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ARTICLE

Near vision in individuals with Down syndrome: a vision screening study

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Abstract

Objectives Children with Down syndrome are known to have reduced focusing ability for near vision (hypoaccommodation). Through a vision screening study we investigated the correlation between hypoaccommodation and near visual acuity in individuals with Down syndrome.

Methods A cross-sectional vision screening study was conducted on individuals with Down Syndrome. The screening was done in 4 city schools and 1 screening was conducted as a part of the Special Olympics Bharat program. In addition to the conventional vision screening tests, Nott dynamic retinoscopy was also performed. Both adults and children (age < 18 years) were included.

Results A total of 55 participants (33 children: age 6 to 17 years, 22 adults: age 18 to 41 years) with Down syndrome were screened. Twenty-two participants had visual impairment. Accommodative accuracy was assessed in 29 children and 13 adults. Accommodative lag (≥1.00D) was present in 12 children (41.37%) and 7 adults (53.84%). No correlation was found between the lag of accommodation and near visual acuity ($\rho_{Spearman} = 0.15$, $p = 0.54$). LogMAR near visual acuity was inversely correlated ($\rho_{Spearman} = -0.841$, $p < 0.001$) to the near viewing distance.

Conclusion Near visual acuity by itself is not a sensitive indicator of accommodative dysfunction. In addition, a closer viewing distance may not indicate adequate amplitude of accommodation. These findings strongly suggest the need for including dynamic retinoscopy in the clinical practice while examining individuals with Down syndrome.

Introduction

Every hour there are three babies born with Down syndrome in India [1]. Down syndrome (also referred as Down's syndrome) is the most commonly occurring chromosomal disorder

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(Trisomy 21) in newborns [2]. The incidence of Down syndrome in India ranges from 0.81–1.2 in 1000 live births [3, 4]. WHO reports worldwide incidence to be 1 in 1000 to 1 in 1100 [5]. Children with Down syndrome have intellectual disability and are at risk of several physical co-morbidities [6–9] including vision disorders. The commonly observed and reported vision disorders in children with Down syndrome are strabismus, refractive errors, nystagmus, cataract, keratoconus, and blepharitis [10–13]. In the past decade however, it has been reported that a large number of children with Down syndrome, with prevalence of 50% and higher have shown reduced accommodation or hypoaccommodation [10, 14–17].

Children are generally expected to have very good accommodation. Under this assumption, children are not usually tested for their accommodation in a routine clinical eye examination. Therefore, hypoaccommodation largely remains undiagnosed or under diagnosed particularly in children with Down syndrome who may also have difficulty in articulating their vision problems. When detected, hypoaccommodation is managed and in some children "cured" with bifocal spectacles [18, 19].

Bifocal spectacle prescription is a common management procedure for presbyopic adults. These adults have reduced accommodation due to the aging change of the crystalline lens. The symptoms in adults include near vision difficulties and the signs include poor near visual acuity and longer working/viewing distance in holding their reading materials [20]. While individuals with Down syndrome may not be symptomatic, the question we ask is do the signs reveal the difficulty? i.e., is there poor near visual acuity or longer working distance while looking at the printed materials in these individuals? Hypoaccommodation would have greater defocus for near therefore one can hypothesize that this would result in poorer near visual acuity or an increase in near working distance. We aimed to study this hypothesis by determining the correlation between near visual acuity, near working distance with hypoaccommodation in individuals with Down syndrome. While several studies have looked at distance visual acuity, near visual acuity as such has not been much commented upon.

A medical record review at our Institute in the past 5 years showed only 125 children with Down syndrome (DS) to have been examined (25 patients per year), which is abysmally low when compared to the incidence of DS in the city of Hyderabad [3, 21]. In addition, none of these children had bifocals prescribed, it is not known if these children required bifocals or not, as no indication of their accommodation status was mentioned. Near visual acuity was also commonly not documented in many patients from our records. Therefore, a prospective vision screening study was undertaken.

