Life Strategies, Work and Health in People with Usher Syndrome
To Johan, Clara and Julina
Life Strategies, Work and Health in People with Usher Syndrome
Abstract


Introduction: People with Usher syndrome (USH) have an inherited disorder causing congenital deafness or hearing loss combined with progressive vision loss and, in some cases, balance problems. Previous research has shown that people with USH have poor physical and psychological health. Research has, however, demonstrated that there are in-group health differences that have not yet been explored and that there is a lack of studies on life strategies as well as health in relation to working life.

Aim: The aim was to explore the relationship between work and health in people with USH1 and 2 and to explore the experiences of life strategies in people with USH 2.

Methods: A quantitative and a qualitative explorative design was employed comprising two cross-sectional studies of the relationship between work and health in people with USH2 (n=67) and USH1 (n=47). The third study, of life strategies, comprised focus group interviews with people with USH2a (n=14) analyzed by content analysis. The fourth study explored the lived experiences of working life by interviewing people with USH2 (n=7) using an interpretative phenomenological approach.

Results: Study 1 and 2 demonstrated significant differences in health between working and nonworking people. Nonworking people showed significantly poorer health. In study 3, a variety of strategies to prevent and resolve challenges in life, as well as to comfort oneself was identified. The lived experiences of working life in people with USH2 (study 4) disclosed feelings of satisfaction, striving towards a work life balance. Work also disclosed feelings of limitations and uncertainty about the future.

Conclusion: The results, which are discussed in relation to the Meikirsh model of health, demonstrated a complexity. Individual life strategies, psychological flexibility and environmental aspects, such as social determinants have to be considered to receive a comprehensive picture of the relationship between work and health in people with USH. This thesis provides new insights into the health of people with USH.

Keywords: Deafblindness, health, life strategies, lived experience, Usher syndrome, work

Mattias Ehn, The Swedish Institute for Disability Research
Örebro University, SE-701 82 Örebro, Sweden, mattias.ehn@oru.se
Included Studies

The thesis is based on the following four studies, which are referred to in the text by their study numbers.

Study 1

Study 2

Study 3

Study 4

Reprints were made available with the permission of the publishers.
Abbreviations

ADL  Activity of daily living
AUC  Area under the curve
CI   Confidence interval
dB   Decibel
HET  Health on Equal Terms questionnaire
ICF  International Classification of Functioning, Disability and Health
HI   Hearing impairment
NA   Not Applicable
OECD Organization for Economic Co-operation and Development
OR   Odds ratio
PF   Psychological flexibility
PTA  Pure tone average
PTA4 Pure tone average for four frequencies
RP   Retinitis pigmentosa
USH  Usher syndrome
USH1 Usher syndrome type 1
USH2 Usher syndrome type 2
USH3 Usher syndrome type 3
VA   Visual acuity
VF   Visual field
WHO  World health Organization
# Table of Contents

PREFACE................................................................................................................................. 13

INTRODUCTION....................................................................................................................... 15
Usher syndrome ...................................................................................................................... 15
Deafblindness ......................................................................................................................... 16
Disability.................................................................................................................................. 17
Health ...................................................................................................................................... 18
  Social determinants ................................................................------------------------------- 21
  Life demands .................................................................................................................. 22
  Individual potentials ................................................................................................. 22
  Life strategies - psychological flexibility as an acquired potential .................... 24
Work and health .................................................................................................................... 25
  Work, health and disability ...................................................................................... 26

RATIONALE............................................................................................................................ 28

AIM .......................................................................................................................................... 29

METHODS.................................................................................................................................. 30
Usher register .......................................................................................................................... 32
  Hearing ............................................................................................................................ 32
  Vision ............................................................................................................................... 32
  Health survey ............................................................................................................... 33
Participants ............................................................................................................................. 34
  Study 1 ........................................................................................................................... 34
  Study 2 ........................................................................................................................... 34
  Study 3 ........................................................................................................................... 34
  Study 4 ........................................................................................................................... 35
Data collection ........................................................................................................................ 37
  The Health on Equal Terms questionnaire .............................................................. 37
  Focus group interviews ............................................................................................ 38
  Individual interviews ................................................................................................. 38
Data analysis .......................................................................................................................... 39
  Statistics ....................................................................................................................... 39
  Qualitative content analysis .................................................................................... 40
  Interpretative phenomenological analysis ............................................................. 41

ETHICAL CONSIDERATIONS ................................................................................................. 42
Preface

In 2005, I started my employment with the deafblind counselling and support team of the Stockholm County Council. In my work as a clinical psychologist, I have had the opportunity to see many people with deafblindness and among them, people with Usher syndrome. Although my primary clinical experience is based on psychology, the support required by our clients almost always considers challenges that are best interpreted by a biopsychosocial framework and best met by an interdisciplinary team of competent professionals, of which I have had the opportunity to be a part.

I found that deafblindness and research on deafblindness in particular was a small field, and when I searched for evidence-based groundwork, there was none to find, aside from the clinical experiences of my colleagues. I realized that if there is to be any research done in this field, I could not wait for others to do the job. Therefore, when the opportunity of doing my PhD occurred, I took the chance.

During my PhD studies, I have had the opportunity to form and discuss the studies conducted in this thesis with a team consisting of passionate members representing different scientific backgrounds and with an openness to the approach, perspectives, and attitudes of members from other disciplines aimed towards interdisciplinary research. The mutual respect and striving to find a common language have been enhanced through the regular meetings of the research group and the composition of the supervisors, thus leading to what I believe is reciprocal learning (cf. Danermark, 2019).

The starting point for the first studies in this thesis was the collection of results found in cross-sectional studies of register data by Wahlqvist, Möller, Möller, & Danermark (2013; 2016b) which show in-group health differences that had not yet been explored. By an unconditional exploration of register data, my repetitions of analysis suddenly showed one variable that seemed to be of more importance than others, and my interest in further exploring work as a promoting health factor for persons with USH was raised.

What followed will be presented in the present thesis.
**Introduction**

This thesis will focus on health in relation to working life and the strategies of how people with Usher syndrome, a syndrome causing deafblindness, handle their life situation. First, in the introduction, Usher syndrome, deafblindness and disability will be described, followed by sections discussing health concepts and work and health, and finally, the relationship between work, health and disability.

**Usher syndrome**

Usher Syndrome (USH) is an autosomal recessive disorder that affects hearing, vision and, in some cases, vestibular function (Kimberling & Möller, 1995; Pennings, 2004; Sadeghi, Kimberling, Tranebjoerg, & Möller, 2004). To date, 13 genes have been identified as causing USH (Mathur & Yang, 2015), and there are three clinical types of the disorder (USH1-3) (Millan et al., 2011). There is a progression in vision loss and, in some cases, hearing loss (Bonnet & El-Amraoui, 2012), leading to a recurrent change in life conditions that requires adaptation. The process of adjustment can be complex as vision and hearing are highly compensatory senses in all environmental interactions and communication (C. Möller, 2003). In this project, two of the three clinical subgroups are in focus: people with USH1, who mainly use sign language in their communication, and people with USH2, who mainly communicate with spoken language. An overview of the differences between the three clinical groups is presented in table 1.

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
</table>
| USH1 | Profound congenital hearing loss  
Progressive vision loss due to retinitis pigmentosa  
Bilateral vestibular areflexia causing balance difficulties |
| USH2 | Congenital Moderate/severe hearing loss  
Progressive vision loss due to retinitis pigmentosa |
| USH3 | Congenital progressive hearing loss leading to profound deafness  
Progressive vision loss due to retinitis pigmentosa  
Progressive vestibular deficiency leading to balance problems |
Research has identified that persons with USH report more physical and psychological health problems and lower levels of social trust, as well as more difficult financial situations, compared to a reference population (Wahlqvist et al., 2013; 2016a; 2016b). Studies have also demonstrated high levels of stress (Högner, 2015), as well as depression and loneliness related to poor health-related quality of life (Dean, Orford, Staines, McGee, & Smith, 2017). Persons with USH express a lack of knowledge from society about living with a progressive combined vision and hearing loss/deafness due to USH (Ellis & Hodges, 2013; Evans, 2017). Although studies of people with USH have demonstrated a situation of poor health, research has also revealed a high in-group heterogeneity (Wahlqvist et al. 2013; 2016a; 2016b) that has not yet been explored. Furthermore, a scoping review has highlighted that although there are a few studies reporting poor health in people with USH, there is still a lack of studies on health and well-being and their relation to psychosocial determinants in people with USH (Arcous et al., 2019).

**Deafblindness**

The combination and severity of limitations in hearing and vision associated with USH leads to a dual sensory loss, causing functional limitations in line with what has been defined in the Nordic definition of deafblindness (NVC 2016). The Nordic definition states as follows:

“Deafblindness is a combined vision and hearing impairment of such severity that it is hard for the impaired senses to compensate for each other. Thus, deafblindness is a distinct disability. To varying degrees, deafblindness limits activities and restricts full participation in society. It affects social life, communication, access to information, orientation and the ability to move around freely and safely. To help compensate for the combined vision and hearing impairment, especially the tactile sense becomes important...” (Nordic Centre for Welfare and Social Issues, 2016).

The Nordic definition of deafblindness is based on the International Classification of Functioning, Disability and Health (ICF) (World Health Organization, 2001), a biopsychosocial model in which disability is defined as difficulties experienced by individuals interacting with a social and physical environment. Disability is defined by the WHO as including difficulties
with functioning, impairments in body structure and body function. These difficulties could be caused by injury or disease, but disability also includes a limitation in activities and restrictions in participation. Contextual factors, such as environment and personality, interact with these components (World Health Organization, 2001). In this thesis, the ICF will be used as a model for clarifying important concepts used in disability science and not used as a tool for classification.

The psychosocial situation of persons with deafblindness has been studied, showing psychological distress (Bodsworth, Clare, Simblett, & Deafblind UK, 2011) and stigmatization (Hersch, 2013). K. Möller (2008) states that the functional limitations in hearing and vision present in deafblindness lead to a vulnerable situation due to the difficulties related to accessing information and face-to-face interaction with other people. Isolation and social exclusion are common consequences, as are restrictions in terms of activity and an increased risk of physical harm (K. Möller, 2008). Living with deafblindness has been associated with ontological insecurity (Danermark & Möller, 2008), and people with deafblindness are often described in terms of their vulnerability rather than in terms of their resilience (Simcock, 2017). A scoping review has shown that the interaction of personal and environmental factors is of importance for the participation of people with deafblindness (Jaiswal, Aldersey, Wittich, Mirza, & Finlayson, 2018). The authors also stressed the need for studies focusing on the lived experiences of people with deafblindness to increase understanding, improve services, and enhance participation for this population (Jaiswal et al., 2018). Although there is a lack of studies focusing on people with deafblindness, studies of health in people with deafblindness are more common than those of specific deafblind diagnoses such as USH. Furthermore, studies of deafblindness show high heterogeneity within the group, and it is therefore important to focus on specific groups or the diagnoses of people with deafblindness. More specifically, there is a lack of studies on how people with USH handle their life situation, as well as studies that can cast light on differences in health found in people with USH.

**Disability**

As described above, the Nordic definition of deafblindness is in line with the definition of disability defined in the ICF (World Health Organization,
2001). Fifteen percent of the population in the world has some type of disability, and persons with disabilities have a higher risk of morbidity and early mortality than the general population both in low- and high-income countries, according to the World health organization (WHO) (2016). Persons with disabilities also have more health problems, lower education and lack employment, health care and rehabilitation (World Health Organization, 2011).

In public health studies, the term disability is often used as the outcome of poor health, while a group of persons with different impairment as default, in most studies referred to as having a disability, are merely viewed as a minority group that might experience health problems (McDonald & Raymaker, 2013). Research has, however, shown that having an impairment does not have to be directly related to perceived poor health, which is described as “the disability paradox” (Fellinghauer, Reinhardt, Stucki, & Bickenbach, 2012). According to the disability paradox, it is the limitations of a person’s activities and participation that affect their perceived health. Environment and personal factors interact and have a significant impact on how persons with impairments perceive their health and quality of life (Fellinghauer et al., 2012). According to the ICF definition, people with deafblindness and USH could be described in terms of having a disability, but this does not disclose any information about the individuals’ perceived health or well-being.

**Health**

The World Health Organization; WHO (1948) defines health as “a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity.” This definition is controversial, and recent definitions have emphasized that health is more complex, including individual factors, activity, participation and environmental factors (Bircher & Kuruvilla, 2014; Nordenfelt, 2007). In this doctoral project, the Meikirch model of health (Bircher & Kuruvilla, 2014) is adopted since it, as a framework, has the ability to include biopsychosocial aspects that are relevant in disability science. Bircher & Kuruvilla (2014) define health as follows:
“Health is a state of wellbeing emergent from conducive interactions between individuals’ potentials, life demands, and social and environmental determinants. Health results throughout the life course when individuals’ potentials and social and environmental determinants suffice to respond satisfactorily to the demands of life. Life demands can be physiological, psychological, or environmental and vary across individual and context, but in every case unsatisfactory responses lead to disease” (Bircher & Kuruvilla, 2014).

The Meikirch model of health not only reflects the complexity of health and well-being but also conceptualizes this complexity, emphasizing that health varies over time in a life span. Individuals’ acquired potentials and life demands are associated with the environment and society in meeting health needs (Bircher & Kuruvilla, 2014) In this thesis, the metaphor of a jigsaw puzzle will be used to clarify the different concepts of the Meikirch model, in which an interpretation of how the pieces are attached to each other will be made.

The Meikirch model jigsaw puzzle consists of four pieces (figure 1) with different shapes and colors. The pieces are social determinants (green), life demands (red) and two pieces (blue) that refer to the biologically given potentials and personally acquired potentials that together form an individual’s potential (blue + blue).
According to the Meikirch model (Bircher & Kuruvilla, 2014), as interpreted in jigsaw puzzle terms, health will occur when a person’s individual potential (blue) and social determinants (green) are sufficient to meet the life demands (red), and the pieces of the puzzle fit together to form a complete puzzle of health (figure 2).

