PERSPECTIVE

Deafblindness in Children: Time to Act in India

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Deafblindness or dual sensory impairment (DSI) is a distinctive disability, which encompasses varying degrees of hearing impairment and low vision. The impact of deafblindness is not merely an addition of the impact of the two disabilities, but it is manifold. Deafblind children have cognitive, speech, motor and social delay. They can have multiple medical needs, which should be identified and met. It is estimated that there could be more than 500,000 deafblind adults and children in India. Doctors, general public, strategists and law makers need to have heightened awareness of its various aspects. It is time India acts for long-term welfare of its deafblind children, including their disability rights and access to a structured health and education system, thus increasing their chance of becoming independent employable adults.

Keywords: *Developmental delay, Disability, Dual sensory impairment*.

Deafblindness is a distinctive disability denoting varying degrees of hearing impairment and low vision. Congenital deafblindness is defined as deafblindness with an onset from birth or before the age of two years [1]. In children, deafblindness causes overall developmental delay affecting cognition, speech and language, motor and social delay. This condition being unique, is a challenge to educate such children in single disability schools. A deafblind child has additional medical needs and at times complex neurological involvement. Such children are unable to compensate for loss of one sense with the other [2,3]. Deafblindness is included in International Classification of Diseases (ICD) 10 classification [4] and in International Classification of Functioning, Disability and Health [5]. Deafblindness occurring as a part of syndromes is included in ICD 11 classification [6].

DIAGNOSTIC CRITERIA

World Health Organization (WHO) classifies deafness into four categories: mild, moderate, severe and profound hearing loss [7]. With mild hearing loss of 26 to 40 decibel (dB), a child will have trouble hearing and understanding soft speech or sound coming from a distance with or without background noise. A child with moderate hearing loss of 41 to 60 dB will have trouble hearing and understanding conversation level at close distance. With severe hearing loss of 61 to 80 dB, a child will hear only very loud speech or loud environmental sounds, *i.e.*, sirens of fire engines or doors slamming. A child with profound hearing loss of more than 81 dB will perceive only loud sounds as vibrations. WHO defines blindness by including visual acuity and excludes field of vision. Better corrected visual acuity (BCVA) better than 3/60, but less than 6/60 in the better eye, and field of vision of ≤10 degree in the better eye is graded as blindness in India. WHO grades this as low vision. In India, low vision is defined as BCVA better than 6/60, but less than 6/18 in better eye [9,10].

PREVALENCE AND ETIOLOGY

In India, there are no organized data about deafblindness, and estimates are based on community projects. It is estimated that there could be more than 500,000 deafblind people in India [10]. In a study conducted in Nigerian single-disability schools, of the 273 students examined, 19 (7%) had deafblindness of which over 60% (12/19) were previously undetected [11]. Buphthalmos, cataract, vitamin A deficiency, optic atrophy, anophthalmos/ microphthalmos and cortical blindness were the causes of blindness noted. Similarly, a study by three agencies providing integrated deafblind services in Montreal, Canada reported a prevalence for deafblindness at 15/ 100 000 with 5.7% for under 18 years of age [12]. Causes of deafblindness are listed in ***Box* 1.** In a series of 190 people (127 adults and 63 children) with deafblindness, rubella syndrome (28%) and Down syndrome (8%) were the largest group among those over 17 years of age; whereas among children, CHARGE association (16%, *n*=13) was

BOX I ETIOLOGY OF DEAFBLINDNESS IN CHILDREN

Prenatal causes

Rubella, Cytomegalovirus, Toxoplasma, Herpes, HIV, Fetal alcohol syndrome, Hydrocephalus

Perinatal causes

Asphyxia, Prematurity and Small for gestational age, Hyperbilirubinemia

Postnatal causes

Meningitis, Encephalitis, Stroke, Head injury, Metabolic disorders

Genetic syndromes

CHARGE association, Ushers syndrome, Down syndrome, Goldenhar syndrome, Stickler syndrome, Alstorm syndrome, Alport syndrome, Apert syndrome, Cockayne syndrome, Crouzon syndrome, Cornelia de Lange syndrome, Norrie disease, Pierre Robin syndrome, Sturge Weber syndrome, Waardenburg syndrome, Turner syndrome

CHARGE: Coloboma, Heart defects, Atresia choanae, Growth retardation, Genital abnormalities, and Ear abnormalities

the largest group [13]. Prevalence among children and adults was 1:19,000 and 1:34,000, respectively.

IMPACT OF DEAFBLINDNESS

Hearing and vision are our distant senses, and smell, taste and touch are the near ones. Young children learn through visual and auditory input. About 80% of learning happens through our distant senses. Initiation of motor patterns and conceptual development starts early in life by motivation, and enhanced by sounds that are heard. Speech, language and social expectations are all learnt through listening and watching. Movements bring the child in contact with people and objects. If the visual input is poor, motor development is likely to lag, and learning social body language and gestures also becomes a challenge. Poor motor development has an adverse effect on cognition, as the child is unable to get around and explore. When locomotion starts, incidental learning occurs in infants through exploration of environment, which happens effortlessly. Secondary learning occurs by listening. Direct learning occurs by hands on experience. In deafblind children, majority of the learning is direct – by use of touch. Secondary learning becomes difficult and incidental learning usually does not occur as the inputs from the auditory and visual system are inadequate.

The severity of speech and language delay depends on the severity of hearing loss. Communication is essential for cognitive development, and any associated adverse factors can impair cognition. The distant senses connect infants to the world and beyond their personal body space. Therefore, a deafblind child will be unable to read sign language from a distance, thus limiting opportunities, and is instead reliant on caregivers to access, interpret and organize information. They need to be in close proximity to their parents, siblings, caregivers or teachers; if not, they feel isolated.

