UPPER LIMB REDUCTION DEFICIENCIES IN SWEDISH CHILDREN

Classification, prevalence and function
with myoelectric prostheses

Liselotte Hermansson

Stockholm and Örebro 2004

From the Department of Woman and Child Health
Karolinska Institutet, Stockholm, Sweden

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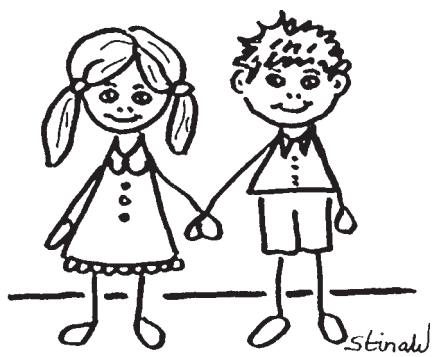
To my beloved family
Lars, Maria and Johan



ABSTRACT

Upper limb reduction deficiency (ULRD) is a rare condition that has been known ever since the 6th century B.C. This is a lifelong deficiency which in an afflicted child can lead to practical limitations, social restrictions and physical problems.

The overall aim of this research was to increase the knowledge about children with upper limb reduction deficiencies from three perspectives: the deficiencies themselves, the use of prostheses and the well-being of the affected children.

To validate information regarding ULRD in the Swedish Register for Congenital Malformations (SRCM), all infants reported to this register during 1973-1987 were re-classified according to a more detailed classification. The result was compared with a clinic-based register at the Limb Deficiency and Arm Prostheses Centre in Örebro, Sweden. The findings indicate that SRCM, with its calculated underestimation of 6%, can be used for studying the prevalence of ULRD in Sweden. However, as SRCM is a surveillance register, the quality of some information seems to be low, making detailed description of cases difficult. Use of the population register data for clinical purposes could therefore result in lower validity. Additional information and follow-up of specific cases are therefore recommended.

The presence of scoliosis and trunk asymmetry was studied in 60 persons with transverse ULRD. Nineteen persons (31%) had a scoliosis of between 10 and 19º and 30 persons had minor curves of between 5 and 10º. There was a significant correlation between leg length inequality and side of the convexity, with the convexity directed towards the side of the shorter leg in 21 of 28 persons. This indicates that children with transverse ULRD may have a transient scoliosis of postural origin of no clinical significance.

A new observation-based test, the Assessment of Capacity for Myoelectric Control (ACMC), which measures a person's capacity to control a myoelectric prosthetic hand during the performance of ordinary daily tasks, was developed. Occupational therapists completed 210 assessments of 75 persons. Rasch rating scale analysis was used for validation and reliability estimations. The results demonstrate internal scale and person response validity.

The external reliability of ACMC was established by scorings from three raters with different degrees of experience on 27 videotapes of client performance. The major finding in this study was that in order to obtain reliable measures from the ACMC the raters have to have some experience of this group of clients. Until the ACMC can adjust for rater severity, the same rater should perform the ACMC when it is used for follow-up or clinical trials.

In a study of 62 children we found that, overall, children with ULRD who have been fitted with a myoelectric prosthetic hand are just as well adjusted psychosocially as their able-bodied peers. There are indications, however, of social stigmata related to the deficiency which have to be considered differently in boys and girls. Most children who have been provided with a myoelectric prosthesis at an early age continue to use the prosthesis.

Keywords: children, upper limb, deficiency, register validation, scoliosis, arm prosthesis, measurement, occupational therapy, psychopathology, depression.

LIST OF PUBLICATIONS

This thesis is based on the following original papers. They will be referred to in the text by their Roman numerals.

I Hermansson L, Bodin L, Wranne L (2001) Upper limb deficiencies in Swedish children – a comparison between a population-based and a clinic-based register, Early Human Development, 63: 131-144.

II Samuelsson L, Hermansson L, Norén L (1997) Scoliosis and trunk asymmetry in upper limb transverse dysmelia, Journal of Pediatric Orthopaedics, 17: 769-772.

III Hermansson L, Fisher A, Bernspång B, Eliasson A (in press) Assessment of capacity for myoelectric control: A new Rasch-built measure of prosthetic hand control, Journal of Rehabilitation Medicine, 36: 1-6.

IV Hermansson L, Bodin L, Eliasson A (submitted) Intra- and interrater reliability of the Assessment of Capacity for Myoelectric Control.

V Hermansson L, Eliasson, A, Engström I (in press) Psychosocial adjustment in Swedish children with upper-limb reduction deficiency and a myoelectric prosthetic hand, Acta Paediatrica, 93: 1-10.

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LIST OF ABBREVIATIONS

ACMC	Assessment of Capacity for Myoelectric Control
ARM	Armreduktionsmissbildning
ATR	Angle of trunk rotation
CBCL	Child Behavior Checklist
CDI	Children's Depression Inventory
CI	Confidence interval
e.g.	For example – *exempli gratia*
i.e.	That is – *id est*
ISO	International Organization for Standardization
LDAPC	Limb Deficiency and Arm Prostheses Centre
LLI	Leg length inequality
LRD/s	Limb reduction deficiency/ies
OT	Occupational therapist/therapy
PUFI	Prosthetic Upper Extremity Functional Index
PUS	Prosthetic Use Scale
SE	Standard error
SRCM	Swedish Register for Congenital Malformations
SIRS	Skills Index Ranking Scale
TD	Terminal device, e.g. hand or hook
TULRD/s	Transverse upper limb reduction deficiency/ies
ULRD/s	Upper limb reduction deficiency/ies
YSR	Youth Self Report

1 INTRODUCTION

Upper limb reduction deficiencies (ULRDs) received a large amount of public interest when the thalidomide disaster became apparent in the beginning of the 1960s (Lenz and Knapp, 1962). As a result of the use of this drug, thousands of children worldwide with limb reduction deficiencies of a certain pattern were born. National surveillance registers were initiated, large resources were assigned to the development of prosthetic limbs, and special rehabilitation centres were established. However, this was not a new phenomenon. Persons with limb reduction deficiencies (LRDs) have been recognized in the early works of Hippocrates (460-377 B.C.) (Löwegren, 1909), and portrayed in art ever since the 6th century B.C. (Fig. 1).

Figure 1. Archaic Greek terracotta vase found in Sicily, 6th century B.C. © Musée d'art et d'histoire, Ville de Genève, dépôt de l'Association Hellas et Roma, inv. no HR 79. Photo: Flora Bevilacqua

In a critical analysis of this figure, it was concluded that it shows a man with a transverse upper and lower LRD (Dasen, 1997). Despite the long history of this type of malformation, few scientific studies have been conducted to elicit the daily life situation of these children and most studies have been focused on individual topics such as the aetiology of the deficiencies or the development of prostheses (Minnes and Stack, 1990). When seeing these children and their parents in the clinic, it is clear that many questions still remain unanswered.

The overall aim of this research was to increase the knowledge about children with upper limb reduction deficiencies from three perspectives: the deficiencies themselves, the use of prostheses and the well-being of the affected children.

1.1 THE DEFICIENCY

1.1.1 Prevalence and aetiology of limb reduction deficiencies

Apart from the thalidomide period, the prevalence of LRD has been relatively stable over the years. In a comprehensive literature review, Ephraim et al conclude that there appears to be a marked similarity in rates of LRD across different countries and reported by different investigators (Ephraim et al., 2003). According to their review, the prevalence of LRD in different countries reported from studies of various hospital-based and population-based surveillance registers is 2-7/10,000 births. In Sweden, the prevalence at birth of LRD reported from the Swedish Register for Congenital Malformations (SRCM) is estimated to be 6.3/10,000 live births (Källén, Rahmani and Winberg, 1984). Upper limb reduction deficiencies constitute approximately two-thirds of limb reduction deficiencies. In British Columbia the prevalence of ULRD was estimated to be 3.8/10,000 live births (Froster and Baird, 1992).

In Sweden, however, no exact figure has been reported for the number of children with ULRD. Information from the SRCM is not detailed enough to allow a true estimation of how many children are born with ULRD, or of the laterality and clinical level of the deficiencies. There is also some uncertainty as to how well the SRCM covers the true situation.

The aetiology of limb reduction deficiency is in most cases unknown. Many theories concerning the origin of the malformation have been discussed over the centuries. In a comprehensive summary of current knowledge the different potential causes of LRD are presented (Brown et al., 1996). Today, most researchers agree that different types of reduction deficiency have different aetiologies.

With the increased knowledge of the embryology of normal limb development, the possible genetic origin of limb malformation is being discussed. However, limb deficiency may not be induced by genetic dysfunction alone; teratogens and other factors may inhibit normal gene expression or act on cells and tissues directly, thereby producing phenocopies that appear to be genetic in origin, but are not (Sadler, 1998). One other factor that affects the development of the limb is the foetal vascular supply. Van Allen (1981) made a comprehensive description of potential vascular incidents that may affect the normally developing tissues and eventually induce limb defects. She suggests that the extent, timing and mechanism of injury all determine the nature of the defect produced.

Disruption of newly formed vessels can be one cause of isolated transverse limb defects. Such defects have been reported, for example, as an effect of early chorionic villus sampling (Golden, Ryan and Holmes, 2003). A theory regarding the Subclavian Artery Supply Disruption Sequence as the basis for isolated transverse ULRD has also been proposed (Bouwes Bavinck and Weaver, 1986). This theory, further, explains the origin of Poland, Klippel-Feil, and Möbius anomalies, syndromes that often include ULRD (Bouwes Bavinck and Weaver, 1986; Weaver, 1998). The characteristic findings that support this aetiology are the presence of fingernails, and of distal phalangeal tuft (Fig. 2).