Materials (subjects) and methods

Vision screening was carried out in four schools for children with special needs in the city of Hyderabad, Telangana state and at Special Olympics Bharat, Visakhapatnam, Andhra Pradesh state. Children with intellectual disabilities typically go to special schools and inclusive education is a rarity in India unlike the developed countries [22, 23]. All the screening was conducted in the respective school premises. Only children with DS as identified by their school records were enrolled for the study. Prior to the screening, the school was informed about the research study, and the protocol and consent forms were circulated to the schools, which informed the parents. On the day of the screening, children whose parents agreed to be a part of the study were enrolled. Additional screening was performed alongside Special Olympics Bharat held in Visakhapatnam, Andhra Pradesh, India. Participants in the Special Olympics Bharat who were identified as having DS through the diagnosis mentioned in their medical certificates were enrolled. Informed signed consent was obtained from either parents or the legal guardians. The research protocol adhered to the tenets of the Declaration of Helsinki for research involving human subjects and the Institutional Review Board of L V Prasad Eye Institute approved the protocol.

Three optometrists (authors) conducted the one-day vision screenings, one of them (PNS) participated in the Special Olympics program at Visakhapatnam with a different team of optometrists trained to conduct the screening tests. Screening tests included case history, distance and near visual acuity, objective and subjective refraction, measure of accommodative lag, anterior segment examination with torch light and ocular motility examination. Distance visual acuity of the participant was tested using logMAR letter charts or logMAR LEA symbols. Near acuity was measured binocularly with word reading chart, number chart (from the near vision test book) or LEA symbols as per the ability of the participants and at their chosen comfortable viewing distance, which was measured and recorded. If the participant was non-verbal, they were asked to write the letters or point to the symbols on the key cards when possible. The near acuities were later converted into logMAR after taking into account the near working distances (see Table 1). In participants who could not perform any of the acuity tasks, LEA grating acuity was used. Light perception and projection of light direction

Table 1 Calculation of logMAR near visual acuity for different charts at different distances

Example 1: N16 read at 25 cm with Bailey-Lovie Word reading chart, the equivalent LogMAR is calculated as below:
$N_8 = 1.0$ M, therefore $N_{16} = 16/8 = 2.0$ M   At working distance of 0.25 m: MAR $= 2/0.25 = 8'$, LogMAR $= \log_{10}(8) = 0.9$
Example 2: 6/6 RS*(Reduced Snellen) chart read at 30 cm, the equivalent LogMAR is calculated as below:
Measured size of 6/6RS = 1 mm. Size of 1.0 M = 1.45 mm. Therefore 6/6 RS = 1/1.45 = 0.69 M   At working distance 0.3 m: MAR $= 0.69/0.3 = 2.3'$, LogMAR $= \log_{10}(2.3) = 0.36$

*The most commonly used near visual acuity chart in India is Near Vision Test Book (variety of suppliers), and the Reduced Snellen optotypes in this booklet are also commonly used when language is a barrier. The calculation for the conversion from this chart is based on direct measurement of the optotype size

was tested if a participant was non-responsive to any of these tests.

Objective and subjective refraction was performed when possible. The measured refractive error from the better eye was analyzed based on the criteria used by Das et al. [24]. for children with special needs. The criterion defines hypermetropia as spherical equivalent (SE) ≥ + 2.00D, myopia as SE ≤ −0.50D and astigmatism as ≥0.75D. In cases of only binocular acuity or no acuity being obtained, the eye with least (in magnitude) ametropia was considered for the analysis. For the purpose of measuring the accommodative lag, Nott dynamic retinoscopy [25] procedure was used with dim room illumination. In this procedure, the participant observed a target at 40 cm. The target was a non-illuminated "smiley face" toy that can produce sound when turned. This target (stimuli features varied in size from 4' to 103' of visual angle) was found to be more attention grabbing than a letter or picture (see Supplementary information). The target was mounted on a stand or was held in place by one of the examiners along with a measuring tape from the participant's eye or spectacle frame as the case may be. A single examiner (PNS), masked to the acuity measurements, performed the accommodative lag measurement. The examiner was aligned as close as possible to the same level or slightly below the toy to minimize off-axis retinoscopy. The examiner moved away from the participant when a 'with' reflex was observed and towards the participant when an 'against' reflex was observed and stopped at the distance at which neutrality was obtained. The appropriate lag (distance behind the target) or lead (distance in front of the target) of accommodation was calculated from the dioptric difference between the target distance (40 cm) and neutrality. In cases where the distance was beyond 110 cm, it was not possible to measure the lag of accommodation and it was recorded as >110 cm. While earlier studies [14, 15] have measured lag with targets held at much closer distances (e.g., 16 cm, 12.5 cm) for children with DS we chose to do it at a standard working distance of 40 cm adhering to the common clinical practice.

