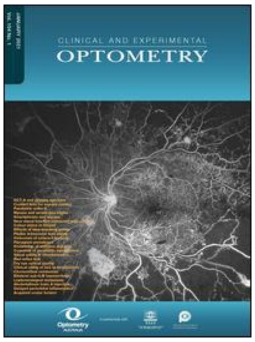

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CLINICAL COMMUNICATION

Cone dystrophy, childhood vision impairment and education: are clinical measures of visual function adequate to support a child through education?

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Introduction

This case report highlights the need to evaluate the adequacy of standard measures alone to assess the impact of vision impairment on learning within the classroom environment. Performance with other visual measures such as reading speed, print size requirements and contrast sensitivity are not routinely measured for children with a vision impairment. These additional functional measurements of visual performance can be used to identify areas causing accessibility deficits within the classroom. Use of functional measures to determine vision ability can contribute to improved support for the child, thereby improving their access to the curriculum alongside their normally sighted peers.

Retinal dystrophies result in progressive degeneration of photoreceptors and consequent gradual loss of visual acuity and visual fields. The impact of retinal dystrophies on daily living activities has been investigated within adults, with difficulties found to be related to standard clinical measures of visual function, such as visual acuity, contrast sensitivity, electrophysiological findings and visual fields.[1,2] Clinical measures have also been found to correlate with observed task performance and self-reported performance.[1] However, these adult populations investigated the effect on activities such as employment, travelling at night and driving which are not pertinent to children in education with a vision impairment.

Clinical measures are essential for diagnosis and vision-impairment level categorisation; this information similarly determines the degree of support a child will receive during their education. Within South Australia, support levels for children with a vision impairment are currently based on visual acuity and visual field extent, which broadly correspond to the World Health Organisation definitions of Blindness and Vision Impairment. This approach does not consider the functional impact of a child's specific visual impairment or reflect how the individual's visual diagnosis impacts their overall visual performance within the classroom. Factors such as room illumination, print size and contrast can influence visual function and capacity for the child to access educational materials.

This case report describes a 13-year-old girl (AR) with a cone dystrophy who has just transitioned from a specialist vision impairment school into a large mainstream high school in South Australia. It investigates differences in the standard visual function tests and compares these metrics to AR's visual performance, including self-reported impact of the vision impairment within the school environment for AR.

Case report

Medical history

AR was delivered at 35 weeks and at a routine post-natal 12-week check, nystagmus was noted. At 6 months, cone dystrophy was diagnosed based on visual electrophysiological measures. AR has a distant family history of Retinitis Pigmentosa on her maternal grandfathers' side. No genetic testing has been conducted to date. A comprehensive assessment of visual function was performed to evaluate AR's current level of performance, and she was interviewed regarding self-reported visual ability, using the Cardiff Visual Ability Questionnaire for Children,[3] and the impact it has on her educational learning.

Clinical/diagnostic tests

AR's refractive error was as follows: right eye: −2.25/-4.50×15 and left eye: −1.75/-2.00×160. Visual acuity was measured using the Freiburg Visual Acuity Test (FrACT).[4] FrACT is a computer-based visual acuity test, widely used in clinical trials and can accurately quantify visual acuity in the lower range.[5] It employs a Landolt C, making it suitable for young children, and visual acuities can be measured with a crowded or uncrowded target – which can indicate difficulty with visual clutter. Monocular crowded visual acuities were as follows: Right: 1.04 LogMAR (6/60^{-}) and Left: 0.98 LogMAR (6/48). AR has manifest nystagmus, with a left preference alternating exotropia.

A central 30-2 threshold visual field test (Humphrey HFA-II, Carl Zeiss) revealed generalised reduction in sensitivity for the right eye −7.60 dB and the left eye −8.70 dB, with a high false negative test index. Fundus photography, fundus autofluorescence and macular optical coherence tomography (Figure 1) were performed using the Zeiss Cirrus 5000 (Carl Zeiss). Optical coherence tomography and retinal photography revealed retinal atrophy with inner retinal thinning and focal macula hyperfluorescence (left greater than right). See Supplementary Material for full reports.

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Supplemental data for this article can be accessed here.

