

These are currently being re-edited.

# **STRABISMUS LECTURE NOTES**

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## **INTRODUCTION**

In the 1930s, Bielschowsky wrote: “not only the novice but also many well trained oculists consider motor anomalies to be the most difficult field in Ophthalmology because of the great variety of signs and symptoms which frequently cannot be reconciled with one another.....in examining and treating motor anomalies, one never loses an uneasy feeling of incompetence until one has become thoroughly familiar with the physiologic fundamentals from which the signs and symptoms of those anomalies are to be derived”.

Seventy years later this is as true as ever. Before beginning to seriously study strabismus, you must revise your basic optics; you must understand hyperopia, and the basic physiology and anatomy underlying the ocular motor system.

Without this basic knowledge, you will not understand strabismus!

### RECOMMENDED BEGINNER BOOKS IN STRABISMUS

1. Rosenbaum and Santiago. (the best).
  2. Two recent smaller texts are by Pratt - Johnson & Tilson [2<sup>nd</sup> Edition], and by Hoyt & Good.
  3. Section 6 ‘Pediatric Ophthalmology and Strabismus’, American Academy “Basic/Clinical Science Course”. An EXCELLENT introduction and superb reference lists.
  4. SURGICAL ATLASES. The most recent and the best is by Wright.
  5. Tychsen in Adler.
- 3, 4 & 5 are minimum requirements. 1 has chapters on orbital pulleys not found elsewhere.

## **CHAPTER 1 OVERVIEW OF STRABISMUS 1 - MOSTLY SENSORY**

### **SUMMARY**

Amblyopia - detection and treatment. Vision testing in various age groups. How to refract and significance of refraction findings. How to treat amblyopia. Suppression and ARC are the very clever sensory responses to strabismus.

### **NUTSHELL:**

The best possible results in childhood strabismus are achieved by:

- \* early Amblyopia detection
- \* conscientious and disciplined amblyopia treatment
- \* optimal glasses (especially full plus in ET)
- \* having the child as straight as possible as soon as possible after amblyopia treatment is optimised

### **SENSORY ASPECTS OF STRABISMUS**

Some concepts you must know:

When the eyes are misaligned, the visual system in children can adapt by developing one or more of:

A. AMBLYOPIA and/or B. SUPPRESSION & ARC

### A. AMBLYOPIA

**IN THE 0-45 YEAR AGE GROUP AMBLYOPIA IS THE GREATEST CAUSE OF VISUAL MORBIDITY, AND GREATER THAN ALL THE OTHER CAUSES PUT TOGETHER.** In ‘exotic’ diseases such as congenital glaucoma, congenital cataract and ectopia lentis the commonest cause of visual loss are amblyopia and not the primary pathology!

### **FEATURES:**

1. Poor best corrected vision in an eye, which on physical examination looks normal.
2. Partly or completely reversible.

The psychophysics, pathophysiology and structural changes seen in amblyopia are incompletely understood, and include:

- \* Loss of Y cells and decreased dopamine levels in the retina.
- \* Unequal development of the lamellae in the LGN.
- \* Abnormal development of ocular dominance columns in the occipital cortex.
- \* PET and firm scans show reduced activity of the primary visual cortex when visual stimulation occurs through the amblyopic as opposed to the normal eye.

- \* decreased ability to accommodate [hence full plus required]
- \* some have afferent pupil defect with sensitive pupillography
- \*normal retinal nerve fibre layer
- \*possibly abnormal discs

An adult who is forced to use an amblyopic eye will complain bitterly about the visual acuity of, say, a 6/9 eye and may prefer to use a cataractous 6/24 eye rather than an amblyopic 6/9 eye. The vision through an amblyopic eye is not equivalent to just defocus (not equivalent to, say, uncorrected myopia); there is loss of contrast, abnormal contour interaction, and many other changes.

The commonest causes of amblyopia are poor image and poor aim.

1. POOR IMAGE. Anisometropia, aniso - astigmatism, unilateral and bilateral high ametropia [all of these are refractive amblyopia], media opacity [= deprivation amblyopia].

2. POOR AIM - strabismus. In general, strabismic amblyopia is more difficult to treat than amblyopia due to asymmetric refraction.

#### BILATERAL AMBLYOPIA

this happens with bilateral form deprivation eg. Bilateral high ametropias or bilateral dense congenital cataract which is not fixed early.

Using very sensitive psychophysical techniques, one can show that there is a defect present in apparently normal "other" eyes of most [?all] apparently unioocular amblyopes, including:

1. Abnormal contrast sensitivity
2. Delayed acuity development.
3. Crowding is a factor in apparently good "other" eyes (more so than in the eyes of children who do not have any amblyopia or strabismus). It is likely that each eye needs the other to develop optimally and that *poor development of the binocular columns in the striate cortex manifests itself as the bilateral and usually very asymmetric disease that most of us recognise as unilateral amblyopia.*

#### CRITICAL PERIODS

There are probably several "critical periods" relevant to different parts of the visual system. There is a critical period for the development of visual acuity, for the development of binocularity, and for the development of amblyopia, and a different critical period for the treatment of amblyopia.

Some critical periods are:

1. VEP techniques strongly suggest that sensory binocularity can be documented by 5-6 weeks of age. If the eyes are not usually straight by this age, then it is possible that the optimal anatomical substrate for binocularity will not form.
2. OKN shows monocular nasal / temporal asymmetry in all neonates (abnormal when the target is moved nasal to temporal; normal when moved temporal to nasal). This asymmetry begins to mature into the symmetric adult form at 3-5 months. Patients with congenital strabismus do not 'mature' and they keep this monocular OKN asymmetry.
3. In the first few weeks of life unilateral or bilateral media opacity is not amblyogenic - it may be up to six weeks before we have to remove bilateral cataracts (Wright).
4. At the other end of the time scale, there are now many well-documented reports where amblyopia is reversed in adults and the plasticity may never be lost in some patients. L-Dopa may improve amblyopia in human adults.

#### B. SUPPRESSION AND ABNORMAL RETINAL CORRESPONDENCE (ARC)

When there is a motor misalignment, two different sensory things happen at the same time:

1. any one object in the environment has two different retinal loci, and
2. the foveas are looking in different directions.

The first effect produces two retinal images, which we call diplopia. The second effect produces confusion about straight-ahead-ness, which we call confusion.

In order to overcome diplopia, the visual system in childhood suppresses one image 'on demand'. If there is any motor asymmetry (eg unilateral oblique dysfunction) or sensory asymmetry (eg 1 DC anisoastigmatism) then the suppression will tend to become profoundly established in one eye only (= Amblyopia) [the relationship between suppression and amblyopia may or may not be this simple].

In order to overcome confusion, we have to somehow change the original projection of the different parts of the retina (reorient the spatial "map"). The usual point - to - point projections on the retinas can't work - it is these projections that produce the confusion. What may happen is that the fovea of the fixing eye now corresponds to an area of retina in the deviating eye rather than a point. Any point in this area, when stimulated, will correspond to the fovea of the fixing eye and produce the same sense of straight-ahead-ness as the fixing eye's fovea. This "new" point- to - area correspondence is "good" and represents a satisfactory sensory adaptation to a bad motor situation and allows some degree of binocularity. This point- to- area correspondence is not compatible with the highest

quality of sensory status as measured by stereopsis.

The profoundness, stability and ease of reversibility of ARC can vary. Worth 4 dots ARC [apparent 'fusion' in face of an obvious tropic] is more profound and stable than Bagolini lens ARC. Most cases of ARC are not profound and are easily reversible when the eyes are straightened. Straightening the eyes in an adult with a tropia and profound ARC may rarely cause PARADOXICAL DIPLOPIA if the ARC is particularly well established.

So far we have postulated two different sensory mechanisms (suppression - amblyopia to overcome diplopia, and ARC to overcome confusion about straight-ahead-ness) to explain the ways in which the visual system might react to the two primary effects imposed on it by misalignment. It is important to appreciate that these adaptations usually both happen together at the same time, and alongside each other.

If these sensory adaptations become profoundly established and there is some asymmetry in the motor or sensory systems, then they can become established in the monocular state. The commonest situation is that suppression "becomes" amblyopia. ARC can also "become" eccentric fixation (but more commonly just remains as ARC in the presence of amblyopia). Eccentric fixation is possibly an end result of ARC.

*It is possible that ARC, eccentric fixation and suppression-amblyopia are primary cortical defects in some patients, which in turn cause strabismus or poor acuity.*

#### TREATMENT OF STRABISMUS: OVERVIEW

Strabismus has both a sensory and a motor component and both have to be treated. Usually we treat the sensory aspect first. If the motor status is very weird [eg 90 Δ ET] we may have to treat that first to achieve optimal sensory treatment. Firstly we have to assess visual acuity.

**ASSESSING ACUITY IN THE PRE VERBAL CHILD** In the infant the first in-office assessment we do is to look for symmetric or asymmetric objections to cover (the latter implying amblyopia). A cover test can be difficult but should always be attempted eg. To a bright and noisy mechanised toy or video across the room.

If there is an ocular deviation then the quality of fixation in each eye is usually a good guide.

If there is no ocular deviation then one can sometimes be produced with a 15^ BO prism over each eye. This will not work if the child looks AT the prisms and not through the prisms.

Fixation preference is a reasonably good but imperfect guide to amblyopia because we know that many adults demonstrate fixation preference (= ocular dominance) without there being any amblyopia! In a child there is no way of differentiating between ocular dominance and amblyopia!

A scheme widely used to describe and assess infantile visual acuity is "GCM" - telling us whether the fixation of each eye tested separately is good (G), central (C) and whether or not it is maintained (M) when the cover is removed. GCM OU means no amblyopia. Lesser assessments of acuity may be GC, not M through a blink; or GC not M (at all).

**CROSS FIXING.** This has classically been taken to indicate absence of amblyopia. Cross fixation is INEVITABLE if the medial recti are tight (large angle congenital ET) and indicates tight medial recti more than it does equal acuity. *Cross fixing with fixation switch consistently at the midline*, however, nearly always means equal vision. Cross fixing with fixation switch consistently to one side of the midline means amblyopia of one eye. Example: 50Δ ET and child consistently switches fixation 20° to left of midline. The left eye is used for all targets to the right, and as targets are moved from right to left the tight LMR is stretched. As the target passes the midline (heading left), fixation should switch to the right eye. If the right eye is markedly amblyopic, the sensory advantage to maintaining left eye fixation overcomes the motor disadvantage of the LMR tightness. The "20°" is a useful measure of this sensori-motor 'tension'.

4. Overplussing. If a child is wearing full + (eg R+3, L+6) and has (L) amblyopia, how much extra + over the R lens is required to produce L fixation for distance fixation? If it's +6, the L amblyopia is profound. If +1, it is mild (equivalent to 6/12 or so).

5. **OLDER CHILDREN.** In an older child we use a proper visual acuity chart. For amblyopia, whole lines (or single optotypes surrounded by interaction bars) are more important than single letters. Sometimes single letters are all that we get (and are still useful information, possibly equivalent to a potential acuity).

Recommended tests: Snellen or equivalent such as Logmar for children who can recognise letters, and "Lea" or HOVT chart for younger ones.

5. Other "newer" techniques to assess visual acuity in preverbal children "objectively" such as various preferential looking techniques are an important part of the repertoire but are rarely required. "SWEEP VEP" which gives an acuity measurement in 12 seconds might be better and is readily available. VERNIER ACUITY may be more discriminating than grating acuity in preverbal children and is applicable to some VEP- style tests.

6. Amblyopic vision is better in the mesopic conditions often found in ophthalmology offices! *Test your patients with the lights ON.*

If amblyopia is present, it is generally agreed that the risk / hassle / benefit ratio is such that it must be treated if it is more than two Snellen lines, closely observed if it is less than two Snellen lines. Even if there is organic disease present then there may be overlying amblyopia and in this circumstance, amblyopia treatment must be offered and continued unless/ until it is shown to be ineffective.

The first treatment is to give the child the correct refraction, based on cycloplegic refraction.

#### WHICH CYCLOPLEGIC?

You need to document good cycloplegia 30+ minutes after one drop each of local anesthetic and 0.5% Cyclopentolate. In infants wait 15 minutes between eyes to lessen risk of systemic toxicity; in dark eyes children use 1%. Check the retinoscopy result with distance and then with near fixation - they should be the same. If the cycloplegic is inadequate then you should wait a little longer because the onset of cycloplegia can be 40-45 minutes. An alternative is Atropine 0.5% tid for 2-3 days, or Homatropine 2% overnight X2 the night before the next visit. If the refraction is optically significant or if there is anisohyperopia or anisoastigmatism then spectacles should be prescribed.

#### WHAT DOES "OPTICALLY SIGNIFICANT" MEAN?

If an eye is amblyopic then "optically significant" usually means:

1. any hyperopia > 2DS
2. If there is sphere, any cyl
3. If there is no sphere, any cyl > 1.50 DC

*Dynamic retinoscopy is a necessary tool in deciding if an amblyopic eye can accommodate without the help of low plus and sometimes plus is indicated with hyperopia even less than +2. +4DS OU would be significant if someone could show that wearing these lenses lessened the risk of subsequent amblyopia or strabismus. Atkinson believes she has shown this. Ingram believes he has shown that prophylactic spectacle wearing in such hyperopes does not help tendency to strabismus and may prevent emmetropisation.*

Dynamic retinoscopy is very useful in children with moderate plus. If a child at any age accommodates well and equally for distance and near with dynamic retinoscopy then there is no need for assistance with plus IRRESPECTIVE of the known cycloplegic refraction.

