

# Vision and vision problems in children with Down's syndrome: a research update

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Photos courtesy Mike O'Carroll, our parents and children



## Our study group

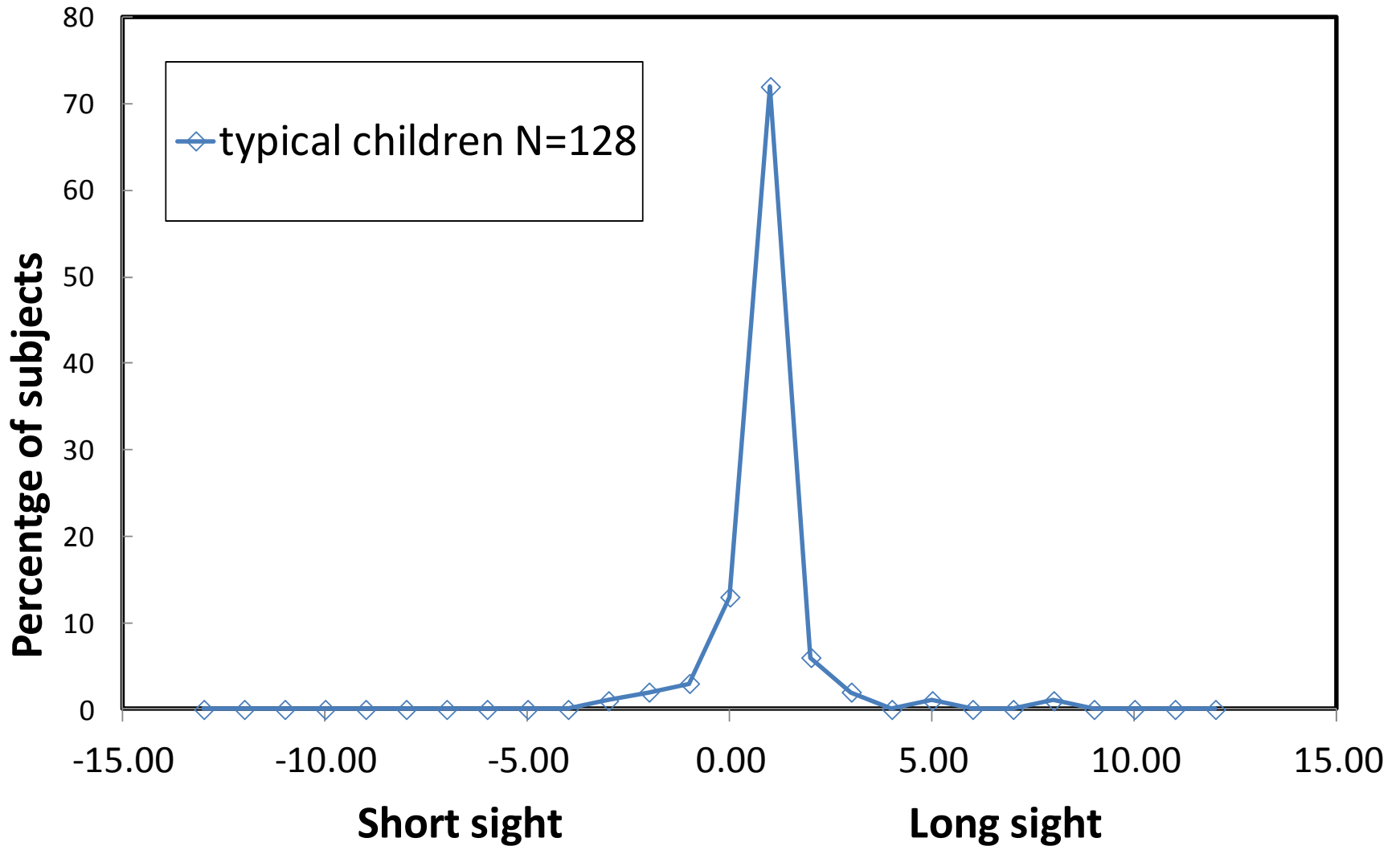
- cohort first established in 1992
- recruitment through paediatricians, ophthalmologists, health visitors, other parents, website
- current cohort = 250+ ages 2 months to 25 years
- longitudinal monitoring by home /school / clinic visits
- lab-based studies

