Impact on participation and service for persons with deafblindness
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 Syndrome
USH I  Usher Syndrome 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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SAMMANFATTNING PÅ SVENSKA/Swedish summary

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 länderna används sedan år 2007 termen dövblindhet som en kombination av syn- och hörselfunktionsnedsättningar som begränsar aktiviteter och inskränker delaktighet i samhället i sådan grad att det behövs särskild service, anpassningar i omgivningen och/eller teknologi.


I ICF:s struktur ges möjlighet att beskriva många tänkbara påverkansstrukturer där olika komponenter befinner sig i ett interaktivt samband med varandra. I ICF fokuseras vidare situationer med personer och inte personen som sådan.


Syften
Avhandlingens syften är att belysa, utvärdera och förklara: dels mekanismer som möj-
ligen inskränker delaktighet för personer med dövblindhet och dels mekanismer som
möjligen 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, kroppsfunction, 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 flertalet 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.
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* 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. Hälso risiker 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 gymnasieutbildning 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 omgången. 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 upplevde bristande hänsyn.

Studie IV granskade med hjälp av patientjournals 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 kroppsstruktur och kropps funktion rapporterades. Avvikelseerna 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 tioårsperioden, så fanns det ingen korrelation mellan insats och grad eller takt på syn försämringarna. 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 journalantekningarna. Tidsspannet var ca 50 år. Genom att lägga samman många olika datakällor framkom intressanta mönster där såväl delaktighet som servicens inverkan förändrades över tid.


Studie 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 höras 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 karakter 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 emphasis 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}
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.
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](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

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**FIGURE 1** Based on ICIDH (WHO 1980). ICDH should be seen as a process where the entrance has to be disease or disorder.
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\(^1\) 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

\(^1\) 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 sjukvårdslag, 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 leg-
islation 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 par-
ticipate 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 re-
 sponsibilities 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 men-
tally retarded or have a condition resembling autism; 2. has a considerable and perma-
ent intellectual functional impairment after brain damage when an adult, the impair-
ment 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 age-
ing, 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 spe-
cial service for children and young people who need to live away from their pa-
rental 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
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>Study</th>
<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>
<td>96 references</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>
<td>33 persons (age 19-92, 27 females) (62 % responded)</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</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>
<td>910 notes/2 departments/20 years/9 patients</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>
<td>627 notes/3 departments/≈ 50 years/3 persons and interviews 3 occasions</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.


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 3 Study I–V Methods, Analysis, Age groups* and ICF component**.

<table>
<thead>
<tr>
<th>Study</th>
<th>Methods and time for data collection</th>
<th>Analysis</th>
<th>Age group</th>
<th>ICF</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 E</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 E</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 framework. 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,
Are there any relations between the components in this situation? Does a certain component have impact on something else in the situation?

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 Etiikprüvning 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 turnover 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 reported (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 behaviour 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 mechanisms.

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 afferent 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 itself, 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 deafblindness.

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-
environment. 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 consequence 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>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>e1+, e3, e3+</td>
<td></td>
</tr>
<tr>
<td>Sisson et al (1993)</td>
<td>2</td>
<td>C/Y</td>
<td>Pre</td>
<td>d2</td>
<td>e585+</td>
<td></td>
</tr>
<tr>
<td>Stein et al (1982)</td>
<td>101</td>
<td>C/Y</td>
<td>Pre</td>
<td>d3, d8</td>
<td>e585</td>
<td></td>
</tr>
<tr>
<td>Torrie (1978)</td>
<td>17</td>
<td>C/Y+W</td>
<td>Post</td>
<td>d7</td>
<td>e3</td>
<td></td>
</tr>
<tr>
<td>Study</td>
<td>Study Code</td>
<td>Life Period</td>
<td>Onset of Deafblindness</td>
<td>ICF Component</td>
<td>Factors</td>
<td></td>
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<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>747457</td>
<td>C/Y+W+E</td>
<td>Pre + Post</td>
<td>d1, d6, d8</td>
<td>e3, e5</td>
<td></td>
</tr>
<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></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>Study</td>
<td>Type</td>
<td>Sample Size</td>
<td>Age</td>
<td>Design</td>
<td>Methodology</td>
<td>Summary</td>
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<tr>
<td>Bruce (2005)</td>
<td>No</td>
<td>C/Y Pre</td>
<td>e3+</td>
<td>Personal</td>
<td>Development</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>
<td></td>
</tr>
<tr>
<td>Capella-McDonnell (2005)</td>
<td>E Post d3, d7 e5</td>
<td>447</td>
<td></td>
<td>Personal</td>
<td>Retrospective cross-sectional study of persons with sensory impairment for persons aged 55+</td>
<td>Retrospective cross-sectional study of persons with sensory impairment for persons aged 55+ 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>
</tr>
<tr>
<td>Chia et al (2006)</td>
<td>E Post e1+, e5+</td>
<td>2015</td>
<td></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>
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</tr>
<tr>
<td>Chou &amp; Chi (2004)</td>
<td>E Post d4 e2, e4+</td>
<td>130</td>
<td></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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</tr>
<tr>
<td>Study</td>
<td>Design</td>
<td>Pop.</td>
<td>Measures</td>
<td>Findings</td>
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<tr>
<td>Crews &amp; Campbell (2004)</td>
<td>E Post d2,</td>
<td></td>
<td></td>
<td>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.</td>
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<tr>
<td>Damen et al (2005)</td>
<td>W+E Post d1,</td>
<td></td>
<td></td>
<td>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.</td>
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<tr>
<td>Damen et al (2006)</td>
<td>C/Y Post d1,</td>
<td></td>
<td>Quality of life</td>
<td>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.</td>
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</tbody>
</table>
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.

Janssen (2004) 4 C/Y Pre d1,d2, d3, e3+ Skills  
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.

Janssen et al (2007) 1 W Pre Skills  
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.

Lee et al (2007) 1244 W+E Pre, Post 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.
<table>
<thead>
<tr>
<th>Author(s) (Year)</th>
<th>Design</th>
<th>Time</th>
<th>Sensory Loss</th>
<th>Research Focus</th>
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</thead>
<tbody>
<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>
</tr>
<tr>
<td>Nikolopoulos et al (2006)</td>
<td>C/Y</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>
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</tr>
<tr>
<td>Schneider (2006)</td>
<td>8 W Post</td>
<td>Personal</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>
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<tr>
<td>Reference</td>
<td>Year</td>
<td>Method</td>
<td>Sample Size</td>
<td>Data</td>
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<tr>
<td>Shaw (2005)</td>
<td>2005</td>
<td>d1, e2+, d3, e3+, d6, d7</td>
<td>(n=80)</td>
<td>Object was to study grandparent’s role in communication development for children with deaf-blindness. Grandparents were found to be a resource for the child’s language acquisition. Grandmothers were most involved.</td>
</tr>
<tr>
<td>Sigafoos et al (2008)</td>
<td>2008</td>
<td>d3 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>
</tr>
<tr>
<td>Soper (2006)</td>
<td>2006</td>
<td>5</td>
<td></td>
<td>Post</td>
</tr>
<tr>
<td>Sukontharungsee et al (2006)</td>
<td>2006</td>
<td>C/Y Pre+ C/Y Post</td>
<td>35</td>
<td>d3 e470, d4 e580, d6 e585, d8</td>
</tr>
<tr>
<td>Vervloed et al (2006)</td>
<td>2006</td>
<td>C/Y Pre</td>
<td>1</td>
<td>d1, e1, d3, d7 e3+</td>
</tr>
</tbody>
</table>
The Impact of Genetic Hearing Impairment

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Chapter 8
The impact of combined vision and hearing impairment and of deafblindness

Kerstin Möller

In addition to having a hearing impairment, some people also have a reduction of their vision. The combination of auditory and visual impairments generates joint consequences that have an impact on a person's entire life situation. In this chapter, the term deafblindness is used to signify a functional and social condition, in which a person, due to lack of or reduction in seeing and hearing functions, needs (or will later need) to use touch, smell and taste, and/or to compensate cognitively (memory, conclusion, etc.) when communicating, receiving information and orienting themselves.

The purpose of this chapter is to give an account of the psychosocial effects of deafblindness in a life perspective. The chapter begins with a short description of people with deafblindness. Opinion is divided about how to define the group, and a brief account of major features characterizing this discussion is provided. A short historical presentation follows, centring on education and rehabilitation. The subsequent section on participation focuses on areas such as learning, communication, mobility, interpersonal interactions and relationships, and major life areas such as work, leisure and recreation. Environmental factors are crucial to opportunities for participation of people with deafblindness. Sounds, light and other circumstances in the environment, assistive devices, individual support and services could make possible, facilitate, impede or prevent participation by people with deafblindness. In addition to issues considered in the ICF, some further crucial matters related to deafblindness are discussed. The chapter concludes with a discussion. Issues related to the various domains are reported first for the entire group, then for people with prelingual onset of deafblindness and, finally, for those with postlingual onset.
A brief account of people with deafblindness

There are a number of genetic disorders that may cause combined reduction of vision and hearing, the most common being Usher syndrome. Moreover, during the course of life, various conditions, such as trauma, infections, viruses, drug injuries and tumours can cause loss of vision and hearing. Age-related vision and hearing impairments also occur. People with deafblindness sometimes also have additional impairments (McInnes and Trefftz, 1982; Stein, Palmer and Weinberg, 1982; Fredericks and Baldwin, 1987).

The aetiologies of deafblindness in 57 pupils, 5–20 years of age in 1998/99, were compared with aetiologies in 49 pupils in 1986/87. In addition, 55 preschool children with deafness in 1998 were compared to 68 preschool children with deafness in 1988. When conducting this study, Admiraal and Huygen (2000) found several cases of unusual hereditary syndromes. Occurrences of acquired deafness – in particular congenital rubella – had decreased during the period, whereas occurrences of prenatal causes had increased. Moreover, the proportion of pupils with multiple impairments had increased from 25 to 38%. During the period 1972–79, Stein, Palmer and Weinberg (1982) studied 141 children (aged 0–16) with deafblindness and found a large proportion with additional impairments. Testing the children’s hearing, they furthermore found children who could be excluded from the list of those with deafblindness.

People with deafblindness rarely have complete loss of both vision and hearing. Given the definition of deafblindness used, according to some analyses no more than 6% had both deafness and blindness: the vast majority had residual vision and/or hearing (Fredericks and Baldwin, 1987; Wolf-Schein, 1989). Among people with combined vision and hearing impairments, however, change and deterioration of remaining functions are frequent. People with combined vision and hearing impairments – i.e. deafblindness – therefore constitute a heterogeneous group (Ingraham, Daugherty and Gorrafa, 1995) which is usually subdivided according to age of onset or the main impairment.

The onset of deafblindness, before or during the development of language skills (prelingual onset), has an impact on a child’s potential to develop psychologically and socially. This is often referred to as congenital deafblindness. Onset when language has been acquired (postlingual onset) is accompanied by altered life conditions and is usually called acquired deafblindness. People with onset of deafblindness in old age are usually referred to as the elderly group. Some individuals may have contextually conditioned deafblindness – depending on the function in question in combination with specific circumstances – and hence need services aimed at people with deafblindness. People with severe vision or hearing loss and people with medical diagnoses
in which reduction in vision or hearing may occur, should be considered deafblindness risk groups. There are furthermore accounts of a few cases of factitious deafblindness (Miner and Feldman, 1998).

**Definition of deafblindness**

The concept of deafblindness does not refer to the medical terms deaf and blind, respectively, but is based on a combination of functional impairments that generate psychosocial outcomes. In the debate on how to define deafblindness, some authors stress issues related to services and education whereas others stress the degree of impairment. The situation is further complicated by the heterogeneity of the group and the way various educational systems and systems of services define their scopes of authority (McInnes and Treffry, 1982; Fredericks and Baldwin, 1987; Reed and Fontan, 1987; Wolf-Schein, 1989; Vernon and Duncan, 1990; Ingraham, Daugherty and Gorrafa, 1995; Marchant, 1996; Brennan, 1997; Aitken, 2000; Rönnberg and Borg, 2001; Bruce, 2002). In this chapter an extensive interpretation of the concept is used, relating to impairments in combination with psychosocial consequences. This understanding of the concept means including, for instance, children with Usher syndrome and elderly people with residual vision and hearing.

**Historical perspective**

The possibility of teaching and educating children with deafblindness seems to have been recognized for the first time by l’Abbé Deschamps, who wrote in 1779 that the first step was to give the pupils some idea of what was being pursued. He was convinced that it was possible to make pupils understand that when we want something we move our lips to ask for it. The next step was to consider how to proceed with education. In 1795 Lorenzo Hervas y Panduro outlined a method using signs of touch, smell or taste (Enersv’estdt, 1996).

From the beginning of the nineteenth century there are some reports on teaching pupils with deafblindness. The pioneers and their pupils became world-famous. Some of them were associated with schools for pupils with deafness and others with schools for pupils with blindness. The idea of using fingerspelling was taken from the education of the deaf, and the idea of using letters in relief from the education of the blind (Enersv’estdt, 1996).

The first special school in Sweden for pupils with deafblindness was set up in 1886. The headmistress, Elisabeth Anrep-Nordin, was a teacher with professional training in special education for the deaf. Before the school opened she travelled to the USA and Germany to study. The Swedish school was based on the assumptions that pupils with deafblindness were, firstly, good by nature and, secondly, able to learn. The school had three educational goals: to teach Christianity, to train skills necessary to perform a job and to
master a language. These were similar to the goals of regular schools at that time. The educational, individualized, theoretical approach was inspired by Fröbel in Germany. Methods of language teaching were akin to methods for the deaf (Liljedahl, 1993). After the 1880 Milan conference deaf education in Sweden changed from the signing to the oral method (Erikkson, 1998). However, pupils with deafblindness were allowed to use signs - and had therefore to be kept separate from pupils with deafness, as they might otherwise exercise a bad influence. At the outset, the idea was to teach pupils to learn to talk, as they were supposed to return to their home district after school. Ragnhild Kaata, a child with deafblindness from Norway, was trained in a school for the deaf and learned to speak by holding her hand over the mouth of the speaker (Liljedahl, 1993; Enerstvedt, 1996). Handicraft, and especially weaving, had multiple purposes in the school for pupils with deafblindness in Sweden: obedience, learning a profession and keeping the hands busy to prevent low mood. A further goal of handicraft production was to obtain money for the school (Liljedahl, 1993).

The study by Liljedahl, referred to above, reveals that there are differences in early opinions about educability of pupils with deafblindness. The first school for pupils with deafblindness in Sweden was based on the assumption that pupils were educable, but Goode (1994) maintains that early opinions were that pupils with prelingual onset of deafblindness were uneducable.

Some authors describe a more recent change in the approach to people with deafblindness from the 1970s to the 1980s. While, for instance, deafblindness was earlier written and talked about as a quality of the child, now the child is referred to as a child with impairments. Such an alteration in attitudes also affects the sort of community a person with deafblindness is expected to live in. The new approach is based on the idea that children with deafblindness are expected to be together with their age peers, and adults with deafblindness are expected to live in their communities and not in protected institutional environments (Fredericks and Baldwin, 1987; Marchant, 1996). These changed attitudes have had an impact on the locations used for training activities. Earlier, training took place in special premises at institutions, but now rehabilitation increasingly takes place in natural environments, for example on the pavement going to school, in the fast-food restaurant, etc. (Gee, Harrell and Rosenberg, 1987).

In the 1960s, there were epidemics of rubella in the USA and Europe and as a result more children with deafblindness of prelingual onset were born than had ever previously been the case (Vernon and Duncan, 1990; vanDijk and Nelson, 1997). McInnes and Treffry (1982) made an important contribution to the new attitudes previously mentioned. The rubella epidemic generated a need for special education. Europeans looked for support from the USA. For example, van Dijk reported receiving support in particular from the
Perkins School for the Blind. According to van Dijk, knowledge acquired at this school was later to influence most western European countries (van Dijk and Nelson, 1997). See, further, Enservedet(1996) for a historical overview of education of children with deafblindness.

**Activities and participation (disabilities)**

**Learning and applying knowledge (d1)**

Deafblindness, as such, severely affects the educational context and is very demanding as far as educational, auxiliary means and attitudes are concerned whether or not the person with deafblindness has additional impairments such as intellectual disability. Pupils with deafblindness may receive special education in segregated schools or be integrated into normal schools. They may also follow general education at all levels.

Most of the spontaneous learning acquired by seeing and hearing children is disturbed by loss or substantial reduction of both distance senses. The very initial learning processes, as well as establishing relationships with parents and others, are seriously affected by prelingual onset of deafblindness (Mar and Sall, 1995; Marks, 1998). Concluding a summary of theories on van Dijk's methods concerning children with prelingual onset of deafblindness, MacFarland (1995) noted that initially this is all about relating and security, just as it is for all other children.

**Communication (d3)**

The point of departure in this section is the assumption that all human beings communicate, either intentionally (with or without formal language) or unintentionally. One consequence common to everyone with deafblindness is that communication is affected. Expressive communication (to express oneself) as well as - and in particular - receptive communication (to receive messages) is affected. Languages, spoken or signed, as well as communicative modalities, vary as a function of age at onset and the degree of hearing and vision losses, as well as other factors. Some people have grown up in the Deaf community and have sign language as their first language. Others have oral language as their first language (Pollard, Miner and Cioffi, 2000). The language skills of people with deafblindness range from unintentional communication to the skills of people with a university degree. Research summaries on communication with people who have deafblindness demonstrate, among other things, the heterogeneity of the group and the variety of communication methods used (Engleman, Griffin and Wheeler, 1998; Rönberg and Borg, 2001). Communicating with people with deafblindness is very demanding in terms of the interaction partner's knowledge, skills and
attitudes. However, research on communication stress on the part of the communication partners seems to be lacking.

All people with deafblindness need individualized and professional assessment of their communicative skills and appropriate individualized plans of rehabilitation, which will possibly have to be reconsidered later on in relation to altered and further reduced seeing and hearing functions (Godfrey and Costello, 1995).

Non-symbolic communication here connotes transferring messages without using symbols such as words, signs or graphics. Instead, bodily movements, facial expressions, sounds, breathing, glances, change in voice, etc. are used (Siegel-Causey and Downing, 1987). These authors deliberately do not use the expression ‘prelingual communication’ but rather use ‘non-symbolic communication’ because some people never reach the prelingual or speech level in communication.

Communication based on speech or on sign language has been very controversial as far as children with early onset of deafness are concerned, and this applies also to those children with deafness and reduced vision. Tadoma is a method where the person with deafblindness seeks to tactually feel the speech, for instance by putting a hand over the speaker’s mouth or on his or her throat. The use of the Tadoma method has been evaluated (Reed et al., 1985, 1990). Use of sign language in the tactual version is dealt with, in particular, in studies by Reed et al. (1995) and Mesch (1998). Reed and colleagues have also evaluated the use of finger-spelling in the tactual version (Reed et al., 1990). Earlier it was not considered necessary for children with prelingual onset of deafblindness to learn formal signing. However, in accordance with the altered approach to deafblindness, and as these children begin to move around in the community, formal signs are increasingly introduced (van Dijk and Nelson, 1997).

Mutual glances, as a natural beginning of mutual communication and interaction between child-carer and child, will not appear in children with prelingual onset of deafblindness (Chen and Haney, 1995). There is an imminent danger that the child will go into a condition of introversion with autism-like symptoms. If communication is at an unintentional level, it is necessary to establish a mutual emotional involvement. From this point it is possible to be guided by the expressions of the person with deafblindness (Naftstad and Rödstrøe, 1999). The more introverted the child or adult becomes, the more difficult it is to discover and interpret the communicative signals they send out and to respond to them in a way that they can understand (Siegel-Causey and Downing, 1987; Chen and Haney, 1995; van Dijk and Nelson, 1997; Bruce, 2003). When the child has grasped that their expression has evoked a response, intentional communication can commence (Siegel-Causey and Downing, 1987; Chen and Haney, 1995).
Several authors give accounts of intentional non-symbolic communication with people who have deafblindness. At the end of the 1970s, Goode (1994) made participant observations of two children with deafblindness and without language. He demonstrated how the children communicated in a number of ways with bodily gestures, sounds and glances. One of the children (Christina) lived in a state institution. According to Goode, at that time opinion was that children with deafblindness and intellectual disability (caused by rubella) were not educable, and hence no efforts were made to interpret the children’s expressions. Goode’s ambition to understand and share experiences with such a child was met with much scepticism and distrust. The other child (Bianca) lived with her parents as a much-valued member of the family. Bianca’s parents made efforts to interpret her communicative expressions (stamping her foot to have some more fruit, for example) and comply with her wishes. At the same time, however, there were conflicts between Bianca’s parents and her teachers because the teachers were of the opinion that Bianca could not communicate since she could not talk.

Half of the young people in Petroff’s (2001) study communicated on a non-symbolic level. Most of those who used spoken language also received spoken language, and they also had a larger vocabulary than those using sign language. Petroff furthermore found a relationship between the communication method and walking. Youngsters unable to communicate using language (signed or spoken) were also unable to walk independently, whereas those with language also walked independently.

