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Dear colleagues and friends!!

We as ophthalmologists have witnessed a rapid and striking technological evolution with aims that are beyond just 6/6 vision. During the last few years, there has been upheaval in surgical instrumentation, imaging and intraocular lenses. But there is a single sub-speciality which is minimally or least affected by our recent armamentarium i.e. pediatric ophthalmology and strabismus.

The basic principles to examine the pediatric eye have remained grossly the same. But the residents and most academicians need access to a recapitulation of these basic examination techniques. The present issue highlights these techniques along with a pinch of newer diagnostic tools. The anti VEGFs have carved their way into pediatrics through preterm babies and stereovision is the new goal of a strabismologist. We have experts teaching us the basic surgical steps as well small cases highlighting largely accepted norms.

In summary, this issue transforms a whole chunk of pediatrics in ophthalmology to a booklet in your hand.

Happy reading!!

Thanks

Dr. (Prof.) Subhash C. Dadeya
Secretary - Delhi Ophthalmological Society
Room No 114, 1st Floor, OPD Block,
Guru Nanak Eye Centre, Maharaja Ranjit Singh Marg,
New Delhi - 110002
Email: dadeyass@gmail.com, dadeya868@gmail.com
Mobile: 9968604336, 9810575899
WhatsApp: 8448871622
CONFEREE HIGHLIGHTS

› LIVE SURGERY
› SCIENTIFIC DELIBERATIONS IN CATARACT & REFRACTIVE SURGERY, RETINA, CORNEA, GLAUCOMA, SQUINT & NEURO-OPHTHALMOLOGY AND OCULOPLASTY
› WET LABS
› YOUNG OPHTHALMOLOGIST SESSION
› INTERESTING CASES
› FREE PAPER
› DEBATES
› POSTER SESSIONS
› E-VIDEOS
› EARLY BIRD PRIZES
› AN EXCITING FELLOWSHIP DINNER
› QUIZ
Greetings from Delhi Ophthalmological Society!

On behalf of the Executive Committee of Delhi Ophthalmological Society, it is our great pleasure to invite you to the Winter Conference of Delhi Ophthalmological Society (DOS) to be held on 17th and 18th November, 2018 at India Habitat Center, New Delhi, India. This Conference will bring together practitioners, researchers and educators from India and abroad who are engaged in the field of ophthalmic academic work.

The Delhi Ophthalmological Society, with over 9000 members across India is committed to furthering the cause of continuing medical education and promoting camaraderie and fellowship amongst ophthalmologists worldwide. The DOS conference is a much-awaited, time-honored academic fiesta, which attracts faculty, trade exhibitors and delegates from all over the world.

The conference will be a two-day celebration of ophthalmology featuring the best works in the fields of Cataract, Refractive Surgery, Cornea, Glaucoma, Strabismus, Neuro-ophthalmology and Cataract surgery. The diverse program will feature plenary lectures, symposia, instruction course, wet labs and live surgery demonstration, Epistew, Video and Interesting Cases. A Session for young ophthalmologists will be a special addition to our diverse program.

As you will venture out of the confines of the state of the art conference hall and the world of ophthalmology, the vibrant Delhi winters, along with its legendary hospitality and bustling energy will enthrall you. After the academic extravaganza, will be the captivating cultural program and fellowship dinner, true to DOS traditions.

We would be delighted to have you present at this conference, and to hear what the experts, gurus and researchers have to share about technology advancements and their impact on Ophthalmology. We would also love to hear your thoughts and opinions to this direction.

We look forward to greeting you at the Winter Conference of Delhi Ophthalmological Society, at India Habitat Centre Delhi from 17th to 18th November, 2018.

Prof. S.K. Khokhar  
President, DOS

Prof. Subhash Dadaya  
Secretary, DOS

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REGISTRATION FEE

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**IMPORTANT NOTES**

* Inclusive Tax  
* **Proof of residency required from HICD along with the registration form of the conference.**  
* *Registration for Spouse/child must be done separately for each person.  

Bank charge as applicable on all online transaction.

For Spot Registrations, Complete Kit subject to availability.

- Wearing of identity badge is mandatory at all times.
- Entry to Scientific Sessions, Exhibition Area, Felicitations Ceremony will be restricted to Registered Delegates only.
- Last date will be replaced at the registration counter for a fee of Rs. 300/-.
- Pre-registration closes on 30th October. Past President of DOS or AOS and Senior Member (> 70 years) will be registered complimentary provided their registration form is received till 30th October.
- DOS life members Senior Citizen above >70 years are requested to send the registration from along with a proof of age.
- Cancellation & Refunds: Cancellation is permitted up to 30th October 2018 only against a written request submitted to the conference secretariat and 60% of the registration fee would be deducted as processing charges. No Cancellation requests will be accepted thereafter.
- Attendance certificate will not be issued to Associate Delegates, Trade Delegates and Optometrists.

Conference Organising Committee

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Address for All Correspondence

Dr. Subhash C. Dadaya  
Room No. 114, 1st Floor, OPD Block, Sana Narain Eye Centre, Maharaaj Ranjit Singh Marg, New Delhi-11 9002
Ph. +91-11-23210810  | Email: info@dosis18.org / dosirecords@gmail.com  | Web: www.dosis18.org / www.dosisonline.org
Esotropias are one of the most discussed and researched entities of Strabismus. With this article we aim to discuss the most common of the esotropias that is the accommodative esotropia. Experts around the world were questioned about their practice patterns in treating patients with accommodative esotropia.

(FM): Frank J. Martin, Clinical Professor University of Sydney VMO Sydney Children’s Hospitals Network and Sydney Eye Hospital
(JS): John Sloper, FRCS, FRCophth, Honorary Consultant, Strabismus and Paediatric Service, Moorfields Eye Hospital, City Road, LONDON.
(DS): Derek T Sprunger, MD, FAAO, MAAPPOS. Associate Clinical Professor, IUSOM, Indianapolis, in USA
(SL): Scott Larson, Scott A. Larson, MD. William E. Scott Professor of Pediatric Ophthalmology Department of Ophthalmology and Visual Sciences, University of IOWA. USA
(SP): Stacy L. Pinealess, MD, MS, Jerome and Joan Snyder Chair in Ophthalmology, Director, Ophthalmology Residency Program, Pediatric Ophthalmology, Adult Strabismus, Neuro-Ophthalmology, Stein Eye Institute, UCLA Department of Ophthalmology, Los Angeles, California, USA.
(RG): Maria Rosario Gomez De Liaño Sanchez, Professor of Ophthalmology at Universidad Complutense de Madrid (Spain), Head of the Department of Motility Unit at Hospital Clínico San Carlos (Madrid - Spain), ISA President (International Strabismological Association), IPOSC Secretary - Treasurer (International Pediatric Ophthalmology and Strabismus Council).
(SD): Dr. Subhash Dadeyya, Director Professor of Ophthalmology, Guru Nanak Eye Center, Maulana Azad Medical College, New Delhi, India. President Strabismus and Pediatric Oph Society of India 2017-18 and Secretary, Delhi Ophthalmological Society.

The questions have been prepared by (SK): Dr. Savleen Kaur, Senior Research Associate, Advanced Eye Center, Post Graduate Institute of Medical Education and Research, Chandigarh, India. Executive Editor DOS Times.
(SK): Do you see esotropia more commonly than exotropia in your clinical practice?

(FM): In our clinical practice we see an equal number of esotropes and exotropes.

(JS): Yes, considerable more frequently.

(DS): Intermittent is most common patient in my clinical practice.

(SL): Yes. The majority of children I see with strabismus have accommodative esotropia. I see many who have Northern European heritage who are more likely to be hyperopic.

(SP): No, I see more exotropia.

(RG): Yes, I do.

(SD): No, in our setting esotropia is more commonly seen than esotropia.

(SK): At what age and with what symptoms do these patients present to you?

(FM): Congenital esotropes present between the age of 3-6 months. Their parents or a health professional has observed their strabismus. Very occasionally they present later as the child’s family had been given incorrect information that strabismus in the first year of life is not unusual. Accommodative esotropes usually present between the age of 2½-4 years. Their parents or carers observe that the child has intermittent esotropia that is noticeable when the child is fixating on a near object. The strabismus tends to be more obvious when the child is tired. Accommodative esotropia may at times present in infancy as an esotropia with an accommodative element. Exotropia may present as intermittent exotropia at any time between the age of 6 months to 4-5 years. The child is observed to have intermittent exotropia that manifests at distance fixation. The child very often has a tendency to close one eye in sunlight. Constant exotropia may present at any age. There is often a history of premature birth, developmental delay, or poor health. Parents may also observe that these children have poor vision in one eye.

(JS): A few children present in the first year of life, but more at age 2 to 4 years. They can present at any age. Parents usually bring them because they have noticed the appearance of a squint.

(DS): All ages for esotropia and exotropia

(SL): The peak age is between 2 and 3 years old. Most of the time the family has noticed intermittent crossing of the eyes.

(SP): Patients either present as infants less than 6 months with infantile onset esotropia, at a later stage such as 2-4 years of age with accommodative esotropia, or later with diplopia.

(RG): Being a referential centre Infantile ET comes too late-usually between 8 -12 months. Acquired ET presents at all ages and often after receiving different treatments.

(SD): Most cases of Accommodative esotropia usually present between age of 3-5 years however in our setting some patients report at later age. Strabismus is the most common symptom for which these patients are brought to the hospital by parents.

(SK): How do you like to classify accommodative esotropia?

(FM): We like to classify accommodative esotropia as fully accommodative with or without a high AC/A ratio or partially accommodative with or without a high AC/A ratio.

(JS): Based on age of onset, the presence of refractive error and its effect on the angle of squint; the presence of any normal or anomalous binocular function and any difference between near and distance angle and binocularity. But there are many children who don’t fit into a neat classification and I try to understand the pathophysiology of each individual child using the above features.

(DS): I classify them as A) Fully accommodative (eyes aligned with spectacle) vs partially accommodative (eyes not aligned with spectacles). B) High AC/A) Eyes aligned at distance with spectacles and esotropia at near) vs Normal AC/A.

(SL): Fully accommodative and partially accommodative. Distance/Near Disparity with or without high AC/A.

(SP): I tend to classify them as fully accommodative or partially accommodative and then group by normal or high AC/A ratio. Of course, there are the rarer cases that are non-refractive and only present at near but those are not typically seen.

(SD): I classify them as fully accommodative and partially accommodative with high AC/A and Normal AC/A ratio.

(SK): Which cycloplegic do you prefer for children with esotropia?

(FM): We prefer to use Cyclopentolate 1%. In most children one drop is sufficient to achieve cycloplegia; however, in some children with dark brown eyes two drops are needed. We like to wait at least 30 minutes before refracting the child.

(JS): I usually use cyclopentolate 1% unless the child’s irides are very dark or cyclopentolate doesn’t fully relax their accommodation. For these children I use atropine 1% twice a day for 3 days before the refraction.

(DS): Cyclopentolate 1% for > 1 year of age; cyclopentolate 0.5% for < 1 year.

(SL): Cyclopentolate 1%.

(SP): I prefer Cyclogy1 1%.

(RG): Cyclopentolate 1% below 9 months cyclopentolate 0.5%. Atropine 1% if refractive ET/ high AC/A ratio.

(SD): I prefer 1% atropine till 7 year of age and 1% Cyclopentolate after 7 year of age.
What are the most important clinical considerations while examining patients with esotropia?

The most important consideration is the visual acuity in each eye. We measure the esotropia both at near and distance fixation. Ocular motility is checked looking for any restriction to exclude a restrictive problem such as Duane syndrome or a cranial nerve palsy. Careful optic nerve and retinal examination is critical to exclude underlying pathology. It is important to ensure the child is fully cyclopleged for an accurate refraction. In children who appear to have a high AC/A ratio we use +3.00D lenses to ascertain the response at near fixation. Visual acuity is assessed using an age appropriate test. Accurate visual acuity is very important for amblyopia management.

Excluding lateral rectus weakness and assessing acuity and any potential for binocular function. Also determining the full cycloplegic refractive error and assessing its effect on the esotropia.

Whether the eyes are aligned with spectacle correction (from full cycloplegic refraction) and presence or absence of amblyopia.

Amblyopia is the major impediment to developing binocular vision. A careful visual acuity assessment is critical.

For me, the most important initial consideration is refractive error. Once we correct refractive error, then it is important to determine whether there is any amblyopia. Once refractive error and amblyopia are treated, if there is still esotropia manifest with correction, then we start to consider near: distance disparity. If the patient is fusing at distance but not at near, then bifocal spectacles can be added.

1. To rule out any systemic and ocular aetiology.
2. Refractive value.
3. Amblyopia. It is really a concomitant ET, or it has limitations or if is mainly Nystagmus with ET.
4. Vertical associated strabismus.
5. Head turn.
6. Evolution over time.

Visual acuity, refractive error, amblyopia, AC/A ratio, ocular motility to rule out VIth nerve palsy and fundus examination are important clinical considerations while examining patients with esotropia.

Do you prefer a partial or full correction with glasses? Do you prefer a step wise increase in power of the glasses?

In children with accommodative esotropia we generally prescribe the full correction. If the child is unable to tolerate their glasses, we use either a short term or long term cycloplegic agent to allow the child to relax into their hyperopic correction.

I give full cycloplegic correction from the start. The number of squint operations performed for esotropia in the UK is now half of what it was 25 years ago because we are much better at fully correction hypermetropia in children with esotropia.

Full correction with no stepwise increase.

The major obstacle to binocular vision after amblyopia is persistent strabismus. I would like the child to get as straight as possible as soon as possible. I prefer prescribing full cycloplegic refraction and have not found benefit in prescribing less than the full plus at the outset of treatment.

Typically prescribe the full correction. I only treat with partial plus if the patient does not tolerate the full hyperopic correction.

Full correction if it is accommodative and no increase if possible.

I prefer full correction.
In which type of patients with esotropia do you prefer bifocals or progressive addition lenses?

We prescribe bifocals or progressive additional lenses only if the child is straight at distance and has residual esotropia at near fixation. In the partially accommodative esotrope we usually repeat refraction 2-3 months after the initial pair of glasses are prescribed as repeat refraction may uncover additional hyperopia. We prefer to use bifocals and specify to the dispensing optician that the bifocals should be a large segment with the upper part of the segment bisecting the pupil.

I use bifocals in children with convergence excess esotropia (binocular for distance and esotropic for near) when parents prefer bifocals to surgery. I don’t use progressive lenses in children.

If there is an apparent distance / near disparity from a high AC/A ratio. Most children in this situation will get better alignment at near with single vision glasses. For those children in their full hyperopic diopters and their alignment improves with +3.00 lenses at near viewing over their glasses, I would consider bifocals then. I warn the parents at the outset that we may have to change the lenses more than once in the beginning, so this is not a surprise for them. Luckily most opticians in our area are willing to change the lenses at no additional cost if we do the change in the first few months of purchase.

I prefer flat top bifocals, so the power of the bifocal is high. I find progressive-add bifocals are more likely to fail given the add power is at the bottom of the lens. Children are much more likely to ignore the progressive bifocals.

I start with refractive correction and if the child’s strabismus is not fully corrected when they have fully adapted to their full hypermetropic correction, I prefer part time occlusion and atropine 1% penalisation if non-compliant with patch.

My goal with amblyopia treatment is equal vision at the youngest age possible. For this reason, I recommend full time patching in children if their vision has not improved with the glasses. I prefer full time patching to gain vision and part time patching to maintain vision. With that said, I try to make the parents a partner in the decision-making for their child so if they make an informed decision to do part time patching or atropine at the outset, I support them.

I prefer flat top bifocals, so the power of the bifocal is high. I find progressive-add bifocals are more likely to fail given the add power is at the bottom of the lens. Children are much more likely to ignore the progressive bifocals.

I only treat with bifocals if the patient is fusing at distance but not at near.

AC/A ratio with no or very small strabismus at distance, the first glasses are Bifocals then I go for progressive lenses.

I prefer bifocals if there is residual esotropia at near while eyes are ortho at distance.

When do you like to follow up a child with accommodative esotrope after prescribing him/her glasses?

We generally follow-up within 2-3 months after issuing the prescription. If the child’s strabismus is not fully corrected with the hyperopic correction, we will frequently repeat the refraction.

Usually six weeks after first glasses prescription to check that they are wearing the glasses properly and to assess their effect.

I like to follow up after 2 months.

For the initial glasses, I check within 2 months. Once the child has been stable for a year and there are not significant changes to the glasses, I usually see the children yearly.

I usually like to follow up after two to three months.

4-6 weeks but not only to see improvement of the deviation but amblyopia and fusion may also improve over time. Some patients have initial improvement with the glasses and deviation recurs after months.

After 4-6 weeks.

How do you treat Amblyopia in these patients of accommodative esotropia?

Our management of amblyopia is no different to that in other patients. We follow the PEDIG guidelines and prefer to use face occlusion to pharmacological penalisation. Occasionally we will use a filter occlusion Bangerter filter.

By patching as a first choice, usually initially for two hours a day increasing if needed. I generally reserve atropine penalisation for a child who won’t wear a patch.

I prefer part time occlusion and atropine 1% penalisation if non-compliant with patch.

My goal with amblyopia treatment is equal vision at the youngest age possible. For this reason, I recommend full time patching in children if their vision has not improved with the glasses. I prefer full time patching to gain vision and part time patching to maintain vision. With that said, I try to make the parents a partner in the decision-making for their child so if they make an informed decision to do part time patching or atropine at the outset, I support them.

I start with refractive correction and if the amblyopia does not improve, I start 2 hours per day of patching.

Mild to moderate amblyopia is treated by partial occlusion; Severe cases by total occlusion, then depends on the rhythm of recovery, deviation and refraction I combine it with penalisation, atropine, filters etc.

Refractive correction followed by patching and if required television and mobile games.

Which patients with accommodative esotropia are the best candidates for surgery?

The best candidates for surgery are children with partial accommodative esotropia. The surgical outcome in these children is usually extremely good as they have been binocular during the early part of their life before the accommodative esotropia manifested.

Those with a recent onset of esotropia who do not fully correct when they have fully adapted to their full hypermetropic correction because they have the potential to recover binocular function. I only operate on children with no binocular potential if they still have a cosmetically poor squint when they are wearing their full hypermetropic correction.
(DS): Those with residual deviation while using spectacles and no amblyopia.

(SL): Those who partially respond to the glasses could be considered. Ideally, they have equal vision before surgery. This includes those who may respond with good distance alignment but still have crossing at near.

(SP): I treat any patient with more than 10-12 PD of esotropia with their full hyperopic correction with surgery.

(RG): Early onset cases, normal retinal correspondence, Decompensated microtropia or any other case with good potential of binocularity.

(SD): Cases of partially accommodative esotropia.

(SK): According to you what is the best time to operate these patients?

(FM): If the child’s accommodative esotropia is fully controlled with glasses, there is absolutely no indication for surgery. We will operate on children with partial accommodative esotropia at any age if the esotropia is uncosmetic. Early surgery also improves the opportunity for regaining binocular visual function. In children with accommodative esotropia with high AC/A ratio where the eyes are straight at distance and esotropia continues to manifest at near, we will operate on these children after the age of 13-14 years to help take them out of bifocals or multifocals. We have found that by waiting until the child’s accommodative component has significantly decreased in the teenage years, we rarely have to consider surgery in this group.

(JS): If I think that there is a chance of recovering binocular function when the squint is of recent onset I operate as soon as practicable once they are fully adapted to their glasses. Otherwise it is when the child and parent wish for surgery.

(DS): As soon as possible after determining that spectacles alone do not align eyes.

(SL): When amblyopia treatment has reached an end point (equal vision or as good as can be obtained with treatment) and there are two strabismus measurements that correspond within about 5 prism diopters, surgery can be considered.

(SP): As soon as I am sure that they are still esotropic with their full cycloplegic refraction and amblyopia is treated.

(RG): When the deviation is almost stable and there are no signs of spontaneous recovery.

(SD): In fully accommodative esotropia surgery is not recommended, however in cases of partially accommodative esotropia and non-accommodative esotropia surgery should be performed for cosmetically unacceptable deviation after treating the amblyopia when child is cooperative for examination.

(SK): Do you prefer a bilateral symmetrical surgery in these patients?

(FM): In children with accommodative esotropia we prefer symmetrical surgery – bimedial rectus muscle recession.

(JS): No, I generally do a recess/resect procedure on one eye unless there is a significantly greater angle for near, when I do bimedial recessions.

(DS): Yes, if no amblyopia. No if amblyopia present.

(SL): Not necessarily. I often do bilateral symmetrical surgery but often also use recess/resect. I honestly don’t know that one is superior than the other. However, I offer pre-operative prism adaptation for those who partially respond to glasses, in that case, I use bilateral symmetrical surgery. I also use bilateral surgery for Esotropia associated with a pattern or for distance near disparity. If the family doesn’t want to do pre-operative prism adaptation and the angle of esotropia is less than 35 prism diopters, I would consider a recess/resect procedure. Also, if there is significant residual amblyopia, I prefer to do a recess/resect procedure in the weaker eye.

(SP): Yes, I prefer a Bilateral symmetric surgery

(RG): In Acquired Accommodative ET it will depend on the amount of deviation (for large deviations I prefer R+R3); If the deviation is greater at near distance- Bimedial recessions. If the AC/A is very large and deviation at distance is very small I prefer faden surgery alone or combined with unilateral MR recession. Small deviations up to 25 PD, no vertical surgery is needed and children below age of 3-4; I usually treat them with Botox. In cases of Infantile ET in children my preferred technique is Bimedial recessions or Botox if deviation is up to 30 PD.

