

Accommodation deficit in children with Down syndrome: practical considerations for the optometrist

Julie-Anne Little

School of Biomedical Sciences,
University of Ulster, Coleraine,
Northern Ireland, UK

Abstract: Down syndrome is the most common cause of intellectual impairment, and life expectancy in this group has increased in recent decades, meaning that health care is increasingly focused on quality of life and the management of treatable illnesses. There are frequent problems associated with vision in Down syndrome, including refractive errors, strabismus, reduced vision, and reduced accommodative ability. This review will discuss the importance of accommodative ability; describe the prevalence and nature of accommodative deficits in Down syndrome, which are found in approximately 55%–76% of individuals; discuss the management of this deficit with the prescription of bifocal correction; and summarize the possible etiologies of hypoaccommodation in Down syndrome. Finally, the review will consider practical considerations for the optometrist managing accommodative deficits in patients with Down syndrome.

Keywords: Down syndrome, accommodation, accommodative deficits, dynamic retinoscopy, bifocals, refractive error

Introduction

Down syndrome is the most common cause of intellectual disability in humans, affecting approximately one in 500–600 births in the United Kingdom.¹ In addition to a higher risk of systemic diseases,² such as hypothyroidism, osteoporosis, and cardiac problems,³ there are frequent visual deficits associated with Down syndrome. These include refractive errors,^{4–7} strabismus,^{8,9} reduced vision,^{10,11} nystagmus,^{12,13} cataract,^{14,15} and reduced accommodative ability.^{9,16–18}

There has been a substantial increase in life expectancy in Down syndrome over the last number of decades, with average life expectancy reported in 2002 at 59 years in Australia,¹⁹ and 49 years in the United States.²⁰ Quality of life and the management of treatable illnesses are increasingly the focus of medical care in Down syndrome,²¹ especially with increased risk of dementia and respiratory failure.²² It has been reported that vision disorders can negatively impact cognitive function in individuals with Down syndrome.²³ Consequently, it is important for optometrists in primary care to have knowledge of this condition to be equipped for examination and the effective management of patients with Down syndrome in order to maximize these patients' visual capacity.

One particular aspect of visual function that can be effectively managed by optometrists is accommodative deficits. Woodhouse et al¹⁶ initially established that significant accommodative deficits were present in children with Down syndrome, and further work has established that this is not just a consequence of other ocular

Correspondence: Julie-Anne Little
School of Biomedical Sciences, University
of Ulster, Coleraine BT52 1SA,
Northern Ireland, United Kingdom
Tel +44 28 7012 4374
Email ja.little@ulster.ac.uk

conditions, such as uncorrected refractive error or poor visual acuity (VA). Accommodative deficits are particularly important as near tasks are central to development in childhood. Children with Down syndrome are considered visual learners,²⁴ and vision is a key sense contributing to the achievement of developmental milestones and accessing educational and recreational material. This article will describe the profile of accommodative deficits in Down syndrome and the management of this with bifocal correction. As accommodation and refractive error are closely linked, the review will begin by summarizing what is known about refractive error in this population.

Refractive errors in Down syndrome

Refractive errors are common in Down syndrome, and longitudinal refractive assessment on infants and young children report a failure to emmetropize.^{4,5,25} Large magnitudes of refractive error are common in Down syndrome, particularly hyperopic refractive errors,²⁶ and Woodhouse et al²⁷ note that refractive errors present in infancy in Down syndrome are often not only retained, but they increase. Prevalence of hyperopia has been reported to be as much as 55%,⁷ depending on the definitions used. Astigmatism is also more prevalent, with Little et al²⁸ reporting a 41% rate of astigmatism >0.50 DC and significantly steeper corneas. Mirrored oblique astigmatism is a frequent finding.²⁸ Again, depending on classification and the age of the children examined, myopia prevalence is reported to range between 8% and 25%,^{4,7,29} and Bromham et al²⁹ noted an association between congenital heart defects and the presence of myopia. It is important that refractive error is measured and monitored in Down syndrome and refractive correction prescribed when significant.

