Institution: Cardiff University

Unit of Assessment: UoA3

Title of case study:
Improving eye care and learning potential for children with Down's syndrome

1. Summary of the impact (indicative maximum 100 words)

Cardiff University has established the world's largest single cohort of children and young adults with Down's syndrome in which visual deficits have been characterised. Cardiff research studies over 20 years have informed evidence-based guidelines published by the Department of Health and national practitioner bodies for the recognition and practical management of common visual problems in children with Down's syndrome. Consequently, eye care specialists now recommend bifocals for these children. The Cardiff research has also supported changes to teaching methods and resources for children with Down's syndrome. Collectively these children are now benefiting from better eye care and improved learning and educational opportunities.

2. Underpinning research (indicative maximum 500 words)

Beginning in 1992, with grant support from the Down's Syndrome Association and the Medical Research Council, Dr Margaret Woodhouse (1974 to present, Lecture and Senior Lecturer) and her team from Cardiff University began to build up a unique study group of children and young people with Down's syndrome. As Director of the University's Special Optometry Assessment Clinic (founded in 1993), Woodhouse has driven the expansion and monitoring of the cohort over the past 20 years. This clinic acts as a flagship service for the treatment of visual deficits in children and young people. It is the only clinic of its kind in primary healthcare in England and Wales, seeing over 200 children with Down's syndrome each year. The Cardiff longitudinal database now comprises over 250 children and young adults with Down's syndrome ranging in age from 1 to 25 years; it is the largest database of its kind in the world. Longitudinal data and studies from this research programme have contributed to a series of important research findings, detailed below.

The causes of poor vision in children with Down's syndrome

Woodhouse and colleagues showed that in the early weeks of life the occurrence and distribution of refractive errors are similar in children with Down's syndrome to those of typical infants. However, the process known as 'emmetropisation', which reduces these errors in typical infants, fails in infants with Down's syndrome; refractive errors do not diminish with age in these children. The Cardiff team showed that by the time they go to school, ten times more children with Down's syndrome need glasses compared with typical children [3.1]. Further data analysis of the same cohort (n=182) showed that the prevalence of oblique corneal astigmatism, which causes blurred vision and which is uncommon in typical children, increases to an incidence of 45% in teenagers with Down's syndrome [3.2].

However, refractive errors are not the only cause of poor vision in children with Down's syndrome. The Cardiff team through research examinations (using the Cardiff Acuity Test developed by Woodhouse in 2005 and licenced (royalty-free) across 38 countries – ca. 150 licenses p.a. - specifically to assess the vision of infants and children who have difficulties in communication) showed that up to three-quarters of children with Down's syndrome display inaccurate accommodation (i.e., inability to focus through shape adaptations of the lens). By the age of 12 months, typical children have accurate eye accommodation that rarely deteriorates through childhood; the accommodation defect in children with Down's syndrome persists even when they wear spectacles to correct fully long or short-sight [3.3].

The case for bifocals

The Cardiff researchers discovered that bifocal spectacles corrected defective accommodation in children with Down's syndrome. Children wearing bifocal lenses achieved accurate accommodation and preferred to wear their bifocals full-time rather than changing back to conventional spectacles for out-of-school activities [3.4]. The team also found that a significant proportion (35%) of children with Down's syndrome prescribed bifocals began to accommodate accurately after wearing bifocals for roughly three years; these children returned successfully to single vision lenses [3.5].

A persistent visual deficit

However, it appears that children with Down's syndrome have an additional visual deficit that is not correctable with glasses. Recognising that vision tests rely to some extent on the cognitive ability of the subjects, the Cardiff team (in collaboration with US investigators) led research using electro-encephalography (EEG) to measure visually stimulated brain activity. These studies revealed that visual deficits in children with Down's syndrome cannot be explained by poor cognitive performance alone [3.6]. The deficits remain even when the child's visual function is fully optically corrected and assessed by entirely objective means [3.6]. By the time children with Down's syndrome enter school, their visual capabilities are significantly poorer than their classroom peers.

In summary, prior to the Cardiff research the extent and natural history of refractive errors in children with Down's syndrome was unknown. For example, focusing for near tasks had not been measured at all in children with Down's syndrome, the assumption being in clinical practice that, as in typical children, accommodation would be good. The Cardiff research has redefined the landscape for visual deficits in children with Down's syndrome.

3. References to the research (indicative maximum of six references)

[3.1] **Woodhouse, J.M.**, Pakeman, V.H., Cregg, M., Saunders, K.J., Parker, M., Fraser, W.I., Lobo, S. and Sastry, P. Refractive Errors in Young Children with Down Syndrome. Optometry Vision Sci. (1997) 74: 845-851. http://journals.lww.com/optvissci/Abstract/1997/10000/Refractive_Errors_in_Young_Children_with_Down.23.aspx

[3.2] Al-Bagdady, M., **Murphy, P.J.** and **Woodhouse, J.M.** Development and distribution of refractive error in children with Down's syndrome. Brit. J. Ophthalmol. (2011) 95: 1091-1097. http://dx.doi.org/10.1136/bjo.2010.185827