Figure 2. Health occurs when individual potentials and social determinants suffice to life demands.

The pieces of the jigsaw puzzle might have different shapes and sizes, as long as they adapt and a complete puzzle of health occurs. However, if the pieces do not fit due to having a mismatched size or shape, then a situation of incongruence in life demands (red), social determinants (green) and individual potentials (blue) will occur, which is defined as ill health (Bircher & Kuruvilla, 2014).

The different concepts or pieces of the jigsaw puzzle included in the Meikirch model of health will be further described below and illustrated with examples from previous research in deafblindness or USH. The concept of environment (i.e., food, water, shelter, or climate) in the Meikirch model, which indeed affects peoples’ health, will not be further discussed in this thesis.
Social determinants
The green piece of the jigsaw puzzle.
The social determinants that interact with an individual in a society can be exemplified, such as social networks, health care, lifestyle, opportunities and conditions of education and employment, norms and attitudes or legislation. All of these are examples of processes that could be influenced by policy and have been shown to be important for health outcomes in public health studies (Dahlgren & Whitehead, 2017; Diderichsen et al., 2012). Studies have also shown that the relative difference in health among people in a society is often due to social inequalities and to understand health inequalities, the effect of different social determinants must be considered (Marmot, 2005).

In Relation to USH:
In a welfare state, the health of people with USH are affected by social determinants, such as norms, which are manifested by attitudes that affect how people with disability in general are seen/valued; an example is the risk of stigmatization (Hersch, 2013). Legislation can enable the right to specialized health care and rehabilitation and can promote education or employment in people with USH, as in the general population (Diderichsen et al., 2012). Ellis & Hodges (2013) describe the importance of specialized rehabilitation, sign-language interpreters, guides, supported employment or accessibility aids for well-being in people with USH. The importance of social networks such as friends and family has also been emphasized (Ellis & Hodges, 2013; Evans, 2017), and social support has been shown to be closely linked to perceived quality of life in people with USH (Dean et al., 2017).
Life demands
The red piece of the jigsaw puzzle.
Life demands can be physical, psychological or social (Bircher & Kuruvilla, 2014) and can vary between individuals and over time. In this thesis, one of the life demands, working life, will receive special attention. Despite this focus, in the following section, life demands will be illustrated with examples from research on USH and deafblindness from a broader perspective.

In relation to USH:
Most people with deafblindness, including those with USH, encounter situations that include demands relating to receiving information, face-to-face interaction or mobility/orientation, which are demands shown to be difficult to meet among people with deafblindness (K. Möller, 2008). Concrete examples embracing such demands in people with USH are evident in working life, education and leisure activities, as well as social or family life (Ellis & Hodges, 2013; Evans, 2017). However, work in terms of an important life demand for health has not been studied in relation to deafblindness or people with USH.

Individual potentials
The blue pieces of the jigsaw puzzle.
According to Bircher & Kuruvilla (2014), all people have a set of biologically given potentials; that is, biological or genetic variations that facilitate or restrict an individual’s potential. Examples are differences in sex, age, or physiological and psychological prerequisites, for example, sensory abilities, such as degree of hearing or vision. In addition to the biologically given potentials, people have a set of personally acquired potentials. Some of the acquired potentials can be defined as life strategies that a person develops when handling obstacles in life. In this thesis, personally acquired potentials in terms of life strategies have been partially interpreted terms of psychological flexibility/inflexibility, as described by Hayes, Strosahl and Wilson (2012); for details, see the next section. These acquired potentials can vary over time in the life span (Bircher & Kuruvilla, 2014).
In relation to USH:
People with USH all have an autosomal recessive inherited disorder causing Usher syndrome (Mathur & Yang, 2015). They have sensory limitations in terms of hearing loss or deafness and a progressive vision disorder (C. Möller, 2003). People with USH1 lack vestibular function. In terms of biologically given potentials, vision will deteriorate over time but demonstrate individual differences (Bonnet & El-Amraoui, 2012). The biologically given potential, in terms of genetics, hearing, and vision, has been extensively studied, as presented in a previous section of this thesis.

When using the Meikirch model of health to conceptualize the health of people with USH, it becomes clear that one piece of the jigsaw puzzle, namely, the biologically given potentials, has received much focus in research, while two of the other pieces of the health puzzle, namely, the personally acquired potentials and life demands, have received less attention. By extending studies and include more of the jigsaw puzzle pieces, based on the Meikirch model of (Bircher & Kuruvilla, 2014), a more comprehensive picture of the health of people with USH may be obtained.

In the following section, life strategies, in terms of acquired potential, will be presented, followed by a section discussing working life, which is interpreted as a life demand.
Life strategies - psychological flexibility as an acquired potential

As described above, the Meikirch model of health (Bircher & Kuruvilla, 2014) highlights that one factor affecting health is the individual’s potential. This potential is derived from the person’s biologically given and personally acquired potentials. The acquired potentials include aspects of the ability to handle or the life strategies related to handling the demands of life. One model that could be helpful in describing a person’s acquired potential in terms of life strategies is the model of psychological flexibility (Hayes et al., 2012). Psychological flexibility (PF) embraces a variety of human abilities that facilitate identifying and adapting to situational demands. According to the model, behavior or mind-sets need to shift when social or personal functioning is compromised. Balance in important life domains is maintained by awareness, openness and commitment to behaviors in line with deeply held values (Kashdan & Rottenberg, 2010). PF has been described as a process of being fully in contact with the present moment as a conscious human being and maintaining or changing behaviour in line with one’s own chosen values (Hayes et al., 2012). PF entails the following six core flexibility/inflexibility processes:

- **Cognitive defusio**: the ability to change one’s relation to unwanted thoughts;
- **Acceptance**: the ability to embrace private events without experiential avoidance;
- **Being present**: the ability to be in contact with the present moment;
- **Self as context**: the ability to experience events and to separate experiences from content;
- **Value**: qualities and desires that can be manifested in purposive action; and
- **Committed action**: an ongoing increase in effective behavior patterns towards chosen values (Hayes et al., 2012).

The six interrelated core processes form the two aspects of PF, namely, commitment-behavior activating processes and mindfulness-acceptance processes (Hayes et al., 2012).

There is a general lack of studies on life strategies of people with deafblindness (Jaiswal et al., 2018; Simcock, 2017). By using the model of psychological flexibility (Hayes et al., 2012), it is possible to develop an understanding of the life strategies of people with USH and thereby receive insights on individual acquired potentials in the health jigsaw puzzle of people with USH.
In the following section, work as an aspect of life demands (Bircher & Kuruvilla, 2014) will be presented.

**Work and health**

The relationship between work and health has been studied extensively in the general population, and working life has, in addition to the economy, been shown to be one of the most important arenas for meeting the psychological and social needs of people who are of working age (Blustein, 2008). The balance between work and leisure time is important, and among persons who reported a poor work-life balance, health problems were more common (Lunau, Bambra, Eikemo, van der Wel, & Dragano, 2014). Job strain, tiring working positions, and temporary job contracts were strongly associated with a higher likelihood of reported health problems (Bambra, Lunau, Van der Wel, Eikemo, & Dragano, 2014). Other factors, such as moderate working time and welfare state support, have been shown to be protective in maintaining a work-life balance (Lunau et al., 2014). The greatest health risks are, however, outside the labor market, where meta-analyses have shown that unemployment is a cause of poor health (Paul & Moser, 2009). Furthermore, research shows that unemployment is correlated with a higher risk of morbidity and mortality (Bambra & Eikemo, 2009; Dorling, 2009). A significant rise in suicide behavior has been found in countries where unemployment is increasing (Nordt, Warnke, Seifritz, & Kawohl, 2015; Stuckler, Basu, Suhrcke, Coutts, & McKee, 2009).

The information above shows that there are many studies on the relationship between work and health in the general population and that work constitutes an important piece of the health puzzle as an important life demand for health, according to the Meikirch model (Bircher & Kuruvilla, 2014). However, there is still a need for an overview of the relationship between work and health in people with disabilities, which will be presented in the following section.
Work, health and disability

The UN Convention on the Rights of Persons with Disabilities (Article 27) recognizes their right to work on an equal basis with others, which implies inclusion and accessibility (United Nations, 2006). It also states that governments should promote the realization of this right to work. The labor market is often a challenge for persons with impairments, and reports by the Organization for Economic Co-operation and Development (OECD) show that persons with impairments often rely on disability benefits (OECD, 2009a, 2009b). In Sweden, 12% of people who are of working age report having an impairment, and among them, two-thirds have a reduced work capacity (Statistics Sweden, 2019). Among people with impairments, only 60% are employed compared to 80% of the general population; while employment rates have increased during the last five years in the general population, the rate has remained at the same level among people with impairments (Statistics Sweden, 2019). A higher quality of life, a more positive attitude towards disability and a higher self-efficacy have been found in work-active people with impairments compared to those who were unemployed or retired (Martins, 2015).

The literature in the field of deafblindness and USH, including aspects of working life, is sparse (Ellis & Hodges, 2013; Högner, 2015), and besides a study from US showing an employment rate of 33 percent of people with dual sensory loss (Varadaraj et al., 2019), there are no studies on work in relation to health in people with USH or in any other group of people with deafblindness. There are, however, a few studies that focus on people with single sensory losses. Rydberg, Gellerstedt and Danermark (2010) have shown that persons with deafness who use sign language face great challenges on the labor market, including great differences between deaf persons and the general population regarding level of education, position in the labor market and disposable income (Rydberg et al., 2010). A review of the literature regarding the working life of people with hearing impairment who use oral communication has shown that high levels of stress and fatigue are common (Punch, 2016). Among people with visual impairment, the labor force rate is low and the unemployment rate is significantly higher compared to both the general population and persons with other impairments (Benoit, Jansson, Jansenberger, & Phillips, 2013; Sherrod, Vitale, Frick, & Ramulu, 2014). People with visual impairments not only report difficulties in finding and remaining employment but also experience prejudice and everyday disability-related assumptions by colleagues, employers and other
people in positions of power. Barriers such as negative attitudes among employers, problems finding support and difficulties in travelling to work were also reported. Despite these barriers, people with visual impairment show resilience in achieving goals related to work by obtaining and sustaining employment against all odds (French, 2017).

The situation of people with disabilities in general and single sensory losses in particular show that work for people with disabilities includes both challenges and opportunities. However, the work situation of people with deaf-blindness and USH has not yet been explored. More research on the work life and health of this group can cast light on a life demand that, for other groups of people, has been shown to be an important piece of the health puzzle.
Rationale

There is a lack of knowledge about the relationship between work and health in people with USH; although studies have explored health problems and have described people with USH as being vulnerable, there is still a lack of studies that examine the life strategies of this group and of people with deafblindness in general. Such knowledge is needed, first, to raise awareness about people with USH and their health in relation to their working life, and second, to contribute to the development of services for people with USH and the empowerment of people with USH through an increased knowledge of experiences about maintaining health through life strategies.
Aim

The general aim of the current thesis was to explore the relationship between work and health in people with USH1 and USH2, as well as the life strategies of people with USH2.

The study specific aims were as follows:

1. To explore the relationship between working and disability pension groups and physical and psychological health in persons with USH2.

2. To explore the relationship between health, work, and social determinants, such as social trust and financial situations, in persons with USH1 in comparison with a reference group.

3. To explore the life strategies of people with Usher syndrome type 2a.

4. To explore lived experiences with working life from the perspective of people with deafblindness due to Usher syndrome type 2.
Methods

The thesis has both a quantitative and qualitative explorative design. The quantitative studies consist of two retrospective cross-sectional studies based on register data from persons with USH1 and USH2. The first qualitative study is based on the content analysis of focus group interviews with people with USH2a, and the last study is designed as an interpretative phenomenological analysis of individual semi-structured interviews with people with USH2 to deepen the understanding of the first two studies. For an overview of the design, methods and study participants, see table 2.
Table 2. Overview of design, methods and study participants in studies 1-4.

<table>
<thead>
<tr>
<th></th>
<th>Study 1 (published)</th>
<th>Study 2 (published)</th>
<th>Study 3 (published)</th>
<th>Study 4 (manuscript)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Design</strong></td>
<td>Quantitative explorative</td>
<td>Quantitative explorative</td>
<td>Qualitative explorative</td>
<td>Qualitative explorative</td>
</tr>
<tr>
<td><strong>Aim</strong></td>
<td>To explore the relationship between working and disability pension groups and physical and psychological health in persons with USH2</td>
<td>To explore the relation between health, work, and social determinants such as social trust and financial situation in persons with USH1 in comparison with a reference group</td>
<td>To explore life strategies in people with Usher syndrome type 2a</td>
<td>To explore lived experiences with working life from the perspective of people with deafblindness, due to Usher syndrome type 2</td>
</tr>
<tr>
<td><strong>Participants</strong></td>
<td>67</td>
<td>47</td>
<td>14</td>
<td>7</td>
</tr>
<tr>
<td><strong>Mean Age, years (range)</strong></td>
<td>48 (18-65)</td>
<td>44 (20-64)</td>
<td>41 (21-65)</td>
<td>44 (38-50)</td>
</tr>
<tr>
<td><strong>Women, %</strong></td>
<td>53</td>
<td>53</td>
<td>29</td>
<td>57</td>
</tr>
<tr>
<td><strong>Data collection</strong></td>
<td>Register data</td>
<td>Register data</td>
<td>Focus group interviews</td>
<td>Individual interviews</td>
</tr>
<tr>
<td><strong>Data analysis</strong></td>
<td>Non-parametric chi-2 significance testing</td>
<td>Multiple logistic regression analysis</td>
<td>Qualitative inductive content analysis</td>
<td>Interpretative phenomenological analysis</td>
</tr>
</tbody>
</table>
Usher register

The participants in the studies were all registered in the Swedish Usher register, located at the Audiological Research Center, Örebro University Hospital. The register comprises data from approximately 400 persons with USH types 1-3. It was initialized in 1987 and comprises records related to ancestry, genetic diagnoses, data on hearing, vision, and vestibular function, and other relevant medical records. The register has been updated regularly and contains approximately 40% of the known USH population in Sweden. The register comprises data on individuals with a clinical form of USH types 1-3, and approximately 60% of these individuals also have a genetically confirmed diagnosis of USH. The proportions in the register between the different types are USH1 at 40%, USH2 at 45% and USH3 at 15%. Self-assessed data from surveys covering physical and psychological health, lifestyle factors and living conditions have been added to the register (Wahlqvist et al. 2013; 2016a; 2016b), which will be described in more detail below. All records in the register were collected after authorization by the individuals in the register. In studies 1-3, hearing and vision assessments and clinical or genetic diagnoses were retrieved from the register. No medical data on hearing or vision from the register was used in study 4.