Accommodative function

The accommodative ability of the eye is vital for the human visual system to effectively see over a range of near distances. The retina is provided with a clear, sharp image through changes to the shape and thickness of the crystalline lens by contraction of the ciliary muscle. Accommodation develops early in infancy, with accurate accommodation displayed by the age of 6 months of life.^{30,31} Throughout childhood and early adulthood, accommodative response is typically unconscious and innate to our visual system, occurring in conjunction with vergence eye movements and pupil constriction. Early work from Donders³² in the 1860s and Duane³³ demonstrated large accommodative amplitudes in childhood. Consequently, accommodative function is not

routinely measured in children, as there is still the traditional clinical view that this is unnecessary. However, more recent research has challenged the convention that children always have ample reserves of accommodation.³⁴ With the advent of portable electronic technology increasing the burden of near work educationally and recreationally, it could be increasingly important to assess accommodation to investigate near visual status.^{35,36}

Uncorrected hyperopia places an obvious burden on accommodative performance, but while the status of accommodative function has been assessed in myopia as a precursor for myopia development,^{37,38} there has been limited work involving hyperopic status and accommodative deficits, and the exact magnitude of hyperopia leading to accommodative deficits and interplay with binocular functions is not well understood. Consequently, there is a lack of clinical guidance regarding prescribing for hyperopia, and considerable variation between the prescribing habits of individual optometrists. Interestingly, recent work has noted that even low amounts of uncorrected hyperopia can reduce visual performance,³⁹ and Shankar et al⁴⁰ reported that uncorrected hyperopes show evidence of avoiding reading tasks. However, it is important to note that this association does not imply causation – ie, we cannot determine whether the presence of uncorrected hyperopia leads to reading avoidance, or whether there are other factors contributing to this situation.

While optometrists are often familiar with the measurement of amplitude of accommodation, an individual's "typical" accommodative response to a target placed at a near distance may be more indicative of their typical function and accommodative exertion. This can be assessed through the technique of dynamic retinoscopy. Unlike the amplitude of accommodation measures where the examiner is dependent on the observer to report blurring of the target, dynamic retinoscopy is an objective technique, which can be undertaken for targets at several near test distances, and compared with normative findings. The accommodative response is the actual amount of accommodation exerted by the crystalline lens. The accommodative demand is the dioptric distance of the target from the eye, and for reasons of aberrations, depth of focus, etc (see Charman⁴¹), the accommodative response tends to be less than the demand. Rouse et al⁴² reported that an accommodative lag of up to 0.75 D could be considered "normal" for a child and, more recently, McClelland and Saunders⁴³ report that a lag of up to 1.00 D for a 25 cm target is within normal limits for a typically developing child.

The accommodative deficit in Down syndrome

The prevalence of accommodative deficits in Down syndrome is approximately 55%–76%.^{4,6,17,44} For children with Down syndrome, a subjective assessment of amplitude of accommodation is often not appropriate, and several groups have established the use of Nott dynamic retinoscopy as an effective, objective means at assessing accommodative status.^{43–45} These studies have used a slightly different criterion for categorizing accommodative deficits in Down syndrome, ranging from 0.75 to >1.00 D lag, but nevertheless, this approach is attractive, due to the objective nature of the test and the relatively straightforward task of gazing at a near target required of the observer. Details of how to carry out this technique are given in the next section of this review. Figure 1, adapted from Woodhouse et al,¹⁷ illustrates the deficit in accommodative response for a group of young children with Down syndrome (5 to 85 months) compared with age-matched typically developing children. The line of equality denotes the accommodative response matching the accommodative demand, and it is evident that individuals with Down syndrome, unlike controls, have a significantly reduced response at all test distances.

The convention of Woodhouse et al^{17,44} and Al-Bagdady et al⁴⁶ is to measure accommodation by dynamic retinoscopy at three accommodative demands, 4 D, 6 D, and 10 D. Other authors have assessed accommodation using different methods (eg, Haugen et al⁴), but all have consistently reported underaccommodation in Down syndrome, and also that as the accommodative demand increases, the accommodative deficit increases too.