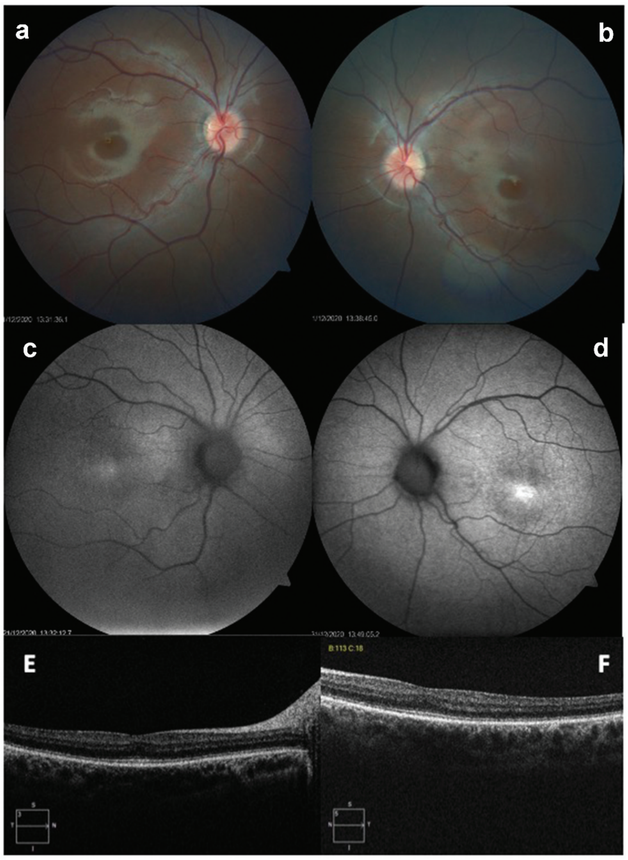

Figure 1. Figures (A) and (B) show the right and left eye fundus photographs with no marked retinal anomalies. Figures (C) and (D) show fundus autofluorescence with hyperfluorescence (left greater than right) indicating retinal pigment epithelial stress. Figures (E) and (F) show macular scans with normal foveal architecture.

Functional vision tests

Binocular crowded visual acuities were measured under differing ambient light levels using Lux LightMeter App. Under photopic illumination (450 lux), an acuity of 0.85 LogMAR (~6/45) was measured, which improved under mesopic illumination (10 lux) to 0.78 LogMAR (~6/38). Resolution threshold contrast sensitivity (CS) was measured using FrACT Contrast C,[4] and revealed a reduced measure of 0.71 LogCS compared to a typical value of 1.93 LogCS. Binocular colour vision, using a standard sized D15, revealed a Tritan defect as previously reported in macular dystrophies.[6]

Reading analysis was performed using the MNREAD App.[7] MNREAD was designed to assess aspects of reading such as reading acuity, critical print size (the smallest font size read at maximum speed), accessibility to printed material and maximum reading speed.[8] Detailed information of MNREAD and the metrics it measures can be found in supplementary material.

All reading measurements were taken at 30 cm. Using normal polarity (black writing on white background), AR achieved a reading acuity of 22-point (0.90 LogMAR) and critical print size of 29-point (1.02 LogMAR). With reverse polarity (white writing on black background), AR achieved a smaller reading acuity of 15-point (0.73 LogMAR) and a critical print size of 23 point (0.92 LogMAR). Additionally, at the recommended minimum font size of 23-point print with reverse polarity, AR's reading speed was 92.1 words per minute, compared to 56.8 words per minute with normal polarity. Correspondingly, Reading Accessibility Index was higher for reversed polarity (0.34) compared to normal polarity (0.31) indicating an improved reading performance with reversed polarity text (Figure 2, Table 1 and Supplementary Material).

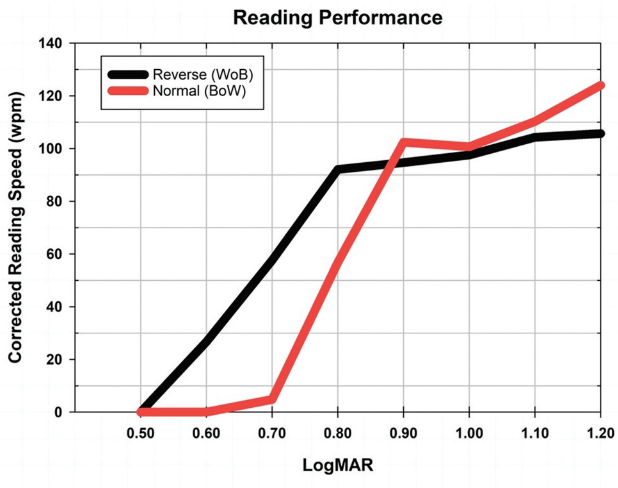

Figure 2. The effects of reversed polarity (White text on Black background (WoB) Black line and normal polarity (Black text on White background (BoW) Red line on reading speed). The sharp drop in reading speed is apparent once critical print size is reached. With the Reversed polarity AR was able to read smaller text and critical print size was smaller.

