Visual Assessment in a Developmentally Disabled Population: Marsden Eye Survey

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Abstract

The aim of this study was to assess the visual function, ocular conditions and general diagnoses of 328 Marsden residents who were seen at the Marsden Eye Clinic between July 1985 and July 1997.

A questionnaire was sent to all parents and guardians regarding past medical history, pregnancy and birth history as well as family history. Medical records and Eye Clinic notes were reviewed and correlated with the questionnaire.

Results showed that a significant number (9%) of the Marsden residents are blind (with both eyes open) and many visually impaired, with best vision less than 6/12 (49.9% - 53.3%). Strabismus, nystagmus and refractive error feature predominantly in this group, as well as organic pathology, such as cataract and corneal scarring.

This remarkably stable population has a significantly greater incidence of visual impairment than the general Australian population.

Keywords:
Developmental delay, visual impairment, general disability, strabismus, refractive error, ocular abnormality.

Introduction

Marsden Hospital, located in Westmead, Sydney, is a residential centre for mentally and physically handicapped patients. When it opened in November 1969, it was the first purpose-built hospital in New South Wales for disabled children. Many of the original children, now adults, have remained at Marsden since its opening and reflect a remarkably stable population. These Marsden patients make up part of the 1.86% of Australians who have an intellectual disability[1].

Method

This current study, the Marsden Eye Survey (MES), commenced in July 1985, focuses on the visual function, ocular conditions and general diagnoses of 328 residents assessed at the Eye Clinic. Questionnaires were sent to all parents and guardians regarding background information on each patient at Marsden seen in the 12 year period. Information regarding pregnancy, birth, early childhood and illnesses, and family history were included in the questionnaire. The response rate was 53%. As the average age of the patients is now thirty, many parents have since passed on or were simply unable to be contacted.

Table 1.
The percentage of affected individuals in each category.

Category of General Diag.	% ofpopulation
Prenatal	5.5
Perinatal	8.5
Postnatal	9.5
Genetic	29.5
Unknown	47.0

Table 2.
The most common general conditions.

Condition	Number of Cases
Down syndrome	35
Postnatal infection	20
Rubella	13
Prematurity	11
Birth trauma	8
Angelman syndrome	6
Tuberous sclerosis	5

Table 3.
Predominant associated conditions found in the MES.

** Over half of this group were deaf because of Rubella.*

Condition	% of population
Epilepsy	43.0
Spastic Quadriplegia	12.8
Microcephaly	8.8
Deafness *	4.3
Hydrocephalus	3.0

Results

The youngest resident at Marsden is 13 years of age and the eldest is 45 years, with the average age being 30 years, which follows on from that found by Dr Graham Henry in his study of the patients at Marsden 17 years ago, when the average age was 13 years[2]. Since our study began in July 1985, only 13 residents have died, either from the progressive nature of their disease or pneumonia, and one in an accident. The number of males outnumbered the number of females at 1.8:1. 1995 figures of the Australian Institute of Health and Welfare show that, generally the number of Australian males with a primary disability type of acquired brain injury outweighs that of females by 1.8 : 1.[3]

14.6% of the Marsden patients had a family history of mental retardation and 1.8% had consanguinous parents. 73% were mobile, 22.5% wheelchair bound and 4.5% unknown as it had not been recorded earlier in the case notes and they are no longer at Marsden.

When the general disability diagnosis was known, it was grouped into one of five categories:

a)Prenatal causes: teratogenic, intrauterine infection (e.g. Rubella), trauma, structural abnormalities of the brain.

b)Perinatal i.e. at the time of birth and up to 4 weeks after birth: prematurity (<36/40), birth trauma and asphyxia, kernicterus, intracranial haemorrhage, hydrocephalus.

c)Postnatal, i.e. 4 weeks after birth: infections such meningitis, encephalitis, near-miss sudden infant death syndrome (SIDS), septicaemia, trauma, reactions to immunisation.

d)Genetic: chromosomal disorders (e.g.Down syndrome, fragile X), familial conditions (e.g. Tuberous sclerosis)

e)Unknown aetiology.

The incidence for each category is given in Table 1.

The high percentage in the unknown category reflects the fact that only if the diagnosis was definite, i.e. well documented in the history, was it grouped according to the cause. Further to this, the most common causes of developmental delay amongst the Marsden residents are summarised in Table 2.

Interestingly, 6 patients over the last few years have been detected as having Angelman (Happy Puppet) syndrome whereas previously their diagnosis was unknown. This reflects the greater sophistication of chromosomal testing in recent years, particularly the emergence of the Fluorescence In-Situ Hybridisation (FISH) technique, which looks for a specific gene mutation. It is now known that Angleman syndrome is caused by a mutation of chromosome 15.

It has been observed throughout the study that, in addition to their primary diagnosis, many of the Marsden patients have other significant disabilities. The most common of these conditions are summarised in Table 3.

The visual function and ocular condition of all residents was assessed. Visual acuity was tested using the Catford Drum, Sheridan Gardiner Single Letters and the linear chart. Visual acuity standards were divided into three levels:<6/60, 6/18-6/60, and 6/12 or better. Vision was recorded right eye (RE) and left eye (LE) if possible, otherwise with both eyes open (BEO). Approximately half of the patients had best recorded vision as 6/12 or better in either eye (RE = 50%, LE = 50%) or with BEO (46.7%). A study by Jan Erby[4] showed 50.2% of the same group to have best recorded vision at this level. Between 27.3% (RE) and 30.6% (LE) had monocular vision of 6/18-6/60 and 38.1% with BEO (Erby 43.2%). Visual acuity of less than 6/60 was present in 22.7% RE, 19.4% LE and 15.2% BEO, which is quite different from that found by Erby (6.7%). Since her survey, 18 years ago, many of the patients have had either

progressive neurological impairment, progressive eye disease including glaucoma and cataracts, or ocular trauma including that from self-injurous behaviour.

