# UNDERSTANDING STRABISMUS

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*Foreword*
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CHAPTER

# 4

# Essential Infantile Esotropia

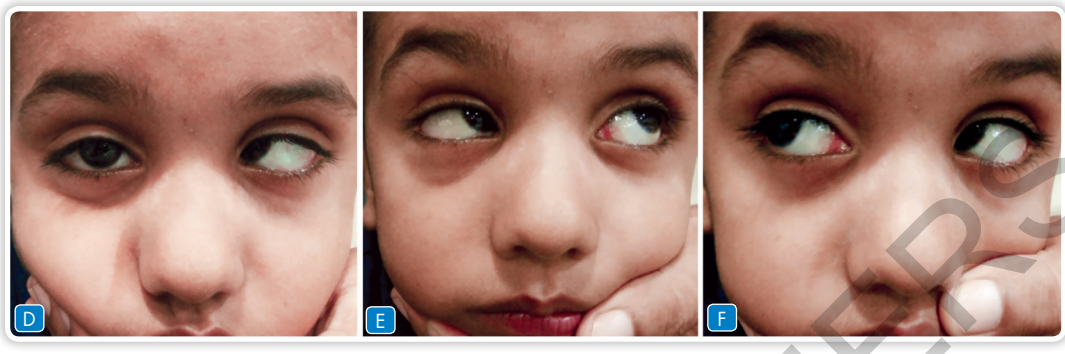


## ESSENTIAL INFANTILE ESOTROPIA (FIGS 4.1A TO C)

The term Esotropia is derived from 2 Greek words: *ésò*, meaning inward, and *trépò*, meaning turn, or crossed eyes, while one eye looks straight ahead, the other eye is turned in towards the nose. This can begin as early as in infancy, childhood, or even in adulthood. Greenberg et al reported an annual age- and gender-adjusted childhood esotropia incidence of 111 per 100,000 patients younger than 19 years. This rate corresponds to a cumulative prevalence of approximately 2% of all children younger than 6 years, with a significant decrease in older ages.

Esotropia with an onset before the age of one year is considered to be a congenital esotropia. In most cases, true essential infantile esotropia is noted at 2 months of age. It is thought to affect about 1% of full-term, healthy newborns and a much higher percentage of newborns with perinatal complications due to prematurity or hypoxic/ischemic encephalopathy. Its classical features are given below and it has to be distinguished from other causes of esotropia that have a different management.


**Figs 4.1A to C:** (A) Large angle left ET with face turn. (B and C) On straightening the head, alternate ET




**Figs 4.1D to F:** Alternate esotropia with bilateral inferior oblique overaction.

## Clinical Features (Figs 4.1D to F)

**Age of onset:** It is between 2 months - one year; the child presenting with a large angle deviation of 30–70 PD and cross fixation (right eye being used for looking to the left and left eye being used for looking towards the right).

## Risk Factors for Developing Infantile Esotropia (ET)

An ophthalmologist should evaluate all children with a history of prematurity, hydrocephalus, seizures, developmental delay, intraventricular hemorrhage and a family history of strabismus.

**Pathophysiology:** Its exact cause remains unknown but following pathogenic mechanisms have been proposed:

i. Excessive tonic convergence.
ii. An inborn and irreversible defect of fusion (Worth). As such, it is a primary dysfunction in the normal development of binocular sensitivity.
iii. A few authors have implicated practically everything from and between the extraocular muscles to the visual cortex in the causation of infantile esotropia.

**Associated with:**

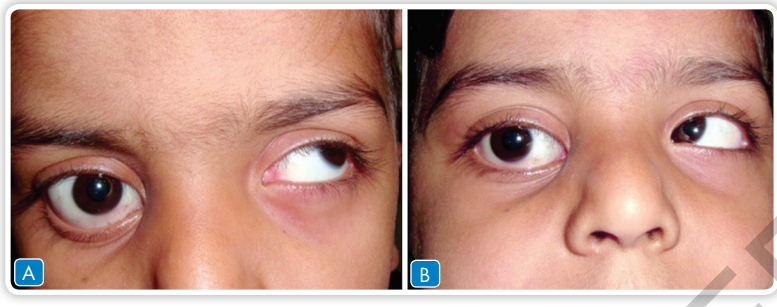
Dissociated vertical deviation (DVD): Less chances of binocular vision, inferior oblique overaction.

