinate (daytime)
66. Consistently indicates need to urinate with time to get to toilet (daytime)
67. Takes self into bathroom to urinate (daytime)
68. Consistently stays dry day and night

O. Management of Bowel (Score = 1 if child has previously mastered skill)

0 1

69. Indicates need to be changed
70. Occasionally indicates need to use toilet (daytime)
71. Consistently indicates need to use toilet with time to get to toilet (daytime)
72. Distinguishes between need for urination and bowel movements
73. Takes self into bathroom for bowel movements, has no bowel accidents

SELF-CARE DOMAIN SUM

PLEASE BE SURE YOU HAVE ANSWERED ALL ITEMS.

Comments

Parts II and III: Caregiver Assistance and Modification

Circle the appropriate score for Caregiver Assistance and Modification for each item.

SELF-CARE DOMAIN

- A. **Eating:** eating and drinking regular meal; do not include cutting steak, opening containers or serving food from serving dishes
- B. **Grooming:** brushing teeth, brushing or combing hair and caring for nose
- C. **Bathing:** washing and drying face and hands, taking a bath or shower; do not include getting in and out of a tub or shower, water preparation, or washing back or hair
- D. **Dressing Upper Body:** all indoor clothes, not including back fasteners; include help putting on or taking off splint or artificial limb; do not include getting clothes from closet or drawers
- E. **Dressing Lower Body:** all indoor clothes; include putting on or taking off brace or artificial limb; do not include getting clothes from closet or drawers
- F. **Toileting:** clothes, toilet management or external device use, and hygiene; do not include toilet transfers, monitoring schedule, or cleaning up after accidents
- G. **Bladder Management:** control of bladder day and night, clean-up after accidents, monitoring schedule
- H. **Bowel Management:** control of bowel day and night, clean-up after accidents, monitoring schedule

Self-Care Totals

SELF-CARE SUM

Self-Care
Modification
Frequencies

MOBILITY DOMAIN

- A. **Chair/Toilet Transfers:** child's wheelchair, adult-sized chair, adult-sized toilet
- B. **Car Transfers:** mobility within car/van, seat belt use, transfers, and opening and closing doors
- C. **Bed Mobility/Transfers:** getting in and out and changing positions in child's own bed
- D. **Tub Transfers:** getting in and out of adult-sized tub
- E. **Indoor Locomotion:** 50 feet (3-4 rooms); do not include opening doors or carrying objects
- F. **Outdoor Locomotion:** 150 feet (15 car lengths) on level surfaces; focus on physical ability to move outdoors (do not consider compliance or safety issues such as crossing streets)
- G. **Stairs:** climb and descend a full flight of stairs (12-15 steps)

Mobility Totals

MOBILITY SUM

Mobility
Modification
Frequencies

SOCIAL FUNCTION DOMAIN

- A. **Functional Comprehension:** understanding of requests and instructions
- B. **Functional Expression:** ability to provide information about own activities and make own needs known; include clarity of articulation
- C. **Joint Problem Solving:** include communication of problem and working with caregiver or other adult to find a solution; include only ordinary problems occurring during daily activities; (for example, lost toy, conflict over clothing choices.)
- D. **Peer Play:** ability to plan and carry out joint activities with a familiar peer
- E. **Safety:** caution in routine daily safety situations, including stairs, sharp or hot objects and traffic