Some reports deal with difficulties in communication encountered by people with deafblindness who have a language. Sign languages are visual languages that can be adjusted and modified for users with limited peripheral vision or for tactual use (Godfrey and Costello, 1995; Mesch, 1998). Some examples of adjustments are: the speaker is at a certain distance; contrasting colours in the background and contrasting colours on speaker’s/interpreter’s clothing; lighting and positioning in relation to light (Gioffi, 1996). People with deafness since childhood can have poor knowledge of the language spoken and written in their country. Such shortcomings should not be confused with intellectual disability or low intelligence, but are related to the deafblindness of these people and to the kind of modified education they have (or have not) received (Godfrey and Costello, 1995; Brennan, 1997). Miner (1995) observed that it is important for children with Usher syndrome to learn sign language in its tactual version, considering these children’s problem with dark adaptation, and this also applies to others with fluctuating or progressive deterioration of vision. She furthermore maintains that people with Usher syndrome may also need to use sign language in tactual version after cochlear implantation, in order both to express themselves and to
receive information. Depending upon their particular background, people with deafblindness associate with either hearing or deaf subcultures. Considering the sensitivity to the issue of vision within the Deaf community, it is understandable that transition from visual to tactual sign language is made stepwise and can be traumatic (Brennan, 1997).

Godfrey and Costello (1995) give examples of communication methods for people with deafblindness who have spoken language as their first language and whose hearing is deteriorating, as is the case in Usher syndrome type 3. Miner (1997, 1999) considered the same issue in people with decreasing vision, as is the case with Usher syndrome type 2. Increasing difficulties in receiving spoken language may occur if these people previously used to combine residual hearing function with lip-reading. Godfrey and Costello point to the possibility of writing letters on the palm, perhaps in combination with using signs in tactual version, Braille, etc. (Godfrey and Costello, 1995).

It is clear that people with deafblindness sometimes use more than one modality of language and communication. In one study, 7 out of 13 people used 2 methods and 4 of them (all deaf) used both sign language and spoken language. All those making use of just one method of communication used sign language. Generally, the participants in this study reported that they had difficulties in communicating in about half of communication circumstances, and this was so in communicating about thoughts as well as feelings (Rönnberg, Samuelsson and Borg, 2002). Murdoch (1994) video-recorded interaction between children with deafblindness and one hearing and seeing partner, and observed that the partner did not always respond by using the same method as the child and also that several communication modes were used simultaneously.

**Mobility (d4)**

Mobility - and in particular mobility in unfamiliar surroundings - is difficult for people with deafblindness. The pathological picture of some of the syndromes generating deafblindness (for instance, Usher syndrome type 1) includes disordered balance (Kimberling and Möller, 1995). Some people with deafblindness and intellectual disability can also have mobility difficulties even in familiar settings indoors. Lancioni and Mantini (1999) described a woman with total blindness who frequently lost her orientation while moving around indoors because of her balance disorder. People in her situation may need specific technical devices and/or personal guides to find their way.

Miner (1995) described a young woman with Usher syndrome type 1, who had had a cochlear implant, having mobility training in darkness. The trainer did not understand that the woman's hearing with the cochlear
implant did not give her sufficient information about the traffic situation to guarantee safe mobility. The woman felt that her fears and difficulties in crossing the street were belittled. This emphasizes the importance of instructors in mobility having knowledge of deafblindness.

In some cases people with postlingual onset of deafblindness are able to use public transport. The possibilities depend on their remaining seeing and hearing functions, their ability to absorb information from other senses (for instance, feeling the wind), their ability to orient themselves and to solve problems, and good knowledge of the language spoken around them, including the ability to express themselves in an intelligible way (Vernon and Duncan, 1990; Cioffi, 1996). In an experiment, a person with deafblindness asked for assistance, with the help of a written note, to cross a street at a street crossing. Pedestrians not offering assistance in most cases stated that they had not noticed that this person needed help (Sauerburger and Jones, 1997; Franklin and Bourquin, 2000).

The kind of medical examination required for a driver's licence is not, in all countries, sufficient to exclude people with social blindness related to decreasing peripheral vision (Tedder, 1987). People who do not know about their visual impairment (for example, in cases of late diagnosis of Usher syndrome) can find themselves involved in traffic accidents when driving (Hicks, 1978). Tedder (1987) points out the need to counsel young people and adults on responsible attitudes to driving in relation to their reduced vision. Giving up driving is one of the most difficult experiences for a person with deafblindness. When they have to stop driving, further consequences are triggered off within all life areas, not only those related to transportation, and stress and grief can result (Brennan, 1997).

**Domestic life (d6)**

In the 1970s, the general position in the USA and Europe was that people with deafblindness should be cared for in institutions. Now, their life is normalized to an increasing extent and they live in less sheltered environments: with parents or independently. No studies dealing with housing etc. for people with postlingual onset of deafblindness have been found. Research in progress, as well as personal contacts with people with postlingual onset of deafblindness in a number of European countries and in other parts of the world, indicates that these people usually live in their own homes and are (or have been) working. Sometimes they also have a family and children, with or without deafblindness.

In a study by Pteroff (2001) more than half of the youngsters lived with their parents and just a few lived independently. Various studies indicate that parents of children with prelingual onset of deafblindness do not think that their child will live independently in the future (Wolf-Schein, 1989; Pteroff,
2001). Moreover, parents of children or young people with postlingual onset of deafblindness, for instance with Usher syndrome, vary in their opinion about whether their children could manage a household by themselves (Miner, 1995).

From a gender perspective, difficulties in independently managing household tasks, such as cooking and cleaning, might be a worse loss for a woman than for a man. If so, such practical barriers have an emotional significance similar to that of being no longer able to drive (Miner, 1995).

**Interpersonal interactions and relationships (d7)**

Common to all people with deafblindness is the fact that problems of communication make interpersonal interactions more difficult, particularly for people with prelingual onset of deafblindness.

Mar and Sall (1995) conducted intervention research, focusing on three children with deafblindness. The aim was to examine the children's engagement in social activities with their age peers without impairments, with the purpose of implementing improvements. Two of the children had intellectual disability in addition to deafblindness and communicated intentionally on a non-symbolic level (gestures, facial expressions, etc.). The third child expressed himself through spoken language and could also receive speech with the help of a hearing aid and an FM system. Initially the study found that the children – including the child with spoken language – had relatively few friends. Any friends were as a rule associated with specific situations, such as the church or the immediate neighbourhood. The child with speech, who was in a regular class at the local school, did not have adequate social contacts at school either. During the course of the study, the children's interaction with their age peers and new acquaintances increased but it was not possible to decide whether this was due to the intervention or other circumstances. Petroff (2001) also indicated occurrence of social isolation among young people with deafblindness.

One study of siblings' interactions with their brothers and sisters with deafblindness indicated unequal roles, with the child without deafblindness frequently taking a supporting role. The more difficulties siblings had in communicating with each other, the more joint activities were negatively affected. Many siblings did not play with the brother or sister with deafblindness, or even avoided them. When playing games with friends, more than half of the siblings did not include their brother or sister with deafblindness in these activities. Nor did siblings adjust their behaviour by, for instance, providing information about things going on in the surroundings or during transportation. Seeing and hearing siblings were furthermore usually self-taught about ways of communicating with their sibling with deafblindness.
They also expressed wishes to meet other children who had siblings with deafblindness. Writers stress the need of further research into this area and the need for information and training of siblings by professionals, in particular support to improve communication with their siblings with deafblindness (Heller et al., 1999).

Regarding postlingual onset of deafblindness, there are contradictory reports about interaction within the family. There are some reports about families with children with Usher syndrome, for example, where parents learnt sign language (Torrie, 1978; Miner, 1995), but there are also reports of parents who can barely or not at all communicate with their children (Miner, 1995; Brennan, 1997). Young people and adults with Usher syndrome may encounter increasing difficulties in reading their parent’s lips when their vision deteriorates and, if the parents do not know sign language, communication is made more difficult (Miner, 1995).

Relationships between children or young people with Usher syndrome and their age peers may be affected by decreasing peripheral vision, associated with Usher syndrome, when a person with deafblindness does not notice that another person has opened a conversation. The person with deafblindness could then be thought of as rude or stupid and could even be abused (Vernon and Duncan, 1990; Miner, 1995). Pupils with deafblindness are reported to be socially isolated from age peers when integrated into regular schools and also when attending special schools (Ingraham, Daugherty and Gorrafa, 1995; Miner, 1995). Attention has also been paid to matters concerning information, training skills and giving support to age peers of young people with deafblindness (Brennan, 1997; Goetz and O’Farrell, 1999). Social interaction for pupils with deafblindness can be improved through powerful support and information to their peers (Mar and Sall, 1995).

People with Usher syndrome (types 1 and 2) have reported that friends withdrew when their seeing functions deteriorated (Miner, 1997, 1999).

A number of researchers stress the importance of creating opportunities for people with deafblindness to meet with others who also have deafblindness (Hammer, 1978; Fillman, Leguire and Sheridan, 1989; Vernon and Duncan, 1990; Miner, 1995; Brennan, 1997).

Miner (1995) gives accounts of difficulties encountered by seeing and hearing children when parents have Usher syndrome type 1. The children have felt the responsibility of interpreting for their parent(s) to be too much for their age (Miner, 1995).

Tedder (1987) maintains that there are risks of physical and psychological violence as well as broken relationships, but the research examined in this chapter provided no information about incidents of violence.
Major life areas (d8)

Education (d810)

Petroff (2001) conducted a survey in the USA, directed at parents of young people (aged 18–24) with deafblindness who left school in 1996. The youngsters’ last educational placement had been segregated special education. In this study half of the pupils had left school because they had reached the maximum age and barely half had graduated with a diploma. After leaving school more than half of the pupils were still illiterate. Miner (1997) maintains that pupils with Usher syndrome type 2, and having hearing loss, either attend special schools for pupils with deafness or are integrated into regular local schools.

A number of studies have demonstrated that pupils with deafblindness do not always have access to specially trained teachers, and also how such circumstances affect the pupils’ possibilities of learning (Ingram, Daugherty and Gorrafa, 1995; Kirchner and Diament, 1999a; Bruce, 2002). Ingram and colleagues give an example of how substantial support can be provided for pupils with deafblindness, guided by pupils’ present and future needs. Cooperation between various authorities, as well as flexible solutions to financing support services, were required and implemented. Teachers needed to prepare lessons several days in advance, as the lessons had to be interpreted and educational material had to be written out in Braille, for instance. Special resource teachers played important roles in developing education. Authors repeatedly stress the importance of considering the specific needs of each and every pupil. The pupils referred to in this example were subsequently moved from special school settings to their local general school. When considering educational placement, issues regarding risks of isolation also have to be taken into account (Ingraham, Daugherty and Gorrafa, 1995).

In Petroff’s study (2001), 17% of pupils had received post-secondary education but none of them attended a 4-year college. Participants were provided with trained deafblind counsellors at school. Other studies report people with deafblindness having post-secondary and university education (Enerstvedt, 1996; Rönberg, Samuelsson and Borg, 2002).

As a result of their deafblindness, many young people and adults have inadequate all-round education and therefore lack knowledge of, for instance, matters related to sexuality. Even people who have acquired deafblindness later in life, and people who have been married and have brought up children, can need special adult education. The Helen Keller National Center in the USA provides an example of tailored sexual education. Information is provided in spoken language, in sign language and also in written form using the most appropriate means: for instance Braille, photographs, large letters. Models of female and male anatomy are used to
name parts of the body, to prepare for gynaecological examination and to provide information about instruments and procedures that will be used during examination, as well as information about safe sex and contraceptive counselling. The education could also deal with masturbation methods, and information on when and where masturbation is appropriate. Students are also informed about parts of the body that are appropriate or not appropriate for touching. If they need help with hygiene and personal care, they are informed about circumstances when it may be right for assistants and interpreters to give tactile support, and when not. Information on relational issues and pregnancy is also included. The authors emphasize that sexual education of people with deafblindness must be viewed in a life perspective and be provided throughout the course of the person’s life (Ingraham et al., 2000).

Work (d840)

Matters regarding the labour market and vocational training are rarely dealt with. Research into the occurrence of employment among adults with deafblindness, the kind of jobs they have, experiences of work, relations to workmates, etc. is still lacking.

As with the issue of housing, parents of youngsters with deafblindness seem not to believe that their children will be able to manage a job (Wolf-Schein, 1989; Petroff, 2001).

One study of supported employment demonstrates how work preferences can be tested and considered before choosing work tasks and work placement for a person with prelingual onset of deafblindness and without language (Parsons et al., 1998). Petroff (2001) writes that only 8% of the young people in his study were reported to have had vocational training in competitive or supported work settings as part of their secondary education. In general, long-term vocational rehabilitation, adjustment of the workplace, technical devices, etc. are important preconditions for the prospective employment (Vernon and Duncan, 1990; Miner, 1995; Petroff, 2001). Bourquin, Mascia and Rusenski (2002) examined a service programme for adults with postlingual onset of deafblindness aiming at employment. The quantity and quality of services delivered were usually dependent upon issues that were not related to the vocational rehabilitation process as such, but rather to matters such as transport to and from any future workplace which frequently impeded success. Marchant (1996) gave an account of another kind of workplace-related support: workmates of a man with deafblindness were given information about deafblindness and were taught sign language, and subsequently accepted the man.

In spite of various measures to promote employment, most people with deafblindness are unemployed: more than 80% of the participants in Petroff's (2001) study.
For people with postlingual onset of deafblindness, it is imperative to obtain an accurate diagnosis as soon as possible. Vocational counselling and choice of profession are important (Davenport et al., 1978), and also complicate matters because of difficulties in making prognoses about when and how rapidly vision and/or hearing will deteriorate (Miner, 1995). There are cases of people with Usher syndrome who have invested much time and money in vocational training, which they later had to abandon when their vision deteriorated. They had not been informed about their diagnosis or prognosis (Hicks, 1978). Vernon (1969) mentions that many people with Usher syndrome are forced to stop working before retirement age because of progressive visual impairment. There has been no research focusing on the labour market experiences of people with postlingual onset of deafblindness and hence we do not know much about this.

**Leisure and recreation (d920)**

Access to leisure-time activities for people with deafblindness can be dependent on services (for instance, guides or transportation), as well as the skills of the specific person. Several studies deal with the importance of physical activities and the limited opportunities for people with deafblindness to engage in such sports as bowling, swimming, walks, etc. (Hammer, 1978; Vernon and Duncan, 1990; Petroff, 2001). Children with deafblindness were included in one study dealing with physical activities and fitness of children with visual impairments, and findings were then compared to those of children without visual impairments. Children with a double impairment were significantly worse off (Liberman and Hugh, 2001).

The most frequent leisure-time activities in a study of adults with postlingual onset of deafblindness were walking, swimming, computer work and reading. Even if most of them participated in some activity, 60% were not satisfied with their current level of participation in recreational activities. They wanted, for instance, social activities, activities requiring good physical shape, dancing and boat trips (Liberman and Stuart, 2002). The low level of participation was due to lack of transportation, co-participants, suitable programmes, guides or time, and to negative attitudes (Reed and Fontan, 1987; Petroff, 2001; Liberman and Stuart, 2002).

People with Usher syndrome and similar visual impairments encounter specific problems because of their decreasing peripheral visual field, light sensitivity and contrast sensitivity (Miner, 1995). Engagement in associations where speakers make use of sign language can also cause problems: people with decreased peripheral vision may have difficulty in tracking the shifting turns of speakers, and can therefore find themselves becoming outsiders (Miner, 1995). Furthermore, they may withdraw from evening activities because of difficulties with dark adaptation (Vernon and Duncan, 1990).
Environmental factors

Environmental factors can facilitate as well as act as barriers for people with deafblindness.

Products and technology (e1)

Technical devices - in addition to hearing aids, FM systems and cochlear implantation to compensate for hearing impairments, and glasses and a white stick in the case of visual impairments - can provide assistance for people with deafblindness. Assistive devices specific to deafblindness and unusual devices are at the centre of the following discussion.

Research and development of vibratory devices to facilitate perception are in progress (Borg, 1997; Borg, Neovius and Kjellander, 2001; Borg, Rönnberg and Neovius, 2001). These devices have also been used to facilitate indoor mobility (Lancioni and Mantini, 1999). When deafblind subjects were asked to rank assistive devices, optical and vibrotactile signals were the most preferred (Rönnberg, Samuelsson and Borg, 2002).

An auxiliary means of independent mobility is the use of guide dogs specially trained for people with deafblindness. Dogs can also be trained to pay attention to and localize sounds. However, 'hearing dogs' cannot be used for mobility and vice versa (Vernon and Duncan, 1990).

An example of low-tech assistive equipment is the use of photographs. Books with photographs of a pupil's everyday surroundings and of motivating events - favourite activities such as playing in the pool - have been used for interactive exchange (Goetz and O'Farrell, 1999).

For people with deafblindness who know how to write, textphone (stationary) and Tellatuch are useful devices. Tellatuch is a small portable apparatus on which messages to a person with deafblindness are written using a keyboard while the receiver reads messages in Braille (Vernon and Duncan, 1990). It has provided possibilities of obtaining information through the internet and making contact by e-mail, all of which is a new research field, not yet entered (Miner, 1999).

Assistive devices are useful only when they work. Devices that are difficult to use, or broken or otherwise non-functional, can have disastrous consequences, as has been demonstrated in a study of pupils (Giangreco, Edelman and Nelson, 1998).

Natural environment and human-made changes to environment (e2)

In several causes of deafblindness, for instance Usher syndrome, seeing functions alter when light conditions change; for instance, dazzle from snow or bright sunshine, haze, mist, dusk and darkness can cause problems. The
use of ultraviolet filters cannot fully compensate for such changes in the environment. Sensitivity to contrasts and light can make a green board with white chalk most difficult to perceive, and a white board can be dazzling (Fillman, Leguire and Sheridan, 1989).

Support and relationships (e3)

Research dealing with support and relationships is included in the earlier section on 'Interpersonal interactions and relationships'. Here, the question of professional support is considered.

Some authors have demonstrated the positive impact of interveners (a kind of personal assistant). Programmes with interveners for children as well as adults with prelingual onset of deafblindness have been evaluated. Progress was reported for people with deafblindness, and their parents also viewed the measure in a positive way (Watkins et al., 1994; Hammer and Carlsson, 1996).

Some reports deal with the significance of interpreters and the specific requirements for interpreters interpreting for people with deafblindness as compared to interpreting between spoken language and sign language for people with deafness (Petronio, 1988; Bourquin, 1996). People with deafblindness have specific needs that vary from person to person and sometimes between various settings of interpretation. When interpreting for people with deafblindness, the interpreter has to be flexible and aware of a number of conditions that have to be taken into consideration (Petronio, 1988). Even if interpretation is visual, the scope of signs and also the particular signs used have to be adjusted and the interpretation has to be supplemented with information about events and objects in the environment, etc. (Petronio, 1988).

Attitudes (e4)

Unlike people with hearing impairments, people with deafness who are members of the Deaf community usually do not view their deafness as a disability but rather as part of their identity and a source of pride and support (Miner, 1995, 1997). Visual impairments are, however, particularly traumatic to people with deafness because of the importance of vision for their communication and perception. Moreover, as Gioffi (1996) puts it, there is an almost phobic fear of blindness in the Deaf community. Young people in the Deaf community may deny any visual impairment and the need for specific adaptations due to loss of vision, use of white stick, mobility training, etc. (Miner, 1995; Gioffi, 1996).

There are contradictory reports on relationships and interaction between people with deafblindness and people with only deafness. Brennan (1997) maintains that, even though most people with deafness know someone with
deafblindness, they usually have no patience with people who use tactual sign language. Vernon and Duncan (1990), on the other hand, point out that people with deafness easily understand the kind of problems that light sensitivity generates for those with Usher syndrome and can be supportive if they are informed in advance.

Given that deafblindness is relatively rare, most professionals may not have much knowledge of it, or about how to use sign language. Frequently professionals turn to parents or the guide and not to the person with deafblindness. If the parents do not know sign language, the chances are that the person with deafblindness is treated like a child and people talk above her or his head (Miner, 1995). Well-educated people, people with high professional positions, parents and others who have deafblindness report experiences of other people looking down on them or patronizing them (Miner, 1995).

Services, systems and policies (c5)

People with deafblindness are to a large extent dependent upon services and support in order to live an active and participating life. The present literature review reveals considerable inadequacies in these respects.

Petroff's (2001) study reports on services received by young people during their last year at school. Only 60% had received speech and language services - a low number considering that a large proportion of the youngsters did not communicate using formal language. Only about 30% had received orientation and mobility services, even though about 50% of the youngsters in the group under study moved around on their own.

As far as preparations for adult life are concerned, 40% of the young people in Petroff's (2001) study lacked a written, individual plan of transition and almost a quarter had none whatsoever.

Several authors report insufficient coordination of services; moreover, people who supplied services lacked knowledge about deafblindness (Wolf-Schein, 1989; Goode, 1994; Miner, 1995, 1997; Giangreco et al., 1999; Kirchner and Diament, 1999b; Petroff, 2001; Bourquin, Mascia and Rusenski, 2002; Janssen, Riksen-Walraven and van Dijk, 2002). Giangreco (2000) summarizes various methods of providing services. Some reports point out problems with large staff turnover and burnout symptoms among those professionals working closest to people with deafblindness. In a 4-year project, comprising 358 professionals working with 18 pupils with deafblindness, only 29 remained for the entire project and, of these, 10 did not come into contact with the pupil more than about once a month (Giangreco et al., 1999) Giangreco and colleagues also point out risks of conflict between specially trained personnel and, for instance, teachers of children integrated into regular schools (Goode, 1994; Giangreco, Edelman and Nelson, 1998).
Personal factors

Psychological and mental conditions

Murdoch found several difficulties in testing the mental development of children with deafblindness: the instruments assumed that the child could either hear or see (Murdoch, 1994).