We first measured what we termed "presenting lag". The presenting lag was performed in a similar fashion as described above in the Nott dynamic retinoscopy but was performed with the participants own spectacle prescription (if present) or in case of uncorrected refractive error, without the refractive correction. The distance at which the first neutrality was obtained was measured (least hyperopic meridian). This measure gives the "uncorrected" lag/lead that a participant experiences during their near tasks. Later we measured the lag of accommodation with the objective refraction in place in a trial frame. In case of uncorrected hyperopic participants when the neutrality distance was beyond 110 cm, the presenting lag was re-

calculated by adding the measured lag of accommodation to their hyperopic refractive error (e.g., if a participant with an uncorrected + 3.00DS hyperopic refractive error exhibits a lag of accommodation of + 1.00DS over their objective refractive correction, then the presenting lag of accommodation for that participant was calculated as + 4.00DS). "High accommodative lag" was said to be present if participants exhibited a lag value ≥1.00D [26] in the eye with better acuity or least ametropic error (when acuity value was unavailable).

Participants in the school screenings requiring additional tests were referred to our tertiary eye care center. Participants requiring a spectacle prescription were also referred to the tertiary eye care center for cycloplegic examination. Those who participated in the Special Olympics–Opening Eyes program had a comprehensive eye examination and when required, spectacles were dispensed free of cost to them.

Data analysis: Descriptive statistical data analysis was performed using SPSS version 20.0 (IBM, SPSS). The measured outcome variables were checked for normality (one sample Kolmogorov-Smirnov test). If normally distributed, parametric tests were used else non-parametric test were applied to check for correlation between visual acuity and lag of accommodation.

Results

The vision screening was performed on a total of 55 participants (Table 2) that included 33 children (age < 18 years). No parent or guardian refused participation. Only 14 participants (25.4%) were prior users of spectacles out of which 1 had an ill-fitting spectacle frame. Distance visual acuity was measurable for 53 participants. The distance acuity charts used included logMAR letters (36 participants), LEA symbols (4 participants) and LEA gratings (13 participants). Monocular testing was possible only with 38 participants. Therefore the binocular presenting visual acuity was considered for the analysis. The mean presenting visual acuity (Fig. 1) recorded binocularly was 0.56 ± 0.34 logMAR (20/80) and twenty two participants (41.5%) had presenting visual acuity worse than 0.5 logMAR (20/60), which is the WHO definition for low vision [27]. The near visual acuity ranged between 0.24 and 1.74 logMAR (Snellen equivalent of 20/32 to 20/1099 at 40 cm).

A total of 34 (61.81%) participants (children = 20, adults = 14) were referred for further management of their ocular conditions (media opacities, glaucoma suspect, strabismus evaluation, watering and lid anomalies). Four out of six participants from Special Olympics Bharat-Opening Eyes program were prescribed spectacles, two of them were given bifocals.

Table 2 Demographics and refractive error of the study participants

Age	Years	
Mean (SD)	18 (9)	
Range	3.5–41.0	
Gender	n (%)	
Male	42 (76.4)	
Female	13 (23.6)	
Levels of intellectual disability*	n (%)	
Mild	15 (38.4)	
Moderate	19 (48.7)	
Severe	04 (10.2)	
Profound	01 (2.5)	
Refractive error**	n (%)	Range
Emmetropia	23 (43.4)	−0.38 DS to + 1.75 DS
Myopia	21 (39.5)	−0.50 DS to −12.88 DS
Hypermetropia	09 (17.0)	+2.00 DS to +4.50 DS
Astigmatism	30 (56.6)	−0.75 DC to −3.50 DC