Figure 2. Transverse upper limb reduction deficiency with finger buds and nails.

Another disruption caused by embolic occlusion of a vessel, e.g. the brachial artery (Hoyme et al., 1982), may result in ischaemia and necrosis of distal tissues. Also, external pressure occluding vessels, as in amniotic bands (Wiedrich, 1998) and strangulation with the umbilical cord, may cause pressure necrosis. An adverse position in utero may lead to vessel compression at sites of pressure points (Van Allen, 1981).

1.1.2 Classification of limb reduction deficiency

The method used for classification of limb reduction deficiencies is always guided by the aims of making the classification. For instance, in searches for possible aetiological explanations of the deficiencies, as in surveillance registers, the malformations are classified into homogeneous subgroups with a common pathogenetic mechanism or aetiology. In contrast, in studies of the epidemiology of limb reduction deficiency conducted for clinical purposes, classification according to details of the malformation, its laterality and the number of affected limbs are essential. This has led to the development and use of several classification systems.

In most classification systems the limbs are divided into groups. The deficiencies are arranged according to severity in a possible teratological sequence based on embryological failures (Henkel and Willert, 1969; Swanson, 1976; Lösch et al., 1984). The work by Temtamy and McKusick (1978) has been cited in many attempts to make a classification of LRD. However, as indicated by Stoll (1998), the origin of LRD cannot be explained in only one way, and hence it is not possible to have a classification based on morphogenesis alone. The only way to classify limb defects, according to Stoll, is to use a descriptive system. Another aspect to consider when choosing a classification system is that it should be of practical value in the everyday management of ULRD (Watson, 2000).

A working group from the International Society for Prosthetics and Orthotics (Kay et al., 1975) recognized the problem of the use of different terminology and classifications, and suggested that deficiencies should be described in the simplest yet most precise language that would be understandable by all in the English-speaking world and easily translatable to other languages. The deficiencies should be divided into either “transverse” or “longitudinal”.

According to the system proposed by Kay et al (Kay et al., 1975), the transverse type of deficiency comprises limbs that have developed proximo-distally to a certain level beyond which no skeletal remnants exist. These deficiencies shall be classified by naming the side of the deficiency and the level at which the limb ends, e.g. the forearm (see Study I, Fig. 2). Long bones are divided into thirds, and small bones in the hand are classified as totally or partially absent. For example, the deficiency depicted in Figure 3 is a transverse deficiency, left, forearm, upper third.

Figure 3. Child with transverse upper limb reduction deficiency, left, forearm upper third.

The longitudinal type of deficiency comprises limbs in which one or more bones are partially or totally absent, but distal parts of the affected limb may be present. In these cases the limbs

are classified by naming the side and the missing or partially missing bones. In the hand, corresponding parts may be classified as “rays” (see Study I, Fig. 3). Children with longitudinal deficiency often have a small or weak but yet functioning grip. Hence they are not fitted with a prosthesis.

This attempt at a uniform classification has now become an international standard (ISO 8548-1:1989 (E)) (International Organization for Standardization, 1989) and is being widely accepted and used (Cobben, Hiemstra and Robinson, 1994). The present thesis will focus on the situation of children with transverse ULRD (TULRD).

1.2 CONSEQUENCES OF TRANSVERSE UPPER LIMB REDUCTION DEFICIENCY

Transverse upper limb reduction deficiency that is present at birth or as the result of amputation in childhood is a lifelong condition that can lead to practical limitations, social restrictions and physical problems.

1.2.1 Practical limitations

The loss of a hand has major consequences in the execution of daily tasks. In most activities we use both hands, separately or together, to perform tasks. Further, in the society the norm for doing things is by using both hands. Even if one hand is the principal (the dominant) and the other is the helper (the non-dominant) hand, both hands are usually involved in the task performance. Thus, for a child with TULRD, many tasks are difficult if not impossible to perform according to the standard.

For example, to hold on to something, such as a swing or a ride-on toy, to climb a ladder or a climbing frame, or to ride a tricycle or a bicycle, the child needs both hands to stabilize the body and prevent him/her from falling off. In these situations, children with TULRD have great difficulties and need either to avoid doing these things, or to do them with support from an adult. However, in order to participate in all areas of life these activities are important for young children. To not be able to crochet or knit, for example (Gardsäter, 2004) in the same way as her peers can be devastating for a six-year-old child.

1.2.2 Social consequences

A highly visible cosmetic and functional impairment does indeed have many social consequences for the TULRD child. The non-symmetrical appearance with an unusual stump makes most children with TULRD suffer from peoples’ looks and questions. To avoid this, it is typical for these children to hide the deficient arm or hand in a pocket or under a long sleeve. This, in turn, leads to functional impairment in social situations. The children use only the non-deficient hand to perform tasks and thus cannot do all the things that their peers can do. Among friends, the children sometimes ask for help, thus making them dependent on other people. If they try to solve the problem themselves by using alternative techniques for doing things, other peoples’ attention is drawn to

the deficiency. This attention from other people and the distress of not being able to do things as well and as quickly as their peers have been reported to be "micro-stressors" leading to highly depressive symptoms (Varni et al., 1989b) and low general self-esteem (Varni et al., 1989a) in children with LRD. Furthermore, micro-stressors are predictors of low perceived physical appearance (Varni and Setoguchi, 1991), which, in turn, leads to low self-esteem and depressive symptoms. This has been reported as the "new hidden morbidity" in paediatric practice (Varni and Setoguchi, 1992). In those studies, however, the type and level of deficiency were not homogeneous. Children with transverse and longitudinal, upper and lower limb, deficiencies were included and mixed. Nor was the clinical management of the children specified.

Factors that might lessen the chronic strain of living with LRD are social support (Varni et al., 1989b), with classmate social support being the strongest alleviator (Varni et al., 1991), and perceived physical appearance (Varni et al., 1989a). Furthermore, access to a functional prosthesis is a factor that may affect psychosocial adjustment by facilitating independence (Tyc, 1992).

1.2.3 Physical consequences

It is well known but rarely considered in TULRD and amputations that unilateral work leads to over-use of the unaffected arm (Jones and Davidson, 1999). Uncompensated, high-level TULRD leads to asymmetrical body positions during play and work, which in turn may cause physical problems. To be able to use the deficient arm, most children with TULRD have to bend over or rotate the trunk to compensate for the shortness of the arm. This is most apparent when the children are riding a bicycle or working at a table. Consequently, many adolescents with TULRD complain of back-pain.

In TULRD, besides the lack of symmetry and function other non-limb malformations are rare (Stenninger and Hermansson, unpublished manuscript). Occasionally, TULRD occurs as part of a known syndrome or in association with other medical conditions (Froster-Iskenius and Baird, 1989). However, throughout growth, children with TULRD may encounter different physical problems related to the deficiency.

In rare cases of TULRD, and only in the humerus, skeletal overgrowth of the distal part of the bone may lead to pain and infection, and, eventually, penetration of the skin by the bony spike. This condition, generated by local mechanical stimuli at the end of the stump, is caused by excessive modelling activity identical to a wound-healing sequence seen during healing of fractures. By plugging the medullary canal of the humerus, the healing process can be disrupted and the subsequent overgrowth prevented (Davids, 1998; de Smet and Fabry, 1999). Marquardt first introduced this "stump capping procedure", where autogenous bone transplant was used to prevent the skeletal overgrowth and eventually to assist in suspension of a prosthesis (Marquardt, 1989).

In several studies abnormalities of the spine have been found to occur in association with ULRD. An increased incidence of scoliosis (48%) compared with that in the

general population has been reported in longitudinal ULRD (Makley and Heiple, 1970). In mixed cases of longitudinal and transverse ULRD, the incidence of idiopathic scoliosis was 16% (Powers et al., 1983). Powers et al, (1983) concluded that all patients with ULRD warrant close observation throughout growth for the development of scoliosis. This is an issue of great concern, especially for the parents. No information is available, however, on the incidence of scoliosis in persons with isolated transverse deficiencies.

1.3 MANAGEMENT OF CHILDREN WITH UPPER LIMB REDUCTION DEFICIENCY

To give birth to a child with TULRD is a situation of emotional chaos for most parents. The parents' immediate reaction is to feel apprehension about the child's future and imagine all things that will be impossible for the child; will he be able to play like other children, to meet a partner, to support himself and raise a family? Hence, the first priority for the medical professionals is to support the parents.

To assist in a healthy attachment between child and caretaker, most practitioners agree that the parents of a child with ULRD should receive satisfactory initial information and support (Sörbye, 1989; Setoguchi, 1991). Genetic counselling should be considered (Cobben et al., 1994; Sadler, 1998), and a clinical assessment to make a plan for future interventions should be made. Depending on the type, level and laterality of the deficiency, different interventions are plausible: (i) a prosthesis; (ii) constructive hand surgery; or (iii) technical aids/compensatory techniques.
Once the parents have received adequate information that will allow them to make a decision about future intervention, further plans are made for the child.

1.3.1 Prostheses

In the fifteenth and sixteenth centuries, artificial hands and arms were developed to replace limbs lost to gangrene or battle injury. Ambroise Paré demonstrated an articulated artificial hand and arm in his book "A Universal Surgery" in 1561 (Lyons and Petrucelli, 1978). A similar prosthesis, a Stibbert, was described and the production was discussed by Vincente Putti in 1933 (Putti, 2003). In the old days, prostheses were made for adult males, to compensate for the loss of a grip and to enable soldiers to get back and fight. Today, prostheses are made both to compensate for reduced function and to restore a normal appearance, in persons of all ages and gender.

