insight

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SANKARA NETHRALAYA

Sankara Nethralaya – The Temple of the Eye.

It was in 1976 when addressing a group of doctors, His Holiness Sri Jayendra Saraswathi, the Sankaracharya of the Kanchi Kamakoti Peetam spoke of the need to create a hospital with a missionary spirit. His words marked the beginning of a long journey to do God's own work. On the command of His Holiness, **Dr. Sengamedu Srinivasa Badrinath**, along with a group of philanthropists founded a charitable not-for-profit eye hospital.

Sankara Nethralaya today has grown into a super specialty institution for ophthalmic care and receives patients from all over the country and abroad. It has gained international excellence and is acclaimed for its quality care and compassion. The Sankara Nethralaya family today has over 1400 individuals with one vision – to propagate the Nethralaya philosophy; the place of our work is an Alaya and Work will be our worship, which we shall do with sincerity, dedication and utmost love with a missionary spirit.

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Not Childs' Play!

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I started my career as a fellowship trained pediatric ophthalmologist at the cusp of the new millennium in 1999. Then there were only very few like me and the question which was frequently asked was what is there in Pediatric ophthalmology? Fifteen years down the line, I can confidently report that probably none of my junior colleagues have had to face that question.

In this millennium, Pediatric Ophthalmology has evolved in India as a standalone speciality encompassing both comprehensive eye care in children as well as adult strabismus. Interest in the speciality has grown with more institutes offering long term fellowships and a matching increase in applications. But it still falls way short of requirements of our country with 407 million children under 16 years of age. In a survey, Murthy *et al.* found that less than a third of respondent hospitals provided Pediatric eye care. Many centers providing services were hobbled by the absence of key components of Pediatric eye care like equipment, anesthesia facilities and support team consisting of optometrists, anesthetists and counselors. However, it is heartening to see that advanced eye care hospitals did much better and provided services on par with the developed world. In this millennium, I have great hopes that we will bring down the disparity in the Pediatric eye care between cities and smaller centers. The solution perhaps lies in having more general ophthalmologists with interest in Pediatric eye care along with specialists who can take care of more complex problems.

This issue reflects millennial changes in thinking and approach to some common problems which have generated interest and controversy among Ophthalmologists. Last words on these issues have probably not been said as yet but Sally Brown got it right in 1965 when she said "With nothing more than a simple eye patch we have brought amblyopia to its knees".

New and complex strabismus surgeries: exciting times!

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Strabismus surgeries have, in recent times, progressed at a rapid rate, with the introduction of a multitude of remarkable techniques that have defied the rules of classic surgery. With the advent of newer imaging modalities, and a better understanding of disease pathophysiology, surgical techniques are more targeted toward the specific muscles involved and its pathology. This article endeavors to outline some of the newer complex surgeries, their effectiveness and the basis behind them.

Surgeries for "Heavy Eye Syndrome"

"Heavy eye syndrome" or "myopic strabismus fixus convergence", is an acquired restrictive strabismus occurring in patients with pathologic myopia.[1] Patients often manifest extreme esotropia, hypotropia and restricted ocular motility. Simple recessions and resections have a limited role in this entity.

What's new ?

After the advent of orbital imaging, MRI studies showed supero-temporal prolapse of the elongated posterior portion of the eyeball, leading to an inferior shift of the lateral rectus (LR) and nasal shift of the superior rectus (SR) (Figure 1).[2,3]

Based on this new-found explanation, there was a paradigm shift in surgical techniques, which aimed more at vector mechanics, rather that simple muscle strengthening/weakening.

Loop myopexy

In 2001, Yokoyama[4] first proposed a new surgical idea, when he suggested to union the LR and SR together, and use it as a muscle pulley to push the herniated eyeball back into the muscle cone.

Technique

This technique is performed by isolating both the SR and LR muscles through either a superotemporal fornix based conjunctival incision or a limbalperitomy. Once the muscles are isolated, a 5-0 non-absorbable suture is passed through the lateral one-half of the SR and the superior one-half of the LR at ~14–16 mm posterior to the limbus and securely tied. Scleral fixation is not necessary. This eliminates the risk of scleral perforation. This technique can be done either with muscle splitting (e.g. partial Jensen's)[5–7] or without muscle splitting.[8–10] Other variants of this surgery include scleral fixation[11] and use of type 240 silicone band with Watzke sleeve[12] as a tethering aid instead of suture material. If preoperative forced duction testing (FDT) is positive or if the esotropia is long standing with likely contracture of the medial rectus (MR),

recession with the possible use of an adjustable suture can improve outcomes.[10,13] However, in the absence of these findings, MR recession may be staged for a second surgery.

Surgical success of <20 prism diopters (PD) postoperatively is seen in 73%, according to Shenoy et al.,[14] with mean improvement in esotropia of 60 PD and hypotropia of 9 PD.

Muscle transplantation

True muscle transplantation has been proposed for very large angle strabismus patients like heavy eye syndrome, especially when only one eye is planned to be operated upon.[15–17] This is of value if the FDT shows a tight MR in cases of long-standing esotropia, and can be combined with loop myopexy. Successful outcomes have been reported with >120 PD of esotropia and 40 PD of hypotropia getting corrected to <20 PD esotropia and 20 PD hypotropia after this procedure.[18]

Technique[18]

First the MR is dissected and separated via a fornix incision in the lower nasal quadrant. A non-absorbable 6-0 prolene suture is tied at its muscle insertion. The muscle is then incised from its insertion. Next the LR is hooked. Two 6-0 vicryl sutures are placed away from the insertion as is done in a routine resection of rectus muscle and another pair of 6-0 vicrylis placed at the insertion. The muscle is then incised from its insertion and the posteriorly (distally) placed 6-0 vicryl sutures are passed through the original insertion as in a routine rectus muscle resection. The stump is then cut and placed at the MR site and the distal end of this stump is sutured with the proximal end of MR with the 6-0 prolene already placed on the MR. The now elongated muscle is sutured from the original insertion site of MR as is done in a routine rectus muscle recession.

Paralytic strabismus

Complete muscle palsies, where there is no residual muscle function, do not respond to regular strengthening procedures. Recently, a whole gamut of surgeries has been advocated, with the basis of transposing the other available functioning muscles to serve the purpose of the palsied muscle.

Complete sixth nerve palsy
Vertical rectus transpositions

Vertical rectus transpositions (VRT), coupled with the antagonist muscle weakening, has showed promising results. The main purpose of a VRT is
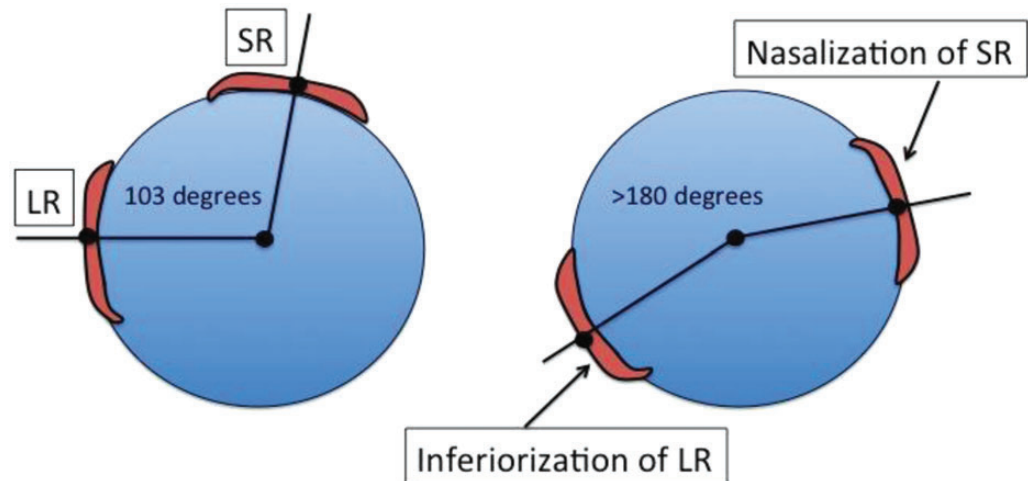
Figure 1: Shows the inferior shift of LR and nasal shift of SR, increasing the angle between them to . 180° (normal angle: 103°).

to allow better rotation of the eye into the field of the palsied muscle by creating tone through the transposed muscles in primary position.[19]

They also increase the area of binocular vision and shift the binocular field toward the palsied gaze. The transposed muscles do not have active innervation in the field of gaze of the palsy, thereby allowing the antagonist weakening procedure to have better effect.[20]

The most commonly performed VRT is transposition of the SR and IR to the palsied LR. Rosenbaum describes reattachment of the temporal border of the transposed muscle adjacent to the LR.[21] The nasal border is reattached following the spiral of Tillaux.

Results

VRT of both vertical recti without antagonist recession or posterior fixation can correct on average 32 PD of esotropia.[22] The binocular field increases from 25° to 41–51°.

Augmentation of VRT:

1. The partially transposed muscle can be resected symmetrically prior to transposition.[24–26] This augments the surgery by another 10 PD.[25]

2. Foster's posterior fixation suture:
    As originally described by Foster,[27] a single-armed permanent suture can be placed in the sclera 16 mm from the limbus at superior border of the LR. This suture is then passed 8 mm from the insertion of the SR and IR muscle incorporating ~25% of the transposed muscle. FDT at the conclusion of the transposition with posterior fixation should be free. This can correct an average of 40–55 PD of esotropia in primary and increases the degrees of binocular field to 71°.[27,28]

3. Mehendale[29] describes a loop myopexy to close the gap in the transposition procedure in place of the posterior fixation suture. This corrects ~30–35 PD of esotropia for only the SR.

4. Botulinumtoxin:
    Injection of botulinum toxin into the ipsilateral MR at the conclusion of VRT or in the immediate postoperative period[30,31] can increase the surgical effect to 30–50 PD.

5. To prevent induced torsion:
    The 12 o'clock and 6 o'clock positions on the cornea are marked preoperatively.[32] Assessing the markings on table following transposition may reveal a torsional shift, which may accompany an induced vertical deviation. Posterior fixation sutures can then be loosen edintraoperatively to relieve the torsion created and prevent vertical misalignment.

Nishida's partial tendon transposition procedure

This is a ciliary vessel sparing surgery, where a 6-0 polypropylene monofilament fiber suture or a 5-0 polyester braided fiber suture is inserted through the temporal muscular margin of each vertical rectus muscle at a distance of 8–10 mm behind the muscle insertion, at approximately one-third of the width from the edge. The sutures should avoid the ciliary vessels in their bite. The scleral bite is then taken at a distance of 10–12 mm behind the limbus at the superotemporal or inferotemporal quadrant, thustransposing the vertical muscles without tenotomy or muscle splitting.The surgical correction by muscle transposition alone ranged from 24 to 36 PD, and that by muscle transposition and recession of the MR muscle ranged from 50 to 62 PD [33] (Figure 2a and b).
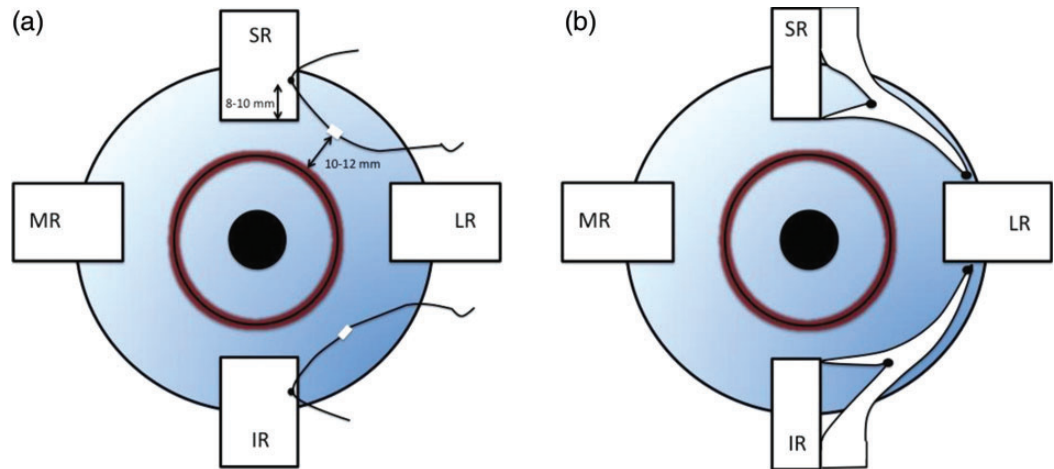
Figure 2: Nishida's Procedure showing aligning of SR and IR to the LR vector.

Complete third nerve palsy
Periorbital fixation of LR

This technique involves complete disinsertion of the LR tendon away from the globe and neutralizing its action by fixing it to the lateral orbital wall.

Technique

After exposing and isolating the LR, blunt dissection is performed to expose the adjacent periosteum ~5 mm posterior to the orbital rim, outside the muscle cone. A 6-0 non-absorbable monofilament suture is used to tag the LR, which is then disinserted from the globe. It is then attached to the adjacent orbital periosteum with two periosteal bites. In a study by Velez et al., this procedure was performed in 7 subjects of complete oculomotor nerve palsy, and their postop deviation was within 12 pd[34] (Figure 3).

Periosteal anchoring of MR

This procedure mechanically pulls the globe away from abduction and fixes the eye in the desired position of alignment with the help of a non-
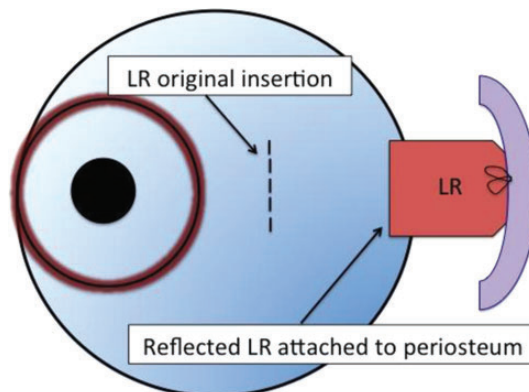

Figure 3: Surgial technique showing the LR muscle fixed to periosteum of the lateral orbital wall.

absorbable suture between the MR insertion and the periosteum of the anterior lacrimal crest.

Technique

Once the MR is isolated, a 15-mm long vertical transcutaneous incision is made directly over the anterior lacrimal crest down to bone depth. The insertion of medial palpebral ligament (MPL) is exposed by blunt dissection. The two arms of a double armed 5-0 non-absorbable coated braided polyester suture is passed through the superior and inferior part of MPL insertion at the anterior lacrimal crest including periosteum. The suture ends are tied to each other. Then either an empty semicircular needle[35] or an artery forceps[36] is passed under the skin and subcutaneous tissue toward the conjunctival side and used as a guide to bring out the two needles of the suture onto the conjunctival side. The sutures are then tied at the insertion of the MR directly onto the sclera, at the same time as the assistant rotates the globe medially (Figure 4). A slight overcorrection is desirable, as the sutures tend to get loosened with time.A case series by Sharma et al.,[36] of 4 subjects with a mean of 90 pd exotropia, showed satisfactory outcomes of <10 pd exotropia at 6 months following this technique, combined with supramaximal LR recession.

Y split and medial transposition of LR muscle

This was first suggested by Taylor[37] and later modified by Kaufmann[38] and Gokyigit and colleagues.[39]

Technique

A 300° conjunctivallimbalperitomy is made. The LR muscle is split up to 15 mm toward the posterior septum. One or two full-thickness locking bites are placed at the edge of the muscle halves using non-absorbable 6-0 polyester sutures. The muscle halves are disinserted and the sutures on
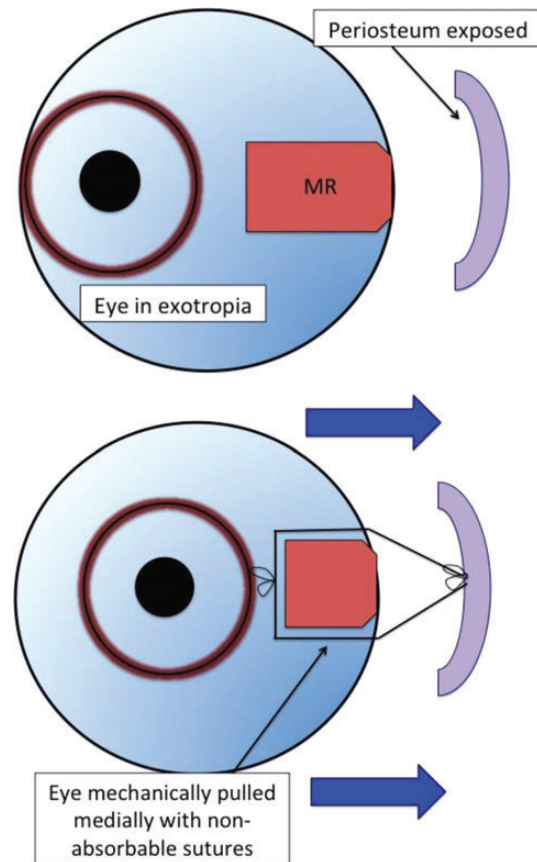
Periosteum exposed

MR

Eye in exotropia

Eye mechanically pulled medially with non-absorbable sutures

Figure 4: Steps of the technique of periosteal anchoring of MR with empty semi-circular needle.

the muscle halves are passed through the hole of the Gass hook. Then the upper half of the muscle is passed under the superior oblique tendon behind the insertion with the help of the Gass hook and the inferior half of the muscle is passed between the sclera and both the IR and inferior oblique muscles. The lower and upper halves of the LR muscle are reattached 1 mm posterior to the inferior and superior border of the MR insertion, respectively (Figure 5). The mean
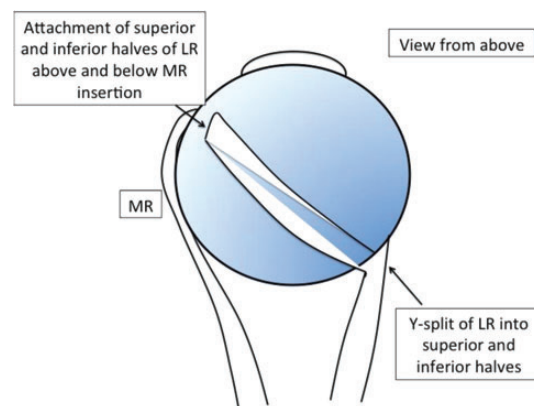

Attachment of superior and inferior halves of LR above and below MR insertion

View from above

MR

Y-split of LR into superior and inferior halves

Figure 5:

improvement in exotropia ranges from 73 PD to 8 PD postoperatively.[39] The same procedure has also been described on adjustable sutures, with successful outcomes.[40] Combining this technique with MR resection showed promising results, as compared to Y-splitting alone.[41]

Restrictive strabismus
VRT for Duane's retraction syndrome
VRT has been described for Duane's retraction syndrome (DRS) also, with highly successful outcomes.[26] In a study by Akar et al.,[28] 40 eyes with esotropic DRS underwent VRT. The deviation was reduced by a mean of 95%; Abnormal head posture was eliminated in 86%; Abduction improved by 42%, and a useful binocular single field of 67% was achieved at the end of 1 year.

