Basic Concepts of Primary Exodeviations

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**Introduction**

Strabismus occurs among 1-4% of the children worldwide and strabismic amблиопia is a major cause of unilateral visual impairment among all age groups. Though it has been reported by many that esodeviations are seen 3 times more commonly than exotropia in European and American countries, exodeviations or outward deviation of eyes, are more commonly met with in Asia, Africa and the Middle-East with ratio varying from 2:1 to 3:1. Nepal has a reported incidence of exotropia in 76% of strabismus. In India exotropia is more commonly seen than esotropia. Jenkins has proposed the concept that the more equatorial a country is, the higher the incidence of exotropia. The dazzle produced by bright light is supposed to disrupt fusion, inducing tropia. Primary exotropia is more common among females. It is more symptomatic than esotropia since the intermittent form is the commonest and the phoria to tropia phase produces lot of sensory phenomena like diplopia and blurring of vision.

Small asymptomatic exophorias are not uncommon in our clinical practice, in the normal population. Variable transient forms of exotropias are seen in the newborn which disappear by 4-5 months. Exodeviations are much more common in the intermittent form unlike in esotropia. Primary exodeviations are the most common of exotropias, which constitute a progressive spectrum of deviations from exophoria to constant exotropia, through a period of intermittency i.e. intermittent exotropia. The progression from phoria to tropia is influenced by the decreased tonic convergence with increasing age, the development of suppression, loss of accommodative power and increasing divergence of orbit with aging. Von Noorden has reported that 75% of the intermittent exotropes progressed to constant exotropia. Intermittent exotropia constitute 50-90% of exodeviations.

**Causes of primary exotropia**

Primary exodeviations are caused mainly by a combination of innervative and mechanical factors, added to which there is an interplay of other factors. An innervational imbalance which upsets the reciprocal relationship between active convergence and divergence mechanism has been the basis of classification of Duane, who considered divergence an active process and put forth his theory more than 100 years ago. Later, Weiss and Bielschowsky, showed that anatomical and mechanical factors like growth and depth of orbit, insertion of extraocular muscles etc. also play a role, this being supported by the fact that there is a higher prevalence of exodeviations in craniofacial dysostosis like Crouzon’s disease due to shallow and laterally directed orbits. Current concepts combine these two theories. There are other factors which contribute to progression to constant tropia.

**Defective fusion capability**

Worth has pointed out the role of absence of fusion faculty in causation of any strabismus.

**Refractive errors**

Constant understimulation of convergence in uncorrected myopes, anisomyopes and anisoastigmatic
facilitate progression due to absence of fusional reflexes. Even high hypermetropes who have a low AC/A ratio, give up attempts to clear vision and can lead to development of exotropia.

Bitemporal hemiretinal suppression – that is, the ability to suppress temporal field of vision has been postulated to favour progression.

AC/A ratio - when high produces a true divergence excess exotropia and when low, convergence insufficiency.

Hereditary factors-Knapp, Burian, Spivey have all reported the higher incidence of intermittent exotropia in families. The refractive status also may be inherited.

Proprioceptive reflexes-Mitsui et al have shown that proprioceptive reflexes from extra-ocular muscles have a role. The demonstration of Magician’s forceps phenomenon is in favour of this postulation.\footnote{After anaesthetizing the eye, the fixing or dominant eye is held by the nasal limbus with a forceps and forcibly adducted. It is seen that the divergent eye makes an adduction movement with the disappearance of the strabismus. Care should be taken to see that the other eye does not take up fixation.}

**Classification**

There are different classification systems in primary exotropia, all of them taking the distance near disparity into consideration.

**Duane’s Classification**-

I Basic type where deviation for distance and near are equal

II Divergence Excess type where distance deviation is more than near deviation

III Convergence Insufficiency type where near deviation is greater than distance deviation.

**Burian’s Classification**- takes into consideration, the exuberant fusional convergence reflexes which may mask a basic deviation for near i.e. produces a simulated divergence excess pattern. This is the more generally accepted classification. Thus

I **Divergence Excess pattern** where distance deviation is >10pd larger than at near.

