Chapter 1

Introduction
Young individuals who were born with limb deficiencies experience challenges in daily life on different levels of functioning. Either or both psychosocial or physical challenges stimulate the common efforts of young individuals, their families, and healthcare professionals to search for enabling solutions. A particular group of the young individuals with limb deficiencies was the focus of this current research: young individuals with congenital upper limb reduction deficiencies (ULRD). Aspects of the functioning of young individuals with ULRD were addressed in this thesis according to the International Classification of Functioning, Disability, and Health (ICF).

The ICF is a comprehensive model used in healthcare and by researchers to describe health and disability. Combining psychological and physical aspects of functioning, the ICF provides an overall description of a person’s well-being by considering several components: body functions and structures, activities and participation, and personal and environmental factors. The ICF components can influence each other. For instance, ULRD is a consequence of the failure of formation of upper limb body structures. Depending on the degree to which the limb structure is affected, children with ULRD suffer from a more or less impaired upper limb. Consequently, functioning might be hindered since the hands play a major role in exploring and learning, are indispensable in activities and participation, and facilitate non-verbal communication. To successfully rehabilitate young individuals with ULRD, it is important to focus on all ICF components.

The themes that inspired the research in this thesis were: (1) determining the actual numbers of young individuals with limb defects, (2) identification of difficulties in activities of daily living due to ULRD and solutions used to facilitate the difficulties, (3) assessment of the impact of solutions on the psychosocial and physical functioning, and (4) finding an adequate functional test to measure hand function in young individuals with ULRD. As such, the thesis addresses aspects of ICF components and is structured in two main research directions. The first part of this thesis is dedicated to studying the epidemiology of limb defects; use and impact of help accessories, including prostheses and adaptive devices; and rehabilitation care on functioning of young individuals with ULRD. Data are provided regarding several ICF components, including body structures and functions, activities and participation, and personal and environmental factors. The second part focuses on an existing measurement test, the Southampton Hand Assessment Procedure (SHAP) that is used to evaluate hand function (body functions). SHAP comprises tasks simulating activities of daily living (activities). Studies were conducted to determine the learning effects of SHAP tasks, to
adapt SHAP for use in children and test the reliability of this adapted version, and to propose an alternative scoring system for SHAP.

I. Young individuals with ULRD–epidemiology, functioning, prostheses, and adaptive devices

In order to understand the magnitude of the affected population, it is important to have epidemiological estimates of children with congenital limb defects (CLD), including children with ULRD. In Europe, 36 cases per 10,000 births were reported for CLD from 2003–2007. The reported birth prevalence of ULRD varied from 3.0 per 10,000 births in Australia from 1980–1990 to 5.3 per 10,000 births in Finland from 1993–2005. In the Netherlands, however, an updated and detailed description of the birth prevalence for CLD is lacking. This constitutes the research question:

*What is the birth prevalence of CLD in the northern Netherlands? (Chapter 2)*

In rehabilitation care, prostheses are considered good treatment solutions for young individuals with ULRD to improve their functioning. Between 24–49% of young individuals with ULRD refuse to wear prostheses, whereas others continue to wear it. The reasons for prosthesis rejection have been learned from parents and healthcare professionals caring for children with ULRD; these reasons include a lack of functional gain and cosmesis, discomfort, and parental dissatisfaction with prostheses and rehabilitation. On the other hand, reasons for wearing a prosthesis as well as children’s opinions about rehabilitation are underexplored. To address this issue, this thesis addresses answers to the questions:

*Why do children, early, and late adolescents with below-elbow ULRD choose to wear or not wear prosthesis? How do they view their rehabilitation experience? (Chapter 3)*

The purposes for prescribing a prosthesis are well-intended, primarily to improve the functioning of young individuals with ULRD. However, considering the rejection rates for prostheses, the question arises “are prostheses the only or the best solution?” Few studies describe the existence of alternative devices to prostheses; these alternatives are known as terminal, assistive, or adaptive devices. The adaptive devices are meant to facilitate the activities of daily living and recreation/play that further facilitate participation. There is,
however, no information about the use of or satisfaction with adaptive devices in young individuals with ULRD. Therefore, this thesis compared adaptive devices to prostheses; prostheses were not considered adaptive devices. The following research questions have been approached:

How many adaptive devices are actually used? Are children with ULRD more satisfied and socially better-adjusted with adaptive devices than with prostheses? (Chapter 4)

II. ULRD–functional tests: SHAP and SHAP-C

Young individuals with ULRD function well in activities of daily living with or without using prostheses. Nevertheless, evaluation of impaired hand or upper limb function in activities of daily living helps rehabilitation professionals monitor changes in functioning and adjust the treatment of young individuals with ULRD. It would be ideal for clinicians to be able to assess functional abilities of any type of hand (normal, impaired, or prosthetic). The ability to discriminate between functional scores for specific hand grips would focus the treatment or prosthetic training on improving the less functional grips. Existing tests are rather limited to evaluating only specific impaired hands; for example, spastic hands due to cerebral palsy or prosthetic hands due to ULRD or amputation. Some tests require preliminary training of the assessors.

