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Dear DOS Members,

New beginnings can sometimes be daunting, everything is alien and yet fraught with possibility. Interacting with new people, taking on new responsibilities of an organisation as dynamic as DOS, could have been overwhelming. Instead, thanks to the absolutely rock-solid support of my mentors, friends, students and readers like you, I am looking forward to the colossal task.

The past editorial board added many feathers to the cap of DOS with an excellent amalgam of scientific papers, discussions and reports. We begin as a new team, but strive towards academic excellence; the ultimate goal of the DOS. We attempt to extend and spearhead this educational brilliance through the various issues of DOS times in the coming months. We promise to deliver scientific discussions which increase our understanding, knowledge and management of ophthalmic problems.

We are trying to transform DOS Times to make it more standardised, unique, resourceful and relevant. We hope that you will soon begin to see a change in our perspective and accomplishments. We plan to change some of our patterns and guidelines. We wish to make the articles more informative and recent. We also endeavour to offer you access to original research from the various subspecialties in ophthalmology. Having spent most of my career studying and mastering the art of childhood eye diseases, I venture to bring before you the first ‘focus’ issue of the DOS times – a special edition in paediatric ophthalmology. Children are not small adults. Although this subspecialty is as vast as ophthalmology itself, there are only a handful of dedicated paediatric ophthalmologists in India today. Unhealthy children are a burden in terms of budget and energy to the society. Various screening programs can prevent many causes of reversible blindness which are low in cost as compared to other sub-specialities. Hence, we dedicate this issue to paediatric ophthalmology. This issue is an assortment of cases, reviews and reports from across the world in paediatric ophthalmology. We expand the spectrum of this domain to include excellent write ups from the stalwarts of paediatric ophthalmology. We welcome any pointers and feedbacks to make DOS times outshine and progress even more.

I also take this opportunity to welcome you all to the joint DOS-SPOSI International Congress on 9th and 10th December at India Habitat Centre, Delhi. The congress will focus on the global perspectives in strabismus and paediatric ophthalmology with symposiums by AAPOS, ESA, ISA, APSPOS, IPOSC and IOA, AOIS, OPAI, APSOOP, ACOIN, QOIC. I am sure it will be an academic fiesta with an opportunity to interact with the glitterati in paediatric ophthalmology. The preparations for I-DOS-QOIC Joint International Conference from 4-7 January, 2018 at Dubai are in full swing. On behalf of the Executive, it gives us great pleasure to invite you and your family to participate in this wonderful academic and fun filled International Conference.

We are a union of thousands of ophthalmologists, who strive for better clinical outcomes. Yet we are divided by choices, subspecialties, regions and personal agendas. Let us all aim to work towards our objective. To promote healthy teaching, surgical skill transfer, ethical conduct and state of the art cutting edge information should be our purpose and ambition. I have imagined and re-imagined what my ideas of DOS are. I had a dream and vision for DOS. I will ensure that DOS grows from strength to strength and scales great heights as an organization committed to academic excellence, fulfill everything we were founded for; reform our existing norms; provide you with the best of best.

I ask for your help and support, as always, because it is only with your cooperation that I have managed to achieve whatever little that I have. The affection and regard that I have received from all of you gives me the courage to stand up for our collective rights and responsibilities. I look forward to our continued association.

Wishing you all a pleasant reading.

Warm regards

Dr. (Prof.) Subhash C. Dadeya
Secretary - Delhi Ophthalmological Society
Room No 205, 2nd Floor, OPD Block,
Guru Nanak Eye Centre, Maharaja Ranjit Singh Marg,
New Delhi - 110002
Email: dadeyassi@gmail.com
Mobile: 9968604336, 9810575999
Joint DOS- SPOSI
INTERNATIONAL CONGRESS
9th & 10th Dec., 2017
Venue: India Habitat Centre, Delhi

CONFERENCE HIGHLIGHTS
- Symposia of AAPOS, ESA, APSPOS, ISA, IPOS, IOA, AIOS, OPAI, APSOOP, ACOIN, QOIC
- Live Surgery
- Scientific deliberations in Cataract & Refractive Surgery, Retina, Cornea, Glaucoma, Squint & Neuro-ophthalmology and Oculoplasty
- Breakfast with experts
- Wet Labs
- CME Credit hours
- Debates
- Poster Sessions
- E-Videos
- Early Bird Prizes
- An exciting Fellowship Dinner
- Quiz
Invitation

Dear Colleagues,

On behalf of the organizing committee, it is our pleasure to invite you to our Joint SPOS-DOS International Congress to be held on 9th and 10th, 2017.

This year the theme of the conference is global perspectives in Strabismus and Pediatric Ophthalmology. The scientific sessions promise a weighted amalgam of innovative research and timely lectures on a broad spectrum of topics relevant to your ophthalmic practice.

With the support of the international symposium by AAPCO, ISA, ISA, IDA, ARPSIS, IPOS, AOCS, COMA, APSOAR, ACCON, etc., the conference provides an unparalleled avenue for continuing medical education and updates on recent diagnostic and therapeutic modalities, in all facets of current ophthalmology practice. It is also a unique opportunity to interact with the world-renowned experts in the field of Strabismus and Paediatric Ophthalmology, for roundtable discussions and hands-on teaching sessions, which will cater to the needs of the audience from all over the world.

The Delhi Ophthalmic Society, with over 6700 members and SPOS with over 550 members across India, are committed to furthering the cause of continuing medical education and promoting camaraderie and fellowship amongst ophthalmologists worldwide. The DOS conference is a must-attend event, with numerous academic sessions, which attract faculty, trade exhibitors and delegates from all over the world.

As you will venture out of the confines of the city and the world of ophthalmology, the vibrant Delhi wintertime, along with its legendary hospitality and bustling energy, will enthral you. As always, there will be scientific sessions, workshops, the cutting-edge photobiography, video presentations, and a variety of live surgery sessions from the operating rooms of leading ophthalmic institutions of Delhi. After the academic extravaganza, the conference will culminate in a captivating cultural program and a 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 say about technology advancements and their impact on Ophthalmology. We also would love to hear your thoughts and opinions on this direction.

We look forward to your esteemed presence at the Joint Strabismus and Paediatric Ophthalmology Conference of DOS and SPSI.

Respectfully Yours,

Prof. Kamlesh
President, DOS

Prof. Subhash C. Dadeda
Secretary, DOS

REGISTRATION FEE

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IMPORTANT NOTES

*Inclusive tax. **Proof of residency required from ICC along with the registration form of the conference. ***Registration for Special/Child must be done separately for each person.

For more information, please visit our website at www.dosonline.org

Address for all Correspondence:

Dr. Subhash C. Dadeda
Room No 206, 2nd Floor, OPD Block, Guru Nanak Eye Centre, Mahesraja Ranjit Singh Marg, New Delhi - 110002, Delhi
Ph: +91-11-65705229 Email: dscdadeda@gmail.com Web: www.dosonline.org
Intermittent exotropia (IXT) is an entity that every strabismologist comes across in their day to day clinic. The incidence is estimated to be 32 per 100,000; ranging from 1 in every 30 cases in the eastern countries to about 1 in every 100 in the western countries. It is first noted as an intermittent strabismus or exophoria in children and the patients can usually control their deviation at this stage. The ability to do so can be measured based on various described control scores. Although assessment of control can facilitate management decisions, yet intermittent exotropia is a difficult condition to master because of its variability and uncertain natural history. There are a variety of conservative options available as well surgical treatment strategies in cases of progression. However, even surgical options are known to cause recurrence. Dunlap has stated that few problems in ophthalmology are more frustrating than surgical treatment of intermittent exotropia. To correct an exotropic child for few months or a few years, only to watch his eyes turn to their divergent state is an unpleasant experience for both the child’s parents and the ophthalmologist. It is especially distressing to observe such a failure even after the second surgical procedure, which involves the remaining two horizontal recti. As randomised prospective trials on intermittent exotropia are rare, we asked a panel of eminent strabismologists from around the globe about their opinion and views on various aspects of the diagnosis and management of IXT through this questionnaire.

**Controversies in Intermittent Exotropia**

Dr. Seyhan B. Özkan, Turkey
Dr. Michael X. Repka, USA
Dr. Frank Martin, Australia
Dr. Scott A. Larson, USA

Dr. Federico G. Velez, USA
Dr. Kamlesh, India
Dr. Subhash Dadey, India
Dr. Savleen Kaur, India

(SBO): Dr. Seyhan B. Özkan, Professor, Adnan Menderes University Medical School, Department of Ophthalmology, Aydin, Turkey and President International Strabismus Association.
(MR): Dr. Michael X. Repka, Wilmer Eye Institute, Johns Hopkins School of Medicine Baltimore, Maryland, USA.
(FM): Dr. Frank Martin, Clinical Professor. Medical officer at the Sydney Children’s Hospitals Network, Australia and President International Pediatric Ophthalmology and Strabismus Council.
(SL): Dr. Scott A. Larson, Professor of Paediatric Ophthalmology, Department of Ophthalmology and Visual Sciences, University of Iowa, USA.
(FV): Dr. Federico G. Velez. Associate Clinical Professor of Ophthalmology. (Pediatric Ophthalmology and Adult Strabismus- Jules Stein Eye Institute; Doheny Eye Institute; Olive View-UCLA Medical Center). UCLA School of Medicine. Los Angeles, California, USA.
(K): Dr. Kamlesh, Director and Head of Ophthalmology, Guru Nanak Eye Center, Maulana Azad Medical College, New Delhi, India and President, Delhi Ophthalmological Society.
(SD): Dr. Subhash Dadey, Director Professor of Ophthalmology, Guru Nanak Eye Center, Maulana Azad Medical College, New Delhi, India. President Strabismus and Pediatric Oph Society of India and Secretary, Delhi Ophthalmological Society).
(SK): Dr. Savleen Kaur, Senior Research Associate, Advanced Eye Center, Post Graduate Institute of Medical Education and Research, Chandigarh, India. Executive Editor DOS Times.
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<th>SK:</th>
<th>How many cases of IXT do you routinely see in your clinic?</th>
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<td>SBO:</td>
<td>We did not make a statistical survey in our clinic on the number of the cases with IXT but on observational basis the routine number of IXT is not different than that of the literature results as 1:3 of esodeviations. The Turkish population represents Caucasians and the incidence of esodeviations are not as high as in some Asian countries.</td>
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<td>MR:</td>
<td>There are probably 4-5 patients per session with IXT, and probably one new IXT patient every clinical day. Many of the children are seen for a second opinion on what treatment to do as I suspect this discussion will demonstrate that the decision is not simple. There is also possibly one adult with the phenomenon at least weekly in search of care.</td>
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<td>FM:</td>
<td>The common presenting complaint in children is the complaint of parents about the appearance of their child. In adults, the main complaint is again the appearance and problems of eye contact in social life. As the patients with IXT suppresses while squinting, most of the time they do not have asthenopic symptoms as the patients do with decompensating exophorias who present with diplopia. However, some patients learn to control their deviation by voluntary convergence and this group is highly bothered with asthenopic symptoms and visual blur. The most essential points for examination are same as the routine evaluation of any patient with strabismus that includes visual acuity, cover test prism cover test, ocular motility, evaluation of alphabetical pattern, measurement of near point of convergence, assessment of fusion and stereopsis, dynamic retinoscopy, cycloplegic refraction and fundus examination. The additional points to assess are: how well the deviation is controlled at near/distance and far distance, binocular visual acuity, the prism cover test at far distance, measurement of the deviation on side gazes and one hour patch test followed by plus 3 diopters lens test. We perform the last two tests on subsequent visits.</td>
</tr>
<tr>
<td>SK:</td>
<td>What is the most common presenting complaint in your setting and what are the most essential points for examination in such patients?</td>
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<tr>
<td>SBO:</td>
<td>The most common presenting symptom in children is the complaint of parents about the appearance of their child. In adults, the main complaint is again the appearance and problems of eye contact in social life. As the patients with IXT suppresses while squinting, most of the time they do not have asthenopic symptoms as the patients do with decompensating exophorias who present with diplopia. However, some patients learn to control their deviation by voluntary convergence and this group is highly bothered with asthenopic symptoms and visual blur. The most essential points for examination are same as the routine evaluation of any patient with strabismus that includes visual acuity, cover test prism cover test, ocular motility, evaluation of alphabetical pattern, measurement of near point of convergence, assessment of fusion and stereopsis, dynamic retinoscopy, cycloplegic refraction and fundus examination. The additional points to assess are: how well the deviation is controlled at near/distance and far distance, binocular visual acuity, the prism cover test at far distance, measurement of the deviation on side gazes and one hour patch test followed by plus 3 diopters lens test. We perform the last two tests on subsequent visits.</td>
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<td>SL:</td>
<td>I conduct six clinics per week and would see at least five children with IXT in each clinic.</td>
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<td>SD:</td>
<td>Approximately 30-40% of all our strabismus cases are IXT. This represents about 8-12 per day. We cover a referral area of 3 million people. This problem is the second most common strabismus problem seen in our university clinic after accommodative esotropia.</td>
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<td>SL:</td>
<td>Most of the complaints are from the parents. They notice the eye drifting and they are concerned that the vision is being damaged. Other children and adults complain of their appearance, eye strain, headaches and intermittent diplopia. The examination should first include an assessment of the stereovision and fusional capability. We do this with Randot or stereo fly testing and the Worth 4 dot test. The examiner should then assess the level of control of the deviation. This is incorporated in the cover testing. With the patient fixating on an object, the testing occurs at both distance and near. The examiner occludes an eye and after uncovering the eye assess the facility and rapidity with which the patient recovers from the esotropia. Then ocular alignment (in the 9 cardinal gaze positions plus head tilts if there is any vertical deviation) and an assessment of ocular ductions can be performed. This is followed by visual acuity testing at both distance and near. Next one would do a cycloplegic refraction and an assessment of ocular health including an examination of the anterior and posterior segments of the eye with attention to the macula and optic nerve. Other testing for distance/near disparity could include the use of 45-minute occlusion test and ocular alignment at near with plus 3.00 lenses.</td>
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<tr>
<td>SM:</td>
<td>Most commonly; the parents see the drifting and squinting.</td>
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| SL: | Most common complaints are intermittent outward deviation of eyes followed by asthenopic symptoms. Children often present with diplopophobia. As in all patients of strabismus, complete and thorough examination should be done in all the patients of IXT. However, few important points of examination in these patients are: a) BCVA should be given after cycloplegic refraction, and deviation should be measured with glasses. b) Deviation measured should be the 'static one’ i.e. that measured by breaking the fusion and suspending the accommodation. At our institute, we
use overnight patching of one eye for this purpose. c) Deviation should be measured in all the gazes to look for any pattern and lateral incommitance. d) Both near and distance deviation should be measured. e) Binocular status must be checked.

SD: Patients of IXT are usually asymptomatic. This can be explained due to the presence of well-developed sensory adaptations. As the age of onset period of intermittent exotropia (2-5 years of age) is the period of visual function maturation for children, binocular function is often affected. As a rule, during the phoric phase of intermittent exotropia, the eyes are perfectly aligned and the patient has excellent bifoveal fusion with excellent stereo acuity ranging between 40 and 60 seconds of arc.

During the tropic phase, when the exodeviation manifests itself, outward drifting of eye is the most common complaint. Patients with late onset exotropia, after the age of 6-7 years, may experience diplopia because the exotropia develops after the loss of plasticity that allows suppression.

The various symptoms seen in IXT are: asthenopic symptoms, transient diplopia, micropsia and diplophotophobia. The examination, in addition to routine examination of visual acuity, refraction, cover test for near and distance, motility examination and tests for stereopsis, following special tests are required in relation to IXT.

Measuring the angle of deviation by a prolonged alternate cover. If there is significant angle variability or a significant distance/near discrepancy after prolonged alternate cover testing, only then a patch test is indicated. We must understand the basic principles that plus 3.00 lenses suspend normal accommodative convergence, whereas monocular occlusion relaxes fusional convergence mechanisms, therefore monocular occlusion should be used before plus 3.00 D lenses to measure near deviation, to avoid misdiagnosing a high AC/A ratio.

I rely on Kusher's Method for assessment of control. It is very important to carry out repeated examination in these children.

SK: How do you measure deviation in such patients and what is the role of Post patch test and plus 3 dioptre lens test?

SBO: The deviation is measured with and without glasses at near and distance as well as far distance. I first ask the patient to look at far distance before dissociating the patients’ eyes with cover test. The first thing that I do is to evaluate the control level first with a quick cover test without much dissociation effect. Then I make a more dissociative cover test and make prism cover measurements at near, distance and far distance. The near point of convergence is measured in all patients. Especially in adult patients I also measure the visual acuity binocularly. Those who use voluntary convergence have a decrease in visual acuity when they control their deviation and that can easily be observed during binocular visual acuity examination. The voluntary convergence, which is highly bothersome, should not be confused by a good control. Post patch test is essential to unmask the real near deviation by eliminating the fusional convergence and plus 3 diptters test to unmask the accommodative convergence. Plus 3 diptters lens test must be done following monocular occlusion and the patients should not be allowed to remain binocular during these evaluations. The existence of the exodeviation similar with the distance deviation indicates a normal AC/A ratio and the need for a recess-resect surgery. The increase of near deviation with plus 3 lenses indicates a high AC/A ratio and these patients may need bifocals after surgery.

MR: Prior to performing any strabismus measurements or even measure visual acuity, it is ideal to watch the patient for a few minutes to see if there is spontaneous deviation of the eyes and how much of the time it occurs under casual viewing. Also noting what maneuvers with the eyes or the head that a child uses to restore alignment can be informative. I would assess control at distance based on the control score we have used in PEDIG studies and described by Mohney and Holmes. It is important to recognize that there is tremendous variability of the measurements of control during the day, so this should not be an absolute determinant. It is also important to remember that far distance (or remote) viewing will unmask many intermittent exotropia.

Post patch test is used to determine the maximum distance angle as well as to determine if the smaller near angle when present is due to tenacious proximal fusion. It is reassuring to go forward with the surgery and not fear an overcorrection. The plus 3.00 diopter test is used when the deviation at near does not increase with the patch to see, if the small near angle is due to a high AC/A ratio. If that is found, caution is needed when deciding to operate on such patients as they are at risk for a postoperative overcorrection and diplopia.

FM: The deviation is measured with the prism bar cover / uncover test. It is difficult to perform this test in very young children as they will not fixate in the distance or far distance. The post patch test differentiates true divergence excess intermittent exotropia from pseudo divergence excess intermittent exotropia. This test is performed when the child is old enough to cooperate for accurate prism bar measurements. The 3 dioptre lens test unmask the deviation that is controlled by convergence / accommodation. We do not place much emphasis on this test.

SL: We have orthoptists assisting the physicians in the evaluation of all strabismus patients. They begin with a cover-uncover test to determine the amount of tropia present and to assess the level of control. If there is a large latent component, then a simultaneous prism and cover test is used to quantify the alignment. This is followed by an alternate prism and cover test. If there is a variable alignment or a larger deviation at distance, the deviation at near is measured with a plus 3.00 lens before each eye. The
45-minute occlusion of one eye is then done next with care not to allow the child to have binocular vision when the patch is removed. The alternate prism and cover test is repeated after 45 minutes of monocular patching. The examination is finished with a cycloplegic refraction.

**FV:** Diagnosis can be made by the alternate prism cover test. Patching and plus 3 D tests are very helpful, but time consuming as well as hard to do for all patients.

**K:** As mentioned above, we measure deviation after patching one of the eyes. Deviation is measured in all the gazes. Also, both near and distance deviations are measured. Post patch test is the single most important test for measuring exact deviation in such cases to determine Full (static) deviation to determine distance/near disparity (True /Simulated divergence excess and to determine the type of surgical procedure.

Plus 3.0 D test has been devised to diagnose the patients of divergence excess type who have true divergence excess due to high AC/A ratio. This test uses the lens gradient method to measure the AC/A ratio. These patients are the ones who will continue to have a distance/near disparity post-operatively, and may require bifocal spectacles after surgery for a consecutive esotropia at near. This test should be resorted to in patients who have a distance deviation greater than near deviation of 10 prism diopters or more after the patch test. After the patch test while still dissociated, re-measure the deviation at near with a plus 3.0 add. If the exodeviation at near increases by 20 prism diopters or more the diagnosis of high AC/A ratio true divergence excess intermittent exotropia is made.

**SD:** Routine alternate cover prism testing gives variable results in IXT; therefore, it is important to carry out a prolonged alternate cover. It suspends tonic fusional convergence. If there is significant angle variability or a significant distance/near discrepancy after prolonged alternate cover testing, only then a patch test is indicated.

Patch Test - It is used for differencing cases of true divergence excess from pseudo-divergence excess. It’s another use is to control the tonic fusional convergence. We do the patch test after 24 hours of mono-occlusion to get the best results.

Plus 3.0 D near add test (lens gradient method) - This test is useful in cases of true divergence excess due to high AC/A ratio. This test uses the lens gradient method to measure the AC/A ratio. After the mono-occusion patch test, we measure the deviation at near with a plus 3.0 D add. In cases of high AC/A ratio true divergence excess intermittent exotropia, the deviation at near increases by 15 diopter or more.

**SK:** What is the best way to monitor progression in these patients? Do you routinely use any scoring system for these patients?

**SBO:** It was recently demonstrated that the evaluation of the ‘control’ of the deviation was not consistent even with the same examiner on the same patient at different times of the day. We do not use any scoring system to decide on surgery, but we certainly look for all the aspects in clinical scoring systems. The control level is documented as ‘spontaneous control’, ‘control with blinking’ and ‘control with refixation’ at near, distance and far distance in all the patients.

The general rule is to document the worsening of the control of deviation in three consecutive visits. If there is a decrease in stereoaucity or conversion into a constant exotropia, the decision for surgery is more rapid and verification of the decompenation is done in more frequent visits.

**MR:** I would recommend monitoring the IXT patient with good visual acuity about every 6 months. Always consider that this patient might have an optic neuropathy. We use the PEDIG control score (0 to 5), but also recognize the remarkable variability of this measurement and perhaps do not make a surgical decision based on a single observation, but rather examine a couple of times.

**FM:** The best way to monitor progression in these patients is to listen to the parents and ask them how much of the time they are aware of their child’s exotropic deviation. In children who are old enough to cooperate, we also use the Lang stereopsis test as a monitor of control of the deviation. If the child starts to lose stereopsis, this is generally an indication that control is deteriorating. We do not use any particular scoring system. We, however, do note in the records where the children have good, fair or poor control, depending on whether the deviation after being unmasked with the cover test recovers spontaneously, with a blink or fails to recover.

**SL:** I use the parent’s assessment of control as the prime factor in deciding in when to proceed to more treatment. If there is a disparity in the assessment level of control between my assessment and the parent’s assessment, particularly when I believe the child has worse control than the parents see, I work to come to common ground with the parents by teaching them to monitor their children closely. I don’t record a formal scoring system in the patient chart. However, I use most of the elements from formal scoring system which include the rapidity of recovery from the exotropia and the behaviours that were needed to recover from the exotropia. This could include spontaneous recovery, blinking or near fixation.

**FV:** Control reported by parents is the best way to monitor progression. Measuring the near and distance stereopsis is also very essential.

**K:** Signs of progression of Intermittent Exotropia are: Progression of tropic phase, exotropia occurs more than 50% of waking hours, secondary convergence insufficiency, increase in size of basic deviation, development of suppression (no diplopia).

We routinely use the following system to judge the control.

Good Control: Patient “breaks” only after cover testing and resumes fusion rapidly without need for a blink or refixation. Fair Control: Patient blinks...
or refixes to control the deviation after disruption with cover testing. Poor Control: Patient who breaks spontaneously without any form of fusion disruption. SD:

Best way is to take the information provided by parents in account. Recently Kushner et al have introduced the concept of assessment of control of intermittent exotropia. It is a very good method and it helps to evaluate the patients in a better way and acts as a guide to monitor deterioration and progression of intermittent exotropia.

**Subjective Methods**

*Home Control:* The parents may be told to keep a chart noting the control of deviation at home in terms of the percentage of waking hours the manifest deviation is noticed at home.

*Office Control:* Good Control: Patient “breaks” only after cover testing and resumes fusion rapidly without need for a blink or refixation. Fair Control: Patient blinks or refixes to control the deviation after disruption with cover testing. Poor Control: Patient who breaks spontaneously without any form of fusion disruption.

**Objective Methods include measurement of Distance and near stereopsis.**

*Distance Stereo acuity:* It provides an objective assessment of both control of the deviation and the deterioration of fusion that occurs early in this disorder. Normal distance stereoaucity indicates good control with little or no suppression. The Mentor B-Vat II BVS assesses distance stereoaucity using both contour circles and the random dot E test from 240 to 15 seconds of arc disparity.

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**SK:** What is your first line of management in these patients-conservative or surgical?

**SBO:** In all patients as a first line of management, I prefer to be on the conservative side and try to increase the control of the deviation. The first golden rule is not to make the patient worse. Most of these patients may develop and keep good level of stereopsis despite that they have day-fatigue exotropia at distance. Post-surgical monofixation syndrome is my major concern in childhood.

**MR:** I tend to be conservative and observe for a few months at least. I also want to be sure that there is no evidence of an optic neuropathy. It looks bad if you rush to surgery and this was for a sensory exotropia from optic nerve compression or developing oculomotor nerve paresis.

**FM:** We tend to adopt a conservative management in children with intermittent exotropia. At the initial consultation, we outline to the child’s parents that the child may require glasses if there is significant refractive error, part-time occlusion or orthoptic treatment. Generally, we do not commence orthoptic treatment until the child is aged 4½-5½ years and old enough to cooperate as otherwise, it becomes a frustrating experience.

**SL:** In the past, I believed patching helped IXT. I prescribed it to many. I was a participant in the PEDIG trial for alternate patching of IXT in children. The results of this study have significantly changed my behaviour and I do not recommend patching very often any more. I am part of the minus lens glasses study for IXT with PEDIG as well. We will have those results in a few years. I have been less impressed with minus lens therapy in kids that don’t have a high AC/A ratio than I was for patching and I am concerned about inducing myopia in a minority of kids. I discuss patching and glasses with families, but they often move to surgery.

**FV:** The first line of treatment can be both conservative and surgical depending on the presentation of the patient.

**K:** Non-surgical treatment for intermittent exotropia is not very effective but it may be preferred in patients with small deviations (<15 PD), convergence insufficiency type of IXT, very young patients in whom surgical overcorrection could lead to amblyopia or loss of bifoveal fixation and in patients who otherwise cannot be taken up for surgery. Any patient showing signs of progression should be operated.

**SD:** The treatment for IDS is primarily surgical, non-surgical methods can be tried as a temporary measure in the following situations. 1. Patients with small (<20 PD) deviations 2. Very young patients in whom surgical overcorrection could lead to amblyopia or loss of bifoveal fixation 3. Patients waiting for surgery or in whom surgery is contraindicated 4. Patients with a high AC/A ratio may be responsive to non-surgical methods.

**SK:** What are the conservative measures that you advise for IXT when the control is good- orthoptic exercises, occlusion, botulinum toxin A therapy, binocular vision training or watchful waiting?

**SBO:** The first major step is the correction of refractive error. Refractive correction is the major tool to control the level of accommodation and convergence as well as obtaining good and equal visual acuity. The first step is to balance the fixation preference. Any level of amblyopia provokes decompensation and requires occlusion of the dominant eye. Even though I find an equal level of visual acuity, if there is a fixation preference, I prescribe patching which most of the time helps to control the deviation. As the orthoptic exercise, I only use pencil push up exercises if the near point of convergence is remote or if there is difficulty to maintain convergence. I tell the parents about the possible scenarios and inform the necessity of follow up and the necessity of parental observation. The collaboration with the teachers usually give reliable observational results as they have the opportunity to observe the child during the whole day from distance. If the control is getting worse in an adult patient with moderate angle of deviation, I advise botulinum toxin injection before considering any surgery. In children, because of the off-label use of botulinum toxin, we prefer to use it to rescue over/undercorrections in IXT.

**MR:** Monitoring is foremost and probably what I use most often with patients. I would suggest home orthoptic exercises for school age children that are interested...
Some ophthalmologists use occlusion therapy for non-surgical treatment consists of following options:

K: My impression with orthoptic exercises or binocular vision training is that they don't help. Although I think there is a need for more studies in this area. I don't use Botox for this problem, so most often the decision is to observe or do surgery.

SL: Where there is good control, we generally use orthoptic eye exercises to improve the control. If the child can learn to recognise when the eye has deviated and spontaneously recovers from the exotropic position, then we feel we have made great progress and are well on the way to longstanding good results without the need for surgery.

FM: For non-surgical management, we observe; followed by patching for one hour and glasses.

Non-surgical treatment consists of following options: Correction of Refractive error, Overcorrecting minus lens therapy, Part time occlusion, Prismotherapy, Orthoptics and botulinum toxin injection.

Anisometropia, astigmatism, myopia and even hyperopia can impair fusion and can lead to occurrence of manifest deviation, therefore it is mandatory to do a proper cycloplegic refraction in each and every case. It is particularly useful in myopes as correction of refractive error keeps the deviation in control in few patients. In cases of high AC/A ratio, overcorrecting with minus lenses is helpful because stimulating accommodative convergence can reduce an exodeviation. Part time occlusion is temporary method and useful in young children or can be used to postpone surgical intervention in responsive patients. In some cases, part time occlusion of the non-deviating eye for four to six hours daily may convert an intermittent exotropia to a phoria. Alternate occlusion may be used in patients with equal fixation preferences. If the angle of deviation is decreased, the occlusion should be continued and assessment should be made every 4 months until no further change occurs. In case there is no improvement for 4 months, it is discontinued. Prisms are rarely a long-term solution in patients with intermittent exotropia, but can be used to improve fusional control. For smaller deviations of up to 20 PD, base in prisms may be used to assist control and relieve asthenopic symptoms. Convergence exercises are indicated for symptomatic patients with convergence insufficiency because of the high risk of overcorrection with surgery. The goal is to increase the ranges of fusional convergence and divergence.

Some ophthalmologists use occlusion therapy for preoperative antisuppression but the efficiency remains debatable. Alternate patching has been advised to improve control and reduce suppression.

These measures can delay surgery or can be beneficial for smaller deviations, but are not a substitute for surgery. Botulinum toxin injection into the lateral recti has been reported as an alternative to surgery but with varying success rates.

SL: I have nearly stopped doing patching for IXT since our PEDIG trial results became available. I do use minus lenses in cases of High AC/A but not in true computer-based activities, although the data are sparse to confirm a benefit. I do patching only if there is evidence of amblyopia, but I do tell patients about the PEDIG trial which showed a minor benefit in a few patients.

FM: Some patients are interested in trying exercises and those will try vergence exercises (e.g., pencil pushups) for a period of time. I am unimpressed with their outcome in most cases. Over minus lenses are an interesting option and seem to help a few patients. There is some evidence that it can work, often with about minus 3.00D added to the cycloplegic refraction. PEDIG is currently conducting a randomized trial of over minus by minus 2.50D in children when the hyperopia is 1.00 or less after an earlier pilot study showed some potential for this dose.

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divergence excess type IXT. I haven’t seen benefit in my patients with convergence exercises.

FV: These modalities cannot be used as a substitute. Yes, I do prescribe pencil push up exercises.

K: No, we do not use these methods as a substitute for the surgery. Rather these methods are used as temporary measures till surgery is performed. Convergence exercises are prescribed by us in convergence insufficiency type of IXT. Orthoptics are not a substitute for surgery. Orthoptics don’t make any significant difference to surgical outcome but may be useful pre/post operatively for: Antisuppression therapy, diplopia awareness and fusional convergence training.

SD: Treatment of IXT is surgical and only surgical. There is no substitute for surgery. Knapp summarized the opinion of most strabismologists by stating that orthoptics should not be used as a substitute for surgery but rather as a supplement. The aim is to make the patient aware of manifest deviation and to improve the patient’s control over it. Convergence exercises are preferred in convergence insufficiency type of IXT.

SK: How do you decide which patient needs surgery and what are the goals of surgery?

SBO: In children, my major concern is to avoid amblyopia and to preserve stereopsis. If I see any risk about losing the binocularity which is usually manifested by decreased control of near deviation or in cooperative ones with the documentation of diminished stereopsis scores, I suggest surgery. The goal of surgery is to correct the alignment by preserving binocular function. The worst scenario is to end up with monofixation syndrome, loss of stereopsis and development of amblyopia in a child who had stereopsis despite moderately controlled IXT. The goals of surgery in adult patients are again to correct the alignment, to reduce the asthenopic symptoms related to the convergence effort and to increase their quality of life by eliminating the psychosocial impact of squint. In case of a recent decompensation of a well-controlled IXT, my first choice is botulinum toxin injection in adult patients. If botulinum toxin treatment does not provide a satisfactory result, or if the patient does not accept botulinum toxin injection for any reason, surgery is planned.

MR: Symptoms are my key determinant of when surgery is needed. Diplopia, squinting, covering an eye or children in school commenting about the patient and having difficulty with social issues. If we knew the surgery to be always successful I might lower my threshold but IXT surgery does not always have a perfect outcome with very frequent recurrences. The goal of surgery is functional binocularity plus an angle that stays small or completely controlled.

FM: Generally, we try to avoid surgery until the child is out of the amblyogenic age group (5-6 years). At times, we do need to perform earlier surgery if the child meets our indications and criteria for surgery. The indications include: exotropia present more than 50% of the time, asthenopic symptoms from the child trying to control the IXT, the child is being teased at school about the IXT and at times we will go ahead with surgery if the parents are extremely concerned about the child’s strabismus (cosmesis). At each consultation we monitor the child, not only for control of the IXT but also for amblyopia and loss of stereopsis. The goal of surgery is to achieve a good cosmetic outcome, even after surgery children still break down to exophoria or IXT. The most stable postsurgical results are children who end up with a micro esotropia. This, however, can sacrifice their previously good stereopsis.

SL: The decision to go to surgery is a cooperative one between the parents and myself. I rarely push surgery if the family is not keen to do it. The goal of course would be to eliminate the IXT completely. However, often the result is much better control with very infrequent exotropia. I believe this could be a successful outcome even though cover testing shows a recurrent IXT.

FV: The indications for surgery are poor control; diplopia and worsening stereopsis. The goal of surgery is an initial overcorrection.

K: As mentioned earlier a patient who shows signs of progression or in other words when a patient shows loss of control, he/she should be operated. The goal of strabismus surgery for intermittent exotropia is to restore alignment and to preserve or restore binocular function. We target overcorrection of 8-10 PD in all patients above 4 years of age.

SD: Following may be the indications for surgical intervention:

- Poor control: Poor control may be judged both by home control and office control. Exotropia occurring at least 50% of waking hours may be manifestation of poor control calling for surgical intervention.
- Deterioration of control: Serial observations are required to assess deterioration/progression. Following are the signs of progression:
  - a) Increase in the frequency of the manifest phase of squint
  - b) Development of secondary convergence insufficiency
  - c) Increase in size of the basic deviation
  - d) Development of suppression as indicated by absence of diplopia during manifest phase
  - e) Progressive deterioration of distance & near stereopsis.
- Severe asthenopia: Most patients are usually asymptomatic because of the development of hemiretinal suppression. In some cases, however, such as in the convergence insufficiency type of exodeviation, severe asthenopic symptoms are especially bothersome at near. Asthenopia may be a consequence of deteriorating control. If a trial of vigorous orthoptic exercises is unsuccessful, definitive surgery will be required.
- Bother some diplopia is usually an indication for surgical correction.

By surgical intervention in intermittent exotropia, we aim to achieve both cosmetic as well as functional success. We should aim for short term over correction.
for long term desirable results in children (4-10 PD), to counter the tendency of eyes to drift over time. In young adults, we should aim for orthophoria so as to restore alignment and to preserve or restore binocular function.

**SK:** Do you believe there is a relation between age and response to surgery and thus do you have any age preference for surgery? What is the best age to operate these patients?

**SBO:** The general tendency is not to operate IXT before 4 years. However, there is a group that may certainly require surgery below 4 years of age with signs of converting into a constant exotropia. In those cases, waiting too long may result with the loss of binocular capacity. Unless early surgery is required, the rule of thumb is to allow the necessary time for the visual system to recover by providing the optimal circumstances with refractive correction, amblyopia treatment or convergence exercises where necessary. The correct timing for surgery depends on the patient and it is the time to operate when deterioration of the control of the deviation occurs.