(SD): In most of cases I prefer bilateral symmetrical medial rectus recession.

(SK): What is your procedure of choice-recession with or without augmentation, faden/posterior fixation suture?

(FM): We prefer bimedial rectus muscle recession. We have found that there is no need for augmentation.

(JS): Straightforward medial rectus recession and lateral rectus resection or bimedial rectus recessions.

(DS): Bilateral medial rectus recession alone.

(SL): For distance/near disparity I would operate for the near angle or the prism adapted angle for those with a High AC/A. If they have a distance near disparity without a high AC/A, I would use a posterior fixation suture.

(SP): I typically do a bilateral medial rectus recession for the average between the distance and near angles and will add posterior fixation sutures if there is a high AC/A ratio.

(RG): I prefer faden for very cases with very High AC/A ratio, usually bridge technique.

(SD): Bilateral medial rectus recession.
For all post-op esotropia patients, I hope for a small (SL): Alignment (no E or X).
(DS): If I think they have the potential to regain binocular (JS): function I aim for orthotropia. If not, I aim to leave them slightly esotropic in their glasses.

For fully controlled accommodative esotropia, (FM): Our goal at surgery is to regain binocular visual function. We aim for orthotropia or a very small residual esotropia (within 10 prism dioptries) which would still allow the child to regain binocular visual function.

Generally, after age 9 -10 weaken them by 0.75 D (SP): We do not recommend refractive surgery before the age of 21 years. At that age we ensure that before the child undergoes refractive surgery, their refraction has been stable for at least 2-3 years. Surgery can be successful if hypermetropia falls in correctable range.

Orthoptic exercises (clear and misty) can be helpful in giving the child an understanding how they can “flex” their accommodation/convergence relationship and learn to hold their eyes straight on occasions, such as for school photographs without the need to wear their glasses.

If we are discussing the first scenario, I don’t know what is exotropic in the full cycloplegic refraction. I know others use this term for post-operative exotropia. If I think they have the potential to regain binocular function I aim for orthotropia. If not, I aim to leave them slightly esotropic in their glasses.

Within 8 prism dioptries.

What is your early postoperative goal in such patients?

What are the risk factors for consecutive exotropia in these patients?

What is your take on hyperopic refractive surgery in these patients?

How do you wean glasses in patients with accommodative esotropia?

The risk factors are uncorrected amblyopia, not decreasing the hyperopic correction at appropriate interval and most importantly operating on children with fully accommodative esotropia. The child with partial accommodative esotropia we tend not to over- weaken the medial rectus muscles.

As the child grows older there is a tendency to become less hyperopic and the hyperopic correction can gradually be decreased. As control of their accommodative esotropia increases we slowly decrease the hyperopic correction giving the minimum amount required to maintain binocularity both at distance and at near. Older children will often present for review complaining of some blur of their distance vision. This is usually a good indication that we can safely decrease the hyperopic correction. In children with good control, we encourage them to wear their glasses less for outdoors and recreational activities. We insist that they continue to use their glasses for schoolwork. Orthoptic exercises (clear and misty) can be helpful in giving the child an understanding how they can “flex” their accommodation/convergence relationship and learn to hold their eyes straight on occasions, such as for school photographs without the need to wear their glasses.

I do not recommend weaning.

I don’t recommend weaning complete off (small hyperopia, small astigmatism, and small anisometropia).

How do you wean glasses in patients with accommodative esotropia?

Uncorrected refractive error, amblyopia, surgery on accommodative esotropia.

We do not recommend refractive surgery before the age of 21 years. At that age we ensure that before the child undergoes refractive surgery, their refraction has been stable for at least 2-3 years. Surgery can be successful if hypermetropia falls in correctable range.

We have found that contact lenses are an excellent alternative to glasses and can be worn very successfully in children from a young age (5-6 years).

We encourage them to wear their glasses less for outdoors and recreational activities. We insist that they continue to use their glasses for schoolwork. Orthoptic exercises (clear and misty) can be helpful in giving the child an understanding how they can “flex” their accommodation/convergence relationship and learn to hold their eyes straight on occasions, such as for school photographs without the need to wear their glasses.

Uncorrected refractive error, amblyopia, surgery on fully accommodative esotropia.

I do not recommend refractive surgery before the age of 21 years. Response to surgery after 21 is satisfactory.

I don’t recommend it. The refractive correction required often changes with time and it is important to keep then full corrected.

I don’t recommend refractive surgery before 21 years old. Response to surgery after 21 is satisfactory.

I know some young adults may want to get out of their glasses who still need them to maintain straight eyes and they don’t want to do contact lenses. I think it is not a bad option for those >20 change of glasses. I will do if I think the refraction can be weaned completely off (small hyperopia, small astigmatism, and small anisometropia).

I don’t recommend it. The refractive correction required often changes with time and it is important to keep then full corrected.

We do not recommend refractive surgery before the age of 21 years. At that age we ensure that before the child undergoes refractive surgery, their refraction has been stable for at least 2-3 years. Surgery can be successful if hypermetropia falls in correctable range.

We have found that contact lenses are an excellent alternative to glasses and can be worn very successfully in children from a young age (5-6 years).
years. I have seen it done successfully.

(SP): I have not used it so do not have any experience with it.

(RG): I usually do not suggest refractive surgery in children. Over life I speak with them on the possibility of a future refractive surgery. Small moderate hyperopes may decrease and may need a refractive surgery if Accommodative ET persists. High hyperopia usually does not decrease over years and some have too shallow anterior chambers and cannot have an ICL so in those patients I am cautious with my conversations.

(SD): I don’t recommend refractive surgery in these patients.

(SK): What do you think is the natural course of patients with accommodative esotropia?

(FM): This is one group of patients who do extremely well with conservative management (correction of refractive error, bifocals where indicated and management of any amblyopia). Maintaining excellent communication with the family as to what to expect from the start is extremely important. Many of the children with accommodative esotropia are very intelligent and learn to understand why they have strabismus and what techniques they can use to help control the accommodative esotropia. Fully accommodative esotropes usually depend on optical correction for their control throughout their life. The hyperopic correction over time can generally be decreased. Many become successful contact lens wearers and look forward to refractive surgery when they are older.

Those with a high AC/A ratio can usually be weaned out of their bifocal or multifocal by the age of 13-14 years. If they continue to rely on their near correction, then surgery to weaken the medial rectus muscle can be considered after the age of 13-14 years.

Partial accommodative esotropes, especially those with a low hyperopic correction generally improve over time and many can manage their activities apart from close work without the need for glasses. I have found that children with accommodative esotropia of any form with a high hyperopic correction usually prefer to continue to wear their glasses throughout life.

(JS): Most will stay stable provided they continue to wear full correction for their hypermetropia.

(DS): The usual course is to become less hyperopic over time and need less correction for good alignment.

(SL): A majority will have less hyperopia over time. A substantial minority will be straight without glasses at some point before age 12 years but many will need glasses into young adulthood or adulthood to keep straight eyes.

(SP): I think that the component that is corrected with spectacles slowly improves as the child grows and typically resolves by the mid-teens.

(RG): Some improve spontaneously over years - partially accommodative ET of smaller amount and greater at near.

(SD): Majority of patients become less hyperopic during course of time.

(SK): Do you have any experience in botulinum toxin in esotropia with an accommodative component?

(FM): I have not managed accommodative esotropia with Botulinum Toxin. I have seen a few patients for a second opinion where the Botulinum Toxin has proved to be unsuccessful. I would not recommend the use of Botulinum Toxin which is an intervention with risk attached to manage a problem that can be safely and successfully managed with optical correction.

(JS): No, I don’t have any experience.

(DS): I have no experience with injectable toxins for this clinical entity.

(SL): No.

(SP): No, I have not used it for this situation.

(RG): It is a very good option for ET until 25/30 PD and for residual deviations.

(SD): No, I don’t used botulinum toxin in esotropia with an accommodative component.

Compiled by:
Dr Savleen Kaur
Senior Research Associate, PGIMER, Chandigarh
Email: mailsavleen@gmail.com
Surgical Timing in Infantile Esotropia

Dr. Suma Ganesh MS, DNB, Dr. Soveeta Rath DNB, FICO

Department of Pediatric Ophthalmology, Strabismus and Neuro Ophthalmology, Dr Shroff Charity Eye Hospital, Daryaganj, New Delhi, India

Summary: Infantile esotropia is classically defined as an esotropia with an onset before the age of 6 months. It affects 1 in every 100 to 500 persons. The most beneficial timing of surgery of infantile esotropia still remains controversial. Helveston describes intervention between 4-6 months of age as “Ultra-early”, surgery before the age of 2 years as “Early” and any intervention after 2 years as “Late” surgery. Basic research proves that alignment of eyes during the critical period of development of sensory system ensures development of binocular vision. The benefit of prompt surgery must be balanced with the rate of reoperations and spontaneous resolution while deciding on the surgical management of Infantile esotropia and the same must be meticulously counselled to parents.

Infantile esotropia (IE) is classically defined as an esotropia with an onset before the age of 6 months. It affects 1 in every 100 to 500 persons. It presents with a large angle of strabismus (>30 prism diopters (PD), no or mild amblyopia, hypermetropia < 2 dioptres, cross fixation and pseudo limitation of abduction, latent nystagmus, dissociated vertical deviation (DVE) and absent or reduced binocular vision, in the absence of any neurological disorder. Affected children exhibit abnormal stereopsis and naso-temporal asymmetries of smooth pursuit, optokinetic responses and abnormal visual motion processing. Multiple factors like sufficient patient cooperation for evaluation and planning, horizontal deviation stability, estimation of vertical deviation (if any), any oblique muscle overaction, absence of accommodative factors, presence of alternating fixation determine the appropriate surgical option and timing. Although surgical correction remains the gold standard management option for infantile esotropia, the most beneficial timing of surgery still remains controversial. This review focuses on appropriate timing of surgical repair in infantile esotropia to attain the best motor as well as sensory outcomes.

MANAGEMENT OF INFANTILE ESOTROPIA

Management of IE begins with assessment of visual acuity, refractive error, amblyopia. Though not very common, yet amblyopia and refractive error has to be treated first before planning any surgical intervention.

The Congenital Esotropia Observation Study (CEOS) found that infants presenting with esotropia before 20 weeks of age have a greater variation of size and character of the deviation. Yet in 98% of these infants esotropia persists, especially those who have large-magnitude (≥20° or 40 PD) esotropia with onset after 10 weeks of age and refractive error ≤3.00 diopters, warranting early surgery.

Surgical options

The available surgical approaches for correction of infantile esotropia includes:

1. Bilateral medial rectus (MR) recession
2. Unilateral MR recession combined with lateral rectus (LR) resection
3. Bilateral MR recession with unilateral LR resection
4. Bilateral MR recession with bilateral LR resection

However, in many cases single surgery does not result in complete motor outcome and a second surgery for correction of residual esotropia is warranted. In some children development of dissociated vertical deviation and inferior oblique overaction usually develops later (after 2 years of age) desiring second surgery for the treatment of resultant vertical deviation then.

Surgical dosage

In children with IE, bilateral medial rectus recession is the most commonly preferred surgical approach although size of the growing eyeball and unstable angle of deviation have always been a deciding factor. The dosage which is usually followed varies with the angle of deviation. In these children amount of recession to be performed is usually measured from limbus rather than insertion.

WHY IS TIMING IMPORTANT FOR INFANTILE ESOTROPIA

The question of timing of surgery has always been a matter of debate related to optimal timing of surgery. The unresolved debate dates back to 1939 when Chavasse proposed that the ability of developing fusion in infants with congenital squint was possible...
The ultimate goal of treatment of IE is

- To align the visual axes
- To promote the development of binocular vision

The figure below shows the amount of recession for different angle

<table>
<thead>
<tr>
<th>Angle of Deviation</th>
<th>Recession</th>
</tr>
</thead>
<tbody>
<tr>
<td>Small 8.5-9 mm 20-30 PD</td>
<td></td>
</tr>
<tr>
<td>Medium 9.5-10 mm 30-45 PD</td>
<td></td>
</tr>
<tr>
<td>Large 10.5-11 mm 45 PD</td>
<td></td>
</tr>
<tr>
<td>Larger 12-12.5 mm 50-60 PD and greater</td>
<td></td>
</tr>
</tbody>
</table>

Authors have reported various time frame appropriate for surgery to achieve good motor and sensory outcomes. Based on age of child at intervention, the different terms regarding surgical timing used by Helveston are:

- Ultra early-surgery between 4-6 months of age
- Early-surgery before the age of 2 years
- Late-any intervention after 2 years

if the deviation could be fully corrected before the age of two years.

Helveston’s proposal of ultra-early surgical timing was supported by Wright, Ing and Birch et al. Surgical intervention before the second birthday has been considered as ‘Early’ intervention by Shirabe, Simonz and Magli et al. Intervention beyond 2 years of age is considered ‘Late’ or ‘Standard’ treatment regimen. However the controversy lingers and there has been no consensus on exact timing as noted by Shaffiq and Elliot. As per current US standard, age of first surgery for IE is approximately 12 to 18 months, whereas in many European countries surgery is performed at the age of 2 or 3 years.

INDICATIONS FOR EARLY SURGERY

Based on multiple studies, following are the indications where early surgery can be advocated without adverse motor or refractive outcomes. Absence of nystantus or head bobbing
- Absence of any previous extraocular muscle surgery
- Absence of any structural ocular:
  - Absence of following birth history:
    - Gestational age < 34 weeks
    - Birth weight < 1500 grams
    - Ventilator treatment in the newborn period
    - History of meningitis or any major medical event.

Sensory and Motor Outcomes: Early Vs Late Surgery:

SENSORY: Stereopsis is considered the gold standard in evaluation of binocular status in patients with strabismus. This parameter has thus been assessed by multiple studies in the light of surgical timing of IE. Surgery around 1 year of age is associated with better stereopsis has been reported in multiple studies. Other group of authors found that a higher percentage of children operated before 2 years of age also achieved good stereo-acuity than those operated beyond 2 years of age. However the attainment of stereopsis following early surgery has also been questioned by others. Central and peripheral fusion has also been evaluated as the other parameter for measurement of binocularity.

The following table (Table 1) describes some major studies which report on the basic sensory outcomes following early surgery for IE.

It has also been reported that early surgery during the first year of life was found to improve the nasotemporal asymmetries of both motion detection and mVEPs.

Though all above studies focus on potential benefit of improved sensory outcomes following early surgery, stereopsis beyond intermediate grade has been achieved only by few of these.

MOTOR: The literature is replete with reports suggesting enhanced binocular vision following prompt surgery for IE, however concerns affecting motor outcomes include:

1. Instability of angle of deviation and accuracy of measurement
2. Chances of reoperation
3. Tendency of spontaneous resolution
4. Onset of DVD
5. Need of hyperopic glasses
6. Instability of angle of deviation and accuracy of measurement: It is believed that surgical outcomes usually get
affected while operating on unstable and imprecise angle of deviation in children with IE. However Birch et al reports comparable motor as well as sensory outcomes at 1 year post-operative follow up both for stable and unstable angle of deviation. Unstable deviation was considered if there was >10 PD variation at 2 preoperative visits at an interval of 6 weeks.

Studies have reported good post operative alignment (<10PD of orthophoria) following early intervention for IE only with single surgery. These provide an inference that neither the angle instability nor does the accuracy of measurement affects motor outcomes adversely.

Reoperation rates: Several studies has shown that surgery at a very early age increases the number of reoperations. In the ELISS study, the mean number of operations was 1.18 in the early group and 0.99 in the late group. In a meta-regression analysis of the ELISS and 12 other studies it was found that reoperation rates were 60-80% for children first operated around age 1 and 25% for children operated around age 4.28 In another recent study by Magli et al, patients who underwent surgery ≤2 years of age had a reoperation rate of 12.1% vs a reoperation rate of 8.6% in children operated on at >2 years of age.

The cause for second surgery was either residual or recurrent esotropia or consecutive exotropia. Onset of new vertical deviation or DVD and inferior oblique overaction were the other indications for another surgery.

Spontaneous resolution: The other issue in a small subset of children with IE is tendency of spontaneous resolution. Surgery on patients in whom the esotropia may spontaneously resolve may lead to consecutive exotropia. The tendency of spontaneous resolution depends mainly on constancy of deviation rather than the size of deviation has been described by Fu et al.

Thus the benefit of prompt surgery has to be balanced with the rate of reoperations and spontaneous resolution while deciding on the surgical management of IE and the same has to be meticulously counselled to parents.

Dissociated vertical deviation: Shin and Paik identified association of DVD with later surgery and large preoperative angle by multivariate analysis in a retrospective review of 90 patients of IE who had undergone surgery.40 Surgery before 24 months of age for IE reduces the incidence and the severity of spontaneous DVD, especially in cases with a large amount of esodeviation, lowering the need for a second operation to correct DVD was also reported by Yagashaki et al.41

Hyperopic glasses: Pre operative hyperopic glass prescription is an important determinant of surgical planning and outcomes. ELISS study found that spherical equivalent of the glasses was 3.0 D in the early vs. 2.9

<table>
<thead>
<tr>
<th>Author Year</th>
<th>No of Children</th>
<th>Timing of Surgery</th>
<th>Steropsis Achieved/Not</th>
<th>Tests Used</th>
<th>Fusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birch 1990</td>
<td>81</td>
<td>Before 1 year</td>
<td>35%</td>
<td>Randot</td>
<td>Not available (NA)</td>
</tr>
<tr>
<td>Charles 1992</td>
<td>24</td>
<td>9-23 months</td>
<td>19.04%</td>
<td>Frisby/Titmus</td>
<td>NA</td>
</tr>
<tr>
<td>Wright 1994</td>
<td>7</td>
<td>13-19 weeks</td>
<td>71.4%</td>
<td>Randot</td>
<td>NA</td>
</tr>
<tr>
<td>Ing 1995</td>
<td>16</td>
<td>4 months</td>
<td>68.7%</td>
<td>Titmus/Randot</td>
<td>68.7%</td>
</tr>
<tr>
<td>Birch 1995</td>
<td>73</td>
<td>5-16 months</td>
<td>41.1%</td>
<td>Random dot</td>
<td>NA</td>
</tr>
<tr>
<td>Birch 1998</td>
<td>80</td>
<td>Before 15 months</td>
<td>30%</td>
<td>Random dot</td>
<td>NA</td>
</tr>
<tr>
<td>Shirabe 2000</td>
<td>9</td>
<td>Before 8 months</td>
<td>55.5%</td>
<td>Titmus Randot stereo test (RDS) Dynamic RDS</td>
<td>44.4%</td>
</tr>
<tr>
<td>Simonz 2005</td>
<td>231</td>
<td>6-24 months</td>
<td>13.5% vs 3.9%</td>
<td>Housefly</td>
<td>NA</td>
</tr>
<tr>
<td>Birch 2006</td>
<td>50</td>
<td>0-6 months</td>
<td>38% vs 16%</td>
<td>Randot</td>
<td>78% vs 61%</td>
</tr>
<tr>
<td>German 2014</td>
<td>38</td>
<td>13-39 months</td>
<td>50%</td>
<td>Randot/Titmus</td>
<td>NA</td>
</tr>
<tr>
<td>Magli 2016</td>
<td>188</td>
<td>Before and after 2 years</td>
<td>50.7% in &lt; 2 years 36.1% in &gt; 2 years</td>
<td>Lang and Titmus</td>
<td>78.3% 63.8%</td>
</tr>
</tbody>
</table>
D in the late group. Glasses had been prescribed previously in 19.1% children in the early group vs. 24.3% in the late group. 25 Birch et al. reports postoperative hyperopia of +4.00 D to +5.50 D was found in 9% of those in the early surgery group and 6% of those in the standard surgery. Hyperopia as a baseline parameter did not vary between groups with regard to surgical timing.

OTHER FACTORS

Social concern: Besides above parameters, social stigma has always been a prominent determinant while discussing about strabismus. There are chances of poor parent-child relation due to the cosmetic appearance of the child. Moreover, straight eyes following early surgery alleviates psychomotor development of the growing child.

Type of surgery: There has been no consensus so far regarding the best surgical approach for IE. Although various techniques have been proposed by different studies, none have a clear advantage over other. Magli et al. analyzed 9 different surgical approaches and found that a six muscle approach (Bilateral MR recession+ LR resection+ Inferior oblique weakening) had 11 times higher chance of improving binocular vision compared to 3 muscle approach (Bilateral MR recession+ Unilateral LR resection).

Duration of misalignment: It has also been found that duration of misalignment determines binocular vision development. Better stereocuity is seen with stable long-term eye alignment.

Early Vs Late: Risks And Benefits:

An extensive literature review now provides enough evidence favoring prompt and early surgical management for infantile esotropia. Yet, late surgery is still being practised by several proponents. In the following table we summarize the advantages and disadvantages of early vs. late surgery.

Although above issues may pose some difficulties in planning early surgery the major factor that outweighs these problems are preservation and restoration of binocular vision by early surgery.

