UPPER LIMB REDUCTION DEFICIENCIES IN SWEDISH CHILDREN

Classification, prevalence and function with myoelectric prostheses

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Stockholm and Örebro 2004
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Stockholm and Örebro 2004
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 psychologically 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.


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

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).

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.

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

![Figure 3. Child with transverse upper limb reduction deficiency, left, forearm upper third.](image)
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.

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 PUF1 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).

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.

Figure 7. Adaptive equipment, a so-called “spatula”.

Figure 7. Adaptive equipment, a so-called “spatula”.
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.

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

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 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.

\begin{table}[h]
\centering
\begin{tabular}{lccc}
\hline
\textbf{Side/Type} & \textbf{Left} & \textbf{Bilateral} & \textbf{Right} & \textbf{Total} \\
\hline
\textbf{Transverse} & 60.7\% & 7.9\% & 31.4\% & 290 (55.3\%) \\
\textbf{Longitudinal} & 30.8\% & 30.8\% & 38.5\% & 234 (44.7\%) \\
\textbf{Total} & 47.3\% & 18.2\% & 34.5\% & 524 (100\%) \\
\hline
\end{tabular}
\caption{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)}
\end{table}
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 ≥10°, 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.
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 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.

**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.
3.2.2 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.

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.
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.
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 allra delar av världen. Förekomsten av LRD är relativt stabil – en nyligen gjord sammenställning 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 beskrivts 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.


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 armprotestenheten, Universitetssjukhuset i Örebro. Resultatet 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 skolios1 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 ryggraden, 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 myoelektrisk2 protes (ACMC) under utförande av vardagliga aktiviteter har utvecklats. För detta genomfördes 210 bedömningar av barn och vuxna med myoelektrisk protes. Resultatet 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.

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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 myoelektrisk 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.

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

I would like to express my sincere gratitude to all those who have contributed to the preparation of this thesis, especially to:

My main supervisor, Associate Professor Ann-Christin Eliasson, for all professional guidance, constructive criticism and support. Your creative mind, free from traditional thinking, and your ability to think on new lines have been a great source of inspiration for me.

Professor Lennart Bodin, my secondary supervisor, for your great support even in the darkest of moments. With your wide scientific experience and your statistical expertise you have always been there for me, ready to support and advise me.

Professor Hans Forssberg, my other secondary supervisor, for taking me into the Neuropediatric Research Unit. This unit is characterised by your open mind for research in many different areas of neuropaediatrics, and I am proud to have been a postgraduate student there.

My late supervisor, Lars Wranne, who introduced me to postgraduate studies and paediatrics. His death in 1995 was a great loss to me, resulting in a couple of years without progress in this work.

My co-authors Lars Samuelsson, for sharing with me his knowledge in paediatric orthopaedics and entering the project of scoliosis in TULRD, and Lars Norén, for fruitful collaboration.

My co-authors Professor Anne Fisher and Professor Birgitta Bernspång from Umeå University. Thank you for introducing me to Rasch analysis and for giving me a deeper knowledge of occupational therapy theories and models.

My co-author Professor Ingemar Engström, for support and for kindly sharing with me his considerable knowledge in child-psychiatrics.

Rolf Sörbye, former head of the Department of Neurophysiology, for being a role model and mentor for my clinical work. Your international pioneer work in the early 1970s had an enormous impact on the life of many children around the world. Without your contributions this thesis would not have been accomplished.

Kristina Waldenlöv, colleague and dear friend, for creative clinical discussions through the years, resulting in development of the care of our patients. Thank you for all happy laughs, all your support and your contributions to the production of this thesis.

Sven-Olov Pettersson, CPO, for friendship and fruitful co-operation over the years. You introduced me to the field of prosthetic technology, and I promise I will never try and make a prosthesis again!
Professor Jens Schollin, former head of the Department of Pediatrics, and Jürgen Bensch, neuropaediatrician, for creative discussions, support and encouragement over the years.

Colleagues, friends and staff – past and present – in the multi-disciplinary team at the Limb Deficiency and Arm Prostheses Centre, for encouragement and help with collecting material.

All the children and their parents who willingly participated in the studies.

“Protes-tantera”, Ingela Andersson and Anna-Carin Öjteg, and, again, Kristina Waldenlör, for many years of creative teamwork and highly valued friendship. You have all contributed to the preparation of this thesis.

Katarina Helgesson, hand surgeon and friend, for sharing with me your considerable knowledge in hand malformations, and for encouragement, support and inspiring discussions over the years.

Lena Krumlinde-Sundholm, colleague and friend, who was my fellow doctoral student and partner in our search for the “true measure”.

The staff at the Medical Library, Örebro University Hospital, for providing me with the books and papers I have needed, Maud Marsden, Uppsala, for excellent linguistic revision of this work, and Anders Magnusson for statistical advice and help with the SPSS.

The staff at the Department of Medical Photography, Örebro University Hospital for providing me with the photographs I have needed.

Hampus Hillerström, Geneva, for helping me with the communication with Musée d’art et d’histoire, Ville de Genève.

My fellow doctoral students in the Research Pavilion, Örebro University Hospital, for educational discussions and for great support and friendship, and my colleagues at the Neuropediatric Research Unit, Karolinska Institutet, for support and fellowship.

The staff at the Department of Pediatrics, Örebro University Hospital, for encouragement and support over the years.

All my friends and hunting companions, for providing me with silence, fresh air, beautiful scenery and contemplation on life and research.

My dear parents, my father David and my late mother Margaretha, my step-mother Gudrun, my sisters Eva and Cecilia, and my brother Bosse, who have always encouraged me and supported me.

Last, but not least, the most important persons in my life, my wonderful family – Lars, Maria and Johan. I love you!
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Upper limb deficiencies in Swedish children—a comparison between a population-based and a clinic-based register

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Received 7 November 2000; received in revised form 23 March 2001; accepted 27 March 2001

Abstract

\textbf{Objective:} To validate information in the Swedish Register for Congenital Malformations (SRCM). \textbf{Methods:} A comparison was made with a clinic-based register kept at the Limb Deficiency and Arm Prosthesis Centre (LDAPC). The report frequency and the quality of the information in SRCM were analysed. Cases were classified according to a detailed, clinically relevant classification, the ISO 8548-1:89 method. \textbf{Outcome measures:} The completeness of SRCM was first estimated. The Kappa statistic was then used to assess the agreement between the two registers regarding individual categories and across all categories. \textbf{Results:} For the period 1973–1987, we found 125 cases of upper limb reduction deficiencies (ULRD) in the clinic-based register, of which 117 was found in the national register. The completeness of SRCM was thus estimated to be 94\% (95\% confidence interval 89\%–98\%). The inter-register agreement varied from almost perfect agreement in laterality of deficiency (Kappa 0.98) to substantial agreement in type and level of deficiency (Kappa 0.72–0.79). For specific levels of transverse deficiency, however, the agreement varied between –0.05 and 0.66. \textbf{Conclusions:} The results 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

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PII: S0378-3782(01)00163-3
clinical purposes could therefore result in lower validity. Additional information and follow-up of specific cases are therefore recommended. © 2001 Elsevier Science Ireland Ltd. All rights reserved.

Keywords: Upper limb; Deficiency; Children; Register validation

1. Introduction

To study the epidemiology of upper limb reduction deficiencies (ULRD) for clinical purposes, detailed descriptions of the reduction, side and number of affected limbs are essential. This is difficult to achieve since not all children with ULRD are available for detailed studies. However, a national register may cover the population. But there is some uncertainty in using register data with respect to the sensitivity in the frequency (completeness) of reporting. Also, the information concerning the limb deficiency in the register may lack details, making an accurate description difficult. By comparing register data with clinical data, validation may be achieved.

Limb reduction deficiencies (LRD) have been recognised and described for more than 2000 years [1]. The thalidomide tragedy in the beginning of the 1960s [2], when an unexpectedly large number of children with a certain pattern of LRD were born, prompted the establishment of many surveillance registers. These are hospital-based, reporting from a number of selected hospitals, or population-based, covering the total population in a certain area [3]. The surveillance registers report from a population of births [4–9] or from a population of livebirths [10–13]. According to Källén et al. [3], no clear difference in rate of ascertainment has been shown between hospital-based and population-based registers. Other registers based on specialised clinics, such as limb-fitting centres or hand-surgery clinics [14–16], are biased by the selection of cases. There is no surveillance function applicable to these registers since the time between birth and reporting to the register is too long.

Since most studies on limb deficiencies aim at estimating the prevalence of a deficiency and describing its features, they utilise either existing or newly designed methods for classification. Some are based on morphology [17–21]. Others are based on a hypothesis in search for possible aetiological factors [22–24]. By adjusting an existing classification or creating their own, most authors thereby make comparison between reports difficult. Recently, the Classification Committee of the International Clearing-house for Birth Defects Monitoring Systems proposed a classification [25] for congenital anomaly surveillance systems. The Committee suggests that to classify limb defects, a descriptive classification is the only way. An additional type of classification, similar to the descriptive morphological type, is that based on clinical principles [26]. An attempt at a uniform, clinically applicable classification, based on these principles, was suggested by a working group from the International Society for Prosthetics and Orthotics [27]. This classification is not intended for surveillance registers but for specialised clinic registers, such as hand surgery clinic registers or limb fitting centre registers. It is based on more detailed descriptions of the anomalies than those needed for the surveillance registers. This classification was later accepted as an international standard, ISO 8548-1:1989 (E) [28]. Because of the clinical applicability, we used the ISO-method for classification of deficiencies in this study.
In Sweden there are two national systems for registering birth defects. The Medical Birth Record System (MBRS) is based on a standardised delivery record system. It contains a variety of information on virtually all deliveries and infants born in Sweden since 1973. Each infant can have a maximum of seven diagnoses, including that of any malformation. The malformation diagnoses are given as ICD codes; during 1973–1986 they were given as ICD-8, with an extra digit for further specification.

The other register, the Swedish Register of Congenital Malformations (SRCM), was established to allow early detection of changes in the incidence and prevalence of LRD, and to identify hitherto unknown but specific malformations or patterns of malformations [29]. The important aspect for this register is to look for deviations over time. Any such apparent deviations will need to be substantiated, and if confirmed, will need further investigation into possible cause. The SRCM largely have a public health function to warn of hazards to reproductive health. This register has been operating since 1965. All liveborn and stillborn infants with malformations observed within 1 week of delivery are reported to the register. There is a potential under-representation in this register of children with multiple limb defects, some with a lethal syndrome and some antenatally diagnosed and terminated. For international comparisons, this register uses the International Clearinghouse for Birth Defects Monitoring Systems classification [25].

In 1977, Ericson et al. [30] made a comparison of the SRCM and MBRS record systems, which suggested that there might be some under-representation in the SRCM register. They found that in 1973 and 1974, of the 82 cases with reduction malformations of the upper limbs, 56% were reported to both registers, 11% only to the MBRS and 33% only to the SRCM. Currently, the quality of diagnoses in the SRCM are controlled regularly by comparison to other sources such as patient files and other registers. The completeness of the SRCM is not important for the purpose of surveillance. However, to estimate the size of the population of ULRD in Sweden for clinical purposes, information on the completeness of the register used for the estimation is essential. No recent information on the under-representation of the SRCM is available, though.

Since the first successful fitting of a myo-electrically controlled hand prosthesis on a pre-school child in 1971 [31], children with ULRD from all over Sweden have been referred to the Limb Deficiency and Arm Prosthesis Centre (LDAPC) at Örebro Medical Centre Hospital. For 6 years, the LDAPC in Örebro was the only limb-fitting centre providing myo-electrically controlled hands for children in Sweden. The clinic now serves children with upper and lower limb deficiencies of different levels and sites. The LDAPC register covers all children with limb deficiencies present at birth referred there since 1971. The objectives of this clinic-based register are different from those of the population-based register, and are directed towards the care of individual patients. Hence, both major and minor defects are represented in the register. Presumably, the majority of the deficiencies in this register are single transversal due to the original objectives of the clinic. Children with longitudinal and multiple defects may thus be under-represented. In this register, the deficiencies are classified and recorded according to the ISO 8548-1:89 [28] method.

The present study was undertaken to compare data gathered from two different registers—the SRCM and the LDAPC registers. The frequency of reporting to the
SRCM, the reported laterality and level of deficiency were investigated in terms of sensitivity. Data on type, laterality and level of deficiency were analysed to measure the agreement between the two registers. To our knowledge, no previous systematic investigation on the agreement between a clinic-based register and the SRCM has been performed. Neither has the ISO-method for classification been used on a population-based register previously.

2. Material and methods

2.1. The registers

The Swedish Register of Congenital Malformations is a population-based national register [29]. To this register, paediatricians are to report all liveborn and stillborn infants who show signs or symptoms of malformations in the perinatal period. The notification forms are sent in monthly by the paediatrician, and a preliminary analysis and classification are made. Verbal descriptions, often supplemented with drawings or photographs, autopsy reports and X-ray examination reports, are scrutinised, coded, and entered into the computerised register. Further information about the child may be required, and thus added by direct contact with the hospital concerned. The register is closed for reporting, and a final classification is made 6 months after the birth. In this study, all reports in the SRCM coded as ULRD for the years 1973–1987 were included.

The LDAPC at Örebro Medical Centre Hospital keeps a clinic-based register. To this register, approximately 8–10 new children with upper limb deficiency present at birth, referred to the LDAPC from different parts of Sweden, are added yearly. Before entry in the register, every child is examined and classified by the medical supervisor at the LDAPC and an occupational therapist-in the children of the present study, one of the authors (LH). For the classification, physical examination and, in most cases, X-ray examination are performed. The classification is confirmed by a new examination at each follow-up. The present study covered children in the LDAPC register with ULRD born during 1973–1987.

2.2. Classification

At the LDAPC, the children are classified according to the method described by Kay et al. [27], later known as the ISO 8548-1:89 [28] method. In the SRCM register, however, the ULRD malformations are classified into homogeneous subgroups with a common pathogenetic mechanism or aetiology [29], making an immediate comparison with the LDAPC register impossible. However, the National Board of Health and Welfare keeps the notification forms on which the SRCM classification is made, and these forms made it possible to reclassify the SRCM cases in accordance with the ISO 8548-1:89, allowing comparison with the LDAPC cases. Thus, one of the authors (LH) manually evaluated all cases with ULRD found in the SRCM. Children who were also entered in the LDAPC register were not identifiable in the SRCM at the time for the classification, and thus the LDAPC cases in the SRCM were classified blindly.
The ISO 8548-1:89 method is focused on the clinical perspective. Based on the previous work by Kay et al. [27], it states that the deficiency shall be classified with regard to upper versus lower extremity and right versus left side. It shall then be further classified according to type, i.e. into either “transversal” (Fig. 1) or “longitudinal” (Fig. 2). The ISO classification follows the hierarchical structure of the limb, i.e. the proximal parts are always described prior to the distal parts.

Limbs with a transverse type of deficiency (Fig. 1) are those that have developed proximo-distally to a certain level, beyond which no skeletal remnants exist. These deficiencies are classified by the level at which they end. The long bones are subdivided into thirds, namely upper, middle or lower, and the small bones of the hand into either totally absent or partially absent. This gives a total of 15 levels, by which each case with a transverse deficiency can be classified.

In limbs with longitudinal deficiencies (Fig. 2), one or more bones are partially or totally absent, but distal parts of the limb may be present. In these cases, the limbs are classified by naming each missing or partially missing bone. In the present study, the longitudinal deficiencies are represented by the most proximal bone, partial or total affected. This gives 15 levels, by which each case with a longitudinal deficiency can be classified.

Fig. 1. Transversal deficiency—right upper limb, forearm, lower third.
In the statistical calculations of overall agreement, however, the number of levels was reduced. This was accomplished by aggregating levels according to the hierarchical structure of the limb, two or three neighboring levels joined into one category.

2.3. Technique of comparison

The presence or absence of a child with ULRD in the SRCM was checked, using the date and place of delivery combined with the mother’s personal identity number. When an infant was found in the LDAPC register but not in the SRCM, the case was regarded as non-reported. For infants present in both registers, agreement between the two classifications was checked. Side, type and level of deficiency were compared. In patients with longitudinal deficiency, the most proximal portion of the deficiency was recorded as level.