Hearing

The audiological data were obtained from audiograms in the Usher register. The audiograms were collected serially over 30 years. The audiometric data used were retrieved as close as possible to the time of the studies. The hearing was assessed by standardized pure-tone audiometry (0.5-8 kHz) (PTA) with a calculation of the pure tone average for the frequencies 0.5, 1, 2, and 4 kHz (PTA4). Thresholds in the register were classified according to European standards (Stephens, 2001). Thresholds were classified as mild (20-40 dB), moderate (40-70 dB), severe (70-95 dB) or profound (>95 dB). Data on the participants’ hearing are presented in table 3.

Vision

Visual data were retrieved from the USH register (visual acuity and visual field limitations). As with the audiological data, the visual data used were retrieved as close as possible to the studies. The best corrected visual acuity (VA) (the ability to discriminate details) was measured by Snellen chart-based standard tests, given in decimals, where 1.0 indicates normal VA and
0.05 indicates a severely reduced VA (in the US, VA <=0.1 is defined as legal blindness).

Visual field (VF) tests were performed by Goldmann perimetry and categorized into five phenotypes (1–5): category 1, normal visual field; category 2, presence of a partial or complete ring scotoma (the latter either extending or not extending into the periphery); category 3, concentric central field loss with a remaining peripheral island less than one-half of the field circumference; category 4, marked concentric loss <10 degrees; and category 5, no visual field at all (blind) (Grover, Fishman, Anderson, Alexander, & Derlacki, 1997). Data on visual acuity and visual field are presented in table 3.

Health survey
The Health on Equal Terms questionnaire (HET) was sent to people with USH2 and USH1 in 2008 and 2012, respectively. All 122 people with USH2 aged 18-84 years registered in the Usher register were invited to answer the HET questionnaire in 2008, and in 2012, all 87 people with USH1 aged 18-84 years were invited. The data collection procedures are presented in detail in Wahlqvist et al. (2013; 2016b). To increase the accessibility of the questionnaire, the second data collection contained translations in sign-language, and the questionnaire was available in large print (Wahlqvist et al., 2016b).

The HET questionnaire has regularly been conducted by the Public Health Agency on a random sample of the Swedish population since 2004. The survey comprises 75 questions on physical and psychological health, experiences of health care, living habits, social relations, participation in different activities, working life, and alcohol and drug use (Boström & Nyqvist, 2010). The HET questionnaire is a synthesis of different commonly used health questionnaires in Sweden. It has been revised and evaluated to obtain a high construct validity and good metric capacity to discriminate between variables when used in the Swedish population (Boström & Nyqvist, 2010).
Participants

Study 1
In study 1, all 67 persons with USH2 of working age (18-65 years) who responded to the HET survey were included in this study. A total of 34 were working (working group), and 33 had a full (100%) disability pension (non-working group). The background data of the participants are presented in table 3.

Study 2
In study 2, all 47 persons with USH1 of working age (18-65 years) who had responded to the HET survey in the year 2012 were included in the study. Of the responders, 23 were work active (working group), and 24 were not work active, had a 100% disability pension, sick leave or unemployment (nonworking group). The background data of the participants are presented in table 3. To compare the data from people with USH1 with a random sample from a reference population, reference group data were retrieved from an external database administered by the Public Health Agency of Sweden. The data from the reference population included 3247 individuals aged 18-65 years, of whom 3049 were working and 198 were nonworking.

Study 3
From the 58 people of all ages with a genetically confirmed diagnosis of USH2a who had registered in the Usher register as of 2013, a purposeful sample was made, striving to achieve heterogeneity in terms of age and sex, as well as geographic distribution. Thirty-two persons of working age (18-65 years) and capable of participating in a focus group using verbal communication were invited to participate in study 3. Of the 32 persons invited, 14 agreed to participate, 5 declined, 1 person initially accepted but was unable to attend, and 12 persons did not respond to the invitation. All 14 individuals (4 women and 10 men) who agreed to participate were included in the study. All participants travelled to participate in the interviews, which were conducted at an audiological research center in Sweden. The background data of the participants are presented in table 3.
Study 4
In 2018, the Usher register included 170 persons of all ages with a diagnosis of USH2. A purposeful sample of 14 participants was made, striving towards homogeneity, where all participants had a clinical diagnose of USH2, were aged 35-50 years and had a level of hearing loss where they were able to use verbal communication in an interview setting. Participants who were work active (50% or more) for the last three years were invited to participate in the study. The inclusion criteria were intentionally narrow due to the small sample size, which is common in an interpretative phenomenological analysis (IPA) (Smith, Flowers, & Larkin, 2009). A total of eight persons agreed to participate. One person did not meet the inclusion criteria of work activity. All of the other seven participants, four women and three men, were included in the study. Background data on age, self-reported education, work activity and work-related support were collected prior to individual interviews. The background data are presented in table 3.
Table 3. Background data of participants study 1-4.

<table>
<thead>
<tr>
<th></th>
<th>Study 1 (USH2)</th>
<th>Study 2 (USH1)</th>
<th>Study 3 (USH2A)</th>
<th>Study 4 (USH2)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Work activity</td>
<td>Working</td>
<td>Non-working</td>
<td>Working</td>
<td>Non-working</td>
</tr>
<tr>
<td>N</td>
<td>34</td>
<td>33</td>
<td>23</td>
<td>24</td>
</tr>
<tr>
<td>Women, n (%)</td>
<td>18 (53)</td>
<td>18 (54)</td>
<td>10 (43)</td>
<td>15 (63)</td>
</tr>
<tr>
<td>Age mean (year)</td>
<td>44</td>
<td>52</td>
<td>37</td>
<td>50</td>
</tr>
<tr>
<td>Hearing loss diagnosis age, year mean (SD)</td>
<td>4 (2)</td>
<td>5 (2)</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>Vision loss diagnosis age, year mean (SD)</td>
<td>25 (11)</td>
<td>25 (10)</td>
<td>15 (9)</td>
<td>10 (4)</td>
</tr>
<tr>
<td>Hearing loss PTA4 mean (SD)</td>
<td>72 dB (12 dB)</td>
<td>71 dB (15 dB)</td>
<td>Profound deaf</td>
<td>Profound deaf</td>
</tr>
<tr>
<td>Visual field median (category)</td>
<td>&lt;10° (4)</td>
<td>&lt;10° (4)</td>
<td>10-30° (3)</td>
<td>10-30° (3)</td>
</tr>
<tr>
<td>Best corrected visual acuity mean (SD)</td>
<td>0.3 (0.2)</td>
<td>0.4 (0.3)</td>
<td>0.6 (0.3)</td>
<td>0.5 (0.3)</td>
</tr>
</tbody>
</table>
Data collection

The Health on Equal Terms questionnaire

In studies 1 and 2, HET survey data collected in previous projects (Wahlqvist et al., 2013; 2016b) were retrieved from the Usher register.

In study 1, the answers from all 36 questions concerning physical and psychological health were used. These questions were chosen because previous results have indicated significantly poorer health in the USH2 group compared to the reference population (Wahlqvist et al., 2013). The questions used encompassed health (general health, number of days with bad physical or psychological health, and number of days with reduced activities of daily living). Some of the questions were graded on a four- or five-point scale, while the questions about diabetes, asthma, allergy, high blood pressure, shoulder or neck pain, headache, and fatigue had dichotomized answers. Psychological health was collected by 12 questions concerning the ability to concentrate, feelings of worthlessness, and the inability to appreciate the day as experienced over the course of the previous two weeks. Two questions pertaining to suicidal behavior were also included.

In study 2, 12 questions concerning physical and psychological health, social trust and financial situation were used. These questions were chosen because previous studies (Wahlqvist et al., 2016b) have indicated significant differences, not only in psychological and physical health, as examined in study 1, but also regarding financial situations and levels of social trust between people with USH1 compared to a reference population. The selected health questions focused on the number of days with poor physical or psychological health and the number of days with reduced activities of daily living (ADL) in a 30-day period. Two questions pertaining to suicidal behavior (suicide thoughts and suicide attempts) were included, as well as five questions about social trust indicators (refraining from going out alone, not receiving help when needed, no general trust in most people, having no one to share innermost thoughts with, being offended) and two questions about the respondent’s financial situation (economic difficulties and not having a cash margin of 15,000 SEK for an unforeseen situation). The answers of the variables were graded on a three- or four-point scale, and the number of days ranged from 0-30.
Focus group interviews
In study 3, focus group interviews were used to elicit data. In focus groups, participants are encouraged to interact, talk, comment on statements and ask questions to obtain a range of perceptions (Kitzinger, 1994). The 14 participants (4 women and 10 men) diagnosed with USH2a were divided into three focus groups: women (n=4), younger men (n=5) and older men (n=5), where the limited size of the groups facilitated functioning communication. The following questions were posed based on the aim of the study to start the focus group discussion: “How do you cope with your life situation and how do you manage difficulties? If you were not successful in dealing with a situation, in what way would you have liked to do things?” Thereafter, the moderator helped the participants take turns and verbalized nonverbal communication. The interviews were moderated by a group leader with the support of an experienced researcher. The interviews were held in the oral Swedish language and lasted for approximately two hours with a short break. The interviews were filmed and audio recorded. To optimize communication, a hearing loop was used, and light adjustments and window blinds were used.

Individual interviews
Individual interviews were used in study 4. The individual interviews enabled the participants to share their lived experiences in their own terms. In accordance with the literature (Smith et al., 2009), the focus was on the participant’s involvement, understanding and attempts to find a sense of meaning. The interview schedule included open-ended questions based on themes of importance for the research purpose, such as description of work tasks and questions about the participant’s experiences of health in relation to their working life. The interview schedule served as a map for the focus of the interview, where the aim was to attain information about the lived experiences related to working life and health both on a descriptive and analytic level. Probes were posed for clarification and encouragement towards more analytic reasoning. The interviewer had the ability to go off of the schedule when the interviewee presented other themes relevant and related to the purpose of the study, in line with the literature (Smith et al., 2009).

All interviews were conducted by the author of the thesis. The interviews were held in the oral Swedish language and lasted for approximately 1-1,5
hours. Three interviews were conducted at the workplace of the interviewee, two were conducted at different counselling and support teams for people with deafblindness, one was conducted in the home of the interviewee, and one was conducted at a research center. All interviews were audio recorded. Some of the participants used a hearing loop to improve the sound quality in their hearing aids. Locality, when selected by the interviewer, was selected to reduce noise and dazzle and equipped to enable proper illumination.

Data analysis

Statistics
In studies 1 and 2, the responses to the items from the questionnaire pertaining to physical and psychological health, social trust, and financial situation were all dichotomized as “Problem” and “No problem” in accordance with Boström et al. (2010). All “Yes” responses on the three- and four-point scales were coded as “Problem”, and all “No” responses were coded as “No problem”. The question concerning “the number of poor health days” was also dichotomized in accordance with Boström & Nykvist (2010), where 15 or more days was considered poor physical or psychological health. To compare groups of binomial data, i.e., a nominal scale level, nonparametric statistical analysis was employed (Field, 2013). Pearson’s chi-square testing was performed in study 1 to compare the answers to the health questions between the two groups (USH2 working group and USH2 non-working group), and a significance level of p <0.05 was used.

In study 2, multiple regression models were performed to analyze the differences between the four separate groups with different sample sizes of binomial data (Field, 2013). The approach enables the exploration of the association and odds ratio (OR) between health variables (psychological, physical, social trust and financial situation) and the groups of persons with USH and the two reference population groups while controlling for age and gender. A series of multiple logistic regression models were examined to explore the association between the variables of (1) health, (2) social trust, (3) financial situation, and the groups of (1) USH nonwork, (2) USH1 work, (3) reference nonwork, and (4) reference work, while controlling for age and gender. The models were evaluated by OR to examine the odds related to the groups of USH1 nonwork, USH1 work, reference nonwork and ref-
ference work, where the reference work group served as the reference category in the analyses (had a value of 1). The differences between the groups are described in terms of risk, i.e., OR, where a value >1 indicates an increased risk. Furthermore, the increased risk is presented with a corresponding 95% confidence intervals (95% CI).

The sensitivity at 90% specificity shows how capable the model is of identifying individuals with experience of the dependent variable, while at the same time only incorrectly classifying one out of ten individuals without experience of the dependent variable as having such experience. The area under the curve (AUC) was used to examine the overall strength of the models. An AUC value of .60 or higher may be considered relevant, while a value of .80 in general indicates a well-discriminating model (Steyerberg et al., 2010). The IBM SPSS 22 statistical program was used for all computer analyses (studies 1 and 2).

