Impact on participation and service for persons with deafblindness
Studies from the Swedish Institute for Disability Research 26

Kerstin Möller

Impact on participation and service for persons with deafblindness
ABSTRACT


Persons with deafblindness experience difficulties in daily life and they experience service to sometimes barrier. The overall aim of this thesis is therefore to discover, evaluate and explain: 1. mechanisms that might have impact on participation restrictions for people who have visual and hearing impairment i.e. deafblindness and 2. mechanisms that might barrier service to these people. Service is used as an umbrella term for health care, education and certain service for persons with disabilities. Materials from multiple sources have been used: literature (Study I No 96 papers). Interviews (Study I and V) with 32 and 3 adults with deafblindness respectively. Questionnaires (Study II and III): answered by 33 and 34 adults and youth with deafblindness. Patient records (Study IV and V): records from 9 and 3 adult females with USH I respectively. Materials mostly retrospectively cover the period from 2005 and about 40–50 years. Both quantitative and qualitative methods were used. International Classification of Functioning, Disability and Health (ICF) were consequently used as a framework to describe as well as a tool to analyze mechanisms. Further, the Ecological approach, Disability as a laminated system and Life course approach were used in order to evaluate and explain mechanisms. The conclusions that can be drawn from an ecological, laminated and life course approach are: Participation restrictions for people with deafblindness are far-reaching and are embedded in a complex process of interaction between the person with deafblindness and the environment. Services entail systematical barriers. In order to improve service it is extremely important to understand the role of participation restrictions in deafblindness. Primary activity limitation is to not see and hear enough for comprehension. Hence, not taking part in the visible and audible world is primary participation restriction. Performing activities without basic information includes risk. One important aspect of deafblindness is exposure. Persons with deafblindness require rehabilitation in a life perspective. In order to increase people’s participation and protection requirement of individually adapted support and assistive devices is necessary. ICF and the UN convention support service alterations.

Keywords: Deafblindness, ICF, participation restriction, service barrier, Usher Syndrome
ORIGINAl STUDIES

The present thesis is based on the following five studies, which will be referred to in the text by their roman numerals:


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ABBREVIATIONS

AD   Audiology Department
ADL  Activities of daily living
CI   Cochlear Implant
DBU  Deafblind Youth (in Sweden)
FSDB Swedish Association for the Deafblind
HsL  Health and Medical Services Act
IADL Instrumental activities of daily living
ICIDH International Classification of Disease, Disability and Handicap
ICF  International Classification of Functioning, Disability and Health (2001)
LSS  Act concerning Support and Service for Persons with Certain Functional Impairments
LVC  Low Vision Clinic
NUD  The Nordic Staff Training Centre for Deafblind Services
OD   Ophthalmology Department
OMIM Online Mendelian Inheritance in Man
Post Post-lingual denote deafblindness onset after language acquisition and is equal with acquired deafblindness
Pre  Pre-lingual denote onset before or during language skills and is equal with congenital deafblindness
RP   Retinitis pigmentosa
SoL  Social Services Act
UN   United Nations
Update Updated literature review 2002–2007
USH  Usher Syndrom
USH I Usher Syndrom Type I
WHO  World Health Organization
The work on this thesis was carried out at the Swedish Institute of Disability Research (SIDR), School of Health and Medical Science, Örebro University, Sweden. I wish to express my deepest gratitude to everyone who has supported me in different ways in my research. Many people have inspired and helped me. I cannot mention you all. However I have not forgotten your contributions and your friendliness.

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Inverkan på delaktighet och service till personer med dövblindhet

Introduktion

Det råder delade meningar om vad som menas med dövblindhet. En allmän uppfattning är att döv respektive blind betyder att en person vare sig kan se eller höra något alls. Bland professionella som arbetar med service till personer med dövblindhet anpassas ibland definitionen till det regelverk som omger servicen. I en del fall används termen dövblind för att markera att personen är "handikappad". I de nordiska länarna används sedan år 2007 termen dövblindhet som en kombination av syn- och hörselnedsättningar som begränsar aktiviteter och inskränker delaktighet i samhället i sådan grad att det behövs särskild service, anpassningar i omgivningen och/eller teknologi.


Syften
Avhandlingens syften är att belysa, utvärdera och förklara: dels mekanismer som möj-
ligen inskränker delaktighet för personer med dövblindhet och dels mekanismer som
mogligen hindrar service till dessa personer.

Teoretiska överväganden
För att kunna upptäcka, studera och värdera mekanismer som påverkar delaktighet
och service är studien inspirerad av teorin om det ekologiska systemet, teorin om funk-
tionshinder som ett laminerat system och av livsloppsstudier. Den ekologiska teorin
handlar om samspel mellan människa och miljö, t.ex. på det sätt som tidigare beskrevs i
förhållande mellan signaler, kroppsliga strukturer, kroppsfunktion, aktivitet och del-
aktighet. Teorin om det laminerade systemet utgår bland annat från att det finns olika
nivåer i verkligheten och att det för det första finns specifika mekanismer på respektive
nivå men, för det andra, att dessa mekanismer interagerar med varandra i en komplex
helhet. Perspektivet med ett laminerat system har i ett flertal studier inkorporerat en
tidsdimension men den har aldrig tillämpats inom funktionshindersområdet så att ti-
den är en framträdande del i analysen. I denna avhandling betraktas tid som ett ”lami-
nat” där händelse läggs till händelse och på så sätt bildar tidsmässiga mönster av hän-
delser som är socialt och kulturellt sammanvävda. Genom att studera situationer som
laminat och ur livslopps perspektiv kan mönster och förändringar i mekanismerna stu-
deras över tid.

Material och metoder
I de fem delstudierna har olika typer av material, litteratur, intervjuer, frågeformulär
och patientjournaler använts. Den största delen utgörs av primärt källmaterial. Många
olika datainsamlings- och analysmetoder har använts. Se vidare tabell 1.
<table>
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* [http://web.unife.it/progretti/gendeaf/psychosocial/download/Kerstin_bibliography.pdf](http://web.unife.it/progretti/gendeaf/psychosocial/download/Kerstin_bibliography.pdf)
Resultat

De fem delstudierna visar ett komplex mönster av mekanismer med faktiskt eller potentiellt inflytande på inskränkningar i delaktighet.


Studie II visade att personer med dövblindhet har stora aktivitetsbegränsningar och ännu större delaktighetsinskränkningar. Det framkom att förändringar i möjligheterna att se och höra kan förändras på mycket kort tid, eftersom många personer med små funktionsrester är känsliga för förändringar i omgivningen, till exempel ljud och ljus. Även i denna studie rapporteras om att personer med dövblindhet drar sig tillbaka från aktiviteter de ”kan” utföra. Olika aspekter på tid belystes också, t.ex. att många upplevde att det tar längre tid att utföra uppgifter i takt med att funktionerna försämras. Hälsorisker som beror på dövblindhet, t.ex. att inte få nödvändig information från omgivningen och frågetecken som rättigheter och skyldigheter för personer med dövblindhet togs också upp.

I Studie III beskrevs olika faktorer i gymnasiutbildning för elever med dövblindhet. Det förekom både faktorer som bidrar till delaktighet och sådana som begränsar. Hindren för delaktighet i skolan fanns i skolans fysiska miljö, i den pedagogiska miljön och i det sociala umgånget. I denna studie framkom också att vissa elever drog sig tillbaka, vilket kan tolkas som att det fanns brister i erkännandet av dem och att elever upplevt bristande hänsyn.

Studie IV granskade med hjälp av patientjournaler ögon/syn-sjukvården retrospektivt under 20 år för nio kvinnliga patienter med USH I. En tredjedel av de åtgärder vården rapporterat i journalerna var undersökningar. Enbart undersökningar av kroppssstruktur och kroppsfunktion rapporterades. Avvikelserna i patienternas ögon och nedsättningarna av synen ökade väsentligt under den studerade perioden. Vårdens öv-
riga åtgärder riktade sig till största delen till omgivningsfaktorer, till exempel i form av intyg eller hjälpmedel. Studie IV visade vidare att av den stora mängd undersökningar som genomfördes under den studerade 20-årsperioden så fanns inga undersökningar av hur personen genomförde aktiviteter i sin egen miljö, utan endast kapacitet i en optimalt tillrättalagd situation. Även om mängden åtgärder ökade under den senare tionårsperioden, så fanns det ingen korrelation mellan insats och grad eller takt på syn försvåringsarna. I stort sett inga åtgärder riktade sig till personernas psykosociala hälsa. Vården som var fördelad på ett stort antal landsting rapporterade ett stort antal resurser, i genomsnitt 9 åtgärder/person och år i 20 år. Eftersom undersökningar, patientens synförmåga och åtgärder inte korrelerade så indikerar denna studie ineffektivitet.

I studie V användes både intervjumaterial och patientjournaler för att undersöka delaktighet och service hos tre kvinnor med USH I. Denna retrospektiva studie utgick från nutid, och gick bakåt till tidiga barndomsminnen och de första journalanteckningarna. Tidsspannet var ca 50 år. Genom att lägga samman många olika datakällor framkom intressanta mönster där såväl delaktighet som servicens inverkan förändrades över tid.


Studierna I, III och V visar att professionella oftast saknar kunskap om hur kombinationen av funktionsnedsättningarna påverkar aktivitet och delaktighet för personer med dessa funktionshinder. Studie I och IV indikerar stor personalomsättning och studie I att det lätt blir konflikter mellan experter och personal i den vanliga verksamheten.

Studie I och V visade att vården under många år inte upplyste personen själv om sin diagnos. Detta innebar delaktighetsinskränkning som också påverkade relationerna i familjen och försvårade möjligheterna att förbereda sig för det framtida livet med USH. Negativa attityder som brist på hänsyn hos professionella inom utbildning framkom också i studie III.
Slutsatser

De slutsatser som kan dras från ekologisk, laminerad och livslopps perspektiv beskrivna med ICF termer är:

- **Delaktighetsinskränkningar** för personer med dövblindhet är omfattande och är inkluderade i komplexa processer av interaktion mellan personen med dövblindhet och dennes omgivning. Service som syftar till att underlätta dessa personers delaktighet innehåller ibland systematiska hinder.

- Tidigare definitioner av dövblindhet koncentrerar sig på sociala symtom. Eftersom dövblindhetens natur inte granskats i dessa definitioner, så kan inte dövblindhetens symtom förstås i dess fulla vidd. Till exempel så förknippas dövblindhet främst med aktivitetsbegränsningar i det dagliga livet. För att förbättra servicen så är det ytterst viktigt att förstå **delaktighetsinskränkningarnas** roll i detta funktionshinder.

- Dövblindhet är ett funktionshinder som framträder när synliga och hörbara signaler inte passerar genom kroppsstrukturer och kroppsfunktion vilket leder till att aktiviteterna se/titta och höra/lyssna blir begränsade. Ett kännetecken på dövblindhet är delaktighetsinskränkning i information från synliga och hörbara signaler.

- Dövblindhet betyder att den information som andra människor får från det som syns och hörs, inte alls eller i begränsad omfattning, kan ses och höaras av personer med dövblindhet. Grundläggande information kan ofta inte förstås så att den blir begripligt i relation till den aktuella kontexten. Det föreslås att aktivitetsbegränsning och delaktighetsinskränkning delas in i **primär** och sekundär begränsning respektive inskränkning. Primär aktivitetsbegränsning är att inte kunna se och höra tillräckligt för att det ska bli begripligt och sålunda att inte kunna ta del av det synliga och hörbara är primär delaktighetsinskränkning. Resultatet av primär delaktighetsinskränkning är ofta sekundär aktivitetsbegränsning, som är begränsning i andra aktiviteter än att se och höra. Det betyder att begränsningarnas karaktär i till exempel kommunikation, dagligt liv hushållsarbete och att röra sig fritt beror på den primära delaktighetsinskränkning och generellt inte på oförmåga att utföra dessa aktiviteter. Sekundär aktivitetsbegränsning kan vidare skapa sekundär delaktighetsinskränkning.
• För personer med dövblindhet kan möjligheter och faror i den nära omgivningen inte fullt ut upptäckas eller förstås. Att genomföra aktiviteter utan grundläggande information medför risk. En viktig aspekt av dövblindhet är *skyddslöshet*. För personer med dövblindhet betyder skyddslöshet att liv och lem riskeras. En psykologisk konsekvens är att dövblindhet förknippas med ontologisk osäkerhet, vilket är ett resultat av att den information som personen får, inte alltid är tillförlitlig på grund av de primära aktivitetsbegränsningarna. För att undvika att detta leder till social isolering måste betydligt mer resurser satsas på personliga faktorer och sociala omgivningsfaktorer.

• Syn och hörselnedsättningar kan ha olika psykosocial betydelse för personen med dövblindhet, t.ex. så upplevs synnedsättning särskilt svårt för personer med medfödd dövhet eftersom den kan upplevas som ett hot mot deras sociala roll. Personer med dövblindhet drar sig ofta tillbaka om de har möjlighet till ett sådant val. Dövblindhet medför ökad risk för isolering och exkludering i sociala sammanhang. Detta betyder att dessa personer möjlighet att bidra som medborgare hotas vilket är allvarlig delaktighetsinskränkning.


• *Service* till personer med dövblindhet saknas ibland, är inte alltid tillfredsställande och tar inte *personen som helhet* med i beräkningen. Det finns också exempel på service som över tid har förändrats från att ha varit en underlättande faktor till att bli ett hinder. Service som personen tidigare inte fått, eller om den inte varit tillfredsställande så bildar lager av hinder som också har inflytande i det dagliga livet och i förhållande till kontakt med service i nutid. Det finns vidare indikationer på bristfälligt samarbete mellan olika service.
Service till personer med dövblindhet måste ta hänsyn till bio-psyko-sociala aspekter av detta komplexa och sällsynta funktionshinder. Personer med dövblindhet behöver rehabilitering i ett livsperspektiv som tar hänsyn till syndromets komplexitet och prognos. Rehabilitering behöver i större utsträckning än vad som tycks vara fallet idag förstå dövblindhetens natur, betydelsen av den fysiska, sociala och kulturella omgivningen och personliga faktorer för att främja delaktighet och förebygga risker. För att öka dessa personers delaktighet och behov av skydd så behöver de individuellt anpassat stöd och hjälpmedel.

Dessa utmaningar påkallar utveckling av lagstiftning, service management och service i direktkontakt med personen. FN:s konvention om rättigheter för personer med funktionshinder inkluderar grundläggande principer för att säkra liv och full delaktighet vilket innebär skydd, främjande och individuell utveckling. ICF och FN konventionen understödjer de nödvändiga förändringarna.
INTRODUCTION

It began with a letter.

“We are a group of deafblind who live in... We experience a lot of problems and difficulties which we believe can be avoided if you, the decision makers know more about us and our problems. We also believe that the municipality and the county council need to collaborate in questions that concern us in order to get know-how and thereby be able to come up with good solutions.”

(Citation from a letter sent to a municipality and a county council, September 15th 1995. Translated by G. Asplin)

This letter was sent to different authorities that perform service for people with deafblindness. It became the starting point for a number of investigations (Möller, K. 1999, 2001, 2002; Möller & Samuelsson, 1998).

The citation is interesting in several aspects. Experiences of problems and specifically problems relating to, service, lack of knowledge and lack of collaboration. The writers present themselves differently according to the two impairments, which indicate that impairment may have different meaning for them. The object of this thesis is to elucidate these issues in different ways.

Deafblindness

In everyday terms, deaf and blind denote individual impairments and deafblind denote combination of the impairments. There are however other definitions of deafblindness.

Three-sensed and multi-sensory-deprived

Historically, the primary education system in Sweden, referred to children with deafblindness as “three-sensed” (Liljedahl, 1993), which emphasised the senses left. In the eighties McInnes & Treffry (1982) tried to replace the word deafblind with multi-sensory-deprived (MSD). Denoting deafblindness solely by degree of impairment is in general not used any more.

Handicap or disability

Among professional’s deafblind denote a certain “handicap” or disability (Aitken, 2000; Fredericks & Baldwin, 1987; McInnes & Treffry, 1982; Wolf-Schein, 1989). The Nordic countries had between the years of 1980–2006 a common definition of deafblindness of this type. The Swedish Association of the Deafblind (http://www.fsdb.org
Disability that require specific service etc.