2.4. Analysis

To examine the validity of the SRCM, the sensitivity (completeness) of the register was estimated. The sensitivity refers to the coverage in the SRCM of cases identified in
the clinic-based register with respect to the frequency, laterality and level of the deficiency. Cases of limb deficiencies in the LDAPC register that were found in the SRCM were considered true positive cases (TP), while those in the LDAPC register not found in the SRCM were considered as false negative (FN). Sensitivity was then estimated as the ratio TP/(TP + FN) [32]. The estimate was supplemented with 95% confidence intervals (CI). In addition, we also applied a simple capture–recapture model [33] to estimate the total number of ULRD during 1973–1987. The estimate was also supplemented with a 95% confidence interval.

When the ISO method is applied for classifying different types of deficiencies, however, the sensitivity is not of primary interest. Here, the main focus is on the agreement between the classifications of the ULRD in the two registers. This agreement can be measured by the Kappa (k) statistic [34]. Kappa quantifies agreement beyond that of chance. It usually ranges between zero and 1.0. For the interpretation of Kappa, Landis and Koch [35] have suggested the following limits: 0.40 and below represents poor agreement beyond chance, 0.40–0.75 represents fair to good agreement, and 0.75 and above represents excellent agreement. In the present study, Kappa was calculated both to estimate the agreement in the individual categories and to serve as a composite measure of agreement across all categories. In the latter case, a weighted Kappa, i.e. a generalised version in which the relative seriousness of each disagreement was quantified, was used. We used a linear function to model the decrease in agreement across categories, that is, disagreement on two steps of the hierarchical scale was considered as twice the error of a one-step disagreement. As a complement to Kappa, we also calculated the proportion of cases with perfect agreement [34].

3. Results

A total of 125 children with upper limb reduction defects were registered at the LDAPC during 1973–1987. During the same period, 617 infants registered as having limb reduction deficiencies were identified in the SRCM.

3.1. Reporting frequency

Of the 125 children in the LDAPC register, eight were not found in the SRCM (Table 1) and were thus classified as false negative cases. The sensitivity of the SRCM for registering all true limb deficiencies varied from 67% to 100% during 1973–1987. For the whole period it was estimated as 94%, with a 95% CI of 89–98%. The capture–recapture estimate of the total number of ULRD was 659 with a 95% CI of 632–686, figures which correspond very well to the sensitivity figures. Hence, information on 117 children was available in both the SRCM and the LDAPC register (Table 1). In two cases in the LDAPC register, the reported information was insufficient in terms of type, laterality or level of deficiency. Further comparisons were thus made in 115 cases.
Table 1
Accuracy of reporting to the Swedish Register of Congenital Malformations (SRCM) compared with the register of the Limb Deficiency and Arm Prosthesis Centre (LDAPC) for 1973–1987

<table>
<thead>
<tr>
<th>Year</th>
<th>LDAPC and SRCM (true positive)</th>
<th>LDAPC only (false negative)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1973</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>1974</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>1975</td>
<td>7</td>
<td>1</td>
</tr>
<tr>
<td>1976</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>1977</td>
<td>9</td>
<td></td>
</tr>
<tr>
<td>1978</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>1979</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>1980</td>
<td>11</td>
<td></td>
</tr>
<tr>
<td>1981</td>
<td>9</td>
<td></td>
</tr>
<tr>
<td>1982</td>
<td>8</td>
<td>1</td>
</tr>
<tr>
<td>1983</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>1984</td>
<td>9</td>
<td></td>
</tr>
<tr>
<td>1985</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>1986</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>1987</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>117</td>
<td>8</td>
</tr>
</tbody>
</table>

3.2. Laterality

In the LDAPC register, there were 78 left-sided, four bilateral and 33 right-sided deficiencies. One case, recorded as a left-sided deficiency in the LDAPC file, was recorded as a right-sided deficiency in the SRCM (Table 2). The agreement in laterality between the two registers, measured by Kappa, was 0.98, which is considered as almost perfect [35]. The proportion of cases with perfect agreement was also high, 99%.

3.3. Type of deficiency

The agreement in type of deficiency between the registers (Table 3) measured by Kappa was 0.78, which is judged as substantial [35]. Further analysis was made in those cases where there was full agreement in type of deficiency between the registers.

Table 2
Agreement between the Swedish Register of Congenital Malformations (SRCM), and the Limb Deficiency and Arm Prosthesis Centre (LDAPC) register: distribution of side

<table>
<thead>
<tr>
<th>LDAPC</th>
<th>SRCM</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right</td>
<td>33 B 4 77</td>
<td>115</td>
</tr>
<tr>
<td>Left</td>
<td>1 B 4 77</td>
<td></td>
</tr>
</tbody>
</table>

Note: Perfect agreement in bold.
Table 3
Agreement between the Swedish Register of Congenital Malformations (SRCM), and the Limb Deficiency and Arm Prosthesis Centre (LDAP) register: distribution of type of deficiency

<table>
<thead>
<tr>
<th>LDAP</th>
<th>SRCM</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Transversal</td>
<td></td>
</tr>
<tr>
<td>Transversal</td>
<td>104</td>
<td>3</td>
</tr>
<tr>
<td>Longitudinal</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>Total</td>
<td>106</td>
<td>13</td>
</tr>
</tbody>
</table>

Note: Bilateral cases (n = 4, see Table 2) registered twice; perfect agreement in bold.

3.4. Level of deficiency

Among the 10 cases with true longitudinal deficiency (Table 3), perfect agreement regarding the level of deficiency was found in four cases. On analysis of the six cases showing disagreement, the cross-classified results for the two registers showed that in four cases, the higher portion of the deficiency was not entered on the SRCM notification forms. In one case, the deficiency was reported at a higher level, and in another case, one portion of the extremity (the ulna) was wrongly reported as missing. The overall weighted Kappa was 0.79, substantially high, though with a very wide confidence interval on account of the small number of observations.

Among the cases of true transverse deficiencies (n = 104, Table 3), there was none with a deficiency at the shoulder and upper-arm level (Table 4). Perfect agreement was

Table 4
Agreement between the Swedish Register of Congenital Malformations (SRCM) and the Limb Deficiency and Arm Prosthesis Centre (LDAP) register: distribution of level of transverse deficiency (n = 104)

<table>
<thead>
<tr>
<th>LDAP</th>
<th>SRCM</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1a</td>
<td>1b</td>
</tr>
<tr>
<td>1a ARMP</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>1b FATO</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>2a FAUP</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>2b FAMI</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>3a FALO</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>3b CATO</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>4a CAPA</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>4b MCTO</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>5a MCPA</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>5b PHTO</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>5c PHPA</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

Note: Abbreviations, see Appendix A. N.a. = not available as the marginal frequency was equal to zero; perfect agreement in bold.

Classification codes, where the numbers 1–5 indicate main classifications used to calculate overall Kappa, and the letters a–c indicate a more detailed level.

Kappa calculated for the individual level. Overall weighted Kappa is 0.72, see text.
noted in 48/104 cases, or 46%. The cross-classified results for the two registers showed that on the SRM notification forms, the deficiency was reported at a more distal level in 33 cases, compared with the level in the LDAPC register. However, the difference between the registers amounted to one (22 cases), two (nine cases) or more (two cases) levels. Further, the cross-classified results showed that in 23 cases, the deficiency was reported at a more proximal level on the SRM notification forms than in the LDAPC register. In these cases, the difference was one (12 cases), two (five cases) or three (six cases) levels. The overall weighted Kappa was 0.72, with a 95% confidence interval of 0.63–0.81. On the other hand, when calculated with respect to individual levels, Kappa showed considerable variation, ranging from −0.05 (FALO) to 0.66 (PHPA). Thus, on the individual level the disagreement was notable.

4. Discussion

In this study, a comparison between a population-based register, the SRM, and a clinic-based register at the LDAPC was performed. The report frequency to the SRM concerning ULRD was estimated to be 94%, and the quality of information in the SRM varied from almost perfect to substantial. Although the agreement on levels was substantial, the agreement on more detailed individual levels of transverse deficiency varied considerably. In particular, the result for deficiencies at the lower third forearm level indicates serious lack of agreement and a severe risk of misclassification. On analysis of these results, we found several possible explanations for the disagreement.

The comparisons in this study were made on a selected sample from the SRM, which may have had an effect on the results. However, the LDAPC register consists of entries for children from all Sweden reported to the SRM and referred to the LDAPC by different physicians from the whole country. In this sense, the LDAPC register is a sample from the whole of Sweden, although it is not strictly random. During the period 1973–1987, 19% of the children in the SRM were found in the LDAPC register. Most cases had unilateral transverse deficiencies at the forearm level. A possible reason for this is that referrals to the LDAPC are made with the intention that the child in question will benefit from a myo-electric prosthesis, which is the case mostly in transverse deficiencies. Results based on comparisons of longitudinal deficiencies are unreliable because of the small number of cases.

When Ericson et al. [30] compared the SRM with the other national register of birth defects, the Medical Birth Record System, they estimated the under-notification of LRD in the SRM to be 11%. In the present study, when ULRD in the SRM were compared with those in a clinic-based register, the under-notification was lower, 6% (95% CI 2–11%). The difference between these two studies may be explained by diagnostic differences between the registers and the quality of the MBRS. In the previous study by Ericson et al., some normal children were found to be reported to the MBRS register as having ULRD. These children were not found in the SRM. The under-notification of especially multiple ULRD in the SRM may also be explained by the fact that the individual malformations are not registered when they are a part of a defined syndrome. When the false negative cases in this study were reviewed, two cases were found
reported to the SRCM but not registered as limb reduction deficiencies. These were clinically diagnosed as having Poland’s syndrome. Furthermore, three cases of longitudinal, multiple limb deficiencies (one case of ulnar club-hand syndrome and two cases of ULRD in proximal femoral focal deficiency syndrome) had not been reported.

As part of the development of the standard, the ISO 8548-1:89 method for classification was tried by members of the group who initiated it. It was recognized that more extensive trials were desirable. During 1974, a preliminary trial was carried through [27]. No inter-rater reliability test has been performed, however. Neither has it previously been applied to a population-based register. The results for the classification of level of transverse deficiencies in the hand illustrate some of the difficulties encountered. It seems as if nature does not follow the structural order as presented in ISO 8548-1:89. At our clinic, we have never seen a child with total absence of fingers only. When there is absence of fingers, there is also partial absence of metacarpals. And conversely, when all the metacarpals are present, there is always a part of the fingers present. This may explain some of the disagreements in the classification of level (MCPA, PHTO and PHPA: five cases) among the transverse deficiencies between the SRCM and the LDAPC register (Table 4). Also, this is the reason why the Kappa statistic is not available for the PHTO level.

Other differences in the classification of deficiencies in the hand, and thus low agreement in individual Kappa statistics, may derive from the qualifications of the examiner. As stated by Källén et al. [3], the precision in describing the abnormalities very much depends on this factor. In the present case, it will depend on the experience of the paediatrician in identifying the embryonic structures in the hand. These structures might have a potential to grow, and thus later be classified at another level. We found 16 cases classified in the SRCM as any level from lower forearm level to part of the metacarpals, which, in the LDAPC register, were either part of the carpus, total absence of the metacarpus or part of the metacarpus.

According to Pritchett [36,37], the distal growth plates in the forearm contribute to 75–85% of the total forearm growth. Thus, children with a transverse forearm deficiency may have only one-fifth of the growth potential of the non-deficient arm, with which it is compared in the initial classification. With a total forearm length of 5 cm, a deficiency classified as a mid-forearm deficiency, for example, will have a length of 2 or 3 cm. When the total length of the non-deficient arm reaches 20 cm, the arm deficiency will be 5 to 6 cm—and will thus be reclassified as an upper-forearm deficiency. On the other hand, in cases of an absent forearm, growth plates may be present but not identifiable at birth. These limbs may attain a total length of 15–25% of that of the contralateral side. Thus, the time of the classification may to some extent explain the disagreement. There may also be differences in the individual growth potential in the remaining distal growth plates.

Accordingly, we suggest that in 49 of the 56 cases of transverse deficiencies that were classified differently in the two registers, the discrepancy can be explained either by the method of classification (five cases), differences in the skill and experience of the classifiers (16 cases) or a difference in time between the classifications (28 cases). The seven remaining cases cannot be explained by these factors and are thus to be considered as severe mis-classifications.
The definition of longitudinal deficiencies by the ISO standard is a very detailed procedure. In this study, the proximal portion of the deficiency decided the level of the longitudinal deficiencies. However, this definition proved to be sensitive enough to detect some disagreements in the definition of level of deficiency. Our findings indicate that reporters to the SRCM seem to neglect the higher portions of the deficiencies in their notification forms, in both transverse and longitudinal cases. This must be taken into consideration in the future when conclusions regarding these deficiencies are drawn from the SRCM. It should be realised, however, that the SRCM is a surveillance register whereby an attempt is made to identify changes in the occurrence of specific types of malformations, and is less concerned with actual clinical details such as the level of a transverse reduction. If a teratogenic agent that causes transverse arm reductions appears, it is rather unlikely that it will cause such reductions at one specific level.

In the analysis of the overall agreement between the diagnosis and classifications in the two registers, we have used the weighted Kappa, since it has the advantage of dividing discrepancies in classifications into different degrees of severity according to a linear ordinal scale. The ordinary Kappa statistic, without weighting, treats all discrepancies as equal in this respect. A specific reason for using the weighted Kappa is that reclassification of the level of deficiency (i.e. upper, mid- or lower third) is not unusual as the children grow. Hence, the data on level of deficiency in the SRCM may be incorrect in that respect, and upper third to mid-third discrepancies were considered to be less severe than upper third to lower third. Weighted Kappa assumes that the categories are ordered, and we therefore used weighted Kappa only in those cases where there was natural ordering of the categories.

A calculation of an overall measure of agreement with Kappa requires that the measurements be represented by relatively few distinct values. We accomplished this by aggregating the original classification into fewer subgroups. The reclassification was based solely on medical grounds and had, as its starting point, the aggregation of diagnoses that comprised neighbouring parts of the arm, going from upper arm to lower arm and hand. In this sense, the clinical classification approached the classification systems used in surveillance systems.

In conclusion, the results of this study are based on information from two registers with different aims and with different data collection processes. Our findings indicate that although the SRCM is a surveillance register, it can be used as a valuable additional data base for studying the epidemiology of ULRD in Sweden. It has an estimated rate of ascertainment of 94%, and thus gives an underestimation of 6%. In general, the agreement between the SRCM, the population-based register, and the clinic-based LDAPC register, was high as measured by the Kappa statistic. There were exceptions, however, especially in the distribution of levels of transverse deficiency. 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.

Acknowledgements

We are indebted to Professor Bengt Källén, Tornblad Institute of Embryology, University of Lund, Sweden, for giving us access to the register data and for valuable
help and discussions during different stages of the project. We would also like to thank Hans Forsberg and Ann-Christine Eliasson, Department of Woman and Child Health, Karolinska Institute, and Anders Ericson, National Board of Health and Welfare, Stockholm, Sweden, for advice and comments on the manuscript. The study was supported by grants from the Norrbacka-Eugenia Foundation, the Solstickan Foundation and Örebro County Council.

Appendix A. Table abbreviations

<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>ARMI</td>
<td>upper arm, middle third</td>
</tr>
<tr>
<td>FATO</td>
<td>forearm, total absence</td>
</tr>
<tr>
<td>FAUP</td>
<td>forearm, upper third</td>
</tr>
<tr>
<td>FAMI</td>
<td>forearm, middle third</td>
</tr>
<tr>
<td>FAPO</td>
<td>forearm, lower third</td>
</tr>
<tr>
<td>CATO</td>
<td>carpal, total absence</td>
</tr>
<tr>
<td>CAPA</td>
<td>carpal, partial absence</td>
</tr>
<tr>
<td>MCTO</td>
<td>metacarpal, total absence</td>
</tr>
<tr>
<td>MCPA</td>
<td>metacarpal, partial absence</td>
</tr>
<tr>
<td>PHTO</td>
<td>phalangeal (finger or thumb), total absence</td>
</tr>
<tr>
<td>PHPA</td>
<td>phalangeal, partial absence</td>
</tr>
</tbody>
</table>

References

Scoliosis and Trunk Asymmetry in Upper Limb Transverse Dysmelia

*Lars Samuelsson, M.D., Ph.D., †Lise-Lotte Hermansson, O.T., and §Lars Norén, M.D.