Hyperopia tends to increase during childhood in the strabismus population [? more so in amblyopic eyes] but not in the 'normal' population where the natural history of hyperopia is to emmetropise! Anisometropia and astigmatism tend to decrease but sometimes will increase and thus the cycloplegic refraction needs to be regularly reassessed - you must do a refraction at least twice a year!

#### DYNAMIC RETINOSCOPY (DR)

This is an INVALUABLE examination technique, which provides UNIQUE information about what is optically significant for your particular patient.

If with distance fixation, DR is plus (gross) and for a near target DR is low minus and maintained over several seconds then we know that this child "handles" all his / her plus well, accommodating appropriately for distance and near (irrespective of what the actual plus is!).

If with distance fixation DR is plus and for near does NOT go minus we know that there is too much plus for this child to handle, the child is at risk of refractive Amblyopia, and the child needs plus lenses to wear. Thus sometimes it can be shown that a 10 month old child does not need any correction of +6, and another needs correction of +4.

**OCCLUSION** After the correct refraction has been dispensed it must be worn for some time to allow for spontaneous improvement of acuity. This can take months. The next step in the treatment of amblyopia is to occlude the better eye. Full time patching (>50% of waking hours, 7 days a week) is still the best technique. For VA between 6/12 and 6/24 a recent study has shown that 2 hours a day is as good as 6 hours a day.

"Opticlude" and "Ortopad" [exclusively from OPSM] are the only readily available patches here. "Elastopad lite" by Elastoplast, and "Pro Ophtha" by Beiersdorf are less available. When you initially patch, each cycle of treatment should be one week per year of life and you need to review after each treatment cycle to assess progress. So for a six-month-old baby the instruction might be to patch one eye for the three days before the next visit.

You should get significant response after 2-3 cycles and a pretty complete response in 3-6 cycles. If not, something is wrong and you should check the refraction again and look for underlying pathology that you may have missed easily, such as optic nerve hypoplasia. Checking the refraction is easy - one drop of Ophthaine, one of Cyclogyl,

wait 30-40 minutes and with a +1.50 DS lens stuck with Blu Tac to the child's glasses do a retinoscopic over-refraction.

Incomplete responses to amblyopia treatment are seen more frequently in high degrees of anisohyperopia or anisoastigmatism, or if treatment is commenced late (but you should try at least one half-cycle of occlusion in school holidays even up to the age of 10 or 12). In older children or in children whose amblyopic eye has particularly poor vision instruct parents to keep the child @ home indoors for the first few days of patching and to go outdoors only under adult supervision.

Once the very best result has been achieved then the frequency of patching should be halved (say, to every second day or afternoons only every day - let the parents decide which is the least disruptive to the child's and family's life-style) and the child checked again after one cycle to see that this lesser regime is adequate. If so, then it can be halved again etc. Once the best result has been achieved, you can also try "lesser" techniques of occlusion such as spectacle occlusion, part time occlusion, nasal sector occlusion etc. Strabismic amblyopia is more likely to require continuing treatment.

Atropine 'Occlusion'.

This is easier for many (not all) parents and children. It is probably preferable in straight eyed amblyopes because continuous sensory fusion is possible and opaque occlusion may be more strabismogenic [not proven].

It should only be used if effective. Effectivity can be proved or implied:

Proved: If cyclopentolate to the fixing eye produces fixation switch for distance, Atropine occlusion will be effective. If only for near, it may be effective.

Implied: The following table shows the likely acuity in the fixing eye for distance if it is underplussed by the stated amount. If there is a several line difference, Atropine occlusion (with an intentionally underplussed lens for the duration of the occlusion) can be used with confidence.

Average amount of hyperopia required in cyclopleged normal eye to reduce acuity to that of amblyopic eye.

Acuity Hyperopia

6/9 +0.50

6/12 +0.75

6/18 +1.25

6/21 +2

6/30 +2.75

6/36 +3.25

6/60 +4.25

6/120 +6

6/240 +7.5

#### IMPORTANT: PRECIPITATING/ CHANGING A TROPIA WITH OCCLUSION

Warn the parents that occlusion treatment carries this small risk and that you are prepared to and competent to handle the possible adverse sequelae. It is, of course happening because a fragile binocular situation is being interrupted rather severely by monocular occlusion.

In one group of 67 patients with strabismic amblyopia requiring occlusion therapy, more than half the patients had a change in deviation following occlusion therapy, mostly an increase of esotropia, but 14% had a decrease.

## CHAPTER 2

### OVERVIEW OF STRABISMUS 2 - MOTOR

#### SUMMARY

This chapter discusses motor examination. It is important to always have accommodation controlled during the examination so that your findings are consistent from one examination to the next, from one patient to the next and can be compared with those in the literature. Prisms must always be used for measurement.

The principles of planning surgery are discussed - which groups of strabismics will benefit from surgery, which may benefit from surgery, and the overall expectations of surgery are discussed. Each individual muscle is discussed with relation to surgery on it.

The development of motor control is very complex. One of the factors that seems to be very important is smooth pursuit. With each eye tested separately, all infants under the age of 4-5 months show monocular OKN (or smooth

pursuit) asymmetry. When the stripes (or an interesting target) go from temporal to nasal the OKN (or smooth pursuit) is normal, but when it goes from nasal to temporal the OKN (or smooth pursuit) is clearly abnormal. Mnemonic: N-T OKN (or smooth pursuit is NOT ok. By the age of 5 months this neonatal OKN (or smooth pursuit alone) response has begun to mature and symmetrise. This maturation does not occur in children who go on to develop congenital strabismus, and they and some of their first degree relatives maintain this immature OKN (or smooth pursuit alone).

Such children never develop the potential for high quality sensory and motor fusion and have bilateral monocular visual systems (rather than a binocular one).

These children often develop esotropia because:

i) Caucasian children are typically hyperopic as infants (>25% are >+4 in the first week of life).

ii) The N-T pursuit anomaly encourages fixation in adduction.

In the first few months of life, children's eyes are sometimes "all over the place". A group of 43 orthoptists in the UK collected data on their own babies for more than twelve months. One of the children developed a constant esotropia with an accommodative element that developed intermittently from apparently normal alignment in the first few months. In the first two months of life, only five out of the 43 babies were never seen to have a misalignment. The other 38 were noted to have strabismus from time-to-time, the commonest being an esotropia with a possible accommodative component.

#### MEASUREMENT AND TREATMENT OF STRABISMUS

We have to accurately measure the motor deviation. We first need to assess the distance deviation with accommodation controlled and then the near deviation with accommodation controlled.

#### WHAT DOES "ACCOMMODATION CONTROLLED" MEAN?

? Firstly, it means that the child is wearing all the optically significant refraction.

? In the case of esotropia this means all the plus that the cycloplegic [or manifest if available] refraction demonstrates and that repeat cycloplegic or manifest refractions demonstrate - in esotropia this is so important that it cannot be emphasised enough.

When the child with esotropia comes back for the first time with glasses, the examiner must show the child cannot accept any more plus. An older child can be tested without cycloplegia - an extra +0.75DS should blur by 2-3 lines. In a younger child cycloplegia should demonstrate net emmetropia in the overrefraction. It is vital that this be checked again just before surgery is booked. Some very clever and experienced doctors (eg Jampolsky) believe that it is sensible to overplus children by a small amount (no more than +0.5) if that will make the difference between presence and absence of motor control. For the benefit of motor control the child is paying the price of being a -0.5 myope which is a trivial price to pay for the benefit of being straight and staying straight (less chance of strabismus amblyopia, less chance of deterioration of strabismus).

"ACCOMMODATION CONTROLLED" also means that the child really is looking at the details of the distance and near targets which are tested and not just vaguely staring in that direction. An older child is asked to read a Snellen chart whilst the deviation is being assessed ("What's the letter next to the A, the one below the T", etc). A younger child will look at a noisy toy especially one that moves or has bright lights ("can you see the teddy's nose, what colour are his shoes,...").

A near target has got to be something bright and interesting that contains a lot of small detail, and while measuring the deviation be constantly asking questions about the detail in the near target or ask the child to draw on parts of it.

#### HOW DO WE MEASURE THE DEVIATION?

As accurately as possible, with prisms in every case. Cover test is best but sometimes prisms are used to equalise the light reflexes (= Krimsky or "K").

With big deviations you will have to hold one prism over each eye (or stack one on top on another). Because prism are calibrated as inverse tangents and not as degrees they do not add up arithmetically - 30PD over one eye and 40PD over the other eye add up the 80PD. The errors in stacking prisms are greater than this!

EVERYONE who uses prisms to measure deviations must read the article by Guyton in Ophthalmology, March 1983, "OPHTHALMIC PRISMS MEASUREMENT ERRORS AND HOW TO MINIMISE THEM" and photocopy the tables and keep them nearby.

An associated article is in the AJO, October 1983; "ARTEFACTS INTRODUCED BY SPECTACLE LENSES IN THE MEASUREMENT OF STRABISMUS DEVIATIONS". This discusses the peripheral prismatic effects of spectacle lenses in the measurement of a deviation. With plus lenses the true deviation is greater than the measured deviation (opposite for minus lenses). With glasses greater than +/- 4 DS the differences are important.

Light reflexes are a useful estimate of an angle, but should never be relied on to plan surgery. When measured in the frontal plane (which is what we do) ONE MM OF DISPLACEMENT IS EQUAL TO 20-22 PD (and not the 7 degrees that Hirschberg postulated in 1886 and which does apply if measurements are made in the iris plane).

You should measure horizontal deviations in up and down gaze to see if there is an A or V pattern. You need to measure in lateral gaze to see if there is any lateral incomitance. You need to look for any associated vertical deviation and if present you must measure it in the "H" positions of gaze (see later). You need to assess the ocular rotations (versions first and ductions if there is an underaction).

Document over/under actions as mm of displacement of the 6 of 12 o'clock limbus in a vertical deviation (eg RIO OA +2 should mean that in the extreme up/left version the right limbus at 6 o'clock is 2mm higher than the same point on the left eye). Similarly for horizontal deviations measure, say, from the 3 or 9 o'clock limbus to the nearest canthus (or the caruncle in case of the MR) and document it in mm. This means that your 2+ will mean the same as my 2+. Use a ruler, watch for parallax errors, or simply remember that a corneal radius is 6mm, and you can usually be accurate to 1/3 radius displacement.

You must watch a strabismus specialist examine a large number of patients, and then have your examinations critically evaluated. You need to document your findings in a way that makes sense to anyone that reads your notes at another time (including you!). The method of measuring the deviation must be documented - whether by light reflexes alone (Hirschberg H), light reflexes symmetrised by prisms (Krimsky -K), by prisms and cover test (the usual technique and not specified) or by a dissociative technique.

#### WHAT DO WE DO THEN?

Surgery is sometimes considered if:

- ? our data is adequate and consistent and
- ? our child is healthy and does not have unrecognised neurological disease, and
- ? we have checked the refraction recently and
- ? there is a cosmetic defect requiring reconstructive surgery or
- ? there are symptoms (diplopia, monocular eye closure, abnormal head posture, etc)

A child who is referred by an optometrist and who has not seen a doctor should be referred for evaluation if there is any question about neurological or intellectual development.

#### WHEN DO WE SCHEDULE SURGERY?

It depends what we are aiming for:

1. If the child has a congenital esotropia and is in the first two years of life (and the sensory aspects have been optimised) then we know that having the child straight by the age of 2 may lead to a better functional result (=more likely to stay straight) than if straightened later. The aim is some crude binocular vision (eg small or no angle esotropia with W4D 'fusion' and ARC).
2. There is some evidence (Birch) that constant tropia of any sort for 3 or 4 months is sufficient to permanently interfere with the potential of high quality sensory motor fusion. In all cases of strabismus (whether congenital or acquired) surgery should be offered within 3 or 4 months of constant tropia being documented.
3. There is some evidence that any deviation reduced to less than 10pd (or maybe even 20pd) results in better binocularity than previously existed, with the development of stable ARC, some stereopsis, and less likelihood of motor deterioration.

#### IMPROVED BINOCULARITY

It is reasonable to point out to parents just what "binocular functions" and "improved binocularity" means. A child with poor binocularity and straight eyes will grow into an adult with essentially no defect except on artificial tests done by an ophthalmologist. Only if the child wants to be a jet pilot, a crane driver or a microsurgeon will this deficit be important. What improved binocularity means in practice is better motor stability and the main reason why we measure aspects of binocularity is to give prognostic advice. Better motor stability means less risk of subsequent strabismic amblyopia!

There have been some studies that have shown improved eye-hand coordination and less clumsiness after a deviation has been corrected. These changes are frequently reported by parents but these qualities are difficult to study in a controlled manner. Some adults who have deviations corrected will tell you that their visual functioning is better (and indeed you may show regained Bagolini lens fusion in these patients) and they are aware of better peripheral field if they were esotropes.

4. Other cases considered for surgery are for RECONSTRUCTIVE (cosmetic) reasons or have DIPLOPIA and the parents or the patient determine urgency of surgery.