Surgery for scarred conjunctiva
Restrictive strabismus due to fibrotic conjunctiva following periocular surgery such as pterygium-removal, can cause debilitating diplopia despite being a small-angle strabismus.

Amniotic membrane transplantation
Amniotic membrane transplantation seems to help prevent recurrence of adhesions in patients with restrictive strabismus caused by conjunctival scarring, fat adherence syndrome or rectus muscle contracture.[42]

Conjunctival mini-flap surgery
This was first introduced by Akura et al.[43] The procedure can be done under topical anesthesia and involves dissection of conjunctival hyperplasia and adhesiolysis. A tongue shaped flap is then fashioned from the adjacent virgin conjunctiva, and it is rotated and sutured such that it fully covers the bare sclera.

Minimally invasive strabismus surgery
Propounded by Gobin,[44] and later popularized by Mojon,[45] this is a technique of minimal access, wherein the conjunctival incision sites are essentially key-hole, placed strategically adjacent to the muscles to be operated. This reduces the post-operative irritation, dellen formation and improves cosmesis on first postoperative day. This principle of access has been adapted and developed further to allow to perform all types of strabismus surgeries, namely, rectus muscle recessions, resections, plications, reoperations, retro equatorial myopexias, transpositions, oblique muscle recessions and plications, and adjustable sutures. The incisions are 1–2 mm radial parainsertinal cuts, and may range in number anywhere between 2 and 6, depending on the type of surgery[45] (Figure 6). Another technique of trans-conjunctival muscle reinsertion has also been described by him, where the needle is passed
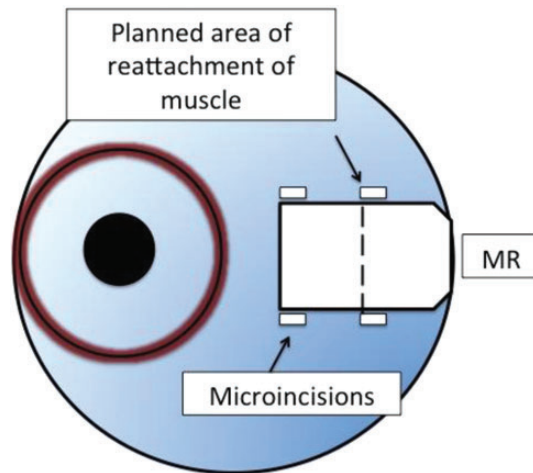
Figure 6: Surgical technique shows minimally invasive strabismus surgery for simple muscle recession, with two key-hole openings made at the original insertion of the muscle, and two more at the desired insertion posteriorly. The sutures are guided through a tunnel created subconjunctivally.

through the conjunctiva, and the sutures retrieved from the subconjunctival space.[46]

Conclusion

Great strides are being made in the field of strabismus surgery in recent years, with iconic innovators changing the way we look at strabismus surgery. There have been paradigm shifts in the approach to complex strabismus, based on the solid foundation of reasoning and better understanding of disease pathophysiology. Novel techniques have been aimed at improving consistency and predictability in outcomes. As a strabismologist, these are exciting times indeed!

References

1. **Taylor R**, Whale K, Raines M. The heavy eye phenomenon: orthoptic and ophthalmic characteristics. *German J Ophthalmol* 1995; **4**: 252–255.
2. **Aoki Y**, Nishida Y, Hayashi O, *et al.* Magnetic resonance imaging measurements of extraocular muscle path shift and posterior eyeball prolapsed from the muscle cone in acquired esotropia with high myopia. *Am J Ophthalmol* 2003; **136**: 482–489.
3. **Yokoyama T**, Tabuchi H, Ataka S, *et al.* The mechanism of development in progressive esotropia with high myopia. In: de Faber JT, ed. *Transactions of the 26th meeting European Strabismological Association.* Barcelona: Swets & Zeitlinger; 2000: pp. 218–221.
4. **Yokoyama T**, Ataka S, Tabuchi H, Shiraki K, Miki T. Treatment of progressive esotropia caused by high myopia–a new surgical procedure based on its pathogenesis. In: de Faber J-T, ed. *Transactions: 27th Meeting, European Strabismological Association*, Florence, Italy, 2001. Lisse (Netherlands): Swets & Zeitlinger; 2002: pp. 145–148.
5. **Larsen PC**, Gole GA. Partial Jensen's procedure for the treatment of myopic strabismus fixus. *J AAPOS* 2004; **8**: 393–395.
6. **Ahadzadeghan I**, Akbary MR, Ameri A, *et al.* Muscle belly union for treatment for myopic strabismus fixus. *Strabismus* 2009; **17**: 57–62.
7. **Ho TH**, Lin MC, Sheu SJ. Surgical treatment of acquired esotropia inpatients with high myopia. *J Chin Med Assoc* 2012; **75**: 416–419.
8. **Wong I**, Leo SW, Khoo BK. Loop my opexy for treatment of myopic strabismus fixus. *J AAPOS* 2005; **9**: 589–591.
9. **Rowe FJ**, Noonan CP. Surgical treatment for progressive esotropia in the setting of high myopia. *J AAPOS* 2006; **10**: 596–597.
10. **Durian JM**, Maddula S, March IB. Treatment of 'heavy eye syndrome' using simple loop myopexy. *J AAPOS* 2010; **14**: 39–41.
11. **Leo SW**, Del Monte MA. Surgical correction of myopic strabismus fixus by modified loop transposition with scleral myopexy. *J AAPOS* 2007; **11**: 95.
12. **Shenoy BH**, Sachdeva V, Kekunnaya R. Silicone band loop myopexy in the treatment of myopic strabismus fixus: surgical outcome of a novel modification. *Br J Ophthalmol* 2015; **99**(1): 36–40.
13. **Morad Y**, Pras E, Goldich Y, *et al.* Surgical treatment of esotropia associated with high myopia: unilateral versus bilateral surgery. *Eur J Ophthalmol* 2010; **20**: 653–658.
14. **Bhamy Hariprasad Shenoy**, Virender Sachdeva, Ramesh Kekunnaya. Silicone band loop myopexy in the treatment of myopic strabismus fixus: surgical outcome of a novel modification. *Br J Ophthalmol* 2015; **99**: 36–40.
15. **Hiatt RL.** Extraocular muscle transplantation. *J Pediatr Ophthalmol* 1977; **14**: 79–92.
16. **Amitava AK**, Goswami AK, Mishra A. Large-angle strabismus and primary true muscle transplantation. *J Pediatr Ophthalmol Strabismus* 2005; **42**: 211–215.
17. **Diamond GR.** True transposition procedures. *J Pediatr Ophthalmol Strabismus* 1990; **27**: 153–156.
18. **Jethani J**, Amin S. Loop myopexy with true muscle transplantation for very large angle heavy eye syndrome patient. *Indian J Ophthalmol* 2015; **63**: 71–12.
19. **Gunton KB.** Vertical rectus transpositions in sixth nerve palsies. *Curr Opin Ophthalmol* 2015; **26**(5): 366–370.
20. **Miller JM**, Demer JL, Rosenbaum AL. Effect of transposition surgery on rectus muscle paths by magnetic resonance imaging. *Ophthalmology* 1993; **100**: 475–487.
21. **Rosenbaum AL.** Costenbader Lecture. The efficacy of rectus muscle transposition surgery in esotropic Duane syndrome and VI nerve palsy. *J AAPOS* 2004; **8**: 409–419.
22. **Bansal S**, Khan J, Marsh IB. Unaugmented vertical muscle transposition surgery for chronic sixth nerve paralysis. *Strabismus* 2006; **14**: 177–181.
23. **Yamaguchi M**, Yokoyama T, Shiraki K. Surgical procedure for correcting globe dislocation in highly myopic strabismus. *Am J Ophthalmol* 2010; **149**: 341–46.
24. **Brooks SE**, Olitsky SE, de BRG. Augmented Hummelsheim procedure for paralytic strabismus. *J Pediatr Ophthalmol Strabismus* 2000; **37**: 189–195.
25. **Couser NL**, Lenhart PD, Hutchinson AK. Augmented Hummelsheim procedure to treat complete abducens nerve palsy. *J AAPOS* 2012; **16**: 331–335.
26. **Hendler K**, Pineles SL, Demer JL, *et al.* Adjustable augment edrectus muscle & transposition surgery with or without ciliary vessel sparing for abduction deficiencies. *Strabismus* 2014; **22**: 74–80.
27. **Foster RS.** Vertical muscle transposition augmented with lateral fixation. *J AAPOS* 1997; **1**: 20–30.
28. **Akar S**, Gokyigit B, Pekel G, *et al.* Vertical muscle transposition augmented & with lateral fixation (Foster) suture for Duane syndrome and sixth nerve palsy. *Eye (Lond)* 2013; **27**: 1188–1195.
29. **Mehendale RA**, Dagi LR, Wu C. Superior rectus transposition and medial rectus recession for Duane syndrome and sixth nerve palsy. *Arch Ophthalmol* 2012; **130**: 195–201.

30. **Flanders M,** Qahtani F, Gans M, Beneish R. Vertical rectus muscle transposition and botulinum toxin for complete sixth nerve palsy. *Can J Ophthalmol* 2001; **36**: 18–25.

31. **Rosenbaum AL,** Kushner BJ, Kirschen D. Vertical rectus muscle transposition and botulinum toxin (Oculinum) to medial rectus for abducens palsy. *Arch Ophthalmol* 1989; **107**: 820–23.

32. **Holmes JM,** Hatt SR, Leske DA. Intraoperative monitoring of torsion to prevent vertical deviations during augmented vertical rectus transposition surgery. *J AAPOS* 2012; **16**: 136–140.

33. **Muraki S,** Nishida Y, Ohji M. Surgical results of a muscle transposition procedure for abducens palsy without tenotomy and muscle splitting. *Am J Ophthalmol* 2013; **156**(4): 819–824.

34. **Federico G Velez,** Neepa Thacker, Michelle T Britt, Deborah Alcorn, R Scott Foster, Arthur L Rosenbaum. Rectus muscle orbital wall fixation: a reversible profound weakening procedure. *J AAPOS* 2004; **8**: 473–480.

35. **Kuldeep Kumar Srivastava,** Kannan Sundaresh, Perumalsamy Vijayalakshmi. A new surgical technique for ocular fixation in congenital third nerve palsy. *J AAPOS* 2004 **84**.

36. **Pradeep Sharma MD,** Madhurjya Gogoi MD, Sachin Kedar MD, Rahul Bhola MD. Periosteal fixation in third-nerve palsy. *J AAPOS* 2006; **10**: 324–327.

37. **Taylor JN.** Surgical management of oculomotor nerve palsy with lateral rectus transplantation to the medial side of the globe. *Aust N Z J Ophthalmol* 1989; **17**: 27–31.

38. **Kaufmann H.** "Lateralis splitting" in total oculomotor paralysis with trochlear nerve paralysis. *Fortschr Ophthalmol* 1991; **88**: 314–16.

39. **Gokyigit B,** Akar S, Satana B, Demirok A, Yilmaz OF. Medial transposition of a split lateral rectus muscle for complete oculomotor nerve palsy. *J AAPOS* 2013; **17**: 402–410.

40. **Ankoor S Shah,** Sanjay P Prabhu, Mohammad Ali A Sadiq, Iason S Mantagos, David G. Adjustable nasal transposition of split lateral rectus musclefor third nerve palsy. *JAMA Ophthalmol* 2014; **132**(8): 963–969.

41. **Jaspreet Sukhija,** Savleen Kaur, Usha Singh. Nasal lateral rectus transposition combined with medial rectus surgery for complete oculomotor nerve palsy. *J AAPOS* 2014; **18**: 395–396.

42. **Yi Ning J Strube,** Francisco Conte, Claudia Faria, Samuel Yiu, Kenneth W Wright. Amniotic membrane transplantation for restrictive strabismus. *Ophthalmology* 2011; **118**: 1175–1179.

43. **Akura J,** Kaneda S, Matsuura K, Setogawa A, Takeda K, Honda S. Measures for preventing recurrence after pterygium surgery. *Cornea* 2001; **20**(7): 703–707.

44. **Gobin MH,** Bierlaagh JJM. Chirurgie horizontal eetcyclo vertical esimultanée du strabisme. Centrum voorStrabologie: Anvers, Belgium; 1994.

45. **DS Mojon;** Review: minimally invasive strabismus surgery. *Eye* 2015; **29**: 225–233.

46. **Mojon DS.** A new transconjunctival muscle reinsertion technique for minimally invasive strabismus surgery. *J Pediatr Ophthalmol Strabismus* 2010; **47**: 292–296.

30 *Glorious Years*

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Thanks to all the past editors of Insight

Progressive myopia: an update

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Progressive myopia is a major concern to eyecare professionals all over the world. A simple search on popular search engines for "myopia progression" yields over 380,000 results. A Pubmed search yields. Myopia is a global health problem associated with not only vision impairment but also blinding complications. It is a significant economic burden too.[1] In Singapore mean annual cost of myopia for a child 7–9 years of age is $148. In US annual direct cost of correcting distance vision impairment due to refractive errors is between US $3.9 and US$7.2 billion.[2]

Myopia also poses a significant medical burden with increased incidence of glaucoma and cataracts in those myopic individuals.[3] There is also a higher incidence of blinding retinal complications due to choroidal neovascular membranes, not to mention the impaired quality of life.

Prevalence
Studies in adults have found myopia prevalence ranging from 19.4% in Taiwan to 41.8% in Japan with figures from Singapore and China falling in between.[4–9] The population-based studies in children have found myopia prevalence ranging from 1.2% in Nepal to 42.4% in China.[10,11] Studies from the early 2000s from India have quoted prevalence figures of 7.4% by Murthy *et al.* and 4.1% by Dandona *et al.*[12,13]

A recent study by Saxena *et al.* looked at prevalence of myopia in Delhi. Amongst a total of 9884 school children screened the prevalence of myopia was 13.1% with only one-fourth of those wearing appropriate spectacles.[14] A look at figures from Asian countries have shown a sharp increase in prevalence in the last 20 years.

Risk factors for myopia progression
In this section, various risk factors that have been studied will be discussed.

Outdoor activities
Rose *et al.* in an Australian study found that children with low outdoor time and high near work were two to three times more likely to be myopic compared to those performing low near work and high outdoor activities.[15] Dirani *et al.* found a significant negative association between myopia and outdoor activity.[16] For each hour increase in outdoor activity per day, spherical equivalent (SE) increased by 0.17 D and axial length (AL) decreased by 0.06 mm. The Orinda Longitudinal Study of Myopia (OLSM) found that children who became myopic by the eighth grade spent less time in sports and outdoor activity (hours per week) at the third grade compared to those who did not become myopic.[17] More recently, the Guanzhou Outdoor activity longitudinal study results provided proof of principle that increasing the amount of time children spent outdoors through the school system can decrease the number of children who become myopic.[18]

Researchers have also found that it is not the sports but the exposure to the outdoors that appears to be protective. Chicks reared in less ambient light became myopic.[19] Higher light intensity outdoors could make the depth of field greater and reduce image blur. Studies have also shown that there is release of dopamine from the retina when stimulated by light and dopamine is known to inhibit eye growth.[20] Spectral composition of the light rather than the intensity seems to be a more important factor.[21]

Near work
The summary of findings regarding near work and myopia increase as found in The Sydney Myopia Study (SMS) and the Singapore Cohort Study for the risk factors for myopia (SCORM) are as follows:[22,23]

Children who read continuously for more than 30 min had a higher incidence of myopia. Children who performed near work at <30 cm distance were 2.5 times likely to be more myopic. Children who read more than two books per week were also three times likely to have higher myopia. Children who read more than 2 h per day were 1.5 times likely to have higher myopia. For every book read per week the AL elongation was more by 0.04 mm. However, several other studies failed to find any significant correlation between near work and myopia

Education
Higher educational level, higher achievement and earlier schooling were also found to be associated with an increase in myopia prevalence.[24–26] It is unclear if this can be considered as implying that it is the near work that was more important than the education itself.