OUR DATA: (UNPUBLISHED)

A retrospective chart review of children who presented with infantile esotropia at out tertiary eye care center between 2012-2017 showed 54 children underwent surgery for IE before 2 years of age (“early group”) whereas 126 were operated after their second birthday (“late group”). The mean preoperative deviation was comparable in both the groups (49.07PD vs 46.41PD). In the early group, mean age at surgery was 16 months and after a mean follow up of 36 months, 41 (75.92%) achieved an alignment within 10PD of orthophoria and 13/54 (24.07%) needed reoperation for residual esotropia and consecutive exotropia. The mean age at surgery for those operated >2 years of age was 60 months. Ninety eight (76.5%) children achieved good alignment within 10PD orthophoria whereas 28 (22.2%) needed second surgery. Stereoaucity between 3000-200 seconds of arc was attained in 55% children in the early group versus only 27% in the late group in those who were cooperative for evaluation at final follow up.

To conclude this data from the north Indian cohort also supports the current literature that early surgery results in comparable motor but better sensory outcomes irrespective of stability of deviation.

CONCLUSION

Infantile esotropia and its management still remains a controversial issue though there is no dearth of literature related to surgical outcomes. Majority of studies favor surgery before 2 years of age or earlier to enhance the potential of binocularity, in the absence of any adverse motor or refractive outcomes. However, it is noted that only coarse stereopsis and peripheral fusion are achieved in most of them. Another concern that has to be borne by surgeons is post-operative stability of deviation on a long term basis is essential to maintain binocularity. Preoperative instability of deviation does not influence the surgical outcomes. Thus, prompt surgical intervention during the critical period based on the rationale of restoration of cortical connections has to be followed for management of IE. Yet, the surgical decision is based on the clinical examination and a meticulous orthoptic evaluation by the surgeon. Optimal surgical timing definitely varies as per clinical scenario.

REFERENCES

33. Çerman E, Eraslan M, Öğüt MS. The relationship of age when motor alignment is achieved and the subsequent development of stereopsis in infantile esotropia. JAAPOS. 2014;18:222-5.
Ocular nystagmus is a regular, rhythmic and involuntary to and fro movement of the eye which leads to the disruption of the steady fixation. Nystagmus cycle consists of a slow ocular movement away from fixation which is followed by a corrective movement towards fixation in the opposite direction. These oscillations may either consist of slow eye movements (pendular nystagmus) or as a combination of slow and fast movements (jerk nystagmus). Pendular nystagmus is a smooth sinusoidal oscillation which is equal on both sides whereas in jerk nystagmus the eyes appear to beat in one direction.

Infantile nystagmus syndrome (INS) is a common type of early-onset nystagmus. The incidence of this disease is about one in thousand and there is a male preponderance. The pathophysiology of this syndrome is still unclear. It is also called congenital nystagmus, however, it is rarely seen at birth and presents in the first few months of life. The Classification of Eye Movements Abnormalities and Strabismus (CEMAS) Working Group have proposed a classification to bring about clarity to the eye movement and strabismus literature. They have two criteria for infantile nystagmus syndrome: infantile-onset and accelerating slow phases on ocular motor recordings. According to CEMAS, INS is a different clinical entity from other forms of early-onset nystagmus like fusion maldevelopment syndromes (manifest latent nystagmus), spasms nutans and the ones localizing to brainstem or cerebellum.

**CLINICAL FEATURES**

This condition usually begins at 2 to 3 months of age. Clinically the nystagmus is binocular and conjugate. It is uniplanar and usually horizontal, rarely it can be vertical or torsional. It can have distinctive waveforms: pendular, jerk or mixed. These waveforms may vary in different gaze positions and can vary at different times. The nystagmus is accentuated by fixation effort and improves on eye closure. It is dampened by convergence effort, hence the near vision is usually better. A null zone is often present where the amplitude or frequency of both is minimum. Hence, the patient usually assumes a head posture to bring the eyes to the null zone. Table 1 enlists the clinical features of infantile nystagmus syndrome. Congenital periodic alternating nystagmus (PAN) is a primary-position nystagmus that periodically switches direction after approximately 120 seconds. Between each cycle there is a quiet phase of a few seconds before the nystagmus begins to beat in the opposite direction. Acquired forms of PAN often occur with pathology involving the cerebellum, such as cerebellar degenerations, tumours and multiple sclerosis. Arnold Chiari malformation can also present with PAN. Hence these conditions should be always ruled out in a patient presenting with PAN.

### Table 1: Clinical characteristics of infantile nystagmus syndrome

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Almost always bilateral and conjugate in direction and frequency</td>
<td></td>
</tr>
<tr>
<td>2. Uniplanar: usually horizontal. Rarely can be vertical or torsional</td>
<td></td>
</tr>
<tr>
<td>3. Similar amplitude on both sides. An accelerating slow phase is present.</td>
<td></td>
</tr>
<tr>
<td>4. Dampered by convergence, so near vision is better</td>
<td></td>
</tr>
<tr>
<td>5. No oscillopsia</td>
<td></td>
</tr>
<tr>
<td>6. A null zone is usually present and the head posture is assumed to bring the eyes in null zone.</td>
<td></td>
</tr>
<tr>
<td>7. Disappears during sleep or general anaesthesia</td>
<td></td>
</tr>
<tr>
<td>8. Distinctive waveforms are present: jerk /pendular/mixed</td>
<td></td>
</tr>
</tbody>
</table>

The prevalence of anterior visual pathway abnormalities has been noted in patients having INS. It ranges from 38 per cent to 91 percent. These include congenital cataracts, albinism and various optic nerve disorders.

### PATHOGENESIS

The pathophysiology of INS remains unclear; however many theories have been proposed to support the pathogenesis of this condition. These theories can be grouped into two categories: those which consider INS resulting from a primary defect in the ocular motor control system and those which consider it to result from a defect in the visual sensory system.

### DIAGNOSIS AND CLINICAL EXAMINATION

As per CEMAS classification, the waveform analysis of eye movements is required for the diagnosis. The nystagmus movements must be distinguished from nystagmoid movements which include oculo-palatal myoclonus, opsdodonus, ocular bobbing and Heimann-Bielchowsky’s unilateral vertical bobbing. A detailed history regarding the onset of nystagmus, abnormal head posture, developmental delay, family history of nystagmus must be taken. A detailed ocular examination must be done to rule out the presence of any anterior visual pathway defect. The nystagmus waveform, frequency, amplitude, direction and plane of oscillation must be recorded. Presence or absence of null zone must be looked for. A dilated fundus examination is essential to rule our retinal pathologies. INS must be distinguished from FMS with which it can be confused. Table 2 lists salient differences between INS and FMS. An ERG may be done to rule out retinal degeneration. A magnetic resonance imaging may be ordered if any neurologic pathology is suspected.
Management of INS involves symptomatic therapy for nystagmus. Any underlying systemic and ocular disorders must be looked for and treated accordingly. Asymptomatic forms with good vision do not require specific therapy, however, these patients must be screened for refractive errors and should be screened accordingly. The patients having a significant visual impairment and head postures must be treated must for the same. Following are the management options of INS.

### Non-surgical therapy

The use of optical devices has been tried in the treatment of nystagmus. The use of over minus lenses stimulates accommodative convergence and thus can dampen nystagmus. Base-out prisms can be additionally used to induce fusional convergence by using 7-prism dioptre base out prism in front of each eye. Contact lenses have been used to achieve fusional control in these patients. Soft contact lenses may additionally dampen nystagmus through a sensory feedback mechanism by stimulating the ophthalmic division of oculomotor nerve. Additionally, acupuncture applied to the sternocleidomastoid muscle may also work to reduce the nystagmus. However, in a study conducted at our centre, the effect was found to be shortlived.

Pharmacologic therapy for nystagmus has also been tried. Both memantine and gabapentin have been tried for nystagmus, both of them reduced the intensity but had no effect on visual acuity. Oral and topical carbonic anhydrase inhibitors have also been tried. In a study conducted by Hertle et al. to compare Topical brinzolamide (Azopt) versus placebo in the treatment of INS, topical Azopt significantly improved; INS waveform characteristics in the primary position null zone, group mean values of the nystagmus acuity function across gaze and group mean ETDRS binocular letter visual acuity. However further studies are needed in this area to establish anything concrete. Baclofen prescribed at 30 mg per day in three divided doses is useful and commonly decreases acquired PAN. However, congenital PAN appears to respond less so to baclofen. Similarly, baclofen has no effect on INS.

### Surgical Management

The principle for nystagmus surgery in INS is to correct the abnormal head posture by shifting the null zone to the primary position. This can be done with the help of surgery on horizontal recti muscles. Kestenbaum devised the first surgical approach using recession – resection of all four horizontal recti. Anderson advocated only recessions of yoke muscles to correct face turn. We follow Augmented Anderson approach to correct up to 30 degrees of face turn (Figure 1). This involves 9 mm of medial rectus recession and 12 mm of lateral rectus recession on yoke muscles depending on the direction of face turn. Hertle Dell’Osso procedure which involves disinsertion of all recti and their reinsertion has been found to be effective for short durations in decreasing amplitude of nystagmus in patients having no eccentric null.

In cases of vertical nystagmus having a chin elevation of depression of 25 degrees or more, a vertical Kestenbaum surgery can be done which involves the same principle as that of Kestenbaum procedure. For torsional nystagmus, three modalities have been described: the vertical shifting of horizontal recti, horizontal shifting of vertical recti.

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**Table 2: Differences between infantile nystagmus syndrome (manifest nystagmus) and Fusional maldevelopment syndrome (manifest latent nystagmus)**

<table>
<thead>
<tr>
<th>Manifest nystagmus</th>
<th>Manifest latent nystagmus</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Biphasic, mostly planar</td>
<td>Mostly jerk or mixed</td>
</tr>
<tr>
<td>2. No change on abduction</td>
<td>Increased on abduction</td>
</tr>
<tr>
<td>3. Accelerating slow phase</td>
<td>Decelerating slow phase</td>
</tr>
<tr>
<td>4. No change in covering one eye</td>
<td>Increase on covering one eye</td>
</tr>
<tr>
<td>5. Direction independent of fixing eye but may change on the either side of null zone</td>
<td>Fast phase always towards fixing eye</td>
</tr>
<tr>
<td>6. Less commonly associated with infantile esotropia (10-50%)</td>
<td>Nearly always associated with infantile esotropia (95%)</td>
</tr>
<tr>
<td>7. Binocular visual acuity same as uniocular</td>
<td>Binocular visual acuity better than monocular vision.</td>
</tr>
</tbody>
</table>

**Table 3: Treatment options for nystagmus**

**Non-surgical treatment**

1. Optical devices: glasses, contact lenses, prisms
2. Occlusion and pleasoptics
3. Medical therapy
4. Acupuncture
5. Auditory biofeedback
6. Botulinum toxin

**Surgical treatment**

1. To shift the null position to primary: Modified Kestenbaum surgery
2. To induce adduction / convergence: Bimedial rectus recession / faden
3. To reduce the amplitude of nystagmus: supramaximal recession or faden on all horizontal recti.

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**Figure 1:** Showing 7-year child having INS presenting with 30 degree faceturn (1A) corrected by Augmented Anderson procedure (1B).
Review Article

and lastly selective advancement and recession of the anterior fibres of the four obliques. Table 3 enlists the management options for nystagmus.

CONCLUSION

INS remains a challenging disorder for the paediatric ophthalmologists. The aetiology still remains obscure. Care must be taken to uncover any associated neurological and visual pathways disorder. Proper refractive correction and amblyopia management must be assured. Timely management in cases of patients having face turn can assure prevention of torticollis. The surgical procedures to dampen the nystagmus have not been able to provide satisfactory results. With newer studies in the area of pharmacological therapy for nystagmus being undertaken, the future holds promise for the medical treatment of this disease.

REFERENCES

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nerior oblique surgery remains a challenge for strabismologist. Various types of Inferior oblique weakening procedures in the past decade have been useful addition to our armamentarium of surgical procedures. Technical advances have translated a better outcome, however the art has to be mastered & skill need to be acquired. This article will provide an insight into the surgical skill required for various types of commonly performed Inferior oblique surgeries

**Anatomy:**

The inferior oblique originates at the periosteum of the maxillary bone near the lacrimal fossa and inserts into the posterior globe near the inferior border of the lateral rectus, passing under the inferior rectus. With the eye in the primary position, the inferior oblique makes a 51° angle with the visual axis and acts an excycloductor. The secondary and tertiary actions are elevation and abduction, respectively. The inferior oblique is innervated by the lower portion of the coulometer nerve.

Surgery is performed more commonly on the inferior oblique muscle than on any other cyclovertical muscle. Surgical access to the inferior oblique muscle and procedures designed to alter the effect of contraction of the inferior oblique muscle on the globe are relatively straightforward and complications are infrequent in experienced hands.

The anatomy of the inferior oblique muscle is somewhat atypical compared to the other extraocular muscles, which is important for understanding the effects of the various surgical procedures on eye position and ocular motility. In particular, innervation of the muscle occurs through the neurofibrovascular bundle (NFVB), which attaches midway between the origin and the insertion 2 mm temporal to the inferior rectus muscle. After anterior transposition of the muscle, the NFVB ligament will mechanically act as the muscle's new origin.

**BRIEF HISTORY**

The first studies of the effects of inferior oblique weakening that include anterior transposition were published in the 1940s. Accordingly, most operations on the inferior oblique muscle are designed to diminish its function. Weakening procedures on the inferior oblique muscle are numerous and include myotomy, myectomy, recession, marginal myotomy, disinsertion, anterior transposition, Hang Back recession and denervation and extirpation.

**INDICATIONS**

1. Primary inferior oblique overaction.
2. Superior oblique palsy.
3. V, Y, or X pattern strabismus with or without craniosynostosis.
4. Duane’s syndrome patients with upshoot.
5. chin-down head position in patients with nystagmus.
6. Inferior rectus aplasia.

**PRIMARY INFERIOR OBLIQUE SURGICAL PROCEDURES**

**Recession**

The inferior oblique muscle can be recessed along its anatomical path by as much as 14 mm. The inferior oblique recession is commonly used as a primary weakening procedure in patients with over elevation in adduction. 10 mm Inferior oblique recession is a simple, safe and effective method for the cosmetic and functional treatment of horizontal deviation and V pattern with primary inferior oblique overaction.

**SURGICAL TECHNIQUE**

- After inserting the speculum and applying the fixation suture, retract globe in extreme elevation and adduction.
- Make incision in infero temporal bulbar conjunctiva 7 to 9mm from the limbus a point between the lateral rectus and inferior rectus.
- The incision extended downwards and backwards.
- Blunt dissection of facial attachments is done.
- Muscle is isolated over by muscle hook facilitating direct visualization of the inferior vortex vein underneath.
- Do not hook LR/IR fibres.
- Care should also be taken to hook the entire muscle and be aware of the possibility of multiple congenital insertions.
- 6 " vicryl is inserted into the anterior aspect of the inferior oblique muscle near its insertion and the suture is looked.
- The inferior oblique is severed from its insertion 3 mm from the globe.
- The inferior oblique is sutured 7 mm lateral and 7 mm inferior end of the lateral rectus insertion (Figure 1).
- This method ensured that there was no bunching of the muscle and the muscle remained in its plane of action. When needed, this surgery was combined with the appropriate horizontal muscle surgery either in the same sitting or at a second sitting.
- Most commonly performed Inferior Oblique Surgery is by: Parks (IR) or Fink's (LR) technique.

**Anterior transposition**

Many current inferior oblique recession techniques include various ranges of transposition of the insertion and are categorized under “anterior transpositions.” The main difference between these procedures and true recessions is that anterior transposition alters the path of the muscle, which is generally more effective at weakening the inferior oblique. Anterior transposition of the inferior oblique muscle has been
widely used for the treatment of increased adduction associated with DVD (Table 1).

It is achieved by placing the IO insertion at any point anterior to its normal course can be placed just below the lower border of LR muscle or temporal to the IR insertion.

It is stronger weakening procedure as compared to recession.

It causes antielevation syndrome (AES). Hence this procedure when performed unilaterally is reserved for patients with significant hypertropia in upgaze and patients of SO paresis with at least 25PD hypertropia in primary position.

Anterior nasal transposition

Anterior nasal transposition (ANT) is a relatively new procedure, in which the insertion is transposed to a location over the nasal half of the inferior rectus muscle typically 2 mm nasal to the nasal border of the inferior rectus muscle and 2 mm posterior to the inferior rectus insertion. The inferior oblique is thus transformed from an extorter into an intorter and from an elevator to a depressor. The ANT can be used to eliminate or reduce severe excyclotorsion. An advantage of this procedure over temporal anterior transposition is that it avoids AES.

Indications for ANT are
1. Bilateral 4th nerve palsy
2. Over elevation combined with severe excyclotorsion.

Myectomy

Inferior oblique myectomy typically involves the surgical removal of a segment of muscle between the NFVB and the insertion of the muscle. The muscle segment is cauterized prior to removal. The primary advantage is convenience as this procedure is faster than a recession. The primary advantage is convenience as this procedure is faster than a recession. The advantage is the possibility that the muscle may reattach and lead to recurrent overaction.

SECONDARY INFERIOR OBLIQUE SURGICAL PROCEDURES

Residual or recurrent symptoms of inferior oblique overaction are common. Further weakening of the inferior oblique muscle can be accomplished by a further recession (re-recession). However, if the muscle was already maximally recessed or previously transposed, or if severe weakening is the objective, re-recession may not be an option.

Traditional drastic inferior oblique weakening procedures, such as denervation with extirpation or myectomy are disadvantageous; because they render the muscle definitively nonfunctional, prohibiting any further change or adjustment to the inferior oblique muscle.

Nasal myectomy. In this procedure, a 5-mm section of the muscle is excised along the muscle belly between the NFVB and the origin. The most recently described weakening operations on the inferior oblique muscle are nasal myectomy and anterior and nasal transposition.

Both anterior transposition and myectomy reduced the function of the overactive inferior oblique muscle, but eyes with anterior transposition tended to be more effective.

Hang Back recessions

Although hang-back recession has widely been used as a weakening procedure on extraocular muscles, its effectiveness has mostly been studied for rectus muscles. A surgical technique for recessing the inferior oblique muscle by Hang Back technique its effectiveness in V-pattern strabismus with inferior oblique overaction (IOOA) has been described by Kamlesh et al. and they concluded 10 mm Inferior oblique recession is a simple, safe and effective method for the cosmetic and functional treatment of horizontal deviation and V pattern with primary inferior oblique overaction. The surgical technique consisted of free suspension of one or both inferior oblique muscles 10 mm along their physiological path using

Figure 1
Table 1

<table>
<thead>
<tr>
<th>Overaction</th>
<th>Transposition Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>+4</td>
<td>At inferior rectus</td>
</tr>
<tr>
<td>+3</td>
<td>1 mm to 2 mm posterior to IR insertion</td>
</tr>
<tr>
<td>+2</td>
<td>3 mm to 4 mm posterior to IR insertion</td>
</tr>
<tr>
<td>+1</td>
<td>4 mm posterior and 2 mm lateral to IR insertion</td>
</tr>
<tr>
<td>DVD and IOOA</td>
<td>Full anteriorization</td>
</tr>
</tbody>
</table>

**Advantages**
1. It is landmark free surgery
2. Easily gradable from 6-12 mm
3. Minimal manipulation
4. Muscle is always available for reoperation and adjustments

**CONTRAINDICATION**
1. V pattern strabismus without IOOA
2. X pattern in patients with long standing exotropia causing overshoot in upgaze simulating IOOA.
3. Pseudo IOOA in craniofacial disorders.
4. Restrictive pathologies like after retinal detachment surgery, where IOOA may have associated incyclotorsion.

**COMPLICATIONS**
There are several possible complications of inferior oblique surgery that can occur:
1. The most common may be recurrent or persistent inferior oblique overaction, for which a secondary, more profound weakening procedure is often performed.

**Figure 2**
6-0 polyglactin 910 sutures bridging the cut ends of muscle (Figure 2). There are studies on various inferior oblique muscle weakening procedures. However, none of them are full proof and each case has to be treated depending upon the situation. IO will retract into its sheath and shall stay recessed according to amount of recession done.
transposition into a less profound, traditional recession or alternatively one of several superior oblique weakening procedures.

2. Damage to vortex vein: the vortex vein is vulnerable in most inferior oblique recession and transposition procedures. Damaging the vortex vein may result in excessive hemorrhaging (bleeding).

3. Retrobulbar hemorrhages have been reported.

4. Transient pupillary dilation.

**DISCUSSION**

A wide variety of surgical procedures can weaken the action of the inferior oblique muscle. However recession, anterior transposition and Hang Back recession are commonly used.

10 mm Inferior oblique recession is a simple, safe and effective method for the cosmetic and functional treatment of V pattern with primary inferior oblique overaction.

Kamlesh et al concluded 10 mm Inferior oblique recession by hang back technique is a simple, safe and effective method for the cosmetic and functional treatment of horizontal deviation and V pattern with primary inferior oblique overaction.

One challenge in understanding (and thus predicting) the effects of these techniques is that most inferior oblique procedures not only weaken the inferior oblique muscle but also modify, quantitatively and qualitatively, the field of action. A well-known example is the limitation of elevation in abduction after anterior transposition and the concurring apparent inferior oblique overaction in the contralateral eye, or AES. Placing the new insertion that is less temporal, or even nasal (as in the ANT procedure), to the inferior rectus border avoids the limitations of elevation in abduction.

As excyclorotation is the primary action of the inferior oblique in its natural position, some surgical procedures of the inferior oblique may alleviate torsional deviations. Regardless whether the preoperative torsional deviation is the reason for surgery, the potential for torsional effects of any inferior oblique procedure warrants attention. Preferably, torsional changes should also be measured with the double-Maddox rod test or fundus imaging.