Social Function Totals

SOCIAL FUNCTION SUM

Social Function
Modification
Frequencies

Pediatric Evaluation of Disability Inventory

VERSION 1.0

Name _____	Test Date _____	Age _____
ID# _____	Respondent/Interviewer _____	

SCORE SUMMARY

Composite Scores

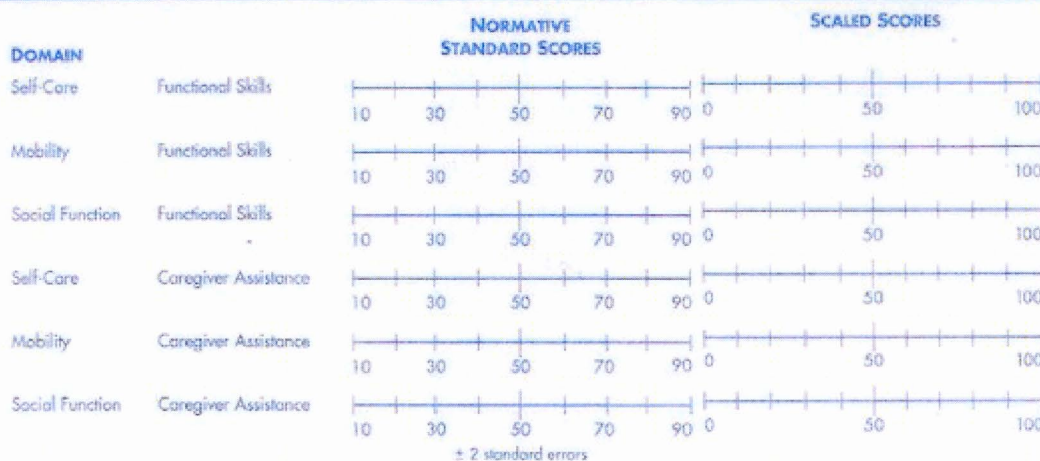
		RAW SCORE	NORMATIVE STANDARD SCORE	STANDARD ERROR	SCALED SCORE	STANDARD ERROR	FIT SCORE*
DOMAIN	Self-Care Functional Skills						
	Mobility Functional Skills						
	Social Function Functional Skills						
	Self-Care Caregiver Assistance						
	Mobility Caregiver Assistance						
	Social Function Caregiver Assistance						

*Obtainable only through use of software program

MODIFICATION FREQUENCIES

SELF-CARE (8 ITEMS)				MOBILITY (7 ITEMS)				SOCIAL FUNCTION (5 ITEMS)			
None	Child	Rehab	Extensive	None	Child	Rehab	Extensive	None	Child	Rehab	Extensive

Score Profile



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 PED Research Group: Stephen M. Holey, Ph.D., P.T., Wendy J. Cohen, Ph.D., OTR/L, Larry H. Ludlow, Ph.D., Jane T. Heltwanger, M.A., Ed M., Peter J. Andriello, Ph.D.

APPENDIX C: PERMISSION TO CONDUCT RESEARCH (Human Research Ethics Committee and Department of Education)

UNIVERSITY OF THE WITWATERSRAND, JOHANNESBURG
Division of the Deputy Registrar (Research)

HUMAN RESEARCH ETHICS COMMITTEE (MEDICAL)
R14/49 Ms Frances Geyser

CLEARANCE CERTIFICATE

M110928

PROJECT

Functional Independence in Personal
Management of Children with Arthrogryposis
Multiplex Congenital Aged One to Seven
and a Half years

INVESTIGATORS

Ms Frances Geyser.

DEPARTMENT

Department of Occupational Therapy

DATE CONSIDERED

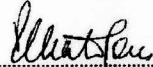
30/09/2011

M1109280DECISION OF THE COMMITTEE*

Approved unconditionally

Unless otherwise specified this ethical clearance is valid for 5 years and may be renewed upon application.

DATE 15/11/2011

CHAIRPERSON 
(Professor PE Cleaton-Jones)

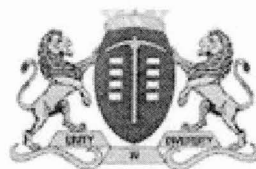
*Guidelines for written 'informed consent' attached where applicable
cc: Supervisor : Mrs Denise Franszen

DECLARATION OF INVESTIGATOR(S)

To be completed in duplicate and ONE COPY returned to the Secretary at Room 10004, 10th Floor, Senate House, University.

I/We fully understand the conditions under which I am/we are authorized to carry out the abovementioned research and I/we guarantee to ensure compliance with these conditions. Should any departure to be contemplated from the research procedure as approved I/we undertake to resubmit the protocol to the Committee. **I agree to a completion of a yearly progress report.**

PLEASE QUOTE THE PROTOCOL NUMBER IN ALL ENQUIRIES...



education
Department: Education
GAUTENG PROVINCE

For administrative use:
Reference no. D2012/203

GDE RESEARCH APPROVAL LETTER

Date:	24 January 2012
Validity of research Approval:	6 February 2012 to 30 September 2012
Name of Researcher:	Geyser F.
Address of Researcher:	P.O. Box 116
	Brakpan
	1540
Telephone Number:	011 024 3201 / 079 196 4912
Email address:	francesgeyser@gmail.com
Research Topic:	Functional independence in personal management of children with Arthrogryposis Multiplex Congenita aged one to seven and a half years.
Number and type of schools:	TEN LSEN Schools
District/s/HO	ALL Districts except Sedibeng East and Sedibeng West

Re: Approval in Respect of Request to Conduct Research

This letter serves to indicate that approval is hereby granted to the above-mentioned researcher to proceed with research in respect of the study indicated above. The onus rests with the researcher to negotiate appropriate and relevant time schedules with the school/s and/or offices involved to conduct the research. A separate copy of this letter must be presented to both the School (both Principal and SGB) and the District/Head Office Senior Manager confirming that permission has been granted for the research to be conducted.