Parental myopia
The SMS reported that children with one or both parents myopic had to two and eight times higher risk of developing myopia compared with children who had no myopic parents.[23] SCORM cohort showed that having one and two myopic parents

was associated with an increase in AL of 0.14 and 0.32 mm, respectively, compared with no myopic parents.[27]

Peripheral refraction

Theories of peripheral refraction state that due to the oblong shape of the eyeball the peripheral rays of light do not focus on the retina. This resultant hyperopic defocus serves as a stimulus for growth of the eyeball. While this process may be important in emmetropization, it also appears to contribute to eyeball elongation.[28,29] OLSM assessed peripheral refractive error in 822 children aged 5–14 years. This study indicated that myopic children had greater relative hyperopia in the periphery, compared to emmetropes and hyperopes.[30]

Control of myopia progression

Spectacles

1. The rationale for the use of bifocals or progressive addition lenses (PAL) is that they optimize accommodative accuracy for near tasks and minimize the retinal blur. Bifocals seem to slow the myopia progression in children with nearpoint esophoria but not necessarily in children with exophoria.[31,32] The Correction of Myopia Evaluation Trial (COMET) was a multicenter randomized study involving 469 ethnically diverse subjects where participants were randomized to wearing single vision lenses (SVL) or PALS. Mean 3-year increases in myopia were −1.28 D in the PAL group and −1.48 in the SVL group, which was statistically significant. Mean myopic progression in the PAL group with nearpoint esophoria was −1.18 D compared to −1.39D in SVL group.[33]

2. COMET 2 Trial looked at a separate cohort of children with near esophoria and high accommodative lag and placed them in PALs. The overall reduction in myopia over 3 years was only 0.28 D.[34]

3. A meta-analysis by Li *et al.* showed that PALs slowed myopia progression by 0.25 D/year and reduced AL by 0.12 mm/year. Patients that appeared to benefit the most from PALs were those with moderate myopia, Asian ancestry, Near point esophoria and high accommodative lag.[35]

4. Altering peripheral defocus was the focus of work by Sankaridurg *et al.* In their study, 210 Chinese children aged 6–16 years were fitted with one of three novel spectacle lens forms. After 12 months, no statistically significant difference between SE refraction or AL in the control group wearing SVL versus children wearing the novel spectacle lenses was found. In younger children (6–12 years old), with a

parental history of myopia 30% less myopia progression in children wearing one of the lens designs than in children wearing control SVD lenses was found. These novel lenses however are limited in their availability.[36]

Contact lenses

Extending the idea of peripheral refraction to contact lenses, the Dual Focus contact lenses were studied and found to show effectiveness similar to spectacle dual focus lenses.[37]

Orthokeratology

Orthokeratology involves temporarily reshaping the cornea with rigid gas permeable lenses. It involves wearing the lenses overnight. While meta-analysis of all studies using Orthokeratology showed a definite slowing of progression, the use of these lenses is also associated with a slight increase in incidence of epithelial defects and corneal infections. There is also a higher rebound increase in myopia progression after cessation of lens wear.[38] These lenses are of limited availability in India.

Pharmacological therapy

Muscarinic receptor antagonists Atropine and Pirenzpine have been studied for myopia control, the former extensively. The rationale behind the use of these agents is the theory that excessive accommodation leads to myopia. The other mechanisms postulated are affecting the release of neurotransimitter dopamine and synthesis of glycosaminoglycans in the sclera and thereby reducing AL elongation.

The ATOM (Atropine treatment of Myopia) 1 and 2 studies have studied the daily application of Atropine in various concentrations from 1 to 0.01% through various phases. Atropine 1% was the first to be studied. There were concerns about these children needing PALs due to the accommodative difficulties, UV toxicity and rebound progression after cessation of the treatment. Typically, the atropine was administered for 2 years and stopped. The children were followed for a washout period of 1 year following which the atropine was reinstituted for 2 more years. Both AL elongation and cycloplegic refraction were closely followed as indicators of myopia progression. Children were also monitored for side effects such as photophobia and difficulties with accommodation. The recently published 5 year results of the ATOM 2 study shows the efficacy of Atropine 0.01% in the retardation of myopia progression and its safety.[40]

- Pirenzipine which is a selective M1 receptor antagonist was also studied in a randomized control trial in the US Pirenzipine study. 174

children, aged from 8 to 12 years (mixed ethnicity/73% were Caucasian) were enrolled in the study. The mean increase in myopia was −0.26 D/year in the pirenzepine group versus −0.53 D/year in the control group.

Myths and traditional therapies

Undercorrection of myopia and giving plus glasses and witholding minus lenses are harmful practices with no evidence.

Chinese eye Exercise, Acupuncture, Qi-gong ocular exercise, Electrophotomagnitostimulation, eye massage and Drishti Dosha, Triphala, Saptamrita Lauha, Amla, Fennel, Licorice, Yoga eye excercises, one dose one globule Rhus Tox 1M and splashing water, while mentioned in various articles and websites, do not have a place in scientific literature.

Clinical practice

The ideal age to start treatment is between 6 and 8 years. Children with one or two myopic parents, whose myopia started early, who live in an urban environment, with less chance for outdoor play, with more near work, intense school curriculum, perhaps females, shorter working distance with prolonged reading, with near esophoria and/or accommodative lag maybe good candidates for intervention.

If progression of more than 0.5 D SE in last 1 year as per a cycloplegic autorefraction, then these patients may be considered for intervention. A baseline testing for near esophoria and accommodative lag, a cycloplegic autorefraction and AL measurement using a non-contact method such as an IOL master are basic requirements prior to initiation of therapy.

Choice of therapy has been covered in detail earlier. It may be guided by the availability of certain therapies too. Six monthly monitoring is needed. Increase in outdoor activity may be achieved by public health measures and interventions by school authorities through education of teachers and parents.

In conclusion, progression of myopia is a major public health burden.[41] The control of progression continues to be an area of debate and research.

References

1. **Lim MC**, Gazzard G, Sim EL, Tong L, Saw SM. Direct costs of myopia in Singapore. *Eye* 2009; 23: 1086–1089.
2. **Vitale S**, Cotch MF, Sperduto R, Ellwein L. Costs of refractive correction of distance vision impairment in the United States, 1999–2002. *Ophthalmology* 2006; 113: 2163–2170.
3. **Saw SM**, Gazzard G, Shih-Yen EC, Chua WH. Myopia and associated pathological complications. *Ophthalmic Physiol Opt* 2005; 25: 381–391.
4. **Xu L**, Li J, Cui T, *et al*. Refractive error in urban and rural adult Chinese in Beijing. *Ophthalmology* 2005; 112: 1676–1683.
5. **Cheng CY**, Hsu WM, Liu JH, Tsai SY, Chou P. Refractive errors in an elderly Chinese population in Taiwan: the Shihpai Eye Study. *Invest Ophthalmol Vis Sci* 2003; 44: 24.4630–4638.
6. **Sawada A**, Tomidokoro A, Araie M, Iwase A, Yamamoto T. Refractive errors in an elderly Japanese population: the 25. Tajimi study. *Ophthalmology* 2008; 115: 363–370.e3.
7. **Wong TY**, Foster PJ, Hee J, *et al*. Prevalence and risk factors for refractive errors in adult Chinese in Singapore. *Invest Ophthalmol Vis Sci* 2000; 41: 2486–2494.
8. **Saw SM**, Chan YH, Wong WL, *et al*. Prevalence and risk factors for refractive errors in the Singapore Malay Eye Survey. *Ophthalmology* 2008; 115: 1713–1719.
9. **Pan CW**, Wong TY, Lavanya R, *et al*. Prevalence and risk factors for refractive errors in Indians: the Singapore Indian Eye Study (SINDI). *Invest Ophthalmol Vis Sci* 2011; 52: 3166–3173.
10. **Pokharel GP**, Negrel AD, Munoz SR, Ellwein LB. Refractive error study in children: results from Mechi Zone, Nepal. *Am J Ophthalmol* 2000; 129: 436–444.
11. **He M**, Zeng J, Liu Y, Xu J, Pokharel GP, Ellwein LB. Refractive error and visual impairment in urban children in southern china. *Invest Ophthalmol Vis Sci* 2004; 45: 793–799.
12. **Dandona R**, Dandona L, Srinivas M, *et al*. Refractive error in children in a rural population in India. *Invest Ophthalmol Vis Sci* 2002; 43: 615–622.
13. **Murthy GV**, Gupta SK, Ellwein LB, *et al*. Refractive error in children in an urban population in New Delhi. *Invest Ophthalmol Vis Sci* 2002; 43: 623–631.
14. **Saxena R**, Vashist P, *et al*. Prevalence of myopia and its risk factors in urban school children in Delhi: the North India Myopia Study. *PloS One* 2015; 26: 10(2)e0117349.
15. **Rose KA**, Morgan IG, Ip J, *et al*. Outdoor activity reduces 69. The prevalence of myopia in children. *Ophthalmology* 2008; 115: 1279–1285.
16. **Dirani M**, Tong L, Gazzard G, *et al*. Outdoor activity and myopia in Singapore teenage children. *Br J Ophthalmol 70* 2009; 93: 997–1000.
17. **Jones LA**, Sinnott LT, Mutti DO, Mitchell GL, Moeschberger ML, Zadnik K. Parental history of myopia, sports and outdoor activities, and future myopia. *Invest Ophthalmol Vis Sci* 2007; 48: 3524–3532.
18. **He M**, *et al*. Effect of time spent outdoor at school on the development of myopia among children in China; a randomized clinical trial. *AMA*. 2015; 314(11): 1142–1148.
19. **Ashby RS**, Schaeffel F. The effect of bright light on lens compensation in chicks. *Invest Ophthalmol Vis Sci* 2010; 51: 5247–5253.
20. **Rose KA**, Morgan IG, Ip J, *et al*. Outdoor activity reduces the prevalence of myopia in children. *Ophthalmology* 2008; 115: 1279–1285.
21. **Mehdizadeh M**, Nowroozzadeh MH. Outdoor activity and myopia. *Ophthalmology* 2009; 116: 1229–1230; author reply.
22. **Ip JM**, Saw SM, Rose KA, *et al*. Role of near work in myopia: findings in a sample of Australian school children. *Invest Ophthalmol Vis Sci* 2008; 49: 2903–2910.
23. **Saw SM**, Carkeet A, Chia KS, Stone RA, Tan DT. Component dependent risk factors for ocular parameters in Singapore Chinese children. *Ophthalmology* 2002; 109: 2065–2071.
24. **Tay MT**, Au Eong KG, Ng CY, Lim MK. Myopia and educational attainment in 421,116 young Singaporean males. *Ann Acad Med Singapore* 1992; 21: 785–791.
25. **Teasdale TW**, Goldschmidt E. Myopia and its relationship to education, intelligence and height. Preliminary results from an on-going study of Danish draftees. *Acta Ophthalmol Suppl* 1988; 185: 41–43.
26. **Rose KA**, Morgan IG, Smith W, Burlutsky G, Mitchell P, Saw SM. Myopia, lifestyle, and schooling in students of Chinese ethnicity in Singapore and Sydney. *Arch Ophthalmol* 2008; 126: 527–530.
27. **Ip JM**, Huynh SC, Robaei D, *et al*. Ethnic differences in the impact of parental myopia: findings from a population-based study of 12-year-old Australian children. Invest Ophthalmol Vis Sci 2007; 48: 2520–2528.

28. **Mutti D,** Hayes J, Mitchell L, *et al.* Refractive error, axial length, and relative peripheral refractive error before and after the onset of myopia. *Invest Ophthalmol Vis Sci* 2007; **48**: 2510–2519.

29. **Sng CC,** Lin XY, Gazzard G, *et al.* Change in peripheral refraction over time in Singapore Chinese children. *Invest Ophthalmol Vis Sci* 2011; **52**: 7880–7887.

30. **Mutti D,** Sholtz R, Friedman N, Zadnik K. Peripheral refraction and ocular shape in children. *Invest Ophthalmol Vis Sci* 2000; **41**: 1022–1030.

31. **Fulk GW,** Cyert LA, Parker DE. A randomized trial of the effect of single-vision vs. bifocal lenses on myopia progression in children with esophoria. *Optom Vis Sci* 2000; **77**(8): 395–401.

32. **Goss DA,** Jyesugi EF. Effectiveness of bifocal control of childhood myopia progression as a function of near point phoria and binocular cross cylinder. *J Optom Vis Devel* 1995; **26**: 12–17.

33. **Gwiazda J,** Hyman L, Hussein M, *et al.* A randomized clinical trial of progressive addition lenses versus single vision lenses on the progression of myopia in children. *Invest Ophthalmol Vis Sci* 2003; **44**(4): 1492–1500.

34. **Correction of Myopia Evaluation Trial 2 Study Group for the Pediatric Eye Disease Investigator Group.** Progressive-addition lenses versus single-vision lenses for slowing progression of myopia in children with high accommodative lag and near esophoria. *Invest Ophthalmol Vis Sci* 2011; **52**(5): 2749–57.

35. **Li SM,** *et al.* Multifocal vs single vision lenses intervention to slow progression of myopia in school-age children: a meta-analysis. *Surv Ophthalmol* 2011; **56**(5): 451–460.

36. **Sankaridurg P,** *et al.* Spectacle lens designed to reduce progression of myopia: 12 month results. *Optom Vis Sci* 2010; **87**(9): 631–641.

37. **Sankaridurg P,** Holden B, Smith E 3rd, Naduvilath T, Chen X, de la Jara PL, Martinez A, Kwan J, Ho A, Frick K, Ge J. Decrease in rate of myopia progression with a contact lens designed to reduce relative peripheral hyperopia: one-year results. *Invest Ophthalmol Vis Sci* 2011;**52**(13): 9362–9367.

38. **Li SM,** Kang MT, Wu SS, Liu LR, Li H, Chen Z, Wang N. Efficacy, safety and acceptability of orthokeratology on slowing axial elongation in myopic children by meta-analysis. *Curr Eye Res* 2015: 1–9.

39. **Chia A,** Lu QS, Tan D. Five-year clinical trial on atropine for the treatment of myopia 2: myopia control with atropine 0.01% eyedrops. *Ophthalmology* 2015.

40. **Siatkowski RM,** Cotter SA, Crockett RS, Miller JM, Novack GD, Zadnik K; U.S. Pirenzepine Study Group. Two-year multicenter, randomized, double-masked, placebo-controlled, parallel safety and efficacy study of 2% pirenzepine ophthalmic gel in children with myopia. *J AAPOS* 2008; **12**(4): 332–329.

41. **Chen-Wei Pan,** Dharani R, Saw SM. Worldwide prevalence and risk factors for myopia. *Ophthalmic Physiol Opt* 2012; **32**(1): 3–16.

Infantile cataract: where are we now?

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Introduction

Pediatric cataract is one of the major causes of preventable childhood blindness affecting approximately 200,000 children worldwide.[1] In developing countries, the prevalence of blindness from cataractC is higher, about one to four per 10,000 children. Early diagnosis and treatment WWWis essential to prevent the development of stimulus deprivation amblyopia in these children. Cataract surgery in infants poses greater challenges compared to young children. Primary implantation of an intraocular lens remains controversial for infants, and the selection of an appropriate IOL power is difficult. The management of infantile cataract has changed over the last decade. In this study, we present an overview of the changing concepts of cataracts in infants and its management.

Etiology of childhood cataract

The common causes of congenital cataract are genetic, metabolic disorders, prematurity and intrauterine infections. Almost 60% of cases of congenital cataract in developed countries are idiopathic.[2] One-third of cases of congenital cataract are hereditary without any known associated systemic disease.[3] The various causes of congenital cataract are

1. Heredity: These cataracts are usually autosomal dominant but can be autosomal recessive and x linked.

2. Associated with genetic disorders: Seen in downs and turners syndrome.

3. Metabolic disorders: Galactosemia, Hypocalcemia.

4. Intrauterine infections: Toxoplasmosis, Rubella, Cytomegalovirus, Herpes, Varicella and Syphilis.

5. Associated Ocular conditions: Aniridia, Iris coloboma, lens coloboma, Lenticonus, Lentiglobus, Persistent fetal vasculature.

Next generation DNA sequencing shown promising results in identifying mutations leading to metabolic abnormalities and cataracts. This new technology has immense potential in identifying metabolic abnormalities and correct them with appropriate intervention. In future, this technology is likely to become cheaper to allow testing in children with non-syndromic bilateral cataracts with systemic manifestation and delayed development.[4]

Morphology of pediatric cataract

Morphology of pediatric cataract can not only help in the diagnosis of specific associated disorder but also helps in planning the management. Based on morphology, pediatric cataracts can be classified into cataracts involving the entire lens, central cataracts, anterior cataracts, posterior cataracts, punctate lens opacities, coralline cataracts, sutural cataract, wedge shaped cataract and cataracts associated with PFV.

Preoperative evaluation

History taking is an integral part in the evaluation of an infant with congenital cataract. The history should include

Family history of congenital or developmental cataract,

1. Antenatal history of maternal drug intake and fever with rash.

2. Birth history should be specifically looked for as bilateral congenital cataract is more common in preterm, low birthweight, small for gestational age children.[5]

3. Developmental mile stones should be carefully assessed.

4. History of visual interaction of the child with the family members should also be inquired as it helps in determining the severity of visual dysfunction.

5. History of the onset of the opacities, progression and laterality is also important. Unilateral cataracts are isolated but are usually associated with other ocular abnormalities like persistent fetal vasculature, lenticonus, lentiglobus.[6,7]

Examination of the child

Detailed ocular examination of the child can be done either in an outpatient setting if the child is cooperative or under general anesthesia when the child is being taken up for surgery. Quantification of visual acuity of the child as far as possible should be done. In infants, fixation behavior, fixation preference and resistance to occlusion gives us a clue to the visual acuity. In young infants with poorly developed fixation, an undilated distant direct ophthalmoscopy can indicate whether the opacity is visually significant or not. Dense central opacities larger than 3 mm in diameter usually need surgical removal.[8]

Examination of both the eyes has to be done to determine whether the cataract is unilateral or bilateral. Unilateral cataract, even if mild can cause irreversible deep amblyopia if not treated.[9]

Often the first symptom is a white or partially white reflex noted by the parents. Strabismus and nystagmus should be specifically looked for in these children and sometimes these may be the presenting signs. Strabismus is usually seen in children with unilateral cataracts and develops when an irreparable visual loss has already occurred. The presence of manifest nystagmus at age of 2–3 months or elder generally indicates a very poor prognosis.[10] The presence of either strabismus or nystagmus indicates that cataract is visually significant.

Slit lamp biomciroscopy should be done to assess the size, location, density of the opacity. Corneal diameters and intraocular pressure have to be measured with a tonopen or Perkins hand held applanation tonometer. Indirect ophthalmoscopy can reveal persistent fetal vasculature or other posterior segment abnormalities that may affect the visual outcome. In cases where the media opacity precludes examination of the fundus, a B scan ultrasonography has to be performed to rule out other posterior segment pathologies that mimic congenital cataract. These conditions include retinoblastoma, persistent hyperplastic primary vitreous, coats disease, ROP with retrolental fibroplasia, orgainzed vitreous hemorrhage, congenital falciform fold, ocular toxocariasis and retinal hamartomas. Performing cataract surgery in these conditions is disastrous and can lead one into medicolegal problems. These children should be evaluated by a pediatrician to exclude systemic disorders or metabolic causes causing cataract.