For people with prelingual onset of deafblindness, there are some 25 reports about behavioural disorders such as repetitive habits (swinging the body, bringing fingers in front of eyes), self-injury (biting or scratching oneself), aggressive behaviour towards others (scratching, biting, pinching) and - common to most people with deafblindness - introversion (Tweedie, 1974; Yarnall and Dodginson-Ensor, 1980; Rapoff, Altman and Christopherson-Edward, 1980; McInnes and Treffry, 1982; Fewell and Rich, 1987; Fredericks and Baldwin, 1987; Lancia et al., 1991; Myrbakk, 1991; Peine et al., 1991; Romer and Schoenberg, 1991; Luiselli, 1992; Sisson, Hersen and Van Hasselt, 1993; Goode, 1994; Watkins et al., 1994; Chen and Haney, 1995; van Dijk and Nelson, 1997; Nafstad and Rödbroe, 1999, to mention only some). It is easy to be misled and believe that behavioural disorders are associated with prelingual onset of deafblindness. However, most of the research focuses on just one or a few cases of behavioural disorders and does not provide information about prevalence. In Petroff's (2001) study, about half of the youngsters were reported to have problematic or challenging behaviour.

There are also many reports about measures to reduce or halt behavioural disorders. Such measures include throwing water on the child (Peine et al., 1991), smells (Rapoff, Altman and Christopherson-Edward, 1980; Gross, 1994), tastes (McDaniel, Kocim and Barton, 1984), technical devices (Lancia et al., 1991), positive reinforcement and encouragement (Yarnall, 1980; Luiselli and Loilli, 1987; Luiselli, 1988; Myrbakk, 1991; Sisson, Hersen and Van Hasselt, 1993) and medication (Luiselli, 1991).

Some studies, concentrating on environmental factors, point out that certain treatments generate stress in children with prelingual onset of deafblindness. If the stress is not removed, behavioural disorders will result (Tweedie, 1974; McInnes and Treffry, 1982; van Dijk and Nelson, 1997). Misunderstandings or abortive efforts to communicate could lead to frustration and trigger off aggressive or self-injuring behaviour (Janssen, Riksen-Walraven and van Dijk, 2002). Other studies indicate that behavioural disorders can decrease or cease as a result of changes in the behaviour of seeing and hearing adults (van Dijk and Nelson, 1997; Marks, 1998; Janssen, Riksen-Walraven and van Dijk, 2002, 2003).

There are some studies dealing with psychological issues relating to the postlingual onset of deafblindness. Vernon and Hammer (1996) argue that
various measures and tests in the field of psychology are not suitable for people with deafblindness. The difficulties of communication caused by deafblindness are not related to a person’s mental capacity.

There are different opinions on psychiatric illness as a common complication in the case of people with Usher syndrome. There are reports of psychological complications in cases of Usher syndrome, but there is no proof of increased levels of psychiatric complications (Kimberling and Möller, 1995). There are reports of depression, psychosis and suicidal tendencies as elements of an existential crisis for people losing vision and hearing (Miner, 1995, 1996, 1997; Vernon and Hammer, 1996; Hess-Rover et al., 1999). Vernon and Duncan (1990), moreover, maintain that elderly people with deafblindness can develop additional impairments, such as depression and dementia.

**Emotional consequences**

Deafblindness is a highly emotional topic. This is evident from the literature as well as from personal experiences. People with deafblindness, their relatives and professionals all express strong emotions such as grief, guilt, shame, anger and frustration – more or less observably so in the accounts given previously in this chapter.

People with Usher syndrome express emotions relating to loss of independence when their vision deteriorates in a number of ways (Hicks, 1978; Miner, 1995, 1997, 1999). The inadequate behaviour of people with Usher syndrome, such as refusal to use a white stick, guide dog or guide – in spite of obvious needs and these things having been prescribed for them – could be related to denial. Acting-out and promiscuity could, in fact, be about a refusal to deal with grief and pain (Fillman, Leguire and Sheridan, 1989; Vernon and Duncan, 1990). Hammer (1978) argued that people with Usher syndrome who do not demonstrate stress are possibly out of touch with reality. He maintains that attention should be paid to this group. Rönberg, Samuelsson and Borg (2002) found emotional problems in a group of adults with deafblindness. A positive correlation was found between the inability to discover and localize people and events in the surroundings on the one hand, and pessimistic thoughts on the other.

Frequent feelings of loss and grief are common to people with Usher syndrome, as a result of successive changes in seeing and hearing functions (Miner, 1995, 1997, 1999). In addition, practical consequences, such as reduction of independence, loss of driver’s licence or the introduction of a white stick, are emotionally loaded (Brennan, 1997; Miner, 1995, 1997). The ongoing loss brings up past losses that need to be re-examined and reworked, so there is a need for recurrent psychological support and therapy (Miner, 1999). Miner, as well as Vernon and Hammer (1996), points out the need for
therapy in the language and mode suitable to the person with deafblindness. Miner, in particular, stresses the importance of therapists having skills and personal strength to handle their own anxiety. This can be crucial in the case of therapists who themselves have deafness, as they cherish their own vision and fear losing it.

Parents of children with prelingual onset of deafblindness can have strong emotions, such as frustration, due to feeble and hard-to-interpret responses from the children during interaction (Chen and Haney, 1995). Grief and feelings of guilt among parents of children with Usher syndrome are also reported (Torrie, 1978; Fillman, Leguire and Sheridan, 1989; Miner, 1995, 1997). Families of a person with Usher syndrome can need support in talking to each other about their emotions relating to the syndrome (Miner, 1999). The previously mentioned reports of elevated staff turnover furthermore indicate that the care of people with deafblindness is stressful for health-care professionals too (Goode, 1994; Giangreco et al., 1999).

Other social effects

This section deals with issues not included in the previous accounts linked to the ICF that are particularly relevant when deafblindness is considered: chains of consequences, impact on psychology, importance of knowledge about deafblindness, public health matters, emotional consequences, genetic issues and alternative strategies.

Chains of consequences

Characteristically, the consequences of deafblindness are linked in long chains. For example, combined vision and hearing impairment affects communication, which in turn affects relationships with other people, which then causes social isolation. Each link in the chain contributes to further consequences at the same time as it is connected to and dependent upon earlier links. Taken together, chains of consequences generate an impact on life that reaches far beyond any sum of single consequences. A number of references provide examples of outcomes generated by previous consequences.

Children with prelingual onset of deafblindness run the risk of acquiring persistent learned helplessness. According to Marks (1998), learned helplessness emerges when a person cannot control their surroundings or the results of their actions. The risk is that teachers and caregivers foster a dependency by expecting too little from a child.

Miner (1995) described how an elderly woman with Usher syndrome type 1 fell down in the street when she was out walking alone. Hitherto the
woman had usually been escorted by a relative and, by the time of the event described, she had not yet learnt to use a white stick. Several people passing by rushed to help her, but her vision was not sufficient to read their lips and they could not use sign language.

Further examples of chains of consequences are presented in studies of people with postlingual onset of deafblindness (Fillman, Leguere and Sheridan, 1989; Vernon and Duncan, 1990; Miner, 1995, 1997; Cioffi, 1996; Brennan, 1997).

**Knowledge about deafblindness – other people’s skills**

In quite a few references, the knowledge and skills of parents, relatives and professionals are pointed out as being crucial for positive development among people with deafblindness. From the accounts given previously, it is obvious that specific knowledge is needed from the very initial interactions with children with prelingual onset of deafblindness. This is pointed out in a number of studies (Chen and Haney, 1995; Nafstad and Rødbroe, 1999; Bruce, 2002, 2003).

However, there is no research regarding, for instance, the early impact of seeing functions on playing games and education. Furthermore, studies of the scope of knowledge about these issues among educationalists, parents and others coming into contact with children with postlingual onset of deafblindness, for example children with Usher syndrome, are also lacking.

There is rarely specific accommodation for people with deafblindness, and those who need to live in some kind of sheltered accommodation frequently do so within programmes directed to people with other impairments. If personnel and other residents cannot communicate in a manner receivable to the person with deafblindness, he or she becomes more and more isolated (Vernon and Duncan, 1990).

**Public health issues and health threats**

There are specific health threats related to deafblindness. Besides the risk of traffic accidents or of being the victim of criminals (Cioffi, 1996), there are also threats related to lack of information on public health matters, such as food recommendations, warnings concerning use of tobacco and alcohol, warning notes on medicines and so on. No attention has been paid to deafblindness from a public health perspective in the reviewed literature.

Health threats related to labour are noticed in one study, which in addition points out that occupations traditionally regarded as dangerous are not necessarily dangerous to people with deafblindness. It is rather their occupational skills and adjustment to deafblindness that are decisive (Vernon and Duncan, 1990).
Genetics-related issues

Psychosocial aspects of genetically caused deafblindness are dealt with in a few studies, in all cases in relation to Usher syndrome.

Even though Usher syndrome is genetically caused, most children with Usher syndrome are born to seeing and hearing parents. Initially, parents are informed that their child is born with deafness; not until later do they learn that the child also has a serious and progressive visual impairment. This can cause a crisis for the parents, and relational problems associated with the child can emerge. Parents need to learn sign language and to meet other parents in similar situations, as well as children and adults with Usher syndrome, to gain experience (Miner, 1995). Miner’s observations show that rehabilitation in the area of deafblindness needs to encompass not only the person with impairments but also parents, siblings and other family members as well as personnel and officials, as specific knowledge and conduct are required.

Because Usher syndrome is genetically caused, parents as well as the person with Usher syndrome have feelings of guilt, according to Miner (1995, 1997). She also notes that family members cannot always talk to each other about these feelings. There are people who do not find out that they have Usher syndrome until they are adults (at 25, 35 or 40 years of age), although their parents have known this for years. Miner maintains that children with genetically caused syndromes should be informed as soon as the diagnosis is decided, claiming support for this opinion from several studies. Explanations should furthermore be provided that are suited to the child’s cognitive and mental level of development, and questions should be answered in an honest way (Tedder, 1987; Miner, 1995).

Not everyone with Usher syndrome tells their children and grandchildren that they have a genetically hereditary disease and that descendants could be carriers of the defective gene (Miner, 1995). Some people who know that they have Usher syndrome marry and have children early on, whereas others choose not to have children. Harrod (1978) maintained that families that receive genetic information about Usher syndrome must be encouraged to assimilate the facts. Considering such information, they are, according to Harrod, able to judge alternatives and make their own informed choices regarding children. Harrod maintains that no decision is incorrect as long as it is based on information. In previous contacts with people with postlingual onset of deafblindness, I have learned that some people have experienced abusive treatment by physicians, forbidding them to have children because of their heredity.
Alternative strategies and skills

In order to compensate for deafblindness, it may be necessary to develop alternative strategies, for instance various methods of making use of smell, taste, sensitivity of the skin, memory or ability to draw conclusions. Such consequences of deafblindness have not been considered in the literature. However, Goode (1994, p. 17) has taken an interest in alternative strategies and provides some interesting observations. Moreover, he maintains that:

Establishing an understanding (intersubjectivity) with the children in their 'own terms' would be significant not only for our understanding of them but for our efforts at teaching and socializing.

Goode (1974) described a situation during the previously mentioned participant observation. He was going to feed one of the children with deafblindness but did not succeed in holding the spoon in the right position. The child then took his hand with her hand and brought it to the right position. In other words the child, who was considered uneducable, was able to correct him. Goode maintained that she understood routines and could teach them as well. The same child used her mouth to explore her surroundings and Goode gave an account of how she wetted things she wanted to know more about with her saliva. He also described something he calls *esoteric communication*, in which knowledge is transmitted from the person with deafblindness although the receiver cannot explain how this was done. There is also a feeling that the child has known about events before they occur. Such communication cannot be explained scientifically.

People with decreasing peripheral vision can compensate by turning their heads more often without being themselves aware that they do something different from other people (Vernon and Duncan, 1990).

Discussion

The present overview demonstrates that, on the whole, our knowledge of the psychosocial consequences of deafblindness is fragmentary. Basically, questions about the way people with deafblindness live their lives – in particular adults and elderly people – remain unanswered. By and large the reviewed literature is prescriptive rather than descriptive, and does not search for causes. Also, it chiefly relates to children with prelingual onset of deafblindness. Services offered to people with deafblindness follow the same pattern, according to Pollard, Miner and Gioffi (2000).

Few studies are based upon empirical information gathered for scientific purposes. This overview is based exclusively upon references found in scien-
tific databases. In addition to such references, there is a number of more or less scientific reports emanating from conferences and projects.

However, the overview illustrates that the consequences of deafblindness are decisive to the way that life can be lived. Furthermore, the consequences of deafblindness affect not only the person with the impairment, but also their family and other people in their surroundings. Environmental factors play a decisive role when it comes to activities and participation, and sometimes even with regard to the impairment itself (for instance, darkness causing a temporary deterioration in the vision of people with Usher syndrome, or inadequate rehabilitation of those with prelingual onset of deafblindness causing stress and generating behavioural disorders). It has also been shown that specific measures by trained professionals are required to achieve optimal living conditions. However, one report after another reveals that – particularly in cases of prelingual onset of deafblindness – rehabilitation, education and services are insufficient and generate activity limitations and participation restrictions, adding further threats to health.

The psychosocial consequences of deafblindness with onset during old age is not considered in the literature reviewed here. Indeed, elderly people are included in a few studies but attention has not been paid to issues related to old age (Wolf-Schein, 1989; Vernon and Duncan, 1990; Miner, 1995; Rönnberg and Borg, 2001; Liberman and Stuart, 2002; Rönnberg, Samuelsson and Borg, 2002). However, there are a number of reports originating from conferences and projects, specifically dealing with the situation of older people with deafblindness in Norway (Statens sentralteam, 1999), the Netherlands (Sooth, 1996) and the UK (Greaves, 1999).

There is an urgent need for research into the social consequences of deafblindness, particularly various conditions of children and young people with postlingual onset of deafblindness and consequences of deafblindness for adults and elderly people. The few references available concerning adults, such as the studies reported by Petroff (2001) and Miner (1995, 1997), with few exceptions, lack accounts of the relationship between the degree of visual and hearing impairments on the one hand and the psychosocial conditions on the other.

At present, a longitudinal project (5 years) is in progress in the Nordic countries, dealing with the social consequences where alterations in communication are required as a result of deteriorating sight and hearing. This project – which has not generated any scientific papers so far – is an example of much-needed research. Furthermore, research into the emergence and impact of disability movements in the case of people with deafblindness is non-existent.

Another unexplored area is the use of alternative strategies. Instead of focusing on problems associated with deafblindness, we could look upon
people with deafblindness as survivors with unique talents. Associations created by people with deafblindness and emerging deafblind communities are other fields that could be investigated in the future. In our capacity as human beings with vision as well as hearing, we can learn from people with deafblindness if we make the effort to perceive the world as it presents itself to them. A quote from Goode, relating to Christina – a child with prelingual onset of deafblindness in his study – illustrates the importance of a universalistic view when researching about and with people with deafblindness. Goode points out sources of joy and belonging in spite of barriers caused by lack of a common spoken language:

If we can accept and understand the malleability and heterogeneity involved in the expressions of these basic projects in the life world, then we can stop one of our central self-deceptions and move away from a view of humankind that raises us above other creatures or that affirms one ‘kind’ of human as better than another. Chris and I may have had our ‘differences’, but these were differences of degree, not of quality. This is probably the most important ethical lesson I learnt from Christina (Goode, 1994, p. 42).

References


The impact of combined vision and hearing impairment and deafblindness


Deafblindness: a challenge for assessment—
is the ICF a useful tool?

Abstract
The International Classification of Functioning, Disability and Health (ICF) is considered to be a big step forward in relation to the possibilities for describing disabilities. The aim of this paper is to highlight some strengths and limitations in the ICF that have been found during a study of people with deafblindness. Thirty-two adults aged 19–92 years, totally deaf and blind, as well as those with some remaining vision and hearing, were included. Questionnaires and personal interviews were used. The participants had difficulties in activities and participation on almost all investigated items. The following five circumstances could not be taken account of: fast variation in functioning due to different personal and environmental factors; choosing not to do an activity because of a health condition; time loss impacting on quality of life; health risks related to particular impairments; and obligations. The ICF needs to address these questions and needs to be further developed.

Introduction
In May 2001, the assembly of the World Health Organization (WHO) endorsed the International Classification of Functioning, Disability and Health (ICF) (World Health Organization, 2001), which is a second edition of the International Classification of Impairments, Disabilities and Handicaps (ICIDH), which itself dates from 1980 (World Health Organization, 1980). This represents a big step forward in the description of disabilities. The world has also got a common language on this question, which can be used between different cultures, between professionals, and also by people who themselves have problems related to health. How the ICF is constructed will not be described in this article. It has been done by others, including those in the field of audiology (e.g. Stephens & Kerr, 2000). The aim of this article is to highlight some examples of the strengths and limitations of the ICF that have been found when studying people with deafblindness.

In a study of living conditions among people with deafblindness, some other disadvantages, apart from limitations in activities or restrictions in participations, have been found. The concept of consequence has been found to be fruitful in understanding and describing what it means in daily live to have deafblindness. The first version of the ICF, which was edited by the WHO (World Health Organization, 1980), started with a discussion about what the healthcare system can do and what it should do. There was a need to describe the consequences of disease. The answer to that need was the ICIDH. The purpose of the ICIDH was to promote more sensitive and comprehensive assessment of individual problems and to facilitate a more critical evaluation. The ICIDH was criticized by many people and for many different reasons (e.g. Oliver, 1986, 1990; Söder, 1988; Nordenfelt, 1993). The concept of handicap has been taken out of the ICF, since in English it has an alternative meaning in the field of sport. The concepts of consequence and disadvantage have also disappeared from the ICF, but without any explanation.

In the debate about the ICIDH, there has been very little consideration of this. In this article, the concept consequence will be brought back into the analysis.

Materials and methods
The consequences of deafblindness described in this article stem from research on living conditions among adults with deafblindness in the county of Örebro in Sweden during 1997–99 (Möller & Samuelsson, 1998; Möller, 1999).

In the Nordic countries, there has, over the last 20 years, been an agreement on the notation of deafblindness. The Nordic definition is based on serious lack of functioning, and not on assessment by a doctor (Nordiska arbetsgruppen för dövblinda handikappfrågor, 1980). This means that not only people who are completely deaf and blind but also those with serious vision and hearing impairment are included. Here, the concept deafblind will be used in accordance with the Nordic notation.

The population in the study consisted of 27 women and five men between the ages of 19 and 92 years. Six were deaf and blind, and the rest were in the process of losing vision and/or hearing. One person was born deaf and blind, a condition that was formally defined as congenital deafblindness, and the others had acquired the condition. Eight people had a serious sight and hearing impairment that was related to their age. The largest group (approximately 12 people) had Usher’s syndrome.

Nineteen subjects had sign language as their first language, while 13 used spoken language. Six of those using sign language used the language in the tactile form. About half of them had an education level beyond the Swedish general 9-year schooling.

The conceptual framework used to describe the everyday life of the participants was ‘To have, to be, to love’ (Allardt, 1977) and the dimensions activity and participation (ICIDH-2 beta 1) (World Health Organization, 1997). All information about the participants in the study was obtained from self-reports (questionnaires and personal interviews). The parents, together with the

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staff, answered on behalf of the person who was congenitally deafblind, while all of the others reported themselves. Fifty-three persons were invited to participate; 14 did not answer the invitation, and six, mainly elderly (older than 80 years), declined to participate. Örebro Municipal Council was responsible for the project. Örebro County Council and the National Institute for Disabilities in School also took part. The study was carried out in cooperation with the local branch of the deafblind association (FSDB). Persons who themselves had deafblindness took an active part in the design and carrying out of the study.

Results

The participants with deafblindness had difficulties in almost all forms of activities and participation. The classification of difficulties was based on what the participants recounted during the interviews and on my own observations. They had moderate- to-severe difficulties in performing about 66% of the activities. Regarding participation, they had 'participation with restrictions' on about 80% of the items. The sources of the limitations or the restrictions could sometimes be related to the impairment itself, e.g. driving a car. Sometimes the limitations were caused by personal factors, e.g. age. Mostly, limitations were caused by different kinds of factors in the environment. Despite these clear results, there were consequences that could not be grasped by the ICDH-2. These problems still remain in the ICF. In the following sections, the focus will be on five consequences that were identified.

Variations in functioning

First, functioning can vary. In the ICF, impairments caused by problems in body function or structure are declared as significant deviations or losses. There is a possibility in the ICF to measure change in function by registering function on at least at two occasions. This can be enough for impairments that have long improvement or deterioration times. For some of the participants with deafblindness, changes in function took place extremely rapidly, e.g. while walking from one room to another. Some of them had vision problems caused by retinitis pigmentosa (RP), which, among other things, leads to night blindness. If the person was in a room with normal daylight and went from that room to a fairly dark room, their vision could decrease from severe impairment to almost complete blindness. A consequence of this is that it very often became a new functional condition. As the example shows, most of the participants had a span of functioning. The functioning could vary due to factors intrinsic to the person (e.g. tiredness and general condition) and/or to conditions in the environment (e.g. light and noise). To understand the consequences of deafblindness, it was important to register these variations and the circumstances that caused them.

The variation in functioning also affected activities and participation. At best, a person could perhaps do an activity, and at worse, they could not do it at all, e.g. be able to follow a particular lesson at school. In the ICF, it is possible to register effects from the environment, but not these kinds of very fast changes in functioning. The problems arise when a researcher or a clinician registers the level of functioning in the ICF. Should they register the condition under favourable conditions (e.g. in the light room) where the person has better functioning, or in less favourable conditions (e.g. in the dark room), when the functioning could be much lower? Or should they register 'the average'?