**Qualitative content analysis**

In study 3, the focus group interviews were analyzed by qualitative content analysis with the aim of understanding the meaning of the interview text and gaining new insights into the studied phenomenon. In line with the literature (Krippendorff, 2012), the analysis was inductive due to the exploratory design of the study.

All recordings were transcribed verbatim, and the transcripts were compared to the recordings. To obtain a sense of the whole, the recordings and transcripts were listened to and read several times. Meaning units were identified, condensed and labeled with a code (Graneheim & Lundman, 2004). A comparison of the codes was made to find similarities and differences, after which they were abstracted to manifest categories and subcategories. All categories were evaluated in a reflective interpretative process, where all four authors were active in moving back and forth between the meaning units, codes and categories. The process continued until a consensus was obtained. In addition to the manifest categories, a more abstract, latent, thematic content running through the entire dataset was found, resulting in two subthemes and one theme (Graneheim & Lundman, 2004).
**Interpretative phenomenological analysis**

In study 4, the individual interviews were analyzed by interpretative phenomenological analysis with the aim of understanding the participants' lived experiences and interpreting their understanding and attempts to make sense of their experiences (Smith et al., 2009). Recordings from interviews were transcribed verbatim. Each interview was first analyzed separately, divided into general and more specific detailed parts where the authors separately made exploratory comments on a descriptive level. Thereafter, a more interpretative analysis followed, where the authors, who have different theoretical backgrounds, added personal experiences and understanding to the notes. In this iterative process of the particular and the general in the interpretation led to an abstraction of emerging themes at the individual level. The interpreted themes were organized in temporal order, then clustered and analyzed, leading to superordinate themes. The authors compared their analyses or critically reviewed the analysis performed by the first author to reach a common interpretation on each case. The same process was followed for each interview, and finally, the themes from all interviews showed patterns with higher order themes in accordance with Smith et al. (2009). The table of superordinate themes, themes and concrete transcription finally constituted a base for the presentation of the results.
Ethical Considerations

Research on small groups is important but requires special consideration in terms of balancing sometimes contradictory values. Research can be beneficial for a group, the participants of the study or society but also involves risks related to loss of anonymity, problems with accessibility or exhaustion and the fact that participants find it difficult to decline loyalty to the group or researchers. Taking part in research can be demanding, and it is of utmost importance that studies conducted are identified as being of importance from the perspective of both the research and responders (Vetenskapsrådet, 2017).

USH is a rare diagnosis, and therefore, extra consideration of confidentiality is necessary. The recruitment of participants from different parts of Sweden reduced the risk of identification, and data that could disclose identities (names, places) were removed from the result presentations (Vetenskapsrådet, 2002).

The limitation in hearing and vision among participants required special adjustments of informed consent. All printed information and forms regarding informed consent were printed in large print, where participants were informed about the studies, that their participation was voluntary and that they had the right to withdraw from the study at any time without giving reasons. Prior to the interview studies (studies 3 and 4), the information and consent were also repeated verbally. All participants signed an informed consent form.

Hearing and vision difficulties can make participation in research demanding. Extensive questionnaires and interviews must be arranged to ensure accessibility to facilitate participation. In study 2, the questionnaire was adjusted in font and translated to sign-language and filmed to increase accessibility. In all interview settings, special focus was given to illumination and dazzle and to increasing the ability to use hearing aids with a hearing loop. Moreover, in the focus group interviews (study 3), all of the participants were offered the opportunity to be assisted by a specific person, guide or interpreter. One of the participants had an interpreter and another chose to have a family member on hand.
The studies conformed to the criteria of the Declaration of Helsinki for medical research in human subjects. All people in the USH register signed informed consent forms that their data can be used in clinical and genetic research on Usher syndrome, as approved by the Ethics Committee of Linköping University Hospital (1990, 1997). The studies were approved by the Regional Ethics Committee in Uppsala (Nr. 2012/515, 2012/515/2).
Results

Study 1
The aim of this study was to explore the relationship between the working and disability pension groups and physical and psychological health in persons with USH2.

There were no differences in terms of gender, degree of hearing loss, visual acuity, visual field loss or age of diagnosis between the two groups (USH2 working group, n = 34; USH2 non-working group, n = 33). There was a difference in mean age between the USH2 working group (44 years) and the USH2 non-working group (52 years). The participants in the USH2 working group reported significantly better general psychological health compared to the USH2 non-working group. Depression, suicidal thoughts, and suicide attempts were significantly more common among persons in the USH2 non-working group. Loss of faith in oneself, feelings of worthlessness, unhappiness, and inability to handle problems, as well as problems with sleep and concentration, were reported to a significantly higher extent in the USH2 non-working group. No significant differences were found between the

Figure 3. Psychological health in the USH2 working group and the USH2 non-working group (%); * = significant difference, p < 0.05.
groups in terms of the ability to make decisions, of feeling tense or in reported anxiety levels (Figure 3).

However, both the USH2 working and non-working groups reported high levels of physical health problems comprising headache, stress, fatigue, allergy, asthma, and high blood pressure. The results revealed that there were no significant differences between the two groups concerning pain in the neck, shoulders, or hands, eczema, bowel problems, and diabetes. The USH2 nonworking group reported overweight and a decreased ability to run a short distance, as well as incontinence, significantly more often than did the working group (see Figure 4).

![Figure 4. Physical health among the USH2 working group and the USH2 non-working group (%); * = significant difference, p < 0.05.](image)

**Study 2**

The aim of the study was to explore the relationship between health, work, and social determinants, such as social trust and financial situation, in persons with USH1 in comparison with a reference group.
There was no difference in best corrected visual acuity, visual field or age of diagnosis between the USH1 working \((n = 23)\) and nonworking groups \((n = 24)\). There was a difference in the mean age between the USH1 working group \((37\) years\) and the USH1 nonworking group \((50\) years\). There were more women in the USH1 nonworking group \((63\%)\) compared to in the USH1 working group \((43\%)\). The results presented from the multiple logistical regression model were controlled for differences in age and gender while analyzing the association of the USH1 nonworking, USH1 working, reference nonworking, and reference working groups, including the variables of health, social trust indicators, and financial situation.

For physical and psychological health problems, the non-work groups had the strongest association (largest ORs). For suicide attempts, the USH1 groups showed higher associations compared with the reference groups. Overall, the presence of ADL problems was most strongly related to the nonwork groups. The ADL problems variable also showed the highest area under the curve (AUC), which is a measurement used to examine the strength of the models. An AUC value of .60 or higher may be considered relevant, while a higher value of 0.80 in general indicates a well-discriminating model.

In the social trust variables, refraining from going out alone was most strongly associated with the USH1 group. Not receiving help when needed was mainly a concern for the USH1 nonworking group but was followed by the USH1 working group and the reference nonworking group. The same pattern of association appears for the variable of having no one to share innermost feelings with. For the variable of having no general trust in most people, both the USH and reference nonworking groups had larger ORs than the working groups.

For economic difficulties, the reference nonworking group was associated with an increased OR, while there was no such significant association for any of the USH1 groups. However, the OR for a poor cash margin of 15,000 SEK in the case of an unforeseen situation was larger in all the groups compared to the reference working group, with the USH1 nonworking group having the highest OR.
The multiple logistic regression analysis does not demonstrate any individual response patterns within each group. A series of cross-tabs was employed among the different health, social trust and financial situation variables to receive an overview of the individual response patterns. The small sample did not permit any statistical analysis of response patterns, but the results showed that some of the persons who reported having problems related to one health variable often seemed to be the same individual reporting problems related to another health variable. The same pattern was shown among the social trust and financial situation variables. Within the USH1 nonworking group, a few persons reported having problems in most of the health, social trust and financial situation variables.
Table 4. Multiple logistic regression in study. Multiple regression models of the association between USH1 work, USH1 nonwork, reference nonwork, and reference work groups with health variables, social trust variables and financial situation variables.

<table>
<thead>
<tr>
<th>Health variables</th>
<th>Dependent variable</th>
<th>USH1 Non-working group</th>
<th>USH1 Working group</th>
<th>Ref Non-working group</th>
<th>Ref Working group</th>
<th>Sensitivity (at 90% specificity)</th>
<th>AUC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Health variables</td>
<td>Psychological health problem</td>
<td>3.86 (1.56-9.55)</td>
<td>1.82 (0.61-5.47)</td>
<td>2.81 (2.00-3.97)</td>
<td>1</td>
<td>0.19 (0.15-0.35)</td>
<td>0.63 (0.60-0.66)</td>
</tr>
<tr>
<td>Health variables</td>
<td>Suicide thoughts</td>
<td>3.60 (1.30-9.95)</td>
<td>5.54 (2.30-13.35)</td>
<td>2.29 (1.56-3.38)</td>
<td>1</td>
<td>0.22 (0.18-0.40)</td>
<td>0.63 (0.60-0.67)</td>
</tr>
<tr>
<td>Health variables</td>
<td>Suicide attempts</td>
<td>15.23 (5.31-43.69)</td>
<td>4.59 (1.04-20.24)</td>
<td>1.36 (0.58-3.19)</td>
<td>1</td>
<td>0.24 (0.16-0.42)</td>
<td>0.66 (0.60-0.72)</td>
</tr>
<tr>
<td>Health variables</td>
<td>Physical health problems</td>
<td>6.93 (2.92-16.45)</td>
<td>1.82 (0.53-6.24)</td>
<td>3.47 (2.44-4.92)</td>
<td>1</td>
<td>0.22 (0.18-0.40)</td>
<td>0.59 (0.55-0.62)</td>
</tr>
<tr>
<td>Health variables</td>
<td>ADL problems</td>
<td>22.99 (9.37-56.45)</td>
<td>4.69 (1.36-16.19)</td>
<td>10.38 (7.09-15.20)</td>
<td>1</td>
<td>0.43 (0.36-0.79)</td>
<td>0.72 (0.68-0.77)</td>
</tr>
<tr>
<td>Social trust variables</td>
<td>Refraining from going out alone</td>
<td>7.79 (2.89-20.97)</td>
<td>5.19 (2.02-13.36)</td>
<td>0.98 (0.69-1.39)</td>
<td>1</td>
<td>0.24 (0.21-0.44)</td>
<td>0.73 (0.71-0.75)</td>
</tr>
<tr>
<td>Social trust variables</td>
<td>Not receiving help when needed</td>
<td>16.48 (6.72-40.42)</td>
<td>4.64 (1.34-16.07)</td>
<td>3.67 (2.20-6.12)</td>
<td>1</td>
<td>0.28 (0.21-0.50)</td>
<td>0.65 (0.60-0.70)</td>
</tr>
<tr>
<td>Social trust variables</td>
<td>No general trust in most people</td>
<td>4.98 (2.12-11.71)</td>
<td>1.11 (0.43-2.90)</td>
<td>1.82 (1.33-2.48)</td>
<td>1</td>
<td>0.21 (0.18-0.39)</td>
<td>0.62 (0.60-0.64)</td>
</tr>
<tr>
<td>Social trust variables</td>
<td>Having no one to share innermost thoughts with</td>
<td>8.55 (3.44-21.24)</td>
<td>4.31 (1.72-10.82)</td>
<td>2.96 (2.04-4.29)</td>
<td>1</td>
<td>0.20 (0.16-0.37)</td>
<td>0.63 (0.60-0.66)</td>
</tr>
<tr>
<td>Financial situation</td>
<td>Being offended</td>
<td>0.77 (0.22-2.61)</td>
<td>3.04 (1.31-7.06)</td>
<td>1.62 (1.17-2.26)</td>
<td>1</td>
<td>0.21 (0.18-0.39)</td>
<td>0.63 (0.61-0.65)</td>
</tr>
<tr>
<td>Financial situation</td>
<td>Economic difficulties</td>
<td>2.30 (0.84-6.31)</td>
<td>1.14 (0.38-3.41)</td>
<td>2.50 (1.79-3.50)</td>
<td>1</td>
<td>0.19 (0.16-0.36)</td>
<td>0.66 (0.63-0.68)</td>
</tr>
<tr>
<td>Financial situation</td>
<td>Cash margin, unforeseen situation, 15,000 SEK</td>
<td>10.80 (4.59-25.40)</td>
<td>2.83 (1.16-6.89)</td>
<td>3.22 (2.33-4.44)</td>
<td>1</td>
<td>0.28 (0.24-0.52)</td>
<td>0.70 (0.67-0.72)</td>
</tr>
</tbody>
</table>
Study 3
The aim of the study was to explore life strategies in people with Usher syndrome type 2a.

The content analysis resulted in seven manifest categories based on 17 subcategories. In the process of working with the manifest categories, the overarching theme of “being at the helm” emerged, which encompasses the participants’ active, cognitive and emotional striving to be the person in control of their important life domains. The theme was divided into two subthemes: resolving or preventing challenges and comforting oneself. For an overview of the results, see table 5.

Table 5. Overview of the theme, subthemes, categories and subcategories in study 3.

<table>
<thead>
<tr>
<th>Theme</th>
<th>Sub-themes</th>
<th>Categories</th>
<th>Sub-categories</th>
</tr>
</thead>
<tbody>
<tr>
<td>Being at the helm</td>
<td>Resolving or preventing challenges</td>
<td>Remaining active</td>
<td>Business as usual</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Adapting activities</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Using memory and attention</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Using devices</td>
<td>Accessibility aids</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Everyday tools</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Using support</td>
<td>Formal support</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Informal support</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sharing knowledge</td>
<td>Informing in everyday situation</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Educating professionals</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Appreciating the present</td>
<td>Seize the moment</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Enjoy meaningful activities</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Maintaining a positive image</td>
<td>Negotiate who I am</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Stand up for myself</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Boost self-confidence</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Alleviating emotional pain</td>
<td>Self-distance</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Escapism</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Hope</td>
</tr>
</tbody>
</table>
The subtheme of resolving or preventing challenges was formed from the first four categories, reflecting the participants’ ability to manage the practical aspects of their life situation. To remain active by acting in accordance with business as usual and doing things the same way as always was mainly described by younger participants. Both younger and senior participants, however, reported adapting their activities to remain active. They used memory and attention to compensate for sensory limitations. When using devices, the participants were inventive in handling everyday tools in a way that compensated for their limitations in hearing and vision. They also used specialized accessibility aids, although the use of aids was sometimes restricted due to the perceived risk of stigmatization. Using support was a journey in which the participants gradually had to handle the need for informal support by family and friends. They also benefitted from formal support, both in terms of health care and personal support. The use of support helped maintain equality in their relations. Sharing knowledge about deafblindness was required, both in everyday situations and by educating professionals, including experiences related to needing to educate the officers who were the ones who should give support.