**MR:** I do not know, but there are many opinions. There are arguments for delay until older ages in childhood to preserve binocularity and avoid a postoperative permanent monofixation syndrome that may result from an overcorrection compared with earlier to preserve binocularity before it withers away. It would be nice to study this question but the randomization from an overcorrection compared with earlier to preserve binocularity before it withers away. It would be nice to study this question but the randomization of surgery to prevent occurrence of monofixation syndrome from consecutive esotropia. On the other hand, Jampolsky advocates delayed surgery, citing advantages like accurate diagnosis and quantification of the amount of deviation and to avoid consecutive esotropia and development of amblyopia. Success rate from surgery is more in the younger patients as compared to adult patients. We believe patients should be operated around four years of age and the surgery in less than age 4 is reserved for patients in whom rapid loss of control is documented.

**FM:** Our practice has been to try to defer surgery until the child is out of the amblyogenic age group. Our preference for surgery is around 5-6 years. We do not believe there is any relation between age and response to surgery. I prefer unilateral recess-resect surgery unless there is a visual difference with surgery in the poorer seeing eye, when there is a real strong fixation preference, and in all of my adult patients. A PEDIG trial exploring this question failed to show a difference in outcome with the two surgical approaches 3 years after surgery.

**SL:** I have seen a higher success rate from surgery in the younger patients compared with older patients. I believe the PEDIG study on surgery for IXT will also report on their findings in this regard. Stay tuned for that information.

**FV:** Age is not a direct influence. Any patient who is doing poorly or deteriorating regardless of age is a candidate for surgery.

**K:** Small children are uncooperative for examination; thus, true deviation is not measured. Thus, surgery often results in undercorrection. Timing of surgery is controversial. Surgery should be delayed in visually immature infants to avoid overcorrection. It can be delayed up to 4 years for proper preoperative assessment and to avoid amblyopia and loss of stereopsis due to desired initial consecutive esotropia. Early surgery under 4 years only if deterioration occurs in phase of intermittency to maintain binocularity to avoid intractable sensory changes. If there are significant signs of progression we operate that patient irrespective of age.

**SD:** Intermittent exotropia is as much a controversial topic. Among all the factors, the most confusing and most controversial is the age of patient at the time of surgery. Opinions vary widely about the appropriate timing of surgical intervention for patients with intermittent exotropia. Knapp and many other workers advocated early surgical intervention to prevent development of sensory changes that may prove intractable later. However, we must keep in mind that in visually immature children a slight under correction should be attempted to prevent occurrence of monofixation syndrome from consecutive esotropia. In small differences between near and distance deviation, I manipulate the amount of surgery on a recession-resection basis. In pure convergence insufficiency where the patients are orthophoric at distance, I prefer botulinum toxin injection instead of surgery because of the high risk of carrying the problem at distance. If the near deviation is more than the far deviation but there is still manifest deviation at distance I manipulate the surgery by decreasing the lateral rectus recession and increasing the medial rectus resection. The presence of lateral incongruity, the results of patch test and +3 dioptries test influence the planning of surgery.
I think if we do this young enough, we may prevent the under-correction rates are significant with this procedure. I may do a recess/resect or bilateral medial rectus resection for convergence insufficiency type but I have been humbled by the difficulty with this sub-set of patients. In my hands operating on a convergence insufficiency problem in an adult is one fraught with poor outcomes from overcorrection at distance. For this, I use a procedure that is weaker in my hands, medial rectus plications. This has reduced the distance overcorrections and in theory may be easier to fully or partially reverse.

FV: For basic type I prefer unilateral surgery and for all other types bilateral surgery.

K: We decide for surgery depending on distance-near relationship: A. Basic type -1. Monocular recession and resection. 2. Bilateral recession - if deviation is less than 35pd. B. Distance exodeviation more than near: Bilateral LR recession is the choice of surgery. C. Exo more at near than distance: Bilateral MR resection.

SD: For all types of exotropia except the convergence insufficiency type we advise bilateral lateral rectus resections. To avoid lateral incomitance, it is advisable to perform symmetric surgery rather than monocular recession/resection procedures. For convergence insufficiency type our preference is for bimedial resection. 8-mm unilateral lateral rectus recession is an attractive alternative procedure in the treatment of intermittent exotropia with a deviation of 25 to 30 PD.

SK: Do you think treatment in IXT changes the natural course of the disease? Does surgery correct the central and peripheral fusion as well as stereopsis in a patient?

SBO: Yes, I think it changes the natural course, these patients go back and forth in good stereopsis and suppression and the aim is to increase the balance towards the binocularly well controlled state. Lack of appropriate follow up or treatment, leads many of those patients to convert into constant exotropia with amblyopia and loss of binocular vision.

MR: Yes, in some patients, the deviation is cured and cannot be found during long term follow up with very normal binocularity. However, in many patients there is a recurrence of the deviation over time, while in others the eyes are aligned but the stereo is not improved.

FM: The short-term results from surgery for IXT are reasonably good, however, long term results are disappointing. Surgery may help the child maintain central and peripheral fusion as well as stereopsis. Surgery does not correct abnormal sensory function.

SL: I think if we do this young enough, we may prevent peripheral suppression from becoming so habitual that we can change the course of the condition. We do need more data and the PEDIG data will help to clarify this information.

FV: I believe timely surgery prevents further deterioration.

SD: Yes, the success rate of surgery varies from 60 to 90 percent. Surgery at appropriate time helps the patients to maintain fusion as well as stereopsis.

SK: How do you determine the target angle for surgery and do you aim for overcorrection as your early post-operative goal? Do you routinely do the prism adaptation test before surgery?

SBO: We do not perform prism adaptation test in our clinic as it costs high and is too much time consuming. The early post-operative goal is to have and overcorrection of 10 prism dipters. I am also glad to see an overcorrection up to 15 prism dipters but larger esodeviations make me concerned. If the overcorrection persists with decrease of stereopsis I first adjust refractive correction and if it does not work, inject botulinum toxin into the medial rectus muscle.

MR: I measure the distance deviation with prism and alternate cover test. I expect a slight over correction in children with bilateral lateral rectus muscle resections based on my tables for surgery. For adults when I do unilateral recess-resect I aim at the adjustment to have them aligned at near and a very slight to no deviation at distance. I do not use preoperative prism adaptation in the management of IXT or exotropia.

FM: In the preoperative workup, it is important to unmask the maximal angle of the IXT. Surgery aims to correct the maximal angle and even though it is not what one aims for, there is often a small overcorrection immediately after the surgery. We do not routine use the prism adaptation test before surgery.

SL: I aim for a small amount of esotropia, less than 15 PD for the post op angle in the first week in children and in adults that can handle it. I do not routinely do prism adaptation for this condition.

FV: We rarely use the prisms adaptation test. We aim for overcorrection as long term recurrent drifting occurs in 30%.

K: We perform post patch test at least 3 times before proceeding to surgery. Maximum deviation measured for distance after patching is taken as target angle. As mentioned earlier, we aim for 8-10 PD over correction in all our patients except very small children. No, we don’t perform prism adaptation test routinely before surgery in our cases.

SD: Measurements should be performed at fixation distances greater than 6m after unilateral occlusion for 24 hours. We aim for overcorrection of 10 prism dipters. Prism adaptation testing is considered by some to be useful in determining the target angle of surgery whereas others have found no differences in surgical results with their use. We do not perform this test routinely.
SK: What is the long-term stability of surgery in these patients? How do you prognosticate your patients in terms of longevity of your surgical success? Do you believe some patient/ surgical factors make them prone to an exotropic drift?

SBO: Amblyopia, lack of stereopsis or fusion and remote near point of convergence are the high-risk factors for the recurrence of the deviation. Coexisting vertical deviations and especially the presence of ‘A’ pattern deviation are also the major risk factors for recurrence.

MR: About 60% will do well with no significant deviation recurring, but the remainder will have a residual deviation which may grow with time. I do not know which factors are significant.

FM: Patients with intermittent exotropia often have a strong family history of the condition. The long-term results are disappointing. We estimate that approximately 50% of patients who undergo a surgery within 5 years have a recurrence of the IXT. We have found it is important to correct refractive error, especially myopia as this helps maintain control of the IXT. We have not been aware of any patient/surgical factors that make patients more prone to an exotropic drift.

SL: The majority of patients will drift back towards exotropia. If we have < 15 PD of esotropia in the first week or two and a small esophoria a month later, then I believe we will have a better long-term success. If they have any exotropia on either of those visits, then there will likely be more exotropia in the future. If they have good alignment a year out from surgery then they will unlikely break down later but you can never be sure. I try to never label anyone a “cure”.

FV: I believe that patients with basic type of IXT operated with only lateral recessions drift out again even more.

K: We believe that if pre-operative work up of the patient is thorough, and the surgery has been done for the largest measured angle of deviation with a target of 5-10 PD of overcorrection, then most of these patients maintain correction over a long period of time.

SD: Dunlap has stated that few problems in ophthalmology are more frustrating than surgical treatment of intermittent exotropia. To correct an exotropic child for few months or a few years, only to watch his eyes turn to their divergent state is an unpleasant experience for both the child’s parents and the ophthalmologist. It is especially distressing to observe such a failure even after second surgical procedure, which involves remaining two horizontal recti. Around 60 to 70 percent will drift back towards exotropia after 5 years. We don’t associate any particular patient/ surgical factor that make them more prone to an exotropic drift.

SK: What are the poor prognostic indicators for surgical treatment?

SBO: Preoperative lack of stereopsis and amblyopia are the poor prognostic indicators for IXT.

MR: It is difficult to have long term success with unilateral or bilateral sensory exotropia. I think poor preoperative stereo is a factor associated with poorer outcomes, but large studies are needed to confirm that impression. Future analyses of PEDIG IXT surgical trial data may help but the study was not large enough to have sufficient power to look at important risk factors in a definitive manner.

FM: The poor prognostic indicators are the presence of amblyopia. Children who refuse to wear their glasses to correct significant refractive error. Poor preoperative assessment and planning leads to poor prognosis.

SL: Large angle intermittent exotropia in obese myopic adults are the patients that seem to be the hardest patients for us to treat in my part of the world. They require more surgery than other patients.

FV: Absence of stereopsis and presence of amblyopia are poor prognostic factors.

K: Some poor prognostic factors are Large deviations, hyperopia, deviation angle at the initial post-operative period and amblyopia. High AC/A ratio is also a poor prognostic factor as these patients tend to have consecutive esotropia at near.

SD: A high AC/A is an indicator of a poor surgical prognosis and most of these patients have a consecutive esotropia at near. Also, larger angle deviations with amblyopia are poor candidates for surgery.

SK: Do you modify your surgical plan in cases with lateral incomitance?

SBO: Yes, we evaluate lateral incomitance before surgery in all of our cases and modify the surgical plan where necessary. The lateral incomitance which is defined as 20% decrease of the deviation on side gazes, may be due to underacting lateral rectus or a tight medial rectus muscle. Depending on the forced duction test, the lateral rectus recession or the medial rectus resection needs to be reduced to avoid overcorrection.

MR: I do not adjust. Some of these are measurement artefacts and some are real but I have not reduced the surgical dosage.

FM: We have found lateral incomitance to be a rare phenomenon. Sometimes we slightly reduce the dose of the surgery.

SL: Yes. If there is more than 5 prism dipters of lateral incomitance, and the lateral gaze usually has a smaller angle. I would reduce my surgical dose down by a millimetre or two.

FV: Yes. I do modify my surgical plan.

K: In cases of lateral incomitance, one might do either: 1. Recess each LR, more on the side where deviation is greater - for greater reduction of action of LR in that gaze 2. Recess LR on the side of greater deviation and Resect MR of opposite side - for greater effect of MR in that gaze

SD: In cases of lateral incomitance, a reduction in surgical dosage is advisable.

SK: How do you treat a case of IXT with V PHENOMENON?

SBO: The correction of any alphabetical pattern is a must in IXT. Although in V pattern the patients may have a
better control in downgaze, this is usually associate
with torsional abnormalities which has a negative
influence on control of the deviation. These patients
may develop abnormal head posture in an attempt to
control their deviation.

MR: I repair the IXT with lateral rectus muscle recessions.
Rarely is there a reason to perform inferior oblique surgery as some of these are pseudo-overaction of the inferior oblique as described many years ago by Capo et al. They noted the higher position of the fellow eye when not fully adducted because there was more room for ocular elevation. However, if there is torsion in a V pattern, and clear overaction when the eye is completely in addition then a weakening of the inferior oblique muscle can be performed.

FM: This depends on whether there is associated inferior oblique overaction. If the inferior obliques are overacting, then these are weakened at the time of the surgery for IXT. If V exotropia is present without overaction of the inferior oblique muscles, then the pattern is corrected by supracing the lateral rectus muscles at the time of surgery.

SL: Without oblique muscle dysfunction, I use vertical offset of the horizontal rectus muscles. Any oblique muscle dysfunction will lead me to address them directly.

K: Surgical considerations in such cases are: A. Large A or V patterns will have significant oblique muscle dysfunction. B. Surgery is selected to reduce horizontal deviation in primary position. C. Horizontal Muscle Transposition works well if no oblique dysfunction, but it is not substitute for it. D. Inferior oblique weakening or SO tucking correct 15-25 PD of V-pattern. E. Bilateral superior oblique tenotomies correct 35-45 PD of A-pattern. F. Primary & reading positions are functionally the most important positions of gaze.

SD: V pattern may or may not be associated with primary inferior oblique overreaction (100A). In the absence of 100A, the lateral recti may be shifted upwards or the medial recti may be shifted downwards. If 100A is present, then one of the various inferior oblique weakening procedures may be performed along with horizontal muscle surgery.

SK: What is the overall success rate in your cases?

SBO: We did not make a recent survey for IXT in our department but I think it is consistent with the general literature results.

MR: Since our move to the electronic medical record in 2013, I have not been able to search and review the data. That is clearly a deficiency in our practice and I think we , for now, only have the ability to access our patient lists by procedure and diagnosis. This data can be used to create a dashboard that show the success rate of surgery being constantly updated over time. This would make it much easier to examine the impact of practice modifications. One problem I note with any outcome measure is what we want to know. For instance, is surgery the key variable OR healing or other patient variables the important question?

This choice affects the postoperative interval to be chosen for that dashboard. Many might choose 3 months as the primary effect of the surgery, while excluding many late healing and patient factors that have impact during long term follow up.

FM: The short-term success rate is 70-80%. The long-term success rate (over 5 years is approximately 50%). We firmly believe that in managing intermittent exotropia the ophthalmologist when considering surgery should operate as late as possible and do as little as possible.

SL: Success can be defined in multiple ways and at multiple points in time. Most cases will have improved control of the misalignment. Post-op ocular alignment of <10 Prism diopters occurs in 75% of my cases on the 2-month post op visit. About 25% are under corrected and 1- 5% of my cases are overcorrected at this time point. Most of the under corrections will go on to have additional surgery. If I include all cases where I have operated one or two times, > 90% have good alignment 2 months post-surgery. I am in the process of looking at my outcomes 4-5 years out but I don’t have that data currently.

FV: Cure rate is approximately 40% but improvement is seen in 80% in my experience.

K: Based on a case series of 325 cases over a period of 3 years, our immediate success rate in these cases was 92.8%.

SD: Immediate post op success (Ortho ± 8 PD) is approximately 95 % in my cases. Three months post op success is 85- 90 %; one year success is 70-80 % and 5-year success is around 50 %.

SK: How do you treat cases of Post-operative under and over corrections?

SBO: In post-operative, over and under corrections, my first tool is glasses. In those cases with overcorrection, I prescribe full hypermetropic correction and use bifocal glasses if there is a high AC/A ratio. If the problem is under correction, I use over minus glasses. If it does not seem to work I consider botulinum toxin injection during the early post-operative period.

MR: Overcorrections are patched and occasionally placed in prism for about 6 months, trying to wean over time. A small reoperation is done if the problem does not resolve. Clearly patience seems very important with overcorrections as many will resolve. Under corrections are watched for 6 months or more, and surgery is offered at that time for large and socially unacceptable residual deviations. I do not think this resolves very often, if ever.

FM: In the preoperative informed consent progress, it is explained to parents that approximately 30% of children will require more than one procedure to correct the strabismus. We stress to the parents that the need for further surgery may occur soon after the operation if there is significant over and under-correction or may occur years later if the deviation should recur. If there is overcorrection immediately after the surgery, it is important that the surgeon and the team remain calm and reassure the parents that
most of these correct spontaneously over a period of 6 weeks. If the child is complaining of diplopia then we use part-time occlusion. If the child has any significant hyperopia, then it is worth correcting this with glasses to help relax accommodation. We have found that most overcorrections resolve spontaneously with an excellent long-term outcome. If overcorrection persists, then we have found that performing a unilateral medial rectus muscle recession tends to correct the esotropia. Under corrections are more difficult to manage. If there is significant under correction then more surgery can be performed on the muscles that have already been operated on, depending on how much has been done to the muscle. If there is no scope for further surgery on the operated muscles, then the patient is managed as an intermittent exotropia with a dose of surgery depending on the angle of the strabismus.

SL: My under-corrections will go on to have one or two muscles surgery, typically on the unoperated horizontal rectus muscles. Typically, if I have an overcorrection, I will recess the previously resected muscle. It is rare for me to have large persistent over-corrections however those people are usually unhappy and have diplopia. I would consider re-operating in a few months if no improvement is measured. If there is any improvement in the over-correction, I try to convince the patient to wait for more surgery as an exotropic drift is so common.

FV: We begin trial with glasses followed by patching and then surgery.

K: Over correction: Small consecutive Esotropia 10-12 PD is desired. Patient may have diplopia-resolves within 2 weeks, If Esotropia persists for > 2 weeks use of base out prisms is recommended. Persistence after 8 weeks – resurgery- B/LLR advancement. If residual Esotropia for near only- Bifocals, miotics or base out prisms or bimedial recession with Faden

Under correction: A small residual esotropia (<15 PD) should be managed by non-surgical measures such as orthoptics. Patients with a residual esotropia over 15 PD in the first postoperative week will probably not improve. These patients require additional surgery. It is better to wait 8-12 weeks before re-operating on the residual esotropia. If the primary surgery was bilateral lateral rectus recession of 6 mm or less, re-recession of the lateral rectus may be performed. If the primary recession was greater than 7 mm, then perform bilateral medial rectus resections with a conservative approach, as over corrections are common after resecting against a large recession.

Under correction: For Small residual exotropia (20 PD): Nonsurgical treatment should be tried for 6-8 weeks in such cases. However, in cases with large residual exotropia (>20 PD), it is better to wait 8-12 weeks before re-operating. If the primary surgery was bilateral lateral rectus recession of 7 mm or less, re-recession of the lateral rectus may be resorted to. If the primary recession was greater than 7 mm, then perform bilateral medial rectus resections with a conservative approach, as over corrections are common after resecting against a large recession.

SL: I use adjustable sutures less often now than when I was younger. One reason is the difficulty I have found in predicting where the muscle needs to be left before tying it down. I work to leave them about 10-15 ET. I prefer adjustable suture surgery as a routine.

FV: I prefer adjustable suture in adults and most teens. I use hang back surgery for most lateral rectus recessions.

SD: Small over correction is a desirable outcome. If there is over correction up to 20 PD, we should wait and watch as there is high chance of spontaneous resolution. The possibility of postoperative diplopia must be explained to parents of child. Nonsurgical treatment should be tried for 6-8 weeks. If after 6-8 weeks the esotropia persists, then a reoperation should be considered. In case of a limited adduction or lateral incomitance, advancement of the lateral rectus is indicated. Otherwise, bimedial recession is usually the procedure of choice for a consecutive esotropia, especially if the esotropia is greater at near the possibility of lost or slipped lateral rectus muscle should be kept in mind if there is an unusually large over correction with gross limitation of ocular motility on the first postoperative day and surgery should not be delayed in such cases. In adults; up to 25 PD deviation, nonsurgical measures should be tried till 6-8 weeks, if there is no improvement, re-surgery should be planned.

SK: Which is your preferred surgical technique in such cases, adjustable hangback or nonadjustable surgery, Hangback or conventional surgery?

SBO: If the surgical field is convenient and if the patient is a child I prefer conventional sutures. In adults, I use adjustable suture surgery as a routine.

MR: I use adjustable sutures in all adults and most teens. I use hang back surgery for most lateral rectus recessions.

FM: In children we do not use adjustable sutures. We tend to perform recessions using a Hangback technique and conventional resection surgery although we are now starting to think about the use of publications.

SK: Any indications for Hangback surgery? What are the advantages and disadvantages of Hangback Surgery in such cases?

SBO: Large lateral rectus recessions in a small eye is an indication for me for hang back surgery as well as the
ones who will undergo adjustable suture surgery. Too large recessions carry the potential risk of anterior migration of the lateral rectus muscle but this is usually a problem in restrictive strabismus cases. In IXT we did not face anterior migration in any of the reoperations. The major advantage of hangback surgery is to avoid scleral sutures in close proximity with the macular area. We use adjustable suture surgery in adult patients as a routine procedure and we have not faced any problem about the effect of surgery.

**MR:** Lateral rectus recession being performed. I do use conventional surgery for some smaller deviations. I tend to use most of the time for safety and facility, recognizing that different tables are necessary. My feeling is that I use this approach with adjustable surgery and thus feel it can be effective at other times. I recognize others are concerned about where the muscle migrates, typically forward, but I find that effect is also fairly predictable.

**FM:** We use hangback surgery on all horizontal muscle weakening procedures. We find Hangback surgery is safer with a lower risk of perforation of the sclera. We are meticulous with our surgical procedure and the knot to ensure that we do not end up with a slipped or lost muscle. To date, we have been very happy with this form of surgery.

**SL:** I would use Hangback surgery for trainees that are just starting out or who are less skilled. For example, if a trainee perforates the sclera, I have them move to hangback surgery until I can see they have improved their technique. I like to clean the muscle capsule as little as possible so the muscles are harder to hang back in that setting. I believe, there is faster healing but the muscles tend to get hung up on the supporting tissue if they are minimally cleaned and therefore they need to be sewn in the desired location.

**FV:** Difficult surgery, when using adjustable sutures and in cases of high myopia we can use hangback technique. Final position of the muscle however may move forward.

**K:** We do hangback recession in high myopes, children, recessions more than 8 mm, and while doing adjustable recessions. Advantages of hangback surgery include minimal chances of scleral perforation, adjustable surgery can be done and less chances of tenons capsule snaring in large recessions. Disadvantages of hangback surgery include Central posterior muscle bowing and Chances of late over-corrections if muscle reattachment lags suture hydrolysis.

**SD:** I perform hangback squint surgery in most of my patients. Advantages of hang-back recession include decreased incidence of globe perforation, shorter surgical time, better exposure of surgical field and decreased amount of surgically induced astigmatism. Disadvantages include unpredictable results in especially large angle deviations, difficulty in performing recessions of more than 7 mm and occurrence of possible late over corrections.

Compiled by:
Dr Savleen Kaur
Senior Research Associate, PGIMER, Chandigarh
Email: mailsavleen@gmail.com
Duane’s Retraction Syndrome: Diagnosis and Management

Nripen Gaur MD, Pradeep Sharma MD

Dr Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

Abstract: Duane’s retraction syndrome (DRS) is a form of complex strabismus. This clinical entity can have a varied presentation and may be a part of syndromic association as well. Numerous theories have been proposed regarding the etiopathogenesis of this clinical entity, however, the clinical findings are best explained by the paradoxical innervations of the lateral rectus secondary to maldevelopment of sixth nerve nucleus. Apart from this, the genetic basis of this disease is also being unravelled with the ongoing research. The diagnosis and management of this disease require an astute observation and surgical planning and the treatment plan must be individualised. In this article, the authors discuss the management strategies for various clinical forms of DRS.

Duane’s retraction syndrome (DRS) is an unusual congenital form of strabismus. This condition was initially described by Stilling (1887) and Turk (1896) and was later made popular by Alexander Duane. Its frequency in the general population of strabismus patients is 1–4%1. The ophthalmic manifestations and associated non-ocular anomalies, as well as the demographic and epidemiologic aspects of the syndrome, have all been extensively studied in affected cases and their relatives. It is suggested that certain characteristics of the syndrome may vary in different populations. DRS is included in a group of disorders called Congenital Cranial Dysinnervation Disorders (CCDDs)2. These are secondary to some neurologic pathology of congenital origin and have a wide spectrum of phenotypic presentation. This wide spectrum results due to either primary or secondary dysinnervation. The concept of CCDDs was proposed in 2002. Recent studies have supported this concept and the focus is now on identifying the genes that cause CCDDs.

CLINICAL CHARACTERISTICS

DRS is characterised by the limitation of abduction and / or adduction along with a narrowing of palpebral aperture and retraction of the globe and variable upshoot and downshoot of the globe on attempted adduction (Figure 1). Apart from these, the patients may also have a face turn to achieve binocularity. Other ocular findings include nystagmus, anisocoria, ptosis, and epibulbar dermoid. Other findings may include congenital cataract, heterochromia and optic nerve hypoplasia. DRS has also been shown to be associated with gustatory lacrimal reflex or crocodile tears. The clinical features may vary case to case and one may see a myriad of presentations. Several syndromic associations of DRS have been noted out of which the most common ones include Goldenhar syndrome, Klippel – Feil anomaly and Wildervank syndrome.

ETIOPATHOGENESIS

A maldevelopment or any insult to the development of the sixth nerve nucleus at 4 to 8 week of gestation has been shown to be the causative factor in the development of this disease process5. The branches of the third nerve, in turn, are redirected to the lateral rectus. This abnormal or paradoxical innervational impulses to the horizontal recti are the basic pathology of this disease. The lateral rectus (LR) may exhibit an array of affection. It may have a normal, subnormal or absent activity along with an anomalous third nerve innervation. In a few cases, even the medial rectus (MR) may have subnormal innervations owing to the redirection of developing nerve fibres to the sixth nerve. Secondary muscle changes may occur in form of contracture of medial rectus and superior rectus as a result of constant esotropia and upshoots respectively.

DIAGNOSIS

The diagnosis of DRS is usually clinical. Molecular genetic testing is being investigated for this disease, however, it remains in infancy. It involves single gene testing using sequential and deletion/duplication analysis of the CHN1 gene4. This disease has been differentiated into three subtypes on the basis of myography by Huber5. (Table 1) Apart from these it can also be classified based on the primary position deviation into eso DRS, exo DRS and ortho DRS. MRI imaging has shown the absence of abducens in many studies6,7.

DIFFERENTIAL DIAGNOSIS

Major differentials of this condition include sixth nerve palsy, Moebius syndrome, infantile esotropia and congenital

Figure 1: showing bilateral eso-DRS in a patient with Klippel-Feil syndrome, (1B, C) showing clinical photograph of the patient depicting short neck and low hairline.
The primary indication for surgery in DRS is the correction of primary position deviation and face turn. Apart from this, surgery can also be done to ward off cosmetically unacceptable globe retraction, upshoots or downshoots which are present because of severe co-contraction of LR and MR on attempted adduction. If binocular vision is present with cosmetically acceptable ocular appearance, surgery must not be undertaken just for improving the abduction limitation.

**ESO DRS**

Cases of classic eso DRS do not have normal LR function, but anomalous action occurs during adduction. These patients have an esodeviation in the primary position and as such, they assume a corrective face turn. The treatment options include asymmetric MR recessions or transposition surgeries in form of vertical rectus transpositions. Asymmetric MR recession can be done in the good fellow eye to produce a fixation duress without fearing adduction deficiency in the good eye. This not only corrects the primary position esotropia but also decreases the likelihood of contracture of the medial rectus in the affected eye through the fixation duress in the right eye which leads to a continuous inhibition on affected eye’s MR. However, before planning any surgery in DRS, it is imperative to check force duction test to rule out MR contracture in the affected eye as a tight MR should always be recessed. One must take care to never resect lateral rectus or medial rectus in the involved eye. Superior rectus transposition (SRT) or vertical recti transposition (VRT) to lateral rectus to achieve an abduction force can be done if there is no anomalous LR recruitment. The combination of SRT and MR recession has been found to be more effective than MR recession or bilateral MR recession at improving abduction while allowing for a smaller recession to align the eyes and eliminate a compensatory head posture. However, in cases with uncorrected severe anomalous LR recruitment, any transposition procedures are not indicated. And in case the MR and LR are both to be operated adding the SRT is fraught with the danger of anterior segment ischemia.

In cases with minimal anomalous LR activity and normal LR abducting force, a MR recession in the affected eye may suffice. Those with anomalous LR activity should be tackled by LR weakening procedures. The abduction deficit can be tackled by transposition procedures like SRT (Figure 2) and VRT, however we prefer a balanced partial VRT to SRT as the latter has risk of vertical incomitance and inducing torsion. In cases with significant upshoot or downshoot, the co-contraction can be countered by the recession of both LR and MR of the affected eye. Another strategy is lateral rectus recession with Y split. In this procedure the LR is split from its insertion to 18-20 mm posteriorly and the two halves are placed at a distance of 20 mm apart. This procedure has been shown to be effective in tackling severe upshoot and downshoot.

**Exo- DRS**

These patients present with a primary position exotropia and may have upshoots or downshoots along with deviation. Such patients can be tackled by a LR weakening in form of supramaximal.

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**Table 1: Huber’s classification of DRS**

<table>
<thead>
<tr>
<th>Types</th>
<th>Innervation</th>
<th>Clinical presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>On abduction – LR innervation absent&lt;br&gt;On adduction – LR innervation present</td>
<td>Abduction limited&lt;br&gt;Adduction limited due to resistance with retraction</td>
</tr>
<tr>
<td>II</td>
<td>On abduction – LR innervation present&lt;br&gt;On adduction – LR innervation present</td>
<td>Abduction may be normal.&lt;br&gt;Adduction limited with retraction.</td>
</tr>
<tr>
<td>III</td>
<td>On abduction – LR + MR innervation present&lt;br&gt;On adduction – LR + MR innervation present</td>
<td>Limited abduction with retraction&lt;br&gt;Limited adduction with retraction</td>
</tr>
</tbody>
</table>

*LR* = Lateral Rectus; *MR* = Medial Rectus

oculomotor apraxia. However, these conditions can be ruled out based on the absence of characteristics like globe retraction, narrowing of the palpebral aperture on attempted adduction and upshoots or downshoots. A simple patch test can rule out the presence of infantile esotropia.

**WORK UP OF A CASE OF DRS**

A complete work up of DRS includes a good history recording along with a comprehensive ocular examination. The patient must also be investigated for the concurrent systemic anomalies which can be a part of this disease. Any abnormal head posture must be recorded. DRS is usually associated with the presence of a face turn that the patient assumes to gain binocularity. The degree of face turn be recorded. Ocular movements must be examined carefully to record any globe retraction and palpebral aperture narrowing on attempted adduction and the presence or absence of upshoots and downshoots must be recorded. The primary, as well as secondary deviations, should be recorded in all the nine gazes. A sensory examination must be done to check for binocularity and stereopsis. Hess charting can be done to record the field of binocular field of vision. It is important to look for any contracture in medial and lateral rectus by doing force duction tests. The presence of anomalous lateral rectus activity can be detected by the help of force degeneration test of Romero-Apis. This test is extremely useful in confirming cocontraction in adduction and the missing LR innervation on abduction and thus helps in deciding the plan of surgery.

**MANAGEMENT**

The primary indication for surgery in DRS is the correction of primary deviation.

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**Figure 2A**: Showing abduction deficit in patient of left eso-DRS tackled by SRT(2B)
LR recession or a peristeal fixation (PF) of lateral rectus to the medial rectus. An LR recession with Y split is also an option in cases with normal LR activity where the upshoots and downshoots are a problem (Figure 3). It may be noted that the LR recession should be substantial to correct the retraction in adduction, as the Y split per se does not affect the retraction. The PF wards off the anomalous LR activity most substantially. Some authors have even advocated extirpation of the lateral rectus! The resultant lack of abduction can be taken care of by transposition procedures to LR. A study done at our centre has shown LR peristeal fixation to be effective surgery to correct the exodeviation, anomalous head posture and improving abduction in Exo DRS and partial VRT in addition to be effective in improving abduction and binocular single visual field.10.

**ORTHODO DRS**

These patients are orthophoric in primary position. These patients may still have symptoms like palpebral aperture narrowing and upshoots which may be cosmetically disfiguring. The treatment in such cases may lead to a consecutive heterotopia and as such should be tackled only by the experienced surgeons. The author tackles such patients by the symmetrical recession of both lateral and medial rectus in the affected eye in absence of co-contractions with the use of adjustable surgery on one muscle to control the risk of undesirable under or over corrections.

**BILATERAL DRS**

Bilateral DRS is seen in 15% cases and most of them are either eso or ortho DRS11. These can be managed on the basis of similar principles as the unilateral cases. Bilateral eso DRS can be managed by bilateral MR recessions. Bilateral exo DRS are quite rare and bilateral LR recessions have been recommended for its management. LR Y split can be added in cases with marked upshoots.

**CONCLUSION**

DRS can have a variety of clinical presentations, thus the treatment should be tailored-made for the patients. (Table 2) An astute clinical observation examination is required to set the treatment goals and planning. The author’s GCPE (observe, confirm, infer, plan, execute) strategy serves well as treatment recipe in the successful management of these cases.

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**REFERENCES**


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**Table 2: DRS management summary**

<table>
<thead>
<tr>
<th>1. Eso DRS</th>
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<tbody>
<tr>
<td>A</td>
<td>Minimal anomalous action of LR (cocontraction) with normal LR action in abduction</td>
</tr>
<tr>
<td>B</td>
<td>Anomalous LR present (moderate cocontraction)</td>
</tr>
<tr>
<td>C</td>
<td>With upshoot and downshoot</td>
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<table>
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<th>2. Exo DRS</th>
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<tbody>
<tr>
<td>A</td>
<td>LR weakening (periosteal fixation/supramaximal recession with pVRT)</td>
</tr>
<tr>
<td>B</td>
<td>Exo DRS with normal LR activity with upshoot and downshoot</td>
</tr>
</tbody>
</table>

| 3. Ortho DRS | Symmetrical recession of LR (with adjustable sutures) and LR with Y split |

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**Figure 3A:** Showing severe upshoots in a patient with eso-DRS tackled effectively by LR recession with Y-split (3B)

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**Correspondence to:**

Dr Pradeep Sharma, Professor of Ophthalmology, Dr. R. P. Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India.
Abstract: Retinoblastoma is the most common intraocular malignancy of childhood. The treatment of retinoblastoma is multimodal. Since the introduction of intravenous chemotherapy in the mid-90's, it has been the most extensively used eye-saving modality of treatment. Periocular and intravitreal chemotherapy have emerged as treatment for recurrent seeds in retinoblastoma. Intra-arterial chemotherapy is an alternative for advanced and refractory retinoblastoma. Radiation in the form of external beam radiotherapy and plaque radiotherapy are also used in refractory retinoblastoma. Enucleation still continues to play a role in advanced cases in eyes with no useful vision. The management of retinoblastoma is thus aimed at not only to save life, but also to preserve eye, and optimize residual vision.

Retinoblastoma is an ocular malignancy with a well-established genetic mutation. RB1 is a tumor suppressor gene, implicated in the genesis of retinoblastoma, located in the long arm of chromosome 13 (13q). Heritable retinoblastoma constitutes 30-40% of all retinoblastomas, while the rest 60-70% are non-heritable. In heritable retinoblastoma, the mutation is present in the germ cell which is carried in every cell in the body, making the patients prone for, apart from retinoblastoma, other second cancers (most commonly pinealoblastoma, osteosarcoma and soft tissue sarcomas)\(^1\). The incidence of retinoblastoma is 1 in every 15000 to 18000 live births\(^2\). There are an estimated 5000 new cases worldwide annually, with India alone contributing to 1500-2000 cases.

CLINICAL FEATURES

Retinoblastoma is usually diagnosed at an average age of 18 months, with 95% of children diagnosed by 5 years of age. Germine retinoblastomas can present as early as first month and sporadic retinoblastomas are detected at an average age of 24 months\(^3\). Retinoblastoma can be unilateral or bilateral. The most common presenting symptom and sign is leukocoria, and strabismus is the second most common sign. The other common clinical features are as listed in Table 1.

A child with a suspicious retinoblastoma is best examined under anesthesia for a detailed fundus evaluation. Retinoblastoma typically manifests as a unifocal or multifocal, well-circumscribed, dome-shaped retinal mass with dilated retinal vessels. Although initially transparent and difficult to visualize, it grows to become opaque and white. In the exophytic growth pattern, the tumor causes diffuse retinal detachment (Figure 1A), and is frequently associated with small subretinal seeds. In contrast, an endophytic retinoblastoma progressively fills the vitreous cavity to cause vitreous seeding (Figure 1B). At times, the tumor maybe a combination of these two growth patterns. Diffuse infiltrating retinoblastoma is a rare pattern of presentation where there is no obvious mass, only a flat retinal infiltration, and is acalcific. It is generally seen in older children, and the incidence is less than 2%.