Most upper limb prostheses include a terminal device (TD), which replaces the hand, a wrist unit, and a custom-made socket. There are two major groups of upper limb prostheses, those with an active TD, e.g. a myoelectric prosthesis, and those with a passive TD. Depending on the need of the person concerned, the two types can be equally functional.

Myoelectric prostheses

In the 1960s, Russian scientists were the first to introduce a myoelectric prosthesis suitable for clinical use (Childress, 1973). Myoelectric prostheses mostly have a motorized, electric hand as TD. Of the five fingers in the prosthetic hand, usually only two fingers and the thumb are active and oppose to each other (Fig. 4). The remaining two fingers are passive. A cosmetic glove covers the fingers and the motor. Electrodes, located inside the prosthetic socket over muscle bellies, detect electrical activity in the muscles. Through the electrodes, the contracting muscle activates the motor of the TD. Adjustments in the force or velocity of the contraction control the range of opening and closing in the TD. Rechargeable batteries supply the energy to operate the motor in the TD.

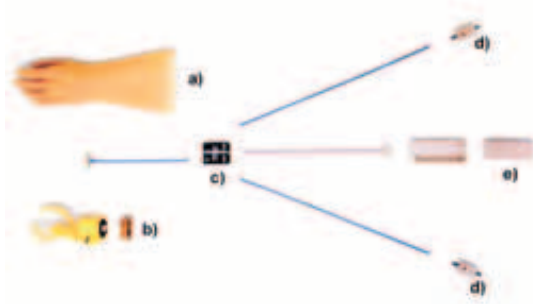

Figure 4. Components in an Otto Bock 2000 myoelectric prosthetic hand. a) cosmetic glove; b) active fingers and wrist unit; c) control module; d) electrodes; and e) battery-pack. Reprinted with permission.

Following the successful myoelectric fitting of a pre-school child in Örebro, Sweden in 1971 (Sörbye, Bartels and Rolander, 1973), child-sized myoelectric hands came into production in Sweden (Sörbye, Hedström, Holmqvist and Randström, 1978). The advantages of a myoelectric prosthesis are that it is self-suspended and self-containing (usually no harness or external power source is needed), the control is independent of the position of the arm, it has a strong grasp, and has a cosmetic appearance. The disadvantages are the high costs and the heavy weight.

Passive prostheses

Prostheses with a passive TD either have a tool for a special task (e.g. a fork or a hammer) or a cosmetic hand. The use of this kind of prosthesis is restricted to its specific function, e.g. for eating or to make the appearance of the wearer more symmetrical and not to draw attention to the deficiency. The advantages of the passive prosthesis are that it is lightweight and durable. Disadvantages are the restriction of having only one specific tool, and the lack of a grasping function.

1.3.1.1 Myoelectric prosthetic fitting

The fitting of the prosthesis is the first step in a long period during which the child has to learn to adjust to the artificial limb, control the TD, and perform daily life tasks using the prosthesis. The overall aim of supplying TULRD children with a prosthesis is that they will come to experience unrestricted participation in everyday life; that is, involvement in formal and informal everyday activities (Law, 2002).

Fifty years ago, the importance of early fitting of a prosthesis was demonstrated in children with body-powered prostheses (Brooks and Shaperman, 1965). Based on his experience from myoelectric fittings in children, Sörbye (1977) suggested that

children with TULRD should initially be fitted with a passive prosthesis at the age of 3-6 months. This was for the child to become accustomed to wearing a prosthesis, to use it for support and symmetry when sitting, crawling and pulling to stand, and subsequently to adapt to the additional length. It was later shown that children and adolescents 5-21 years of age with prosthetic arms adapted to the residual limb length and prosthesis length in the same way as normal children of the same age adapt to the length of their arm (McDonnell et al., 1989). Today, most centres agree on the benefit of initial fitting of a passive prosthesis at the age of 3-9 months (Curran and Hambrey, 1991; Jain, 1996; Hubbard, Kurtz, Heim and Montgomery, 1998).

The age for the first fitting of a myoelectric prosthesis varies somewhat between countries. Sörbye (1989) suggested 2 ½ - 4 years of age. Today, in some countries fitting of a body-powered cable-operated hook at the age of 14-20 months precedes the fitting of a myoelectric prosthesis at 2 ½ - 4 years of age (Curran and Hambrey, 1991; Jain, 1996; Kuyper et al., 2001). In other countries a single-site myoelectric prosthesis is fitted at 10-15 months, with a change to a dual site myoelectric prosthesis at the age of 3-4 years (Hubbard, Kurtz, Heim and Montgomery, 1998).

In Sweden, the Limb Deficiency and Arm Prostheses Centre (LDAPC) at the University Hospital in Örebro provides service for the majority of the TULRD children in Sweden. Here, the procedure established by Dr Sörbye (Sörbye, 1989) is maintained. This means that children are fitted with a passive prosthesis at the age of 3-6 months and a myoelectric prosthesis at the age of 2 ½ - 4 years.

1.3.1.2 Myoelectric prosthetic training

In children with TULRD who receive a prosthesis, training by an occupational therapist (OT) is initiated. The parents are the representatives of the child, and by interviewing them and observing the child a plan for the training is decided upon. The aims of the training are that the child will (1) wear the prosthesis so that it is available when needed, (2) be able to control the prosthesis so that it can be used when required, and (3) be able to use the prosthesis in the performance of daily tasks when appropriate.

1) Regarding the first aim, the parents are the most important persons. They are the ones who will assist the child in establishing a habit of wearing the prosthesis on a regular daily basis. Thus the role of the OT is to support the parents and give instructions as to how to don and doff and to maintain the prosthesis. Also, a plan for increasing the wearing time up to full-day use is agreed upon. This is done when the child is about 3-6 months old and receives the first prosthesis, which usually has a passive hand.

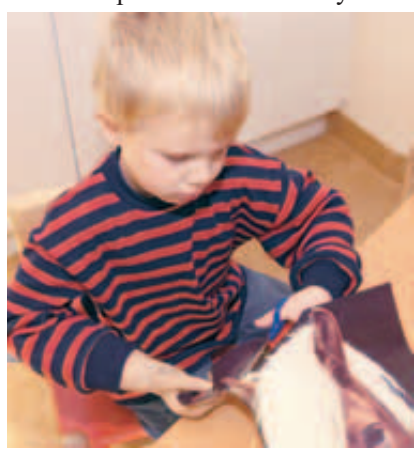

Figure 5. Use training

During the initial fitting and the following years, an ongoing relation with the child and the family (therapeutic rapport) is established. This is a mutual experience of concentration, communication and enjoyment that should have a beneficial effect on the child's performance and follow-through with treatment plans (Tickle-Degnen, 1995).

2) Next, when the child is fitted with an active, myoelectric hand, the control training is initiated. During control training, children are expected to learn to contract muscles in the residual limb and control the function of the prosthesis. The control training is performed during play (Hambrey and Withinshaw, 1990). Children automatically view toys and games as play and therefore as fun, motivating and non-threatening (Tobias and Goldkopf, 1995). By using games chosen by the child, the meaningfulness and purposefulness of the game makes the use of the prosthesis natural. During this play, in bilateral activities where activity in the TD is spontaneously achieved, e.g. when the child is reaching for the handlebars of a tricycle, the OT makes the child aware of the grip and encourages him/her to use it. Once the child begins to get the idea of how the grip is activated, this procedure is repeated in different situations during play. To increase the capacity to control the grip, the encouragement is repeated in activities with varying degrees of difficulty, e.g. with different sizes and shapes of the objects or in different positions in relation to the body (Hermansson, 1991).

3) By the above-described method for control training, the use of the prosthesis is now becoming natural to the child. Gradually, as the capacity for control improves and the child is getting older, the attention is focused more on training in the use of the prosthesis (Fig. 5). Use training is an ongoing issue for many years as the child gets older and new tasks are introduced. In many families this can become a natural component of the regular upbringing of the child. Other families need more support. In the bi-annual follow-up, this is discussed with the family and further support is given to those who need it.

This procedure for prosthetic training is very similar to that described in the late 1950s when the first OT programmes for children with TULRD were introduced (Richardson and Lund, 1959).

1.3.1.3 Outcome assessment of myoelectric prostheses

As myoelectric prosthetic fittings are becoming a more or less standard procedure in children with TULRD, assessment of the outcome of prosthetic training is becoming increasingly important. Two questions that need to be answered are: Are we choosing the right methods for prosthetic training, and are the training methods effective?

Outcome of control training

As already mentioned, the first step to be taken before the prosthesis can be actively used in the performance of daily activities is to learn the ability to control the prosthesis. That is, to learn to operate the TD as easily and spontaneously as any other part of the body, given the restrictions imposed by the prosthetic hand, i.e. the opening width or the motor velocity. To be able to use the prosthesis in any activity requires an ability to control the grip in any position around the body and with any object, and to do this without visual support. However, despite the fact that enhanced control of the myoelectric prosthesis (i.e. myoelectric control) is thought to improve the child's ability to perform essential tasks (Hubbard, Galway and Milner, 1985), no method is available for measuring the outcome of control training, i.e. myoelectric control assessment.

