ESOTROPIA

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DIAGNOSING ESOTROPIA

Esotropia occurs when there is excess convergence than can't be controlled with fusional divergence. The "fault" can be either in the demand on fusion, or the quality of fusion.The six most common causes of esotropia are (percentages from Mohney, 2001)[1]:

- Accommodative esotropia (53%)
- Esotropia associated with CNS abnormalities (17%)
- Acquired non-accommodative esotropia (10%)
- Esotropia associated with vision loss (7%)
- Infantile esotropia (5%)
- Neurological esotropia (2% sixth cranial nerve palsy, and 1% Duane's retraction syndrome)

[1]It is worth noting that Mohney's population was children with esotropia presenting to tertiary care ophthalmology in an area of predominantly European ethnicity. It is likely more biased to CNS abnormalities than a normal optometric clinical population.

Determining the cause of strabismus is particularly important in esotropia, as it directly effects management. For example, *accommodative esotropia* is usually best managed with glasses and amblyopia therapy, whereas management of *infantile esotropia* will usually include surgery. Determining the cause of an esotropia involves describing direction, frequency, magnitude, laterality, comitancy of direction and comitancy of distance, together with (at minimum):

1) Taking a relevant history that establishes the nature of onset and searches for any relevant associations
    - Unusual symptoms that will help you to differentiate out potentially dangerous ETs (and refer them appropriately): Nausea and/or vomiting, unusual headaches, clumsiness, general malaise, trauma, recent weight changes, hormonal changes, growth defects
2) Measuring the effect of plus lenses (e.g. via Raab's +3D test or similar)
3) Swinging flashlight test to check afferent pupil pathways
4) Specifically searching for neuro-muscular adaptations associated with infantile esotropia (e.g. latent nystagmus)
5) Cycloplegic refraction
6) Observation of the external eye with specific scrutiny for any proptosis
    - Proptosis indicates potentially dangerous ET that you should refer appropriately
7) Ocular health examination
    - Best-practice ocular fundoscopy – preferably through dilated pupils – is essential
    - Checking the clarity of the ocular media
8) Checking for unusual signs
    - Nystagmus, torticollis, ptosis, incomitancy, proptosis, pupil abnormalities

DIFFERENTIALS

1) Accommodative Esotropia

Usually presents with a medium (10-50pd) angle of deviation, with an age of onset between 1 and 4 years of age. There is usually significant hyperopia, and correction of the hyperopia significantly decreases the angle of the esotropia. However, if accommodative esotropia is left untreated, it deteriorates (a process involving changes in sarcomere size and number in the extraocular muscles, which makes the medial rectus shorter and the lateral rectus longer, and results in loss of responsiveness to hyperopic correction). Management will be covered in detail in the next section.