II **Basic** – where Distance deviation equals or is within 10 pd of near deviation.

III **Convergence Insufficiency** pattern where near deviation is >10 pd greater than distance deviation.

IV **Simulated or pseudo-divergence excess**- where distance deviation is more than near deviation, but near deviation equals or is within 10 pd of distance deviation after monocular occlusion for 30 to 45 mts. This is due to the tonic fusional convergence reflexes which gives an apparent smaller deviation for near, but which is dissipated by the monocular occlusion.

**Kushner’s classification** takes into account in addition to the fusional convergence reflexes, the accommodative convergence reflex as well, which is eliminated by the use of +3 D lenses after monocular occlusion before measuring deviation again. This gives an etiological evaluation to guide in the management

**Kushner classification**

<table>
<thead>
<tr>
<th>Distance/near disparity</th>
<th>Deviation</th>
<th>+3D lenses</th>
<th>PBCT</th>
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<tbody>
<tr>
<td>Masking of deviation for near by accommodational, fusional or tonic convergence. Fusional and tonic convergence is overcome by prolonged monocular patching and doing PBCT. After eliminating fusional convergence, accommodational convergence is eliminated by adding +3D and PBCT.</td>
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**High AC/A ratio**- Kushner measured the gradient AC/A and reported that >90% of divergence excess show normal AC/A. The small percentage that did have high AC/A ratio can develop overcorrection for near and may need bifocal lenses postoperatively.

**Convergence insufficiency (CI)** is characterized by a reduced NPC, a low AC/A ratio, a high exophoria or intermittent exotropia for near and a reduced positive fusional vergence. They have asthenopic symptoms for near.
**Characteristics of intermittent exotropia (X(T)) DE**

Most DE and basic exotropes are intermittent. Since X(T) is the most prevalent exodeviation-50-90% and since the management is critical before it becomes constant, it is important that we recognize and treat the condition. The age of onset is usually before 18 months. They have an initial stage of phoria which later become intermittent and then progress until about 6 years of age, when it becomes more noticeable. Initially deviation is seen for distance only and later progresses for near also. Only 6% are first observed after 5 years of age. It is more prevalent in females. They have equal and good visual acuity in both eyes, and good stereopsis when ortho and may have ARC when deviated. They have normal NPC and adequate convergence amplitude.

**Factors contributing to Progression**

As we have already seen the contributory factors in the etiology of exotropia influence the progression to from intermittent to the constant exotropic phase. Decreasing tonic convergence with increasing age is a major factor. As the frequency of tropic phase increases, suppression of the temporal half of retina sets in which favours a speedy progression to tropia due to lack of fusion. Loss of accommodative power and increasing divergence of orbits also contribute. Kushner has shown that 75% progress to constant XT.

**Assessing Control of Progression**

Most clinicians follow the child by assessments of control in the clinic. Calhounz et al has described 4 phases of exodeviations.

- **Phase I** - Exophoria for distance, ortho at near-asymptomatic.
- **Phase II** - Intermittent exotropia for distance, orthophoria or exophoria for near-symptomatic for far only
- **Phase III** - Exotropia at distance, Exophoria/intermittent exotropia for near-Binocular vision for near, suppression scotoma for far.
- **Phase IV** - Exotropia for far and near- no binocularity.

**Symptoms**

A common complaint for which child is brought to the clinic is for frequent closure of eyes or one eye in bright light especially in the sun. This is due to the breakdown of fusion in bright light experiencing diplopia. It may be described as a photophobia or rubbing of the eyes by the parent and may be taken as an allergic symptom. Parents also complain of deviation of eyes when watching TV. Transient blurring or diplopia and vague discomfort may be complained of. Asthenopic complaints are not uncommon in early decompensating phase. Micropsia is a rare complaint of overconvergence by accommodation which they utilize for correcting the deviation. Symptoms become less as they become constant.