#### HOW MUCH DEVIATION IS UNCOSMETIC?

Some guidelines are:

- ? vertical deviations need to be about 20pd before they are a cosmetic problem
- ? EXOtropias are detectable at about 8 pd
- ? ESOTropias are detectable at about 15 pd

If a patient considers a deviation of less than the above suggestions as uncosmetic, then BEWARE - go and check

your measurements, look for big angle kappa, look for telecanthus, look for very eccentric fixation take a proper history - "is today's appearance typical?" etc.

Angle Kappa is very easy to assess. Cover one eye and ask your patient to fixate your direct ophthalmoscope light while you are looking through the other end. The corneal reflection is usually slightly nasal and then angle kappa is positive.

Very positive angle kappa will make an esotropia found on cover test look better, telecanthus will make such a patient look worse. Positive kappa will make an exotropia look worse, telecanthus will make exotropia look better. ? HYPOTROPIA is more uncosmetic than hypertropia because the secondary "ptosis" emphasises the deviation.

WHAT SORT OF SURGERY?

A RECESSION works by lessening the passive elastic tension in a muscle and lessening the torque that the muscle can generate. In conjunction with a recession, a RESECTION will create a balancing increased tension in the antagonist of the recessed (weakened) muscle, and this is the way a resection works best.

A bilateral LR resection will work in ET and bilateral MR resect in XT so a resection must work by mechanically shortening a muscle and changing its tension and contractility in a permanent way.

For all muscles passive tension and length have a fairly linear relationship over the 'usual' range of recessions and resections; outside this range excessive slackness or stiffness produces less predictable results.

Unless there is marked amblyopia in one eye, bilateral symmetric recession is preferred to recess/resect surgery.

Recessions are easier, faster and safer. They cause less reaction in the immediate post operative period and there is less scarring if one has to reoperate. There are no figures to show that the results of those who do recess-resect in preference to symmetric recessions are better or worse than those who prefer symmetric recession surgery so long as correct amounts of surgery are done each time.

Resections are best performed in association with a recession and there are many such situations where this is clearly the preferred procedure.

Good reasons for doing a recess-resect include:

? in a functionally monocular patient, it is malpractice to expose the only eye to the tiny risk that any strabismus surgery has if an alternative exists

? if one eye has had a recess-resect procedure then the results are more predictable if the next procedure is a recess-resect procedure on the other eye.

Nearly all other horizontal surgery is probably best approached by bilateral symmetric recession surgery, of sufficient magnitude to correct the deviation. We consult a table prepared by someone we wish to emulate who assesses strabismus with the same care that we do and who has a vast experience which looks good when published, and do that amount of surgery.

It is very important to continue amblyopia and refractive surveillance and treatment after surgery even (and especially) when the eyes are straight Amblyopia is never cured in the operating theatre.

HOW MUCH SURGERY?

There is an old Rule of 5-6-7-8. The maximum MR recess should be 5mm, maximum MR resect 6mm, recess LR 7mm, and resect LR 8mm. This is a very safe approach and will fix a deviation of 30-35pd in a 2-muscle procedure. The MEDIAL RECTUS. The two potential problems with over-recessing the MR are convergence insufficiency (CI) and consecutive exotropia.

If you over-recess the MR you will cause a progressive XT in primary position increasing in its direction of gaze.

A maximum figure for recession is probably 7-8mm. Kushner has recently shown that you can safely recess to a point 1.5mm behind the anatomical equator without causing a late consecutive exotropia.

The LATERAL RECTUS is more friendly and you can usually recess it 11-12mm before you produce a persistent deficiency in rotation or ET in its direction of action. Indeed if you want to "maim" the lateral rectus (in say a third nerve palsy) you have to recess it 15mm or more (behind I.O. insertion!) to really impair abduction.

It is difficult to do very large LR recessions on an adjustable or slingback because the fascial connections around LR may not allow it to "take up the slack" beyond 10-11mm. An LR which has already been recessed 8-10mm can be recessed another 5-7mm on a slingback. The LR has been shortened by being chronically recessed, and will take up the slack of a re-recession.

The SUPERIOR RECTUS is friendly in some circumstances and you can do terrible things to it without producing a deficiency in upgaze in DVD where recessions of 10mm or more are necessary to get a result. In cases other than DVD, restrict recess or resect surgery to no more than 4mm or you will produce lid malposition. Beware of operating on the SR and IO (ie both of the elevators) of the same eye - upgaze problems are more common.

The INFERIOR RECTUS is the least friendly muscle and any surgery should normally be limited to no more than

4mm. Attention must be given to lower lid position after IR surgery and repositioning the capsulo- palpebral head of IR when recessing IR.

#### LARGE RESECTIONS

In general, the largest resection you can do is that which will not produce a restriction of duction. In the case of a blind eye or where you can be sure that lateral gaze diplopia will not be an issue, you can resect a rectus sufficiently to produce a restriction to 2/3 of the usual duction.

In an eye with a very LARGE DEVIATION the muscle to be resected will have been very stretched (eg MR in XT, LR in ET) and in such large deviations it may be possible to do resections of >10mm without producing a restriction of duction.

#### SO, TO SUMMARISE:

1. Document and treat the sensory aspects first. Refract carefully and give maximum plus in esotropia
2. Document the motor disorder as accurately as you can. Repeat the observations and only act further if they are consistent.
3. Operate if you can improve the patient.

### CHAPTER 3

#### COMPLICATIONS OF STRABISMUS SURGERY

##### SUMMARY

This chapter discusses perforation, anterior segment ischaemia, the slipped/lost muscle, and conjunctival granuloma. Specific complications related to surgery on IR,LR,SR; fat damage, complications of Botox, unexpected diplopia and scleral dellen.

##### 1. PERFORATION

The incidence of perforation is in the order of one in 1,000.

The incidence of perforation causing visual loss is in the order of 1 in 80,000 in a recent AAPOS study. Compare this with the risk of dying in a car accident in Victoria this year – 400 dead per 4,000,000 population = 1 in 10,000 people will die.

##### HOW TO AVOID IT

1. Use only spatula needles such as Ethicon S29
2. Do sling back sutures whenever possible so that the needles pass through the insertions where the sclera is thickest.
3. In very high myopes and other cases where the sclera is very thin do marginal myotomies for recession and duplication or tucks instead of resection.

##### TREATMENT

1. Give subconjunctival antibiotics.
2. Have a retina specialist assess the patient ASAP – preferably while the patient is still in the operating theatre.

##### 2.ANTERIOR SEGMENT ISCHAEMIA (ASI)

The patient is at risk of anterior segment ischaemia if you have to operate on adjacent muscles. The anterior ciliary vessels are most prominent in the vertical recti, the long posterior ciliaries providing ‘backup’ to the horizontal circulation. ASI has been documented in an otherwise healthy elderly patient after surgery to the two verticals alone (no surgery at any previous or subsequent time to other recti).

Elderly patients, patients with rheological disorders and patients with previous retinal detachment surgery are at higher risk. There have been scattered case reports of ASI happening despite an interval of decades between surgery on adjacent muscles - presumably in these cases collateral development was inadequate.

##### TECHNIQUES TO AVOID ASI

- A) Stage your surgery - wait at least six months before operating on a muscle adjacent to one you have just operated on to allow the best chance for collaterals to develop. Some believe that anterior ciliary collaterals do not develop and that any collaterals that do develop come from conjunctiva or episclera.
- B) Use techniques that place the anterior ciliary arteries at lesser risk eg marginal myotomy instead of recession and duplication or lineal tuck instead of resection.
- C) Avoid limbal peritomy in at-risk cases - Fisher has shown quite clearly that this embarrasses the anterior circulation more so than does a Fornix incision.
- D) Routinely dissect off the anterior ciliary vessels (at least one per muscle) if you are going to recess or resect the vertical rectus in an older person or in any patient who may subsequently require horizontal surgery.

##### TREATMENT

Steroids (topical and oral) may or may not help but are often used. Hyperbaric oxygen is used if available and otherwise safe.

### 3. SLIPPED / LOST MUSCLE

Is it just slipped within its capsule and attached to the globe some long distance back or has it been truly lost and not attached to the globe anywhere?

The SLIPPED muscle is quite common and is usually due to an inadequate suturing technique. When you insert the muscle suture, you MUST take a FULL THICKNESS bite of at least 2mm of the muscle / tendon from behind forward so that you NEVER take a bite of muscle capsule alone.

The slipped muscle presents with a markedly restricted duction but saccadic velocities are OK, force generation tests with forceps seem OK and a CT or MRI scan shows the muscle to be in contact with the globe. In such case careful dissection followed by advancement and/or resection [YOU HAVE TO 'FIND THE MEAT'] will improve the case. A stretched scar looks the same as a muscle slipped within its tendon, and is due to stretching of the muscle-sclera union after the vicryl suture has lost its tensile strength 3+ months after the surgery (a slipped muscle may be apparent much earlier). If this is suspected, then non-absorbable suture must be used for the re-op or the same complication is likely to happen again.

The LOST muscle is more difficult and a transplant procedure is required to fix it.

### 4. COMPLICATIONS SPECIFIC TO MUSCLES

#### A: INFERIOR RECTUS

The commonest problem is lower lid retraction as a side effect of inferior rectus recession. This is COMMON and is avoided by

? recognising the white band approximately 8mm behind IR insertion as being the origin of the capsulopalpebral head (CPH) of IR

? you need to maintain this relation of the CPH to the insertion by careful and generous dissection of the CPH off the outer surface of IR before IR recession

? After the IR recession has been done, accurately replacing the CPH (usually with resuturing) the same distance behind the IR insertion where is originally was.

Another common problem with IR is post-operative augmentation of the recession. Days to weeks after IR recession the effect seems to increase, and a good result is lost. Fells refers to it as post-operative stretching of IR. Others point out that the IO is continually pulling on the IR. I always use non-absorbable suture (eg Novafil or Mersilene) on the IR.

#### B: LATERAL RECTUS

The commonest problem is inadvertently including IO in a LR resection. This will often cause an apparent SR underaction worse on abduction, which may appear to resemble a contralateral fourth. Routinely doing a careful sweep with a muscle hook along the undersurface of LR before disinserting it to break the frequent fascial connections between IO and LR is important.

Another problem with LR is that in LR recession the LR won't go back far enough if you are doing an adjustable or slingback recession. The fascial connections to LR are plentiful (and include a constant band from the LSP!) and will tend to limit an adjustable recession to about 9-10mm. If you need to do a larger one (eg. In a Third Nerve palsy) you need to pin the muscle to the sclera as a fixed recession (BEWARE - you are close to macula here and don't produce a macular hole!)

#### C: SUPERIOR RECTUS

Lid changes are frequent. Careful and generous (behind the vortex veins) dissection of the outer surface (related to LPS) is important to lessen the lid change.

A constant frenulum on the undersurface connecting SR to SO must be actively broken or this will limit a SR recession.

SR recession in the elderly may produce unexpected upgaze palsy.

#### D: INFERIOR OBLIQUE

##### POPPING THE FAT

Never do "the big dip" and ALWAYS PICK UP THE POSTERIOR EDGE OF IO UNDER DIRECT VISION. Use an iris repositor to flatten the curve of the sclera that always interferes with your visualising the posterior edge of IO, and ONLY WHEN YOU CAN SEE THE BACK EDGE SHOULD YOU USE A MUSCLE HOOK TO PICK UP IO. Use the hook as close as possible to the posterior edge.

If you pop the fat, finish the surgery, excise any prolapsing fat the same way you do with a blepharoplasty and try to sew the rent in tenons (not possible in the elderly).

Other IO - associated problems are;

? persistent overcorrection after surgery, esp in the elderly, and esp with anterior transposition

? persistent undercorrection after surgery - may be due leaving a slip of IO intact.

## 5. COMPLICATIONS OF BOTULINUM

A small number of patients get a vertical tropia after injection of a horizontal rectus. Sometimes this does not recover and less than 1% of patients require specific treatment for this.

Failed injection - a common cause of this is a weak signal even when you "know" you're in the right spot. Check the forehead electrode! Translate the whole needle up or down a few mm. **YOU CAN GET A SIGNAL IF YOU ARE IN THE VITREOUS.** Wiggle the needle up and down to check the eyeball doesn't move with it before you inject.

Rarely you can get diplopia after lid injections for facial muscle spasm. The commonest is an ipsilateral hypotropia with incyclotropia due to a presumed local dehiscence in the orbital septum allowing seepage of toxin backward to affect IO. These cases all resolve.

## 6. PARADOXIAL DIPLOPIA

You do a beautiful 5mm BMR for an adult 35pd ET and the patient is straight to cover test but now has crossed diplopia. You may have predicted this by asking the patient if he had any diplopia when the ET was fully corrected with prisms for its measurement.

It is a good idea with ALL adult strabismus to check how much leeway you have with prismatic correction as a guide to the surgical result. So, in the above example how much prismatic over - or under- correction is required to produce diplopia? If you can't produce diplopia or if the margins are wide (20+pd) then you will rarely run into problems post operatively.

If prismatic correction produces diplopia or you find that very small over - or under - corrections will, then you must use a pre-operative prism adaptation test to see how troublesome this diplopia will be, and then you must use adjustables if/when you operate.

## 7. DELLEN

Corneal and scleral dellen can be seen. Corneal dellen are best avoided by not doing a limbal peritomy.

# CHAPTER 4

## CONGENITAL (INFANTILE) ESOTROPIA (CET)

### SUMMARY

Congenital strabismus usually manifests itself as CET, sometimes as infantile exotropia, and sometimes as 'pure' DVD. Asymmetry of monocular smooth pursuit is pathognomonic of congenital strabismus of all types, and is close to the 'core defect'.