Anderson et al¹⁸ used a photorefractive method to assess the dynamic accommodative response in Down syndrome and reported that only 13.5% of participants had a typical response. They also reported larger accommodative microfluctuations in comparison to controls.

Further research has investigated associations with other aspects of vision, but no clear pattern has emerged. Nandakumar and Leat⁴⁷ did not find a link between underaccommodation and hyperopia in Down syndrome, whereas Stewart et al⁹ reported a relation between poorer accommodative responses and high levels of hyperopia. These authors also reported that those with accommodative deficits were more likely to be strabismic, but Haugen and Høvdning⁸ failed to demonstrate this relationship in their study.

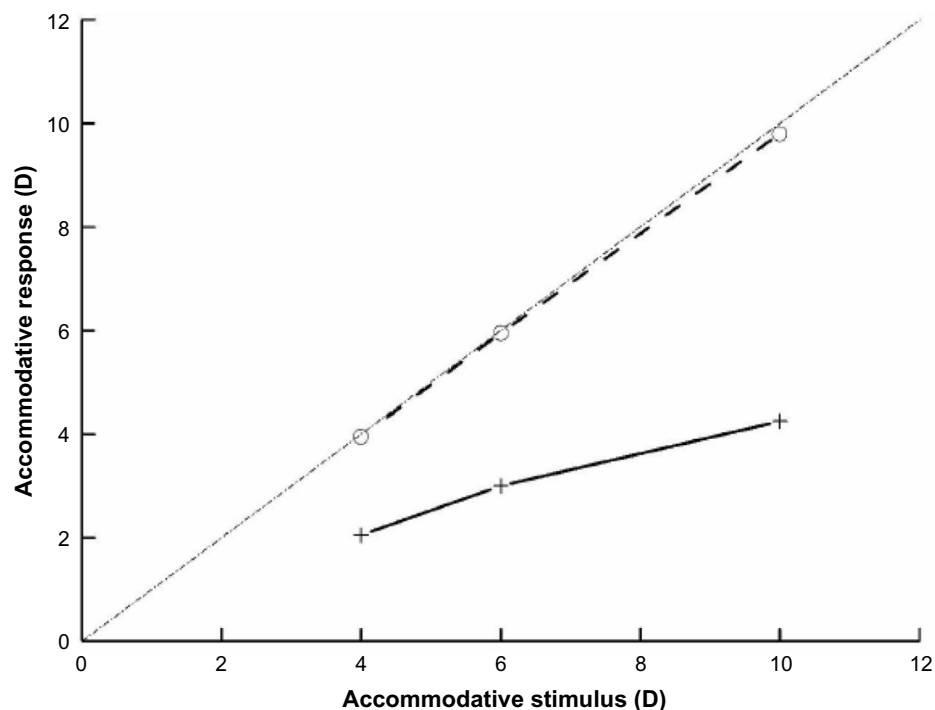


Figure 1 Scatterplot demonstrating accommodative deficit in Down syndrome.

Notes: Data points illustrate the mean accommodative response for three near targets at 4 D, 6 D, and 10 D demand for typically developing control children (aged 1 to 45 months) (O) and children with Down's syndrome (aged 5 to 85 months) (+). Reproduced from Woodhouse JM, Clegg M, Gunter HL, et al. The effect of age, size of target, and cognitive factors on accommodative responses of children with Down syndrome. *Invest Ophthalmol Vis Sci.* 2000;41(9):2479–2485.¹⁷

Abbreviation: D, diopter.

The clinical measurement of accommodation using dynamic retinoscopy

Nott dynamic retinoscopy uses the fact that if an individual is accommodating accurately on a target at near, when one observes the retinoscopy reflex holding the retinoscope beside the target, the reflex perceived will be “neutral”. If the individual has a lag of accommodative response, the reflex perceived will be a “with” movement. The examiner can then move the retinoscope away from the individual and target until a neutral reflex is observed and this distance can be recorded to quantitatively determine the lag of accommodation. Occasionally, an individual may demonstrate a “lead” of accommodation (with an “against” movement noted), and so in this case, the examiner would move the retinoscope closer to the eye until a neutral reflex is observed. To accurately determine accommodative performance, the individual will need to wear their full refractive correction for this procedure.

