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Review article

Definitions of deafblindness and congenital deafblindness

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ABSTRACT

In order to compile knowledge on deafblindness (DB) and congenital deafblindness (CDB), one important factor is comparison of results between different scientific studies. In an attempt to do a systematic review of the literature on cognitive assessment and CDB, considerable difficulties in determining eligibility of the studies were encountered due to heterogeneity in definitions and inclusion criteria used in the articles. The present systematic review aims to provide both an overview of this terminological and methodological heterogeneity and suggestions for better future research practices. A systematic review of definitions used in ($N = 30$) studies employing psychological assessment of people with CDB served as a sample of the scientific literature on DB and CDB. Absent or heterogeneous definitions and inclusion criteria regarding both DB and CDB are evident in the sample. Fifty percent of the studies reported no definition of DB and 76.7% reported no definition of CDB. Main discrepancies are: (1) medical/functional versus ability/functioning definitions regarding DB; and (2) different criteria for onset of DB in the case of defining CDB (e.g. age versus developmental level). The results of this study call attention to a scientifically inadequate approach to the study of DB and CDB. Findings indicate that clear guidelines for sample descriptions of the DB and/or CDB populations are needed. It is suggested that studies including DB and CDB participants provide the following information: definitions of DB and CDB used; severity of sensory impairments; level of sensory ability in relation to mobility, access to information, and communication; age at onset of DB; and communication as well as language ability at onset of DB.

Contents

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1. Introduction

The scientific fields of deafblindness (DB) and congenital deafblindness (CDB) are presently young and relatively incoherent. The few researchers that venture upon the study of DB in general and CDB in particular are still trying to overcome some important fundamental challenges regarding epistemology and methodology. Recently, researchers have addressed terminological issues, such as how to label the condition referred to as *deafblindness*, *dual sensory impairment*, *combined vision and hearing loss*, and the like (Wittich, Southall, Sikora, Watanabe, & Gagné, 2013). The present study adds to this discussion, by addressing the epistemological differences underlying the terminology and the connected methodological problems of sample delimitation and sample description.

The issue of defining DB has been addressed by others in the literature. According to Saunders and Echt (2007), a clear definition of dual sensory loss in general (i.e. DB) is lacking. This lack of clarity is supported by discrepancies in terminology and inclusion practice. One discrepancy regarding definitions of DB is between those based on sensory impairment measures and those based on the resulting functioning. The former correspond roughly to the legal definitions of deafness and blindness in the United States and take the medical/functional degree of the two separate sensory impairments as the defining factors, adhering to international standards of diagnoses (e.g. ICD-10). The latter stress the total outcome of DB on ability and functioning in relation to communication, access to information, and mobility (Dammeyer, 2012) and correspond to the Nordic definition of deafblindness, often labelled "functional" (Danermark & Möller, 2008; Rönnberg, Samuelsson, & Borg, 2002).

There is also a discrepancy in the literature regarding the use of the term 'functional'. Some authors use the term 'functional' in the medical sense, which is comparable to the definition of *Impairment* as damage to structure or function of body parts in the International Classification of Functioning, Disability and Health: ICF (WHO, 2001). In our study, these will be labelled medical/functional. Others use the term in the Nordic sense, in line with the ICF definitions of *Functioning* and *Disability* as the outcome of the interplay between body functions and structure on the one hand, and activity limitations and participation restrictions on the other. These will be labelled ability/functioning. The labels 'medical/functional' and 'ability/functioning' are used here in accordance with the distinction between *Impairment* and *Disability* in the ICF.

Disagreements concerning the differentiation of CDB from acquired deafblindness (ADB) pose another discrepancy in the literature. CDB is DB with early onset. Some authors define CDB as DB with onset before a certain chronological age, e.g. "from birth" (Dammeyer, 2009; Möller, 2003; Prain, McVilly, & Ramcharan, 2012) or "before 2" (Dalby et al., 2009a; Guthrie et al., 2011). Others define it as DB with onset relative to communication development and/or before language acquisition (Dammeyer, 2011; Rødbroe & Janssen, 2006). In the latter case, CDB is sometimes labelled "pre-lingual deafblindness" (Dammeyer, 2011).

Furthermore, the severity of DB is assessed in different ways in the literature. Adhering to a medical/functional definition of DB, Dalby et al. (2009a,2009b) have devised The Deaf-blind Severity Index (DdSI). This index "uses two variables on functional vision and hearing to create a severity score ranging from no impairment in either sense (0) to severe impairment in both senses (6)" (Dalby et al., 2009a, p. 11). Others take the time of onset to be an important indication of severity of DB (e.g. Dammeyer, 2012). The impact of DB on the development of communication and language acquisition is considered more severe for those with an earlier onset of DB.

Finally, some take the medical aetiology of DB, such as congenital syndromes like CHARGE Syndrome, an acronym for Coloboma, Hart anomaly, choanal Atresia, Retarded growth, Genital, and Ear anomalies (Thelin & Fussner, 2005; Vervloed, Hoevenaars-van den Boom, Knoors, van Ravenswaaij, & Admiraal, 2006), or Congenital Rubella Syndrome, CRS, (Armstrong, 2010; Dammeyer, 2010a) to be the defining factor or inclusion criteria in studies of deafblindness.

This leaves two unresolved issues: (1) both DB and CDB are defined in different ways in the scientific literature and, consequently, (2) no consensus regarding inclusion criteria is established. These discrepancies in the field regarding definitions and the methodological heterogeneity make it difficult to compare studies in order to compile knowledge about DB and CDB and the disabling dynamics of the conditions, which in turn hinders scientific progress. The present literature study aims to provide an overview of this terminological and methodological heterogeneity.

In connection with a separate systematic review on psychological assessment in relation to CDB (Ask Larsen & Damen, unpublished results), we encountered the practical problem of how to identify studies that had people with CDB in their samples, and the analyses that are presented here were made to overcome this problem. We will use the sample of reviewed studies from that study as an exemplary case when addressing the above mentioned problems in CDB research.

In summary, at least five parameters influence how DB and CDB are defined in the literature. DB is defined according to (1) level of sensory impairment (medical/functional definitions) or (2) sensory functioning and disability (ability/functioning definitions), whereas CDB is distinguished from ADB according to (3) onset age or (4) onset relative to communication development and language acquisition. In addition, (5) medical aetiology of DB is used both as an indication of DB and, in some cases, as an indication of CDB. We conducted a systematic review in order to examine these parameters in studies on