CNS or developmental disorders, manifest latent nystagmus.

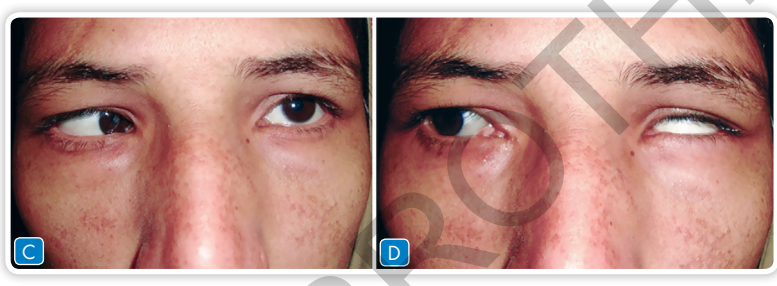
## DISSOCIATED VERTICAL DEVIATION (FIG. 4.2A TO D)

*In this condition, there is upturning of the non-fixing eye in cases of horizontal strabismus when fusion or fixation of both eyes is temporarily suspended.*

**Etiology:** Instability of central control of ocular alignment when there is a weak or absent sensory fusion. Its essential feature is a disregard for the Hering's Law, i.e. the eyes tend to move independently of one another.




**Figs 4.2A and B:** DVD: Fixing with the right eye, the left eye drifts upwards in both abduction and adduction




**Figs 4.2C and D:** Fixing with the left eye, a large angle Right ET. B: Fixing with the right eye, a left DVD/inferior oblique over-action

## Clinical Features

1. In the presence of a horizontal strabismus, the covered eye drifts upwards independent of the fixing eye. This is mostly seen in essential infantile esotropia but it may be seen in any strabismus; DVD is to strabismus as fever is to infection. Its presence indicates that the eye is strabismic with poor sensory fusion. Hence it drifts upwards under the cover.
2. This up-drift is the same in adduction, primary position and in abduction, differentiating it from the inferior oblique over-action, which is also fairly common in essential infantile esotropia; it is readily differentiated from DVD as it is present only in adduction. However, the two may co-exist in the same patient. If one eye goes up higher in adduction than abduction, then inferior oblique over-action is also present along with DVD.
3. Since central suppression is well developed, patients do not complain of diplopia as the eye drifts upwards.
4. The diagnosis is made by observing that each eye goes up under cover, regardless of whether the test is made in straight ahead gaze, right or left.
5. It should also be differentiated from true hypertropia, when one eye is always up under cover and the other is always down under the cover. In addition, DVD is associated with latent occlusion nystagmus, in which when one is covered, the other eye develops a latent jerky nystagmus.

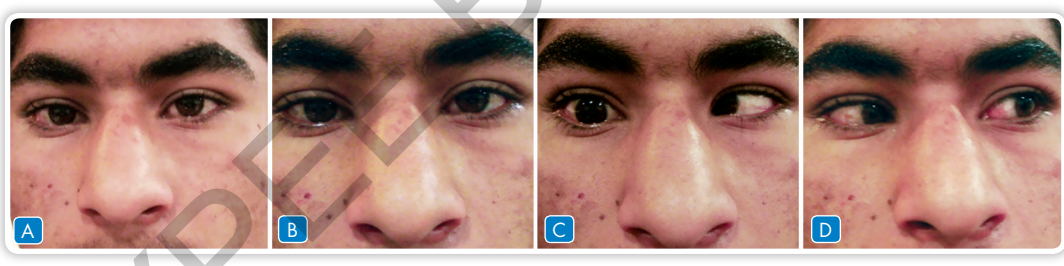
## Management of DVD

Since the cause of a DVD is horizontal misalignment and secondarily absent sensory fusion, the first consideration in surgical correction of DVD is to fully correct the horizontal strabismus. Once the sensory fusion and binocularity develops, DVD lessens on its own with the passage of time; it rarely persists into adult life if managed properly in childhood.