Currently, there is good understanding of the mechanical functionality of the NFVB after inferior oblique anterior transposition. However, not all aspects of the effects of inferior oblique muscle surgery can be explained in terms of the geometry of the new insertion. This, in turn, indicates that greater study is needed regarding the response of connective tissue pulleys and pulley bands to inferior oblique surgery.

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5. Kumar K, MS Hari Narayan Prasad, MD, Sumit Monga, MS, FRCS, Rahul Bhola, MD; JAAPOS; 2008;12,4: 401–404.


**Correspondence to:**

Dr. Subhash Dadeya
Director-Professor Ophthalmology, Guru Nanak Eye Centre, Maulana Azad Medical College, Delhi, India.
Stereoacuity is derived from the words ‘stereo’ meaning solid and ‘opsis’ meaning sight. Stereovision or binocular stereopsis is the perception of depth after the transmission of visual information from both the eyes. The range over which fusion has occurred is known as the Panum’s Fusional Area. An imaginary line connecting points corresponding to zero retinal image disparity is known as a horopter. The left and right retinal images are fused for objects located along the horopter and within Panum’s area. Beyond Panum’s area, either in front or behind, diplopia occurs (Figure 1). Stereoaucity, which is reciprocal of stereo threshold is the stereopsis that can be resolved by minimal horizontal separation. Mean stereoaucity in a person with normal binocularity is around 20±10 arc sec. Any impairment in stereopsis leads to poor quality of vision, which can influence daily activities like difficulty in playing sports or performing visuo-motor tasks, which in turns leads to limited career options in adults.

Stereopsis develops during the first year of the life, followed by further improvement up to 18 months. It starts from 3-4 months of age, and rapidly develops during next 4-12 months. The right and left ocular dominance columns develop in harmony if normal visual input reaches from both eyes after birth. Fine stereopsis is possible after development of both ocular dominance columns. Lack of visual input by any eye results in suppression of ocular dominance column of that side. Normal alignment of two eyes with good vision, along with a good cortical and ocular motor function is necessary for normal stereovision. In strabismus, misalignment of the two eyes result in formation of retinal images at non-corresponding locations, which finally leads to abnormal stereovision. To restore normal stereovision after strabismus surgery is always a challenge for the strabismologists, especially in infantile and accommodative esotropia. Achievement of stereoaucity after strabismus surgery is usually associated with everlasting alignment of the two eyes together and less risk of amblyopia.

**Table 1: Various principles of dissociation used in stereoaucity testing**

<table>
<thead>
<tr>
<th>Principle</th>
<th>Details</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Haploscopic</td>
<td>Dissociation by using angled mirror in front of each eye.</td>
<td>Synaptophore, Stereoscope</td>
</tr>
<tr>
<td>Anaglyph</td>
<td>Dissociation by using coloured glasses</td>
<td>TNO test</td>
</tr>
<tr>
<td>Vectographic</td>
<td>Dissociation by using glasses with polarized filters. It prevents from the exasperation of seeing red-blue tints.</td>
<td>Titmus fly test, Randot stereopsis test</td>
</tr>
<tr>
<td>Panographic</td>
<td>Real depth stereogram which uses cylinder or prisms.</td>
<td>Lang’s test, Frisy devis distance stereotest (FD2 test)</td>
</tr>
</tbody>
</table>

**ESOTROPIA (ET)**

Infantile and accommodative esotropias are the most common forms of esodeviation seen in practice. Both are associated with abnormal binocular functions and stereopsis. Infantile ET has an onset at around 6 months of age, which is the critical period for the development of stereopsis. Studies have shown that the abnormal stereoaucity in infantile ET is due to...
Accomodative ET has a late onset at around 18-48 months of age and is commonly associated with hyperopic refractive error and/or abnormal accommodative convergence/accommodation (AC/A) ratio. The deviation is intermittent in initial stage that gradually progresses to become constant. Despite late onset, abnormalities of binocular sensory functions are associated with accommodative ET. According to literature, although stereoacuity is most susceptible to disruption by 3-4 months of age, it continues to be affected by abnormal binocular experience even beyond 4 years of age. Abnormal stereoacuity has been noted in both intermittent as well as constant accommodative ET. Despite the intermittency in deviation, 40% cases exhibit abnormal stereoacuity in accommodative ET. The stereodefects in accommodative esotropia may be primary in nature; either genetic or pre-existing before the onset of ET. Infact, hyperopia along with pre-existing abnormal stereoacuity precipitate accommodative ET.

A study by Fawcett S et al, has evaluated the effect of several factors on stereoacuity in accommodative ET. According to them, age of onset has minimal effect on stereoacuity outcomes with better stereoacuity documented in children with onset ≤24 months than in those with early onset at 7-17 months. The strongest predictor of stereoacuity outcome was the duration of constant misalignment (>4 months). Intermittent misalignment of <4 months duration had a better outcomes. Other studies have found high AC/A ratio to be associated with poor stereoacuity. This is because cases with high AC/A ratios are difficult to treat, treatment is often delayed and this leads to prolonged misalignment.

These observations go in favour of prompt and aggressive management of both infantile and accommodative ET to restore binocular functions.

**EXOTROPIA (XT)**

Intermittent exotropia is a common exodeviation affecting nearly 1% of the population. As the name suggests, it is intermittent to begin with that later deteriorates into a constant XT. Distance stereoacuity is the first to be affected. As the disease progresses, the deviation starts to manifest for both distance as well near and this leads to disruption of near stereoacuity as well. Sharma P et al, have suggested a cut-off- 40 arc sec for near and 400 arc sec for distance stereoacuity- as an indication for prompting surgical intervention in intermittent XT.

**STEREOACUITY FOLLOWING STRABISMUS SURGERY**

**Esotropia (ET)**

It is important to keep in mind that strabismus surgery is not just a cosmetic surgery. The ultimate target after all strabismus surgery is to restore normal stereovision in addition to the binocular vision. The presence or loss of stereovision is an important deciding factor in the success of strabismus surgery. Therefore, it is crucial to assess stereoacuity post-surgery to evaluate the success of the procedure. The table below summarizes the near and distance stereovision tests available in clinical settings.

<table>
<thead>
<tr>
<th>Stereopsis test for near</th>
<th>Principle</th>
<th>Available disparities (s of arc)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. TNO (The Netherlands Organization) test (Figure 2)</td>
<td>Anaglyph</td>
<td>480-15</td>
</tr>
<tr>
<td>2. Titmus fly test (Figure 3)</td>
<td>Vectographic</td>
<td>3500-40</td>
</tr>
<tr>
<td>3. Randot stereopsis test (Figure 4)</td>
<td>Vectographic</td>
<td>660-20</td>
</tr>
<tr>
<td>4. Lang’s test (Figure 5)</td>
<td>Panographic</td>
<td>1200-200</td>
</tr>
<tr>
<td>5. Frisby devis near stereotest (Figure 6)</td>
<td>Panographic</td>
<td>600-5</td>
</tr>
<tr>
<td>6. Synaptophore</td>
<td>Haploscopic</td>
<td>720-90</td>
</tr>
<tr>
<td>7. Lang 2 pencil test</td>
<td></td>
<td>5000-3000</td>
</tr>
</tbody>
</table>

**Stereopsis test for distance**

<table>
<thead>
<tr>
<th>Stereopsis test for distance</th>
<th>Principle</th>
<th>Available disparities (s of arc)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Frisby devis distance stereotest (FD2 test) (Figure 7)</td>
<td>Panographic</td>
<td>5-50</td>
</tr>
<tr>
<td>2. Distance randot test (Figure 8)</td>
<td>Vectographic</td>
<td>400-60</td>
</tr>
<tr>
<td>3. AO-Vectographic Project-O-Chart Slide test</td>
<td>Vectographic</td>
<td>480-30</td>
</tr>
</tbody>
</table>

These observations go in favour of prompt and aggressive management of both infantile and accommodative ET to restore binocular functions. The table below summarizes the near and distance stereovision tests available in clinical settings.
factor for the timing and outcomes of the strabismus surgery. The ideal time to intervene for infantile ET is 4-6 months and for intermittent divergent strabismus is 4-6 years. Proper assessment of visual acuity and stereoacuity should be done prior to any strabismus surgery by using various methods described earlier.

In infantile and accommodative ET, several studies have been done to look for the outcomes of stereoacuity after strabismus surgery. An early intervention and alignment within 8 prism dioptre (pd) is considered essential for good stereovision. As mentioned earlier, duration of misalignment plays an important role in disruption of stereoacuity in ET. Thus, an early intervention is expected to improve the stereoacuity outcomes in such cases by decreasing the duration of misalignment. However, the studies prove otherwise. A study published by Birch EE et al. concluded that in infantile esotropia despite an early intervention and good refractive correction only < 0.5% patients gained normal stereoacuity by 5 years of age. No stereoacuity was achieved in 60% cases. This suggests that there are other factors that interplay and contribute to poor stereoacuity outcomes in infantile ET. These factors are inability of the current surgical techniques to predict post-operative alignment and inability to achieve a post-operative residual deviation within 4-6pd in every case, which in turn affects the development of ocular dominance columns. Also, an intervention much earlier (2-3 months) than the current recommendations may be needed.

Accommodative ET is mostly managed with optical correction and surgical intervention may be needed in around 30% cases. Stereoacuity outcomes are anticipated to be better in accommodative esotropia than infantile esotropia due to following reasons: late age of onset of accommodative esotropia (mostly after 18 months of age) and presence of intermittency of deviation early in the course of the disease. But despite surgical alignment of strabismus, stereo deficits persist in many cases. In a prospective study by Birch EE et al., only 18% cases with accommodative ET had normal Randot stereoacuity during a follow-up of 4-11 years. Rest had either subnormal or nil stereoacuity.

Poor stereoacuity at immediate post-operative period in both infantile and accommodative ET is a poor predictor. Nil stereoacuity in immediate post-operative period is associated with greater risk of permanent loss of stereoacuity and recurrence of deviation requiring resurgeries. Also, poor stereoacuity is associated with greater risk of amblyopia and poor quality of life.
EXOTROPIA (XT)

Intermittent XT affects distance stereovision more than the near stereovision. Stereovision assessment is now used as an objective clinical method to look for the progression in intermittent XT. There are no specific guidelines concerning the timing for surgical intervention. Various studies have been done to look for the change in stereovision after strabismus surgery in intermittent exotropia. In a prospective study of 31 intermittent XT followed up for 6 months by Sharma et al., distance stereoacuity after strabismus surgery in intermittent exotropia were done to look for the change in surgical intervention. Various studies have been done to look for the change in stereovision after strabismus surgery in intermittent exotropia. In a prospective study of 31 intermittent XT followed up for 6 months by Sharma et al. distance as well as near stereovision showed significant improvement after surgery. Distance stereoacuity improved in around 87% patients, while no change was noted in 6.5% cases. Worsening of stereovision was seen in 2 cases who developed consecutive esotropia. The percentage of patients with poor near stereovision (<40 arc sec) improved from 81% to 42% post-operatively. In another study of 30 cases of intermittent XT by Singh et al., distance as well as near stereovision improved significantly from 50 s of arc to 17.5 s of arc, and 240 s of arc to 90 s of arc respectively 6 months following surgery. They concluded that pre-operative stereovision is an important predictor of stereovision outcomes. In a prospective case control study, Singh et al., reported that a distance stereovision worse than 70 s of arc (FD2) is poor prognostic sign for post-operative stereovision improvement. They also gave a cut-off of worsening of distance stereovision > 20 arc (FD2) as an indication for surgery.

CONCLUSION

Rapid stereovision development is seen in first two years of life. Any interruption of binocular input during this period leads to subnormal stereopsis. Both early-onset esotropia as well as exotropia are associated with stereo deficits. A major contributing factor is the greater duration of misalignment. Therefore, early management of strabismus (optical or surgical) is considered indigenous to successful restoration of binocular sensory status. However, studies reveal that despite early intervention recovery of stereovision remains a challenge, especially due to the role several other factors influencing its outcome. Better pre-operative stereovision is predictor of good post-operative stereovision outcomes with lower rate of recurrent deviations.

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Correspondence to:
Dr. Rohit Saxena
Professor of Ophthalmology
Dr. R.P. Centre for Ophthalmic Sciences
All India Institute of Medical Sciences
Ansari Nagar, New Delhi, India.
The incidence of congenital nasolacrimal duct obstruction (CNLDO) ranges from 5%-20%, and this condition usually resolves by 3-4 weeks after birth due to the negative pressure created by crying, sucking and nasal breathing by the neonate. Before one year of age, the CNLDO can be managed with conservative treatment like lacrimal sac compressions with or without antibiotic eyedrops. After one year of age, these children are commonly subjected to a conventional probing at most of the places, but in few centers, the primary/first probing is performed under the nasal endoscopic guidance. The primary probing provides >90% success rate making it a reasonably satisfying procedure to perform but in few, the conventional probing fails, or the CNLDO persists.

Definition of persistent CNLDO or failed probing: If the symptoms of CNLDO recur within 6 weeks of primary probing, the condition is called as persistent CNLDO or failed probing.

The treatment options for a persistent CNLDO are:

1. Nasal endoscopy guided lacrimal probing ± inferior turbinate medialisation
2. Balloon catheter dacryoplasty/ dilatation (BCD) of nasolacrimal duct (NLD)
3. Lacrimal intubation with silicone stents (mono or bicanalicular)
4. Dacryocystorhinostomy (DCR)- external or endonasal

Nasal Endoscopy Guided Lacrimal Probing ± Inferior Turbinate Medialisation

Procedure: A 2.7 mm pediatric Hopkins II telescope of 0° or 30° is used for the visualization of the inferior turbinate (IT), meatus and the floor of the nose. The Bowman’s lacrimal probe is passed similarly as in the conventional probing, and the tip of the probe is observed to appear from the inferior meatus region (Figure 1).

In case of an impacted turbinate, where there is no space between the lateral nasal wall and IT or the inferior meatus is very narrow, the IT is medialized by passing a periosteum elevator between the two structures and pulling it into the nasal cavity, thereby enlarging the inferior meatus. This allows the 2.7 mm telescope to go into the inferior meatus and observe for the opening of NLD. The mucosa surrounding the inferior meatus is preferably decongested for better visualization and minimizes the bleeding. A few surgeons also prefer a serial dilatation of the NLD.

Advantages: The nasal endoscopy provides visualization of the tip of the lacrimal probe, hence prevents the formation of a false passage and even it is formed, it helps in the redirection of the probe to its desired location. The nasal endoscopy also provides the information about the nature of IT and width of the inferior meatus.

Disadvantages: Require specialized skills, learning curve, and expensive equipment.

Success rate- 80-95%.

Balloon Catheter Dacryoplasty (BCD)

This is an effective treatment option for persistent CNLDO or as a primary procedure in children older than 48 months. Lacrimal probing alone is much less effective in children with stenosis, obstructions, or diffuse narrowing of the nasolacrimal duct proximal to the level of the valve of Hasner. The BCD of the NLD achieves true dilatation, is technically much easier
to perform than silicone intubation and obviates the need to remove the tube or concerns about the child pulling out the tube. A high success rate can be achieved with BCD in cases of failed probings and failed silicone intubation, as well. Hence the indications for BCD would be:

1. Failed probing at any age.
2. Failed silicone intubation at any age.
3. As a primary procedure in children with trisomy 21 at any age.

Children with trisomy 21 have an inadequate response to probing or silicone intubation. Silicone intubation should be reserved for patients with canalicular stenosis or used in combination with BCD in patients who have failed a primary BCD.

**Procedure:** After surgical preparation, the superior punctum is gently dilated. After ensuring an adequate nasal decongestion, the desired size Bowman probe is used to perform conventional probing. The tip of the probe can be directly visualized with an endoscope or indirectly confirmed with a metal-to-metal touch. This is to ensure that the probe has perforated through all the obstructions and has entered the nose. The probe is then removed and the probe of the balloon catheter is placed in precisely similar fashion.

The lacrimal balloon catheter is a cylindrical balloon along the length of the probe making its handling and feel like a Bowman probe. It is available in 2 sizes - a 2-mm balloon catheter (≤30 months of age) and a 3-mm balloon catheter (>30 months of age). The deflated balloon catheter (coated externally with a viscoelastic) is pushed through the upper punctum, canaliculus, lacrimal sac, and down the nasolacrimal duct into the nasal cavity. Once in position, the end is connected to the inflation device. The inflation is performed twice for 90 and 60 seconds sequentially, at a pressure of 8 atmospheres. At first, the distal nasolacrimal duct is dilated followed by the deflation of the balloon catheter and pulling it proximally towards the lacrimal sac-proximal NLD junction. The success rate of BCD can be up to 75% (age 24-48 months) and 82% (12-24 months).

**Advantages:** relatively non-invasive, single-step procedure, no need for the additional procedure (stent removal etc.)

**Disadvantages:** require specialized skills, availability, learning curve, expensive equipment.

**Success rate:** 75-100%.

**LACRIMAL INTUBATION WITH SILICONE STENTS (MONO OR BICANALICULAR)**

Silicone intubation of the lacrimal drainage system has been used for the children having persistent CNLDO or are older (>48 months of age) at presentation. The intubation system can be bicanalicular (common) or monocanalicular. In both, the end of the silicone stent is retrieved from the nose (inferior meatus), and one needs to keep the stent in-situ for 3 months.

The outcomes of monocanalicular intubation (with Masterka, Monoka-Crawford) and bicanalicular intubation are comparable. The advantage of the monocanalicular stent is the easy removal under topical anesthesia in the clinic without the need of sedation or general anesthesia.

**Procedure:** After surgical preparation, both upper and lower lacrimal puncta are gently dilated. After ensuring an adequate nasal decongestion, the desired size of Bowman's lacrimal probe is passed, and its tip is visualized with nasal endoscopy in the inferior meatus region. The Bowman's lacrimal probe is then removed, and the metal bodkin of Crawford's lacrimal probe is passed from either of punctum. The passage of the olive tip of the metal bodkin requires a sufficiently dilated lacrimal punctum. The further passage of bodkin is similarly as of a lacrimal probe. Once the olive tip is visualized in the inferior meatus, Crawford’s hook is passed, and the olive tip is hooked safely. Now the metal bodkin is pulled out of the nasal cavity. Now the other metal bodkin is passed similarly and retrieved from the nasal cavity. The whole lacrimal tract is now intubated with the silicone stents. The metal bodkins are detached from the silicone stents, and the stents are tied with each other, and the knot is placed in the inferior meatus (Figure 2).

Care is taken to keep the stent loop loose enough to prevent the cheese-wiring of the puncta. This is done by opening the eyelids apart at the end of the
procedure (Figure 3).

**Advantages:** failed BCD, canalicular stenosis, cheaper than BCD, provide constant dilatation of NLD.

**Disadvantages:** premature stent prolapse, extrusion or loss, punctum cheese wiring, nasal or conjunctival mucosal granuloma, need for removal (under GA- bicanalicular)

**Success rate:** 80-98%.

**DACRYOCYSTORHINOSTOMY (DCR)**

This procedure is considered as the final surgical procedure for the persistent CNLDO patients in whom the lacrimal sac is opened up into the nasal cavity, in the middle turbinate region via a bony osteotomy. This procedure has a success rate in pediatric age group- external (93.5%) and endoscopic endonasal (95.6%). This is advisably performed in patients after the BCD and nasolacrimal duct intubation.

**Critical points in pediatric DCR**

1. The postoperative fibrosis is a crucial feature in the healing of the nasal mucosa and lacrimal sac mucosa leading to the formation of a functional ostium

2. The bones are softer than adults, ethmoid sinuses are not fully developed, and nasal cavity is of smaller size.

3. The skull base is lower; hence the superior edge of bony osteotomy should be strictly by the opening of the common internal opening of common canaliculus in the sac.

4. The overall size of the sac is small and intraoperative bleeding is crucial. The intraoperative use of mitomycin-C and lacrimal intubation is helpful in improving the overall success rate of the procedure.

**BIBLIOGRAPHY**


**Correspondence to:**

Dr. Manpreet Singh
Department of Ophthalmology, Advanced Eye Centre, Post Graduate Institute of Medical Education and Research, Chandigarh, India
Pediatric Cataract Surgery: What Roles do Study Methodology and Surgical Techniques Play?

Dr. Sumit Monga

Consultant, Pediatric Ophthalmology, Strabismus and Neuro-Ophthalmology Service, Centre for Sight group of Eye Hospitals, Delhi NCR, India

The advances in technology and refinements in surgical techniques, over past two decades, have catapulted pediatric cataract surgery into a new era. This has led to vastly improved surgical and functional outcomes. Various innovative studies have contributed to the current understanding and evolution of surgical techniques that have helped to partly overcome the challenges of pediatric cataract management. However, pediatric cataract surgery has been associated with many complex issues that have been debated in literature. We believe that possibly the information on key aspects of pediatric cataract surgery may have been influenced by study methodologies and surgical techniques that have been reported in the literature.

SURGICAL TECHNIQUE

Pediatric cataract surgery has evolved from the procedure of discussion and aspiration in 1930’s, lensectomy in 1970’s, extra-capsular cataract extraction in 1980s to the present surgical technique of anterior continuous capsulorhexis with phaco-aspiration of lens matter with primary posterior capsulorrhesis with limited anterior vitrectomy.

The use of mechanized vitrectomy instrumentation to selectively perform a primary posterior capsulotomy and vitrectomy combined with IOL implantation resulted in decreasing the scourge of visual axis opacification and has led to fewer reoperations in younger children. In the literature, the rate of PCO is up to 100% when the posterior capsule remains intact. There is a strong relationship between age and incidence of PCO. The rate of PCO membranes is high in young children, reflecting greater tissue reactivity of lens epithelial cells (LECs).