Thirdly in 2007, the Nordic Staff Training Centre adapted a new Nordic definition for Deafblind Service (NUD) as follows:

“Deafblindness is a distinct disability. Deafblindness is a combined vision and hearing disability. It limits activities of a person and restricts full participation in society to such a degree that society is required to facilitate specific services, environmental alterations and/or technology” (www.nud.dk [accessed 22 March 2008]).

This definition includes five clarification comments, which briefly extracted highlights the following issues: the importance of vision and hearing in getting information; the need for specific alteration depends on many factors; that the degree of disability may vary in different situations; varying needs for co-creating alteration; and that specific know-how related to deafblindness is needed in service delivery and environmental alterations (ibid.).

Alternative way of perceiving the world

Finally, based on self-reports within deaf studies, deafblindness is regarded to be an alternative way of perceiving the world, though often as “isolated island” (Barnett, 2002). Self-reports by persons with deafblindness show strategies and habits used of “negotiating a place in a hostile world” (Schneider, 2006).

Heterogeneity of the group

The group may be limited due to degree of impairment, degree of daily impact or according to regulations of the service (Aitken, 2000; Fredericks & Baldwin, 1987; McInnes & Treffry, 1982; Wolf-Schein, 1989). The group is very heterogeneous and is often subdivided according to age of observed impact. In pre-lingual onset impact of both impairments comes before or during development of language skills. Professionals usually call this subgroup congenital deafblindness. Post-lingual onset is often among professionals called acquired deafblindness. The third group is elderly people that have got combined visual and hearing impairment at old ages. Persons regarded to have deafblindness rarely have complete loss of both vision and hearing (Wolf-Schein, 1989).
Etiology of combined visual/hearing impairment- deafblindness

Combinations of visual and hearing impairment are caused by a number of heterogeneous diseases and disorders. Visual and hearing impairment is the most common dual sensory impairment, and 30 percent of children with hearing impairment have been found to have visual impairment (Nikolopoulos et al. 2006).

Pre-lingual deafblindness is extremely rare. (1 in 10.000) (Möller, C. 2007). Genetic syndromes, premature birth, congenital virus infections, are the most common causes. At least 20 different genetic syndromes are known to cause pre-lingual deafblindness. Some of which have been genetically identified (Möller, C. 2007). The rarity of these conditions and difficulties in assessment increase the risk of wrong diagnosis, which also may be “hidden” due to other dysfunctions and, thus attributed to other conditions (McInnes & Treffry, 1982; Möller, C. 2007).

Developing severe visual and hearing impairment (post-lingual deafblindness) later in life is also rare. The aetiology of post-lingual deafblindness is as in pre-lingual most often genetic. More than 50 hereditary syndromes are known to cause acquired deafblindness, in 40 syndromes the gene has been localised and in 20 syndromes the gene has been cloned. (Möller, C. 2007).

Usher Syndrome

Usher syndrome (USH) is a genetic disorder with autosomal recessive inheritance. USH is the most common cause of deafblindness before older ages (Kimberling & Möller, 1995; Sadeghi, 2005). The syndrome is divided in three distinct clinical types (type I–III). Different gene mutations and clinical features distinguish these types.

USH affects the structure of the cochlea, the vestibular organ (type I and III) and the retina bilaterally (Kimberling & Möller, 1995).

In the inner ear (cochlea and the labyrinth) the hair cells are damaged. In the eye (retina) the rod and cone cells are gradually undergoing degeneration/deviation. The disorder in the eye, Retinitis Pigmentosa (RP), comprises several genetic disorders affecting the retina (Hartong et al. 2006).

USH type I is associated with profound hearing impairment (deafness) while type II and III are associated with moderate to severe (type II) and in type III usually progressive (Kimberling & Möller, 1995; Sadeghi et al. 2004).

Vestibular function is absent bilaterally in USH type I. This will result in delayed walking age (> 18 months) and clumsiness especially in dimly lit situations or darkness. (Möller, C. 2007) Type II has normal vestibular function while type III has progressive loss of vestibular function.

All three types of USH entail progressive visual impairment (Kimberling & Möller, 1995). In USH the deviation starts in the periphery where the rod cells are situated.
Cone cells situated in the centre are affected later (Möller, C. 2007). RP in USH has been demonstrated to have a rather slow progression (Sadeghi et al 2006). The RP in the three types of USH do not differ as much as auditory or vestibular function. In many cases the progression of visual impairment can be the same in type I, II and III. There are however in large materials some variations (see below).

The RP in Usher includes different visual impairments such as glare sensitivity and night blindness both due to impaired light adaptation and impaired contrast sensitivity. Visual field impairment follows the degree of retinal deviation. Thus, since light adaptation is also impaired, the limitations in seeing broad views in soft light become worse than in fitful light. Furthermore glare sensitivity is a hinder in very light environments. Thus, light may cause a barrier both when too soft and bright. The first symptom of visual impairment in USH is known to be night-blindness. Visual field impairment, which starts in the midperiphery, occurs later with a gradual loss of visual acuity. A common sequel (> 80%) is sub capsular cataract (Sadeghi et al 2006).

Between the ages of 30 and 40 the visual field is usually restricted to 10 degrees, and visual acuity is 0,5–0,3 (Sadeghi et al 2006). The visual acuity can remain normal even in individuals with advanced RP with a small island of remaining visual field (Hartong et al 2006). Visual acuity < 0,1 (legal blindness) is estimated to be 25% at the age of ≈ 50 and 50% at the age of ≈ 60 (Sadeghi et al 2006). The likelihood of losing total vision in both eyes is rare in USH although these patients may present with severe visual impairment (Grover et al 1999; Hartong et al 2006; Sadeghi et al 2006). Decline in contrast sensitivity can account for poor subjective vision in those people who have good high contrast visual acuity (Lodha et al 2003). The visual impairment has been found to be worse and increase with age in people with USH I compared to those with USH II (Edwards et al 1998; Pennings et al 2004; Sadeghi et al 2006). Visual impairment in USH III has also been described (Plantinga et al 2006). No evidence of other major organic dysfunctions has been presented during the last 30 years.

In conclusion; USH I involves congenital profound deafness, absent vestibular function (poor balance) and later in life (from 4–5 years) symptoms of retinitis pigmentosa (RP).

Nine different genes associated with USH have so far been identified in Online Mendelian Inheritance in Man (OMIM), see table 1.

<table>
<thead>
<tr>
<th>Locus name</th>
<th>Locus</th>
<th>Gene</th>
</tr>
</thead>
<tbody>
<tr>
<td>USH1B</td>
<td>11q13.5</td>
<td>MYO7A</td>
</tr>
<tr>
<td>USH1C</td>
<td>11p15.1</td>
<td>Harmonin</td>
</tr>
<tr>
<td>USH1D</td>
<td>10q21-q22</td>
<td>Cadherin-23</td>
</tr>
<tr>
<td>USH1E</td>
<td>21q21</td>
<td>Unknown</td>
</tr>
<tr>
<td>USH1F</td>
<td>10q21-q22</td>
<td>Protocadherin 15</td>
</tr>
<tr>
<td>USH1G</td>
<td>17q24-q25</td>
<td>SANS</td>
</tr>
<tr>
<td>USH2A</td>
<td>1q41</td>
<td>Usherin</td>
</tr>
<tr>
<td>USH2B</td>
<td>3p24.2-23</td>
<td>Unknown</td>
</tr>
<tr>
<td>USH2C</td>
<td>5q14.3-21.3</td>
<td>VLGR1</td>
</tr>
<tr>
<td>USH3</td>
<td>3q21-q25</td>
<td>Clarin-1</td>
</tr>
</tbody>
</table>

The overall prevalence of USH in Sweden has been estimated to 3.3/100000 (Sadeghi et al 2004) and is thus the most common cause of deafblindness (Möller, C. 2007). A global prevalence of USH is estimated to be 250,000 and in Sweden the estimated prevalence of Usher syndrome is 600 individuals of whom 250 have USH I.

USH is so far incurable. Today habilitation in children with USH I have changed dramatically with the introduction of Cochlear Implant (CI). Children with USH I in Sweden are fitted with 2 implants at the age of 6–12 months. The aim is to hear spoken language and to develop speech. From around 1997 nearly 95% of all children with congenital profound deafness (including USH I) have received cochlear implants (Möller, C. 2007). Treatment of RP have so far been large doses of Vitamin A, which have been reported to slow down the degeneration of retina (Berson et al 2004a, 2004b; Hodge et al 2006). Rapid progress of gene identification and cloning might in the future lead to medical and gene therapy (Möller, C. 2007).

**From handicap in ICIDH to functioning in ICF**

Attitudes towards the impact disease and injuries have in daily life have altered during the space of time, this is roughly outlined from 2005 and 50 years back. There have been changes from an individual bodily focus to relationships between the body and the context e.g. environment and personal factors.

In the sixties in developed countries, it was observed that health care cost increased despite the fact that the rate of serious diseases decreased and large vaccination programs eradicated some diseases. It became apparent that people could live longer with diseases and disorders (WHO, 1980). Information from diagnosis does not predict many of the important health care and service planning outcomes that health planners need to know in order to make informed judgments about resource allocation and cost-effectiveness (Üstün et al 2003 p. 4).
The first attempt to create an international classification of certain consequences of diseases was taken by WHO and especially Dr Wood, a rheumatologist in Manchester, UK. This was an important event that increased theory building in disability (Nordenfelt, 2003). The name of this classification was; International Classification of Impairments, Disabilities and Handicaps (ICIDH), which was published by WHO in 1980, but never endorsed.  

In the introduction to ICIDH, WHO write:

“The simplest requirement of a health care system is that some beneficial change in the individual’s situation or status should result from contact with the system. If no such change can be detected, then the value of a given health care process is seriously open to questions.”


WHO raised a further three critical questions, also based on Cochrane’s writings on effectiveness and efficiency in health service: 1. What are the full ranges of problems that lead people to make contact with a health care system? 2. How does the system respond to the contact? 3. What is the outcome of the contacts?

These were important questions since the medical model of health care failed to answer these questions (WHO, 1980). Hope for solving these problems was placed on ICIDH. Problems people with illness experience were according to ICIDH regarded to be caused by disease.

“Sickness interferes with the individual’s ability to discharge those functions and obligations that are expected of him. In other words the sick person is unable to sustain his accustomed social role…”


In this model it is necessary that an individual have a disease or disorder, which is the starting point.

ICIDH is furthermore a model that is based on hierarchical and causal conceptions see Figure 1.
FIGURE 1 Based on ICIDH (WHO 1980). ICIDH should be seen as a process where the entrance has to be disease or disorder.

The impairment level is concerned with abnormalities of body structure and appearance and with organ or system function, resulting from any cause; in principle, impairments represent disturbances at the organ level. The disability level reflects the consequences of impairment in terms of functional performance and activity by the individual: disabilities thus represent disturbances at the level of the person. The handicap level concerns the disadvantages experienced by the individual as a result of impairments and disabilities; handicaps thus reflect interaction with and adaptation to the individual’s surroundings (WHO, 1980 p.11). The notion of handicap in ICIDH reflect disadvantage in relation to fulfill one’s normal role in life. ICIDH represents images of the able body to be normative in the meaning that it is better than the body with impairment.

The definition of disability in the Americans with Disabilities Act (ADA) from 1990 is an example of this kind of disability concept. It defines disability as:

“…a physical or mental impairment that substantially limits one or more of the major life activities…” (Americans with disabilities act, 1990 http://www.ada.gov/pubs/ada.htm#Anchor-Sec-47857 [accessed 22 March 2008])

ICIDH was the subject of debate (Nordenfelt, 1993; Oliver, 1996; Söder, 1989) and given severe criticism especially by the disability rights movement because of the normative use of the term handicap (Hurst, 2003).

By 1993 WHO initiated a revision process of ICIDH (Bickenbach et al 1999; Üstün et al 2001). The process lasted for 7 years and included several field trials in various cultures. Finally by 2001 when WHO endorsed a new model of human functioning including disability: International Classification of Functioning, Disability and Health, known as ICF (WHO, 2001). This classification is a radical shift from emphasizing
people’s diseases to focusing on their level of functioning in all aspects of human health and some health-relevant components (WHO, 2001 p.8) ICF encompass possibilities to describe both positive and negative aspects of different health domains.

ICF is a multipurpose classification, which may be used in various disciplines. One of the aims is to provide a scientific basis for understanding and studying health and health-related states, outcomes and determinants (WHO, 2002).

ICF is based on the presumption that variation of health and illness during life is human common, named as universalism (Bickenbach et al 1999; Üstün et al 2003). During life the health states vary for all humans (Zola 1989). Therefore ICF is not only about people with disabilities, it is for all people. The properties of ICF are supposed to encompass all aspects of human health and some health-relevant components of well-being (WHO, 2001).

WHO stress that ICF does not classify people but provides possibilities to describe situations with regard to human functioning. In order to describe situations environment and personal factors is included (ibid.).

ICF is based on an integration of the medical and the social model. ICF is expressed to be a bio-psycho-social approach in order to capture various perspectives of functioning. ICF is further a complement to the medical perspective (Üstün TB et al 2003; WHO, 2001).

ICF consists of two parts with four and two components respectively. Components in the first part are: body structure; body function; activity and participation. Components in the second part are: environment and personal factors.

Body structure is anatomy e.g. ear and eye. Body function includes both physical and psychological functions e.g. hearing and vision. In the classification these two components share the same negative term, which is impairment. In this study, in order to distinguish between negative aspects of body structure and body function, the term deviation signifies negative aspects of body structure. Activity is about doing. What a person with a health condition performs is what he/she actually does and capacity is what a person might do in an adjusted environment. Activity may for example be to listen or hear and to watch or see. Participation in ICF is a person’s involvement in a

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1 In ICF one term, impairment, signify negative aspects of both body structure and body function. This makes it more difficult to distinguish mechanisms that influence the relationships between body structure and body function. In this thesis this is important, therefore the term impairment will be used to denote negative aspects of body function. The term deviation will be used to denote negative aspects of body structure. In ICF section 4.1 item 5 the term deviation is used (WHO, 2001 p.16) and in Annex 1, (ibid. p.190) the term abnormal is used. Abnormal is here avoided since the term ‘normal’ in the relation to disability have been criticized for being used in a normative way.
life situation and represents the societal perspective of functioning. WHO propose that participation incorporate “taking part, being included or engaged in an area of life, being accepted, or having access to needed resources” (WHO, 2001 p.19), which for instance may be to take part in audible and visible information. “Information that reflects the person’s feeling of involvement or satisfaction with the level of functioning is currently not coded in ICF” (WHO 2001 p.206), which is relevant since this is information related to the personal component. The concept of participation will be further elucidated below. The negative terms are *activity limitation* and *participation restriction*.

Two umbrella terms encompass all components in the first part. *Functioning* denotes positive aspects and *disability* denotes negative aspects.

The second part is *context*. Environment include physical, social and societal environment. In the physical environment there are products such as hearing aids and glasses. In the natural environment there are e.g. sound and light. The environmental component can also be used to describe support, relationships and attitudes, which are social matters. ICF also entails societal environmental tasks such as service, systems and policies. Factors in environment may play a positive role, which is labeled *facilitate* as well as play negative roles, which is labeled *barrier*.

Personal factors are e.g. age, gender, background, past and present experiences, coping style etc (WHO, 2001 p.23-24). The personal factor component is not included in the coding system and does not have a negative term.

Though ICF is not a theory, it has been used in research and is gradually accepted as a concept in health and disability issues (Bales et al 2006; Bruyère et al 2005). There have however been some critical remarks directed at different studies. The most important so far has lead to development of ICF adapted for children and youth (Simeonsson, 2003; Simeonsson et al 2003).

The international field trials during the revision process showed cultural diversity concerning activity and participation and difficulties to distinguish between individual and social perspective (Room et al 2001 p.338). Difficulties to discriminate between activity and participation were elucidated because performing something in general includes involvement in the task. ICF therefore include possibilities to use activity and participation components together or separately. Perenboom and Chorus (2003) measured 9 existing survey instruments in relation to participation in ICF. Their study revealed that more discussion is needed in order to be able to get an unambiguous picture to distinguish between activity and participation.

Several studies have elucidated experience of participation and conceptions of participation to be a component of its own. For instance, Eriksson and Granlund (2004) define participation as consisting of three dimensions: perceptions of participation, activity/behavior, and pre-requisites for participation. Pre-requisites are viewed as factors
in the environment. Furthermore, they argue that participation and self-determination can be viewed as partially overlapping. They write that self-determination includes both the inner characteristics of a person, and the actions of that person.