Patients with advanced tumor can have anterior extension of the tumor with anterior chamber cells, neovascularization of iris and glaucoma (Figure 1C), or an orbital cellulitis-like picture (Figure 1D). Retinoblastoma which has extended outside the confines of the eye is known as orbital retinoblastoma and this can occur when the tumor invades either the optic nerve, or full thickness of the sclera and beyond, and the patient generally presents with proptosis.

GROUPING AND STAGING

The grouping system is for retinoblastoma confined to the eye, where eye salvage is the end-point, whereas the staging system is for predicting survival in patients

Table 1: Clinical features of Retinoblastoma

<table>
<thead>
<tr>
<th>Feature</th>
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<tbody>
<tr>
<td>Leukocoria</td>
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<tr>
<td>Strabismus</td>
</tr>
<tr>
<td>Poor vision</td>
</tr>
<tr>
<td>Red painful eye</td>
</tr>
<tr>
<td>Vitreous hemorrhage</td>
</tr>
<tr>
<td>Phthisis bulbi</td>
</tr>
<tr>
<td>Sterile orbital cellulitis</td>
</tr>
<tr>
<td>Proptosis</td>
</tr>
</tbody>
</table>

Figure 1: Clinical presentation of retinoblastoma (A) Exophytic growth pattern with diffuse subretinal fluid (B) Endophytic growth pattern with diffuse vitreous seeds (C) Advanced retinoblastoma with neovascular glaucoma (D) Advanced retinoblastoma presenting as sterile orbital cellulitis.
with retinoblastoma. International Classification of Retinoblastoma (ICRB) was devised in 2003 and includes both grouping and staging\(^1\). The grouping is based on the tumor size, location, severity and presence of subretinal and vitreous seeds (Table 2).

**MANAGEMENT**

The management of a child with retinoblastoma is aimed at achieving the three sequential goals of life salvage, eye salvage, and optimal vision. The management involves the identification of the tumor group and stage, decision-making regarding the appropriate therapeutic measure, and meticulous follow-up for monitoring the treatment progress and detection of any recurrence.

**IMAGING**

While the diagnosis of retinoblastoma is mostly clinical, ancillary tests like ultrasonography, fluorescein angiography (FA), optical coherence tomography (OCT), computed tomography (CT) and magnetic resonance imaging (MRI) aid in the documentation of the disease and differentiation of pseudoretinoblastomas from retinoblastoma. CT scan also helps to diagnose extraocular extension, while MRI is most appropriate to detect optic nerve invasion and to screen for pinealoblastoma in heritable retinoblastoma.

**INTRAVENOUS CHEMOTHERAPY**

Currently, IVC is the most widely used treatment in India (Table 3). Used as a combination triple drug therapy of vincristine, etoposide and carboplatin, chemotherapy with focal consolidation achieves excellent success rates in the primary management of retinoblastoma. Chemotherapy alone can achieve an impressive tumor control in less advanced cases, with success rates of 100%, 93% and 90% in ICRB groups A, B and C, respectively (Figures 2A-D)\(^4,5,6\). Rates of regression of retinoblastoma and eye salvage with standard triple-drug chemotherapy have been suboptimal for ICRB group D and E tumors.

Periocular injection of carboplatin or topotecan injection results in higher intravitreal drug level. Transscleral penetration of posterior sub-Tenon carboplatin leads to augmented vitreous concentration. High-dose chemotherapy with concurrent periocular chemotherapy can lead to higher eye salvage in group D and E eyes.

<table>
<thead>
<tr>
<th>Table 2: International Classification of Retinoblastoma</th>
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</thead>
<tbody>
<tr>
<td><strong>Group A</strong>: Small tumor</td>
</tr>
<tr>
<td><strong>Group B</strong>: Larger tumor</td>
</tr>
<tr>
<td><strong>Group C</strong>: Focal seeds</td>
</tr>
<tr>
<td><strong>Group D</strong>: Diffuse seeds</td>
</tr>
<tr>
<td><strong>Group E</strong>: Extensive retinoblastoma</td>
</tr>
</tbody>
</table>

**Staging:**

- **Stage 0**: Unilateral or bilateral retinoblastoma and no enucleation
- **Stage I**: Enucleation with complete histological resection
- **Stage II**: Enucleation with microscopic tumor residual (anterior chamber, choroid, optic nerve, sclera)
- **Stage III**: Regional extension
  - A. Overt orbital disease
  - B. Preauricular or cervical lymph node extension
- **Stage IV**: Metastatic disease
  - A. Hematogenous metastasis
    - 1. Single lesion
    - 2. Multiple lesions
  - B. CNS extension
    - 1. Prechiasmatic lesion
    - 2. CNS mass
    - 3. Leptomeningeal disease

*Figure 2: Standard-dose chemotherapy in retinoblastoma (A) A group B eye (B) After 6 cycles of standard-dose chemotherapy (C) A group C eye with focal vitreous seeds (D) After 6 cycles of standard-dose chemotherapy.*
INTRA-ARTERIAL CHEMOTHERAPY

Suzuki and Kaneko described the technique of ‘selective ophthalmic artery infusion’ (SOAI) of the chemotherapeutic drugs in 2004 by the balloon technique. In 2006, Abramson and Gobin pioneered direct intra-arterial (ophthalmic artery) infusion or superselective intra-arterial chemotherapy or “chemosurgery”. The decision to treat with IAC is undertaken in consultation with an ocular oncology team, an endovascular neurosurgeon and a paediatric oncologist. The procedure is performed under general anesthesia using a sterile technique (Figures 3A-D). Through a transfemoral approach, the ipsilateral internal carotid artery is catheterized with a 4F pediatric guide catheter. The arterial anatomy is visualized with serial angiography runs, and the ostium of the ophthalmic artery is super selectively catheterized. Each chemotherapy dose is administered in a pulsatile fashion over 30 minutes.

IAC has emerged as an effective treatment for advanced retinoblastoma. It is increasingly being used in tumors as a primary treatment, especially in unilateral retinoblastoma. It can be used as a secondary therapy for those cases which have recurred or have not responded adequately to IVC. Shields et al observed 94% globe salvage in group D eyes. IAC also seems to be more effective in eyes that have failed to respond to previous therapies.

Table 3: Intravenous Chemotherapy

<table>
<thead>
<tr>
<th>Drug</th>
<th>SD-VEC (≥3 years of age)</th>
<th>SD-VEC (&lt; 3 years of age)</th>
<th>HD-VEC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vincristine*</td>
<td>1.5 mg/m2</td>
<td>0.05 mg/kg</td>
<td>0.025 mg/Kg</td>
</tr>
<tr>
<td>Etoposide</td>
<td>150 mg/m2</td>
<td>5 mg/kg</td>
<td>12 mg/Kg</td>
</tr>
<tr>
<td>Carboplatin</td>
<td>560 mg/m2</td>
<td>18.6 mg/kg</td>
<td>28 mg/Kg</td>
</tr>
</tbody>
</table>

*maximum dose < 2 mg

Indications:
(1) Primary tumor
(2) Recurrent tumor
(3) Recurrent subretinal seeds
(4) As adjuvant therapy in post-enucleation patients with high-risk features (discussed elsewhere)
(5) Orbital retinoblastoma
(6) As palliative therapy in metastatic retinoblastoma

Advantages:
(1) Long-term tumor control
(2) Reduces incidence of pinealoblastoma
(3) Reduces incidence of second cancers
(4) Reduces incidence of systemic metastasis

Disadvantages:
(1) Systemic side-effects including thrombocytopenia, leucopenia and anemia
(2) Allergic reactions to carboplatin and etoposide
(3) Long-term effects include hearing loss, renal toxicity and secondary leukemia

INTRAVITREAL CHEMOTHERAPY

Vitreous seeds are aggregates of tumor cells found in the avascular vitreous, which are relatively resistant to the effect of intravenous chemotherapy due to lack of blood supply. Intravitreal chemotherapy (IVitC) provides a high concentration of the chemotherapeutic drug in the vitreous. Melphalan is now the most extensively used drug to control the vitreous disease in retinoblastoma by a potentially safe technique to perform intravitreal injections to prevent extraocular extension of the tumor (Figure 4A-D). The authors have used topotecan in achieving vitreous seed regression in 36 eyes.

RADIATION THERAPY

Retinoblastoma is a highly radiosensitive tumor, and radiation therapy can be curative. Radiation in the form of External Beam Radiotherapy (EBRT) was the most popular globe-salvage therapy in retinoblastoma before the introduction of chemotherapy in 1990s. Although it is no longer the primary modality of treatment for retinoblastoma due to the associated complications, it is used as a part of multimodal treatment for advanced retinoblastoma.

Episcleral plaque radiotherapy is a form of brachytherapy wherein the source of radiation is placed on the episclera adjacent to the tumor, and the tumor absorbs radiation, sparing...
other healthy ocular tissues from the ill-effects of radiation\textsuperscript{14}. Radioisotopes like Iodine-125 and Ruthenium-106 that emit radiation are used for the treatment of small recurrent retinoblastoma.

**FOCAL THERAPY**

The use of cryotherapy, Transpupillary thermotherapy (TTT), and laser therapy in the treatment of retinoblastoma is for consolidation of the tumor, once it attains a considerably lower volume after chemoreduction. Transscleral cryotherapy involves freezing the tumor under visualization using indirect ophthalmoscopy. In TTT, the hyperthermia generated by infrared radiation at sub-photocoagulation levels destroys the tumor. Photocoagulation using argon green laser (532 nm) delivered with an indirect laser delivery system causes tumor apoptosis.

**ENUCLEATION**

Enucleation is the oldest form of treatment for retinoblastoma, and is still indicated in advanced cases. Unilateral disease with no salvageable vision is best treated by enucleation and the patient can be cured of the disease for life. Enucleation is a simple procedure, although special precautions need to be taken when handling an eye with retinoblastoma to avoid accidental perforation that can potentially cause orbital seeding of the tumor.

An enucleated eyeball is always submitted for pathology to assess for high risk factors (HRF). In a landmark paper by Honavar et al, the need for adjuvant chemotherapy has been emphasized to reduce the risk of secondary orbital recurrence and systemic metastasis\textsuperscript{12}. The incidence of metastasis was 4\% in those who received adjuvant therapy, compared with 24\% in those who did not. Hence when HRF is positive, adjuvant treatment with chemotherapy and/or EBRT is indicated (Table 4). Adjuvant chemotherapy consists of a combination of vincristine, etoposide and carboplatin given 4-weekly for 6 cycles.

**ORBITAL RETINOBLASTOMA**

Orbital retinoblastoma is an advanced form of retinoblastoma seen mostly in developing countries of Asia and Africa. The incidence varies among different countries, and is in the range of 18-40\%\textsuperscript{13}. The presence of orbital disease is generally known to carry a poor prognosis. Orbital disease increases the risk of systemic metastasis by 10-27 times and the mortality rates range from 25 to 100\%. However, with an intensive multimodal management and careful monitoring, patients with orbital disease are known to do well (Figures 5A-D).

**PRENATAL GENETICS**

To prevent transmission of the disease from parents to offspring, genetic testing for germine mutations can be done at specialized laboratories. RB1 is the only gene that is implicated in retinoblastoma. Peripheral blood lymphocytes or tumor tissue, when available, are sampled for the detection of the mutation. Preimplantation genetic testing for carriers of mutation involves the identification of RB1 mutation in a blastomere (8-cell embryo) which is obtained by in vitro fertilization (IVF) technique. The small material is amplified by polymerase chain reaction (PCR) and the blastomeres without the RB1 mutation maybe implanted for a successful pregnancy.

**CONCLUSION**

The management of retinoblastoma revolves around having a sound knowledge of the disease, choosing the best treatment for the patient among the various available options and careful monitoring for recurrences. Retinoblastoma has a very high cure rate, and is best managed in an integrated retinoblastoma clinic under the watchful monitoring of an expert ocular oncologist.

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**Table 4: High Risk Features on pathology where adjuvant therapy is indicated**

<table>
<thead>
<tr>
<th>Anterior segment invasion</th>
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<tr>
<td>Giliary body infiltration</td>
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<tr>
<td>Massive choroidal invasion (invasion ≥ 3 mm in basal diameter or thickness)</td>
</tr>
<tr>
<td>Full thickness scleral extension</td>
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<tr>
<td>Extra-scleral extension</td>
</tr>
<tr>
<td>Retrolaminar optic nerve invasion</td>
</tr>
<tr>
<td>Optic nerve invasion at line of transection</td>
</tr>
<tr>
<td>Combination of optic nerve infiltration till any level (pre-laminar/ laminar/ retrolaminar) and choroidal infiltration (any thickness)</td>
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REFERENCES


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Dr. Subhash Dadeya MD
Secretary – DOS
dadeyass@gmail.com
011-65705229
+91-9868604336
Superior Oblique Palsy: Diagnosis and Management

Rebika Dhiman MD, Anita Ganger MD, Mayank Jain MBBS, Rohit Saxena MD

Dr Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

Abstract: Superior oblique palsy (SOP) is the most common cyclovertical muscle palsy encountered in ophthalmic practice. All cases of acute onset SOP need to be thoroughly evaluated for the underlying etiology. Neuroimaging should be advised in young onset SOP, and/or as deemed necessary by the treating ophthalmologist. Superior oblique surgery is one of the most challenging surgeries. However, when performed with appropriate clinical indication may result in optimal clinical outcome.

Superior oblique palsy (SOP) is the most common cause of ocular torticollis in children, and the most frequent cyclo-vertical muscle palsy encountered in ophthalmic practice. It may be congenital or acquired. Superior oblique muscle (SO) is supplied by the fourth cranial nerve, also known as the trochlear nerve, that is purely motor in function. Superior oblique muscle is primarily an intorter, a depressor and abductor. Hence, a superior oblique palsy causes:

- Excyclotorsion,
- Hypertropia,
- Esotropia of the involved eye.

There is a considerable variation in the treatment of this disorder. High-definition radiological studies have given us a better understanding of the patho-physiology of SOP, influencing its diagnosis and treatment. Hence, in the light of recent literature, this article highlights the current perspective in the diagnosis and management of superior oblique palsy.

The search of published literature for this review article had been completed using Ovid, Medline, Embase, Pubmed over the last 5 decades along with the checking of cross references. English language articles with full text access were included and electronic literature search was performed using fourth nerve palsy, trochlear nerve, superior oblique palsy and management as key words. While reviewing the literature, parameters evaluated were: applied neuroanatomy, etiology, medical and surgical management for trochlear nerve palsy.

Applied Neuroanatomy

Trochlear nerve is the longest and the thinnest of all cranial nerves. Unlike the other cranial nerves, trochlear nerve crosses completely to the opposite side during its course and arises from the dorsal aspect of the midbrain.

Nucleus- The fourth nerve nucleus lies in the lower midbrain at the level of the inferior colliculus, near the midline, just anterior to the cerebral aqueduct. There is one nucleus on each side corresponding to the cell bodies of the neurons comprising each trochlear nerve.

Fasciculus- In contrast to the third nerve fascicles, which course anteriorly, the fourth nerve fascicles course posteriorly around the cerebral aqueduct, decussate in the anterior medullary velum and cross the midline to emerge from the brainstem contralaterally just past the vertical midline.

Subarachnoid space- The subarachnoid segment of the trochlear nerve then courses around the midbrain and passes anteriorly along the edge of the cerebellar tentorium.

Cavernous sinus (Intracavernous)- It then travels in the lateral wall of the contralateral cavernous sinus, below the oculomotor nerve and above the first division of the fifth cranial nerve, on its way to the superior orbital fissure.

Orbit (Infrarorbital)- After entering the orbit through the lateral part of the superior orbital fissure, the nerve passes medially and ends by supplying the superior oblique muscle.

The involvement of the fourth cranial nerve anywhere along its course or of the superior oblique muscle causes superior oblique palsy.

Etiology

- Congenital- Congenital fourth nerve palsies are particularly common in clinical practice due to the slender size and long circuitous anatomical course taken by the developing fourth cranial nerve. High definition magnetic resonance imaging (MRI) studies have identified two groups of congenital SOP. The most frequent type, present in 73% of cases, is a Congenital Cranial Dysinnervation Syndrome (CCDS) where the trochlear nerve is absent and results in secondary atrophy of the superior oblique muscle. The second type has a normal trochlear nerve and size of the superior oblique muscle, but presumably has an abnormal superior oblique tendon (e.g., tendon laxity).

- Traumatic- Trauma frequently causes bilateral SOP due to an impact in the anterior medullary velum, where the two

Figure 1: MRI image of the orbit (coronal section) showing a reduced bulk of the muscle on the affected side as compared to the fellow eye in a case of a congenital left SOP.
nerves decussate. It could be a mild concussion or a direct trauma to the trochlea.

- Idiopathic
- Vascular - Microvascular palsies are thought to occur most often in adults 50 years of age or older with existing vascular risk factors.
- Neuroligic

**CLINICAL FEATURES**

Symptoms - In acquired SOP, patient complains of vertical or torsional diplopia maximum in downgaze. If the chief complaint is torsion, bilateral palsy should be considered. Other non-specific complaints may include asthenopia or cervical discomfort.

**Signs** - Superior oblique palsy is characterized by hypertropia of the involved eye that increases in the opposite field of gaze (i.e., in adduction of the paretic eye), and during ipsilateral head tilt. Consequently, patients maintain a head tilt to the opposite side that enables them to compensate for the hypertropia. The abnormalities in the version movement observed are underaction of ipsilateral superior oblique and overaction of ipsilateral inferior oblique with correlative excyclotorsion.

**DIAGNOSIS**

I. Park's three-step test. Bielschowsky described characteristic changes in the hypertropia with head tilt as a means of diagnosing paresis of the superior oblique muscle. Parks later described his 3-step test that used this phenomenon in a broader sense to help the clinician identify which paretic cyclovertical muscle could cause such a hyperdeviation. For left SOP (Figure 2),

1. Left hypertropia in primary position
2. Hypertropia increases on dextroversion (worse on opposite gaze)
3. Hypertropia increases on left tilt (worse on ipsilateral tilt)

However, several limitations of these tests have become apparent over time. Various studies have indicated the lack of specificity of 3-step test in diagnosing SOP especially bilateral SOP. A recent study has reported 40% sensitivity for Beilschowsky's head tilt test and 24% sensitivity for Parks 3-step test in diagnosing bilateral superior oblique palsy.

II. Torsion - Torsion can be assessed subjectively (Double Maddox Rod, Synoptophore), or objectively (Indirect ophthalmoscopy; Fundus photograph). Important points to consider are:

1. Congenital SOP frequently has little or no measurable subjective torsion.
2. Complain of torsional diplopia or a torsion ≥10 degrees is suggestive of a bilateral SOP.

III. Diplopia charting

IV. Hess/Lees' Charting

V. Oblique traction test (Exaggerated forced duction test) is an important clinical test to identify lax superior oblique tendon in SOP that may be corrected by superior oblique tuck.

VI. Neuroimaging - The decision to perform neuroimaging in patients who present acutely with ocular motor cranial palsy is based on age at presentation, which nerve is involved and pertinent past medical history. All patients <50 years with acute ocular motor cranial nerve involvement need neuroimaging to identify the underlying cause. Although the presence of vasculopathic risk factors in patients >50 is a significant predictor for a presumed microvascular cause, a substantial proportion of patients with other causes were found in a recent study regardless of vasculopathic risk factors. Hence, neuroimaging is now recommended in all patients presenting with acute onset ocular motor neuroopathy.

Bilateral superior oblique palsy: Bilateral SOP should be suspected in a patient with one or more of the following features:

- SOP following closed head trauma
- Subjective complain of torsion
- Alternating hypertropia on head tilt
- V-pattern esotropia
- Chin down head posture
- Objective torsion ≥10 degrees

**CONGENITAL SUPERIOR OBLIQUE PALSY**

Despite its congenital origin, this condition may first come to medical attention during teenage or adult years, when diplopia, asthenopia, or headache develops from a combination of spontaneous decompensation and superior rectus contracture. A characteristic facial asymmetry, especially shallowing of the mid-facial region, is usually seen on the side of the tilt (Figure 4). Table 1 shows the differentiating points between congenital and acquired SOP.
There are significant clinical differences between the two groups of congenital SOP. In the first type with an absent trochlear nerve, a greater percentage have a head tilt, which is present within the first year of life, significant facial asymmetry, and more hypertropia on ipsilateral head tilt. Conversely, the second type of congenital SOP with a normal trochlear nerve and muscle have more overaction of the inferior oblique muscle and higher frequency of dissociated vertical deviation.

DIFFERENTIAL DIAGNOSIS

1. Primary inferior oblique overaction
2. Brown syndrome
3. Skew deviation
4. Thyroid associated ophthalmopathy

MANAGEMENT

Medical management

Treatment initially involves medical management of systemic predisposing factors and conservative measures to obviate symptoms followed later by surgical intervention in non-resolving trochlear nerve palsy. The patient should be evaluated at each follow up with a complete squint work-up, diplopia charting and Hess-charting.

Ischemic neuropathies usually resolve in 6 weeks to 3 months\(^2\) and can be treated conservatively. Underlying systemic conditions should be adequately controlled and managed to prevent further ocular morbidity. Mononeuropathies with aneurysmal compressions or other compressive lesions require neurosurgical referral and intervention.

Oclusion of the normal eye during the period of observation eliminates the troublesome diplopia and past-pointing associated with acute palsies. In children under the age of 6 sustained patching can result in amblyopia, hence close follow-up is recommended\(^23\). Adult patients can safely patch either eye without concern for amblyopia. Ocular motility exercises are sometimes recommended following an ischemic palsy, but there is no clinical data to support the same\(^24\).

In non-recovering or residual trochlear nerve palsies, with a stable deviation for at least 6-12 months, surgical intervention may be considered.

Surgical management

Superior oblique surgery is one of the most challenging surgeries and requires thorough knowledge of anatomy, extensive experience, and appropriate preoperative decision-making in view of possible complications and unpredictable outcomes. Its management has been systematized by Von Noorden’s modification of the Knapp’s classification\(^25,26\). Main indications for surgery in a fourth nerve palsy are a significant head tilt, diplopia, decrease in binocular vision and dissociated vertical deviation. A significant head tilt is the main indication for surgery in children younger than age 5, as it is thought that the uncorrected torticollis will lead to progressive facial asymmetry. The surgical procedure employed aims at achieving adequate alignment of the two eyes in the primary gaze. The goals and limitations of the surgery should be clearly understood by the patient to avoid disappointment. Several staged procedures may be required to achieve an optimal correction. Figure 5 describes the various types of surgical intervention for SOP and their indications.

1. Ipsilateral superior oblique tuck—Several studies advocate the strengthening of the SO muscle as the primary procedure to correct SOP when a lax SO tendon is identified, or when there is significant head tilt. SO tuck is titrated based on the intraoperative tension of the SO muscle. Figure 5A describes the strengthening of the SO muscle with an end point of inducing mild post-operative Brown syndrome, which subsequently is expected to either disappears or becomes insignificant over time\(^27,28,29\). Bhola et al recommended isolated SO tendon for SOP and their indications.

2. Harada-Ito procedure is also a strengthening procedure for SO, employed for the correction of excyclotorsion especially in bilateral SOP (Figure 6). The original method of correction was to anteriorize the anterior half of the SO tendon without disinsertion\(^31\). Fells modified...
this technique in 1974, whereby the anterior half of the tendon was disinserted and moved forward and laterally. Metz and Lerner later reported using the adjustable suture technique with this procedure. Harada-Ito procedure is on the basis that the anterior fibers of the SO contribute to the torsional action while the posterior fibers contribute to the vertical action. It corrects around 10 degrees of excyclotorsion in primary gaze and 15-20 degrees in downgaze.

3. Ipsilateral inferior oblique recession (IOR) can be performed especially in the absence of SO tendon laxity, and in the presence of ipsilateral IO overaction. In their retrospective series, Kaeser et al compared the surgical results of inferior oblique recession alone, with those of inferior oblique recession combined with superior oblique tuck in patients with congenital superior oblique palsy and found that inferior oblique recession alone is an appropriate procedure with a low incidence of iatrogenic Brown syndrome.

4. Anterior and nasal transposition (ANT) of ipsilateral IO (Figure 7). In the absence of significant vertical deviation in the primary gaze but significant torsional diplopia, anterior and nasal transposition of the inferior oblique may be undertaken as described by Stager. This procedure converts the IO muscle from an extorter and elevator in adduction to an intorter and tonic depressor in adduction. Extreme ANT may induce exotropia in the primary position. Adjustable ANT has been described for a case of SOP to obviate torsional diplopia.

5. Recession of ipsilateral superior rectus (SR) can be considered in cases where the superior rectus is tight due to long standing hypertropia.

6. Recession of contralateral inferior rectus (IR) is an effective treatment for superior oblique palsy with hypertropia greatest across the lower fields of gaze (Knapp Class V SOP). The critical surgical step for patients with fourth nerve palsy is to decide whether to perform a single-muscle or two-muscle surgery. The decision depends on the magnitude of hypertropia in primary position and side gaze, and the presence of torsion.

CONCLUSION
Superior oblique palsy is a common occurrence in clinical practice. It is
essential to rule out an underlying etiology in acute onset SOP followed by the adequate referral for the same. One should not hesitate to order neuroimaging at the slightest doubt of an intracranial lesion. Superior oblique muscle surgery may result in good clinical outcomes when performed with appropriate clinical indications and for selected cases. The choice of surgical method should be individualized in order to avoid complications.

REFERENCES

Retinopathy of prematurity (ROP) is an important cause of preventable childhood blindness in our country. The ROP classification, screening and treatment guidelines are well established. High quality neonatal care can prevent ROP and effective ROP screening programs are essential to detect treatable ROP in time. Laser treatment of avascular retina ensures good disease regression. Anti-VEGF drugs have proven useful in selected cases of ROP, though safety concerns still exist. Early referral of advancing ROP for surgical management can give good outcomes. Though it is essential to establish a ROP screening program in every NICU, yet lack of awareness/services for screening and treatment compounded with poor neonatal care is leading to a huge increase in ROP blindness in India.

ROP CLASSIFICATION

The international classification of retinopathy of prematurity (ICROP) group classifies ROP into zones and stages on the basis of location of disease on retina and extent of vascular proliferation.

ZONES OF ROP (FIGURE 1)

- **Zone 1** - Circle drawn from centre of disc with a radius of twice the distance from disc to macula.
- **Zone 2** - From nasal edge of zone 1 to ora nasally and upto equator temporally.
- **Zone 3** - Temporal crescent of retina anterior to zone II.

Typically, smaller the zone, more severe is the disease which occurs as there is a large area of avascular retina. The disease is most commonly seen in zone 2.

STAGES OF ROP (FIGURE 2)

- **Stage 1** - Demarcation line between the vascular and avascular retina
- **Stage 2** - Demarcation ridge between the vascular and avascular retina
- **Stage 3** - Extraretinal neovascularization and proliferation
- **Stage 4A** - Subtotal retinal detachment not involving macula
- **Stage 4B** - Subtotal retinal detachment involving macula
- **Stage 5** - Total retinal detachment

**Plus disease** - It is characterized by dilated tortuous posterior pole vessels, iris vascular engorgement, pupillary rigidity and vitreous haze (Figure 3). Plus disease is a good indicator of the activity of the disease and worsening plus disease can signify disease progression, while reduction of plus disease may signify disease regression. Poor pupillary...
dilation must always raise suspicion of plus disease.

Pre-plus disease- The dilation and tortuosity of the posterior pole vessels is less than that observed in classical plus disease, but is more than normal. Pre-plus disease can progress to plus disease if the disease worsens.

Aggressive Posterior ROP (AP-ROP)- This is a severe variant of ROP which has an unpredictable course and can often lead to rapid progression and blindness. It is characterized by a featureless junction between the vascular and avascular retina, often associated with multiple avascular loops, which can confuse beginners about the disease stage. However, notably plus disease is out of proportion to the severity seen at the junction. A high index of suspicion for APROP should be maintained and it should be treated promptly.

ROP SCREENING

Since ROP has a well-defined course and can occur in many high risk premature babies, an effective screening program is the only way to detect ROP in time and ensure prompt treatment. It is essential that every neonatal intensive care unit (NICU) should have a screening programme.

When to Screen?- Most screening programs now follow the National Neonatology Forum (NNF) of India ROP screening guidelines, which suggest that we should screen all preterm babies <34 weeks gestational age or <1750 g birth weight. Bigger babies of 34–36 weeks gestational age or 1750–2000g birth weight should also be screened if they have risk factors for developing ROP.

Whom to Screen?- NNF suggests that first retinal examination should be performed at not later than 4 weeks of age or 30 days of life. In infants born <28 weeks gestation age or <1200 grams birth weight, they should be screened by 2-3 weeks of age, primarily to detect APROP.

Where to screen?- The ideal setting is always the NICU where the baby is admitted or comes for routine follow-up visits. The pediatrician can monitor vitals of the baby and deal with any emergency. However, in many eye hospitals, screening is being done on an outpatient basis as well - though in such a scenario emergency equipment should be kept ready and a pediatrician on call should be available.

How to Screen?- Screening is a stressful procedure for the baby and proper preparation is necessary. It is very important to take written informed consent from the parents and provide them details of the procedure. The babies are kept fasting for 30-60 minutes prior to the procedure. Pupillary dilatation should be started an hour prior to screening using a combination of tropicamide (0.5%) and phenylephrine (2.5%) drops, instilled 2-3 times about 10 minutes apart. Beware of the non-dilating pupil, which should raise suspicion of severe plus disease and APROP.

Screening is performed under topical anesthesia, assisted by a pediatric speculum and pediatric scleral depressor. The procedure is to examine the anterior segment (especially for pupillary dilation and tunica vasculosa lentis) and then the posterior segment, starting with the posterior pole (to look for plus disease) and then identify the disease in all clock hours of the retinal periphery. It is very essential to properly document all the ROP findings correctly in a record sheet.

Further follow-up screening is done at 1-2 weeks (or lesser) depending on the zone, stage of ROP and post-conceptional age. Screening is usually stopped when complete retinal vascularization occurs (mature retina) or complete regression of ROP is noted.

Role of Digital ROP screening?- Digital screening has become popular now due to the relative ease of performing ROP screening by trained technicians/nurses, without the need for an ophthalmologist. The main advantage is photo documentation which can be used for training, referral and tele-screening purposes. These hand-held cameras (e.g. Retcam; Figure 4) provide wide field angle retinal imaging up to 130 degrees, and can be easily transported to areas like NICU or OT. It is also easier to explain to the parents the disease of their child using these photos. It provides advantage of performing fundus fluorescein angiography7 in select cases as well.

Its diagnostic accuracy compares well with indirect ophthalmoscopy, and many successful tele-screening programs are running globally where screening can be performed at peripheral centres and experts in a reading centre can opine about treatment and follow-up. The Stanford University Network for Diagnosis of Retinopathy of Prematurity (SUNDROP) program9 and Karnataka Internet Assisted Diagnosis of Retinopathy of Prematurity (KIDROP) program9 are successful examples of such digital tele-screening programs.

How long to follow-up?- Long term follow up is essential, regardless of whether they have undergone treatment, as these babies are at risk for developing visual disorders later in life such as strabismus, amblyopia, myopia and cataract. The importance of follow-up and visual rehabilitation must be stressed to the parents.

ROP TREATMENT

It is very important to treat ROP in a timely manner to prevent blindness. A screening programme is the only way to detect treatable ROP in time.

When to treat?- The Early Treatment For Retinopathy of Prematurity Cooperative Group (ETROP) study10 divided ROP into two types.
• Type 1 ROP - defined as zone I, any stage ROP with plus disease; zone I, stage 3 ROP without plus disease; or zone II, stage 2 or 3 ROP with plus disease. This disease needs prompt treatment.

• Type 2 ROP - defined as zone I, stage 1 or 2 ROP without plus disease or zone II, stage 3 ROP without plus disease. This can be followed up and treated if it converts to type 1 ROP.

How to treat?: Various treatment options are available for management of ROP like laser, Anti-VEGF drugs and vitreoretinal surgery.

A) Laser Treatment – Laser treatment by indirect ophthalmoscopic delivery is the gold standard for treatment and is very effective in causing disease regression (Figure 5). The aim of treatment is to ablate entire anterior avascular retina up to ora serrata in a near confluent burn pattern. Some disadvantages of the laser treatment are pain experienced during the procedure, visual field loss in lasered area, induction of significant myopia and sometimes poor results in zone 1 aggressive disease. Moreover, laser treatment of ROP requires properly trained ophthalmologist, who can perform the procedure quickly and adequately, with minimal systemic stress to the baby.

B) Anti-VEGF drugs - Recently anti-VEGF drugs like Bevacizumab and Ranibizumab have emerged as a popular treatment modality, especially for Zone 1 APROP. The Bevacizumab Eliminates the Angiogenic Threat of Retinopathy of Prematurity (BEAT ROP) study has shown the benefits of these drugs in zone 1 stage 3 plus disease. Intravitreal Anti-VEGF drugs (usually given in half adult dose) help to quickly dilate the pupils, reduce neovascularization inducing effective disease regression, and allows normal vascularization to proceed into the periphery. Some possible indications where these drugs may be used are –
• In cases with severe iris new vessels, plus disease and poor pupillary dilation preventing laser treatment - these drugs can ensure early pupillary dilation allowing complete laser treatment.
• In severe zone 1 disease with extensive neovascularization which might not be controlled by laser alone - these drugs can serve as useful adjunct to laser treatment.

In very small zone 1 disease with macular non-perfusion - these drugs might provide benefit of progressive vascularization into the periphery, improving visual fields. Though it is a quick procedure, with benefits of better visual field and lesser myopia, its long term safety still needs further evaluation. There is proof of systemic absorption of the drug, and it might prevent normal angiogenesis of organs which are still developing in preterm babies. There is still lack of evidence about the correct dose and number of injections which can be given. There is a high chance of late recurrence, which might need prompt treatment, thus longer follow-up is warranted. Thus, its indiscriminate use is not warranted and proper consent and case selection are essential.

C. ROP Surgery - Advanced ROP with retinal detachment (stage 4-5) needs surgical intervention. This further highlights the importance of timely detection and treatment to prevent disease progression. Timely surgery in stage 4 ROP can have excellent outcomes, therefore in cases of progressive ROP, despite the laser/ AntiVEGF prompt early referral to a higher surgical centre is necessary. ROP surgery needs high-risk informed consent, preoperative clearance by pediatric and anesthesia team (since these babies are often very sick and preterm) and experienced surgical teams to perform surgery on very small eyes. An experienced anesthesia team and reliable NICU backup are very essential.

Though previously socalled buckling was performed, now modern 25/27G lens sparing small gauge vitrectomy has good anatomical and functional results in stage IV ROP. Lens sparing vitrectomy is the procedure of choice in stage 4A and subtypes of 4B. Surgery in bilateral cases of Stage 5 ROP is performed with the hope of obtaining some navigable vision, but often the results are poor.

CONCLUSION

Retinopathy of prematurity is rapidly emerging as an important cause of childhood blindness in India. The poor awareness about ROP among the pediatricians, ophthalmologists and parents, is leading to an uncontrolled epidemic of ROP blindness. It is essential that an ROP program is established in every NICU with partnership between ophthalmologists and pediatricians. We need to remember that the child is not born with ROP, therefore timely screening, referral and laser treatment are the key to success in preventing ROP-related blindness.

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Diagnosis and Management of Abducens Palsy

Geetha Srinivasan MS
ICARE Eye Hospital and Research Centre, Noida, India

Abstract: Sixth cranial nerve palsy is the most common of the extra ocular muscle palsies. The incidence is reported as 11.1/100000. A patient with an isolated 6th Nerve palsy presents with a complaint of diplopia in primary position or on abduction of the paretic eye, depending on the magnitude of limitation of movement. While in the elderly the cause is very often vasculopathies like diabetes, hypertension and atherosclerosis, viral and unknown etiologies are common in young adults and in children brain infections and neoplasms like a brainstem glioma must be ruled out.

Sixth cranial nerve palsy is the most common of the extra ocular muscle palsies. The incidence is reported as 11.1/1000001. It is also the most common cause of an abduction deficit, though restrictive and inflammatory conditions among others can also cause abduction deficits. The long tortuous course of the nerve intracranially from the pons to the orbit and the peripheral part of the nerve near the clivus makes it a target of intracranial lesions and high intracranial pressure. (Figure 1). So a lateral rectus paresis calls for a thorough evaluation of the fundus to rule out papilloedema. Sixth nerve palsy can occur at any age. However, the causes and the approach to management of sixth nerve palsy vary with age.

The nerve may be involved in isolation or may be affected with other structures along its path leading to a variety of syndromes.