The subtheme of comforting oneself was formed from the three latter categories representing different aspects that reflect the emotional aspects of the participants’ struggle to manage their life situation. Being able to appreciate the present, to seize the moment and to enjoy meaningful activities, rather than focusing on the uncertain future, was considered helpful by the participants. The participants strived to maintain a positive image of themselves. The subcategory of negotiating who I am included a gradual change of identity where the deafblindness was accepted one step at a time. To be able to stand up for oneself, raising one’s voice was deemed important. However, sometimes a boosted self-confidence by way of external attributes was helpful in keeping up a positive self-image. To alleviate emotional pain, the participants found that strategies of self-distancing and being able to use humor were ways to distance themselves from situations where mobility or communication failed. Escapism was another strategy that, at least in a short-term perspective, helped them handle difficult emotions. To have hope in what the future might bring, in terms of a possible cure or being confident of one’s capacity to handle uncertainty in the future was helpful for the participants. One participant said:
“One important thing that becomes clear here [in the discussion] is that if you manage to do something, whatever it is, it gives you a sense of satisfaction, no matter how nice or unattractive the result turns out. It sorts of refreshes me.” (person 10)

**Study 4**

This study aimed to explore lived experiences with working life from the perspective of people with deafblindness due to Usher syndrome type 2.

The interpretative phenomenological analysis of the seven individual interviews yielded four superordinate themes derived from 11 themes. For an overview, see table 6. A brief summary of the superordinate themes and themes, illustrated by a quotation, will be presented below.

*Table 6. Superordinate themes and themes in study 4.*

<table>
<thead>
<tr>
<th>Superordinate themes</th>
<th>Themes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Feelings of satisfaction</td>
<td>Feeling a sense of belonging</td>
</tr>
<tr>
<td></td>
<td>Feeling recognized</td>
</tr>
<tr>
<td></td>
<td>Feeling content with own competence</td>
</tr>
<tr>
<td>A commitment that needs balancing</td>
<td>Taking on responsibility for recovery</td>
</tr>
<tr>
<td></td>
<td>Accepting the need for adaptations</td>
</tr>
<tr>
<td></td>
<td>Prioritizing work tasks</td>
</tr>
<tr>
<td></td>
<td>Making every effort</td>
</tr>
<tr>
<td>Facing limitations</td>
<td>Feelings of exhaustion</td>
</tr>
<tr>
<td></td>
<td>Feelings of insufficiency</td>
</tr>
<tr>
<td>Feelings of uncertainty</td>
<td>Feeling insecurity on a daily basis</td>
</tr>
<tr>
<td></td>
<td>Perceiving the future as unpredictable</td>
</tr>
</tbody>
</table>

**Feelings of satisfaction**

The sense of belonging at the workplace, feelings of being appreciated and recognized as an important and competent work force offered the participants a deep feeling of satisfaction. This feeling, at times, compensated for the difficulties caused by deafblindness, it increased self-esteem, and it served as an inner motivation to continue striving to maintain their working life.
A commitment that needs balancing

Work had a physical, cognitive and emotional expense, which was related to deafblindness and could be a health hazard. By taking responsibility for recovery, accepting the need for adaptations, prioritizing work tasks and making a great effort, the participants strived to find a balance between work and health.

Facing limitations

The demands of working life included experiences in which the restrictions in hearing and vision due to deafblindness were no longer possible to neglect or compensate for. Despite the satisfying work or high commitment, there were experiences of having to suddenly or gradually face one’s own limitations. This could result in feelings of insufficiency or feelings of exhaustion. Being forced to face one’s limitations was associated with negative thoughts and emotions, lowered self-esteem and the motivation to continue work. It was therefore common to try to neglect or avoid such thoughts for as long as possible. However, finally accepting the limitations and stopping fighting them, could produce feelings of relief.

Feelings of uncertainty

Working life was not only a source of satisfaction, but the restrictions in vision and hearing with a constant progression led to feelings of uncertainty both on a daily base and for the unknown, as respondents often feared their unpredictable future. Not being able to foresee what will happen or feeling insecure in one’s daily working life threatened stability and security, and when working life as a platform for satisfaction, commitment and a positive self-image was put at risk, the uncertainty could reach an existential dimension.

One person in the study concluded her experiences as follows:

“If I didn’t have a job, then I would feel really, really bad. It is fantastic to have a job and colleagues. An everyday place, which is nice. Then, it feels really stupid, to feel that you can’t master the situation completely, because I have an enormous sense of duty and want to do right. So, this has been the greatest burden through the years, to have the feeling that I can’t do what I want and need to do. That’s why I have worked extra hard. Mmm, yes...so... How do I feel today, when I have diminished this, and I really can do what I have to do? Pretty good. Right now, I am feeling good. “(P5)
Discussion

The general aim of the thesis was to explore the relationship between work and health in people with USH1 and USH2, as well as the life strategies of people with USH2.

The results showed a poorer psychological health in people with USH1 and USH2 who were not work active compared to those who were work active (studies 1 and 2). Those who were work active also reported having health problems, however, to a lower extent than those who did not work. The fourth study revealed that working life was an important source of satisfaction for the interviewed participants with USH2. However, they also disclosed experiences of limitations and uncertainty related to their working life. Although they had a commitment to work, they were striving for a balance between work and leisure time to maintain or regain their health (study 4). To be able to remain active and to have the ability to appreciate the present through the enjoyment of meaningful activities was described in the focus group interviews with people with USH2. They strived to take command over their own lives, prevented and resolved challenges and found ways of soothing themselves (study 3).

The results of this thesis will be discussed in relation to each other, in relation to relevant research and theoretically by the use of models of psychological flexibility (Hayes et al., 2012) and the theoretical framework of the Meikirch model of health (Bircher & Kuruvilla, 2014).

To work or not to work

A substantial portion of the participants with USH2 (49%) and USH1 (51%) were not work active. i.e., had a disability pension, were on sick-leave or were unemployed (studies 1 and 2). Differences in the level of hearing or vision impairment, age and gender did not explain work activity (studies 1 and 2). These results are consistent with what has been found in a report from the UK by Ellis and Hodges (2013). The results in studies 1 and 2 regarding work activity are also somewhat in line with what has been found among people with different disabilities in Sweden, where 60% are employed (Statistics Sweden, 2019). However, the Swedish statistics do not reveal any impairment-specific figures. An American study showed that
among people with dual sensory loss, only 33% were employed (Varadaraj et al., 2019), figures that are in line with what have been found in a Canadians study of legal blindness (Benoit et al., 2013). It is interesting to note that people with USH that have a dual sensory loss seem to have a better employment rate, close to 50%, compared to American figures on dual sensory loss (Varadaraj et al., 2019) and Canadian figures on single sensory loss, as described by Benoit et al. (2013). This outcome might be understood by country-specific factors related to differences in support available from welfare systems (c. f. Diderichsen et al., 2012).

However, another possible explanation could be the progressive course of visual impairment in USH, as participants in study 4 revealed that they have had their employment for many years, long before their vision loss become prominent. It is plausible that the combination of the onset of visual problems in combination with long-term employment might have had a positive impact on their chances of employment compared to other groups of people with visual impairment with an earlier onset. According to this reasoning, differences in onset and progression are put in terms of the Meikirch model, important aspects of an individual’s biologically given potential to meet the needs of their working life as a life demand.

The interplay between work and health

Having work was a source of great satisfaction based on feeling competent, feeling recognized and a sense of belonging (study 4) and was associated with lower rates of psychological health problems (studies 1 and 2). The participants with USH2 emphasized the importance of their belonging to a work group; some participants underlined work as being the most important or even the only arena for social interaction outside of their family (study 4). These results are in line with what has been shown by Blustein (2008) with regards to the general population, that working life is one of the most important arenas for meeting the psychological and social needs for people who are of working age. The results of work-active people with USH2 (studies 1 and 4) are consistent in that, rather than losing faith in themselves (study 1), they show confidence (studies 3 and 4), perceive their self-value (study 4) rather than their worthlessness (study 1) and emphasize their competence (study 4) rather than their difficulties in handling problems (study 1).
The positive outcomes, i.e., feelings of satisfaction, could, in light of the Meikirch model (Bircher & Kuruvilla, 2014), be interpreted as when people with USH have a well-functioning working life (life demand) and their individual potential is sufficient for that work, they have a positive state of health. Based on this reasoning, the importance seems to be finding a balance between work and the individual’s potential. Moreover, based on this reasoning work might even have a reinforcing effect on an individual’s acquired potentials, expressed as feelings of recognition, perceived competence, and sense of belonging, as identified in study 4. The compensatory effects of using routines, being competent, feeling related and self-confident, herein interpreted as acquired potentials and described in studies 3 and 4, might to some degree buffer the limitations related to biologically given potentials, i.e., restrictions in vision and hearing.

Social networks and support from society were deemed important to remaining work active and managing everyday situations among people with USH2 (studies 3 and 4). Interpreting our results in terms of social determinants (Diderichsen et al., 2012), working conditions (study 4), social networks (studies 3 and 4), economy (studies 2 and 4), and opportunity for active living habits (studies 1, 3 and 4), along with functioning informal and formal support (studies 2, 3 and 4), were all important in relation to remaining active in general (study 3) or work active (studies 1, 2 and 4) and health among people with USH. Furthermore, work-active people with USH1 who reported no problems with levels of social trust or financial situations were those who reported low levels of psychological health problems (study 2). Our results correspond to what has previously been reported to be important in relation to health and well-being in individuals with USH in terms of specialized support (Ellis & Hodges, 2013), social networks (Evans, 2017), and social support in general (Dean et al., 2017). However, there are no studies specifically focusing on the social determinants of health in people with USH (Arcous et al., 2019). Nevertheless, in light of our results, it is plausible to interpret social determinants’ potential to function as lubricant in the interplay between work as a life demand and an individual’s potential (c.f Bircher & Kuruvilla, 2014).

The interplay between being nonworking and having ill-health
The results of poor psychological health among nonworking people with USH (study 1, 2) are in line with what has previously been reported among
people with USH, i.e., negative effects of being outside the labor market due to long-term sick leave (Ellis & Hodges, 2013). The poor psychological health of people with USH who were not working (studies 1 and 2) is also consistent with what has been found in a meta-analysis of unemployed people in general, with a significant increase in psychological health problems such as anxiety, depression, poor self-esteem, lack of subjective well-being and psychosomatic symptoms (Paul & Moser, 2009). The meta-analysis not only demonstrated positive correlations between unemployment and symptoms of distress but also showed that unemployment actually caused psychological health problems. No such analysis of correlations was possible from the limited data of people with USH in studies 1 and 2.

In the general population, the unemployment rate has been shown to correlate positively with suicide mortality (Nordt et al., 2015; Stuckler et al., 2009). Similarly, among the nonworking people with USH in studies 1 and 2, one out of five (20%) reported having made at least one suicide attempt. The situation of suicide in people with single sensory loss due to visual impairment shows an increased risk of early mortality (Meyer-Rochow, Hakko, Ojamo, Uusitalo, & Timonen, 2015). In the general population, there is a strong positive correlation between suicide attempts and mortality, making suicide attempts an extremely important predictor of the risk for suicide (Christiansen & Jensen, 2007; Skogman, Alsen, & Ojehagen, 2004). Hence, it is of the utmost importance that psychological health problems, including issues of suicide, are addressed for people with USH.

In light of the lived experiences of competence, a sense of belonging and recognition found in relation to work in study 4, it is not unlikely that some of the results of significantly poorer psychological health among the nonworking people with USH who reported feeling worthless, having a loss of faith in one-self and difficulties handling problems (study 1) could be understood by the absence of satisfaction and commitment associated with work (study 4). The results from studies 1 and 2 related to not having a job can be interpreted as an incongruent situation, where the life demand does not suffice the individual’s acquired potentials activating a situation of psychological distress, as reported among people with unemployment described above (Paul & Moser, 2009). The results concerning not having work (studies 1 and 2), having work with too-high demands (study 4) or having a work life in association with other strenuous life demands, i.e., family life (study
4) might, from a theoretical perspective, all lead to a failure in balance between life demands of individual potentials and social determinants. This imbalance could lead to poor health and, in the worst cases, suicidal behavior (c.f. Bircher & Kuruvilla, 2014). The situation of poor psychological health found among nonworking people with USH calls for further attention in clinical practice as well as in further research.

**Life strategies and psychological flexibility**

Among people with USH2, uncertainty and unpredictability, as well as insecurity, are common themes, both in relation to working life (study 4) and as an overall challenge related to living with USH2 (study 3). These results are in line with what Ellis & Hodges (2013) found among people with USH, where participants disclosed a situation of ongoing change in function due to their diagnosis and the constant challenge of trying to create predictability in an unpredictable situation. However, the authors also found great ingroup differences in how people perceived challenges (Ellis & Hodges, 2013).

The capability to actually handle challenges was the main focus in study 3, showing a variety of life strategies to prevent and resolve challenges, as well as for comforting oneself. In a work life context, life strategies were manifested in the ability to taking responsibility for recovery, prioritizing work tasks, and accepting adaptations (study 4). In addition to disclosing different strategies, the participants in studies 3 and 4 show that strategies vary due to differences in age and progression in hearing or vision.

