

## Deafblindness and dual sensory loss research: Current status and future directions

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### Abstract

Deafblindness is more than the addition of hearing impairment plus vision impairment. The absence or impairment of both distance senses gives a condition which is more disabling than the sum of each. Deafblindness is rare among young people but becomes frequent at higher ages. Deafblindness can be either congenital or acquired. The heterogeneity of the population has been reported to be huge. Different levels of vision and hearing loss, different use of language modality, different kinds and severity of additional disabilities, and different medical aetiology are some of the variables splitting the group. Research

in deafblindness is still in it is advent due to a number of limitations and a lack of current scientific interest. Some of the challenges in deafblindness research are: lack of consensus on the definition of deafblindness; rareness of the condition which makes it difficult to even gather just a small group to study; heterogeneity of the population; difficulties with using traditional functional assessment procedures; communication barriers; and the difficulties of interpretation of deafblind behavior. This editorial calls for more interest in deafblindness in general and for more international cooperation and innovative studies to overcome existing barriers: Cooperation on data collection to form big enough sample sizes; development of reliable and valid tests and assessment tools; development of new research methods and approaches.

**Key words:** Deafblindness; Dual sensory loss; Dual sensory impairment; Definition; Methodology

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**Core tip:** Research in deafblindness may have the potential to generate knowledge about several basic questions: How sensory loss affects human development such as mental wellbeing, language development, and cognition. The nature of tactile language and tactile perception. How medical genetics are linked to combined vision and hearing loss. However, research in deafblindness is still in it is advent due to a number of limitations and a lack of current scientific interest. This editorial calls for more interest in deafblindness in general and for more international cooperation and innovative studies to overcome existing barriers.

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## DEAFBLINDNESS

Deafblindness - otherwise known as dual sensory loss - refers to the combination of vision and hearing impairment. Deafblindness is often researched as two different subgroups - those with congenital vision and hearing impairment and those with acquired dual sensory impairment - because of their different developmental conditions. People with congenital deafblindness have to develop language despite being deaf and blind, while people with acquired deafblindness have to maintain language abilities when becoming deafblind.

Scientific interests in deafblindness have been present throughout history. The scientific interest in deafblindness can in one point be dated back to around 1860 with the case-story of Helen Keller and her teacher Ann Sullivan<sup>[1]</sup>, which shed light on the uniqueness and challenges in being deafblind. Deafblindness is more than the addition of hearing impairment plus vision impairment. The absence or impairment of both distance senses gives a condition which is more disabling than the sum of each. The equation  $1 + 1 = 3$  illustrates the situation where vision cannot compensate for loss of vision and vice versa. Therefore, people with residual hearing and/or vision are also labeled as deafblind.

Heterogeneity of aetiologies portrays the populations of congenital deafblindness. More than 30 causes of congenital (pre-lingual) deafblindness have been identified in a recent population study<sup>[2]</sup>. The large number of different aetiologies includes intoxications and infections such as Rubella virus or Cytomegalovirus infection of the fetus or meningitis infection postnatal. A large number of genetic and chromosomal disorders involve vision and hearing impairment such as CHARGE syndrome which is one of the most frequent congenital conditions. CHARGE syndrome is a genetic disorder characterized by coloboma of the eye, heart defects, atresia of the nasal choanae, retardation of growth and/or development, genital and/or urinary abnormalities, as well as ear abnormalities and deafness<sup>[3]</sup>.

Several causes are also known for acquired (post-lingual) deafblindness with Usher syndrome being the most dominant (about half of all cases) among people below 60 years of age<sup>[2]</sup>. Usher syndrome is an autosomal recessive disorder characterized by congenital or progressive hearing loss and progressive vision impairment due to the eye disease Retinitis Pigmentosa and involve several subtypes<sup>[4]</sup>. Other causes are head injuries, tumors, blindness following diabetes, as well as hearing impairment caused by noise exposure. In old age, the major causes to vision impairment are age-related macular degeneration, cataracts, and glaucoma<sup>[5]</sup>. Hearing impairment in old

age are often attributed to presbycusis (age-related sensorineural hearing loss)<sup>[6]</sup>.

Research in the field of acquired deafblindness has mainly been concerned with mental health and life outcome consequences among the elderly. Some of the reported findings have been, that acquired deafblindness is associated with a much higher prevalence of depression, cognitive decline, and difficulties with activities of daily living (for a review see Dammeyer<sup>[7]</sup> 2014).

Research themes with respect to congenital deafblindness have mainly concerned the study of how language and communicative development can be supported by using pre-lingual tactile support. It has, for instance, been explored how the caretaker can sustain and expand the social interaction by responding to the child's expressions of tempo, rhythm, intensity, and emotions within the tactile modality<sup>[8]</sup>. These studies are mostly case-based studies with a focus on qualifying intervention methods (for a review see Dammeyer<sup>[7]</sup> 2014).