The following conditions apply to GDE research. The researcher may proceed with the above study subject to the conditions listed below being met. Approval may be withdrawn should any of the conditions listed below be flouted:

1. The District/Head Office Senior Manager's concerned must be presented with a copy of this letter that would indicate that the said researcher/s has/have been granted permission from the Gauteng Department of Education to conduct the research study.
2. The District/Head Office Senior Manager's must be approached separately, and in writing, for permission to involve District/Head Office Officials in the project.

Making education a societal priority

Office of the Director: Knowledge Management and Research

9th Floor, 111 Commissioner Street, Johannesburg, 2001
P.O. Box 7710, Johannesburg, 2000 Tel: (011) 355 0506
Email: David.Makhado@gauteng.gov.za
Website: www.education.gag.gov.za

Makhado

3. A copy of this letter must be forwarded to the school principal and the chairperson of the School Governing Body (SGB) that would indicate that the researcher/s have been granted permission from the Gauteng Department of Education to conduct the research study.
4. A letter / document that outlines the purpose of the research and the anticipated outcomes of such research must be made available to the principals, SGBs and District/Head Office Senior Managers of the schools and districts/offices concerned, respectively.
5. The Researcher will make every effort obtain the goodwill and co-operation of all the GDE officials, principals, and chairpersons of the SGBs, teachers and learners involved. Persons who offer their co-operation will not receive additional remuneration from the Department while those that opt not to participate will not be penalised in any way.
6. Research may only be conducted after school hours so that the normal school programme is not interrupted. The Principal (if at a school) and/or Director (if at a district/head office) must be consulted about an appropriate time when the researcher/s may carry out their research at the sites that they manage.
7. Research may only commence from the second week of February and must be concluded before the beginning of the last quarter of the academic year.
8. Items 6 and 7 will not apply to any research effort being undertaken on behalf of the GDE. Such research will have been commissioned and be paid for by the Gauteng Department of Education.
9. It is the researcher's responsibility to obtain written parental consent of all learners that are expected to participate in the study.
10. The researcher is responsible for supplying and utilising his/her own research resources, such as stationery, photocopies, transport, faxes and telephones and should not depend on the goodwill of the institutions and/or the offices visited for supplying such resources.
11. The names of the GDE officials, schools, principals, parents, teachers and learners that participate in the study may not appear in the research report without the written consent of each of these individuals and/or organisations.
12. On completion of the study the researcher must supply the Director: Knowledge Management & Research with one Hard Cover bound and an electronic copy of the research.
13. The researcher may be expected to provide short presentations on the purpose, findings and recommendations of his/her research to both GDE officials and the schools concerned.
14. Should the researcher have been involved with research at a school and/or a district/head office level, the Director concerned must also be supplied with a brief summary of the purpose, findings and recommendations of the research study.

The Gauteng Department of Education wishes you well in this important undertaking and looks forward to examining the findings of your research study.

Kind regards

David Makhado

Dr David Makhado

Director: Knowledge Management and Research

2012/01/24

Office of the Director: Knowledge Management and Research

9th Floor, 111 Commissioner Street, Johannesburg, 2001
P.O. Box 7710, Johannesburg, 2000 Tel: (011) 365 0506
Email: David.Makhado@gauteng.gov.za
Website: www.education.gpg.gov.za

APPENDIX D: PERMISSION TO CONDUCT RESEARCH (Hospitals)



GAUTENG PROVINCE

HEALTH
REPUBLIC OF SOUTH AFRICA

CHARLOTTE MAXEKE JOHANNESBURG ACADEMIC HOSPITAL

Office of the CEO
Enquiries:
Ms. L. Mngomezulu
(011) 488-3793
(011) 488-3753
07th May 2012

Ms. Frances Geyser
Occupational Therapist
CMJAH

Dear Ms. Geyser

RE: "Functional independence in personal management of children with arthrogryposis multiplex congenita aged one to seven and half years"

Permission is granted for you to conduct the above research as described in your request provided:

1. Charlotte Maxeke Johannesburg Academic hospital will not in anyway incur or inherit costs as a result of the said study.
2. Your study shall not disrupt services at the study sites.
3. Strict confidentiality shall be observed at all times.
4. Informed consent shall be solicited from patients participating in your study.