Study conducted at Örebro Medical Center Hospital, Örebro, Sweden

Summary: The incidence of scoliosis and trunk asymmetry were studied in 60 patients with upper limb dysmelia of the transverse type. The evaluations were based on radiographic measurements of the spine and scoliometer readings of the angle of trunk rotation (ATR), which were correlated with the side and level of the limb deficiency and also with leg length-inequality (LLI). Nineteen patients (31%) had a scoliosis between 10° and 19°, whereas the curves were between 5° and 10° in another 30 patients and the remainder had no measurable curve. The scoliosis seemed at least partly to be of postural origin as LLI significantly correlated with the direction of the curves, but there was no correlation between the Cobb angles and the magnitude of LLI. The scoliometer readings did not correlate with the scoliosis or LLI. Our findings indicate that most patients with upper limb transverse amputations do not develop a significant scoliosis. A scoliometer screen has a low positive predictive rate for scoliosis and the diagnosis of scoliosis requires a radiograph of the spine. Key Words: Ectromelia—Leg-length inequality—Limb deficiency—Scoliosis—Trunk asymmetry.

Dysmelia is reported to be associated with scoliosis in 16–48% of cases (2,8,9,11). However, most patients studied have congenital limb deficiencies of the longitudinal type only or combined with the transverse type. Our aim was to study the type and degree of scoliosis in dysmelia patients with transverse amputations of the upper limb specifically. We also wanted to assess the value of a scoliometer screening procedure in identifying scoliosis associated with dysmelia.

PATIENTS AND METHODS

Sixty patients with upper limb deficiencies of the transverse type according to the classification accepted by WHO in 1988 (6,7,12) were included in the study. With few exceptions, the patients had been fitted with a myoelectric arm prosthesis at the arm prosthetic unit at Örebro Medical Center Hospital and were seen between November 1993 and November 1995. There were 30 male and 30 female patients with a mean age of 13 years (range, 7–48). The amputation level was humeral in three patients, forearm in 41, and distal to the wrist in the remaining 16 patients. Thirty-nine patients had a left-sided limb deficiency, 20 had a right-sided one, and one patient had bilateral forearm amputations.

A posteroanterior standing radiograph was obtained in all patients and the scoliosis was measured by the Cobb method. Any congenital deformation of the spine or ribs including numerical variations were recorded. Leg-length inequality (LLI) was estimated according to Friberg (3) by measuring the difference between the heights of the highest articular points of the femoral heads.

The angle of trunk rotation (ATR) was measured by one of the authors (L.S.) at the upper thoracic, mid-thoracic, lower thoracic, and lumbar regions using a scoliometer (Pedihealth ky, Oulu, Finland) with the patients in a standing forward-flexed position (1,4). No compensation for clinically estimated LLI was made. The level and degree of maximal truncal asymmetry and the side of the corresponding rib bump or lumbar prominence were recorded. This examination was performed in 46 patients both with and without the myoelectric prosthesis applied and in three patients without the prosthesis, whereas 11 were not measured at all. Clinical data on all patients with a scoliosis of >10° are shown in Table 1.

Statistical analysis

Continuous variables were tested using the Student t test, e.g., scoliometer readings. Fisher’s exact test was used to study the relationship between curve direction, side and level of amputation, direction of scoliometer readings, and LLI. Multiple regression was used to study the correlation between Cobb angles, scoliometer readings, LLI, age, and amputation level. A 5% level of significance was accepted.

The study was approved by the ethics committee of Örebro County Council.
TABLE 1. Clinical data of 19 patients with scoliosis and transverse upper limb dysmelia

<table>
<thead>
<tr>
<th>Patient/sex age (yr)</th>
<th>Deficiency side and level</th>
<th>Scoliosis direction/level/Cobb angle</th>
<th>Scolimeter angle prosthesis (high side)</th>
<th>Leg length difference (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/5/48 Left transverse</td>
<td>Left thoracolumbar curve 11°</td>
<td>n.a.</td>
<td>n.a.</td>
<td>n.a.</td>
</tr>
<tr>
<td>2/9/23 Right transverse</td>
<td>Right thoracic curve 15°</td>
<td>4° (left)</td>
<td>5° (left)</td>
<td>n.a.</td>
</tr>
<tr>
<td>3/19/2 Left transverse</td>
<td>Left thoracic curve 11°</td>
<td>8° (left)</td>
<td>9° (left)</td>
<td>n.a.</td>
</tr>
<tr>
<td>4/9/21 Right transverse</td>
<td>Right thoracolumbar curve 19°</td>
<td>7° (left)</td>
<td>3° (left)</td>
<td>n.a.</td>
</tr>
<tr>
<td>5/8/19 Left transverse</td>
<td>Left lumbar curve 11°</td>
<td>n.a.</td>
<td>n.a.</td>
<td>n.a.</td>
</tr>
<tr>
<td>6/8/19 Right transverse</td>
<td>Right lumbar curve 17°</td>
<td>n.a.</td>
<td>n.a.</td>
<td>n.a.</td>
</tr>
<tr>
<td>7/6/17 Left transverse</td>
<td>Left thoracolumbar curve 17°</td>
<td>n.a.</td>
<td>n.a.</td>
<td>n.a.</td>
</tr>
<tr>
<td>8/8/14 Right transverse</td>
<td>Right thoracolumbar curve 11°</td>
<td>4° (right)</td>
<td>7° (right)</td>
<td>left leg 10 mm longer</td>
</tr>
<tr>
<td>9/19/14 Left transverse</td>
<td>Left lumbar curve 10°</td>
<td>3° (right)</td>
<td>n.a.</td>
<td>right leg 2 mm longer</td>
</tr>
<tr>
<td>10/5/13 Left transverse</td>
<td>Left thoracic curve 16°</td>
<td>n.a.</td>
<td>n.a.</td>
<td>no difference</td>
</tr>
<tr>
<td>11/8/13 Left transverse</td>
<td>Left thoracic curve 16°</td>
<td>5° (right)</td>
<td>4° (right)</td>
<td>right leg 4 mm longer</td>
</tr>
<tr>
<td>12/5/13 Left transverse</td>
<td>Left thoracolumbar curve 13°</td>
<td>7° (right)</td>
<td>7° (right)</td>
<td>n.a.</td>
</tr>
<tr>
<td>13/7/10 Left transverse</td>
<td>Left thoracic curve 10°</td>
<td>7° (right)</td>
<td>7° (right)</td>
<td>right leg 5 mm longer</td>
</tr>
<tr>
<td>14/9/10 Left transverse</td>
<td>Left thoracolumbar curve 17°</td>
<td>5° (right)</td>
<td>n.a.</td>
<td>right leg 3 mm longer</td>
</tr>
<tr>
<td>15/5/10 Left transverse</td>
<td>Right thoracolumbar curve 16°</td>
<td>0° (level)</td>
<td>3° (right)</td>
<td>left leg 5 mm longer</td>
</tr>
<tr>
<td>16/8/9 Right transverse</td>
<td>Right thoracic curve 11°</td>
<td>3° (right)</td>
<td>4° (right)</td>
<td>no difference</td>
</tr>
<tr>
<td>17/5/8 Right transverse</td>
<td>Right thoracolumbar curve 10°</td>
<td>4° (left)</td>
<td>7° (left)</td>
<td>n.a.</td>
</tr>
<tr>
<td>18/5/8 Right transverse</td>
<td>Right thoracic curve 10°</td>
<td>3° (left)</td>
<td>3° (left)</td>
<td>no difference</td>
</tr>
<tr>
<td>19/5/8 Right transverse</td>
<td>Right thoracic curve 18°</td>
<td>9° (right)</td>
<td>9° (right)</td>
<td>no difference</td>
</tr>
</tbody>
</table>

n.a., not assessed

RESULTS

There were no congenital vertebral anomalies or structural abnormalities of the ribs, but there were two patients with 11 ribs and two patients with 13 ribs. Nineteen patients (31%) had a scoliosis of >10° with a mean of 11° (range, 10°—13°) and 30 patients had minor curves between 5 and 10°. In the 19 patients with scoliosis by definition, the thoracic curves were right convex in four cases and left convex in four. The thoracolumbar curves were right convex in four and left convex in four cases, and there were one right and two left convex lumbar curves. Age was positively correlated with the magnitude of scoliosis, but the increase with age was of no clinical importance.

There was no correlation between scoliosis magnitude, level or direction, and dysmemic side or level. The convexity of the spine was opposite the dysmemic side in 33 cases (56%), whereas 26 were on the same side, minor curves included. In patients with scoliosis of >10°, 11 (58%) had the convexity opposite the dysmemic side and eight on the same side.

The ATR was measured in 46 patients both with and without prosthesis. Three patients did not have their prosthesis available at examination and 11 patients were not examined with the scolimeter. The median ATR measured without the prosthesis applied was 5° (range, 0°—10°), and with the prosthesis applied, 4° (range, 0°—9°). In the forward-flexed position the elevated side of the trunk was opposite the dysmemic side in half of our patients. In the 19 patients with scoliosis >10°, the elevated side was opposite the dysmemic side in 11 cases (59%). There was one scolimeter reading of 0° indicating a straight spine in a 10-year-old boy, his radiograph showing a right thoracic curve of 10°. There was no correlation between the scolimeter readings with and without prosthesis, the ATR being less without the prosthesis in 12 patients, unchanged in 16 patients, and higher in 15 patients. However, the ATR measured without the arm prosthesis applied was higher in patients with...
amputations distal to the wrist compared to more proximal amputation levels.

In 40 patients the spinal radiograph also permitted measurement of LLI with a median value of 0.4 cm (range, 0-1.7). The left leg was the longer in 20 patients, the right leg in 8 patients, and in 12 patients the legs were of equal length. The LLI did not correlate with the dysmorphic side nor was the magnitude of LLI correlated with the degree of scoliosis or the ATR readings. There was, however, a significant correlation between the leg length and the direction of the scoliotic curve according to Fisher's exact test (p = 0.02). The convexity of the curve was toward the side of the short leg in 21 of 28 patients and toward the side of the long leg in the remaining seven patients. In the forward-bent position without the prosthesis applied, the rib hump or lumbar prominence was on the same side as the longer leg in 15 patients and on the opposite side in the remaining 10 cases. The side and level of the arm amputation, the side of the lumbar prominence or rib hump, and the magnitude of the ATR readings did not correlate with the LLI.

DISCUSSION

A simple and effective classification of types and levels of congenital limb deficiencies is necessary for both treatment and research purposes. We used the terminology of the International Organization for Standardization accepted by WHO, based on anatomic structures of the bony parts of the limbs suggested by Kay (7) and Swanson (12). From our experience, patients with longitudinal amputations are sometimes wrongly classified as transverse hemimelia, which complicates the analysis of specific questions and makes previous reports less useful for comparison.

Our findings confirm earlier reports (2,8,9,11) that upper limb dysmelia is associated with scoliosis, but the magnitude of the curves was small. None of our 25 patients older than 12 years was expected to require brace or surgical treatment for scoliosis. We found the incidence of scoliosis almost twice as high as the one reported by Powers et al. (11). In contrast to their two patients with transverse hemimelia showing significant curves of 50 and 65°, none of our patients had a scoliosis >19°. This might be due both to differences in classification and the amount of patients studied.

The convexity of the curve was evenly distributed between the right and left side and was not associated with trunk asymmetry according to scoliometer readings and the very slight correlation between degree of scoliosis and age differs from adolescent idiopathic scoliosis (AIS). Like Hult (5) we found that the right leg was shorter approximately twice as often as the left one. The finding that 21 of 28 patients had a scoliosis on the contralateral side of the long leg shows that the curve at least partly was of a static or postural origin even if the absolute values of LLI in centimeters were small. The fact that the magnitude of the scoliotic curve was not correlated to the magnitude of LLI indicates that unknown factors contribute to the development and magnitude of scoliosis in transverse dysmelia.

Owing to the postural imbalance caused by less weight of the dysmorphic arm, discrepancies in shoulder and scapular height are easily observed, but the relationship between trunk asymmetry and scoliosis is less well known in this population. Noninvasive methods of screening for spinal deformity rely on surface topography and there is a physiologic variation of the shape of the trunk (10,14). In AIS, the correlation between the Cobb angle, on one hand, and the scoliometer reading and Moiré pattern, on the other, is well established. Noninvasive screening using these techniques is effective in identifying patients who might need treatment for idiopathic scoliosis (1,15). Opinions differ regarding the positive predictive rate of the ATR at different angles, but >5-7° justifies referral for a spine radiograph (1,4). Our finding that there is a lack of correlation between Cobb angles and ATR readings also indicates that the nature of the scoliosis in dysmelia is different from AIS. The trunk symmetry of a patient with a congenital transverse upper limb amputation is probably affected by the lesser weight of the dysmorphic arm and hypertrophic muscles partly caused by inactivity, but the effect of a scoliosis is unknown. These factors probably partly explain the low positive predictive rate of a scoliometer screening in identifying a scoliosis. A spinal radiograph seems to be necessary to assess the degree of scoliosis in dysmotic patients because the drawbacks that exist with the scoliometer procedure probably also exist with other surface topographic methods such as Moiré screening and other similar but computerized techniques.

The observed trunk asymmetry associated with limb deficiency is reported to be associated with pain in the neck and shoulder region (13), but this question is beyond the scope of the current report. Patients with limb deficiencies distal to the wrist had higher scoliometer readings without the prosthesis applied than more proximal amputations. One possible explanation for this observation might be that the weight of the dysmorphic arm and prosthesis is relatively higher compared to forearm-deficient patients and they become more decompensated when bending forward on scoliometer screening without their prosthesis.

In conclusion, our findings suggest that the frequent but mild scoliosis associated with transverse dysmelia of the upper limb is of no clinical importance in most cases. The scoliosis seem to be at least partly of postural origin as LLI significantly correlated with the direction of the curves. A scoliometer screen fails to identify the significant curves.

Acknowledgment: The study was supported by grants from Östrobo County Council. We also thank Svante Hugosson and Anders Magnusson for assistance with the statistical analysis and Anna-Carin Ojefors for assistance in the clinical follow-up examinations.

REFERENCES

ASSESSMENT OF CAPACITY FOR MYOELECTRIC CONTROL: A NEW RASCH-BUILT MEASURE OF PROSTHETIC HAND CONTROL

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Objective: To report the results from a Rasch rating scale analysis of the Assessment of Capacity for Myoelectric Control (ACMC) implemented to evaluate internal scale validity, person response validity, separation reliability, targeting and responsiveness of the measures over time.

Design: Longitudinal data (18 months) from a prospective study of development of capacity for myoelectric control in children and adults were used for the analysis.

Patients: A consecutive sample of 75 subjects (43 males, 32 females) with upper limb reduction deficiency or amputation and myoelectric prosthetic hands referred for occupational therapy from September 2000 to March 2002. Participants’ ages ranged from 2 to 87 years.

Methods: Outcome measure was the ACMC. Occupational therapists completed 210 assessments at an arm prosthesis centre in Sweden. A two-faceted rating scale analysis of the data was performed.

Results: All 30 ACMC items and 96.2% of participants demonstrated goodness-of-fit to the rating scale model for the ACMC. Separation and SE values suggested adequate reliability of the item and person estimates.

Conclusion: The items demonstrated internal scale validity and the participants demonstrated person response validity. The ACMC was well targeted and sensitive enough to detect expected change in ability.

Key words: psychometrics, measurement, arm prosthesis, occupational therapy.

J Rehabil Med 2004; 36: 1–6

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Submitted January 15, 2004; accepted September 1, 2004

INTRODUCTION

Persons with upper limb amputations are often fitted with prostheses in order to enhance function and appearance, and to facilitate performance of activities of daily living (ADLs) (1). Besides the conventional, body-powered prosthesis, fitting of a myoelectrically controlled prosthesis is a well-established practice for persons with both acquired upper limb amputations (2) and limb deficiencies present at birth (3, 4). As part of the rehabilitation of these patients, occupational therapists provide training in natural contexts so that competence in prosthetic use in everyday life can be ensured. The overall aim is for these persons to use the artificial limbs in such a way that they realize unrestricted possibilities for age-relevant performance of ADLs and participation in society.