It must be noted that observation is a major tool used when assessing a population such as that of Marsden. Concentration and co-operation are very limited, making formal testing extremely difficult at times. Because of the difficulty in quantifying accurate visual acuity objectively, vision was also recorded subjectively as either good or poor. "Good" visual acuity meant functional vision allowing the person to move around, recognise people or accurately reach for objects, and "poor" meant blindly having to reach for objects and feel their way when walking. 82.6% had good vision, this group containing 38 people whose vision could not be tested due to lack of co-operation. 16.8% had poor vision, including 9% who had no perception of light with both eyes open. Reasons for poor vision and blindness are summarised in Table 4.

Congenital anophthalmos was present in 2 cases and 6 enucleations were also recorded. 9% of the Marsden population is blind. Current Australian Bureau of Statistics figures show that 0.09% of the Australian population is blind, with a further 1.42% having partial visual impairment[5]. These numbers highlight the fact that reduced vision is significantly more prevalent in this handicapped population. Squint was evident in nearly half of those seen (49.4%), with slightly more exotropias than esotropias (25.9% : 22.9%). 25% of patients had nystagmus with 18% being horizontal in direction. Eye muscle movement disorders were minimal, with no pattern detected.

Refraction and fundoscopy were also performed, using either Cyclogel 1% or Mydriacyl 0.5%. Refractive error was not corrected if it fell within plano to 3.00D of hypermetropia or myopia, or <2.00D of astigmatism, which is a judgement based on clinical experience. Glasses given for those lower amounts generally are not worn and often no obvious visual benefit is gained. It has been observed many times that a patient with uncorrected high myopia or hypermetropia is very mobile and accurate when reaching for an object.

Results showed the highest incidence of refractive error to be myopia and astigmatism (Table 5). 29% of patients have either previously worn or currently wear glasses, 61% have never worn them and 10% are unknown. It should be stressed that there is a need to check the glasses being worn as it is not uncommon to find someone else's glasses being worn by mistake.

Cause of Poor Vision/Blindness	Number of Cases
Cortical visual impairment	15
Optic atrophy	8
Congenital abnormality *	8
Cataract	5
Rubella oculopathy	4
Retrolental fibroplasia	2
Glaucoma	2
Trauma	1
Optic nerve hypoplasia	1
Retinal dystrophy	1

Table 4.
Causes of poor vision and blindness found in the MES.

**Congenital abnormality includes microphthalmos and coloboma.*

Refractive Error	Right Eye	Left Eye
Plano to +/-3.00D	67.4%	68.2%
Myopia>-3.00D	22.2%	21.6%
Hypermetropia>3.00D	10.4%	10.2%
Astigmatism<2.00D	26.3%	26.5%
Astigmatism>2.00D	20.7%	18.6%

Table 5.
Percentages of type of refractive error found in the MES.
** Note: Some patients had both a spherical and cylindrical error combined - therefore totals are greater than 100%*

Throughout the orthoptic and ophthalmological assessments, may ocular abnormalities were noted, the six most common of which are summarised in Table 6 below.

Ocular Abnormality	% of Population
Cataract	17.7
Corneal scarring	7.9
Keratoconus	5.8
Glaucoma	4.8
Retinal detachment	3.4
Uveitis	1.0

Table 6.
The most common ocular abnormalities found in the MES.

Often, corneal scarring was secondary to previous cataract surgery, which led to uveitis and then a phthisical eye. This was particularly evident in the settling of Rubella oculopathy. Keratoconus was noted in 5.8% of patients. Interestingly, unilateral cataract formation has been observed to develop in 2 patients who had keratoconus and episodes of hydrops. It is considered by the authors that keratoconus is often a non-specific finding reflecting the fact that severe rubbing of the eyes causes a significant amount of trauma to induce keratoconus and occasionally a secondary cataract. It is also noted that acute hydrops is often a recurrent problem in this group of developmentally delayed patients.

Conclusion

Overall, the MES showed that this remarkably stable population of developmentally delayed individuals has a significantly greater incidence of visual impairment than the general Australian population. Often the general diagnosis is elusive, but with improved genetic testing, more patients and their families are able to be given a cause for the disability. This is an important consideration as often siblings are at the age of having children of their own. If genetic factors are involved, there are obvious implications of having further affected children.

Approximately 50% of patients had vision that was less than 6/12, but subjectively the majority had good functional vision (82.6%). Squint is a common feature, as is nystagmus. Myopia and astigmatism are the most prevalent refractive errors but glasses are infrequently prescribed.

The ocular status of each patient at Marsden has a significant impact on their daily living skills. Those with poorer vision need extra help and there is a stronger emphasis on stimulating other senses. Music therapy, ball rooms, touch rooms, carpeted walls and tactile toys are an integral part of those wards with the partially sighted and blind residents. At Marsden a homelife environment exists. This means day outings, picnics, and a variety of special functions. Those patients who are better sighted will visually gain more from these activities, but with extra intervention from staff, all patients are helped to ensure maximal gain. When assessing the vision of this population, the visual needs of each individual must be taken into account, and the functional level considered. Acknowledging the overall daily visual requirements of each patient is necessary, and realising that the effects of visual loss can sometimes be compensated for in other ways.

To know the ocular and visual status of a population such as that of Marsden is essential. Maximising the quality of life in the developmentally delayed is often the responsibility of the health professional. To quote Dr Graham Henry :"At grass roots level, if you cannot see you cannot feed or educate yourself and the earlier this is known to helpers [carers], the better".[2]

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