'Don'ts'

Second, it is not easy to register in a situation where the person chooses not to do an activity. Measures of activities and participation register what people do. However, what people do not do can also be of great importance for their wellbeing. For instance, a person who would like to perform a certain activity has to relinquish doing it because it is too humiliating, and so forth. The participants in the study often reported 'don'ts'. The occasion could be a gathering with relatives, attending union meetings, or even attending a funeral. An example from the study was an elderly woman with some vision and hearing remaining, who previously used to play cards now and then with her friends. This was still possible, and she was welcomed by her friends, but she chose not to attend, because as she said, 'I can't hear them and it is not fun anymore'. The ICF does not catch restrictions caused by the use of a person's 'free will'.

Time loss

Third, losing time is another important factor in the living conditions of the participants. Loss of time was a very obvious experience during the research. I observed the huge loss of time, and, in addition, it was reported to me during the interviews by participants with deafblindness, by relatives, and by professionals. Those who had deafblindness often reported that they could not do more than one thing at a time; for example, they were not able to cook and talk with their children at the same time. People with deafblindness also spent a lot of time in contact with the health and welfare system. They had many different contacts, and each contact generated several phone calls, including waiting time in each, e.g. to book the interpreter or to book the transportation. The professionals who I interviewed reported that they booked twice as much time for an appointment with a person with deafblindness than for other clients. In the ICF, it is possible to record limitations in activities caused by taking a longer time to do the activity than normal. For people who have deafblindness, most activities took a very long time, and because of limitations in the welfare system, they also lost time in waiting for service. Losing time had a severe impact on their quality of life. However, in the ICF it is not possible to add all this time loss to make a 'losing time consequence'.

Health risks

Fourth, there were shortcomings because of health risks due directly to deafblindness. In the ICF, there is no provision to record health risks that occur as a consequence of impairment. Sometimes the participants had other diseases which, in combination with the deafblindness, could constitute a risk. For instance, there was one completely deaf and blind participant who also has asthma and was allergic to animals—she could not see or hear if a dog was close to her.

Some individuals had, as mentioned above, some residual vision or hearing. However, it was very hard for them to see or hear, and it cost them a lot of energy. Those individuals often reported tiredness and headache. Both tiredness and headache can be recorded in the ICF, but in these cases the tiredness and the headache were caused by the effort of seeing and hearing. Some of the health risks directly resulted from inadequate knowledge of the consequences of deafblindness. Frequently, the participants did not have grounded knowledge, could not read warning signs, or were not informed about risks.
Duties and obligations
The fifth shortcoming of the ICF has to do with obligations. The standard rules relating to equality of opportunity for persons with disabilities (United Nations, 1994) mention not only rights, but obligations as well. It became very obvious during my study of deafblindness that people around a person with deafblindness, and the professionals they met, seldom talked about obligations. I think that people should have as much opportunity as possible to take responsibility in their own life and for the whole society. I think that if we expect a person to fulfil obligations, this leads us to look at them as equal human beings. This raises the issue of such taking of responsibility by people with deafblindness or with some other disability. Do we expect them to contribute? Or do we think that they only have needs? In the future, I hope that duties and obligations will be included in the ICF.

Discussion and conclusion
The revision of the ICIDH was an ongoing process for many years, involving practitioners and researchers from all over the world. The WHO has done a good job in increasing the range of possibilities for describing disability. With the ICF, the world has got a common language. One important point is that the environment is now included in the framework. The ICF is intended to be a bio-psychosocial model and tries to achieve a synthesis of different perspectives, which is also important, since impairment and disability concern both body and society. If there are limitations in the body function or structure, there are usually implications for the broader society. The activities and participation components in the ICF may also contribute to the political struggle for equality of opportunity for persons with disabilities, since the ICF highlights two important ingredients for realizing human rights. Another important aspect of the ICF is universalism. The ICF is a model not only for people with disabilities, but also for all people. Zola (1989) and Bickenbach, Chatterji, Badley & Ustün (1999) argue that universalism as a theory serves persons with disability more effectively than a civil rights or ‘minority group’ approach. I hope that the ICF will be used. The remarks I have made in this paper do not imply that we should stop using the ICF—on the contrary, only by using it can we push the process forward.

For persons with deafblindness, it is possible to use the ICF to show the great impact of deafblindness on their living conditions, as it affects almost all their activities and participation. Despite this, there are some important shortcomings in the ICF. It is difficult to use when: first, a function varies rapidly due to different personal and environmental factors; second, a person chooses not to do an activity due to a health risk; third, the loss of time can have an important impact on the quality of life; fourth, there are health risks related to particular impairments; and fifth, obligations are also important in the drive for equality.

In its present state, the ICF does not serve as a sufficiently precise tool to describe functioning and the consequences of reductions in functions. This will in turn lead to great uncertainties regarding measures to improve the wellbeing of the person. The taxonomic structure of the framework is not developed enough to describe a highly complex social process. One reason for this could be that the taxonomy seems to have been developed with a less complex level of reality (e.g. physiology, biology) in mind, and not the more complex social level. The phenomenon that the new framework is trying to describe has to be seen as a process. The framework hardly catches this. Furthermore, the health condition (impairment and/or disability) can lead to secondary conditions. In this article, secondary conditions are shown by the ‘don’t’, ‘health risks’ and ‘time loss’. It is hard to be precise about the circumstances under which secondary conditions are created, as the process cannot be described in the ICF. There is a risk that health insurance companies, welfare providers and others will only look only at what can be described through the ICF and draw wrong conclusions about the consequences caused by health conditions. This in turn could lead to disadvantages for the persons who have disabilities. There is need for research into the use of the ICF for different purposes and in different fields. I hope that a future version of the ICF will come closer to the stated goals: forming a scientific basis for understanding and studying health and health-related states, establishing a common language for describing health, permitting comparisons of data, and providing a coding scheme.

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Social Recognition, Participation, and the Dynamic Between the Environment and Personal Factors of Students With Deafblindness

The study describes environmental and personal factors that, from the student perspective, impede participation in education in secondary upper schools by students with postlingual deafblindness. The discussion is framed by the International Classification of Functioning, Disability, and Health. The researchers use the theory of social recognition as a theoretical tool in understanding the dynamics between personal factors and environment in the context of secondary upper-school education. Thirty-four students with deafblindness responded to a questionnaire; the survey’s findings indicate experiences of barriers in the natural and social environments that restrict participation. Experience of considerateness—such as concern for the special requirements of students with deafblindness—and experience of the lack of considerateness are the most important factors. Negative roles adapted by some students for themselves may be interpreted as resulting from a lack of recognition, in the form of denigration or insults.

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Changes affecting the cohort of children and youth with deafness or hearing impairment (e.g., the increased frequency of neonatal screening, cochlear implantation, and schooling in general education settings) will affect these students’ learning. Luckner, Muir, Howell, Sebald, and Young (2005) emphasize that the main priority of new research and training in the field of deaf education is to teach administrators about services appropriate for students with hearing loss. A way to approach this priority is to start with a discussion of participation. Over the last decade, the concept of participation has increasingly become the focus in discussions about people with disabilities. One of the main reasons for this is the highly advanced position held by participation in the International Classification of Functioning, Disability, and Health, or ICF (World Health Organization, 2001).

There are other, yet similar, ways to approach participation. For instance, Eriksson and Granlund (2004) define participation as consisting of three dimensions: perceptions of participation, activity/behavior, and prerequisites for participation. Prerequisites are viewed as factors in the environment. Furthermore, Eriksson and Granlund 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. Others have also highlighted the prerequisites offered by the environment in combination with the prerequisites of the individual (e.g., Oliver, 1996).

In a study of students with different types and degrees of disability, Eriks-son and Granlund (2004) concluded that the student’s conception of participation does not depend on the type of disability, with one exception: the deafblind student. They found that persons with deafblindness are significantly (p ≤ .05) distinguished from other groups with disabilities in three ways: Study participants with deafblindness more frequently mention self-determination and conditions in the living environment, as well as prerequisites for taking part in activities, as a part of their conception of participation than do other groups. In another study (Almqvist & Granlund, 2005), students with disabilities, their teachers, their parents, and special education consultants (N = 472) responded to questionnaires. Almqvist and Granlund highlighted issues of interaction, autonomy and locus of control, availability in the environment, and participation in activities in school. They found that the conception of participation was based on the total number of positive factors, rather than a single special factor. Further research is needed to understand the relationship between types and degrees of disability, on the one hand, and participation, on the other. However, the fact that students with deafblindness are singled out as the only disability group that shows such a correlation is of great interest.

In Sweden, the goal of the School Act is “a school for all.” How this can be implemented is a matter of power and attitudes and is part of a struggle to create a strong democratic future for students and communities, as Skrtic (2005) puts it. He discusses the necessary shift from blaming school failure on students, at one end of the spectrum, through medicalizing and objectifying discourses, to a fundamentally different kind of organization, that is, a nonbureaucratic problem-solving organization. The problem with focusing on students rather than on attitudes and structures in the education system has been the focus of several studies.

In his review, Haug (2004) draws the conclusion that there is little research on integration and inclusion in special education, especially in the Nordic countries. He also states that there is little research into the experiences of integration and inclusion. Haug refers to a study by Grue (2001) in which the informants had physical disabilities. According to the students in Grue’s study, many of them suffered experiences indicating that their school failed to understand their disability and thus had neither routines nor rules for including them. Some students said that instead of focusing on their requirements—that is, on physical adaptations—their school behaved as if they had learning disabilities.

**Definition of Deafblindness**

Deafblindness is an unclear concept, most frequently referring not to a total absence of hearing and vision but to some reduction in the body function of hearing and vision. The Nordic countries have since 1980 defined deafblindness as a distinct disability, stating that “a person is deafblind when (s)he has a severe degree of combined visual and auditory impairment. Some deafblind people are totally deaf and blind, while others have residual hearing and residual vision” (Bättre livs villkor för dövblindna i Nor- den, 1980, pp. 78–79). However, this definition has recently been amended to incorporate the basic ideas and vocabulary of the ICF. For the purpose of defining the population and sample in the present study, we have adopted the Nordic definition from 1980.

**Research on Students With Deafblindness: A Literature Review**

Students with deafblindness are a vulnerable group. Research reveals a history often showing an exposure that violates their right to an adequate education (for a comprehensive overview, see Möller, 2005). Most research focuses on deafblindness with prelingual onset. Knowledge about students with postlingual deafblindness (the most common diagnosis in this group is Usher syndrome) is fragmentary and often anecdotal. One reason for this is that the group is quite small.

Kirchner and Diament (1999a, 1999b) demonstrated that students with deafblindness run the risk of failing to receive the support they need because they are not considered as students with deafblindness. This finding indicates the role of an adequate level of knowledge of deafblindness among persons in the student’s immediate educational environment. It seems also to be the case that, for persons with a reduction in the function of two senses, the sense first diagnosed or that is most seriously limited draws attention—and not that the combination of hearing and vision reduction per se has implications for activity and participation. A consequence of this is that students with deafblindness do not always have access to the support they require—especially, for example, trained teachers—and this is irrespective of whether a student is placed in a standard or a special school.
Olesen and Jansbøl (2005) conducted a 5-year follow-up study of the living experiences of 20 adults (ages 17 to 63 years) who had Usher syndrome (8 had Type I and 12 had Type II) from four of the five Nordic countries. The authors found that several of their informants had experiences of both mainstream schools and schools for the deaf. A common theme was that the informants felt great relief when they went to a special school with young people similar to themselves. These individuals’ narratives of their experiences of education bear witness to their struggle for recognition. Those who had attended mainstream schools reported social isolation and a sometimes very demanding situation. There were often many types of problems. For instance, they lacked information and guidance about their disability and the existing compensatory measures. Furthermore, they lacked the opportunity to inform others about their situation, and they missed counseling and support.

Regarding the transition from special to mainstream schools, Ingraham, Daugherty, and Gorrafa (1995) reported in a case study of three gifted students (ages 12 to 13 years) that there was a great need for investigation, support, service, coordination, and counseling. The students were transferred from a special school (a boarding school) to a mainstream school. Support services and financing were coordinated among several agencies, at both the federal and local levels. Ingraham and colleagues noted, for instance, that teachers were required to prepare lessons and assignments several days in advance in order to enable information to be transcribed in an accessible format. The support included vocationally related efforts aimed at, among other things, allowing the students to keep possession of technical aids when they moved from one educational level to another or between schools. Although the transition was an educational success, from a social viewpoint it had its drawbacks. As in the study by Olesen and Jansbøl (2005), the students reported isolation and the feeling of being outsiders.

In light of these findings, it appears that special arrangements are needed to promote social inclusion. In their case study of four students with deafblindness in inclusive classrooms, Goetz and O’Farrell (1999) recommended an adapted multimedia communications system to facilitate social interactions between and among classmates, teachers, and staff. This must be based on the continuous input of information about the student’s needs and abilities. Goetz and O’Farrell also reported on classmates standardly involved in commitments to activities that include and support the student with deafblindness.

Physical education is a subject of great importance. However, it can sometimes render a disability very obvious. Students with disabilities usually wish to pass as nonspecial students, but this is very difficult in physical education. Lieberman and Houston-Wilson (1999) highlight major barriers to inclusion in physical education. Their study focused primarily on students with visual impairments, yet the results are applicable to students with deafblindness. Data were collected from surveys completed by physical education teachers (N = 170). Lieberman and Houston-Wilson found that teachers’ barriers included a lack of professional preparation; curricula and activities provided in general physical education classes; the pace of the lessons; fear; overprotectiveness; and limited expectations. Students’ barriers included parental overprotectiveness, a lack of opportunities, and a lack of confidence. Administrative barriers included inadequate time, a lack of appropriate equipment, and “blanket” medical excuses.

Some students with deafblindness face additional impairments or other obstacles, such as related life crises and depression, particularly when vision or hearing deteriorates (Miner, 1999).

To sum up, knowledge about students with deafblindness is limited. Eriksson and Granlund (2004) have shown that students with deafblindness value prerequisites for education more highly than any other group of students with disabilities. Eriksson and Granlund also found that self-determination and the living environment were aspects specifically noted by students with deafblindness. Research also indicates that in order to obtain greater participation, a number of conditions must be fulfilled, such as the possibility of having access to oral and written information and mobility, conditions that for most other students are a matter of course. Many studies have dealt with contextual factors, and the results indicate that several factors exist that are crucial to participation in education by students with deafblindness. Subsequently, the possibility of influence on participation by students with deafblindness themselves will be increased.

Method

Research Questions

The aim of the present study is to describe factors that pose barriers to participation in education in secondary upper schools by students with deafblindness, from the perspective of the student themselves. We posed three research questions:

- How do environmental factors influence participation?
- What is the role of personal factors?
To what extent are students with deafblindness subject to social recognition or misrecognition (or both)?

**The ICF: the Conceptual Framework**

The present study's conceptual framework is the International Classification of Functioning, Disability, and Health, or ICF (World Health Organization, 2001). Two major parts, *functioning* and *contextual factors*, constitute the model. These are, in turn, divided into a total of six components with a number of subcomponents. Such subcomponents can be described in both positive (neutral) and negative ways. The four central components in Part 1, *functioning*, are *body structures*, *body functions*, *activities*, and *participation*. The terms *deviation* and *impairment* cover the negative aspects of body structure and body function. *Limitation* and *restriction* are the terms for negative aspects of activity and participation. Part 2 of the ICF, *contextual factors*, consists of the two components *environmental factors* (external) and *personal factors* (internal). The environmental factor is assumed to play both a *facilitating* role and an *impeding* role (i.e., it can be a barrier). The umbrella terms *functioning* and *disability* are viewed as outcomes of interaction between health conditions (diseases, disorders, and injuries) and contextual factors. The relationships among the components of the ICF are illustrated diagrammatically in Figure 1.

Body structures are anatomical parts of the body, and body functions are physiological and psychological functions of body systems. Activity is the execution of a task or action by an individual. Participation in the ICF is a person’s involvement in a life situation and represents the social perspective of functioning. Participation includes involvement in a life situation, and the latter concept has several dimensions: *taking part*, *being included*, *being accepted*, *being engaged in an area of life*, and *having access to needed resources*. Information that reflects the person’s subjective experience of involvement is not incorporated into the component of participation. Furthermore, the environment is, in the ICF, divided into six areas:

1. Products and technology, which may be usual (e.g., buses and buildings) or specialized (e.g., hearing aids and books printed in Braille).
2. The natural environment and human-created environmental changes (e.g., light or sound).
3. Support and relationships (e.g., a counselor for the deafblind individual and his or her friends).
4. Attitudes, which include values, beliefs, ideology, and habits (e.g., considerateness).
5. Service, systems, and policies (e.g., pedagogy and interpreting).
6. Personal factors, which relate to the individual, and include age, gender, life experiences, and coping strategies. This is the only area with no negative term, domain, or category. The subjective experiences of participation and emotions are, in the present study, regarded as personal factors.

The relationship among the six components is interactive; hence, the influence in terms of causality is a complex one for which the ICF gives no guidelines for description. The role of the ICF is to provide a conceptual framework rather than with a theory. In the present study, we focus on participation in school education. In the ICF (World Health Organization, 2001), this implies gaining admission to school, engaging in all school-related responsibilities and privileges, and learning the course material, subjects, and other curriculum requirements in a primary or secondary education program, including attending school regularly, working cooperatively with other students, taking direction from...
Social Recognition

Among other factors, participation includes a very important subjective dimension. It has to do with a sense of being included and being accepted. Axel Honneth has developed this dimension in his discussion of recognition (see, e.g., Honneth, 1995). Recognition also includes an environmental aspect—how people in your environment treat you. In that way, the concept works as a “bridge” between the inner dimension of participation and the outer dimension, the human environmental aspect. Honneth has developed a model of recognition characterized by a categorical imperative of recognition involved in all human interaction: If I want to be recognized, I have to recognize the other. Lack of recognition by others distorts the development of a sense of self (Honneth, 1995, chapters 5, 6). Although he does not address the situation of people with disabilities, an account of Honneth’s approach in the context of disability studies is found in Abberley (2002).

The important observation that one can achieve positive relations to self only by being recognized by others highlights the question of social relations. Honneth (1995) makes operational relations of recognition and the process of recognition in three different categories.

The first category centers on the individual and his or her specific needs and emotions. Recognition evolves in primary relationships between the individual and his or her career, lovers, family members, friends, etc., and is crucial to the individual’s basic self-confidence as an autonomous being. Lack of recognition affects an individual’s physical integrity, exhibited in its extreme forms as abuse and rape.

The second process of recognition focuses on an individual’s position as a legal subject in law. Recognition evolves in legal relations and is expressed as rights—in the present period of history, often described in terms of civil, political, and social rights—crucial to acquiring a sense of possessing universal dignity as a human being and thereby to acquiring self-respect. Lack of recognition is accordingly expressed in terms such as the denial of rights and exclusion.

The third process of recognition centers on the individual and her or his individuality, traits, and abilities. Recognition evolves in communities of shared values that in the present historical period of individualization must be characterized by solidarity—a mutually felt concern for what is individual and unique about another person and his or her way of life—and is crucial to a sense of self-esteem. Lack of recognition is expressed by means such as denigration and insult. As human beings, all individuals need to be recognized through each of the three processes and modes of relations of recognition and thus to establish positive relations with themselves in terms of self-confidence, self-respect, and self-esteem. When a lack of recognition is experienced, harm is done; important to Honneth (1995) is the impact of such negative experiences in triggering collective actions and struggles for recognition. For the purposes of the present discussion, this brief sketch will serve as a reminder of the importance on a personal level of experiences such as harm, disrespect, and lack of recognition.

Honneth’s Three Relations of Recognition in the Case of People With Disabilities

To numerous people with deafblindness, the first category of recognition is to a greater or lesser extent permanently present in relations with assistants and caregivers: It involves physical integrity and fragile balances between autonomy and dependency.

For self-confidence to evolve and be maintained in such relations is very demanding, as many accounts testify. The second category is also highly relevant in the case of people with deafblindness. The history of this kind of recognition process through law illuminates the varied conditions under which people with deafblindness are to develop self-respect. The third category of recognition is familiar in studies focusing on stigmatization in Erving Goffman’s sense.

To Goffman (1968), stigmatizing is precisely the process in which the negative valuation of a characteristic within a “local moral order” causes people with this characteristic to, for instance, develop strategies to protect their sense of self—their self-esteem. It is certainly no easy task to obtain the solidarity required for the elimination of the lack of recognition, such as the stigmatizing of people with deafblindness; it is necessary to have much greater, proven knowledge about the processes of recognition regarding people with deafblindness.

In short, the ICF will provide a conceptual repertoire of concepts, and thus an opportunity to organize a description of a situation and to communicate the findings. The theory of social recognition is fundamental to a deeper understanding of the described situation for students with deafblindness. According to Honneth (1995), recognition has three types of relations connected to three different
processes. The first type of relation is accumulated in the primary relationships, that is, in the family. Detailed examination of this type of relation lies outside the scope of the present article; however, regarding the two other types of relation—that is, being included in an educational setting and being a member of a shared community—certain interesting and important features of such recognition, as well as lack of recognition, have been revealed.

**Questionnaire, Inclusion Criteria, and Sample**

For the present study, we used data from an investigation by the first-named author about secondary education for students with deafblindness (Möller, 2001). A questionnaire was developed in cooperation with the Association of the Swedish Deafblind (FSDB). Although one of the authors has insight into the topic, it was considered important to involve the FSDB for two main reasons: Doing so increased the legitimacy of the study, and the comments made by representatives of the target group contributed to a better-developed questionnaire. The question of legitimacy is extremely important with respect to the target group, for which the issue of confidence is very important.