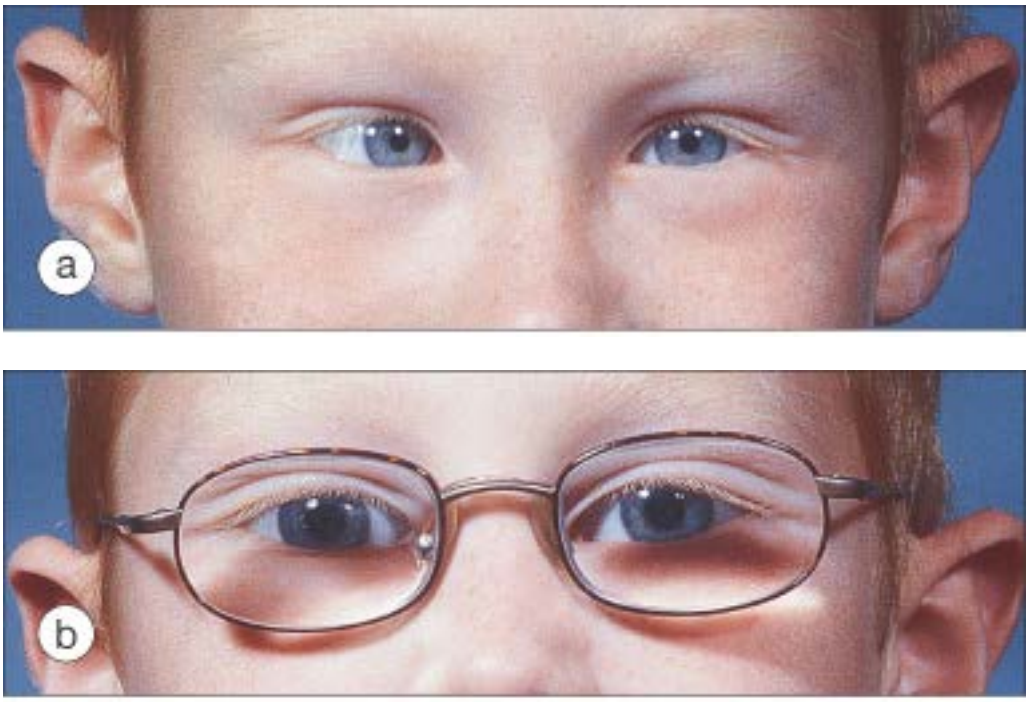

***Figure 13.1**: Accommodative esotropia untreated (above) and managed with plus spectacles (below)*

Accommodative esotropia is commonly, although somewhat artificially, divided into three sub-types:

- Refractive accommodative esotropia has significant hyperopia as a major causative factor, although AC/A ratio is rarely normal
- Non-refractive accommodative esotropia has high AC/A ratio as the major causative factor – any refractive error (including myopia) can be present, but most commonly there is age appropriate hyperopia
- Mixed accommodative esotropia has both significant hyperopia and high AC/A ratio

2) Esotropia associated with CNS abnormalities

These may be congenital abnormalities such as cerebral palsy, developmental delay, autistic behaviour, spasmus nutans, Down Syndrome, infantile spasms, hydrocephalus. Or they may be acquired abnormalities such as cerebral haemorrhage, MVA, meningitis. The acquired abnormalities can be life-threatening emergencies – so refer urgently based on unusual symptoms described above.

3) Acquired Non-Accommodative Esotropia

The median age at diagnosis is 4 years, with a range from 10 months – 18 years (Jacobs et al 2011). The cause of acquired non-accommodative esotropia is debated:

- Are they degenerated accommodative esotropias that haven't been treated properly? (Fawcett and Birch, 2003)
- Or do they have a primary muscle aetiology that will respond to nothing other than surgery? (Gobin, 2001)

Maybe they are some of each. Regardless, it is probably worth trying to correct any hyperopia you find, together with vision therapy to decrease suppression and improve fusion. But don't be surprised if they need surgery, and don't wait too long (Mohney 2001b). Jacobs et al (2011) found that 73% underwent surgery, most achieved gross stereopsis, and 6% achieved stereoacuity of 50" or better.

4) Esotropia Associated with a Vision Loss

Blind eyes can't fuse. Binocular blindness will usually lead to a wandering nystagmus, but monocular blindness will often lead to a strabismus - may be ET or XT depending on innate tendency. Search for the cause, and refer or manage as indicated.

5) Infantile Esotropia Complex

Infantile esotropia presents as a collection of neuro-muscular behaviours, so can be classified as a "complex". These behaviours include the eye misalignment, poor fusion, latent nystagmus (LN), Monocular Naso-Temporal Smooth Pursuit Asymmetry (MNTSPA), Dissociated Vertical Deviation (DVD), and oblique muscle dysfunction. Children suffering cerebral lesions around the time of birth have 30-60 times higher risk of developing infantile esotropia complex (Tyschen, Arch Ophthalmol 2012).

The strabismus component can be any size (e.g. PEDIG (AM J Ophthalmol 2002) found only 49% were larger than 40pd). This misalignment usually becomes apparent between 3 and 9 months of age, and doesn't change with plus lenses.

People who have had infantile esotropia are usually stereoblind for life. However, there is often no amblyopia as 50% of cases alternate, using cross fixation to steady fixation.

Infantile esotropia may spontaneously resolve. This is most likely if the deviation is less than 40pd, and is intermittent or variable. Resolution is also more likely in children younger than 20 weeks of age. Otherwise, they typically require surgery. Tyschen (2005) and Costenbader (1961) observed that 1 in 5 children could develop gross stereopsis if surgically aligned by 1 year of age. Shorter duration of misalignment particularly correlated with better stereopsis (aim for 60 days or less) – so refer without delay.

6) Sixth Cranial Nerve Palsy

This presents as a large esotropia in primary gaze with three distinct aspects:

- Larger at distance, smaller at near
- ET increases when looks to affected side (may present with a head turn to avoid this)
- Loss of ABduction in the affected eye

CN6- can be congenital or acquired. They can be idiopathic or be caused by vascular, traumatic or neoplastic incidences – referral is needed to determine the cause (blood tests and scans are required).