Student’s conception of participation depended not on the type of disability, with one exception: deafblind students (Eriksson & Granlund, 2004). Persons with deafblindness were significantly distinguished from other groups with disabilities in three aspects. Students with deafblindness mentioned more frequently self-determination and conditions in the living environment as well as pre-requisites for taking part in activities as a part of their conceptions of participation than did other groups. In another study (Almqvist & Granlund, 2005) pupils with disabilities, their teachers, parents and special education consultants responded to questionnaires about participation. Results indicate the total number of positive factors rather than a single special factor contributes to participation. Among the different factors, interaction, autonomy and locus of control, availability in the environment and participation in activities in school were mentioned.

The concept of participation was analyzed in the domain of upper secondary school education for pupils with intellectual disabilities (Molin, 2004). His study covers social and societal level of participation. At societal level, school officials show different approaches towards their students. The “qualification perspective” was to obtain a good life as an adult, which was to learn to comply with the demands regarding qualification in society. The other perspective Molin labeled the “security perspective”. In this perspective the pre-assumption was that the pupils should be taken care of with a day activity center in mind. Molin conclude as long as the pupils are contented and there is an easily accessible path out into a day activity centre, there is no need for these pupils, with intellectual disabilities, to confront external circumstances.

At the social level of participation Molin (ibid.) showed that pupils exhibited different patterns of participation. This could for instance be solitary activities or interplay. Also in relation to the teacher and other pupils, some pupils communicated more often with the teacher whilst others more often communicated with one another. Regarding involvement, Molin showed differences in what the pupils were involved. Regarding autonomy and power, some pupils exhibited the will and capacity to make independent choices, but this was sometimes prevented by external conditions i.e. negative attitudes of persons in authority. Molin did not examine intellectual capacity or body function within the students. However, results indicate that those with higher degree of intellectual capacity behaved in ways that indicate that they didn’t want to belong to this group but to those in “the big school”. Molin emphasizes complexity in the concept of participation.
In ICF it is stated that ICF as a classification does not model the “process” of functioning and disability (WHO, 2001 p.25). It is however possible to use ICF for description of processes since the model provides a multi-perspective approach in an interactive and evolutionary process.

WHO gives some examples of how ICF can be used as a process model. “A person’s functioning and disability is conceived as a dynamic interaction between health conditions (diseases, disorders, injuries, traumas etc.) and contextual factors” (WHO, 2001 p.10). This interaction can be viewed as a process or a result depending on the user. The WHO figure of ICF (Figure 2) shows arrows between the components indicating that there may be mechanisms.

**FIGURE 2 Interactions between the components of ICF.**
(WHO, 2001 Short version p.26.)

**Service**

Service is used as an umbrella term covering public service in Sweden and in particular health care, education and support and services for persons with certain functional impairments.

**Health care**

Different approaches towards disease and health have impact on health care for persons with deafblindness.
One approach starts with disease and is in ICF labeled medical model (WHO, 2001 p. 28). This approach is the basis for diagnosis, treatment and cure. Different parts of the human body function together but can be studied and treated separately (Medin & Alexandersson, 2000). Diseases in this sense are distinguished in medical classifications (WHO, 1992) mainly according to the sequence from etiology to pathology to manifestation (WHO, 1980). Professionals are prepared to ask patient about symptoms, execute relevant examinations, make evaluative statements and treat the body if treatment is available. Such evaluation requires comparison to established norms (Arnold & Janssen Breen, 2006). Disease is equal with not normal functioning bodily organ. “Health is normal functioning where the normality is statistical and the function is biological” (Boorse, 1977 p.542).

A second approach entails more than disease or absence of disease. That is health, which is valued to be something more. There are different denotations of health. WHO’s definition:

“Health is a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity.”

http://whqlibdoc.who.int/hist/official_records/constitution.pdf [accessed 25 August 2007]

This approach is normative since it regards health to be something positive and disease negative, that is undesirable or bad (Nordenfelt, 2007). Holistic approach to health is more than the sum of its pieces. Furthermore, the pieces must be understood in relation to the whole (Medin & Alexandersson, 2000).

The Hospital Act (SFS 1959:112) and earlier legislation of health care exclusively included institutional care. The Medical Care Act (SFS 1962:242) regulated the county council’s duties concerning medical care of disease and injury. The present Health and Medical Services Act (HsL) (SFS 1982:763; Health and Medical services act 1982 1§) states: “The term health and medical services in this act refers to measures for the medical prevention, investigation and treatment of disease and injury.”

The goals of HsL (SFS 1982:763; Health and Medical services act 1982) are expressed as follows: “Health and medical services are aimed at assuring the entire population of good health and care on equal terms.” This indicates a more inclusive approach than the first paragraph. The following three services are included in county council’s responsibility:

1. Habilitation and rehabilitation;
2. Assistive devices
Interpreting services for everyday situations for persons deaf from childhood, the deafblind, persons becoming deaf during adulthood and persons with hearing impairment.

In preparation of HsL a holistic approach was in the bill proposed “to consider the patient as a whole” (Prop. 1981/82:97 Hälso- och sjukvårds lag, m.m. [bill: Health- and Medical Care Act etc.], 1982). This was however rejected by the parliament.

Obligations to inform the patient about his/her health condition and state was a new approach (ibid. p.2), in line with the policy of transferring responsibility for the disease from the professional to the individual (Serner, 1980).

The main structure of medical health service is provided by local self-government (county councils and municipalities) with a considerable degree of autonomy vis-à-vis the state but regulated by legislation, the Health and Medical Service Act.

Health care concerning diseases that cause visual impairment or hearing impairment is usually provided as outpatient care at specialized departments. The mission of the Department of Ophthalmology (OD) is to provide medical treatment. Rehabilitation of severe visual impairment and administration of assistive devices is delivered at the Low Vision Clinic (LVC). Service by LVC requires referral from OD. The Audiology Department (AD) is responsible for the medical treatment, rehabilitation as well as administration of assistive devices. Thus unify medical treatment and rehabilitation.

**Education**

Swedish policy to educate in basics and foster citizens goes back to 1686 and this policy was intimately connected to the Swedish Lutheran church. In 1842, the state was given ultimate responsibility for fostering and education of the citizens. This was an act, which regulated the municipalities’ responsibility to provide education rather than an act for compulsory school attendance (Pärsson, 1997). The current legislation regarding education is from 1985. Its policy is for there to be a school for all (SFS 1985:1100, 1985; Education act, 1985). Both the Education Act and curriculum for education in compulsory school and upper secondary school include students with impairments. Children with deafness have been included in the compulsory education system in Sweden since 1889 (Pärsson, 1997) and children with blindness were included from 1896 (Liljedahl, 1993).

Due to advances in medical science conceptions arose that all diseases were curable or that the people could be rehabilitated and this lead to specific education for pupils with impairments. Special education for students with impairments aimed to foster the child into Christianity. The idea was that the child’s problems should decrease and thereby lessen the economic burden on society. The child was the problem and the individual was going to be made normal (Peterson, 2000). This idea has the same ideologi-
cal base as handicap in ICIDH described above. Management of the special schools was a responsibility for county councils and subsidized (SOU 1998:66, 1998). Schools for the deaf that were earlier a responsibility for the county council was transferred to the state 1938 (Pärsson, 1997).

Education for students with deafness was initially performed in sign language. The policy changed however to oral method in Sweden and several other European countries. This was due to influences from the Congress of Teachers of the Deaf in Milan 1880. The idea of the oral method was to make children with deafness normal, in normative sense, with hearing persons as the norm. The oral method aimed at teaching students to talk and to lip-read what other people said and sign language was in generally prohibited. In practice some teachers turned a blind eye to the fact that deaf children signed, especially during 1970–1980 where some teachers began to reinforce their speech by performing some signs. This signed Swedish, technique follow Swedish and not sign language grammar. In 1981, sign language was recognized and education for students with deafness was supposed to be given in sign language. Students with hearing impairment are reinforced with assistive devices and signed Swedish if required (Pärsson, 1997).

Other services for persons with disability

In ancient time persons with disability, who due to their impairment could not earn a living, were regarded as poor and sometimes worthy poor, with the right to beg. In the countryside the poor were moved from farm to farm or were disposed of by auction and the pauper was expected to do one’s share of work. This system was repealed 1918. County councils were obliged to set up institutional care for the crippled, with Government grants. Within institutional care, control and rehabilitation was integrated but vis-à-vis society as a whole it was segregated. An important political shift was the development of the Swedish Welfare State from about 1920. It contained major government responsibilities for people with difficulties. Critics argue that the Welfare State became oppressors rather than realizing solidarity, equality, security (Lindqvist & Hetzler, 2004).

The Scandinavian normality policy was developed during the fifties and sixties, which proposed to reorganize the institutions and aimed to facilitate a normal life for people who not were normal. The purpose was not to transform the persons but to make alterations in the environment instead (Tideman, 2000). This was the beginning of the relative handicap denotation (Söder, 1989).

In Sweden, the word handicap has not been associated with normal in the normative sense. It has instead been used to signify the relative meaning, which calls for altera-
tions in the environment in order to make it possible to live a life like others e.g. out from the larger institutions mentioned above.

The Swedish policy regarding persons with disability was in the Welfare state’s legislation legal in Social Services Act (SoL) from 1980 and updated 2001 (SFS 2001:453, 2001; Social Services Act, 2001). The municipality is ultimately responsible for ensuring that persons staying within its boundaries receive the support and assistance they need (ibid. 1:2§). Persons unable to provide for their needs are entitled to assistance from the social welfare committee (ibid.). Individuals should through assistance from SoL be assured of a reasonable standard of living (ibid. 4:1§). People who, for physical, mental or other reasons, encounter difficulties in their everyday lives should be enabled to participate in the life of the community and to live like others (ibid. 5:7§). Entitlement in the later section points towards a higher degree of quality of life than the previous.

The act concerning support and service for persons with certain disabilities (LSS) (SFS 1993:387, 1993; Act Concerning Support and Service for Persons with Certain Functional Impairments, 1993) states rights for persons while SoL states certain responsibilities of the municipality. LSS shall promote equality in living conditions and full participation in the community life (ibid. section 5).

The special support and service in LSS is delimited to include those who: 1. are mentally retarded or have a condition resembling autism; 2. has a considerable and permanent intellectual functional impairment after brain damage when an adult, the impairment being caused by external force or a physical illness or; 3. has some other lasting physical or mental functional impairments, which are manifestly not due to normal ageing, if these impairments are major and cause considerable difficulties in daily life and, consequently, an extensive need for support or service.” (ibid. section 1). LSS entails ten different services:

1. Advice and other personal support;
2. Help from a personal assistant or financial support for such help (SFS 1993:389, 1993; Assistance Benefit Act, 1993);
3. Escort service;
4. Help from a personal contact;
5. Relief service in the home;
6. Short stay away from the home;
7. Short period of supervision for schoolchildren over the age of 12 outside their own home in conjunction with the school day and during the holidays;
8. Arrangements for living in a family home or residential arrangements with special service for children and young people who need to live away from their parental home;
9. Residential arrangements with special service for adults or some other specially adapted residential arrangements for adults; and
10. Daily activities for people of a working age who have no gainful employment. (This service is only available for circle 1 and 2) (SFS 1993:387, 1993; Act Concerning Support and Service for Persons with Certain Functional Impairments, 1993)

Personal assistance is a personally designed service provided by a limited number of professionals and where the person themselves has a great degree of influence in the service. It is however limited to those who need assistance with personal hygiene, meals, dressing and undressing, communicating with others or other help that requires extensive knowledge about persons with a functional impairment (basic needs).

There are further services available for people with disabilities: transportation service; adaptation in the home, economic support etc., which not will be elucidated in the Studies.

**Present state of knowledge**

Deafblindness denotes different matters: the impairments, handicap and disability or an alternative way of perceiving the world. None of these denotations is based on an ecological approach.

Restriction in participation for persons with deafblindness has been reported in children/youths, people of working age, elderly and for persons with pre-lingual as well as persons with post-lingual onset of deafblindness. One of these studies uses ICF (Crews & Campbell, 2004). Relationships between body and activity and between activity and participation in certain contexts have however not been explored.

Barriers in services for persons with deafblindness have been reported in different life periods and for both onset types. However studies concerning relation between service barriers, service policy and service management were not found. Few papers relate service barriers to problems in the interaction between persons with deafblindness and service for the specific disability (Giangreco, 2000; Petroff, 1999). Restrictions in participation and barriers experienced by persons with deafblindness have not been reported from a life course perspective. Neither have any disability studies that combines ICF and life course perspective been found.

This means that there is very little if any knowledge about mechanisms that restrict participation for persons with deafblindness and mechanism that barrier service to these people.
**AIMS**

The general aims of the thesis are to discover, evaluate and explain:

1. mechanisms that might have impact on participation restrictions for people who have visual and hearing impairment i.e. deafblindness and
2. mechanisms that might barrier service to these people.

The aims of the five Studies are:

**Study I** To given an account of the psychosocial effects of deafblindness in a life perspective.

**Study II** To highlight some examples of the strengths and limitations of ICF, that have been found when studying people with deafblindness

**Study III** To describe factors that pose barriers to participation in education in upper secondary schools for students with deafblindness, from the student’s perspective.

**Study IV** To explore ophthalmic health care in female patients with USH I during a time period of 20 years and to evaluate the relationship between the ophthalmic health care and functioning and disability of the patients from an integrated health perspective.

**Study V** To discuss mechanisms that have impact on participation in a life course perspective in some females with USH I in relation to rehabilitation and support.
THEORETICAL CONSIDERATIONS

Theories in science guide the researcher into what to study. The choice of theory was not clear in the beginning of this study. Different theories were deliberated. Charmaz (2007) encourages researchers to consider a range of theoretical ideas and then choose the one that offer the best fit.

Human senses in an ecological approach

Human beings are always situated in an environment (Gibson, 1966, 1979). Waves in the environment are physical phenomena and some waves provide light, which is necessary for visible features. Sound is another type of physical wave caused by events in the environment. Human beings have sensory systems that make it possible to perceive oneself as well as the environment. Eyes, ears and other anatomical parts are included in the whole body structure system that is the ground for the body function system both of which interact with the environment. The possibility to perceive something visible or audible depends however on both conditions in the environment and factors within the person. Environmental factors that have an affect are the degree and quality of the signals. The receiving person is required to have a certain level of physical body fitness with respect to their vision and hearing organs that receive signals as this affects the condition of sight and hearing functions. It is not enough having the ability to receive signals from the surroundings but also necessary to be able to process and interpret them. By doing so the visual and audible signals are made comprehensible in a cognitive process.

Visible information becomes significant and the individual discover the environment from distinct patterns and by dynamic alterations (ibid.).

Eysenck and Keane (2000) distinguish between down-to-up-processes i.e. processes caused by external stimuli and up-and-down-processes, caused by the individual his or her self. Perception is information gained by the sensory body structure and transformed to experience of objects (ibid.) e.g. to see and hear another person who says hello. To see and to hear is hence interaction between the human body including its functions and features in the environment (Lundh, 1992). Visible and or audible information that has been perceived is then transmitted from the receiving eye or ear by the afferent nerve system up to the brain. The information is processed in the cognitive system, which is to recognize, remember etc in order to make sense of the visible or audible features.
To see or hear is to perform *activities*. What a certain individual sees or hears in the situation depends besides the general conditions mentioned above but also on necessary and or possible environmental alterations e.g. glasses, hearing aids that *facilitate* and on *personal factors* such as willingness to see and hear. What people do and are involved in include features, which have visual (Gibson, 1979) and or audible (Noble & Hetu, 1994) significant meaning. The individual gives perceived sounds meaning. In this process sounds are identified based on alteration in frequency, loudness and sometimes alteration in location (sound source move) as well as context such as time and previous experiences. If the person wants to see and hear and is able to see and hear, then there is a possibility to understand the significance in the visible and audible information. That is to participate in the information gained from what the person has seen and heard.

### Disability the necessarily laminated system

Within the meta-theory system critical realism (Bhaskar, 1978) disability has been the fore of some studies (Danermark, 2002; Williams, 1999). Thomas (1999b) discusses disability from a feminist approach. Her views on disability are close to critical realism even if she never uses the word (Bhaskar & Danermark, 2006).