Fifteen years ago, a first attempt to create a method for assessment of myoelectric control was made. This was a stepwise description of different qualities of myoelectric control, the Skills Index Ranking Scale (SIRS; Fig. 6), assessed during the performance of daily activities (Hermansson, 1991). Using this method, OTs were able to document the progress of children with myoelectric prostheses, and to make a rough estimation of the outcome. However, since the size of the difference between the steps in the SIRS was unknown, we could only describe the level of the child on the scale, but never measure the difference from one point in time to another, i.e. the real outcome.

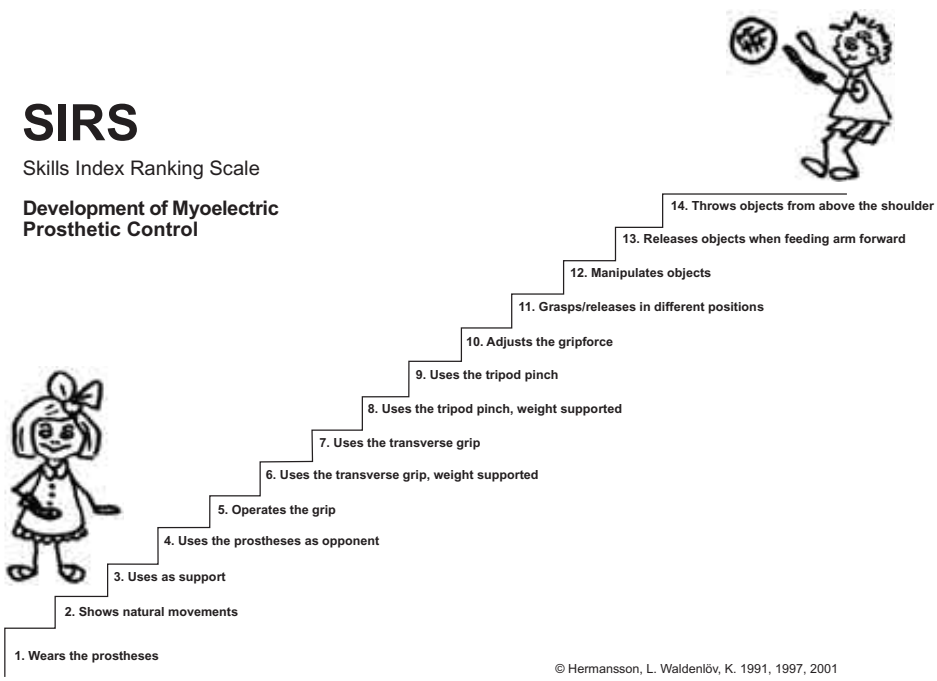

Figure 6. Skills Index Ranking Scale

Outcome of use training

The aim of use training is to teach the child to learn to use the prosthesis in the performance of everyday tasks. To evaluate the outcome of this training, the Prosthetic Upper Extremity Functional Index (PUFI) (Wright et al., 2001) can be implemented. The PUFI is a self-reporting questionnaire in which the older child or the parents answer questions about the performance of everyday tasks, and the use of the prosthesis when carrying out these tasks.

1.3.1.4 Aspects of instrument development

The interest in, and indeed the need for the development of outcome assessments in rehabilitation have prompted discussions on what statistical methods should be used in instrument development. Basically, the choice of method depends upon whether one wants to measure, or to order, the ability in question. Measuring implies that the ability is measurable by an objective abstraction of equal units of this ability. Ordering, on the other hand, implies that the ability is classified in an ordered manner. Most scales in the physical sciences represent measurements, whereas most scales in the human sciences are in fact merely orderings (Bond and Fox, 2001).

It has long been recognized as untrue to the statistical properties of qualitative data, e.g. data on the ability to perform ordinary tasks, to create measures of these. However, by 1952 a Danish mathematician, George Rasch (1901 – 1980), had laid down the basic foundations for a new psychometric method applicable to instrument development, where ordinal data could be converted to linear measures, and later introduced as the Rasch analyses (Wright and Stone, 1979). This method has proved to be very useful both in rehabilitation and paediatrics (Haley, Ludlow and Coster, 1993; Penta, Thonnard and Tesio, 1998; Duncan et al., 2003; Fisher, 2003).

As in other aspects of human science, learning of the capacity for myoelectric control is an ongoing, linear, and not a stepwise, process. In attempts to describe this process today, we are limited by the number of steps, or items, on the SIRS. We can only place a child at a level that we have previously described. All development that takes place in between these steps is obscured to us. Hence, to develop an instrument for assessment of myoelectric control, we need to make a measurement scale.

1.3.2 Surgery

In children with TULRD, when there is a possibility of achieving grasp function, that is in all cases where there are partial hand deficiencies, hand surgery is considered. The surgical methods include, for example, separation of syndactylies or deepening of finger webs to make the grip wider, phalangeal transplantation from toes, transposition of fingers, or microvascular free toe-to-hand transplantation to give two-digit opposition (Flatt, 1994; Watson, 2000).

1.3.3 Compensatory techniques/technical aids

When the deficiency leaves no possibility for surgical grip construction, and fitting of a prosthesis is no option, learning special techniques or using adaptive equipment is a way for the child to be able to perform tasks independently. One example may be to make a so-called "spatula" (Fig. 7).

Figure 7. Adaptive equipment, a so-called "spatula".

In a situation in which the deficiency makes normal performance impossible, children with TULRD mostly learn alternative techniques by themselves. However, in view of the physical effort with which certain tasks are performed by means of such techniques, and the risk of future damage to the body, use of technical aids or adaptive equipment is strongly recommended.

2 AIMS OF THE INVESTIGATION

The overall aim of this project was to increase the knowledge about children with upper limb reduction deficiencies from three perspectives: the deficiency, the use of prostheses and the well-being of the child. The specific objectives were

- to estimate the number of Swedish children born each year with ULRD, and to examine children with transverse ULRD for the presence of scoliosis (Studies I and II).

- to develop an instrument that measures control of myoelectric prosthetic functions (Studies III and IV).

- to study the well-being of children with TULRD in relation to the use of myoelectric prostheses (Study V).

3 MATERIAL AND METHODS

3.1 SUBJECTS

In this investigation, 135 persons with either congenital or acquired TULRD (73 males, 62 females; 2 to 57 years of age) participated in one or more of the separate studies (Table 1). Specifically, 68 persons participated in one study, 48 persons in two studies, 17 persons in three studies, and two persons in four studies. In Study I, all reports coded as ULRD for the years 1973 to 1987 in the SRCM and in the LDAPC register were used. In Studies II to V the participants had all been, and most of them were still, patients at the LDAPC in Örebro.

Table 1. The distribution of participants in the different studies.

Study	Number of patients	Gender (male/ female)	Age in years	Number of patients only in this study	Overlap in study groups
I		Data from registers			
II	60	30/30	7-48	35	13 also in study III, 4 also in study IV, 23 also in study V
III	75	43/32	2-57	21	13 also in study II, 22 also in study IV, 39 also in study V
IV	26	11/15	2-40	3	4 also in study II, 22 also in study III, 11 also in study V
V	62	31/31	8-18	9	23 also in study II, 39 also in study III, 11 also in study IV

3.2 DATA COLLECTION

To cover the broad aim of this research, several different methods for data collection were used.

3.2.1 The deficiency (Studies I and II)

3.2.1.1 Prevalence of ULRD

To estimate the number of Swedish children born each year with ULRD, information from the SRCM was used. To this registry, all infants born in Sweden with signs or symptoms of malformation are to be reported. However, since the information in the SRCM is classified into homogeneous subgroups with a common pathogenetic mechanism or aetiology, it is not immediately useful for this purpose. To obtain clinically useful data on the number of children with ULRD and on the laterality, type and level of deficiency, we need more precise information. In our clinic, we have used

the ISO 8548-1:89 method (International Organization for Standardization, 1989) for classification and have found it very useful for categorization of the children according to their deficiency. Hence, all notification cards in the SRCM for a 15-year period were scrutinized and reclassified according to the ISO 8548-1:89 method.

In a study by Ericson, Källén and Winberg (1977) the SRCM was compared with another national register of birth defects, the Medical Birth Registry, to estimate the validity of the SRCM. The under-notification was estimated to be 11%. However, specific information on ULRD in the SRCM has not been validated. Therefore, to validate the information regarding ULRD in the SRCM, we used the clinic-based register in the LDAPC at Örebro University Hospital. To this register, children from different parts of Sweden referred to the LDAPC are added. All children are classified according to the ISO 8548-1:89 method (International Organization for Standardization, 1989) before they are entered into the register, thus simplifying the comparison with the SRCM. The presence or absence of a child with ULRD in the SRCM register was checked, and if a child was present in both registers the type, side and level of deficiency were compared between the two registers.

3.2.1.2 Scoliosis in TULRD

To examine a person with TULRD for the presence of scoliosis, several methods may be considered. Firstly, the conventional way is to use a standing postero-anterior radiograph and measure the curvature by the standard Cobb method (Cobb, 1948). Secondly, since scoliosis is not simply a lateral deviation of the spine – there is also a rotational component of the trunk leaving a typical rib prominence on the convex side – the forward bending position is a method that has been considered the most sensitive clinical test for evaluating scoliosis (Kahanovitz and Levine, 1982). In this position, in the case of a scoliosis a prominent rib hump is seen on the side of the convexity.