When fitted with a myoelectric prosthesis, an additional issue arises related to the person’s capacity for myoelectric control. That is, if the person does not display good myoelectric control, he or she will probably have more difficulty in performing essential ADLs (5). Unlike conventional prostheses, myoelectrically controlled prostheses have motorized operation of the opening and closing of the terminal device. Electrodes, located inside the prosthetic socket over muscle bellies, allow detection of electrical activity in the muscles. The contracting muscle activates the motor of the terminal device, and adjustments in the force or speed of the contraction control the range of opening and closing in the hand or hook.

When the occupational therapist provides training in natural contexts, several additional qualitative aspects of myoelectric control must be considered. For example, especially for persons fitted for the first time, the myoelectric prosthetic arm is often heavy. This can result in difficulties in contracting the correct muscle and activating the motor in the desired direction when the arm is not supported, for example when it is resting on a table. Moreover, there is no positional feedback from the myoelectric prosthetic hand to inform the user whether the prosthetic hand is open or closed. Hence, most people need to look at their prosthetic hands to determine their positions. To be able spontaneously to use the myoelectric prosthetic hand in ADLs, new prosthetic users must, therefore, learn to operate the hand without visual feedback. Although improved myoelectric control is thought to enhance the person’s ability to perform essential tasks, there is no standardized method for measuring qualitative aspects of myoelectric control in daily life in an individual subject.

In order to meet the need for a valid, reliable and sensitive evaluation, the Assessment of Capacity for Myoelectric Control (ACMC) was developed. The ACMC is administered and scored based on clinical observations of the myoelectric prosthesis user as he or she is gripping, holding and releasing daily life objects when, for example, preparing a chosen meal, or, in the case of a child, playing. The items comprising the ACMC (see Appendix) describe different levels of difficulty of control of the...
myoelectric hand. Each item is scored on a 4-point ordinal scale. To convert these ordinal ratings into linear measures of myoelectric control (6), the ACMC has been developed using Rasch analysis methods (7, 8). The family of Rasch measurement models (7) has been used increasingly in rehabilitation to develop linear measures of ability (9–12). The 30 items included in the current version of the ACMC were previously piloted and then revised based on an earlier Rasch analysis of the tool. Items in the earlier version found to be redundant or misfitting to the Rasch rating scale model of the ACMC were eliminated. The aim of the present study was to evaluate the revised tool for aspects of validity and reliability.

The specific research questions addressed were as follows: (i) Do the ACMC demonstrate internal scale validity as evidenced by goodness-of-fit to a Rasch rating scale model?; (ii) Do the people using myoelectric prostheses show person response validity as indicated by goodness-of-fit to the Rasch rating scale model?; (iii) Are the item difficulty and person ability measures associated with separation indices greater than 2.0?; (iv) Are the items on the ACMC targeted in terms of difficulty to the abilities of the persons tested?; and, (v) Are the person ability measures sensitive measures of expected change?

METHODS

Participants

In this study, data collected prospectively from September 2000 to March 2002 were analysed. Seventy-five persons participated (43 males, 32 females, aged 2–57 years, mean age 8 years, interquartile range 5–14 years), the final analysis was based on 210 assessments. More specifically, the participants were all persons who were tested 1–9 times (median = 2) over an 18-month period as they were developing their overall capacity for myoelectric control both through normal growth and development, and ongoing treatment. All participants had a congenital upper limb deficiency (n = 64) or acquired amputation (n = 11) and had been fitted with a myoelectrically controlled prosthesis. They had been referred for fitting of artificial limbs by primary and secondary health care centres throughout Sweden.

The participants entered the study consecutively during the study period. For some of the participants (n = 11), this was the first myoelectric fitting. Other participants had had myoelectric prostheses for a period of between 3 months and 18 years prior to entering the study. Clinically, all participants had transverse upper limb deficiency, 30 participants had right-sided limb deficiencies (transhumeral = 2; transradial = 21; hand = 7), and 45 had left-sided limb deficiencies (transhumeral = 3; transradial = 31; hand = 10). Depending on their ages, the participants had been fitted with myoelectric prosthetic hands of different sizes, and depending on the level of their deficiency, they had been fitted with additional body-powered or friction-regulated prosthetic joints (i.e. wrist, elbow or shoulder joints). All participants exhibited developmentally typical physical and mental behaviour. The data were generated by assessments made by 3 occupational therapists and 2 occupational therapy students.

Instrumentation

A basic premise of the ACMC is that the evaluation is made during the performance of everyday tasks. In contrast to many other instruments, the ACMC does not require the use of specific tasks or tools. According to the ACMC manual (13), any task, easy or difficult, can be used to evaluate the capacity for control, as long as the task requires active use of both hands (i.e. the unaffected hand and the prosthetic hand). A person with a prosthetic hand would not naturally use the prosthesis as a dominant hand. Only the performance of simple actions requiring the use of the prosthesis is considered in the evaluations. Hence, any tasks chosen as purposeful and meaningful to the individual may be used for the assessment. For example, a person might be observed preparing a simple meal, making a bed, doing crafts or playing with different toys. The persons are encouraged to accomplish the tasks spontaneously in their usual way (i.e. by using the prosthetic hand as they are used to, as an active assisting hand or as a passive support or stabilizer of objects). If they need help to be able to perform the task successfully, the occupational therapist adjusts the task (e.g. by helping a young child to read recipes and measure the ingredients for cooking). It is the person’s capacity to control the myoelectric prosthesis that is evaluated, not the person’s independence or quality of task performance.

As the persons perform chosen tasks and actively or passively use their prosthetic hand, the occupational therapist assesses their capacity for control of their myoelectric prosthesis by rating their performances on items representing different aspects of quality of myoelectric control. The 30 items in the ACMC are classified into 4 groups: (i) gripping (12 items), (ii) holding (6 items), (iii) releasing (10 items) and (iv) coordinating between hands (2 items) (Appendix). Each person’s performance is rated with scores ranging from 0 to 3, where 0 = not capable, 1 = sometimes capable, capacity not established, 2 = capable on request and 3 = spontaneously capable (13).

Procedure

Before its initiation, this study was approved by the local county council ethics committee review board. Informed consent was also obtained from all participants and, when appropriate, their parents. The 5 raters were initially trained in the administration of the ACMC by the first author and received an administration manual (13). When data collection was initiated, the participants were evaluated during regular visits to the occupational therapy clinic for planning, re-evaluation and/or training. The rater was to score each item of the ACMC after having observed the participant performing the task. If a particular item was not part of the task performed, the item was to be recorded as missing. That is, it occasionally is possible that a person can be observed performing a task where there is no opportunity to observe the person’s relative level of myoelectric control. While this situation is most likely to arise with more difficult items on the ACMC, we felt that it was most appropriate to make no assumptions about level of control if the action was required by the task and therefore not observed. The advantage of using the family of Rasch models is that they allow for missing data (14).

Statistics

The data were entered into the computer by the first author and analysed using the FACETS Rasch measurement computer program (version 3.1) according to a 2-facet rating scale model with 4 response categories. Rasch analysis methods are discussed in detail elsewhere (7, 8, 11, 14). The goal was to construct a unidimensional scale that yields valid measures of person capacity to control myoelectric prostheses.

The ACMC raw scores, the FACETS program estimates an ability measure for each person and a difficulty calibration for each item, locating both persons and items on the same common linear metric. Measures and calibrations are expressed in logits (log-odds probability units). The estimated measures and calibrations are accompanied by goodness-of-fit statistics that can be used to determine whether persons and items meet the expectations of the model. For items, fit statistics are used to determine whether each item fits a single underlying unidimensional construct; when at least 95% of the items demonstrate acceptable goodness-of-fit, the scale can be said to possess internal scale validity (7, 14). For persons, fit statistics are used to verify that the persons have responded to the items in an expected manner (14). That is, persons who have higher myoelectric control are expected to pass more difficult items than persons with less myoelectric control, and all persons are expected to be more likely to pass easy items than difficult items. When at least 95% of the persons demonstrate acceptable goodness-of-fit, the people can be said to have valid patterns of response or person response validity.

The acceptable range for item and person mean-square (Mnsq) fit statistics in observational rating scales is 0.6 to 1.4 (15). The standardized fit statistic (z) is used to evaluate the significance of the Mnsq values. The commonly accepted interpretation of z is that values greater than –2 or less than 2 indicate compatibility with the relevant Rasch model (8). The measures and calibrations are also accompanied by...
standard errors (SEs), which indicate the precision of each calibration and measure. Separation, an index of the spread of the person ability measures and item calibrations relative to their precision (16), is used to estimate the number of strata that are distinguished by the test (14). Both SE and separation are indices of reliability.

In order to ensure that repeated assessments of the same persons did not result in a violation of local independence, we initially obtained item difficulty estimates based on the first assessment ($n = 75$) for all persons. We then obtained item difficulty estimates for all 210 assessments. Calculation of the standardized difference ($Z$) revealed stable item calibration values ($Z < 2.0, p < 0.05$) (7, 14). These results were further confirmed by Pearson product moment correlations between the 2 sets of item calibrations, $r = 0.99$. Hence, all 210 assessments were analysed together.

**Evaluation of change**

To evaluate whether or not the ACMC myoelectric control ability measures are sensitive measures of expected change, the participants were divided into 2 clinical groups: (i) participants who had been using a myoelectric prosthesis prior to this study; the ability measures of these persons were expected to remain relatively constant over sessions (time); (ii) participants who were fitted for the first time with a myoelectric prosthesis; these participants were expected to display improvements in capacity for myoelectric control over time, with an increase in their ability measures by session. All participants from each group who had been assessed at least 6 times were selected. At least 6 assessments per subject were considered needed in order to see a clear pattern between and within individuals.

**RESULTS**

**Internal scale validity**

All 30 ACMC items demonstrated acceptable goodness-of-fit ($MnSq \geq 0.6$ and $\leq 1.4$ and $z > -2$ and $< 2$), indicating that the items met the model expectation of unidimensionality (8, 15).

**Person response validity**

Only 8 individual assessments (3.8%) failed to demonstrate acceptable goodness-of-fit ($MnSq < 0.6$ or $> 1.4$ and $z < -2$ or $\geq 2$); 5% would be expected to misfit due to chance (8, 17). Thus 96.2% of the participant assessments showed valid response patterns across items as expected by the Rasch rating scale model of the ACMC. Further examination of the data revealed no consistent pattern of misfit associated with age, gender, aetiology, level or side of deficiency, or level of ability to control the prostheses among the 3.8% who failed to exhibit acceptable goodness-of-fit. We concluded therefore, that the failure of those 3.8% of the participants to fit the model was due to random error.

**Separation**

The separation index for items was 11.59, indicating that the participants separated the items into 16 statistically distinct item strata. The associated mean SE for items was 0.17 logit (range 0.12–0.28 logit). The participant separation index was 3.79, indicating that the items separated the 210 assessments into 5 statistically distinct strata based on the persons’ myoelectric control capacity. The associated mean SE for the person ability measures was 0.59 logit (range 0.30–1.87 logit).

**Targeting**

The map of the person ability measures in relation to the item difficulty calibrations (Fig. 1) shows that the items were well targeted to the abilities of the persons. Only 12 persons had ability measures of $\geq 4.0$ logits. They also had overall higher SEs associated with the estimates of their ability measures. Seven persons had ability measures $< 4.0$ logits. They also had overall higher SEs associated with the estimates of their ability measures.
DISCUSSION

In this study, aspects of validity and reliability of a newly developed instrument, the ACMC, were evaluated. The results revealed that the ACMC items met the Rasch rating scale model expectations for unidimensionality indicating that the ACMC demonstrated internal scale validity (7, 14). Similarly, persons with myoelectric prostheses met the model expectations for person response validity. Separation of items and persons into distinct strata, as well as reasonable SEs, support the reliability of the estimated item difficulty and person ability measures. The finding that the mean SEs of persons not at the upper and lower ends of the myoelectric control continuum were less than 0.54 logits further supported the reliability of the person ability estimates.

The limits of the range measurable with the ACMC is indicated by the range of item difficulty (Fig. 1). While in this study the ACMC does not demonstrate a ceiling effect (where persons with higher ability obtain maximum possible scores), persons with higher ability in another sample may achieve the maximum score. It is not possible to study improvements in these persons, or to differentiate between them, without increasing the range of the instrument. For further improvement of the ACMC, adding more difficult items will be considered. A floor effect (where persons with low ability obtain no scores) is not likely to occur in the ACMC. This is demonstrated by the results from this study, where persons with no previous experience from myoelectric control were tested and none obtained minimum possible score.

All participants who were attending the occupational therapy clinic during the study period (18 months) were consecutively enrolled in the study. Most of the subjects (64%) attended the clinic more than once, and were thus repeatedly assessed. In accordance with other studies (18), this repeated testing of the sample was used effectively to increase the sample size so as to ensure more stable estimates of item difficulties (19). Several factors converge to suggest that the use of repeated measurement did not violate the assumption of local independence. Firstly, the item calibration values remained stable between those based on the first evaluation of each participant and those based on all 210 evaluations. Secondly, most participants were tested several times, decreasing the probability that repeated testing over time in a few individuals would bias the results. Furthermore, the repeated testing of participants as they developed increasing control ability. Earlier reports have demonstrated that when some items becoming easier than others. However, in this study, it was the occupational therapist who rated the items. The participants knew only that they were under observation and all ACMC items could be assessed (21). In this study, however, some raters have confused difficult items with missing items. Failure of some raters to leave some items becoming easier than others. However, in this study, it was the occupational therapist who rated the items. The participants knew only that they were under observation and being evaluated. This may have influenced their overall use of their prostheses, but it seems unlikely that such effects would increase with repeated testing. Future research would be needed to know for sure.

As reported earlier, items potentially observable but impossible for the participant to perform should be scored 0 rather than missing. All ACMC items except 3 can be observed in any task. More specifically, as long as the person performs a task where he or she moves about in the room, all ACMC items could be assessed (21). In this study, however, some raters have confused difficult items with missing items. Failure of some raters to leave hard items blank rather than score them as 0 could account for higher SE among those participants. Since it is not possible to differentiate retrospectively between those items that were too difficult and erroneously left blank, and those left blank because they were not observable, revision of the ACMC manual to clarify the importance of scoring all observable items and then differentiate retrospectively between those items that were too difficult and erroneously left blank, and those left blank because they were not observable, revision of the ACMC manual to clarify the importance of scoring all observable items and then implementing further research is warranted. Further research will also be important to further examine aspects of validity and
reliability of the ability estimates. For example, the items may not be equally difficult to perform in different situations or tasks. The independence of the item difficulty relative to the type of task (e.g., feeding, cooking, doing crafts) will need to be evaluated with a larger sample. Likewise, rater severity and reliability will need to be addressed in future studies. Finally, to further validate the ACMC measures, responsiveness to treatment and prosthesis adjustment, predictive power and sensitivity to change, are also needed.

The results of this study have important implications for occupational therapists working in the field of upper limb prosthetics and for the children and adults with myoelectric hand prostheses receiving therapy. One of the main issues in clinical practice is to ensure that occupational therapy interventions are cost-effective. In the past, there was a lack of occupational therapy methodology for determining a person’s current level of myoelectric control, which is vital for planning, re-evaluating, or measuring the outcomes of prosthetic control training. The ACMC has the potential to help therapists and prosthetic users to monitor individual patterns of prosthetic skill development, and evaluate intervention efficacy. The context in which these assessments are made (in the performance of purposeful and meaningful occupations) allows evaluation of spontaneous myoelectric control reflecting the most important aspects of prosthetic use. Our preliminary results indicate that by using the ACMC, it may be possible systematically to study the development of myoelectric control and search for factors that lead to an increase or decrease in myoelectric control.

