Visual performance and ocular abnormalities in deaf children and young adults: a literature review

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Abstract

Visual defects are common in deaf individuals. Refractive error and ocular motor abnormalities are frequently reported, with hyperopia, myopia, astigmatism and anomalies of binocular vision all showing a greater prevalence in deaf individuals compared with the general population. Less is known about near visual function in deaf individuals and it appears to have been relatively neglected in the literature to date. Comparisons between studies are problematic due to differences in methodology and population characteristics. Increased understanding of visual defects emphasises the need to improve screening and treatments for deaf children, particularly because any untreated defect has the potential to impair the development of language, with consequences for education more generally.

Key words Deaf, vision, reading, refraction, binocular vision.

Overview

Deaf people are thought to view the visual world very differently from people with normal hearing, due to adaptation to their hearing loss and consequential changes to their communication strategy. For example, deaf people who use sign language must be able to discriminate quickly between facial expressions in order to interpret signed sentences. In a large study of hearing impaired students, over a quarter were found to have visual defects, the most common being refractive error (Gogate et al. 2009). Therefore, assessment and treatment of visual defects, especially refractive errors and binocular vision anomalies, are essential to allow the best possible social and professional adjustment for deaf individuals. In the current paper, we review the literature concerned with visual function in deaf children and young adults aged 1-21 and suggest areas where further research is desirable. We show that differences in methodology and population characteristics have at times led to conflicting findings. Many terms have been used for visual and ocular deficits associated with deafness. We will use *visual defects* to refer to those conditions usually detected in optometric practice and *ocular abnormalities* to refer to conditions usually detected in hospital ophthalmology clinics.

The review process involved a comprehensive electronic literature search from various data bases: OneFile, Health Reference Center Academic, Social Sciences Citation Index (Web of Science), SciVerse ScienceDirect (Elsevier), Science Citation Index Expanded (Web of Science), Medline (NLM), MLA International Bibliography, American Psychological Association (APA), Project MUSE, ERIC (U.S. Dept. of Education), Oxford Journals (Oxford University Press), SpringerLink, SAGE Journals, Wiley Online Library, PMC (PubMed Central), Nature.com (Nature Publishing Group), Google Scholar. The following key words and combinations of words were used: deaf children vision, vision and deafness, deaf vision, eye and deafness, ophthalmic and deaf, optometry and deaf, refraction and deaf, vision and hearing, ophthalmological and deaf, ophthalmological and hearing, vision and ear, deaf and blind, eye and deaf, deaf vision and reading, reading and deaf, vision reading and deaf, near vision and deaf, near vision and hearing impaired.

Introduction

In the UK there are approximately 1 per 1000 children born each year with hearing impairment (Fortnum et al. 2001). Fortnum et al. (2001) defined hearing impairment as a hearing loss in the better ear of more than 40dB averaged over 0.5, 1, 2, and 4 kHz. Visual defects and ocular abnormalities have consistently been documented as being more prevalent in deaf individuals (Table 1) than comparative groups of hearing individuals (Pollard & Neumaier 1974).

Insert Table 1 about here

Refractive error is common in deaf individuals including children with uncomplicated deafness (i.e. no evidence of family history, congenital, or deafness caused by infective or metabolic disease) even allowing for emmetropisation. There is little consensus as to whether refractive errors are more frequent in the congenitally deaf than in those who acquire deafness at a later stage of life (e.g. Guy et al., 2003).

Ophthalmological screening regimes have been implemented for deaf children in an attempt to maximise visual abilities and minimise social and educational disadvantages (Siatkowski et al. 1993; Guy et al. 2003; Hanioglu-Kargi et al. 2003). Despite the awareness that visual abilities are essential in a non-hearing world, it would seem that very little attention has previously been given to near visual function, and in particular reading. Perfetti & Sandak (2000) suggest that the use of phonology (the study of how sounds are organized and used in languages) is associated with higher levels of reading skills among deaf readers and that "the effectiveness of the visual channel is not an issue". On the other hand, Martin et al (2012) suggested that deaf children who have reduced dynamic visual acuities may have reduced vestibular responses and reading difficulties. Children with congenital vestibular abnormalities displayed gross motor developmental problems that the authors suggested may impede the usual ocular motor/vestibular relationship. This in turn could impact on visual stability and hence acquisition of reading (Martin et al. 2012).

Due to the difficulty in recruiting deaf participants, several studies have found themselves reliant on retrospective examination of medical data (Table 1). This methodology reduces the validity of the data (Woodruff 1986) and is reliant on observations gathered from many different sources, giving results that are at best hypothesis generating (Hess 2004).