**Sensory Adaptations**

Though bifoveal fusion with good stereopsis is seen during orthophoric phase, various sensory phenomena occur during decompensation.

Temporal Hemiretinal suppression-during the tropic phase they may develop temporal hemiretinal suppression. Dual retinal correspondence with ARC during the tropic phase and NRC during orthophoric phase is not uncommon. They may also have panoramic viewing with extension of field during the tropic phase. Monofixation and amblyopia are very rarely seen. Patients who develop deviation after visual maturation, experience diplopia.

Since the clinic control does not take into account the duration of tropic phase and the fact that there is no standardization as to when to intervene, a novel method was put forth by H. Haggerty and Richardson.
The Newcastle Control Score seems to be useful in grading the severity of intermittent exotropia and as a criteria for surgical intervention.

The Newcastle Control Score takes into consideration the subjective and objective criteria to grade severity and quantify progress. The score is the sum total of scores obtained in home control and in the clinic for near and far. Patients with a score of 3 or more are considered to need surgical intervention. It is a consistent method of rating severity and enables one to easily monitor progress of X(T).

### Newcastle Control Score

<table>
<thead>
<tr>
<th>Score</th>
<th>Component</th>
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<tbody>
<tr>
<td>0</td>
<td>Squint / monocular eye closure never noticed</td>
</tr>
<tr>
<td>1</td>
<td>Squint / monocular closure seen occasionally (&lt;50% of time) for distance</td>
</tr>
<tr>
<td>2</td>
<td>Squint / monocular eye closure seen frequently (&gt;50% of time) for distance</td>
</tr>
<tr>
<td>3</td>
<td>Squint / monocular closure seen for distance &amp; near fixation</td>
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**Clinic control near**

<table>
<thead>
<tr>
<th>Score</th>
<th>Component</th>
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</thead>
<tbody>
<tr>
<td>0</td>
<td>Manifest only after CT and resumes fusion without need for blink or refixation</td>
</tr>
<tr>
<td>1</td>
<td>Blink or refixate to control after CT</td>
</tr>
<tr>
<td>2</td>
<td>Manifest spontaneously or with any form of fusion disruption without recovery</td>
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**Clinic control distance**

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<tr>
<td>0</td>
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The score varies from 0-7; score of 3 or more significant

Differential diagnosis-Unlike pseudoET, pseudoXT, is not common and if a mother complains of eye deviating out, it is unlikely that it is pseudoexotropia. But there are a few conditions which can give an apparent XT. Hypertelioism, large angle kappa due to a temporally dragged macula due to ROP or a toxoplasmic scar are some.

**Congenital XT**

Congenital IIIrd nerve palsy often resolves to an extent with minimal or no ptosis and may have to be differentiated. Exotropic Duane's syndrome i.e., Duane type II is another condition to be differentiated. Transient exotropia of the newborn disappears by 4-5 months.

**Evaluation**

History of complaints as in any strabismus, age of onset, any glasses, previous surgery, family history, any neurological problems -are all important. Special attention is given to the duration of control at home-whether dissociated for > 50% of the time. Refraction under full cycloplegia is important. Some children may show pseudomyopia due to overaccommodation to control tropia.

**Evaluation of deviation**

Three factors are important in assessment. Fixation for near and distance, fusional vergence and accommodation. The cover/uncover test, cover test, and the alternate cover tests are invaluable in that they detect presence or absence of bifoveal fusion and characterize vergence movement if present- the information of which is indispensable in diagnosis and management of X(T).The cover tests are done in the primary position, for distance and near.

The Uncover Test-The importance of this test is that in the presence of 6/6 vision in both eyes the presence of a real life fusional vergence is positive proof of bifoveal fixation. The character of refusion movement is observed e.g. whether fusing without a blink etc. This inference does not need further confirmation by sensory tests and can be used for management decisions.

**Alternate cover test**

The fusional vergence in some exophorics may be very strong and difficult to dissipate. Here alternate cover test is done switching between the eyes rapidly to
dissociate any fusion in between. Even if the deviation could not be detected on the first visit, if a mother says his eye deviates outwards on watching TV etc, every effort should be taken to dissociate and do cover test again.