*The level of intellectual disability is based on the IQ report given by the schools when available ($n = 39$)

**Hypermetropia and myopia includes participants having simple and compound astigmatism. Spherical equivalent values are reported in the range for all the refractive errors other than astigmatism
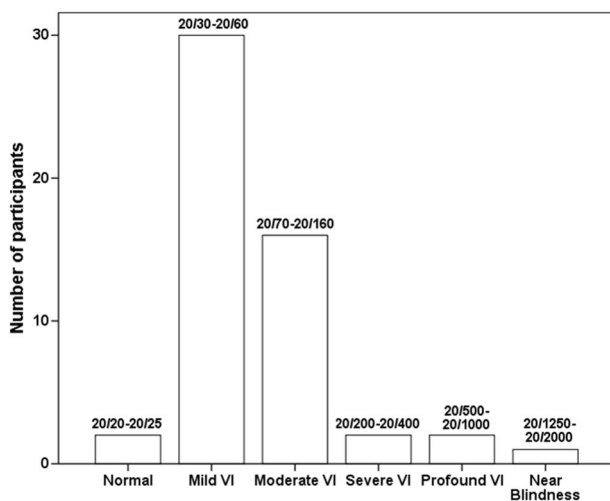

Fig. 1 Distribution of visual impairment levels for all the 53 participants based on presenting visual acuity as per WHO criteria [27]

Fig. 2 Box plot showing distribution of presenting lag and lag of accommodation in diopters in children and adults with Down syndrome [uncorrected myopes ($n = 13$) and adults over 35 years ($n = 4$) were excluded from the analysis]

The distribution of the estimated refractive errors is shown in Table 2. On classifying the astigmatism we found with the rule astigmatism was more common in 22 (73.3%) participants followed by against the rule astigmatism seen in 7 participants (23.3%) and one with oblique astigmatism.

Accommodative lag

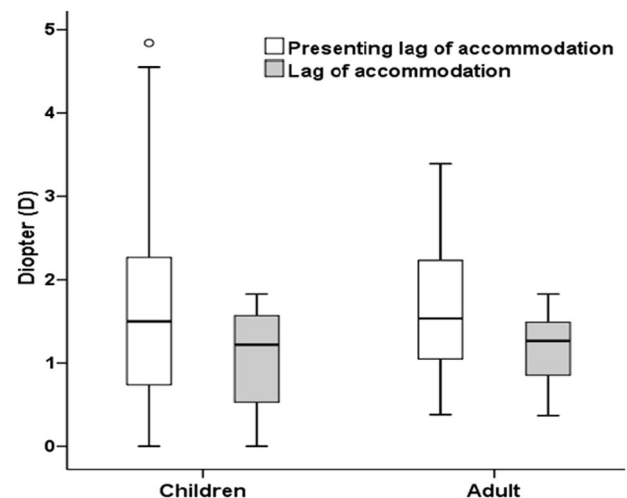Accommodative status was assessed in 29 children and 17 adults (Fig. 2). Four adults above 35 years had high lag as expected due to possible pre-presbyopia and were excluded from further analysis. Hence, the data from the remaining 13 adults was used for analyzing. High presenting lag of accommodation was observed in 16 (55.2%) children [median lag: 2.37D, IQR: 3.32D–1.50D] and 8 (61.5%) adults [median lag: 2.01D, IQR: 2.44D–1.59D]. High accommodative lag was present in 12 (41.37%) children [median lag: 1.52D, IQR: 1.35D–1.70D] and 7 (53.84%) adults [median lag: 1.50D, IQR: 1.62D–1.39D].

Correlation between visual acuity and presenting lag of accommodation

Correlation between visual acuity (both distance and near visual acuity) and presenting lag of accommodation was investigated. All the uncorrected myopes ($n = 13$) and adult over 35 years ($n = 4$) were excluded from this analysis. Analysis was done for participants with high presenting lag and without high presenting lag. No significant difference was found between the two groups for both distance (Mann Whitney U test, $P = 0.531$) and near (Mann Whitney U test, $P = 0.70$) visual acuities. No correlation was observed between the presenting lag of accommodation and measured near visual acuity, for groups with high presenting lag of accommodation ($\rho_{Spearman} = 0.20$, $P = 0.48$) and even when both the groups were pooled together ($\rho_{Spearman} = 0.15$, $P = 0.54$). A similar trend was found for lag of accommodation as well.

