Low-vision aids improve the visual performance of children with bilateral chorioretinal coloboma

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PURPOSE To quantify the improvements in visual performance for both distance and near tasks attained by children with bilateral chorioretinal coloboma (CRC) with use of low-vision aids (LVAs).

METHODS This was a hospital-based, cross-sectional, interventional case series of children with bilateral CRC. Demographic data were collected through a structured questionnaire and review of medical records. Distance and near best-corrected visual acuity, contrast sensitivity, and reading speed were evaluated with refractive correction alone and with the use of LVAs (Keplerian telescopes for distance; handheld magnifiers and a tinted lens [400 nm filter] for near). Effects are presented as medians of differences with 95% binomial-exact confidence intervals.

RESULTS Six children were included (median age, 11.5 years; range, 7-17 years), of whom 5 were already using LVAs on a daily basis. The use of a Keplerian telescope achieved a significant median improvement in distance best-corrected visual acuity of 0.75 logMAR (95% CI, 0.20-1.20). Contrast sensitivity was also improved across all tested spatial frequencies. Use of near LVAs resulted in a significant median improvement in near reading acuity of 0.47 logRAD (95% CI, 0.28-0.90). Critical print size and reading speed at N10 were also improved.

CONCLUSIONS LVAs enable meaningful improvements in the visual performance of children with bilateral CRC, allowing noteworthy increases in distance and near visual acuities as well as good reading speeds at small print sizes. (J AAPOS 2018;■:1-5)

Ocular colobomata are rare congenital abnormalities caused by defective optic fissure closure, which occur in isolation, or associated with other ocular (eg, the microphthalmia/anophthalmia/coloboma [MAC] spectrum) or systemic diseases (eg, CHARGE syndrome).[1] The uveal tract, retina, and optic nerve may be involved. Involvement of the retinal pigment epithelium, the retina, and the choroid, with or without extension to the macula and/or the optic nerve, is called chorioretinal coloboma (CRC).[2] CRC often causes significant visual impairment[3] that cannot be improved by medical or surgical treatment aside from amblyopia management, although surgical interventions may be warranted for cosmetic purposes[4] or to manage CRC comorbidities (eg, cataracts) or complications (eg, retinal detachment).[5,6]

Bilateral visual impairment can negatively affect childhood development and learning.[7] Reading can be improved with larger print sizes and appropriate refractive correction. Low vision aids (LVAs) include optical, nonoptical, and electronic devices that maximize visual perception by increasing magnification and/or improving contrast. These aids have the potential to increase the child's ability to perform high-acuity activities[8] and are a fundamental component of most low vision rehabilitation strategies.[9]

A community-based report has previously ascertained the need for LVAs in visually impaired children with bilateral MAC and provided gross measures of benefit.[10] However, data quantifying the improvements attained are lacking. This study aimed to quantify how much the visual performance for both distance and near tasks can be improved with the use of optical and nonoptical LVAs. We recruited children with bilateral CRC and tested whether distance visual acuity and contrast sensitivity improved with the use of a Keplerian telescope (KT) and whether reading performance improved with use of a handheld magnifier (HHM) with and without a tinted lens (400 nm filter [400F]).

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Subjects and Methods

This study was approved by the Institutional Review Board of Centro Hospitalar e Universitário de Coimbra (CHUC) and was performed in accordance with the tenets of the Declaration of Helsinki. All children that participated in the study gave their assent, and their parents or legal guardians provided informed oral and written consent.

Children ≤17 years of age diagnosed with bilateral CRC were recruited from the Low Vision Clinic at CHUC. Patients with unilateral CRC were excluded, as were bilaterally affected patients with best-corrected visual acuity in the better-seeing eye of 0 logMAR or better (Snellen equivalent 20/20). Patients were also excluded if they were <5 years of age or unable to complete the study protocol (eg, had not learned how to read). Demographic data were collected through a structured questionnaire; subjects' medical records were also reviewed.