The early findings by Hubbard et al. (5) indicate that the capacity to control a myoelectric prosthesis, as measured by ACMC, is fundamental to the future use of the functions of the prosthetic hand. More research is needed systematically to examine the relationship between the capacity for myoelectric control and the person’s actual use of the prosthesis in everyday life, and if good myoelectric control leads to greater habitual use for performing essential daily life tasks (22).

In conclusion, the ACMC seems to yield valid interval and reliable level measures that can be used to evaluate the capacity to control a myoelectric prosthesis in both children and adults. It represents an innovative evaluation method, and may improve evaluation of the effectiveness of rehabilitation of individuals with externally powered prostheses. The results of this study also show that evaluations based on individuals’ performances of natural tasks are possible and should, therefore, be encouraged.

ACKNOWLEDGEMENTS

We acknowledge the help of Kristina Waldénlov, registered occupational therapist at the Limb Deficiency and Arm Prosthesis Centre, Örebro University Hospital, who participated in the early process of developing items for the scale and in the data collection for this study. We are also grateful to Lisa Sjöberg and Malin Janson, registered occupational therapists at the same centre in Örebro, for contributing to the data collection. We thank Lennart Bodin, statistician, Unit of Medical Statistics and Epidemiology, Örebro University Hospital, Örebro, and Department of Statistics, Örebro University, Örebro, for advice and comments on the manuscript. Financial support was received from the Norrbacka-Eugenius Foundation, the Solstücken Foundation, the RBU Foundation, the Folke Bernadotte Foundation and Örebro County Council.

REFERENCES

### APPENDIX. ASSESSMENT OF CAPACITY FOR MYOELECTRIC CONTROL ITEMS

<table>
<thead>
<tr>
<th>Gripping</th>
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<tbody>
<tr>
<td>4</td>
<td>Grips with weight supported</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Positions the hand and grips</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Uses the tripod pinch grip with weight supported</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>Positions the hand and uses the tripod pinch grip</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>Adjusts grip force to avoid crushing</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>Grips with the arm in different positions</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>Grips through iterative refinement, manipulates</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>Grips object by feeding arm forwards</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>Grips object moving towards the hand</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>Grips with no visual feedback</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>Adjusts grip force with no visual feedback</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>Grips behind the back to manipulate object</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>Holds with arm supported</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>Holds without support</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>Holds without crushing</td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>Holds with the arm moving</td>
<td></td>
</tr>
<tr>
<td>21</td>
<td>Holds with no visual feedback</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>Holds with the arm moving, no visual feedback</td>
<td></td>
</tr>
<tr>
<td>Releasing</td>
<td></td>
<td></td>
</tr>
<tr>
<td>26</td>
<td>Releases with arm supported</td>
<td></td>
</tr>
<tr>
<td>27</td>
<td>Positions the hand and releases</td>
<td></td>
</tr>
<tr>
<td>28</td>
<td>Releases grip without dropping object</td>
<td></td>
</tr>
<tr>
<td>29</td>
<td>Releases the grip with arm in different positions</td>
<td></td>
</tr>
<tr>
<td>30</td>
<td>Releases through iterative refinement, manipulation</td>
<td></td>
</tr>
<tr>
<td>31</td>
<td>Releases object with arm swinging low</td>
<td></td>
</tr>
<tr>
<td>32</td>
<td>Releases object by feeding arm forwards at, or above, shoulder height</td>
<td></td>
</tr>
<tr>
<td>33</td>
<td>Releases synchronized with the other hand</td>
<td></td>
</tr>
<tr>
<td>34</td>
<td>Releases with no visual feedback</td>
<td></td>
</tr>
<tr>
<td>35</td>
<td>Releases behind the back to manipulate object</td>
<td></td>
</tr>
<tr>
<td>Co-ordinating</td>
<td></td>
<td></td>
</tr>
<tr>
<td>36</td>
<td>Co-ordinates grip using both hands</td>
<td></td>
</tr>
<tr>
<td>37</td>
<td>Co-ordinates release using both hands</td>
<td></td>
</tr>
</tbody>
</table>
Intra- and Interrater Reliability of the Assessment of Capacity for Myoelectric Control

Liselotte M. Hermansson¹, ², Lennart Bodin³, and Ann-Christin Eliasson²

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ABSTRACT

Objective: To examine the reliability of the Assessment of Capacity for Myoelectric Control (ACMC) in children and adults with a myoelectric prosthetic hand.

Design: Intrarater and interrater reliability estimated from reported assessments by three different raters.

Patients: A sample of convenience of 26 subjects (11 males, 15 females) with upper limb reduction deficiency or amputation and myoelectric prosthetic hands were video-filmed during a regular clinical visit for ACMC. Participants’ ages ranged from 2 to 40 years.

Methods: After instructions, three occupational therapists with no, 10 weeks’, and 15 years’ clinical experience from myoelectric prosthesis training and follow-up independently rated the 30 ACMC items in each patient. The ratings were repeated after 2-4 weeks. Inter- and intrarater reliability in items was examined by using weighted κ statistics and Rasch-measurement analyses.

Results: The average intrarater agreement in items was excellent (κ 0.81) in the more experienced raters. Fit statistics showed too much variation in the least experienced rater, who also had only good (κ 0.65) agreement in items. The stability of rater calibrations between first and second assessment varied from 0.01 to 0.16 logits in severity. The mean interrater agreement in items was fair, κ 0.60, between the more experienced raters and κ 0.47 between the less experienced raters. Rater calibration severity ranged from –0.37 to 0.28 logits.

Conclusion: Overall, the agreement was higher in the more experienced raters, indicating that reliable measures of the ACMC require clinical experience from myoelectric prosthesis training.
INTRODUCTION

The purpose of this study was to examine the reliability of the Assessment of Capacity for Myoelectric Control (ACMC) [1], a recently developed observation-based assessment that measures a person’s capacity to control a myoelectric prosthetic hand during the performance of ordinary daily tasks.

Reliability refers to the consistency of measurements when the procedure is repeated on a population of individuals or groups [2]. The need for standardized, observational assessments of the performance of a person with a myoelectric prosthetic hand has been pointed out [3-5]. However, 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. The capacity to control a myoelectric prosthetic hand is essential for the future use of the hand in daily life [6], and in the light of this fact the ACMC was developed.

The ACMC is a test based on clinical observations of the clients, which can be made when the client is performing any task involving the use of two hands. The 30 items comprising the ACMC represent different levels of capacity for control of the myoelectric hand when gripping, holding, and releasing daily life objects. An earlier study has demonstrated the hierarchical order of the items, showing how they range from easy to hard [1] making it possible to evaluate clients with varying degrees of ability. By Rasch measurement analysis [7, 8] the data are converted into linear measures.

In the previous study mentioned above, it was shown that the ACMC was sensitive enough to evaluate changes over time in groups of clients with myoelectric prosthetic hands. However, when occupational therapists use the ACMC in their work, it is important to determine whether the ACMC can score consistently, both within and between raters. Thus, for further use of the ACMC in clinical practice, the reliability of the instrument needed to be determined.

The validity and internal reliability of the ACMC were investigated in the above study [1]. There has been no study, however, in which the rater reliability in use of the ACMC has been estimated. The present study was therefore undertaken to evaluate this instrument regarding intra- and inter-rater reliability. The specific research questions addressed were as follows: (a) Do the raters display consistent scoring in repeated assessments?; (b) Is the scoring consistent between raters?; and (c) Are there any indications of a pronounced difference between inexperienced and well-trained raters?

METHODS

Design
To evaluate the reliability of the ACMC, 25 persons were video-filmed once and one person was video-filmed twice within a three-month period. They were filmed during a regular visit to the limb-fitting center for training in or follow-up of the use of a myoelectric prosthesis. Three independent raters made the assessments on the basis of the persons’ performance as seen on the videos. Each subject-video was rated in the same order by the three raters. For the
intrarater evaluation, all assessments were repeated in the same order three to four weeks later by each of the raters; thus there were two sessions of ACMC assessments in this analysis.

The local Ethics Committee approved the study. In addition, oral consent was received from the subjects and, in younger subjects, their parents.

**Subjects**
The subjects comprised a sample of convenience of 11 males and 15 females (mean age, 10 years; range, 2-40 years) with a myoelectrically controlled prosthesis. They were recruited from patients attending the limb-fitting center during the period August to December 2002. An effort was made to recruit patients with varying degrees of capacity for myoelectric control. The subjects had had the prosthesis for a mean period of 6 years (range 0-20 years).

**Raters**
To represent new users of the instrument, one randomly assigned occupational therapy student (rater A) and one occupational therapy student with 10 weeks’ practice at the limb-fitting center (rater B), both in their last year of education, were trained in the ACMC method. To represent experienced users, one of the most experienced occupational therapists (rater C) at the limb-fitting center, with previous training in the ACMC method, was assigned for this study. All raters received the same information and they all had a copy of the ACMC manual [9].

**Instrumentation**
The ACMC is scored on the basis of observations of the myoelectric prosthesis user as he or she is performing everyday occupations. Any task, easy or hard, can be used to evaluate the capacity for control as long as the task requires active use of both hands (i.e., the unaffected hand and the prosthetic hand). During the assessment, the persons are encouraged to accomplish the tasks spontaneously in their usual way (i.e., by using the prosthetic hand as they are used to, as an active assisting hand or as a passive support or stabilizer of objects). The occupational therapist assesses their capacity for control of their myoelectric prosthesis by rating their performances on 30 items representing different aspects of quality of myoelectric control. The 30 items in the ACMC are classified into four groups: (a) gripping (12 items), (b) holding (6 items), (c) releasing (10 items), and (d) co-ordinating between hands (2 items) [1].

Each person's performance is rated with scores ranging from zero to 3, where zero = not capable, 1 = sometimes capable, capacity not established, 2 = capable on request, and 3 = spontaneously capable. Only those items that are observed during the test session are scored. In accordance with Rasch measurement models, items not observed are recorded as missing; the estimation of item and person statistics when using Rasch models allows for missing data [10]. To convert the ordinal ratings into linear measures, Rasch measurement analysis according to a rating scale model with four response categories is performed [10].

**Data analysis**
The data were analysed in two ways. First, analysis of individual ACMC items concerning inter- and intrarater reliability was performed using the weighted *Kappa* statistic with weights
according to the quadratic model [11]. The Kappa estimates were supplemented with 95 % confidence intervals. The guidelines for the interpretation of Kappa proposed by Fleiss [11] were used to interpret the strength of the agreement. Agreement below Kappa 0.40 was considered poor, and it was judged that those items would probably need further definition to enhance the agreement. To summarize the item estimates of Kappa we calculated their average and will refer to this in the following as average Kappa.

Next, each subject’s assessments (27 subjects x 3 raters x 2 sessions = 162 assessments) were analyzed using the many faceted Rasch (MFR) analysis [12], applying the computer software FACETS (version 3.49) according to a three-facet rating scale model [13]. The following three facets were considered in the analysis: (a) the capacity of the persons, (b) the severity of the rater, and (c) the difficulty of the items. The Rasch analyses are reported with estimates of measures for the subject’s capacity and calibrations for the rater’s severity, supplemented with the standard error of the calibrations. Measures and calibrations are expressed in logits (log-odds-probability units). In Rasch analysis, goodness-of-fit statistics are used to indicate the degree to which each rater’s ordering of persons is consistent with the estimated subject ability measures (intrarater reliability) [8]. In this study the criteria for acceptable rater reliability were 0.6 ≤ mean-square (MnSq) residuals ≤ 1.4 and/or -2 < z < 2, the same as were used in the development of the ACMC [1]. Another way of analyzing intrarater reliability is to use the rater severity calibrations and look at the stability of calibrations over time. This has been done in several studies in larger populations [14, 15] but is not readily applicable to the sample in the present study. However, the comparison may add some valuable information and was therefore carried out nevertheless.

The model was applied in two different settings. The first aimed at a global analysis of both intra- and interrater reliability. Each of the two sessions was analyzed separately in order to obtain comparable results. Parameters for each one of the three raters and the two sessions were estimated and evaluated. By means of fit statistics and comparisons of calibrations from each session, intrarater reliability was estimated. The second formulation of the Rasch analysis simplified the model to include only one rater, in order to obtain individual measures for each subject for each rater and time (session). This analysis was repeated for each one of the three raters. The session-wise differences in the resulting subject measures were plotted against their average for the two sessions to obtain a ‘Bland-Altman plot’ [16].

RESULTS

Since the ACMC allows for missing items, some items were scored in almost every subject, whereas other items were often left blank. Overall, the less experienced raters (raters A and B) left many more blanks than the experienced rater (rater C; Fig. 1). In addition, during the first session rater A did not realise that one of the videotapes (subject #12) was not fully rewinded. Hence, she missed some information and scored many blanks in that subject. All three raters scored most items more frequently during the second session than during the first, indicating that by repeating the assessments the raters had improved their ability to observe the items.
Figure 1. Number of assessments performed by three raters for 30 items of the ACMC protocol, session 2 (no items of numbers 1-3, 15, or 23-25).

**Intrarater agreement**

Analysis of the individual ACMC items showed that the intrarater agreement in the more experienced raters (raters B and C; average Kappa 0.81 for both raters) was higher than in the least experienced rater (rater A; average Kappa 0.65) (Table 1). In addition, the rater goodness-of-fit statistics (Table 2) indicated that rater A had too many variations in her ratings (MnSq >1.4). According to the fit statistics for session 1, raters B and C, in contrast, showed consistency in scoring and thus strong intrarater reliability. In session 2, however, rater B tended to limit her use of the rating scale; MnSq <0.6 indicating fewer variations than expected. The result from the Rasch analysis (Table 2), moreover, showed that the severity calibration difference between session 1 and session 2 was smallest for rater B, whereas raters A and C had somewhat larger differences, though of different directions. Although not readily applicable to this data set, the differences in severity calibration between sessions 1 and 2 are well within the limits reported by others [15], indicating stability in rater severity.

Another aspect of intrarater agreement is illustrated in Figure 2. Here, the individual measures for each subject are analyzed in the ‘Bland-Altman plot’. For rater C there is a constant difference, close to zero, between individual measures in the two sessions, whereas this is not the case for the less experienced raters. Rater A, the most inexperienced, scored higher at the second session in the least able persons, and scored lower at the second session for the most able persons. The shift was substantial, as seen from the slope of the regression line. Rater B had a similar performance but in the opposite direction.
Table 1. Weighted *Kappa* with 95% confidence interval (within parenthesis) of intrarater agreement for ACMC ratings. Duplicate assessments by each rater.