The life-altering consequences of constant change in hearing and vision among people with deafblindness has previously been highlighted in a report by Gullacksen, Göransson, Rönnblom, Koppen, & Rud Jörgensen (2011). In their life adjustment model, the authors highlight the importance of taking a process-oriented perspective when studying health in people with deafblindness. The life adjustment model emphasizes an ongoing process in which both personal and social factors are important for how people with deafblindness handle the progression of hearing and vision loss (Gullacksen et al., 2011). Our studies did not have the opportunity to follow people over time, but the retrospective perspectives and individual differences in narratives in study 3 reveal that age and progression of vision loss were closely...
associated with differences in some strategies used in handling everyday situations, thereby indicating a development over time. This development calls for further studies.

The ability to use life strategies (studies 3 and 4) when facing obstacles and uncertainty can be described in terms of psychological flexibility (Hayes et al., 2012). When people with USH are psychologically flexible, they are both in contact with the present moment and maintain or change their behavior in line with their own chosen values (c.f. Hayes et al., 2012). In light of the Meikirch model (Bircher & Kuruvilla, 2014), a range of life strategies reveals psychological flexibility (personally acquired potentials) with the ability to shift strategies due to the progression of hearing and vision (biologically given potentials). The people with USH2 who participated in study 3 were, in line with this rezoning, interpreted as having the ability to buffer biologically given potentials by personally acquired potentials.

**Health as an act of balance**

People with USH2 who were work active showed a strong commitment to work and showed a variety of strategies to balance their work by recovery, accepting adaptations or prioritizing work tasks, aiming to restore a balance between their work, leisure time and health (study 4). The work life balance, also known as the association between working life and leisure time, shows that people reporting a poor work-life balance often report health problems (Lunau et al., 2014). Some of the participants in our study reported job strain, constantly feeling tired and exhaustion (study 4), as well as stress and fatigue (study 1). Högner (2015) showed that people with USH who were work active reported high levels of stress related to employment. Job strain, tiring working positions, and uncertainty related to temporary job contracts have been shown to be strongly associated with a higher likelihood of reported health problems in the general population (Bambra et al., 2014). The participants with USH2 did not report having temporary job contracts, but they disclosed a feeling of uncertainty regarding their own possibility of continuing to work in the future due to their progressive impairment (study 4).

Many of the participants reported making every effort to keep their position or to compensate for limitations due to their visual or hearing impairment, sometimes by working extra hour outside regular working time (study 4).
Moderate working time and welfare state support, common in Scandinavian countries, is said to be beneficial for a work-life balance (Lunau et al., 2014). In view of the extra effort spent by some of the participants, it is likely that the results can be interpreted such that working time regulations and other general welfare services associated with the welfare state that facilitate work-life balance for people in general do not have the same significant impact on the work-life balance of people with USH2.

Fatigue was an aspect of health problems that was reported by people with USH2 (studies 1 and 4), where more than 70% reported having problems (study 1). The results revealed a working life situation of constantly being tired and on the edge of exhaustion or fatigue (study 4). A systematic review has shown that among people with single sensory loss due to hearing impairment, fatigue is common (Punch, 2016). Hua, Anderzén-Carlsson, Widén, & Lyxell (2015) similarly showed that fatigue among people with a hearing impairment was a consequence of the effort and concentration needed in work related to communication situations. Fatigue calls for extra recovery, and as a consequence, has a negative impact on leisure activities and on family life. For some people with hearing impairment, fatigue is associated with long-term sick leave (Svinndal, Solheim, Rise, & Jensen, 2018). The situation discovered in people with a hearing impairment was shared by people with USH2, where the need for recovery not only balanced the strenuous working life but was a prerequisite for continuing working (study 4). Nevertheless, for some, the work-related strain, tiredness and eventual exhaustion led to fatigue, depression and, in due course, sick leave (study 4).

The results from study 4 indicate that work has both positive and negative health outcomes in people with USH and that finding a balance has been shown to be a challenge. By using all the pieces of the Meikirch model jigsaw puzzle (Bircher & Kuruvilla, 2014), as described in the Introduction section, we can, by putting the different pieces together, identify health or ill health. The progression in vision loss (biologically given potential) in USH forms a piece of the jigsaw puzzle that constantly changes in shape and size. Another piece of the jigsaw puzzle that can shift form is the individual’s life strategies, i.e., the individual’s acquired potential. Work among people with USH has shown to be an important piece of the jigsaw puzzle of health, as a life demand that sometimes has the perfect shape and size but that is sometimes too small and other times is too large to fit into the puzzle. The
last piece of the puzzle, the social determinants, is often associated with support from society, which is indeed a piece that can shift in size or shape. When people with USH have a situation in which the pieces of the jigsaw puzzle fit together, there is an opportunity for health (c. f. Bircher & Kuruvilla, 2014). Any change in shape by one of the pieces, will, according to this reasoning, need to be compensated by shifts in any or all of the other pieces to maintain a balance in health.

**Methodological Discussion**

An explorative design of all four studies was regarded as appropriate in view of the fact that the literature on work and health, as well as life strategies among people with USH, is sparse. To explore the different aspects of health and work, both quantitative and qualitative methods were employed. The first two studies were quantitative and raised new questions best countered by the use of qualitative methods. The use of a set of multiple methods enabled triangulation (Polit & Beck, 2013) and the scrutiny of different aspects of health and work enabled a deepened understanding of health and work in Usher syndrome type 2. However, in the process discussing all of the included studies, it became evident that variables in the HET questionnaires represent the poor side of the health continuum rather than the good side of health. In this project, the absence of poor health has therefore been regarded as an indicator of health. With the use of a different questionnaire, this interpretation would not have been necessary. However, the quantitative data were already collected in a previous project, and HET is a well-established questionnaire with high construct validity and reliability (Boström & Nyqvist, 2010). The data from study 1 were collected 12 years ago. It is plausible that shifts in the welfare system during the last decade would have affected the results if they were collected today. However, one benefit from using collected data is from an ethical perspective, i.e., this use is better than exposing people with USH to yet another data collection.

The number of participants in the quantitative studies may be considered low, but the total number of people with USH in Sweden is small. The USH2 study had a response rate of 80%, representing approximately 40% of the total population of known individuals of working age in Sweden. Based on this knowledge, the sample size can be regarded as relatively good and has a positive impact on the internal validity. The USH1 study had a somewhat lower response rate, 71%, which represents approximately 20% of the
known individuals with USH1 of working age in Sweden. The response rate may be considered high when considering the visual difficulties that might limit a person’s ability to respond to an extensive questionnaire. To the best of our knowledge, the number of participants in studies 1 and 2 is by far the largest in work- and health-related studies in people with USH, and the consistency in results between the two studies may have a positive impact on external validity in terms of the possibility to generalize the findings to other clinical groups of people with USH, as well as other groups of individuals with similar hearing and vision progression.

The statistical analysis of small groups is a challenge as small groups are often associated with low statistical power and limited ability to obtain less than strong effects. Hence, results that would have been statistically significant in a study with larger sample sizes might not have been identified in studies 1 and 2. Our sample is based on a register, and there is a risk of selection biases as nonresponders (study 2) had somewhat lower vision levels than responders; thus, the results may reflect a group of persons with slightly better vision than average for persons of working age with USH1, which would negatively affect the internal validity. Both quantitative studies revealed significant correlation; however, since they were cross-sectional studies, no conclusions regarding causality can be established.

The qualitative design of studies 3 and 4 was regarded as appropriate as both studies aimed to explore experiences. The method of focus group interviews with people with USH2a used in study 3 enabled interaction between the participants, eliciting a variety of perceptions. The credibility in the two qualitative studies was reached, for example, by audio recording, as well as by professional transcription. Furthermore, the involvement of all authors, representing different professions with extensive clinical and research experience, in the content analysis (study 3) and the interpretative phenomenological analysis (study 4) strengthened the credibility and confirmability (Graneheim & Lundman, 2004; Smith et al., 2009). The trustworthiness was also increased by all participants having a clinically confirmed diagnosis. The group of persons with USH2 was relatively small, and ensuring the attendance of persons of different ages and gender from various parts of Sweden in study 3 and 4 was challenging. Thus, one limitation of the study is the small number of women who participated in study 3. The ability to assess transferability to other groups of people with deafblindness...
is enhanced through the rich presentation of results and background data (Graneheim & Lundman, 2004). The fourth study could have been designed in another way by using questionnaires, enabling participation and using advanced statistical analysis. An extensive interview study including both working and nonworking people could have also provided interesting insights.

The use of cross-sectional studies enabled the inclusion of many participants, and the qualitative studies offered narratives that it would not have been possible to attain with the use of questionnaires and statistical inferences. Thus, the variety used reveals a picture that is both wide and deep. The cross-sectional studies show significant correlations between work and health, and the phenomenological study of lived experiences of work life not only shows health benefits congruent with the results of the quantitative studies but also permits insight into the nuances of health that is only available by narratives best studied through interviews.
Conclusion

The general aim of the current thesis was to explore the relationship between work and health in people with USH1 and USH2, as well as the life strategies of people with USH2. The thesis has highlighted feelings of great satisfaction and commitment to work among people with USH and has shown that work-active people with USH report significantly better health compared to people who are not work active.

The quantitative results in the thesis have demonstrated a significant difference in psychological health between working and nonworking people with USH, with work active reporting significantly less psychological health problems than those who are not work active.

The levels of social trust and financial situations were poor in people with USH1 who were not work-active. Nonworking people with USH1 and USH2 reported poor psychological health, where the most striking difference between working and nonworking people with USH was that one out of five nonworkers reported having made at least one suicide attempt. The relation found between not working and poor health highlights that the nonworkers are a group that needs particular attention in health care and rehabilitation, and their needs call for further studies.

Work activity was not only related to positive health outcomes, such as satisfaction, but also revealed feelings of limitations and uncertainty about the future. People with USH2, however, showed a commitment to maintaining a work-life balance and to actively reducing the risk of health hazards related to their working life. The ability to find a balance was not only highlighted in the context of work but was also found in the study of life strategies pertaining to challenging everyday situations. People with USH2, by taking the helm, showed a variety of strategies used to prevent and resolve challenges as well as to comfort themselves. These strategies were helpful in striving to be in contact with the present moment and maintaining or changing behaviors in line with their own chosen values.

People with USH have demonstrated that personally acquired potentials, such as life strategies and being psychologically flexible, increased their individual potential to handle life demands. Moreover, by studying health
through one specific life demand, namely, work, in relation to social determinants and individual potentials, a nuanced picture of health in people with USH emerged. This thesis has thereby provided new insights into health in people with USH, and these insights may be valid for other groups of people with deafblindness. Perhaps the current study reflects sides of health that will be valid for anyone of us in our life span.

**Future Research**

A future continuation of the exploration of the lived work experiences in other groups of people with USH would enable a more comprehensive picture of the working life of people with USH. The HET questionnaire used in studies 1 and 2 is designed to be used in longitudinal studies. Another future study could be the exploration of experiences related to not being work active. Such studies are essential since they would enable insights into the lived experiences of a group of people who in the present thesis have been described as having the most psychological health problems and highest levels of reported suicide behavior. In addition to descriptive studies, the results from the study on life strategies (study 3) revealed psychological flexibility. However, there is a need for studies focusing on interventions that can address psychological inflexibility. Such studies would enable the development of evidence-based guidelines for practitioners.
Svensk sammanfattning/Swedish summary

Föreliggande doktorsavhandling handlar om hälsa och arbete hos personer med Ushers syndrom och de livsstrategier personer med Ushers syndrom typ 2 använder för att hantera sin livssituation.

Introduktion


Tidigare forskning har visat att personer med USH uppvisar en försämrad psykisk och fysisk hälsa. Det finns emellertid stora skillnader i upplevd hälsa inom gruppen som inte kan förklaras av skillnader i hörsel och syn. För att förstå skillnader i upplevd hälsa mellan olika personer med USH kan ett vidgat, biopsykosocialt hälsobegrepp användas, som även omfattar personliga faktorer som livsstrategier och omgivningsfaktorer t.ex. anknytning till arbetsliv.

Det finns ett flertal studier som belyser arbetets betydelse för hälsa och att arbetslivet är en av de viktigaste arenorna för att tillgodose sociala och psykiska behov hos människor i allmänhet. Samtidigt visar forskning att det finns starka orsakssamband mellan arbetslöshet och upplevd ohälsa. FN:s konvention om rättigheter för personer med funktionshinder (artikel 27) tillerkänner personer med funktionsnedsättning samma rätt till arbete som andra i befolkningen. Svensk statistik visar dock att bland personer med
funktionsnedsättning har endast 60% ett arbete jämfört med 80% i befolkningen som helhet. Ingen motsvarande statistik finns för personer med dövblindhet i Sverige, men amerikanska och kanadensiska studier visar att bland personer som har en kombinerad syn och hörseledsättning så har cirka 33% ett arbete.


Det finns ett flertal studier som fokuserat på det som i Meikirsh hälsomodell definieras som biologiskt givna förutsättningar, såsom genetik, hörse- och synförmåga hos personer med USH. Emellertid är forskning som studerat hälsa utifrån livskrav, som till exempel arbete, sällsynt. Likaså är studier som fokuserar på personligt utvecklade förutsättningar, såsom livsstrategier, sällsynt förekommande.

**Syfte**

Det övergripande syftet med denna avhandling var att studera samband mellan arbete och hälsa hos personer med Usher syndrom typ 1 och 2 samt att studera livsstrategier hos personer med Usher syndrom typ 2a.