Clinical Characteristics

A patient with an isolated Sixth Nerve palsy presents with a complaint of diplopia in primary position or on abduction of the paretic eye, depending on the magnitude of limitation of movement. In mild lateral rectus paresis, the patient may have an intermittent diplopia with esophoria. The symptoms are usually of acute onset. However, very young children rarely complain of double vision. The parents observe that the child’s eye is squinting or that he is shutting one eye during visual tasks.

A history of trauma, systemic illness, fever, vomiting, weakness of arms or limbs, neurosurgery, headache etc should be elicited. This is important because involvement of the nerve due to neurological and other systemic causes should be excluded before arriving at a diagnosis of idiopathic palsy. An idiopathic palsy should always be a diagnosis of exclusion.

Evaluation of a Case

After recording visual acuity and assessing the pupils, cover and alternate cover testing would reveal an esotropia more for distance than near, with limitation of abduction in the paretic eye (Figure 2). The greatest esotropia is seen when the patient attempts to abduct the paretic eye. As mentioned, in mild cases only a latent deviation (esophoria) is seen.

Attempting to abduct the eye maximally increases the innervational effort and may widen the palpebral fissure. (Figure 3) There may be a V pattern strabismus due to decrease in abduction and unopposed action of medial rectus in downgaze.

In bilateral involvement, there may be no change in the esodeviation with either eye fixing.

A compensatory head posture to negate or decrease the diplopia is often seen. The face turn is typically towards the direction of the paralyzed muscle and away from the action of the lateral rectus. (Figure 4). However, this is not a consistent sign and may be missed if not specifically looked for. This is associated with horizontal diplopia, which is worse on levo/
dextroversion, depending on the eye affected. Involvement of other extraocular muscles should be looked for.

Hess and diplopia charting corroborate the clinical findings and the former quantifies the weakness in the nerve (Figure 5a, 5b).

**Causes:** While in the elderly the cause is very often vasculopathies like diabetes, hypertension and atherosclerosis, viral and unknown etiologies are common in young adults and in children brain infections and neoplasms like a brainstem glioma (commonest) must be ruled out. (Table 1 and 2). Posterior fossa tumors like ependymoma and medulloblastoma, clivus chordoma, parasellar tumors with cavernous sinus involvement are some of the causes of sixth nerve palsy in children.

Newborns can have a transient 6th Nerve paresis, which usually resolves spontaneously (Figure 6). The incidence has been reported to be 0.4% by Galbraith et al. However, Duane’s retraction syndrome and infantile esotropia must be kept in mind and excluded while evaluating these infants. Overall, the incidence according to etiology has been reported variously as Idiopathic 8-30%; Miscellaneous 10-30%; Trauma 3-30%; Aneurysm 0-6%; and Ischemia 0-36%.

**Non-Isolated sixth nerve palsy:**

The sixth cranial nerve can be affected along its course in the brainstem, subarachnoid space, petrous apex, cavernous sinus or orbit. Involvement in each of these locations would have different manifestations that help localise the lesion.

**Brainstem**

1. Millard Gublar syndrome: associated with ipsilateral 7th nerve paralysis and contralateral hemiparesis.
2. Raymond syndrome: Sixth nerve paresis with contralateral hemiparesis
3. Foville syndrome.

**SUBARACHNOID SPACE**

Any condition that raises intracranial pressure can lead to downward displacement of the brainstem and compression of the nerve at its exit from the pons and in Dorello’s canal. This manifests as a sixth nerve palsy. This is quite commonly seen in pseudotumor cerebri with associated papilloedema and visual field changes.

<table>
<thead>
<tr>
<th>Causes</th>
<th>Neoplasms</th>
<th>Systemic disease</th>
<th>Neurological disorders</th>
<th>Others</th>
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**Table 1: Causes of 6 Cranial nerve palsies in adults**

<table>
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<tr>
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<td>metastasis</td>
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<td>Pontine glioma</td>
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<td>Diabetes mellitus</td>
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<tr>
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<tr>
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<td>Inflammatory causes</td>
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<td>Miscellaneous causes</td>
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**Table 2: Causes of 6 Cranial Nerve Palsy in children**

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<tr>
<th>Infections</th>
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<td>Influenza virus</td>
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<td>Meningoencephalitis</td>
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<td>Infections</td>
<td>Streptococcal disease</td>
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<td>Astrocytoma</td>
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<td>Chondrosarcoma</td>
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<td>Head trauma</td>
<td>Craniopharyngioma</td>
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<td>Head trauma</td>
<td>Ependymoma</td>
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<td>Gioma</td>
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<td>Pinealoma</td>
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<tr>
<td>Head trauma</td>
<td>Neuroblastoma (secondary)</td>
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<tr>
<td>Systemic disease</td>
<td>Leukemia</td>
</tr>
<tr>
<td>Systemic disease</td>
<td>Aneurysm</td>
</tr>
<tr>
<td>Systemic disease</td>
<td>Arterio-venous malformation</td>
</tr>
<tr>
<td>Neurological disorders</td>
<td>Arnold Chiari malformation</td>
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<tr>
<td>Neurological disorders</td>
<td>Demyelinating disease</td>
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<tr>
<td>Neurological disorders</td>
<td>Increased intracranial pressure</td>
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<td>Neurological disorders</td>
<td>Migraine</td>
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<td>Neurological disorders</td>
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<td>Immunological</td>
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<tr>
<td>Neurological disorders</td>
<td>Infectious</td>
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</tbody>
</table>

**Pretros apex**

Inflammatory or infective lesions of the petrous apex due to ear infections lead to 6th Nerve palsy with ipsilateral hearing loss with facial nerve involvement. Trauma, Cerebellopontine tumors, nasopharyngeal carcinoma with spread to cavernous sinus are some other conditions that involve the 6th nerve palsy (Figure 7).

**Cavernous sinus:** The palsy is unlikely to be isolated if the cause is a lesion in the cavernous sinus. The other oculomotor nerves, carotid oculosympathetic plexus are also involved in most cases.
Orbit: Orbital lesions are associated with proptosis. There may be associated congestion of the conjunctival vessels and chemosis. Other oculomotor nerves may also be involved.

Recurrence: Isolated palsies are known to recur; though the incidence of recurrence is not high. In one series of 782 patients more than 50 years, 7 patients showed a recurrence. The main causes identified in those patients were mass lesions in the parasellar or petrous apex cavernous sinus regions.

Benign 6th nerve palsy of childhood may be recurrent either on the same side or contralateral eye. However, any recurrence should be viewed with suspicion and an indicator of an etiology that can increase morbidity and mortality. Imaging is mandatory.

INVESTIGATIONS

The work up of a patient with an isolated 6th Nerve palsy depends on the age and the clinical circumstances of the patient.

OPHTHALMOLOGIC

Hess charting: Serial charting gives us an objective record of the amount of weakness in the lateral rectus muscle.

Diplopia charting: Shows maximum uncrossed horizontal diplopia in the direction of paretic lateral rectus.

While these tests are usually not needed to establish the diagnosis, having a baseline helps to monitor progress objectively.

Imaging of the orbit (when indicated): In cases where orbital restrictive pathology is suspected. (Figure 8).

SYSTEMIC

In the elderly, the following investigations are indicated.

1. A hemogram with sedimentation rate
2. Blood pressure,
3. Blood sugar
4. Serum lipid profile

   • In patients, older than 50 years with isolated 6th Nerve palsy and diagnosed vasculopathic illness, imaging is not indicated at initial visit
   • In patients who do not show any improvement in 3 months or if there are associated neurological symptoms, whatever the age, imaging is indicated. Non vasculopathic causes need to be ruled out in these cases. It is important to remember that even patients with history of vasculopathic disease may be harbouring a neoplastic or infiltrative disease. Any progression of the abduction deficit even in a patient with a microvascular disease should be investigated thoroughly and imaging is to be done.
   • In younger patients with acquired isolated 6th Nerve palsy imaging is indicated in all and urgent imaging is indicated if there is papilloedema.

DIFFERENTIAL DIAGNOSIS

Though the diagnosis is often straightforward, few conditions may mimic 6th Nerve palsy.

1. Duane’s retraction syndrome Type 1 needs exclusion. Assessment of extraocular movements to detect the limitation of movement can pose a challenge in infants sometimes. Of course, the lid and palpebral changes...
characteristic of DRS confirm the diagnosis.

2. Infantile esotropia with crossed fixation. Patching one eye will demonstrate normal abduction.

3. Divergence paralysis: Seen in the elderly usually, though it has also been reported in young adults (Figure 9).

4. Fibrosis syndromes: Not very difficult to differentiate, because they are usually associated with ptosis and limitation of other extraocular movements.

5. Thyroid eye disease usually presents with inferior and medial rectus involvement. Blood tests can confirm diagnosis.

6. Myasthenia gravis can be a mimic. Variable ptosis is characteristic. Laboratory tests of Anticholine esterase antibody and Repetitive nerve stimulation test can aid diagnosis.


8. Spasm of near reflex: Is usually associated with miosis of pupil, diminution of vision and variable esotropia with pseudo myopia and nystagmus.

9. Inflammatory disorders involving the globe/orbit: Cause restrictive myopathy.

Follow up: Many cases show spontaneous recovery, without the need for imaging. Initially the follow up is 2 weekly and later monthly till it resolves. The standard dictum is to wait for 6 months to allow spontaneous recovery if the aetiology/pathology so allows.

**TREATMENT**

**Nonsurgical**

**Oclusion:** In children prevention of amblyopia by patching one eye is advised. The patching may be alternate, allowing for eye movements and prevention of secondary contracture of the medial rectus in the paretic eye. In adults patching of either eye is advised to prevent diplopia.

**Botulinum toxin (BTX-A):** While awaiting recovery, Botulinum toxin injection in the antagonist medial rectus can be given as a temporizing measure. It is usually given after about one month of the onset. Besides tackling the problem of diplopia, it is also believed to prevent contracture of the medial rectus. However, it does not appear to influence the percentage of patients requiring surgery. Transient vertical deviations are seen in 5-30% and ptosis in 15-30% after BTX-A.

**Prisms:** In those patients who have a small residual esotropia, prisms are a useful option either as a temporizing option while awaiting recovery in patients who need the binocularity.
or later as a permanent option when recovery is suboptimal after 6 months. Fresnel prisms can also be tried in those with larger deviations.

**Surgery:** Aim of surgery is to maximize binocular vision, treat the diplopia by improving abduction and comitance and improve the compensatory head posture. Surgical options in a lateral rectus palsy are dictated by the amount of force that the muscle can generate.

**Preoperative assessment:** Forced Duction test and Active Force generation test should be done pre-and intraoperatively to assess the residual lateral rectus function.

Based on these tests, surgery is planned.

1. Horizontal muscle surgery: LR resection if esotropia is <20PD and LR function is good with no MR contracture.

   If LR function is moderate (-1 to -2) Unilateral MR recession (maybe adjustable) and LR resection +/- contralateral MR recession if >40PD esotropia in primary position.

2. Vertical transpositions If LR function is poor (-3 to -4) with or without a tight MR, full tendon /partial tendon SR + IR transposition with or without botulinum toxin to MR. The aim of the tendon transfer is to reduce the esotropia in primary position. Abduction deficit generally persists even after surgery.

The split tendon transfers (Hummelshaim and Jense procedure) have the advantage of minimizing the risk of anterior segment ischemia. The Hummelshaim procedure is not a preferred choice because of complete disinsertion of the vertical recti and higher risk of anterior segment ischemia. Some advocate Superior Rectus (SR) transposition with MR recession as an equally effective procedure.

**REFERENCES**

Diagnosis and Management of Third Nerve Palsy

Birsen Gokyigit MD

HSU Beyoglu Educational and Training Eye Hospital, Istanbul, Turkey.

Cranial nerve third innervates five extra ocular muscles ‘medial rectus, inferior rectus, inferior oblique, superior rectus and levator palpebra’ and carries parasympathetic out flow to the ciliary ganglion which are responsible for pupillary constriction and accommodation. Palsy may be partial or complete.

Clinical signs in complete palsy are ptosis, restriction of ocular movement except for abduction and the pupil may be fixed and dilated. Eye is found in exo hypotropia position with incyclotorsion. In partial palsies, we notice ptosis and upgaze restriction in superior division palsy and restriction in downgaze, restricted adduction and pupil dilatation in inferior division palsy.

Partial paralysis may give different clinical signs according to the localization of the lesion or according to the involved muscle. Even though, the palsy can occur in inferior or superior branch of the third nerve, sometimes it affects only one muscle. Because of this reason partial palsies may mimic many other types of strabismus. Aberrant regeneration or oculomotor synkinesis, loss of accommodation, loss of near light reflex can occur in old cases.

Common Causes of 3rd nerve palsy are congenital, vasculopathy, neoplasm and traumatic in children. Important parameters in history include diabetes mellitus (DM), hypertension, hyperlipidemia, herpes zoster, leukemia and new onset headache.

TREATMENT

Basically, the treatment of nerve palsies involves treating the primary problem. If the primary pathology is treated, motility turns to normal very soon1-4. Botulinum toxin injection as a surgical procedure can help us both in acute and chronic cases. In acute cases, injection is to be made to antagonist muscle to induce temporary palsy and/or to prevent secondary fibrosis. In old cases, injection is to be given to antagonist muscle pre-operatively or intra-operatively to gain more effectiveness or to release the contracture; or to be performed in post operative stage to help to enlarge the binocular visual field (Picture 1).

SUPERIOR RECTUS MUSCLE PALSY

Superior rectus palsy can be seen in congenital cases or secondary to trauma. In acute cases, we have a defective elevation in abduction with normal elevation in adduction. There can be elevation deficit both in abduction, adduction and primary position in chronic cases. Overaction of ipsilateral inferior rectus and contralateral inferior oblique and encycloptorsion is also noted. There is hypotropia with ptosis in primary position and absence of Bell phenomenon amongst other findings. There is ocular torticollis as the face is turned upward, the chin is elevated the head is inclined toward the sound eye with pseudo ptosis.

Differential diagnosis includes: Double Elevator Palsy (DEP), mechanical causes like contracture, fibrosis, high myopia, myositis, endocrine orbitopathy and blow-out fracture; and structural abnormality in differential diagnosis.

There are several choices for treatment. These are inferior rectus recession, superior rectus resection, vertical transposition of horizontal rectus muscles, inferior rectus faden operation and nasal transposition of superior oblique5,6.

Recent treatment modality is novel inferior rectus recession which doesn’t cause lower lid retraction operation - introduced in Brazil & USA in 2006 and in literature in 20137,8. During the operation, we separate ciliary artery with few superficial muscle fibers and connective tissues from deeper muscle fibers.

Figure 1: The sectoral diagrams of novel inferior rectus recession operation. 1- Surface Conjunctiva 2- subconjunctival tissues with adjacent structures 3a-10% surface layer of tendon, 3b. - 90%deeper layer of tendon, 4a- 10% Surface layer of IR muscle, 4b- 90% deeper layer of IR muscle, 5- Sclera,- 6- Disinserted 90% deep layer.
are performed for aberrant innervation of the inferior oblique muscles surgery of the superior oblique and myectomy medial rectus muscle (MR) and tenotomy lateral rectus muscle (LR), resection of the when needed. Total tenotomy of the Lateral rectus recession may be added medial rectus resection and vertical recti divergence, Duane’s syndrome type II and supranuclear disorders.

Surgical treatment choices include medial rectus resection and vertical recti transposition to the medial rectus area. Lateral rectus recession may be added when needed. Total tenotomy of the lateral rectus muscle (LR), resection of the medial rectus muscle (MR) and tenotomy of the superior oblique and myectomy of the inferior oblique muscles surgery are performed for aberrant innervation treatment.

**MEDIAL RECTUS MUSCLE PALSY**

There is exotropia with adduction deficit in isolated medial rectus paralysis. It is very rare but if suspected needs a very careful examination to determine the difference from similar diseases like internuclear ophthalmoplegia, synergistic divergence, Duane’s syndrome type II and supranuclear disorders.

Surgical treatment choices include medial rectus resection and vertical recti transposition to the medial rectus area. Lateral rectus recession may be added when needed. Total tenotomy of the lateral rectus muscle (LR), resection of the medial rectus muscle (MR) and tenotomy of the superior oblique and myectomy of the inferior oblique muscles surgery are performed for aberrant innervation treatment.

**INFERIOR RECTUS MUSCLE PALSY**

In a patient with inferior rectus palsy, we note down gaze limitation especially in abduction, incyclotorsion and hypertropia. Etiology might be either congenital or following trauma. Vascular diseases and myasthenia gravis are the other reasons.

For surgical treatments choices are inferior rectus muscle resection alone, inferior rectus plication, superior rectus recession, reverse Knapp procedure and/or inferior oblique muscle anterior transposition.

**INFERIOR OBLIQUE MUSCLE PALSY**

Inferior oblique (IO) muscle paralysis is also seen very rarely and has to be differentiated from Brown syndrome. In IO palsy, there is negative force duction with a pattern strabismus. Limited elevation in adduction, marked superior oblique overaction and positive head tilt test are their other clinical signs. Etiologies are similar with the other muscle palsies.

Surgical plan is created according to amount of hypotropia. If hypotropia is less than 10 pd in primary position, ipsilateral superior oblique tendon expander operation or superior oblique split tendon elongation operation is performed. If hypotropia is over 10 pd in primary position, we need to add contralateral superior rectus recession operation to above surgeries. Inferior oblique plication may be effective in selective cases.

In the presence of partial or weak nerve third palsy rules are: to perform maximum LR recession, strengthening MR (i.e. weakening the strong muscle, strengthening the weak muscle). If these muscle surgery is ineffective; to apply transposition procedure. Lastly if there is ptosis, ptosis surgery in patients after checking the Bell phenomenon.

**COMPLETE 3RD NERVE PARALYSIS**

In complete cranial third palsy, paralyzed eye is in a position of abduction, slight depression, and intorsion. Additionally, there is ptosis of the upper lid, small degree of proptosis, dilated and non-reactive pupil and paralysis of accommodation in affected eye. Aberrant regeneration may occur in congenital or old cases.

Simple recession-resection procedure was preferred in the past. But the results of these operations were not successful over time, because of the ineffective medial rectus. Adding superior oblique nasal transposition and long duration fixation suture has improved the results.

Recent surgical approach is defined by two goals: 1) weakening of the lateral rectus and fixation of medial rectus to the medial orbital tissues 2) Using the power in lateral rectus to change the globe position to more nasal. Good or the healthy eye surgery can be done for enlargement of binocular visual field.

Myectomy of lateral rectus, or disinsertion and orbital fixation of lateral rectus with medial rectus resection; or to use titanium t- plate as platform for globe alignment operation are the other choices for surgical treatment which were published ten-fifteen years ago.

Inferior nasal transposition of lateral rectus operation is another choice. Similarly splitting the lateral rectus and transposing the arms to medial side of the globe near the vortex veins was tried but the results were not good. Lateral rectus transposition to the nasal pole of superior rectus muscle or medial rectus nasal orbital wall fixation have limited effectiveness.

We published the technique of medial transposition of Y split lateral rectus introduced by me 7 years ago. In this operation, lateral rectus arms are transposed just near medial rectus insertion. Surgical procedure involves “Splitting the lateral rectus muscle for all length posterior from the insertion and reattach the superior arm to the medial
rectus superior pole and inferior arm to the medial rectus inferior pole to the its insertion. Resect or plicate the medial rectus muscle maximally if necessary. During the technique, adjustment is applied only on operation table. The Figure 2 from original paper, shows post operative inferior and superior aspects. One of our young cases preoperative, early postoperative and following 4 years of operation-late postoperative gaze positions photos seen in the Picture 2. Picture 3 shows another patient’s pre-and post operative gaze positions. Adjustable suture, augmentation suture and simplified modifications were published with very successful results following our publication. In later studies MRI findings have also been published. The technique has choroidal effusion as transient complication but it improves in a couple of weeks. We had only one case who had similar complaint. His diagnosis was not choroidal effusion but posterior scleritis and it can occur following any reoperation of strabismus.

CONCLUSION

Complete oculomotor paralysis can be corrected surgically. Perfect results can be achieved with proper approach in selected patients at the appropriate time. The excellent result expected from the eye surgeon is that the eyes are in a primary position, with the widest possible binocular viewing area and good cosmetic appearance. Complications should always be anticipated and must be followed meticulously.

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Correspondence to:
Dr. Birsen Gokyigit
Assoc. Prof. of Ophthalmology
HSU Beyoglu Educational and Training Eye Hospital, Turkey
Lost Muscle in Strabismus Surgery – How to Handle the Problem?

Seyhan B. Özkan

Adnan Menderes University Medical School, Department of Ophthalmology, AYDIN, Turkey

Summary: Lost muscle is accepted as one of the most serious complications in strabismus surgery. Prevention is far more important than the treatment since the majority of these develop due to the problems with the surgical technique. If an extraocular muscle is lost, it must be searched with respect to the tissues and if it cannot be found transposition surgery may be considered. Botulinum toxin A is helpful to reduce the contracture of the antagonist muscle where necessary.

Loss of an extraocular muscle (EOM) may occur related to strabismus surgery, pterygium surgery and retinal detachment surgery. Orbital surgery, endoscopic sinus surgery and trauma are the other main causes of this problem. Lost muscle is most commonly seen during strabismus surgery and it is a nightmare complication for the surgeon. The term “lost muscle” is something to be questioned as the EOM is still there in its anatomical location. Most of the time the problem is the “lost surgeon” rather than the “lost muscle”. The former is much more harmful for the patient indicating an inexperienced surgeon without a good knowledge of the anatomy of the surgical field. In this article, etiology and types of lost muscle, preventive measures, clinical presentation and methods of management will be evaluated.

CLINICAL TYPES
The types of lost EOM are evaluated in four groups:
- Slipped EOM within its capsule
- Cut and lost EOM
- Broken (snapped) EOM
- Inadvertently transected EOM

SLIPPED EOM WITHIN ITS CAPSULE
The extraocular muscle slips within its capsule. The sutures do not include the muscle – tendon fibres but only the capsule. The recognition of the slipped muscle is at the postoperative stage (Figure 1). It is demonstrated that stretched scar tissue formation may cause consecutive deviations and the stretched scar tissue and slipped EOM may be confused in diagnosis. In stretched scars there is minimal or no limitation of versions, less separation of the tendons from sclera, and thicker appearance of the scar segments. Magnetic resonance imaging (MRI) demonstrates the EOM in distant location to limbus in slipped EOM, in contrast to stretched scar formation.

CUT AND LOST EOM
The muscle is lost during the surgery. If the problem occurs during recession there is a long muscle with its tendon. However, if it occurs after resection, there is a short muscle without tendon. If the latter is the case it is much more difficult to re-localize the muscle (Figure 2).

Figure 1: In slipped extraocular muscle the sutures do not include the muscle tendon fibres but only the capsule.

Figure 2: The EOM muscle may be lost during recession where there is a long tendon and the muscle. If the EOM muscle is lost after resection, there is a short muscle with almost no tendon.

BROKEN (SNAPPED) EOM
The break usually occurs at the tendon – muscle junction and the major etiological factor is excessive pulling of the EOM; which is called as “pulled - in - two syndrome”.

INADVERTENTLY TRANSECTED EOM
The most frequently transected EOM are the inferior rectus and lateral rectus muscles that develop during inferior oblique surgery. Superior rectus transection may occur during superior oblique surgery. The main etiological factor for this problem is blind hooking.
careful removal of the anterior Tenon’s capsule, avoiding dissection over the EOM area without hooking the muscle, avoiding unnecessary posterior dissection of fascial structures and avoiding the use of serrated instruments for sutures are the major preventive measures to reduce the risk of a lost muscle in strabismus surgery. In reoperations, the risk of lost muscle is higher because of the adhesions and scar tissue. Especially in previously recessed muscles there are some adhesions at the original insertion area and dissection of those adhesions carry the risk of inadvertent cutting of the muscle tissue. Severing the tissues from the sclera must be done after hooking the muscle and locating the EOM insertion.

**CLINICAL FINDINGS**

A large over or under correction with marked limitation of ductions or marked incomitance in the field of action of the operated EOM are the signs that suggest a lost muscle problem. A mild exophthalmos or widening of the palpebral fissure may develop in some of the cases.

Medial rectus (MR) muscle is the most commonly lost EOM. Unlike other rectus muscles, MR has no fascial attachments with the oblique muscles. When the MR muscle is lost, it recoils easily posterior to the penetration of the Tenon’s capsule.

If a lost muscle is suggested, MRI may be helpful for locating the muscle. In case of any doubt about the innervation of the posterior fibres, kinematic MRI is required to demonstrate the functional capacity of the posterior fibres. Forced generation test can easily be performed in an office setting and saccadic velocity measurements give reliable results if available.

**MANAGEMENT**

When lost muscle is recognized during surgery, the first rule is not to fall into panic and to be calm. The anaesthesiologist must be warned about the problem and the anesthesia must be deepened in order to prevent any contraction of the muscle that may cause the muscle to move backwards further. The anaesthesiologist must be informed not to use atropine as the oculocardiac reflex may be helpful to differentiate the EOM tissue.

Using high magnification is very useful to differentiate the tissues. The major rule is not to make more harm to the patient. The most devastating problem that may occur in lost muscle is penetration of the posterior Tenon’s capsule by blind fishing and addition of the fat adherence syndrome which converts the problem into a much more complicated one. If it is not possible to find the EOM, transposition might be considered. However, it is not an easy decision to consider transposition during primary surgery if two recti have already been operated because of the anterior segment ischemia risk.

When a lost muscle is suspected in post operative period, early intervention is essential to increase the chance for finding the lost muscle. However, in some cases the patient may present at a later stage with an excessive inflammatory reaction. In such cases, it is preferable to wait for six weeks till the inflammation resolves. It is stated that the contracture of the unopposed antagonist can occur as early as 2 weeks’ time. We found botulinum toxin A (BTXA) very effective to prevent the contracture of the antagonist during the waiting period for surgical intervention. It also enables the eye to stay at primary position during wound healing. It also enables the surrounding soft tissues to attach to a more anterior point to the globe representing another additional advantage of BTXA injection (Figure 4).

During search for a lost muscle the globe should not be pulled towards the opposite direction (Figure 5). Retropulsion of the globe increases the chance for locating the EOM. During the search for a lost muscle, all the clues of fascial structures that may be attached with the muscle must be used. It is a useful strategy to find the oblique muscles first if the lost muscle is superior, inferior or lateral rectus muscles. Their intermuscular septa may hold the rectus muscles. Trypan blue dye 0.1% may be helpful to differentiate some tissues and it stains fibrotic tissue, muscle sheath and tendon but not muscle fibers.

One of the most common mistakes is to search for the muscle at the scleral site. However scleral plane may take the surgeon even closer to the optic nerve. When an EOM is lost it moves posteriorly within the Tenon’s capsule at the orbital site, so the surgeon must be aware of the correct plane to look for the muscle (Figure 6).

If the problem is a slipped muscle within the capsule the search must be done through the muscle capsule (Figure 7). The relocation of MR muscle...
is reported between 10% and 86% in lost muscle and 92% and 100% in cases of slipped muscles.\textsuperscript{6,7,9,10}

If standard conjunctival approach fails some alternative methods are reported. These methods are transcutaneous medial orbitotomy, trans nasal endoscopic approach, transcaruncular incision and anterior orbitotomy.\textsuperscript{11,12} (Figure 8). These methods require experience and their use instead of muscle transposition surgery must be considered with their potential risk of complications.

It is known that a lost and found muscle may function even years after surgery. However, these muscles may have contracture and it may not be possible to suture them at their original insertion.

**CONCLUSIONS**

In conclusion, for a correct management and not to be the “lost surgeon” a good knowledge of the anatomy is essential. The majority of lost muscle complications develop because of the mistakes about the basic rules of surgery. Blind fishing during attempts to find an EOM may result with penetration of posterior Tenon’s capsule and development of adherence syndrome which is much worse than a pure lost muscle. BTXA has a significant role in management of lost muscle by preventing the contracture of the antagonist. An acceptable outcome is possible in most of the cases either with retrieval of the lost muscle or muscle transposition procedures.

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Correspondence to:
Dr. Seyhan B. Özkan,
Adnan Menderes University Medical School,
Department of Ophthalmology, AYDIN, Turkey
Squint Surgery is not just “Cosmetic”, but Functional and Restorative Procedure

Sumit Monga MS, FRCS
Centre for Sight Group of Eye Hospitals, Delhi, India

Summary: Times are changing, so are the health care trends. One of the paradigm shifts in healthcare management has been the facility of medical insurance to the patients for their healthcare needs, reducing their financial burden. However, when it comes to squint surgery, the insurance claims are often declined, the reason assigned being that squint surgery is purely cosmetic and hence the costs need not be reimbursed. Needless to say, this deters many patients, especially children, to avail the benefits of squint surgery. This view is not much different from the general mindset prevalent in society that squint needs to be tackled mainly for “cosmetic” reasons. This short article attempts to dispel the notion of squint surgery being a cosmetic procedure alone.

Strabismus, abnormal ocular alignment, is one of the most common ocular problems in children, affecting 5% of the preschool population and around 4% of adult population. The ocular misalignment can be vertical, horizontal, torsional or a combination of the three. Strabismus can be treated with conservative therapy such as glasses, prisms, patching and/or orthoptic exercises, with a majority of the cases eventually requiring correction with eye muscle surgery. When the patient does not suffer from diplopia, squint surgery has long been regarded as a cosmetic procedure. The term ‘cosmetic’ originated in the early 17th century as a noun denoting the art of beautifying the body. The English variant of the term derives from French ‘cosmétique’ or Greek ‘kosmetikos’. Both of these mean “to arrange” or “adorn”.

The primary goal of strabismus surgery is to improve the alignment of the visual axis of the eyes. It offers both functional and restorative benefits (Table 1). In children, untreated misalignment of eyes impairs visual acuity development and leads to amblyopia. In other words, the alignment of eyes helps in visual development and binocular coordination of eyes and helps to maintain it. The corrective benefit of squint surgery on the facial appearance of affected individuals is noticeable; however, the widespread belief that corrective squint surgery is a purely cosmetic procedure is incorrect. This is because, first, it seeks to restore the normality disrupted by a disease process rather than to add glamour. This directly influences the social and psychological issues directly related to their condition, which impact all aspects of their lives: education, social relationships, employment prospects, and self-esteem. Therefore, a distinction needs to be made between squint surgery, which restores normal eye alignment (and thus aids normal functional performance of eyes) and other cosmetic procedures, available for enhancing one’s look. In the figures below, different case scenarios are presented to highlight the functional and restorative benefits of squint surgery (Figures 1 to 5).

In a study performed by Olitsky et al., patients without strabismus had their photos digitally altered to create the appearance of strabismus. Study subjects were then asked to judge the photos and rate personality characteristics and

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Figure 1: Functional and restorative benefit. (a) This young boy, with infantile esotropia, was often teased in school and was often not invited to his friend’s birthday parties, as he looked different. (b) Squint surgery was able to restore his normal appearance. He also developed some stereovision. An earlier intervention (around 6 months of age) would have, probably, resulted in greater functional benefit.

Figure 2: Restorative benefit. (a) This gentleman, with infantile esotropia, was dissuaded from squint surgery in childhood, by relatives and family physician. (b) Alignment restored by squint surgery in adulthood. But, binocular vision deficit was irreparable.

Figure 3: Functional and restorative benefit. (a) Case of decompensated intermittent exo-tropia with asthenopic symptoms. (b) Restoration of alignment with resultant increase in binocular cooperation, decrease in asthenopic symptoms and increase in binocular field of vision.
**Table 1: Benefits of Squint Surgery**

**Functional**
- Re-establish binocular single vision (Eliminate diplopia, if any)
- Promote stereo-acuity
- Improve ocular strain related to intermittent deviations
- Decrease visual confusion
- Improve abnormal head posture
- Restore peripheral visual field

**Restorative**
- Restores normal eye alignment and facial appearance (positive psychosocial impact)

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**CONCLUSIONS**

To conclude, the perception that squint surgery, which is usually regarded as cosmetic and as such may be less deserving of funding in health care systems, needs to be changed. The current evidence shows that the effect of successful strabismus surgery has long lasting effects both on the functional and psychological well being of the patients and far beyond a cosmetic effect. It would therefore be sensible to redefine the so called cosmetic squint surgery as “reconstructive/restorative” with the aim being to restore parallel visual axes, which is the “normal” condition.
Skew Deviations: The Brain Throws a Wild Pitch

P.K. Pandey MD, Annu Joon, Divya Kishore, Kartik Rana, Anupam

Guru Nanak Eye Center, New Delhi

Evolutionary Underpinnings

Light from the sky above and gravity from below are two physical constants quintessential for any form of life to exist on this planet. Balance and verticality are pivotal for survival. The two vestibules and the two eyes serve as balance organs in lockstep to maintain verticality in the roll plane, lest the animal fall and be preyed upon. In lower lateral eyed animals, the two-parallel monocular visual systems are fully crossed like pyramidal tracts, the right side of brain receiving inputs from the left eye and vice versa. There is panoramic vision but no binocularity. In lateral eyed animals, a body tilt along long axis generates rotation of both eyes that is vertical, right ward tilt causes right eye to be lower in space than the left eye resulting in compensatory skew deviation with upward rotation of lower eye and vice versa. The conjugate torsional eye movement known as ocular counter roll is most prominent in frontal eyed binocular animals like man. Skew is thus a vestigial remnant of primitive otolithic righting reflex released under pathologic conditions.

What is a Skew / Ocular Tilt Reaction?

For balance, bilateral weighted otolithic inputs are leveraged to modulate extraocular muscles (EOM) and postural tonus in the roll plane. Skew originates from imbalance of vestibulo-ocular reflex (VOR) projections from the two utricles to the ocular motor neurons. The utriculo-ocular reflex, manifests as vertical divergence, head tilt and conjugate torsion to the same direction constituting 'Ocular Tilt Reaction'. Skew is the vertical component which may be most prominent or very subtle. Ocular tilt reaction can be physiological / pathological, tonic / phasic, complete / incomplete. Any component may be present or absent in differing proportions giving diverse presentation to skew deviations, for example, only extorsion may be present, intorsion may be trace or even absent as we shall see later.

Vestibulo-Ocular / Otolithic Pathways

The otolithic pathways serving tonic ocular tilt reaction arise mainly in the two utricles and to a much lesser extent in saccules. The VOR pathways for phasic ocular tilt reaction arise in the anterior (ASC) and posterior (PSC) semicircular canals and serve in a push-pull lockstep fashion. The ASC activates ipsilateral superior rectus (SR) and contralateral inferior oblique (IO) while inhibiting ipsilateral inferior rectus (IR) and contralateral superior oblique (SO). The PSC activates ipsilateral superior oblique and contralateral inferior rectus and inhibits ipsilateral inferior oblique and contralateral superior rectus. The VOR pathways travel in the vestibular nerve synapse at the level of vestibular nuclei in the pons, cross over to the opposite side at the level of 6th cranial nerve nucleus and ascend in the medial longitudinal fasciculus till riMLF Interstitial nucleus of Cajal is very vital for vertical VOR pathways. From riMLF they ascend to thalamus and to vestibular cortex in the temporal lobe, there are many connections to the cerebellum at the level of vestibular nuclei (Figure 1).

The three planes and three dimensions of Skew Deviations - There are three planes - roll (occipito-frontal), pitch (interauricular), and the yaw (vertical) plane. OTR is manifested in roll, whereas laterally alternating skew is in the pitch and acute acquired comitant esotropia accompanying neurological disease in the Yaw plane respectively.

Etiopathogenesis

Lesion anywhere along the otolithic pathways can give rise to motility abnormalities. Fellow travellers often help localise the site, as the site age and activity of the lesion are the sole determinants of the clinical presentation in neurological disease, etiology of the lesion has a very little role in the presentation. The OTR can be ipsiversive or contraversive depending upon location of the lesion, is it before or after crossing of VOR pathways at the level of vestibular nuclei in the pons. A selective lesion of the ASC produces intorsion of contralateral eye and hypotropia of ipsilateral eye due to underaction of ipsilateral
SR and overaction of contralateral SO and resultant hypertropia (HT) simulating SO palsy. A lesion selectively involving PSC produces hypotropia and extorsion of ipsilateral eye, may simulate IO palsy. A lesion involving both ASC and PSC pathways will result in accentuated vertical (Skew), conjugate torsion and head tilt to the same direction constituting complete OTR. OTRs can be thus quite variable complete/ incomplete, asymmetrical/ symmetrical, comitant or incomitant, tonic / phasic, physiologic / pathologic.

**ETIOLOGIES**

Skew deviations share diverse etiologies from most benign to most malignant including tuberculomas, stroke, infarction, tumors, trauma, hydrocephalus, Arnold Chiari malformations, epilepsy and drugs. The list is exhaustive.