In CET, despite cross-fixation, amblyopia is present in about a third of patients. 70+% patients get excellent results from one surgery. Surgery should be performed so that the child is straightened by however many surgeries are necessary before the age of two. There is uncertainty as to whether straightening the child before twelve months or before six months gives even better results.

Apparent CET can be due to early onset accommodative esotropia and this must be excluded.

### FEATURES OF CET

The features of CET are:

1. Onset before 6 months
2. Stable large angle esotropia (>30pd)
3. may be associated with:
  - defective abduction -excessive adduction
  - oblique dysfunction DVD
- LMLN
4. Initial alternation
5. Limited potential for normal binocular vision (BV)
6. Normal CNS

The cause (s) of CET are unknown. A small minority of CET patients have a family history of strabismus (usually not CET). CT changes, associated neurological and developmental problems and BSER changes reported by some have not been substantiated by others and may reflect variations in local referral patterns. Strathdee (Sydney) has found that prematurity and possible post maturity are factors associated with congenital esotropia. Neurological problems in the perinatal period make CET much more likely eg seen in 100% of those with IVH and hydrocephalus.

### COMMON ASSOCIATIONS

1. AMBLYOPIA >35% ... Even when cross fixation is present! Scoot has pointed out that in cross fixation, attention to the point at which alteration consistently occurs is an accurate determinant of the equality of vision. If amblyopia is present cross fixation does not take place at the midline; the sound eye will abduct to follow a target

lateral to the midline before the amblyopic eye picks up fixation.

2. DVD 50% Onset seems to be greater in the second year of life. Early surgery for CET has been reported to lead to increased frequency of DVD (Moorfields) and to less frequent DVD (Toronto). Large ET with increased MR tone may mask DVD in untreated CET and may thus factitiously suggest a delayed onset of DVD. DVD is a manifestation of blockage of congenital torsional nystagmus. Eg, left fixation with congenital torsional nystagmus causes tensing of the obliques which causes a Hering - like effect on the other eye of (usually) elevation and extorsion.

3. IO OA 70+ Average age of appearance of IO OA is 3.

4. DVD & IO OA together.

5. LMLN 25% This is a special type of congenital nystagmus where the fast phase beats to the fixing eye (eg. L fixing, fast phase to L). There is overlapping between CET-with-LMLN and nystagmus blocking syndrome (NBS).

6. ABNORMAL SMOOTH PURSUIT (MOST) When each eye is tested separately, pursuit is normal when a target is moved T to N and abnormal when moved N to T. This asymmetry is normal in the first few months of life and then gradually disappears in 95% of infants. Conversely the presence of asymmetric monocular pursuit in someone with ET presenting at an older age fairly reliably places it as CET. This asymmetry also has a sensory component.

#### CONTINENTAL EUROPEAN TECHNIQUES

Management in some parts of Europe can be quite different. Early treatment may be with alternate or binasal occlusion. Jampolsky (US) also promotes the use of alternate occlusion in order to prevent the development of abnormal binocular interactions.

Sometimes the child is kept fully atropinized in order to tolerate low plus glasses.

Surgery is delayed until about 3 or 4 years and may include anteriorization of the obliques whose dysfunction is considered to be critical to the loss of fusion that is felt to be necessary for the development of CET.

None of these techniques have been subjected to a formal trial. Currently the European Strabismus Association is conducting a multicentre trial to see whether surgery before the age of 2 gives better results than later surgery. A retrospective US study by Ing has shown that it probably does.

#### EARLY ONSET ACCOMMODATIVE ET (AET) - NOT RARE

All children with apparent CET need a cycloplegic refraction. Those with early onset AET are most likely to have a variable angle. I ALWAYS try some anti-accommodative treatment (glasses or Phospholine) if the child has more than +3DS, and persist with anti-accommodative treatment if it has any effect. Trying Phospholine drops for a week is a very simple therapeutic trial.

Failure to recognise early onset AET will result in a high proportion of surgical over - and undercorrections.

Recognising the possibility of early onset AET but ignoring correct treatment because the child is "too young" and 'it's too difficult' is very wrong.

#### DIFFERENTIAL DIAGNOSIS

1. Pseudo Esotropia

Check the parents' appearance and if the parents do not have telecanthus you can reassure them that the child will indeed 'grow out' of his/her apparent esotropia. Always arrange some type of follow-up eg. 'Send me a flash photo if you think he/she might be worse' because some of these children will have true intermittent esotropia.

2. Duane's

The pathognomonic retraction can be difficult to pick in an infant.

3. NBS

4. Congenital 6th

5. ET with CNS anomalies (Down's albinism, CP MR, etc)

6. Sensory ET eg retinoblastoma

#### SURGERY

You have been able to examine the child to your satisfaction on at least 2 different occasions and have found the angle to be stable (within 5pd) when measured with prism (either Krinsky or cover test). The plus is less than 3 (and/or anti-accommodative therapy has been to have no effect on the measured angle). There is no more amblyopia. The child is physically and developmentally OK.

NEVER OPERATE UNLESS ALL THE DATA IS "IN". IT IS FAR BETTER TO DELAY SURGERY UNTIL YOU ARE SURE OF WHAT YOU ARE DOING THAN TO OPERATE ON INCOMPLETE OR TENTATIVE DATA, for the results must surely be inadequate and less reliable than the usual imperfect results. An unstable or varying CET should not be operated on because neither you nor I nor anyone else knows how much surgery to do for such a case. The commonest causes for such instability are un- or under- corrected plus and neurological / developmental problems. Sometimes a variable ET indicates underlying fusional divergence and a small varying CET may even resolve spontaneously!

WHEN ALL THE DATA IS "IN" you go to a table prepared by someone whose published results you wish to emulate and do that amount of surgery. The table published by Calhoun is for BMR

25pd - 4mm 35pd - 5mm

50pd - 6mm 75pd - 7mm

.....and interpolate where appropriate.

The commonest deviation is around 35-45pd. and 5-5.5mm BMR are the most usually needed treatments. If a second procedure is required, LR resect OU is the most reliable.

#### WHEN TO OPERATE

Some of the data on very early surgery is very encouraging, eg. Wright 16pts straightened by the age of 5 months. All have 2y+ follow-up. With surgery, 12/16 were within 10pd of ortho. 6 of these 12 were initially XT post-op. 5 of these 12 have demonstrated stereo of 80 to 3000". These are impressive results and if repeated by others may increase the urgency with which we seek to straighten these patients.

Birch has data that suggests that surgery should be done within 3-4 months of constant esotropia for best results.

I usually operate when I am sure of the data - if the parents wait a while before they see a doctor who then procrastinate for a month or two and the child then has to wait a month to see me and I then need to see the child two or three times to be sure of the consistency of the angle and the absence of amblyopia.... It's uncommon to be 'ready' for surgery before age of 9 months.

My 2-3 anecdotal cases of random dot stereo in CET were straightened by me at 5 months of age. Many more of my cases are orthotropic with some fusional amplitude.

#### STABILITY OF RESULTS

In the days following CET surgery, XT is sometimes seen. CET results are best assessed at 4-6w after surgery. Of patients who have successful straightening surgery that is stable for at least six months, about 50% end up needing a second procedure for either consecutive exotropia (10-20%), oblique dysfunction, DVD or recurrent esotropia (if glasses are/become inadequate).

#### BOTOX

Scott and others have been treating CET with Botox. Scott routinely treats all CET patients with 1.25u Botox to both medial recti, usually in the office with local anaesthesia and restraint, and he claims that about 20% of CETs are CURED with one injection. The other can be offered repeat injections or surgery.

Those aged 1-3 have light Ketamine anaesthesia. If botulinum works as well as surgery the need for multiple minor procedures (injection with/without brief anaesthesia) will have to be balanced with the need for fewer but more substantial (surgical) interventions.

The amblyogenic potential of the (usually) non-obstructive ptosis seen in 10-15% of MR injections seems to be low.

#### POSTOPERATIVE MANAGEMENT

##### 1. OVER - CORRECTION

The natural history of overcorrection is not clear. If small it may be stable, get better or get worse. Any unc cosmetic consecutive XT should have a second surgery within months. MR advancement should be done if over-recession has produced lateral incomitance, otherwise do LR recess OU.

##### 2. UNDER-CORRECTION

If there is any hyperopia then anti-accommodative treatment must be urgently (re-) instituted even if it was shown to be inadequate or ineffective preoperatively. Any residual non-accommodative ET greater than 20pd. should be considered for repeat surgery. If the first operation was BMR, LR resect OU is more predictable than re-recession MR OU. If recess-resect has been done as a first procedure then the logical second procedure is recess-resect on the other eye.

##### 3. RECURRENT ET

30+% of patients with CET who are surgically straightened later develop a recurrent ET. MOST will be accommodative and glasses must be instituted with some urgency.

In other one series, if the ET recurred within 3 months of surgery the average plus was +4, and if later (average 27 months) the average was +2. In the early group nearly all the patients had warranted a trial of anti-accommodative therapy preoperatively which was considered to be ineffective.

##### 4. AMBLYOPIA

Parents frequently believe that amblyopia will be fixed surgically and must be specifically told that it will not. Amblyopia after the eyes are surgically straightened may be more difficult to detect and parental compliance with treatment more difficult to achieve.

#### EXPECTATIONS AFTER SUCCESSFUL SURGERY

After successful CET surgery, further surgery WILL be required in 50% of these successful patients (Wills).

Further surgery is performed in 30% for uncosmetic 100A, in 25% for recurrent ET not controlled with glasses, in 15% for DVD, and in 10% for XT.

There is considerable selection bias in any one series, which is not appreciated by the reader or even the author!

#### VERY IMPORTANT REFERENCES

Tychsen in Adler

## CHAPTER 5

### ACQUIRED ESOTROPIAS

#### SUMMARY

Accommodative esotropia (AET) is a manifestation of high hyperopia, and/or high AC/A ratio, and is (initially) fully correctable with glasses. After some time a non-accommodative component develops, due to shortening of the medial rectus which cannot be corrected with glasses (the strabismus 'anatomises').

Sometimes 'normal' levels of hyperopia seem to precipitate ET. In these cases there must be some other reason why the motor fusion is poor – cyclovertical disturbance, pulley displacements, smooth pursuit asymmetry, ...

This chapter discusses when to operate on deteriorated AET, the principles of prism adaptation testing, how much surgery to do, operating for near esotropia only, and what the expectations of surgery are.

Acquired esotropias are usually accommodative to some degree. There are three groups of accommodative esotropia patients to consider:

#### HIGH HYPEROPIA AND NORMAL AC/A RATIO (about 1/2)

This is a normal physiological response to an abnormal refractive situation. Often called REFRACTIVE esotropia. Glasses correct the whole deviation.

Usual plus: >4

#### LOW / NORMAL HYPEROPIA AND HIGH AC/A RATIO (1/2)

This is an abnormal physiological response to a normal clinical situation.

Treatment: Glasses. Usual plus: 2.5 (range 0-9)

#### DETERIORATED AET

These are patients who have developed or will develop a non-accommodative component to their ET. One cause of deterioration is wearing inadequate plus or a delay in giving plus.

Treatment: Glasses with full plus first then surgery if the residual ET is noticed.

Hyperopia and/or abnormal AC/A ratio will result in an ET if the fusional divergence is or becomes inadequate. Typically in childhood the ET is intermittent at first. The frequency and duration (and eventually the angle) of ET increase over weeks or months. Secondary structural changes may ensue to cause a shorter MR OU and a non-accommodative (=deteriorated) component to the ET.

Another group of patients is deteriorated congenital ET patients who easily develop AET with small amounts of hyperopia. In the literature these are sometimes included with others AETs, but are probably best considered as a separate group.

Another small group is acquired ET patients who do not have an accommodative element to their ET. Some of these are deteriorated microtropia. Some are late presentations of 'congenital' esotropia.

The commonest age of onset of AET is 2-3, but the range is from a few months to 8-10 yrs, and even into adulthood.

#### AC/A RATIO

there are easy and less easy ways of assessing this.

#### THE EASY ( and inaccurate) WAY:

If, with accommodation controlled, the near ET (=ET') is 10 or more PD than the ET, then the AC/A ratio is considered high. If the difference is 20 or more Pd it is truly abnormal. AC/A ratio cannot be easily separated from similar conditions such as convergence excess or abnormal CA/C ratio. This lack of differentiation may not be important as the treatment is probably the same.

#### LESS EASY (and more accurate) WAYS:

There are two techniques, gradient and heterophoria. Some authors still measure the AC/A ratio formally in these ways, but the figures don't always mean much because they depend on the techniques used. There is also some variability in AC/A ratios in some/many patients.

#### HOW IMPORTANT IS HIGH AC/A RATIO?

If the AC/A ratio is high then an AET is more likely to deteriorate. Parks first stated this 30 years ago. The deterioration in high AC/A patients was about 50%; in normal AC/A patients about 25%.

Scott and Raab have recently challenged this.

Scott showed that the most significant factors in deterioration were:

? delay between recognition of ET and prescription of optical correction

? early onset (first year of life)

? moderate hyperopia (3-4 DS as c/f. Under 2.5 or over 4.25) Raab has shown only a slight trend to AC/A being significant in his patients. My smaller personal series is consistent with Park's findings.