Correlation between viewing distance and presenting lag of accommodation

The near viewing distance showed a significant negative correlation ($\rho_{Spearman} = -0.841$, $P < 0.001$) to the logMAR near visual acuity (Fig. 3). This shows those with poor near visual acuity would tend to bring objects closer to view. Though statistically not significant, there was a moderate negative correlation observed between the near working distance and the lag of accommodation ($\rho_{Spearman} = -0.42$, $P = 0.23$). No correlation was observed between the presenting lag of accommodation and working distance ($\rho_{Spearman} = -0.008$, $P = 0.979$).

Discussion

We undertook a vision screening study in individuals with Down syndrome and found evidence for a high prevalence of hypoaccommodation (lag greater than 1.00D) in our study participants. With "presenting" lag criteria the prevalence was 55.2% in children and 61.5% in adults. With the "usual" lag of accommodation it was about 41% for children and 54% for adults. Earlier studies [10, 14–17] have found a higher prevalence in children, however reports on adults is scarce.

The primary focus of this study was to investigate the near visual acuity, near working distance and lag of accommodation relationship in participants with DS. The evidence that we found indicates that participants can have good near visual acuity despite having hypoaccommodation. This is contrary to what we hypothesized and also contradicts the findings of Lindstedt (1983) [28]. Lindstedt's study [28] triggered the investigation of
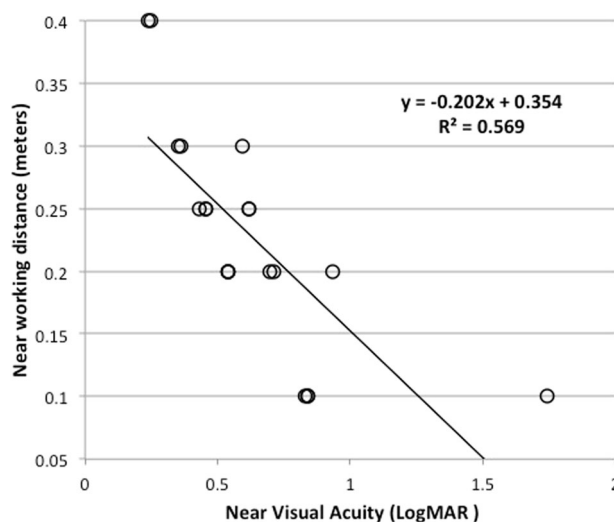

Fig. 3 Scatter plot showing correlation between near visual acuity (in logMAR) and near working distance [uncorrected myopes ($n = 13$) and adults over 35 years ($n = 4$) were excluded from the analysis]

accommodation research in Down syndrome when an observation for near visual acuity was found to be reduced, speculating a deficiency in accommodation [14–16, 29]. While we observed accommodation is not adequate for a large number of participants with DS, it did not correlate to the near visual acuity. Lindstedt's study [28] showed the ratio between distance to near acuity to be less than 1 in some patients with DS. In our study, we calculated this ratio and we did find 9 participants (31.03%) to have less than 1. However, we did not observe any significant correlation between this ratio and lag of accommodation (or presenting lag of accommodation).

This outcome i.e., lack of correlation between lag of accommodation to near acuity is important since it demonstrates that near visual acuity is not a sensitive detector for accommodative disorder. In addition, we also observed that those who have hypoaccommodation also tend to hold the reading material closer (also confirmed by clinical observations-Woodhouse personal communication, 2017), contradicting the common clinical belief of poor accommodation being associated with a longer near working distance (like in the cases of presbyopia). This finding is also important since it indicates a child holding a reading material at a closer distance may still have poor accommodation. It is common clinical practice to assume that children will have good accommodation especially when they read at a closer distance. Such an observation prevents the clinician to suspect any accommodative disorder in the child, especially when no symptoms are reported.