Bhaskar and Danermark (ibid.) have developed a disability theory, which they call a necessarily laminated system. Bhaskar and Danermark regard disability to be a system that essentially refers to several different levels of reality. They also take into account that multiplicity and complexity derives from multiple contexts of sociality and that deriving from scale of social being. Social events are then understood in terms of four dimensions:

“1. material transactions with nature;
2. social interactions between agents;
3. social structure proper; and
4. the stratification of embodied personalities of agents.”

(Bhaskar & Danermark 2006 p.289)

They mean that all social events involve each of these dimensions and therefore social events and social systems in general are laminated. Furthermore explanations involving mechanisms at several or all of these levels are laminated explanations. All dimensions do not necessarily have to be mentioned every time, though they are there and have potential impact. They argue that it is important to understand and have in mind that there are in reality a large number of mechanisms contributing to the situation otherwise a risk of a fragmentary understanding of the situation. They call this “A non reductionist approach”. They further stress that order of sequence and geo-history is
also important. The layers in disability sciences are bodily, psychological and social levels. Although, these layers can in turn be further divided. The relative importance and specific role of mechanisms varies and their actual role is always an empirical question (Bhaskar & Danermark 2006).

Life course

To study life course means to study populations and individual life over time in relation to cultural and societal phenomena located in time and space. Process is the central element and is defined in terms of its functional relationship to the context and person. This perspective is a way to conceptualize the interaction of age, period, place and cohort. Giele and Elder (1998) mean that life course science comprises a combined social system approach that makes it possible to address person dynamics and system dynamics simultaneously. Priestley (2003) understands life course approach in research to be a central organizing principle in modern societies, rather than as a description of individual life careers.

Life course approach includes five components, labeled as themes. The themes that are connected to each other in an interactive relationship that may help to understand life course in a social way. The themes are: location in time and place; linked lives; human agency and timing. Location in time and place refers to the social structure and culture. Linked lives are the result of the interaction of individuals. Human agency is embodied in the active pursuit of personal goals and the sense of self. Timing covers the chronologically ordered events of an individual’s life that simultaneously combine personal, group and historical markers (Giele & Elder Jr, 1998).

This scientific approach was introduced in the 1960s and was controversial. Communication was impeded by difficulties of translating ideas across field. Later on the debate was about different ways of using the same terms. The term development meant different phenomena in different sciences. The claim in life course approach that development is historically and culturally contingent and that individual’s adaptation takes different forms is no longer so controversial at is was (Colby, 1998). Developing life course, research were generally focused on a macro perspective that is the whole society. Later on, life course approach has also been applied on a micro level using small groups or face-to-face relationships (Giele & Elder Jr, 1998).

Life course perspective in disability research is a way to, for instance, study relationships between educational policy and persons who were educated during that educational policy. The role of social institutions is to manage and support successful transition from one life stage to the next. Welfare institutions can be seen as societal response for reproducing new generations and ensuring their successful passage through
life. These responses include managing and containing those who ‘fail’ to make ‘proper’ life course transitions. The underlying idea is the ‘normal’ life course. In the case of disability it is important to elucidate the course of these institutions and the impact they have on individual lives (Priestley, 2003, Corker, 2001).

To sum up, the ecological approach and the necessarily laminated system of disability are used to analyse mechanisms and their role in creating restrictions in participation and barriers in service to persons with deafblindness. Furthermore, in this study the point of departure for analysis is the individual, i.e. a bottom-up approach. The societal systems’ impact on services and persons with deafblindness in a life course perspective lay to ground for analysis of the relations between deafblindness, service and societal system taking time and place into account. This means that the study is rather inspired by life course approach than a complete life course study.
MATERIALS

Materials were selected in several steps by strategic selection (Merriam, 1988). Table 2 below shows type of material, data collection, criteria for inclusion and number of papers/informants that were included.

TABLE 2 Materials in Study I–V.

<table>
<thead>
<tr>
<th>Type of material</th>
<th>Number of possible sources</th>
<th>Cause of exclusion</th>
<th>Remaining</th>
<th>Used in the Study</th>
</tr>
</thead>
<tbody>
<tr>
<td>Study I</td>
<td>Knowledge review</td>
<td>&gt; 400</td>
<td>Did not fulfil criteria = 200</td>
<td>227</td>
</tr>
<tr>
<td>Study II</td>
<td>Interviews on two occasions, (Questionnaire)</td>
<td>56 of whom 3 were not invited</td>
<td>14 did not respond 6 declined</td>
<td>33 persons 1 declined interview</td>
</tr>
<tr>
<td>Study III</td>
<td>Posted questionnaire</td>
<td>60 persons in estimated population,* of these 46 persons were traced (77%)</td>
<td>4 respondents did not fulfill criteria</td>
<td>34 persons</td>
</tr>
<tr>
<td>Study IV</td>
<td>Patient records</td>
<td>Females with confirmed diagnosis of USH I in age cohort born 1946–1960 = 21 persons</td>
<td>12 patient records not complete from both OD and LVC during 1985-2004</td>
<td>9 patient records from two departments = 18 records during 20 years = 910 notes</td>
</tr>
<tr>
<td>Study V</td>
<td>Interviews at 3 occasions. Patient records from OD, LVC and AD</td>
<td>12 persons with USH 2 persons decline 1 male 1 not possible to trace 1 not USH I 1 not working age</td>
<td>3 females with USH I in working age less materials</td>
<td>3 females with USH I in working age most materials available</td>
</tr>
</tbody>
</table>

* The entire population is not known.
Preparation for Study I included a literature review of nine databases (AHMED, CINAHL, EBSCO, ERIC, MEDLINE, MLA, Psych. Info, Soc. Abstr. and Web of Science) in April and May 2001 followed by a complementary search in spring 2003. The search words used were ‘deaf-blind*’, ‘deafblind*’, ‘visual and hearing impairment (hard of hearing or deafness)’. Some additional references were found during the process by reading reference lists in the papers. The list was delimited in several steps. Firstly, reports studying medicine exclusively were excluded. Secondly, studies reported before the year of 1980 were in general excluded, except for those of great importance. These included studies that were mentioned in several of the other references and which included aspects of impact that have not been fully elucidated yet. Thirdly, conference reports and articles in languages other than English were excluded. References were evaluated by reading abstract or the full text.

The remaining 227 references were categorized according to title and abstract into a) category of life course (C=children, W=working age and E=elderly or H=historical matters), b) onset of the deafblindness (pre-lingual onset and post-lingual onset). The result of the search was published on the GENDEAF website. [http://audiology.unife.it/www.gendeaf.org/psychosocial/download/Kerstin_bibliography.pdf](http://audiology.unife.it/www.gendeaf.org/psychosocial/download/Kerstin_bibliography.pdf) [accessed 22 March 2008]]

The 227 references were further reduced to 96, which were included in Study I. The criteria for inclusion in Study I was that the papers should add new information. Thirty-two of the references did not include any empirical material. The 61 references with empirical material are shown in Appendix 1. Update of literature review 2002-2007 (Update) show further literature in Appendix 2

With regard to Study II, professionals from different services identified a total of 56 adults with deafblindness. These adults were then invited to participate. Three persons were however not open about having a second impairment in their relationship with Low Vision Clinic or Audiology Department and was therefore not invited by these clinics.

The nine females with a genetic or clinical diagnosis of USH I in Study IV were selected from a national database of USH. There were 21 persons in the database in the age and sex cohort: born between 1946 and 1960 and females. The criteria for selection were a) further confirmed diagnosis and b) patient records during the follow up time 1985–2004 from both Ophthalmology Departments and Low Vision Clinics. The 9 females selected were compared with the remaining 12 in the following aspects: degree of visual field impairment and visual acuity impairment (group wise in three age groups, 25–39, 35–49 and 45–59, because of progression of visual impairment) and degree of hearing impairment. Differences in degree of visual field and hearing impairment between the groups were not significant. In the selected groups there were how-
ever two persons with higher degree of visual acuity impairment than in the other group.

Sources have different values depending on its authenticity, i.e. if the source is a primary or secondary source (Merriam, 1988). The sources in Study II–V were primary sources, although in Study V, secondary sources were also used. Different terms were therefore used when referring to a source. Primary sources included what the informants reported, what the professionals at AD, OD and LVC documented with regard to results of examinations and observations made during their interaction with the informant. Primary sources were referred to as follows: tell, write and observe. Secondary sources included what the informant and other people (e.g. informant’s mother) told the professional and what the professional documented in the patient records. Secondary sources were referred to as follows: report and note.

Interviews (Study II and V) about experiences were valued to be primary sources. Data in Study IV was valued to be of primary source since the data included patient records.
METHODS

Different methods were used in the studies: literature review (Study I); interviews (Study II and V); questionnaire (Study II and III) and individual patient records (Study IV). Design of the evaluation in Study IV was of managerial type (Øvretveit, 1998). Table 6 shows methods, analysis method, age group, ICF components and service if any service was reported.
<table>
<thead>
<tr>
<th>Study</th>
<th>Methods and time for data collection</th>
<th>Analysis</th>
<th>Age group</th>
<th>ICF component*</th>
</tr>
</thead>
<tbody>
<tr>
<td>IV</td>
<td>Retrospective assessment of patient records of health state and of ophthalmic health care. Patient records 1985–2004.</td>
<td>Content analysis of the text in patient records into four themes; (1) managerial; (2) examination; (3) health state; and (4) actions with ICF as a frame. Descriptive statistics and Spearman’s rho.</td>
<td>C/Y W</td>
<td>s, b, d, e and personal</td>
</tr>
<tr>
<td>V</td>
<td>Interviews and patient records. Patient records notes were from first notation to 2005, which is ≈ 40–50 years.</td>
<td>Selected examples of situations with impact on participation and service. ICF is used as a framework. Analysis by content analysis with Disability as a laminated system, Bhaskar &amp; Danermark (2006) Life course approach, Priestley (2003) in mind.</td>
<td>C/Y W</td>
<td>s, b, d, e and personal</td>
</tr>
</tbody>
</table>

* Age group: C/Y = childhood/youth; W = working age; E = elderly.
** ICF component described by its code letter: s = body structure; b = body function; d = activity and participation; e = environment and personal for the personal factor.
*** Self-reported
Throughout the study ICF is used as a framework, a tool in order to study influence i.e. mechanisms. This means that ICF is used as a model for disabling processes rather than as a classification. Few codes are however used in Study I, in Appendix 1 and in Appendix 2.

The questionnaire in Study II was developed in collaboration with members of the Association of the Swedish Deafblind (FSDB). How questions should be asked and the layout of the questionnaire was elucidated. A pilot version was first developed and tested on five members. Communication and environmental considerations in these collaboration situations will be described further in the discussion. The questionnaire in Study III was performed in collaboration with members in the Swedish Deafblind Youth (DBU).

In the qualitative research interviews in II, and V, knowledge was obtained through interplay between interviewer and interviewee (Charmaz, 2007).

Interviews were performed using different themes (Kvale, 1997). The theme adopted in interviews performed in 1997 in Study II was: to have, to be and to love (Allardt, 1977). The idea was to find out present issues related to quality of life in relation to their impairments and service. The participants however often reported about previous experiences, which broadened the information gathered. Results of interviews in the 1st investigation’s report (Möller, K. 1999) were performed using ICIDH-2 (WHO 1999) as a framework2. This material was then reanalyzed during the preparation of Study II according to ICF (WHO, 2001). The third interview was performed in 2004 using an interview guide with five central themes: 1. How do you see?; 2. How do you hear and how is your balance?; 3. Which other senses do you use and what do you do in order to get information about things that you can’t see or hear?; 4. What impact does your deafblindness have in your daily life? 5. How do the social services (health care etc.) work?

The first two interview occasions were tape recorded and transcribed verbatim. The third interview occasion was performed using video-film and also transcribed verbatim. Each session took about two hours, with short breaks after about one hour.

Professional interpreters skilled in interpreting for persons with deafblindness facilitated interactions in preparation of questionnaires as well as in interviews.

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2 With kind permission from Dr Üstun, WHO.
Coding

Once the text was transcribed, coding of interviews and patient records were performed using the following steps:

1. Transcribed texts from the interview and reproduced records were read in their entirety to obtain an overall picture.
2. Statements in the text were coded deductively using ICF as a frame.
3. In the patient records four themes: health care system, procedure examination, patient’s functioning and disability and procedure action were also coded. Within the themes coding was performed inductively. All texts were coded, both into tree nodes and in free nodes, which is possible in NVivo2.
4. Text that was coded was printed in order to check if coding was pertinent, some recoding was then performed.

In Study I, text from the whole paper or abstract was scrutinized and activity/participation and environment were coded into ICF components at one level that is chapter level (e.g. d1) and some at two level (e.g. d810). Appendix 1 and Appendix 2 show a more systematic description of components found within the papers than was reported in Study I.

In Study II interviews were used. A citation from one of the interviews is as follows:

“A friend said that her parents had told her that people get a lot of information from the radio. There is even information that not is printed in newspapers. I was very surprised. We miss a lot of what is happening in society.”

(Möller, K. 1999 p.70).

This informant is a female with USH I. She was at the time of the interview about 35–40 years old. This citation was categorized into three components: environment (Not all information broadcast on radio was accessible for people with hearing impairment); participation restriction (miss information); and personal components (even information that wasn’t printed in newspapers, surprised her).

Answers from the questionnaire in Study III were regarded to encompass and have relevance to different components in ICF. One question in the survey was for instance: “If the lesson was interpreted? Could you in these cases see what the interpreter signed?” Alternatives were: always, sometimes, seldom and never. This question was considered to be about environment. The responses to these and similar questions were compared with how the person answered the question concerning degree of visual and hearing impairment. Students with blindness reported not relevant.
In Study IV patient case records from both Ophthalmology Department and Low Vision Clinic were rewritten verbatim, note-by-note. Each note include, the event’s date, name and title of the professional, usually cause of the event, examinations performed, results of examinations, intervention (which in Study IV were labeled procedures) and occasionally some other reports about the patient. Other papers in the patient records that were stored in the folder: letters; copies of certificates etc. were not included.

When it was time for Study V most materials (data) were already coded. Records from AD were coded earlier following the same principles as with notes used in Study IV. The third interview was coded following the same principles as with first and second interview occasions.

For the purpose of coding different software was used and databases created. References (Study I) and the first and second interview occasions (Study II) were stored by using Microsoft Access (version 1997). Closed questions in Study III, were stored in (SPSS version 10) and open-ended questions were stored in Access. Notes in Study IV were rewritten into Access and then categorized in a qualitative database QSR NVivo (2.0). The third interview occasion was written in Microsoft Word. This material and the other qualitative data were transformed (to rich text format) and stored in NVivo.

Analysis

To analyse complex phenomena mean that a phenomenon is described in categories that are abstract in both a theoretical and empirical sense (Danermark, 2008). To abstract is to get the general conception rather than the specific i.e. concrete of something. Categories represent qualities that are central in order to understand a phenomenon. A way to find mechanisms with possible impact is to ask: what are the necessary conditions for this mechanism to have any impact on the phenomenon? The fundamental conditions for a phenomenon to be what it is differ (ibid.). Furthermore, mechanisms can reinforce, counteract and neutralize each other. Absence of mechanisms may also have impact. Mechanisms have potential power and do not disappear just because its impact not is empirically manifested (Danermark, 2002; Danermark et al 2002). Danermark (2008) emphasizes that it is important to distinguish between neutralizing and removing a mechanism.

In this study the six components in ICF are qualitative categories. Arrows in the ICF figure (Figure 2) are indicating a possible impact on the phenomenon, i.e. mechanisms. In the analysis it was investigated if there were any mechanisms facilitating or hindering e.g. participation.

In ICF it is situations and not persons that are described. Therefore situation with some events was placed in focus. Questions were asked to the situation (Charmaz,
Matters in a situation could however sometimes be described, but not fully understood. This problem was approached in two ways. First, situations were categorized chronologically. Situations followed each other in courses of events. New questions were then asked. Are there any relations between this situation and previous situations? Second, disability is understood as a laminated system (Bhaskar & Danermark, 2006). This means that there might be relationships between for instance different categories. Relations may also be loaded with power to accomplish something in something else. For instance there is relationship between sound in the environment and ear. There has to be sounds in order to hear something. Sound may in general have impact on ears in ways that makes it possible to hear. This is a mechanism that may have impact on human ears and thereby on hearing function. Deviation in the ear like damage haircells is something that has negative impact on possibilities to hear. This means that the deviation is a mechanism. In the final analytic work significance of mechanisms was evaluated. Is this mechanism important? How important is it? What are the impacts of it? These were examples of questions raised in the analysis.