Object permanence is the ability to know that objects do continue to exist, even if they cannot be seen or heard. It is harder to establish in deafblind children as they are unable to hold the image of an object they are unable to see. This ability is related to naming and categorizing objects, which links conceptual and speech development, thus affecting development of cognition and communication.

Developmental delay due to congenital infections, genetic syndromes, prematurity and infections occurring in early life carry a higher risk of causing deafblindness associated with complex needs. Approximately 90% have one or more health related problems, 75% have two or more, and 50% have three or more [14]. A study indicated that 66% had cognitive disability, 57% physical disability, 38% had complex health needs, 9% behavior challenges and 30% other difficulties [15].

CHALLENGES IN EVALUATION

A combination of limited incidental learning and complex neurology makes intellectual testing a challenge in children with deafblidness. This evaluation can be performed only by highly specialized psychologists and educators/teachers. There are only a few specific developmental assessment tests for deafblind children. Most formal psychometric tests and behavioral tests are not applicable for these children. INSITE scale is a developmental assessment skill designed for young children with multisensory impairment. The Callier Azusa scale is a comprehensive developmental scale, designed to measure the progress of deafblind children specifically. This scale includes motor development, perceptual abilities, daily living skills, language development and socialization [16].

MANAGEMENT ISSUES

Multidisciplinary inputs can be achieved in the following stages: (*i*) early identification can be enabled by the education of parents and health practitioners; (*ii*) prompt referral to the nearest community rehabilitation center; (*iii*) early assessment and intervention by identification of high-risk children, providing support and intervention facilities through assistance with home aids and center-based services and therapies, and providing counseling for families; (*iv*) individualized education plans to

monitor progress; and (*v*) continued review and planning for further education.

Caregivers must provide the child access to a world beyond the limited reach of eyes, ears, and fingertips. A conversation can begin with an adult who simply notices what the child is paying attention to, and finds a way to let the child know that their interest is shared. Once this shared interest is established, it can be built, which can increase the possibility for a conversational interaction. Conversations can be continued by pausing the interaction sequence to allow time for response, as these children respond slowly. Respecting the child's own timing is crucial to establishing successful interactions. Often it is helpful to accompany introduction of spoken or signed words with simple gestures and/or objects that serve as symbols for activities. Doing so may help the child develop anticipation. A deafblind child needs as much language stimulation as any other child. Meaningful communication and education methods are: touch cues, gestures, object or picture symbols, sign language, finger spelling, Braille writing and reading, Tadoma method of speech reading, large print reading, and lip reading speech.

Technology can tackle the challenges of deafblindness and enable communication. The development of assistive devices for communication is crucial. This should happen in both national and indigenous languages, which is a crucial area of research. Hearing aids (or cochlear implants, where indicated, for severe to profound hearing loss) should be provided where possible as they facilitate communication and learning. Telescopic spectacles and light emitting diode illuminated magnifiers can fully exploit visual potential. Audiobooks and electronic communication boards can help children harness listening and reading skills. Braille books can boost effective tactile learning. In addition, motor development should be assisted with tools such as mobility canes.

PREVENTION

The advocated primary prevention measures are: (*i*) immunization against rubella to prevent congenital rubella syndrome; (*ii*) education of all women of childbearing age about healthy habits and avoiding alcohol and drug use; and (*iii*) accessible antenatal health care for all pregnant women. Secondary prevention entails symptom awareness, and treating and managing various causes of deafblindness by training and education of primary care workers, medical students and relevant specialists for early diagnosis. Tertiary prevention to soften impact of illness involves appropriate support at schools for the deafblind, providing them with technological knowledge, and vocational rehabilitation.

GOVERNMENT'S ROLE

India abides with United Nations convention on 'persons with disabilities.' Deafblindness has been included as one of the multiple disabilities under The Rights for Persons with Disabilities Act of 2016. This is a definite step forward in uplifting care of deafblind children and adults. However, a lot more needs to be done. Deafblind children and adults continue to be discriminated against and denied education as a basic human right. Distinct steps need to be taken by the government in order that deafblind children and adults do not face this basic discrimination. Given that the numbers of deafblind people are only estimated, and as a means of enforcing the Rights for Persons with Disabilities Act 2016, it is time strategists and politicians consider adding this disability as a separate category in census 2021. Education of medical officers from primary care centers, auxiliary nurse midwives, anganwadi workers and teachers will help towards early diagnosis. Deafblindness as a preventive and social medicine topic could be introduced in the medical curriculum for graduates undergoing medical training. This could be rolled out by the health ministry to the syllabus coordinators across the upcoming as well as existing universities in India. This topic should also form part of curriculum for postgraduate courses in Pediatrics, Ophthalmology and Otolaryngology. Right to early diagnosis as a part of Rashtriya Bal Swasthya Karyakram (RBSK) will be a key to early intervention and rehabilitation. In RBSK, deafness and blindness exist as separate entities in categories such as defects at birth, diseases of childhood including developmental delay and disabilities. It is essential that education should convey that the two disabilities can exist together.

The Right of Children to Free and Compulsory Education Act (2009) directs free compulsory education for all children from 6-14 years. Sarva Shiksha Abhiyan (SSA) is the main vehicle for execution of Rights to Education (RTE) Act. One of the important components of SSA is inclusive education of children with special needs. All deafblind children should be included under this category.

CONCLUSION

Deafblindness diagnosed before two years of age poses unique management challenges. Strategists and politicians need a strong will and an understanding to consider deafblindness as a distinct category in the national census and survey counts. This along with preventive strategies, require strong leadership, perseverance, will and sustained input from government.

Funding: None; *Competing Interests*: None stated.

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