Syftena i de fyra delstudierna var:

1. Att utforska skillnader i fysisk och psykisk hälsa mellan personer med USH2 som arbetar och de som har sjuksäkring.
2. Att utforska relationen mellan hälsa, arbete och sociala determinanter såsom social tillit och ekonomisk situation hos personer med USH1 i jämförelse med en referensgrupp.

3. Att utforska livsstrategier hos personer med USH2 a.

4. Att utforska levda erfarenheter av arbete hos personer med USH2.

**Metod**

Avhandlingens fyra delstudier har en explorativ design. Både kvantitativa och kvalitativa metoder har använts. De två kvantitativa studierna utgjordes av tvärsnittsstudier med syfte att utforska samband mellan arbete och hälsa hos personer USH2 (Studie 1) och USH1 (studie 2). Den första kvalitativa studien (studie 3) baserades på fokusgruppsintervjuer som analyserades med induktiv innehållsanalys. För att följa upp studie 1 och möjliggöra en fördjupad analys av resultaten i studie 1 genomfördes individuella semi-strukturerade intervjuer med personer med USH2 (Studie 4) som analyserade med tolkande fenomenologisk analys.

Alla deltagare i de fyra delstudierna rekryterades från det svenska Usher registret. Registret omfattar data om klinisk och genetisk diagnos, hörsel, synskärpa och synfält samt hälsodata insamlade med enkäten Hälsa på lika villkor.

Studie 1 omfattade registerdata från 67 personer med USH2 (34 personer med arbete och 33 personer utan arbete). I studie 2 hämtades registerdata rörande 47 personer med USH1 (23 personer med arbete och 24 personer utan arbete). I Studie 3 deltog 14 personer i fokusgruppsintervjuer och i studie 4 deltog sju personer i individuella intervjuer.

I de två första studierna hade studiedeltagarna tidigare besvarat ett omfattande frågeformulär: Hälsa på lika villkor, som innefattar frågor som rör fysisk och psykisk hälsa, livnadsvanor, familjesituation och arbete. I studie 3 genomfördes fokusgruppsintervjuer där deltagarna under ledning av samtalsledare diskuterade olika sätt att hantera sin livssituation. I studie 4 genomfördes 7 enskilda kvalitativa intervjuer om levda erfarenheter av arbete och dess betydelse för hälsa. En intervjuguide användes.
Icke-parametrisk Pehrson Chi-2 testning användes för att jämföra hälsovariabler mellan grupper i studie 1. I studie 2 jämfördes hälsodata från personer med och utan arbete med data insamlade från en referensgrupp bestående av 3247 personer ur den svenska befolkningen. En multipel logistik regressionsmodell användes för att jämföra de olika grupperna. I studie 3 analyserades fokusgruppsintervjuerna med hjälp av induktiv innehållsanalys. I studie 4 analyserades de levda erfarenheterna med en tolkande fenomenologisk analys, som även inkluderar att intervjuarens tidigare erfarenheter aktivt används som ett steg i den tolkande analysprocessen.

Resultat

Studie 1:
Personer med USH2 som arbetade hade en signifikant bättre psykisk hälsa än de som hade sjukersättning. Självmordstankar och självmordsförsök var signifikant vanligare hos personer som hade sjukersättning. Problem med stress och utmattning var vanliga och förekom ungefär lika ofta i båda grupperna.

Studie 2:
I studie 2 framkom att personer med USH1 som saknade arbete hade sämre social tillit och ekonomisk situation samt oftare rapporterade sämre fysisk och psykisk hälsa, jämfört med personer med USH1 som hade arbete. Självmordsförsök var vanligare hos personer med USH1 som saknade arbete. Vid en jämförelse mellan personer med USH1 och referensgrupper med och utan arbete framkom samma hälsomönster, men magnituden av skattade problem var genomgående större hos personer med USH1.

Studie 3:
Innehållsanalysen resulterade i 7 manifesta kategorier och 17 underkategorier. Därutöver framkom ett överordnat tema: ”Att hålla handen på rodret”. Temat kunde delas in i två underteman där det första beskrev hur deltagarna förebyggde och hanterade uppkomna utmaningar i livet. Till exempel beskrevs strategier såsom att fortsätta vara aktiv, att använda hjälpmedel, att använda stöd och att dela kunskap med andra. Det andra temat fokuserade på mer känslomässigt inriktade sätt att trösta och lugna sig själv. Exempelvis beskrev deltagarna att de uppskattade nuet, strävade efter att behålla en positiv självbild, och att de strävade efter att minska känslomässig smärta.
Studie 4:

Slutsatser
Avhandlingen har visat att arbete är relaterat till upplevd tillfredsställelse och ett starkt engagemang hos personer med USH2, men även att personer med USH1 och USH2 som arbetar har bättre hälsa, än de som inte arbetar. De båda kvantitativa studierna i avhandlingen visade signifikanta skillnader i upplevd psykisk hälsa mellan de som arbetar och de som inte arbetade, där yrkesverksamma rapporterade signifikant färre psykiska hälsoproblem än de som inte var yrkesverksamma.

Den sociala tilliten och ekonomiska situationen var signifikant sämre hos personer med USH1 som saknade arbete och den psykiska hälsan var signifikant sämre hos de som saknade arbete såväl hos personer med USH1 och USH2. Ett av de viktigaste fyndena avseende psykisk hälsa var skillnader i rapporterade självmordsförsök. Bland de som saknade arbete hade en av fem, vid minst ett tillfälle försökt ta sitt liv. Det tydliga sambandet mellan hälsoproblem och att inte ha ett arbete understryker att detta är en grupp som särskilt behöver uppmärksammas inom vård, rehabilitering och habilitering. Fördjupade studier behövs för att bättre förstå orsaker till ohälsa i denna grupp samt för att utveckla stöd som kan bidra till bättre hälsa.

Att ha ett arbete var dock inte enbart associerat till positiva hälsoupplevelser. Arbetslivet var också förknippat med upplevelser av begränsningar och ovisshet inför framtiden. Personer med USH2 visade ett starkt engagemang och strävade efter att bibehålla en hälsosam jämvikt i arbetslivet. De tog aktiver ansvar för att minskar arbetsrelaterade hälsorisker. Avhandlingen visar vidare att detta engagemang inte bara återfanns i arbetslivet utan även i...
de livsstrategier som personer med USH2 uppvisade i krävande vardagliga situationer. Genom att ”hålla handen på rodret”, framkom en mångfald av strategier för att förebygga och hantera utmaningar samt för att hantera negativa känslor. Livsstrategierna var till hjälp för personerna för att finna en för dem värdefull riktning i livet genom att både vara medvetet närvarande i nuet och samtidigt behålla eller förändra sina beteenden i linje med viktiga värden i livet.

I ljuset av Meikirsh hälsomodell visar resultatet av avhandlingen att personer med USH2 genom personligt förvärvade egenskaper, såsom livsstrategier ökat sina personliga förutsättningar att hantera krav i livet. Genom att studera hälsa utifrån ett specifikt livskrav, arbete, i relation till sociala determinanter och personliga förutsättningar framträder en mångfaseterad bild av hälsa hos personer med USH.

Denna avhandling har möjliggjort nya insikter om hälsa hos personer med USH. Insikter som även kan vara giltiga för andra grupper av personer med dövblindhet. Kanske visar studierna också sidor av hälsa som i ett livsperspektiv kan vara giltiga för oss alla. Förhoppningsvis kan denna avhandling vara till nytta för att öka kunskapen hos personer som i sitt arbete möter personer med USH eller dövblindhet. Den kan i delar fungera som ett underlag för viktiga samtal om hälsa. En förhoppning är att även personer som själva har USH ska finna avhandlingen värdefull och stärkande.

**Framtida forskning**

Att undersöka levda erfarenheter av arbete hos personer med USH1 skulle kunna ge en mer heltäckande bild av arbetslivet hos personer med USH. En uppföljande enkätstudie med ”Hälsa på lika villkor” skulle möjliggöra longitudinell forskning i linje med den som regelbundet sker i den svenska befolkningen. Att fördjupa kunskapen genom studier av levda erfarenheter hos de som inte arbetar är av stor vikt, då de i denna avhandling är de som rapporterar sämst hälsa.

Förutom fler beskrivande studier behövs även forskning som syftar till att utveckla vetenskapligt beprövade riktlinjer för instanser som har uppdraget att ge stöd till personer med USH.
Acknowledgement

To all participants who have contributed in the research, my sincere thanks. Without your generosity, your time spent, and efforts in sharing your rich life experiences there would not have been any research conducted.

Claes Möller my first main supervisor, from our first meeting in Dronninglund in 2005, your passion and compassion for persons with deafblindness has been a source of inspiration to me, first in my clinical work but I am so grateful that you believed in me and invited me to disability science and to Örebro University. Agneta Anderzén-Carlsson, since you became my main supervisor, you have been my dedicated guide to become a researcher. I am so glad for your commitment, all the experience you have shared and our joint delight for the narratives in people with deafblindness. Moa Wahlqvist, during my years as a PhD student, you always had all the answers to my stupid questions, someone with whom I could share my, sometimes mixed feelings of being a doctoral student. I am also grateful that I had the opportunity to inherit and present some of the data you have collected. Your interest, ability to stay calm and humor has at times helped me to keep a sound distance to my project. Agneta, Moa and Claes, without your support my PhD project and this thesis would not have been possible.

My former supervisor Kerstin Möller, I am so happy for our talks, your sincere interest in persons with deafblindness, and your support early in my project. Berth Danermark, also a former supervisor, thank you for sharing the philosophy of science in disability and for your rich knowledge of research in work and disability. Örjan Dahlström and Göran Jutengren, thanks for all support with statistics.

I want to acknowledge all co-workers at the Audiological Research Centre, for always making me feel as a part of the family. Special thanks to, Jonas Birkelöf, Frida Chebat, Hanna Hagsten, Ann-Marie Helgstedt, Elina Mäki-Torkko, Camilla Sandelius, and Tobias Åslund for all the technical and administrative support and for all chats over a cup of coffee. The Deafblind research group, Hans-Erik Frölander, Elin Lundin, Björn Lyxell, Berit Rönnåsen and Parivash Ranjbar thanks for inspiration and discussions. Your support, warmth and our laughs during our regular meetings also made me feel at home in Örebro. Margareta and Claes...
Möller, you opened up your home when I needed a place to stay and for that hospitality, I am very grateful.

Writing a thesis when having a visual impairment is not without challenges. I want to acknowledge Margareta Landin, you have been my devoted librarian, helping me out whenever facing problems with any references at any time or day of the year. Helena Claesson, without your daily technical assistance, revisions of my texts, your encouragement and humor this thesis would not have been possible. I also want to recognize Annika Behm and Debora Elgeholm for all your assistance in making my visually demanding work more accessible.

Anita Dath & Gunilla Kvist, my executives during these years, and all colleagues, Bo Ahlner, Maria Collin-Jensen, Camilla Falkegård, Emilia Goodwill-Keller, Sofia Hansdotter, Kristin Hansson, Lena Jacobson, Guneli Johansson, Christina Offnegård, Catarina Temse-Andersson, Sari Vähänikkilä, Ronnie Widmark, Ulrika Åström-Svedénäs and Lizette Östman, I am so grateful for all experiences you have shared with me, and most important, for the satisfaction of us spending our working days together during the years. Thanks to your support, encouragement and flexibility it has been possible to combine my research with clinical work.

I want to acknowledge Lena Göransson and Anne Jalakas at The Swedish National Resource Centre for deafblindness for inspiration and for making my research public. Helene Engh and Lil Falkensson and all former colleagues in the Swedish national expert team for diagnosing deafblindness, thank you for all the diagnostic challenges and your encouragement that inspired to do research.

Maria Hugo-Lindén, Caroline Larsson and Camilla Ehnfors a special thanks for your patience and help when navigating through the University bureaucracy.

For all the financial support associated with my project, I want to acknowledge The Swedish institute for disability Research, Örebro University and The Swedish Research Council. Furthermore, I want to recognize Region Stockholm, Habilitering & Hälsa, Centrum för Psykiatriforsknning, Tysta skolan and Svenskt Audiologiskt Sällskap for all economic support.
Peter Dehlbom, my fellow student, thanks for always listening, giving feedback and comfort during our lunches. Mattias Sjöberg, thanks for being my most long lasting friend that I can always talk to and rely on regardless what project I am doing. Emil Östberg, thanks for taking care of my physical health and for listening to my research problems during our jogging tours.

My mother Berit and father Gunnar, without you there would not have been any Mattias and thus, no thesis. With the love and support from you and your spouses Tommy and Maria, along with my siblings Karin, Lina and cousin Anna, the logistics of working hard and late hours during the doctoral studies has always been facilitated smoothly. My brother Martin, our almost daily reflective phone calls always helps me through the day. I want to acknowledge my extended family, Christian, Gunilla, Karin, Kjell, Susanne and Tommy, for all your direct and indirect support in my family life and for your reassurance.

My grandmother Birgit, thank you for always believing in my abilities, I miss you.

Johan, Clara and Julina I love you! Thank you for your never-ending patience. You are the fundamental pieces in my jigsaw puzzle. I love to regain a better work life balance and to spend more time with you now.
References


Kitzinger, J. (1994). The methodology of focus groups: the importance of interaction between research participants. *Sociology of Health and Illness, 16*(1), 103-121.