Good near visual acuity even if viewing the reading material at a closer distance, despite poor accommodation is unexpected. This paradox can perhaps be explained by factors such as pupil size and depth of focus. These factors

were not considered in this study. As closer working distance results in pupillary miosis, a better depth of focus can be achieved with some added benefit of relative distance magnification. It could also be possible that participants with DS may have accommodated differently for the target that was presented to measure the lag of accommodation and to that used for near visual acuity measurement. Future studies could aim to measure or sample continuous accommodative responses when a near visual acuity task is given to a participant. It could be possible that there are short bursts of accurate accommodation to focus the target clearly to read, but an accurate steady state accommodative response could be suboptimal. Such instabilities have been observed in infants [30].

The secondary outcomes showed that more than half of the participants ($n = 30$, 54.5%) had refractive errors, within which a large proportion of them ($n = 22$, 40%) did not have the needed spectacle correction. All children with special needs require visual assessment, as the prevalence of uncorrected refractive error is higher in this population [24, 31–33]. Unaddressed or inadequate refractive error correction in children with special needs and/or multiple disabilities is not uncommon among Indian population [34–36]. This is the case even in developed countries such as UK [32]. We found 41.5% of the participants in this study to have low vision. A pair of spectacles can easily rectify low vision caused by uncorrected refractive error. While no participant was found wearing bifocal spectacles, 14 had spectacles (including one who had an ill-fitting frame with broken nose pads and severe indentation marks on his nose. The teacher of this child was counseled about this child's condition, and was asked to request his parents to change his spectacles immediately). In a previous study [32] it was reported that uncorrected refractive error was the leading ocular condition followed by strabismus in individuals with DS. Previous studies have also reported myopia and astigmatism to be most common in individuals with DS [10, 13]. Our study also found a similar trend (Table 2). Previous studies [10, 37] reported oblique astigmatism to be common in individuals with DS, the prevailing hypothesis being oblique astigmatism is caused by the upward slanting of the palpebral fissure [10, 38]. We found with the rule astigmatism to be more common (73.3%) in participants followed by against the rule astigmatism (23.3%). It is unclear what might cause the lower occurrence of oblique astigmatism in our cohort. A larger sample size will be needed to investigate this trend further.

In the management of patients with DS, single vision spectacles are typically prescribed first. The status of accommodation is rechecked with the prescription at a follow-up appointment. If the lag of accommodation persists, bifocals are then prescribed [19]. In our study, we did not have adaptation time factored in to determine if the participants will accommodate accurately with just their single vision refractive correction. We acknowledge this as a limitation towards looking at the actual prevalence for hypoaccommodation. In addition, presenting lag values calculated for participants with uncorrected hyperopic refractive error could be exaggerated since it assumes their hyperopic refractive error is not compensated. Nevertheless, our computation of the 'presenting' lag points out that a large number of individuals (about 70.5%) may have difficulty performing near tasks and would need spectacle correction either for their hyperopic error alone or along with a bifocal correction for near vision tasks.

It has been shown that with adequate bifocal correction children with DS can start accommodating accurately [19]. The mechanism for this improvement is not known. If bifocals can 'kick start' the hypoaccommodated eye, it could be possible that home vision therapy exercises can also be attempted as one of the treatment modalities for hypoaccommodation in children with DS. Accommodative exercises recommended for symptomatic individuals with accommodation infacility and/or accommodation insufficiency has shown promising results in individuals without any systemic conditions [39]. Exploring vision therapy for individuals with DS may also be worthwhile.

In conclusion, our study found a majority of participants with Down syndrome to exhibit hypoaccommodation. The clinical measure of near visual acuity or the viewing distance may not be a good indicator of the accommodation status. Hence, performing dynamic retinoscopy should be a common clinical practice while examining these individuals.

Summary

What was known before

- Individuals with Down Syndrome tends to have a high lag of accommodation.
- Closer near viewing distance could indicate good accommodation, particularly in children.
- Measurement of near visual acuity is generally considered for prescribing near vision spectacles.

What this study adds

- Good near visual acuity and reading at closer distance may not always be indicators of adequate amplitude of accommodation.
- Accommodative accuracy should be checked even if individuals with Down syndrome have good near acuity.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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