The monocular estimate method dynamic retinoscopy is a variation on the Nott method where plus (or minus) lenses are employed to neutralize the reflex rather than moving the retinoscope. This method, while useful, is arguably more complex to perform on an individual with Down

syndrome, as the lenses are often a distraction to attention to the target.

Figure 2 demonstrates the Nott dynamic retinoscopy technique with the Ulster–Cardiff accommodation cube (PA Vision Ltd, Ramsgate, UK). This instrument was recently developed to enable clinicians to easily measure accommodative function with Nott dynamic retinoscopy. The illuminated cube provides four cartoon targets of differing resolutions, to ensure that a target appropriate for a range of visual acuities can be accessed.

Accommodative norms have been reported by McClelland and Saunders⁴³ for three target distances – 25 cm (4 diopter (D) demand), 16.67 cm (6D demand), and 10 cm (10D demand) for children aged between 4 and 15 years. McClelland & Saunders⁴³ report the following lower 95% confidence limits for these three measurements, which gives the threshold beyond which a lag would be deemed to be significant: for the 10D demand, lag significant beyond 19.9 cm from the child; for the 6D demand, lag significant beyond 24.3 cm from the child; and for the 4D demand, lag significant beyond 34.0 cm from the child. The target distance of 25 cm (4D demand) is the most commonly used: so, for this demand, a significant lag of accommodation would be determined from dynamic retinoscopy if the examiner noted a “neutral” reflex at a distance of 34 cm or greater from the child. Adyanthaya et al,⁴⁸ in a retrospective analysis of clinical visual assessments in children with Down syndrome, noted that where accommodative deficits were found, they tended to be obvious lags of accommodation.

Management of the accommodative deficit

Accommodative deficits in Down syndrome can be managed with the prescription of bifocal correction. This pragmatic approach ameliorates the accommodative deficit and ensures near vision is optimized. In some cases, single vision near glasses are prescribed, but there are several reports of the success of bifocal correction in Down syndrome and that children with Down syndrome rapidly adapt to bifocals and use the appropriate portion for tasks in daily life. A recent study reported on the increased compliance with bifocal correction in Down syndrome compared with single vision correction,⁴⁸ which is in agreement with earlier work.^{6,47} There have been a number of longitudinal studies that have investigated bifocal prescribing, and have reported significantly improved accommodation, to accurate levels, when the accuracy of focus was measured through the bifocal segment.^{6,46,47,49} These studies

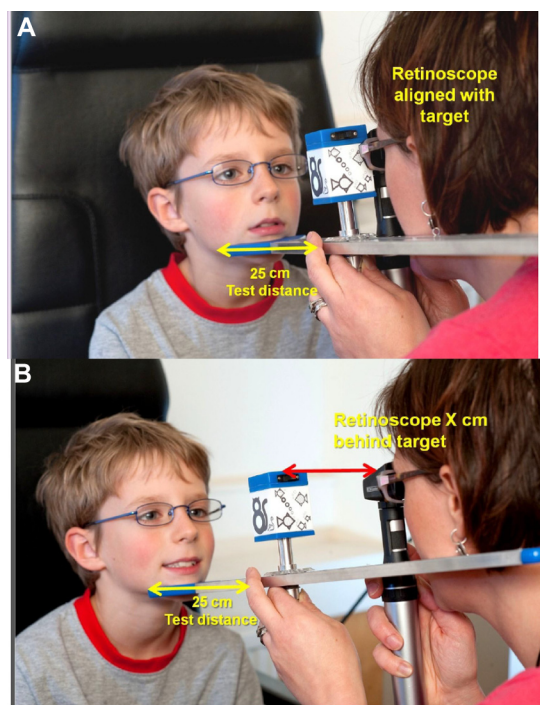


Figure 2 Photographs demonstrating the Nott dynamic retinoscopy technique. **Notes:** (A) Initial position of the examiner with the retinoscope aligned with the target for dynamic retinoscopy. (B) The examiner has moved the retinoscope further away from the eye to find a neutral reflex. The rule ensures that alignment is maintained near the observer's visual gaze to minimize off-axis errors.

assessed accommodative ability with dynamic retinoscopy and prescribed bifocals when a significant accommodative deficit was found. Nandakumar and Leat⁴⁷ and Nandakumar et al⁴⁹ also reported improvement in literacy skills once bifocal correction was given.