The Southampton Hand Assessment Procedure (SHAP) is a promising test for unilateral hand function, including normal, impaired, or prosthetic hands. The scores generated by SHAP give insight into the functional status of six hand grips (spherical, tripod, power, lateral, tip, and extension) and a general functional score. SHAP uses an objective data collection method, self-timing by the subject, and requires 20–30 minutes to be completed (SHAP kit, Figure 1). The reliability of SHAP was good in healthy adults; currently, SHAP is applicable for adults only.

To adapt and use SHAP reliably in children with an impaired hand requires research. However, before adapting SHAP for children, more work is needed to determine the psychometric properties of SHAP in adult prosthetic users, as recommended by the Upper Limb Prosthetics Outcome Measures group (ULPOM).
Novice prosthetic users are particularly influenced by learning effects due to carryover information from previous measurement sessions.\textsuperscript{30} SHAP is used in rehabilitation to monitor the training of novices using a prosthesis. Clinicians at University Medical Center Groningen often noted improvement in SHAP functional scores that did not represent the actual performance of the prosthetic user, possibly due to learning effects of SHAP tasks. Apart from the learning effects, each SHAP task has a time limit, and exceeding this time limit affects the SHAP scores. Whether these time limits are appropriate for novice prosthetic users is yet to be investigated. This thesis addresses the following questions:

- Are SHAP tasks affected by learning effects? Are the time limits for each SHAP task adequate for novice prosthetic users? (Chapter 5)

Modifying SHAP for use in children (SHAP-C) considers several steps. The SHAP kit consists of 26 tasks performed with specific objects (Figure 1) that have to be downsized to accommodate a pediatric hand and prosthesis. The SHAP protocol used to guide proper measurement requires adjustments as well, for instance, the data collection method.

\textbf{Figure 1.} SHAP kit content with the objects used to perform the tasks (adult version)

SHAP scores range from 0–100, with 100 representing optimal function. The scores are calculated relative to a normative dataset from unimpaired adults.\textsuperscript{27,31} These norms are not representative for the pediatric population, because children are on a different motor-learning
stage and hand capabilities of children are less developed compared to adults.\textsuperscript{32,33} To have estimates for normal function of the pediatric hand, a normative dataset for SHAP-C needs to be collected from unimpaired children.

Another important requirement is to test the reliability of the adapted test. However, prior to determining reliability in children with ULRD or other type of hand impairments, it is more logical to first determine if SHAP-C is reliable in healthy children. The first steps to creating a version of SHAP for children have been presented:

\textit{Adjustment of SHAP for use in children, determination of normative values, and the reliability of SHAP-C. (Chapter 6)}

The last study in this thesis is the critical observations collected during the above-mentioned studies about SHAP and identifies problems with its scoring system. The SHAP scores are generated on a website after filling in the collected times for each task.\textsuperscript{34} The web-based system then provides a functionality profile with six scores for the hand grips and a general score known as the index of function (IOF). To understand how each task influences the hand grip and IOF scores, the underlying formulas need to be understood. The existing literature about SHAP\textsuperscript{27,31,34,35} is partly unclear and raises questions. What are the exact consequences of exceeding the task time limits on the hand grip and IOF scores? In addition, the SHAP formulas are quite complicated and subject to multiple interpretations. Direct access to the normative data or a detailed explanation of the formulas was not possible due to intellectual property rights. Nevertheless, SHAP has great potential to be an accurate and objective method for evaluating hand function.\textsuperscript{28,29,36} Researchers should strive to provide clinicians with such a test. Furthermore, clinicians would benefit from having a clear and simple understanding of the calculations of SHAP scores. Accordingly, a study is presented that addresses the following issue:

\textit{Identification of issues with the current SHAP scoring system and provision of a simpler calculation method. (Chapter 7)}

The last chapter, chapter 8, places the main results of this thesis into perspective and provides suggestions for healthcare researchers and providers.
Outline of the thesis


Chapter 3 investigates reasons for wearing or not wearing a prosthesis and rehabilitation care experiences in young individuals with below-elbow ULRD, their parents, and professionals.

Chapter 4 provides the first data regarding adaptive devices as alternative solutions to prosthetic use in young individuals with ULRD by determining their satisfaction and social adjustment with adaptive devices compared with prostheses.

Chapter 5 provides documentation of the learning effects of the SHAP in novice prosthetic users; as SHAP consists of several tasks, each with its own time limit, this chapter also evaluates the appropriateness of time limits each SHAP task for novice prosthetic users.

Chapter 6 focuses on how SHAP needs to be adjusted for use in children and determines the first normative data for and reliability of a SHAP version for children.

Chapter 7 addresses issues observed with the SHAP scoring system and proposes a new, simplified method for calculating SHAP scores.

Chapter 8 provides perspective regarding the main results with implications for healthcare and further research.
References