## Visual defects in DS

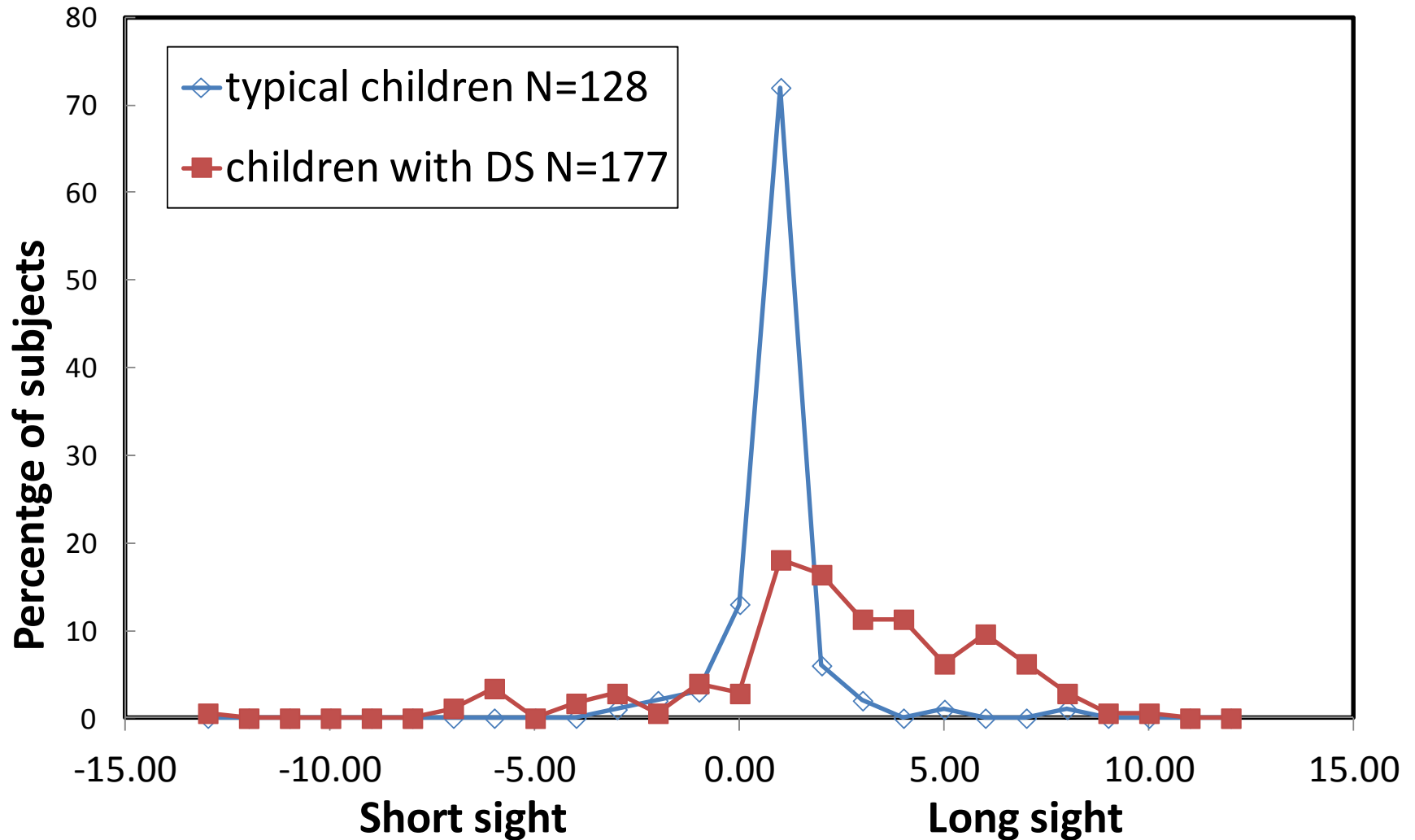
- 60% will require spectacles in childhood for long or short-sight
- 75% will require help with poor focusing
- 100% have below-normal vision
- 25% will have squint
- 15% will have nystagmus
- 15% will develop keratoconus (distorted cornea) in adolescence