Nandakumar and Leat⁴⁷ used a bespoke reading addition depending on the amount of accommodative lag. Our clinic, in common with Woodhouse et al^{17,44} and Al-Bagdady et al,⁴⁶ prescribes a +2.50 DS near addition when an accommodative deficit outside the normative values is found. The position of the bifocal is important to ensure that the segment is accessed appropriately. For children with Down syndrome, the segment top is positioned at the pupil center and a flat-top bifocal design is recommended.⁶

Recently, Al-Bagdady et al⁴⁶ reported that the prescription of bifocals seemed to act as a treatment for accommodative deficits for a number of children with Down syndrome. In a longitudinal investigation, they reported that some children appeared to recover accommodative function, demonstrating accurate accommodation through the distance portion of their bifocal. These individuals were then returned to single vision distance glasses. This is intriguing, as it suggests that the accommodative deficit can be overcome once an individual is “shown” clear near vision, and learns to use their accommodation effectively. However, in their bifocal study in Down syndrome, Nandakumar and Leat⁴⁷ and Nandakumar et al⁴⁹ did not report any children where accommodative function recovered. Consequently, it is as yet unresolved as to whether bifocal correction could be considered an active treatment for accommodative deficits, or a passive correction, and these studies differed in their convention for refraction under cycloplegia or noncycloplegic methods. However, it is established that the accuracy of focus is improved with the prescription of bifocals in Down syndrome, and this provides significant benefits in terms of spectacle compliance and improving educational outcomes.

What is the etiology of accommodative deficits found in Down syndrome?

It is as yet unresolved why individuals with Down syndrome exhibit a failure to accommodate to near tasks. There are a number of explanations for this, including a mechanical or presbyopic-type deficit, or a sensory deficit.

It is known that structural aspects of the eye are different in Down syndrome, with a steeper, thinner cornea,^{28,50} and thinner crystalline lens.⁵⁰ This could mean that even with typical innervation of the ciliary muscle, the lens does not

change shape and increases in power to provide an appropriate accommodative response. However, Watt et al⁵¹ used derived lens power data from Haugen et al⁵⁰ to calculate the predicted reduction in response that a thinner lens could yield (based on the percentage change in lens power), and they only found a small theoretical 0.23 D deficit in accommodative response. Another possibility is that the crystalline lens in Down syndrome is stiffer, such as occurs in presbyopia, which could mean that the lens does not produce a response despite an input of effort. However, this is not supported by Clegg et al,⁵² who demonstrated that the accommodative response did not saturate, as would be found in presbyopia. In addition, Anderson et al¹⁸ noted increased microfluctuations in Down syndrome, and suggested that could indicate a more flexible crystalline lens.

Alternatively, sensory explanations of the deficit could involve differences in the accommodative/convergence relationship. The fact that esotropias are much more common in Down syndrome^{8,9} could support an abnormal neural control mechanism between the two systems. Recent work from our group has simultaneously investigated, for the first time, accommodation and convergence using binocular photorefractive measurements.⁵³ This has shown that accommodative deficits occur in conjunction with accurate and appropriate vergence eye movements, substantiating the theory of a difference between the neural control mechanisms for accommodation and convergence. The accurate vergence eye movements noted also refute the notion that accommodative deficits are merely an attentional issue. Further work could involve eliciting the role of blur and disparity cues in accommodative responses and examination of the crystalline lens under accommodative effort *in vivo*.