When primary posterior capsulotomy is not combined with anterior vitrectomy, the incidence of posterior capsule closure is up to 60%. The main reason for the occurrence of VAO after performing a PCCC could be the increased LEC activity and also the presence of an intact anterior vitreous face, which acts as a scaffold for LEC migration. Reported rates of VAO after vitrectomy are less than 6%. The importance of anterior vitrectomy in pediatric surgery is emphasized in in cataract surgery in patients younger than 7 years by several authors.

In a study, the technique of optic capture through the posterior capsulorrhexis has been shown to prevent PCO. However, Vasavada et al found that optic capture without anterior vitrectomy did not always ensure a clear visual axis. Eyes with an obscured visual axis had reticular fibrosis of the anterior vitreous face in the first 2 months after surgery. Hence, vitreous Opacification could be a primary response of the anterior vitreous face when it occurs with the IOL optic rather than a secondary scaffold response caused by proliferating LECs, inflammatory cells, and exudate deposits.

As per the current understanding, one can make the following recommendations. In children younger than 5 years, PCC with anterior vitrectomy is advisable. Posterior CCC without anterior vitrectomy may be considered in children between 5 years and 7 years. In older children, maintaining intact posterior capsules is advised.

PRIMARY IOL IMPLANTATION IN CHILDREN

IOLs are being used increasingly for the optical correction of aphakia in infants following cataract surgery. In children older than 2 years of age, primary IOL implantation of foldable Acrylic IOLs is the current standard of care. There is growing evidence in literature to support the use of primary intracocular lens implantation in children less than 2 years of age.

While the surgical technique has mostly remained similar for all studies, the study methodology was variable. Most of the reports are retrospective, non-comparative series while others have compared the outcomes of primary IOL implantation with the group receiving contact lens for visual rehabilitation. The number of patients in these series failed to provide the statistical power necessary to adequately assess the outcomes of IOL implantation. Few studies depicted in Table 1 are prospective, out of only one is a randomized trial comparing outcomes in the IOL versus contact lens group. The reporting of results in the retrospective and non-randomized studies could have been marred by fallacies of inadequate sample size and lack of standardized protocols.

The primary outcome measure in majority of these studies has been the complication rate and overall safety profile of IOLs in young children. Visual axis opacification has been the most frequent complication in all studies, but its incidence rate has differed between them. One glaring explanation could be the variable follow up, as posterior capsular opacification (PCO) can have a delayed onset. Particularly, the use of Acrylic foldable IOLs has been implicated with delayed and milder variety of PCO.

Many studies, at the start of the decade, had some cases in which Polymethyl- methacrylate (PMMA) IOLs were implanted. Research has conclusively established that IOL material and design has a direct influence on PCO rates. Acrylic foldable IOLs have been documented lesser PCO rates and severity of PCO compared to PMMA and silicone foldable IOLs. This trend
has been corroborated by demonstrating increased biocompatibility and improved IOL design of Acrylic foldable IOLs. Further, placement of in-the-bag has been shown to be associated with lesser postoperative inflammation and PCO rates. Table 1 reveals that the information on type of fixation of IOLs either in sulcus or bag, either has been missed or the exact number of cases with each type of fixation has not been mentioned. Moreover, the presence of associated ocular anomalies, like persistent fetal vasculature, in almost half of the studies could have influenced the VAO rates. Hence, the differential reporting of visual axis opacification and re-operation rates in various studies could partly be explained by the above-mentioned factors. It is worth noting that in the recent study by IATS group, despite following meticulous surgical protocols based on current knowledge, the re-operation rates in the IOL group were reasonably high.10 Clearly, there are some undetermined factors, particularly in infants, which may lead to visual axis opacification.

We believe that certain aspects of methodology, practiced in this study, could have affected the outcomes. In a previous study, similar visual outcomes and complication rates were reported with the limbal and pars plana approaches for doing PPC in children (mean patient age: 6.3 years ± 2SD). But the latter approach would be more challenging in infants in view of inadequately developed pars plana. Moreover, the limbal approach for PPC also obviates the need for raising a conjunctival flap. This may seem more relevant, especially in view of the need for conjunctival preservation for any future glaucoma filtering surgery in these patients. Use of a vitrector for cortical clean up, as has been used in this study, may be challenging in congenital cataracts. It would have been useful if the authors had clarified whether there was any difference in between the two groups (IOL vs. Aphakia group), with respect to presence of residual lens matter, capsular contraction rates, posterior synechiae, IOL deposits or IOL displacement in their series. All of these can directly influence the rate of AEs and the need for AIS in the postoperative period. Additionally, the authors’ postoperative protocol included instilling topical steroids four times a day, which does not seem to corroborate with the amount of expected postoperative inflammation in infants. In our experience, topical steroids instilled 8-10 times/day and tapered over 6-8 weeks provides satisfactory outcomes.

The authors’ view that “surgery to remove the re-proliferated lens epithelial cells is straightforward” appears to be technically true, but the psychological trauma that the parents and the young

<table>
<thead>
<tr>
<th>Authors, Year of Publication</th>
<th>Type of Study</th>
<th>Laterality of Cataract</th>
<th>Coexisting Ocular / Systemic Pathology</th>
<th>N (No. of eyes)</th>
<th>Age Group (Wks)</th>
<th>Mean FU (mo)</th>
<th>Type of IOL</th>
<th>IOL power Calc formula</th>
<th>Type of IOL</th>
<th>Fixation</th>
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<tr>
<td>Lambert SR et al, 1999</td>
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<td>Holladay SRK II or I</td>
<td>Bag</td>
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<td>3-22</td>
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<td>SRK II or SRKT</td>
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<td>Retrospective</td>
<td>UL, BL</td>
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<td>29</td>
<td>0.8-43.2</td>
<td>33.4 ± 16.1 months</td>
<td>AcrySof single piece</td>
<td>Holladay</td>
<td>Bag (86.2%)</td>
<td>Sulcus (13.6%)</td>
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<td>Retrospective</td>
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<td>HSM-PMMA, AcrySof</td>
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<td>Ashworth JL, 2007</td>
<td>Retrospective</td>
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<td>25</td>
<td>1-51</td>
<td>44.36 months</td>
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<td>SRK T</td>
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<td>45</td>
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<td>4-24</td>
<td>12</td>
<td>AcrySof</td>
<td>Holladay I</td>
<td>Bag (93%), Sulcus (7%)</td>
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</table>

Table 1: Studies on Primary IOL implantation in Children less than 2 years of age: Methodology
child undergo with an additional surgery cannot be overlooked. The limited data available from other retrospective studies, evaluating the safety profiles of primary IOL implantation in infantile subset of patients, revealed much less re-operation rates.

An attempt has been made to document the visual outcomes of primary IOL implantation in monocular aphakia in infancy, first by a pilot study and then by a randomized clinical trial by the Infant Aphakia treatment study group. The results of the latter study, at 1 year of age, showed that there was no significant difference between the median visual acuity in the treated eyes with IOL and the CL group after cataract surgery. However, in the youngest group of children with AL < 19 mm. In a recent study, Nihalini B R et al concluded for eyes with significant deviation in prediction error (>0.5 D), there was usually an undercorrection, except with Hoffer Q, which was almost as likely to overcorrect as undercorrect. This may be explained by the higher proportion of short eyes (AL < 22 mm; 69 eyes) in their series, and because Hoffer Q was formulated for shorter eyes.

A potential source of error in IOL power selection in children than adults is inaccuracy of axial length and/or keratometry power measurements. Most studies have either used applanation or immersion technique for axial length calculation under general anesthesia. Anaplanation technique is thought to artificially reduce the AL and can contribute to the significant amount of additional error, especially for high powered IOLs. Additionally, most studies except one failed to mention whether they tried to minimize the inter-observer variability, by deploying single observer for biometry and refractive error measurements.

With the trend towards implanting IOLs in infants with shorter ALs (Table 1), there is a felt need to understand the accuracy and the differences between prediction formulas at the lower extremes of axial length and higher keratometry values. While in few studies, the age stratification of younger children was not done, in others an attempt was made to dichotomize children according to the age at the time of surgery. As most of the ocular growth occurs during the first 2 years of life, most studies have taken the same as the cut off for grouping children. In two major studies, the number of eyes in < 2 year age group constituted about one-fifth of the total, the percentages

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**IOL POWER CALCULATION AND MYOPIC SHIFT**

Other issues that have received attention, in literature, are the need for accuracy in post-operative target refraction and trend of emmetropisation of refractive errors. The need for pediatric IOL calculation formulae has been felt for long time, primarily because all available formulas were derived from considerations regarding the adult eye. Unlike adults, pediatric eyes undergo rapid growth and significant refractive changes in the early years. Moreover, in most pediatric cases, the desired postoperative refraction is different from Plano. All these factors add to the complexity of the IOL power calculations in children.

Various IOL formulae designed for adult eyes are being used to predict IOL power in pediatric eyes, which have shown varying degrees of accuracy. Neely et al found that the SRK II regression formula gave the least amount of variability, whereas the Hoffer Q gave the greatest amount, particularly among the youngest group of children with AL < 19 mm. In a recent study, Nihalini B R et al concluded for eyes with significant deviation in prediction error (>0.5 D), there was usually an undercorrection, except with Hoffer Q, which was almost as likely to overcorrect as undercorrect. This may be explained by the higher proportion of short eyes (AL < 22 mm; 69 eyes) in their series, and because Hoffer Q was formulated for shorter eyes.

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**Table 2: Studies on Primary IOL implantation in Children less than 2 years of age: Results**

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<td>Visual Axis Opacification(VAO)</td>
<td>VAO rate= 37.9%, Greater VAO with associated ocular Anomalies</td>
<td>37.9%</td>
</tr>
<tr>
<td>Lundvall A8, 2006</td>
<td>Complications and visual results</td>
<td>VAO rate=67%; Better visual outcomes in BL cases compared to UL cases</td>
<td>70%</td>
</tr>
<tr>
<td>Ashworth IL-9, 2007</td>
<td>Refractive Outcome</td>
<td>Satisfactory mean refractive outcomes, but wide range of errors</td>
<td>-</td>
</tr>
<tr>
<td>Ram J et al, 2010</td>
<td>Safety profile of IOLs</td>
<td>VAO rate 13.3%, No major refractive surprises</td>
<td>28%</td>
</tr>
<tr>
<td>IATS11, 2010</td>
<td>Grating visual acuity at 1 year of age, Adverse events</td>
<td>Similar visual outcome in IOL and CL ; VAO rate 72%, Higher reoperation rates in IOL group</td>
<td>63%</td>
</tr>
<tr>
<td>Gupta A et al, 2011</td>
<td>Safety profile of IOLs</td>
<td>VAO rate (6.7%); adverse events comparable in age group &lt; 6 months and beyond</td>
<td>NA</td>
</tr>
</tbody>
</table>
being 22.7% in the study by Neely et al and 16.2% in the study by Nihalini B R et al. In other studies, the cut off for ‘younger children’ varies between 12 months, 18 months and 36 months. This disparity and fewer number of cases, limits our understanding about refractive outcomes in the younger, especially the infantile age group. However, these studies concur that there is a greater variability of refractive outcomes, post cataract surgery, in younger children compared to the older children. In the work by Eibschitz et al, an analytical comparison of predicted implant power using in the pediatric range of axial length and keratometry values was performed. Significant differences in intraocular lens power prediction were found among the Holladay Q, Holladay I, and SRK II formulas. This explains the higher degree of prediction error that was documented in the age group <2 years, in two recent studies.

It has been reported there is a trend towards larger postoperative prediction errors in younger children, compared to adults. The refractive outcome of pediatric patients depends on the assumed anterior chamber depth (ACD), effective lens position of IOL and the effects of axial displacement of IOL. The effect is even more pronounced in pediatric eyes as these short eyes require higher power IOLs. The standardized assumptions about ACD and vaulting characteristics of an IOL within the bag may not be accurate for pediatric eyes owing to shallow anterior chambers and the particular postoperative dynamics of posterior capsule contraction, vitreous pressure, haptic angulation, effects of primary posterior capsulotomy and vitrectomy on lens position and, later, the sometimes significant re-proliferation of lens material. So it is imperative that studies, evaluating IOL calculation formulae in children, should include information on these parameters so as to facilitate better comparison between them.

It has been documented that axial elongation of the globe in a pseudophakic eye leads to myopic shift, akin to what happens in normally developing eyes. Younger children are more prone to have larger and more unpredictable myopic shifts. Further, the myopic shifts have been shown to be variable and no consistent correlations with pre-operative axial length or IOL power have been found.

One basic discrepancy, in the reporting of myopic shifts, in various studies has been the variable timing of the initial refraction and the length of follow up period. The initial refraction may vary from 2 weeks to 12 weeks, and the mean follow up has ranged from 1 to 7 years. This has led to disagreement on the amount of myopic shift, in different largely retrospective studies. Further the heterogeneity of study groups, in terms of age groups and number of eyes included, may be a determining factor.

It has been reported that pseudophakic eyes have tendency to grow longer than the fellow phakic eyes. Interestingly, in pseudophakic eyes apart from the normal process of axial elongation with age, there can possibly be factors like secondary glaucoma or amblyopic visual deprivation which could influence the amount of myopic shift. Further, there could be other undetermined reasons, including genetic factors that could contribute to the myopic shift. In the study by Greiner et al, the axial elongation in pseudophakic eyes was reduced in comparison to the phakic eye. It was worth noting that all eyes in this study had sulcus fixation of IOLs, compared to in-the-bag implantation in most other studies. However, the exact mechanism of sulcus fixation contributing to the myopic shift needs to be evaluated. Further, an interesting proposition was made by Nischal KK et al, who wondered whether the reduction of peripheral visual input by anterior capsular fibrosis in pseudophakic eyes might explain the refractive and axial length change following pediatric cataract surgery. It is possible that with improved understanding of IOL power calculation and prediction of post-operative refraction in younger children improved results may be achieved in future studies.

GLAUCOMA

Secondary glaucoma is one of the most vexing problems after congenital cataract surgery. It can emerge years later and can jeopardize vision. Some earlier studies had studied children and eyes with a co-existence of conditions associated with cataracts that will also develop glaucoma, such as Lowe’s syndrome, aniridia, and trauma. However, more useful information could be attained only after studying “uncomplicated” cases of surgical aphakia. There is a wide variation in the reported incidence of glaucoma, after congenital cataract extraction. It ranges between 6% and 58.7% of children, depending on the population studied and the length of follow-up. Initial reports by Chandler, Phelps and Arafat drew attention to this problem. Magnusson et al studied Swedish children born with cataracts and found a 12% incidence of aphakic glaucoma (mean follow up 9.6 years). This figure is probably more reliable since the study was conducted within the confines of a fixed geographic location with centralized data collection. In the Infant Aphakia Treatment study, preliminary results showed that glaucoma developed in 5% eyes in the contact lens group and 12% eyes in the IOL group. But the follow up was too short to derive a true incidence.

Various factors have been postulated to influence the risk of developing postoperative glaucoma viz, age at detection of cataract, age at cataract surgery, cataract surgery procedure, primary intraocular lens implantation, significant postoperative uveitis, and microphthalmos. However, such information is available from reports of selected individual case series, which may be subject to bias and confounding. The data, procured from British Congenital Cataract Study, was an attempt to derive results from population based research. Its finding suggested that detection of cataract was the only significant factor associated with the development of glaucoma after surgery for congenital cataract. It is imperative to believe that early detection of cataract would directly translate to early age of cataract surgery.

Literature review points out to a bimodal pattern of onset for aphakic glaucoma. The first onset peak is noticed within the first weeks to months following cataract surgery. This early-onset glaucoma is frequently associated with pupillary block, shallowing of the anterior chamber, and angle closure. It is well known that smaller eyes and eyes with reduced corneal diameter are more prone to develop angle closure in postoperative period. However, the unavailability of data about preoperative gonioscopic findings, in cases undergoing pediatric cataract extraction, limits our understanding about the pre-existing state of the angle and its predisposition to insult in the postoperative period. Residual lens material contributes to the development of glaucoma by forming Elschng pearls that may cause pupillary block and induce inflammatory adhesions in the angle and at the pupil edge which can cause pupillary block and angle closure.
closure. It has been observed that sulcus-fixation of the IOL is a risk factor for the development of pupillary block. But, it is notable that most studies reporting the incidence of glaucoma following pediatric primary IOL implantation have failed to specify the differentiation of in-the-bag versus sulcus fixation. The possible mechanism of glaucoma can occur due to the pupillary block leading to extensive synechial closure or the forward displacement of iris tissue by the haptics of sulcus fixated IOL causing crowding of structures in the angle.

The age of onset of delayed-onset, open-angle glaucoma may occur years following the cataract. Because of the late onset occurring five or more years following the cataract surgery, it is not likely that inflammation, use of postoperative corticosteroids, or any other portion of the cataract surgery is the cause of this problem. Asrani et al., in a meta-analysis of 377 eyes with primary lens implantation, found only one eye with open angle glaucoma. They have suggested that when a primary IOL is used, there is a reduced incidence of delayed-onset glaucoma. However, it is worth noting that implants are infrequently placed in eyes with microcornea and the surgery in this series was relatively delayed (mean age at surgery: 5.06 years). Hence, two important risk factors, microcornea and early age of surgery, which could have influenced the results otherwise, were conspicuous by their absence.

Although it is clear that pediatric cataract surgery places the eye at risk for glaucoma, the exact mechanism remains elusive. The volume of the lens and its dynamic role in accommodation are more prominent in younger eyes. Surgical removal of the lens early in life can alter normal development of the filtration angle. Morphologic studies of developing eyes of children have shown that the angle recess of the iridocorneal angle is expected to move toward the periphery, exposing the scleral spur and the ciliary body band. It has been speculated that the absence of the lens early in life alters or causes an arrest in development of the filtration angle, or it may be a lack of accommodation and pull of the ciliary muscles on the trabecular meshwork that in some way permits the meshwork to become compact and dysfunctional. It may be that some chemical substance may diffuse from the posterior eye into the anterior chamber and change the facility of outflow of the eyes. Clearly, there are some undefined genetic and mechanical factors that may contribute to the development of glaucoma after pediatric cataract surgery.

On one hand, early cataract surgery has been advocated to prevent amblyopia and on the other hand, the possibility of increased complication rate after early cataract surgery looms large. This leads to the dilemma, for which there are no clear answers. At present, the clinician may be best advised to do a balancing act of performing early cataract surgery, avoiding the neonatal period, followed by a careful surveillance, to detect postoperative glaucoma. The latter must continue for long term basis. Probably future prospective studies, in which children would be randomized to cataract surgery at different ages within the critical period, would enlighten us about the issue of optimum timing.

REFERENCES
Childhood Glaucoma—The CGRN Classification

Dr. Deepika Dhingra, Dr. Savleen Kaur, Dr. Sushmita Kaushik

Advanced Eye Centre, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Childhood glaucoma classification began with the term buphthalmos, or ox-eyed, which was used to describe the secondary effect of elevated intraocular pressure (IOP) on the elastic infantile eye. Buphthalmos was subdivided into simple buphthalmos due to a primary mechanism, now known as primary congenital glaucoma, and buphthalmos associated with other developmental anomalies. The more complex anatomic classification was given by Hoskins, which divided the developmental abnormalities into trabecular meshwork, iris, and/or cornea. The Shaffer-Weiss classification introduced the categories of isolated congenital (infantile) glaucoma, glaucomas associated with congenital anomalies, and acquired glaucoma. Walton proposed an exhaustive listing of all disorders known to be associated with childhood glaucoma.

Unlike the adult definition of glaucoma which focuses on the pathology of the optic nerve to make the diagnosis, subdividing glaucoma in children is more difficult because the optic nerve can be difficult to evaluate properly in the presence of corneal opacity.

The definition of childhood glaucoma is IOP-related damage to the eye, rather than being based solely on optic-nerve criteria as suggested by an expert panel of glaucoma specialists in the world. The childhood glaucoma research network (CGRN) composed of clinicians and scientists who specialize in treating children with glaucoma, has provided definitions of childhood glaucoma, glaucoma suspect, and a new classification system of pediatric glaucomas.

According to CGRN classification, childhood age is based on national criteria:
- US: younger than 18 years
- EU, UK, UNICEF: 16 years or younger

DEFINITIONS OF GLAUCOMA AND GLAUCOMA SUSPECT

Glaucoma: IOP related damage to the eye; at least 2 criteria required for diagnosis:
1. IOP > 21 mm Hg; however, investigator discretion is required if there is data of examination under anesthesia alone due to variable effects of anesthesia on all methods of IOP assessment.
2. Optic disc cupping: progressive increase in cup-disc ratio.
3. Cup disc asymmetry of ≥ 0.2 when the optic discs are similar size, or focal rim thinning.
4. Corneal findings: Haab’s striae or diameter ≥ 11 mm in newborn, > 12 mm in child < 1 year, or > 13 mm at any age.
5. Progressive myopia, myopic shift, or an increase in ocular dimensions out of keeping with normal growth.
6. Reproducible visual field defect consistent with glaucomatous optic neuropathy with no other observable reason for defect.

Glaucoma suspect: No IOP related damage; at least 1 criterion required for diagnosis:
1. IOP > 21 mm Hg on 2 separate occasions.
2. Suspicious optic disc appearance for glaucoma, i.e., increased cup disc ratio for size of optic disc.
3. Suspicious visual field for glaucoma.
4. Increased corneal diameter or axial length in the setting of normal IOP.