[3.3] **Woodhouse, J.M.**, Cregg, M., Gunter, H.L., Sanders, D.P., Saunders, K.J., Pakeman, V.H., Parker, M., **Fraser, W.I.** and Sastry P. The effect of age, size of target and cognitive factors on accommodative responses of children with Down syndrome. Invest. Ophth. Vis. Sci. (2000) 41: 2479-2485. http://www.iovs.org/content/41/9/2479.full

[3.4] Stewart, R.E., **Woodhouse, J.M.** and Trojanowska, L.D. In Focus: The use of bifocal spectacles with children with Down's syndrome. Ophthalm. Physiol. Opt. (2005) 25: 514-522. http://dx.doi.org/10.1111/j.1475-1313.2005.00326.x

[3.5] Al-Bagdady, M., Stewart, R.E., Watts, P., **Murphy, P.J.** and **Woodhouse, J. M.** Bifocals and Down's syndrome: correction or treatment? Ophthalm. Physiol. Opt. (2009) 29: 416-421. http://dx.doi.org/10.1111/j.1475-1313.2009.00646.x

[3.6] John, F.M., Bromham, N.R., **Woodhouse, J.M.** and Candy, T.R. Spatial vision deficits in infants and children with Down syndrome. Invest. Ophth. Vis. Sci. (2004) 45: 1566-1572. http://dx.doi.org/10.1167/iovs.03-0951

Since 1994 Woodhouse and her team have received over £800K external research funds to support the work with some examples below. (Note: Cardiff University staff in **bold**).

- Welsh Assembly Government. Oct 2004-06. Ocular morphology in people with Down's syndrome (PI: **Woodhouse).** £62,000.
- National Eye Research Centre. April 2000-03. Factors Limiting Visual Acuity and Contrast Sensitivity in Children with Down's syndrome (PI: **Woodhouse**). £24,000.
- PPP Healthcare Medical Trust. April 2000-03. The impact and management of visual defects in children with Down's syndrome (PI: **Woodhouse**; CIs: **Fraser, Bennett-Gates**). £204,000.
- National Lottery Charities Board with Mencap. Oct 1999-2002. Visual dysfunction in children with Down's syndrome: clinical and molecular studies of myopia (PI: **Woodhouse**; CIs: **Erichsen, Guggenheim, Owen**). £251,000.
- Medical Research Council. 1994-97. Visual and cognitive development in infants and young children with Down's syndrome (PI: **Woodhouse**, CI: **Fraser**). £195,805.

4. Details of the impact (indicative maximum 750 words)

Educational guidance in the teaching and learning of children with Down's syndrome

Historically, children with Down's syndrome struggle with literacy skills, including exercises that require the ability to 'write on lines' which in particular was previously assumed to be due to poor fine motor control. The Cardiff work challenged this notion, showing that the problem stems from their poor vision. From this research the Cardiff team recommended that darker lines would improve the classroom performance of children with Down's syndrome. Teachers and parents have confirmed that children with Down's syndrome are better able to follow lines if they are darkened*,"Big and Bold"* [5.1]. In direct correspondence with Woodhouse, one parent states: *"One of [Ben's] targets at school was to write on the line which he couldn't do. It turned out the line was in pencil so I suggested that perhaps he couldn't see… they then drew the lines in pen and instantly he could write on the lines!"*

Cardiff's studies also showed that most children with Down's syndrome who were prescribed bifocals made an active choice to wear them all the time, not just in the classroom. The benefit of wearing bifocals is summed up in an email from the mother of Ethan, aged 3, a regular attendee of the Special Assessment Clinic in Cardiff: *"… he took to the bifocals straight away and hasn't put his old glasses back on since wearing them. [He is] steadier on his feet, has better communication and attention when looking at flash cards and so on."*

A recent study by an independent Canadian research team examined educational practices in children with Down's syndrome. The study confirmed the benefits of the Cardiff research-led recommendations that bifocals improved focusing for children and the wearing of bifocals led to a significant improvement in literacy and visual perceptual skills [5.2].

Cardiff's research findings and recommendations have been embraced by the Down's Syndrome Association, the only organisation in England, Wales and Northern Ireland which supports people with Down's syndrome at every stage of life. Since 2008 the Association has used the Cardiff research in its educational guidance [5.1] on the modification of teaching materials and practices, and the need for bifocal spectacles. This guidance is provided to schools and is also available for download [5.3]. Between February 2012 and September 2013 the guidance has been downloaded 9561 times [5.1]. The All Party Parliamentary Educational Advisory Group on Down's Syndrome has published a report [5.4] on educational practices that includes the Cardiff-research findings on visual deficits, the need for bifocals and of problems with writing on lines. The DSA Educational packs [5.3] that underpinned by the Cardiff research [5.1] are cited.