Discussion

This case highlights the benefits of using functional and performance-based measures of visual ability for children in education. Results from AR's clinical measures were consistent with a previously diagnosed cone dystrophy.

Childhood vision impairment is often categorised and support levels determined according to visual acuity and visual fields, but by providing additional measures of visual performance such as reading speed, print size

Table 1. Summary measures of reading ability with normal polarity (black print on white background) and reversed polarity (white text on black background) for AR. Reading acuity is the smallest print size that can be read before making errors. The critical print size is the print size at which subsequent smaller print sizes were read at 1.96 standard deviations slower than the mean of the preceding print sizes. The Reading Accessibility Index is a normalised value of an individual's ability to access commonly encountered print material (0 to 1 scale) where 1 = ability to read all print, and 0 = no ability to read commonly printed material.

Reading test parameters at 30 cm	Normal polarity	Reversed polarity
Reading acuity LogMAR (Print size)	0.90 (22)	0.73 (15)
Critical Print Size LogMAR (Print size)	1.02 (29)	0.92 (23)
Corrected Reading Speed at 23-point print (words per minute)	56.8	92.1
Reading Accessibility Index	0.312	0.342

requirements or the effect of contrast, a more holistic picture of the individual's abilities and needs can be utilised to improve their learning performance within the classroom.

Results from functional vision tests were used to inform class teachers of expected accessibility deficits and strategies to help overcome these. Recommended adaptions included required print size (including recommendations for electronic documents with reverse polarity/colour inversion), contrast requirements, lighting requirements based on visual acuity measures from different ambient lighting, management of visual fatigue due to nystagmus, impact of colour vision deficit, and extra time requirements based on reading speeds. A detailed report including functional measure results, interpretation of results and suggested strategies was provided to AR's parents and her school support team. (Recommendation examples included in supplementary material).

Reading, writing, and literacy skills are essential foundations to learning, and access to appropriate print size material can have a significant effect on a child's ability to learn at the same rate as their peers.[9] Critical Print Size can be used to determine an appropriate print size to enable optimum reading capacity.[7] AR's critical print size was 29-point with normal polarity, which can then be used to inform AR's teachers and carers of the minimum font size required to facilitate comfortable reading for extended periods. AR was able to read smaller sized print if it was brought closer, effectively magnifying the print. However, for sustained and extended near work in the classroom environment, using a closer working distance is difficult to sustain, particularly in the presence of nystagmus. When reading analysis was performed using reverse polarity, AR was able to read smaller print size at a faster rate and reading accessibility index improved, indicating better reading performance with reversed polarity text. This information informs teachers that AR's preference for learning material is electronic documents, so she can enlarge font to a comfortable size and reverse the polarity. MNREAD charts have published reference baseline measures of reading speeds,[10] which enables the provision of advice regarding time allowances for class work and assessments.

In South Australia, recommended print size in combination with other factors, such as reading speed, impact of visual fatigue and likelihood of disease progression, is used to determine long-term provisions for support. Long-term support includes introduction to Braille, audio-based learning and screen reading software programs, and Orientation and Mobility requirements to ensure future independence.

During the interview, AR acknowledged that a more comprehensive assessment of visual function had helped to improve her accessibility to educational materials, and the positive impact it had made transitioning to secondary education. AR commented that:

> "Because I have trouble with bright light and glare, the polarity is reversed on electronic documents so it's easier, and more comfortable, for me to read."

> "I am allowed extra time for homework and exams because it takes me longer to scan and read, especially if I have a lot of reading to do."

These comments support the use of reversed polarity text for AR and reading speed analysis provided a quantitative measure that enabled AR to be given the appropriate extra reading time for her work.

This case highlights the impact of specialist support and the need for investigative assessments which include functional aspects of vision impairment, so adequate provisions can be made for children to access their educational needs. While clinical tests are important to confirm diagnosis, subsequent assessments should be performed to assess the individual's functional visual performance which may not correlate directly with clinical tests such as visual acuity and visual fields. These measures can be used to help provide information around optimum accessibility requirements and future requirements that may be needed. This is especially important for children transitioning to high school where children with vision impairment can face additional accessibility obstacles and have difficulty accessing learning material at the same rate as children without a vision impairment.[9]

Future direction would benefit from a national standardised functional vision assessment to determine visual ability for children with a vision impairment. This comprehensive measure of functional visual performance, repeated periodically through education to provide for changes in vision and educational requirements, would optimise support and independent accessibility throughout education.

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Disclosure statement

No potential conflict of interest was reported by the authors.

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