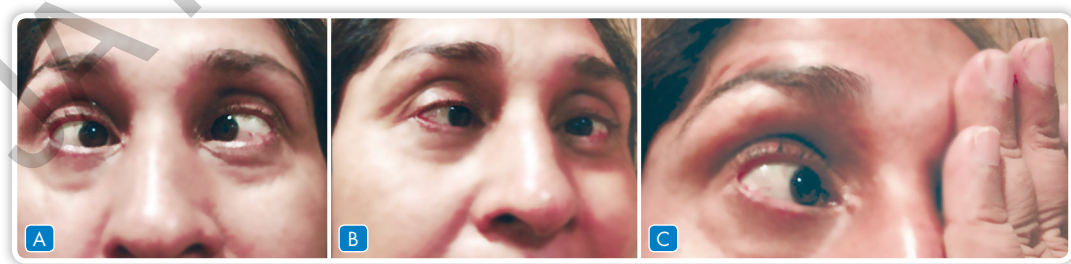
There are many options to correct DVD surgically but easier procedure is a small recession (4–5 mm) of superior rectus combined with a posterior fixation suture, Faden, applied to the sclera posterior to the equator of the eye (12 mm from insertion of superior rectus muscle). The advantage of this technique is that the action of superior rectus is reduced in its field of action and vertical alignment is minimally disturbed in the primary position.

## Differential Diagnosis of Essential Infantile Esotropia

1. *Secondary Esotropia (visual loss):* Corneal opacity, congenital cataract, development anomaly of retina or optic nerve, intraocular tumor.
2. *Early Onset Accommodative Esotropia:* Hypermetropia > 2–3 D.
3. *Bilateral 6th Nerve Palsy (Figs 4.3A to D):* Congenital, traumatic, Mobius syndrome: VI, VII, IX, XII cranial nerve palsies.
4. *Strabismus Fixus (Figs 4.4A to C):* an ankylosing band of fibrous tissue between medial orbital wall and medial rectus muscle prevents abduction.




**Figs 4.3A to D:** Sixth nerve palsy: (A) Face turn to the right; (B) A right ET in straight ahead position; (C) Limited abduction of the right eye; (D) Full abduction of the left eye




**Figs 4.4A to C:** Strabismus fixus: (A) Looking straight ahead, bilateral esotropia; (B) Absent abduction of either eye on turning the head; or (C) on covering the left eye and asking the patient to look to the right (abduct)

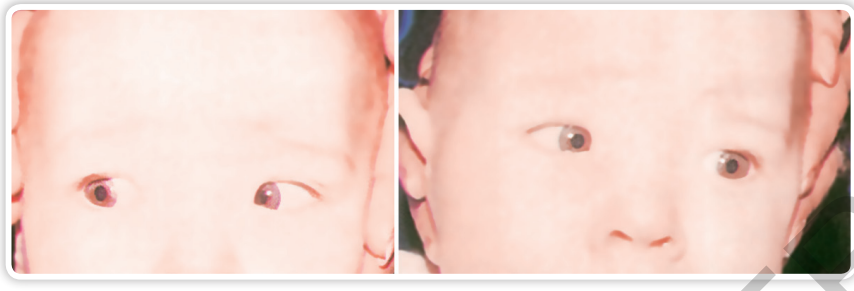


**Fig. 4.5:** Demonstrating abduction by doll's eye maneuver

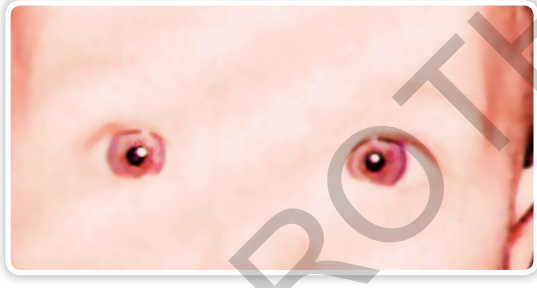


**Fig. 4.6:** Pseudostrabismus: flat nasal bridge, epicanthic folds

5. *Nystagmus Blockade Syndrome:* In abduction, the infant has marked nystagmus, which is blocked in adduction hence the child fixates in adduction.
6. *Bilateral Duane's Syndrome:* Narrowing and retraction of lid fissures in adduction is present.