To measure the rotation of the trunk, two non-invasive methods have been developed, moiré topography (Willner, 1979) and scoliometry. In an earlier study (Nissinen et al., 1993), scoliometry was found to be preferable to moiré topography in screening for scoliosis. By scoliometry, the angle of trunk rotation (ATR) is measured in the forward bending position with an inclinometer adjusted on the waterscale placed on the trunk on the concave side (Bunnell, 1984). However, the validity of this method in children with TULRD has not been established. As part of the present study we therefore tested the validity of scoliometry as a screening tool for scoliosis in children with TULRD.

A secondary, or functional, type of scoliosis is present when the lateral deviation of the spine is caused by another condition, usually leg length inequality (LLI) or muscle spasm (Keim, 1978; Staheli, 1992). This type of scoliosis is considered to be a non-structural condition produced compensatorily by the difference in leg length. The pelvis dips down on the short side, causing a transient scoliosis that is corrected in sitting, walking and lying down. A correlation between side of decreased leg length and side of lumbar convexity has been shown (Nissinen et al., 1993). To measure LLI, different methods have been

suggested, e.g. measuring the leg with the patient lying down (Keim, 1978), or simply putting a board of known thickness under the heel of the shorter leg, during standing, until horizontal symmetry is attained (Nissinen et al., 1993). Another method (Friberg, 1983) is to measure the difference in height between the highest articular points of the femoral heads from standing radiographs. Since this seemed to be a more precise method, we chose this in the subjects in whom it was possible from the information in the radiograph.

3.2.2 The use of prostheses (Studies III and IV)

3.2.2.1 Development of an instrument – evidence of validity

In order to meet the need for a valid, reliable and sensitive evaluation instrument, the Assessment of Capacity for Myoelectric Control (ACMC) was developed. As will be reported below, the structure of the ACMC was developed in three phases, two pilot trials and a final test that is presented in Study III. The same statistical method, Rasch analysis (Wright and Stone, 1979), was used for both the preliminary analyses and the final testing. Data collection was initiated after the first, and perhaps the most important, part of the instrument development – the selection of items (American Educational Research Association, American Psychological Association and National Council on Measurement in Education, 1999).

The list of items is an expression of the underlying capacity to be measured. As mentioned in the Introduction, we had an idea of how the capacity for myoelectric control was progressing; thus the capacity was expressed as the SIRS (Hermansson, 1991). The results of the first Rasch analysis showed that SIRS represented a unidimensional construct but was not sensitive enough to detect changes in the capacity for control. Hence, by analysing children of different abilities when they were operating their myoelectric prosthetic hand during the performance of daily tasks this list of items was expanded from 14 to 37 items. The different levels of quality of control that were observed and documented were discussed with another experienced OT. The resulting 37 items were then pilot-tested and revised on the basis of a new Rasch analysis. In this analysis, items that were found to be redundant or misfitting to the Rasch rating scale model of the ACMC were eliminated. Hence, there were 30 items in the final list (see appendix in Study III). The items are scored on a four-point ordinal scale as follows: zero, not capable; 1, sometimes capable, capacity not established; 2, capable at request; and 3, spontaneously capable.

To validate the use of the ACMC in persons of different ages and with different aetiologies, we gathered data from persons of all ages with myoelectric hands who had either congenital or acquired TULRD. For validation of outcome assessments, and to ensure more stable estimates of item difficulties, a sample size of 150 is recommended (range 108-243) (Linacre, 1994b). In developing an instrument for assessment of myoelectric prosthesis users, however, this number is almost impossible to achieve. As described in earlier studies, the prevalence of TULRD is low and not all affected persons are eligible for a myoelectric prosthesis. Hence, the number of patients even in a major centre such as the LDAPC in Örebro is small. However, in other studies

repeated testing of participants has been performed to increase the sample size (Tham, Bernspång and Fisher, 1999). Since the data collection period was 18 months, and we expected that most participants would make progress during this time and/or be assessed by different OTs, we decided to increase the sample size in this study by repeating assessments.

3.2.2.2 Reliability of the ACMC instrument

Reliability refers to the consistency of measurements when the procedure is repeated on a population of individuals or groups (American Educational Research Association et al., 1999). Assessment of the reliability, i.e., the information about measurement error, is essential to the proper evaluation and use of an instrument, since measurement errors reduce the usefulness of measures. Thus, for further use of the ACMC in clinical practice, the reliability of the instrument needed to be determined.

When determining the reliability of an assessment method, there are two main factors to be considered. First, internal factors that may lead to inconsistency, such as the client's motivation or the consistency of the client's application of the capacity, need to be considered. For the ACMC, the internal reliability was determined by Rasch analysis in Study III.

Next, external factors that may influence the measurement, such as the types of forms used for recording information, types of tasks, or rater subjectivity, also need to be considered. These factors are regarded as potential systematic errors because they influence the measurement in a consistent direction. Hence, the external reliability of the ACMC should be studied. Since the ACMC is administered in a clinical setting with no specification as to which task should be performed, the influence of different tasks was difficult to assess. However, the rater reliability could be established.

One problem in deciding the agreement between raters is that the analysis can demonstrate consistency only among the rank orders of clients or scores. It does not tell us anything about differences in severity or leniency between raters, i.e. rater discrepancy in difficulty levels (Bond and Fox, 2001). This, however, is something that needs to be considered, since most raters add their subjectivity to their ratings. Besides items and persons estimates, the Many Faceted Rasch analysis (Linacre, 1994a) provides estimates on raters. Hence, by Rasch analyses, the individual rater severity can be estimated.

According to the standards of the American Psychological Association et al. (American Educational Research Association et al., 1999), the ideal approach to the study of reliability entails independent replication of the entire measurement process. Thus, to study the rater reliability in ACMC, we decided to use video-films. Twenty-five clients of varying age and ability were video-filmed once during a regular visit to the LDAPC in Örebro and one client was video-filmed twice with a three-month interval, while they were doing varying daily tasks.

The precision of a rater's score may be influenced by the rater's experience of the area in which the client is to be assessed. In order to study this, three raters with different degrees of experience were assigned for this evaluation. Two raters were OT students in their last year of education, of whom one had had 10 weeks of practice in this area of clinical work and the other had no experience. The third rater was one of the most experienced OTs in the LDAPC in Örebro. Data were obtained by the raters' assessments of the clients' performance on the videos. For intra-rater evaluation, the assessments were repeated by all three raters in the same order three to four weeks later.

3.2.3 The well-being of the child (Study V)

Well-being is a personal, subjective, trait, and there are many methods that can be used to capture and describe this feeling. In our study of children with TULRD, we focused attention on how the children adapted psychosocially to their situation, as compared with observations on their peers.

We wanted to study the situation of children 8 – 18 years of age, but considered children below 11 years of age too young to answer questionnaires about their psychosocial situation and mental health, and we therefore decided to ask the parents of children younger than 11 years to answer questionnaires on behalf of their children. However, since the size of the population and hence the number of children eligible for this study were small, we included parents of all children and not only those of children aged 8-11 years. Thus, the study comprised two groups: parents of all children 8-18 years old, and adolescents aged 11-18. In order to study the well-being in relation to use of a myoelectric prosthesis, we included only those who were using or had been using a myoelectric prosthesis.

To be able to compare the outcome of our study with results of other studies on children with TULRD, we used methods for data collection that have been applied in earlier studies of children with LRD. The questionnaires Child Behavior Checklist (CBCL) and Youth Self Report (YSR) (Achenbach, 1991a-b), and the Children's Depression Inventory (CDI) (Kovacs, 2001), have been widely used and translated into more than 50 languages, including Swedish (Larsson and Melin, 1992; Larsson and Frisk, 1999; Broberg et al., 2001; Ivarsson, Svalander and Litlere, 2003).

Sixty-two parents completed the Swedish version of the CBCL and 37 adolescents completed the Swedish versions of the YSR and CDI. Data collection was achieved in two ways: One group (56 parents, 32 adolescents) completed the surveys in connection with a visit to the clinic. The other group (6 parents, 5 adolescents) completed the surveys at home and mailed them to the clinic.

To study the use of myoelectric prostheses in children 8-18 years old, both parents and adolescents reported how much the prosthesis had been used in the last six months and in the last two weeks on the Prosthetic Use Scale (PUS). The PUS is a clinical tool for follow-up of the use of prostheses. The reports on the PUS by parents and adolescents were compared.

3.3 DATA ANALYSES

Agreement between nominal and ordinal variables was estimated with the *Kappa* statistic, using different weight models (**Studies I, IV and V**).

Analysis of differences between the study group and hypothesized mean values from the reference population was performed by Student's one-sample t-test (**Study V**), whereas analysis of differences in mean values concerning continuous variables was done by Student's t-test for independent groups when two groups were concerned (**Study II**) and by one-way analysis of variance (ANOVA) when three or more groups were being compared (**Study V**).

For estimation of sensitivity and calculation of 95% confidence intervals (CI), analysis of proportions was applied (**Study I)**.

The completeness and coverage of two different registers were estimated in a capture-recapture model (**Study I**).

The relationship between categorical data was analysed by Fisher's exact test (**Study II**), and the relationship between continuous variables was assessed in a multivariate analysis using multiple linear regression (**Study II**). For bivariate analysis of item calibrations, Pearson's product moment correlation was calculated (**Study III**). 'Bland-Altman' plots illustrated the relationship between differences and averages in pair-wise readings and measures (**Study IV**).

For these analyses the software SPSS (Statistical Package for Social Sciences) version 11 was used.

For the Rating scale analysis, Rasch measurement analyses were performed, using the software FACETS (version 3.1 and 3.49). In **Study III** a two-facet rating scale model with four response categories was used, and in **Study IV** a three-facet rating scale model with the same response categories.