There are few studies that include direct comparisons between deaf groups and a matched hearing control group (Pollard & Neumaier 1974). Instead the majority of studies have chosen to compare their data with previous studies on a hearing population (Regenbogen & Godel 1985; Leguire et al. 1992; Guy et al. 2003; Hanioglu-Kargi et al. 2003). The majority of studies have reported simply age range and gender. However, some studies have divided gender and ages into year groupings (Pollard & Neumaier 1974; Mohindra 1976). One study, conducted in Washington DC, USA (Suchman 1967) specified racial grouping without attributing deafness or visual dysfunction to this factor. The racial grouping may or may not be important, but the majority of studies have not directly addressed this issue and have been ethnically biased to the country of origin (Table 1).

Visual defects - refractive and binocular vision abnormalities

Refractive and binocular vision abnormalities have typically been the most commonly reported. Studies have shown the prevalence of hyperopia, myopia and astigmatism to be between 18% and 39% (Pollard & Neumaier 1974; Mohindra 1976; Regenbogen & Godel 1985; Guy et al. 2003; Gogate et al. 2009) and binocular vision abnormalities (e.g. strabismus) between 5.3% and 18% (Regenbogem & Golden 1985; Hanioglu-Kargi et al. 2003).

Insert Table 2 about here

Various methodologies and classification criteria have been used in the assessment of vision / visual acuity (Table 2). For example, Bist et al. (2011) assessed vision and visual acuity with a Snellen tumbling "E" test chart as these do not require literacy. Whilst most research has used traditional Snellen charts at 6 metres there has been little use of log MAR assessment despite it being acknowledged as a superior measurement (Lovie-Kitchin 2008). Younger children's distance visual acuity has been assessed with a variety of tests including Sheridan Gardiner cards, Kay pictures, Lea Crowded Symbols (near vision), and for pre-verbal children, Cardiff preferential looking cards (Armitage et al. 1995; Guy et al. 2003). Crowed Kay pictures and Lea pictures are considered the most appropriate tests for younger children with the LogMAR crowded acuity test and the Sonsken LogMAR chart being the tests of choice for children over 3 years (Saunders 2010). The reliance on Snellen acuity charts as compared to the Log MAR system may be at least in part due to the location and the clinical nature of the majority of studies where Snellen charts are more commonly available.

Near vision assessments in deaf individuals are a rarity within the literature and when they have been undertaken the reduced Snellen tumbling 'E' letter charts have typically been used (Regenbogen & Godel 1985). For example, Hanioglu-Kargi et al. (2003) assessed with a Snellen Reduced E near chart and Khadehar et al. (2009) with near Lea symbols, finding 15 participants to have defective near vision though no definition of defect was given. Although measurement of near vision was detailed in Khadehar et al.'s (2009) methodology, no near vision results were published or discussed. It is evident that many of the deaf studies from developing countries (Khadehar et al. 2009; Gogate et al. 2009; Abah et al. 2011) have greater reliance on non-reading "illiterate" tests possibly indicating the greater difficulties these children have in acquiring basic reading skills when compared to their hearing counterparts or simply that the levels of literacy are much lower in these countries.

Refractive error has often been assessed objectively using retinoscopy both with, (Mohindra 1976; Regenbogen & Godel 1985; Leguire et al. 1992; Siatkowski et al. 1993) and without cycloplegia. Evidence of subjective non-cycloplegic refractions

having been performed is limited (Suchman 1967; Pollard & Neumaier 1974). This is consistent with the accepted viewpoint that cyloplegic refractions are the most accurate method of assessing refraction for children because of the control of accommodative effort (Fotouhi et al. 2012).   Inclusion criteria for refractive errors have considerable variation. For example, Guy et al. (2003) set inclusion for spherical ametropia at ≥ 4.00 D (dioptres) whilst Armitage et al. (1995) included hyperopia of ≥ 1.50D with esotropia (≥ 3.00D without esotropia). Outlined below are a few of the most commonly observed refractive and binocular vision abnormalities as documented in deaf individuals.

Hyperopia

Hyperopic ametropia associated with deafness is the most commonly reported refractive error (Alexander 1973: Mohindra 1976; Regenbogen & Godel 1985; Siatkowski et al. 1993; Armitage et al. 1995; Abah et al. 20011) with the prevalence varying between 8% (≥2.25D; Pollard & Neumaier 1974 - non-cycloplegic refraction) and 31.5% (≥2.50D; Siatkowski et al. 1993; cycloplegic refraction) as compared to between 4% (≥2.00D; Fan et al. 2004) and 12.8% (≥1.25D; Kleinstein et al. 2003) in a normal hearing population for cycloplegic refractions and 7.7% (≥1.50D; Junghans et al. 2002) for non-cycloplegic refractions.