**Measurement of deviation**

Due to variable angle, routine PBCT may not be reliable. Prolonged alternate cover test to suspend tonic fusional convergence is necessary. Measurement should be done for near, distance, ie 6 meters and for distant far i.e. > 50 ft.

Distant far deviation It has been found that measurement of distant far as when looking out of the window gives larger deviation than for routine 6 meters and when surgery is planned the maximum deviation for distant far should be targeted.

**Patching**

If after prolonged alternate cover test there is a disparity of > 10 pd between distance near measurements, then a patch test i.e. measurement of deviation for far and near after monocular occlusion for 30-45 minutes should be done. This is to overcome the influence of fusional convergence reflexes. Controversy exists regarding duration of time of patching. Kushner in a comparative study has found no difference by patching for more than 30-45 minutes.

**+3D add test**

After patch test while still dissociated, +3 D add given to both eyes and deviation measured. If XT at near increases by 20 pd or more it is a case of DE due to high AC/A ratio. 9

**Lateral and vertical incomitance**

Ocular movements and deviations in different gaze positions are evaluated. Size of deviation may differ in lateral gaze. 11 It has been found that 6-25 % show a smaller deviation in lateral gaze. As shown by Repka et al it may be an artifact in measuring deviation in lateral gaze or a true lateral incomitance, in which case the LR recession has to be less on that side or overcorrections can occur. Vertical incomitance with A, V or X pattern may be seen, V pattern being the most common. Oblique overaction may or may not be present.

**Binocularity**

Is tested with Worth 4 dot test-distance test showing central fusion and near testing showing peripheral fusion. Bagolini’s glasses show sensory status under natural viewing conditions and is a better assessment for suppression, suppression scotoma and ARC. BSV can be assessed at the synoptophore and stereoacuity measured.

**Bagolini’s glasses**

Objective methods of assessing distance stereoacuity-by Mentor B-VAT random dot and contour circle tests are supposed to be good in assessing binocularity because near stereopsis does not correlate with the degree of control. 3
Management Decisions in X(T) present a challenge unlike other childhood onset strabismus. Timing of interference is controversial due to frequent fusional capabilities. Decisions to intervene are controlled by the size of the deviation and control of deviation which is the most important.

Nonsurgical methods of Treatment of X(T) are not found to be effective in achieving a cure. But deviations < 20 pd and the very young children are better managed nonsurgically

Maximize visual acuity: Anisometropia, astigmatism, myopia and even hypermetropia can impair fusion. If visual acuity is not affected by a hypermetropia, it is better left alone to stimulate accommodation unless > 4-5 D. Myopia is fully or overcorrected. Treatment of amblyopia may be necessary.

Overcorrecting minus lenses may be used to stimulate accommodative convergence thus favouring fusion. It is particularly useful in patients with high AC/A ratio. Young children tolerate 2-3 D or more. When there is a mixed astigmatism transpose in such a way that maximum minus sphere is given. e.g. +1 D/-2 Dcyl at 180 may be given as -1/+2 Dcyl at 90 if visual acuity is not compromised.

Orthoptic therapy is meant as a supplement to surgery and not a substitute. It makes the patient aware of manifest deviation and teaches him to control it. Diplopia awareness, antisuppression therapy and convergence exercises are the modes of orthoptic therapy used in the convergence insufficiency type of XT. Red lens and TV trainers may be used to eliminate suppression scotoma. Occlusion / penalization of the dominant eye or alternate eyes to prevent or eliminate suppression scotoma can be done with caution before surgery.

Prisms- Base in prisms may be instituted to enforce bifoveal stimulation in deviations <20 pd. and in undercorrections. In larger deviations, prisms are used only for short term preoperative enforcement of fusion. Prisms should be used with caution since in the presence of ARC, the deviation may increase due to prism adaptation.