**Ethical considerations**

Research involving human participants is regulated by ethical considerations. Ethical considerations (Ds 2001:62 Etikprövning av forskning som avser människor [Ethics for Research about Human Beings], 2001) include: to participate in research should be of one's own free will; the participation should be based on well-informed consent, which they at any time can withdraw; the research team has to ensure integrity, confidentiality and report the results to the informants. Furthermore there are special considerations when the research includes persons who have disease, are elderly or else have difficulties to be answerable. In these cases the researcher has to consider the legal protection of the informants (ibid.).

To participate in these studies were voluntarily. Information about the projects were received, written, oral and translated into sign language by professional interpreters. Information about the projects was also performed at meetings for the association for the Deafblind (FSDB). Informants gave their informed consent.

The Research Ethics Committee at Örebro University Hospital had approved the project.
SUMMARY OF THE RESULTS

Study I

Title: The impact of combined vision and hearing impairment and of deafblindness

Definition of deafblindness is often referred to as a combination of functional impairments and psychosocial impact. In the debate about how to define deafblindness some authors refer to regulation of service while others stress the degree of the impairments. How the group is defined and whether the combination of impairments has been taken into consideration or not in official statistics has great impact on the number of persons found.

Great heterogeneity in the group with visual and hearing impairment was reported and very few were regarded as totally lacking both functions. Most studies about psychosocial impact do not report degree of impairment or measurements of capacity and performance. Difficulties in testing persons with deafblindness were reported and that many instruments, especially psychological instruments were not suitable.

Differences between persons receiving services for people with deafblindness are: onset; age group, degree of impairment; communication level and skills, language and communication mode. Communications skills range from unintentional communication to skills at university level. Additional impairments were reported especially in persons with pre-lingual onset.

Impairment aggravation was reported to bring practical consequences. Withdrawal from activities was reported both for those with pre-lingual onset and those with post-lingual onset. Difficulties in mobility in unfamiliar surroundings were reported for persons with post-lingual onset. Persons with pre-lingual onset sometimes had problems in mobility and orientation in their familiar surrounding, irrespective of additional impairments.

Deafblindness was reported as something emotionally demanding for persons with post-lingual onset. People with inherited deafness e.g. those with USH I were reported not to view deafness as a disability. However visual impairment was valued as being traumatic. Frequent feelings of loss and grief, were found in people with successive progression of impairments. A positive correlation was found between inability to discover and localize people and events in the surroundings and pessimistic thoughts.

Social isolation was reported and isolation tends to increase with progression of impairment. Friends withdrawing were reported especially in the case of USH. However supportive attitudes were also reported. Children with pre-lingual onset tend not to get opportunities for making friends.
There were no reports concerning emotions for those with pre-lingual onset. However, there were numerous reports about behaviour problems among children and adults with pre-lingual onset. Prevalence of behaviour problems was however not verified. Many studies in the eighties and beginning of the nineties reported behaviour problems in persons with pre-lingual onset. Intervention was then directed to stop these behaviours.

Most reports about service report barriers such as insufficient service and some indicate inadequacies in service. In the case of facilitating services this was specifically adapted, professionals have specific knowledge and skills or get it by the reported intervention. Follow up periods of intervention were however mostly from some months to a year. An exception was a four-year follow up of intervention where different aspects of service were elucidated. Care of people with deafblindness was found to be stressful. Many professionals lack knowledge about deafblindness and thereby do not know how to provide the service. There were increased risk for conflicts between those professionals with expertise in the field of deafblindness and regular professionals. Burnout symptoms and a high level of staff turn over were also found.

More than half of the studies with empirical material concern children with pre-lingual onset. Few studies include elderly and do not consider specific issues related to old ages. Literature was rather prescriptive than descriptive and does seldom search for explanations. Few studies were conducted for scientific purposes. Majority of the studies were professional reports. On the whole our knowledge of psychosocial impact of deafblindness is fragmentary. How people with deafblindness live their lives remains in general unanswered.

Update literature review of 2002–2007 literature (Update)

In the additional literature review of literature 2002–2007 the references found shows in Appendix 2.

Six different studies about the situation for elderly people with deafblindness were found. There are positive correlations between age and prevalence of visual and hearing impairment (Chia et al. 2006). One paper show significant increased risk of mortality especially for women (Lee et al. 2007). Greater difficulties in performing daily life tasks were found (Brennan et al. 2005) but a prospective cohort study did not show increased risk in the long run (Lin et al. 2004). The only reference found that uses ICF in describing situations for persons with combination of visual and hearing impairment, show less health state, more falling and limitations in mobility, self care and domestic life. Social life was also restricted (Crews & Campbell, 2004). Depression was not correlated to dual sensory loss (Chou & Chi, 2004) or not only correlated to these im-
pairments (Capella-McDonnell, 2005). Decrease in health related quality of life is re-
ported (Chia et al 2006).

Studies in the twenty-first century and even some thirty years ago indicate that stress and frustration awaken inappropriate behaviours among persons with pre-
lingual onset of deafblindness. However some studies indicates that inappropriate be-
haviour decreases or ceases as a result of changes in social environment, that is adapted teaching methods (Janssen, et al 2002; 2003; Bruce, 2002).

Two papers show self-reported experiences from the perspective of persons with deafblindness (Barnett, 2002; Schneider, 2006). The previous did not find evidence for a deafblind culture analogue to deaf culture in United Kingdom. Deaf culture is identity based on an affiliation with a particular set of beliefs with emphasize on sign language. This study is conducted within “Deaf studies” who not position themselves to be within the disability research field (Barnett, 2002). The study showed touch (tactile sense) to be a common significant experience among persons with deafblindness. Schneider’s (2006) study on the other hand show: feelings of interactional powerlessness and active strategies to minimize this by doing: things differently; managing support and relationships; surviving others’ perceptions; and presenting sides of self.

Ninety-three patients with USH from seven European countries participated in a study of maintaining independence. Patients with USH I tend to need more help than USH II and the amount of needed help grows when patients get older (Damen et al 2005).

Recently found reviews (Nikolopoulos et al 2006; Sigafoos et al 2008) confirm the picture of methodological weakness, such as not report degree of impairment, which also was found in Study I.

**Study II**

Title: Is ICF a useful tool?

The 33 informants were found to have limitations in almost all items of activities and participation. Difficulties were classified based on what the participants reported during the interviews and from observing them. They had moderate to severe difficulties in performing approximately 66 percent of the activities. With regard to participation they showed restrictions in approximately 80 percent of the items. The sources of the limitations or the restrictions could at times be addressed to the impairment itself. Sometimes the limitations were caused by personal factors. However, limitations and restrictions were generally caused by different factors in the environment. In spite of this significant result there were consequences that were difficult to describe by the
ICIDH-2 (WHO 1999). Five of these issues were elucidated in Study II where ICF was used.

Firstly, there was the problem of how to describe rapid variations. It was found that informants, especially those with residual visual and hearing function, experienced rapid shifting in their degree of visual or hearing impairments, e.g. moving from a space with broad daylight to a space with less light.

Secondly, there were difficulties in adopting ICF to describe withdrawal from activities that informants, earlier performed, still ‘could’ perform but had given up.

Thirdly, different aspects of time were important. Loss of time, connected to activity e.g. takes longer to perform a task; could only do one thing at a time. “Loss of time” was also connected to environmental factors e.g. a lot of time spent in contact with health care and welfare system, communicating through the use of interpreters. When all the “time loss” was added up it appeared to be an impact factor, a factor that is not included in ICF.

Fourthly, there was the aspect of health risks, a consequence directly related to the nature of deafblindness. This was exemplified by: a person who except for the deafblindness also had allergy. Exhaustion, tiredness and headache caused by high degree of energy consuming when performing seeing or hearing was also reported. Another health risk elucidated was difficulties in gaining access to information in health promotion.

Finally this study brought up the issue of duties and obligation as an attitudinal aspect of expectations on people with deafblindness.

ICF was found to provide possibilities to show the great impact in all activities and participation when people have deafblindness thus the earlier mentioned remarks shows that ICF was not valued to be a sufficiently precise tool especially not in the social level of disability. The study ends up with the conclusion that ICF should be further developed.

Study III

Title: Social Recognition, Participation and the Dynamics Between the Environment and Personal Factors of Students with Deafblindness.

Many different factors contribute to participation or restrictions in participation. Experiences of environmental factors that facilitated were e.g.: light in the classroom, 90% (natural and product environment), see what the interpreter signed, 85% (combination of service environment and skills, personal factor). Less facilitation was reported for teacher showing which of classmates had answered a question, repeated what that
student said or signed, or handed around the microphone (47% facilitate) and adapted schoolbooks (24% facilitate).

Results is discussed according to Honneth’s (1997) theory of recognition, which have been applied by Danermark and Coniavitis Gellerstedt (2004) on disability with emphasis on persons with hearing impairment.

The study indicates experiences of barriers in the natural, as well as in the social environments, which restrict participation. Experience of considerateness, such as e.g. concern of the special requirements for students with deafblindness and specifically shortage of considerateness was the most important factor. Indications of a negative role played by some students (personal factor) for themselves e.g. withdrawal from activities may be interpreted as being due to lack of recognition, such as denigration or insult.

**Study IV**

Title: Long-term ophthalmic health care in Ushers Syndrome type I.

Results were presented into four themes: health care system; procedure examination; patient functioning and disability; and procedure action.

In health care systems there were several health care districts that provided the care. Low Vision Clinics (LVC) was responsible for the majority of notes. The number of professionals was relatively even between the two actors of the health care. A high degree of resource allocation was found. A considerable number of procedures were found; in total 1611. The average annual invested resources were 9 procedures per patient.

Examinations comprised thirty-seven percent of the procedures, which exclusively were oriented towards body structure and capacity. Ophthalmology Departments (OD) conducted almost eighty percent of the examinations and there was no difference between the first 10-year period and the second ten years. Measurement of performance, environmental and personal factors were not found.

In general, all patients showed deviation in the structure of the eye and impairments of several visual functions. In the beginning of the follow up period these impairments were severe in some of the patients. Findings showed aggravation of visual impairment in all nine patients during the 20 year follow up period. Differences between informants in pace and degree of deviation and impairment development were found. The fact that USH I was an aggravated phenomenon was clearly demonstrated in the examination of the patient’s vision.

The other type of procedure, actions, comprised sixty-three percent of all the procedures. In contrast to examinations, LVC was the more active actor conducting sev-
enty-five percent of all the action procedures. It was notable that the number of actions increased, from 224 in the first period to 539 in the second 10-year period. No correlation was found between procedures performed and to pace or degree of impairment in the patients with USH I. The overwhelming part of the actions comprised actions towards environmental factors, i.e. eighty-nine percent towards assistive devices or other service. LVC reported 77% of these actions.

The study indicates that the ophthalmic health care was characterized by inefficiency. ICF ought to be incorporated in ophthalmic health care strategy.

**Study V**

**Title:** Participation in people with Deafblindness – an ICF and the Life-Course perspective.

Study V shows complexity regarding participation in females with USH I. Complexity stems from inner mechanism (body structure and body function), mechanisms from outside (environment) and mechanisms in interplay between inner and outer factors. Furthermore both the world and the individual in the world change over time.

The three informants’ profound hearing impairment was detected in infancy since they did not respond to sounds in their environment. They were provided with rehabilitation: hearing training; training to lip-read what people said; and speech training. Visual impairment was detected when they were in school age. The children were also provided with hearing aids. Parents were urgently requested to perform the training. Informants also had speech training in school. It was however later reported in AD records that these efforts failed. One informant however reported that she was skilled in lip-reading. USH was diagnosed before informants were fifteen years old. However, the informants were not informed about the diagnosis. Diagnosis and by that means prognosis were not used as a potential for special rehabilitation, education etc. Informants attended school for the deaf, during the oral-education period where sign language not was permitted. Other children introduced informants to sign language. So other children with deafness brought them into a language and they were included in a deaf community.

Mothers must have however learnt sign since note in AD and OD records says that the informant’s mothers interpreted using sign language.

Visual field impairment was found to have a negative impact on possibilities to receive sign language. One informant had more rapid aggravation of visual acuity impairment than the two other informants. From about the age of 25 she had to receive sign language in tactile modality. The usual method using visual modality was no longer useful. The examples above, illustrate the dynamics and the need for a life course
perspective demonstrating the complexity of the interplay between numbers of mecha-
nisms.

The informant with the worst visual impairment was provided with an assistant when she was about 30 years old. The second informant who lived in another municipality was denied the same service. Instead she was offered other services, which usually were designed for the elderly, although she was in her thirties. The third informant, who lived in another part of the country, was denied both kinds of services. This resulted in the informant being unable to perform household work without insecurity and high risk. These tasks were also energy consuming and took considerable time for them. This example illustrates another type of important mechanisms, such as knowledge, attitudes and behaviors of the service providers.

Adults with deafness in the Deaf community in Sweden have changed their policy and attitudes towards deafness during the informants’ life course.

All informants reported that they learnt to cycle when they were children and that they frequently cycled. In the neighbourhood where the informants lived it was very common that adults cycle to work and leisure activities. Visual field impairment had an impact on activity and thereby also on participation in leisure activities when informants were teenagers. Study shows experiences of variation in visual function, reported to be due to environmental alteration and due to bodily factors. They all report withdrawal from activities after incidents. It appears that the emotion of insecurity and unpredictability were two very strong mechanisms that limited activity. In addition to the above-mentioned problems, there was a problem with lack of general knowledge. Not having good general-knowledge was a personal mechanism, which could be a severe barrier.

There were strong indications that services have failed to efficiently rehabilitate and educate the informants and arbitrariness was found with regard to provision of support. ICF and Life course perspective has brought an understanding of mechanisms in relation to participation. Rehabilitation based on ICF and supplemented with life course perspective should be encouraged and further developed in this context.
DISCUSSION

The empirical part illustrates participation restriction for persons with deafblindness in different life periods especially working age and children/youths (Study I–III and V). The updated literature review (Update) shows studies, which concern elderly persons with deafblindness. Restriction in participation for persons with post-lingual onset is shown in Study II, III and V. Study I show participation restriction in persons with pre- and post-lingual onset. All the studies reveal barriers in service for persons with deafblindness. In order to evaluate and explain the mechanisms that might have impact on the results, ICF is used as process model together with inspiration from the human ecological approach, disability as a laminated system and a life course approach.

Deafblindness

Several denotations of deafblindness were presented in the introduction. In general their starting point is symptoms, such as difficulties in daily life. None of these however deals with the nature of the disability. Furthermore none of these denotations approach the matter from the human ecological perspective (Gibson, 1966) or theories about mechanisms (Bhaskar & Danermark, 2006; Danermark, 2002).

According to Gibson (1966) there are features in the environment that the human body percept and by affordance assign some meaning. A precondition for this process is that the person can receive information from the source (environment). Reduced information from sources increases risk and uncertainty (Shannon, 1948). Although, Shannon suggested caution when applying his theory to all kind of fields, from a deafblindness perspective this approach contributes to an understanding of the nature of deafblindness.

Inspired from the theories by Gibson (1966) and Bhaskar & Danermark (2006) and using ICF as process model together with results in Study I–V a scientific concept of deafblindness is created. It is a complex disability that entails six components at several levels in interrelation with each other. The process briefly described from a functioning perspective starts with environmental component at natural level and ends with coherence and comprehensible messages at social level. Human body structure, ears and eyes pass signals. The internal cognitive body function processes signals pass afferrent and efferent nerve functions. These functions enable activities to see and hear, that respond to external and internal signals. Seeing and hearing are therefore dependent both on quality and degree of environmental and body function signals. Furthermore for an individual to understand the message in its coherence carried by the signals also
depends on the individual’s personal experiences. Describing these processes from a disability perspective is as follows: When visible and audible signals do not pass through body structure and body function, seeing/watching and hearing/listening activities become limited. This implies participation restrictions since the information cannot be interpreted in an intelligible way and related to the actual context. Furthermore, this means that deafblindness is a bio-psycho-social and contextual matter.