Practical considerations for the optometrist

While this review has focused on accommodative deficits, visual management of the individual with Down syndrome will compromise several other aspects of assessment. Along with an understanding of the refractive error profile in Down syndrome, it is worth noting that VA is often reduced, even with optimal refractive correction. Children with Down syndrome rarely achieve “6/6” or “20/20” levels of VA. Figure 3 illustrates this with data from Little et al.⁵⁴ This figure demonstrates VA (plotted against age) measured with the Lea symbols VA chart in a group of children with Down syndrome aged 6–16 years. These were compared with normative data from age-matched typically developing children, indicated by the black solid and dashed lines in Figure 3. These results

indicate that when using the Lea symbols VA chart, acuity that falls outside the fifth centile (the lowest dashed line) of these normative results is considered significantly poorer, and it is evident from Figure 3 that the majority of children with Down syndrome had acuity below the normative values.

The following sections are key aspects of visual assessment for consideration of accommodative deficits in the patient with Down syndrome.

Establish visual acuity (preferably monocular) distance and near where possible

It is important to obtain a measure of VA, both binocularly and monocularly, where possible. There are a variety of alternatives to the conventional VA charts used for adults – eg, the Keeler logMAR crowded acuity test, the Lea symbols acuity chart, or the crowded Kay picture test. Getting an assessment of near VA can also be useful. A clinician may find it takes more than one visit to be confident that a reliable measure of VA is elicited. For all types of visual assessment, it can be useful to document the degree of cooperation obtained. A simple qualitative score of good/moderate/poor cooperation can be a useful reference for interpreting visual findings at future visits.

Undertake dynamic retinoscopy to assess accommodative ability

As described earlier, the technique of dynamic retinoscopy is straightforward and quick, and with practice, a clinician can efficiently determine accommodative function.

Undertake cycloplegic refraction to know the full magnitude of hyperopia

Cycloplegic refraction is recommended to elicit knowledge of the full hyperopic correction and will likely produce more reliable results for the skilled retinoscopist. For Caucasian patients, one drop of 1% cyclopentolate hydrochloride is usually sufficient to yield effective cycloplegia, and after a time lapse of 30 minutes, cycloplegic retinoscopy can be performed. Even where no deficit in accommodation is found, children with Down syndrome are likely to benefit from the correction of hyperopic refractive errors, so consideration of refractive findings is important. Once the decision to prescribe has been made, our clinic typically employs the full hyperopic refractive correction. As emmetropization does not occur, the more frequent assessment of refractive error is indicated, typically on a 6-monthly or annual basis, depending on the child's age and necessary visual management. Astigmatism can be a frequent finding, and

