

BASIC MEDICAL SURVEILLANCE ESSENTIALS FOR PEOPLE WITH DOWN SYNDROME.

OPHTHALMIC PROBLEMS

(Revised 2012)

One of a set of guidelines drawn up by the Down Syndrome Medical Interest Group (DSMIG(UK))

1. There is a high prevalence of ocular disorder among people with Down syndrome. Refractive errors and/or squint may be present from an early age and persist into childhood (1,2,3). The majority of children with Down syndrome have reduced accommodation at near (2,4,5). Compared to the general population there is a tenfold increase in congenital cataract (6) and infantile glaucoma may also occur (7). Nystagmus is present in at least 10% (8) . Cataracts and keratoconus may develop in teenage years or later and studies suggest that these are approximately 4 times more common than in the adult general population (9). If untreated most of these disorders are a significant cause of preventable secondary handicap at all ages. Therefore there should be extra vigilance at all ages.
2. As with all children newborns with Down syndrome should be examined for congenital cataract and other eye anomalies by a trained person and this should be repeated at 6 weeks (10)
3. Visual behaviour must be monitored by the child's paediatrician particularly before the first formal ophthalmologic review. Those who start to squint or show other abnormalities of gaze, visual behaviour or attention should be referred for ophthalmological review.*
4. Between 18 months and 2 years all children with Down syndrome should have formal ocular/visual assessment by an orthoptist and ophthalmologist/optometrist in accordance with local arrangements. This should include orthoptic assessment, refraction, and fundus examination. At least one third will have ocular/visual defects by this age (1,11,12). Those with deviation from normal should be kept under appropriate specialist review. Refractive errors, most commonly hypermetropia, which often reduce spontaneously in other children, are likely to persist beyond infancy (3, 13). Correction for hypermetropia may be helpful at a younger age than that for typically developing children especially since the majority will have defective accommodation (2, 4, 5).
5. Those with no abnormality at first review should nevertheless have further full ocular/visual assessment including refraction around age 4 years (14, 10). At this age at least 50% are likely to have refractive errors (1).
6. After age 4, due to the increased prevalence of disorders, eye checks should be at least 2 yearly throughout life by professionals with appropriate skills and expertise in managing this client group (14, 15). These may be optometrists (hospital or high-street based) or ophthalmologists. If hypermetropia is not present at age 4 it is not likely to occur later on, but myopia may develop at any age (3, 13).
7. Children and adults with Down syndrome should be expected to respond to standard vision testing procedures at appropriate developmental age but a distraction free environment and extra time may be necessary to optimise performance. Distance and near functioning vision should be checked at every review whenever developmentally possible and a prescription for near correction or bifocals considered at all ages * (13, 16, 17). Detail vision is likely to

remain poorer than expected throughout life even when appropriate spectacles are worn (18,19)

8. Many High Street opticians/optometrists give an excellent service particularly for older children, but younger children and those who are difficult to examine in this setting should be seen in a specialist clinic.
9. Blepharitis has been reported to occur in up to 30% of children with Down syndrome (8,20) and can be managed in the usual way (21). Nasolacrimal duct obstruction also occurs commonly (20, 22) and may need specialist referral. *
10. In view of the high prevalence of ocular disorders (see item 1) and the communication difficulties encountered in this client group any child or adult with pain, and/or changing vision, and/or red eye should be referred in the normal way.