**SYMPTOMS**

Skew deviations rarely cause symptoms by themselves since there is tilt in the subjective visual vertical (SVV), which patient is not aware. There could be symptoms due to fellow travellers. Rarely symptoms like vertical/ horizontal diplopia, oscillopsia, ataxia may be experienced.

**DIFFERENTIAL DIAGNOSIS**

Incomitant OTRs can be confused with superior / inferior oblique palsies, inferior division 3rd nerve palsy, oculary myasthenia gravis, thyroid eye disease, mitochondrial cytopathies, chronic progressive external ophthalmoplegia, the list is long. Vertical deviation and conjugate torsion disappear in supine position in OTR but not in oblique muscle palsies and upright / supine test should be performed in all such cases suspected of harbouring a skew. An incomitant OTR due to selective damage to otolithic pathways corresponding to contralateral ASC may produce a superior oblique palsy (SOP) sporting a positive Bielschowsky test, meeting the diagnostic criteria for SOP. Head tilt test is positive as the vestibule on the opposite side (hypotropic side) is damaged, leading to worsening of HT on ipsilateral tilt to hypotropic side. A similar damage to ipsilateral PSC pathways can result in inferior oblique palsy. The hypertropic eye is intorted due to overaction of the superior oblique and hypotropia eye is extorted due to overaction of the inferior oblique in OTR as opposed to oblique muscle palsies. Associated 3rd, 4th and 6th nerve palsies could coexist concealing the clinical picture. Similarly, asymmetrical visual inputs to two eyes in infancy can lead to breakdown in binocularity and lead to dissociated vertical deviations (DVD), primary oblique muscle overaction and pursuit asymmetry in infantile strabismus. See-saw nystagmus and DVD can be conceptualised as inverse OTRs. Other conditions must be distinguished by attendant clinical clues.

The Roll Plane, the Ocular tilt reaction- The lesion could be anywhere along vertical VOR pathways, Fellow travellers may include internuclear Ophthalmoplegia (paresis), pretectal syndrome, ataxia, vertigo, vertical upbeat / downbeat nystagmus etc.

**ILLUSTRATIVE CASE REPORT**

A 50-year-old man with history of atrial myxoma, presented with left ischemic INO, left HT and right head tilt. Objective ocular torsion revealed conjugate intorsion of left eye, extorsion of right eye, constituting classical OTR (Figure 2). Patient complained of oscillopsia, vertigo and occasional vertical and horizontal diplopia. Three step test was positive for left superior oblique palsy suggesting selective damage to otolithic pathways corresponding to right ASC. The vertical tropia and conjugate torsion improved significantly in supine position alluding to the diagnosis of an OTR rather than superior oblique palsy. An ischemic micro- infarct along the left MLF was suspected, however MRI did not reveal any lesion. Patient was followed up for recovery as large number of skews may recover with time, however significant recovery did not take place. Surgery in such cases is congealed as weakening an inferior oblique for example may add to intorsion and worsen his symptoms, surgery on the vertical recti is only viable option.

The Pitch Plane, The Laterally Alternating Skew deviations-Manifests as reversing HT in lateral gaze positions. The abducting eye is usually higher but reverse may also be true. The forward body pitch is sub served by ASC and backward body pitch by PSC, the corresponding otolithic pathways are segregated in the brainstem. A selective damage to bilateral ASC pathways will result in the dominance of PSC pathways and result in bilateral abducting HT, A pattern and intorsion whereas a dominance of ASC pathways will catapult into adducting HT, V pattern and extorsion, head tilt test will be negative in symmetrical injury. There may be a primary position HT if damage is asymmetrical.

**ILLUSTRATIVE CASE REPORT**

A 20-year-old girl presented with occasional vertigo and vertical horizontal diplopia, she betrayed bilateral abducting HT, PP HT of left eye, downbeat nystagmus, bilateral intorsion and A pattern. MRI brain showed large tuberculoma at the level of the pons having bilateral involvement. She showed good response to anti-tubercular treatment. (Figure 3).

Acute Acquired Comitant Esotropia (AAE) accompanying serious neurological disease – Posterior
fossa lesions such as Arnold Chiari malformations and vermal tumors in particular may cause AACE as otolithic pathways sub serving horizontal rather than vertical vergence are affected. The esotropia is greater in distance and motor fusion is grossly compromised, some may also have associated divergence palsy. This horizontal form of skew deviation may have its physiological substrate in translational VOR. Associated fellow travellers like nystagmus, ataxia, papilloedema should be carefully looked for and neuro imaging ordered, lest a life-threatening condition be missed.

ILLUSTRATIVE CASE REPORT

A 12-year-old boy presented with acute onset horizontal diplopia, isolated AACE with V pattern and conjugate torsion, there was no papilloedema, MRI brain revealed disseminated CNS tuberculomas. Patient responded to antitubercular therapy and esotropia as well as conjugate torsion resolved completely with treatment (Figure 4). Skews may simulate comitant/incomitant strabismus including SOP/IOP/AACE. Mostly acquired, skews share diverse etiology from most benign to most malignant. Look for signs of CNS involvement, include evaluation of subjective / objective torsion & head position dependent changes as 4th & 5th step in motility assessment. SUMMARY - Keep low threshold for suspicion, order neuroimaging and neurology opinion. Wait for resolution. Surgical options different with variable outcomes as hypertropic eye is intorted and hypotropia eye extorted. Muscle sequelae may not be prominent. Better understanding of skew deviations may enhance our understanding of acquired strabismus and give us insight as to when to order neuroimaging and seek neurology opinion. Ophthalmologist may be the first one to have a close encounter with a skew as initial presentation and thus be held responsible for a missed diagnosis and/or botched up treatment. Skew should be kept in the differential diagnosis of acquired strabismus or motility disorder.

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Pediatric Cataract: Evaluation and Management

Manisha Mishra, Anju Rastogi, Shweta Dhiman

Guru Nanak Eye Center, New Delhi

Summary: Management of congenital and childhood cataracts remains a challenge. Increased intraoperative difficulties, low scleral rigidity, high vitreous pressure, propensity for increased postoperative inflammation, changing refractive state of the eye, very high incidence of visual axis opacification and a tendency to develop amblyopia, all add to the difficulty in achieving a good visual outcome in the pediatric patient.

Pediatric cataract is an important cause of vision impairment and blindness because of the impact on child’s development, education, future work opportunities as well as quality of life. Management of congenital and childhood cataracts remains a challenge. Low scleral rigidity, increased elasticity of the anterior capsule, and high vitreous pressure are among the major obstacles that interfere with the highly demanding cataract surgery in children. Increased postoperative inflammation, a changing refractive state, higher re-surgery rate and an inherent risk of amblyopia makes the management further challenging.

Pre-operative evaluation

1. History - The first complaint is often leukocoria, strabismus, nystagmus, inability to recognize mother or closing of eyes in bright light. A detailed history is taken that includes asking about
   • Age of onset and duration of symptoms
   • Antenatal history including that of any drug intake and fever with rash
   • Perinatal history of low birth weight, pre-term delivery is important as bilateral cataracts are more common in these patients
   • Family history of congenital cataract and consanguinity
   • History of any trauma or previous surgery
2. Ocular examination-
   a) Vision:
      i. Preverbal children - vision can be assessed in the following ways
         • Fixation pattern
         • Optokinetic nystagmus
         • Preferential looking tests like Tellers acuity test and Cardiff acuity test
      ii. Verbal children - vision can be assessed in the following ways
         • Landoff C test
         • HOTV Test
         • Tumbling E test
         • Allen’s test
         • Snellen’s letter chart
   b) Anterior segment biomicroscopy - done to assess the size, location, density of the opacity. Corneal diameters and intraocular pressure have to be measured with a tonopen or Perkins hand held applanation tonometer.
   c) Dilated fundus examination
   d) Ultrasound B Scan if fundus is not visible
   e) Ultrasound A Scan
   f) Presence of strabismus and nystagmus
3. Laboratory investigations - It is important in cases of bilateral cataract
   a) Hemoglobin
   b) TORCH
   c) Plasma calcium/ phosphorus/ phosphatase
   d) Urine reducing substance

Systemic evaluation - A detailed systemic evaluation is to be done as congenital cataract can be associated with galactosemia, Lowe’s syndrome, Alport syndrome, Down’s syndrome etc.

Timing of cataract surgery

a) In visually significant opacities –
   • Unilateral cataracts should be operated before 6 weeks of age
   • Bilateral cataracts should be operated before 8 weeks of age
   • In bilateral symmetric cataracts second eye should be operated within 1 week of the first eye
b) When there is significant asymmetry, the worse eye is operated first
   c) In visually non-significant opacities
      • Surgery should be avoided for 1-4 years due to difficult pre-operative and post-operative assessment and management.

Prognostic factors

Pediatric cataract causes decreased vision and interferes with the normal development of the child. Later age of onset, early cataract surgery, choice of aphakic correction, good parental motivation and amblyopia management are of utmost importance in achieving good outcomes.

A number of factors may influence the surgical outcome adversely which include presence of a unilateral cataract (as they are detected at a later age), nystagmus, strabismus, any associated ocular defects like microphthalmos, persistent fetal vasculature and poor parental compliance.
**IOL POWER CALCULATION**

Calculation of accurate IOL (Intraocular Lens) power is a major hurdle for long term care of patients undergoing pediatric cataract surgery. Accuracy of the calculated IOL power is affected by the short eyes and the steep keratometric values. Selecting a fixed-power lens for implantation into an eye that is still growing requires a complicated decision-making process. Implantation of an IOL at the emmetropic power risks a significant myopia at ocular maturity. However, if we aim for early hyperopia with the expectation that the eye will become emmetropic during adulthood, then this may predispose the child to amblyopia. Both approaches have pros and cons; the best solution probably lies in finding a compromise.

**AXIAL LENGTH MEASUREMENT**

A small difference in axial length measurement may result in clinically significant residual refractive error. For example, a 0.1 mm difference in axial length may result in 0.25–0.75 D difference in IOL calculation. In shorter eyes of children, the error could be magnified to 14 D/mm. Axial length can be measured using ultrasonography or optical biometry. A-scan ultrasonography is the conventional method for measurement of axial length in children. Ultrasound can be performed using application or immersion techniques. The application technique may introduce a measurement error in recorded axial length (shorter axial length obtained) by the slight indentation of the corneal surface and the lack of fixation in a patient under anesthesia. With the immersion technique, care must be taken to ensure that the ultrasound beam is perpendicular to the retina by ensuring that the retinal spike is displayed as a straight, steeply rising echo spike. A recent prospective study on 50 cataractous eyes of 50 children (mean age, 3.87 ± 3.72 years) by Trivedi and Wilson found that the axial length was shorter in eyes with contact ultrasonography compared to immersion ultrasonography (21.36 ± 3.04 mm vs. 21.63 ± 3.09 mm respectively; P < 0.001).

Partial coherence interferometry for axial length measurement has been shown to be very accurate but requires patient cooperation, and thus, may not be a viable option in infants, young children, and uncooperative children.

**KERATOMETRY**

In a young child keratometric values are obtained under general anesthesia using a hand held autokeratometer. Keratometry steeply reduces in the first 6 months, i.e., -0.40 D/month, -0.14 D/month in the next 6 months, and -0.08 D/month in the 2nd year. Corneal curvature reaches the adult range at about 3 years of age. Keratometry readings without spectacle are preferred though technically difficult, as keratometry with the spectacles is known to deform the globe and give an unreliable reading.

**ACCURACY OF IOL CALCULATION FORMULAE**

The Hoffer Q and Holladay 2 formulæ were reported to be more accurate for shorter adult eyes. The accuracy of different IOL calculation formulæ in children has been previously studied. Neely et al. used SRK II, SRK/T, Hoffer Q and Holladay 1 to recalculate the IOL power for 101 patients who had undergone surgery at mean age of 4.8 years and found that the mean prediction error was 0.3 ± 1.5 D. There was no significant difference in the predictability of the lens power between these four formulæ and the newer theoretical formula did not out-perform the older regression formulæ. In this study, there was greater variability in shorter eyes (less than 19 mm), and in children less than 2 years old. The least variable was the SRK II and the greatest was Hoffer Q. Another study found that Hoffer Q was more predictable than the other formulæ especially in young children (<2 years old) and in shorter eyes (<22 mm).

Dahan et al. have suggested under correcting biometry reading by 10% in children between 2 to 8 years. For children younger than 2 years, perform biometry and under-correct by 20% or use the axial length only. IOL power suggested for 21 mm is (22.00D), 20 mm (24.00D), 19 mm (26.00D), 18 mm (27.00D) and for 17 mm axial length 28.00D.

Enedy et al. recommended post op goal as described in the following (Table-1).

**INTRA OPERATIVE APHAKIC REFRACITION**

Intra operative aphakic refraction can be used to calculate the IOL power. Spherical equivalent from the aphakic refraction can singularly predict the emmetropic IOL power without the need for axial length measurement and keratometry.

**CURRENT SURGICAL TECHNIQUE**

Pediatric cataract surgery is evolving with advances in microsurgical techniques. Currently there is an increasing trend toward IOL implantation in children with evidence of better visual outcomes in infants managed with IOL implantation. The aim of the surgical technique is to provide a long-term clear visual axis by preventing development of VAO or secondary membrane.

Lensectomy and vitrectomy: This was introduced in 1975 and can be done by 2 routes. The pars plana route was advocated by Peyman in 1978 while limbal route was described by Calhoun and Harley in 1980. This method leaves lack of adequate capsular support for PCIOL implantation. It can be done in cases where IOL implantation is contraindicated like microcornea and microphthalmos, uveitis, glaucoma, aniridia and persistent hyperplastic primary vitreous.

Lens aspiration with IOL implantation: Current surgical steps are described under the following heads:

1. **Wound construction**: Tunnel clear corneal incisions have replaced limbal incisions. The preferred location is superior rather than temporal. Brown et al recommended tight suturing in pediatric patients as vitrectomy done in pediatric patients collapses the sclera making the already elastic sclera even less rigid.

2. Increased intravitreal pressure: The ophthalmic visco surgical devices such as 1.4% sodium hyaluronate allows the surgeon to neutralize the positive vitreous pressure. A bolus of thioptopente and hyperventilation helps reduce the intravitreal pressure.

3. Anterior capsular management: The anterior capsule in children is more rigid.
elastie and has more tensile strength. A cohesive OVD is recommended to facilitate capsulorhexis and prevent its peripheral extension. Capsular staining aids in better visualization and also makes it less elastic in children. An adequate size and location of the ACCC is of utmost importance in long term centration of the IOL.\(^{29}\)

Gimbel and Neuhann in 1992 introduced the technique of continuous curvilinear capsulorhexis which is now the gold standard for use in pediatric cataract patients. Ideally, the technique provides for any size of smooth, circular capsular opening with a strong capsular rim that resists tearing even when stretched during lens material removal or IOL implantation.\(^{30,31}\)

Vitrectorhexis is an alternative to manual ACCC. Dr. M. Edward Wilson demonstrated the use of a vitrectorhexis to create the opening in the anterior capsule in a case of infantile cataract. While a manual capsulorhexis is typically used in pediatric cataract patients more than a year old, in very young infants, a vitrector provides improved control and a smooth rhexis.\(^{30,31}\)

Radiofrequency diathermy capsulotomy, developed by Kloti and colleagues in 1984, has been used as an alternative to CCC for cataract surgery in children.\(^{32}\)

The Fugo blade has also been recently introduced as a plasma knife that can be used to perform an anterior capsulotomy.\(^{33,34}\)

Hydrodissection and lens matter aspiration: Multi-quadrant hydrodissection (at least three quadrants) is the preferred method, following which bimanual lens aspiration is completed to remove the lens matter.\(^{35}\)

Intraocular lens implantation: Whether IOL implantation should be done as a primary procedure has been debated by pediatric ophthalmologists for several years. The infant aphakia treatment study (IATS) was designed to answer this question. This was a prospective randomized multi-centric trial comparing infants (<7 months) who underwent cataract surgery for unilateral cataract with either IOL implantation or were left aphakic and were fitted with a contact lens. The main outcome variable was visual acuity at 1 year and 4 ½ years of age. The investigators also looked at complications, re-surgery rates and strabismus and stereo-opsis and compliance to occlusion. IATS found that there was no difference in visual acuity at either 1 year of age or at 4 ½ years between the two groups. But alarmingly adverse events like membrane proliferation into the visual axis, corectopia were almost 10 times more common in infants with IOL implantation compared to aphakic infants.\(^{35}\)

Monocular cataracts in infants, where only way of visual rehabilitation is contact lens, is often problematic in developing countries because of poor hygiene, socioeconomic factors and non-availability of contact lenses in smaller towns. So, these infants will probably do better with IOL. The following factors need to be considered while selecting an IOL for implantation in pediatric eye.

**Material of the IOL:** In terms of the type of IOL and the safety profile, foldable hydrophobic single piece acrylic IOL material is preferred.\(^{37}\)

**Size of Haptics:** One of major problems in choosing an IOL for a child relates to the changing eye size during growth. The average size of currently available adult-sized, in-the-bag IOLs is 12mm. However, for children <2 years the “ideal” size of IOL should be 10-11mm. The haptics should also be able to adapt to capsular shrinking as well as capsular growth.

**IOL Design and Material:** Square-edge single piece hydrophobic acrylic IOLs of optic size 5.75-6.25mm is considered ideal in pediatric age group. If the optic is too small dysphotopic phenomenon can be produced because of distribution of light around the optic edge in mesopic conditions.

**IOL Filter:** Implantation of IOL in children leaves retina exposed to harmful effects of blue light, therefore a blue filter IOL would be more beneficial.

So the ideal IOL for children will be hydrophobic acrylic, one piece, biconvex with 5.75-6.25 mm optic diameter with closed loop haptic.\(^{38,42}\)

PMMA lenses are not commonly used now due the need for a longer incision and hence greater post-operative astigmatism which can be amblyogenic in children. However, they are useful in certain scenarios like in post-traumatic cataract, for optic capture of the IOL in PCCC and are also used in developing nations due to financial constraints.

Posterior capsular management and posterior vitrectomy: Anterior vitreous acts as a scaffold for proliferation of the lens epithelial cells thus causing VAO. Thus, posterior capsulotomy (PCCC) and limited anterior vitrectomy (LAV) is recommended. The goal of a vitrectomy is to remove the central anterior vitreous without removing the peripheral or posterior vitreous.

VAO is the most common complication after pediatric cataract surgery with or without IOL implantation. The aim of the surgical technique is to provide a long term clear axis by preventing development of visual axis opacification.\(^{43,46}\)

Traditionally, forceps have been used to perform PCCC. Manual posterior capsulorhexis remains the gold standard because it yields a smooth, round edge and also resists capsule tearing. It has a long learning curve and achieving the appropriate sized opening still remains a challenge.

Vitrectorhexis is an alternative to manual PCCC. The PCCC and LAV may be performed before or after in-the-bag placement of the IOL. If performed before placement of the IOL, care must be taken when placing the lens in the capsular bag and not to extend the posterior capsulotomy. Performing posterior capsulotomy after implantation of IOL ensures in-the-bag IOL fixation.\(^{47,48}\)

The desirable size of posterior capsulorhexis is 3-3.5 mm.

Posterior capsulotomy and anterior vitrectomy can be done using the anterior limbal approach or the pars plana approach.

1. **Pars plana approach:** The unique anatomy of the pediatric eyes requires modification of the surgical technique. A major anatomic constraint is imposed by the relative size of the pars plana. In newborn eyes, the pars plana region is incompletely developed so that the anterior retina lies just behind the pars plicata. To avoid iatrogenic retinal breaks entry incision are made through or anterior to the region of pars plicata. The distance of the sclerotomy from the limbus depends on the age of the patient.

2. **Limbal approach:** Limbal based approach is preferred by anterior segment surgeons. It avoids the adverse effect of pars plana approach on the growth of a young eye and avoids retina related complications.\(^{49}\)

Other approaches to prevent posterior capsular opacification: In the year 1994, Gimbel H V described that posterior capsulorhexis with optic capture may be done for secondary membrane formation eliminating the need for vitrectomy. However, other
studies have shown that optic capture performed alone (without vitrectomy) does not eliminate the risk for VAO even though it was associated with better centration of the IOL. In order to perform optic capture, a well centered PCCC of adequate size is a pre-requisite, which is technically demanding50.

Small gauge vitrectomy in pediatric cataract surgery: In recent years, there has been a continuous refinement in the field of vitrectomy devices with introduction of small gauge vitrectomy systems. Fujii et al introduced the first commercially available transconjunctival, sutureless 25-gauge vitrectomy system (Bausch and Lomb, Rochester, NY, USA). This system is based on microcannulax for three-port pars plana vitrectomy. The cannula is inserted through the conjunctiva and sclera with a sharp trocar. During insertion of the cannula, the conjunctiva is displaced to misalign the scleral and conjunctival openings. The main purpose of the cannula is to maintain the misalignment of these openings to facilitate self-sealing closure of wounds after cannula removal. The main advantage of beveled incision is prevention of wound leakage. The smaller instrumentation does not require conjunctival peritomy or suturing of sclerotomies. The advantages of small-gauge PPV include reduction in operative time, inflammation, reduced astigmatism, patient discomfort and recovery time51-54.

CONTROVERSIES IN THE MANAGEMENT OF PEDIATRIC CATARACT

Primary IOL implantation: Primary IOL implantation is the recommended practice in children older than 2 years. In children below 2 years it is controversial. However, with advancements in microsurgical techniques, more and more surgeons are implanting lenses in younger infants.

IOL power calculation formulae: Hoffer Q and Holladay 2 are more predictable than the other formulae especially in young children (<2 years old) and in shorter eyes (<22 mm).

Post-operative refractive goal: Immediate postoperative refractive error and its correction are important factors in preventing amblyopia that may seriously affect the visual outcome of an excellent surgery. Most of the authors prefer to aim for an initial postoperative undercorrection (hyperopia) that varies with the following factors.

1. **Age at cataract surgery**: The closer to birth the implantation is performed, the more marked the undercorrection.
2. **Status of the fellow eye**: It is important to determine the refractive status of the fellow eye to minimize the aniseikonia.
3. **Expected compliance**: It is better to leave less hypermetropic if the child and/or family is not expected to be compliant.

**Anterior capsular management: manual CCC or vitrectorhexis??** A well performed manual CCC provides a capsular opening with strong capsular rim which resists radial extension. However, manual CCC is especially difficult to perform in infants (<1 year of age) because the capsule is highly elastic. Here vitrectorhexis is a good option as owing to the highly elastic capsule, the edge remains regular and resists radial tearing. In the older children, the capsulotomy edge of a vitrectorhexis is scalloped and never as regular as a CCC.

**Posterior capsular management in young children**: In children younger than 5 years, PCCC with anterior vitrectomy is mandatory. Posterior CCC without anterior vitrectomy is done in children between 5 to 8 years. In older children, maintaining intact posterior capsules is advised. Subsequent Nd: YAG capsulotomy should be considered in the presence of PCO.

**Posterior capsular management: manual CCC or vitrectorhexis?** Manual PCCC has a long learning curve and achieving the appropriate sized opening still remains a challenge. Vitrectorhexis after IOL implantation is an easy to learn alternative to manual PCCC in pediatric cataract surgery. It is more predictable and reproducible, with a short learning curve and lesser surgical time.

**Timing of anterior vitrectomy**: LAV can be done before or after placement of the IOL in the bag. Vitrectomy done after IOL implantation ensures in-the-bag placement of the IOL while doing it before may increase the chances of decentration of the IOL.

**Multifocal IOL in pediatric patients**: Jacobi et al. studied pediatric patients aged 2-14 years with multifocal IOL implantation with more than 1 year of follow-up. They found that only 22% children reported permanent use of an additional near correction. The remaining children were either using distance correction only (44%) or no glasses at all (33%). It was concluded that multifocal IOL implantation is a viable alternative to monofocal pseudophakia in this age group. However, we do not recommend multifocal IOL implantation in children due to reduced contrast sensitivity, problems like glare, need for proper centration, changing refractive status of the eye and inaccuracies in biometry55.

POST OPERATIVE VISUAL REHABILITATION

1. **Spectacles** have the advantage of low cost and that they can be conveniently changed. However, they cause problems like glare, need for proper binocular vision. However, they have the disadvantage of higher cost, lens loss, intolerance to contact lens, difficulty in insertion in children and infectious keratitis.

AMBLYOPIA MANAGEMENT56

This is one of the most critical steps in post-operative management of a young child for better visual outcome. This is achieved by occlusion therapy. The amount of patching required is dependent on age.

**Beyond 1 year of age**

| Table 2: Schedule for occlusion according to age |
|-----------------|------------------|-----------------|
| **Age In Months** | **Patching Required** |
| 0-1 Month | No Patching Required |
| 1-2 Months | 1-2 Hours/Day |
| 2-4 | 2-3 Hours/Day |
| 4-6 | Up To 50% of Waking Hours |
| 6-12 | Up To 80% of Waking Hours |

of occlusion of the amblyopic eye can be increased with the general rule-patching of amblyopic eye and normal as a ratio of age. Careful supervision and monitoring is mandatory.

COMPLICATIONS OF PEDIATRIC CATARACT SURGERY

1. **Visual axis opacification**: It is the most common complication of infantile cataract surgery and
Recent Trends and Advances

Conclusions

Improved education of primary health care workers and the public will help in early detection and timely treatment of pediatric cataract in the future. Surgical management of pediatric cataracts is different from adult cataracts. The reduced sclar rigidity, elastic lens capsules and positive vitreous pressure make surgical manipulations more difficult. The high rates of posterior capsular opacifications make PCCC and anterior vitrectomy mandatory in the younger age group. Ocular growth makes selection of IOL power a difficult choice. However, outcomes have improved greatly in the last few decades and with better microsurgical instrumentation, techniques and improved understanding of pediatric eye growth.

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CONCLUSIONS

Vision screening programs and improved education of primary health
Recent Trends and Advances


Intraocular Lens Power Calculation for Pediatric Cataract

Shubhangi Bhave MS, Rohini Khurana Juneja MS

Drishti Eye Clinic & Squint Centre, Nagpur, India.

Abstract: Intraocular Lens (IOL) power calculation of a small eye in a growing child is quiet challenging. To implant an IOL of fixed power into an eye that is still growing is difficult. An optimum IOL power needs to be considered, which best benefits the child’s eye at that time as well as in future as the child grows. The younger the child at the time of surgery, more difficult becomes the task. During initial days of IOL implantation for paediatric cataract surgery, adult IOL powers were used, leading to hyperopia, which in turn caused or aggravated amblyopia.

Intraocular Lens (IOL) power calculation of a small eye in a growing child is quiet challenging. To implant an IOL of fixed power into an eye that is still growing is difficult. An optimum IOL power needs to be considered, which best benefits the child’s eye at that time as well as in future as the child grows. The younger the child at the time of surgery, more difficult becomes the task. During initial days of IOL implantation for paediatric cataract surgery, adult IOL powers were used, leading to hyperopia, which in turn caused or aggravated amblyopia.

ANATOMIC CONSIDERATIONS

The Human eye undergoes rapid growth during first year of life.

- During the first 6 weeks of life the cornea flattens from a mean of 51D to 44D. Keratometry becomes stable by 12 to 18 months (Figure 1).
- The axial length increases from a mean of 17mm at birth to 20mm by 1 year of age, 22mm at 3 years and a presumed endpoint of 23mm at 13 years (Figure 2).
- The lens power decreases from 34D at birth to 28D by 6 months of age. It drops by 10D in first year and then by 3-4D from 2-10 years.

Since the corneal curvature assumes its adult dimensions by 8 weeks of age, a decrease in lens power is believed to compensate for the increase in axial length of the infant eye during the first year of life.

IOL POWER CALCULATION

Biometry: A-scan ultrasound and keratometry measurements can be very difficult or unattainable especially in very young children in the OPD. Examinations under anaesthesia should be preferably done.

Axial length measurement can be done either by immersion or contact method. We prefer the immersion method as the tip does not indent the cornea so there are less chances of error in the readings. It also depends on the ultrasound velocity settings. Axial length of 25mm is best measured with 1550m/sec and 20mm with 1560m/sec. Accurate measurement may be obtained by setting an average velocity of 1532m/sec and then correcting for axial length by adding Corrected AL Factor (CALF) of 0.32 to it. Repeated measurements are taken until three equal readings are obtained. Errors in axial length measurement are the most significant errors in IOL power calculations accounting to 2.5D/mm. However, this error jumps to 3.75D/mm in very short eyes (20mm).

Figure 1: Change in keratometry with age

Figure 2: Change in axial length with age
Recent trends and advances

Keratometry readings should be taken with hand held auto keratometer under anaesthesia, whenever possible. Eyelid speculum should not be used as it alters the curvature. The K-readings in the newborn are ignored and replaced by the average adult reading of 44D.

IOL power calculation formula is an unresolved issue in paediatric cataracts. The Holladay formula is considered most accurate for eyes with axial length between 22 and 26 mm. The Hoffer Q formula is considered most accurate for short eyes (<24.5mm). The SRK/T formula is considered optimal for long eyes (>26mm)\(^4,5\).

Important points to be kept in mind include the velocity that needs to be used for specific eyes (phakic/ aphakic/ pseudophakic), the A-constant for the specific IOL to be used, and the characteristics of a good A-scan tracing with a spike from each layer of eye.

TARGET POSTOPERATIVE REFRACTION

Ideally one should aim for a refractive state that would cause minimal amblyopia in childhood (due to high hypermetropia), while inducing least refractive error (myopia) in adulthood. There is no common opinion regarding the ideal postoperative refraction in infants and children after IOL implantation.

- Initial Emmetropia after the surgery will remove the need of spectacle correction and chances of amblyopia. However, the patient will eventually develop high myopia in adulthood.
- Initial high hypermetropia would compensate for the myopic shift in the adulthood but will leave the child amblyopic in the early phase with immediate spectacle correction along with the amblyopia therapy.
- Slight initial under correction will make the child slightly hyperopic during childhood with easily treatable myopia during adulthood (this is what most surgeons try to achieve).

FACTORS AFFECTING TARGET POSTOPERATIVE REFRACTION

Age at the time of surgery

Lesser is the age of the child at the time of cataract surgery, more will be the under correction needed. What undercorrection is to be done at what age to reduce the myopic shift in future is calculated as shown in (Table 1).

On the basis of percentage - Dahan suggested that final aim of the refraction should be an under correction of 20% in infants and 10% in toddlers\(^6\) or on the basis of axial length alone (Table 2).

Status of fellow eye: More hyperopia can be left when surgery is done bilaterally as the chances of amblyopia would be lesser even in cases of poor compliance with glasses. In case of contralateral pseudophakia, the refractive status of the other eye has to be kept in mind.

Visual Acuity: In dense amblyopia it is better to leave less hyperopia or even emmetropia to help vision recovery by occlusion therapy.

Expected compliance: It is better to leave less refractive error if the child and/or family are expected to comply poorly with glasses, contact lenses and amblyopia therapy.

Parent’s refractive error: It has been noted that 30 to 40% of children become myopic if both the parents have myopia, while the percentage reduces to 20-25% in others. Such children can be left more hyperopic in the beginning to reduce the later myopic shift\(^7\).

Site of IOL implantation: The power of the IOL which was initially intended for capsular bag placement should be reduced by 1D to 2D if it needs to be placed in the sulcus.

IOL power: Higher the IOL power more is the under correction needed. For example, at the age of 6 months, if one child has an emmetropic power of 50D and another child has emmetropic power of 40D, the first child will need a higher residual refraction.

KEY POINTS

- Target hypermetropia with treatable amblyopia to avoid myopic shift in children
- Careful measurements to avoid errors in keratometry and axial length calculations.

### Table 1: IOL Power on the basis of age

<table>
<thead>
<tr>
<th>Age</th>
<th>Residual Refraction</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;1 year</td>
<td>+10D to +7mm</td>
</tr>
<tr>
<td>1-2 years</td>
<td>+6D</td>
</tr>
<tr>
<td>2-4 years</td>
<td>+5D</td>
</tr>
<tr>
<td>4-5 years</td>
<td>+4D</td>
</tr>
<tr>
<td>5-6 years</td>
<td>+3D</td>
</tr>
<tr>
<td>6-7 years</td>
<td>+2D</td>
</tr>
<tr>
<td>7-8 years</td>
<td>+1.5D</td>
</tr>
<tr>
<td>8-10 years</td>
<td>+1D</td>
</tr>
<tr>
<td>10-14 years</td>
<td>+0.5D</td>
</tr>
<tr>
<td>&gt;14 years</td>
<td>+0D</td>
</tr>
</tbody>
</table>

### Table 2: IOL power on the basis of axial length

<table>
<thead>
<tr>
<th>Axial length</th>
<th>IOL power</th>
</tr>
</thead>
<tbody>
<tr>
<td>17mm</td>
<td>28D</td>
</tr>
<tr>
<td>18mm</td>
<td>27D</td>
</tr>
<tr>
<td>19mm</td>
<td>26D</td>
</tr>
<tr>
<td>20mm</td>
<td>24D</td>
</tr>
<tr>
<td>21mm</td>
<td>22D</td>
</tr>
</tbody>
</table>

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Correspondence to:
Dr. Shubhangi Sudhir Bhave,
Consultant Ophthalmologist & Paediatric Ophthalmologist
Drishti Eye Clinic & Squint Centre,
Nagpur, Maharashtra, India.
Abstract: Pediatric cataract can sometimes put us in a spot of bother, managing these difficult situations wisely with proper technique can lay the premonitions to rest. The early identification with few tell-tale signs can help us in confidently tackling the condition during surgery.

DIFFICULT SITUATIONS

Pre-existing Posterior capsule defect

Posterior capsular defect can not only be seen in traumatic cataracts but can also be a pre-existing defect in association with posterior polar cataract, persistent fetal vasculature, lenticonus or lentiglobus. The incidence varies from 2.2-6.75% \cite{1,2} and has higher unilateral association \cite{3}. The defect may be due to the developmentally weak posterior capsule or traction of regressing hyaloid artery. Once the defect is there, the fluid vitreous starts hydrating the lens and with accommodation, there is egress of the lens material into the Berger’s space. Posterior capsular defect may present to us with differential opacification i.e. whiter at center than at periphery and parent often noticing a white dot initially, which has increased in size gradually. The other tell-tale sign we should look for, is membranous looking cataract with flattening of anterior capsule (deep anterior chamber), white dots \cite{4} and fish tail sign (due to lens matter in Berger’s space) \cite{5}. The diagnosis can almost be established with ultrasound biomicroscopy using 35MHz probe though sometimes it might be difficult in early stages where it just looks like a posterior subcapsular cataract (Figure 1A-D).

Management: The anterior capsulorhexis can be difficult due to the partially absorbed nature of the lens. Preferably, anterior continuous curvilinear capsulorhexis should be attempted under high molecular weight viscoelastic substance either using Utrata forceps or 23G intravitreal forceps. This is the most important step as the likelihood of placing lens in sulcus is high in such cases. Hydro dissection and delineation should be avoided. The bottle height/ intraocular pressure is kept low and preferably, vitrectomy cutter is kept in irrigation-aspiration cut (I-A cut) mode and the lens matter is aspirated keeping vacuum at 400 mmHg, aspiration flow rate at 50 cc/min and cut rate at 100 cpm (cuts per minute). Anterior vitrectomy is invariably required to clean the anterior vitreous. The placement of Intra-ocular lens in the bag can be difficult as there is high risk of extension of the posterior capsular defect but can be achieved in long standing cases where fibrosis of the margin of defect is present. The three-piece lens can be placed in sulcus and posterior optic capture may be attempted in an adequately sized anterior capsulorhexis.

Juvenile idiopathic Arthritis associated Uveitis (JIA)

Pediatric cataract is most common in Juvenile idiopathic arthritis associated uveitis (71\%) \cite{6}. The cataract formation is mainly due to severe intra-ocular inflammation and use of topical steroid. These patients generally present with posterior subcapsular cataract associated with posterior synechiae, iris bombe and even peripheral anterior synechiae\cite{7}. Long standing cases presenting with band shaped keratopathy and hypotony, generally have a poor visual prognosis\cite{8} (Figure 2A-D).

Management: The preoperative evaluation should ensure there is no anterior chamber activity i.e. child is in remission at least for a period of 3 months\cite{9}. Hence slit lamp examination to look for cells (<1+) and flare along with fundus examination is of utmost importance. The pupils are often non-dilating and ultrasound is required for posterior segment evaluation. The ultrasound biomicroscopy (UBM) helps us to see the extent of the pupillary membrane, which sometimes can be mistaken for cataract. The child should be started on topical steroid 1% prednisolone acetate 1 week prior and oral steroids 1mg/kg at least 3 days prior to surgery\cite{10}. The role of immunosuppressive agents has been documented to be of benefit in these patients and lower dose methotrexate can be started in consultation with the rheumatologist\cite{11}. Intra-operatively, removal of pupillary membrane should be done with intra-vitreal forceps before staining with dye. The pupil can be released of posterior synechiae using cannula of OVD (Ophthalnic Viscosurgical device) and simultaneously injecting the viscodispenser agent.