The Down's Syndrome Association uses the Cardiff research findings in its campaigns to benefit children with Down's syndrome [5.1]. For example, in February 2012 the Down's Syndrome Association has used the Cardiff research findings in its newsletter to its 7,500 UK members. The "Focus on Vision" theme of the Association's 2013 Awareness Week (18-23 March 2013) specifically sought to raise attention among schools, families and professional groups of the issues arising from the Cardiff research; the work of Woodhouse and her team was explicitly acknowledged on the Awareness Week web pages. As part of the week's campaigning, the Association distributed its educational packages to all 286 Local Education Authorities in England and Wales [5.1] and to 138 related charities. A recent poster campaign by the Association focused on the specialist eye care needs of children with Down's syndrome was published in Optometry Today (March 2013), which has a circulation of 20,000 within the optometry profession. Cardiff's research has formed a key part of guidance provided to parent support groups throughout the UK and Ireland, along with local professional associations [5.1], and is also reaching Down's syndrome support associations beyond the UK, for example in USA [5.5].

Eye care practice guidelines for children with Down's syndrome

The Cardiff research on inaccurate accommodation in children with Down's syndrome and the successful correction of this deficit using bifocal spectacles has led directly to changes in the UK eye care management of these children. Information for parents published in 2010 by the Department of Health acknowledges the visual impairments and Cardiff's research [5.6]. They recommended eye tests for children with Down's syndrome to include the endorsed measurements of their visual accommodation and the prescribing of bifocal spectacles to correct any deficit. This has changed the way eye-care practitioners manage children with Down's syndrome [5.7].

The Down's Syndrome Medical Interest Group (DSMIG) publishes evidence-based guidelines for

minimum standards of health surveillance aimed at UK and international paediatricians. It has a membership of over 150 clinicians. Dr Woodhouse was invited to become the group's first non-medical health care practitioner member in recognition of her contributions in the field of healthcare for children with Down's syndrome. The group use the Cardiff research in its 'Vision' guidelines [5.8,5.9] (updated in 2012). The DSMIG guidelines and the research of the Cardiff team are adopted by Paediatric Opthalmologists in their practice [5.10].

In summary, the unique structure provided by Cardiff University's Special Optometry Assessment Clinic has enabled the conduct of distinctive research that has led to the measurement of visual accommodation being introduced into the recommended UK eye-care management for children with Down's syndrome and to the prescribing of bifocal spectacles to these children. Furthermore, the Cardiff research showed that while persistent deficit in visual acuity in these children is not due to reduced cognitive ability it negatively impacted upon educational/classroom experiences, findings which have led to changes internationally in educational guidance in the teaching and learning practices for children with Down's syndrome.

5. Sources to corroborate the impact (indicative maximum of 10 references)

[5.1] Statement from Chief Executive Down's Syndrome Association. How Cardiff's research findings and recommendations have been embraced by the Down's Syndrome Association in the Association's educational guidance aimed at teachers and parents, and in the Association's optical health advice aimed at professional healthcare providers and parents.

[5.2] Independent educational research confirming the Cardiff research-based recommendations for the use of bifocals to improve in literacy and visual perceptual skills in children with Down's Syndrome. Nandakumar, K., Leat, S.J. Bifocals in children with Down syndrome (BiDS) - visual acuity, accommodation and early literacy skills. Acta Ophthalmol. (2010) 88: 196-204. http://onlinelibrary.wiley.com/doi/10.1111/j.1755-3768.2010.01944.x/abstract

[5.3] Links to Down's Syndrome Association support packages for primary and secondary education: http://www.downs-syndrome.org.uk/information/for-professionals/education.html. For example the primary school strategies that can be adopted:
http://www.downs-syndrome.org.uk/images/stories/DSA-documents/Publications/education/including_pupils_primary.pdf
with pp 3 and pp 14 acknowledging Woodhouse and Cardiff University research.

[5.4] All Party Parliamentary Educational Advisory Group Report (2012) indicates the challenge of visual deficits in Down's Syndrome children and the need for bifocals pp 8-9. The DSA Educational packs are referenced pp 31. http://appg-ds.org/web_images/APPG_Final2013.pdf

[5.5] USA State Down's Syndrome Association - Arkansas (USA) promoting the bifocal research findings of Cardiff and Woodhouse (p 3).
http://www.ardownsyndrome.org/Newsletters/2008_Vol.%201-No.%204.pdf

[5.6] Department of Health (2010). 'Information for parents: Down syndrome'. Raises as one of the topics Visual Impairment (p 55) and cites (p 55) the Cardiff team and the prescribing of bifocals. https://www.education.gov.uk/publications/eOrderingDownload/ES13-2010.pdf

[5.7] Statement from Senior Orthoptist, Leeds Teaching Hospitals - how Cardiff research has influenced eye-care practitioners managing children with Down's syndrome.

[5.8] Guidelines (2012) from DSMIG citing the Cardiff Research (pp 1-2), the prescribing of bifocals (bullet 7) as a good practice point. http://www.dsmig.org.uk/library/articles/guideline-vision-5.pdf

[5.9] Statement from paediatrician member of the Down's Syndrome Medical Interest Group (DSMIG). DSMIG's use of the Cardiff research in its vision guidelines, and impact of Cardiff research upon vision healthcare for children with Down's syndrome.

[5.10] Statement from paediatric ophthalmologist, Fellow of The Royal College of Ophthalmologists. Influence of Cardiff research upon practice.

All documents, testimony and webpages saved as PDFs are available from the HEI on request.