The questionnaire was constructed to cover a number of components relevant to the target group in functioning and contextual factors. Issues of body structures and body functions were not included under the heading of functioning, which instead concentrated on participation and environment. Contextual factors were personal factors such as sex, age, education, and environment factors such as sound, light, assistive technology, experience of attitudes, and treatment. There were also questions about whether persons in the deafblind students’ immediate environment knew about their deafblindness, and how open the deafblind students were to informing these persons about their disability. The questionnaire consisted of 55 questions.

The respondents all had a significant combination of visual and hearing impairment. They had all received support and special services related to their disability and or were members of Swedish Deafblind Youth, a national organization of young people with deafblindness that is affiliated with the FSDB.

The inclusion criteria were postlingual deafblindness, a year of birth between 1969 and 1984, and a history of school attendance at the secondary upper level. These criteria were chosen in order to include people who had completed their education and had thus formed a perspective on their educational experiences.

Since no national register of the target population exists, the total possible number of informants remains unknown. Earlier reports have indicated a prevalence of 4.5 to 5.2/age cohort before adulthood (Dövblinda i Sverige, 1984; Mehlis & Tuftesson, 1989). The most frequent cause of deafblindness in Sweden, particularly in the younger age groups, is Usher syndrome. Sadeghi, Kimberling, Tranebjerg, and Möller (2004) estimate the prevalence of Usher syndrome in the investigated age cohorts (years of birth 1969–1984) to be 3.5/year, which suggests a total of 53 persons. Sadeghi and colleagues’ study indicates a lower incidence than do Dövblinda i Sverige and Mehlis and Tuftesson. We therefore estimate the incidence of disease or injuries leading to deafblindness to be 4 per year in the group relevant to the present study. This leads to an approximation of the number of students with deafblindness in Sweden of about 60.

We were able to trace 46 persons with deafblindness (77% of the estimated population), of whom 34 (57%) responded to the questionnaire, a group that hence constitutes a majority of the estimated total population.

In Sweden, the duration of standard compulsory schooling is 9 years; secondary upper school, which is voluntary, lasts 3 years. In the special schools the periods are 10 and 4 years, respectively.

The curriculum for secondary upper education, regardless of whether the school is standard or special, can be characterized as having either a focus on theoretical subjects, such as additional foreign languages, or a focus on vocational training, for example, barbering or housepainting. Both types of programs convey admission to higher education. Additional, there is a special type of curriculum, the individual program, for those who do not meet the national standard of the regular curriculum. The individual program does not convey admission to higher education.

For the present study, informed consent was obtained, and the respondents were promised that full integrity would be maintained and that the results would not be published in such a way that could reveal individual identities. Simple Swedish was used throughout the questionnaire, which was printed in large fonts and, as needed, in Braille. It was also possible for informants to contact one of the authors by text telephone for further information.

**Characteristics of the Respondents**

There was a roughly equal distribution between the genders among the survey respondents. Average age was 25.0
years (SD 4.6). There was also a roughly even distribution between students who were deaf and students who were hard of hearing; sign language was the most common mode of communication (see Table 1).

Regarding educational experiences, most of the respondents had attended special schools during compulsory schooling. At the secondary upper school level, most had attended a special school and taken a theoretical program of study. Responses were mixed concerning how long it had been since they had finished their secondary education: For some it was more than a decade ago, while for most it was relatively recently (see Table 2). This finding is addressed in the Discussion section.

Data Analysis

We used quantitative analysis for the present study. Most of the data are described in simple univariate tables, while in some cases a bivariate analysis is applied. The low number of respondents and the ordinal data level do not allow for any multivariate analysis. Since the aim was to make a study including the whole population, there were no sampling procedures. Hence, no test for significance was conducted. The results offered in the present article are, therefore, restricted to the survey respondents. However, since the respondents most probably constitute the majority of the total target population, they provide a basis for reliable description of the situation for students with deafblindness in Sweden.

Watching and listening are two activities mostly presupposed in the regular educational environment. However, students with deafblindness have limitations in both of these basic activities and hence require special devices, adaptation of the built environment, and treatment to assist their perception. In order to ascertain suitable adaptations at the individual level, Ingraham and colleagues (1995) compiled a checklist:

- What was the cause of the visual and hearing impairment (etiology/diagnosis)?
- At what age did the student’s impairment manifest itself?
- How severe is the student’s disability?
- What type of educational intervention did the student receive in the past?
- What type of support service is available? (p. 261)

Because the present study was conducted at the group level, it paid no

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>56</td>
</tr>
<tr>
<td>Male</td>
<td>44</td>
</tr>
<tr>
<td><strong>Self-reported disability</strong></td>
<td></td>
</tr>
<tr>
<td>Deafness and visual impairment</td>
<td>56</td>
</tr>
<tr>
<td>Hearing impairment and visual impairment</td>
<td>38</td>
</tr>
<tr>
<td>Blindness and hearing impairment</td>
<td>6</td>
</tr>
<tr>
<td><strong>Communication: language and modality</strong></td>
<td></td>
</tr>
<tr>
<td>Sign language, in visual modality</td>
<td>59</td>
</tr>
<tr>
<td>Spoken language</td>
<td>18</td>
</tr>
<tr>
<td>Mix of spoken language and signs</td>
<td>15</td>
</tr>
<tr>
<td>Sign language, in tactile modality</td>
<td>9</td>
</tr>
</tbody>
</table>

Notes. N = 34. Not all sets of percentages equal 100 because of rounding.

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Type of school</strong></td>
<td></td>
</tr>
<tr>
<td>Special school for students with deafness or hearing impairment</td>
<td>79</td>
</tr>
<tr>
<td>Standard school</td>
<td>21</td>
</tr>
<tr>
<td><strong>Type of program</strong></td>
<td></td>
</tr>
<tr>
<td>Theoretical</td>
<td>59</td>
</tr>
<tr>
<td>Vocational</td>
<td>21</td>
</tr>
<tr>
<td>Individual</td>
<td>6</td>
</tr>
<tr>
<td>Not known</td>
<td>15</td>
</tr>
<tr>
<td><strong>Number of years since respondent finished education</strong></td>
<td></td>
</tr>
<tr>
<td>Still attending</td>
<td>29</td>
</tr>
<tr>
<td>Finished 0–5 years ago</td>
<td>35</td>
</tr>
<tr>
<td>Finished 6–10 years ago</td>
<td>15</td>
</tr>
<tr>
<td>Finished 11–15 years ago</td>
<td>21</td>
</tr>
</tbody>
</table>

Notes. N = 34. Not all sets of percentages equal 100 because of rounding.
attention to the first question, and relatively little to the third.

Results

Contextual Factors

Environmental Factors

Indications of opportunities to participate in lessons and use educational materials (e.g., books) were reported by the respondents. They were asked whether there were hearing devices such as the FM loop, whether the classrooms were sufficiently well illuminated, and whether they had access to adapted schoolbooks. They were also asked if the teachers adapted their pedagogy—for example, slowed down their rate of speech or signing or turned the microphone around—which may also facilitate participation by students with deafblindness. In compiling the data, we defined a factor as facilitating if the students answered always or sometimes. Table 3 shows the proportion of students who responded that the factors were facilitating. Students who because of their respective levels of impairment took no part in one or more of the activities referred to in Table 3 did, however, respond to the question with not relevant or not necessary.

Teaching strategies—for example, facing the students while talking, adjusting the communication tempo, and supporting the students’ interaction during the lessons—were not the only prerequisites for participation. The possibilities both to travel to and from the school and to walk independently and safely within the area of the school are other indications of prerequisites for taking part. Students expressed their wishes for an inclusive design of the school and its surroundings in comments such as “I wish for better lighting in the dining hall and the corridors” and “There is a need for more and better marks and indications within the school.”

Personal Factors

The second type of contextual factors is personal factors. The students were asked about their experiences of other people’s considerateness. This reflects their perception of the behavior and attitudes among people in their immediate educational environment. When a person does not show considerateness to the special requirements of a student with deafblindness, it can be interpreted as disrespect, lack of recognition, and even not being worthy of thoughtfulness. Table 4 shows that the students perceived that the greatest considerateness was shown by teachers and classmates, and less was shown by others.

Another personal factor is not fulfilled the secondary education (applied to the so-called dropouts). Twenty-one percent of the respondents (six students) failed to complete their secondary education. Their dropping out was motivated by criticism of the teachers, being uncomfortable at school, and school fatigue. One of the students was offered a job, while another transferred to a different form of education.

In regard to satisfaction with the choice of school, 80% of the survey respondents were very satisfied or quite satisfied with their choice of school, a finding that should be compared with the initial self-determination of choice of school (see Table 5). Nearly 70% were very happy or quite happy with their time at their school.

Participation

Education includes participation in different activities in the school, of which two have been selected. The first is

| Table 3 |
| Respondents’ Opinions on Whether Environmental Factors Were Facilitating |
| Environmental factor | Facilitating (%) |
| Light in the classroom (N = 31) | 90 |
| See what the interpreter signed (N = 26) | 85 |
| Could lipread the teachers’ lips or see what they signed (N = 29) | 83 |
| Transport service, to and from school (N = 28) | 64 |
| Light and color in other parts of the school (N = 34) | 60 |
| Teacher/interpreter signed at a pace they could follow (N = 26) | 58 |
| Teacher showed which of the classmates answered a question, repeated what that student said or signed, or handed around the microphone (N = 32) | 47 |
| Telecoil (N = 13) | 46 |
| Adapted schoolbooks (N = 29) | 24 |

| Table 4 |
| Respondents’ Experience of Considerateness From Different Groups, by Percentage |
| Teachers (N = 32) | Other school personnel (N = 30) | Classmates (N = 32) | Other students at the school (N = 33) |
| Always | 9 | 10 | 16 | 6 |
| Often | 53 | 37 | 31 | 30 |
| Seldom | 25 | 37 | 34 | 30 |
| Never | 13 | 17 | 19 | 33 |

Note: Not all sets of percentages equal 100 because of rounding.
physical activities, which is a critical issue (Lieberman & Houston-Wilson, 1999). Forty-four percent of survey respondents reported that they did not participate in physical education. Seven percent of these nonparticipants said this was because they were exempted, 33% because they did not want to participate and 60% because it was not possible, even though they wished to take part.

Even though most of those who did not attend wished they had been able to do so, there were a number of barriers to taking part in physical education. Among students who reported having participated in physical education are also found those who reported partial restrictions. For example, one student said, “I had as many hours of physical education as my classmates, but alone. I wanted to participate within the class, but was denied by the teacher.”

Thirty percent of the students had earlier experiences of change of school type—back and forth between standard school and special school—during their time at school before the secondary upper school level, or in the shift between compulsory education and secondary-level education. There were students who attended standard school at the compulsory level yet were denied the opportunity to continue there. They were thus obliged to transfer, which was why they chose a particular school and program. Those who were both at the school of their choice and in the program of their choice were placed in the first category in Table 5, those who chose the particular school but not the program were placed in the second category in Table 5, and so forth.

Six of the nine students who reported that they had no options to choose between or were not allowed to choose attended the special school.

A hard of hearing student with visual impairment who was attending a special school commented on his choice:

I wanted to continue at the nearby upper secondary school, but they could not offer the support I needed. I applied to the National School for Hard of Hearing Students, but I was not accepted. The only possibility that existed was the National School for the Deaf. I was forced to learn sign language, which was extremely tiring during the first year, but now I am very happy about the choice and very satisfied with the education.

**Considerateness**

**Table 5**  
Respondents’ Self-Reports of Options for Own Choice of School and Program  

<table>
<thead>
<tr>
<th>Option</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Could choose both school and program</td>
<td>6</td>
</tr>
<tr>
<td>Could only choose school</td>
<td>15</td>
</tr>
<tr>
<td>Could only choose program</td>
<td>47</td>
</tr>
<tr>
<td>Had no options to choose between</td>
<td>12</td>
</tr>
<tr>
<td>Was not allowed to choose</td>
<td>15</td>
</tr>
<tr>
<td>Other response</td>
<td>3</td>
</tr>
<tr>
<td>No response</td>
<td>3</td>
</tr>
</tbody>
</table>

*Notes. N = 34. Not all sets of percentages equal 100 because of rounding.*

Considerateness is interpreted as a way of showing respect, inclusiveness, and recognition; not showing considerateness is interpreted as showing lack of respect and being exclusionary.

In order further to explore the question of considerateness, a simple additive index was calculated of the considerateness shown by the four groups of people encountered by the student with deafblindness in the educational environment (see Figure 2). Hence, there was one group of students for whom 4 was the lowest possible value (no considerateness at all from any of the categories). Based on the considerateness values, two groups were constituted according to the median. The first group, labeled no considerateness ($n = 16$), ranged from 4 to 9, and the second group, labeled considerateness ($n = 13$), ranged from 10 to 16.

There was a tendency toward polarization. Ten of the students reported a feeling of a rather low level of considerateness (≤ 9) from people they encountered in the educational environment, while 11 students reported a high degree of considerateness (results of 10 or higher).

It was found that among the respondents more men (57%) encountered considerateness (i.e., belonged to the considerateness group) than did women (33%). Experience of a feeling of considerateness was also more common among hard of hearing students with vision impairment (55%) than among deaf students with vision impairment (38%). It is furthermore important to note that there was no difference regarding the feeling of considerateness in relation to the period when the students attended the school, that is, 50% of those who attended school at the time of the investigation belonged to the considerateness group, compared to 50% of those who had left more than 10 years previously. That means no improvement over time regarding considerateness can be estimated from these results. However, surprisingly, it was found that of those respondents who attended a special school (Upper Sec-
ondary School for the Deaf and Upper Secondary School for the Hard of Hearing), only 40% belonged to the considerateness group, while of those students who attended standard schools, 67% belonged to the considerateness group.

A specific illustration of the feeling of considerateness was found in the answers to the question “Did the teacher or the interpreter used to show what your classmates answered, i.e., show the turn taking; show, themselves, what other students signed; hand around the microphone, or repeat what the other students answered?” Of those belonging to the considerateness group only 1 (of 13) answered “seldom” (no one answered “never”). Among the 16 students belonging to the no considerateness group, 10 (63%) answered “never.”

Considerateness is also reflected in how much the students liked their education. Eight-five percent of the considerateness group felt very happy with the education they received, while 50% among the no considerateness group were happy.

**Knowledge of Deafblindness**

During the work with the questionnaire and discussions with students with deafblindness, the issue of who in the students’ environment knew about their deafblindness was frequently raised. It seemed to be a “hush-hush” matter for many of the students. Therefore, the respondents were asked to list which categories of people in the educational environment they thought knew of their deafblindness. Six categories are included: teachers, other school personnel, classmates, other schoolmates, doctors, and other health personnel. As was done with considerateness, an additive index was constructed; the higher the value, the greater the number of categories of people who knew about their deafblindness (yes = 1, no = 0). The median is between three and four categories that knew; the group of students who reported that 3 or fewer categories knew is labeled less known; those who reported that four or more categories knew is labeled well known (see Figure 3).

It should be noted that nine students reported fewer than three categories. It was more common for the hard of hearing students than among the deaf students for people in several categories to know. Among the hard of hearing students, 68% reported that four or more categories knew, compared to 53% of the deaf students. Another result was that among those attending standard schools, 86% reported that many categories knew, compared to 56% of those attending special schools. A comment from a deaf student with visual impairment illustrates this finding:

> The staff at the school should have good knowledge about deafblindness. Since this was not the case when

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**Figure 2**

Experience of Degree of Considerateness

![Figure 2](image-url)

**Notes.** Considerateness shown by people in four categories (teachers, other school personnel, classmates, and other schoolmates) was measured. Intensity was gauged as a value between 4 and 16. Va., Value. N = 29.
I attended the school, I did not tell anyone about my vision impairment. However, today I have accepted my vision impairment and recognize myself as deafblind. It is a relief to have the support I need.

One can also argue that knowledge about a student’s deafblindness is a precondition for showing considerateness, and that there is thus a strong correlation between these two variables. Among the students who belonged to the considerateness group, 85% said that many categories of people in their environment knew about their deafblindness. However, among those students who reported no considerateness, only half said that many categories of people knew. A tentative conclusion is that knowledge seems to be a precondition, but not sufficient, for showing considerateness.

Discussion

It should be emphasized that the present study is based on responses from 34 students with postlingual deafblindness. This might seem to be a small number of respondents from which to draw any firm conclusions. However, one must bear in mind that the population being described is indeed very small, and that the people who participated in the study probably constitute the majority of the entire population of the students with postlingual deafblindness in Sweden, the target population for this study, and at least 50% of it. Nevertheless, the fact remains that, given the apparently small number of respondents, care must be taken with the interpretation of the results. Further, although considerateness and recognition are general phenomena, we cannot generalize our results to students with prelingual deafblindness and those with additional cognitive impairments. The factor brought to the fore in this study, “considerateness,” is one of the most important issues; it seems to reflect a dimension in education that appears with inherent consistency in the analysis since it correlates systematically with a number of other factors.

Another factor of which to be aware in interpreting the results is that some of the respondents left school a number of years ago. There might be issues of “skewing” with regard to retrospective questions: It is a well-known fact that individuals’
memories and descriptions are often given in the light of later experiences, some time after those events under description actually occurred. This is a possible shortcoming, yet there are some interesting findings that support the notion that the retrospective bias might not indeed distort the result. No specific tendencies in reporting considerateness, environmental knowledge, and other factors in relation to when the students finished school were observed. While this does not in itself demonstrate a lack of bias (bias probably exists), it nevertheless shows a stable pattern over time, which has to be taken seriously.

One aspect of participation of importance for students with deafblindness (Eriksson & Granlund, 2004) is prerequisites for participation, or as the ICF puts it, “having access to needed resources” (World Health Organization, 2001, p. 19). We found that there were a number of environmental factors that some of the students considered as constituting a barrier to participation. Levels of light and color in other parts of the school other than the classroom, hearing devices, adapted school books, slower signing, and the teacher’s awareness of the need for special attention in communication were among the factors that seem to be in need of improvement. Since such factors are prerequisites for participation, they must be given due attention; if the education system fails to provide such prerequisites, the whole education is jeopardized. While it could be assumed that this situation has improved over the last decade, our data fail to confirm any such improvement. No changes over time were revealed; on the contrary, it was striking how consistent the pattern of reporting actually remained. Students who left school about a decade ago had more or less the same experiences regarding prerequisites as those who were still studying at the time of the investigation.

The issue of diagnosis has been criticized in several studies on special education because it has been used to stigmatize students with impairments. However, we concur with Williams (2006) that “corporeal realities” play a role. It is considered that the type of impairment and degree to which the body is impaired play a significant role in education, when the given student has deafblindness. Examples include the fact that students with deafness require teaching media that uses sign language. To this group of students with deafness, visual adaptations or sign language in tactile modality must be added. Knowledge of the diagnosis gives helpful information about, for instance, how vision and hearing function. The diagnosis of Usher syndrome involves natal or very early hearing impairment (in some cases, total hearing loss), as well as visual impairment that leads to total night blindness, impaired light adaptation, impaired contrast sensitivity, a gradually restricted visual field, and gradually impaired visual acuity. Students with Usher syndrome may be dazzled by the whiteboard and by white paper. The deterioration of contrast vision leads to problems in communication activity, specifically in visually following what another person is signing. If the person signing is not dressed in clothes whose color contrasts with the color of his or her hands, or if the background has no nondazzling contrast color to facilitate communication, then there may be limitations in the receptive side of the communication. This is an example in which the impairment is significant, yet not enough in itself to create restrictions on participation.

Information on the degree of the impairment may also facilitate cooperation. Students with blindness may benefit from talking books, which other students with deafblindness cannot always use because of the degree of their hearing impairment. A personal factor such as skill in reading Braille is another important factor.

To be overlooked is to experience lack of recognition. Participation in education is a legal relationship, which includes the possibility of availing oneself of equal rights. Several of the barriers in the environment we have mentioned can also be viewed as the type of lack of recognition with detrimental effects on the self-respect of the individual with deafblindness.

Another dimension of participation (Eriksson & Granlund, 2004) concerns perceptions of participation. These have to do with being included and being accepted, and with the subjective feeling of belonging. A very important aspect of this is the student’s feeling of considerateness, regarded in the present study as a personal factor. For a deafblind person to have the feeling that people in the proximate environment show considerateness is for that person to be part of an ongoing situation, a subjective element of being included.

The importance of considerateness is also reflected in the students’ feelings about their secondary education. Of those students who reported positively on considerateness, only 2 (of 13) felt unhappy with their time at school, compared with half of the group (7 of 14) who reported no considerateness. The feeling of being considered seems to be a key factor in satisfaction with one’s education.

A third aspect of participation (Eriksson & Granlund, 2004) is activity and behavior. We have already discussed the behavior of the persons in the students’ educational environment, with the focus being on the behavior of the student with deafblindness. To inform persons in their environment about their disability is...
another aspect of recognition. It has been demonstrated that a precondi-
tion for showing considerateness, and hence recognition, seems to involve
knowledge about the other person’s
disability, in this case deafblindness
(Kirchner & Diament, 1999a, 1999b).
However, it was also noted that this
was in itself not enough to overcome
ignorance. Many of those who knew of
the students’ deafblindness showed
no considerateness, which indicates a
process of disrespect and stigmatiza-
tion. This process could be an efficient
barrier for students with deafblindness
in informing persons in their educa-
tional environment about their disabil-
ity. Compared to hard of hearing
students, deaf students seemed to be
more reluctant to inform others of
their visual impairment. Yet again, the
issue of being included in a commu-
nity comes to the fore.