# **Refractive error** (long and short sight)

# Typical children



# Children with Down's syndrome

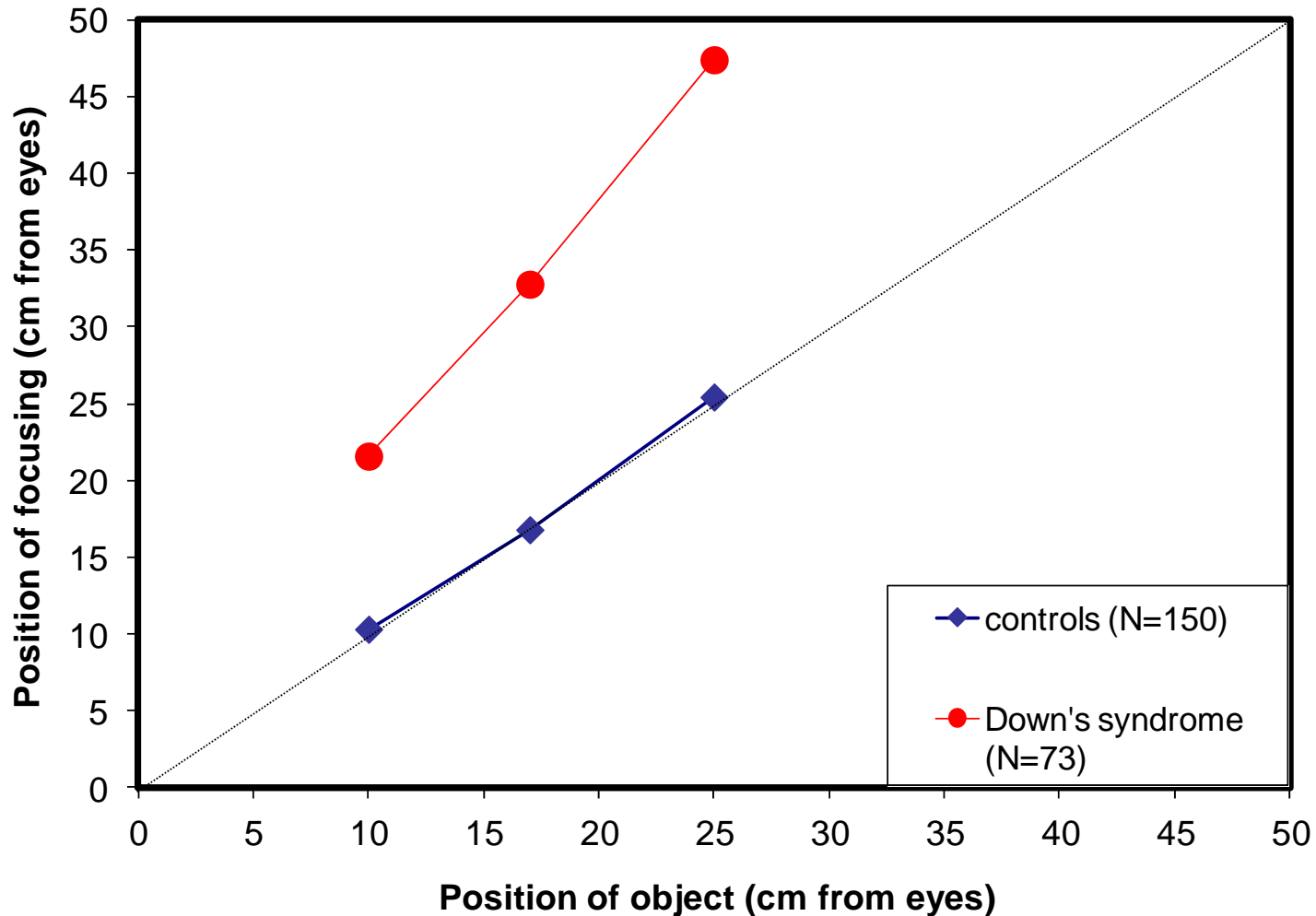


**Children learn at near**

**Measuring near focusing - accommodation**

# Children's focusing

(norms replotted from McClelland JF, Saunders KJ. *Optom Vis Sci.* 2004  
81:929-33)





- Among children with DS, 75% show under-accommodation
- *Children's long and short sight were fully-corrected when these measurements were taken*

## **In cross-sectional studies**

- There was no difference in the accommodative response between corrected and uncorrected hypermetropes (long-sight,  $p = 0.991$ )

## **In longitudinal studies**

- There was no difference in the accommodative response in individual long-sighted children before and after spectacle provision ( $p = 0.181$ ,  $N=9$ )

(Cregg , Woodhouse, *et al* . IOVS 2001: 42, 55-63)

- There was no difference in the accommodative response in individual children before and after provision of up-to-date spectacles ( $p=0.323$ ,  $N=7$ )

(Nandakumar and Leat, OVS 2009: 86, 196-207)

# Bifocal spectacle trial

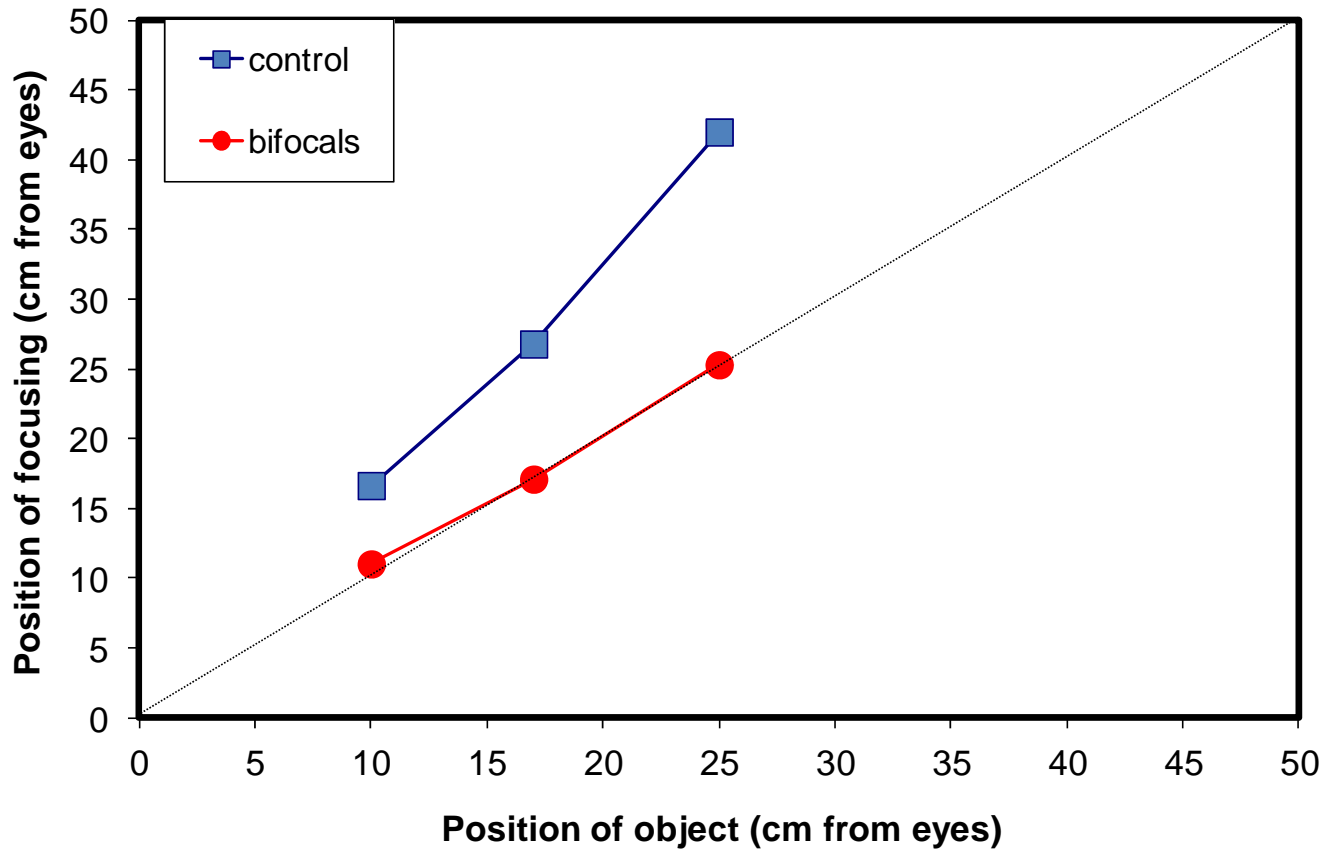
- Two groups of 17 children with focusing defect, matched for all relevant factors
- Bifocal group – given specs with a +2.50D add (for use in school only) and had single vision specs for home
- Control group - had new ‘single vision’ specs for school use and kept their current specs for home