<table>
<thead>
<tr>
<th>Item category</th>
<th>Item no.</th>
<th>Rater A</th>
<th>Rater B</th>
<th>Rater C</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gripping</td>
<td>4</td>
<td>0.49 (0.12 – 0.86)</td>
<td>0.89 (0.78 – 1.01)</td>
<td>0.44 (0.00 – 0.89)</td>
</tr>
<tr>
<td></td>
<td>5</td>
<td>0.32 (0.07 – 0.57)</td>
<td>0.94 (0.84 – 1.03)</td>
<td>0.53 (0.17 – 0.88)</td>
</tr>
<tr>
<td></td>
<td>6</td>
<td>0.82 (0.66 – 0.98)</td>
<td>0.06 (-0.40 – 0.52)</td>
<td>0.77 (0.61 – 0.94)</td>
</tr>
<tr>
<td></td>
<td>7</td>
<td>0.80 (0.60 – 1.00)</td>
<td>0.92 (0.82 – 1.01)</td>
<td>0.76 (0.56 – 0.96)</td>
</tr>
<tr>
<td></td>
<td>8</td>
<td>0.62 (0.38 – 0.87)</td>
<td>0.95 (0.89 – 1.02)</td>
<td>0.90 (0.81 – 0.99)</td>
</tr>
<tr>
<td></td>
<td>9</td>
<td>0.71 (0.43 – 1.00)</td>
<td>0.84 (0.75 – 0.93)</td>
<td>0.85 (0.69 – 1.00)</td>
</tr>
<tr>
<td></td>
<td>10</td>
<td>0.95 (0.91 – 1.00)</td>
<td>0.89 (0.80 – 0.97)</td>
<td>0.84 (0.67 – 1.01)</td>
</tr>
<tr>
<td></td>
<td>11</td>
<td>n.d.</td>
<td>n.d.</td>
<td>0.81 (0.52 – 1.10)</td>
</tr>
<tr>
<td></td>
<td>12</td>
<td>n.d.</td>
<td>n.d.</td>
<td>0.74 (0.42 – 1.06)</td>
</tr>
<tr>
<td></td>
<td>13</td>
<td>n.d.</td>
<td>n.d.</td>
<td>0.80 (0.61 – 0.99)</td>
</tr>
<tr>
<td></td>
<td>14</td>
<td>n.d.</td>
<td>n.d.</td>
<td>0.80 (0.61 – 1.00)</td>
</tr>
<tr>
<td></td>
<td>16</td>
<td>0.81 (0.65 – 0.97)</td>
<td>0.64 (0.20 – 1.08)</td>
<td>0.87 (0.66 – 1.08)</td>
</tr>
<tr>
<td>Holding</td>
<td>17</td>
<td>0.41 (0.05 – 0.77)</td>
<td>0.90 (0.79 – 1.01)</td>
<td>0.80 (0.55 – 1.04)</td>
</tr>
<tr>
<td></td>
<td>18</td>
<td>0.34 (-0.09 – 0.77)</td>
<td>0.94 (0.87 – 1.02)</td>
<td>0.69 (0.37 – 1.00)</td>
</tr>
<tr>
<td></td>
<td>19</td>
<td>0.58 (0.10 – 1.06)</td>
<td>0.87 (0.77 – 0.97)</td>
<td>0.91 (0.84 – 0.99)</td>
</tr>
<tr>
<td></td>
<td>20</td>
<td>0.68 (0.36 – 1.00)</td>
<td>0.90 (0.81 – 0.99)</td>
<td>0.80 (0.63 – 0.96)</td>
</tr>
<tr>
<td></td>
<td>21</td>
<td>0.63 (0.21 – 1.05)</td>
<td>0.90 (0.77 – 1.03)</td>
<td>0.87 (0.74 – 1.00)</td>
</tr>
<tr>
<td></td>
<td>22</td>
<td>0.66 (0.31 – 1.01)</td>
<td>0.79 (0.56 – 1.02)</td>
<td>0.87 (0.71 – 1.02)</td>
</tr>
<tr>
<td>Releasing</td>
<td>26</td>
<td>0.32 (-0.04 – 0.67)</td>
<td>0.93 (0.83 – 1.03)</td>
<td>0.77 (0.48 – 1.06)</td>
</tr>
<tr>
<td></td>
<td>27</td>
<td>0.76 (0.48 – 1.03)</td>
<td>0.93 (0.85 – 1.00)</td>
<td>0.84 (0.76 – 0.93)</td>
</tr>
<tr>
<td></td>
<td>28</td>
<td>0.61 (0.32 – 0.91)</td>
<td>0.83 (0.74 – 0.93)</td>
<td>0.93 (0.87 – 0.98)</td>
</tr>
<tr>
<td></td>
<td>29</td>
<td>0.47 (0.09 – 0.85)</td>
<td>0.86 (0.75 – 0.96)</td>
<td>0.95 (0.90 – 1.00)</td>
</tr>
<tr>
<td></td>
<td>30</td>
<td>0.83 (0.63 – 1.03)</td>
<td>0.85 (0.75 – 0.96)</td>
<td>0.93 (0.84 – 1.03)</td>
</tr>
<tr>
<td></td>
<td>31</td>
<td>0.42 (-0.04 – 0.89)</td>
<td>0.80 (0.53 – 1.07)</td>
<td>0.94 (0.87 – 1.01)</td>
</tr>
<tr>
<td></td>
<td>32</td>
<td>0.88 (0.70 – 1.07)</td>
<td>0.50 (-0.25 – 1.25)</td>
<td>0.87 (0.65 – 1.09)</td>
</tr>
<tr>
<td></td>
<td>33</td>
<td>0.85 (0.64 – 1.06)</td>
<td>0.87 (0.76 – 0.98)</td>
<td>0.80 (0.63 – 0.97)</td>
</tr>
<tr>
<td></td>
<td>34</td>
<td>n.d.</td>
<td>0.91 (0.76 – 1.07)</td>
<td>0.86 (0.72 – 1.01)</td>
</tr>
<tr>
<td></td>
<td>35</td>
<td>0.70 (0.40 – 1.00)</td>
<td>0.64 (0.20 – 1.08)</td>
<td>0.83 (0.64 – 1.03)</td>
</tr>
<tr>
<td>Coordinating</td>
<td>36</td>
<td>0.84 (0.67 – 1.02)</td>
<td>0.78 (0.60 – 0.96)</td>
<td>0.79 (0.60 – 0.97)</td>
</tr>
<tr>
<td></td>
<td>37</td>
<td>0.79 (0.61 – 0.98)</td>
<td>0.77 (0.59 – 0.94)</td>
<td>0.79 (0.60 – 0.97)</td>
</tr>
<tr>
<td>Average</td>
<td></td>
<td>0.65</td>
<td>0.81</td>
<td>0.81</td>
</tr>
<tr>
<td><em>Kappa</em></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

n.d. Items could not be analyzed because of the small number of ratings on these items.
Table 2. Rater severity calibration difference for ACMC in two sessions.

<table>
<thead>
<tr>
<th>Rater</th>
<th>Session 1 (S1)</th>
<th>Session 2 (S2)</th>
<th>Calibration difference S1 – S2 (logits)‡</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Rater severity (logits)</td>
<td>SE (logits)</td>
<td>Infit</td>
</tr>
<tr>
<td>B</td>
<td>0.27</td>
<td>0.06</td>
<td>0.79</td>
</tr>
<tr>
<td>A</td>
<td>0.24</td>
<td>0.09</td>
<td>1.62*</td>
</tr>
<tr>
<td>C</td>
<td>-0.51</td>
<td>0.06</td>
<td>0.82</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>B</td>
<td>0.28</td>
<td>0.05</td>
<td>0.45†</td>
</tr>
<tr>
<td>A</td>
<td>0.08</td>
<td>0.08</td>
<td>2.05*</td>
</tr>
<tr>
<td>C</td>
<td>-0.37</td>
<td>0.05</td>
<td>0.75</td>
</tr>
</tbody>
</table>

* Rater who assigned unexpectedly high or low scores; † Rater who tended to limit her use of the range of the rating scale; ‡ Negative difference in rater severity calibration indicates greater rater severity in session 2.

Figure 2. Intrarater reliability illustrated by a 'Bland-Altman plot'. Pairwise differences in subject measures (logits) between two different sessions are plotted against their means. Linear regression lines are given to show the trend. The horizontal trend located at 0 indicates no systematic difference between sessions. Rater A (inexperienced), rater B (some experience), and rater C (experienced).
The intrarater agreement in individual items in rater A ranged from Kappa 0.32 to 0.95, in rater B from Kappa 0.06 to 0.95, and in rater C from Kappa 0.44 to 0.95 (Table 1). In all raters, the lower intrarater item agreement was noted in the easiest gripping items (items 4, 5, and 6). In raters A and B, the highest intrarater item agreement was also in gripping items (numbers 8 and 10), whereas in rater C the highest intrarater item agreement was found in a releasing item (number 29). For items 5, 18 and 26 in rater A, and item number 6 in rater B, intrarater Kappa was ≤0.40. In rater C there was no item with intrarater Kappa ≤0.40. This indicates that in inexperienced raters items 5, 6, 18 and 26 are more likely to be inconsistently rated.

Interrater agreement
Because of the missing information for rater A, session 1, the results from the second session were used for analyses of the interrater agreement.

Overall, in individual items the agreement between rater B and rater C (average Kappa 0.60) was higher than that between rater A and rater B (average Kappa 0.44). The agreement in individual items between raters A and B ranged from Kappa – 0.01 to 0.71, and between raters B and C from Kappa 0.04 to 0.84 (Table 3). Again, the lowest interrater agreement was found for the easier gripping items (items 4 and 6). The highest agreement between both raters A and B and raters B and C was noted for the releasing item number 30. In raters A and B there were 8 items with interrater Kappa ≤0.40 (items 4, 6, 9, 16, 17, 19, 22 and 35). In three of these items (6, 16, and 35) the agreement was also low between raters B and C. For all other items the interrater Kappa value for B and C was >0.40 (Table 3).

The Rasch analysis for interrater agreement was based on only three raters, for which reason we concentrated on the analysis of the individual measures for each subject, and these are illustrated in the ‘Bland-Altman plot’ in Figure 3. The figure shows that rater A differed in a systematic way from both rater B and rater C, since the difference between the raters was dependent on the size of the individual measures. The difference between raters B and C was less systematic and on average close to zero, a preferred result in comparisons of this kind [16].
Table 3. Weighted Kappa with 95% confidence interval (within parenthesis) of interrater agreement for ACMC ratings.

<table>
<thead>
<tr>
<th>Item category</th>
<th>Item no.</th>
<th>Raters A and B</th>
<th>Raters B and C</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gripping</td>
<td>4</td>
<td>0.11 (-0.21 – 0.44)</td>
<td>0.47 (0.15 – 0.78)</td>
</tr>
<tr>
<td></td>
<td>5</td>
<td>0.51 (0.27 – 0.76)</td>
<td>0.53 (0.26 – 0.81)</td>
</tr>
<tr>
<td></td>
<td>6</td>
<td>0.38 (-0.09 – 0.84)</td>
<td>0.04 (-0.41 – 0.50)</td>
</tr>
<tr>
<td></td>
<td>7</td>
<td>0.66 (0.43 – 0.89)</td>
<td>0.81 (0.65 – 0.97)</td>
</tr>
<tr>
<td></td>
<td>8</td>
<td>0.63 (0.38 – 0.88)</td>
<td>0.80 (0.65 – 0.95)</td>
</tr>
<tr>
<td></td>
<td>9</td>
<td>0.30 (0.02 – 0.89)</td>
<td>0.73 (0.58 – 0.88)</td>
</tr>
<tr>
<td></td>
<td>10</td>
<td>0.69 (0.50 – 0.87)</td>
<td>0.77 (0.60 – 0.93)</td>
</tr>
<tr>
<td></td>
<td>11</td>
<td>n.d.</td>
<td>n.d.</td>
</tr>
<tr>
<td></td>
<td>12</td>
<td>n.d.</td>
<td>0.67 (0.36 – 0.98)</td>
</tr>
<tr>
<td></td>
<td>13</td>
<td>0.43 (0.09 – 0.77)</td>
<td>0.52 (0.21 – 0.82)</td>
</tr>
<tr>
<td></td>
<td>14</td>
<td>n.d.</td>
<td>0.43 (-0.04 – 0.89)</td>
</tr>
<tr>
<td></td>
<td>16</td>
<td>0.33 (-0.05 – 0.71)</td>
<td>0.33 (-0.02 – 0.68)</td>
</tr>
<tr>
<td>Holding</td>
<td>17</td>
<td>0.28 (0.02 – 0.54)</td>
<td>0.52 (0.21 – 0.83)</td>
</tr>
<tr>
<td></td>
<td>18</td>
<td>0.54 (0.28 – 0.79)</td>
<td>0.70 (0.50 – 0.90)</td>
</tr>
<tr>
<td></td>
<td>19</td>
<td>0.24 (-0.18 – 0.66)</td>
<td>0.59 (0.37 – 0.81)</td>
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<tr>
<td></td>
<td>20</td>
<td>0.42 (0.07 – 0.76)</td>
<td>0.64 (0.40 – 0.88)</td>
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<tr>
<td></td>
<td>21</td>
<td>0.43 (0.18 – 0.68)</td>
<td>0.75 (0.57 – 0.93)</td>
</tr>
<tr>
<td></td>
<td>22</td>
<td>-0.01 (-0.39 – 0.37)</td>
<td>0.73 (0.55 – 0.91)</td>
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<tr>
<td>Releasing</td>
<td>26</td>
<td>0.53 (0.27 – 0.78)</td>
<td>0.63 (0.33 – 0.93)</td>
</tr>
<tr>
<td></td>
<td>27</td>
<td>0.66 (0.46 – 0.87)</td>
<td>0.67 (0.48 – 0.85)</td>
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<td></td>
<td>28</td>
<td>0.55 (0.28 – 0.82)</td>
<td>0.74 (0.57 – 0.90)</td>
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<tr>
<td></td>
<td>29</td>
<td>0.43 (0.14 – 0.73)</td>
<td>0.77 (0.61 – 0.92)</td>
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<td>30</td>
<td>0.71 (0.54 – 0.88)</td>
<td>0.84 (0.75 – 0.93)</td>
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<tr>
<td></td>
<td>31</td>
<td>0.50 (0.30 – 0.70)</td>
<td>0.51 (0.07 – 0.94)</td>
</tr>
<tr>
<td></td>
<td>32</td>
<td>n.d.</td>
<td>0.78 (0.50 – 1.06)</td>
</tr>
<tr>
<td></td>
<td>33</td>
<td>0.47 (0.17 – 0.77)</td>
<td>0.46 (0.12 – 0.80)</td>
</tr>
<tr>
<td></td>
<td>34</td>
<td>n.d.</td>
<td>0.73 (0.50 – 0.96)</td>
</tr>
<tr>
<td></td>
<td>35</td>
<td>0.33 (-0.05 – 0.71)</td>
<td>0.37 (-0.02 – 0.76)</td>
</tr>
<tr>
<td>Coordinating</td>
<td>36</td>
<td>0.48 (0.20 – 0.77)</td>
<td>0.44 (0.12 – 0.75)</td>
</tr>
<tr>
<td></td>
<td>37</td>
<td>0.49 (0.19 – 0.78)</td>
<td>0.46 (0.15 – 0.78)</td>
</tr>
</tbody>
</table>

Average Kappa 0.44 0.60

n.d. Items could not be analyzed because of the small number of ratings on these items.
DISCUSSION

In this study we found an intrarater reliability that among the more experienced raters was on average almost perfect, and in the rater with no clinical experience was substantial. The interrater agreement, however, was moderate between both groups of raters. These results indicate that however small, the clinical experience in rater B meant that she was in greater agreement with the more experienced rater than with her fellow student. Also, not surprisingly, it was evident that a substantial training period and clinical experience are necessary for consistent use of the ACMC.

In comparison with other Rasch derived instruments such as, for example, the Assessment of Motor and Process Skills [17], the methods used for rater reliability analyses in this study (Kappa statistics and Rasch analysis) derived from different psychometric traditions. We found both methods very useful, since they added different perspectives to the study. Besides the overall rater agreement, Kappa statistics identified certain ACMC items that need further clarification for use by less experienced raters (Table 1). In the same way, besides fit statistics from the Rasch analyses, the Bland-Altman plot, demonstrating the stability in subject measures obtained by rater C (Fig. 2) added useful information.
The video-recordings used for the analyses had some shortcomings. In ACMC assessments in clinical practice, the patients are observed by the occupational therapist during the performance of different tasks. In this study, the assessments were made on the basis of information from the videos only. This meant that since the information available for the rater was limited to what was in the video, some items may have been difficult to identify. This may explain the lack of scoring on some of the items. For example, the video-operator may have focused on the hands, zoomed in to them, and thus missed the information on how the client was using his/her sight to compensate for the lack of sensation. This is clearly demonstrated by the small number of ratings on items representing use of visual feedback for control of the prosthetic hand (items 13, 14, 16, 34 and 35) (Fig. 1). In the videos, during the task performances the easiest gripping items (numbers 4, 5, and 6) were not always shown in the most able persons. In these cases it seems as if the raters used different strategies, as demonstrated by the low intra- and interrater agreement in these items. Scoring of items 11 and 12 also appeared to be very difficult, especially for the less experienced raters. Revision of the ACMC manual to clarify the importance of scoring all observable items and then implementing further research is warranted.

The differences between inexperienced and well-trained raters that have been demonstrated in this study clearly illustrate the importance of training and experience for consistent ratings both within and between raters. Firstly, the more experienced the rater, the fewer the items that were inconsistently rated, as seen in Table 1. Secondly, the most experienced rater showed no systematic difference between sessions 1 and 2 (Fig. 2), and thirdly, there was a systematic difference both between raters A and B and between raters A and C in subject measures (Fig. 3). Moreover, the less experienced raters had more missing items than the more experienced rater. The results suggest that more than 10 weeks of practice are required for consistent ACMC ratings.