If you are not recognized as the
person you are—a student with deaf-
blindness—in the community to
which you wish to belong, your self-
esteeem will be adversely affected.
There will also be a severe impact on
the possibility of establishing good so-
cial relationships with peers and the
risk of being stigmatized. It could not
be expected that people with low self-
esteeem would inform other people
about their deafblindness if they were
aware that this was stigmatized. This
also demonstrates how the different
aspects of participation interact with
other factors.

The importance of consideratenes-
seems also to have consequences
for the students’ individual behavior,
such as withdrawal and reluctance to
inform others about their disability;
these, in turn, demonstrate the inter-
action between environmental, per-
sonal, and participational factors.

Without discussion of the causes of
the students’ own respective deci-
sions, there is a risk of falling into a
new “blaming the student” trap. The
schoolbook example and the experi-
ences reported by students with
physical disabilities (Grue, 2001) do,
however, show that knowledge about
the impact of disability on activities
in education is a necessary field of
knowledge for all persons involved in
education. Research and training of
administrators (Luckner et al., 2005)
should therefore consider all of the
components of the ICF and the compo-
nents in order fully to recognize the
students.

However, observations have to be
followed up by studies in depth. Since
we found indications of mechanisms
at different levels of reality that restrict
participation, we consider that an in-
terdisciplinary research approach
(Bhaskar & Danermark, 2006) includ-
ing the education system, pedagogy,
and the students is required.

Note
Data for the present study were col-
lected for an investigation of educa-
tion at the secondary upper level for
students with deafblindness at the
Swedish National Agency for Educa-
tion. Many thanks for the agency’s
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clusion].undersaktar om S
SOCIAL RECOGNITION AND STUDENTS WITH DEAFBLINDNESS

PAPER III


Long-term ophthalmic health care in Ushers Syndrome type I from an ICF perspective

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Abstract

Purpose: The aim was to explore ophthalmic health care in female patients with Usher Syndrome type I (USH I) over 20 years and to evaluate the relationship between the ophthalmic health care and the health state of the patients from a health perspective.

Methods: A retrospective study of records from Ophthalmology Departments (OD) and Low Vision Clinics (LVC) from 1985-2005. Assessment of the reports was performed based on International Classification of Functioning, Disability and Health (ICF). Findings were analysed by manifest content analysis with ICF as a framework and using four themes: health care system, procedure examinations, patient’s functioning and disability and procedure actions.

Results: The records of 9 female patients (aged 25 – 39 years 1985) with USH I were selected from the national database of USH. A great number of notes were collected (OD 344 and LVC 566). Procedure examinations were exclusively oriented towards body structure and function. All patients showed aggravated visual impairment over and above the hearing and vestibular impairment. Procedures actions were oriented towards environmental factors. No correlation was found between procedures performed and patient’s experience of disability.

Conclusions: The high degree of resource allocation was not correlated to the patients’ impairment. The study indicates that the ophthalmic health care was characterized by inefficiency. This conclusion is very serious since patients very likely face severe disability and emotional difficulties. ICF ought to be incorporated in ophthalmic health care strategy in order to improve the health care.
**Introduction**

Human activities involve features with visual and audible meaning. Thus the ability to see and hear is crucial. Visual and hearing impairment in combination will therefore have great impact in daily life from both a medical and a health perspective. People with loss of both visual and hearing function (deafblindness) require health care that incorporates treatment, rehabilitation, service for people with disabilities and specific assistive devices, in order to manage daily life. The efficiency of this health care is therefore of great importance. We have so far not found studies that consider health care in a health perspective for people with deafblindness.

**Usher syndrome, type I**

Usher syndrome (USH) is a genetic disorder with autosomal recessive inheritance that entails both visual and hearing impairments and is the most common cause of deafblindness before older ages.

The syndrome is divided in three distinct clinical types, Usher Syndrome type I-III\(^2,3\), which are distinguished by different genetic mutations. Nine genes associated with USH have so far been identified\(^4\). The overall prevalence of USH was observed to be 3.3/100000 in Sweden\(^5\) and is estimated to be the most common cause of early onset of deafblindness. The prevalence of Usher Syndrome type I (USH I) is estimated to be 1.6/100000, and is the most frequent type in Sweden\(^5\). Research in USH has generally focused on prevalence, clinical and genetic diagnostics\(^6,7\).

USH affects the structure of the cochlea, the structure of the vestibular organ (type I and III)\(^2\), and the structure of the retina bilaterally. In the cochlea and the labyrinth the hair cells are damaged and in the retina the rods and cones are gradually undergoing degeneration/deviation (further on the term deviation is used to signify body structure impairment). The disorder in the eye is named Retinitis Pigmentosa (RP), which comprises several genetic disorders affecting the retina\(^8\). The RP in USH has been demonstrated to have a rather slow progression\(^6\).

USH I cause congenital profound deafness, bilateral vestibular areflexia and progressive visual impairment\(^2\). The first symptom of USH I is a congenital profound hearing
impairment/deafness. The first symptom of the visual impairment is night-blindness. The
mean age for diagnosis of USH I in a Swedish study is 14 years (range 2-57 years). Visual
field impairment, which starts in the midperiphery, occurs later with a gradual loss of visual
acuity. A common sequel is subcapsular cataract (> 80%). Between the ages of 30 and 40 the
visual field is usually restricted to 10 degrees, and visual acuity is 0.5 – 0.3. The visual acuity
can remain normal even in individuals with advanced RP with a small island of remaining
visual field. Visual acuity ≤ 0.1 (legal blindness) is estimated to be 25% at the age of ≈ 50
and 50% at the age of ≈60. The likelihood of losing total vision in both eyes is rare in USH I
although these patients may present with severe visual impairment. The visual impairments
have been found to be worse and increase with age in people with USH I compared to those
with USH II.

Children with USH I have inborn deafness, which means that they have never experienced
sound. It is reported that they often have a delayed walking age (1½ - 2 years) due to the
vestibular impairment, are insecure in darkness, and are often clumsy and bump in to
things. Since they have deafness they usually speak sign language and are generally
educated in special residential schools for the deaf. It is often in school they discover that they
are different from other people with deafness, which is reported to be the first of many
emotional crises. Restrictions in participation, especially in various social settings is
reported to be increased due to aggravated impairment of visual acuity. For an overview
of impact on communication see Rönnberg et al. Studies that highlight the impact of
environmental factors, such as darkness or glare, experiences of isolation from the Deaf
community and personal factors, such as refusal to use a white cane is also found. It is
reported that independence decreases along with increasing age, especially in USH I.

When providing health services for people with deafblindness additional knowledge and
skills are required i.e. in communication, about Deaf culture and community standards and in
adaptive technology. It is likely to assume that persons with USH I face extensive impact
in daily life: several activity limitations; several participation restrictions; and emotional
challenge. It is therefore likely to assume that they, especially as adults when visual
impairment aggravates would have comprehensive needs of health care that is rehabilitation
and support. Since these persons have congenital deafness and aggravated visual impairment
Ophthalmic health care would probably play a significant role.
International Classification of Functioning, Disability and Health

International Classification of Functioning, Disability and Health (ICF)\(^7\) will be used as a frame. ICF can be used for a wide range of purposes. It is possible to describe health and health-related impact by using body, individual and social perspectives. Furthermore ICF provide possibilities to describe the interplay between bio-psycho-social aspects of impairment and the context, which in ICF are environment and personal factors. Though ICF is not a theory it has been used in research and is gradually accepted as a concept in health and disability issues\(^9\).

ICF is a universal classification of disability and health and is a radical shift from emphasizing people’s disabilities to focus on their level of functioning in the domain of health. ICF is a complement to an etiological framework that focuses on diseases, disorders and other health conditions, i.e. a bio-medical perspective. The aim of both approaches is for the individual to achieve a life where they can exploit his or her opportunities to the fullest possible degree\(^7\).

ICF includes six components that are bound together in an interactive relationship. Disability is valued as a negative outcome and functioning is a positive outcome of the interaction between an individual with a health condition and the contextual factors. The components are: body function (including both physical and psychological functions) and its negative aspect impairment; body structure (the anatomy) and its negative aspect is deviation; activity (is execution of a task or action by an individual) and its negative aspect limitation; involvement in life situations is participation and the negative aspect is restriction. The context is divided into two components: environmental factors (including the physical, social and societal environment); and into personal factors (e.g. age, gender, background, past and present experiences, coping style etc.)\(^7\).
Functioning denotes the positive aspects of body function, body structure, activity and participation and disability is the complementary term that encompasses impairments, limitations in activities and restrictions in participation. The context plays a critical role in these relations because these factors have the power to facilitate or function as a barrier.

Medical health care in Sweden

Medical health care in Sweden is public and provided by local self-government (county councils and municipalities) with a considerable degree of autonomy. The service is regulated by legislation. Health care concerning diseases that cause visual impairment is usually provided as outpatient care at specialized departments. Ophthalmology Departments (OD) provide diagnostics and medical treatment. Low Vision Clinics (LVC) supplies patients with rehabilitation and assistive devices. Access to LVC requires a referral from the OD. Additional services for people with disabilities are provided by various other agencies. Both OD and LVC are obliged to document the care in written case notes in patient’s records. Notes should be dated and verified by name, profession and signature.

The underlying problems related to the management of the health care are significant because managers are responsible for the health care and have the power to accomplish organizational alterations. Managerial evaluation is usually concerned with ensuring that the
service is delivered properly and that available resources are used to their best effect. In this study we have delimited us to investigate only the ophthalmology and low vision care. This is maybe the most important part of the health care provided for people with USH I. As will be demonstrated later in this article this care is very extensively provided. Furthermore, this type of care has so far not been scrutinized and assessed from an ICF perspective.

To sum up this article focuses on the health service provided for people with deafblindness. The background is the growing awareness of and concern for, firstly, the resource allocation and secondly, the emphasis on participation as the most important aspect of disability. The latter will be elaborated within the conceptual framework of WHO’s International Classification of Functioning, Disability and Health (ICF). By combining these two aspects – resource allocation and participation – it might be able to assess whether the resources within the health care sector are optimised from an ICF perspective. The approach in this study is to investigate the services people with deafblindness have been provided by the Ophthalmology Departments and the Low Vision Clinics in Sweden. The actions, taken by these agents – here labelled procedures – will be categorised according to ICF. Procedures refer to examinations and interventions. Functioning and disability of the patients over the 20-year period the investigation covers is also recorded. Hence, the study will be able to demonstrate the relationships between the procedures, and functioning and disability.
The aim of this study was 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.

The main questions were:

- How were the ophthalmic health care systems structured: staff and professions?
- Which procedures were used within the ophthalmic health care system?
- What was the number of different procedures?
- What was the relationship between the procedures: examinations and actions?
- How did the patient’s functioning and disability develop?
- What was the relationship between the health care provided and the patient’s functioning and disability?
Materials

The material analysed was from the written case notes in patient records during the study period, from 1985 to 2005 (20 years) by both OD and LVC.

Nine female patients were selected from a national database of Usher Syndrome\textsuperscript{21}. 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 cohort. Differences in degree of visual field and hearing impairment between the groups were not significant. Median age at inclusion 1985 was 28 years (range 25 – 39 years) and at the end of the study period 48 years (range 45 – 59 years). In 5 patients the clinical diagnosis was confirmed by genetic analysis. The initial contact with the departments and clinics was established before 1985.

These patients have a congenital profound deafness; therefore records from the Audiology/Hearing Centre or Department were excluded. Other medical files were also excluded.
Methods

Since the material comprised text from the records, a text analysis method was used. The information in patient records is considered to be legally and medically accurate and reliable. This may also be the only way to compare health states over time. The re-typed records were stored in a qualitative database (QSR NVivo 2.0), which was used in the analysis process.

The following four themes, that represent an ideal image of the health care process, were adopted:

1. Health care system,
2. Procedure examination,
3. Patient’s functioning and disability
4. Procedure actions.

Health care system has the aim to gain an overview of the ophthalmic health care that provided the care. This theme was used to describe managerial issues. In this topic, the number of notes, ophthalmic health care units, professionals and professions were counted. The term health care system was used as the phrase has been used by WHO when performance and effectiveness of health care related to ICF are in focus.

Procedure examination has been used to determine the ophthalmic care’s assessment to obtain information about the patient’s functioning and disability. Examinations cover all inspections and examinations of the patient, which were reported in the records.

Patient’s functioning and disability is the reported outcome of the above-mentioned examinations.

Procedure action in the fourth step is judged to be the ophthalmic health care response to the patient’s functioning and disability. Actions cover the other procedures, which were reported in the records.

Text in the records, with other content than these four themes was not included in this study. This was for instance where the interaction with the health care and the patient took place, if professionals other than OD and LVC participated, the patient’s emotional expressions etc.
We used manifest content analysis, which describes the visible and obvious in the text. The meaning unit is the specific note at a certain occasion. The meaning units in the text were condensed and abstracted into categories and then into codes within the categories. Categories were created inductively except for categories of the degree of impairment.

The second author assessed the reports of the retina, the visual field and the visual acuity and put them into categories.

The retina had been assessed by inspection of the posterior eye. The retinal deviation that is reported in 'Patient’s functioning and disability’ is of the best eye. The degree of retinal deviation was categorized into four categories by the above-mentioned author. The categories include: (1) thin blood vessels, (2) pigmentation outside the papillomacular area, (3) the papillomacular area, macula affected, (4) total pigmentation with less than a papilla diameter area of normal retina left.

Examination of the visual field had been assessed by Goldman perimetry. The visual field reported in 'Patient’s functioning and disability’ is of the best eye assessed by the largest object and best luminance (V:4). Categorization of visual field was made by using a system applied from Grover et al. (Table 3).

Examinations of visual acuity had been assessed by Snellen charts or by refraction. The visual acuity reported in 'Patient’s functioning and disability’ is from the best eye with best correction. Categories of visual acuity were used in accordance with Sadeghi et al. (Table 3).

The study was based on informed consent and the Research Ethics Committee at Örebro University Hospital had approved the project.
Results

The presentation of the results will follow the ideal model of the process described in Methods.

HEALTH CARE SYSTEM

In total eleven health care districts in six county councils had provided the ophthalmic health care. Of the notes collected, 62% came from LVC. The structure of the care provided is shown in Table 1.

Table 1  Structure of the provided health care

<table>
<thead>
<tr>
<th></th>
<th>Ophthalmology Department</th>
<th>Low Vision Clinic</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of Professionals</td>
<td>35</td>
<td>43</td>
<td>78</td>
</tr>
<tr>
<td>No. of Professionals</td>
<td>3</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>No. of Notes</td>
<td>344</td>
<td>566</td>
<td>910</td>
</tr>
<tr>
<td>No. of Procedures</td>
<td>724</td>
<td>877</td>
<td>1611</td>
</tr>
</tbody>
</table>

The number of professionals was relatively even between the two agents of the health care. In OD there were three different professions (Oculist, Nurse and Counsellor) involved and in LVC seven professions (Optician, Low vision therapist, Mobility teacher, Counsellor, Occupational therapist, Therapist of assistive devices and Administrator).

PROCEDURE: EXAMINATIONS

As we saw in Table 1 a considerable number of procedures were found; in total 1611. Of these procedures, examinations comprised 37%. OD conducted a large majority of these, 79%, and there was no difference between the first 10-year period and the second ten years. This could be compared with LVC’s, which increased their number of examinations from 46 to 78 in the second 10-year period. (See table 2)
Examination of Body structure and Body function

If we take a closer look at the examinations we find that most of them were examinations of Body functions (72%). And the rest were examinations of Body structure (28%). Of the latter OD reported 95%. These examinations comprised inspection of anterior and posterior eye; ophthalmotonometry and examinations concerning cataract extraction. Examination of body structure was likewise reported in both 10-years periods.

A closer look at the examinations of the Body function shows that Visual acuity was the most common examination (65%). The second most common examination of body function was visual field (21%).

The distribution between different types of examinations of body function is shown in Table 2.

Table 2 Number of examinations of body function, distribution between OD and LVC and into the two decades

<table>
<thead>
<tr>
<th></th>
<th>Ophthalmology Departments</th>
<th>Low Vision Clinics</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1985-1994</td>
<td>n</td>
<td>Min - Max (per Pat.)</td>
</tr>
<tr>
<td>Visual acuity</td>
<td>82</td>
<td>(3 - 21)</td>
<td>81</td>
</tr>
<tr>
<td>Visual field</td>
<td>48</td>
<td>(0 - 10)</td>
<td>40</td>
</tr>
<tr>
<td>Contrast vision</td>
<td>5</td>
<td>(0 - 4)</td>
<td>12</td>
</tr>
<tr>
<td>Colour vision</td>
<td>13</td>
<td>(0 - 4)</td>
<td>11</td>
</tr>
<tr>
<td>Light adaptation</td>
<td>5</td>
<td>(0 - 3)</td>
<td>8</td>
</tr>
<tr>
<td>Electrorheinography</td>
<td>2</td>
<td>(0 - 1)</td>
<td>2</td>
</tr>
<tr>
<td>All examinations of body function</td>
<td>155</td>
<td>154</td>
<td>42</td>
</tr>
</tbody>
</table>

Examination of activity/ participation and environmental and personal factors

It is notable that there were no reports concerning examination of activity/participation such as learning and applying knowledge, general tasks and demands, communication, mobility, self-care, domestic life, interpersonal interaction and relationships, major life areas or community, social and civic life.

Furthermore there were no findings of examinations of environmental and personal factors in the records during the follow up period.
In summary, examinations in the ophthalmology health care are exclusively oriented toward body structure and function and with OD being the more active of the two agents involved in the health care.

PATIENT’S FUNCTIONING AND DISABILITY

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.

Body structure

At the beginning of the study period the patients were aged 25-39 and all of the patients showed retinal deviation. Seven of them had pigmentation outside the papillomacular area, which were classed as category 2 and the remaining two had less than a papilla diameter of normal retina left classed as category 3. Ten years later two more patients had also reached category 3. At the end of the study period all of the participants except one were classed having category 3.

Deviation of the eye was found regarding bilateral cataract. At the beginning of the study period six of the patients suffered from cataracts.
Body function

Decreased visual fields were found in all patients from the beginning of the study period. Two of them had some peripheral scotoma and the other seven had tunnel vision. Further decrease of visual fields during the follow up was found in all but two patients and by 2004 all but one had marked concentric loss of visual field \(< 10^\circ\).

Decreased visual acuity (\(\leq 0.5\)) was found in all patients at the beginning of the study period. Further decrease in visual acuity during the follow up period was found in all but two patients. By 2004 four had a visual acuity of 0.1 (legal blindness) or worse and three patients had a visual acuity below 0.05.

Activity; participation; environmental and personal factors

Since no reports of examination of activity, participation or of the roles of the environmental and personal factors were found it was not possible to show limitations in activity or restrictions in participation.
In summary, the fact that USH I is an aggravated phenomenon is clearly demonstrated in the examination of the patients vision. All patients showed aggravated impairment of vision during the study period.

PROCEDURE: ACTION

The other type of procedure, actions, comprised 63% of all the procedures. In contrast to the other type of procedures (examinations) LVC is the more active agent conducting 75% of all the action procedures. It is notable that the number of actions increased, from 224, three times, to 539 in the second 10-year period.

For a further analysis we have coded the actions into four main categories according to ICF. These are actions towards the body; activity/participation; environmental factors and personal factors. An uneven distribution of the actions was found, with 89% of them oriented towards environmental factors.

**Actions toward the body (structure and function)**

Actions towards the body, i.e. treatments, comprised 4% of the actions, a rather small share of the total actions. All treatments were reported by ODs. Ten reported actions of cataract extraction were found in five of the patients and 35 reported actions whereby (all) patients were prescribed with vitamin E and/or A.

**Actions toward activity/participation**

The second type of action was oriented toward activity and participation and comprised 6% of the actions. LVC reported all of them. These actions included; Braille practice, mobility training and training to handle assistive devices. However, thirty-two of the 56 actions reported were found in one patient’s case notes and was found to be mostly Braille training. The remaining 24 actions were; mobility training; training to handle assistive devices and Braille training.
Actions toward environmental factors

As already mentioned the overwhelming part of the actions comprised actions towards environmental factors 89%, e.g. assistive devices or other service. LVC reported 77% of these actions. In Table 4 we show the distribution of the different types of actions toward environmental factors.

Simple certificates comprise doctor’s certificate on sick leave. Extensive certificate comprise doctor’s certificate on entitlement of pension and other labour-intensive certificates.

Both agents reported conferences and administration. Half of the conferences reported by OD and 45% of the conferences reported by LVC were conferences without the patient present. Administration comprised 234 actions. OD reported 86 actions towards administration, which were 34% of their actions. LVC reported 148 actions towards administration and this was 19% of their actions. Administration increased in the second 10-year period.

We can see that it was only OD which reported various certificates; simple certificates of e.g. illness, extensive certificates e.g. grant a disablement pension etc and referrals to other departments. On the other hand it was only LVC, which provided assistive devices and made adaptations in the patient’s home.