For persons with some residual visual or hearing function left the possibility to understand a message depends on: possibilities to see or hear; environmental factors and personal factors. If both impairments are total, then possibilities for the person to see and hear are totally limited irrespective of degree and quality of visible and audible signals. Signals can be transformed to perception of touch, which is the tactile sense. Thus the message can be delivered in tactile mode by an interpreter or it can be written in Braille. This means that to participate in the information in a message does not necessarily demand seeing or hearing. However it is also necessary that the message is delivered adapted to the receiving person’s knowledge and skills.

Further on the discussion, the above-mentioned concept of deafblindness will be used in order to evaluate and explain participation restrictions and barriers of service for these people. However in the Studies I-V different definitions of deafblindness are used. The denotation deafblind or deafblindness does not properly cover the complex relationships since it only designate the body function impairments. It is however out of the scope of this thesis to create a term that will be accepted by professionals and associations for persons with this disability.

**Mechanisms that restrict participation**

Participation, which in ICF is involvement in life situations, is approached in the meaning of taking part as the essence. Participation is further understood as something related to different roles at social and societal levels. For instance; taking part in education as a student, taking part in ones own affairs at individual level e.g. knowing if one’s clothes are clean, if food and beverage is edible. Participation at social level includes interaction with other people directly, face-to-face or by other forms of media such as letter, newspaper, television etc. Participation at societal level is in this study, mostly delimited to the role as patient, student or client.

**Body structure and Body function**

An example of a process that starts by mechanisms in the social environment and interferes with body structure is when two persons that both carry the same allele gene have a common child and this child develops Usher syndrome (Kimberling & Möller, 1995).
Study IV shows aggravation over time in the retina and consequently visual field impairment and gradually visual acuity impairment over a period of 20 years for some females with USH I. These findings are in accordance with Sadeghi et al (2006).

Differences between informants in the pace and degree of deviation and impairment development have also been reported before (Pennings, 2004; Sadeghi, 2005).

The mechanisms behind these variations have however yet not been found, although research about the role of environmental and personal factors proceeds.

Deafblindness at human body level is understood to bring increased risk for life, personal security and health (Study II and V). Striving to see, hear and comprehend was (Study II) reported to bring exhaustion, tiredness and headache. Study II also indicates increased health risks due to difficulties in gaining access to health promotion information. Update showed poorer health state, increased risk of falling and increased mortality rate especially in elderly women with deafblindness (Crews & Campbell, 2004; Lee et al 2007). Risk of falling and fracture is especially high for persons with USH I that together with the deafblindness also have bilateral areflexia (Study I, IV and V).

Study II described experiences of rapid variation in seeing and hearing function for instance when light and sounds changed. This indicates stronger dependency on these environmental conditions than for persons in general.

Visual field impairment was found to have negative impact for receiving sign language (Study V), which leads to activity limitation and participation restriction. The large amount of procedure examination directed towards body structure and body function in Study IV, did however not result in any noticeable procedure action towards activity and participation

**Activity and participation**

Activity and participation are explained from an outside perspective while personal experiences will be discussed below as a personal factor component.

Most of the definitions of deafblind(ness) in the Introduction include difficulties to perform activities. This is confirmed in Study I, V and Update. Study V and Update show activity limitations in performing self-care, and domestic life, which to a very limited extent has been reported before. However relationships between activity limitation and participation restrictions in deafblindness have not been explained in earlier studies. In order to increase our understanding of these relationships, activity limitation and participation restriction has been divided into primary and secondary. The division is important because the two types of participation restriction are related to different mechanisms. Primary participation restriction depends on seeing and hearing activity limitations (which is primary activity limitation). Primary activity limitation have to be
solved by facilitating interventions such as glasses and hearing aids, light and sound. If these interventions are not enough, primary participation restriction remains. Furthermore primary participation restriction has negative impact on activities other than seeing and hearing in a second step e.g. preparing food, eating or walking. These secondary activity limitations may have negative impact on participation other than taking part in that, which is visual and audible, which is secondary participation restriction. Consequences of secondary participation restriction are separation and isolation from the physical environment that is not experienced by touch, smell, taste or airwaves and thereby also social isolation. For instance, not discovering or localizing people and events in the near proximity result in social exclusion. Hence, a solution may be support like interpretation into sign language tactile mode or written information transformed into Braille.

Another example of the fruitfulness of discussing in terms of primary and secondary participation restriction may be seen when a person with deafblindness cease to take part in a certain activity, e.g. the decision to give up cycling (Study V) was not because of the inability to cycle in it self, but to the restriction in seeing and hearing.

Not being able to interpret the available information in an intelligible way in the context created such insecurity and risks that the person decided to give up cycling. In this example there existed a possibility to decide to not carry out the activity, but in real life there are a number of situations where such options do not exist.

Environment

Environment encompasses all levels from e.g. light and sound, glasses and hearing aids, support, attitudes, service and policy. Physical, social and societal environmental factors are included in this section except factors concerning service.

There are several examples in Study I–III and V of conditions in the environment that affect participation. For instance, not receiving adapted schoolbooks, or access to transportation service etc. Both persons with pre- and post lingual deafblind experienced social isolation although caused by different mechanisms. Study I shows that children with pre-lingual onset of deafblindness rarely get opportunities to make friends and in post-lingual onset, friends tend to withdraw, which is also reported by (Schneider, 2006).

Personal component

Personal component is among other things, experiences of deafblindness from the person’s perspective.

Persons with congenital profound hearing impairment (deaf) were found to have different attitude towards their seeing and hearing limitations. Limitations in hearing
were reported by some informants in Study III but by none in Study V. To understand this mechanism several issues were scrutinized. Informants in Study V have USH I i.e. they do not have experience of receiving information from sounds. For those with moderate to severe hearing impairment the situation is different. When informants in Study V began school for deaf, they were introduced to a language, sign language, and included in the deaf community. In their case being deaf may rather be a question of belonging to the deaf community (Berbrier, 1998). This was one of the observations that were raised due to the letter mentioned at the beginning.

Seeing is something persons with USH I have experience of, so losing sight has meaning. Developing limitations in seeing is generally emotionally demanding but for the informants in Study V it was catastrophic in a double sense. Communication with other people occurs by visual input and signs, which goes back and forth. Anything that affects this interaction has significant meaning.

An explanation of the social withdrawal was that they reported it was emotionally demanding to perform activities in situations where their restriction in participating becomes obvious both to themselves and to other members in their social environment.

The aggravated visual impairment is experienced as an ongoing loss, which is a threat to the social role ‘able deaf’ and threatens the possibility to communicate with other people. Other authors (Brennan, 1997; Miner, 1999; Vernon, 1998) have also noticed such experiences. Furthermore, reduced information brings uncertainty, which fits well with results in Study I and V. Since information may not be reliable this affects self-image, because they cannot trust what they see and or hear. This may also contribute to withdrawal. Uncertainty and feelings of vulnerability are also in concordance with increased risks.

In USH person’s ability to see the signs and to lip-read are lost little by little. This means that the impairment/limitation mechanism neutralizes these skills and the person has to develop new skills. Learning to receive messages in a new modality can be tiresome which contributes to withdrawal.

Study V shows that persons with diagnosis, which lead to deafblindness were not informed about their diagnosis. Their parents also often adapted this policy. However these persons found out as adults. This may have created distrust and feelings of suspicion towards service and may have affected family relationships because the parents were informed but didn’t inform their child and could thus not prepare the child for his/her future requirements in a proper way.

From the perspective of the person with deafblindness, they experienced themselves as an isolated island (Barnett, 2002) because the impairment separated the human from environment and thereby self in the environment. This is in concordance with conclusion with the findings of Study V.
Schneider (2006) report experiences of environment to be hostile. This is explained as personal experiences, separation and isolation from the physical environment, which is a secondary participation restriction.

*Other sources of restriction*

The dimension of time is not explicitly addressed in ICF. However several findings in Study I–V have to be related to time in order to be understood. Time has different meanings, such as lamination of experiences. Different aspects of time are reports of loss of time, activities taking longer to perform and more energy consuming (Study II, Study V), which are related to body and participation.

Timing (Giele & Elder Jr, 1998) becomes different depending on onset. For persons with pre-lingual onset of deafblindness, primary participation restrictions have impact on language acquisition and thereby further learning (Siegal-Causey and Downing, 1987; Chen and Haney, 1995; Nafstad & Rödbroe, 1999). For persons with post-lingual onset social life becomes restricted later in life and carries on. However there are reports in Study V that some adults with USH I did not acquire any language before school start. This is in line with deaf experiences (Corker, 2001). These experiences are however related to location in time and space.

The period when the informants attended the regional schools for the deaf was a transition time. The deaf community at the beginning of the period claimed that linguistic development would be improved by using signed Swedish (Fredäng, 2003). Some years later, when informants were educated in upper secondary school at the National school for the deaf, the Deaf community further altered their attitudes: disproof of the idea to make children with deafness to be like hearing people; emphasizing deaf people’s abilities and strongly claiming their right to sign language. The attitude was: deaf people as an able minority (Fredäng, 2003). This advocacy policy was successful and the possibility for participation in society increased. Sign language was recognized to be the first language of deaf people in 1981. This was a radical shift in the society. The emphasis on abilities was a contrast to the previous accepted stigmatization of deaf as helpless and not normal, i.e. handicapped (Fredäng, 2003). These attitudes are contextually bound in space and time and are contemporary with the period when the females with USH I in Study IV and V grew up. However, “ableism” among deaf people may lead to stigmatization of persons with impairment within the group. Reports of isolation and exclusion in Study I-III and V indicate that this has been the case.

In general secondary participation restrictions and many activity limitations can be reduced by facilitating environmental factors and by personal factors. This was however not the aim of this thesis and will therefore not be elaborated further.
Mechanisms that barrier service

Services are regarded to be important environmental factors for persons with deaf-blindness.

Health care

In health care, prioritization is necessary and in general life-threatening conditions take precedence over those that are less life threatening. The increase risk of life, personal security and health that were mentioned earlier are for many diagnoses that lead to deafblindness not due to the condition. However, none of the studies indicate that persons with deafblindness are prioritized less. On average, the annual invested resources were 9 procedures (Study IV), which indicate quite the reverse, great investment of resources. Study IV indicates low degree of efficiency in health care since no correlation between procedures performed and pace or degree of impairment was found. Matters of efficiency were the starting point for developing a classification concerning impairment, disability and handicap, ICIDH (WHO 1980). This problem is not only a task for health care management, but also a matter for the patients who require good health care and rehabilitation, which is efficiency from patient perspective.

Study V show that the three informants with USH I were provided hearing rehabilitation. This is an example of a mechanism that starts promising but that in the end form a barrier. The policy was to provide hearing aids and training in hearing for children with profound hearing impairment, which with ICF terms is total. This was based on research. Wedenberg (1951) assumed that it was possible to teach children with severe hearing impairment to hear (Ahlsén, 1991). It was for instance, children with profound impairment in high frequencies and severe impairment in low frequencies, which is the case in USH I. His research led to oral-preschools and hearing aids for these children. Wedenberg’s theory was however rejected by some professionals 1965 (personal communication Risberg). Ericson and Risberg (1977) showed that loud low frequency auditory stimuli had impact on vibratory cutaneous tactile senses rather than on hearing. The three informants in Study V however continued to have speech training and hearing aids until they were teenagers (about 1970–1975) when they themselves decided to not use hearing aids. Audio-otologist reported in informant’s records that despite great efforts with training both hearing and speech skills had failed. This may have implications in service today. Persons with for instance USH I, who were subjected to lies, failure in rehabilitation etc. may distrust professionals working in service. Furthermore these adults may now distrust Cochlea implant (CI), which is another type of assistive device. CI is beneficial in terms of improvements in quality of life for persons with deafblindness (Damen et al; 2006, Soper, 2006). Quality of life show better results for persons with USH I who have been provided with CI than those without CI (Da-
Examples of rehabilitation failure in Study I and V call for caution with regard to present day’s rehabilitation. It is important to understand that CI is a great assistive device but it must not be interpreted to be equal with cure. CI may serve as both facilitating, when it works, and barrier, if it doesn’t work. In situations when persons with deafblindness, cannot use CI, or when CI is out of order or if battery is not available may really affect these persons negatively. Therefore rehabilitation of persons with deafblindness must take the person as a whole in a life perspective into account. Miner (1995) strongly argues that children with USH I, also those with CI, ought to learn sign language tactile modality in order to secure future communication.

In Sweden health care is highly specialized. Health care is, primarily managed according to the medical model. This means that all patients with deafblindness need to have contact with at least two departments. However the separated body structure deviations becomes a unified primary participation restriction, which calls for unified understanding and thus collaboration. There is a need to approach these patients as a whole and this call for a holistic model. The current legislation, the Health and Medical Act state that services in the act refers to measures for medical prevention, investigation and treatment of diseases and injuries i.e. medical model.

ICF, which includes both functioning and disability, has potential to extend health care policy from the medical model to include the holistic model. Study IV is a retrospective study, which also includes the years, 2001–2004, after endorsement of ICF. However the pattern of reported health care in patient records showed examinations within the medical service field and absence of examinations of performance of activity and participation in the patient’s own environment. This indicates that ICF has not yet been implemented in ophthalmic health care for females with USH I in Sweden.

The policy whereby physicians are not obliged to inform patients about their disease was found in Study V and which professionals also report from USA (Miner, 1995) and other Nordic countries (Olesen & Jansbol, 2005b). The consequences of this policy were also discussed earlier related to patient’s confidence in professionals. Also in relation to rehabilitation this may have prevented patients from preparing for future life challenges. However this policy has been changed. In the current Health and medical care act it is obligatory to inform patients about his/her health condition (Prop. 1981/82:97, 1982).

This was part of a new attitude towards patients. People were considered to be responsible for their own health. This progress has however special implications when the patient, as a whole, on top of their impairment also have primary and secondary participation restrictions.
Education

Education from the student’s perspective was the focus in Study III and some self-reports in Study V also concern experiences of education. In both studies there are reports of barriers. In face-to-face interaction there are indications of negative attitudes, which form a barrier, for instance professionals not showing considerateness (Study III). Pre-requisite for participation, such as e.g. not having access to adapted schoolbooks (Study III) was also reported.

Petroff (1999) referred to in Study I, show that many persons with deafblindness don’t receive specialized service in school. Study I also show that: professionals lack knowledge; working with students with deafblindness can be stressful; and indications of conflicts between regular professionals and experts. These findings are mechanism at management level in the education system.

Study III and V indicate shortcomings in dissemination of knowledge between the health care, the school health care and other professionals at the school, such as teachers.

Study V shows persons with USH I who experience lack of general knowledge. Could this be understood as an effect of education that did not recognize them as a whole?

Other services for persons with disability

Study V shows that access to services for persons with disabilities were not equal. This is despite far-reaching legislation that emphasizes participation for persons with disabilities.

The social services act (SoL) that regulate municipalities responsibility for societal provision of service to persons with disabilities use the word unable to limit inclusion in service, which mean limitations in activities (SFS 2001:453, 2001; Social Services Act, 2001). The other act LSS uses the term functional impairments, which allude to the body function component (SFS 1993:387, 1993; Act Concerning Support and Service for Persons with Certain Functional Impairments, 1993). Persons with deafblindness have severe to total impairments, which have severe to total impact on activity. However, except for seeing and hearing the other activity limitations persons with deafblindness (like USH I) experience are of secondary limitation type. Being a person with deafblindness is to constantly encounter insecurity and risks. At body level there are situations threatening life or health, at psychological level ontological insecurity and isolation and at social level being a “non citizen”. These risks, may however not be regarded as inability. Both acts mention participation in their goals: To be enabled to participate in the life of the community (SoL) and shall promote equality in living conditions and full participation in the life of the community (LSS). Both acts also mention
participation in the community, which is participation as citizen, patient, student or client. Furthermore participation is valued as something good without specifying what restrictions in participation signify. This study shows that some persons that have deafblindness: experience sensory impairments; primary activity limitation; primary participation restriction; secondary activity limitation, secondary participation restriction; risks and are still not provided with SoL or LSS service. In these cases welfare professionals are sticklers for details. It takes common sense and self-government to understand that participation restrictions due to not getting visual and audible information leads to inability in the meaning of performing tasks safely and with reasonable energy consumption. Capricious decisions by professionals regarding allowance of service or denial is not surprising but iniquitous.

**Barrier presence of deafblindness**

Service Face-to-face is the level where interaction between professionals and persons with deafblindness takes place. Letters, emails and telephone calls between service and the patient/student/client are also included in face-to-face interaction.