Statistical significance was evaluated against a limit of 0.05 for the p value.

4 SUMMARY OF RESULTS

4.1.1 Study I

In Study I we found that 125 children who were born during the period 1973-1987 were registered as having ULRD in the clinic-based register at the LDAPC. Eight of these children were not found in the population-based register (SRCM). Thus we estimated that the completeness of the SRCM was 94% (95% CI 89-98%).

By re-classification of the cases in the SRCM, further comparisons of the children in the registers were possible. Comparable data were obtainable in 115 cases. From this comparison, the agreement regarding laterality between the registers could be considered as almost perfect (*kappa* 0.98) and the proportion of cases with perfect agreement was 99%. The inter-register agreement in classification of the type and level of deficiency was substantial (*kappa* 0.72 to 0.79).

4.1.2 Study II

In Study II, the presence of scoliosis and trunk asymmetry was studied in 60 persons with TULRD. None of these persons had vertebral anomalies or structural abnormalities of the ribs, but four persons had a deviating number of ribs (two had 11 ribs and two had 13 ribs). Nineteen persons (31%) had a scoliosis of between 10 and 19º and 30 persons had minor curves of between 5 and 10º.

Among the 60 participants, besides the degree of scoliosis the ATR was determined in 46 persons and the degree of LLI was measured in 40 persons. Surprisingly, there was no correlation between degree of scoliosis and ATR. This indicates that a common screening method such as scoliometry is not a useful tool for screening children with TULRD for scoliosis. There was a significant correlation, however, between LLI and side of the convexity, with the convexity directed towards the side of the shorter leg in 21 of 28 persons. This indicates that the scoliosis is of postural origin.

The most important conclusion to be drawn from this study is that deviations in the spine in children with TULRD do not immediately imply that they have a structural scoliosis but rather that they have a transient scoliosis of postural origin of no clinical significance.

4.1.3 Study III

In Study III, to validate the ACMC the performance on the ACMC items was evaluated in 75 persons, 2 – 57 years of age. The persons were assessed one to 9 times (median two times), resulting in 210 assessments.

Validity concerning content, response processes and internal structures of the ACMC were obtained. Firstly, the goodness-of-fit of the items (100%) demonstrated that the relationship among items conformed to the construct, i.e., the items formed a unidimensional construct. Secondly, the person fit statistics demonstrate that >95%

of the persons showed valid response patterns across items that were consistent with the intention of the instrument. Thirdly, the targeting of the persons' ability to the difficulty of the items (Fig. 1, Study III) showed that the items match the ability of the persons.
Item calibrations were stable and correlation analysis confirmed the relation between the two sets of item calibrations, demonstrating no violation of local independence from use of repeated assessments.

The separation of persons into five, and items into 16, distinct strata, as well as a finding of reasonable *SE* (standard error) values for both persons and items, suggest adequate internal reliability. The results from plotting of ACMC measures over time in ten persons (Fig. 2, Study III) demonstrate that the ACMC is sensitive to change.

The results from this study show that the ACMC is a sensitive, valid and reliable instrument for measurement of myoelectric prosthesis control functions in both children and adults with congenital or acquired TULRD.

4.1.4 Study IV

In Study IV, the external reliability of ACMC was established by scorings from three raters with different degrees of experience on 27 videotapes of client performance.

The results show that the most experienced rater had excellent intra-rater agreement, with no systematic difference in her ratings between session 1 and session 2. The student with some clinical experience had excellent intra-rater agreement but tended to limit her use of the rating scale in session 2. Also, there was a slight systematic difference in her assessments between session 1 and session 2, with results pointing in different directions depending on the client's ability. The rater with no previous experience of users of myoelectric prostheses displayed a low intra-rater agreement. Rasch analysis showed that she gave unexpectedly high or low scores and hence misfitted to the model for ACMC. Her calibrated severity in ratings decreased from session 1 to session 2; the difference in her assessments between sessions 1 and 2 pointed to more severe ratings for the more able clients in session 2 (Fig. 2, Study IV).

As expected from the rater calibrations, the inter-rater agreement was only fair. However, the two raters with clinical experience displayed only minor systematic differences between their client measures, indicating that the less experienced rater assigned higher scores to the more able clients and lower scores to the less able clients compared with the most experienced rater. Furthermore, both experienced raters deviated systematically from the rater with no clinical experience, with differences pointing in the same direction (Fig. 3, Study IV).

Hence, the major finding in this study was that in order to obtain reliable measures from the ACMC the raters have to have some experience of this group of clients. Until the ACMC can adjust for rater severity, the same rater should perform the ACMC when it is used for follow-up or clinical trials.

4.1.5 Study V

In Study V, 62 parents and 37 adolescents answered questionnaires regarding the children's prosthetic use, competencies, problems and affective state. The outcome of the ratings on myoelectric prosthetic use on the PUS shows that most of the children (n=49) use their prosthesis to varying degrees. Further examination of the children who did not use the myoelectric prosthesis (n=13) revealed that some of the girls used, instead, a passive, cosmetic prosthesis, whereas most of the boys with partial hand deficiency had abandoned the prosthesis and managed by using their small hand.

Important findings in this study are that, overall, children with TULRD who have been supplied with a myoelectric prosthetic hand exhibit social competence and behaviour/emotional problems similar to Swedish standardized norms. In this group of children, however, the frequency of withdrawn behaviour was significantly higher than the norm. Also, girls and older children displayed lower social competence and social activity, respectively. The relation between prosthetic use and psychosocial adaptation differed in boys and girls.

5 DISCUSSION AND IMPLICATIONS OF FINDINGS

5.1 THE DEFICIENCY

5.1.1 Prevalence of ULRD

By applying the results from Study 1 to the material from the SRCM for the period 1973-1987, we can now estimate the number of Swedish children born each year with ULRD.

Including the children reported in Study I, the reclassification of all infants reported to the SRCM during the study period (1973-1987) as having ULRD resulted in information regarding 617 infants. Taking into account the calculated underestimation of 6%, it was concluded that 656 children with ULRD were born during this period. This corresponds well to the capture-recapture estimate of the total number of children with ULRD, 659 (95% CI 632-686). By a detailed scrutiny of the notification cards, additional information regarding the children in the SRCM was obtained. Out of the 617 children, there were 524 who survived the perinatal period. In this section, data on these children will be presented. This information has not been given previously in this thesis.

With consideration paid to the under-reporting of 6% (95% CI 2-11%) to the SRCM, 557 (95% CI 535-589) live-born children with ULRD were delivered in Sweden during this 15-year period; and the average number of children born annually with ULRD, among live-births, was thus 37 (95% CI 36 – 39). With the average birth-rate in Sweden of 100 000 children/year, this highly corresponds to the prevalence of ULRD of 3.8/10 000 live births reported by Froster and Baird (1992).

5.1.2 Characteristics of ULRD

The results of the reclassification of all cases in the SRCM indicated that in a majority (47.3%) of the live-born infants with ULRD entered in the SRCM the deficiency was left-sided, in 34.5% it was right-sided and in 18.2% it was bilateral (Table 2). The results from Study I showed almost perfect agreement between the SRCM and the LDAPC register regarding the laterality of the cases (*kappa* 0.98). Thus, the distribution by laterality in Table 2 is highly probable.

Table 2. Live-born infants with upper limb reduction deficiency in Sweden 1973-1987, distributed according to side and type of deficiency (under-estimation 6% not taken into account)

Side/Type	Left	Bilateral	Right	Total
Transverse	60.7%	7.9%	31.4%	290 (55.3%)
Longitudinal	30.8%	30.8%	38.5%	234 (44.7%)
Total	47.3%	18.2%	34.5%	524 (100%)

By reclassification of the 524 infants in the SRCM, it was found, further, that 290 children had TULRD and 234 children had longitudinal ULRD (Table 2). According to Study I, there was substantial agreement between the SRCM and the LDAPC register concerning the type of deficiency (*kappa* 0.78).
Detailed information on ULRD is, however, still lacking. The results from the estimation of agreement between the LDAPC and SRCM registers indicate that the information in the SRCM is not detailed enough to make a precise description of the deficiencies and hence to provide an estimation of the prevalence of different levels of ULRD.

However, through the distribution of the cases from the SRCM regarding type and laterality of ULRD, some interesting information emerged. It was found, for instance, that the left-sided dominance is even greater among the transverse deficiencies (60.7%) compared to the longitudinal deficiencies (30.8%). This has not been shown earlier, although it has been estimated in earlier studies that the left side predominates in transverse deficiencies.

The finding that there were few bilateral cases among the TULRDs (7.9%) compared to the longitudinal ULRDs (30.8%) supports the theory that the two types of ULRD have different aetiologies.

5.1.3 Classification of ULRD

A secondary finding in Study I was that the ISO method (International Organization for Standardization, 1989) is probably inadequate for classification of transverse deficiencies in the hand. In the clinic-based register we found no case of total absence of fingers alone. Total absence of fingers was always associated with partial absence of metacarpals 2-4 or 2-5. Likewise, when all metacarpals were present, there was always part of the thumb and sometimes also the little finger. When the ISO classification is strictly applied, a case with a hand of this kind should be classified as a “cleft hand”, i.e. a longitudinal deficiency.