# Accommodation

At the outset there was no difference in the accommodation between the two groups ( $p=0.851$ )

- control group: mean accommodative deficit =  $3.37D \pm 1.31$
- bifocal group: mean accommodative deficit =  $3.44D \pm 1.07$

# After 5 months of spectacle wear



## Outcomes of trial

- Children wore their bifocals successfully
- Children in the bifocal group focused more accurately
- Teachers reported better concentration and writing skills in the children wearing bifocals (Stewart, Woodhouse, Trojanowska, OPO, 2005; 25, 514-522)

## Corroborative data

- Independent study in Waterloo, Canada showed significant improvement in literacy and visual perceptual skills with bifocals (Nandakumar, Leat, *Acta Ophthalmologica*, 2010; 88, 196-204)

## Prescribing bifocals

- We prescribe bifocals routinely for all children who consistently show a focusing deficit and who are old enough to sit on a chair to do near tasks
- We use straight-topped D-28 bifocals, fitted with the segment top at, or just below, pupil centre.



# Bifocal wear can be temporary

- 40 children were prescribed bifocals
- 26 (65%) children improved accommodation through the distance part of the lens
- 14 (35%) children became accurate through the distance part of the lens and **have returned to single vision lenses**

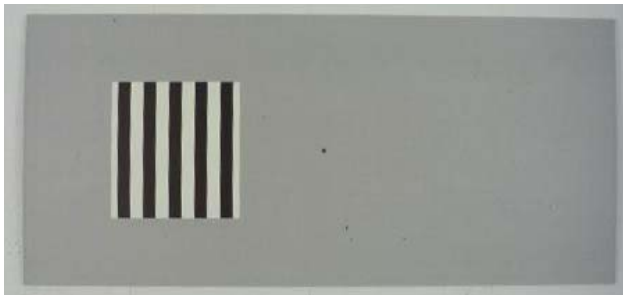
(Al-Bagdady, Woodhouse, *et al* OPO 2009:29: 416–421)