#### THE NATURAL HISTORY OF HIGH AC/A RATIO...

.....is to improve in >1/2 of cases between 7-12 years of age. This is very important information and any series on treatment of AC/A ration must therefore have a control group. (This is usually not the case!) If your patient has an unc cosmetic near ET but is straight for distance cc (and stays straight for distance) then it is likely that the near deviation will improve between the ages of 7-12 years, or will require surgery beyond that age if it has not improved.

#### TREATMENT

The mainstay of treatment is to wear full plus, especially in younger children (say under 5), and to keep wearing full plus.

It is important to give your young patients the best chance for the best result. You can control an ET better with glasses (if they work) than with Sx. You can control the sensory development with glasses and with patching. You need to do all of these up until age 7-8 to give your younger patients the best possible result.

In older patients less-than-full plus may be adequate to control the ET and ET', but this must always be preceded by a period of wearing full plus.

Only about 1/4 of your patients will end up "growing out of their glasses" and the presence of high AC/A ratio and the amount of initial hyperopia seems not to influence the "improvement" rate.

You should determine the full plus with a cycloplegic refraction done at the first visit, and prescribe glasses based on this. You must check the plus in the spectacles is maximal by doing an overrefraction with cyclo and this must be checked every few months. Hyperopia usually increases during childhood up to the age of about 7 in the strabismus population and then sometimes declines.

In kids of 4 or more a "quick" check is that an extra +.75DS will blur the Snellen acuity by 2-3 lines. If it does not then the child is underplussed. It is just as important not overplus, and if -.50 overrefraction improves the vision by a line then you must reduce the plus. This is especially important in high plus where BVD means that what the child is wearing may not be what you want. Manifest or cycloplegic overrefraction is necessary in these group.

Amblyopia is common in all ETs (about 1/2) and must be treated according to accepted guidelines.

#### THERE IS SOME URGENCY IN ALL THIS TREATMENT

Scott has shown that the longer an acquired ET is left un- or under- corrected by glasses, the more likely it is that structural changes and a secondary non-accommodative will occur. Parents will tell you that this deterioration takes place over a few weeks on some kids. If an acquired ET is quickly and fully corrected with glasses, then there is a much better chance of long term stability = good binocularity. If a recently deteriorated accommodative ET is surgically straightened within weeks - Ditto.

I don't routinely use bifocals because Parks shows 30+ years ago that the natural history of high AC/A ratio was to spontaneously improve in >50% of patients and this figure is about the same as any bifocal series. The mooted "advantages" of maintaining bifoveality more of the time with bifocals make sense to me but have never been proven, though they have been used in high AC/A ratio since late 19th century!

Those who do use bifocals use them in only one circumstance - when the full plus gives orthotropia at distance and there is an ET which when fully corrected by the bifocal add shows sensory fusion eg/ some stereo.

I DO use bifocals if a child gets near diplopia from high AC/A ratio (rare), and hardly ever otherwise.

#### MIOTICS?

If you want to try miotics, use Phospholine 0.06% every night OU for a week. If you get a full response (ie the eyes straighten for distance and for near) use it less frequently. If you do not get a full response abandon it. If you get a full response continue with the drops but also try glasses again from time to time- one day you will succeed in switching from drops to glasses.

There is a place for Phospholine in treating small undercorrections after ET surgery.

In a very young child who needs significant plus but will not wear glasses, there is a place for miotics for a month or two.

There may be a place for miotics for a while in persisting near esotropia in older children. If the P.I. straightens the child, it may allow fusional divergence to develop as the P.I. is wearing off, ie the P.I. is used as a vergence "exercise".

#### THE UNCORRECTED STRAIGHT EYED HYPEROPE

What do we do for 6 month old baby who is straight and +4 DS OU? A 2 year old who is +4 DS OU?

Will giving some or the full plus lessen the risk of developing esotropia. The evidence and literature is a little confusing and suggests:

1. Giving full plus early may lessen the risk of subsequently developing accommodating esotropia.
2. Giving full plus early may lessen the natural tendency to emmetropise.

Thus, giving full plus may itself have some morbidity.

If you find that a child has significant uncorrected hyperopia (+4 or more) then my recommendations are as follows

1. If there is intermittent esotropia that you can document in the office then you should give full plus
2. If esotropia is infrequent and transient by history and you can rarely, if ever, detect it than I would not give plus
3. If the child never has esotropia but there is a family history of esotropia then there is a moderate risk of development of esotropia the child should be reassessed by you every few months.
4. If there is no family history of esotropia and you can detect some motor fusion or sensory fusion then you should review the child if the parents notice any deterioration.
5. Check with dynamic retinoscopy that the child accommodates adequately for near.

#### WHEN TO DISCUSS SURGERY

For undercorrected ET for distance when wearing full plus, which is stable over at least two visits a few weeks apart, and which is unc cosmetic, surgery must be considered fairly quickly, according to the usual guidelines and tables.

#### WHEN SHOULD YOU OPERATE?

When you can be quite confident that you can improve on the natural history in a significant way ie.

Non controversial : moderate-large ET, say 20+ PD, which is unc cosmetic

Non controversial : leave small angle ET alone, say <12 PD

You should also leave alone cosmetically fine ET no matter what the cover test angle is. So, if a 40pd ET happens to have significant angle kappa and look fine, never operate. Treat amblyopia, keep in full plus to lessen deterioration of the angle, but never operate.

#### WHEN SHOULD YOU NOT OPERATE?

1. If your data is insecure
2. If you find variable ET you might wish to try a prism adaptation test and operate on the angle you induce. See the big paper in Archives September '90.
3. In small angle ET (a/a)
4. ET with positive angle kappa such that the child is cosmetically OK (a/a) - this is a difficult situation to improve on.

#### WHEN SHOULD YOU OPERATE TENTATIVELY?

1. In brain damaged children. Aim to leave small angle eso.
2. In children who have shown unusual responses to strabismus surgery in the past.
3. Variable measurements - Think myasthenia. Think undercorrected hyperopia.

#### WHAT DO YOU AIM FOR IN SURGERY?

In ET without amblyopia, the result you want is orthotropia with glasses. In ET with monocular poor vision the result you want is a cosmetically acceptable small ET to lessen the risk of consecutive XT.

#### WHAT RESULTS SHOULD YOU EXPECT?

You should expect 70%+% straight after one operation. After the surgery, monitoring refraction and amblyopia is at least as important as before surgery but often more difficult to accomplish because parents are sometimes less diligent in co-operating with you on these matters - as far as they are concerned their child is "fixed" because the child is now straight and any amblyopia or refractive error was fixed in the operating room! - make sure they understand this is not the case.

#### PRISM ADAPTATION STUDY

See: Archives 9/90 for the results of this excellent study. The library has a copy of the manual of this study, which provides an excellent model for the treatment of acquired esotropia. When residual acquired ET wearing full plus is treated firstly with a prism to fully correct the ET, some patients will 'eat up' the prism and the angle will increase. If the prism is increased stepwise until there is no further increase and the "does" of bimedial recession is based on this newer larger angle of ET then the overall success rate of surgery in acquired ET increases to about 90% - a very impressive result!

A less well appreciated conclusion is a sensory one. If sensory fusion could be demonstrated and the patient was prismatically straightened, those patients did better after surgery (irrespective of the surgical dose!). Approx one third of the patients who had sensory fusion could not be demonstrated preoperatively. It thus seems that the largest factor in success is probably whether or not the patient is capable of some sensory fusion.

## GENERAL SURGICAL GUIDLINES

For those cases where orthotropia is sought and:

### - NORMAL AC/A RATIO

The amount of surgery is non controversial. Parks' or similar tables will produce good results.

### - HIGH AC/A RATIO

Surgery needs to be augmented. The three main approaches offer different ways of augmenting the usual bimedial recession:-

1. Increased bimedial recession. Parks introduced this 30 years ago. He increases the amount of surgery in a BMR by 1mm per muscle if the AC/A ratio is high.
2. Faden sutures. These are especially popular in Europe as the method of correcting the high AC/A ratio. Kushner (Archives 6/87) compared these two ways of augmenting bimedial recession for deteriorated accommodative ET with high AC/A ratio. The Faden patients had more over and under corrections than the increased BMR Group.
3. Operate for the near angle (using the usual tables).

### OPERATING FOR ET' ONLY

Several authors have operated on the near component only using the usual tables for the near deviation even if the patient is ortho for distance CC. The first few papers look good. Eg. Jampolsky Arch April 1974.

My own experience is small but also good and involves children 11-12 or older who have an uncosmetic ET' only, sometimes with near diplopia. The feared overcorrections have occurred infrequently because these patients have some fusional vergence.

### OPERATING TO "GET YOUR CHILD OUT OF GLASSES"

This is done by some. You can do it if the child absolutely refuses to wear glasses and continues to refuse and the parents give informed consent. I have never met such a child/parent combination.

For many years Gobin has been operating on the angle without glasses and claims good results. His rationale is as follows:

1. The natural history of hyperopia is to emmetropise.
2. The child needs to be horizontally straightened, hence needs a bimedial recession.
3. All motor factors that interfere with motor fusion need to be improved. The most frequently found factor is oblique dysfunction. This needs to be strenuously looked for a surgically corrected at the same time. This approach is not considered mainstream. This author believes that it has enormous merit but needs to be studied in a prospective controlled manner and this has not yet been done.

The ultimate paper on accommodative squints is still by Parks, AMA Archives of Ophthalmology, March 1958, p. 364, "Abnormal Accommodative Convergence in Squint". It's a long paper but the summary and tables must be understood

### OTHER ACQUIRED ESOTROPIAS

1. The acquired esotropia of High Myopia which seems to be due to compression of the Lateral Rectus Muscle against the Lateral Orbital Wall by an enlarging globe.
2. Divergence insufficiency type of acquired Esotropia.  $ET > ET'$  and sometimes  $ET' = 0$ . In the elderly this is usually due to microvascular disease and may be the late result of sixth nerve palsy(ies). In younger patients it seems to be unrelated to any CNS Pathology. In my experience younger patients with this are likely to be quite neurotic about the problem by the time they see me. BO Fresnels for driving often fix the problem.

### 3. INTERMITTENT ESOTROPIAS.

There are several different types:

- A. Cyclic Esotropia - 24 hour cycle. Very rare
- B. As part of accommodative spasm with acquired myopia and miosis.
- C. In un-or under-corrected hyperopes with Esophoria. Rx is to give full plus if the problem bothers the patient or if it is getting worse.

## CHAPTER 6

### ALPHABET PATTERNS IN STRABISMUS

#### SUMMARY

Vertical incomitance is common in horizontal strabismus ( $> 1/3$  of cases). Orbital and other factors are the commoner causes or associations. In fixing horizontal strabismus, it is important to lessen vertical incomitance in order to achieve a more stable longterm result.

$>1/3$  of horizontal deviations in childhood are vertically incomitant and are associated with an A or V pattern (or similar). They are often referred to as alphabet patterns.

Alphabet patterns are due to:

1. Oblique dysfunction due to abnormal oblique muscle anatomy.
2. Heterotopic pulleys which cause vertical displacement of the horizontal muscles or horizontal displacement of the vertical recti.
3. Intorsion or extorsion of the whole orbit.

#### COMMON PATTERNS

1. V. Increased ET on downgaze (dg) or increased XT on upgaze (ug) - the commonest pattern.
2. A ..... increased ET on ug or increased XT on dg. Less common patterns include 'X' (more XT (less ET) on ug and dg) and 'Y' (more XT (less ET) on dg only) and diamond pattern (more ET on ug and dg) which tends to be seen only after multiple surgeries or in thyroid cases.

#### WHY DO THESE HAPPEN?

##### 1. ORBITAL FACTORS

Orbital Morphology is an important factor in causing these patterns and it does so by influencing oblique function, eg. a shortened floor of orbit (an extreme eg would be Crouzons) will necessarily mean the I.O. origin will be postero-placed and the IO mechanics and function will differ from that in a 'normal' orbit. Specifically, it means that the angle and mechanical actions of IO will be different to the SO in the same orbit - they may not be matched and oblique dysfunction is expected. Another example might be plagiocephaly - the trochlea (= the origin of SO) is posteroplaced and thus the mechanical action of SO will not 'match' that of IO. In 1962 Fink showed that in 1/5 cadavers the obliques were angled 'wrong' and this would predispose to oblique dysfunction.

Jampolsky emphasises that on upgaze the IO is an abductor and the SR an adductor (ditto for SO/IR on downgaze). Any disturbance in this balance may produce an A or V pattern. Recessing both inferior recti may produce an A pattern according to this scheme, and, indeed, it often does!

The oblique dysfunctions secondary to orbital twisting are common in some genetic pools (not ours) EG the Mongoloid slanted orbit where the lateral canthi are higher than the medial are associated with intorted orbits and eyeballs and A patterns.

##### 2. INNERVATIONAL FACTORS

Abnormal innervation may explain some cases - on upgaze or on convergence there may be inappropriate co-contraction of inferior obliques (purely speculative).

##### 3. ABNORMAL MUSCLE ANATOMY

Demer has shown that the effective origin of the recti is from the pulley that begins at the level of the optic nerve - globe junction and extends forward by several mm. Displacement of the pulleys produces abnormal muscle function, eg, if the RMR pulley is displaced upwards, then on left gaze the right eye will elevate as well as adduct. An as-yet undetermined percentage of cases of apparent oblique dysfunction are due to heterotopic pulleys.