Laboratory workup

Most children with congenital cataract do not need systemic work up. Unilateral, familial, isolated cataracts with no systemic association do not need any systemic investigations. Trauma should be ruled out in all cases of unilateral cataract. A child with peculiar facies or systemic malformations like microcephaly, deafness, cardiac abnormalities, developmental delayneed systemic workup. The work up usually includes fasting blood sugar, urine for reducing substances for galactosemia, aminoacids for Lowes syndrome. Plasma phosphorous, calcium levels, RBC transferase and galactokinase levels have to be assessed. TORCH titers have to be done to rule out infectious causes of cataract.[11,12]

Management

Indication for cataract surgery in infants depends on the extent of its effect on the visual function. Mere presence of a lenticular opacity does not warrant surgical removal. Cataract which is incomplete at birth, peripheral lens opacities, punctate opacities with intervening clear zones, opacities <3 mm in diameter can be kept under close follow up.[13] Associated amblyopia in these children

should be treated by appropriate glasses and patching. Small opacities can be managed by prescribing mydriatric agents to achieve a larger area of clear visual axis.

Timing of surgery

Extraction of unilateral congenital cataracts by 4–6 weeks and bilateral cataracts within 6–8 weeks of life can prevent the development of strabismus, nystagmus and amblyopia.[14]

How does an infant eye differ from the adult eye?

A child's eye is unique and is different from an adult eye. The eyes are smaller in size at birth and have steeper corneas. The normal new born eye has a mean axial length of 16.6–17 mm.[15] It reaches a mean adult value of 23.6 mm at 15 years age. More than half of this growth in axial length occurs before 1 year age and most axial elongation occurs during the first 2 years of life. The corneal curvature reduces from 51.2 D in new borns to 43.5 D in adults.[16] The sclera is thin and less rigid, the lens capsule is more elastic, and there is a risk of severe postoperative inflammatory response.

Biometry in infants

With advances in surgical techniques and instrumentation, several surgeons are implanting IOLs in infants. Refractive growth after IOL implantation in infants cannot be predicted accurately and current IOL formulae vary in their predictive outcomes. If r target postoperative emmetropia, amblyopia treatment is easier but this strategy results in high myopia in later life. If we aim for hyperopia, amblyopia therapy and refractive correction in initial phase is difficult but thisstrategy has the advantage of potentially achieving either emmetropia or low myopia later in adulthood. The amount of hyperopia will vary depending on the age of the child at the time of surgery. Most surgeons prefer to leave infants with hyperopia as it's easy to titrate as children grow. IATS recommended an hyperopia ranging from +6 D to +8 D depending on the age of infant at the time of surgery. It is important to counsel parents regarding need for glasses postoperatively as well as perhaps through the life.

Three important things to be considered when determining the IOL power to be implanted in infants are

1. Anticipated refractive shift.

2. Age of the patient.

3. Target refraction in the immediate postoperative period.

Children who are younger at the time of surgery, have a significantly greater myopic shift and greater variance in predictive refractive change than older children. Crouch *et al.* in a study of 52 eyes undergoing cataract surgery with

IOL implantation found a mean myopic shift of 3.66 D in children operated on at 3–4 weeks age which reduced to 0.38 D in children operated on at 15–18 years age.[17] Most pseudophakic eyes grow normally and so a significant shift after IOL implantation is expected in these children.[18]

Postoperative refractive goal in infants
In infants, implantation of IOL still remains controversial and several surgeons prefer to leave the infants aphakic after cataract surgery.[19] In the Infant Aphakia Treatment Study, the target refractive error after IOL implantation was +8 for infants 4–6 weeks of age and +6 for infants between 6 weeks to 6 months age.[20]

Keratometry and axial length measurements in children are usually less accurate compared to adults. These measurements are often obtained under anesthesia in infants who do not cooperate for fixation. Mittelviefhaus et al. in their study have shown that lack of fixation in children under general anesthesia can result in inaccurate keratometry measurements.[21] However, the reliability can be increased by averaging several readings per eye.

Axial length is a more significant source of error in IOL power calculation. Inaccurate axial length measurement can account for 4–14 diopters for each millimeter difference in IOL power.[22] Errors are often magnified because of shorter axial length. Immersion biometry is more predictable than contact method for IOL power calculation in infants.[23] But the limitation of the immersion scan is that it cannot be used in small eyes and globe with shallow anterior chamber and other ocular anomalies as in infants. Partial coherence interferometry can be used to measure axial length in cooperative children with reliability and accuracy.[24] Advantages over conventional ultrasound include high reproducibility, contact free measurements, observer independence of the measurements. The disadvantage is that it cannot be used in total cataracts which are more often encountered in children.

IOL formula
Furthermore, no time tested formula exists for calculation of IOL power in infants. The accuracy of each formula depends on optimized values and measures of the formula components, including factors such as actual anterior chamber depth, lens thickness, vertex distance, and use of a personalized surgeon factor or A-constant. The anterior segment of an infant eye is significantly smaller, eyes with congenital cataract may have greater anatomic variation in anterior segment structures and the anterior segment of an infant is proportionally larger to the posterior segment compared to an adult eye. The capsular bag of an infant eye is smaller and will contract earlier, which may

result in greater posterior IOL displacement. Implantation of high-power IOLs in these eyes, can increase the measurement and calculation errors as well as the errors induced by changes in IOL position. Nihalani and Vanderveen in a retrospective study of 135 pediatric eyes that underwent cataract surgery and primary IOL implantation found mean predictability of four formulae was comparable, with 57% of infants having a prediction error of more than 0.5 diopters.[25] Greater prediction errors were seen in children <2 years, axial length <22 mm and mean keratometry readings >43.5. The SRK II, SRK T and Holladay formulae tended to overcorrect whereas Hoffer Q had an equal number of undercorrection and overcorrections. Kekunnaya et al. in their study on IOL power calculation in children <2 years age found prediction errors were larger for all formulas but SRK II had the least prediction error.[26] In the Infant Aphakia treatment study to determine the predictability of IOL power calculation formulae in infants eyes, overall median absolute prediction error values appeared to be similar for the Holladay 1, Holladay 2 and SRK/T formulae (1.2 D, 1.4 D and 1.3 D, respectively), and in paired comparisons of SRK/T versus other formulae, the median paired differences in absolute prediction error was more than zero, indicating greater accuracy for the SRK/T formula. The study concluded that Holladay I and SRK T formula gave good comparable results and have the best predictive value for infant eyes. The greatest prediction errors in their study were seen in eyes with axial length of 18 mm or less.[27]

Type of IOL to be implanted
There is a large debate regarding the type of IOL to be implanted in infants. IOL implantation during childhood may be associated with better visual outcomes but in IOL implantation in infancy, these potential advantages are offset by a higher incidence of intraoperative and postoperative adverse events. Additional intraocular surgeries are often required to treat these adverse events which are associated with risks, costs and parental stress. Although it is agreed that cataract surgery during early infancy is associated with the best visual outcomes, it remains undetermined whether primary IOL implantation is advisable in this age group. Ram et al. compared outcomes of hydrophobic acrylic and PMMA lenses in children <1 year age and reported that complication rates were comparable in both the groups. PMMA lenses may require early surgical intervention for PCO.[28] The single piece acrylic hydrophobic IOL is a soft IOL and can be implanted in the smaller capsular bag as in infants with relative ease. It also has the advantage of requiring a smaller incision thereby allowing corneal incision leaving conjunctiva intact. However, single piece IOL cannot be placed

in sulcus. Silicon IOLs have also be implanted in young children, but are associated with more capsular contraction.[29]

Are IOLs good for infants?

This question has been debated by Pediatric ophthalmologists for several years. There was little evidence to support the claim either way. The infant aphakia study was designed to answer this question. This was a prospective randomized multicentric trial comparing infants who underwent cataract surgery for unilateral cataract with either IOL implantation or were left aphakic and were fitted with a contact lens. The main outcome variable was visual acuity at 1 year and 4 ½ years of age. The investigators also looked at complications, resurgery rates and strabismus and stereopsis and compliance to occlusion. IATS found that there was no difference in visual acuity at either 1 year of age or at 4 ½ years between the two groups. But alarmingly adverse events like membrane proliferation into the visual axis, corectopia were almost 10 times more common in infants with IOL implantation compared to aphakic infants.[30] In aphakic eyes, the margins of the anterior and posterior capsular bag usually fuse together preventing lens material from migrating out of the Sommerring ring into the pupillary space. Whereas in pseudophakic eyes, lens epithelial cells are able to migrate into the pupillary space because the IOL interfers with the fusion of the lens capsular remnanats. Hence, not surprisingly, the commonest indication for resurgery in IOL group was to clear the visual axis. Additional intraocular surgeries were 3 ½ times more in pseudophakic infants compared to aphakic infants.[31] The risk of glaucoma was same in both the groups. The development of stereopsis did not differ depending on the type of optical rehabilitation. In conclusion, the study did not demonstrate any visual benefit from implanting an IOL at the time of unilateral cataract surgery in infants <7 months of age and the children who had IOL implantation had more adverse events and required more reoperations to clear visual axis opacities. The investigators concluded by saying "When operating on an infant younger than 7 months of age with a unilateral cataract, we recommend leaving the eye aphakic and focusing the eye with a contact lens. Primary IOL implantation should be reserved for those infants where, in the opinion of the surgeon, the cost and handling of a contact lens would be some burdensome as to result in significant periods of uncorrected aphakia."[32] In the context of our country, however the IATS conclusions need to be interpreted in a slightly different light. Monocular cataracts in infants, where only way of visual rehabilitation is contact lens, is often problematic in developing countries because of poor hygiene,

socioeconomic factors and non-availability of contact lenses in smaller towns. So these infants will probably do better with IOL. Only rider is that lens be placed in the bag with appropriate capsular management and anterior vitrectomy to ensure clear visual axis. Other important factor is patient selection and we do recommend IOL in infants who have otherwise anatomically normal eye with no anterior segment dysgenesis or other anomalies. Bilateral cataracts in infants however aphakia can be easily managed with aphakic glasses as well as contact lenses. Attending surgeon is best placed to take that decision customising it according to patient's profile and his/her skills. IATS has certainly provided information for better informed decisions.

Complications following surgery

Postoperative inflammatory response in children can result in fibrinous and pigment deposits on the IOL. Inflammatory response can be really exaggerated in infants with Rubella syndrome.

Posterior capsular opacification is the most common complication after pediatric cataract surgery. Primary posterior capsulotomy with anterior vitrectomy combined with hydrophobic acrylic IOL in the bag, can prevent or delay the occurrence of VAO. PCO if develops can be treated by NdYag laser capsulotomy or membraectomy depending on the child's cooperation and the thickness of PCO.

Secondary glaucoma is the most feared complication of infantile cataract surgery. IATS showed that IOL implantation does not seem to protect against the development of glaucoma.[33] Mataftsihave reported that glaucoma after pediatric cataract surgery is associated with surgery with in first 1 month of life and additional surgical procedures but not with primary IOL implantation.[32] Parents must be counselled regarding glaucoma and need for regular follow up. Rapid myopic shift and increased axial length points towards glaucoma and high index of suspicion should be maintained in these children.

Retinal detachment is a rare late postoperative complication of pediatric cataract surgery.

Postoperative visual rehabilitation

Visual rehabilitation in children after surgery can be achieved by aphakic glasses, contact lenses, IOL implantation. Aphakic glasses are efficient method of visual rehabilitation in infants especially in bilateral cataracts. Contact lenses are particularly useful in unilateral cataract. Silcon soft lenses or rigid gas permeable lenses are commonly used. Speegschatz *et al.* in their study of 157 aphakic subjects found that initial rehabilitation with aphakic glasses and secondary IOL implantation at a later date has the advantage of predictable postoperative refraction and fewer complications.[34] Visual rehabilitation can be done in the immediate

postoperative period by aphakic glasses in bilateral cases and contact lenses in unilateral cases. When fitting an infant with aphakia with contact lens the problem of appropriate power of the contact lens arises. Silicon elastomer contact lens is the preferred contact lens for the treatment of aphakia in infants. It is easy to fit and can be used as an extended wear contact lens. Rigid gas permeable lens is also an option given the advantage of cost and good oxygenation for cornea. The preoperative axial length can be used to determine the contact lens power to be used. Martin *et al.* in their study reported the power of the contact lens depending on the preoperative axial length as: 0–6 months, +29 D; 7–17 months, +26 D; 18–28 months, +23 D and 29–34 months, +18 D.[5] Moore noted that the mean spherical equivalent refractive error for these patients was +28.5 D at 6 months, +26.5 D at 12 months, +23 D at 24 months and +21.5 D at 36 months.[35] Trivedi *et al.* recommends 32-D CL when the preoperative AL is <17 mm, a 29-D Cl when the preoperative AL is between 17 and 18.5 mm, a 26-D CL when the preoperative AL is 18.5–19.5 D, a 23-D CL when the preoperative AL is between 19.5 and 20 mm (21 mm) and a 20-D CL for an AL of 20–21 mm (20 D for >21 mm).[36] Secondary IOL implantation can be reserved as an option for later visual rehabilitation in these children. Nihalani *et al.* in their study of secondary IOL implantation in children left aphakic during initial cataract surgery found satisfactory visual and refractory outcomes in children receiving secondary IOL implantation. However, immediate postoperative inflammation and corneal edema was more in eyes with sulcus implanted IOL compared to in the bag IOL.[37]

Last but not the least, several of these infants will need continued monitoring for amblyopia and patching and strabismus. They will need appropriate refractive correction moving from a single vision glass to a bifocal or progressive as their visual needs change. It is important not to miss these unglamorous factors in follow up visits otherwise amblyopia can trump a beautifully done surgery in a matter of few months.

Conclusion

Cataract surgery in infants is a specially challenging subset among all cataract surgeries in children in terms of surgical technique, formula to be used, biometry and postoperative visual rehabilitation. But it is rewarding, if we are able to rehabilitate them either with glasses or primary IOL implantation or contact lenses. Surgeon has the onus of deciding on the best course looking ahead not few years but few decades.

References

1. **Holmes JM**, Leske DA, Burke JP, Hodge DO. Birth prevalence of visually significant infantile cataract in a defined U.S. population. *Ophthalmic Epidemiol* 2003; **10**: 67–74.

2. **Lim Z**, Rubab S, Chan YH, Levin AV. Pediatric cataract: the Toronto experience-etiology. *Am J Ophthalmol* 2010; **149**: 887–892.

3. **Merin S**, Crawford JS. The etiology of congenital cataracts. A survey of 386 cases. *Can J Ophthalmol* 1971; **6**: 178–182.

4. **Gillespie RL**, Sullivan J, Ashworth J, Bhaskar S, Williams S, Biswas S, Kehdi E, Ramsden SC, Clayton-Smith J, Black GC, Lloyd IC. Personalized diagnosis and management of congenital cataract by next-generation sequencing. *Ophthalmology* 2014; **12**: 2124–2137.

5. **Haargaard B**, Wohlfahrt J, Rosenberg T, Fledelius HC, Melbye M. Risk factors for idiopathic congenital/infantile cataract. *Invest Ophthalmol Vis Sci* 2005; **46**: 3067–3073.

6. **Amaya L**, Taylor D, Russell-Eggitti , Nischal KK, Lengyel D. The morphology and natural history of childhood cataracts. *Surv Ophthalmol* 2003; **48**: 125–144.

7. **Haargaard B**, Wohlfahrt J, Fledelius HC, Rosenberg T, Melbye M. A nationwide Danish study of 1027 cases of congenital/infantile cataracts: etiological and clinical classifications. *Ophthalmology* 2004; **111**: 2292–2298.

8. **Arkin M**, Azar D, Fraioli A. Infantile cataracts. *Int Ophthalmol Clin* 1992; **32**: 107–120.

9. **Zetterström C**, Lundvall A, Kugelberg M. Cataracts in children. *J Cataract Refract Surg* 2005; **31**: 824–840.

10. **Gelbart SS**, Hoyt CS, Jastrebski EM. Long term visual results in bilateral congenital cataracts. *Am J Ophthalmol* 1982; **93**: 615–621.

11. **Raghu H**, Subhan S, Jose RJ, Gangopadhyay N, Bhende J, Sharma S. Herpes simplex virus-1–associated congenital cataract. *Am J Ophthalmol* 2004;**138**:313–314.

12. **Vijayalakshmi P**, Kakkar G, Samprathi A, Banushree R. Ocular manifestations of congenital rubella syndrome in a developing country. *Indian J Ophthalmol* 2002; **50**: 307–311.

13. **Choi J**, Kim JH, Kim SJ, Yu YS. Clinical characteristics, course, and visual prognosis of partial cataracts that seem to be visually insignificant in children. *J AAPOS* 2012; **16**: 161–167.

14. **Birch EE**, Cheng C, Stager Dr, Weakley Dr. The critical period for surgical treatment of dense congenital bilateral cataracts. *J AAPOS* 2009; **13**: 67–71.

15. **Swan KC**, Wilkins JH. Extraocular muscle surgery in early infancy–anatomicalfactors. *J Pediatr Ophthalmol Strabismus* 1984; **21**: 44–49.

16. **Eibschitz-Tsimhoni M**, Archer SM, Del Monte MA. Intraocular lens power calculation in children. *Surv Ophthalmol* 2007; **52**: 474–482.

17. **Crouch ER**, Crouch ER Jr, Pressman SH. Prospective analysis of pediatricpseudophakia: myopic shift and postoperative outcomes. *J AAPOS* 2002; **6**: 277–282.

18. **Hutchinson AK**, Drews-Botsch C, Lambert SR. Myopic shift after intraocular lens implantation during childhood. *Ophthalmology* 1997; **104**: 1752–1757.

19. **Lambert SR**, Lynn M, Drews-Botsch C, *et al.* Intraocularlens implantation during infancy: perceptions of parents and the American Association for Pediatric Ophthalmology and Strabismus members. *J AAPOS* 2003; **7**: 400–405.

20. **Eibschitz-Tsimhoni M**, Archer SM, Del Monte MA. Intraocular lens power calculation in children. *Surv Ophthalmol* 2007; **52**: 474–482.

21. **Mittelviefhaus H**, Gentner C. Errors in keratometry for intraocularlen simplantation in infants. *Ophthalmology* 2000; **97**: 186–188.