CGRN CLASSIFICATION OF CHILDHOOD GLAUCOMA

Primary childhood glaucoma
IA. Primary Congenital Glaucoma (PCG) (Figure 1)
1. Isolated angle anomalies (+/- mild congenital iris anomalies)
2. Meets glaucoma definition (usually with ocular enlargement)
3. Subcategories based on age of onset
   a. Neonatal or newborn onset (0-1 month)
   b. Infantile onset (>1-24 months)
   c. Juvenile onset or late-recognized (>2 years)

Figure 1: Primary Congenital Glaucoma. A-C: Newborn glaucoma; A: Central Haab’s striae; B: Persistent corneal opacity in a rubella positive neonate after IOP control; C: Bilateral large eye with right eye central corneal opacity and optical iridectomy. D-E: Infantile Glaucoma
D: Bilateral large eyes with clearer cornea; E: Bilateral glaucoma with left eye larger in size than right eye and haab striae in left eye. F: Late onset PCG presented at 5 years of age with bilateral large cornea (L>R) and advanced cupping in left eye.
Recent trends and advances

Kaushik S. et al. Childhood Glaucoma the CGRN Classification

Secondary childhood glaucoma

A: Glaucoma Associated with Non-Acquired Ocular Anomalies (Figure 3)
Includes conditions of predominantly ocular anomalies present at birth which may or may not be associated with systemic signs and meets glaucoma definition

Includes:
- Axenfeld-Rieger anomaly (syndrome if systemic associations)
- Peters anomaly (Syndrome if systemic associations)
- Ectropion uveae
- Congenital iris hypoplasia
- Aniridia
- Persistent fetal vasculature/PFV (if glaucoma present before cataract surgery)
- Oculodermal melanocytosis (nevus of Ota)
- Posterior polymorphous dystrophy
- Microphthalmos, Microcornea
- Ectopia lentis
- Simple ectopia lentis (no systemic associations, possible Type 1 FBN mutation).

B: Glaucoma Associated with Non-Acquired Systemic Disease or Syndrome (Figure 4)

Includes conditions predominantly of systemic disease present at birth which may be associated with ocular signs and meets the definition of glaucoma; namely
- Chromosomal disorders such as Trisomy 21 (Down syndrome)
- Connective tissue disorders

4. Spontaneously arrested cases with normal IOP but typical signs of PCG may be classified as PCG

1B. Juvenile Open Angle Glaucoma (JOAG) (Figure 2)
1. No ocular enlargement
RECENT TRENDS AND ADVANCES

Figure 6: Glaucoma secondary to cataract surgery. A: Unilateral glaucoma after cataract surgery in right eye; B: Iris-fixated intraocular lens (IOL) in a case of microcornea; C: Iris-fixated IOL in a case of microcornea with corneal decompensation and glaucoma; D: Iris-fixated IOL with shallow anterior chamber and glaucoma.

Dhingra et al. Childhood glaucoma classification

C. Glaucoma Associated with Acquired Conditions (Figure 5)

1. Meets glaucoma definition after the acquired condition is recognized (except congenital cataract surgery)
2. Acquired conditions
3. Based on gonioscopy results:
   - Open angle glaucoma (>50% open or acute angle closure).
   - Angle closure glaucoma (<50% open or acute angle closure).

Includes:
- Uveitis
- Trauma (hyphema, angle recession, ectopia lentis)
- Steroid induced
- Tumors (benign/malignant, ocular/orbital)
- Retinopathy of Prematurity
- Prior ocular surgery other than cataract surgery.

D. Glaucoma Following Congenital Cataract Surgery (Figure 6).

Meets glaucoma definition after cataract surgery performed.

Excludes acquired cataract or cataract in the setting of a syndrome with a known glaucoma relationship, such as Lowe syndrome, congenital rubella syndrome, aniridia, or persistent fetal vasculature.

SUGGESTED READING


Correspondence to:
Dr. Sushmita Kaushik
Glaucoma Services, Advanced Eye Centre
Department of Ophthalmology, PGIMER, Chandigarh, India

Figure 7: Shows an algorithm for classification of childhood glaucoma

- Marfan syndrome
- Weil-Marchesani syndrome
- Stickler syndrome
- Metabolic disorders
- Homocystinuria
- Lowe syndrome
- Mucopolysaccharidoses
- Phacomatoses
- Neurofibromatosis (NF-1, NF-2)
- Sturge-Weber syndrome
- Klippel-Trenaunay-Weber syndrome
- Rubinstein-Taybi
- Congenital rubella

C. Glaucoma Associated with Acquired Conditions (Figure 5)

1. Meets glaucoma definition after the acquired condition is recognized (except congenital cataract surgery)
2. Acquired conditions
3. Based on gonioscopy results:
   - Open angle glaucoma (>50% open or acute angle closure)
Intra-vitreal Anti-vascular Endothelial Growth Factor Drugs for Treatment of Retinopathy of Prematurity

Dr. Bruttendu Moharana MS, Dr. Savleen Kaur MS, Dr. Mangat Ram Dogra MS

Advanced Eye Centre, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Retinopathy of prematurity (ROP) is a proliferative vitreoretinopathy in preterm infants. It is one of the major avoidable causes of childhood blindness affecting countries of all economic strata. ROP causes lifelong irreversible significant visual loss in the infants. It is well established that low birth weight and small gestational age are good clinical predictive factors for development of ROP. Other risk factors include high level of oxygen supplementation, hypercapnia, anaemia, chronic lung disease, blood transfusion, sepsis, intraventricular haemorrhage, multiple birth etc.

ROP has become a major public health problem in middle income country like India. Increasing number of neonatal intensive care units (NICU) and special new-born care unit (SNCU) has led to survival of more and more very low birth weight (VLBW) infants. But suboptimal neonatal care, lack of awareness, screening and treatment programme has caused the absolute number of infants with ROP to rise.

Pathogenesis of ROP

ROP is a two-phase disease. Phase I is characterised by delayed retinal vascular growth due to hyperoxia. Neonatal hypoxia results in apoptosis and capillary regression. This interrupt normal vascularisation and leads to retinal ischemia. Phase I occurs from birth to 30-31 weeks of gestation. The metabolically active but nonperfused immature retina becomes hypoxic and stimulates over-production of VEGF. This marks phase II which begins around 31-32 weeks of gestation. Excess VEGF leads to formation of abnormal new vessels, fibrovascular proliferation and later tractional retinal detachment. Fetal IGF 1 (insulin like growth factors) and various other angiogenic cytokines also play a role in pathogenesis of ROP.

Gold Standard Treatment of ROP

Treatment is primarily aimed at ablating the avascular, hypoxic retina and converting it into anoxic retina and thereby preventing excess VEGF secretion and the complications that follows. Cryotherapy and laser photocoagulation to the avascular retina is the traditional treatment modality with laser treatment being the gold standard (Figure 1). In ETROP study, laser ablation of peripheral retinal significantly reduced progression of the disease, although the visual outcome was poor especially in zone I disease. Also, there is permanent loss of peripheral visual field and development of high myopia later in life. Progression to retinal detachment occurred in 12% of eyes in the ETROP study even with adequate peripheral ablation. This may be related to either the individual skills of the surgeon who applies laser treatment, or vitreal VEGF levels.

Figure 1: Male neonate, 1000gm birth weight, 28-week gestational age presents at 34-week PMA. BE APROP with plus disease in posterior zone II (image on the left). BE laser photocoagulation done. Marked regression of ROP at 4-week after laser (image on the right).
which cannot be reduced by retinal laser photocoagulation. Although VEGF is mainly secreted in the avascular retina, vitreal macrophages as a second source of VEGF may be responsible for the lack of the effectiveness of laser therapy for ROP.

ANTI-VEGF THERAPY IN ROP

VEGF is a potent mitogen for vascular endothelial cells and necessary for the physiological angiogenesis. It is regulated by tissue hypoxia. It also promotes pathological angiogenesis. Blocking the action of VEGF is expected to reduce the neovascular activity associated with ROP. Treatment with cryotherapy or laser photocoagulation indirectly reduces VEGF levels by ablating the peripheral avascular hypoxic retina and converting it into anoxic retina. On the other hand, anti-VEGF agents don’t have the disadvantage of tissue destruction. Also, these drugs decrease VEGF levels both in the retina and vitreous.

Various anti-VEGF drugs in use are pegaptanib sodium (Macugen), ranibizumab (Lucentis), bevacizumab (Avastin) and aflibercept (Eylea). Pegaptanib sodium, ranibizumab and aflibercept are approved by the US Food and Drug Administration (FDA) for intraocular use in the treatment of adult ocular neovascular diseases. Bevacizumab has not yet been approved by FDA for the same.

However, the safety and efficacy of these drugs in preterm neonates is yet uncertain. There is lack of consensus on criteria, dosage, frequency of administration and follow-up schedule. VEGF is essential for organ development of neonates. There is no conclusive data on any late systemic complications and neuro-developmental issues after treatment with anti-VEGF drugs. In this review, we discuss the efficacy and safety of anti-VEGF drugs used as monotherapy or along with cryotherapy or laser photocoagulation.

TRIALS USING ANTI-VEGFs IN ROP

Use of pegaptanib along with conventional laser (combined treatment) has significant favourable outcome compared to only cryotherapy or laser photocoagulation. In patients with stage 3 ROP in zone I and posterior zone II, the combined modality of treatment showed faster regression of plus disease and development of peripheral retinal blood vessels and lower rate of recurrence of neovascularisation. Pegaptanib stimulated development of peripheral retinal blood vessels rather than preventing it. Although, it is well established that anti-VEGF agents impede peripheral retinal vascular development leading to persistent peripheral avascular retina and late recurrence of the disease.
Though recurrence rate was lower, the interval from treatment to recurrence was significantly higher in eyes receiving combined treatment. This highlights the need for long term follow-up in patients who receive anti-VEGF therapy. No local or systemic complications were noted.

BEAT-ROP (Bevacizumab Eliminates the Angiogenic Threat of ROP)\textsuperscript{11} is a prospective, controlled, randomized, stratified, multicentre trial to assess intravitreal bevacizumab monotherapy for zone I or zone II posterior stage 3+ (i.e., stage 3 with plus disease) retinopathy of prematurity. Infants were randomly assigned to receive intravitreal bevacizumab (0.625 mg in 0.025 ml of solution) or conventional laser therapy. Intravitreal bevacizumab monotherapy, as compared with conventional laser therapy, in infants with stage 3+ retinopathy of prematurity showed a significant benefit for zone I but not zone II disease. At 54 week of postmenstrual age (PMA), the rate of recurrence for zone I disease was significantly higher with conventional laser therapy than with intravitreal bevacizumab. There was no significant difference in recurrence rate in posterior zone II disease. Development of peripheral retinal vessels continued after treatment with intravitreal bevacizumab, but conventional laser therapy led to permanent destruction of the peripheral retina. APROP regression rate was significantly better after bevacizumab monotherapy compared with laser photocoagulation at follow-up of 80 weeks\textsuperscript{12}.

The use of intravitreal ranibizumab for the treatment of ROP has been described in several case reports and series with variable results. Intravitreal ranibizumab induced rapid, complete regression of high-risk posterior ROP with continued growth of peripheral retinal vessels. The potential for recurrent ROP requires vigilant monitoring. Subsequent peripheral laser for ROP recurrences may spare the posterior retina from the destruction caused by photocoagulation\textsuperscript{13}.

Both bevacizumab and ranibizumab have similar responses in neovascularization and plus disease regression, and none of the eyes had recurrence of ROP after an initial good response. After one-year follow-up, there was no recurrence of disease if there was initial good response to either drug. Multiple other studies show higher recurrence rate in group treated with ranibizumab\textsuperscript{15,16}. Aflibercept, which inhibits all isoforms of VEGF-A may provide longer duration of intraocular effect\textsuperscript{17}. No randomised study is available to compare the efficacy of ranibizumab or bevacizumab with aflibercept.

DISEASE RECURRENCE

Recurrence occurs much later after anti-VEGF treatment than conventional laser photocoagulation. Time interval from anti-VEGF injection and disease recurrence varies with different studies. Hwang et al\textsuperscript{18} reported 1-year recurrence free interval after with either bevacizumab or ranibizumab. Castellano et al\textsuperscript{19} in a small case series showed 3-year recurrence free interval after single ranibizumab injection. However multiple other case reports and series have reported early and late recurrence after anti-VEGF treatment\textsuperscript{20,21,22}. Wong et al\textsuperscript{23} reported reactivation after ranibizumab injection between 5 to 7 weeks after injection whereas recurrence free interval of 9 to 10 months with bevacizumab injection. Chan et al\textsuperscript{24} reported reactivation between 4 to 8 weeks after treatment with ranibizumab. Hu et al\textsuperscript{25} have reported recurrence at 35 weeks post-injection of bevacizumab.

SAFETY OF ANTI-VEGF AGENTS

VEGF plays many positive roles related to neural, vascular, and lung development. The long-term systemic complications of anti-VEGF agents are not known\textsuperscript{26}. BEAT-ROP suggests that, bevacizumab being a large molecule cannot penetrate the intact retina or escape the eye except in very small amounts. The study was not large enough to assess if bevacizumab injection results in significant mortality and morbidity. There were five deaths in the bevacizumab arm of the study. Though statistically not significant, it is of concern that four out of these five deaths were related to respiratory problem. Other studies have shown that bevacizumab has detectable concentration in the serum of the neonate after intra-vitreal injection\textsuperscript{27}. Bevacizumab has longer serum half-life than ranibizumab. So theoretically, ranibizumab is a safer option for the neonates.

High myopia after laser photocoagulation is a well-known complication. In BEAT-ROP study high myopia was found in eyes that received laser treatment than compared to the eyes that received intravitreal bevacizumab. The difference is related to anterior segment development that is present with intravitreal bevacizumab but minimal or absent following laser ablation of peripheral retina.

Neurodevelopmental delay is another area of concern while treating ROP with anti-VEGF. Rien et al\textsuperscript{27} studied sixty-one patients who were treated with bevacizumab. At two years after treatment, no difference on neurodevelopment for those who received only bevacizumab versus only laser treatment were found. Those infants who required rescue therapy with laser or bevacizumab injection after initial unsuccessful treatment showed some detrimental neurodevelopmental effects. The reason for this is not investigated in the study. It could be due to a greater number of procedures requiring more sedation or anaesthesia. Morin et al\textsuperscript{28} in an observational study found higher odds of severe neurodevelopmental disabilities in infants treated with bevacizumab. In contrast, two-year follow-up of patients enrolled in BEAT-ROP didn’t show any adverse neurodevelopmental effect in the bevacizumab arm. There were no significant differences between the bevacizumab and laser therapy groups in weight, length, head circumference, cerebral palsy, or Bayley scores\textsuperscript{29}. There are two potential explanations for the difference. First, in the non-randomised observational study\textsuperscript{28} the sicker infants would preferentially be treated with bevacizumab. Second, the observational study was much larger and thus had greater power to identify differences. Although the BEAT-ROP study did not identify any concerning trends toward worse systemic outcomes among infants treated with bevacizumab, it was not adequately powered to identify significant differences.

CONCLUSION

1. Anti-VEGF agents have been claimed to have multiple advantages over conventional laser photocoagulation for treatment of ROP. These are:
   A. They are less destructive than laser treatment as they don’t ablate the peripheral retina.
   B. There is no permanent loss of peripheral field.
   C. There is less chance of development of myopia.
   D. Anti-VEGF drugs are easily accessible and easier and faster to administer compared to laser photocoagulation.
E. The short duration of treatment is highly important if laser photocoagulation cannot be performed in very sick neonates. It also can be used in eyes with small pupil or hazy media.

2. The possible local complications related to anti-VEGF injection are lens injury, retinal detachment, and infection. The concerns are related to the systemic safety issues of anti-VEGF, which are yet to be studied in randomised controlled trials with a large cohort. Studies are needed to establish safe yet effective dose of anti-VEGF.

3. Late recurrence of disease is major area of concern with anti VEGF therapy when used alone. More studies are needed to determine an appropriate follow-up schedule after anti-VEGF treatment.

4. Insufficient data precludes strong conclusion favouring monotherapy of anti-VEGF agents or combination therapy with laser photocoagulation in preterm infants with ROP. However, these drugs could be used as a rescue treatment in sick infants or eyes with non-dilating pupil or poor media after informing the caretakers all risks associated with anti-VEGF drugs.

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Ambylopia is classically described as unilateral or bilateral reduction in the best corrected visual acuity due to form vision deprivation and/or abnormal binocular interaction in the absence of any identifiable organic pathology. Visual neurodevelopment is immature at birth and appropriate integration of binocular information at the cortical level requires adequate visual stimulation during the critical period of development. Critical period of visual development is the period of cortical brain plasticity and extends up to 7-8 years of age. Conditions that interfere with binocular sensory fusion such as significant refractive error, anisometropia, stimulus deprivation and ocular misalignment cause abnormal visual development resulting in amblyopia. The standard treatment regimen for unilateral amblyopia is based on eliminating the competitive advantage of the dominant eye by providing appropriate refractive correction and occlusion therapy or penalization. However, maximum response to treatment is seen during the critical period and, if left untreated past this period of cortical plasticity, amblyopia can result in permanently impaired vision.

Changes in excitatory-inhibitory balance regulate the plasticity of neural networks. A number of current researches aim at evolving strategies to modulate brain plasticity in adulthood as well.

CORTICAL BRAIN PLASTICITY AND ITS MODULATION

Duffy et al. conducted animal experiments with bicuculline, a GABA receptor blocker and suggested that a reduction of GABA may be a crucial step for restoration of plasticity in adulthood. GABA neurons express parvalbumin and become emwrapped by perineuronal nets. It has been seen that maturation of these parvalbumin cells corresponds to the closing of the critical period. Techniques that promote parvalbumin cell function or delay perineuronal net formation can push plasticity onset into adulthood. The involvement of perineuronal nets was demonstrated by Pizzorusso et al., when an injection of the enzyme chondroitinase into the visual cortex of adult rats dissolved the extracellular matrix and destroyed perineuronal nets to reactivate ocular plasticity. After patching a rat’s good eye, the researchers witnessed the recovery of normal vision. Cortical circuits for both the left and right eyes began firing together, just as they are during the early phase of childhood development. The Hensch lab has demonstrated that brain plasticity is modulated by cholinergic transmission, and that it may be possible to reactivate plasticity by manipulating cholinergic pathways. In adult mice with amblyopia, vision was restored to normal acuity levels by administration of acetylcholinesterase inhibitors. Thus, based on the laboratory evidence one can potentially accomplish effective amblyopia therapy even after the “critical period” has passed.

PHARMACOLOGICAL AGENTS

Several different pharmacological agents have been identified to have clinical application in the treatment of amblyopia. Penalisation with atropine is the most common of all these. We are describing four new drugs whose effects are relatively promising.

DOPAMINE

Dopamine is a neurotransmitter that has been evaluated for its ability to enhance cortical plasticity. Dopamine is active in the retina and the cortex but does not cross the blood–brain barrier. A catecholamine precursor to dopamine, known as levodopa, does cross the blood–brain barrier and is converted to dopamine in the brain. Levodopa, in combination with a peripheral decarboxylase inhibitor – Carbidopa, has been used in various human studies on amblyopia. Carbidopa inhibits the peripheral conversion of levodopa to dopamine and therefore increases the availability of levodopa in the central nervous system. Levodopa–Carbidopa is usually administered in a 4:1 ratio. Most studies advocate a low dose of 1.5 to 2 mg/kg levodopa for a 3-7 week period.

Yang et al. have shown that older amblyopic children treated with carbidopa-levodopa had visual cortical activation. Levodopa administration has also been shown to improve pattern VEP and decrease fixation point scotomas. The Pediatric Eye Disease Investigator Group (PEDIG) evaluated the role of levodopa in amblyopia and found that despite initial improvement in visual acuity, there may be partial regression after stopping the medication.

CITICOLINE

Citicoline is a molecule that prevents neurological cell damage by maintaining the anatomical and functional integrity of cell membrane. It has been used in cases of ischaemic and traumatic insults to the neuronal tissue and also as an adjunct to levodopa in Parkinson’s disease. Citicoline improves membrane ATPase activity and modulates the turnover of catecholamine and acetylcholine. Oral administration of citicoline combined with occlusion therapy seems to show that there are more stable effects of this medication on the results of occlusion therapy compare to occlusion therapy alone. And in patients beyond the critical period of visual plasticity, citicoline showed visual acuity improvement, although the improvement was not
Recent Trends and Advances

**Donepezil**

The neurotransmitter Acetylcholine (Ach) is involved in many cognitive functions, including attention and learning. The role of Ach in facilitating plasticity has been demonstrated in animal models by increasing the level of norepinephrine and dopamine levels in CNS. It has been shown to inhibit apoptosis in neurodegenerative models, thereby potentiating neuroplasticity.

**Fluoxetine**

Fluoxetine is a selective serotonin reuptake inhibitor which increases extracellular serotonin and noradrenaline levels. It has been shown by Maya Vetencourt et al. to reactivate cortical plasticity in ambyopic rats, promoting full recovery of visual function. However, there have been no clinical studies involving fluoxetine in amblyopia. Fluoxetine can have systemic side effects such as irritability, behavioural changes, restlessness, and agitation. This drug should be prescribed with caution in patients with impaired liver and renal function, in case of diabetes and bipolar disorders.