Distance best-corrected visual acuity was determined using the Early Treatment Diabetic Retinopathy Study (ETDRS) chart (CSV-1000ETDRS, VectorVision, Greenville, OH), and results were recorded in logMAR notation. Failure to read any letters was assigned a value of 1.7 logMAR.[11] Using the best correction in a trial frame, each eye was tested separately. Only the distance best-corrected visual acuity of the preferred eye was tested with the aid of a monocular handheld KT (931965, 931975, 931975; ImproVision, Schweizer, Germany). KTs have a weak positive objective lens and a strong positive eyepiece lens that are separated by the sum of their focal lengths; they improve visual acuity by increasing angular magnification subtended by objects.[12] These are lightweight devices about 4–8 cm in length that are usually worn around the neck and held in front of the preferred eye to focus on objects at intermediate and far distances while the contralateral eye is closed. Testing started with a 4× magnification KT; higher magnification KTs (6×, 8×) were tested until a visual acuity of 0 logMAR was attained or if no further improvement was achieved.

Contrast sensitivity function was tested with the CSV-1000E (VectorVision), which is a printed chart, forced-choice paradigm test. It is a validated tool that has been used in various clinical studies.[13,14] The test presents sine wave targets of varying spatial frequency (3, 6, 12, and 18 cpd) at a distance of 2.5 m. Contrast was adjusted using the method of limits with eight discrete levels of contrast per spatial frequency; the lowest contrast available on the chart was 0.5%. We used the table list on the company's website (http://www.vectorvision.com/csv1000-norms/) to translate scores (ranging from 0 to 8) to logCS units for statistical analysis. All patients were tested without glare and monocularly, using the eye with the better-corrected visual acuity, with refractive correction and with the KT magnification with which they had achieved the best visual acuity.

Binocular near best-corrected visual acuity was measured using LEA numbers (270900, Good-Lite, Elgin, IL), and results were recorded in metric (M) notation. According to the techniques of Lovie-Kicthin and Whittaker,[15] the HHM with the most suited magnification (214002, ImproVision, Schweizer, Germany) was chosen. We also aimed to test whether the use of a 400F would provide additional benefit, because yellow filters have been shown to improve achromatic contrast[16] and reading performance.[17]

To assess reading performance, we used the Radner-Coimbra Reading Charts, a validated Portuguese version of the Radner Reading Charts (RRC), which have been shown to reliably assess the reading performance of low vision patients.[18,19] RRC consist of three charts with 14 sentences. The logarithm of reading acuity determination (logRAD) is the reading equivalent of logMAR.[19] The difference in size between two RRC sentences is 0.1 logRAD and, at a testing distance of 40 cm, print size ranges from 1.2 to −0.2 logRAD. Patients were assessed binocularly in three different testing conditions: (1) refractive correction only; (2) refractive correction and HHM; and (3) HHM with a 400F (Multilens, Sweden) over the refractive correction. Briefly, the reading test was performed at an exact distance of 40 cm on a reading stand with standardized lighting conditions (luminance was 80−90 cd/m^2). Testing was performed as previously described.[19] All testing procedures were recorded for later analysis. Reading time was measured with a stopwatch. Children included in this study were in different educational stages (from elementary to high school). Thus, we could not reliably set a time threshold (typically 30 sec per sentence for adult populations) to terminate the reading assessment. Instead, reading acuity, defined as the smallest print size at which some words can still be recognized, was computed in logRAD through the following formula: reading acuity = 1.3 − (sentences read × 0.1) + (number of syllables of incorrectly read words × 0.005). Reading speed was calculated as the quotient between the number of words in a sentence (14 words) by the time needed to read it (in seconds), multiplied by 60 to render the result in words per minute (wpm). Critical print size is the print size at which reading speed starts to deteriorate and was identified as the print size (in logRAD) of the sentence at which all the following (smaller) sentences were read at a speed 1.96 times the standard deviation below the average of the (larger) preceding sentences.[20]

When patients were not familiar with the LVAs used for testing, a training period of 10 minutes on different material was allowed. Near best-corrected visual acuity with LEA numbers was determined before the reading assessment was performed, and the KT magnification needed to achieve best visual acuity was ascertained before evaluating contrast sensitivity function. Otherwise, the order in which each task (and each testing condition) was performed was randomized for each patient to minimize bias due to learning effect or fatigue.

Finally, the anterior segment and the fundus were photographed using a Canon EOS 5D camera (DS126091) mounted on an FF450*plus* fundus camera (Zeiss, Germany).

Statistical Analysis

Demographic data are described with standard descriptive methods. Improvements are reported as the difference in outcome measures while using LVAs compared with the use of refractive correction only. Medians with 95% binomial-exact (conservative) confidence intervals are presented. Correlation between near visual acuity assessment with LEA numbers and near reading acuity with RRC was assessed using Spearman's rank correlation. For