Figure 1A): Preoperative clinical picture not able to identify pre-existing posterior capsular defect. 1B): Ultrasound biomicroscopy picture showing pre-existing posterior capsular defect. 1C): Posterior capsulorhexis. 1D): Intraocular lens placed in bag.
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If the pupillary dilation is insufficient iris hooks or Malyugin ring can be used. Preferably iris manipulation should be kept to minimum to avoid post-operative reaction and break in blood aqueous barrier. The anterior capsule may become fibrous requiring use of intra-vitreal scissor to make an opening and completion of the rhexis. The anterior capsulorhexis should be on the larger side just covering the IOL margin to avoid iris forming posterior synechiae in optical zone. The posterior capsulorhexis and anterior vitrectomy is preferred even in children older than 8 years as there is high risk of visual axis opacification (55-100%)12,13. The decision to put intraocular lens is debatable but should be tried as these patients are often intolerable to contact lens because of the formation of band shaped keratopathy. The results have improved with modern minimally invasive techniques and foldable intraocular lens. The heparin coated hydrophobic acrylic foldable IOL is preferred. The need to remove all the viscoelastic substance needs to be kept to minimum to avoid post-operative fibrinous reaction. The tapering of steroid should be slow and should be done over a period of 8 to 12 weeks along with short acting mydriatic and cycloplegic (tropicamide 1% with 2.5% phenylephrine TDS) to avoid synechiae formation15. Long term follow up is generally required to look for cystoid macular edema and need for secondary procedures like membranectomy and YAG capsulotomy (56-100%)16.

SPIEROPHAKIA

Spherophakia was first described by Hartridge in 1886, it is due the defective development of the lens zonules which results in increased length, weakness, absence and non-attachment of posterior zonules to the ciliary processes16. Hence, zonules do not exert sufficient pressure to flatten the developing lens resulting in fetal spherical conformation. The arrest of lens development usually occurs between the fifth and sixth month of intrauterine life16. It usually presents in first or second decade of life with progressive myopia, angle closure glaucoma due to pupillary block, anteriorly or posteriorly dislocated lens and subluxation19. Spherophakia is usually associated with systemic disorders such as Weill-Marchesani’s syndrome, Marfan’s syndrome, homocystinemia, Alport’s syndrome and Klinefelter’s syndrome20 (Figure 3A-E).

Management: The acute angle closure glaucoma secondary to pupillary block is worsened by miotics and relieved by cycloplegics also known as ‘Inverse glaucoma’. On ultrasound biomicroscopy (UBM), shallow anterior chamber, steep anterior lens curvature, iridolenticular contact, elongated zonules, increased distance between the lens equator and the ciliary processes can be seen21. Intraocular bimanual irrigation and aspiration using low bottle height and vitreotomy cutter kept in irrigation-aspiration cut (I-A cut) mode, the lens matter is aspirated keeping vacuum at 400 mmHg, aspiration flow rate at 50 cc/min and cut rate at 100 cpm (cuts per minute)22. Anterior chamber intraocular lens (ACIOL) or Scleral fixed intraocular lens (SFIOL) with or without trabeculectomy can be done.

PERSISTENT FETAL VASCULATURE

Persistent fetal vasculature (PFV) is the term coined by ‘Goldberg’. The lens is supplied by hyaloid artery in dolquet’s canal and when this persists beyond 7 month of gestation it results in persistent fetal vasculature23. This generally presents as eccentric posterior fibrovascular plaque hence initially was called as “Persistent Hyperplastic Primary Vitreous (PHPV)”. This sometimes may be associated with tunica vasculosa lentis, iridohyaloid blood vessels or persistent pupillary membrane. The presence of microphthamols and elongated ciliary body is pathognomonic of persistent fetal vasculature although it can rarely present in normal and buphthalmic eyes24. The patients with anterior PFV generally have good visual prognosis, in contrast to patients with posterior PFV in whom visual potential is often limited by coexisting retinal and optic nerve abnormalities. If the persistent fetal vasculature is not obscuring the visual axis during the first year of life, the prognosis for patient’s vision is good, if surgery and treatment for amblyopia of the affected eye is done at the earliest (Figure 4A-D).

Management: Ultrasound shows a stalk connecting the optic nerve head to the posterior capsule of lens but this finding can sometimes be missed. Colour Doppler imaging is an informative screening and diagnostic tool that shows characteristic flow patterns in PFV. There may be risk for the development of vitreous haemorrhage during and even after surgical repair. UBM preoperatively might help us identify elongated ciliary processes and posterior plaque or defects. Intra-operatively, presence of
Recent trends and advances of the cases. The lens subluxation may lead to the pupil becoming pinpoint or sluggish in about 60% of cases with limitation of dilatation. Transillumination may be positive in about 60% of cases. The pupil is found to be normal while the lens is displaced in the opposite directions as chance of developing amblyopia.

Other ocular features that may be associated include megalocornea, persistent pupillary membrane, polycoria, axial myopia, cataract, glaucoma, retinal detachment, retinitis pigmentosa, retrobulbar cyst, coloboma of iris, lens and choroid, anterior uveitis, phacolytic glaucoma and displacement of lens in the anterior or posterior chamber.

The pathomechanism of this condition is controversial. Several theories have been suggested including mechanical interference of zonular development due to persistent remnants of tunica vasculosa lentis. Occurrence of skeletal mesodermal abnormalities, like in Marfan’s syndrome, suggest a mesodermal origin of ectopia lentis. Presence of iris stromal hypoplasia and persistent pupillary membrane in many patients also suggests a mesodermal dysgenesis. Luebbers proposed the neuroectodermal origin of this condition based on iris trans-illumination defects due to maldevelopment of pigmentary epithelium of iris and absence or hypoplasia of dilator pupillae muscle resulting in corectopia and poor dilatation.

Management: Pre-operatively, dilated fundus exam, ultrasound and specular count are essential. Due to the poorly dilating pupil, there might be a need for iris hooks intra-operatively. After doing the intra-lenticular lens aspiration and anterior vitrectomy, pupilloplasty is done to center the intraocular lens (IOL) (Figure 5A-5F). Then choice can be made between an iris claw lens, anterior chamber lens or a scleral fixated IOL.

Conclusion: Children under general anaesthesia need to be tackled with care. The proper identification, pre-operative evaluation and intra-operative decisions in difficult cases can make the surgical outcomes excellent.

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Correspondence to:
Dr. Sudarshan Khokhar,
Professor of Ophthalmology
Dr. R.P. Centre for Ophthalmic Sciences,
All India Institute of Medical Sciences,
New Delhi, India.
**What is New in Treatment of Amblyopia?**

Surabhi Shalini¹, Subhash Dadeya²


Abstract: Amblyopia is the most common cause of monocular vision loss in children and population under 40 years, with an estimated prevalence of 1-5%. Amblyopia is caused by a prolonged period of abnormal retinal stimulation due to strabismus (ocular misalignment), anisometropia (refractive imbalance), or both (combined) and leads to functional deficits, including reduced contrast sensitivity, poor spatial localization, poor stereovision, and foveal crowding. The present treatments for amblyopia are predominately monocular, aiming to improve the vision in the amblyopic eye. Recent evidence shows that amblyopes possess binocular cortical mechanisms for both threshold and suprathreshold stimuli. Hence, there is an ongoing search for binocular stimulation methods. The purpose is to present stimuli of varying contrast to either eye leading to a concomitant improvement in monocular acuity of the amblyopic eye with the reduction in suppression and strengthening of binocular fusion.

**PATHOPHYSIOLOGY**

Amblyopia is the most common cause of monocular vision loss in children and population under 40 years, with an estimated prevalence of 1-5%⁰⁻¹⁴. It has initially been described as “amblyopia ex anopsia” meaning amblyopia from non-seeing. The mechanism of retinal and cortical interaction varies in different amblyogenic condition. In strabismus, the foveae in the two eyes are directed towards different visual objects. As the foveae have a common visual direction, it provokes visual confusion, retinal rivalry, diplopia and eventually, a decrease of visual acuity of the deviated eye (strabismic amblyopia). However, it is visual confusion and not double vision that is amblyogenic. In anisometropia, the foveae receive images from the same visual object; however, the images from the more ametropic eye are out of focus (anisometropic amblyopia). As the details are clearly focused in the better eye, there is no stimulus to the further effort of accommodation to produce clear images in the worse eye.

Critical period is a time in early life, during which the visual system shows lability of deprivation and ability for reversal of the effect of deprivation. This critical period begins at about 4 months of age, probably passes its peak by 2 years and is well down by 5 years and thereafter undergoes a slow decline to cease by about 12 years of age. The time of onset of the critical period is not uniform for all visual functions, which explains the varied defects seen in amblyopes.

**Treatment**

It is crucial to detect and treat amblyopia as early as possible. Amblyopia can be treated effectively in the first decade of life, greater success being achieved when therapeutic measures are instituted at the earliest as the success rates of amblyopia treatment may decline with increasing age. However, all children should be considered for treatment of amblyopia regardless of age.

**GOALS OF AMBLYOPIA THERAPY**

- Make the patient use the amblyopic eye
- Various treatment modalities have been described for amblyopia. These are: Refractive correction, Occlusion therapy, Penalization, Pharmacotherapy, Home vision therapy, Refractive Surgery, Pleoptics, CAM stimulator, Red filter, Acupuncture, Transcranial magnetic brain stimulation, Television games, Mobile games, Omega fatty acids and Smart glasses. However, none of them is perfect.

**REFRACTIVE CORRECTION**

An essential component in amblyopia treatment is to provide clear foveal image to the amblyopic eye. Refractive correction alone improves visual acuity in one third of patients with anisometropic amblyopia and about two-thirds in ametropic amblyopes⁵⁻⁷. Hence, it is mandatory to carry out cycloplegic refraction in every case of amblyopia before starting any adjunctive treatment.

**OCCLUSION**

Occlusion is considered the gold standard for amblyopia treatment. There is no shortcut or substitute for occlusion in amblyopia therapy till date. However, variability exists in number of hours prescribed to treat amblyopic patients. The success rate of occlusion therapy varies from 30%-92% in various reports. The variation in success rate is due to various factors like patient selection, treatment duration, age, definition of amblyopia used in study and type of amblyopia.

Different authors have recommended different amount of patching.

A recent study by Scott et al⁸ demonstrated that full time occlusion provided excellent visual acuity results. 88% of the patients, who achieved a visual acuity of 20/20 at the end of treatment, maintained that level after an average follow up of 15.5 years. This has been supported by Dorey et al⁹ and Cleary¹⁰. Study done at our centre by Shalini et al. in children aged 4-12 years showed 6 hours of occlusion shows improvement in visual acuity in moderate and severe amblyopia. The means improvement over the baseline visual acuity was 2.2 (SD-1.23).

Hug¹¹ compared full time occlusion with part time occlusion on 3-7 year children. A higher percentage of those treated with
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Full time achieved 20/30 VA in amblyopic eye than part time occlusion patients over a shorter duration of treatment.

Recent recommendations of PEDIG support part time occlusion and rate it similar in efficacy to full time occlusion as ATS213.

ATS 2B concluded that in moderate amblyopia, prescribing two hours of daily patching produces an improvement in visual acuity that is of similar magnitude to the improvement produced by prescribing six hours of daily patching in children 3 to less than 7 years of age.

ATS 2A concluded that for severe amblyopia, prescribing six hours of daily patching produces an improvement in visual acuity that is of similar magnitude to the improvement produced by prescribing full-time daily patching in children 3 to less than 7 years of age.

Another problem with full time occlusion, particularly in severe amblyopia is the functional debilitation that it causes, especially early in treatment. Poor vision during occlusion of the sound eye presents a significant deterrence to good compliance. The child may require additional supervision to avoid accidental injury. Occlusion also causes fusion disruption and increase in angle of deviation. Other than cosmetic unacceptability, allergic skin rash may also hamper patient willingness of putting the patch. This can be minimized with use of skin cream to irritated areas14. All these adverse effects are seen in both full time and part time occlusion, but are more pronounced with full time occlusion.

Penalization

In view of the difficulties encountered during occlusion therapy and the occasional complication of occlusion amblyopia, alternative methods of treatment of amblyopia have been explored. Classically, penalization has been used as a second treatment when occlusion was not complied with, or for post-occlusion as a maintenance treatment.

Indications for penalization are:
1. Moderate amblyopia in uncooperative patient
2. Anisometropic amblyopia
3. Maintenance therapy
4. Occlusion failure
5. Occlusion nystagmus

ATS 115 was initiated to address whether occlusion or atropine penalization is best initial treatment for moderate amblyopia (20/40 - 20/100). It was concluded that both the treatments were well tolerated and the effect of each treatment seemed consistent. The Atropine Group had statistically significant higher degree of acceptability. Patching has a potential advantage of a more rapid improvement in visual acuity and possibly a slightly better acuity outcome, whereas atropine has the potential advantage of easier administration and lower cost. Atropine or patching for a six months period produced a similar improvement in amblyopia two years after treatment. Improvement from baseline to 6 months was 3.16 lines in the patching group and 2.84 in atropine group.

Since compliance is the most important determinant to success in amblyopia therapy, clinical experience has found that atropine penalization has a high acceptability. The PEDIG assessed the acceptance of both types of treatment (occlusion and penalization) by means of a questionnaire. Although atropine was better accepted; the difference in acceptance was small. It has the advantage over occlusion by providing a wider visual field for both eyes, which may have safety and other functional implications. There is also evidence that it improves the ability to see with both eyes.

The PEDIG compared daily atropine to weekend atropine for moderate amblyopia in a randomized clinical trial and the study concluded that weekend atropine provides an improvement in visual acuity of a magnitude similar to that of improvement provided by daily atropine in treating amblyopia in the 3-7 years age Group14.

Conclusions - in this study atropine penalization has been shown to be as effective as occlusion therapy in the treatment of amblyopia. Patient acceptance of atropine penalization was superior to that for occlusion therapy as was shown by the compliance rate. Atropine treatment was also advantageous in that compliance could be readily checked by inspection.

Pharmacological Therapy

Most recently, PEDIG performed a randomized, placebo-controlled study of levodopa in treatment of amblyopia. They concluded that the combination of levodopa-carbidopa and occlusion improves visual function more than levodopa-carbidopa alone in amblyopic children16. Average dose levels of 0.95/0.24 mg/kg and 1.94/0.49 mg/kg of levodopa-carbidopa were found to be well tolerated and efficacious at temporarily improving visual acuity in amblyopic eyes of children17.

Study by Dadeya et al18 evaluated the role of levodopa/carbidopa in the treatment of amblyopia and concluded that there was more than two lines improvement in visual acuity, especially in children younger than eight years of age.

ATS 1419 was done to study the dosage of levodopa as a treatment of residual amblyopia in children 8-18 years old. The study intervention consisted of continuing 2 hours of daily patching plus the addition of levodopa in one of two doses randomly assigned with equal probability (0.51 or 0.76 mg/kg/tid, referred to as lower dose and higher dose, respectively). The lower dose had been used in most prior studies. The study medication was administered for 8 weeks with one additional week for tapering of treatment. Levodopa was prepared in capsules combined with carbidopa 0.17 mg/kg/tid. Carbidopa was combined with levodopa to reduce side-effects associated with levodopa alone. The mean improvement in amblyopic eye visual acuity from baseline to the 9-week primary outcome visit was +4 (+4) letters in the 16 subjects in the lower dose Group and +6 (+6) letters in the 17 subjects in the higher dose Group (mean difference between Groups = -2 letters).

ATS 1720 compared the efficacy and safety of oral levodopa and patching versus oral placebo and patching at 18 weeks, after 16 weeks of treatment for amblyopia in children 7 to <13 years old and it concluded that children 7 to 12 years of age with residual amblyopia after patching therapy, oral levodopa while continuing to patch 2 hours daily does not produce a clinically or statistically meaningful improvement in VA compared with placebo and patching.

Citicholine (CDP Choline) is an essential intermediate for phosphatidylcholine synthesis. It increases cerebral blood flow. It also shows neural restorative effect, via action on dopaminergic pathway of central nervous systems.

Near Visual Activities

Near visual activities are often prescribed during patching for amblyopia based on the assumption that those activities stimulate the visual system. In the recent times their role again has...
Recent Trends and Advances involving eye-hand coordination in the effect of near activities or activities and also to several case series reporting of the previous randomized pilot study. The investigators did not notice any period of 1.62±1.20 years. Lines (0.33±0.26 log MAR), in a follow-up from baseline by an average of 3.2±2.5 success rate with visual acuity improving strabismic or combined amblyopia (both age 4.86 years) with anisometropic, visual activities with part time patching retrospective study published in 2008 in the treatment of amblyopia. As a matter of fact, it has in the past, being regarded by many as notorious popular mode of entertainment, having last slightly longer than the actual time of stimulation. As binocular game play was shown to be superior to monocular game play in visual acuity and stereopsis improvement, researchers began to develop dichoptic games.

Hess et al. examined the potential of treating amblyopic adults using mobile game. In this study dichoptic mobile game Tetris was used on iPod to treat 14 amblyopic patients aged 13 to 50 years were examined. The dichoptic video game treatment was conducted at home and visual function assessed before and after treatment.

On this basis they concluded that the home-based dichoptic iPod mobile game approach represents a viable treatment for adults with amblyopia.

Birch et al. also did similar study to find out the effectiveness of a novel home-based binocular amblyopia treatment. They treated children (4–12 years of age) who wore anaglyphic glasses to play binocular games on an iPod platform for 4 h/w for 4 weeks. The first 25 children were assigned to sham games and then 50 children to binocular games. They concluded that Binocular iPad treatment rapidly improved visual acuity, and visual acuity was stable for at least 3 months following the cessation of treatment.

These Games achieve their therapeutic effect by presenting a different image to each eye, thus rewarding the patient when both eyes work together to win the game. For instance, in the Stereoblock game, some of the blocks seen by the amblyopic eye are in high contrast, while other blocks

ROLE OF REFRACTIVE SURGERY

LASIK is an alternative method for correcting high myopic and hyperopic anisometropia. Recent trends show that refractive surgery is effective and safe alternate treatment modality in desperate cases of conventional therapy failure cases of anisometric amblyopia. Acupuncture is a potentially useful complementary treatment modality that may provide sustainable adjunctive effect to refractive correction for anisometric amblyopia in young children.

Although the treatment effect of acupuncture appears promising, the mechanism underlying its success as a treatment for amblyopia remains unclear. Acupuncture at vision-related acupoints may modulate the activity of the visual cortex. Moreover, acupuncture has been shown to be effective in increasing blood flow to the cerebral and ocular vasculatures (including the choroid), stimulating the expression of retinal nerve growth factors and leading to metabolic changes in the central nervous system.

TRANSCRANIAL MAGNETIC BRAIN STIMULATION

Transcranial magnetic stimulation (TMS) is a non-invasive method for stimulating parts of the brain by use of weak electric current that are induced into the tissue by use of rapidly changing magnetic currents. TMS produces effects that last slightly longer than the actual time of stimulation.

MOBILE GAMES

It has been shown that loss of binocularity is one of the defining features of amblyopia (Mckee, Levi, & Movshon). Therefore, the focus of research in this area has shifted from monocular interventions that involve patching of the fellow eye to approaches that directly target binocular visual function and as the primary therapeutic step. This has led to increased interest in the development of amblyopia treatments that directly address binocular dysfunction by promoting binocular vision and reducing inhibitory interactions within the visual cortex.

The first attempt to provide the combination of short-term occlusion (20 min), controlled visual stimulation and attentive game play (noughts and crosses) was the CAM treatment (Campbell et al). Its beneficial effects were later isolated to the short term nature of the occlusion and the attentive game play (Mitchell, Howell, & Keith). There is no doubt that perceptual learning combined with short-term patching is much better than longer-term patching with passive stimulation in terms of improving monocular acuity (Li et al), however its usefulness for re-establishing binocular vision and stereopsis is less clear. A number of hybrid-binocular treatments have been suggested, which are all directed to recovering monocular function under binocular viewing. The aim is to involve the fixing eye in recovery of vision through intensive training/detection of targets presented exclusively to the amblyopic eye. The iBit system (Cleary et al), the “Push–Pull” (Ooiemail et al).

As binocular game play was shown to be superior to monocular game play in visual acuity and stereopsis improvement, researchers began to develop dichoptic games.

Role of television/video games

Television has always been a very popular mode of entertainment, having being regarded by many as notorious for engaging children and adults alike in spending idle time in front of it. In spite of that, its popularity refuses to die down. As a matter of fact, it has in the past, found utility in administering amblyopia therapy.

In 1977, Saladin and Bohman noted that orthoptic therapy often entails the repetitive usage of detailed and complicated training procedures and amblyopia and antisuppression therapy in particular may require weeks or months of training, so as to maintain a high level of patient interest and cooperation throughout the training procedure. They reported an anaglyphic TV. ping pong antisuppression trainer.

In study conducted by Dadeya et al it was concluded that the television games along with full time patching, are effective in the treatment of amblyopia.
in lower contrast are seen by the healthy eye. A pilot study done at our centre was done to evaluate the efficacy of 2 hours of occlusion along with mobile game exercises as a form of near visual activity in the treatment of amblyopia.

### Role of omega fatty acids

Amblyopia can be considered the result of a lack of normal plasticity. Visual cortical dominance by the better eye leads to correspondent visual deprivation of the representations related to the eye with worse acuity. Knowledge of neuroplasticity and the factors that control the opening and closure of critical periods will lead to new therapeutic strategies which may allow for greater recovery of visual functions in both children and adults with amblyopia.

Omega fatty acids are important structural components of membrane lipids in central nervous system. There are two long chains polyunsaturated fatty acids, DHA and ARA. During third trimester of human foetal development there is tenfold increase in brain size which is accompanied by 30 fold increase in DHA content and a 15 fold increase in ARA content.

Several randomized trials are conducted and found specific benefit of omega fatty acids supplementation for retinal maturation, visual acuity development, or cognitive development.

Preliminary results from a study in England show that school performance improved among a group of students receiving omega-3 fatty acids. In an Australian study, 396 children between the ages 6 and 12 who were given a drink with omega-3 fatty acids and other nutrients (iron, zinc, folic acid and vitamins A, B6, B12 and C) showed higher scores on tests measuring verbal intelligence, reading, learning and memory after six months and one year than a control group of students who did not receive the nutritional drink. This study was also conducted with 394 children in Indonesia. The results showed higher test scores for boys and girls in children in Indonesia. The results showed higher test scores for boys and girls.

This study was also conducted with 394 children who went from six hours of patching per day to no patching. Additionally, patients with a history of successfully treated amblyopia need continued close monitoring for a recurrence of amblyopia.

A study of moderate and severe amblyopia treatment found approximately 25% of patients under age seven had a recurrence of amblyopia within the first year of stopping treatment, and children ages seven to 12 had a 7% chance of recurrence (worsening of two lines of visual acuity). This recurrence is more common in patients with severe amblyopia who went from six hours of patching per day to no patching. Additionally, patients with a history of successfully treated amblyopia need continued close monitoring for a recurrence of amblyopia.

To help prevent this recurrence, patients should be weaned off patching therapy. Residual amblyopia is another treatment challenge, considering vision does not improve sufficiently with one treatment for some patients.

### Lacunae in existing amblyopia therapy

Besides the fact that these treatment modalities have dominated the pediatric ophthalmologist world for ages, patient compliance is poor because of the many side effects of patching and drops. Furthermore, patching can cause many psychosocial problems when trying to force a child to comply with wearing their eye patch. Kids don’t want to wear their eye patch because it impacts their quality of life and atropine drops can cause light sensitivity and disorientation.

Even the patient who has done occlusion therapy with good compliance may improve visual acuity in only 27-32% of cases; leading to suppression, stereo blindness and a deficient depth perception in the rest.

### NEWER STRATEGIES

Recent studies have shown that amblyopia is a binocular problem caused by active suppression that converts a structurally intact binocular system into a functionally monocular system. Evidence suggests that focusing on binocular treatments for amblyopia may prove to be beneficial in both improving vision as well as possibly improving binocularity.

These treatments are based on three principles:

- Monocular perceptual learning (PL),
- Monocular videogame play (VGP) and
- Dichoptic PL/VGP.

A dichoptic treatment presents a stimulus to each eye separately and the brain is forced to integrate the images into a single perception. In these treatment modalities; under binocular conditions, the signal strength coming into the patient’s good eye is reduced enough so that it cannot suppress the amblyopic eye. The result is binocular perception in a patient with otherwise deep suppression of the amblyopic eye. Over time, the viewing conditions are changed and the image seen by the good eye is suppressed less until both eyes see approximately the same image. There are many popular softwares that are developing now-a-days promoting binocular vision therapy.
Recent trends and advances and that additional treatment might be still present in treated amblyopes, who together. Published reports indicate that visual acuity of the amblyopic eye, techniques that could be applied to a animals. This research not only aims to found to reverse amblyopia in adult (Prozac), which has previously been published). Although part of patching for amblyopia in children 3 to < 7 years old. Ophthalmology. 2008; 115(11):2071–2078.


Hess RF, Babu RJ, Cavagneri Set et al. The iPod binocular home-based treatment for amblyopia in adults: efficacy and compliance, Clinical and experimental ophthalmology 2014; 97: 389-398


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**Do you want to be a Pediatric Ophthalmologist?**

**Take Control of your Future: Scope of Pediatric Ophthalmology**

Savleen Kaur MS

*Advanced Eye Center, Post Graduate Institute of Medical Education and Research, Chandigarh, India*

As I walked into my first paediatric clinic posting in my first year of residency; the chaotic crying noises and several screaming kids made me want to cry. Ten minutes into it and I told my colleague “I can’t do this. I can never have that much patience”. I reminded myself most of the days, “It’s just a day; this shall also pass”. I restricted myself to a long history taking and I kept trying to make the mothers realise that their kid was a monster whom they could not control. I kept yelling “You have to make them stop crying like you do at home”. I copied down retinoscopy values like a donkey; writing ‘uncooperative’ most of the time in my strabismus examination. I nodded deceptively in agreement every time my seniors increased or decreased the spectacle prescription.

And then I met this kid. A four-year-old I operated for a bilateral total white cataract. He kept howling preoperatively; before induction of anesthesia and postoperatively. Till I removed his postoperative dressing. I took it out and he stopped crying. He looked at me and stared at me for a good two minutes. And then he looked at his mother, looked around the room and smiled. He could see. It dawned on me that this kid, like many others is going to do everything I say. I had the power to shape his life and his future. Today after only a few years into this speciality I realise not only did I change the kids’ future; I also changed mine.

Choosing paediatric ophthalmology demands a lot of patience and stamina with a holistic approach of the child’s visual system. It can be boring for some; with no lucrative treatment options and minimal changes in paradigms. Do not choose this branch because someone said so. But if you don’t know what it is all about, how will you even decide that you don’t want to do it? I myself have a very limited experience in the field mostly under an excellent direction and guidance by my teachers. My objective behind this piece of writing is to give you an insight on the work life of a pediatric ophthalmologist.

A paediatric ophthalmologist is not only about strabismus or spectacles. It’s a whole lot of ophthalmology shrunken in one small clinic. You need to know the basics of optics, refractive errors, spectacle prescriptions, normal visual and growth milestones in a child; genetic disorders and a lot of paediatrics too. One needs to be aware of a vast era of abnormalities ranging from allergic eye diseases; to epiphora; strabismus; paediatric cataract; glaucoma; uveitis; retinoblastoma and retinopathy of prematurity to name a few. So, don’t restrict yourself to just what you are taught and what you see people doing. Expand and make your own realm.

It is a branch that is ever increasing in demand, with only few selective people pursuing it. Most of the times, a pediatric ophthalmologist is dependent on adult cataract surgery or other realms due to work restraints or for livelihood. How many pediatric ophthalmologists you know who practice just on pediatric eye diseases? Doing pediatric surgery will make you an equally good and sometimes a better adult cataract surgeon. In the 3D world that we are heading into; strabismus surgery is becoming functionally more necessary than being only cosmetic. Retinopathy of prematurity is an ever-increasing epidemic and in a country like India, parents choose the girl child to rather go blind than be enucleated for retinoblastoma. Pediatric ophthalmologists are very few in proportion to cataract and refractive surgeons. So, what are the tricks of the trade? Imagine the most difficult thing you had to do as a fellow in ophthalmology. Application tonometry or gonioscopy in the first year; retinoscopy or indirect ophthalmoscopy? May be a tough task in life like riding a bike or car; reconciling with an offended colleague or raising your own child? Could you overcome that? If yes, how difficult will it be to make a kid talk to you. If I have learnt something from my teachers, it is that intelligence is nothing but an infinite capacity for hard work. If you believe in yourself, you can do it. Believe me, as a pediatric ophthalmologist; you get instant gratification and good prognostic cases, make the kids laugh; the parents indebted.

<table>
<thead>
<tr>
<th>Get rid of your white coat sometimes if needed or decorate it with a patch</th>
</tr>
</thead>
<tbody>
<tr>
<td>Use as many different toys as you can each time/ have kids’ videos on mobile</td>
</tr>
<tr>
<td>Examine the child in the mother’s lap in a usual chair rather than examining chair</td>
</tr>
<tr>
<td>Shake the child’s hand before starting examination (with a candy perhaps)</td>
</tr>
<tr>
<td>Build a relationship with the child first-he/she will let you examine at ease</td>
</tr>
<tr>
<td>Try examining only one kid at a time-their attention breaks easily</td>
</tr>
<tr>
<td>Keep your walls coloured and as less equipment in the room as possible; don’t make kids listen to other kids crying while dilation</td>
</tr>
<tr>
<td>Learn to make funny noises!</td>
</tr>
<tr>
<td>Expand the visual acuity charts you have</td>
</tr>
<tr>
<td>Learn not to shout-even when not in clinic</td>
</tr>
<tr>
<td>Educate the mother and trust her- she can be a better doctor than you!</td>
</tr>
</tbody>
</table>

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*Dos Times - July-August 2017*
and most of all— you get to laugh and to have fun.

Even while being a resident if you have thoughts like “I am not good with kids” doesn’t mean you cannot pursue a career in pediatric ophthalmology. You can be a pediatric ophthalmologist even if you do not have much passion and patience towards kids. This is something that can be trained. Also, it is said there is no good borne-surgical hand, most of things come with mere training and perseverance. The teacher of teachers, Prof Amod Gupta; taught us all the most important quality needed to be a physician- that is ‘empathy’. I tried to remember it on every clinic day. That’s all it takes to be a pediatric ophthalmologist too.

I am no expert, we are all learners here. I am not promising you any good money to begin with. But I can promise you gratification if you choose this career with a change in your own personality. You may have to make the medical fraternity appreciate and perceive the burden of childhood ophthalmic problems. May be you must alert everyone to the fact that a child must be screened early for something as simple as a refractive error to a malignant condition like retinoblastoma. Maybe you have to become more of a social worker and counsellor sometimes because even making them wear spectacles could be a social stigma. You know, 153 million people can be cured of their blindness worldwide by wearing glasses alone.

The first step towards getting somewhere is to decide you’re not going to stay where you are.

—John Pierpont “J.P.” Morgan

So, what does being a pediatric ophthalmologist mean? To me, being a pediatric ophthalmologist means being a teacher, a counselor, a guide, an empathetic doctor and a brilliant surgeon! A touch of humanity, a pinch of compassion, a bit of patience and loads of hard work- is this not why we became doctors in the first place?

Dedicated to all my teachers

Acknowledgment (For education material): Ghada Zein El- Abedin Rajab MD. Lecturer of Ophthalmology, Menoufia, University Hospitals, Egypt.

Speciality Societies / Conference avenues
- Strabismus and pediatric ophthalmology society of India [www.sposiindia.org]
- American Association for Pediatric Ophthalmology and Strabismus [www.aapos.org]
- Global pediatric ophthalmology congress [www.pediatricophthalmologyconferenceseries.com]

Fellowship opportunities
- http://www.sankaranethralaya.org
- http://www.aravind.org

International
- Commonwealth fellowships-http://cehc.lshtm.ac.uk/clinical-fellows/
- http://www.uovs.com/education/fellowships/paediatrics-strabismus
- http://soevision.org/fellowship-platform/
- https://www.willseye.org/fellowship-pediatric-ophthalmology-for-foreign-medical-graduates
- International council of Ophthalmology fellowships including fellowship for ROP
- http://www.icoph.org/refocusing_education/fellowships.html

Must read journals
- Journal of the American Association of pediatric ophthalmology and strabismus
- Journal of pediatric ophthalmology and strabismus
- Indian journal of pediatric ophthalmology and strabismus

Web education sources
- https://www.aao.org/clinical-education/pediatric-ophthalmology-education-center
- http://www.eophtha.com
- Cybersight organization: provides online courses, live lectures (webinars), and consultation. Pediatric ophthalmology is an integral part of this website. https://cybersight.org/
- Eyeforum by University of Iowa http://webeye.ophth.uiowa.edu/eyeforum/index.htm
- Canadian neuroophthalmology group: Neuro-ophthalmology is intimated to the pediatric ophthalmology. http://www.neuroophthalmology.ca/

* Paid sources;
- American Academy one network. https://www.aao.org/international/programs/about-one-network
- Facebook, telegram, and Yahoo groups and virtual universities
- Telegram of free ophthalmic downloading; Facebook; e.g. Piece of ophthalmology videos
- Yahoo group: e.g. FRCophthyahoogroup- Asian ophthalmology yahoo group.
- Virtual university for postgraduate teaching e.g. Muthusammy
Botulinum Toxin in Strabismus

T.S. Surendran MS, Supraja Kasturirangan

Sankara Nethralaya, Chennai, Tamil Nadu, India.

Abstract: *C. botulinum* elaborates eight antigenically distinguishable exotoxins (A, B, C1, C2, D, E, F and G). Type A is the most potent toxin, followed by types B and F toxin. The uses of botulinum in strabismus are manifold and highlighted in this article.

**INTRODUCTION**

*C. botulinum* elaborates eight antigenically distinguishable exotoxins (A, B, C1, C2, D, E, F and G). Type A is the most potent toxin, followed by types B and F toxin. Types A, B and E are commonly associated with systemic botulism in humans.

All botulinum neurotoxins are produced as relatively inactive, single polypeptide chains with a molecular mass of about 150 kDa with a high degree of amino acid sequence homology among the toxin types. The polypeptide chain consists of a heavy (H) chain and a light (L) chain of roughly 100 and 50 kDa respectively, linked by a disulphide bond. The botulin toxin neurotoxin complex is also associated with various other nontoxic proteins, which may also have hemagglutinating properties.

**MECHANISM OF ACTION**

All the serotypes interfere with neural transmission by blocking the release of acetylcholine, which is the principal neurotransmitter at the neuromuscular junction. Intramuscular administration of botulinum toxin acts at the neuromuscular junction to cause muscle paralysis by inhibiting the release of acetylcholine from presynaptic motor neurons. Botulinum toxins act at four different sites in the body: The neuromuscular junction, autonomic ganglia, postganglionic parasympathetic nerve endings and postganglionic sympathetic nerve endings that release acetylcholine. The heavy (H) chain of the toxin binds selectively and irreversibly to high affinity receptors at the presynaptic surface of cholinergic neurons, and the toxin-receptor complex is taken up into the cell by endocytosis. The disulphide bond between the two chains is cleaved and the toxin escapes into the cytoplasm. The light (L) chain interact with different proteins (synaptosomal associated protein (SNAP) 25, vesicle associated membrane protein and syntaxin) in the nerve terminals to prevent fusion of acetylcholine vesicles with the cell membrane. The affected nerve terminals do not degenerate, but the blockage of neurotransmitter release is irreversible. Function can be recovered by the sprouting of new terminals in a process that takes two to three months. The toxin lasts for nearly 8-12 weeks.

**INDICATIONS**

1. Paralytic strabismus-
   a. 6th nerve palsy
   b. 4th nerve palsy
   c. 3rd nerve palsy
2. Childhood strabismus-
   a. Infantile esotropia
   b. Accommodative esotropia
   c. Cyclic esotropia
   d. Intermittent exotropia
   e. Cerebral palsy
3. Adult strabismus-
   a. Strabismus already operated multiple times
   b. Post retinal detachment strabismus
   c. Thyroid ophthalmopathy
   d. Post-operative adjustment
   e. Intrinsic muscle disorders
   f. Nystagmus
   g. Post cataract strabismus surgery
4. Any strabismus for which the patient is not fit for anesthesia due to any systemic illness
5. When multiple muscles need to be operated on in a single eye, one of the muscles can be given injection of botulinum toxin to prevent anterior segment ischemia.