##### 4. TRUE SO PALSY

The best (and possibly only reliable) way to diagnose this condition is to do a coronal CT and demonstrate relative atrophy. Patients who have true atrophy typically develop floppy tendons. This can only be diagnosed on forced duction test under anaesthesia.

5. Tight muscles (e.g. tight LR in large long standing exotropias or 'tight' medial recti in Duane's type 1) can also seem to cause alphabet patterns (X and V respectively).

\*\* Do not call it an A or V unless the patient is wearing the correct refraction, the measurements are made on a distance accommodative target, and you have seen it on more than one occasion (these are the criteria the rest of the world uses).

\*\* If you test for alphabet patterns:

? Without the full correction

? For near

then you will always over diagnose these patterns especially V pattern.

#### WHY ARE THESE IMPORTANT

1. COSMESIS - Sometimes the oblique dysfunction is cosmetically weird.

##### 2. BARRIER TO FUSION

If you have a child who is horizontally straight (or potentially so with their own fusional vergence), then the vertical and torsional incomitance that these alphabet patterns cause may not allow fusion to occur.

#### WHEN TO OPERATE

Alphabet patterns are nearly always operated on "En Passant" - only if and when the associated horizontal deviation needs to be and is being fixed. Whilst operating for a horizontal deviation, I will modify the horizontal surgery and/or add oblique surgery in EVERY case of alphabet pattern (no matter how mild) to try and improve comitance which may improve long term motor and sensory stability.

## PRINCIPLES OF SURGICAL MANAGEMENT

Because the significance of pulley displacements and the possible need for orbital imaging in strabismus is new and evolving, and because these factors are highly relevant to the genesis and therefore the treatment of alphabet patterns, the treatment recommendations here will evolve quickly in the first decade of this 21st century – WATCH THIS SPACE.

The recommendations here are mainstream. Those that are not will be in italics.

In both A and V patterns, first select the surgery you would do for the deviation in the primary position and modify it or augment it as suggested below:

### 1. MAIN MESSAGES

\*\* Only weaken obliques if they OA and the other obliques UA and there is appropriate fundus torsion.

\*\* Beware SO tenotomy - don't produce a symptomatic fourth nerve palsy.

1a. IO OA/SO UA and fundus extorsion

Do IO weakening.

1b. IO OA and SO is normal or OA, or no fundus extorsion.

Do vertical shift of horizontals.

### 2. NO OBLIQUE DYSFUNCTION - UNCOMMON

Rx: Vertical shift of horizontals 5mm (1/2 tendon width). This is the only technique, which has been critically evaluated.

It works by changing the vector of the pull. If the terminal quarter or so of the MR is angled upward then the adducting vector is lessened because there is now a vertical vector as well. The insertion of the muscle should be moved in the direction in which one wants to most decrease its horizontal action, therefore:

? Move MR to the Apex of the V (also applies to A patterns), and this will open up the 'point' of the V or A because the pull of the MR is now shared between an adducting vector and a vertical vector.

? Move LR to the mouth of the V or A, and similarly the abducting pull of the LR will be lessened because it is now 'shared' with a vertical vector. The same type of shift seems to work in recess-resect surgery. This 5mm vertical shift of horizontals will fix about 15-20 PD of V and will never overcorrect the V.

One study suggests that whole tendon shift gives a bigger effect.

If a patient does not have the full "???????" (IOOA and SOUA and fundus extorsion) then I will do coronal scans centred around the optic nerve-globe junction. If there is clear vertical displacement of one of the horizontal recti, then that becomes part of the surgical treatment. The exact technique for shifting the pulley is evolving. It will probably have to be a full tendon width transposition augmented with a "Foster" suture.

### 3. LARGE PATTERN OF $\geq$ 25-30 PD

Need to do oblique weakening and vertical shift of horizontals at the same time.

#### A PATTERNS

1. If there is SO OA and IO UA and fundus intorsion you must weaken the SO to lessen the chance of recurrence of the A pattern (especially in XT).

2. If there is not the "full hand" then vertical shift of horizontals will be inadequate.

3. With SO weakening procedures, you must be very careful not to produce bilateral fourths with a whole new set of difficult-to-fix problems.

The SO weakening technique depends on the FDT.

FDT negative (can elevate  $>$  5mm in adduction): Posterior  $\frac{3}{4}$  tenotomy - you approach the SO temporally and excise about 1cm of the back  $\frac{3}{4}$  of the tendon at the insertion.

FDT positive: (definite reduction of elevation in adduction): Do either:

a. SO recession (say, to the nasal border of SR just behind the equator so you are still in the line of SO, otherwise you can turn SO into an elevator!).

OR

b. Using a 'spacer'. Wright has popularised the use of '240' silicone retinal buckle sewn in between two cut ends of SO tendon using 5/0 Dacron. See his atlas for details.

### 4. LARGE A PATTERNS - OVER 40 PD

I would do vertical shift of the horizontals combined with an SO weakening procedure as described above.

#### X PATTERNS

This is normally seen in long-standing XT, and is due to the tight LR syndrome. This is fixed by correction of the XT and specific treatment of the alphabet pattern is not necessary.

#### REVERSAL OF ALPHABET PATTERNS

Of 540 patients who Von Noorden operated for alphabet pattern, 17% had reversal of the alphabet pattern. The use of superior oblique tenotomy for the treatment of A pattern resulted in reversal of alphabet pattern 27% of the time.

His study also showed that reversal was less likely to occur if a V Pattern was associated with both overacting IO and underacting SO (and similarly for A Pattern).

## CHAPTER 7

### SUPERIOR OBLIQUE PALSY

#### Summary

The diagnosis of fourth nerve palsy (or simulating condition) can often be made from the history (vertical +/- torsional diplopia +/- head tilt). Careful examination is necessary to allow classification into one of the different patterns of fourth nerve palsy. The surgical management is very dependent on the correct classification.

#### THE HISTORY

##### 1. DIPLOPIA

Is often 'classical' (vertical, worse in the field of action of the muscle and lessened by head tilt). Sometimes presents as non-specific reading difficulty. You need to specifically ask for torsional diplopia patients rarely volunteer this as a spontaneous symptom. Ask them what happens to light poles or the line down the middle of the road - whether it looks like a "V" or like a tram line.

##### 2. PRESUMED CONGENITAL FOURTHS

This comprises two separate conditions:

a. True congenital fourths. This is possibly the result of birth trauma. Coronal CTs will show an atrophic belly of the SO. Forced duction test will show a long floppy tendon. These patients often decompensate in the teenage or adult years due to slow progression of the deviation which eventually exceeds the very large vertical fusional vergence amplitude that these patients typically develop (can be as much as 20<sup>Δ</sup>). These patients often have a head tilt in childhood photos, often have hemifacial asymmetry, and often have neck strain symptoms as adults.

b. Simulated congenital fourth nerve palsy. These patients look just like the above but the SO belly is not atrophic on CT and there is no floppy tendon. Some of these are due to heterotopic pulleys of the horizontal recti. Some of these are due to the oblique dysfunction that is seen in subtle disorders of orbital anatomy as mentioned in the section on alphabet patterns.

##### 3. TRAUMA

There is a history of head injury, nearly always with loss of consciousness. Some of these patients will have other problems at the upper end of the brain stem esp upgaze palsies and convergence problems. About ½ if these patients improve after initial presentation and you should wait 12 months before you plan surgery. Compared with other groups of patients, these patients are more likely to have incomitance greatest in the direction of action of SO and more likely to have a bilateral SO palsy.

##### 4. THE ELDERLY

These cases are presumed microvascular. Sinister intracranial disease causing an isolated 4th is extremely rare and neuro-imaging is not required in isolated 4th in the elderly. Most of these patients improve, and those that don't are more likely to be fairly comitant and respond to prisms. Usually small angle (<10<sup>Δ</sup>).

#### EXAMINATION:

An accurate examination is VITAL for an accurate description which in turn is necessary for accurate and predictable treatment. I recommend:

1. Measure the primary position vertical tropia for a distance target such as a Snellen letter. Measure the horizontal deviation in primary position, up and down gaze.

2. Repeat the vertical measurement with a Maddox rod (now measuring a vertical phoria). If phoria and tropia are very different the patient will probably have intermittent and variable symptoms and the treatment should be aimed at the larger angle, which has been demonstrated by the Maddox rod.

3. Measure the 'H' positions - primary, left gaze (LG), left and up (LUG), left and down (LDG), right gaze (RG), right and up gaze (RUG) and right and down gaze (RDG). I do this with the patient holding a Maddox rod over one eye; I move a torch with one hand about a meter from the patient in the extreme positions of the 'H'. In the other hand I hold a vertical prism bar to measure the vertical deviations.

4. Measure head tilt deviations. The patient tilts the Maddox rod parallel with the head tilt and the vertical deviation is measured perpendicular to the tilted position.

5. Measure TORSION using two red Maddox rods for the primary position and on DG. Exyclo >10 degrees in any position OR exyclo which increases by 100% from primary position to DG means bilateral fourth. Look for extorsion with the indirect ophthalmoscope. With the indirect the fovea seems to be level with the upper pole of the disc. If you imagine the disc to be the centre of the retina, if the retina looks extorted then it is.

6. Measure the range of single vision (SV). With the patient's thumb held horizontally at arm's length the patient slowly moves the arm up and down until double is seen while you measure from the side with a protractor. Do the

same measurement with the patient's thumb held vertically and moved to left and to right on the 'horizon'. These measurements should be repeatable to within 5 degrees. Many patients with fourths will have a horizontal deviation as well (usually a V Exo or an ET on downgaze). If these deviations are small (and they nearly always are) ignore them, and with correction of the cyclovertical deviation the patient will comfortably fuse this horizontal deviation.

#### V-PATTERN

These are common. They are secondary to the extorsion of the globe. If the globe is extorted, the inferior rectus will adduct on depression - hence V. If there is a V pattern greater a 20° strongly suspect a bilateral fourth.

#### WHEN TO OPERATE

In acquired fourths, wait until the situation is accurately documented to have stopped improving both by symptoms and signs, and to have been stable for at least 3 months. If acquired after a head injury wait at least 12 months after the injury before scheduling surgery. If the measurements start to get worse contractures have begun to develop and you need to schedule the patient's surgery soon. In all cases you should operate when the symptoms demand it and the surgical prognosis warrants it. Symptoms can include diplopia, a sore neck from chin down for reading, an uncosmetic head posture, problems going down stairs etc.

#### WHAT TO DO: IMPORTANT GUIDELINES!!

If you have examined your patient properly, the next decision is always straight forward.

#### THREE BASIC PRINCIPLES:

1. If the situation is incomitant, do incomitant treatment, ie. operate on the obliques.
  2. If the situation is fairly comitant, do comitant treatment (prisms or vertical rectus surgery).
  3. If there is significant torsion, which is not fusible with prisms in free space, you have to operate on an oblique.
- There are two main schemes for classification of fourths, the old Knapp classification and the newer Scott-Kraft classification, which is similar. I introduce a 3rd (simpler and different) classification:

#### 1. GROUP 1:

Small/moderate unilateral with incomitance greater in direction of superior oblique.

#### 2. GROUP 2:

Small/moderate unilateral with.... Inferior oblique.

#### 3. GROUP 3:

Large unilaterals.

#### 4. Bilateral.

#### 5. OTHER.

#### GROUP 1 & 2 - COMMONEST.

Features: Incomitant ++, primary position deviation <15°, largest deviation in the field of action of SO or IO, little/no deviation in opposite gaze. This is an incomitant situation and should usually be treated with incomitant surgery (IO or SO). One muscle will be enough.

GROUP 1: Incomitance greater in direction of action of SO. If the deviation is greatest in the field of the superior oblique, the best operation is an SO strengthening procedure sufficient to produce a mild Browns'. If you are not competent to do this, do contralateral IR recess and nasally transpose (to lessen excyclo). IO weakening will not produce results as good as the above procedures.

#### FLOPPY S.O. TENDON

Some cases of presumed congenital SO palsy will have a floppy SO tendon on forced duction testing (FDT). You need to do a few to get a 'feel' for normal. For a left eye, you grab the limbus at approx 2 and 8 with two pairs of Hoskins forceps and push the eye up and in. A normal FDT will see the 5 o'clock limbus just hidden, and a floppy one will see the Hoskins forceps disappear. If the result is uncertain the tendon is normal. SO tightening should be part of the operation in these patients to get good results.

#### FULL TENDON S.O. STRENGTHENING PROCEDURES

There are several procedures that all seem to be equivalent, e.g. tuck with a tucker, plication or folding, and advancement with/without resection. With any procedure you have to show intraoperatively that you are producing a mild Browns'. Put your sutures in a bow and then remove all your instruments and the speculum and do a forced duction test to show that you have produced a mild Browns'. You should be able to elevate the eye 2mm above the horizontal intercanthal line when adducted. I do plication either nasally or at the insertion with 6/0 Novafil and find that a 8-10mm plication gives good results in most patients.

If you do produce an unacceptable Browns' don't rush to undo it unless it is getting worse. A take-down of a SO plication/tuck is usually not completely reversible. In the first 1-3 days after SO surgery you may get an apparent SR palsy with marked hypotropia due (presumably) to SR neuropraxis from the handling of SR.

Do not do SO strengthening if your patient needs upgaze, eg. if wheelchair bound or a security guard required to look at a bank of monitors on a wall. In this circumstance do an IR recession and Harada, or an IO weakening.