In several studies [14, 15] the variability between raters in terms of severity or leniency has been demonstrated. Raters seem to establish an individual profile of severity and usually tend to keep this consistent across clients and protocols [14]. The results from this study, with rater severity varying, in session 1 from –0.51 (lenient) to 0.27 (severe) logits, and in session 2 from –0.37 to 0.28 logits (Table 2), and with a calibration difference of less than 0.16 logits, are in line with those findings. This variation between raters in their manner of rating, and in their variability in calibration severity, may partly explain the low interrater agreement (Kappa 0.47 and 0.60) shown in this study. There was no logic, however, in the difference in severity between raters with different degrees of experience (Table 2), which indicates that variability in severity is more dependent on rater personality than on experience.

The impact of tasks and rater severity on subject ability measures has been described earlier [8]. Besides the judgement of the specific rater, the items may not be equally difficult to perform in different situations or tasks (e.g. feeding, cooking, doing crafts). These are factors that need to be considered and will require evaluation with a larger sample.

The results from this study have given indications of how much experience is needed for reliable measures with the ACMC. A further study on larger populations to address both rater calibration stability and rater severity is in progress.
In conclusion, until the ACMC can adjust for rater severity we recommend that for clinical trials or follow-up, the same rater should perform the ACMC. The assessment method requires training and practice.

ACKNOWLEDGEMENTS

Financial support was granted from the Norrbacka-Eugenia Foundation, the Frimurare-Barnhuset Foundation, the Research Unit at Örebro University Hospital, and the Research Committee of Örebro County Council.
REFERENCES


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Psychosocial adjustment in Swedish children with upper-limb reduction deficiency and a myoelectric prosthetic hand

L Hermansson1,2, A-C Eliasson2 and I Engström3

Aim: To study psychosocial adjustment and mental health in children with upper-limb reduction deficiency and a myoelectric prosthetic hand. Methods: Sixty-two parents of children aged 8 to 18 y old answered a questionnaire concerning competence and behaviour/emotional problems in their children. Of the 62 children, 37 adolescents aged 11 to 18 y old answered questionnaires concerning competence, problems and mood state. The results were compared with Swedish normative data. The children were divided into five groups based on degree of myoelectric prosthetic use. Results: Children with upper-limb reduction deficiency and a myoelectric prosthetic hand showed social competence and behaviour/emotional problems similar to Swedish standardized norms. However, withdrawn behaviour was significantly higher in all children, social competence was significantly lower in girls, and social activities were significantly lower in older children with upper-limb reduction deficiency. There was a significant difference between prosthetic use groups. Non-users had significantly more delinquent behaviour problems than full-time users. There was an interaction between gender and prosthetic use in their affect on competence and behaviour/emotional problems, yielding two contrasting patterns.

Conclusion: Children with upper-limb reduction deficiency and a myoelectric prosthetic hand are 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.

Key words: Arm prostheses, depression, psychopathology, upper-limb deformities

Children with upper-limb reduction deficiencies are often fitted with myoelectric prosthetic hands to compensate for functional and cosmetic discrepancies. While it is clear that an artificial arm can compensate for parts of the arm function, less is known about how the children with these devices adjust psychosocially and how their mental health is to be perceived.

Limb reduction deficiencies (LRD) are rare conditions that have been recognized and described through art and literature for more than 2000 years (1). The yearly prevalence of LRD is estimated at 6.7/10000 births (2–4). According to a study in Finland (2), as many as 80% of these children may have upper-limb reduction deficiencies (ULRD) (69% upper limb only, 11% both upper and lower limb). With a mortality rate of 20% (3) and an underestimation of 5% (5), the yearly prevalence of children with ULRD in Sweden is estimated to be 0.05%.

Few scientific studies have been carried out in Sweden. Also, there is no information on the type of intervention that the children in these studies have received. The available studies have shown that the chronic strain of living with ULRD has a potentially negative impact on the psychological adjustment. This has been found with regard to self-esteem (6), depressive symptoms (7) and low perceived physical appearance (8) in these children. The findings have been discussed in terms of a “new hidden morbidity” in paediatric practice (9).

Factors that might potentially mediate the relationship between chronic disorders and psychological functioning have been described. One major factor in children with limb deficiencies is social support, especially from classmates (10, 11), but family support also seems to have a positive effect on depressive symptoms (7), psychosocial adaptation (12) and per-
ceived physical appearance (8). In addition, high perceived physical appearance has been found to be a predictor of low depressive symptomatology and high general self-esteem (8,13). Access to a functional prosthesis is a factor that is considered to affect adjustment to a deficiency by facilitating independence (14). This indicates that supplying ULRD children with prosthetic hands with a cosmetic appearance and a functional grasp, as in a myoelectric prosthetic hand, combined with subsequent training and support in using the prosthesis, may have a beneficial impact on the children’s psychosocial health. This has not, however, been confirmed in empirical studies.

An indication of the potential effect of myoelectric prostheses is that users of such prostheses, whether they use them actively or passively, seem to have greater levels of perceived social support than non-users (15).

As mentioned above, previous findings have linked low levels of perceived social support with depressive symptomatology (7) in this population. This indicates that children using a myoelectric prosthesis may experience less pronounced depressive symptoms than has been suggested previously. From this perspective, it may be of considerable value when investigating the depressive symptoms in children with a myoelectric prosthetic device to compare them with a normative sample. The aim of the present study was thus to investigate the psychosocial adjustment and mental health in children with ULRD and myoelectric prosthetic hands, in comparison with standardized norms.

The specific objectives of the study were: a) to investigate the social competence, behaviour/emotional problems and depressive symptoms in Swedish children with ULRD who have been fitted with a myoelectric prosthetic hand at an early age; b) to determine whether social competence and any behaviour/emotional problems and depressive symptoms differ between children with ULRD who have been fitted with a myoelectric prosthetic hand and normative samples; and c) to determine whether social competence and any behaviour/emotional problems differ between children with various levels of prosthetic use.

Participants and methods

Participants

Sixty-six children and adolescents, aged 8 to 18 y, with ULRD and a history of myoelectric prosthetic use were identified by the Limb Deficiency and Arm Prosthesis Centre (LDAPC) at O¨ rebro University Hospital, Sweden. The sample in this study comprised all children eligible during the relevant period of the study (December 2001 to June 2003). The families had all been referred soon after the child’s birth by paediatricians throughout Sweden for fitting of a prosthetic hand in the child. Between the ages of 1 and 3 y, all children had had a myoelectric prosthesis fitted and had undergone subsequent training and follow-up according to the limb-fitting scheme at the LDAPC in O¨ rebro. Fifty-eight families were regularly attending the clinic for medical check-ups, occupational therapy and prosthetic maintenance. Eight families were no longer patients at the clinic. They had been transferred to other clinics in the country or discharged at their own request after discontinuing use of the prosthesis after at least 3 y at the LDAPC. Information in the patients’ files made it possible, however, to contact them by letter and telephone, and they were included in the study. During the study period, two families failed to attend the scheduled visit at the LDAPC and were therefore never asked to participate. Hence, 64 children and adolescents and their parents (mother or father) were asked to participate. The children and adolescents were asked if they agreed to their parents answering questions about their social competence and behaviour/emotional problems. In addition, the adolescents (11 to 17 y) were asked if they agreed to participate themselves. The children (8 to 10 y) were considered too young to answer the questionnaires described below.

Initially, all parents (n = 64) and all but two (girls, 12 and 17 y) adolescents (n = 38) agreed to participate in the study. The girls agreed, however, to participation of their parents. Hence, all children and adolescents agreed to their parents’ participation on their behalf. Eventually, two families failed to participate (3%): one family with a 9-y-old boy who was attending another clinic, and one family with a 16-y-old boy with a partial hand deficiency who was no longer using a prosthesis. Finally, the parent-reported study group consisted of 62 children (31 girls, 31 boys) and the children’s self-reporting group consisted of 37 adolescents (18 girls, 19 boys). The mean age of the children in the parent-reporting group was 12.6 y (median 12 y). The mean age in the children’s self-reporting group was 14.8 y (median 16 y). Fifty-seven children had deficiencies present at birth and five children had acquired amputation. More than two-thirds (69.4%) of the deficiencies were on the left side and one-third (30.6%) were on the right side, and the majority (66%) were at the transradial level. One child had bilateral deficiencies with a prosthesis on the right arm.

Measurements

Child behavior checklist (CBCL). The parents completed the CBCL (16), which has two parts. Part I consists of seven competence areas with 2–8 items in each, and part II covers 118 problem/behaviour items, all rated for the past 6 mo. As judged from earlier results, the CBCL is a useful screening instrument in identifying behavioural and emotional problems and social competence in children and adolescents with limb deficiencies (9).

One father of an adolescent girl failed to answer enough questions on the CBCL to make the result
useful. Hence, the total mean social competence and behavioural/emotional problem scores for 61 ULRD children are reported. For the CBCL, Swedish normative data are available for children aged 6 to 17 y (17). Forty-nine children and adolescents with ULRD had relevant ages (8 to 17 y) for comparisons with the CBCL normative data.

Youth self report (YSR). The YSR measures self-reported competence items and problems during the past 6 mo (18). This was completed by the adolescents. As with the CBCL, the YSR consists of two parts, 118 behaviour/problem items and seven competence items.

Among the children in the self-reporting study group, one adolescent boy failed to answer enough questions on the YSR to make the result useful. Accordingly, the total means of answers by 36 ULRD adolescents in the YSR are reported. YSR has been validated for Swedish children, and normative data for adolescents 13 to 19 y old are available (19). Twenty-seven adolescents had relevant ages (13 to 18 y) for comparisons with the YSR normative data.

Children’s depression inventory (CDI). The adolescents also completed the CDI (20). This is a 27-item, self-rated, symptom-oriented scale for children aged 7 to 18 y intended to measure the affective state. The CDI has been validated and standardized in several studies. It is one of the most commonly used self-report questionnaires designed to measure childhood depression, and normative Swedish data are available for children aged 8 to 13 y (21) and for adolescents aged 13 to 18 y (22). All 37 adolescents (11 to 18 y) had relevant ages for comparisons with the CDI normative data.

To study the differences in social competence and behaviour/emotional problems between children with various frequencies of prosthetic use, the children were categorized into groups based on level of prosthetic use.

Prosthetic use scale (PUS). The PUS consists of five levels of prosthetic use decided by the wearing pattern and the number of hours for which the prostheses is used: 1) full time; 2) part time; 3) occasional; 4) sporadic; and 5) non-user. For this study, parents and adolescents were asked to report on the PUS how often the myoelectric prostheses had been used during the last 6 mo. The agreement between the parents and adolescents, measured by kappa, was 0.82, which is considered as almost perfect (23). Thus, for the analyses of the CBCL, the parents’ ratings on the PUS were used. According to the parents, a majority of the children (58.1%) used their prostheses every day (35.5% full time and 22.6% part time), 12.9% used it occasionally, 8.1% used it sporadically and 21% no longer used the prostheses. In boys, the PUS was dependent on functional level, indicating that boys with partial hand deficiency used the prostheses significantly (p = 0.016) less than boys with transradial deficiency. In girls, there was no significant difference in prosthetic use at different functional levels.

Procedure

The study was approved by the Örebro County Council Research Ethics Committee. Informed consent was obtained from all participants.

The families were first informed about the study by a notice published in the biannual “Newsletter” from the LDAPC. Prior to data collection, all identified families received a letter with further information about the study, in which they were told that they could expect to be contacted by the researcher by telephone (discharged patients) or at their scheduled visit at the clinic. Since not all participants were attending the LDAPC, two data collection procedures were used. 1) At the time of a scheduled clinic appointment, the researcher met both child and parent, and verbal consent was obtained from both of them. These participants (56 families, 88%) were subsequently given more information about the study and had the opportunity to ask questions, and they were then asked to complete the questionnaires. In one case, this procedure was transferred to a local therapist at a secondary clinic in Sweden so that she could assist with data collection from the family who were no longer attending the LDAPC. 2) Seven families were contacted by telephone, and verbal consent was obtained from the parent and the child. These participants (6 families, 9%) were then given more information about the study procedure and the questionnaires. Also, more information about the study was mailed to the families together with the questionnaires. The researcher was available by telephone at all times to answer any questions regarding the administered instruments.

Statistical analyses

In accordance with the manuals for each instrument, the data for the dependent variables social competence (CBCL, three scales; YSR, two scales), behavioural/emotional problems (CBCL/YSR, eight/nine syndrome scales) and depressive symptoms (CDI, five factor scales) were expressed as indices derived from a summation of the scores from 2–20 questions each. This corresponds to the way in which the normative data were derived. Subsequently, these data were compared with the normative data. Student’s one-sample t-tests with hypothesized mean values obtained from the normative data were performed to test for differences between data for the ULRD sample and the normative data.

To detect any possible effects of age or gender on social competence, emotional/behavioural problems or depressive symptoms in the ULRD sample, one-way analyses of variance (ANOVA) was performed. Because of the small number of participants, further ANOVA to study the effects of prosthetic use were carried out using the parent-rated CBCL only. These
analyses were performed in two different ways, i.e. both the observed data and the calculated individual differences between the observed data and the normative values were analysed. This ANOVA was done with the main factors age, gender and prosthetic use and two-way interactions between the main factors. P-values lower than 0.05 were accepted as statistically significant. The magnitudes of the effects were measured according to Cohen’s criteria for effect sizes (23), where 0.10 = small effect size, 0.25 = medium effect size, and 0.40 = large effect size (24).

Table 1. Means and standard deviations (within parentheses) of social competence scores in children with upper-limb reduction deficiency compared to Swedish standardized normative data, measured by the Child Behavior Checklist.

<table>
<thead>
<tr>
<th>Area</th>
<th>All 4 (n = 61)</th>
<th>Allb (n = 49)</th>
<th>Swedish norms 4</th>
<th>Swedish norms 5</th>
<th>Girlsb  (n = 25)</th>
<th>Boysb  (n = 24)</th>
<th>Swedish norms 5</th>
<th>8–11 y (n = 29)</th>
<th>Swedish norms 5</th>
<th>12–16 y (n = 20)</th>
<th>Swedish norms 5</th>
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<tbody>
<tr>
<td>Total score</td>
<td>16.5 (5.4)</td>
<td>16.4 (3.3)</td>
<td>17.0 (3.4)</td>
<td>15.8 (3.4)</td>
<td>17.1 (3.2)</td>
<td>17.1 (3.1)</td>
<td>16.9 (3.5)</td>
<td>16.9 (2.9)</td>
<td>16.7 (3.3)</td>
<td>15.8 (3.7)</td>
<td>17.2 (3.4)</td>
</tr>
<tr>
<td>Activities</td>
<td>5.1 (1.6)</td>
<td>5.2 (1.6)</td>
<td>5.3 (2.0)</td>
<td>5.0 (1.7)</td>
<td>5.2 (1.9)</td>
<td>5.3 (1.4)</td>
<td>5.1 (2.0)</td>
<td>5.5 (1.4)</td>
<td>4.9 (1.9)</td>
<td>4.7 (1.6)</td>
<td>5.4 (2.2)</td>
</tr>
<tr>
<td>Social</td>
<td>6.5 (1.7)</td>
<td>6.4 (1.7)</td>
<td>6.7 (1.8)</td>
<td>6.0 (1.5)</td>
<td>6.6 (1.8)</td>
<td>6.8 (1.7)</td>
<td>6.7 (1.9)</td>
<td>6.4 (1.6)</td>
<td>6.6 (1.8)</td>
<td>6.5 (1.8)</td>
<td>6.7 (1.9)</td>
</tr>
<tr>
<td>School</td>
<td>4.9 (1.0)</td>
<td>4.9 (1.0)</td>
<td>5.1 (0.8)</td>
<td>4.9 (1.1)</td>
<td>5.2 (0.7)</td>
<td>5.0 (1.0)</td>
<td>4.9 (0.8)</td>
<td>5.1 (0.8)</td>
<td>5.0 (0.7)</td>
<td>4.6 (1.3)</td>
<td>5.1 (0.8)</td>
</tr>
</tbody>
</table>

Note: high numbers are positive, indicating more competence.
* p < 0.05 (Student’s t-test with two-tailed levels of significance).