***Figure 13.2**: CN6 palsy*

7) Duane's Esotropia

There are three different types of Duane's Syndrome, but Type 1 is by far the most common. Duane's Type 1 shows a limitation of ABduction of one or both eyes, giving an incomitant esotropia that could be mistaken for a sixth nerve palsy. The distinguishing feature between Duane's Type 1 and sixth nerve palsy is that Duane's Type 1 shows retraction of the globe (and/or ptosis) in ADduction.

Fifteen percent are bilateral. Duane's syndrome is more common in females (4:1). Some have associated congenital abnormalities – consider referral to a paediatrician.

Use the primary gaze state as the basis to decide ophthalmic treatment:

- A patient who is straight in primary gaze probably doesn't need treatment
- A patient with significant misalignment in primary gaze, or a significant head turn to achieve alignment, will probably benefit from treatment with prism or surgery

***Figure 13.3**: Young girl with Duane's Syndrome Type 1 – note that the left eye retracts on right gaze, and the left eye fails to follow the target on left gaze.*

8) Other Esotropia

Rarely, but significantly, esotropia can present as something else. Major vision and life threatening possibilities are orbital lesions such as rhabdomyosarcoma and optic pathway glioma. The details of these diseases are beyond the scope of this course, but note the need to look for proptosis and refer for imaging if found.

MANAGING ACCOMMODATIVE ESOTROPIA

It is convenient to break the management of accommodative esotropia into four stages.

STAGE 1 – PREVENT FURTHER MOTOR ADAPTATIONS

Prescribe glasses that get the eyes as straight as possible ASAP. Regardless of whether the accommodative esotropia is refractive, non-refractive or mixed sub-type, this is done by decreasing accommodation. Consider these prescribing principles (from Berk et al (JAAPOS 2004) and Ludwig et al inc Parks (JAAPOS 2005)):

- Use the full cycloplegic correction if needed, but a lesser amount of plus that achieves *constant* binocular alignment can sometimes be enough
- The correction on the good eye will control the deviation
- The correction on the strabismic eye allows you to treat the amblyopia (stage 2)

Figure 13.4*: Left image: R esotropia, OS fixating (and so controlling accommodation). Right image: eyes straightened by decreasing accommodation in the fixating (left) eye*

STAGE 2 – TREAT MONOCULAR SENSORY ADAPTATIONS

Treat amblyopia to achieve best possible visual acuity in each eye alone – details covered in Amblyopia chapter.

STAGE 3 – TREAT MOTOR ADAPTATIONS AND BINOCULAR SENSORY ADAPTATIONS

Restore binocular alignment as best as possible. This may need a higher hyperopic correction, or a near addition in bifocals (35mm D seg, set high to pupil margin), vision therapy (e.g. improving vergence reserves), and/or surgery.

- Bifocals are most commonly needed when there is a high AC/A ratio, i.e. in the non-refractive or mixed sub-types of accommodative esotropia

Achieve best possible binocularity. Birch et al (JAAPOS 2004) establish that this is needed for long-term stability of alignment. This usually requires vision therapy (more amblyopia patching, more amblyopia penalisation, anti-suppression exercises).

STAGE 4 – PROMOTE EMMETROPIZATION

Several studies have established that there is an appropriate way and time to wean patients off their hyperopic corrections (e.g. Lambert et al 2003, Hutcheson et al 2003, Lambert & Lynn 2006). That is, when you achieve stable alignment, usually sometime in primary/elementary school, start decreasing plus 0.50D (binocularly) at a time.

- This is a type of passive vision therapy –fusional skills are improved a little each time the plus lenses are reduced.
- This is a long-term project – only 8% of children with accommodative esotropia were able to achieve alignment without their glasses 5 years after diagnosis
- Lambert et al (2003) were able to help 60% of children with accommodative esotropia to discontinue glasses 10 years after diagnosis (other authors who have tried to rush the process only achieve 20% - e.g. Mohney et al 2011)
- Children born prematurely or with higher initial hyperopia were significantly less likely to be able to discontinue their glasses
- Children with better accommodation can more successfully reduce their plus corrections (Somer et al in AmJOphthalmol 2006) – remember that MEM retinoscopy is THE best way to establish accommodation skills (Hunter in Surv Ophthalmol 2001)

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