**CONTRAINDICATIONS**

1. Restrictive strabismus when the muscle is scarred - severe restrictions caused by scarred inelastic muscles favour surgery, because botulinum toxin depends on reciprocal agonist-antagonist readjustments of length-tension elasticity, scar tissue restricts this readjustment.
2. Contraindicated in the presence of infection at the proposed injection site(s)
3. In individuals with known hypersensitivity to any botulinum toxin preparation or to any of the components in the formulation.

**TECHNIQUE**

- It can be done under-
  - EMG guidance
  - Direct visualisation of the muscle by making a snip incision on the conjunctiva overlying the muscle

**EMG guided Botulinum toxin injection**

1. Informed consent- mention about over/undercorrection, spatial disorientation, possible diplopia, repeat injections
2. Equipment needed-
   a. Amplifier
   b. Needle with intact tip and good coating of teflon
3. Anaesthesia-
   a. Topical with proparacaine in a minimum of a drop a minute for 3 doses + vasoconstrictor drop (E.g. Epinephrine 1%) to blanch the conjunctival blood
**Dosage of botulinum**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Dosage</th>
</tr>
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<tbody>
<tr>
<td>Horizontal strabismus</td>
<td></td>
</tr>
<tr>
<td>Under 25 PD</td>
<td>2.5 units</td>
</tr>
<tr>
<td>Over 25 PD</td>
<td>2.5-5 units</td>
</tr>
<tr>
<td>Medial rectus for concomitant deviation</td>
<td>1.0-2.0 units</td>
</tr>
<tr>
<td>Surgery</td>
<td>2.5 units</td>
</tr>
<tr>
<td>Lateral rectus for concomitant deviation</td>
<td>1.0-2.0 units</td>
</tr>
<tr>
<td>Healed palsy with MR</td>
<td>2.5 units</td>
</tr>
<tr>
<td>Contracture</td>
<td></td>
</tr>
<tr>
<td>Vertical muscles</td>
<td></td>
</tr>
<tr>
<td>Inferior rectus (IR) for thyroid</td>
<td>5.0 units</td>
</tr>
<tr>
<td>IR for concomitant deviation</td>
<td>2.5 units</td>
</tr>
<tr>
<td>Superior rectus</td>
<td>2.0 units</td>
</tr>
<tr>
<td>Inferior Oblique</td>
<td>2.5 units</td>
</tr>
<tr>
<td>Children with infantile esotropia or exotropia (bilateral injections)</td>
<td>2.5 units</td>
</tr>
<tr>
<td>Weak muscles: myasthenia, external ophthalmoplegia, aberrant degeneration, cerebral palsy</td>
<td>1.0-2.0 units</td>
</tr>
<tr>
<td>Retrobulbar injection for nystagmus</td>
<td>25.0 units</td>
</tr>
</tbody>
</table>

*Courtesy - Clinical Strabismus Management, principles and surgical techniques, Arthue L Rosenbaum, Alvina Pauline Santiago, Chapter 32*

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While listening to the EMG signals, advance the needle tip towards the area giving off the loudest sound. When a cracking sharp EMG signal is heard, inject the fluid slowly. It is a good sign if the EMG sound diminishes with the injection, indicating that the solution pushed the nearby muscle fibres away from the tip. Leave the tip there for 15-30 seconds until the pressure of the solution diminishes. Withdraw the needle slowly while the patient maintains primary gaze. Make a note of how reliable the EMG response and injection were, for future reference.

In our experience dosage can also be calculated depending on the angle of deviation - For example 2 units of Botox for 20 PD of deviation, 3 units for 30 PD of deviation and so on.

**COMPLICATIONS**

**Corneal irritation**

Sub-conjunctival hemorrhage from needle insertion.

Discomfort at time of injection.

Prolonged discomfort lasting a few hours. This is usually associated with a needle touching the orbital peristeum or encountering scar tissue from a prior operation. There is no really good treatment except avoidance and analgesic medication.

Effect of the drug on adjacent muscles. The levator appears to be especially susceptible. These effects on adjacent muscles typically occur only when the injected muscle itself receives a strongly weakening effect.

Injection not into the muscle.

Disorientation of the patient. Past-pointing and spatial disorientation occur with paralysis of a fixing eye. When this is a problem, that eye should be covered. Adaptation to the paralysis typically occurs after the third or fourth day.

Diplopia. Frequently an eye is turned by the injection temporarily to a position where no suppression occurs. Oclusion of the eye is indicated.

**REFERENCES**

Abstract: The goal of the strabismus surgery is to achieve both motor alignment and sensory improvement with minimum number of procedures. Weakening procedure on the recti constitute an important surgical modality for correcting strabismus. Although many surgical techniques are available for weakening of recti muscle, the need for a safe, easy and effective surgical procedure led surgeons to experiment with synthetic absorbable sutures. These high tensile strength absorbable sutures form the basis of suspension recession techniques. Hang back muscle recession technique and its various modifications are now being extensively used because of the multiple advantages that they offer along with increased safety and a lower complication rate.

Earlier myotomies were performed to weaken the extraocular muscles. These efforts were generally unsuccessful and surgeons were frustrated by the random retraction and reattachment of the cut muscles to the globe. In search of more predictable procedures, Jameson in 1922 introduced the technique on which all conventional recessions are based. The conventional recession technique involved direct suturing of the muscle to the sclera posterior to their insertion at the site of recession. This procedure allowed accurate grading of the surgery permitting a definite understanding of the location of reattachment of the recessed muscle. However, this technique alone neither guarantees precise muscle reattachment nor perfect ocular alignment. Below are the problems associated with conventional muscle recession surgery:

a) Inadvertent perforation of sclera resulting in retinal detachment, endophthalmitis or phthisis bulbi.

b) No provision for customization.

c) Problematic in large recession, in high myopes, and infants.

d) More chances of tenons capsule snaring in large recessions.

TECHNIQUE

Gobin in 1960s first used the term, loop recession. He used two 5-0 silk sutures attached to the ends of the muscle when recessing the medial rectus muscle more than 5 mm. In this procedure, the detached muscle was sutured to the sclera at two points separated by a distance equal to the width of the original insertion and located 5 mm behind it. The loose ends of the suture ends were tied over a 2-mm diameter probe and the muscle was allowed to recess another 5 mm for a total recession of 10 mm. This technique of loop recession underwent many modifications and in the early 1970's with the advent of high tensile strength synthetic absorbable sutures, the suspension recession or hangback techniques were introduced (Figure 1). Mills and coworkers conducted the first study of nonadjustable hang-back recession on patients with either horizontal or vertical strabismus. Although not controlled, the study found that 55 of 62 (89%) patients had a successful outcome, which was defined as any alignment that did not require reoperation.

This procedure was advocated as a safe, easy and effective alternative to conventional muscle recession technique. Success here depends upon attachment of the cut muscle to the sclera before complete hydrolysis of the suture material occurs.

Synthetic absorbable sutures such as vicryl are not completely absorbed until 90 days after intramuscular implantation. Firm muscle scleral union occurs within 1-2 months after surgery. Advantages of hangback muscle recession are as follows:

- Reduced risk of scleral perforation.
- Easy intraoperative adjustment in case of over action (spring-back balance test).
- Convenient site for scleral suturing.
- Less chance of snaring tenon's capsule.
- Simplified technique - good for residents and general ophthalmologists.
- Avoid excessive manipulations of the eye.

Hang-back muscle recession also formed an integral part of adjustable suturing technique, a procedure that was introduced by Jampolsky in 1970's. Adjustable suture modification to traditional surgery was introduced to improve surgical outcome and reduce frequency of reoperations by eliminating undesirable early post-operative under or over-corrections.

The hemi hang-back muscle recession was described by Potter et al in 1992. The technique is derived from three procedures mentioned in the literature: conventional scleral suturing, hang-back muscle recession and loop suspension recession technique. In the hemi-hang-back muscle recession technique, the muscle is reattached and suspended from the sclera posterior to the original insertion. This procedure is illustrated in Figure 2. After disinserting the muscle, it is...
Techniques

Kamlesh et al. Hangback Recession

suspended, with the help of sutures, from a point midway between the original insertion and desired muscle insertion site. This technique is recommended for management of difficult strabismus cases as in paralytic strabismus; large deviations or following maximal recessions with residual large deviations and in cases with good vision in one eye when the deviating eye has already undergone maximal surgery. Although there are many advantages of hangback recession, there are some possible disadvantages also:

a) Central posterior muscle bowing
b) Late over-corrections may occur if muscle reattachment lags suture hydrolysis and the muscle slips posteriorly.12-14

c) Possibility of a sideslip of the sutures up or down over the surface of the globe with vertical movement of the eye, producing temporary upshoots or downshoots

d) Extraocular muscle might attach to the globe above or below the horizontal meridian, thereby producing an A or V pattern strabismus or possibly permanent upshoots or downshoots

To tackle these disadvantages, Macleod et al15 described a modified suspension recession technique - an anchored hang-back muscle recession technique. As shown in the Figure 1, the central posterior bowing noted with hangback recession led to enhanced recession making the results less predictable. In addition, the muscle cannot be displaced accurately upwards or downwards as may be required for correction of A and V patterns or vertical deviations. The tendency of the muscle to bunch centrally leads to lack of control over the final vertical position of the muscle thus hindering the correction of coexisting vertical deviation of A and V pattern. Thus, anchored hang-back muscle recession is designed to overcome the problems of hang-back muscle recession while maintaining the ease and safety of the latter procedure. This technique is illustrated step by step in Figure 3. The muscle is exposed using a paralimbal fornix based incision and carefully hooked (Figure 3A-C). Locking bites are taken to secure the muscle with 6-0 vicryl suture (Figure 3D-G). The muscle is then severed from the original insertion and markings taken for the site of desired insertion site (Figure 3H-J). Then a superficial anchoring bite is taken at the desired site of muscle recession and the muscle is suspended from the original muscle insertion using the same sutures (Figure 3K-O). The conjunctival flap is then sutured back (Figure 3P).

Although anchored hang-back muscle recession appears to overcome most of the disadvantages of the hangback muscle recession, it lost the major advantage of customization in terms of post or intraoperative adjustment offered by the hang-back recession.

In order to integrate the advantage of both techniques; Kamlesh et al16 in 2002 described a new procedure 'Modified Anchored Hang-back muscle recession' technique. In this technique, after disinserting the muscle, a superficial scleral anchoring bite is placed midway between the intended recession site and the original insertion (Figure 4: A-I). This procedure on one hand maintains the width of the muscle preventing central bowing and on the other hand allows for intra and postoperative customization.

NEW DEVELOPMENTS

Over the last decade hang-back muscle recession has undoubtedly proven to be a safe, easy and effective
surgery in both routine as well as complicated strabismus procedures for both horizontal and vertical recti muscles. Modified anchored hang-back surgery has added a new armamentarium to the existing surgical techniques as a procedure overcoming the disadvantages of hang-back surgery and at the same time offering an opportunity for intra as well as postoperative customization.

Recently Awadein et al compared non-absorbable versus absorbable sutures in hangback medial rectus muscle resection. They concluded that using nonabsorbable suture for hang-back medial rectus recessions; greatly reduces the incidence of over-corrections that can occur when absorbable suture dissolves.

Hang back recession was carried out on the inferior oblique muscle for the first time in the history of strabismus surgery by Kamlesh et al. They evaluated 15 patients of V pattern strabismus with inferior oblique overaction and performed 10 mm Hangback recession of the inferior oblique muscle. In this study, a mean V pattern correction of 20.2 prism diopters was obtained. 85.37% patients, the V pattern was found to be fully corrected. The mean inferior oblique overaction correction was found to be 19.04 prism dioptres. No significant complications were encountered. This is the only study, which has evaluated the outcome of suspension recession technique on the inferior oblique muscle.

**CONCLUSION**

Hang-back muscle recession offers an exciting and challenging alternative to conventional recession for the correction of strabismus.

**REFERENCES**


**Correspondence to:**
Dr. Kamlesh,
Director and Head of Ophthalmology,
Guru Nanak Eye Center,
Maulana Azad Medical College,
New Delhi, India.
Summary: The one and a half syndrome is a rare clinical disorder of extraocular movements characterized by a conjugate horizontal gaze palsy of one side with an internuclear ophthalmoplegia on the other side. It occurs due to a single unilateral lesion of the dorsal pontine tegmentum involving the ipsilateral paramedian pontine reticular formation or the abducens nucleus with interruption of ipsilateral median longitudinal fasciculus causing failure of adduction of ipsilateral eye. The main causes are demyelination and stroke. Uncommon causes include tumors, AV malformations, basilar artery aneurysms, vasculitis, brainstem tuberculoma and neurocysticercosis.

CASE

A 25-year-old female was referred from the inpatient neurology department for an ophthalmic evaluation. She complained of an inability to move the eyes for the past 2 weeks and vertigo on looking to the left. On examination, the patient was conscious, cooperative, responding to verbal commands. She had a nasogastric tube in situ due to associated dysphagia. Bedside visual acuity was brisk finger counting at 6 meters in both eyes. Pupils were normal in size and reacting normally to light. Ocular motility examination revealed a left exotropia in primary position. There was a total loss of horizontal movements in the right eye with loss of adduction in left eye (Figure 1). On attempted right gaze, there was widening of palpebral fissure in both eyes and she was able to get the left eye just short of midline. The only preserved horizontal movement was abduction in left eye accompanied by abducting nystagmus. The loss of movements was not overcome by Vestibulo ocular reflex. Convergence was normal. Vertical eye movements were within normal limits. Anterior segment and fundus were within normal limits.

INVESTIGATIONS

MRI brain revealed a hypo intense lesion in the dorsal pons on the right side in T2 weighted images indicative of a right sided pontine haemorrhage (Figure 2).

INTERVENTION

The patient was under follow up for 4 weeks under Neurology Department. Dysphagia improved completely though there was little recovery of ocular movements. The patient was diplopia free in primary position.

COMMENT

Supranuclear horizontal eye movements are mainly controlled in the pons. The final common pathway is the abducens nucleus and paramedian pontine reticular formation. The combination of ipsilateral horizontal gaze palsy and internuclear ophthalmoplegia (INO) is known as one and a half syndrome. This peculiar syndrome was first described by Freeman et al and later dubbed as one and a half syndrome by Fisher.

The syndrome is usually due to a single unilateral lesion of the paramedian pontine reticular formation or the abducens nucleus on one side (causing the conjugate gaze palsy), with interruption of adjacent median longitudinal fasciculus. Reported causes of this syndrome include demyelination as in multiple sclerosis, stroke, tumors, AV malformations, basilar artery aneurysm and rarely vasculitis, and neurocysticercosis. Brainstem tuberculoma as a cause of one and a half syndrome has been reported only twice with complete resolution of symptoms following antitubercular treatment.

Pontine hemorrhages represent only 5% of intracranial haemorrhage. The prognosis and clinical picture and recovery largely depend on the volume of insult. While the massive variety produces the classic picture of coma, quadriplegia, decerebrate posturing and eventual demise, the hemipontine syndromes have preserved consciousness with gaze paresis.
and motor sparing. Nuclear magnetic resonance imaging allows a definite diagnosis during the acute phase without recourse to invasive investigations.

In primary position, the eyes may be orthotropic or there may be an exotropia of the contralateral eye also called paralytic pontine exotropia. There may be associated dissociated abduction nystagmus and skew deviation. Recovery of eye movement abnormalities occurs with multiple sclerosis and infarction though it may be incomplete and variable.

REFERENCES


Correspondence to:
Dr. Deepali Garg Mathur,
Senior Consultant, Squint and Paediatric Ophthalmology,
Max Multispeciality Hospital, New Delhi, India.
Myopic Strabismus Fixus

Shailja Tibrewal MS, Regina Lalramhluni MS, Rolli Khurana MS, Suma Ganesh MS

Dr. Shroff Charity Eye Hospital, New Delhi, India.

Abstract: Myopic strabismus convergens is a rare restrictive strabismus characterized by large esotropia and limitation of abduction and elevation. It is described as both congenital and acquired forms. This report presents a case of acquired myopic strabismus convergens occurring in a 35-year-old high myope. Various theories have been proposed regarding pathogenesis of this entity in high myopia. Recent MRI studies have helped in understanding the disease process and led to the development of newer surgical modalities. Careful preoperative evaluation and MRI of the orbit ensures correct diagnosis and choice of appropriate surgical procedure.

A 35-year-old male presented to the strabismus clinic of Dr. Shroff Charity eye hospital in May 2017 with complaints of inward deviation of left eye since childhood (Figure 1). He had been using high myopic glasses for the past 30 years. The deviation was progressing with age and had increased after he underwent cataract surgery with intraocular lens implantation in his right eye two years back. He also had a history of cataract surgery without intraocular lens implantation of left eye one year later after right eye cataract surgery. He is a known asthmatic since the past 5-6 years.

His uncorrected visual acuity in right eye was 20/40 and in left eye was finger counting at 1 meter with eccentric viewing due to inability to move the left eye outwards up to midline. His best corrected vision in right eye was 20/30 with a refraction of +1.00DC @ 180 degree. Refraction in left eye was not possible due to extreme position of the globe. Slit lamp evaluation of the anterior segment revealed pseudophakia and aphakia in the right and left eye respectively. Fundus examination of both eyes showed posterior staphyloma, chorio-retinal atrophic patches in the posterior pole and peripapillary area and mild pallor of the optic disc. The axial length in right eye was 35.72 mm and left eye was 36.06 mm.

Orthoptic examination revealed bilateral convergent strabismus fixus. The patient maintained a small right face turn while reading the distance chart. The left eye was almost immobile with negligible movement in abduction, elevation and depression. The right eye showed limitation of elevation (-3) and abduction (-2). The strabismus was estimated by Hirschberg reflex to be around 15 degrees esotropia in his right eye and >70 degree esotropia and 10 degree hypotropia in his left eye (Figure 2).

Figure 1: Primary gaze photograph showing inward deviation of both eyes, the left eye being in an extreme position.

Figure 2: Nine gaze clinical photograph of the patient showing extremely large angle esotropia and left hypotropia with limitation of movement (left > right).

Magnetic resonance imaging of the orbits revealed bilateral posterior staphyloma with elongation of the globes in supero-temporal direction. As a result, the lateral rectus was seen plastered to the lateral orbital wall (Figure 3). Coronal

Figure 3: Axial scan through mid-orbit showing posterior staphyloma and superotemporal elongation of the globes (asterisk). The lateral rectus is plastered to the lateral orbital wall (bold arrow).
sections of the MRI showed that the superior rectus (SR) muscle path was deviated nasally and lateral rectus (LR) muscle path was deviated inferiorly in both eyes (Figure 4).

Based on history, clinical findings, axial length, and MRI reports, a diagnosis of myopic strabismus fixus convergens was made. Bilateral modified Loop myopexy of LR and SR with medial rectus recession is planned for the correction of deviation. A discussion of the various pathological mechanism and treatment modalities for myopic strabismus fixus follows below.

COMMENT

Myopic strabismus fixus (MSF) convergens is a rare type of acquired strabismus seen in high myopes wherein one or both eyes are fixed in extreme adducting position. In the more common form, however, the patient presents with esotropia and hypotropia of the affected eye with limitation of elevation and abduction. Congenital type of strabismus fixus is thought to be caused due to congenital fibrosis of extraocular muscles and subsequent loss of their elasticity. Villasecca and Martinez were the first to describe strabismus fixus as an acquired condition due to fibrosis of the medial rectus as a consequence of lateral rectus paralysis.

Thereafter several theories have been proposed regarding the pathogenesis of this type of motility derangement. Bagolini et al suggested myopathic paralysis of lateral rectus due to compression against orbital wall and enlarged globe. Hugomier and Magnard proposed a theory of myositis of the lateral rectus muscle. Aydin et al proposed the long globe theory wherein there is mechanical limitation especially to abduction due to the large globe filling the entire orbit and subsequently long standing esotropia causing foreshortening of the medial rectus muscle. Additionally, certain case reports of strabismus convergence fixus occurring in cases with amyloidosis of lateral rectus muscle and mitochondrial myopathy and non-myopic subjects have led to proposition of alternate mechanisms.

Recently, the more widely accepted theory for strabismus fixus in high myopia is the displacement of the paths of the lateral rectus and superior rectus muscle as a result of differential elongation of the globe in the supero-temporal region. Herzau and Loannakis were the first to observe an abnormal path of the LR in MSF intra-operatively in 1996. This was later demonstrated in magnetic resonance scans of the orbit of patients with myopic strabismus fixus by Krzizok et al.

Based on the above theory, Krzizok et al. treated MSF by performing a large recession of medial rectus and fixing the lateral rectus with posterior fixation suture in physiological meridian with non-absorbable suture after resection. However, certain patients with MSF also displayed restriction of elevation and consequent hypotropia. It was Yokoyama et al who in 2001 demonstrated the nasal-ward shift in path of SR along with inferior shift in path of LR in high myopia. Several other studies thereafter found similar pathology and supported Yokoyama’s theory.

Surgical modalities for treatment of MSF can be grossly divided into two groups. Firstly, those procedures that alter the muscle forces which include the classical recess-resect procedure. The second group consists of procedures that aim to correct the deviated muscle paths by either muscle belly union, myopexy of LR at equator or transposition surgery. The choice of the procedure is guided by the amount of limitation of abduction and elevation, angle of esodeviation and the amount of deviation of LR and SR path deviation based on MRI evaluations. In general, belly union or transposition procedures are preferred when the limitation of movement is so severe that the eye fails to cross midline. Yokoyama et al were the first to perform a full loop myopexy of lateral rectus and superior rectus 15 mm behind the insertions using a polyester suture. Suture loop myopexy poses the potential complications of muscle cheese wiring and/or strangulation of the anterior ciliary vessels. Therefore, other materials have been used to prevent the above. Wong et al did loop myopexy of lateral rectus and superior rectus using a silicone 240 band and tightened it with a sleeve. Shih et al used Gore-Tex sling to unite the LR-SR bellies. Shenoy et al modified loop myopexy by Wong by passing the 240-silicone band through a scleral tunnel. This additional step secures the silicone band to the sclera, thereby preventing its anterior migration.

For the present case, the plan of bilateral modified loop myopexy by scleral fixation along with bimedial rectus recession has been made since the angle of deviation is extremely large and the LR and SR muscle paths are deviated significantly on both sides. To conclude, myopic strabismus fixus is a fairly complex strabismus entity. MRI of the orbit forms an important part of diagnosis and further plan of management. Surgical procedure should be decided based on the amount of deviations, grade of limitation of movement and anatomical factors. Recent advances in the understanding of the pathophysiology of the condition have led to development of successful treatment modalities.

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Figure 4: Coronal scan through the orbit showing inferior displacement of lateral rectus (bold arrow) muscle path and nasal displacement of superior rectus (asterisk) and inferior rectus muscle paths.
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Correspondence to: Dr. Shailja Tibrewal, Consultant, Pediatric Ophthalmology and Strabismus Services, Dr Shroff Charity Eye Hospital, Daryaganj, New Delhi, India.
An Interesting case of Head Tilt

Elizabeth Joseph K, Joseph John
Little Flower Hospital, Angamaly, Kerala, India.

Summary: We report the case of a two year old child who had 3 episodes of acquired Brown’s syndrome. He had abnormal head posture in the form of face turn to left and chin elevation. He had left hypertropia, restriction of levoelevation of right eye. Imaging studies revealed bulky right superior oblique tendon and sinusitis. ANA, RA factor were negative. We treated with systemic and periorbital steroids as well as systemic antibiotics to which the child responded remarkably well.

Acquired Brown’s syndrome is not so uncommon. Abrams MS has reported progressive Brown’s syndrome in a two-year-old girl. Depras A et al diagnosed a case of idiopathic acquired Brown’s syndrome in a ten-year-old boy. Rao VB et al has reported acquired Brown’s syndrome in 6 patients who had cysticercosis. Kraft SP et al. has described acquired Brown’s syndrome in six patients aged between 2-8 years. Acquired Brown’s syndrome is also described in a nine-year-old boy with post-streptococcal reactive arthritis and in a child with juvenile idiopathic arthritis.

CASE REPORT

A 2-year-old child presented to us with abnormal head posture (Figure 1) of three days duration. He had fever and upper respiratory tract infection 1 week prior to this episode.

On examination, visual acuity was 6/7.5 both eyes with Cardiff chart. He had a face turn to left with chin elevation. Corneal reflex test showed left hypertropia of 10 degrees (Figure 2). We noticed -4 limitation of laevoelevation of right eye (Figure 3). Lang test showed stereoacuity of 550 seconds of arc. Krimsky test showed 20 prism diopters of left hypertropia.
The left hypertropia was more on laevoversion and on head tilt to left.

Pupil was 3mm round, direct and consensual reflexes were brisk in both eyes. There was fundus incyclotorsion in right eye, but was otherwise normal. Forced duction test was positive indicating restrictive strabismus. Family album scan showed normal head posture, orthophoria and full ocular movements in the past (Figure 4).

Hematology shows relative lymphocytosis, normal peripheral smear. Erythrocyte sedimentation rate (ESR) is 25mm/ 1st hour. Antinuclear antibody (ANA), Rheumatoid factor (RA) are negative. CT Brain and Orbits show bulky right superior oblique tendon, mild surrounding fat stranding in the region of trochlea and right maxillary sinusitis (Figure 5).

We consulted with Pediatrician, Pediatric rheumatologist, Neurologist, Otorhinolaryngologist and started treatment with intravenous hydrocortisone for 3 days and crystalline penicillin for 5 days. He did not show any improvement after 3 days. We gave triamcinolone injection 1ml in the right paratrochlear area. 2 weeks later the child showed remarkable improvement with no limitation of laevoelevation of right eye (Figure 6). On follow up at 4 weeks, the results were maintained (Figure 7).

A year later, he came back with face turn to left and chin elevation. He had left hypertropia of 10 degrees and -4 limitation of laevoelevation of right eye. Prism cover test showed 16 prism dipters left hypertropia. Investigations revealed normal peripheral smear and ESR 16mm/ 1st hour with negative RA factor negative. CT scan showed normal paranasal sinus and no evidence of thickening of right superior oblique tendon. We treated with intravenous hydrocortisone, crystalline penicillin as well as triamcinolone injection in the right paratrochlear area. The child started to improve and 2 weeks later he recovered completely.

1 year later, he again came back with the same head posture (Figure 8), left hypertropia, -4 limitation of laevoelevation of right eye (Figure 9). Stereoscopic acuity was 550 seconds of arc with Lang’s test.

ESR was 15mm/ 1st hour, ANA and RA factor negative.

MRI at this stage showed bulky right superior oblique tendon (Figure 10).

We treated with paratrochlear triamcinolone injection and systemic antibiotics. 1 week later since there was no improvement, we gave intravenous hydrocortisone for 3 days. He responded very well (Figures 11,12) and we tapered the steroid. Two weeks later he had full recovery (Figures 13,14).

DISCUSSION

Patients with acquired Brown’s syndrome should be evaluated medically for coexisting systemic disease. If a disorder, such as rheumatoid arthritis
or sinusitis, is identified, that has to be treated accordingly. Once systemic disease is excluded, patients who have acquired Brown syndrome with signs of inflammation can be treated with anti-inflammatory medication. Oral ibuprofen is a good first-line choice. Local steroid injections in the area of the trochlea and oral corticosteroids can be used for inflammation. Once the inflammatory disease process is controlled, patients with inflammatory Brown syndrome may show spontaneous resolution.

REFERENCES


Correspondence to:
Dr. Elizabeth Joseph,
Chief, Pediatric Ophthalmology and Strabismus,
Little Flower Hospital,
Angamaly, Kerala, India.
A comitant esotropia, with no limitation of ocular movements, is generally thought to be benign and does not warrant any neurological workup. However, there are reports of intracranial lesions presenting with comitant nonaccommodative esotropia without neurological deficit in early stage. Sometimes acute-onset nonaccommodative esotropia may be the only presenting sign of an intracranial neoplasm. A careful history and a meticulous clinical evaluation will pick up the red flags in an atypical case and prompt the need for neuroimaging to confirm the cause.

CASE REPORT

A 9-year-old boy with Right esotropia was referred from Middle East for squint surgery (Figure 1,2). According to the parents, esotropia was first noticed at the age of 4 years and since then he has been on occlusion therapy. He seemed to be a very active, playful child with no systemic diseases. Visual acuity was 6/7.5, N6 OD and 6/6, N6 OS respectively. Worth Four Dot Test showed alternate suppression. Stereopsis was nonrecordable. Pupils were normal in size and reaction. Ocular movements appeared full OU. Alternate Prism Bar Cover Test showed 40 PD esotropia for distance and 20 PD esotropia for near with either eye fixing separately. Cycloplegic retinoscopy was +1.00DS OU. Dilated fundus examination showed large discs with pallor. Systemic examination did not reveal any focal neurological deficit. Optical Coherence Tomography (OCT) showed thinning of temporal retinal nerve fibre layer (RNFL) both eyes. MRI Brain showed hyperintense lesions extending from bilateral optic tracts to tectum of midbrain up to pons. MR Spectroscopy showed increased Choline with reduction in NAA (N-Acetyl Aspartate) suggestive of glioma.

Figure 1,2

Figure 3: Extraocular movements of the child

Summary: We report a case of CNS Glioma masquerading as Acquired Comitant Nonaccommodative Esotropia (ANAET) in a 9-year-old boy who was referred to us for squint surgery. He had visual acuity of 6/7.5, N6 OD and 6/6, N6 OS respectively. Worth Four Dot Test showed alternate suppression. Stereopsis was nonrecordable. Pupils were normal in size and reaction. Ocular movements appeared full OU. Alternate Prism Bar Cover Test showed 40 PD esotropia for distance and 20 PD esotropia for near with either eye fixing separately. Cycloplegic retinoscopy was +1.00DS OU. Dilated fundus examination showed large discs with pallor. Systemic examination did not reveal any focal neurological deficit. Optical Coherence Tomography (OCT) showed thinning of temporal retinal nerve fibre layer (RNFL) both eyes. MRI Brain showed hyperintense lesions extending from bilateral optic tracts to tectum of midbrain up to pons. MR Spectroscopy showed increased Choline with reduction in NAA (N-Acetyl Aspartate) suggestive of glioma.

Glioma Masquerading as Acquired Non-accommodative Comitant Esotropia in a Child

Neena R., A. Giridhar

Giridhar Eye Institute, Kochi, Kerala, India.
Undilated fundus showed large discs with temporal pallor both eyes.

**Investigations:** Family album showed that child had straight eyes in early childhood and the sudden onset of squint at 4 yrs. of age (Figure 4). OCT RNFL showed thinning in temporal quadrants OU and inferior quadrant OD (Figure 5).

MRI Brain and Orbits were done which showed predominantly white matter lesions involving bilateral optic tracts, extending to tectum of midbrain up to pons on left side, mild enhancement of both thalami with subtle contrast enhancement and no diffusion restriction (Figure 6). There was mild hyperintensity noted extending to right temporal white matter lateral to temporal horn (Figure 7). MR Spectroscopy showed increased choline with reduction in NAA (N-acetyl aspartate) (Figure 8).

**Diagnosis:** CNS Glioma; possibly Low grade.

**Management:** Patient was referred to Paediatric Neurosurgeon and Paediatric Oncologist; found to have Neurofibromatosis-Type I. He was advised to undergo biopsy and chemotherapy but parents refused. He is currently on follow up.

**DISCUSSION**

Acquired nonaccommodative esotropia (ANATED) is a less common disorder of ocular alignment characterized by a nonaccommodative esodeviation which can occur in older children, adults, and even the elderly. Onset can be acute and associated with diplopia when it is called Acute acquired comitant esotropia (AACE) or it can result from deterioration of existing, previously controlled, esotropia. AACE is classically divided into five different subtypes. The Swan type (type I) occurs after a period of interrupted binocularity. Type II AACE, known as Burian-Franceschetti has minimal hypermetropia and diplopia that is often associated with physical or psychological stress. The Bielschowsky type (type III) is associated with patients with myopia, convergence spasm and divergence paralysis. Type IV / Refractive-accommodative type is characterized by high hypermetropia that can be adequately controlled with the refractive correction alone. Type Va, lesser common entity, is associated with intracranial pathology, most commonly a posterior fossa lesion.

Red Flags in Our Case which prompted us to image:
1) Sudden Onset of Esotropia at the age of 4.
2) Normal Alignment Prior to 4 Years Noted in Family Pictures.
3) Large Distance Near Disparity
4) Temporal Pallor OU (possibly due to involvement of optic tracts)
5) OCT Showing RNFL thinning

Many brain tumors such as cerebellar astrocytomas, medulloblastomas, pontine gliomas, astrocytoma of the corpus callosum (with hydrocephalus) and Arnold Chiari Malformation are associated with acute acquired comitant esotropia in childhood without any neurological deficit. Sometimes acute-onset nonaccommodative esotropia is...
the only presenting sign of intracranial neoplasm. Tumors involving the corpus callosum and posterior fossa, especially gliomas give rise to no distinctive signs and are typically known to present with only acute-onset comitant esotropia with no other systemic signs such as papilledema. It is important in these cases of acquired esotropia to rule out the possibility of intracranial tumor, Arnold Chiari malformation or other neurological abnormalities.

N-acetyl aspartate (NAA) is an indicator of neuronal density. Gliomas have disruption of neuronal integrity with axonal loss and thus reduced NAA. Choline is a compound involved in membrane synthesis and degradation. Choline peak correlates with cell density and increased choline is seen with increased membrane turnover. As grade of glioma increases, choline levels increase.

The exact mechanism responsible for acute comitant esotropia in patients with brain tumors is not clear. Comitant strabismus might result from involvement of supranuclear mesencephalic structures, which control vergence eye movements. Some have ascribed acquired comitant esotropia to infranuclear insults, such as varying degrees of bilateral sixth nerve paresis. Spread of comitance is another suggested mechanism.

Although most children with this form of esotropia are otherwise healthy, central nervous system lesions must be considered and ruled out before planning surgery. If you miss a brain tumor in a child, it may prove costly. So, we feel that neuroimaging should be done in any patient presenting with acquired comitant nonaccommodative esotropia, especially when there is a high index of suspicion.

REFERENCES

10. Campos EC. Why do the eyes cross? A review and discussion of the nature and origin of essential infantile esotropia, microstrabismus, accommodative esotropia, and acute comitant
A 56-year lady presented with poor vision in right eye since childhood. She had been refused therapy by various ophthalmologists for her ‘Lazy Eye’ since she was considered past the ‘treatable age’. A detailed ophthalmological assessment of her visual status, Best Corrected Visual Acuity for distance and near was done. She was found to have anisomyopic amblyopia. Full-time occlusion therapy was started. Within 6 weeks, her near vision improved from worse than N18 to N6 while her distance vision improved from 0.1 to 0.8 (decimal fraction) in further 3 months. Her progress was monitored for 2 years, and no regression in visual acuity was noted.

Amblyopia or lazy eye, is a disorder of the visual system in which visual loss is out of proportion to any structural abnormality in the eye. It results from disuse of an eye, either due to an inadequate foveal or greater peripheral retinal stimulation (where there is a lesser concentration of cones), or due to an abnormal binocular interaction resulting from variable visual inputs from both fovea. Human brain is designed to allow both eyes to function together to explore space. If signals from one eye are blurred or absent, brain blocks visual input from that eye. In the neuronal visual pathway, the synapses are broken due to disuse of the amblyopic eye. This may occur due to constant strabismus, disproportionately high refractive error in one eye, a combination of both factors, or blocked vision in an eye due to a droopy upper lid, media opacity like cornea/vitreous or congenital cataract.

Amblyopia has been estimated to affect 1–5% of the population. It is generally believed to be fully treatable only till the age of 6-8 years; beyond that age only some visual improvement is considered to be possible. Many clinicians refuse therapy after the age of 8-12 years.

An adult person with unilateral amblyopia is at three times greater risk and a child, 17 times that of a normal person for losing vision in the better eye. There are anecdotal evidences of spontaneous improvement of vision in an amblyopic eye after loss of vision in the good eye. This spontaneous improvement of visual acuity to a usable level (6/24 or better) is relatively low (<17%) unless a complete visual loss occurs in the better eye.

Full-time occlusion therapy for treating amblyopia has been practiced since decades. The case discussed here shows that it can result in improvement of visual acuity at any age.