80  Mattias Ehn  Life strategies, work and health in persons with Usher syndrome
Vetenskapsrådet. (2002). Forskningsetiska principer inom humanistisk-
samhällsvetenskaplig forskning. Retrieved from http://www.co-
dex.vr.se/texts/HSFR.pdf

Vetenskapsrådet. (2017). Good Research Practice Retrieved from
https://www.vr.se/download/18.5639980c162791bbfe697882/1555334908942/Good-Re-
search-Practice_VR_2017.pdf

from http://www.who.int/about/definition/en/print.html

World Health Organization. (2001). International Classification of Func-
tioning, Disability and Health (ICF). Retrieved from
http://www.who.int/classifications/icf/en/


trieved from http://www.who.int/mediacentre/factsheets/fs352/en/
Studies from the Swedish Institute for Disability Research

1. Varieties of reading disability
   Stefan Gustafson

2. Cognitive functions in drivers with brain injury – anticipation and adaptation
   Anna Lundqvist

3. Cognitive deafness
   Ulf Andersson

4. Att lära sig leva med förvärvad hörselnedsättning sett ur par-perspektiv
   Carin Fredriksson
   ISBN 91-7373-105-6, 2001

5. Signs, Symptoms, and Disability Related to the Musculo-Skeletal System
   Gunnar Lundberg

6. Participation – Ideology and Everyday Life
   Anette Kjellberg

7. Föräldrar med funktionshinder – om barn, föräldraskap och familjeliv
   Marie Gustavsson Holmström
   ISBN 91-7203-500-5, 2002

8. Active wheelchair use in daily life
   Kersti Samuelsson

9. Två kön eller inget alls. Politiska intentioner och vardagslivets realiteter i den arbetslivsinriktade rehabiliteringen
   Marie Jansson
10. **Audiological and cognitive long-term sequelae from closed head injury**  
    Per-Olof Bergemalm  
    ISBN 91-7668-384-2, 2004

11. **Att vara i särklass – om delaktighet och utanförskap i gymnasiesärskolan**  
    Martin Molin  
    ISBN 91-85295-46-9, 2004

12. **Rättvis idrottsundervisning för elever med rörelsehinder – dilemma kring omfördelning och erkännande**  
    Kajsa Jerlinder  
    Licentiate Degree, 2005

    Per-Inge Carlsson  
    ISBN 91-7668-426-1, 2005

14. **Hearing and cognition in speech comprehension. Methods and applications**  
    Mathias Hällgren  
    ISBN 91-85297-93-3, 2005

15. **Living with deteriorating and hereditary disease: experiences over ten years of persons with muscular dystrophy and their next of kin**  
    Katrin Boström  
    ISBN 91-7668-427-x, 2005

16. **Disease and disability in early rheumatoid arthritis**  
    Ingrid Thyberg  
    ISBN 91-85299-16-2, 2005

17. **"Varför får jag icke följa med dit fram?" Medborgarskapet och den offentliga debatten om dövstumma och blinda 1860-1914**  
    Staffan Bengtsson  
    ISBN 91-85457-06-X, 2005

18. **Modalities of Mind. Modality-specific and nonmodality-specific aspects of working memory for sign and speech**  
    Mary Rudner  
    ISBN 91-85457-10-8, 2005
19. **Facing the Illusion Piece by Piece. Face recognition for persons with learning disability**  
Henrik Danielsson  
ISBN 91-85497-09-6, 2006

20. **Vuxna med förvärvad traumatisk hjärnskada – omställningsprocesser och konsekvenser i vardagslivet. En studie av femton personers upplevelser och erfarenheter av att leva med förvärvad traumatisk hjärnskada**  
Thomas Strandberg  

21. **Nycklar till kommunikation. Kommunikation mellan vuxna personer med grav förvärvad hjärnskada och personernas närstående, anhöriga och personal**  
Pia Käcker  

22. **”Aspergern, det är jag”. En intervjustudie om att leva med Asperger syndrom**  
Gunvor Larsson Abbad  

23. **Sounds of silence – Phonological awareness and written language in children with and without speech**  
Janna Ferreira  

24. **Postponed Plans: Prospective Memory and Intellectual Disability**  
Anna Levén  

25. **Consequences of brain tumours from the perspective of the patients and of their next of kin**  
Tanja Edvardsson  

26. **Impact on participation and service for persons with deafblindness**  
Kerstin Möller  

27. **Approaches to Audiological Rehabilitation with Hearing Aids: studies on prefitting strategies and assessment of outcomes**  
Marie Öberg  
28. Social Interaction and Participation in Activities of Everyday Life Among Persons with Schizophrenia

Maria Yilmaz
Licentiate Degree, 2009

29. Focus on Chronic Disease through Different Lenses of Expertise Towards Implementation of Patient-Focused Decision Support Preventing Disability: The example of Early Rheumatoid Arthritis

Örjan Dahlström

30. Children with Cochlear Implants: Cognition and Reading Ability

Malin Wass

31. Restricted participation: Unaccompanied children in interpreter-mediated asylum hearings in Sweden

Olga Keselman

32. Deaf people and labour market in Sweden. Education – Employment – Economy

Emelie Rydberg

33. Social rättvisa i inkluderande idrottsundervisning för elever med rörelsehinder – en utopi?

Kajsa Jerlinder
ISBN: 978-91-7668-726-0, 2010

34. Erfarenheter av rehabiliteringsprocessen mot ett arbetsliv – brukarens och de professionellas perspektiv

Helene Hillborg

35. Knowing me, knowing you – Mentalization abilities of children who use augmentative and alternative communication

Annette Sundqvist
36. **Lärare, socialsekreterare och barn som far illa – om sociala representationer och interprofessionell samverkan**  
   Per Germundsson  
   **ISBN:** 978-91-7668-787-1, 2011

37. **Fats in Mind**  
   *Effects of Omega-3 Fatty Acids on Cognition and Behaviour in Childhood*  
   Ulrika Birberg Thornberg  
   **ISBN:** 978-91-7393-164-9, 2011

38. **"Jobbet är kommunikation”**  
   *Om användning av arbetshjälpmedel för personer med hörselnedsättning*  
   Sif Bjarnason  

39. **Applying the ICF-CY to identify everyday life situations of children and youth with disabilities**  
   Margareta Adolfsson  

40. **Tinnitus – an acceptance-based approach**  
   Vendela Zetterqvist  
   **ISBN:** 978-91-7393-040-6, 2011

41. **Applicability of the ICF-CY to describe functioning and environment of children with disabilities**  
   Nina Klang  
   **ISBN:** 978-91-7668-864-9, 2012

42. **Bringing more to participation**  
   *Participation in school activities of persons with Disability within the framework of the International Classification of Functioning, Disability and Health for Children and Youth (ICF-CY)*  
   Gregor Maxwell  
   **ISBN:** 978-91-628-8484-0, 2012

43. **From Eye to Us.**  
   *Prerequisites for and levels of participation in mainstream school of persons with Autism Spectrum Conditions*  
   Marita Falkmer  
   **ISBN:** 978-91-637-2091-8, 2013
44. **Otosclerosis, clinical long-term perspectives**  
Ylva Dahlin-Redfors  

45. **Tinnitus in Context - A Contemporary Contextual Behavioral Approach**  
Hugo Hesser  

46. **Hearing and middle ear status in children and young adults with cleft palate**  
Traci Flynn  

47. **Utrymme för deltagande, beslutsprocesser i möten mellan patienter med ospecifica ländryggsbesvär och sjukgymnaster i primär vård**  
Iréne Josephson  

48. **"Man vill ju klara sig själv” Studievardagen för studenter med Asperger syndrom i högre studier**  
Ann Simmeborn Fleischer  

49. **Cognitive erosion and its implications in Alzheimer’s disease**  
Selina Mårdh  

50. **Hörselscreening av en population med utvecklingsstörning**  
Utvärdering av psychoakustisk testmetod och av OAE-registrering som komplementär metod  
Eva Andersson  

51. **Skolformens komplexitet – elevers erfarenheter av skolvardag och tillhörighet i gymnasiesärskolan**  
Therése Mineur  

52. **Evaluating the process of change:**  
**Studies on patient journey, hearing disability acceptance and stages-of-change**  
Vinaya Kumar Channapatna Manchaiah  
53. **Cognition in hearing aid users: Memory for everyday speech**  
Hoi Ning (Elaine) Ng  

54. **Representing sounds and spellings Phonological decline and compensatory working memory in acquired hearing impairment**  
Elisabet Classon  

55. **Assessment of participation in people with a mild intellectual disability**  
Patrik Arvidsson  

56. **Barnperspektiv i barnavårdsutredningar – med barns hälsa och barns upplevelser i fokus**  
Elin Hultman  

57. **Internet Interventions for Hearing Loss Examining rehabilitation Self-report measures and Internet use in hearing-aid users**  
Elisabet Sundewall Thorén  

58. **Exploring Cognitive Spare Capacity: Executive Processing of Degraded Speech**  
Sushmit Mishra  

59. **Supported employment i en svensk kontext – förutsättningar när personer med funktionsnedsättning når, får och behåller ett arbete**  
Johanna Gustafsson  

60. **Effects of Specific Cochlear Pathologies on the Auditory Functions: Modelling, Simulations and Clinical Implications**  
Amin Saremi  
61. **Children with profound intellectual and multiple disabilities and their participation in family activities**
   Anna Karin Axelsson

62. **Lexical and Semantic Development in Children With Cochlear Implants**
   Ulrika Löfkvist

63. **Rethinking sound. Computer-assisted reading intervention with a phonics approach for deaf and hard of hearing children using cochlear implants or hearing aids**
   Cecilia Nakeva von Mentzer

64. **Assessing cognitive spare capacity as a measure of listening effort using the Auditory Inference Span Test**
   Niklas Rönnberg

65. **Employees with Aided Hearing Impairment: An Interdisciplinary Perspective**
   Håkan Hua

66. **Prosthetic and Orthotic Services in Developing Countries**
   Lina Magnusson

67. **Dealing with digits - Arithmetic, memory and phonology in deaf signers**
   Josefine Andin

68. **Time is of the essence in speech recognition: Get it fast or think about it**
   Shahram Moradi

69. **Effects of hearing loss on traffic safety and mobility**
   Birgitta Thorslund
70. Aspekter på lärande vid dövblindhet - möjligheter och begränsningar för personer med Alström syndrom
   Berit Rönnäsen

71. Memory and communication in typically developing infants and children with Autism Spectrum Disorder: Behavioral and electrophysiological indices
   Emelie Nordqvist

72. Cognitive capacities and composite cognitive skills in individuals with Usher syndrome type 1 and 2
   Cecilia Henricson

73. Functioning and Disability in Adults with Hearing Loss Preparatory studies in the ICF Core Sets for Hearing Loss project
   Sarah Granberg

74. Speech recognition and memory processes in native and non-native language perception
   Lisa Kilman

75. To include or not to include: Teachers’ social representations of inclusion of students with Asperger diagnosis
   Ann-Charlotte Linton

76. Health and People with Usher syndrome
   Moa Wahlqvist

77. Children with mild intellectual disability and their families – needs for support, service utilisation and experiences of support
   Lena Olsson
78. Deafblindness
Theory-of-mind, cognitive functioning and social network in Alström syndrome
Hans-Erik Frölander

79. Signs for Developing Reading
Sign Language and Reading Development in Deaf and Hard-of-Hearing Children
Emil Holmer

80. Speech masking speech in everyday communication
The role of inhibitory control and working memory capacity
Victoria Stenbäck

81. Neural and Cognitive Effects of Hearing Loss on Speech Processing
Eline Borch Petersen

82. A biopsychosocial approach to functioning, oral health and specialist dental health care in children with disabilities – Swedish and international perspectives
Johanna Norderyd

83. Samverkansprojekt, och sen då? – en uppföljande studie av samverkansprocessen kring barn och unga som far illa eller riskerar att fara illa
Ulrika Englund

84. Aural rehabilitation programs for hearing aid users
Evaluating and clinically applying educational programs, supported via telephone and/or the internet and professionally guided by an audiologist
Milijana Malmberg

85. Tolkning vid förmedlade samtal via Bildtelefoni.net – interaktion och gemensamt meningsskapande
Camilla Warnicke
86. **Vardagslivets aktiviteter. Handlingen som terapeutiskt redskap för personer med svårare psykiska funktionshinder – Analys av arbetsterapeuters berättelser med utgångspunkt i G H Meads teori om social handling**
   Marianne Boström

87. **Creative disability classification systems: The case of Greece, 1990-2015**
   Antonia Pavli

88. **Ej utgiven**

89. **Perception of disturbing sounds**
   *Investigations of people with hearing loss and normal hearing*
   Åsa Skagerstrand
   ISBN: 978-91-7529-229-8, 2018

90. **Peer Interaction in Preschool: Necessary, but not Sufficient**
   *The Influence of Social Interaction on the Link between Behaviour Difficulties and Engagement among Children with and without Need of Special Support*
   Madeleine Sjöman
   ISBN: 978-91-88339-13-3, 2018

91. **Prosodic and Phonological Ability in Children with Developmental Language Disorder and Children with Hearing Impairment In the context of Word and Nonword Repetition**
   Simon Sundström

92. **From detection to intervention**
   *Psychological aspects of online hearing rehabilitation*
   Peter Molander
   ISBN: 978-91-7685-226-2, 2018
93. Participation in and outside school
Self-ratings by Swedish adolescents with and without impairments and long-term health conditions
Frida Lygnegård
ISBN: 978-91-85835-92-8, 2018

94. Återgång i arbete efter förvärvad hjärnskada
– livskvalitet, möjligheter och hinder
Marie Matérne
ISBN: 978-91-7529-267-0, 2018

95. Climbing up the hearing rehabilitation ladder
Elisabeth Ingo

96. Situationen på den svenska arbetsmarknaden för personer med funktionsnedsättning
En sekundäranalyse om sysselsättning, yrkesnivå, diskriminering, stöd och anpassning för personer med olika typer av funktionsnedsättning
Tomas Boman
ISBN: 978-91-7529-302-8, 2019

97. Different is cool!
Self-efficacy and participation of students with and without disabilities in school-based Physical Education
Karin Bertills

98. Fonologisk utveckling hos barn med otitbenägenhet
En longitudinell studie i åldrarna 3;6 till 5;6 år
Helena Stålnacke