Myopia

This is the second most frequently reported visual defect. It is acknowledged in the literature that myopia increases with age in hearing individuals (Coleman 1970; Saw et al. 2005), yet even controlling for age as a factor, there is still a greater prevalence of myopia in deaf and hearing-impaired children and young adults (Leguire et al. 1992). In fact the prevalence of myopic in the deaf has ranged from 6% (>1.00D; Hanioglu-Kargi et al. 2003) to 20.9% (≥ 4.00D; Guy et al. 2003).

Astigmatism

There appears to be a greater prevalence of astigmatism in the deaf and hearing impaired, with Pollard & Neumaier (1974) reporting 7.3% in their deaf participants compared to 1.4% in their group of hearing children. Compared to other visual defects, studies have shown far greater agreement with criteria for astigmatism, ranging from ≥1.00D to ≥1.50D (Pollard & Neumaier 1974; Siatkowski et al. 1993; Armitage et al. 1995; Guy et al. 2003), although Hanioglu-Karg et al. (2003) used a ≥ 2.00D criterion and reported prevalence in the deaf of 14.4%. Woodruff (1986) in his retrospective study suggested that higher levels of astigmatism (>1.00D) may be associated with congenital rubella, although no associations with disease process or level of deafness have been suggested elsewhere. Mohindra (1976) subdivided her astigmatic participants into 'with the rule' (steeper corneal curvature vertically) and 'against the rule' (steeper curvature horizontally). Corneal curvature was measured using keratometry, and there were twice the number of 'with the rule' astigmats than 'against the rule', though no relationship to deafness was described. A higher prevalence of with the rule astigmatism is in accordance with studies in a normal population (Khabazkhoob et al. 2010). Woodruff (1986) also reviewed corneal curvature suggesting congenital rubella subjects show greater curvature and a high prevalence of microphthalmia.

Amblyopia

A greater prevalence of amblyopia has consistently been shown in individuals who are deaf compared to individuals with normal hearing, with acuity levels for inclusion ranging from < 6/9 (20/30) (Hanioglu-Kargi et al. 2003) to < 6/60 (20/200) (Gogate et al. 2009) and prevalence ranging between normal levels (Leguire et al. 1992) and 14.4% (Hanioglu-Kargi et al. 2003). The increased occurrence of amblyopia has variously been attributed to ocular pathology, strabismus, cataracts and anisometropia.

Anisometropia

Anisometropia also has an increased prevalence in the deaf. Definitions of anisometropia have been extremely variable. For example, Pollard & Neumaier

(1974) set a criterion of 1.25D differential between eyes whilst Hanioglu-Kargi (2003) used $\geq$ 2.00D and Regenbogen & Godel (1985) $\geq$ 3.00D.

Binocular vision abnormalities

Strabismus (heterotropia) and heterophoria have commonly been measured with a simple cover / uncover test (Suchman 1967; Guy et al. 2003). Heterophoria has occasionally been quantified using an alternating cover test in association with a prism bar although few studies have reported the magnitude of phoria. Alexander (1973) used a cover/ uncover prism test and Maddox rod to quantify the heterophoria. Whilst these tests were stated in the methods, only strabismic anomalies were published in the results. Alexander found 11% of 572 deaf children with strabismus, 16 children having accommodative esotropia with a further 29 being non-accommodative. Mohindra (1976) used the cover test for distance and near, reporting results for the distance cover test only of a prevalence of 9% strabismus and 10% heterophoria. Deviations of > 10 prism dioptres have been considered significant (Leguire et al. 1992; Hanioglu-Kargi et al. 2003) and have been reported as more common in deaf cohorts compared with normal hearing cohorts. Regenbogen & Godel (1985) found a prevalence of 4.6% compared to 1.8% in a normal hearing population whilst Pollard & Neumaier (1974) found no difference with strabismus in 4.9% of their deaf participants compared to 4.8% in a hearing group, although the criteria in their hearing group was "less rigid". Accommodation and associated phoria (fixation disparity) have not featured in the reviewed papers. These assessments would give a greater insight into the coordination of the eyes which is especially important with near vision.

Stereopsis

Stereopis has been measured in early studies using the wings of a toy butterfly and more recently with the Titmus stereo fly, Wirt dot (Mohindra 1976) and TNO tests (Hanioglu-Kargi et al. 2003). Normal stereo acuity has been set at $\leq$100 seconds of arc for the majority of studies. Mohindra (1976), using the stereo fly and Wirt dot tests, found over 70% of the deaf participants with a stereopsis of $\leq$ 100" (seconds of

arc), with 49% having 40" and 19% having reduced stereopsis of > 100". Reduced stereopsis is associated with refractive error and/or an oculomotor abnormality that is in accordance with the greater prevalence of strabismus (Alexander 1973) and amblyopia (Hanioglu-Kargi et al. 2003) in deaf children.