**Citicholine**

Citicholine, once absorbed, crosses the blood–brain barrier and gets incorporated into the cell membrane phospholipids. It has been shown to increase the levels of norepinephrine and dopamine levels in CNS. It has been shown to inhibit apoptosis in neurodegenerative models, thereby potentiating neuroplasticity.

This drug is currently being used for the treatment of Alzheimer’s disease, and for control of cognitive impairment in dementia, schizophrenia, Down’s syndrome, and autism spectrum disorders. The ability of Donepezil to enhance perceptual learning and modulate neural plasticity makes it a potentially promising drug for the treatment of amblyopia even in adults. Studies evaluating the benefit of Donepezil in amblyopia are currently underway.

**Conclusion**

While occlusion therapy remains the primary weapon, our armamentarium for treating amblyopia is expected to expand in the near future with the availability of newer drugs and techniques for neuromodulation. The amblyopic patients who cross the age of cortical plasticity are sustained after discontinuation of the medication.

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**Table 1: A brief review of drugs used for Amblyopia**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Mechanism</th>
<th>Dosage/Duration</th>
<th>Adverse Effects</th>
<th>Remarks</th>
</tr>
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<tbody>
<tr>
<td><strong>Levodopa-carbidopa combination therapy</strong></td>
<td>Levodopa is a precursor of dopamine, and easily crosses blood–brain barrier. Carbidopa is a peripheral decarboxylase inhibitor which prevents peripheral conversion of levodopa to dopamine, thereby increasing the availability of levodopa in the central nervous system. The drug significantly increases the density of NGF-immunoreactive cells and elevate the expression of endogenous NGF in the visual cortex.</td>
<td>Various doses of levodopa have been tried for different durations: single-dose, 1-week, 3-week, and 7-weeks. Some studies used lower doses – 1.5 mg/kg/day and 30 mg/day of levodopa – while others used higher ones – 6–13 mg/kg/day. Levodopa is usually administered along with carbidopa in a 4:1 dose ratio, either in the form of oral tablets or as oral suspension.</td>
<td>Lowers body temperature, headache, nausea, dry mouth, and abdominal cramps</td>
<td>Promising in older children</td>
</tr>
<tr>
<td><strong>Fluoxetine</strong></td>
<td>It acts by altering the cortical expression of various heat shock proteins and neurofilaments which are important for synaptic functions</td>
<td>Fluoxetine can have various side effects such as irritability, behavioral changes, restlessness, and agitation. This drug should be prescribed with caution in patients with impaired liver and renal function, in case of diabetes and bipolar disorders.</td>
<td><strong>Citicholine</strong></td>
<td>Citicholine, once absorbed, crosses the blood–brain barrier and gets incorporated into the cell membrane phospholipids. It has been shown to increase the levels of norepinephrine and dopamine levels in CNS. It has been shown to inhibit apoptosis in neurodegenerative models, thereby potentiating neuroplasticity.</td>
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expected to benefit the most. Regression of effect after stoppage of therapy and occlusion amblyopia in younger patients (≤8 years) remain matters of concern. Further studies are therefore needed to evaluate the full efficacy and side effect profile of these agents.

REFERENCES
10. Pawar PV, Mumbare SS, Patil MS, Ramakrishnan S. Effectiveness of the addition of citicoline to patching in the treatment of amblyopia around visual maturity: a randomized controlled trial.
A 12-year-old female child presented to Pediatric Ophthalmology clinic of Dr. Shroff charity eye hospital with complaints of diminution of vision in right eye noticed since past 15 days. The parents had noted larger size of the right eyeball since birth. The patient had a prior ophthalmic consultation 15 days back and was started on antiglaucoma eye drops in the right eye. She was born of non-consanguineous marriage and the birth history was unremarkable.

On examination, her weight was 31 kgs and height was 137 cms which was slightly below average for her age. She was observed to have pinkish capillary venous malformations consistent with port wine stain on face (Figure 1). Examination of the rest of the skin revealed pigmented nevi on trunk (Figure 2) and limbs (Figure 3). Additionally she had a right sided hemihypertrophy of the lower limbs (Figure 4). There was no history of similar illness in the family.

On ocular examination, her vision in right eye was PL+ with accurate PR and left eye was 6/6. She had right eye exotropia of 20 degree (40 Prism Diopters), right sided RAPD, buphthalmos (Figure 5) and ocular melanocytosis (Figure 6). Her left eye anterior examination was within normal limit. Fundus examination showed total glaucomatous optic atrophy of right eye. The cup-disc ratio of left eye was 0.5:1. The intraocular pressure in right eye was 54mmHg and left eye was 18mmHg. Based on the above findings she was diagnosed to have congenital glaucoma in the right eye in association with phacomatosis pigmentovascularis and Klippel Trenaunay Weber syndrome. She was started on antiglaucoma medications...

Figure 1: Port wine stain on face.
Figure 2: Pigmented nevi on trunk.
Figure 3: Pigmented nevi on lower limbs.
for the right eye and Humphrey visual field was ordered for the left eye.

**DISCUSSION**

Phacomatosis Pigmentovascularis (PPV) is a neurocutaneous syndrome which was first described by Ota in 1947 as “an association of a widespread vascular nevus with an extensive pigmented nevus”. It is a rare disease which presents at birth. Approximately 222 cases have been published worldwide so far. It is found almost exclusively in Asians, Africans and Hispanics and female to male ratio is 1.34 :1.

The vascular nevus in PPV is a cutaneous vascular malformation composed of ectatic capillaries and postcapillary venules in dermis similar to Nevus flammeus or portwine stain. It is seen involving the face and/or body. Histopathological examination reveals increased vessel density but no apparent proliferation of vessels. Though it changes in colour with ageing, it usually does not disappear.

The pigmented nevi in PPV consist of dermal melanocytosis in the form of mongolian spots, nevus of ota and/or blue nevus. It may also fade with ageing but rarely disappear.

The pathogenesis of PPV is believed to be due to an abnormality in the development of melanocytic nevus cells and vasomotor neural cells derived from the neural crest. The genetic mechanism has been explained by the theory of didymosis (twin spotting) which results from somatic recombination.

It is defined as the occurrence of two genetically different clones of cells within a region of normal cells leading to mosaicism. It explains the coexistence of dermal melanocytosis and nevus flammeus.

Hasegawa and Yasuhara classified PPV into four types according to the type of pigmentary lesion associated with nevus flammeus and later a fifth type was described, characterized by cutis marmorata telangiectatica and dermal melanocytosis. Each type was further subdivided according to the presence of cutaneous-only manifestations (subtype a) or if associated with a systemic manifestations (subtype b), the majority of which are ocular, vascular, skeletal and neurological.

In 2005, Happle proposed a new simplified classification utilizing descriptive terms: Phacomatosis Cesioflamea (association of the nevus flammeus and aberrant mongolian spots or blue nevus); Phacomatosis Spilorosea (coexistence of nevus spilus and telangiectatic nevus); Phacomatosis Cesiomarmorata (association of aberrant Mongolian spots or blue nevus with cutis marmorata telangiectatica congenita) and Phacomatosis Pigmentovascularis of non-classifiable type. The most common type is type II a/b or phacomatosis cesioflamma (75% - 86%).

Various systemic associations of PPV have been reported namely oculodermal melanocytosis (ODM), Sturge Weber syndrome (SWS) or Klippel Trenaunay Weber syndrome (KTW) or a combination of these. Features of KTW syndrome that were present in our patient was nevus flammeus and hemihypertrophy of soft tissues and bone. However the patient lacked any venous or lymphatic malformations or AV shunts.

Although there can be a variety of ocular manifestation in PPV like melanosis oculi, iris mammillations, iris hamartomas, prominent vessels in sclera, chronic oedema in the cornea, pigmentary alterations in retina and pigmentary cataract, the most sight threatening one is that of glaucoma. Rarely choroidal melanomas have been reported to develop in intraocular melanocytosis. Hence a periodic detailed retinal evaluation is recommended. The incidence of glaucoma in PPV is not known. The first detailed report in ophthalmology literature was...
by Teekhasaenee C et al in 1997. The pathogenesis of glaucoma in PPV can be threefold; (1) anomaly of trabecular meshwork (2) increased pigmentation if angle and thickened iris processes and (3) increased episcleral pressure due to venous malformation.

The occurrence of both extensive melanocytosis and episcleral venous malformation of the globe strongly predispose to congenital glaucoma. Milder forms may present later in life with raised IOP. A detailed ocular examination immediately after birth, and periodically thereafter, is required to detect glaucoma and prevent its progression. Medical management of glaucoma in PPV may be unsuccessful at times warranting surgical control of the intraocular pressure.

REFERENCES
INFERIOR RECTUS LOSS AND ITS MANAGEMENT FOLLOWING TRAUMA

Dr. Prachi Agashe  
DNB, ICO (UK), FPOS

Aravind Eye Hospital, Madurai, India

Abstract: A 15 year old boy presented with complaints of diplopia since 1 year following trauma to the right eye with a handle of bicycle 1 year back. He gave history of some mass being excised after which he manifested with a large hypertropia. His clinical features were suggestive of a lost inferior rectus in the right eye with limited infraduction. He was managed with an ‘inverse Knapp procedure’ in the right eye where full tendon width of the medial and lateral recti were transposed to the inferior rectus insertion. One year later the patient was orthophoric in primary position but had diplopia in downgaze which was addressed by doing a Faden procedure of the inferior rectus and posterior two third of superior oblique tenectomy of the left eye. Two years after presentation patient was orthophoric in primary and reading position and had 4 PD of hypertropia in extreme downgaze.

A lost inferior rectus muscle is an uncommon problem reported after squint surgeries, trauma or retinal detachment surgeries. The resulting hypertropia with absence of infraduction causes diplopia in primary position and downgaze which are gazes utilized in our daily activities. We report a case of severed inferior rectus managed by inverse Knapp’s procedure on that eye followed by Faden of inferior rectus and posterior two third of superior oblique tenectomy (PTSO) of the normal eye to achieve binocular single vision in primary position and downgaze.

CASE REPORT

A 15 year old boy presented with complaints of diplopia since 1 year following penetrating trauma to the right eye by bicycle handle.

He was operated on that eye with some mass being excised, details of which were unavailable following which he developed hypertropia.

His unaided visual acuity was 6/6 in both the eyes. Anterior segment examination revealed wide palpebral aperture with lower lid retraction of the right eye with inferior conjunctival scarring.

Hirschberg test revealed hypertropia of 15 degrees of the right eye. Ocular motility examination was remarkable with severe limitation of depression in the right eye more marked in abduction with consequent overaction of the left superior oblique (Figure 1). Prism cover test measured a primary deviation of 40 prism dioptres (PD) of right hypertropia for near and distance with exotropia of 12 PD. The hypertropia measured 18 PD in upgaze and was more than 50 PD in downgaze. The hypertropia measured equal on either tilt. Forced duction test (FDT) revealed no restriction to passive depression and the forced generation test demonstrated no tug on active infrauction.

CT scan showed focal atrophy of the inferior rectus near the insertion while the distal portion was thinned out.

The patient underwent surgical correction of squint. Surgical exploration showed fat infiltrated near the inferior rectus insertion and the muscle was not found. With the FDT for superior rectus being negative an ‘inverse Knapp’s procedure’ was performed with full tendons of the medial and lateral recti being transposed near the inferior rectus insertion along the spiral of Tillaux. Three months post operatively the hypertropia significantly reduced and one year post operatively the patient was orthophoric in primary position. The patient had diplopia in down gaze with limitation of depression in the right eye for which the patient underwent Faden procedure on the inferior rectus of the left eye. A 5-0 non absorbable polyester suture was passed 14 mm from the insertion along the medial and lateral border of the muscle fixing it to the sclera. This was accompanied with PTSO to further limit depression of the normal eye.

Two years post operatively the patient was orthophoric for near and distance as well as in reading position but had a small hypertropia of 4 PD in extreme downgaze which the patient was able to fuse with prisms (Figure 2).

DISCUSSION

Extraocular muscle transection or detachment following surgery or trauma is uncommon. Various strategies have been
described in the literature to retrieve lost muscles or perform corrective surgery to deal with the consecutive squint that results.

Traumatic rupture of an extraocular muscle is most commonly found to occur in the medial rectus followed by inferior rectus, superior rectus and lateral rectus in that order. The postulated reason is the close proximity of the medial and inferior rectus to the corneoscleral limbus. Paysse et al described surgical management of two cases of lost inferior rectus using modified Jensen procedure. In one case the inferior rectus was lost during recession and the second case was similar to ours resulting in lost inferior rectus after a dog bite injury. The author reported overcorrection in both cases which probably resulted as the endpoint of their procedure was a mild restriction to elevation while in our case there was no restriction to elevation and we opted out of doing a posterior fixation suture. The diplopia present postoperatively was treated with prisms in one patient and contralateral superior rectus recession in the other patient. In our patient though the infrafusion improved after the transposition procedure, diplopia persisted in downgaze which was managed by Faden procedure of the inferior rectus in the other eye along with PTSO with the aim of limiting infrafusion in the good eye to improve the alignment in downgaze.

Anteropositioning of the inferior oblique is another technique described to correct hypertropia with reduced chances of anterior segment ischemia. However, we chose to shift the vectors towards the weakened muscle by transposing the horizontal recti inferiorly. Also, ours was a young patient with no predisposing factors for anterior segment ischemia and the trauma being old we opted for a full tendon transposition. Flavarjani and Asadi have described anteriorization of the inferior oblique with inferior transposition of the medial rectus muscle in a six year old boy. This child was operated for congenital esotropia and was undergoing surgery for bilateral inferior oblique overaction when the inferior rectus was inadvertently cut and was irretrievable. They reported 8 PD of overcorrection in primary position with no infrafusion deficit.

Plager and Parks presented a series of 25 cases of lost muscles. They reported good results with transposition surgeries in eyes where muscles could not be retrieved with two cases having slight overcorrection. However, our patient did not have overcorrection.

Faden procedure is most commonly utilized for restricting the action of the muscle in the direction of gaze without influencing the primary position deviation. It is most commonly performed on the medial recti; found to be effective mainly in cases of accommodative esotropia with high AC/A ratio. It has also been reported in the management of Grave’s disease, third nerve palsy, sixth nerve palsy, dissociated vertical deviation and orbital trauma. Using the same principle, Faden procedure was done on the inferior rectus of the normal eye along with PTSO with the aim of limiting downgaze in that eye and improving alignment in downgaze (Figure 3,4).

**CONCLUSION**

This report highlights the successful management of a difficult ocular motility problem using a tailor made approach.
REFERENCES


Correspondence to:
Dr. Prachi Agashe
Aravind Eye Hospital, Madurai, India
Bilateral acquired superior oblique palsy with left tonic ocular tilt reaction following resection of a left posterior fossa mass; an esoteric presentation

Dr. Pramod Kumar Pandey MD, Dr. Monika Yadav MBBS, Dr. Divya Kishore MS, Dr. Ipsita Muni MBBS, Dr. Kirti Singh MD

Guru Nanak Eye Centre, Maharaja Ranjit Singh Marg, New Delhi, India

Bilateral acquired superior oblique palsies (SOPs) may be quite distressing to the patient due to torsional and homonymous/heteronymous diplopia emanating from subjective extorsion and V pattern with esotropia in down and extotropia in upgaze respectively. A vertical component to diplopia is often present due to asymmetrical involvement and attendant muscle sequelae. Trauma is the most common etiology, other less common etiologies include hydrocephalous, iatrogenic and tumors. Fourth nerve is particularly prone for damage as it is infratentorial, crosses to the other side in anterior medullary velum, exiting just inferior to inferior colliculus and travelling around the mid brain in the perimesencephalic cistern then coursing between posterior cerebral and superior cerebellar arteries to penetrate the dura below the posterior clinoid to enter the cavernous sinus.

Tonic / phasic ocular tilt reactions (skew deviation, conjugate torsion and head tilt to same side) can result from damage to otolithic pathways anywhere from the vestibules to the vestibular cortex in the parietal lobe, frequently arising from posterior fossa pathology. The medial longitudinal fasciculus (MLF) carrying otolithic pathways is lying ventral and lateral to 4th nerve nuclei in the midbrain tegmentum just ventral to periaqueductal gray, both structures can be involved simultaneously but such lesions have other associated signs and symptoms. Fascicular lesions of 4th nerve may sport associated Horner’s syndrome on the opposite side due to involvement of the central tegmental tract. Tonic ocular tilt reactions (OTR) arising from damage to contralateral otolithic pathways corresponding to anterior semi-circular canal may simulate unilateral SOPs and create diagnostic dilemmas. A bilateral SOP following a neurosurgical procedure with coexisting tonic ocular tilt reaction has not been reported. We report such an esoteric presentation following removal of a posterior fossa mass that was subsequently confirmed as pilocytic astrocytoma.

CASE REPORT

A 20 year old girl was operated for a posterior fossa mass in the left cerebello-pontine angle. Post-operative course was uneventful other than evolution of horizontal and torsional double vision and inward deviation of either eye. There were no other visual symptoms. She was referred for ophthalmological evaluation from a tertiary care neurosurgery treating facility.

On examination she was well oriented in time and space with no major sensory or motor involvement. Her unaided visual acuity was 6/9 OU. She adopted a chin down position due to OTR whereas extorsion in left eye got exaggerated due to SOP. As 4th nerves are infra tentorial with crossing in the anterior medullary velum and long course around the midbrain in the perimesencephalic cistern and between posterior cerebral and superior cerebellar arteries, they are likely to get damaged by trauma, tumor or neurosurgical manipulations in the posterior fossa. Bilateral involvement may be symmetrical with no PP HT or asymmetrical with a PP HT. Torsional symptoms, V pattern with esotropia in PP and down gaze and extotropia in upgaze are frequently encountered. Bilateral extension of >10 degrees is often present.

In the present case diagnosis of Bilateral SOP is unequivocal, only missing link is bilateral extorsion. Conjugate ocular torsion however with intorsion of right eye and extorsion of left eye betrays the presence of a coexisting left OTR. The extorsion in right eye due to SOP went overboard and ended up in intorsion due to OTR whereas extorsion in left eye got exaggerated due to coexisting SOP and OTR, head position dependent changes in torsion further reinforce the presence of OTR.

Discussion

As 4th nerves are infra tentorial with crossing in the anterior medullary velum and long course around the midbrain in the perimesencephalic cistern and between posterior cerebral and superior cerebellar arteries, they are likely to get damaged by trauma, tumor or neurosurgical manipulations in the posterior fossa. Bilateral involvement may be symmetrical with no PP HT or asymmetrical with a PP HT. Torsional symptoms, V pattern with esotropia in PP and down gaze and extotropia in upgaze are frequently encountered. Bilateral extension of >10 degrees is often present.

In the present case diagnosis of Bilateral SOP is unequivocal, only missing link is bilateral extorsion. Conjugate ocular torsion however with intorsion of right eye and extorsion of left eye betrays the presence of a coexisting left OTR. The extorsion in right eye due to SOP went overboard and ended up in intorsion due to OTR whereas extorsion in left eye got exaggerated due to coexisting SOP and OTR, head position dependent changes in torsion further reinforce the presence of OTR.
Bilateral SOP in the present case likely resulted from neurosurgical manipulations in the infratentorial compartment whereas left OTR occurred due to damage to otolithic pathways at the level of the pons before crossing of fibers to opposite side. Acute acquired comitant esotropia infrequently seen with posterior fossa tumors should also be kept in mind, however findings in present case unequivocally establish esotropia being caused by bilateral SOP thus militating against such a diagnosis. Sixth nerve palsy was ruled out as there is no abduction deficit and/or horizontal incomitance.

Evaluation of ocular torsion and head position dependent changes in ocular torsion and vertical alignment should be included as 4th and 5th step in the evaluation of ocular motility disorders.

A SOP should be treated as bilateral till bilaterality is ruled out as upto 20% of bilateral SOPs can be masked and present as unilateral SOP. OTR due to involvement of otolithic pathways corresponding to contralateral anterior semicircular canal may simulate unilateral SOP including the 3 step test, only marker of an OTR may be conjugate torsion. OTR and bilateral SOP may coexist and create diagnostic waterloos. A low threshold of suspicion is in order as such investigations may have very low yield in SOPs.

REFERENCES

Q1. A 55-year-old diabetic develops a partial 3rd-nerve palsy. His eyelid elevates when he looks downward. What is his most likely diagnosis?
   a. Ischemic mononeuropathy
   b. Elevated intracranial pressure
   c. Midbrain stroke
   d. Compressive lesion.

Q2. The far point of the nonaccommodated myopic eye?
   a) Corresponds to fovea
   b) Is behind the eye
   c) Cannot be moved by placing lens in front of eye
   d) Is nearer than the point of focus.

Q3. A one-month-old baby is diagnosed with unilateral anterior polar cataract that is approximately 1.5 mm in diameter. The most appropriate initial management is?
   a) Close observation
   b) Lensectomy
   c) Lensectomy with intraocular lens implant
   d) Chronic dilation.

Q4. The percentage of primary congenital glaucoma that is now known to have a definite genetic component is?
   a 5%
   b 10%
   c 30%
   d 75%.

Q5. Fetal alcohol syndrome includes all of the following except?
   a) Small nose and narrow forehead
   b) Epicanthal fold
   c) Peter’s anomaly
   d) Microphthalmos.

Q6. Identify the syndrome: She can’t look on either side?

Q7. 10 year old male with RE squint since 6 months with BCVA of 6/60. OCT as shown. Diagnosis:
Q8. Identify the condition?