Please liaise with the Head of Department and Unit Manager or Sister in Charge to agree on the dates and time that would suit all parties.

Kindly forward this office with the results of your study on completion of the research.

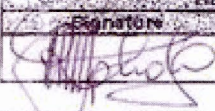
Yours sincerely

Dr. T.E. Selebano
Chief Executive Officer

INITIAL CONSENT BY DEPARTMENTAL HEAD

I Mashudu Mphohoni head of Occupational Therapy
 department of Steve Biko hospital in consultation
 with the Chief Executive Officer / Superintendent of this Hospital grant permission to
 submit an application to conduct a clinical trial/evaluation to the Chairperson (s) of the
 relevant Ethics, Research and Therapeutic Committees of this Hospital.
 The officer conducting the trial/evaluation will be Frances Greyser
 Designation / Rank Snr Occupational Therapist cmjAH

THE HEAD OF THE DEPARTMENT MUST SIGN HERE!

HEAD OF DEPARTMENT			DATE		
Signature	Initial(s)	Surname	Day	Month	Year
	M.R.	MPHONONI	13	08	2012

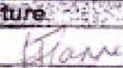
THE APPLICANT MUST SIGN HERE

TRIALIST INVESTIGATOR			DATE		
Signature	Initial(s)	Surname	Day	Month	Year
<u>Greyser</u>	F	Greyser	31	07	2012

THE APPLICANT THAT APPLIES FOR THIS STUDY MUST SEE TO IT THAT THE CHIEF EXECUTIVE OFFICER / S.E.O. OF THE
 HOSPITAL WHERE THE STUDY WILL BE DONE, SIGN HERE FOR THE ETHICAL COMMITTEE RECEIVE
 THIS APPLICATION FORM

APPROVAL BY HOSPITAL CHIEF EXECUTIVE OFFICER:

I Dr. Tanna Chief Executive Officer / superintendent of
Steve Biko Hospital, hereby agree that this trial / evaluation be
 conducted in the Occupational Therapy Department of this hospital.
 The officer conducting the trial will be : Frances Greyser
 The officer controlling supplies will be: N.A.

HOSPITAL C.E.O. / Superintendent			DATE		
Signature	Initial(s)	Surname	Day	Month	Year
	H.	TANNA	16	08	2012



GAUTENG PROVINCE

HEALTH
REPUBLIC OF SOUTH AFRICA

DR. GEORGE MUKHARI HOSPITAL

Enquiries: F.A. Swanepoel
Tel: 012-5293500
Fax: 012-5600776
E-Mail: francois.swanepoel@gauteng.gov.za

Ms. F. Geyser
C/O The Head: Allied

Dear Ms. Geyser

HONORARY APPOINTMENT AT DR. GEORGE MUKHARI HOSPITAL

Approval has been granted for your honorary appointment at this hospital in the department of Occupational Therapy as a Supernumerary Occupational Therapist for a period of one year. Your appointment is subject to the registration with the Health Professionals Council of South Africa since practicing without registration is illegal.

Your appointment is subject to the following conditions:

- I. Appropriate registration with the South African Health Professionals Council.
- II. No remuneration will be payable to you for your services.
- III. You will be subject to all the rules and discipline applicable to full-time employees of the hospital.
- IV. You will be charged against lawsuits due to damages suffered by a third party as a result of an act or omission as determined by Part 12 of the Treasury Regulations.
- V. The Department will not be responsible for any injury or damage that may be suffered by you during your stay at this hospital.

If you have any questions or problems, please contact the Head of your Department who will be delighted to be of assistance to you.

We trust that you will find the experience both useful and pleasant.