## IR RECESSION

At the moment I do the square root of prism dioptres in the primary position as the mm of recession of IR up to a maximum of 4mm (eg.  $10^\Delta = 3\text{mm}$  IR recession).

### GROUP 2:

Incomitance greater in direction of action of IO. If the deviation is greater in the direction of action of IO or if the incomitance IO = SO then do an IO weakening procedure. Many people do 'one size fits all' procedures on the IO. I grade my IO weakening though at this stage I cannot justify this on scientific grounds. For 1-3+ IO IA I do a standard Parks IO recession to the lateral border of the IR 2mm behind its insertion. For 3-4+ IO OA I do an IO recession and anterior displacement level with IR insertion (IO OA 3+) or 2mm anterior to it (IO OA 4+) and the results seem to be 'better' than standard IO recession.

### GROUP 3: LARGE DEVIATIONS.

Primary deviation greater than  $15^\Delta$  - uncommon. These patients will also have a significant vertical deviation out of the field of action of the IO or SO. These patients are nearly always long standing fourths. You need 2 muscles unless a partial correction will be adequate in someone with a presumed congenital fourth where the large vertical fusional amplitudes will 'capture' any small residual deviation.

If the patient has either a SR contracture from "Chronic Hypertropia" or an IR contracture from "Chronic Hypotropia" of the other eye (these contractures are not always demonstrable on an in-office forced duction test) then an adjustable recession of the contracted rectus combined with IO weakening is required to give good results. If no contractures are present the choice of oblique recess-resect

### HUGE HYPERTROPIA - OVER 30 P.D.

Rare. First procedure is IO recess and SO plicate and recess a contracted vertical rectus.

### GROUP 4: Is it a bilateral fourth?

Strongly suspect bilateral fourth if -

? Excyclotropia greater than 10 degrees in any one position and/or increases by >100% on measurement in down-gaze.

? V pattern  $20^\Delta$  or more.

? Alternating hypertropia on head tilt (RH on right tilt, LH on left tilt)..

? In a presumed R fourth, 9 position measurement show LH in ANY position (or RH shown in any position in a L fourth).

? When these features are completely absent the case can still be proved to be bilateral when surgery for a unilateral fourth 'releases' a contralateral fourth = masked bilateral fourth. Sometimes this 'release' can be seen a week after the first surgery, more commonly it is months or years.

When you document or strongly suspect bilateral fourth, your options are:

1. Treat it bilaterally first-up. The 'safest' thing to do is the contralateral IO weakening if there is any IOOA, or a Harada if there is no IOOA.
2. Fix the largest fourth and wait for the contralateral one to become bigger and troublesome and then determine its features before you fix it.

### GROUP 5: OTHERS

5A: Essentially Comitant. Uncommon.

You can use prisms. Sometimes you need different prisms in the distance and near glasses (accommodation has not had any significant effect on the deviation but UG and DG does have an effect). A clever optician can sometimes use different types of bifocal segment to get a prismatic effect that way = Franklin biprism. If surgery is required, do comitant (SR or IR) surgery.

5B: Purely Torsional Diplopia.

Usually bilateral SO palsy. Torsion is >5 degrees and usually >10 degrees. Vertical deviations are < $5^\Delta$  and often 1- $2^\Delta$ . Do Haradas. The Harada is nearly always a simple procedure. Approach the SO tendon on the lateral side of the SR and use an iris repositor on the scleral surface to demonstrate the whole SO insertion. Split the tendon so that you isolate the anterior 1/3 and plicate it forward as far as you can (usually 5mm).

### MY RECENT EXPERIENCE

Of 208 consecutive cases of SO palsy seen privately from 1988 to early 1994, 136 (70%) were unilateral, and 30% (59) were either bilateral (36) or masked bilateral (23).

### UNILATERAL

Aetiology:

Congenital 38 (32%)

Trauma 50 (42%)

Vascular 26 (22%)

Other 4 (3%)

Unknown Cases 18 (13%)

This may not represent the true incidence of each type in the community.

TYPES:

Less than 15° in primary, and ...

Group 1: SO incomitance greater 37 (23%)

Group 2: IO incomitance greater 38 (24%)

IO = SO 61 (39%)

Group 3: Greater than 15° (4%)

11 (7%) unable to classify.

Surgery:

Total No. operated 34 (25%)

Average No. of surgeries per patient 1.3 (S.D. = 0.8).

**BILATERAL**

Aetiology:

Congenital 22 (40%)

Trauma 27 (49%)

Vascular 3 (6%)

Other 3 (6%)

Unknown cases 4 (7%)

Surgery:

Total No. operated 19 (32%)

Average No. of surgeries per patient 1.8 (SD=1.0).

## **CHAPTER 8**

### **DISSOCIATED VERTICAL DEVIATION**

The usual presentation of DVD is a spontaneous uncosmetic updrift of the non fixing eye when the other eye fixes. DVD has this prominent vertical component, and also has less prominent horizontal and torsional components (the eye usually drifts out a little when it goes up). The upward movement is slow and associated with extorsion, and the downward movement is also slow and is associated with intorsion.

There are many incorrect myths about DVD. DVD is fixation linked. It is not linked to inattention. It does not have (and if you understand the cause then it cannot have) a good natural history.

Cause

DVD is a manifestation of nystagmus blocking of the other (fixing) eye. The congenital nystagmus that is part of congenital strabismus commonly has torsional and horizontal component. To block the torsional component of the fixing eye may involve "tensing" of the superior oblique of that eye. This causes a number of Hering sequelae in the other non-fixing eye that typically first cause a small downward shift and then a slow upward and extorting shift. There is a variable horizontal shift as well if the horizontal recti are required to block the nystagmus in the fixing eye.

Presentation

This typically presents in the second or third year of life as an uncosmetic hypertropia. Examination findings will always show that if fixation is forced (eg by occluding one eye), the eye behind the occluder will drift upwards. If there is a large fixation preference then there may be a frequent, almost constant and sometimes constant hypertropia. If the hypertropia is frequent and large, a progressive superior rectus contracture becomes part of the clinical picture, and may eventually dominate the clinical picture.

Inferior oblique overaction is also common in congenital strabismus, and can coexist and be difficult to differentiate from DVD.

### **IS IT DVD OR IO OA?**

In an older child the difference should be easy even when they co-exist, though when they do co-exist accurate measurements can be VERY difficult. IOOA is never present on Abduction. IOOA is always associated with hypotropia of the other eye and the upward deviation of the hypertropic eye can be neutralised with a BU prism over

the OTHER eye. In DVD this will never neutralise the deviation - only a BD prism on the hypertropic DVD eye will neutralise the DVD.

#### MEASUREMENT

DVD can be quantified with the prism cover test. Let us assume the DVD is causing an uncosmetic updrift OD. Increase the amount of base down prism OD until all downward movement OD is abolished when cover is moved from it to OS (the prism sometimes tends to be 'eaten up' somewhat). The end point is sometimes a bit 'soft' and +/3-5 PD. While the BD prism is being held over OD, DVD OS will usually become apparent or greater and should be measured in the same way (while OD is being neutralised get the patient to hold the OD prism).

In a young child with both DVD and IO OA it can be very difficult to quantitate the relative contributions of these components to a hypertropia. The DVD may be incontinent and if so may be the cause of a tilt or turn. It commonly lessens on head tilt to that side (ie. DVD OD less on right tilt 'reverse Bielschowsky'). It may be more/less on left/right/up/down gaze.

#### VARIATIONS

If the superior obliques are tight (with A-pattern) then the DVD may be inhibited on adduction and only present in the adducted position.

#### TREATMENT

##### SR RECESSION.

By the time the DVD is sufficiently uncosmetic to warrant treatment, superior rectus contracture has come to dominate the clinical picture and can always be demonstrated with a forced duction test.

Until recently there was no doubt that the operation of choice is a bilateral large often asymmetric SR recession, and this is still the best proven technique. Unilateral SR recess surgery usually results in an uncosmetic DVD of the unoperated eye unless the first eye is very amblyopic. In Guyton's series of unilateral DVD surgery the mean interval before the second surgery was 10 months. You can justify operating on only one eye if it is most unlikely there will be a switch in fixation preference (eg. a profoundly amblyopic eye).

IO RECESSION. Recessing the IO to a point anterior to the lateral edge of the IR insertion (= anterior transposition) changes its action from an elevator to a depressor of the globe and is a very effective treatment of DVD (especially if there is uncertainty as to the relevant contributions between IOOA and DVD to the patient's problem, as so commonly happens).

#### HOW MUCH SURGERY?

SR RECESS. The commonest DVDs requiring surgery are 15+ PD, and as you can see from this recent table the numbers are very large. What amazes me is the uniform lack of complications such as upgaze limitations and lid problems with these large surgeries.

Prism dioptres.....mm of SR recession

10.....9

11-15.....10

16-20.....12-14

20.....16!

These SR recessions need to be done as a slingback suture. Under-corrections are fairly frequent with lesser numbers. You need to very carefully free the SR from underlying SO (there is always a 'frenulum' about 8-10mm behind SR insertion connecting it to SO) and adjacent intermuscular septum (go back behind the vortex veins) or SR won't retract sufficiently. Studies with stainless steel sutures in the cut end of SR have shown that a slingback recession of 'x' mm does result in the expected recession. IR resect has little/no place in primary DVD surgery but may be effective if SR recess is inadequate.

#### EXPECTATIONS OF SURGERY

Small SR recession commonly results in early failure; when Faden is added, late failure is common. Very large recession as recommended here is associated with approx 30% late failure. There is no information on the late results of anterior transposition of IO.

## CHAPTER 9

### DUANE'S SYNDROME (DS)

#### SUMMARY

The hall mark of Duane's syndrome is abnormal innervation to the lateral rectus and normal innervation to the medial rectus.

#### MAIN FEATURES OF ALL DS:

Retraction on attempted adduction

Limited abduction (often just to midline)

Slight limitation of adduction

In DS 1 the likeliest abnormality is a hypoplastic 6th nerve nucleus, absent abducent nerve and innervation of LR by the 3rd nerve. Two pathological dissections have shown this. In DS 1, therefore, the LR and MR co-fire when the eye tries to adduct.

One paper in the literature has described patients that look just like DS1 but do not have this electrophysiological feature (I have also seen a case). Some cases seem to have dual innervation of LR (sixth and third).

In published series, the next commonest type is DS 3 (limited AB- and adduction). The least common is DS 2 (limited adduction). In my practice (= the local genetic pool) DS 2 is more common than DS 3, and bilateral asymmetric DS is probably also more common than DS 3. Unilateral DS usually affects the left eye. It is much more common in females (locally at least 5:1). Various series have suggested frequent associations with other congenital eyeball problems and other systemic syndromes (especially hearing defects) but in the local genetic pool suggests DS is nearly always an isolated anomaly.

“DS 4” could be considered to include vertical variations of DS with retraction on attempted up/down gaze. In my experience co-innervational problems involving the vertical recti are frequently associated with DS 1-3. You suspect them if:

? there is any vertical deviation in the primary position and especially if there is a slow up/down drift on any of the ocular rotations.

? abduction is better on up or down gaze (V or X pattern).

“DS 5” is “the splits”. Attempted adduction causes a terrible exotropia – so called synergistic divergence. One can also see the splits on upgaze only – so called Y-pattern. Both of these are very rare.

Pseudo-Duane’s. There is a long list that includes known problems (such as medial blowout fracture, thyroid disease, over-resected muscles, post-transposition surgery (especially inappropriate transposition surgery), aberrant regeneration after 3rd or 6th nerve palsy, fibrosed muscles after myositis etc) and UNKNOWN problems such as congenital fascial bands within the orbit that are only discovered when you begin to operate on a ‘DS’ patient. There is a high incidence of DS 1 in patients with Thalidomide Embryopathy, an association of great embryological interest but not seen locally.

SENSORY ASPECTS.

There is usually slight or no amblyopia because typically there is a position of good binocularity. Diplopia is rarely complained of but is usually present in eccentric gaze when asked for.

IS IT OR IS IT NOT A SIXTH NERVE PALSY?

6ths will, of course, not be associated with retraction. In infants the situation can be difficult to assess accurately.

NATURAL HISTORY OF DS

Many cases of DS get worse with time with increasing stiffness or the MR leading to increased ET and increased face turn.

INDICATIONS FOR SURGERY

Some surgical pearls:

1. Recess - never resect; resection will make retraction worse.

2. Never weaken the obliques - they are the proptoters of the globe.

1. AHP (commonest indication for surgery).

Always assess this as accurately as possible (for distance and near) using a protractor while patient is looking at an interesting target. AHP is usually adopted to optimise binocularity and typically in the AHP position the deviation will be zero.

The aim of surgery is to move this position of orthotropia into the primary position. EG: L DS 1, ET 25 in PP and 0 on right gaze. The patient will have a left face turn, the LMR will be quite stiff on pre or intraoperative FDT, and the face turn will be fixed by LMR recession.

Surgical dose: Keep the numbers small (under 5mm) - an undercorrection will improve the situation and the patient will appreciate a modest improvement, whereas an overcorrection can be hard to manage. I usually recess the MR until the FDT allows 80+% abduction.