# Visual acuity and contrast sensitivity

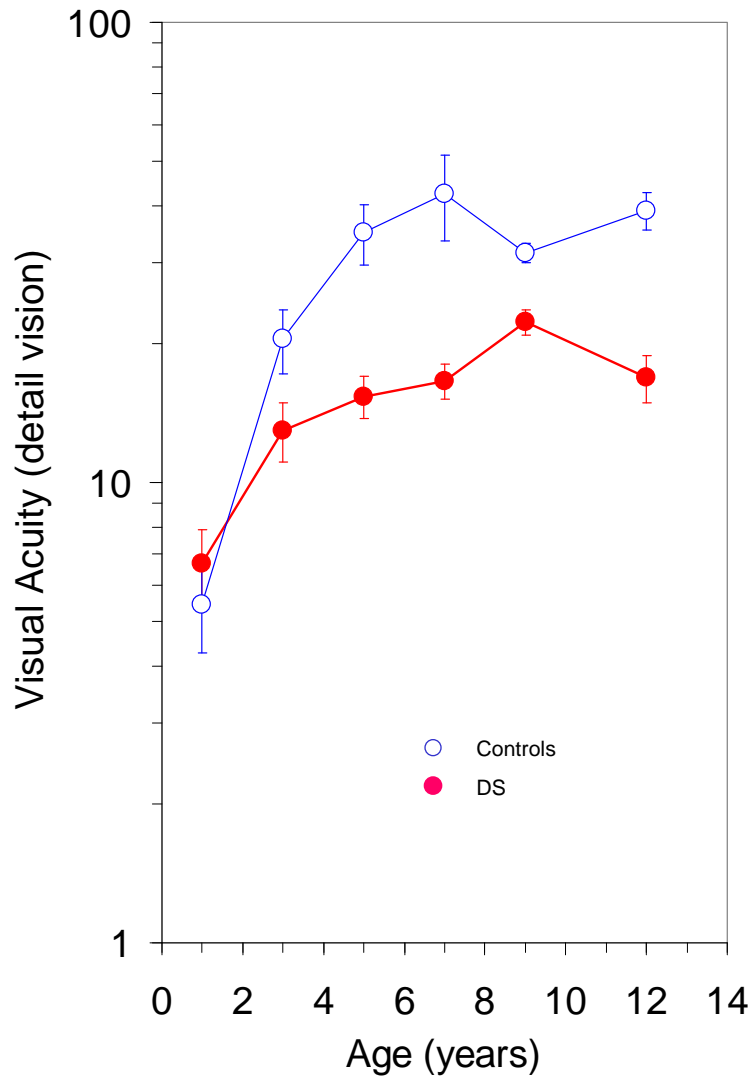


PELLI-ROBSON CONTRAST SENSITIVITY CHART  
Derivated by **HS** - General Color