Surgical Treatment

There exists a controversy regarding timing of surgery in X(T) though the definitive treatment is universally considered surgical. They have a better chance for binocularity and stereopsis. Though Knapp and advocates uphold early surgery in children, they do caution that in visually immature children overcorrection can lead to monofixation and amblyopia. Jampolsky advocates delayed surgery for accuracy in measurement and avoiding consecutive esotropia. Most of the intermittent exotropes progress to constant XT.

Signs of progression are loss of fusional control as evidenced by increase in frequency of tropic phase, development of secondary convergence insufficiency, increase in size of basic deviation, development of suppression symptomatically as evidenced by absence of diplopia during the manifest phase and objectively. The Newcastle Control Score can be followed up and if the score is >3 surgery undertaken.

Type of Surgery Bilateral LR recessions is the standard surgical technique for true divergence excess(DE) XT. In Simulated DE type and basic type either Bilateral recessions or unilateral recession/resection may be undertaken. There are advocates for both procedures. In convergence insufficiency type, binomedial resections are the ideal. In patients with high AC/A ratio, who constitute a small number, posterior fixation – Fadenisation – of MR is done either during lateral rectus recession or during a recess/resect procedure. This fully corrects the distance deviation at the same time minimizing the risk of consecutive esotropia for near. Intentional overcorrection during the immediate postoperative period has been recommended by Cooper as early as 1966. 5-6 pd of overcorrection is considered ideal and immediate postoperative diplopia is considered even therapeutic to prevent suppression. But in young children it can lead to amblyopia. Also in late onset XT, who already experience diplopia, this may produce severe symptomatic diplopia, so better aim for orthophoria in these patients. For overcorrection, bilateral recessions are better as these tend to straighten out during the early postoperative period unlike in recession/resection procedures where it can persist. Adjustable sutures have an upper hand in this regard. In case of lateral incomitance, the LR recession has to be reduced. In small XT, successful unilateral LR recession has been reported.

Constant XT- Only 20% of all exotropes are constant, and are mostly alternating. Many show ARC. Treatment
is surgical. Possibility of diplopia which is usually transient should be explained. In large XT, 3 or more muscle surgery with adjustable sutures ideal.

**A and V patterns** - In the absence of significant oblique overaction, horizontal muscle displacement suffice. If there is Inferior oblique overaction (IOOA) in a V pattern XT, then IO recession is done. In A pattern XT with Superior oblique overaction (SOOA), SO weakening with or without tendon spacers may be necessary. A pattern is rare. An X pattern is not uncommon in longstanding XT due to tethering of lateral rectus and lateral rectus recession suffice.

**Consecutive esotropia** - If the overcorrection lasts beyond 4-6 weeks it becomes a consecutive ET. Upto 10 pd of overcorrection usually straightens out by 6 weeks. Preoperative ambylopia, suppression, resect/resect procedure, all contribute to the development of consecutive ET. In small ET, prisms are given in the immediate postoperative period and limited alternate patching tried. If not responding, surgery will be necessary. Unilateral or bilateral LR advancement or unilateral or bilateral MR recessions are the options.

**Residual XT** - If less than 15 pd, may respond to nonsurgical treatment - fusional exercises, prisms, minus lens etc. >15 pd of deviation will necessitate surgery. Wait upto 12 weeks before reoperating. If < 6 mm bilateral recessions done before, re-recession is better than resection.

**Treatment Outcomes/Goals** - The variability of presentation, type of surgery and lack of standardization as to timing of intervention all contribute to the variable outcome of surgery. The goal of surgery is to restore alignment and to maintain or restore binocular function. Ideal goal is absence of tropia for far and near though success of surgery is defined as alignment within 8 pd of exotropia. Surgery following preoperative occlusion and orthoptic treatment yields the best results.

Kushner and Moton, Morris et al, Scott and associates all have reported an increase in binocularity even in adults as determined by WFD test, or Bagolini's glasses which was in turn, correlated with longterm post-operative alignment. Since undercorrections are common, maximum deviation for distant far should be targeted and longterm follow up undertaken for 4-5 years.

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