On the basis of theories concerning a vascular aetiology of transverse deficiencies, however, the deficiency described here is probably an atypical cleft hand, or, rather, a symbrachydactylous hand (Flatt, 1994). In a recent study (Golden et al., 2003) it was demonstrated that the vascular mechanism underlying a TULRD is more likely to affect one or two middle fingers, rather than all five fingers. Furthermore, according to the centripetal suppression theory of Maisels (Flatt, 1994), in a typical cleft hand the last finger to be present in the hand is the little finger. In symbrachydactyly, in contrast, the single finger is the thumb. Hence, to apply the ISO classification to TULRDs in the hand, revision of the method is warranted.

5.1.4 Scoliosis in TULRD

The results from Study II showed that there are small, but evident, degrees of trunk asymmetry, leg length inequality and scoliosis in children with TULRD. However, a

mean degree of trunk asymmetry of up to 5° in school children (Nissinen et al., 1989) and LLI of up to 1 cm occur so frequently that they may be considered "physiological variations" (Vercauteren et al., 1982). The small spinal curves (5-10°) in half of the group of children with TULRD correspond to earlier reports on this deviation of the spine in 54% of school children.

The high frequency of scoliosis with Cobb angles ≥10° in the children with TULRD (31%) cannot, however, be explained by the comparisons with normally developed children. In Sweden, the prevalence of scoliosis in school girls is estimated to be 3.2%, and in boys 0.5% (Willner, 1990). The lack of correlation of the convexity with side of the deficiency is interesting. In previous studies in persons with TULRD (Waldenlöv et al., unpublished manuscript; Greitemann, Guth and Baumgartner, 1996), the convexity of the spine was in most cases directed towards the deficient side. This difference from earlier results may be explained by the influence of LLI. As reported earlier (Hult, 1954; Friberg, 1983; Nissinen et al., 1993), the lumbar convexity is generally directed towards the shorter leg. This was also found in Study II, where either the rib hump or the lumbar prominence was directed towards the shorter leg. In that study, the right leg was shorter in 50% of the children, indicating that the direction of the convexity should go towards the right side. However, as pointed out earlier, in TULRD the majority of the deficiencies are on the left side. These two factors may interact and thus explain the non-correlation between the direction of the convexity of the spine and the side of the deficiency.

Another interesting finding in this study was that in 11 of 14 persons with scoliosis ≥10º, the rib hump was on the non-deficient side. In an earlier study of trunk asymmetry in children with TULRD (Waldenlöv et al., unpublished manuscript), we found a significant difference in scapula size between the non-deficient and the deficient side. We also found that the children had a non-symmetrical posture, as was also reported by Greitemann et al (1996). The persons with acquired or congenital TULRD seem to have an elevated shoulder on the deficient side.

The most possible explanation for the high prevalence of scoliosis in children with TULRD is a functional correction in an attempt to compensate for the uneven weight distribution.

5.2 THE USE OF PROSTHESES

5.2.1 Assessment of Capacity for Myoelectric Control

In Studies III and IV a test to determine a child's capacity to control the prosthesis in daily tasks was developed. The results were encouraging, though some issues remained for further investigation.

5.2.1.1 Validation of the ACMC

In Study III, we used any bimanual task that the child would like to perform for validation of the ACMC. The items were found to represent the capacity for myoelectric

control and form a hierarchy of increasing difficulty. There is uncertainty, however, as to whether the difficulty of the items remains stable across different situations or types of task. It may be possible, for example, that the difficulty of a specific item is lower in tasks that are considered easy and higher in tasks considered to be more difficult. In a future study, tasks that usually are performed by children of varying age will be specified and described for further validation of the ACMC.

5.2.1.2 Reliability of the ACMC

The internal reliability of the ACMC has been established (Study III) and the external reliability in terms of inter- and intra-rater reliability has been demonstrated (Study IV). The results of both Study III and Study IV indicate, however, that revision of the manual is desirable. Information about the necessity of scoring all observable items needs to be clarified. Further, the definition of certain items (numbers 4, 6, 9, 16, 17, 19, 22 and 35) needs to be improved. This will be addressed in the near future.

In addition, cross-cultural validation and reliability estimations are needed in order to make the use of the ACMC available for clinicians in other countries than Sweden. Furthermore, the issue of rater severity needs to be addressed. To be able to compare the results from two raters on the same client, we need to be able to adjust the client's ability measure according to the rater severity by means of the rater severity calibration. This has been done before in another Rasch-derived instrument (Fisher, 2003) and should further improve the ACMC.

5.2.1.3 Sensitivity of the ACMC

To evaluate the sensitivity of the ACMC, two groups of participants were identified in Study III. The first group comprised participants who had used a myoelectric prosthesis prior to the study and whose ability measures were thus expected to remain relatively stable over sessions. The other group consisted of participants who were being fitted for the first time with a myoelectric prosthesis; their ability measures were expected to increase over time/session. Ten participants who had undergone at least six assessments and who represented persons of different ages (3-39 years old) and gender (4 males, 6 females) were identified. Three of these ten persons were experienced prosthetic users (group I) and seven were new users (group II). In this study, we found that in new users with additional problems (group II b) the ability measures did not increase at the same rate as in the other persons in group II (Study III, Fig. 2).

In order to demonstrate the intra-individual orders of the ACMC measures in the same participants as in Study III, the participants' individual ability measures were plotted in each group. The results showed unexpected peaks and drops in ability measures in some clients (Figs. 8-10). Information from the clinical records of the clients suggests that these fluctuations originated either from the prosthetic socket fit, a tight fit being related to a high ability measure and a loose fit to a low ability measure, or from a break in continuity of prosthetic training or use, resulting in a low ability measure.

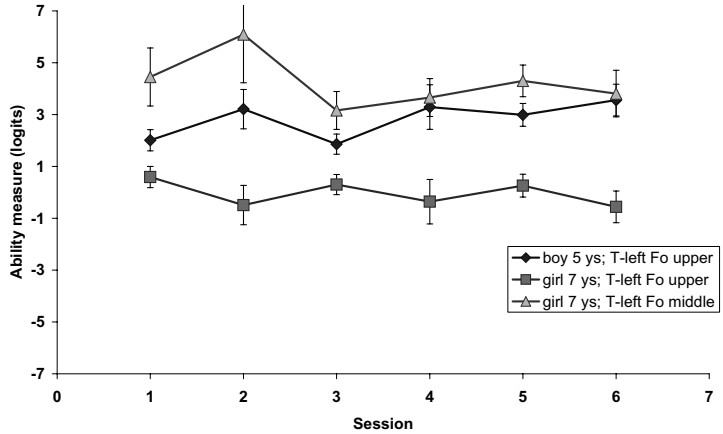

Figure 8. Individual ability measures for experienced prosthetic users (group I).
Note: Capped bars (whiskers) indicate mean standard error of the measurement. T= transverse. Fo= forearm. Upper/middle= level of deficiency.

The results of this plotting of the individual ACMC measures demonstrate that the ACMC is a sensitive and clinically applicable tool, both for evaluating improvements and for detecting changes in ability related to prosthetic fit. To confirm these findings, further controlled studies will be performed in larger groups in order to examine the responsiveness to treatment and prosthetic adjustment.

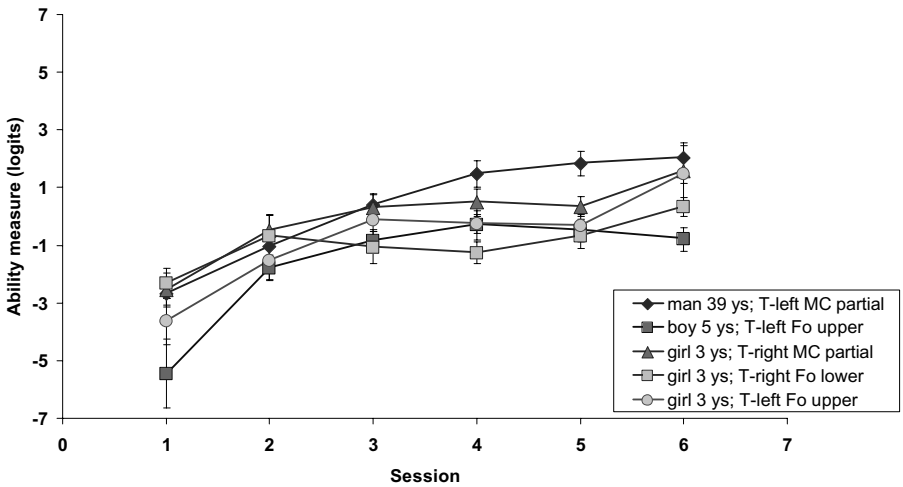

Figure 9. Individual ability measures for new prosthetic users with expected improvements (group II a).
Note: Capped bars (whiskers) indicate mean standard error of the measurement. T= transverse. Fo= forearm. MC= metacarpal. Upper/lower/partial= level of deficiency.

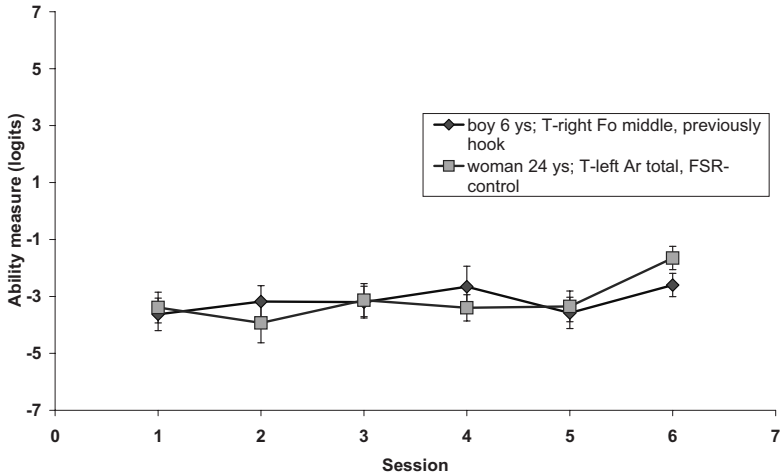

Figure 10. Individual ability measures for new prosthetic users with additional problems (group II b). Note: Capped bars (whiskers) indicate mean standard error of the measurement. T= transverse. Ar= arm. Fo= forearm. Total/middle= level of deficiency. FSR= force sensitive resistor.