### Table 4
Number of actions directed toward environmental factors distribution between OD and LVC and into the two decades

<table>
<thead>
<tr>
<th></th>
<th>Ophthalmology Departments</th>
<th>Low Vision Clinics</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Handling assistive devices</td>
<td>105  (0 - 43)</td>
<td>286  (12 - 53)</td>
<td>391</td>
</tr>
<tr>
<td>Adaptation in the home</td>
<td>50   (0 - 12)</td>
<td>72   (0 - 19)</td>
<td>122</td>
</tr>
<tr>
<td>Referrals</td>
<td>14   (0 - 7)</td>
<td>11   (0 - 5)</td>
<td>25</td>
</tr>
<tr>
<td>Certificate simple</td>
<td>31   (0 - 13)</td>
<td>12   (0 - 5)</td>
<td>43</td>
</tr>
<tr>
<td>Certificate extensive</td>
<td>30   (0 - 9)</td>
<td>16   (0 - 5)</td>
<td>46</td>
</tr>
<tr>
<td>Conference with the patient present</td>
<td>1   (0 - 1)</td>
<td>3   (0 - 2)</td>
<td>3   (0 - 2)</td>
</tr>
<tr>
<td>Conference without the patient</td>
<td>4   (0 - 4)</td>
<td>11   (0 - 5)</td>
<td>6   (0 - 5)</td>
</tr>
<tr>
<td>Administration</td>
<td>35   (0 - 8)</td>
<td>51   (1 - 20)</td>
<td>48   (0 - 18)</td>
</tr>
<tr>
<td>All</td>
<td>115</td>
<td>93</td>
<td>217</td>
</tr>
</tbody>
</table>
**Actions toward personal factors**

The only action towards personal factors was therapeutic dialogue, which was found in the records of both OD and LVC and comprised one percent of the total number of actions. During the 20-year period seven of the patients received this support.

The general trend in the health states of these patients was aggravated visual impairment, we found however no correlation between the changes in functioning and disability, examinations and actions.

In summary, the overwhelming part of the actions in the ophthalmic health care was oriented toward environmental factors and it is LVCs that is the more active of the two agents.
Discussion

The aim of the study was to explore the ophthalmic health care and to evaluate the relationship between the ophthalmic health care and the patient’s functioning and disability in a long term follow up and hence to assess the resource allocation from an ICF perspective.

This is to the best of our knowledge the first study concerning ophthalmic health care in patients with USH I. We are fully aware that ICF has only been available for 3 of the 20-year follow up. Thus by applying the health scope we could explore the reported practice behind the organizational division of the ophthalmic health care.

The findings were based on written materials in patient records from OD and LVC in 11 health care districts in 6 of the 18 counties in Sweden. Health care records are assumed to be a rich source of critical information\textsuperscript{20,22}. The records were generally in good order. Profession and name of the professional accompanied the signature. Pagination showed that no record was missing.

We delimited this study to one diagnosis, USH that causes deafblindness, in order to get patients with similar impairments. In Sweden, USH is the most common cause of deafblindness before older ages. The study was further delimited to USH I since this is the most common type found in Sweden.

The delimitation to one sex/gender was done in order to avoid possible sexual differences in aggravation of the impairments. Males with USH I have been found to have worse impairment than females\textsuperscript{6}. We also wanted to avoid possible differences in treatment due to gender\textsuperscript{25}.

The long-term follow up of 20 years was chosen in order to find out whether the patient’s functioning and disability and the treatments have changed during these years. The account of the body structure; retina and body function; visual field and visual acuity at three different points (inclusion 0, 10 and 20 years) made it possible to evaluate the decrease in visual impairment from inclusion and during the two decades. Delimitation of the follow up period to adult age was done in order to include the period where the progression of the visual impairment is established according to previous studies\textsuperscript{6}.

All these efforts were made to enlarge the trustworthiness of the study. The findings may be valid and representative for ophthalmic health care in other females with USH I since the follow up entailed several health care counties and districts within the counties, many
professionals, a considerable number of case notes and was analyzed over a long period of time 20 years, to follow up.

ICF was used as a conceptual framework in which we applied ICF components to the patient’s functioning and disability and furthermore into what the procedures were dealing with.

Categorization of procedures as well as functioning and disability into ICF components made it possible to, in an integrated health perspective; compare the pattern of examinations with the pattern of actions and to put those in relation to alteration in the health state of the nine patients. This also made it possible to discover mismatch in the relationship between examination, functioning and disability and actions. This method is also suitable for use on other diagnoses as well as health care systems.

The retinal findings as well as visual function findings were divided in categories in order to get a general overview of the deterioration. The advantage of categorization is that it makes it easier to view the results. However a disadvantage is that although categorization of visual acuity and visual field has been used in some studies it has not as far as we know been evaluated. Due to a lack of categorization of retinal deterioration the second author developed one specific for this study.

The studied ophthalmic health care entailed a large amount of resources involving many employees even though there were only 9 patients. Overall, we found that the average annual invested resources were 9 procedures.

There seems to be a division of labour between OD and LVC where the former served the patients mostly in the medical perspective and the latter tend to mostly work in a health perspective. Thus overlapping (e.g. visual examination) was found in the records of both the agents.

The proportion of procedures per note showed a higher degree of intensiveness in OD’s 2.1, compared to 1.5 in the records of LVC (table 1). The great number of professionals (78) indicates staff turnover. This is notable since the syndrome is both complex and progressive, which will likely entail great impact in daily life. These facts require professionals that have knowledge about the impact and specific know-how to work with these patients.

There were fewer notes signed by for instance counsellors. There was also an absence of notes signed by oculist, psychologist, psychiatrist and physiotherapist at LVC.
From a health care system and cost efficiency perspective the time a professional spends on a certain patient at a certain time is also important. The records however did not reveal information about the length of time it took to carry out the procedures.

Our own experience of clinical and other interactions with people with USH I is that working with these patients is time consuming. An appointment with patients with USH I usually take more than twice the allocated time for other categories of patients. Patients with USH I, at least in Sweden, usually speak sign language. If the professional is not fluent in sign language and unable to adapt the signing to the patient’s different requirements i.e. alteration in the signing mode due to the aggravation of the visual impairment, then skilled interpreters are necessary. In Sweden, the health care system provides professional interpreters, usually two on each occasion, which further increases the number of professionals and costs involved in patients care. These overall costs are high.

When we analysed the notes we were given the impression there was a lack of communication between OD and LVC; patients had undergone examinations of visual acuity at both OD and LVC on several occasions within a month or two. This indicates a lack of communication that results in waste of resources.

The number of examinations per patient was not found to be equal. Surprisingly, we found that OD continued to examine the patients during the second decade (table 2). The percentage of reported examinations by OD was equal in the first and second 10-year period. This should be compared with LVC, which nearly doubled their rate of examinations (from 46 in the first 10-year period to 78 in the second). Furthermore, many reported examinations seemed more like ongoing rituals rather than using the limited resources cost-effectively for the patients’ best.

The increased number of actions reported by LVC seems reasonable due to the aggravated impairments found in all the patients. Though in two patients, the number of actions was found to be less in the second decade (table 3). The distribution of actions between the patients was unequal.

Impact of USH I in the retina, body structure, was found in all patients at the beginning of the study period. Impact in the lens of the eye, cataract was found in six patients in the beginning. In all patients the deviation of the body structure of the retina and lens was deteriorating during the follow up period. In concordance, the visual fields, body function, were also impaired resulting in this impairment being further aggravated. The progression of visual field loss showed at the end of the follow up marked concentric impairment of ≤10° in
7 of the patients but none showed totally blindness. This finding is also in concordance with studies of USH I\textsuperscript{6} and of RP\textsuperscript{8}.

Visual acuity, another body function, was impaired in all patients at the beginning of the study period. In all but one patient the impairment aggravated during the follow up period. Although visual acuity was considerably worsened for three of the patients none had total blindness. This result is also found in previous studies\textsuperscript{6}. Visual acuity may be stable until the deviation of the retina has affected the macular area. Two patients showed a faster aggravation of impairment in the visual acuity. The 14-year age difference between the youngest and oldest participant could not explain the differences in degree of impairments within the group, only within individuals, due to the follow up time.

Here, the general picture is that the patients’ eye structure and visual functions decreased in some cases considerably over the period. This should be added to the congenital total impairment in the hearing and vestibular functions. Over and above this it is likely to assume that these 9 female patients with USH I face severe and worsening limitations in activities and restrictions in participation during the follow up period.

Although the number of actions increased over the period, from 369 to 650, we found no correlation (Spearman’s rho) between the aggravated visual field impairment or the decline of visual acuity and the number of actions. Furthermore it is plausible to assume that such a development would trigger a larger number of actions focusing activity/participation and environmental factors. However we found that this was not the case.

The increase of actions is logic due to the aggravated impairments in these patients and we found that 75 % of the actions were reported by LVC. This is also logic due to the division of the ophthalmic health care into OD and LVC and to the changing requirements of the patients when their vision deteriorates.

Altogether 907 actions were judged towards the improvement of environmental factors. This was 92\% of the actions reported by LVC. Among the actions towards environmental factors approximately 26\% was administration. Paperwork was the most frequent action in OD and was increasing from 30\% to 55\% of the environmental actions in the second decade.

We also found a great number of reports with assistive devices and adaptation in the patient’s home as the object, which may facilitate the patients. There were no examinations that verified the requirement of these actions and no follow-ups that reported the outcome of the devices or the adaptation i.e. if they facilitated activity and participation or not. Thus assistive devices also demand resources.
The number of environmental facilities should be compared with the low number (56 reports) of practice (in Braille, mobility etc) especially since 32 of these reports were reported in one patient. Two patients received extensive rehabilitation by other caregivers. This means that for six patients no in-depth low vision rehabilitation was found.

In general many of the different types of examinations and actions were not found in all the records (table 2 and 4). Furthermore the distribution between the percentages of examinations in relation to all procedures varies from 27% to 49%.
Conclusions

The high degree of reported procedures indicates great resource allocation, which is important due to the complexity and progression. It is likely to assume increasing disability for these patients during the follow up period. The higher frequency of procedures by LVC in the second decade is also logic due to the division between the two agents. However, the findings of the health care system reveal distortion in number and type of professionals; professions; examinations and actions, which are not correlated to the development of the syndrome. The distribution of reported resource allocation between the patients was not equivalent with either the pace of aggravation or the degree of the visual impairment. The most important imbalance from an integrated health perspective was the absence of examinations of activity, participation, environmental factors and personal factors. The ophthalmic health care’s records do not reflect the patient as a whole, i.e. bio-psycho-social impact in relation to patient’s physical and social environment and their personal strengths and shortcomings. The ophthalmic health care system was furthermore characterized by: double work; time consuming and expensive work. The indication of inefficiency in the studied ophthalmic health care is worth attention since the syndrome is severe and the invested resources are great both from the perspective of the patient and that of the health care. These conclusions are very serious because patients very likely face severe disability and emotional difficulties.

We suggest that an integrated health perspective, i.e. ICF is incorporated in the ophthalmic health care in the care of patients with USH I in order to improve the health care. The disease is still incurable and those affected are living their lives with three severe impairments, deafness, vestibular areflexia and aggravated visual impairment and it is therefore likely that they will experience severe disability. Reorganization of the ophthalmic health care system, including professions and procedures using a health perspective framework may lead to a higher degree of efficiency without increasing costs. We also call for more in-depth studies of the impact the syndrome has in participation together with environmental and personal factors.
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Participation in People with Deafblindness:
An ICF and the Life-Course Perspective

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Short title: Mechanisms that influence participation

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Abstract

The objective is to discuss mechanisms that have an impact on participation in relation to the rehabilitation and support of females with deafblindness.

The design is a qualitative analysis using ICF and the Life-Course as the framework.

The informants are three females with Usher Syndrome type I and their patient records from Ophthalmology Departments, Low Vision Clinics and Audiology Departments

Methods used are interviews (three occasions) and the retrospective follow-up of service categorized into ICF components in the ≈ 45 – 50-year life span.

Results show that there is a complex play of mechanisms that vary over time. Services have failed efficiently rehabilitate informants. Arbitrariness was found in the provision of support.

Conclusion ICF and the Life-Course perspective have brought about an understanding of mechanisms in relation to participation. Rehabilitation based on ICF and supplemented with the Life-Course perspective should be encouraged and further developed.

Keywords: Deafblindness, mechanisms, ICF, Life-Course
Introduction

To have severe impairment in both visual and hearing functions, deafblindness is very rare. Deafblindness is likely to entail severe limitations in activities as well as restrictions in participation. These persons are assumed to require rehabilitation and support. Rehabilitation entails an assessment of patient needs. The International Classification of Functioning, Disability and Health (ICF) of the WHO is an integrative model of human functioning (World Health Organization, WHO, 2001) that makes it possible to assess the situations experienced by those with this disability.

A tentative ICF-based definition of rehabilitation have been developed by Stucki and colleagues (2007a, 2007b). It is a health strategy that “aims to enable people with health conditions experiencing or likely to experience disability to achieve optimal functioning in interaction with the environment” (Stucki et al., 2007; Stucki & Melvin, 2007). This approach to rehabilitation includes the diagnosis, prognosis and treatment of health conditions. Rehabilitation refers to a multi-sectoral and multi-professional strategy (Stucki & Melvin, 2007). Rehabilitation is one of four health strategies; the others are a preventive strategy, a curative strategy, and a supportive strategy. While the goal of rehabilitation is to restore or optimize functioning, the goal of support is to optimise the quality of life and to preserve autonomy (Stucki et al., 2007a). The strategies are not exclusive, and an individual can become the recipient of more than one strategy simultaneously. Rehabilitation and support are not confined to the health sector since, for instance, education labour and social affairs also use these strategies (Stucki et al., 2007a).

Shortcomings in service (Liberman & Stuart, 2002; Miner, 1995), including health care (Möller et al. submitted) and education (Giangreco et al., 1998; Möller & Danermark, 2007) for people with deafblindness have been found. These findings call for an improved knowledge of the mechanisms involved, especially those mechanisms that have a negative impact on participation, i.e., participation restriction.

This study discusses mechanisms with, in the short or long term, an impact on participation for those diagnosed as having Usher Syndrome; this entails severe hearing and visual impairment. ICF is used as a framework and this is complemented with the life-course perspective (Giele & Elder Jr, 1998; Priestly, 2003; Priestly, 2001). The discussion focuses on aspects of the impact on participation significant as regards habilitation and rehabilitation.
ICF (WHO, 2001) consists of six interrelated components. Five of the components have both a neutral (positive) term, and one denoting negative aspects. Body structure is the anatomy of the human body. Its negative term is, in general, impairment. In order to distinguish negative aspects in body structure from negative aspects of bodily function, the term deviation refers to negative aspects exclusively for body structure, while impairment is mostly related to bodily function. Bodily function refers to both physiological and psychological functions of bodily systems. Activity is understood as what an individual does and his/her capacity to do so. The negative term is activity limitation. Participation in ICF is about involvement in life situations (WHO, 2001, p. 191, short version). Involvement incorporates taking part, being included or engaged in an area of life, being accepted, or having access to needed resources (WHO, 2001, note at p. 19, short version). The negative aspect is labelled participation restriction.

Environment in ICF consists of various factors, such as nature - including sounds and light; products, e.g., hearing aids and spectacles; support and personal relations; and attitudes and services policy. Environmental factors with a supportive impact facilitate while those with negative impact are labelled barrier. The personal factors of an individual can be demographic, such as age and gender, and also include life experience, habits, and coping styles.

Participation in relation to disability has recently increasingly become a central concept in legislation and other policy documents. Participation is regarded as being a tool with the power to achieve other values, such as rights for those with disabilities. Participation has in some studies of students with deafblindness been valued as a multi-dimensional condition, where participation is dependent on the context, i.e., factors in the environment, and on personal factors (Almqvist & Granlund, 2005; Eriksson & Granlund, 2004; Möller & Danermark, 2007).

Perenboom and Chorus (2003) evaluated nine existing survey instruments in relation to participation in ICF. Their conclusion is that further discussion is needed in order for the achievement of an unambiguous picture distinguishing between activity and participation.

ICF is described as a bio-psycho-social framework. ICF is a classification and does not model the ‘process’ of functioning and disability (WHO, 2001, p. 25, short version). ICF provides a set of components for a description of a situation. It is nevertheless clear that the components of ICF are interrelated and the interaction can be seen as a dynamic process. The possibility of further downward spiralling consequences related to disability have been noticed and sometimes called ‘secondary conditions’ which may comprise further impairment, limitations or restrictions (Jette, 2006; Simeonsson et al., 2002).
The empirical illustrations used in this study are selected from three females who have a diagnosis that constitute deafblindness, Usher Syndrome (USH). USH is the most common cause of deafblindness before old age, and entails no other severe medical conditions (Kimberling & Möller, 1995; OMIM, 2007; Sadeghi, 2005). USH is divided into three distinct clinical types, Usher Syndrome type I - III, distinguished by different genetic mutations (Kimberling & Möller, 1995; OMIM, 2007). Usher Syndrome type I (USH I) is estimated to affect 1.6 individuals in 100,000, and is the most frequent type in Sweden (Sadeghi et al., 2004b). USH I has been found to have a more severe prognosis than has USH II, especially in males (Sadeghi et al., 2006). The likelihood of losing total vision in both eyes is low in USH I, although this type may develop severe visual impairment (Sadeghi et al., 2006). The eye condition is Retinitis pigmentosa (RP), an umbrella term for various causes of retinal degeneration. The pace of degeneration in USH is regarded as being relatively slow.

USH I is briefly described below, with ICF as a framework. In the case of USH there is a single gene deviation that brings faulty information (Sadeghi et al., 2004b). The impact of the deviate gene is an inherited deviation within haircells in the body structure of the cochlea in the ears, and of haircells in the structure of the vestibular organs (the labyrinth). The impact of the deviate gene on the eyes is the gradual aggravation of deviation, and the death of rod and cone cells in the retina. The impacts on bodily functions are consequently impairments in: hearing functions, vestibular functions, and – gradually – in visual functions. The impact of the impairments may be limitations in the activities to listen to sounds, to see what is in view, and to maintain balance when standing upright. The impacts of these limitations are restrictions in the possibility of participation in sounds such as spoken messages; in view, for example, of the speaker, and in situations that demand balance.

Described briefly from a life-course perspective, the impact of USH I is as follows. Hearing impairment in USH I is inherited (or developed in early infancy) and is profound; this has an impact on the possibility of acquiring a spoken language. The inherited vestibular impairment in USH I has an impact on the age of starting to walk, which in general is delayed to 18 months of age (Kimberling & Möller, 1995; Möller, 2007).

From clinical experiences the first known impact of visual impairment in USH I is a limitation to seeing in the dark (night-blindness), and is experiencing dazzle in glare. These impacts may not be recognized at their onset since the mean age for the diagnosis of USH I is 14 years of age (Sadeghi et al., 2004b). The deviation of the retina, beginning in the mid-periphery, occurs later. There are initially partial scotomas (blindness) and these gradually become complete. The deviation extends towards the macula structure in the centre, where the visual acu-
ity function is situated. Between the ages of 30 and 40 the visual fields in USH I are usually restricted to 10° (Sadeghi et al., 2006). This should be compared with normal visual field of 180 – 200° (Lennie & Van Hemel, 2002). In the age group 30 – 40, the visual acuity functions in USH I are found to be 0,5 – 0,3 (Sadeghi et al., 2006). This should be compared with normal visual acuity of 0,8 – 1,0 (Lennie & Van Hemel, 2002). Visual acuity ≤ 0,1 (legal blindness) is estimated to be 25 per cent at the age of ≈ 50, and 50 per cent at the age of ≈ 60 in USH I. Visual acuity in people with USH I can, however, remain normal even in individuals with advanced RP who have a small island of remaining visual field (Grover et al., 1999; Hartong et al., 2006). A common sequel to USH in adulthood is bilateral sub-capsular cataract of > 80 per cent (Sadeghi et al., 2006).

It is known that USH is progressive and that independence decreases with age (Damen et al., 2005). It has been established that people with USH face, in their daily lives, many difficulties associated with their dual impairments (Damen et al., 2005; Miner, 1995; Pollard et al., 2000; Vernon, 1969). Patients with USH I have been found to be less independent, thus to require more help than those with USH II (Damen et al., 2005). Among the difficulties experienced, information, communication, and mobility have especially been highlighted (Damen et al., 2005; Miner, 1995). Problems related to social relationships (Pollard et al., 2000) and emotional difficulties have also been described (Miner, 1995; Pollard et al., 2000; Rönnberg et al., 2002; Schneider, 2006). Those with USH I may face social challenges similar to those of the Deaf community (Cioffi, 1996; Miner, 1995). However, in general, knowledge of the impact of USH I on life-course approaches is rare (Möller, 2005) and is usually reported by those with professional experience of working with USH patients in rehabilitation (Miner, 1995).

Studies that combine participation for persons with deafblindness with ICF and the life-course perspective have not yet, to the best of our knowledge, been conducted.
Aim
The aim is to discuss mechanisms that have an impact on participation in a life-course perspective for some females with USH I in relation to rehabilitation and support.

The hypothesis is that there do exist mechanisms with a serious impact on the individual’s life, and it is thus important to address these in the rehabilitation process for persons with USH I. There are further assumptions that such mechanisms may be contemporary, may influence each other, and may vary in degree and quality over time; any present, as well as absent, mechanisms could have an impact, i.e., there is a complex play of mechanisms.

One goal in a broad sense is to contribute to a further understanding of participation as a part of functioning, and of restrictions in participation as part of disability - especially in the case of the rehabilitation of and support provided to females with USH I.
Materials and Methods

Informants with USH I were selected from the Swedish National database of USH (Sadeghi et al., 2004a), consisting of 293 cases of USH. Of these, 140 cases have USH I, confirmed by clinical and/or genetic diagnosis.