In interaction with other persons, the restrictions the person with deafblindness experience become a problem for both because information that can be shared and interpreted is lacking.

In the situation where a third party, an interpreter, is involved this service also affect the communication in different ways. There is a risk that the professional experience the interpreter, which also is a professional, as the communication partner, instead of the person with deafblindness because the interpreter is the one who talk. Furthermore there is also a risk that the information given to the professional is that which has been understood by the interpreter, which one informant in Study V told. The person with deafblindness has difficulties expressing themselves, as she says, using these big words, which mean so many things in one. She is worried that the professional will get the impression that she is more able than she actually is. She is worried that the interpreter uses expressions that are more advanced than the informer is familiar with. Shortcomings in general knowledge should not be confused with intellectual disability (Godfrey & Costello 1995; Brennan 1997). In those cases interpreting may become a mechanism that facilitate and barrier at the same time.

Another type of interaction is written information or phone calls. In these cases other channels are also options, for instance tactile by Braille. Not all persons with USH I know Braille and even if they know Braille they may prefer to read by using their residual vision function.

This account of the services shows many barriers. However the present national goal for disability policy is full participation in society (Prop. 1999/2000:79). Though
neither this plan nor the evaluated services consider the most basic need i.e. protection of life. Instruments that measure Activity in Daily Life measure in general activity with mobility limitations in mind. Some instruments measure seeing activity limitation and others measure hearing activity limitations. However none of these takes life, personal security and health into account from the perspective of deafblindness.

The Convention on the Rights of Persons with Disabilities by UN (2006) includes fundamental principles to ensure life and full participation, which entail protection, promotion and individual progress. Furthermore ICF and the UN convention support necessary alterations of service legislation, management and could be used in education of service professionals in order to promote modern attitudes.

**Discussion about theories**

There are different theories within disability studies. The social model of disability elucidates power and oppression towards people with disabilities. Though they reduce the matter to social and societal issues “on top of the impairment” (Oliver, 1996, 2004). Within feminist approach to disability, issues about power and subordination, a double oppression, has been discussed, indicating that gender plays a role both regarding women who have disability and towards service to females with impairment (Thomas, 1999a, 1999b, 2001). Though there is a difference between Oliver, who argues that the private, which also includes the body should not be included in the analysis of disability and feminist researchers who argue that the body is also political and should be included in the analysis of disability (Meekosha, 1998; Thomas, 1999b; Thomson Garland, 1997; Wendell, 1997). The implication of this debate for this thesis was, firstly to include the body and to focus on one sex – females, in Study IV and V. It has also been argued, from a feminist perspective, that impairment should be valued as “differenceness” rather than something negative (Thomson Garland, 1997).

In research based on phenomenology the body is included and further, the body has different meaning depending on whether the perspective is the patient or the professional. For the patient it is the lived body, through which all experiences of the world come in. For e.g. physician, the patient’s body is the subject of his/hers works (Toombs, 1993). However, to discover relationships between body and environment these theories were not enough.

Gibson (1966) developed an understanding of the relationship between human body and environment – an ecological approach. According to his theory phenomena in the environment are perceptible by the human body (Gibson, 1979) and given significant meaning through cognitive processes. Gibson’s theory emphasises the primary function of perceptions, which is to facilitate interaction between individual and envi-
Gibson’s theory shows relationships between body and environment and indicates relationships between those components and the other three (activity, participation and personal). However Gibson has been criticized for simplifying the processes involved (Eysenck & Keane, 2000 p.53–61). Noble (1983) followed Gibson in an ecological approach of the auditive world. Recognition is possible due to patterns of continuity and of discontinuity. Events make their audible whereabouts due to things that happens and this is an integral feature of their occurrence. Shannon’s theory about passing information from source through channels (Shannon, 1948) was interpreted as in body structure and body function to be channels for passing information. If only parts of the messages are passed on, through the channels, this increase risk and uncertainty, which also was found to be the case regarding deafblindness.

Williams (1999) among others criticise the social model of disability for not including the body. Danermark (2002) introduced the idea of interdisciplinary research. Both Williams and Danermark rely on Critical Realism (Bhaskar, 1978; Danermark et al., 2002). Furthermore disability as a necessary laminated system was developed (Bhaskar & Danermark, 2006) and the theory of a laminated system contributes to an understanding of what kind of impact different mechanisms has. One of the advantages in applying the theory of laminated system is that it is possible to detect impact factors, down up, up down in levels of the laminated system as well as mechanisms at the same level. Presence as well as absence of features and events may have significant meaning.

Discussion about ICF as a frame

ICF has been used as a framework in all five Studies. In this final framework ICF was used as a process model.

The choice in general was not to use codes because ICF was used as a model for describing disabling processes and not as a classification. It is however possible to add codes afterwards, which may be used in the future in an initial core set work in order to identify codes relevant in the case of deafblindness.

All six components have been used and found necessary. However as like mentioned earlier discrimination between body structure and body function require separate negative terms. Even more important was the possibility to discriminate between activity and participation. These components denote different phenomena, especially regarding doing and taking part in, which is important in order to understand the nature of deafblindness. Molin (2004) shows that participation is a phenomenon that can be understood when related to both social and societal levels. Possibilities in ICF to distinguish between personal experiences, described by the personal factors component and participation from an outside perspective, described by participation component
was found to be an important advantage. ICF also includes possibilities to describe participation in relation to different social and societal roles, such as patient, student, client and citizen.

“What a person does or does not choose to do is not related to functioning problem associated with a health condition and should not be coded” (WHO, 2001 p.205). This is correct since the personal factor component is not included in the coding system. This study shows that personal choice, such as withdrawal from activities, was important indications of participation restrictions. Study I, III and V shows that withdrawal is common for persons with deafblindness. This is at present a great problem in Sweden since legislation of service for persons with disabilities allows free scope of interpretation of the rules. In the case of services that use activity limitation and ask questions like: can you do this or do that, for drawing the line for receiving support or not have been found arbitrary for persons with deafblindness. Neither of the acts (SoL or LSS), emphasize, life, personal security or health in relation to disability.

In measurement of activity WHO suggests discrimination between what a person actually do in his or her current environment, which is performance and the individuals ability to execute a task or an action, which is capacity. WHO suggests that capacity is assessed in a ‘standardized’ environment, in order to neutralize impact by varying environmental conditions. Persons with for instance USH I or USH II–III are due to their visual impairment sensitive to light. Therefore a standardized environment in general may not show their capacity. Study V show differences in possibility to perform for instance reading a paper related to inner bodily health state such as fatigue or monthly period.

ICF includes possibility to use it for different purposes. It is also possible to approach the model from other perspectives than an individual. In Study IV evaluation of ophthalmic health care for females with USH I was performed. The study uses ICF to show patterns of examination, patterns of impairment aggravation (as examination results) and patterns of health care in response to their own findings. The results show mismatch and inefficiency.

In Study V and in this thesis framework ICF is used as a process model in order to evaluate and explain relationships between the components. Body structure, body function, activity and participation are phenomena with different nature. By thinking of them as different levels it was possible to detect chain reactions and reactions in different directions. This is described and discussed previously in mechanisms that restrict participation and mechanisms that barrier service.

ICF provide a conceptual base for description of situations that entail persons with or without impairment and facilitating service as well as service barrier. This opportunity made it possible to imagine persons with no present impairment in different situa-
tions in order to discover the differences in relation to the same situation for persons with deafblindness. This was fruitful in order to evaluate and explain mechanisms.

In agreement with Stucki et al (2007) a theory of health needs to be developed and could be preceded by a theory of human functioning. It is very important that ICF is inclusive in relation to medical model and aetiology. The medical model includes expertise in different clinical disciplines. Their procedures and interventions for individuals regarding disease are necessary and are aimed at saving life, treating and curing. Thus these issues are all part of the wider health concept.

The health and medical services act ought to incorporate ICF and bring in, “the person as a whole”. This is in line with rehabilitative and supportive strategy that the health care has besides preventive and curative strategies. Furthermore incorporation of ICF in health care management and education of health care professionals would support a common language between different professions and transferred to patients and their next of kin.

ICF does not however include dimension of time. Study I–V shows that different aspects of time are important for participation restriction and service barrier. For instance Study II show differences in activity limitation due to alteration in environment, Study IV indicates that patients spend time on health care etc. Study V shows that it may take time to do something. ICF as a process model should be supplemented with, time place and events from the life course approach.

**Discussion about Life course approach**

None of the authors (Bhaskar & Danermark, 2006; Danermark, 2002; Williams, 1999), for this study important works, applied their theories to data materials that take location in time and timing into account. Life course approach links, societal and cultural phenomena in time and place with individual’s experiences and sense of self (Giele & Elder Jr, 1998).

The life course perspective has inspired the design of Study IV and V. In Study IV patient’s chronological life and development of the disease was described in relation to documentation of the ophthalmic health care these patients were provided during a period of 20 years. No correlation was found in relation to pace or degree of impairment or to county in Sweden. The life course approach in Study V lead to mapping: years for certain rehabilitation; and educational practice; with informant’s age and attitude changes towards deafness in the deaf community in Sweden. The mapping led to links between rehabilitation policy, oral method without sign language and informants’ childhood. Mapping also led to links between the attitude “ableism” and informants’ youth. This was further linked to the years of body structure deviation of the eyes and
body function impairment of visual function. Thus coping strategies to try to hide seeing limitation and thereby withdrawal is reasonable. Previous experiences in persons with deafblindness ought to be valued as on ongoing time lamination, though Scott and Alwin (1998) call this, cumulative experiences. However past experiences should be considered in present and future service planning.

**Discussion about materials**

This thesis entails material from different sources and covers a considerable time span, approximately 50 years retrospectively from 2005.

Most materials in Study II-V are primary sources and concerns matters in Sweden. In Introduction, legislation and management matters of service, health care, education and services for persons with disability are reported. Literature review in Study I and Update from contains secondary source material. They revealed studies mostly from United States. All these reports are in concordance with findings in Study II, III and V, which increase the trustworthiness of the findings in these studies.

Health care records are assumed to be a rich source of critical information (von Koss Krowchuk et al. 1995; Øvretveit, 1998). Colby (1998) argues that it is of great advantage to reuse existing archival data, in life course research. Especially useful are open ended material that can be recoded and recast in terms consistent with the investigator’s new research questions. Other studies that elucidate health care for persons with deafblindness, such as Study IV have not been found.

The reason for delimitation from all with deafblindness, in Study I and II, to persons with acquired deafblindness in Study III to focus on USH I in Study IV and V, was the difficulties in distinguishing between mechanisms related to limitations and restrictions in seeing and hearing and impact from other impairments. Persons with USH I also have vestibular areflexia but this has rarely been elucidated in the five Studies. USH I is, firstly the most frequent type in Sweden (Sadeghi et al., 2004) secondly, people with USH I usually have total hearing impairment, which has significant social and societal implications. Delimitation to working age in the two later studies was done in order to find mechanisms connected with manifest impairments. Delimitation to females was done due to phenotype differences between the sexes (Sadeghi et al., 2004) and also in order to avoid bias i.e. social impact of life experiences due to gender (Thomas, 1999; Thomson Garland, 1997; Wendell, 1997) and possible impact of gender related to service (Thomas, 2001).

The study is further delimited to those with manifest visual and hearing impairment even though occurrence of factitious deafblindness has been found (Miner & Feldman,
The mechanisms for creation of such disability are different and require different services.

This study has not scrutinized the relationship between deviation in body structure and impairment in body function though this may be important which some genotype phenotype studies of USH show (Pennings et al 2004; Sadeghi, 2005).

There is an amount of data collected and available for later use, for instance about: work, use of alternative strategies, health promotion issues, motherhood, collaboration between different services etc.

**Discussion about methods**

The findings are mostly delimited to mechanisms with impact on restriction in participation for persons with acquired deafblindness and in the two latest studies females with USH I.

Study V and Discussion were inspired by Life course approach rather than being carried out completely as life course studies. However there are some implications in life course studies that are also relevant in this study. The analysis in this framework mostly relies on qualitative data and therefore the question of reliability is rather about trustworthiness. In the discussion, events in the past were linked to present day impact. However it is not easy to obtain accurate and complete information about the past. In retrospective studies the researcher has to rely on people’s present recollection about their past (Scott and Alwin 1998 p.100). People also have their denials etc. In Study V the informants were youngsters, during “the disability rights” period. This may have influence on their willingness to report oppression for instance prohibition of the use of sign language in the compulsory school. However there are assertions that some teachers turned a blind eye to the fact that deaf children used sign language. This shows that the picture is not unambiguous. The oral period in deaf education is however well documented in earlier research (Pärsson 1997; Fredäng 2003) and this increases trustworthiness in this aspect.

Scott and Alwin (1998) recommend combinations of retrospective and prospective design since both methods have their advantages and disadvantages. Recurrent performed interviews can contribute to richer information about the past. Recurrent interviews may however increase the so called Hawthorne effect, which signify that frequent participation in research may have impact on memories etc. In Study V informants were interviewed three times with some years in between the second and third occasion. It is possible that these informants were influenced by “ability” attitude in the deaf community. At the first interview occasion informants with USH I, said that they, as deaf, were not used to talking about problems. At the third interview in Study V, two of the in-
Formants on their own initiative talked about their failings. This may be due to having confidence in interview situation, Hawthorne effect but also due to progress of visual impairment. At the time for the third interview all informants in Study V had marked impact in daily life. Olesen and Jansbøl (2005b) prospectively over 5 years collected data about experiences of living with post-lingual deafblindness (all informants have USH and 8 of 20 have USH I). These authors report that many informants gave emotionally demanding information for the first time at the third to fifth interview occasion.

All data in Study IV and a considerable amount in Study V consist of patient records. This health care documentation comes from Ophthalmology Departments and Low Vision Clinics (Study IV and V) and Audiology Department (Study V). These sources may be regarded as prospectively created data, since notes were made in connection with interaction events from first to last note. However, there may be information that is gathered but not documented in the records. Although such knowledge, if there is any, is not available to allow evaluation of health care based on records.

Most interviews in Study II and all in Study V were performed with support from professional interpreters skilled in interpretation with persons with deafblindness. Two interpreters always worked together and alternated during the interview to perform the interpreting and to support and make corrections. The interpreters were given interview guidelines in advance and preparation meetings were held with them. The interviewee’s were recommended if possible to engage interpreters they had confidence in, which was possible because booking was made well in advance. Interviewee’s were also recommended to choose an optimal place for the interview. In general interviews were performed in informants home. Before the interview started the interviewee was asked to decide where everybody (interviewee, interpreters and interviewer) should be seated. In the third interview (Study V) video camera was used and it was located with focus on the interviewee. From the beginning of the second interview and thereafter the interviewer and interviewee knew each other and shared experiences from previous interactions. Furthermore before the third interview occasion, the last two interviews were studied; in order to find out possible misunderstandings, sort out questions etc. An authorized sign-language interpreter also skilled in interpreting to persons with deafblindness verified quality of interpretation. This evaluation was performed in the following way: randomized video sections from the interviews were scrutinized regarding what interviewer and interviewee expressed and how this was interpreted. Interpreting was found to be excellent.
Future research

Results show many mechanisms with negative impact on participation and on service for persons with deafblindness. Two areas that would increase such knowledge are:

1. This thesis calls for knowledge about service facilitating mechanisms. There are for instance some health care and educational resources for persons with deafblindness at national and regional level. Do these resources facilitate and/or act as a barrier for regular service? What are the mechanisms that facilitate or barrier collaboration and that affect the exchange of information between different service providers?

2. It is important to gain knowledge about facilitating mechanisms especially mechanisms that facilitate participation. This means gaining knowledge about factors that support independence and empowerment.
CONCLUSIONS

The conclusions that can be drawn from an ecological, laminated and life course approach, described with ICF terms are:

- *Participation restrictions* for people with deafblindness are far-reaching and are embedded in a complex process of interaction between the person with deafblindness and the environment. Furthermore, services, which are aimed to facilitate these people’s participation sometimes, entail systematical barriers.

- Earlier definitions of deafblindness concentrate on social symptoms. Since the ontology of deafblindness is not scrutinized in these definitions, the symptoms of deafblindness will not be understood into their full extent. For instance, deafblindness is foremost associated with limitations in daily life. In order to improve the services it is extremely important to understand the role of *participation restrictions* in this disability.