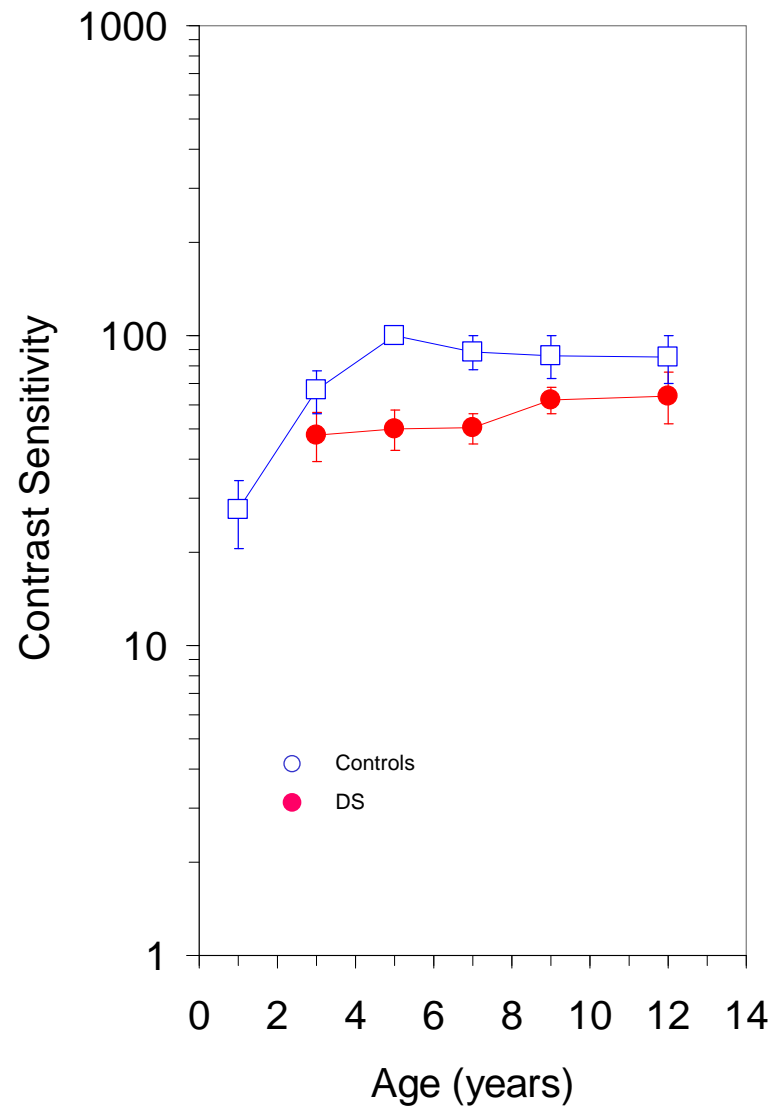




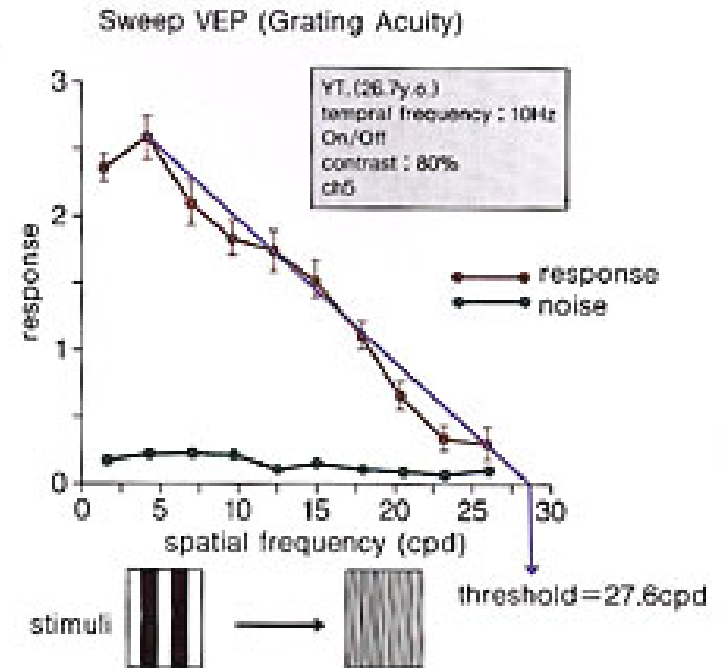
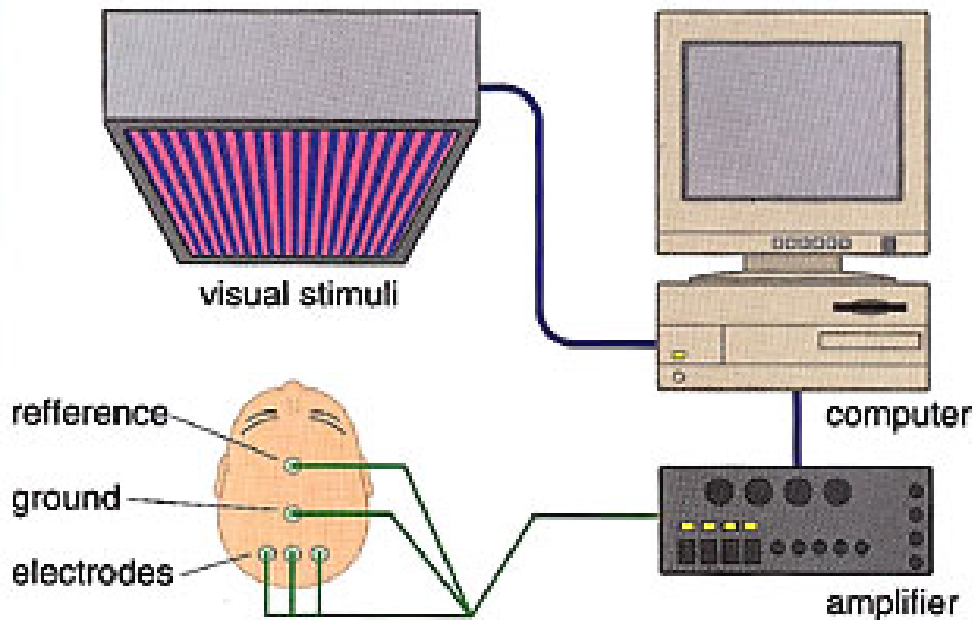
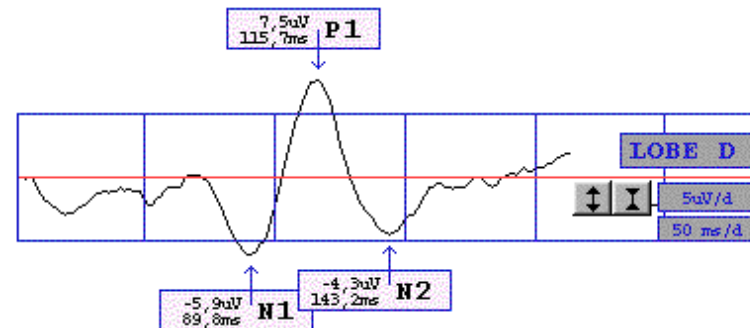
## Behavioural acuity