5.2.2 Prosthetic use

In most prosthetic outcome studies (e.g. Ballance, Wilson and Harder, 1989; Day, 1992; Hubbard, 1992), and also in studies on the psychosocial adjustment in children with ULRD (Varni et al., 1989b), the researchers measure prosthetic use to decide if there are any differences between clients with different patterns of prosthetic use. This measure is conventionally an ordinal scale with numbers indicating the length of time in hours for which the prosthesis is usually worn. In Örebro, we use the Prosthetic Use Scale (PUS), whereby the prosthetic use is recorded on a five-degree scale ranging from full-day use to no use.

To obtain information on whether, and if so to what extent, the children in Study V were using a myoelectric prosthesis, we asked the parents to indicate the children's level of use on the PUS. The result showed that 79% of the children 8-18 years old were still using their myoelectric prosthesis, though to a varying extent. Twenty-one per cent of the children had rejected the prostheses, some in favour of a passive, cosmetic, prosthetic hand and others in order to use their residual hand- or arm-stump.

The length of the residual limb seems to be an important factor for the use or rejection of a myoelectric prosthesis. Postema et al (1999) reported a rejection rate of 34 % in children 1-22 years of age. In their sample, the rejection rate was highest (67%) among children with trans-carpal or more distal TULRDs. This was also shown in another study (Hubbard, Kurtz, Heim and Montgomery, 1998), where the rejection rate in unilateral TULRDs was highest (52%) in children with wrist disarticulations and only 30% in below-elbow TULRDs. Overall, the rejection rate in the LDAPC sample seemed to be low, compared to that in other centres.

As mentioned earlier in this thesis, different limb-fitting centres have slightly different ways of supplying children with prostheses. For example, in some centres the children are provided with a body-powered hook prior to the myoelectric fitting. However, since the control systems in body-powered and myoelectric prostheses are completely different, the change from one system to another may lead to rejection of a prosthesis. This may explain the high rejection rate (31%) in children reported from England (Datta, Kingston and Ronald, 1989).

The myoelectric prosthesis has the advantage of combining function and cosmesis. There are, however, tasks in which the use of the myoelectric hand can be awkward or dysfunctional. As suggested earlier (Crandall and Tomhave, 2002), in these cases the children may be provided with an additional, task-specific, prosthesis, e.g. for horse-back riding, shooting or hockey.

Through the development of the ACMC we are now able to study the relation between prosthetic use and capacity for control, in order to determine whether the length of the prosthesis-wearing time affects the capacity for myoelectric control.

5.3 THE WELL-BEING OF CHILDREN WITH MYOELECTRIC PROSTHESES

5.3.1 Consequences of TULRD

As mentioned earlier, children with TULRD are at risk for severe psychosocial co-morbidity related to the loss of function and physical appearance (Varni and Setoguchi, 1992). In Sweden, to prevent this most children with TULRD are provided with a prosthesis that is functional and has an appearance similar to that of a normal hand, i.e. a myoelectric prosthetic hand.

The results from Study V show that among children 8-18 years old, most children who have been provided with a myoelectric prosthesis at an early age continue to use the prosthesis. These children also exhibit overall social competence and a behaviour/emotional problem score comparable to those in Swedish children in general. This indicates that the provision of a myoelectric prosthesis to children with TULRD can lessen the adverse psychosocial implications of the deficiency.

A significantly high score for withdrawn behaviour in all children with a myoelectric prosthetic hand points, however, to the fact that a prosthesis alone cannot lessen the constraint from living with TULRD. Further studies on other groups of children with TULRD who have not been provided with a prosthesis may add more evidence to this issue.

On the basis of the results of Study V, we suggest that children with TULRD at levels proximal to the metacarpals should be fitted with myoelectric prostheses at the age of 2 ½ - 4 years. The questions of gender and the social impact of the deficiency should be given greater consideration in prosthetic fitting and training.

SUMMARY IN SWEDISH
(POPULÄRVETENSKAPLIG SAMMANFATTNING PÅ SVENSKA)

Reduktionsmissbildning av arm eller ben (LRD) är ett ovanligt tillstånd hos nyfödda. Studier av antik konst och litteratur visar att det förekommit så långt tillbaka i tiden som på 600-talet före Kristus. Det finns rapporter om LRD från alla delar av världen. Förekomsten av LRD är relativt stabil – en nyligen gjord sammanställning visar att det föds mellan 2-7 barn med LRD per 10 000 nyfödda i olika länder. Skillnaden i förekomst kan troligtvis förklaras med olika metoder för rapportering och klassificering av LRD. I Sverige har förekomsten av LRD beskrivits som 6.25/10 000 nyfödda. Armreduktions missbildning (ARM) är ungefär dubbelt så vanlig som benreduktionsmissbildning.

Klassificering av LRD kan variera. Ett system för klassificering av LRD är en internationell standard (ISO 8548-1:89). Enligt det systemet delas LRD in i två huvudgrupper, de transversella (tvärgående) och de longitudinella (längsgående). Det aktuella avhandlingsarbetet berör barn med transversell LRD. Orsakerna till LRD kan vara flera. Vid transversell LRD är de flesta forskare i dag eniga om att en störning i blodförsörjningen till arm- eller benanlaget, eller den färdigutvecklade kroppsdelen, kan vara en orsak till LRD.

Armreduktionsmissbildning är ett livslångt tillstånd som hos ett drabbat barn kan leda till praktiska svårigheter, sociala begränsningar och fysiska problem. Det övergripande syftet med denna avhandling var att öka kunskapen om barn med ARM från tre perspektiv: missbildningen, protesanvändningen och välbefinnandet hos de drabbade barnen.

För att kontrollera information rörande ARM i det svenska Missbildningsregistret (SRCM), omklassificerades alla nyfödda barn som rapporterats till detta register under perioden 1973-1987 i enlighet med ISO-klassifikationen. Resultatet jämfördes med uppgifter i ett register vid dysmeli- och armprotesenheten, Universitetssjukhuset i Örebro. Resultaten visade att 117 av 125 barn fanns i SRCM, dvs. en under-rapportering till SRCM på 6%. Informationen i SRCM var tillräckligt detaljerad för att göra en uppskattning om hur LRD hos svenska barn är fördelad över sida och typ av LRD.

Förekomst av skolios[1] och bålasymmetri studerades hos 60 personer med transversell ARM. Här fann vi att 19 personer (31%) hade en skolios på mellan 10 och 19º. Det fanns ett statistiskt säkerställt samband mellan skillnad i benlängd och sida av ryggkröken, med kröken riktad mot det kortare benets sida hos 21 av 28 personer. Det tyder på att barn med transversell ARM kan ha en flexibel skolios orsakad av skillnad i benlängd som inte behöver någon behandling.

En ny metod för att mäta förmågan att kontrollera en myoelektrisk[2] protes (ACMC) under utförande av vardagliga aktiviteter har utvecklats. För detta genomfördes 210 bedömningar av barn och vuxna med myoelektrisk protes. Resultaten från de personer som testades och de punkter som bedömdes visar att ACMC är användbart för att pröva förmågan att styra en myoelektrisk protes, både hos barn och vuxna.

Samstämmigheten mellan flera bedömare av ACMC prövades genom att tre arbetsterapeuter med varierande erfarenhet av personer med myoelektrisk protes fick göra bedömningar från 27 video-inspelade test-situationer. De tre bedömarna fick samma information och träning innan bedömningen. Resultaten visar att det krävs en viss erfarenhet för att göra samma bedömning, både vid upprepad bedömning av samma person och vid jämförelse med en annan bedömare. Olika arbetsterapeuter är olika stränga i sin bedömning. Det är något som vi behöver ta hänsyn till om ACMC skall användas för bedömning av en person vid olika tillfällen.

För att studera den psykosociala anpassningen hos barn med ARM som har fått myoelektrisk protes gjordes en undersökning av kompetens och beteende hos 62 barn. Vid en jämförelse med uppgifter från svenska barn i allmänhet fann vi att barnen med ARM sammantaget hade en lika god hälsa som andra barn. En mer detaljerad analys tyder dock på ett socialt stigmata relaterat till ARM vilket bör hanteras olika för pojkar och flickor. De flesta barn som fått myoelektris protes som barn fortsätter att använda den (79%).

Sammanfattningsvis, armreduktionsmissbildning är ett ovanligt tillstånd där olika problem kan uppkomma under uppväxten. Dessa behöver beaktas av det professionella team som följer barnet under dess utveckling. Utrustning med protes i tidiga år tycks minska en del av de svårigheter som kan vara relaterade till denna missbildning.

[1]Skolios = sjuklig krökning av ryggraden i sidled, ofta med inslag av rotation i bålen.
[2]Myoelektrisk protes = protes med en muskelstyrd elektrisk proteshand.

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