22. **Eibschitz-Tsimhoni M**, Tsimhoni O, Archer SM, Delmonte MA. Effect of axial length and keratometry measurement error on intraocular lens implant power prediction formula in pediatric patients. *J AAPOS* 2008; **12**: 173–176.

23. **Trivedi RH**, Wilson ME. Prediction error after pediatric cataract surgery with intraocular lens implantation: contact versus immersion A-scan biometry. *J Cataract Refract Surg* 2011; **37**: 501–505.

24. **Hussin HM,** Spry PGD, Majid MA, *et al.* Reliability and validity of the partial coherence interferometry for measurement of ocular axial length in children. *Eye* 2005; **20:** 1021–1024.

25. **Nihalani BR,** VanderVeen DK. Comparison of intraocular lens power calculation formulae in pediatric eyes. *Ophthalmology* 2010; **117:** 1493–1499.

26. **Kekunnaya R,** Gupta A, Sachdeva V, Rao HL, Vaddavalli PK, Om Prakash V. Accuracy of intraocular lens power calculation formulae in children less than two years. *Am J Ophthalmol* 2012; **154:** 13–19.

27. **VanderVeen DK,** Nizam A, Lynn MJ, Bothun ED, McClatchey SK, Weakley DR, DuBois LG, Lambert SR; Infant Aphakia Treatment Study Group. Predictability of intraocular lens calculation and early refractive status: the Infant Aphakia Treatment Study. *Arch Ophthalmol* 2012; **130:** 293–299.

28. **Ram J,** Jain VK, Agarwal A, Kumar J. Hydrophobic acrylic versus polymethylmethacrylate intraocular lens implantation following cataract surgery in the first year of life. *Graefes Arch Clin Exp Ophthalmol* 2014; **252:** 1443–1449.

29. **Pavlovic S,** Jacobi FK, Graef M, Jacobi KW. Silicone intraocular lens implantation in children: preliminary results. *J Cataract Refract Surg* 2000; **26:** 88–95.

30. **Plager DA,** Lynn MJ, Buckley EG, Wilson ME, Lambert SR. Complications, adverse events, and additional intraocular surgery 1 year after cataract surgery in the Infant Aphakia Treatment Study. *Ophthalmology* 2011; **118:** 2330–2334.

31. **Carrigan AK,** DuBois LG, Becker ER, Lambert SR. Cost of intraocular lens versus contact lens treatment after unilateral congenital cataract surgery: retrospective analysis at age 1 year. *Ophthalmology* 2013; **120:** 14–19.

32. **Mataftsi A,** Haidich AB, Kokkali S, *et al.* Postoperative glaucoma following infantile cataract surgery: an individual patient data meta-analysis. *JAMA Ophthalmol* 2014; **132:** 1059–1067.

33. **Trivedi RH,** Wilson ME Jr, Golub RL. Incidence and risk factors for glaucoma after pediatric cataract surgery with and without intraocular lens implantation. *J AAPOS* 2006; **10:** 117–123.

34. **Speeg-Schatz C,** Flament J, Weissrock M. Congenital cataract extraction withprimary aphakia and secondary intraocular lens implantation in the ciliarysulcus. *J Cataract Refract Surg* 2005; 750–756.

35. **Martin NF,** Kracher GP, Stark WJ, Maumenee AE. Extended-wear soft contact lenses for aphakic correction. *Arch Ophthalmol* 1983; **101:** 39–41.

36. **Trivedi R,** Wilson E. Selection of an initial contact lens power for infantile cataract surgery without primary intraocular lens implantation. *Ophthalmology* 2013; **120:** 1973–1976.

37. **Nihalani BR,** Vanderveen DK. Secondary intraocular lens implantation after pediatric aphakia. *J AAPOS* 2011; **15:** 435–440.

Amblyopia: what else beyond patching and critical period?

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Amblyopia has been conventionally defined as "a unilateral or bilateral decrease of visual acuity (VA) caused by pattern deprivation or abnormal binocular interaction, for which no cause could be detected by the physical examination of the eye and which, in some cases, could be reversed by therapeutic measures".[1] Recent understanding emphasizes that, amblyopia could be redefined as a syndrome, "a visual cacophony of deficits in contrast sensitivity, spatial localization, fixation, ocular motility, accommodation, crowding, attention, motion perception and temporal processing in addition to VA loss".[2]

This article aims to review the understanding of amblyopia from the developments in literature in the view of amblyopia mechanisms, treatment and future directions.

Amblyopia is the major cause of defective vision in the young with large population studies showing an amblyopia prevalence of 1.6–3.6% with higher rated in medically underserved population.[3] Amblyopia has traditionally been classified as strabismic (SA), anisometropic (AA), refractive or deprivational according to the accompanying conditions thought to be responsible for the acuity loss.[4] There is converging evidence in literature that suggests that amblyopia might be more correctly classified in terms of the visual and oculomotor disturbances noted.[5]

Amblyopia can be defined as a developmental abnormality of the visual cortex.[6,7] There may be a loss of connections (under-sampling) or a distortion or rearrangement of connections (neural disarray) within the visual cortex, and this depends on the etiology that caused the amblyopia and the central nervous system compensates to the loss by means of the above-mentioned mechanisms.[8] The binocular interactions and the associated deficits are more apparent in the central visual field than the periphery[9,10] and the inhibitory mechanisms are active under binocular viewing.[11]

The art of amblyopia treatment lies in balancing the conflicting demands of monocular and binocular vision. During the past 20 years, different critical periods have been demonstrated for different visual functions during the development of the visual system.[12] Critical period is that sensitive period in the life span of an organism where the skills acquired are indispensable. It is believed that, visual functions processed at higher anatomical levels within the system have a later critical period than functions processed at lower levels.[13]

Binocular function and stereopsis, if disrupted within this sensitive period, are difficult to retrieve at later age. This general principle suggests that the treatment for amblyopia should follow a logical sequence, with treatment for each visual function starting before its critical period is over.[8] Three periods in the development of the visual system in human infants have been postulated by Daw.[8] These include a pre-stereoptic period (0–4 months), onset of stereopsis (4–6 months) and post-stereoptic period (6 months–2years). These functions are all plastic for a period after they first develop.

Based on the understanding of critical period, it could be predicted that anomalous binocular visual experience during early infancy severely disrupts stereopsis. Fawcett et al.[14] used a random dot stereo test to determine the critical period for stereopsis in infantile and accommodative strabismus in children <5 years of age and found that the critical period for susceptibility of stereopsis extends through late infancy and early childhood and continues to at least 4.6 years of age. In more than 200 years, there is better understanding of the neurophysiology and neuropathology of amblyopia than the past, though the treatment has not changed significantly.[15] The mainstay of treatment still consists of forcing the use of the amblyopic eye, most often by occlusion of the sound eye[3] and is probably the oldest treatment modality. Either Saint-Yves in 1722[16] or de Buffon (1743) is credited with the first documentation of occlusion treatment for amblyopia. There has been adequate research on standardizing the occlusion regimens *that ranges* from a few minutes of patching a day to all waking hours, often continuing for many months.[17] More successful treatment strategies are being opted currently through well controlled experiments and neurophysiologic research, and recent randomized controlled trials have tried to answer the basic questions, "which of the two eyes should be occluded, and with what, and for how long?"[18,19]

There is a variety of evidence from the literature that the eye contralateral to the amblyopic eye, referred to as the good eye has subtle but measurable deficits in a wide variety of visual functions.[3] In this context, we (Varadharajan & Hussaindeen, 2012)[20] tried to understand the development of VA in the better eye of unilateral amblyopes. A total of 112 children with amblyopia were included (SA, 14; AA, 51; combined

mechanism (CMA), 47) in this study. Baseline VA in the fellow eye of these children differed significantly from that of age-matched controls up to 8 years of age. Average logMAR acuity reached 0.0 at age 5 years in controls versus age 9 years in patients. Although the mean VA of the fellow eyes improved during treatment, 21% developed temporary occlusion amblyopia. Full-time patching had no additional benefit when compared with part-time patching. We concluded through this study that VA in the fellow eye of children with unilateral amblyopia is reduced at baseline and matures more slowly than in healthy control patients.

Moving on to visual plasticity, it was believed that adult amblyopia is irreversible beyond the sensitive period of development but newer studies suggests that matured amblyopic brain retains a substantial degree of plasticity.[21] The primary sensory cortex also remains plastic in adulthood[22] and perceptual learning (PL) could be of benefit in restoring the visual functions of the adult amblyopic system. PL refers to experience-induced changes in the way perceivers extract information and plays a larger role in complex cognitive tasks. A new approach of binocularly based treatment modality that provides measurement and treatment of suppressive imbalance as a first step is Dichoptic training.[23,24] This leads to restoration of stereoscopic function along with reduction in the monocular acuity deficit.[23] Ultimately, it strengthens the binocular vision by eventually combining binocular information under natural viewing conditions (stimuli of the same contrast in each eye).[24] However, PL as well as Dichoptic training needs a controlled environmental set up as well as large number of trials.

An alternate or a simpler strategy to this approach would be **Action Videogames** (AVG) that can be used as a home-based therapy in any environmental setup. It provides visually enriched environment with extraordinary speed, constant monitoring of the periphery, tracking of fast moving objects, perceptual, cognitive and/or motor skills, ignoring distracters and aiming small moving targets.[25] For training amblyopic eyes, we need a task that trains many visual functions at a time. Video games might be able to fit into such a criterion. AVG exercise has shown to improve VA, potential acuity, spatial attention and stereoacuity in adults with amblyopia.[26]

We tried to understand the efficacy of AVGs in improving the visual functions in adult amblyopia. This study carried out at Sankara Nethralaya had two main goals: to assess whether AVGs can improve visual performance in adult and juvenile amblyopes; to assess the sustainability (retention) of visual functions after cessation of the exercise. A total of 36 subjects (age: 13–31 years) were recruited and allocated into two intervention groups: AVG group ($n = 33$) and Conventional

Patching group ($n = 8$). Crowded VA, uncrowded VA, stereo acuity, contrast sensitivity and degree of suppression were measured in each visit. Visual functions were reassessed after 20th and 40th hour of AVG exercise. Retention of these visual functions was assessed after 1, 5 and 7 months of cessation of the exercise. After 80 h of occlusion, no significant improvement of visual functions was noted in conventional patching group ($P > 0.05$). Twenty-one subjects who completed 40 h of AVG exercise showed substantial improvement of VA (median improvement: 0.16 log MAR; uncrowded: 41.02% and crowded: 27.6%), contrast sensitivity (maximum improvement at four cycles/degree: 113.2%), stereo acuity and depth of suppression. The AA and CMA groups showed speeder recovery of VA as compared to the SA group. Improvement of VA not only sustained fully, but also improved further after 5–7 months of cessation.

These results suggest that AVGs can be administered for active vision training in adult and juvenile amblyopes after the critical period. Ours is the first study to report regarding sustainability of the improvements obtained. Improvement of VA not only sustained well in mild and moderate amblyopes, but also revealed further improvement. Overall improvements in CS almost fully retained for 1 and 5 months post cessation at maximum spatial frequencies and was above 76% after 7 months of cessation. These results show a lot of promise toward newer treatment strategies such as video games and dichoptic training for amblyopia beyond patching and beyond the critical period. Dichoptic training seems to be the future of amblyopia treatment considering the advantages that it renders to both the monocular and binocular visual system in amblyopia. Hence, it is time for a well-planned randomized controlled trial assessing the efficacy of these modern treatment approaches in improving the visual functions in adult and juvenile amblyopes compared to conventional treatment strategies.

References

1. Noorden GKV. Binocular vision and ocular motility; theory and management of strabismus; 1996.
2. Asper L, Crewther D, Crewther SG. Strabismic amblyopia: Part 1: psychophysics. *Clin Exp Optom* 2000; **83**: 49–58.
3. Simons K. Amblyopia characterization, treatment, and prophylaxis. *Surv Ophthalmol* 2005; **50**: 123–166. Review.
4. *American Academy of Ophthalmology Basic & Clinical Science Course (1999–2000) Section VI: Binocular Vision and Ocular Motility*. San Francisco: American Academy of Ophthalmology.
5. McKee SP, Schor CM, *et al.* The classification of amblyopia on the basis of visual and oculomotor performance. *Trans Am Ophthalmol Soc* 1992; **90**: 123–148.
6. Simmers AJ, Ledgeway T, *et al.* The extent of the dorsal extra-striate deficit in amblyopia. *Vis Res* 2006; **46**: 2571–2580.
7. Li X, Dumoulin SO, *et al.* Cortical deficits in human amblyopia: their regional distribution and their relationship to the contrast

detection deficit. *Invest Ophthalmol Vis Sci* 2007; **48**: 1575–1591.

8. Daw NW. Critical periods and amblyopia. *Arch Ophthalmol* 1998; **116**: 502–505. Review.

9. Levi DM, Yap YL, *et al.* Is amblyopia an anomaly of photopicvision? *Clin Vision Sci* 1988; **3**: 243–254.

10. Philipp W, Mayer W, *et al.* Investigation of visual field defects in strabismic and anisometropicamblyopes with the Octopus Program G1. *Graefes Arch Clin Exp Ophthalmol* 1989; **227**: 448–454.

11. Pardhan S, Gilchrist J. Binocular contrast summation and inhibition in amblyopia. The influence of the interocular difference on binocular contrast sensitivity. *Doc Ophthalmol* 1992; **82**: 239–248.

12. Kiorpes L. Critical periods in visual development: implications for amblyopia. *J Vis* 2004; **4**: 31.

13. Levi DM, Carkeet A. Amblyopia: A Consequence of Abnormal Visual Development. In: Simons K (ed.), *Early Visual Development, Normal and Abnormal*, New York: Oxford University Press, Inc.: 1993; 391–408.

14. Fawcett SL, Wang Y-Z, *et al.* The critical period of susceptibility of human stereopsis. *Invest Ophthalmol Vis Sci* 2005; **46**: 521–525.

15. Tulvatana W. Amblyopia treatment reevaluated. *Chula Med J* 2004; **48**: 657–658.

16. Garzia RP. Efficacy of vision therapy in amblyopia: a literature review. *Am J Optom Physiol Opt* 1987; **64**: 393–404.

17. Hiscox FN, Strong N, *et al.* Occlusion for amblyopia: a comprehensive survey of outcome. *Eye* 1992; **6**: 300–304.

18. The Pediatric Eye Disease Investigator Group (PEDIG). A randomized trial of prescribed patching regimens for treatment of severe amblyopia in children. *Ophthalmology* 2003; **110**: 2075–2087.

19. The Pediatric Eye Disease Investigator Group (PEDIG). A randomized trial of patching regimens for treatment of moderate amblyopia in children. *Arch Ophthalmol* 2003; **121**: 603–611.

20. Varadharajan S, Hussaindeen JR. Visual acuity deficits in the fellow eyes of children with unilateral amblyopia. *J AAPOS* 2012; **16**: 41–45.

21. Levi DM, Polat U. Neural plasticity in adults with amblyopia. *Proc Natl Acad Sci USA* 1996; **93**: 6380–6834.

22. Gilbert CD, Sigman M, Crist R. The neural basis of perceptual learning. *Neuron* 2001; **31**: 681–697.

23. Hess RF, Mansouri B, Thompson B. A binocular approach to treating amblyopia: antisuppression therapy. *Optom Vis Sci* 2010; **87**: 697–704.

24. Hess R, Mansouri B, Thompson B. Restoration of binocular vision in amblyopia. *Strabismus* 2011; **19**: 110–118.

25. Achtman RL, Green CS, Bavelier D. Video games as a tool to train visual skills. *Restor Neurol Neurosci* 2008; **26**: 435–446.

26. Li RW, Ngo C, Nguyen J, Levi DM. Video-game play induces plasticity in the visual system of adults with amblyopia. *PLoS Biol* 2011; **9**: 1793.
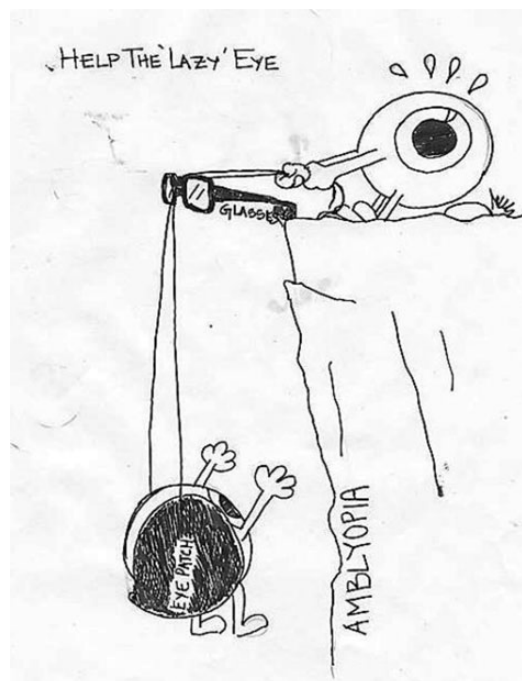
Sketch: Dr Debamalya Das

Surgical management of small angle strabismus

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Small angle deviation refers to deviations <15 prism diopters (PD). Standard rectus muscle recession–resection is designed to correct moderate to large angle strabismus >10 PD. Small angle eso-deviations and vertical deviations cause asthenopic symptoms and diplopia which may be frustrating for the patient and surgeons alike. This can be true for primary deviations and unfortunately even in postoperative patients. Several non-surgical management options to overcome the diplopia include prisms, botuliniuminjections, Bangarters filters and the last resort of self-guided coping mechanism. If these fail or any of these are non-desirable, then the alternative solution would be the surgical intervention. Unfortunately, small angle strabismus surgery is prone for over correction when using standard recess–resect procedure. The reason leading to a list of alternative surgical approaches which include

1. Partial thickness myotomies–marginal myotomy–Z plasty.

2. Graded partial tenotomy of vertical rectus for hypertropia Mini-tenotomy.

3. Mini-plication.

4. Single muscle strabismus surgery.

5. Adjustable fadens/combined recession and resection of a rectus muscle.

Partial thickness myotomy

This technique has been in use over the past 150 years. The common procedure today is marginal myotomy, often performed as 'Z' plasty across one-third to ½ the tendon width. It is used to weaken the already dramatically recessed muscle, when recession is not prudent. It is also indicated in patients with thin sclera or to weaken a rectus muscle that has at or near its insertion an implant, exoplant or encircling element used in retinal detachment or for glaucoma filtration.