*In order to exclude all these conditions, it is very important to* ***demonstrate full abduction*** *in either eye.*

1. Occlude fixing eye and attract child's attention laterally.
2. Doll's eye head maneuver (Fig. 4.5).
3. Note position of eyes during sleep and general anesthesia. Eyes are in slight abduction during deep general anesthesia.

## Examination

1. General appearance: It is important to exclude pseudostrabismus (Fig. 4.6)—large epicanthic folds, broad nasal bridge and a small, flat nose. But corneal reflex is central in both eyes.
2. Vision: Must get an idea of visual acuity in either eye as mentioned in Chapter 1.
3. Head posture and position of eyelids in all positions of gaze.
4. Pupil examination: To have an idea of retina and optic nerve function, to know the visual potential in that eye as well as to exclude secondary esotropia.

5. Slit lamp and fundus examination: To exclude secondary esotropia.
6. Hirschberg's test, Cover/uncover and alternate cover test.
7. Ocular movements in all positions of gaze to find any muscle over- or under-action.
8. Measurement of deviation by Prisms.
9. Cycloplegic refraction: Preferably by atropine to neutralize tonic hypermetropia as well.

**Management (Figs 4.7A and B):** Treatment is surgical correction of strabismus by the age of 18 months. At this age, binocular perception and fusion develops.

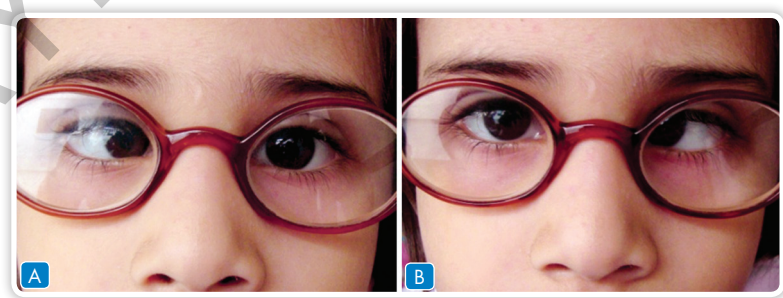
Till that age, child should be prescribed glasses with full astigmatic correction for constant wear in order to avoid amblyopia. Amblyopia is present even though there is cross-fixation and this is due to some degree of hypermetropia as well as astigmatism. Astigmatism of any degree must be fully corrected. Amblyopia must be treated preoperatively as the end point of amblyopia therapy is a freely alternating esotropia with the child fixating with either eye. If surgery is performed for a constant ET without changing it first to an alternating ET by occlusion therapy, it is very difficult to assess postoperatively if amblyopia has been corrected.

In one study, chances of amblyopia in congenital ET are 6% but if surgery is performed without looking for it and treating it preoperatively, chances of amblyopia increase to 50–60%. If the parents refuse amblyopia therapy, then it is important to under-correct ET by 10 PD and warn the parents that eye has a tendency to go out.

**Goal of Surgery**: To reduce ET to within 10 PD to allow binocularity to develop.

- In binocular ET, bimedial recession ± conjunctival recession ± LR resection
- In uniocular cases, recess/resect of the involved eye.
- Always perform FDT at the time of surgery.
- Regular follow-up postoperatively and manage glasses accordingly.

**Botulinum toxin** (BOTOX®) injection into the medial rectus has been explored as an alternative to surgery. Long-term post-injection follow-up up to 7 years showed not only a significant reduction in esotropic angle but also successful binocular alignment (±10 PD) in 89% of the patients.