Three females with USH I were born within a relatively short period; they were aged ≈ 45 – 50 by the year 2005. Diagnosis, cases and materials pertaining to the cases were chosen in several steps by strategic selection (Merriam, 1988), thus the selection was accomplished according to certain criteria (see Discussion).

The database consists of medical information from medical file records. The entire patient records from the Ophthalmology Department (OD), Low Vision Clinic (LVC) and Audiology Departments (AD) are included. Interviews with a number of people with USH were conducted. Interviews with these three informants were performed in 1997 (on two occasions) and 2004 (one occasion).

Records and interviews in several cases of USH I were scrutinized in order to distinguish cases with a rich amount of data in the six sources.

Sources have different values depending on their authenticity; for instance, whether the source is a primary or a secondary one (Merriam, 1988). A primary source here comprises the informant, notes made by professionals at AD, OD and LVC about the results of examinations of the informants, and the professionals’ observations during their interactions with the informants. Hence, primary sources are referred to as: tell, write, and observe. Secondary sources are what the informant and other people (e.g., an informant’s relatives) told a professional, and that which a professional has documented in the patient records. Secondary sources are referred to as: report and note. Table 1 shows the sources.

Life-Course in research is, according to Priestley (2003), understood as an approach that links individuals with space (where) and time (when), and with societal phenomena rather than a description of individual life careers. Adopting this for research into disability entails examining the ways in which the lives of people with disability are understood, organized and governed within societies. Furthermore, these factors differ according to periods in life, to societal periods and to spatial location (Giele & Elder, Jr., 1998).

The analysis was carried out as follows. A passage with a text is, for instance, ‘I have to have correct light and the right reading glasses.’ Light and reading glasses are categorized as environmental factors. The impact is next assessed based, in this particular case, on informa-
tion given by the informant later on in the interview. In this example, light is a barrier if there is too little or too much, while if there is enough light and it come from a suitable direction it is a facilitator. Reading glasses are environmental products that facilitate participation.

We have also included accounts of such elements as legislation, the services system, policy, and attitudes that may for the three informants have an impact on participation.

All interviews were performed by the first author with the support of professional interpreters, skilled in interpreting for people with deafblindness. Both the interviewees and the interviewer are familiar with conversations in this mode.

The interviews were transcribed verbatim. Interviews and text in the patient records from the three departments were stored in case-wise documentation distinguishing among the six sources. The software QSR NVivo 2.0 was used during the coding process for this study.

To maintain the informants’ confidentiality, they were given the aliases Helen, Laura and Olga. Furthermore, a few facts that might easily identify them yet are irrelevant to the purpose have been changed.

The study was based on informed consent; the Research Ethics Committee at Örebro University Hospital approved the project.
Mechanisms with an impact on participation

The oral policy

Hearing impairment, a bodily function, was discovered when Helen and Laura were around one year old and Olga was three. They failed to respond to sounds in their environment since their hearing impairment was profound. The impairment had an impact on listening activities (hearing) and generally on participation in the messages carried by sounds. At this point in the informants’ lives the cause of the hearing impairment was not known.

Children with an inherited hearing impairment were in the late 1950s and early ’60s provided with rehabilitation by the health care authorities. The rehabilitation was as follows: firstly, the parents were urgently requested to teach their child to speak then, by looking at someone who is speaking, to learn to lip-read, trying to find out what the speaking person is saying (Pärsson, 1997). Secondly, they provided the child with hearing aids. The rationale for the oral approach was to make the children as “normal” as possible; at that time, “normal” equated to being like hearing children. The latter step, providing hearing aids and listening training, was assumed to improve hearing (Ahlsén, 1991). At that time such a type of rehabilitation was considered to be a facilitating environmental mechanism at the societal level, counter-acting the “body mechanisms” of hearing impairment. Furthermore, parents’ efforts were also recognized as comprising a facilitating environmental mechanism at the social level. Taking part in the training (participation) was a mechanism at the individual level. What the children learned can be categorized as a personal factor.

The informants had speech training at school, too: they were trained by professionals during both compulsory junior school education and at secondary school. The policy to provide training at school was recognized as a facilitating mechanism at the societal level, since the professionals performed the training in their role as professionals. Although the training was carried out face-to-face with the informants, its execution is a mechanism on the social level.

It is, however, recorded in AD records that when the informants were in their late teens they had not developed intelligible speech, and their lip-reading skills were limited. Thus the informants had severe limitations in communication, both as a receiver and a producer of speech. However, one of the informants, Helen, reports: *I could lip-read so there were no problems.* Divergent reports from professionals and patients are important findings that may reflect different types of knowledge and different experiences. Hearing aids, an environmental
mechanism, did not “cure” the hearing impairment, although there was at that time an optimistic vision of the possibility to “improve hearing by training” (the Wedenberg method reported in Ahlsén, 1991). Thus, total limitations in listening activities and restrictions in taking part in sounds remained throughout the whole period. The consequence of the failure of the rehabilitation policy was contrary to the aim; the “facilitating” mechanisms were in reality barriers to participation.

The ‘hush-hush’ policy

Visual acuity impairment was diagnosed when Olga was six years old and when Helen and Laura were eight. RP in their eyes, and thereby USH, was diagnosed when Olga was ten, Laura, 12, and Helen, 15 years old. Diagnosis is a potential mechanism, for instance, in the possibilities for cure, decisions regarding rehabilitation, and support in decisions made by the individuals as to their choices regarding work, future family, etc. However, the informants were not informed about the diagnosis, either by the health care providers or by their parents. This was the policy of the time, considered to be in line with what was best for the child: a facilitating mechanism. In reality, this ‘hush-hush’ policy of the health-care system acted as an environmental barrier mechanism at the societal level, and the parents’ ‘hush-hush’ a social attitude barrier. Both mechanisms restricted participation in the informant’s own self-care and their opportunities to plan their futures. Furthermore, it has not been found that diagnosis and thus prognosis were used as a potential for special rehabilitation, education, etc. Rehabilitation and services in a broad sense continued as before, although the health care providers and the parents knew that the children had USH I.

Sign language

Children with deafness have since 1889 been obliged to attend school (Pärsson, 1997). The compulsory schools for the deaf in Sweden are managed by the State, each covering a certain region, and are boarding schools. The three informants attended schools for the deaf and all went to the national secondary school for the deaf situated in one town. The opportunity to go to school is a facilitating environmental mechanism, as was the policy to create education for students with this particular special need as a result of hearing impairment.

The education policy, an environmental factor, during the years Helen, Laura and Olga were educated was to teach students to talk and to lip-read what other people said (Pärsson, 1997).
Informants were, as are most children with USH I, born into families with no experience of deafness. Neither the parents nor the children were taught sign language. In general, in education our informants were forbidden to sign during lessons and school break times. Olga reports:

*When I was a child there was nobody in the neighbourhood who could sign; we pointed. When I began school it was a tremendously tough time, but I learned it fast. We kids signed in recess of course and I snatched a sign here and there.*

These experiences indicate a communication barrier mechanism at the social level: in the family. However, our informants were introduced to sign language by other children, playmates at the deaf school. This was a facilitating mechanism for participation in two ways: firstly, it brought them into a language appropriate to their skills and, secondly, they became included into a deaf community.

There are indications that this process also included the relatives. When the informants were between approximately 15 and 25 years old, there are notes in AD and OD records to the effect that the informants’ mother’s interpreted using sign language. To accomplish this, their mothers must have learned signing, an extremely important facilitating environmental mechanism.

**Tactile sign language**

In OD records it was recorded that Helen’s mother had to sign at certain distances, standing increasingly farther away from Helen as time and USH progressed. The first observation made about this factor came from Helen at 21 years old. Helen’s visual field impairment became a significant mechanism with a negative impact on Helen’s capacity to practise sign language. Helen’s visual acuity impairment aggravated more rapidly than did the two other informants’ such that at about 25 years of age her visual acuity impairment was down to near total. This had an extremely important consequence: she later needed to receive sign language in tactile mode. Her skill in receiving sign language in the usual visual mode was no longer useful, thus ordinary sign language interpreting was no longer a facilitating mechanism. From now on both Helen and her interpreter, or anyone who wished to communicate with Helen, needed skills in the tactile modality of sign language. This dynamic process shows how one extremely important facilitating mechanism may suddenly disappear due to the reduction of
the “body” mechanism - vision - and has to be replaced by another mechanism, tactile sign language. However, it requires considerable resources to develop this facilitating mechanism.

The example below, told by Helen, illustrates the dynamics and the need for a life-course perspective, demonstrating the complexity of the interplay among a number of mechanisms:

> When I was a child my family often met our relatives. I could lip-read so there were no problems. At the age of 25, I lost my vision and I could not lip-read any more and I have not met my cousins since then. Some years ago I got a skilled assistant. I wrote to my cousins that we could meet each other again. My assistant can interpret. My cousins wrote back that they didn’t want strangers present when we meet.

Helen’s relationship with her relatives, when seen as a social process, is firstly influenced by her hearing impairment. However, her experience was that she had no restrictions in participation with her cousins. Helen’s limitation in hearing what they say was thus compensated by her skills in lip-reading, a counter-acting mechanism, overriding the hearing impairment mechanism. However, in her twenties, Helen’s visual impairment worsened. She maintained her skill in lip-reading, yet this became passive since the visual impairment developed into the stronger mechanism. Helen was later facilitated with an assistant, a societal mechanism, skilled in tactile sign language, a social mechanism. This facilitating mechanism was simultaneously overruled by another mechanism, the cousins’ attitude (an environmental mechanism) towards having an outsider involved in their communication. Additionally, the cousins never learned sign language, constituting the absence of a facilitating mechanism.

**Service provision**

Helen was provided with an assistant when she was about 30 years old. Laura, who lived in another municipality, was denied the same service; she was instead offered other services, usually designed for the elderly, although she was only in her thirties. The third informant, Olga, who lived in a third place, was denied both kinds of services. Although Helen had worse visual acuity impairment of <0.05, the other two had visual field impairment equal to remnant blindness of <10° of visual field when they applied for services. Olga reported in the latest interview about her experiences when she applied for personal assistance:

> They asked me if I could take a shower, if I can dress, if I can eat. I said yes. Then you can’t get an assistant, they told me.
She further explained to the service providers that it was impossible for her to participate in basic household tasks. During the interviews, Olga chanced to mention causing a small flood, and another time a fire. She had turned on the water, then the stove, before being distracted; she had forgotten them, failing to notice the results until these became obvious and another family member dealt with the repercussions. Given these examples, she was deemed unable to perform household work with confidence or without a high level of risk. Furthermore, such tasks are for her energy- and time-consuming.

This example illustrates another type of important mechanism: the knowledge, attitudes and behaviours exhibited by the service providers.

**Changing attitudes**

Adults with deafness in the deaf community in Sweden have changed their policy and attitudes towards deafness during the informants’ life-course. During the period when the informants were children, Fredäng (2003) writes that the deaf community emphasised the audiological hearing threshold level perceived to constitute the border between those with deafness and those with hearing. Deafness was stigmatized by society; the deaf as a group was also subordinate to these concepts. This attitude was a negative societal mechanism. The period when the informants attended the regional schools for the deaf was a time of transition, whereas at the beginning of the period the deaf community claimed that signing should be valued since it facilitated linguistic development. However, this attitude was also a barrier since it failed to recognize sign language as equal to spoken language. Some years later, when the informants were receiving secondary education at the National School for the Deaf, the deaf community had changed its attitude and strongly claimed its right to sign language. The deaf emphasized their abilities especially as expressed in sign language, together with a rejection of the notion that to force children with deafness into acting as if they were hearing created a positive deaf identity. Sign language was recognized as the first language of deaf people only in 1981. This marked a radical shift in society. The emphasis on abilities is in contrast to the previously accepted stigmatization of the deaf as helpless and not normal, i.e., handicapped (Fredäng, 2003). The ‘new’ attitude was regarded as facilitating people with deafness in general; the advocacy policy was successful, thus the possibilities for their participation in society increased. When in the interviews Helen tells about her childhood, she expresses an attitude indicating that hearing is the norm in a normative sense. Her perception is different from what Laura and Olga relate; they emphasize weak communication before
their introduction to sign language by playmates in school, which for them was a breakthrough in communication. They thereby express a ‘deaf ability’ attitude. Laura and Olga consider themselves to be deaf. To be deaf is not to be disabled. However, having a visual impairment is to have a disabling impairment. Their phrase *I am deaf with visual impairment* should be interpreted in this way. The visual impairment was emotionally loaded, since visual impairment is to lack ability. The informants were provided with a white stick by their Low Vision clinic; it was considered to be an assistive device. However, Laura and Olga refused to use them and for many years kept their sticks in their handbags. The hush-hush policy was an attitudinal societal reinforcement further contributing to difficulties in “coming out” as a person with visual impairment.

**Performing activities**

All informants report that they learned to ride bicycles when they were children and frequently went cycling. In the neighbourhood where the informants lived it was very common for adults to cycle to work and in leisure activities.

Olga reports that when she was a child she was active in skiing, slalom, and climbing mountains in the countryside, interested in playing handball and football, but from the age of about 15 these became highly problematic. Helen stopped playing tennis, which she used to enjoy, when she was about 12. These problems were related to a decreasing visual field, which in turn was related to severe deviation in the retinas. Olga’s visual field impairment had an impact on her activities and thereby also on her participation in leisure activities.

Laura is the one who first got complete scotoma, i.e., tunnel vision (tunnel vision with a visual field of less than \(<10^\circ\) is regarded as blindness.) Laura mentioned:

> It was when I was around 17–18 years old that I stopped cycling. It was due to an accident. It was because an old man crossed the street at a crossing. I didn’t see him and ran into him. He wasn’t hurt, but he got very angry. It was awful. I understood that I had to stop cycling.

Laura’s decision to give up cycling was motivated only by limitations to her sight. Laura did not mention as a reason her profound deafness, which may be explained by the fact that her hearing impairment was congenital. She had never had any experience of hearing, a personal factor. The incident also had a strong impact on her personal security. Insecurity, a personal emotion, as an effect of unpredictable events, was also found in Helen’s and Olga’s narr-
The emotions of insecurity and unpredictability appear to be two very strong mechanisms that limit activity.

The complexity

In the two first interviews, Laura and Olga, at that time not yet 40 years old, read the usual texts in books and newspapers. At the third interview Laura told how she coped when she read the daily paper:

I have to have correct light and the right reading glasses [in the records, the LVC has written that she has four pairs of them, each focusing to a different distance]. If it is too light outside I can put the paper into another position. I do always have to think about how to adapt. It is not possible to just plonk down the paper and spontaneously start to read as others do. I have to think about where to sit. If it doesn’t work I have to move. It also depends on the weather outside, if I have caught a cold, if I am in a rush, in those cases it is even worse. If I’m going to read a book, I often think, I cannot manage now. I can’t read the small letters in the phone book, for instance; the print is too small. I use a magnifying glass, but it is a strain on the eyes. Sometimes I think I can’t stand reading.

This quotation shows that ability to perform the activity of reading is dependent on many mechanisms, both external and internal.

Lack of general knowledge

Additionally to the problems mentioned above, there is a problem with a lack of general knowledge. Both Olga and Laura give many examples of how they often face difficulties in understanding official language in letters from the service providers. Laura says, I can’t use such smart words that signify many things in one word. This problem is also conspicuous when Laura interacts with professionals. Laura says it is difficult for them to detect that my knowledge isn’t good enough because I have interpreters with me. It is the knowledge of the interpreters the doctor hears, not mine. Laura tells about an occasion when she visited a public authority about an important matter.
I didn’t understand what they meant. I had to ask the interpreter to explain. It was so embarrassing.

Lacking good general knowledge is a personal mechanism that could constitute a serious barrier.
Discussion

In this article we have tried to demonstrate the complexity regarding participation for persons with USH I. Furthermore we have tried to show that the ongoing progression of the syndrome, present and past experiences contribute to ongoing needs of rehabilitation and support. The life-course perspective revealed fundamental changes over time in their environments, where the shift from the oral policy to the introduction of sign language seems to be of the utmost importance. However, a person with USH I has to cope not only with changes in the environment, but also with constant changes in their body structure and bodily functioning. There were for the informants some key points in this process; one was the discovery of the syndrome, and another was the forced shift from sign language to tactile sign language. It became possible to reveal with the aid of ICF and a life-course perspective both the complexity of interactions among mechanisms, and the changing preconditions for participation.

ICF is an integrated bio-psycho-social model; each of the other five components in ICF had a direct or an indirect impact on participation. ICF includes three main levels: the biological, the individual, and the social level. Within each of these levels one can distinguish among other levels, those of body structure and bodily function. Within the body structure level there are also different levels, such as protein in the mutated gene, haircells in the cochlea and labyrinth, and rod and cone cells in the retina, as parts of ear, vestibular organ and eyes, relevant in research into USH I. Furthermore, a distinction is made between bodily functions: hearing, balance, and vision, and their activities, which are to listen, maintain balance, and seeing.

An activity is something performed both at the individual level, including the person who performs it, and at social level since each person is an element within a societal environment. Others who interacted with the informants were thereby also enabled to analyze their observations at both individual and social levels. Skills, for instance, were entities attributed both to the environment and to the informants, i.e., a personal factor. The authors found that legislation and policy were societal mechanisms with a significant impact on the rehabilitation and education of the informants. These mechanisms had, furthermore, an impact on the behaviour of the parents: whether or not they learned sign language; whether or not they informed their children of the diagnosis, and hence the complex question of planning the children’s respective futures. To summarise, the five levels applied were: body structure; bodily function; individual, including activity and personal factors; social, including participation and environ-
ental factors such as attitudes and relations, and finally, societal, including environmental factors such as legislation, policy, and services.

We found great variation among mechanisms regarding duration and degree. The mechanism with the longest duration was hearing impairment. It was reported from about one year of age (for Helen and Laura) and three (for Olga), throughout nearly fifty years. Hearing impairment created by body structure mechanisms was the determinant for total listening limitation, for language acquisition, and for rehabilitation and education. Hearing impairment was total; it neither varied in degree nor was influenced for the better or worse by other factors. Limitation in hearing was dependent upon hearing impairment and thus had the same duration and degree. However, the hearing limitation had different meanings for each of the informants and these changed over time, being contextually determined. Limitations in hearing did not always prevent participation in communication since this could be influenced by environmental and personal factors. Facilitating environmental factors included interpretation, while personal factors included skills in lip-reading and in sign language.

Visual impairment was a mechanism with a shorter duration. It was reported from six to 12 years of age; it aggravated over time. Limitation in seeing was dependent upon visual impairment, created by body structure deviations. Seeing was influenced also by other factors: environmental factors with a facilitating impact on seeing included the light source, spectacles, and other assistive devices. Personal factors could also play a positive role, for instance, the use of assistive devices and placing oneself in a certain location, which Laura did in order to promote her seeing. Limitations in seeing did not always prevent any taking part in visual features, but the limitations increased over time. Limitations in seeing have also had an impact on the subjects’ opportunities to take part in audible features, e.g., the lip-reading aspect of hearing.

The combination of limitations in audible and visual features was a mechanism of its own that had an impact both on activities and on participation. The combined mechanisms varied in degree of impact and created other mechanisms, such as insecurity.

We found that the absence of an element could function as a mechanism, e.g., the cousins’ lack of skills in sign language.

Some mechanisms were at a given time recognized as facilitating, yet they in fact played the opposite role. This became obvious when the mechanisms were studied over time from the life-course perspective. The oral policy is an example of such a mechanism. Furthermore, there were often counter-acting mechanisms that were stronger than the initial mechanism. An illustration of this was the facilitating services of an interpreter provided to one of the infor-
nants in order for her to be able to communicate with her cousins. However, the cousins’ negative attitude towards involving a stranger in their interaction was a stronger counteracting mechanism, thus the interaction between the informant and her cousins never came about.

Location in time, i.e., starting education in the oral period then attending school in the transition period towards the acceptance of sign language as the first mode of communication, was important. Location in space was also significant. For instance, the respective regions in which the informants lived played a major part in whether or not they received services. Another aspect of space is being forced to perform an activity only in one certain place, e.g., reading the paper only in one seat in the apartment due to the lighting conditions.

Our study shows that even though the three informants have the same diagnosis and type, the individual variations in bodily functions are considerable. Furthermore, their attitudes toward themselves and their impairments similarly vary; the causes for this are found in biological, social, and societal mechanisms.

In order to provide efficient services to these persons, many complex mechanisms have to be taken into account. It is especially important to note different and divergent opinions among professionals and patients/students/clients since their respective approaches to their life situations are based in different frames of reference. In order to support the person it is necessary first to determine and to understand the mechanisms and the context forming the foundations of their attitudes and opinions.
Conclusions

ICF and the life-course perspective in the analysis of participation for people with USH I have brought about an understanding of how a mechanism recognized as a facilitator constituted, in reality, a barrier. It has also illustrated how a facilitating mechanism shifted over a short period of time from being a facilitator to a barrier. It has revealed how counter-acting negative mechanisms could override facilitating mechanisms. We regard ICF as a useful model of human functioning and disability in rehabilitation (Stucki, 2007a, 2007b). An advantage with ICF is the bio-psycho-social integrative approach. All components of ICF are potential impact factors regarding rehabilitation and support. Activity and participation signify different aspects of functioning. An analytical division into separate components is therefore necessary. We found that ICF supplemented by a life-course perspective was fruitful, since past, present and future features as well as spatial features play an important role. Rehabilitation based on ICF, and supplemented with the life-course perspective, should be encouraged and further developed.
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