- Deafblindness is a disability, which occur when visible and audible signals do not pass through body structure and body function. Thus seeing/watching and hearing/listening activities become limited. A significant aspect of deafblindness is *participation restriction* in information from visible and audible signals. This means that deafblindness as disability is a bio-psycho-social and contextual matter.

- Deafblindness signify that the information other people get from visible and audible features, could not at all or in limited extent be seen or heard for people with deafblindness. Often fundamental information cannot be comprehended in an intelligible way, related to the actual context. It is suggested that activity limitations and participation restriction are divided in *primary and secondary activity limitations* respectively. Primary activity limitation is to not see and hear enough for comprehension. Hence, not taking part in the visible and audible world is primary participation restriction. The results of primary participation restrictions are often secondary activity limitations. That is limitation in other activities than seeing and hearing. This means that the nature of limitation in for instance communication, daily life, mobility, domestic life etc. are due to primary participation restrictions and in general not ability to perform the activity. Secondary activity limitation may create secondary participation restriction.
For people with deafblindness the options and threats in the proximity environment cannot be fully discovered and comprehended. Performing activities without basic information includes risk. One important aspect of deafblindness is exposure. For people with deafblindness exposure sometimes signifies risk of life and limb. A psychological consequences is that deafblindness is associated with ontological insecurity as a result of the fact that the information the deafblind person gets may not be trustworthy because of the primary activity limitations.

In human life, experience after experience became layers of experiences. If a person with deafblindness have not participated and lack experiences this will result in laminated information gap. A result of this is fragmentary knowledge in many fields. Situated in time and space, the social and cultural environments, which persons with deafblindness are in have an impact on the person’s possibilities to coop with and adapt to a situation. This may result in ontological insecurity and a rational way of coping with this is to withdraw. For instance visual and hearing impairment may have different psychosocial impact for people with inherited deafness due to their social roles. Sometimes the social environment show negative attitudes and insufficient knowledge about deafblindness. Personal choice, if this option is available, is often withdrawal from social interaction. At the social level, deafblindness implies risk of isolation and exclusion. This means that these persons potential of contribution as citizens is at risk and a serious participation restriction.

Services for persons with deafblindness are sometimes missing, are not always satisfactory and do not take the person as a whole into account. There are also examples of services that over time shifted from being a facilitating factor to being a barrier. Furthermore, previous lack of, or not satisfactory, services are layers of barrier that have impact on present situations, both in daily life and in relation to present interaction with service providers. There are indications of shortcomings in collaboration between different service providers.

Services for persons with deafblindness must take into account body-psycho-social aspects of this complex and rare disability. Persons with deafblindness require rehabilitation in a life perspective that takes the complexity of the syndrome and its prognosis into account. Furthermore, rehabilitation has to a larger extent than today comprehend the nature of deafblindness, the importance of the physical, social and cultural environment and personal factors in order to promote participation and prevent risk. In order to increase people’s participation and protection requirement of individually adapted support and assistive devices is necessary.
• There is a need for development of service legislation, service management and improvements in face-to-face services. The UN convention on the rights of persons with disabilities includes fundamental principles to ensure life and full participation, which entail protection, promotion and individual progress. ICF and the UN convention support these necessary alterations.
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**APPENDIX 1**

*References in Study 1 that show empirical material*

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<td>d2</td>
<td></td>
<td>e580+ Behaviour</td>
</tr>
<tr>
<td>Rönnberg et al (2002)</td>
<td>13</td>
<td>W+E</td>
<td>Post</td>
<td>d1, d3, d8</td>
<td>e1+</td>
<td>Stress, emotional problem Skills</td>
</tr>
<tr>
<td>Sauerburger &amp; Jones (1997)</td>
<td>69</td>
<td>W</td>
<td>Post</td>
<td>d4</td>
<td></td>
<td>e1+, e3, e3+ Skills</td>
</tr>
<tr>
<td>Sisson et al (1993)</td>
<td>2</td>
<td>C/Y</td>
<td>Pre</td>
<td>d2</td>
<td></td>
<td>e585+ Behaviour</td>
</tr>
<tr>
<td>Stein et al (1982)</td>
<td>101</td>
<td>C/Y</td>
<td>Pre</td>
<td>d3, d8</td>
<td></td>
<td>e585</td>
</tr>
<tr>
<td>Torrie (1978)</td>
<td>17</td>
<td>C/Y+W</td>
<td>Post</td>
<td>d7</td>
<td></td>
<td>e3</td>
</tr>
<tr>
<td>Study</td>
<td>N</td>
<td>Life Period</td>
<td>Onset of Deafblindness</td>
<td>ICF Components</td>
<td>ICF Component</td>
<td>Component</td>
</tr>
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<td>---------------------------</td>
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</tr>
<tr>
<td>Tweedie (1974)</td>
<td>1</td>
<td>C/Y</td>
<td>Pre</td>
<td>d2</td>
<td>e585+</td>
<td>Behaviour</td>
</tr>
<tr>
<td>Watkins et al (1994)</td>
<td>24</td>
<td>C/Y</td>
<td>Pre</td>
<td>d1, d2, d3, d7</td>
<td>e570+</td>
<td>Behaviour</td>
</tr>
<tr>
<td>Vernon (1969)</td>
<td>8</td>
<td>W</td>
<td>Post</td>
<td></td>
<td>e580</td>
<td></td>
</tr>
<tr>
<td>Wolf-Schein (1989)</td>
<td>747</td>
<td>C/Y+W+E</td>
<td>Pre, Post</td>
<td>d1, d6, d8</td>
<td>e3, e5</td>
<td></td>
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<tr>
<td>Yarnall (1980a)</td>
<td>1</td>
<td>C/Y</td>
<td>Pre</td>
<td>d1, d2</td>
<td>e585+</td>
<td>Behaviour</td>
</tr>
<tr>
<td>Yarnall (1980b)</td>
<td>1</td>
<td>C/Y</td>
<td>Pre</td>
<td>d1</td>
<td>e580+</td>
<td>Behaviour</td>
</tr>
</tbody>
</table>

Life period: C=Childhood; W=Working age; E=Elderly.
Onset of deafblindness: Pre= Pre-lingual; Post=Post lingual.
ICF component coded at one or two level, condensate personal factors.
### APPENDIX 2

**Update literature review 2002–2007 (Update)**

<table>
<thead>
<tr>
<th>Reference</th>
<th>No of subj.</th>
<th>Life course</th>
<th>Onset</th>
<th>ICF d</th>
<th>ICF e</th>
<th>ICF personal</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Barnett (2002)</td>
<td>8</td>
<td>W</td>
<td>Post</td>
<td>d3, d4</td>
<td>e1</td>
<td>Emotions Habits Coping style</td>
<td>To identify factors that may indicate presence of a deafblind culture. By mailing lists share experiences from an inside perspective. No culture found, though some common experience. The significance of touch and experiences of being an “isolated island”.</td>
</tr>
<tr>
<td>Bourquin &amp; Sauerburger (2005)</td>
<td>No</td>
<td>W</td>
<td>Post</td>
<td>d3, d4</td>
<td>e1</td>
<td>Personal skills</td>
<td>Discuss how to teach skills people with deafblindness require when they are on their own in public areas.</td>
</tr>
<tr>
<td>Brennan et al (2005)</td>
<td>1082</td>
<td>E</td>
<td>Pre and Post</td>
<td>d3, d4, d5, d6, d8</td>
<td>e1+, e5+</td>
<td>Quality of life</td>
<td>Examine relation between dual sensory impairment and limitations in ADL and IADL tasks. One thousand eighty-two of 5151 had deafblindness. Using longitudinal existing data about elderly age 70+. Found limitations in both ADL and IADL tasks. Greater limitations than those with single impairment.</td>
</tr>
<tr>
<td>Author(s)</td>
<td>Year</td>
<td>Type</td>
<td>Group</td>
<td>Test</td>
<td>Design</td>
<td>Methodology</td>
<td>Findings</td>
</tr>
<tr>
<td>--------------------</td>
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<td>--------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Bruce (2005)</td>
<td>2005</td>
<td>No</td>
<td>C/Y</td>
<td>Pre</td>
<td>e3+</td>
<td>Personal</td>
<td>Discuss progressive distancing to children with pre-lingual deafblindness as a way to develop an understanding of the differences between themselves and others.</td>
</tr>
<tr>
<td>Bruce et al (2007)</td>
<td>2007</td>
<td>7 C/Y Pre d3</td>
<td>Personal</td>
<td>Descriptive the topography, rate, and function of gestures expressed children who are pre-deaf-blind.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Capella-McDonnell (2005)</td>
<td>2005</td>
<td>447 E Post d3, d7 e5</td>
<td>Personal</td>
<td>Retrospective cross-sectional study of persons with sensory impairment for persons aged 55&gt; to find correlation between sensory loss and depression. All in all 9832 persons of whom 447 had dual sensory loss. Depression in this group is not only correlated to the dual sensory loss.</td>
<td></td>
<td></td>
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<tr>
<td>Chia et al (2006)</td>
<td>2006</td>
<td>2015 E Post e1+, e5+</td>
<td>Quality of life</td>
<td>Study correlation between deafblindness in the elderly and its impact on health related quality of life. Interviews and assessment of visual and hearing function. Positive correlation between age and prevalence of hearing impairment and visual impairment. The combination of dual sensory loss and decrease in health related quality of life was also found.</td>
<td></td>
<td></td>
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<tr>
<td>Chou &amp; Chi (2004)</td>
<td>2004</td>
<td>130 E Post d4 e2, e4+</td>
<td></td>
<td>Study of loss of both visual and hearing function causes depression in the elderly in relation to other functional and social variables. Depression does not increase if the person already has visual impairment. Correlation between functional health and depression was not found.</td>
<td></td>
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</tbody>
</table>
Crews & Campbell (2004)  
Retrospective cross-sectional study of health in the elderly 70+, by using ICF as a frame. Those with dual sensory loss reported less health state than those with only one impairment. More falling, heart problems etc were also reported. Limitations were reported for mobility, self care, and domestic life. Social activities were also restricted. Elderly were found to often have additional impairments.

Damen et al (2005) 93 W+E Post d1, d3, d4  
Investigation of access to information, communication and mobility. Patients with USH type I, II and III. Type I tend to need more help. Requirements of help increase when patient gets older.

Damen et al (2006) 28 C/Y +W Post d1, e1+ Quality of life  
Retrospective comparative study of children and adults (USH I) with and without Cochlea implant. A significant benefit of CI was seen in the hearing-specific questionnaire NCIQ. This difference could not be detected in the generic SF12 survey. The Usher Lifestyle Survey indicated that patients with USH I with a CI tend to be able to live an independent life more easily than the profoundly deaf not implanted patients with USH I. EHL and FVS scores varied in both groups. Conclusions: Overall QoL can be enhanced by CI in patients with USH I, although effects are mostly seen in hearing-related QoL items.
<table>
<thead>
<tr>
<th>Author(s)</th>
<th>Year</th>
<th>Study Design</th>
<th>Intervention</th>
<th>Outcome(s)</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>Filipo et al (2004)</td>
<td>31</td>
<td>d3, d7, e1+, e3+, e5+</td>
<td>Quality of life</td>
<td>Case study of attitudes, requirements in the case of patients having dual sensory loss that were provided with Cochlear Implant (CI). Assessment of hearing function, questionnaire, observation and interviews. CI can improve quality of life for persons with deafness and additional impairments. Best results showed adults with deafblindness who communicated at language level.</td>
<td></td>
</tr>
<tr>
<td>Janssen (2004)</td>
<td>4</td>
<td>d1, d2, d3, e3+</td>
<td>C/Y Pre</td>
<td>In a non-experimental prospective study 4 children with deafblindness and their assistant were observed after intervention of personal assistant. Study showed increased improvement in interaction and reduction of inappropriate behavior in the child. Best results were found in those children with routines in every-day life.</td>
<td></td>
</tr>
<tr>
<td>Janssen et al (2007)</td>
<td>1</td>
<td>W Pre</td>
<td>Skills</td>
<td>Examine advantage of encoding tactile information. A 40-year-old woman with deafblindness of congenital rubella syndrome. Control group: 8 adults with typical vision and hearing. Two tasks were completed in immediate succession following a fixed sequence, a perception and a memory task. Results showed that the woman performed the tactile perception task more quickly than did any of the sighted and hearing control individuals without making more errors.</td>
<td></td>
</tr>
<tr>
<td>Lee et al (2007)</td>
<td>1244</td>
<td>W+E Pre, Post</td>
<td>Follow up of association between concurrent visual and hearing impairment by National Health Survey. 1244 of 116796 persons had deafblindness. Moderate to severe deafblindness was found to significant increase risk of mortality especially for women.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Reference</td>
<td>Study Design</td>
<td>Data</td>
<td>Personal Skills</td>
<td>Summary</td>
<td></td>
</tr>
<tr>
<td>-------------------------------</td>
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<tr>
<td>Lin et al (2004)</td>
<td>1636 E</td>
<td>d3, d4, d5, d6</td>
<td>In a prospective cohort study was the aim to study cognitive and functional decrease in elderly females with dual sensory loss. Those with dual sensory loss showed increased risk in the beginning but not in the long run of 4 years. The functional and cognitive aggravation is a result of visual impairment and of dual sensory loss lead to less activity capacity.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nikolopoulos et al (2006)</td>
<td>C/Y</td>
<td></td>
<td></td>
<td>Review of accumulated scientific knowledge on ophthalmic disorders in deaf children. 191 papers used. The overall quality of evidence in the literature concerning deaf children and their ophthalmic problems is very low. Prevalence of ophthalmic problems in deaf children is ≈40% - 60% and may remain undetected for years.</td>
<td></td>
</tr>
<tr>
<td>Schneider (2006)</td>
<td>8 W Post</td>
<td></td>
<td></td>
<td>Developing a theoretical framework to explain everyday experiences of adults becoming deafblind. Grounded theory. Interviews and participating observation. Informants experience feelings of interactional powerlessness and actively engage in trying to minimize their powerlessness by working to negotiate a place in this hostile world via interrelated strategies: doing things differently, managing support relationships, surviving others’ perceptions and presenting sides of self.</td>
<td></td>
</tr>
<tr>
<td>Study</td>
<td>Design</td>
<td>Sample Size</td>
<td>Follow-up</td>
<td>Methods</td>
<td>Findings</td>
</tr>
<tr>
<td>------------------------------</td>
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</tr>
<tr>
<td>Shaw (2005)</td>
<td>d1, d2, d3, d6, d7</td>
<td>e2+, e3+, d6, d7</td>
<td>Object was to study grandparent’s (n=80) role in communication development for children with deaf-blindness. Parents n=143. Grandparents were found to be a resource for the child’s language acquisition. Grandmothers were most involved.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sigafoos et al (2008)</td>
<td>d3</td>
<td>e585</td>
<td>Review of 17 papers about teaching augmentative and alternative communication to deaf-blind individuals. Positive outcome was reported in 90%. Evidence for 11/17 papers was inclusive because of methodological weakness.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Soper (2006)</td>
<td>5</td>
<td>Post</td>
<td>In an exploratory design, experiences of improvement of CI by face-to-face semi structured interviews. Body impairments are briefly described, activity limitations before operation is briefly described, type of CI is not described. Present age or age at operation is not described.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sukontharungsee et al (2006)</td>
<td>35</td>
<td>C/Y Pre+ +W Post</td>
<td>d3 d4, e470, e580, e585, d6 d8</td>
<td>Attempt to examine the population of individuals who are deaf-blind in Thailand. Of 130 reported 35 was estimated to have deafblindness. 34% did not have education. Perceived needs of health care, education and service was found</td>
<td></td>
</tr>
<tr>
<td>Vervloed et al (2006)</td>
<td>1</td>
<td>C/Y Pre</td>
<td>Study of interaction between a boy with deafblindness and his personal assistant from a communication perspective. By videotaped observations. Both child and assistant often missed the others initiative. Adults control over communication restrains the child’s language acquisition.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


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These are the publications from the series *Studies from the Swedish Institute for Disability Research*. Each publication is listed with its author, title, and year of publication. The subjects range from reading disabilities to cognitive functions, participation in daily life, and cognitive and auditory impairments among others. The list also includes a range of perspectives from different areas such as education, health, and social sciences.


25. Edvardsson, Tanja (2008). *Consequences of brain tumours from the perspective of the patients and of their next of kin*.