Q9. Name a topical drug used for the treatment of nystagmus?

Q10. Which drug besides atropine is studied in the ATS as a pharmacological treatment for amblyopia?

Compiled by:

Dr. Savleen Kaur

Senior Research Associate, PGIMER, Chandigarh, India

DOS Times Quiz Rules

1. DOS Times Quiz will now feature as 5 Episodes (Episode 1: July-August, Episode 2: September – October, Episode 3: November – December, Episode 4: January – February, Episode 5: March – April). Entries will have to be emailed before the last date mentioned in the contest questions form. Late entries will not be entertained.

2. Please email (as scanned PDF Only) completed responses for the quiz along with details of the contestant filled in and signed to dostimes10@gmail.com (with cc to dosrecords@gmail.com) or mail to DOS Times Quiz, Dr. Subhash Dadeya, Room No. 114, 1st Floor, OPD Block, Guru Nanak Eye Centre, Maharaja Ranjit Singh Marg, New Delhi.

3. Nonmembers may also send in their entries but will be required to send along with their completed entries, the completed membership application (with the required documents) to enroll as member. Failing this their entries into the contest will not be considered.

4. Contestants are requested to attempt all the 5 episodes of the Quiz contest and send in their applications within the date specified. No entries will be entertained after the last date. The scores of each contestant for all 5 episodes together will be compiled at the end of episode 5 and the winner will be announced in the DOS Annual Conference in April 2019. In the event of more than one winning contestants, a draw of lots will decide the winner. Winner of each episode will also be published in the next episode along with the previous episode answers.

5. Please write to dostimes10@gmail.com or dosrecords@gmail.com for further clarifications if any.

Q. No. Completed Responses for DOS Times Quiz: Episode 1

1. ____________________________ 6. ____________________________
2. ____________________________ 7. ____________________________
3. ____________________________ 8. ____________________________
4. ____________________________ 9. ____________________________
5. ____________________________ 10. ____________________________

Contestant Details

Name: ____________________________ Degree: ____________________________

Designation: ____________________________ Address: ____________________________

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Mobile No: ____________________________ DOS Membership no: ____________________________

Email ID: ____________________________ Signature: ____________________________
DOS CROSSWORD

Episode-1

Correspondence to:
Dr. Savleen Kaur, Senior Research Associate
Advanced Eye Centre, PGIMER, Chandigarh, India

Fill the boxes below with the most appropriate answers by using the hints below

ACROSS
1. Drug Known to Halt Progression of Myopia
2. Largest Trial of IOL Implantation in Infants
3. Inheritance of PHPV
4. Muscle Hook
5. Gene Responsible Congenital for Glaucoma
6. Syndrome Mostly Affecting the Left Eye in Females

DOWN
7. Surgery for Strabismus Fixus
8. Imaging the Fundus of Children by this Instrument/Machine
9. Longest Tendon
10. Unilateral Retinal Disease in Young Boys

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Address for all correspondence
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Secretary - Delhi Ophthalmological Society
Room No 114, 1st Floor, OPD Block,
Guru Nanak Eye Centre, Maharaja Ranjit Singh Marg,
New Delhi - 110002
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SCOPE OF THE JOURNAL
DOS Times covers clinical, experimental and basic science research studies related to medical, ethical and social issues in field of ophthalmology. Articles with clinical interest and implications are given preference.

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A manuscript is reviewed for possible publication with the understanding that it is being submitted to DOS times alone at that point in time and has not been published anywhere, simultaneously submitted, or already accepted for publication elsewhere. Initial screening by the editorial desk assesses the formatting, topicality and importance of the subject, the clarity of presentation, and relevance to the target audience of the journal. Acknowledgement of receipt of all manuscripts will be sent to the corresponding author, once the editorial desk reviews the manuscript for conforming to the requirements of the journal. Manuscripts that are found suitable for publication are sent to two or more expert reviewers for Peer Review through an online reviewer system. The identities of reviewers and authors are kept confidential. Authors and Reviewers are required to declare potential conflicts of interests/ financial interests. The comments and suggestions (acceptance/ rejection/ amendments in manuscript) received from reviewers are conveyed to the corresponding author. Corresponding authors are requested to submit the revised manuscript along with one highlighted copy with revisions highlighted. The final decision on acceptance of the manuscript for publication lies with the Editor-in-chief. This process is repeated till reviewers and editors are satisfied with the manuscript. Manuscripts accepted for publication are copy edited for grammar, punctuation, print style, and format.

CONFLICTS OF INTEREST
All authors must disclose all conflicts of interest they may have with publication of the manuscript or an institution or product that is mentioned in the manuscript and/or is important to the outcome of the study presented.

MANUSCRIPT PREPARATION
Manuscripts under the following subheadings may be submitted:

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<th>Type</th>
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<th>Abstract</th>
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<td>Original article</td>
<td>2500</td>
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<td>Structured 150-200 words</td>
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<td>These include randomized clinical trials, prospective and retrospective observational and interventional studies, questionnaire-based studies, qualitative data based studies, quality of life studies etc. excluding references, abstract, figures and tables.</td>
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<td>Includes comprehensive and systematic literature review and meta-analysis. Review articles can be commissioned either by editorial invitation or by submitted proposals.</td>
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<td>Summary of latest in clinical research, instrumentation and web resources in ophthalmology.</td>
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<tr>
<td>Techniques</td>
<td>1500</td>
<td>30</td>
<td>Unstructured 100 words</td>
</tr>
<tr>
<td>Novel surgical techniques or instrumentation that have the potential to reduce surgical complexity and/or enhance outcomes.</td>
<td></td>
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<tr>
<td>Case reports</td>
<td>1000</td>
<td>10</td>
<td>Unstructured 100 words</td>
</tr>
<tr>
<td>Interesting cases with immense clinical significance / rare case reports</td>
<td></td>
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<tr>
<td>Subheadings: Introduction, case and discussion</td>
<td></td>
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<tr>
<td>Photo-essay/snap shots</td>
<td>500</td>
<td>10</td>
<td>Unstructured summary in 100 words</td>
</tr>
<tr>
<td>Reports of unusual/uncommon clinical case scenarios with good photographic documentation</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>Subheadings: Introduction, case and comment</td>
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</tbody>
</table>

All manuscripts should have the following:
1. Title of the manuscript
2. Type of manuscript
3. Name(s) and surnames of authors with highest academic degree
4. Author affiliations: Department, Institution, and contact details
5. Corresponding author: name, designation and credentials, address, phone, fax, email and digital passport size photograph
6. Information about patient consent and approval for photographs that disclose the identity of the patient.
7. Please submit as word file with embedded figures
8. Figure legend at the bottom of figure
9. Tables with numbering and heading at the top embedded in the text file
10. References as superscripts without brackets numbered consecutively in text. References should be written in standard international format as in Pubmed: Authors. Title of citation quoted. Name of Journal Year of publication; Volume number, Page numbers.
11. Abbreviations spelled out at the first appearance in the text.
12. Generic drug names are to be used in text, tables, and figures. Suppliers of drugs, equipment, and other brand-name material are to be credited in parentheses (company, name, city, state, country).
DOS Travel Fellowship for Partial Financial Assistance to Attend Conferences

Applications are invited for DOS Fellowship for partial financial assistance to attend conference(s).

**Conferences**
- **International**: Eight fellowships per year.
  - Maximum of Rs. 50,000/- per fellowship will be sanctioned

Partial travel fellowship to attend AIOC: Five fellowships per year

The last date of DOS Fellowship for Partial Financial Assistance to Attend Conference(s) for receiving application is 30th September for National Conference.

Maximum of Rs. 10,000/- per fellowship will be awarded out to five one to the winner of the DOS best paper (A C Aggarwal Trophy) for travel to present the paper in the forthcoming AIOC.

**Eligibility**
- DOS Life Members (Delhi Members only)
- 75 or More DCRS Points
- Accepted paper for oral presentation, poster, video or instruction course or invited guest speakers

**Time since last DOS Fellowship**
Preference will be given to member who has not attended conference in last three years. However if no applicant is found suitable the fellowship money will be passed on to next year. Members who has availed DOS fellowship once will not be eligible for next fellowship for a minimum period of three years.

**Authorship**
The fellowship will be given only to presenting author. Presenting author has to obtain certificate from all other co-authors that they are not attending the said conference or not applying for grant for the same conference. (Preference will be given to author where other authors are not attending the same conference). If there is repeatability of same author group in that case preference will be given to new author or new group of authors. Preference will also be given to presenter who is attending the conference for the first time.

**Quality of Paper**
The applicant has to submit abstract along with full text to the DOS Fellowship Committee. The committee will review the paper for its scientific and academic standard. The paper should be certified by the head of the department / institution, that the work has been carried out in the institution. In case of individual practitioner he or she should mention the place of study and give undertaking that work is genuine for invited guest speakers & instruction courses only acceptance letter is required. The fellowship committee while scrutinizing the paper may seek further clarification from the applicant before satisfying itself about the quality and authenticity of the paper. Only Single best paper has to be submitted by the applicant for review (6 copies). Quality of the paper will carry 50% weightage while deciding the final points.

**Poster and Video**
The applicant will need to submit poster and video for review.

**Credit to DOS**
The presenter will acknowledge DOS partial financial assistance in the abstract book / proceedings.
The author will present his or her paper in the immediate next DOS conference and it will be published in DJO / DOS Times.

**Points Awarded**

<table>
<thead>
<tr>
<th>1) Age of the Applicant</th>
<th>Points</th>
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</thead>
<tbody>
<tr>
<td>a) ≤ 35 years</td>
<td>10</td>
</tr>
<tr>
<td>b) 36 to 45 years</td>
<td>07</td>
</tr>
<tr>
<td>c) 45 years plus</td>
<td>05</td>
</tr>
</tbody>
</table>
2) Type of Presentation
   a) Instructor/Co-instructor of Course 12
   b) Free Paper (Oral) / Video 07
   c) Poster 05

3) Institutional Affiliation
   a) Academic Institution 15
   b) Private Practitioner 20

4) The points awarded for DCRS rating in the immediate past year
   a) > 150 10
   b) 75 – 150 5
   c) < 75 Not Eligible

Documents
   • Proof for age. Date of Birth Certificate
   • Original / attested copy of letter of acceptance of paper for oral presentation / video / poster or instruction course / invited talks.
   • Details of announcement of the conference
   • Details of both International & National Conferences attended in previous three years.
   • Copy of letter from other national or international agency / agencies committing to bear partial cost of conference if any.
   • Original air travel boarding passes and photocopy of the attendance certificate of the conference.
   • Fellowship Money will be reimbursed only after submission of all the required documents and verified by the committee.
   • Undertaking from the applicant stating that above given information’s are true.
   • If found guilty the candidate is liable to be barred for future fellowships.

Application should reach Secretary's office and should be addressed to Chairman Travel Grant Fellowship Committee before February 20, June 30, September 30 and December 30 for International Conference and National Conference. The committee will meet thrice in a year in the month of August, November and February within 2 weeks of last date of receipt of applications. The committee will reply within four week of last date of submission in yes/no to the applicant. No fellowship will be given retrospectively.

Dr Subhash Dadeya
Secretary,

Delhi Ophthalmological Society
Room No. 114, OPD Block, 1st Floor,
Maharaja Ranjit Singh Marg,
Guru Nanak Eye Centre,
New Delhi – 110 002
Ph : +91-11-23210810
Email: dosrecords@gmail.com
REVISION OF DOS CREDIT RATING SYSTEM & CALCULATIONS FOR THE AWARDS

REVISIONS of DCRS points:
i. Clinical Case Presentations & Clinical Talk will be evaluated for a total of 100 marks each
ii. Overall meeting arrangement marking will be given out of 20 by outside delegates instead of grading system as was practiced earlier.

BEST CLINICAL CASE PRESENTATION / BEST CLINICAL TALK / BEST CASE PRESENTATION

Average of Marks awarded by all DOS member Delegates attending the monthly clinical meeting

MOST POPULAR DOS CLINICAL MONTHLY MEETING (MINOO SHROFF TROPHY)

The trophy for most popular centre for the DOS Monthly Clinical Meeting will be awarded to the institute with the maximum attendance

BEST DOS MONTHLY CLINICAL MEETING (BODHRAJ TROPHY)

Calculation of overall meeting assessment score

Please Note: For calculation of overall meeting assessment score, only marks awarded by the external DOS member delegates is taken into consideration

Step 1:
❖ The average of the two case presentations & clinical talk out of 300 marks will be calculated and reduced to 30 marks

Step 2:
❖ To this, will be added the overall meeting arrangement marks (for a total of 20). The total will now add up to 50 marks to compute total marking score by each delegate

Step 3:
❖ The average of this marking score (out of a total of 50 marks) of all external DOS member delegates will be calculated. To this will be added the attendance score of outside delegates the institution having maximum numbers of outside delegates will be given 50 marks, the other institutes will be given percentile score proportionately to yield the FINAL SCORE for each centre hosting the DOS Monthly Meeting, based on which for BodhRaj Trophy of best monthly meeting will be awarded.

POINTS TO BE NOTED:
➢ DCRS marking sheets will have details of time in of all delegates & marking sheets according to time of entry will be given to the delegate upon signing in the attendance register
➢ All delegates will be required to hand in their completed DCRS sheets at the conclusion of the meeting, which will be counted by DOS staff, checked and signed by head of the institute/representative holding the monthly meeting and DOS Secretary (/President/DOS executive committee representative) & subsequently sealed in their presence.
➢ All centres are encouraged to provide their meeting programme details 4 weeks ahead of their date of programme.
➢ All presenters of case reports and clinical talk are required to submit their presentations to the DOS SECRETARIAT in two weeks time following the date of the presentation by email (dostimes10@gmail.com) for publication in the monthly meeting korner section of the forthcoming DOS TIMES issues.
➢ PLEASE NOTE THAT ALL DOS monthly meetings SHOULD commence by 11.0 am sharp. The number of speakers in the Clinical symposium should not exceed 3

VIOLATION of the recommendations for DOS Monthly meetings will
Result in PENALIZATION of the final DCRS score of the centre

Hence forth only one institution will be dropped from calendar of monthly meeting based on DCRS rating.
Review Subcommittee for their efforts in reforming the guidelines for the DOS Monthly Clinical Meetings

Dr Sudarshan Khokhar
President – DOS

Dr Subhash Dadeya
General Secretary – DOS
Delhi Ophthalmological Society

(LIFE MEMBERSHIP FORM)

Name (In Block Letters)_____________________________________________________________________________________________

S/D/W/o_________________________________________ Date of Birth________________________

Qualifications____________________________________ Registration No.________________________

Sub Speciality (if any)______________________________________________________________________________________________

ADDRESS
____________________________________________________________________________________________________________
____________________________________________________________________________________________________________
____________________________________________________________________________________________________________

Phone _______________________

Email ____________________________ Mobile No. ______________________________

Proposed by
Dr. ___________________________________________________________________________ Membership No. ________ Signature _________________________

Seconded by
Dr. ___________________________________________________________________________ Membership No. ________ Signature _________________________

[Must submit a photocopy of the Address proof, Pan Card, MBBS/MD/DO, State Medical Council / MCI Certificate for our records.]

Declaration: I hereby declare that the above details are correct. I wish to be Life member. I have carefully read the instructions overleaf. I shall abide by the Rules, Regulation & Bye-Laws of the Society as in force and any subsequent amendment(s) made from time to time

(Life membership fee Rs. 5600/- payable by DD for outstation members. Local Cheques acceptable, payable to Delhi Ophthalmological Society)

Please find enclosed Rs.___________ in words ____________________________________________________ by Cash _________________

Cheque/DD No.________________________________ Dated_______________ Drawn on____________________________

Three specimen signatures for I.D. Card.

FOR OFFICIAL USE ONLY

Dr._____________________________________________________________ has been admitted as Life Member of
the Delhi Ophthalmological Society by the General Body in their meeting held on______________________________

His/her membership No. is _______________. Fee received by Cash/Cheque/DD No.________________ dated________
drawn on _________________________________.

(Secretary DOS)
INSTRUCTIONS

1. The Society reserve all rights to accept or reject the application.
2. No reasons shall be given for any application rejected by the Society.
3. Every new member is entitled to receive the Society’s Bulletin (DOS Times) and quarterly Journal DJO (Delhi Journal of Ophthalmology) of the Society free.
4. Every new member will initially be admitted provisionally and shall be deemed to have become a full member only after formal ratification by the General Body and issue of Ratification order by the Society. Only then he or she will be eligible to vote, or apply for any Fellowship / Award, propose or contest for any election of the Society.
5. To be proposed and seconded by Ratified Life Member only. No application form will be accepted unless it is complete in all respects. Proposed and Seconded by existing Member of the Delhi Ophthalmological Society.
6. Photo ID Card will be issued only after the membership is ratified by the General Body.
7. Resident doctors must submit Delhi address proof with validity after completion of their residency failing which they will be treated as non-Delhi members.
8. Documents to be attached with application form:

   **Copy of Address Proof (Mandatory)**
   Passport/Licence/Voters Identity Card/Ration Card/ Electricity Bill/MTNL (Landline) Telephone Bill
   (Delhi Life Member should either reside or practice in Delhi.

1. **Copy of Degree (MBBS / MD / DNB)**
2. **Copy of Registration Certificate (Medical Council of India or State Medical Council)**
3. **Copy of PAN Card**
4. **One Stamp size Coloured Photograph to be pasted on the Application Form and one stamp size coloured photograph to be attached with form for issue of Laminated Photo Identity Card (to be issued only after the Membership ratification by GBM).**

9. Membership Fee

   **There is life membership on one Time Payment of Rs. 5,600/- only.**
   1. Life membership fee Rs. 5,000/- (This money will be part of corpus of Society)
   2. Admission fee Rs. 600/-

   The application form should be complete in all respects and accompanied by a Demand Draft of Rs. 5,600/- in favour of “Delhi Ophthalmological Society” payable at New Delhi should be sent:

   **Dr. Subhash C. Dadeya**
   Secretary
   **Delhi Ophthalmological Society**
   Room No. 114, 1st Floor, OPD Block, Guru Nanak Eye Centre, Maharaja Ranjit Singh Marg, New Delhi-110002

10. For update address for sending application, please visit website www: dosonline.org
## Associated Syndromes

### Mass
- Term or preterm: ROP
- Triad of leucocoria, lens opacification, and ciliary body

### Associated Microcornea, Microphthalmos, Buphthalmos
- May have under it appear normal, does not grow

### Rare Tumors
- Medulloepithelioma: Arises from ciliary body; clinical
- Incontinentia Pigmenti etc.
- FEVR

### Important Points in History and Examination

<table>
<thead>
<tr>
<th>Provisional Diagnosis and Management Plan</th>
</tr>
</thead>
<tbody>
<tr>
<td>Documentation</td>
</tr>
<tr>
<td>Photography</td>
</tr>
<tr>
<td>Anterior Segment Clinical photography, Retcam for fundus</td>
</tr>
</tbody>
</table>

### Investigations

- CE/ MRI when warranted; avoid CT scan in children <2 years
- Characteristic vitritis, ELISA for toxocara excretory secretory antigen

### Characteristic Features

<table>
<thead>
<tr>
<th>Entity</th>
<th>Presentation</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Retinoblastoma</td>
<td>B/L 12 months; U/L 24 months</td>
<td>USG: mass with calcification</td>
</tr>
<tr>
<td>Medulloepithelioma</td>
<td>UBM to see multicystic irregular internal reflectivity</td>
<td></td>
</tr>
<tr>
<td>Astrocytic hamartoma</td>
<td>Flat tumor, no RD, vessels around</td>
<td></td>
</tr>
</tbody>
</table>

### Common Issues

- Peripheral neovascularization, macular heterotopia, exudation
- Peripheral retina. Organized vitreous membranes, traction on the retina
- Retinal detachment, macular exudation
- Vitreoretinopathy

### Key Points in History

- History of prematurity, oxygen therapy
- Clinical evaluation
- Demographic
- Detailed dilated examination; rule out associated syndromes; surgical management
- Peculiarities in IOL power calculation and role of posterior capsulotomy and anterior vitrectomy

### Complications

<table>
<thead>
<tr>
<th>Cataract U/L or B/L; can vary widely in morphology; important to</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mention lesion location according to vascular bifurcation</td>
</tr>
<tr>
<td>Anterior Segment: Iris, NVI, Pupil margin</td>
</tr>
<tr>
<td>Posterior Segment: Disk and macula</td>
</tr>
<tr>
<td>Lens: Clarity, diameters</td>
</tr>
<tr>
<td>Cornea: Clarity, diameters</td>
</tr>
<tr>
<td>Flow Chart for Evaluation:</td>
</tr>
<tr>
<td>Leucocoria: White Pupillary Reflex</td>
</tr>
</tbody>
</table>

### Associated Syndromes

- Associated with Tuberous Sclerosis and rarely Neurofibromatosis
- Others: Endogenous endophthalmitis, Retinal detachment due to any cause, Vitreous hemorrhage long standing

### Pseudoleucocoria

- Over observant parents may report "squinting of eyes" or abnormal looking eyes on flash photography; important to examine thoroughly and reassure in absence of any pathology

### Astrocytic hamartoma

- Glial tumors from RNFL, associated symptoms

### Malignant Tumors

- Most common malignant intraocular tumor of childhood
- Rhabdomyosarcoma: Involves uveal tract, extraocular muscles, and adjacent orbit; surgical intervention

### Prognosis

- Early detection and treatment are crucial for a better prognosis.