Partial tenotomy/graded tenotomy

In 2000, Alan Scott[1] described a rectus muscle tenotomy procedure called graded rectus muscle tenotomy which he performed under local anesthesia to treat small degrees of vertical strabismus. Biglan[2] and co-workers did the same in 2004 on vertical muscles only. Partial tenotomy can be done as a primary procedure or in reoperations for small angle strabismus to improve diplopia. The procedure was done under sub conjunctival injection of lignocaine, conjunctival incision was made

to expose the tendon and making successive small cuts in the rectus muscle at the insertion until the desired effect is achieved. Over half the tendon was removed starting at one pole, leaving one tendon pole attached to sclera, resulting in the cut tendon slanting back at an angle of 45°. A 60–70% tenotomy, or removing 6–7 mm of tendon, corrects $\sim 4\Delta$ of strabismus.

The slanted tenotomy works by effectively moving the insertion, thus changing the vector of muscle force and potentially inducing incomitance. A vertical deviation could be induced if an upper tenotomy of one medial rectus muscle was performed along with a lower tenotomy on the contralateral medial rectus muscle. Likewise, an A pattern could theoretically be induced, or a V pattern treated, if the upper poles of both medial rectus muscles were removed, effectively moving the insertions down. This induced incomitance caused by the slanted pole tenotomy was substantiated by van der Muelen-Schot et al.[7]

The advantages of this procedure include the ease of the procedure, adjustments can be made intra-operatively, less bleeding, sutureless and the temporal incision reduces the risk of infection. Graded partial tenotomy of vertical rectus muscle may have applications beyond that has been described.

Mini tenotomy

This is usually considered for the treatment of vertical deviation of ≤6 PD, and horizontal deviations <16 PD. Kenneth W[3] described mini-tenotomy, a central tenotomy of 3–mm of tendon insertion, producing a correcting effect of ~ 2–3Δ of strabismus. A correction of up to 4–5Δ can be obtained if binocular surgery was performed. For example, a right hypertropia of 4Δ could be corrected by a right superior rectus tenotomy and left inferior rectus tenotomy. An esotropia of 4–6Δ can be treated with bilateral medial rectus tenotomies. This mini-tenotomy could be adjusted by remeasuring the deviation after operating on one muscle and determine if additional surgery is needed. The size of the tenotomy can be enlarged by resnipping additional tendon fibers for residual deviation.

Surgical technique

Under topical anesthesia following steps can be performed (Figure 1)

(a) Grasp tendon of the rectus muscle centrally with Wright 0.75-tooth tenotomy forceps through the intact conjunctiva.
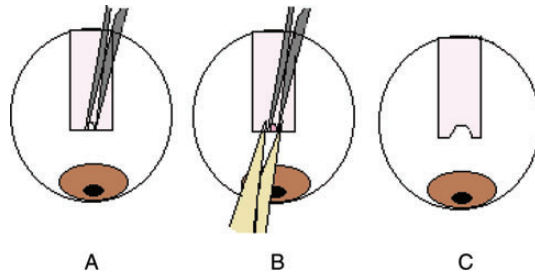
Figure 1:

(b) Cut the central tendon between the forceps and sclera with blunt Westcott scissors.

(c) final outcome shows intact two muscle poles at the extremes of the insertion. These ends maintain the stability and a central approach avoids the ciliary vessels.

In patients in whom the mini-tenotomy fails to alleviate the diplopia standard surgery can be safely done without difficulty.

Mini-plication

The mini-plication described by Kenneth *et al.*[4] was conceived for deviations of 8Δ to 10Δ, which are too small angles for standard surgery. The approximate dose–response of the procedure was about 5Δ to 7Δ correction in patients without previous surgery resection and those with a recession of the antagonist had a correction of 8Δ to 10Δ per muscle. The response can be tailored by placing the securing suture farther back from the muscle insertion.
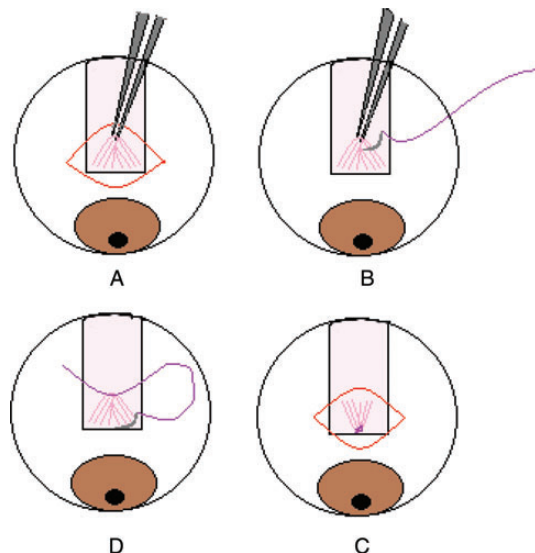

Figure 2:

Surgical technique (Figure 2)

(a) A swan conjunctival incision is made over the muscle insertion, dissection is done and muscle is grasped centrally ~5 mm from insertion.

(b) A double-armed 6-0 polyglactin 910 suture is placed underneath the forceps the suture is tied in a square knot.

(c) The sclera suture is placed anterior to the insertion and tied in place to plicate the central portion of the muscle.

(d) The final outcome showing plication.

The mini-plication has many advantages over standard strabismus surgery that requires hooking and removing the muscle. It is less invasive and preserves the integrity of the muscle insertion and presumably the anterior cilliary vessels, though this was only studied in the full plication procedure. Vicryl suture is used in this procedure as well as in the full plication procedure, because a permanent suture placed anterior to the muscle insertion will erode through the conjunctiva. It can be done under topical anesthesia and can be reversed.

One muscle strabismus surgery

The role of unilateral rectus muscle surgery has been controversial due to the significant number of under correction and or ocular incomittance. Recent studies[5] have shown that unilateral rectus recession or resection is a safe, effective and predictable treatment for small to moderate angle horizontal deviation including exo-deviation between 15 and 20 PD, exo-deviation between 15 and 40 PD, in patients with under-corrections or recurrent strabismus and in patients with convergence or divergence insufficiency. Single muscle surgeries are often resurgeries. Residual esotropia or exotropias of ~15–20 PD respond well with single muscle recessions or resections. Studies show ~60–90% success rate. The advantage of this is that it limits the surgery to one eye.

Combined recession and resection of rectus muscle/adjustable faden procedure

Scott[6] suggested a procedure that was based on modification of the faden principle for improving incomitance in gaze away from the primary position. The faden procedure in itself does not affect primary position alignment and is difficult technically and is ineffective on the lateral rectus due to large arc of contact. He performed a large resection of a rectus muscle and then recessed the muscle, using a standard hang-back/adjustable technique, in a position where the recession amount exceeded the resection amount. The reattachment of the muscle to sclera at a posterior insertion point produced the mechanical effect of

faden operation. The combined recession–resection procedure has a useful role in the management of symptomatic incomitant strabismus. The procedure is particularly valuable when dealing with incomitance on lateral gaze due to limitation of adduction, when the overacting muscle is the contralateral lateral rectus muscle. It seems to be an effective procedure in expanding the field of single binocular vision.

Conclusion

The various causes of small angle strabismus include microtropia, residual deviations after optical and surgical corrections, intermittent squints, sensory micro-strabismus and paretic strabismus. Normal divergence and vertical fusion amplitudes are small, with divergence measuring 4–6 PD (∆) and vertical vergence <2 D. Because of this, even small eso-deviations and vertical deviations can cause significant asthenopic symptoms and diplopia, especially when the deviation is acquired in adulthood. Historically, these small-angle deviations have been treated with prism glasses. Prism spectacles are an excellent option if the patient is wearing spectacles for a refractive error. Prisms do not correct incomitance and for many patients prism spectacles are undesirable.

Management of patients with small to moderate angle horizontal and vertical deviation continues to be challenging for the strabismus surgeon. These lesser invasive strabismus surgery provides a valuable option to our strabismus patients with small deviations. The various surgical techniques described can be used to treat diplopia in patients with small angle strabismus who are not ideal candidates for conservative management. These procedures can be done in the office under topical or local anesthesia and provide an alternative to prisms and the standard strabismus surgery. The key to success is careful patient selection, analyzing the critical factors like the angle type-vertical/horizontal, angle size, angle character–phoria/tropia. Most of the surgeries described are muscle weakening procedure although the weakening effect is limited. The procedures described can be useful addition to a strabismus surgeons repertoire.

References

1. **Scott AB.** Graded rectus muscle tenotomy for small deviations. *Proceedings of the Jampolsky Festschrift*. San Francisco: The Smith-Kettlewell Eye Research Institute; 2000: 215–216.
2. **Albert W,** Biglan , Hye Bin Yim. Graded partial tenotomy of vertical rectus muscle for treatment of hypertropia. *Trans Am Ophthalmol Soc* 2004; **102**: 169–176.
3. **Kenneth W Wright.** Mini-tenotomy procedure to correct diplopia associated with small-angle strabismus. *Trans Am Ophthalmol Soc* 2009; **107**: 97–103.
4. **Rebecca S Leenheer,** Kenneth W Wright. Mini-plication to treat small-angle strabismus: a minimally invasive procedure. *J AAPOS* 2012; **16**: 327–330.
5. **Lihua Wang,** Leonard B Nelson. One muscle strabismus surgery. *Curr Opin Ophthalmol* 2010; **21**: 335–3401.
6. **Scott AB.** Posterior fixation adjustable and without posterior sutures. In: Lennerstrand G, ed. *Proceedings VIIth congress International Strabismological Association*. CRC Press; 1995; pp. 399–402.
7. **van der Muelen-Schot HM,** van der Muelen-Schot SB, Simonsz HJ. Caudal or cranial partial tenotomy of the horizontal rectus muscles in A and V pattern strabismus. *Br J Ophthalmol* 2008; **92**: 245–251.

An approach to Nystagmus management

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When it comes to a patient presenting with nystagmus there are a few issues that come into consideration. The main issues are whether the nystagmus is sensory or motor, whether it is the cause or the effect of vision problem, whether to investigate or not and whether to treat or not. To know whether the nystagmus is sensory or motor we should know the range of vision impairment, if associated diurnal changes, if progressive or stable and the presence of other associated symptoms like photophobia or oculo-digital sign, etc. Once the sensory causes have been addressed then comes the issue of actually treating the nystagmus per se. Treatment of nystagmus actually only implies improving the major two effects of nystagmus namely abnormal head posture due to the presence of a null zone and improvement in vision due to the frequency of the nystagmus. So, all the modalities of treatment address these two aspects.

Non-surgical management of nystagmus

(A) Optical methods

1. Glasses:
   Effort should be made to correct any underlying refractive error which will decrease the nystagmus.

   Retinoscopy may be difficult to perform accurately when the nystagmus amplitude is large and should be performed with the eyes in the null zone if such is present.

2. Contact lenses:
   Contact lenses have been reported to have reduced amplitude and frequency and are helpful in high ammetropias.

   It has the optical advantage of moving synchronously with the eyes so that the visual axis coincides with the optical center of the lens at all times and shows improvement in visual acuity.

   There is a theory that some kind of tactile feedback from the contact lens decreases the nystagmus possibly mediated via trigeminal afferents.

3. Over minus lenses:
   Adding concave glasses to distant correction induces artificial accommodation that is accompanied with secondary convergence. This induced convergence diminishes amplitude and rate of nystagmus thus enhancing vision. Overcorrection with minus lenses stimulates accommodative convergence and may improve visual acuity at distance fixation by nystagmus dampening.

4. Prisms:
   Prisms are used for two purposes in the treatment of nystagmus: (1) to improve visual acuity and (2) to eliminate an anomalous head posture.

   (a) Convergence induced:
   In patients whose nystagmus is suppressed by viewing a near target, convergence prisms will often improve vision. Base-out prisms are prescribed to stimulate fusional convergence, which may be effective in decreasing the amplitude of nystagmus and thus improving visual acuity. The dampening of nystagmus allows "clear vision at a glance," removing the necessity for increased visual concentration and thereby avoiding intensification of the nystagmus resulting from that heightened fixation. Congenital nystagmus responds well to it. Normal binocular vision is a prerequisite of the use of prisms base-out since fusional convergence in response to prism-induced temporal retinal disparity cannot be expected in patients without fusion. In many patients, the disadvantages of prisms outweighs the modest visual benefit gained.

   (b) Induced divergence:
   Some patients with acquired nystagmus and in patients whose nystagmus is worse during near viewing, base-in prisms may help which induce divergence.

   (c) Moving the null point:
   Prisms with base opposite to preferred direction of gaze may be helpful in correcting the head posture. For example, in a patient with head turn to the left, the null zone is in dextroversion and a prism base-in before the right eye and base-out before the left eye will be helpful in correcting the abnormal head posture.

   (d) Preoperative evaluation:
   The prisms are inserted with the base opposite the preferred direction of gaze. For instance, with a head turn to the left, the null zone is in dextroversion, and a prism base-in before the right eye and base-out before the left eye will correct the head turn. Likewise, a compensatory

chin elevation caused by a null zone in deorsumversion will be improved with prisms base-up before each eye. A combination of vertical and horizontal prisms can be used when the null zone is in an oblique position of gaze. Thus, the results of surgery for head turn in nystagmus can be reasonably well predicted on the basis of the patient's response to prisms, and a postoperative residual head turn may be alleviated further with prisms.

(B) Optically coupled device

Rushton and Cox described an optical system that stabilizes images upon the retina. This system consists of a high-positive-power spectacle lens worn in combination with a high-negative-power contact lens. The system rests on the principle that stabilization of images on the retina is achieved if the spectacle lens focuses the primary image close to the center of rotation of the eye. Such images, however, are defocused, and a contact lens is required to extend back the focus onto the retina. Since the contact lens moves with the eye, it does not negate the effect of retinal image stabilization produced by the spectacle lens. With such a system, it is possible to achieve up to 90% stabilization of images upon the retina.

Disadvantage is that it disables all eye movements (including the vestibulo-ocular reflex and vergence), so that it is only useful while the patient is stationary and views monocularly. Field of view is limited. Patients with ataxia or tremor (such as those with multiple sclerosis) have difficulty inserting the contact lens.

(C) Electrical devices

1. Movement based:
   A more recent innovation is to use an electronic circuit to distinguish between the nystagmus oscillations and normal eye movements. This approach is most useful in patients with pendular nystagmus. Eye movements are measured with an infrared sensor and fed to a phase-locked loop that generates a signal similar to the nystagmus but is insensitive to other eye movements, such as saccades. This electronic signal is then used to rotate Risley prisms, through which the patient views the world. When the Risley prisms rotate in synchrony with the patient's nystagmus, they nullify the visual effects of the ocular oscillations. Improvement and miniaturization of a prototype device may eventually yield spectacles that selectively cancel out the visual effects of pathological nystagmus.

2. Biofeedback based:
   Electrical stimulation or vibration over the forehead or neck may suppress congenital nystagmus, again possibly by an action on the trigeminal system, which receives extra ocular proprioception.

(D) Acupuncture

Acupuncture consisting of the insertion of needles into the sternocleidomastoid muscle, has been shown to improve foveation characteristics in congenital nystagmus on a temporary basis.

(E) Botulinum toxin

Injection of botulinum toxin into either the extraocular muscles or the retrobulbar space has been reported to reduce nystagmus and improve vision in some patients. Limitations of this approach are the short period of action (2–3 months), ptosis and diplopia, which may be more annoying to patients than visual symptoms due to the nystagmus. In some patients, the nystagmus may become worse in the non-injected eye, if the patient prefers to view with the injected eye. This is because botulinum toxin weakens all types of eye movement, not just the nystagmus. This paresis of normal movements stimulates the brain to make adaptive changes by increasing innervation that may worsen the nystagmus in the non-injected eye.

(F) Drugs

Advocated to treat congenital nystagmus, and improvement of visual acuity has been reported in some instances.

Drugs not preferred because of their side effects and need for prolonged treatment. Ex: Gabapentin, Scopolamine, BaclofenIsoniazid, Memantine, Carbamazepine, Clonazepam, Barbiturates, Valproate, Alcohol, Trihexyphenidyl, Cannabis, Benztropine, Acetazolamide.

Surgical management

Indications for surgical intervention:

1. Large face turns—more than 40 degrees.

2. Associated with strabismus.

3. Successful prism adaptation.

Principles of surgical management:

(a) To improve head posture—move the eyes toward the null position.
1. Kestenbaum procedure.

2. Augmented Kestenbaum procedure.

3. Modified Anderson procedure—the two muscle recession.

(b) To improve the visual acuity.

1. Four muscle recessions.

Kestenbaum procedure:

In this procedure rule of 13 is followed wherein each eye the amount of surgery performed is 13 mm including the medial and the lateral recti. In each eye, the yoke muscles are recessed and resected according to m the desired shift in position. For example, for a left face turn the left lateral rectus is recessed 7 mm and left medial rectus is resected 6mm (making a total of 13 mm) and the right medial rectus is recessed 5 mm and lateral rectus is resected 8 mm.
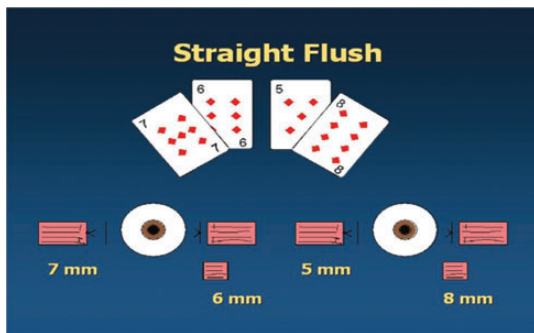
Left face turn

Augmented Kestenbaum procedure:

For larger face turns, there is a modification of the above procedures which can be followed.

Anderson two muscle recession surgery:

This is a more conservative procedure where recessions of only two recti are done on the agonist muscles for mild to moderate degrees of face turn.