Contrast sensitivity (CS)

Contrast sensitivity is mentioned in only one of the reviewed papers (Khandekar et al. 2009) and no methodology or results were published. It would appear unfortunate that assessment of CS has not been conducted as reduced CS can be associated with cataract and retinitis pigmentosa. Research into retinitis pigmentosa which has high association with Ushers syndrome has assessed contrast sensitivity and found reduced sensitivity in this group (Hartong et al. 2006). The lack of CS assessment could reflect the earlier unavailability of clinical CS assessment.

Colour vision

Colour vision has been assessed with the Ishihara Colour Test (Regenbogen & Godel 1974; Mohindra 1976), D15 Test (Khandekar et al. 2009) and Farnsworth-Munsell 100 Hue Test (Mohindra 1976). Mohindra (1976) found 2.1% of females (N=43) and 6.9% of males (N=29) to have colour defects using Ishihara and Farnsworth 100 Hue tests. These levels are consistent with larger scale normative studies and would suggest little variation in the prevalence of colour defects in the deaf (Birch & Platts 1993).

As the research outlined above clearly shows, associations between hyperopia, myopia, astigmatism, binocular anomalies and deafness in children are now well documented. There appears to be no difference in prevalence of these visual defects whether the deafness is congenital or acquired. This is also the case with the degree of hearing impairment, with profoundly and severely deaf children having an equal increase in the likelihood of a visual defect.

Range and severity of hearing impairment and visual performance

Assessment of deafness has centred on congenital sensory neural deafness in which deafness is associated with dysfunction of the vestibulocochlear nerve, inner ear, or central processing centres of the brain. The British Society of Audiology (2004) classifies hearing levels as shown in Table 3.

Insert Table 3 about here

Early studies have qualitatively grouped deafness into broad levels of moderate, severe and profound without quantifying the degree of deafness that was present (Suchman 1967), whilst others have associated hearing levels and ocular defects in greater detail having used subjects from audiology or specific deaf centres. For example, Armitage et al. (1995) assessed 83 children; 46 of them having severe hearing loss (>70dB) and 37 having profound hearing loss (>90dB). They assessed hearing with audiograms and hearing thresholds with octave frequencies of 500, 1000, 2000 and 4000Hz. They found 15 of the severe hearing loss group and 14 of the profound hearing loss group (total 35%) met their criteria for having a visual defect (see Table 2). Stockwell (1952) assessed refractive status in acquired and congenital deaf individuals, finding marginally higher levels of ocular defects in the congenitally deaf group, although 13% of the total cohort had an unknown cause of deafness.

Armitage et al. (1995) also compared ocular defects between congenital and acquired deafness, finding no significant differences between these groups. Moreover, Khandekar et al. (2009) investigated visual defects in the profoundly deaf > 81dB and severely deaf 61-80dB; but did not find any association between visual acuity and contrast sensitivity defects and level of hearing impairment. Leguire et al. (1992), categorised subjects into mild hearing loss (30-45dB), moderate loss (45-60dB), severe loss (60-80 dB) with these being grouped together as hearing impaired, whilst

profound loss (> 80dB) was categorised as individuals being deaf. Visual defects and ocular abnormalities were found in all categories to be more prevalent than in normative data, although the prevalence of refractive defects was similar in the hearing impaired and the deaf groups (hearing impaired 21.6% deaf 24.54%). There was a notable association between increased ocular anomalies and rubella.

In summary, no strong relationship between level of deafness and visual defects has been found (Leguire et al., 1992), with few studies categorizing the level of hearing loss. Whilst the classification criteria differ between studies these have been dependent on the application of international hearing standards or the use of national standards and experimental preferences. Although there may only be a weak association between the level of deafness and refractive and binocular vision abnormalities these defects are significantly more prevalent in deaf children when compared to people with normal hearing.