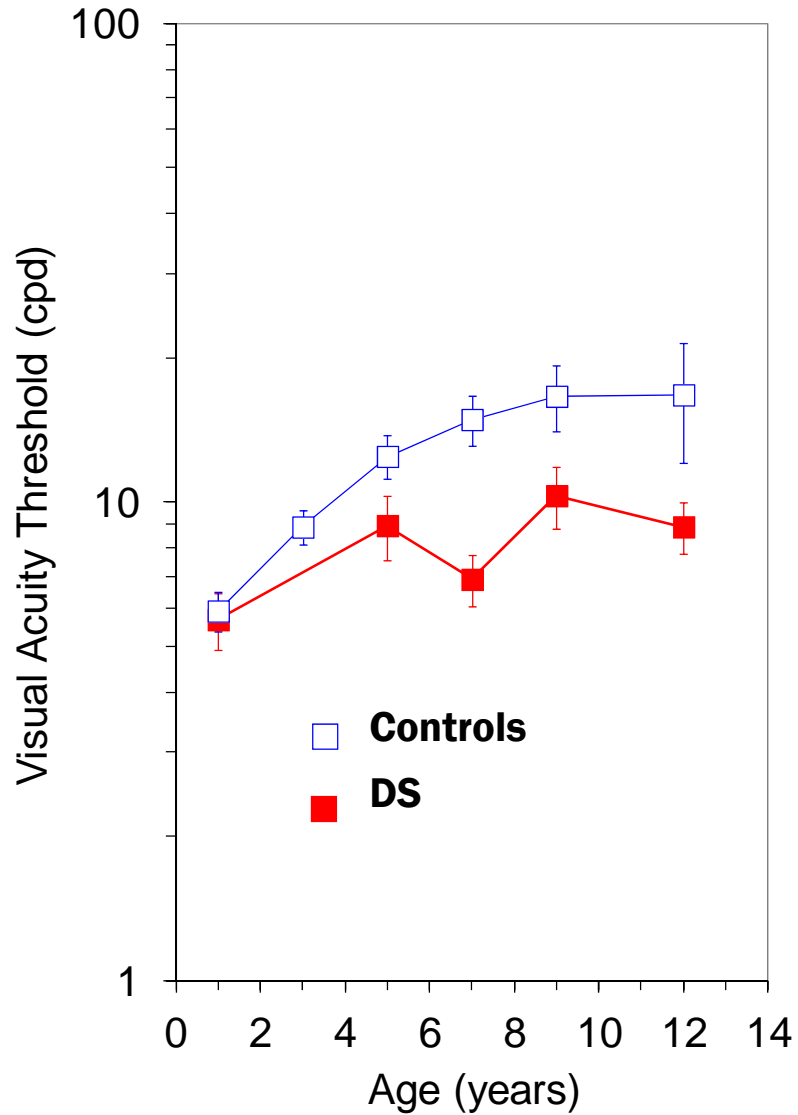
## Behavioural contrast sensitivity



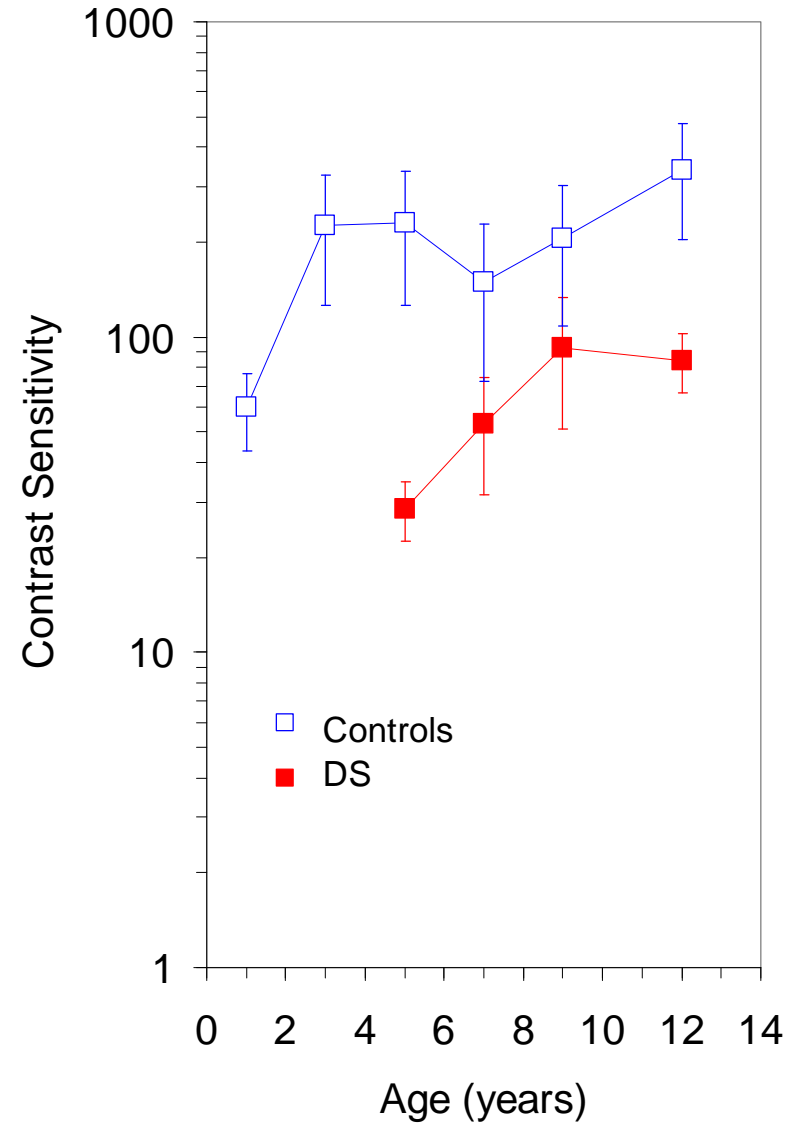
# Visual Evoked Potentials - VEP's



## VEP visual acuity



## VEP contrast sensitivity



# Vision in DS

- Children with DS have poorer acuity and contrast sensitivity than their classroom peers
- Vision is below normal even in children who do not need spectacles, or when correct spectacles are worn

# Squint

- Much more common in children with DS (25-30%) than in the general population (2-4%)
- Managed / treated in the same way
  - Spectacles
  - Surgery
  - Patching



# Nystagmus

- More common in children with DS (15%) than in the general population (0.02%)
- It is NOT part of the syndrome
- Nystagmus is a visually impairing condition