For vertical head positions:

1. Chin down: bilateral superior recti and inferior oblique recessions.

2. Chin up: bilateral inferior recti and superior oblique recessions.

For head tilts:

1. For moderate tilts: superior oblique and fellow inferior rectus weakening.

2. For severe tilts: weakening of the two incyclotorsional muscle in one eye and the two excyclotorsional muscle of the fellow eye.

For improving visual acuity without null position:

1. Four horizontal recti recessions 12– 4 mm.

When there is coexisting strabismus:

1. Best guess dosage.

2. Staged–first correct strabismus and then the null point.

3. Adjustable techniques.

Conclusion:

1. In reality nystagmus has no cure.

2. Treatment plans should be tailored.

3. Goals should be realistic.

4. Recurrences is almost the rule.

Pediatric low vision care services: a new era

Sarika Gopalakrishnan and Sumita Agarkar

Introduction

The major causes of blindness in children vary widely from region to region, being largely determined by socioeconomic development, and the availability of primary health care and low vision care services.[1,3] The prevalence of blindness in children ranges from ~0.3/1000 children in affluent regions to 1.5/1000 in the poorest communities. Reliable population-based data on the causes of blindness in children are difficult to obtain in developing countries.[2] There is an increasing awareness about the needs of students with low vision, particularly in developing countries where programs of integrated education are being developed. However, the appropriate low vision services are usually mandatory in order to improve their residual vision.

Definition of low vision

A child with low vision is one who has impairment of visual functioning even after treatment and/or standard refractive correction, and has a visual acuity of <6/18 to light perception, or a visual field of <10° from the point of fixation in the better eye, but who uses, or is potentially able to use, vision for the planning and/or execution of a task for which vision is essential (WHO, 1992).

Impact of low vision

Functionally, low vision is described as irreversible visual loss and a reduced ability to perform many daily activities, such as recognizing people in the street, reading blackboards, writing at the same speed as peers, and playing with friends.

Causes of low vision

The major causes of visual impairment among children includes Stargardt's disease, Myopic degeneration, Oculocutaneous Albinism, Retinitis Pigmentosa, Maculopathy, Optic Atrophy, Corneal opacities, Rod cone dystrophy and other heredo-macular degenerations.[3]

In the year 2010

The mean number of children seen in the Low Vision Care clinic per year was found to be 1500. The referral criteria included children with low vision and constricted field of vision.

Approach	Before 2010	After 2010
History	General history, family history and birth history	Classroom environment history, academic performance
Vision assessment	Regular method of vision assessment with usual logMAR chart	Functional vision assessment for pre-verbal children with Cardiff acuity card, rudimentary vision kit and LEA symbols
Management	Usually spectacles or optical magnifiers would be the choice	Additionally a wide range of non-optical devices and electronic assistive devices are available
Advice	Generalized case summary would be given	Specific letter to school management is being given based on the needs (e.g. seating arrangements, compensatory time during examinations, use of low vision devices, etc.)
Referral	Most of the times children will be referred to blind school	Children will be referred to integrated or inclusive mode of schooling

New initiatives

1. Awareness on referral criteria among eye care professional including Ophthalmologists was created.

2. The range of optical devices and non-optical devices were extended significantly, so that appropriate magnification was chosen for trial and final prescription.

3. Latest varieties of electronic assistive devices were introduced from different parts of the world.

4. Tools and devices used for daily living activities were included in the stock.

5. All the devices were purchased from all over the world and for lower cost.

6. Primary low vision care service was introduced in all the floors and branches of the tertiary eye care center, so that more number of patients got benefited.

7. Organizational membership was received from Bookshare for accessing DAISY books, which can be useful for patients with print disabilities.

8. Psychological counseling on self-motivation and need for special schooling are being provided to the parents whose children are visually impaired.

9. Braille training and Computer JAWS software demonstration are provided for children with low vision.

10. Smart Cane demonstration for orientation and mobility training.

In the year 2015
The mean number of children seen per year has increased to 3200 which is twice the amount seen 5 years back. The referral criteria have been extended such that pre-schoolers are also referred to the low vision care clinic in order to decide their mode of education and for appropriate guidance.

Transformation in the trend

Sl. No	Visual task	Before 2010	2010–2014	From 2015
1.	Viewing board	Approach magnification	Conventional telescope	ONYX electronic assistive device
2.	Reading tasks	Bifocal spectacles	Dome magnifier	Mouse model CCTV/ portable CCTV
3.	Writing tasks	Cut away stand magnifier	Spectacle magnifier	Quick look zoom CCTV
4.	Watching TV	Approach magnification	SEE TV binocular telescope	Snow 7HD video magnifier
5.	Computer	External screen magnifier	In-built modifications/ JAWS/MAGIC	Newer software/latest technologies
6.	Mobile	Tactile cues for keypad	TALKS	In-built applications/ android accessibility settings
7.	Non-optical devices	Four lined notebooks	Typoscope/letter writer	Thick lined notebooks
8.	Writing tool	Scribe	Writing in larger size	Bold pencils/bold pens
9.	Activities of daily living			
	Time calculation	Help from others	Large dial high-contrast clock	Talking clock
	Color identification	Help from others	Color labeling of pencils/pens	Color recognizer
	Currency identification	Help from others	Notex	Look tel money reader APP
10.	Recreation	Nil	Increased task illumination	Jumbo playing cards

Mode of schooling

1. **Mainstream:** School with normal children without any disability.

2. **Integrated school:** A school where both normally sighted and visually impaired students will be studying in the same class.
    (a) **Resource model:** The school is provided with the resource set up, where a teacher will be available all the day. During the free hours, the students with visual impairment go to the resource room for guidance.

    (b) **Itinerant model:** The Resource teacher (Special Educator) goes to different schools and visually impaired children's place to teach them.

3. **Inclusive school:** Mild to moderate form of all types of differently abled children will be included along with the normal children.

4. **Special school:** A school for visually impaired/ deaf-mutism/mentally retarded/learning disability/any specific disability.

Rights and consideration
Scholarship

1. The Union Ministry of Welfare since 1955 has been operating through the state Governments

and Union territories a scheme of scholarships awarded to challenged person for pursuing education in special schools being run by non-government organizations.

2. The Rate of scholarships is Rs.1000/- per annum for cases hailing from the lower socio-economic status and is renewable from year to year.

3. In case of severely challenged persons who require special arrangements for transportation, an additional monthly allowance of Rs.50/- is sanctioned.

The following allowances and facilities are provided under this scheme

1. Books and Stationary allowance of Rs.400/- per annum.

2. Uniform allowance of Rs. 50/- per annum.

3. Transport allowance of Rs.50/- per month (if a challenged child admitted under the scheme resides in a hostel of the school within the school premises, no transportation charges would be admissible).

4. Reader allowance of Rs.50/- per month in case of blind children after class V.

5. Escort allowance for severely handicapped children with lower extremity disabilities at the rate of Rs.75/- per month.

6. Annual cost of equipment subject to a maximum of Rs.2000/- per student for a period of 5 years.

7. The tuition payable and actually paid by the Government servant is reimbursable subject to Rs.50/- per month per child in the case of multiple disabled children.

Railways

1. 75% concession in the basic fare in the first and second class is allowed to persons with multiple disabilities accompanied by an escort.

2. 50% concession in the first and second class monthly/quarterly season fares both for the individual with disability and his/her escort over suburban and non-suburban section of Indian railways is allowed.

Roadways

1. Most State Governments having state owned and operated transport undertakings or corporations allow subsidized/free bus travel in the city and rural routes, and an escort is charged fifty percent of the fare.

Conclusion

Majority of the students with low vision need low vision services in order to perform better. Accurate refraction is important in the students. Early diagnosis and intervention helps in preventing the vision loss which will lead the students to continue their academic activities without any interference. Many children need additional support from the Government in order to continue their education. The need for awareness on management and referral of children with Low vision has to be improved in this Millennium due to increasing number of children who discontinue their schooling due to Low vision. Awareness among eye care professionals and parents is mandatory for bringing up these children with Low vision.

References

1. **Gilbert Clare AllenFoster.** Childhood blindness in the context of VISION 2020: the right to sight. *Bull World Health Organ* 2001; **79.3**: 227–232.

2. **Sharma MK,** *et al.* A profile of low vision among the blind school students in Lumbini Zone of Nepal. *Nepalese J Ophthalmol* 2010; **2.2**: 127–131.

3. **Shah Mufarriq,** *et al.* Causes of visual impairment in children with low vision. *J Coll Phys Surg Pak* 2011; **21.2**: 88–92.

Case 1:

General history: A 14-year-old female diagnosed with High Myopia and associated retinal finding of dry macula (Myopic Retinal Degeneration) was referred to the Low Vision Care Clinic as she had discontinued school 3 months back. She was accompanied by her parents. There was no family history of parental consanguinity. Her younger sister had myopia (around -3.00D and no visual impairment). There was no other relevant history.

Previous low vision care: nil

Present complaints:

Distance visual tasks: The patient had difficulty seeing the blackboard and recognizing the faces of people who were more than 3 m away. She watched television from a distance of 1 m.

Near visual tasks: The patient was able to manage at a close working distance of around 15 cm.

ADL/mobility: These tasks were independent.

Light sensitivity: There were no complaints regarding light sensitivity.

Additional history: Classroom environment

School: Private school (English medium), State Board of Education syllabus.

Medium of teaching: English.

Class strength: 30.

Academic performance of patient: Average (50–60%).

Reason for discontinuing school: Unable to see blackboard from front bench at a distance of 3 m.
　Parents say teacher wrote letters of size about one finger length (~2.5 inches or 6–7 cms).

Color of board: Black.

Letters: White chalk used, colored chalk: rare.

Class lighting: Sufficient (no glare and no complaints of dim/poor lighting, including cloudy days).

Internal lighting present in class: three fluorescent lights.

External lighting: three large windows.

Seating position: Third row, center.
　(No flexibility in seating, not allowed to copy or reference friends notebook).

Visual requirements: Seeing blackboard.

Examination:

Previous glass prescription:
　OD: -13.50 DS$/-2.00$C $\times 50$,
　OS: -12.00 DS$/-1.25$ DC $\times 45$.

Distance visual acuity with glasses:
　OD: $3/30^{+2}$, $6/60^{+2}$,
　OS: $3/19$, $6/38$,
　B/O: $3/19$, $6/38$.

Near visual acuity with glasses: OD/OS/B/O: N8 @ 15 cm.

Reading speed: 40 wpm.

Unaided near visual acuity: B/O: N5 @ 10 cm

Reading speed: 60 wpm.

Refraction:

Retinoscopy:
　OD: -11.50DS$/-1.00$ DC $\times 60$,
　OS: -11.25 DS$/-1.25$ DC $\times 120$.

Dynamic retinoscopy: (MEM)

Accommodative Lag: +1.00 DS to +1.25DS

Subjective refraction
　OD: -11.50DS$/-1.00$ DC $\times 60$ [3/9.5, 6/19],
　OS: -11.25 DS$/-1.25$ DC $\times 120$ [3/9.5, 6/19].

ADD: OU: Does not prefer ADD

Unaided near visual acuity: N5 with ease @ 10 cms

Low contrast visual acuity: 3/19 [6/38] @ Borderline impairment.

D15: Normal,

Amsler: No scotoma detected,

Confrontation: Normal peripheral fields.

LVD Trial for distance:

Required visual acuity: 6/6

Magnification required: $19/6 = 3 \times$

Trial 1: 3× Monocular Handheld Telescope: 6/6 with ease.
　Telescope training was given—able to localize, hold fixation, scan, track and copy from text on sample blackboard.
　Copies text at speed of 30–40 wpm.

LVD trial for near: Reads up to N8 with correction.
　Reads up to N4 unaided.

Trial 1: Add +1.00DS to +5.00DS tried in 1.00D steps. Patient does not report significant difference/improvement

Trial 2: With 4× Dome Magnifier: N4; Reads with ease @ 40 cms Reading speed: 80 wpm.

Final Rx:

1. New Rx for distance.

2. 3× Monocular handheld telescope for use with blackboard.

3. To use dome magnifier for prolonged reading tasks, to reduce eyestrain, reference atlas, maps, dictionaries or fine print.

4. To use reading stand for prolonged reading tasks to aid posture and maintain working distance.

Other advice: The condition and prognosis were clearly explained to the parents. The parents were reassured about closer working distance and the need to use a magnifier for comfortable reading at close working distance was explained. The child was advised to continue her schooling with the help of low vision care. They were also counseled about available options for computer modification and magnification.

Case 2:
General history: An 8-year-old-Female diagnosed with Congenital Stationary Night Blindness was accompanied by her parents to the low vision care clinic. Her difficulty with dimly lit environment was noticed by the parents at around 1 year of age. She was staying with her parents and two elder brothers. She was currently studying in Std II. Her parents did not have a consanguineous marriage but her mother's age was around 34 when the child was born. Her father who was

working in an automobile assembly company was the source of family income.

Previous history: She had no history of using any low vision devices.

External observation showed she had good fixation but difficulty in searching for a pen among various items placed on the table.

Present complaints:

Distance visual tasks: Difficulty with seeing the blackboard in the classroom (3 m), seeing objects at distance while sightseeing. She was able to manage all near acuity tasks at a closer working distance.

Mobility and ADL: She was able to move around independently but had difficulty in dim light and took long time to adjust to dimly lit surroundings. She had independent Activities of Daily Living within the limitations of her age.

Additional history: Classroom environment:

1. Seated at 3 m from blackboard.

2. Teacher uses white chalk. Colored chalk used occasionally.

3. Allowed to come close to the board.

4. No note-taking until class IV in the school.

5. Artificial room lighting–five fluorescent lights.

6. Class strength: 40 students.

7. Medium of instruction: English.

8. Type of school: Private.

9. Syllabus: State board.

Patient requirement: Seeing blackboard.

Examination:

Visual acuity:

Distance Visual Acuity with Bailey Lovie LogMAR chart: OD/OS/B/o: 3/12 [6/24]

Near visual acuity: N10 @25 cm with the MN Read chart at 10 wpmgood reading skills.

There was no difference between monocular and binocular visual acuities. There was no significant refractive error on objective refraction.

Dynamic retinoscopy: Accommodative lag: +1.00DS.

Cycloplegic refraction: No refractive error requiring correction detected.

Low contrast visual acuity with Bailey Lovie Low Contrast LogMAR chart was 3/15[6/30] (Within normal limits).

Color vision: Able to match a few colors on the D15 panel but could not comprehend the entire test.

Confrontation: Mild peripheral field deficit beyond 50° visual field.

Trial of low vision devices

(1) 4× monocular handheld telescope: 6/6 (no specific preference for right or left eye)

Near

ADD: OD/OS: +2.00DS: N4 @ 20 cm.

The child seemed to be able to be able to read better and reading speed improved to 25 wpm for finer print (>N10 size).

Final R × :

1. New Bifocal R× with +2.00DS ADD.

2. The parents were advised to purchase over the counter binoculars of 3× or more magnification.

3. A letter was given to the school to provide a separate seating arrangement closer to the blackboard at <2 m when required. (Currently, students were not required to copy any notes from the blackboard.)

Advice:

The child was advised to be seated in a separate chair (in front of the front row) closer to board at ∼2 m. Letter was given to school management recommending closer seating arrangement, increased lighting level and compensatory time for completing her examinations. The parents were advised to increase the illumination level at home especially during night time. The child needs to be instructed on caution regarding mobility tasks. Option of low vision devices were explained to parents which will be beneficial in the future. The patient was advised to come for an annual low vision care visit with her school books and class notes.

APPENDIX 1

<u>CLASS ROOM ENVIRONMENT</u>

Standard: Informant:

Mode of education: a) Special school   b) Regular school   c) Private tutor

School name/College name:

Class teacher/Principal name:

Address of the school/ college:

1. Class strength:

2. Seating position: Center/Corner

3. Approx distance from board:

4. Which bench/row:

5. Color of writing material on board:

6. Color of the board:

7. Number of windows in class:

8. Sitting beside window:

9. Amount of light in class: Dimly illuminated/Optimally illuminated/Glaring (too bright)

Academic performance:

Good peer-group interaction: Yes/No   Good parent–teacher interaction: Yes / No

Is there any difficulty in copying from black board: Yes/No

If yes: managing by:

Copying from friends/Going close to the black board/Taking friends note-books to home/Teacher's dictation.

Others:

Using extra hours for writing exams: Yes/No

Use of special assistance: Yes/No

If yes: Scribe/Braille/Talking books/Audio cassettes

Others:

Since how long:

Any difficulty with special assistance:

Is the teacher co-operative and ready to give individual attention: Yes/No/NA:

Will the school management be ready to make changes in the setup: Yes/No/NA

Letter to the class teacher required: Yes/No/NA

APPENDIX 2

LETTER TO SCHOOL MANAGEMENT

<u>LOW VISION CARE CLINIC</u>   Date:

TO WHOMSOEVER IT MAY CONCERN

This is to state that Master/Miss._____ who is studying ____STD in your school (Reference MRD No.: _____) was evaluated in the Low Vision Care Clinic. He was noted to have _____ Visual Impairment in both the eyes due to retinal problem.

This vision will not be sufficient to view the black board comfortably. He needs to be seated closer to the black board (i.e. 1 m distance) or in the front row (without rotation). Most of his learning will be through listening to the classes, so dictating as much as possible will help him in a large way.

Kindly, allow him to copy notes from his friend's notebook in case he is not able to copy from the black board directly. In addition, he can be given compensatory time concession of minimum 30 min for all examinations (school level and for board exams).

Please extend your co-operation so that his ocular condition doesn't interfere with his academic performance.

Thank you for your kind co-operation.

Yours sincerely,
(_____)
Optometrist
Low Vision Care Clinic
Sankara Nethralaya
Chennai
Tamil Nadu
Phone: 044-42271519

NEURO-OPTOMETRY – A NEW EPOCH IN VISION SCIENCE IN INDIA

Neuro-optometry is a branch of vision science that combines Neurology, Neuro-ophthamology and Optometry. It caters the visual needs of patients with a neurologically insulted brain like after a Trauma, Stroke, Post cranial surgery or even Neuro-generative disease like Parkinson's disease, Myasthenia Gravis etc. Neuro-optometry plays an important role in diagnosing, intervening and rehabilitating the visual disturbances caused by brain injury. The clinic focuses to advance the art and science of understanding, diagnosing and rehabilitating the neurologically challenged population thereby aiming to improve their functional vision. The department of Optometry at Sankara Nethralaya has initiated this specialty clinic called the "Neuro-Optometry Clinic" (NOC), first of its kind in India. The clinic gets its patients referred primarily from departments of Neuro-ophthalmology, Pediatric & Strabismology and Oculoplasty.