Yours faithfully

M. Geyser
CHIEF EXECUTIVE OFFICER
DATE: 28/1/2013

Dr George Mukhari Hospital
Mendunza drive
PRETORIA
0001

Private Bag X422
PRETORIA
0001

Telephone number: (012) 529-3202 / (012) 529 3851

31 Jan. 2013 16:48

FAX NO.: 0879444589

FROM: Green Store

MEDICAL ADVISORY COMMITTEE
CHRIS HANI BARAGWANATH ACADEMIC HOSPITAL
PERMISSION TO CONDUCT RESEARCH

Date: 14th February 2012

TITLE OF PROJECT:

**Functional independence in personal management of children with arthrogryposis
multiplex congenita aged one to seven and a half years.**

UNIVERSITY: Witwatersrand

Principal Investigator: FRANCES GEYSER

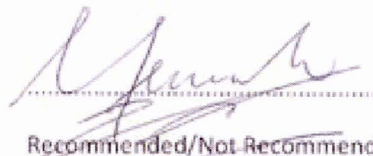
Department: Occupational Therapy

Supervisor (If relevant): Denise Franzsen

Permission Head Department (where research conducted) Yes

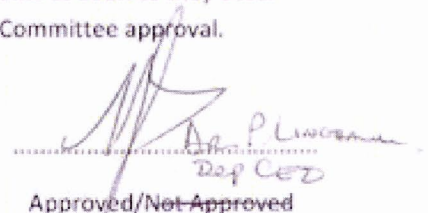
The Medical Advisory Committee recommends/ ~~does not recommend~~ that the said research be conducted at Chris Hani Baragwanath Hospital. The CEO /management of Chris Hani Baragwanath Academic Hospital is accordingly informed and subject to:-

- Permission having been granted by the Committee for Research on Human Subjects of the University of the Witwatersrand.
- the Hospital will not incur extra costs as a result of the research being conducted on its patients within the hospital
- the MAC will be informed of any serious adverse events as soon as they occur
- Permission is granted for the duration of the Ethics Committee approval.



Recommended/Not-Recommended

(On behalf of the MAC)



Approved/Not-Approved

Hospital Management



APPENDIX E: INFORMATION LETTER

INFORMATION LETTER

Hello,

My name is Frances Geyser and I am an occupational therapist working at the Charlotte Maxeke Johannesburg Academic Hospital. I am currently doing my masters in occupational therapy through the University of the Witwatersrand. I am doing a study to determine what children born with hand deformities like arthrogryposis can do for themselves when it comes to executing activities of daily life like dressing, washing, feeding, toileting, brushing teeth etc. This is important for us as occupational therapists to know so that we can help these children to do these things for themselves.

I would like to invite you and your child to take part in this study. I am using a questionnaire called the Paediatric Evaluation of Disability Inventory and would like to observe your child doing some of these activities by themselves like eating, drinking, washing their hands, putting on pants and a shirt and closing buttons and zips. I would also like to ask you questions about what your child can do in terms of activities of daily life, and how much help or modifications they need. This will take approximately 60 minutes.

If you agree to this study, all the information you give me will remain confidential and only I will have access to your child's name or any of your personal information. You can decide to withdraw from this study at any given time without any consequences. Withdrawal will not affect the treatment your child receives in any way.

If the results of the study indicate your child has a problem which is not being addressed I will make the names of appropriate professionals available to you should you wish to consult them about treatment.

Feedback on the study will be available on request otherwise.

If you have any further questions regarding the study please feel free to contact me- Frances Geyser at 079 196 4912/ 011 488 4201. For any ethical queries or complaints please contact the secretary of the Ethics Committee Anisa Keshav at 011 7171234.

Frances Geyser, Occupational Therapist

APPENDIX F: INFORMED CONSENT FORM

INFORMED CONSENT: CAREGIVER

I, _____ (name of caregiver) have read and understood the information letter on the study. I agree to let _____ (name of child) participate, and know that I can withdraw from the study at any stage without any consequences.

Signature of caregiver: _____

Signature of researcher : _____

Witness : _____

Date : _____

I agree that the researcher may contact me _____ at
(Phone number) _____

In order to arrange an appointment for the interview and assessment

APPENDIX G: WRITTEN/VERBAL ASSENT (CHILD)

INFORMED CONSENT: PARTICIPANT

I, _____ (name of participant) have read the letter explaining the study and agree to take part in it. I know that I can decide not to be a part of it at any stage.

Signature of participant : _____

Signature of researcher : _____

Witness : _____

Date : _____