* *good practice point*

References

1. Woodhouse JM, Pakeman VH, Cregg M, Saunders KJ, Parker M, Fraser WI, Sastry P, Lobo S. (1997). Refractive errors in young children with Down syndrome. *Optometry & Vision Science* **74** 844-851
2. Haugen OH, Hovding G. (2001). Strabismus and binocular function in children with Down syndrome: A population-based, longitudinal study. *Acta Ophthalmologica Scandinavica*. **79** 133-139
3. Cregg M, Woodhouse JM, Stewart RE, Pakeman VH, Bromham NR, Gunter HL, Trojanowska LD, Parker M, Fraser WI. (2003). Development of refractive error and strabismus in children with Down syndrome. *Investigative Ophthalmology & Visual Science* **44** 1023-1030
4. Woodhouse JM, Cregg M, Gunter HL, Sanders DP, Saunders KJ, Pakeman VH, Parker M, Fraser WI, Sastry P. (2000). The effect of age, size of target and cognitive factors on accommodative responses of children with Down syndrome. *Investigative Ophthalmology & Visual Science* **41** 2479-2485
5. Cregg M, Woodhouse JM, Pakeman VH, J SK, Gunter HL, Parker M, Fraser WI, Sastry P. (2001). Accommodation and refractive error in children with Down syndrome: cross sectional and longitudinal studies. *Investigative Ophthalmology & Visual Science* **42** 55-63
6. Kallen B, Mastroiacovo P, Robert E. (1996). Major congenital malformations in Down syndrome. *American Journal of Medical Genetics*. **65** 160-166
7. Traboulsi EI, Levine E, Mets MB, Parelhoff ES, O'Neill JF, Gaasterland DE (1988) Infantile glaucoma in Down's syndrome (trisomy 21) *American Journal of Ophthalmology* **105** 389 – 394
8. Pires da Cunha R, Belmiro de Castro Moreira J. (1996). Ocular findings in Down's Syndrome. *American Journal of Ophthalmology* **122** 236-244
9. van Schrojenstein Lantman-de Valk HMJ, Haveman MJ, Crebolder HFJM, (1996) Comorbidity in people with Down's syndrome: a criteria based analysis. *Journal of Intellectual Disability Research*, **40** 385 – 399
10. Rahi JS, Williams C, Bedford H, Elliman D. (2001). Screening and surveillance for ophthalmic disorders and visual deficits in children in the United Kingdom. *British Journal of Ophthalmology* **85** 257-259
11. Wong V, Ho D. (1997) Ocular abnormalities in Down syndrome: an analysis of 140 Chinese children. *Paediatric Neurology* **16**. 4. 311-314
12. Crofts B, unpublished data
13. Haugen OH, Hovding G, Lundstrom I. (2001). Refractive development in children with Down's syndrome: a population based, longitudinal study. *British Journal of Ophthalmology* **85** 714-719

14. Guidelines for Children's Eye Care.(2002) Children's Eye Health Working Party, a joint working party comprising members of the Royal College of Ophthalmologists, College of Optometrists and British Orthoptics Society.
15. Guidance on Frequency of Eye Examinations (2001). Issued by the College of Optometrists
16. *Woodhouse JM*. Bifocal information leaflet.
dsmig.org.uk/library/articles/bifocals.html
17. *Stewart RE, Woodhouse JM, Trojanowska LD*. (2005). In focus: the use of bifocals for children with Down's syndrome. *Ophthal Physiol Opt*. **25**. 6 514-522
18. *Courage ML, Adams RJ, Reyno S, Kwa PG*. (1994). Visual acuity in infants and children with Down syndrome. *Developmental Medicine & Child Neurology* **36** 586-593
19. *John FM, Bromham NR, Woodhouse JM, Candy TR*. (2004). Spatial vision deficits in infants and children with Down syndrome. *Investigative Ophthalmology & Visual Science* **45** 1566-1572
20. *Fierson WM*.(1990).Ophthalmological aspects. In: *Clinical perspectives in Down syndrome* (Ed. Van Dyke D., Lang D., Heide F., Van Duyne S., Soucet M.) Springer-Verlag, New York, pp 26-54
21. *Oxford Eye Hospital*. Advice re lid hygiene.
dsmig.org.uk/library/articles/blepharitis.html
22. *Berk AT, Saatci AO, Ercal MD, Tunc M, Ergin M*. (1996). Ocular findings in 55 patients with Down's syndrome. *Ophthalmic genetics*. **17** 15 – 19

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