# Keratoconus

- Distortion of the cornea causing poor vision
- Cannot be corrected with spectacles
- Prevalence in Down's syndrome reported as 10-30%
- Many people with Down's syndrome successfully wear contact lenses
  - The barriers to contact lenses usually lie with family and carers



# Early diagnosis of keratoconus is now critical

- Collagen cross-linkage therapy can halt progression and is becoming available
- Treatment is viable only while the cornea remains thicker than 400 $\mu$ m



# The cornea is different in Down's syndrome

- The cornea is thinner
  - DS  $475.7 \pm 35.8 \mu\text{m}$
  - DS  $480 \pm 40 \mu\text{m}$

controls  $540.7 \pm 38.4 \mu\text{m}$

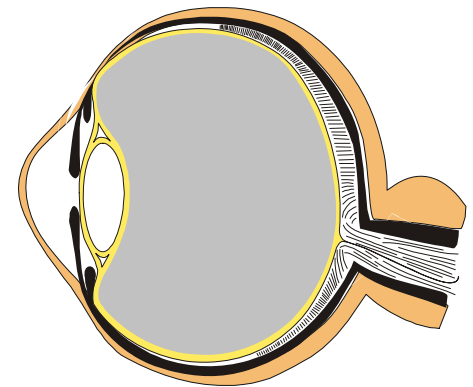
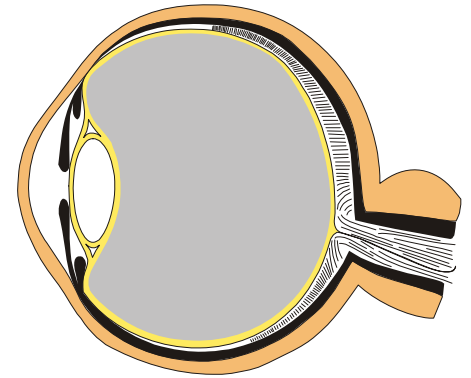
controls  $550 \pm 30 \mu\text{m}^*$

- The cornea is steeper
  - DS  $45.66 \pm 3.10\text{D}$
  - DS  $46.20 \pm 1.95\text{D}$

controls  $42.39 \pm 3.84\text{D}$

controls  $43.4 \pm 1.40\text{D}^*$

\*Haugen OH et al, (2001) Acta Ophthalmol. Scand 79: 616-25



Early diagnosis of keratoconus is difficult

# Clinical implications

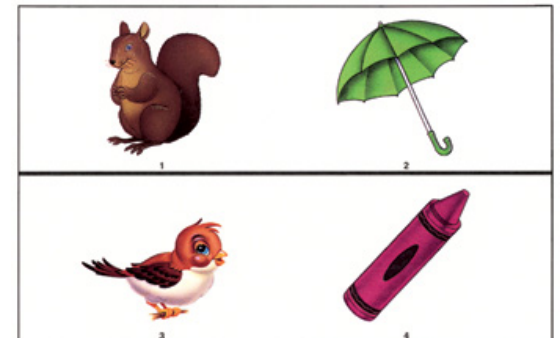
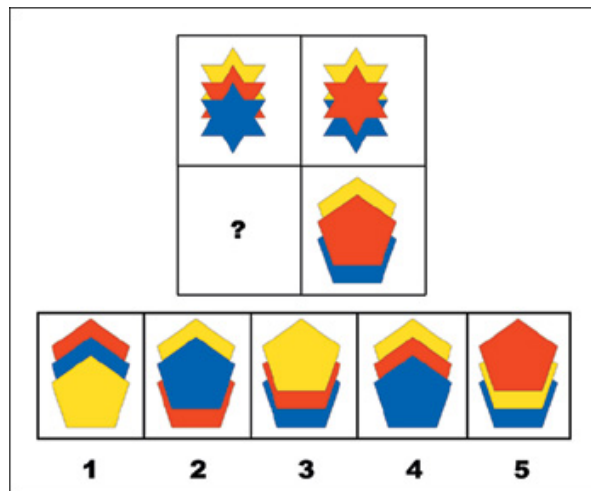
- Children should have a full eye examination by 2 years of age
- Every eye examination should include a check of focusing / near function
- Bifocals should be considered for all children with a focusing deficit
- Parents and teachers must be informed that vision is below normal, and classroom modifications will be needed

# Clinical implications

- Children with squint should be referred in the usual way
- Children with nystagmus should be referred to the Visual Impairment support service of the LEA on diagnosis
- Adolescents should be encouraged to have regular eye examinations with a view to detecting keratoconus

# Question

- When a child with DS is assessed for cognitive development e.g. by an Ed Psych, what difference does it make that the child does not see the test material as well as a typical child?
- The 'norms' for these tests have been established for normally-sighted children – are they applicable to a child with a sight problem?



## Thanks to:

- all of the children who take part in our studies, and their families
- sponsoring organisations:
  - DSA
  - Mencap
  - Community Fund
  - Mencap City Foundation
  - Healthcare Foundation
  - National Eye Research Centre
  - Welsh Assembly Government

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