**Figs 4.7A and B:** Alternate esotropia, not corrected with glasses. However, glasses for the associated hypermetropic astigmatism must be worn to prevent amblyopia

## FURTHER READING


1. Birch E, Stager D, Wright K, Beck R. The natural history of infantile esotropia during the first six months of life. Pediatric Eye Disease Investigator Group. J AAPOS. Dec 1998;2(6):325-8; discussion 329.
2. Birch EE, Fawcett S, Stager DR. Why does early surgical alignment improve stereoacuity outcomes in infantile esotropia? J AAPOS. Feb 2000;4(1):10-4.
3. Birch EE, Stager DR. Monocular acuity and stereopsis in infantile esotropia. Invest Ophthalmol Vis Sci. Nov 1985;26(11):1624-30.
4. Drover JR, Stager DR Sr, Morale SE, Leffler JN, Birch EE. Improvement in motor development following surgery for infantile esotropia. J AAPOS. Apr 2008;12(2):136-40.
5. Friendly DS. Management of infantile esotropia. Int Ophthalmol Clin. 1985;25(4):37-52.
6. Greenberg AE, Mohney BG, Diehl NN, Burke JP. Incidence and types of childhood esotropia: a population-based study. Ophthalmology. Jan 2007;114(1):170-4.
7. Mohindra I, Zwaan J, Held R, et al. Development of acuity and stereopsis in infants with esotropia. Ophthalmology. May 1985;92(5):691-7.
8. Polling JR, Eijkemans MJ, Esser J, Gilles U, Kolling GH, Schulz E, et al. A randomised comparison of bilateral recession versus unilateral recession-resection as surgery for infantile esotropia. Br J Ophthalmol. Jul 2009;93(7):954-7.
9. Rowe FJ, Noonan CP. Botulinum toxin for the treatment of strabismus. Cochrane Database Syst Rev. Apr 15 2009;CD006499.
10. Simonsz HJ, Kolling GH, Unnebrink K. Final report of the early vs. late infantile strabismus surgery study (ELISSS), a controlled, prospective, multicenter study. Strabismus. Dec 2005;13(4):169-99.
11. Tychsen L. Infantile esotropia: Current neurophysiologic concepts. In: Rosenbaum AL, Santiago AP (eds). Clinical Strabismus Management: Principles and Surgical Techniques. 1999:117-138.
12. Uretmen O, Civan BB, Kose S, Yuce B, Egrilmez S. Accommodative esotropia following surgical treatment of infantile esotropia: frequency and risk factors. Acta Ophthalmol. May 2008;86(3):279-83.
13. Weakley DR, Urso RG, Dias CL. Asymmetric inferior oblique overaction and its association with amblyopia in esotropia. Ophthalmology. Apr 1992;99(4):590-3.
14. Wong AM. Timing of surgery for infantile esotropia: sensory and motor outcomes. Can J Ophthalmol. Dec 2008;43(6):643-51.

# UNDERSTANDING STRABISMUS

The management of a patient with strabismus is one of the most challenging tasks for trainee ophthalmic surgeons and general ophthalmologists.

This book is clinically oriented; it decribes the clinical management of all varieties of strabismus in a step-wise and simplified manner which is very easy to follow.

Strabismus surgery is the second most common major ophthalmic surgical procedure after cataract surgery. The basic surgical principles, indications and techniques are fully explained and illustrated with diagrams and figures.

In the last chapter of the book, common clinical cases are discussed regarding their presentation and management. This chapter is actually the whole essence of the book which helps the reader develop a comprehensive knowledge and grasp of the subject.

**Sameera Irfan** FRCS (Edin) graduated from Fatima Jinnah Medical College, Lahore, Pakistan, in 1987. She completed all the pre- and post-fellowships training at Leeds University Hospital, UK. Since 1997, she has been working as a Consultant Oculoplastic Surgeon and Strabismologist. She is an avid researcher and has numerous publications in national and international journals.

She has started a Post-Fellowship Training Program at Mughal Eye Trust Hospital, Lahore, for the last five years where two post-FCPS ophthalmologists receive hands-on training in clinical and surgical skills in the sub-specialties of Oculoplastics, Pediatric Ophthalmology and Ocular Surface Disorders

She has introduced new therapies in Pakistan like corneal tatooing, cyclosporin eyedrops for ocular surface disorders and acetylcysteine eyedrops for filamentary keratitis and chemical burns. Her latest clinical research in amblyopia has revolutionized its management. She has proved that amblyopia can be treated 100% at all ages.

She is also the author of "Understanding Amblyopia", "An Insight into the Basics of Oculoplastics", "Baking Delights" and "Continental Cuisine".

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