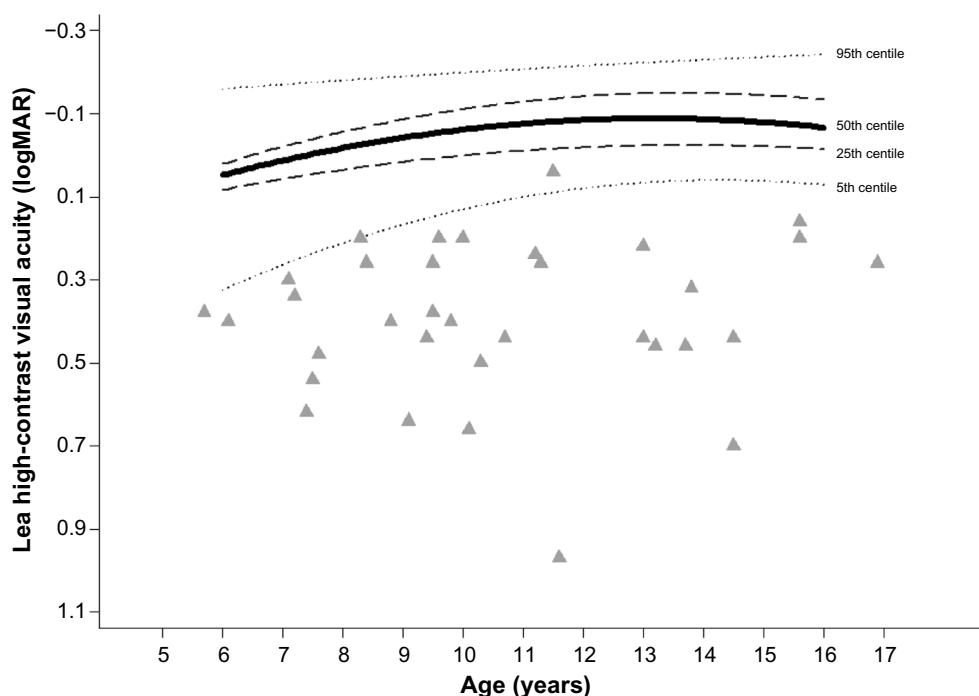


Figure 3 Scatterplot showing the normative data for children aged 6–16 years for the Lea acuity chart, indicated by black lines, and individuals with Down syndrome, indicated by gray triangles (n=29).

Notes: Visual acuity that falls outside the fifth centile of the normal results is considered significantly poorer than the norms. Copyright © 2015. Redrawn from Little JA, McCullough S, McClelland J, Jackson AJ, Saunders KJ. Low-contrast acuity measurement: does it add value in the visual assessment of Down syndrome and cerebral palsy populations? *Invest Ophthalmol Vis Sci.* 2013;54(1):251–257.⁵⁴ The copyright holder is the Association for Research in Vision and Ophthalmology.

accompanying corneal curvature information is valuable to capture and monitor any potential pathological changes in corneal shape, as keratoconus is more frequent in Down syndrome (8%–15%).^{55,56}

If prescribing glasses for the first time, parental and child education will be necessary to maximize compliance and success with spectacle wear. Resources for eye care professionals are available on the Ulster University Vision Resources Webpages, including a useful leaflet directed to parents encouraging spectacle wear.⁵⁷

If necessary, repeat dynamic retinoscopy assessment with full refractive correction once known

If a new or significantly changed refractive correction is prescribed, once adaptation has occurred and the child is compliant with spectacle wear, it will be necessary to re-evaluate accommodation with refractive error in place. It may be that the correction of distant hyperopic refractive error alone is sufficient to promote accurate accommodation, or that the child still exhibits a significant accommodative deficit even with refractive correction in place. In the latter case, near visual correction is then indicated, either with bifocals or another type of near refractive addition.

Consider the prescription of bifocals in cases where significant lag of accommodation exists

If the child requires near refractive addition due to hypoaccommodation, then bifocals are indicated, and recommended convention is a +2.50 DS reading addition using a wide D-segment, flat-top bifocal design, fitted at the pupil center. However, there may be patients with other lifestyle and educational considerations where single vision reading glasses or another progressive-type lens may be prescribed. The simplicity of the bifocal design, rather than a progressive lens design, has the benefit of enabling observation of when the bifocal segment is being viewed through. Finally, it may be the case that the child wears his or her bifocal correction on a full-time basis or, alternatively, some children may still maintain use of another pair of single vision distance glasses for other aspects of their daily life.

Monitor the accommodative status of each child

Whether an accommodative deficit has been found and corrected, or no significant hypoaccommodation has been noted, accommodation should be routinely assessed at the child's

visual assessments. If a bifocal or other form of reading addition has been prescribed for the first time, it is valuable not only to assess accuracy of focus through the bifocal segment at the time of collection, but also as the child adapts to wearing the bifocal. It is also valuable to follow up with the parents to elicit how the child is adapting to the bifocal, and to facilitate communication of the visual status and management of spectacle wear of the child to teachers and other people involved in their daily life, especially where there may be more than one pair of spectacles.

Conclusion

With an understanding of the ocular problems and accommodative deficits frequently found in Down syndrome, and practical tips regarding visual assessment, optometrists should be able to effectively monitor and manage these patients in their clinical practice. Over time, this can be an extremely rewarding and valuable area of clinical practice, fulfilling an important aspect of care for individuals with Down syndrome.

Acknowledgment

The photographs in Figure 2 are the author's own, and permission was granted from parent and child to use this image.

Disclosure

The author reports no conflicts of interest in this work.

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