

Case: INFANT WITH A SQUINT

JP is the first child to Betty [32] and John [34], born at term after a pregnancy marked by some first trimester bleeding.

The Maternal & Child Health Nurse pointed out a possible ocular misalignment at age 3 months. She said that JP was likely to grow out of it and did not recommend referral.

Neither parent had ever noticed a problem with alignment or with visual function. JP always responds to mum's presence and he seems to see well.

Betty had treatment for childhood strabismus [glasses, patching and surgery]. Betty's mum 'encourages' her to take JP to an ophthalmologist. An appointment is made for a local ophthalmologist in 8 weeks time [first available] and Betty brings him to you for a referral a week before. She has noticed that in the last few days the misalignment has got MUCH worse and is now VERY obvious.



You notice the right eye turning in quite badly. You check the light reflexes in the pupils and you see that it is quite central in the left eye, and very decentred in the right.

Q1.

What do you think is going on?

What further information do you need?

Q2.

Which squints get better?

Which ones need urgent referral?

Q3:

Is there a simple office test to recognise misalignment of the eyes?

How often do optometrists or ophthalmologists get it wrong?

Q4:

Is there any rush to treat?

The 3 main treatments for childhood strabismus are glasses, patching, and surgery. Why 3? What does each one do?

Q5:

Will he look normal when he goes to school?

Will he read well? Play sport well? Be able to appreciate our new expensive 3D TV?

A1:

This appearance can be due to a number of conditions.

1. Early onset convergent strabismus is usually [so called] Congenital Esotropia. This is less frequently but more accurately called Infantile Esotropia - it is hardly ever truly congenital, but is typically first noticed at ages 3-5 months. 2.

Accommodative esotropia, which is at least partly glasses responsive, happens in children age 2-5, but can also be seen in the first year of life.

3. Sixth nerve palsy in childhood needs quick paediatric or paediatric neurology referral and usually needs quick imaging [within a few days]. A tiny number of these patients have a serious neurosurgical cause for the sixth, a larger number have a neurologically innocent 'viral' sixth [sometimes a synonym for sixth nerve paresis of unknown/ uncertain cause] which often recovers.

An important historical finding is whether it is *always the same eye* that turns in, or whether *either eye* turns in [parents will often call this 'both eyes' turn in].

Either -eye - turning is called 'alternating'. Alternating esotropia usually means that the vision is developing equally in each eye [or nearly so], and that there is no unilateral neurological lesion. This historical fact essentially excludes:

- i. Sixth nerve palsy, and
- ii. The very rare intra-ocular tumour [retinoblastoma, and the need for urgent referral to a paediatric ophthalmologist].

From the parents you want to know JP's history.

Esotropia is more common in children who are premature or have had a difficult perinatal course. It is more common in children with developmental delay [so you want information about milestones] and those with a family history of strabismus.

From JP you want to know if it is easy to switch him from left eye dominant [right eye turning in] to right eye dominant [left eye turning in]. It's difficult examining babies' eyes: it's sometimes easier to ask Betty 'Is there a photo which shows the left eye turning?'



..and mum has it in her phone. You now confidently know this is unlikely to be due to a sixth or to an intra-ocular tumour, and treatment is semi-urgent rather than urgent.

A2:

Intermittent squints may get better, constant ones don't.

Intermittent esotropia in the first 3 months of life is common [? 30% of babies] and doesn't need referral.

Constant esotropia in the first 3 months of life is rare [<1% of babies] and needs urgent referral.

The MCHN's advice at age 3 months 'that JP was likely to grow out of it' was true if she saw intermittent esotropia, or if she wasn't sure if she saw anything wrong.

Intermittent esotropia beyond age 4 months is rare and needs referral.

Divergent strabismus [exotropia] is far less common in Caucasian infants than is esotropia. In Oriental infants and older children, exotropia is more common than esotropia. The above comments about esotropia also apply to exotropia.

A3:

Use a torch and look at the reflections in the central cornea [over the pupil].

Looking for asymmetry of corneal reflections [centred in one pupil, decentred in the other] is more reliable than looking for symmetry or asymmetry of the nasal sclera.



This child PS clearly has asymmetry of the nasal sclera, and at first glance looks to have right esotropia. But compare him with JP: the light reflections on PS' corneas are probably each centred over the pupil, and PS' eyes are probably straight. This is called pseudostrabismus.

Of children brought to an ophthalmologist because someone suspects strabismus, if an ophthalmologist gives the child the 'all clear', the risk of the child turning out to have strabismus is ~10%, 3-4 times the background rate. It is prudent to always arrange for some type of follow-up of all children with pseudostrabismus e.g. by offering to review all suspicious photos.

A4:

There is always an appropriate rush to have the child seen quickly for a first visit because:

- i. It is difficult for the non-specialist to examine a child's squint with confidence [it is also difficult for a specialist!] and physician anxiety is added to parental anxiety.
- ii. Of rare associations with sinister conditions that can look just like regular strabismus.
- iii. The results of treatment are better if a child is realigned within a few months of constant misalignment.

If there is significant **refractive error** [this is much more likely in a 3 year old than an 8 month old infant] glasses must be worn and this alone often improves alignment and vision.

If with the best glasses there is asymmetry of vision [**amblyopia**], patching the better eye to encourage better vision in the worse eye is required. Blurring the better eye with atropine eye drops has an equivalent effect. Treatment is successful in 75% of children.

Surgery is required to align eyes after treatment with glasses and patching has been completed. Success rates of surgery vary with age at surgery, duration of follow-up, whether esotropia or exotropia, and associated features [e.g.

asymmetric refractive error, any developmental issues and ADHD- like disorders are negative prognostic features].

It is often necessary to use all 3 modalities [glasses, patching and surgery] for many children.

The overall aims of treatment:

1. Good vision in each eye
2. Normal alignment and normal appearance
3. The ability for the brain to assimilate the images from each eye into one, 'binocularity', which at its best means high quality 3D vision.

A5:

Expectations of normal appearance in the Prep year are 90+%. Children who don't look normal at age 5-6 often get negative responses from other children of this age because of this; appearance is important.

Reading and playing sport does not require a perfect visual system. A child needs 6/15 vision to function normally in a school environment, and if 6/24 or worse usually needs some assistance e.g. part time aide.

A 'perfect' visual system allows faster 'maximum reading speed', better performance at small ball sports, and the ability for high quality 3D. Treatment should always aim for 'perfect', and accept 'near perfect' as an outcome.

Box of terms

Squint - any ocular misalignment.

Strabismus - any ocular misalignment

Esotropia – convergent squint

Exotropia – divergent squint

Amblyopia – poor vision in one eye which is reversible with glasses &/or patching &/or correctly aligning the eye[s].