CASE REPORT

A 56 years old lady presented at our tertiary care centre accompanying her 6 years old grandson for the treatment of his lazy eye. While the grandson was being explained the rational of therapy, she queried if it was possible to treat her ‘Lazy Eye’ as well at her age, though she had been refused therapy by various ophthalmologists since her childhood.

A detailed ophthalmological assessment including her visual status, Best Corrected Visual Acuity (BCVA) for distance and near, pupil, colour vision, stereopsis, status of muscle balance by cover-uncover test, slit-lamp and fundoscopy, assessment of foveal fixation by visuoscope was done. She was found to have a BCVA of 0.9 (Decimal fraction on TSCP-700 Chart Projector) OD and Counting finger (CF) OS with a correction of OD = -0.5DS and OS = -5.50DS / -2.00Dcyl@90. She could only read a font much larger than N18 with OS (newspaper headlines only with difficulty), with no improvement for near after an add of +2.5DS into her distance correction. She was prescribed glasses for full-time use and asked to return for follow-up after 1 month.

On first follow-up, her BCVA improved from CF to 0.1 so she was asked to continue with glasses only for a further period of one month. On second follow-up, her BCVA had not shown any improvement. She was prescribed full-time occlusion therapy of her good eye along with active usage of the amblyopic eye by forced reading, writing for 4-5 hours per day. She was explained to start reading a large font that she was comfortable with and shift to smaller fonts every day. The need for regular follow-up was stressed. A written consent form was obtained and commercially available eye patches to be worn over good eye were prescribed for all waking hours daily, to be removed at night while going to sleep. After 2 weeks of starting full-time occlusion therapy, her near vision improved to N18 but there was no improvement in distance vision. On further follow-up, the near vision showed a steady, gradual improvement to N6 with 6 weeks of occlusion therapy and the distance vision also improved to 0.2. With regular follow-up, the distance vision also showed a gradual improvement and after 4 months of full-time occlusion therapy, her BCVA for distance vision was 0.8 while the good eye VA remained at 0.9. After that, a gradual weaning protocol of occlusion therapy was started with 1 day off-patching in first week with 6 days full-time patching, 2 days off-patch in the second week with 5 days full-time patching and follow-up after every 2 weeks. Since the BCVA was maintained at 0.8, weaning schedule was continued till patching was totally off after 7 weeks. She was kept under regular follow-up for 2 years and no regression in BCVA was noted. She was strictly counselled to wear spectacle correction and have a regular follow-up.

REVIEW

In general, the amblyopic adult patients are refused therapy, she queried if it was possible to treat her ‘Lazy Eye’ as well at her age, though she had been refused therapy by various ophthalmologists since her childhood.

A detailed ophthalmological assessment including her visual status, Best Corrected Visual Acuity (BCVA) for distance...
therapy beyond a certain age because of misinterpretation of “Critical Period for Visual Development.” According to Wiesel and Hubel, this period exists from 1-5 years of age. It means that during this period, an individual’s retina and brain are most sensitive to outside environment and stimuli than at other periods of life. This does not mean that the visual cortex becomes unresponsive to visual stimulation once that period is over and the “gates’ leading from retina to visual cortex close tightly; rather these gates remain very slightly open and get rustic by disuse.

Recent findings of neuroplasticity have shown that brain is not a physiologically static organ and it can modify throughout lifetime. Its development does not end after a certain age; it can be stimulated to form new connections between existing brain cells and strengthen older ones in any part of the brain, any time in an individual’s life by strong, persistent and appropriate stimulation. This ability is strong in early childhood when maximum brain growth occurs, slows down with age, but it never stops. The molecule responsible for neuroplasticity is a protein receptor, which is in an “OFF” mode in adults but can be turned “ON” by continued, active brain stimulation. GABA (Gamma Amino Butyric Acid) acts as an excitatory neurotransmitter in immature, developing brains and regulates proliferation of neural progenitor cells, proliferation and elongation of neurons and formation of synapses by releasing Brain-Derived-Neurotrophic Factor. This results in important brain functions like memory, learning, speech, motor control. It not only gradually decreases with age but in mature brains, it has an inhibitory effect by activating GABA-receptors and causing cell arrest in the S-phase (static phase). GABA given exogenously cannot cross the blood-brain barrier. Researchers obtained GABA secreting neurons from young mice while they were in their “critical period” and transplanted into the brains of adult, amblyopic mice. After some time, they found new neural connections forming in the visual pathway and restoration of normal eyesight in those adult, amblyopic recipient mice. Similarly, in other studies, plasticity of brain was shown to improve in specific regions by a specified stimulus. The brain receptors which were turned “Off” with age, could be turned “On” by GABA released in response to a stimulus. An increase in the gray matter volume has been observed in professional typists due to long-term bimanual typing, suggesting that learning can affect not only function but brain structure as well in adults.

Dopamine is another neurotransmitter that stimulates receptors and turns them “On.” It is present in retina and cerebral cortex but does not cross the blood-brain barrier. Its precursor, Levodopa, crosses that barrier and is converted to Dopamine in the brain. These studies prove that neural stem cells (progenitor cells) can be made to generate neurons in various brain areas of mammals. Adults continue to learn throughout life and this is because of continued neurogenesis in the memory area.

Hence, in case of amblyopia the closed, rustic gates in the visual pathway can be fully opened and turned active again but this needs a strong persistent stimulation, without any inhibitory influence. The brain favors neural transmission from the good eye; it is a known fact that the good eye has an inhibitory influence over the amblyopic eye. As shown in this case-report, full-time occlusion of the good eye removed this inhibitory effect over the development of neural connections of the amblyopic eye for the whole duration of therapy. This was combined with the active use of amblyopic eye till neural connections became fully functional. Once that was achieved, the connections were given adequate time to stabilize to avoid regression of amblyopia; this was provided by following a very slow and gradual weaning protocol for occlusion therapy in this case.

This case report proves the concept of neuro-plasticity and shows that amblyopia in adults can be fully treatable. But this needs highly motivated and inspired patients who are ready to cooperate and comply with therapy. They must manage their lives for a period of 2-3 months by keeping their good eye closed and actively using an eye that had been neglected for decades. This is not an easy job either for the patient or the treating ophthalmologist, but this is the only way to ensure full visual recovery by very simple means with no economic burden either on the patient or the health services. Once the visual recovery is achieved, it may be permanent.

REFERENCES


Correspondence to: Dr. Sameera Irlan, FRCS
Pediatric Ophthalmologist & Oculoplastic Surgeon, Lahore, Pakistan.
Utility of Ice Pack Test in Diagnosis of Myasthenia Gravis

Mandeep Tomar MS, Gaurav Sharma MS, Nikhil Verma MBBS, Richa Dhiman MBBS, Vinod Dhiman

Dr. Rajender Prasad Government Medical College, Tanda, Himachal Pradesh, India.

Summary: Ice pack test is a simple clinical test performed for Myasthenia gravis (MG) in the office and is of great value in diagnosing and minimizing costly neuroimaging referrals. A 16 year old male child presented with hoarseness, dysphagia and bilateral ptosis after having local honey and meals in neighbourhood. Marginal Reflex Distance was 1mm in right eye and zero mm in the left. Possibility of botulism was kept. Ice test for 3 minutes was positive. MG was suspected and confirmed by positive edrophonium test. MG suspects should undergo complete ophthalmological examination with ice pack test to differentiate from other neurological mimickers.

Myasthenia gravis is an autoimmune disease in which antibodies bind to acetylcholine receptors or to functionally related molecules in the postsynaptic membrane at the neuromuscular junction. The antibodies induce weakness of skeletal muscles, which is the sole disease manifestation. The weakness can be generalized or localized, is more proximal than distal, and nearly always includes eye muscles, with diplopia and ptosis. With an annual incidence of 8 to 10 cases per 1 million persons and a prevalence of 150 to 250 cases per 1 million, myasthenia gravis and its various subgroups are the major diseases that affect the neuromuscular junction. The Lambert-Eaton myasthenic syndrome and neuromyotonia are additional, rare, presynaptic autoantibody disorders characterized by skeletal-muscle dysfunction. Congenital myasthenic syndromes and toxin-induced conditions (e.g., botulism) can also affect the neuromuscular junction and lead to muscle weakness.

Case report

A 16 years old male child presented in paediatrics routine OPD with insidious onset of inability to speak properly, hoarseness and dysphagia since last 7 days following meals and local honey in the neighbourhood. It was soon followed by asymmetrical drooping of bilateral eyelids since last 3 days. There was no history of associated diplopia and restriction of ocular movements. Child also complained of weakness of both upper limbs which was followed by bilateral lower limbs weakness since the last 1 day. There was history of difficulty in lifting heavy objects and climbing upstairs. There were no diurnal variations, no aggravating and relieving factors, no history of vomiting, diarrhoea, fever or any drug intake.

A provisional diagnosis of botulism with descending paralysis with multiple cranial nerve palsies in the form of 3rd, 5th, 7th and 10th cranial nerve, was kept in paediatrics department, keeping in mind the history of food ingestion associated with it and child was referred to department of ophthalmology for further evaluation.

On ocul ar examination, visual acuity was 6/9 (p) both eyes improving to 6/6 with pin hole. Pupils were reactive and ocular movements were full. There was severe bilateral ptosis with palpebral aperture 8mm and 7mm in right and left eye respectively, marginal reflex distance of 1mm in the right eye and zero in the left. There was a good LPS (levator palpabrae superioris) action. Ice pack test for 3 minutes was done as a part of routine ptosis workup and came out to be positive (Figure 1). After positive ice pack test, a strong possibility of myasthenia gravis was suspected and further investigations were carried out.

Cogan lid twitch sign and lid fatigability test were also positive bilaterally. Neostigmine test was done in neurology department under anaesthetist supervision with monitoring of cardiovascular function, and was found positive (Figure 2).

Anti-acetylcholine esterase antibody levels (binding type) were negative i.e. value<0.15 nmol/l (normal range 0.0-0.4 nmol/L) and anti-muscle specific kinase levels (Musk) were normal. CT thorax to rule out thymoma revealed a normal study. Thyroid profile revealed decreased levels of T3 [77 ng/dl (normal 80-180 ng/dl)], T4 [7nmol/l (normal 58 - 161 nmol/l)] and elevated TSH [6.8mIU/L (normal-0.4-4 mIU/l)]. Lactate dehydrogenase levels were 404 u/l (normal-140-280 u/l), rest of the investigations were normal.

Ultrasonography
of the neck was in view of a midline swelling in the neck which revealed heterogeneous echotexture with diffuse hypoechoegenicity in the thyroid gland suggestive of thyroiditis.

A final diagnosis of generalized myasthenia gravis with ocular involvement was made in consultation with neurology department and child was started on oral pyridostigmine 60 mg 3 times/day and 180 mg (sustained release) bedtime and tablet azathioprine 50 mg once a day. Child was kept under follow up with marked recovery on subsequent visits.

**REVIEW**

Myasthenia gravis (MG) is an autoimmune disorder characterized by reduction in the acetylcholine receptors (AChRs) at neuromuscular junctions due to the effects of autoantibodies. It is characterized by a variable weakness of skeletal muscles, which improves on resting. Weakness is exacerbated by repetitive contraction. Generalized myasthenia involves the bulbar, limb, and respiratory muscles; ocular myasthenia gravis (OMG) is a subtype of MG where the weakness is clinically isolated to the Extraocular muscles (EOMs), levator, and orbicularis oculi. The pattern of involvement is usually symmetric, apart from the eye involvement, which is often markedly asymmetric and involves several eye muscles. In 15% of all patients with myasthenia gravis, symptoms and signs are confined to ocular muscles. Ptosis and diplopia are common initial deficits and ptosis must be measured before and after the test. Although there are no strict guidelines regarding the interpretation of this test it is, usually, considered positive when the upper eyelid elevates by at least 2 mm following ice application. Cooling may reduce cholinesterase (ChE) activity, which increases the amount of available Ach at the neuromuscular junction. There is thus an increase in the efficiency of ACh in eliciting depolarization at the motor end plate. Resolution of ptosis has been reported in over 90% of OMG patients after the ice test. According to one study, the sensitivity and specificity of this test was 76.9% and 98.3%, respectively.

Among electrodiagnostic tests, Repetitive Nerve Stimulation (RNS) (2 to 3 Hz) is the most frequently used electrodiagnostic test for MG. The nerve to be studied is electrically stimulated 6-10 times at 2 or 3 Hz (slow rate) with a supramaximal stimulus and the compound muscle action potential (CMAP) is recorded with surface electrodes. In MG, the number of individual muscle fiber action potentials reduces, the CMAP also reduces in both amplitude and area with a resulting decremental response. In MG, a characteristic decrement (>10%) in muscle action potential amplitude is typically seen by the fourth or fifth response in a series of low-frequency RNS, whereas the amplitude remains the same in normal individuals. This decremental response is seen in only 33% of patients with pure OMG. A decremental response to RNS is not specific and may also be seen in Lambert-Eaton myasthenic syndrome, motor neuron diseases, and myopathies.

Conventional electromyography (EMG) is not useful for the diagnosis of myasthenia, but may be indicated in these patients when concurrent neuromuscular disease is in question. Single fibre EMG (SFEMG) needs more patient cooperation than do these tests. The diagnostic yield in muscle action potential amplitude is significantly higher. Some patients considered to have myasthenia gravis do not show any abnormalities with this technique, particularly those with the pure ocular form.

<table>
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<th>Differences between myasthenia gravis and botulism</th>
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presenting physician, due to abrupt onset of the disease, history of intake of contaminated meals associated with complaints, facial picture mimicking multiple cranial nerve palsy and motor neuropathy, absence of diurnal variations and simultaneous onset of bilateral ptosis. Thorough and meticulous ophthalmological examination with ice pack test helped in this case to make the final diagnosis.

REFERENCES


A Case of Congenital Third Cranial Nerve Palsy

Komal Batra, Sandra Ganesh

Aravind Eye Hospital, Coimbatore, India.

Summary: Congenital third nerve palsy can present with a myriad of features such as ptosis, pupillary involvement, amblyopia, partial or complete loss of function of the extraocular muscles, leading to ocular motility defects. A 23-year-old female presented to us with limitation of movements of her left eye. Based on clinical findings of severe limitation of adduction of the left eye and mild ptosis, a diagnosis of congenital third nerve palsy with residual ptosis and exotropia was made. Surgical correction by fixating the globe to medial orbital wall was attempted, aiming to fix the eye in primary position of gaze.

Third cranial nerve supplies four of the six extraocular muscles and is associated with presence of associated factors such as ptosis, amblyopia, pupillary involvement, aberrant regeneration, lateral rectus muscle contracture leading to ocular motility defects. Third nerve palsy can be isolated or can occur in association with other ocular nerve palsies. Isolated third nerve palsy may be unilateral or bilateral, complete or partial, pupil involving or pupil sparing, and congenital or acquired. Surgery is the mainstay of treatment for management of unrecovered third nerve palsy. Surgical management is challenging and is different for partial and complete third nerve palsy. For partial involvement, surgical intervention is according to the nature and extent of extraocular muscle involvement. For complete third nerve palsy, supramaximal recession-resection of the recti, superior oblique tendon transposition, lateral rectus transposition or globe anchoring procedures are attempted.

CASE REPORT

A 23-year-old lady presented with limitation of movements of her left eye. She had undergone 9mm lateral rectus (LR) recession in her left eye in February 1996 and two Frontalis sling operation were performed elsewhere subsequently for left upper lid ptosis in September 1997 and December 2000 to prevent amblyopia. On examination, her visual acuity was 6/6 in right eye and 6/9 in her left eye improving to 6/6 with glasses. Left eye showed mild upper lid ptosis, 15 degrees of exotropia on Hirschberg's corneal light reflex and absent adduction on cover test. Movements were restricted in all direction of gaze in her left eye with severe limitation of adduction (-4) and inability to move the left eye to midline. There was >50 prism diopters of exotropia of her left eye on modified Krimsky test. Right eye motility was normal (Figure 1). When fixing with involved eye, there was marked secondary deviation in the right eye. There was no evidence of aberrant regeneration. She was diagnosed with congenital oculomotor nerve palsy with residual ptosis and residual exotropia. As she wanted cosmetic correction, she was operated for residual exotropia with her left eye medial rectus (MR) fixated to medial palpebral ligament under GA to fix the globe in primary position of gaze.

Through a medial limbal conjunctival incision, medial rectus (MR) muscle was isolated (Figure 2). A transcutaneous incision was made over the anterior lacrimal crest and insertion of medial palpebral ligament (MPL) was exposed by blunt dissection (Figure 3). Two arms of double armed 5-0 non-absorbable (ethibond) suture were passed through the superior and inferior part of MPL insertion. The suture ends were tied to each other (Figure 4). An empty semicircular needle was used as a guide to bring out the two needles of the suture onto the subconjunctival space (Figure 5). The sutures were then tied tightly at the insertion of the MR directly onto the sclera, at the same time as the assistant rotated the globe medially thus adducting the eye with the aim of slight overcorrection (Figure 6). Post operatively the eye was aligned in adduction with mild hypertropia.
Management of strabismus due to complete third nerve palsy is a surgical challenge. Due to the unopposed action of lateral rectus and superior oblique muscle, the eye is in a divergent and depressed position. In complete third nerve palsy the goal of surgery is realignment of the eye in primary position, compromising the ocular motility of the involved eye. While the globe is aligned in primary gaze, with very little or no horizontal movement of the eye, no meaningful area of binocular single vision can be achieved.

Surgical options for strabismus resulting from third nerve palsy include a very large supramaximal LR recession (14-16 mm) and MR resection (8-14 mm) for correcting the horizontal deviation in primary position as initially described by Helveston. But the conventional recession-resection procedure usually drifts the eye back to exotropia. Superior oblique tendon transposition procedures to medial aspect of globe were advocated by some workers but the procedure is technically difficult and the results were not found to be satisfactory. Y split and lateral rectus transposition to superior and inferior border of medial rectus muscle was described by Taylor et al.

Globe anchoring procedures are the mainstay of treatment in the absence of significant medial rectus function and in the presence of contracture of the lateral rectus muscle. Disinsertion of the lateral rectus muscle and reattachment to the lateral orbital wall or globe fixation of medial rectus muscle to the medial wall of the orbit at anterior lacrimal crest can be performed. This can be achieved by transcutaneous approach as done in this case or precaruncular approach. It can be achieved by using either 5-0 non-absorbable polyester suture as described by Sharma et al, fascia lata, superior oblique tendon or silicon band.

Though the technique of medial rectus fixation to medial palpebral ligament limits the horizontal movements...
of the eye and reduces the field of binocular single vision, it provides a better cosmetic appearance by fixing the eye in primary position of gaze. This procedure is mainly attempted in cases of congenital third nerve palsy (no binocular vision) for cosmetic alignment as there could be intractable diplopia in patients with acquired palsies.

Figure 9: Three-week post-operative photograph showing Left Eye in Primary Position.

REFERENCES


Correspondence to: Dr. Sandra Ganesh, Consultant, Department of Paediatric Ophthalmology and Strabismus, Aravind Eye Hospital Coimbatore, India.
Q1. Identify the test?

(Q1 image)

Q2. The following features are seen in Leber’s congenital amaurosis except:
   A. Nystagmus
   B. Normal fundus at the early stage
   C. Myopic refraction
   D. Arteriolar narrowing in later stage

Q3. All are features of Blepharophimosis syndrome except:
   A. Autosomal dominant inheritance
   B. Epicanthus inversus
   C. Nasal bridge hypoplasia
   D. Mental retardation

Q4. Identify the disorder -

(Q4 image)

Q5. Identify the disorder -

(Q5 image)

Q6. 9-year-old female with inability to move either eye sideways along with drooling of saliva, since childhood. Figure attached. Identify the possible syndrome.

(Q6 image)

Compiled by:
Guru Nanak Eye Centre, New Delhi, India.

Dr. Yashpal Goel MS
Q7. A 2-week-old infant is suspected of having persistent fetal vasculature (PFV). On anterior segment examination, what findings would support this diagnosis?
   A. Congenital bilateral cataracts
   B. Normal axial lengths
   C. Iris hypoplasia
   D. Anteriorly displaced ciliary processes

Q8. Individuals at greater risk for developing anomalous retinal correspondence include which one of the following?
   A. 7-year-old boy with a history of congenital esotropia
   B. 12-year-old girl with a 3-week history of decreased vision in her right eye
   C. 15-year-old boy with a 1-week history of esotropia
   D. 7-year-old girl with a history of exophoria

Q9. Identify the condition depicted by the photographs

Q10. Which of the following statements does not accurately describe the use of indirect ophthalmoscopy to screen for retinopathy of prematurity (ROP)?
   A. Screening should be performed on all premature neonates of less than 30 weeks gestation.
   B. Screening should be repeated biweekly on neonates who demonstrate any stage of ROP on initial examination.
   C. Screening should be performed before hospital discharge, or by 4-6 weeks of age.
   D. Screening should be performed on all premature neonates with a birth weight <1500 g.
ACROSS
2. Treatment for non refractive accommodative esotropia(7)
4. Press-on prism(7)
5. Syndrome most commonly seen with Posterior embryotoxon(8)
8. Chemotherapeutic agent used in retinoblastoma(11)
9. Neurotoxin for treatment of squint(5)
11. Law violated by DVD(6)
13. Benign vascular tumor of infancy(10)

DOWN
1. 3 step test for vertical deviation(5)
2. Superior oblique tendon sheath syndrome(5)
3. Common cause of proptosis in children(10)
5. Lazy eye(9)
6. Syndrome that violates Sherrington law(6)
7. Head tilt test for SOP(12)
10. Surgery for MED(5)
12. Pharmacological treatment for amblyopia(8)
Delhi Ophthalmological Society

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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)

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Cheque/DD No.________________________________ Dated__________ Drawn on_________________

Signature of Applicant with Date

Three specimen signatures for I.D. Card.

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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)
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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)
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   1. Copy of Degree (MBBS / MD / DNB)
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   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).

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   Room No. 205, 2nd Floor, OPD Block, Guru Nanak Eye Centre, Maharaja Ranjit Singh Marg, New Delhi-110002

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DOS TIMES 2017 – 2019 AUTHOR GUIDELINES

MANUSCRIPT SUBMISSION
DOS Times is published once in two months (i.e six issues in a year: July – August, September – October, November – December, January – February, March – April, May – June). Solicited and unsolicited manuscripts of good quality academics are accepted provided that they are not under consideration for publication in any other journal. All submitted manuscripts are subject to editorial review before acceptance.

You may submit your manuscripts along with a covering letter addressed to

Address for all correspondence
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Secretary - Delhi Ophthalmological Society
Room No 205, 2nd Floor, OPD Block,
Guru Nanak Eye Centre, Maharaja Ranjit Singh Marg,
New Delhi - 110002
or by email to dostimes10@gmail.com

In case of any queries please contact Mr. Sunil Kumar, DOS Times assistant @ 011-65705229 or by email (dostimes10@gmail.com).

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.

MANUSCRIPT SUBMISSION AND PROCESSING
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 disclose 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 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>
<th>Word limit</th>
<th>Reference limit</th>
<th>Abstract</th>
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<tr>
<td>Original article</td>
<td>2500</td>
<td>40</td>
<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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<tr>
<td>Review articles</td>
<td>3000</td>
<td>50</td>
<td>Unstructured 150-200 words</td>
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<tr>
<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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<tr>
<td>Perspectives</td>
<td>1500</td>
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<td>Authors will be asked to give opinion on a topic of interest. These should be evidence based and relevant and give perspective and practical applications to existing knowledge.</td>
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<tr>
<td>Recent advances</td>
<td>1500</td>
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<td>Unstructured 100 words</td>
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<td>Summary of latest in clinical research, instrumentation and web resources in ophthalmology.</td>
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Techniques
Novel surgical techniques or instrumentation that have the potential to reduce surgical complexity and/or enhance outcomes.

Case reports
Interesting cases with immense clinical significance / rare case reports
Subheadings: Introduction, case and discussion

Photo-essay/snap shots
Reports of unusual/uncommon clinical case scenarios with good photographic documentation
Subheadings: Introduction, case and comment

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).
<table>
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<tr>
<th>Study</th>
<th>Aim</th>
<th>Age</th>
<th>No. of patients</th>
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<th>Follow up</th>
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<tr>
<td>ATS 1</td>
<td>Occlusion versus Pharmacologic Therapy for Moderate Amblyopia</td>
<td>3-7yrs</td>
<td>419</td>
<td>Randomised comparative trial</td>
<td>2 year</td>
<td>Improvement was initially faster in the patching group, but after 6 months the difference in visual acuity between treatment groups was insignificant; 3.16 lines (patching group) vs 2.84 lines (atropine group). Visual acuity &gt;20/30 and/or improved by ≥3 lines in 79% of occlusion group and 74% of atropine group.</td>
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<td>ATS1-Ext</td>
<td>Atropine vs patching for treatment of moderate amblyopia: follow-up at 15 years of age of a randomized clinical trial</td>
<td>3-7yrs</td>
<td>188</td>
<td>Randomised comparative trial</td>
<td>15 years of age</td>
<td>At 15 years of age, most children treated for moderate amblyopia when younger than 7 years have good visual acuity, although mild residual amblyopia is common. The outcome is similar regardless of initial treatment with atropine or patching.</td>
</tr>
<tr>
<td>ATS 2A</td>
<td>A Randomized Trial Comparing Part-time Versus Minimal-time Patching for Moderate Amblyopia</td>
<td>3-7yrs</td>
<td>175</td>
<td>Randomised comparative trial</td>
<td>4 months</td>
<td>6 hrs of patching equally effective as full-time patching: 86% pts in the 6-hour group and 82% pts in the full-time group had improved by 3 lines from baseline.</td>
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<tr>
<td>ATS 2B</td>
<td>A Randomized Trial Comparing Part-time Versus Full-time Patching for Severe Amblyopia</td>
<td>3-7yrs</td>
<td>189</td>
<td>Randomised comparative trial</td>
<td>4 months</td>
<td>6 hrs of patching as effective as 2 hrs of patching. At 4 months, no difference in amblyopic eye acuity between groups.</td>
</tr>
<tr>
<td>ATS 2C</td>
<td>An Observational Study on Recurrence of Amblyopia After Discontinuation of Treatment</td>
<td>3-7yrs</td>
<td>156</td>
<td>Observational</td>
<td>52 weeks</td>
<td>Recurrence occurred in 35 (24%) of 145 cases and was similar in patients who stopped patching (25%) and in patients who stopped atropine (21%). In patients treated with moderately intense patching (6 to 8 hours per day), recurrence was more common (11 of 26, 42%) when treatment was not reduced prior to cessation than when treatment was reduced to 2 hours per day prior to cessation. Approximately one fourth of successfully-treated amblyopic children experience a recurrence within the first year off treatment.</td>
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<tr>
<td>ATS 3</td>
<td>To evaluate the effectiveness of optical correction alone vs 2-6 hrs/day of patching combined with near visual activities plus atropine.</td>
<td>7-13yrs</td>
<td>507</td>
<td>Observational phase and Randomised trial phase</td>
<td>6 months</td>
<td>In the 7 to &lt;13 year olds, 53% of the treatment group were responders compared with 25% of the optical correction group (P&lt;0.001). In the 13 to &lt;18 year olds, the responder rates were 25% and 23% respectively overall (adjusted P=0.22), but 47% and 20% respectively among patients not previously treated with patching and/or atropine for amblyopia. For patients 7 to &lt;13 years old, prescribing 2 to 6 hours per day of patching with near activities and atropine can improve visual acuity even if the amblyopia has been previously treated. For patients 13 to &lt;18 years old, prescribing patching 2 to 6 hours per day with near activities may improve visual acuity when amblyopia has not been previously treated but appears to be of little benefit if amblyopia was previously treated with patching.</td>
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<td>ATS 4</td>
<td>A Randomized Trial Comparing Daily Atropine Versus Weekend Atropine for Moderate Amblyopia.</td>
<td>3-7 yrs</td>
<td>168</td>
<td>Randomised comparative trial</td>
<td>4 months</td>
<td>Weekend atropine provides an improvement similar to that provided by daily atropine in moderate amblyopia.</td>
</tr>
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<td>ATS 5</td>
<td>Prospective non-comparative trial to evaluate 2 hours of daily patching for amblyopia (Eye glass only phase study)</td>
<td>3-7 yrs</td>
<td>84</td>
<td>Prospective non-comparative trial</td>
<td>Upto 30 weeks</td>
<td>Amblyopia improved in 77% by optical correction and resolved in 27%.</td>
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<tr>
<td>ATS 5</td>
<td>Randomized trial to evaluate 2 hours daily patching for amblyopia (Randomization phase)</td>
<td>3-7 yrs</td>
<td>180</td>
<td>Randomised trial</td>
<td>5 weeks</td>
<td>Refractive correction alone improves visual acuity in many cases and results in resolution of amblyopia in at least one third of 3 to &lt;7-year-old children with untreated anisometropic amblyopia. Following a period of treatment with spectacles, two hours of daily patching combined with one hour of near visual activities modestly improves moderate to severe amblyopia in children 3 to 7 years old.</td>
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<td>ATS 6</td>
<td>A Randomized Trial of Near versus Distance Activities while Patching for Amblyopia in Children 3 to &lt; 7 years old</td>
<td>3-7yrs</td>
<td>425</td>
<td>Randomised comparative trial</td>
<td>17 weeks</td>
<td>No difference in visual acuity improvement between children performing near activities and distance activities during patching.</td>
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<tr>
<td>ATS 7</td>
<td>Bilateral Refractive Amblyopia Treatment Study: Response to Treatment of Previously Untreated Presumed Bilateral Refractive Amblyopia</td>
<td>3-11yrs</td>
<td>113</td>
<td>Randomised comparative trial</td>
<td>1 year</td>
<td>Bilateral refractive amblyopia improves with spectacle correction. Binocular visual acuity of 20/25 or better was achieved by 73%</td>
</tr>
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<td>ATS 8</td>
<td>To compare weekend atropine augmented by a plano lens with weekend atropine alone for moderate amblyopia.</td>
<td>3-7yrs</td>
<td>180</td>
<td>Randomised comparative trial</td>
<td>18 weeks</td>
<td>Augmentation of weekend atropine with a plano lens does not substantially improve amblyopic eye acuity.</td>
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<td>ATS 9</td>
<td>To compare patching with atropine eyedrops in the treatment of moderate amblyopia</td>
<td>7-12yrs</td>
<td>193</td>
<td>Randomised comparative trial</td>
<td>17 weeks</td>
<td>Atropine and patching achieve similar results among older children with unilateral amblyopia.</td>
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<tr>
<td>ATS10</td>
<td>Randomized trial comparing Bangerter filters vs part time occlusion for the treatment of moderate amblyopia</td>
<td>3-10 yrs</td>
<td>186</td>
<td>Randomised comparative trial</td>
<td>24 weeks</td>
<td>Bangerter filters and patching was less than half a line, and there was lower burden of treatment on the child and family. Bangerter filter treatment is a reasonable option to consider for initial treatment of moderate amblyopia</td>
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<tr>
<td>ATS11</td>
<td>Randomized trial to evaluate combined patching &amp; atropine for residual amblyopia</td>
<td>3-10 yrs</td>
<td>55</td>
<td>Randomised trial</td>
<td>10 weeks</td>
<td>Amblyopic eye VA improved similarly in both groups suggestive of no additional benefit of combined treatment in residual amblyopia</td>
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<td>ATS12</td>
<td>A Randomized Trial Comparing Patching with Active Vision Therapy to Patching with Control Vision Therapy as Treatment for Amblyopia in Children 7 to &lt;13 Years Old; to determine the feasibility of conducting a full-scale randomized clinical trial</td>
<td>7-&lt;13 yrs</td>
<td>19</td>
<td>Randomised Trial</td>
<td>17 weeks</td>
<td>16-week treatment trial of vision therapy was feasible with respect to maintaining protocol adherence; however, recruitment under the proposed eligibility criteria, necessitated by the standardized approach to vision therapy, was not successful</td>
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<td>ATS13</td>
<td>Nonrandomized prospective trial of glasses alone for strabismic &amp; strabismic- anisometropic amblyopia</td>
<td>3-7 yrs</td>
<td>146</td>
<td>Non randomised prospective trial</td>
<td>28 weeks</td>
<td>Treatment effect was greater for strabismic amblyopia than for combined mechanism amblyopia</td>
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<tr>
<td>ATS14</td>
<td>A Pilot Study of Levodopa Dosage as Treatment for Residual Amblyopia</td>
<td>8-18 yrs</td>
<td>33</td>
<td>Randomised Trial</td>
<td>10 weeks</td>
<td>The results suggested that levodopa/carbidopa therapy for residual amblyopia in older children and teenagers is well tolerated and may improve visual acuity. There was a suggestion of partial regression of the improvement in visual acuity after treatment was discontinued.</td>
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<tr>
<td>ATS15</td>
<td>Randomized trial of increasing patching for amblyopia</td>
<td>3-8 yrs</td>
<td>169</td>
<td>Randomised trial</td>
<td>12 weeks</td>
<td>More improvement in VA after 10 weeks compared with continuing 2 hours daily.</td>
</tr>
<tr>
<td>ATS16</td>
<td>Augmenting Atropine Treatment for Amblyopia, the effectiveness of adding a plano lens (a lens without any prescription) to weekend atropine treatment after a patient’s visual acuity has stabilized but amblyopia is still present.</td>
<td>3-8 yrs</td>
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<td>Not published</td>
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<td>ATS17</td>
<td>Compare the efficacy and safety of oral levodopa and patching versus oral placebo and patching at 18 weeks, after 16 weeks of treatment followed by a two-week taper of oral medication.</td>
<td>7-13 yrs</td>
<td>139</td>
<td>Randomised Trial</td>
<td>18 weeks</td>
<td>For children 7 to 12 years of age with residual amblyopia after patching therapy, oral levodopa while continuing to patch 2 hours daily does not produce a clinically or statistically meaningful improvement in VA compared with placebo and patching.</td>
</tr>
<tr>
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<td>ATS 18</td>
<td>Compare the effectiveness of 1 hour/day of binocular game play 7 days per week with 2 hours/day patching 7 days per week</td>
<td>5-&lt;12 years</td>
<td>385</td>
<td>Non-inferiority randomised trial</td>
<td>16 weeks</td>
<td>In children aged 5 to younger than 13 years, amblyopic-eye VA improved with binocular game play and with patching, particularly in younger children (age 5 to &lt;7 years) without prior amblyopia treatment. Although the primary noninferiority analysis was indeterminate, a post hoc analysis suggested that VA improvement with this particular binocular iPad treatment was not as good as with 2 hours of prescribed daily patching.</td>
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<tr>
<td>ATS 20</td>
<td>To compare the efficacy of 1 hour/day of binocular game play 5 days per week plus spectacle correction only for treatment of amblyopia</td>
<td>4-13 years</td>
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Delhi Ophthalmological Society

Joint QOIC-DOS International Conference
4th to 7th January, 2018

www.dosonline.org
Respected seniors and dear friends,

Greetings from the Delhi Ophthalmological Society!

On behalf of the DOS Executive, it gives us great pleasure to invite you and your family to participate in the forthcoming Joint QOIC-DOS (International DOS) Conference of 2018.

The 4th International DOS Conference (I-DOS) is scheduled from January 4th - 7th, 2018 in Dubai. Dubai is one of the most iconic shopping and tourist destinations in the world, less than four hours away from New Delhi. January is the best time to experience the glamour and glitz of the desert city, with the Dubai Shopping Festival coinciding with the I-DOS.

In addition to this, the comprehensive package includes a stay in a five star hotel (Le –Meridian), dinner on Dhow Cruise, delicious live BBQ dinner at the Desert Safari with a live show of fire performers. You will also get to enjoy a Dubai City Tour, with a visit to the Burj Khalifa and other tourist attractions. The details of registration fee is attached for your perusal. The air fare is on first cum first basis as it is peak tourist season in Dubai.

We are confident that you will mark your calendar and hope that you will join us for this exceptional academic feast. We invite you to reserve your places and register soon.

The Tour Itinerary and Registration can be accessed online at www.dosonline.org. Please register before August 16, 2017 to avail as limited seats are on offer.

Those who are interested should contact to Mr. Rajeev Malhotra at rajeev@alpord.net +91-9871799152. In case of any problem, please do not hesitate to contact Mr. Praveen Kumar Nagar Office Secretary (+91-11-23210810, 65705229) or Secretary DOS.

We look forward to welcoming you all at the Dubai for a memorable and high-impact Joint QOIC-DOS (International DOS) Conference.

With Best Wishes,

Dr. (Prof.) Kamlesh 
President - DOS

Dr. (Prof.) Subhash C. Dadeya 
Secretary - DOS

Dr. (Prof.) R.V. Azad 
Chairman Sub-Committee International Affairs

Dr. Arun Baweja 
Treasurer - DOS