A number of other research fields are related to people with dual sensory impairment but do not take an explicit focus on deafblindness. Examples are medical research in syndromes which frequently involve dual sensory losses such as Usher Syndrome, CHARGE Syndrome, and Maternal Rubella Syndrome. For instance, one research topic has been the behavioural phenotype in children with CHARGE syndrome<sup>[9]</sup>.

Despite the uniqueness and interest in deafblindness a contradiction exists in scientific research. The number of published research reports with regard to deafblindness is sparse. A search in Pubmed on the terms deafblindness, dual sensory loss, or dual sensory impairment reveals less than 150 hits and the term deafblindness reveals less than 500 hits (November 2014). Most of this research concerns people with acquired deafblindness or specific medical syndromes. Congenital deafblindness has rarely been scientifically explored or reported on. The lack of published research may have at least seven explanations.

First, the rareness of the condition makes it difficult to even gather just a small group to study. Prevalence of congenital deafblindness and acquired deafblindness among people below 60 years of age is below the 0.1 percent level. Acquired deafblindness is more frequent at higher ages (prevalence around 30 percent at age 80) (for a review see Dammeyer<sup>[7]</sup> 2014), and may therefore be more researched.

Second, the heterogeneity of the population makes it difficult to study people with deafblindness as one single group. Different levels of vision and hearing loss, different use of language modality, different kinds and severity of additional disabilities, and different medical aetiology are some of the variables splitting the group. With regard to additional disabilities mental retardation and developmental disorders have been reported as the most frequent in the congenital deafblind group<sup>[10]</sup>.

Third, due to the dual sensory loss, it is generally very difficult to use traditional functional assessment procedures and psychological tests, since these often require full sensory functioning as a prerequisite. Several researchers have concluded that one cannot rely on visual or auditory test items and interpretation of results from standardized norms is dubious, at best, for cognitive, language, and social assessment<sup>[11,12]</sup>.

Forth, it is often difficult to communicate or even cooperate with a person with congenital deafblindness even for a researcher mastering both the local oral, signed, and tactile languages. Communication form and abilities may be very individual to each person. A mix of oral, visual, tactile and alternative communication systems is often used. Similar communicative challenges, but to a lesser degree, are also the case for individuals with acquired deafblindness.

Fifth, interpretation of "deafblind behaviour" can be very challenging. The communication and behaviour of a person with congenital deafblindness must be understood from the "tactile-bodily being in the world"<sup>[13]</sup>. A congenitally deafblind man who hits his head with his hand may be exemplifying a symptom of a mental or behavioural disorder (for example psychosis, anxiety, frustration), or it may be a means of self-stimulation, or communication (telling the caretaker that he needs help, is hungry or misses someone). The question of validity when interpreting deafblind behaviour is challenging but vital to take into consideration in all deafblind research.

Sixth, deafblind research has for the most part been a sub-discipline to deaf education, service and research. In education and rehabilitation the same methods as used among people with a hearing impairment are applied, just adapted for tactile modality. Often tactile adapted visual sign-language has been used. This may lead to "blindness" for the deafblind person's tactile perception which shapes the communication and mind differently compared to people with hearing loss.

Seventh, last, but not least, there is no consensus on the definition of deafblindness in the literature. Overall two types of definitions are employed; a medical definition in terms of audiological and visual criteria, and a functional definition based on self-report and observation, evaluating the individual impact of vision and hearing loss on everyday life activities and the individual's possibilities for participation in society. The lack of a clear definition in research and practice makes it difficult to compare research results across studies<sup>[14]</sup>.

To overcome these barriers in deafblind research, which cover issues related to definition, methodology, theory, and organization of research, a number of different initiatives need to take place. Some of these may be: (1) The individual researcher needs to use a more thorough definition and description of the study population by using well defined terminology and criteria, building on existing research; (2) Development of standardized tests, questionnaires, and other

assessment tools for individuals with deafblindness is important. For example it is important to develop tests which can differentiate symptoms of autism disorder from symptoms related to consequences of dual sensory impairment<sup>[15]</sup>; (3) Many studies use video observation of tactile behaviour and communication. Studying one modality (tactile) with another modality (visual) is not optimal. The methodological challenge is clear; with reliable methods to measure tactile behaviour more directly and with more validity. New technology has to be applied and developed such as automatic motion tracking technology using infrared markers, neuroimaging methods such as event related potentials, and technology measuring touch; and (4) Because of the different methodological challenges the rareness and heterogeneity of deafblindness, an increased cooperation among international research groups is greatly needed to solve the critical challenges and limitations in deafblind research. Cooperation on data collection to form big enough sample sizes may be one aim. Often many variables have to be controlled, such as medical aetiology, cognitive abilities, and communication mode, which is impossible in small sample sizes.

## IN SUMMARY

Research in deafblindness may have the potential to generate knowledge about several basic questions: How sensory loss affects human development such as mental wellbeing, language development, and cognition. The nature of tactile language and tactile perception. How medical genetics are linked to combined vision and hearing loss. However, research in deafblindness is still in its advent due to a number of limitations and a lack of current scientific interest. This editorial calls for more interest in deafblindness in general and for more international cooperation and innovative studies to overcome existing barriers.

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