Results
Presented in the following are the results of comparisons between data from children with ULRD and data from Swedish normative samples concerning social competence and emotional/behavioural problems based on the answers on the CBCL and YSR, and depressive symptoms based on the answers on the CDI.

Social competence
Social competence, measured by the CBCL, part I, total score (Table 1), did not differ significantly between children with ULRD and the Swedish standardized sample (17). On the activity subscale, however, children aged 8 to 11 y old with ULRD had significantly (p = 0.037) higher scores, indicating that these children performed activities more frequently than the Swedish normative sample. In contrast, children aged 12–16 y old with ULRD had lower, though not significantly lower (p = 0.06) scores on the activity subscale. Further, girls with ULRD displayed a tendency (p = 0.08) towards lower social interaction scores on the CBCL, part I, compared with Swedish norms. This finding was supported by the YSR (part I; Table 2) scores on the social competence scale. Here, adolescent girls with ULRD rated themselves as having significantly (p = 0.035) lower social interaction scores than the normative sample of girls (19). There were no differences in social competence depending on level of deficiency.

Emotional/behavioural problems
The total problems score in the ULRD group, as measured by the CBCL, part II (Table 3), did not differ significantly from the Swedish standardized norms (17). The sum of the subscales constituting the broad-band dimensions measuring internalizing and externalizing symptoms were, although increased, not significantly different from the normative data. One exception, however, was the subscale measuring withdrawn behaviour, the score for which was significantly higher (p = 0.037) in the ULRD children than the Swedish normative data. Regarding gender, the girls had a significantly higher total problem score (p = 0.030) and a significantly higher withdrawn behaviour problem score (p = 0.051), and, regarding age, the older children (12 to 16 y) showed significantly higher anxious/depressed behaviour (p = 0.049) and significantly higher attention behaviour problem (p = 0.036) scores compared with the Swedish normative sample (17). In contrast, in the self-reported behavioural and emotional problems, measured by the YSR, part II (Table 4), the

Table 2. Means and standard deviations (within parentheses) of social competence scores in adolescents with upper-limb reduction deficiency compared to Swedish standardized normative data, measured by the Youth Self Report.

<table>
<thead>
<tr>
<th>Area</th>
<th>All  (n = 36)</th>
<th>Allb (n = 27)</th>
<th>Swedish normsb</th>
<th>Swedish normsb</th>
<th>Girlsb (n = 15)</th>
<th>Boysb (n = 12)</th>
<th>Swedish normsb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total competence</td>
<td>11.4 (2.9)</td>
<td>11.2 (2.9)</td>
<td>10.7 (2.9)</td>
<td>12.3 (2.8)</td>
<td>11.8 (2.9)</td>
<td>12.6 (2.9)</td>
<td></td>
</tr>
<tr>
<td>Activities</td>
<td>2.9 (1.4)</td>
<td>2.9 (1.4)</td>
<td>2.7 (1.3)</td>
<td>3.3 (1.4)</td>
<td>3.2 (1.4)</td>
<td>3.1 (1.4)</td>
<td></td>
</tr>
<tr>
<td>Social</td>
<td>6.4 (1.9)</td>
<td>6.2 (1.9)</td>
<td>6.0 (1.6) *</td>
<td>7.0 (1.7)</td>
<td>6.5 (2.3)</td>
<td>7.5 (1.74)</td>
<td></td>
</tr>
</tbody>
</table>

Note: high numbers are positive, indicating more competence.
* p < 0.05 (Student’s t-test with two-tailed levels of significance).
Means and standard deviations (within parentheses) of emotional and behavioural problem scores in children with upper-limb reduction deficiency compared to Swedish norms. No significant differences depending on functional level were detected.

### Depressive symptoms

The adolescents aged 11–12 y with ULRD (n = 10) had a total CDI score of 3.20 (SD 1.55), which was significantly (p < 0.000) lower than the score of 6.25 in the Swedish normative sample (21); i.e. they had less depressive symptoms. In adolescents aged 13–17 y old with ULRD (n = 27), there was a total CDI score of 7.76 (SD 7.78), which was lower but not significantly different from the score of 8.9 in the Swedish normative data (22). Because of the small sample sizes (11–12 y, n = 10; 13–17 y, n = 27), further comparisons in gender-based subgroups were performed only in the older age group (13–17 y). In this group (Table 5), boys with ULRD had overall lower scores, and significantly lower scores for anhedonia (p = 0.019) and interpersonal problems (p = 0.006) factors, compared to the Swedish normative data. Moreover, girls with ULRD had lower scores for most factors but scores that were higher, though not significantly so, than the normative data on anhedonia. Boys with partial hand deficiency had significantly higher scores on interpersonal problems (p = 0.01), whereas in girls there were no differences dependent on functional level.

### Effects of age and gender

Overall, girls and older children with ULRD tended to have lower total competence scores on the CBCL than the corresponding groups in the normative sample (Table 1). However, when age or gender was used as an independent variable in the ANOVAs, there was a tendency to an effect on the total competence, measured by CBCL, part I, but this was not statistically significant. Nor was any effect of age or gender on the CBCL, part I, total competence found when individual factor scores were analysed. Nevertheless, the differences related to age became more evident by this procedure. In this analysis, older children with ULRD had significantly lower scores on the activity (p = 0.005, \( \chi^2 = 0.16 \)) and school (p = 0.05, \( \chi^2 = 0.08 \)) subscales than younger children with ULRD. This was surprising, since in the Swedish normative sample older children scored higher than younger children, especially on the activity subscale. These results thus indicate that older children with ULRD are much less active than their non-deficient peers.

When age or gender was used as an independent variable in analyses of the calculated differences between children with ULRD and the normative sample, no significant differences were found on the score for thought problems was significantly (p = 0.007) lower in ULRD girls, and the score for somatic complaints was significantly (p = 0.010) lower in ULRD boys, compared with the Swedish standardized norms. No significant differences depending on functional level were detected.
CBCL, part II, problem subscales. There was a difference, however ($p = 0.07$, $\chi^2 = 0.07$), between the adjusted values for boys and girls on the delinquent behaviour problem subscale, pointing in different directions based on gender.

**Effects of prosthetic use**

No significant effects of prosthetic use on the CBCL, part I, competence scales were found. However, on the delinquent behaviour problem subscale, there was a significant difference between PUS groups ($p = 0.01$, $\chi^2 = 0.25$). Scheffe’s post hoc test revealed that children with myoelectric prostheses who did not use them (PUS 5, $n = 8$) girls $n = 5$) had significantly more delinquent behaviour problems than those who were using the prosthesis full time (PUS 1, $n = 18$, girls $n = 12$) ($p = 0.03$). When the children were divided into two groups based on gender, there was a significant difference between PUS groups for boys on CBCL, part I, social competence ($p = 0.02$), and for girls on CBCL, part II, total problems score ($p = 0.04$). However, Scheffe’s post hoc test failed to determine which PUS group was different from the others in either the total competence or the total problem scale.

In all, the small number of participants, when divided into subgroups by gender, meant that possible effects on the dependent variables were lost. However, the differences between girls and boys, as shown in Tables 1–5, prompted further comparisons based on gender. The interaction between gender and prosthetic use in their effect on social competence and emotional/behavioural problems resulted in two contrasting patterns. As illustrated in Fig. 1, the total competence score decreased with decreasing prosthetic use in girls, and increased with decreasing prosthetic use in boys. In contrast, the total problems score increased with decreasing prosthetic use in girls, whereas in boys the pattern was less clear (Fig. 2).

**Discussion**

The main finding in this study is that Swedish children with upper-limb reduction deficiencies who have initially been fitted with a myoelectric prosthetic hand...
are doing quite well. The global results show no significant differences between these children and the normative samples. Certain results of the study, however, need to be highlighted.

**Psychosocial adjustment**

The results for some of the CBCL subscales indicate that children with ULRD may have significant problems compared to other, non-disabled children (Table 3). First, all ULRD children, irrespective of age or gender, had higher scores on the withdrawn behaviour subscale. This may be due to attitudes in society towards visible physical differences. From childhood onwards, children with ULRD are aware of people’s looks and questions. Although they become used to this, it is an ongoing stressor, which they very likely try to avoid, perhaps by keeping more to themselves. The finding that girls with ULRD have lower social interaction competence than girls in the normative sample may also be a result of...
this. Further, older children with ULRD showed lower competence on the activities subscale than younger children with ULRD. This was surprising, and is in contrast with the normative data for the activities subscale, according to which older children were more competent than younger ones (17). Altogether, these results suggest social stigmata that may be associated with the deficiency.

The normative study on social competence and problems in Swedish children (17) showed that children from the middle SES groups were rated as more competent than children from upper and lower SES groups. It was also found that children from the lower SES groups had significantly higher problem scores than those in the other SES groups. In the YSR normative data, they found differences due to SES on the competence scales but not on the problem scales (19). ULRD are expected to occur in all social groups; hence, in this study, SES of the parents was not registered. Based on the earlier findings on competence, it is reasonable to assume that children in families with high SES would have more competence in using prostheses. Also, parents with high SES would probably support their children to use the prostheses and not to become delinquent. Regarding the self-reported behavioural problems in this study (Table 4), we were surprised by the finding that most outcomes were more positive for the ULRD children than for the Swedish norm populations and the findings of less depressive symptoms (Table 5) in this population. One possible explanation for this may be that the ULRD children in this study belong to a selected population of children whose parents are prepared to travel long distances and stay away from work in order for their children to receive special treatment. In contrast to the norms, both the ULRD children and their parents in this study have received special treatment. In contrast to the norms, both the ULRD children and their parents in this study have been subjected to comprehensive multi-professional care over the years.

The results of the present study correspond to earlier observations by Singleton (25). She found that mean CBCL scores in juvenile amputees with myoelectric prostheses were normal compared to those of non-handicapped children, but that families had lower levels of adjustment and more negative self-concepts, and there was a decrease in coping skills in older children. The results from the parents’ and the adolescents’ reports on the CBCL/YSR in this study are very similar, supporting the credibility of the results. In contrast, the results from the parent-rated CBCL questionnaire in the present study do not correspond with the earlier results of Varni and Setoguchi (9). In their study of 111 children and adolescents 6 to 17 y of age with limb deficiency, they found significantly lower social competence and significantly more behaviour/emotional problems than in a normative sample. This was not verified in the present study.

**Difference in various levels of prosthetic use and gender**

When studying children with ULRD, as in this study, we have observed an interaction between prosthetic use and gender in their effect on the variables studied. This finding indicates that future analyses of children with ULRD need to be made separately in groups based on gender. Accordingly, we reported the distribution of total competence and total problem scores among different levels of prosthetic use separately for boys and girls.

The present study is, to our knowledge, the first study ever to address the relations between competence, behavioural/emotional problems and prosthetic use in boys and girls with ULRD and a myoelectric prosthetic hand. At first, we expected that the competence scores and the problem scores would follow the mean score, independent of level of prosthetic use. Instead, the results indicate that ULRD and prosthetic use influence the psychosocial adjustment in children differently depending on gender (Figs 1 and 2). The reason for this is not evident. In search of possible explanations, we noted that group 2 (part-time users, n = 6) consisted of boys who were 8–11 (mean 9) y old and thus were possibly in a pre-pubertal crisis. This period is known from clinical practice as a time when many children (mostly boys) question the use of the prosthesis and shift from an earlier compliance with their parents’ wishes concerning the use of the prosthesis to revolting and not using it. This could explain why boys with unexpectedly high problem scores were part-time users (PUS 2, Fig. 2). Also, this may be the reason why these boys scored low on the competence scale (PUS 2, Fig. 1). Another explanation of the differences between boys and girls may originate from experiences of different competencies and problems. A depressed girl, for instance, who does not perceive herself as attractive with the myoelectric hand may choose to convert to a passive, more cosmetically appealing hand. A boy, on the other hand, may perceive himself as more competent, proving his own skills by not using a myoelectric hand. Also, boys and girls perform different activities, using different objects for which the prosthesis may be regarded as a problem by the boys and as a help by the girls.

One further reason for the difference in mean competence and problem scores in this study between boys and girls in the group of children who were no longer using a myoelectric prosthesis (PUS 5) may also be the reason for not using it. All of the boys in this category (n = 3) had partial hand deficiency. The discontinuation of the use of the prosthesis indicates that they had developed competence in using their residual limb and had no further need for a prosthesis. In contrast, the girls in this category (n = 5) had transradial deficiency and had all changed to using passive prostheses, which have a more naturalistic appearance but are less functional.
The impact of prosthetic use on psychosocial adjustment

In Sweden, most children with ULRD are provided with myoelectric prostheses. Very rarely are children fitted with split-hooks. This policy is in contrast to the practice in amputee clinics in the USA, such as CAPP (Child Amputee Prosthetics Project), where the availability of funds plays a major role in determining when and if a myoelectric prosthesis is prescribed (26). According to that report (p. 253), many children in the CAPP wear a body-powered terminal device, i.e. a prosthesis activated by a shoulder harness. Most of the earlier studies on psychosocial adjustment in limb deficiency referred to in the present paper originate from the CAPP.

The mood state of the children aged 13–17 y, once fitted with myoelectric prosthetic hands, as measured by the CIDI, showed scores that did not differ significantly from those of the general population. The total scores as well as the scores of most subscales were lower than the norms, indicating good mental health in this respect also. Similar results were obtained by Varni et al. (7, 10) in children 8–13 y old. In contrast to their findings, children in the present study of ages comparable to those in the Varni studies (11–12 y old) had significantly lower scores for depressive symptoms compared to the Swedish normative data. In one of their studies, Varni et al. (7) found that greater levels of stress generated by chronic strain from limb deficiency and daily microstressors are related to more depressive symptoms. This suggests that children in the present study with myoelectric prostheses experienced lower levels of stress, i.e. less chronic strain from the deficiency and daily microstressors, than the children in Varni et al.’s study.

In the earlier studies, depressive symptoms in ULRD children were found to be affected by social support, both directly and indirectly (through perceived physical appearance). According to Glenn (15), users of myoelectric prosthetic hands experience more social support than non-users. Also, by using a myoelectric, cosmetically appealing, prosthetic hand, perceived physical appearance may increase, and people’s looks are avoided, which in turn affects depressive symptomatology. The results from both this study and the one reported by Singleton (25) indicate that myoelectric prostheses may have a beneficial effect on psychosocial adjustment in children with ULRD, especially girls. However, as indicated by earlier results on the effect of SES on competence and problems, social support may be the primary causal agent, giving rise both to prosthetic use and to fewer depressive symptoms.

In conclusion, the main result of this study is that children and adolescents with ULRD fitted with myoelectric prostheses exhibit good mental health as measured both by instruments assessing general behaviour problems and symptoms and by instruments aiming at describing the mood state and depressive symptoms. Girls and older children, however, display more problems than the group as a whole, indicating that they should be monitored and supported more carefully. We believe that a specialized centre for medical support of children with ULRD is necessary in order to help these children adjust to their deficiency.

Acknowledgements.—We wish to acknowledge our gratitude to the parents and children, without whose contribution this study would not have been accomplished. We would also like to thank Lennart Bodin, statistician, Unit of Medical Statistics and Epidemiology, Örebro University Hospital, Örebro, and Department of Statistics, Örebro University, Örebro, for help with the statistical analyses and advice on the manuscript. We thank Ingela Andersson, medical secretary, and the rest of the staff at the Limb Deficiency and Arm Prosthesis Centre, Örebro University Hospital, Örebro, for help with this study. We also thank Elisabeth Fornell, neuropaediatrician, Department of Woman and Child Health, Karolinska Institute, Stockholm, for advice on the manuscript. Financial support was granted from the Norrbucks-Eugenia Foundation, Queen Silvia’s Jubilation Foundation, the Swedish National Association for Disabled Children and Young People (RBU), the Foundation for Medical Research and the Research Unit at Örebro University Hospital, and the Research Committee of Örebro County Council.

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Received Feb. 26, 2004; revision received May 14, 2004; accepted June 16, 2004