Individuals with brain injuries have varied forms of vague visual symptoms functionally affecting the activities of daily living. The following lists the symptoms of such patients with Traumatic Brain Injury/ Stroke:

- o Diminished vision for far and/or near
- o Diplopia, squinting of eyes, restriction of eye movements
- o Headaches, eyestrain, eyepain
- o Reading difficulties, difficulties in tracking while reading
- o Difficulty to focus from distance to near or vice-versa
- o Dizziness/ nausea, vertigo, visual motion sensitivity, spatial disorientation and balance disorders
- o Convergence and accommodative deficit
- o Visual field defects & Visual midline shift
- o Difficulty in cognitive ability, difficulty in concentration, visual neglect and visual processing
- o Right-left discrimination

- Difficulty in object recognition (Agnosia) and difficulty in manipulation of objects (Apraxia)
- Memory loss
- Psychological problems
- Visual hallucinations
- Light sensitivity and Dry eyes

The Neuro-Optometry Clinic performs various visual function tests that includes detailed history (Details of brain injury, loss of consciousness following trauma, ENT bleeding, classification of brain injury, details of Surgery done, Imaging reports, other rehabilitation procedures provided, specific ocular and non-ocular symptoms), Refraction with binocular balancing techniques, Sensory Motor evaluation, a Comprehensive Binocular Vision Assessment (Manual/ Computerized), tests of Fixations, Pursuits and Saccadic evaluations using subjective and objective test procedures, assessment of Reading speed, tests of Visual Perceptual Skills, Confrontation , Cranial Nerves evaluation, Diplopia and Hess charting (for diplopia), Vestibulo-ocular reflex, tests of Visual Midline Shift, Dynamic visual acuity and Tests of Visual Motion Sensitivity.

VISION REHABILITATION AT Neuro-Optometry Clinic

The clinic follows a specified protocol as followed by the International expertise in the field of Neuro-optometry. The clinic utilizes Special lenses, Fresnel prisms, Addition lenses, Binasal occluders, Field expanders, Yoked prisms, Filters and Bangerter foils to help these patients improve the function of their visual potentials which are affected due to the brain injury. In addition, extensive vision therapy is provided to those with associated binocular vision anomalies, eye movements dysfunctions and poor eye-hand co-ordinations. The brain functions are trained by the tests of visual perceptual skills using perceptual software both in-office and at home and the performance can be tracked for reinforcement.

Reach us at

Neuro-Optometry Clinic, Room No: 517, Fifth Floor, Mahyco Block, SN Main, Chennai: 600006. Contact No: 044 42271517, Intercom: 1517
Email: neurooptometry@snmail.org

Cortical visual impairment

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Cortical visual impairment (CVI) is emerging as one of the important causes of blindness or visual impairment in India. This increasing trend is likely to continue as further advances in the field of neonatology and fetal medicine increase the chances of survival of a premature infant or an infant with multiple congenital abnormalities. Previous studies from developed nations have shown that infants born prematurely and those with low birth weight are at a risk of severe ophthalmic impairment.[1,2]

What is CVI?
The current definition of cerebral or CVI includes all visual dysfunctions caused by damage to or malfunctioning of the retrochiasmatic visual pathways in the absence of damage to the anterior visual pathways, or any major ocular disease.[3]

We need to remember that CVI does not imply total blindness and is a spectrum of vision loss ranging from mild visual impairment to complete visual deprivation.

Pathogenesis
The central nervous system in a newborn is susceptible to various damaging influences such as hypoxia–anoxia, hypoglycemia or a combination of both. Two distinct subgroups of cortical visual loss in terms of the areas of damage and the time of injury have been described.[4] Injury in full term neonates, predominantly involves the striate and peristriate cortex, whereas in preterm neonates, injury involves the subcortical white matter, including the optic radiations. This results in different patterns of damage as seen on neuroimaging and also results in different clinical manifestations.

In a full term neonate, the vascular supply of the brain is derived from the major cerebral arteries and its watershed areas lie at the interfaces between the major cerebral arterial distributions.[5–7] Hence, any hypoxic-ischemic injury produces watershed infarctions in the parieto-occipital and parasagittal cortex, resulting in cortical visual loss. Whereas, in the developing brain, the cortex and underlying white matter receive their blood supply from branches of the blood vessels on the surface of the cerebral hemispheres and the watershed zone lies within the periventricular white matter.[8] Hence, preterm injury to the brain results in injury to the subcortical white matter, resulting in periventricular leukomalacia (PVL).[8] Other causes of injury in neonates include birth trauma, intraventricular hemorrhage and hypoglycemia. Neonatal hypoglycemia, in particular, in known to cause significant visual deficits and neuroimaging in these cases shows restriction on diffusion predominantly in the occipital areas.[9]

Causes
A wide variety of insults to the central nervous system can result in CVI. These include: perinatal conditions such as birth trauma, hypoxia–anoxia, hypoglycemia and post-natal conditions such as sepsis, meningitis, encephalitis, accidental and non-accidental trauma, cerebral malformations such as Chiari malformations, Dandy–Walker complex, hydranencephaly, porencephalic cysts, metabolic and neurodegenerative conditions such as MELAS (metabolic encephalopathy, lactic acidosis and strokelike episodes), Fabry's disease, Leigh's disease, X-linked adrenoleukodystrophy and hydrocephalus.[10]

Clinical presentation and diagnosis
As mentioned previously, CVI can manifest as mild visual loss with a slight delay in achievement of visual developmental milestones or as near total or total blindness. These babies are most frequently brought to the ophthalmologist with complaints of "not seeing the mother's face", poor visual response to light, "shaking of eyes" or nystagmus.

These children often have multiple other disabilities and one must remember to make the child comfortable before starting the examination. Responses to bright light, colored objects and human faces must be assessed. Some of these babies may respond better in a supine position and in dim light.

One must look for the presence of nystagmus, strabismus and ocular motility deficits and pupillary responses. Cycloplegic refraction must be done along with a thorough fundus examination.

Nystagmus is rare or absent in children with CVI. However, roving eye movements or occasional bursts of nystagmus may be seen and the presence of such eye movements signifies severe visual impairment.[4] An intact geniculostriate pathway is a prerequisite for the development of congenital nystagmus and hence nystagmus is absent in extensive posterior pathway disease.[11] However, some children with combined anterior and posterior visual pathway disease may present with CVI and nystagmus.[12]

Strabismus is common in children with CVI. Both esotropia and exotropia have been reported. Esotropia is more common in children with PVL and may be associated with latent nystagmus, dissociated vertical deviation and 'A' pattern.[10] Other studies have shown that cortical visual loss is associated with exotropia more commonly than esotropia. Also, children with more extensive

neurological damage may have congenital exotropia.[10,12]

Other ocular motor abnormalities that have been noted in these children include tonic downgaze, horizontal conjugate gaze deviation and defective smooth pursuits and saccadic eye movements.[10,13]

Fundus examination is extremely important to detect the presence of associated optic atrophy and also to rule out other conditions with similar clinical presentation such as retinal dystrophies, congenital macular scars, sequelae of retinopathy of prematurity, etc.

In cases where the pathological processes affecting the posterior visual pathways and the cerebral cortex have not affected the anterior visual pathway, the fundus is usually normal and the disk appears healthy. However, studies involving children with retrogeniculate vision loss, have described a variety of optic nerve abnormalities including optic atrophy, optic nerve hypoplasia and pseudoglaucomatous cupping.[4,12]

In additional to structural damage, one must keep in mind that these children also have significant functional deficits such as visual field defects, which include generalized field restriction, central scotomas, homonymous hemianopias and altitudinal defects.[10,12]

One must keep in mind that vision is a complex function and has multiple components. These children may show other visual functional abnormalities which may be difficult to quantify, such as difficulties in visual processing, difficulty in simultaneous perception and "crowding" phenomenon. Other peculiar visual traits include a tendency to stare at bright objects, such as fluorescent room lights or the sun. This is known as "light gazing" and is regarded as a sign of severe visual impairment.[14]

These children also, often function better in a familiar environment, may show a better visual performance for objects in motion as compared with static objects and are better at identification of colors as compared to their perception of form.[15] Some children also complain of photophobia.

Diagnosis is confirmed by neuroimaging. While imaging of the infant brain is possible by ultrasound and computed tomography, magnetic resonance imaging (MRI) is the preferred modality. MRI can help to pick up even the subtle changes of PVL such as abnormal dilatation and irregularity of the lateral ventricles, high intensity signals in the periventricular white matter on T2 weighted images and periventricular gliosis. Severe hypoxic injury may lead to encephalomalacia with surrounding gliosis.[10] Other abnormalities seen include thinning of the corpus callosum, altered signals from the thalamus and putamen and cerebellar atrophy.[10,12]

It is not clearly known to what extent PVL and CVI and the associated neurological abnormalities affect visual evoked potentials (VEP), but in general, the absence of a flash VEP is associated with a poor prognosis.[10]

Electroretinogram may help by ruling out retinal dystrophies, which may sometimes present a similar clinical picture.

Management

Early diagnosis and intervention is the key to management in these cases. A team approach integrating the services of a pediatric ophthalmologist, pediatrician, physiotherapist, speech therapist and vision therapist is desirable. Parental counseling is of utmost importance and one must remember that while it is definitely important to provide the parents a realistic picture of their child's condition, encouraging the parents to start rehabilitation and remain positive in their outlook may go a long way toward bettering the child's life.

References

1. Cooke RW, Foulder-Hughes L, Newsham D, Clarke D. Ophthalmic impairment at 7 years of age in children born very preterm. *Arch Dis Child Fetal Neonatal* Ed 2004; **89**: F249–F253.
2. Robaei D, Kifley A, Gole GA, Mitchell P. The impact of modest prematurity on visual function at age 6 years: findings from a population-based study. *Arch Ophthalmol* 2006; **124**: 871–877.
3. Boot FH, Pel JJ, van der Steen J, Evenhuis HM. Cerebral visual impairment: which perceptive visual dysfunctions can be expected in children with brain damage? A systematic review. *Res Dev Disabl* 2010; **31**: 1149–1159.
4. Brodsky MC, Fray KJ, Glasier CM. Perinatal cortical and subcortical visual loss: mechanisms of injury and associated ophthalmologic signs. *Ophthalmology* 2002; **109**: 85–94.
5. Flodmark O, Jan JE, Wong PK. Computed tomography of the brains of children with cortical visual impairment. *Dev Med Child Neurol* 1990; **32**: 611–620.
6. Barkovich A. Pediatric Neuroimaging. Philadelphia: Lippincott Williams & Wilkins, 2000.
7. Volpe J. Neurology of the Newborn, 3rd ed. Philadelphia: W.B. Saunders, 1995; 403–463.
8. Barkovich A. Pediatric Neuroimaging. Philadelphia: Lippincott Williams & Wilkins, 2000.
9. Tam EW, Widjaja E, Blaser SI, *et al.* Occipital lobe injury and cortical visual outcomes after neonatal hypoglycemia. *Pediatrics* 2008; **122**: 507–512.
10. Brodsky M. The Apparently Blind Infant. *In Pediatric Neuro-Ophthalmology, Edition.* New York: Springer, 2010; 1–58.
11. Fielder AR, Evans NM. Is the geniculostriate system a prerequisite for nystagmus? *Eye (Lond)* 1988; **2** (Pt 6): 628–635.
12. Jacobson LK, Dutton GN. Periventricular leukomalacia: an important cause of visual ocular motility dysfunction in children. *Surv Ophthalmol* 2000; **45**: 1–13.
13. Jacobson L, Ygge J, Flodmark O. Nystagmus in periventricular leucomalacia. *Br J Ophthalmol* 1998; **82**: 1026–1032.
14. Jan JE, Groenveld M, Sykanda AM. Light-gazing by visually impaired children. *Dev Med Child Neurol* 1990; **32**: 755–759.
15. Jan JE, Groenveld M, Sykanda AM, Hoyt CS. Behavioural characteristics of children with permanent cortical visual impairment. *Dev Med Child Neurol* 1987; **29**: 571–576.

List of fellows trained by Department of Pediatric Ophthalmology, Sankara Nethralaya

Year	Initial	Name
April 1999	HAM	Dr Hemalini Mehta
April 2001	MK	Dr Madhu Karna
April 2002	AMJ	Dr Aarti Jhamwar
October 2002	KKV	Dr Kavitha Kalaivani Natarajan
October 2003	LK	Dr Lov Kochgaway (Orbis)
	MSH	Dr Manish Sharma (Orbis)
April 2004	ST	Dr Satish Thomas (Orbis)
	RSA	Dr R Sudhindra (Orbis)
October 2004	SKL	Dr Shalini Kaul (Orbis)
	ASV	Dr Anand S Vajawade (Orbis)
April 2005	HLV	Dr Hemalatha Vidyashankar (Orbis)
	RPH	Dr Ramprakash (Orbis)
	SSY	Dr Sharath Shetty (Orbis)
October 2005	CB	Dr Charuta Bhadre (Orbis)
	CSZ	Dr C Sadiq Zamir (Orbis)
	AAT	Dr Amol Anil Tamhane (Orbis)
October 2006	SRK	Dr Sai Rani (Orbis)
	KM	Dr Kaushik Murali (Orbis)
	VAS	Dr Varshini Shankar (SN)
	MRC	Dr Mayee Rishikesh Charudatta (SN)
April 2007	NUV	Dr Naik Uday Vithal (Orbis)
	PSM	Dr Prashant kumar S Murhe (Orbis)
October 2007	AMA	Dr Amit Maitreya (Orbis)
	BCK	Dr Basavaraju C Kadiwal (SN)
April 2008	JGM	Dr Jay Mahida (Orbis)
	BPA	Dr Bhujbal Pritamsingh Appasaheb (Orbis)
October 2008	ATK	Dr Ajit Kumar Sinha (Orbis)
	PNT	Dr Parag N Tupe (Orbis)
October 2009	AGM	Dr Amula Gangaprasad Muthaiah
	PAP	Dr Preeti Ajit Patil
April 2010	HGM	Dr Harsha G Mangalgi
	RBR	Dr Resmi Bhaskar
October 2010	PBP	Dr Prathiba Patil Bapusaheb (SNK)
	SFK	Dr Mohd Sarafraz Khan
	RRA	Dr Renu R Athanikar
April 2011	RSK	Dr R Srikanth
	VAR	Dr Akila Ramkumar
	RMA	Dr Radhimalar Anand
	SVS	Dr Sanil Vijaybhai Shah (SNK)
October 2011	SDN	Dr Sujay Siddharthan
	AKR	Dr Anushree Vijay Kaduskar
	PMH	Dr Preeti M Hurakadli (SNK)
April 2012	SGI	Dr Shama Ali Goliyadi
	NJC	Dr Nidhi J Chopra
October 2012	RJD	Dr Roshani Jaideep Desai
October 2012	STS	Dr Sudipta Sahana
October 2012	SPR	Dr Shalaka Ravindra Paralkar
October 2012	AE	Dr Aarti Elhence
April 2013	MSR	Dr Shruti Rajendra Mittal
April 2013	ARN	Dr Anbarasi Sherren
October 2013	SNA	Dr Soumya Nambiar
October 2013	JSP	Dr Jayesh Suresh Patil
October 2013	KSH	Dr Khyati Jain
April 2014	BRV	Dr Bhatt Riddhi Vijay
April 2014	PPL	Dr Prakash Parimal
April 2014	VVG	Dr Varada Vinay Gokhle

Lessons learnt in Paediatric ophthalmology

Opthalmic and otherwise!

Dr T S Surendran

Vice President,
Medical Research Foundation,
Director,
Paediatric ophthalmology,
Sankara Nethralaya,
Chennai

Correspondence to:
Dr T S Surendran,
Vice President,
Medical Research Foundation,
Sankara Nethralaya,
Chennai
Email: drtss@snmail.org

Many summers back when I joined Sankara Nethralaya, as was the trend then I was trained in vitreoretinal procedures but somewhere down the line it was decided that I should do Paediatric ophthalmology. Thirty seven years later when I look back, it was a momentous decision that changed my life in many ways. I was fortunate to train under doyens of this field like Dr Marshal Parks, Dr Arthur Jampolasky, Dr Alan Scott and Dr Prem Prakash. They were true visionaries and pioneers who changed the way world looked at Paediatric ophthalmology.

Being a Paediatric ophthalmologist has taught me lessons in patience, perseverance and hope. I have learned to communicate with non-verbal children, special children as well as parents and often grandparents too. I have learned to take in my stride when the room furniture gets rearranged by the patient while I talk to the patient. I am not surprised when an enveloped mail to me contains lenses from the trial set which parents have discovered after reaching home. I learnt not to assume anything when an elderly couple brought in a young child who turned out to be their sixteenth offspring. Similarly, I learnt to be non-judgmental when another of my overseas patient requested a squint correction so that he could marry a third time!

In a career spanning 37 years and still counting, there have been many changes in the way we practice ophthalmology, but one thing that has remained unchanged is the team work. Paediatric eye care, in particular, needs a team of optometrists, anaesthetists, paediatricians, nurses and counsellors. Together they make us look good as doctors. I am grateful for working with a dedicated team. Lest I forget, the team also includes parents and caregivers who need reassurance and empathy from us while being honest about the goals of treatment. It is their trust in us that allows us to work with children. It is a responsibility which should not sit lightly. This means constant vigilance, checking whether it is the correct eye, correct patient, correct surgery and then doing it all over again once more!

However, all said and done, if clock were to turn back, I am sure I will choose Paediatric ophthalmology all over again.

Department of Paediatric Ophthalmology

Sankara Nethralaya

From left to right: V Akila Ramkumar, N Kavitha Kalaivani, Sumita Agarkar, T S Surendran, Meenakshi Swaminathan, Srikanth Ramasubramanian