Ocular abnormalities

The retina and the cochlea structures are formed at the same developmental stage and embryonic layer, so any pathological defect within these areas could lead to oculoauditory defects (Armitage et al. 1995; Nikolopoulos et al. 2006), although the associations between various pathological processes and their impact on vision and hearing are not well described. There is little consensus in the literature regarding which diseases should be considered for inclusion in deaf vision studies with generic terms such as 'hereditary' and 'acquired' conditions being the most commonly reported. Some early studies such as Suchman (1967) have examined the external eye and observed the red reflex of the fundus giving little information of posterior segment pathology. Other studies (e.g. Guy et al. 2003) assessed pathological abnormalities in greater detail, having categorised the pathologies into: genetic syndromal, autosomal recessive, autosomal dominant, infective, metabolic, acquired and unknown causes. Sixty three of the 122 children in the study by Guy et al. (2003) had a genetic cause of their deafness, 13 were linked to known oculoauditory syndromes such as Usher's syndrome, Leigh's encephalopathy and Wildervank's syndrome, and 45 had an unknown cause. This greater detail has given better insight

into the associations between deafness, vision and the disease processes, enabling better identification of individuals who may be at risk from these disease processes, whether genetic or acquired, and allowing treatment at an earlier stage of development. In comparison, Regenbogen & Godel (1985) grouped the pathological conditions into broader areas: fundus, macular, external, pigmentary retinal changes, retinitis pigmentosa and optic disc atrophy but without relating the findings to any specific syndrome.

A diverse range of diseases has been related to deafness and vision defects and many of these diseases are very rare. Woodruff (1986) reviewed the case histories of 420 children attending schools for the deaf in Ontario, and reported congenital rubella as the most significant pathology and highlighted its association with an increased prevalence of strabismus and amblyopia, secondary to retinopathy and cataracts. Other studies have also found ocular pathologies associated with rubella (Mohindra 1976; Leguire et al. 1992; Mitchell et al. 2001). Fortunately congenital rubella is now a relatively infrequent cause of deafness particularly within developed countries (Nikolopoulos et al. 2006). Consequently, it is now more common to attribute deafness and visual problems to genetic causes and the more prevalent infective problems, for example: cytomegalovirus, toxoplasmosis and syphilis (Guy et al. 2003; Nikolopoulos et al. 2006). Unfortunately, 'unknown aetiology' is by far the largest pathological category in much of the research. Nikololpoulos et al*., (2006) reviewed in detail the ophthalmological abnormalities associated with deafness, and readers are referred to this paper for a full review.

In conclusion, it is now well established that associations between deafness, ocular pathology and visual performance exist. Assessment of deaf children's vision should always consider ocular abnormalities, together with the refractive and binocular status.

Communication and near vision

Visual defects in the deaf are particularly important due to the social and educational ramifications of having a dual disability (Dammeyer, 2010). The possible effects of visual defects on communication skills has not been adequately researched although it has been well established that deaf children have difficulties in reading and lag behind their hearing peers (Perfetti & Sandak 2000; Musselman 2000; Goldin-Meadow & Mayberry 2001). This developmental delay has often been attributed to a lack of phonic awareness of the words, making comprehension problematic. Surprisingly there has been relatively little assessment of the levels of near vision function and binocular coordination in these children: visual defects appear to have simply not been considered relevant. Indeed, there are a variety of proposed methods in the literature for reading acquisition in deaf children with a large proportion dedicated to phonic defects. Less attention has been given to logographic and orthographic (visual) routes to reading (Perfetti & Sandak 2000; Booth et al. 2000). Whilst phonic understanding of words would appear essential for reading, visual recognition of the words is the starting point for any reading task. Therefore any functional near visual impairment may impede this development.

Conclusion

Research over the past 70 years has established a strong relationship been deafness and ocular abnormalities, Most studies have investigated (almost) exclusively visual acuity when viewing in the distance. Whilst these have shown higher levels of dysfunction in the deaf when compared with normal hearing groups, surprisingly little investigation of near vision function has been made (and when made have not been reported; Mohindra, 1976). Near vision is especially important when considering the altruistic objective of enhancing social and educational abilities.

The visual function of a deaf child has implications for many aspects of the child's social and cognitive development. It is now well established that visual defects are

more prevalent in severely and profoundly deaf children with all levels of deafness showing increased visual defects and ophthalmological abnormalities when compared to hearing children. An understanding of near visual functions is less well established with very few studies investigating these adequately. Information and knowledge are acquired almost exclusively visually in deaf children, whether via sign language, lip reading, facial gestures, reading text, figures or pictorially. The effect of visual defects on communication has until very recently been relatively neglected.

A recent study by Martin et al. (2012) demonstrated that both abnormal dynamic visual acuity and motor impairment are associated with sensorineural hearing loss suggesting a basis for a relationship between reading abilities and deafness. The literature acknowledges the importance of early visual screening in order to maximise deaf children's visual performance. To date, visual function for near vision has not been specifically targeted for screening. Therefore there exists an urgent need for assessment and treatment of vision defects in deaf individuals, particularly those of near vision, because any improvements may enhance the visual development of communication and reading.

References