SACCADIC VELOCITY STUDIES may be useful. If the adducting saccade is < 150 deg/sec it means the LR cofiring is quite powerful and this patient may easily get an overcorrection if the MR is recessed. The same information can be qualitatively implied if there is marked retraction - it indicates significant LR action and risk of exotropia if MR recess is too large.

2. ET/XT in PP:

2A. ET. If unc cosmetic then ipsilateral MR recess 4-5mm will fix it. Do not do more than 5mm MR recess in DS. You need to do bimedial recession if the ET is large.

#### 2B. XT

Small XT - ipsilateral LR recess

Large XT - bilateral LR recess

Surgical dose: if LR is tight on FDT; do enough to almost free the FDT.

### 3. UNCOSMETIC RETRACTION

#### 3A. RETRACTION ALONE

Do same-number surgery on MR and LR - usually 5mm each.

#### 3B. RETRACTION WITH ET/XT

Treat the retraction first and then do the ET/XT surgery on the other eye, eg. retraction with ET 30pd - do ipsilateral retraction surgery (5mm recessions of MR and LR), and then contralateral MR recess of 6mm or so.

### 4. UNCOSMETIC UP/DOWN SHOOT

These are nearly always due to a tight LR, and not to oblique dysfunction. LR weakening will work because the bridle effect does seem to be causing the problem. I do LR recession > 5-6mm to fully free the FDT. If there is a vertical deviation in the Primary position and/or the up/down-shoot is not abrupt, then it may be due to innervational problems involving the SR or IR, and vertical rectus recession may be required to fix the problem. I have recognised this problem but never had a case requiring vertical rectus surgery for up/down-shoot.

### 5. SWINGING DOOR

If a patient fixes with the DS eye then the other eye will dramatically over-shoot on lateral gaze = swinging door.

Some claim to improve these patients with recessing both horizontal recti on the swinging eye.

### EXPECTATIONS OF SURGERY

#### BE MODEST.

Aim to improve, not to cure. Some of your patients will be made worse with surgery. Some of your MR recessions for face turn will fix the face turn but will convert a DS 1 to a DS 3 with almost no range of horizontal movement and a cosmetically unhappy patient. If an MR recess has caused an early over-correction don't wait more than a week or two to partially undo your surgery. Adult patients may have a dramatically improved face turn after surgery but a new area of diplopia, which drives them, crazy, and they will drive you crazy with complaints.

### TRANSPOSITION SURGERY

Some patients are upset by not being able to abduct beyond the midline. These patients can be treated as if they have a sixth nerve palsy and a whole muscle transposition is said to be a safe and effective procedure in improving abduction in DS by about 15+ degrees, but should not be done if there is any suspicion of vertical co-contraction or the retraction may be made much worse.

My own experience is small and not very happy. At UCLA experience is large and positive. At the time of writing, I cannot resolve these differences.

### BOTOX

It makes sense to inject the MR with Botox to lessen the restrictive changes. Experience is anecdotal.

## CHAPTER 10

### CONGENITAL NYSTAGMUS

#### SUMMARY

There are two main types of congenital nystagmus:

“Regular” congenital nystagmus (CN) - that which tends to be associated with eccentric null and convergence null.

Many of these also have strabismus. Typical wave form (increasing slow wave velocity). ‘Infantile’ N is a better synonym for CN. ‘Manifest’ and ‘Motor’ or ‘Central’ N are less acceptable terms for CN. CN is a type of cN that is associated with convergence and eccentric nulls.

1.

2. LMLN - Latent Manifest Latent Nystagmus (terrible name!!) - this type is always associated with strabismus, and has fast phase to fixing eye. In a patient who is binocular with equal vision LMLN will present as LN - Latent Nystagmus (nystagmus only present when covering one eye). In a patient where one eye has strabismus and slightly decreased vision it will present as MLN - Manifest Latent Nystagmus (one eye is amblyopic or suppressed and the eyes behave as if this eye were occluded, with a nystagmus fast phase to the other eye). There is no eccentric null. There may be a preference for fixing in adduction (extreme is Nystagmus Blocking Syndrome) or in intorsion. There may be a conveyance null.

The compensatory mechanisms that patients can use to improve acuity in nystagmus include:

1. Eccentric and convergence nulls in congenital nystagmus.
2. Excess innervation block nystagmus - tensing one or more horizontal recti with a very eccentric face turn and this tense rectus acts as a brake on the nystagmus.
3. Fixation in adduction or intorsion in LMLN.

## NATURAL HISTORY

Are the waveforms stable?

Reinecke studied the waveforms in CN prospectively and showed them to change from large triangular to small amplitude higher frequency pendular to jerk waveforms by the age of 18 months. The large triangular wave forms may have such large amplitudes and slow velocities that they appear to be purposeless eye movements and the child may be thought to be blind, though clearly the child does not function as if he/she is blind.

DOES THE HEAD TILT CHANGE?

The head tilt often improves through childhood. You should not offer surgery for this until the patient is about 7 years old UNLESS the child has a large ametropia (you should straighten the head so the child can look through the middle of the lenses and acuity develop properly) or looks very weird.

DOES THE VISION IMPROVE/GET WORSE?

Reinecke has shown that in adults the vision does seem to improve by up to one line per decade! Hugh Ryan (Melbourne) has also seen this on MANY occasions.

WHAT HAPPENS TO ANY HEAD BOBBING?

This also seems to improve with age in many patients. It is sometimes made temporarily worse after any muscle surgery.

Ocular abnormalities, especially albinism, come dystrophies and marked ametropias are frequent associations that are genetically linked to (but do NOT 'cause') CN. In the local genetic pool MOST patients do not have these associations.

When you examine a patient with cN it is very important to be as detailed as possible in your description. Knowing that a 1mm decentration is approximately equal to 10 degrees when assessed in the frontal plane, try and accurately measure the amplitude of the nystagmus. Get somebody else with a watch to time a 5 second period during which you count every beat of N that you see, divide by 5 and get the frequency. None of these measurements are going to be as accurate as that obtained on eye movement recordings, but trying to be as accurate as you can will force you to become a more accurate observer. It will also encourage you to examine the N for long enough to detect stability or the possibility of periodic alternating N.

CN

1. 'SIMPLE' DEFINITION: CN is a conjugate N that has one or more of the 12 CN waveforms defined by Dell'osso. It can be pendular or jerk. It may seem to have a latent component, which is usually not confirmed on eye movement testing.
2. About 1/3 of patients have strabismus c.f. LMLN where all patients have strabismus (but CN is 5 times commoner than LMLN so most patients with nystagmus and strabismus do in fact have CN).
3. 85% of CN patients have a NULL ZONE where their nystagmus is absent or nearly so.  
? 80% have a convergence null.  
? 60% have an eccentric null.  
? 50% have both convergence and eccentric null.  
? Only 14% have neither null.

The position of the null may be different for stationary, moving, near or distance targets. In a null zone the acuity is better, and if it is a lot better and is eccentric the patient will have torticollis.

A Null Zone is one that has nystagmus on both sides of it. A PSEUDO-NULL can be produced in extreme lateral gaze by having the relevant horizontal rectus so tight and contracted that nystagmus is lessened (muscle acts as a brake). This was first demonstrated by Bagolini who demonstrated both 'active blockage' and null zone as the cause of two different types of head turn. This can be called an excess innervation null. The 'true' null zone is associated with absent innervation.

There is no electrophysiological evidence of what happens to the medial recti in a convergence null - we do not know if it is due to increased innervation or absent innervation.

4. If the CN is jerk type, the direction of the jerk is the direction of the eccentric gaze from the null - eg, jerk right when looking to the right of the null.
5. May be associated with head bobbing which may appear to be apparently compensatory or in-phase with the CN.
6. The OKN response is often abnormal.
7. Positive family history is common and is fairly specific for a particular waveform or group of waveforms in a

particular family.

8. 'Sensory Defect Nystagmus': Ophthalmologists have known that congenital cataracts can be present in someone with no nystagmus, nystagmus can then develop, and when the cataracts are removed the nystagmus can then disappear. This type of patient has not been studied with electrophysiology and the classification of this type of nystagmus is obscure.

Another type of sensory nystagmus is seen as a monocular nystagmus in very amblyopic eyes and typically those that are also very highly myopic. If vertical and monocular it is called the Hiemann Bielschowsky phenomenon.

#### 9. ASTIGMATISM

Patients with cN often have significant astigmatism, which must be recognised and corrected. If the astigmatism is associated with significant torticollis one should offer early Kestenbaum surgery to relieve the torticollis to allow the patient to look through the middle of the spectacle lens.

#### LMLN

1. LMLN is a jerk cN that has one particular waveform and is manifest only with monocular fixation. LMLN has a very typical feature of fast phase to fixing eye and decreasing velocity of slow phase.

LN: It becomes apparent only after actually covering the non-fixing eye with an opaque occluder.

MLN: It is apparent with both eyes open (suppression alone is adequate 'cover').

2. The direction of the jerk is determined by the fixing eye - jerk right when fixing with the right eye, jerk left when fixing with the left eye.

3. It is always associated with strabismus. Whether it is LN or MLN depends on the type of strabismus that is associated with the nystagmus - MLN if a tropia and one eye is suppressed (= 'occlusion'), LN if straight with a phoria.

4. There is No NULL - neither convergence nor eccentric null. Some of these patients prefer to fix with the better eye adducted and get a face turn to that side.

5. Mixed forms are common, and difficult to evaluate, eg. patients with one or more CN waveforms and LMLN as well. Sometimes the preponderance of a particular waveform may depend on whether fixation is for distance or near.

#### SPASMUS NUTANS (SN) : A WARNING

The hallmark of SN is a dissociated nystagmus (and especially a monocular nystagmus) which is associated with a normal CT scan.

Associated but not essential features are:

? head bobbing typically first seen after the age of 6 months;

? the N. is more likely to be intermittent than CN, likely to have a faster frequency and smaller amplitude than the usual types of CN.

SN can be caused/associated with severe CNS disease (especially chiasmal glioma) or be 'idiopathic'. There is no way to clinically differentiate between SN associated with pathology from those cases of SN not associated with intracranial pathology.

With idiopathic SN, there is a good prognosis - the child is usually neurologically and developmentally OK, and the features of SN usually spontaneously resolve over some months. The patients I have seen have usually had a strabismus, which may outlast the SN and require treatment in its own right. One patient with SN developed progressive visual loss and was shown to have a retinal dystrophy.

#### THE ROLE OF THE GENERAL OPHTHALMOLOGIST

1. To recognise the features of CN in order to spare the patient unnecessary neurological workup. You must be aware of Periodic Alternating Nystagmus (PAN) - the history may suggest it (varying torticollis). You should also watch all CN patients for 2 - 3 minutes.

#### 2. ACCURATE REFRACTION

High refractive errors are common in CN. I have now seen several patients with marked torticollis who with accurate refraction had a marked improvement of their torticollis and better vision. Accurate refraction in CN is as important as it is difficult. In older patients contact lenses are worth a try because with N and torticollis it will be impossible for a patient to look through the optical centre of a lens with marked ametropia.

#### 3. PRISMS

If there is a convergence null for near, one can try and produce one for distance with BO prisms and see if this null will be an adequate replacement for an eccentric null. If BO prisms work well, you can then do a large bimedial recession to try and produce convergence 'stress' and improve vision that way (=artificial divergence surgery). Spielman has a large experience; she adapts the BO prism, does MR recess appropriate to the maximum BO prism, and has about 10% consecutive exotropia.

4. Look for associated sensory abnormalities - albinism (commonest +++), achromatopsia, optic nerve hypoplasia, Lebers, etc. These are commonly associated with CN.

#### IMPORTANT AND LITTLE KNOWN FACT:

In CN with these sensory abnormalities, the CN has wave forms indistinguishable from CN without sensory associations suggesting that CN is indeed a motor problem that happens to be linked genetically to these sensory difficulties.

5. Amblyopia is diagnosed in the usual way - with an occluder in front of the eye not being tested. Sometimes this produces a marked latent nystagmus of the tested eye and degrades the vision too much to allow an accurate measurement. In these (uncommon) cases I use +6 or +8 lens as an occluder. If amblyopia requires patching, Simonsz has shown that patching is best continued for 2 days at a time in CN.

#### THE ROLE OF THE STRABISMUS SURGEON

##### TORTICOLLIS SURGERY.

Australia has an important place in the history of CN surgery because Ringland Anderson in 1953 was one of the first in print with torticollis surgery. The principle of torticollis surgery is to rotate the eccentric null into the primary position. If the null is a true rest position null one seems to need a Kestenbaum procedure involving the recess/resect procedures to both eyes as discussed below. If the null is the active blockage null (as described by Bagolini above), then it seems that one needs to do a very large (crippling) recession of the horizontal rectus which is causing this block.

The one way that one can differentiate between these is that with a true rest position null some CN can be demonstrated beyond the null, whereas none can be demonstrated beyond the Bagolini null.

Parks introduced the 5-6-7-8 system doing 13mm of surgery per eye (5=MR recess, 6=MR resect, 7=LR recess and 8=LR resect). This is referred to as the Classic operation,

##### RECOMMENDATIONS FOR MANAGEMENT OF TORTICOLLIS IN CN ARE:

1. Only act if the history and your findings indicate absolute stability of the situation.
2. Measure the torticollis ACCURATELY and REPEATEDLY with a protractor - EYEBALLING IS OUT. Measure for distance and near - the measurements may be very different and 'successful' surgery for a distance torticollis may produce a near torticollis if this difference is not appreciated.
3. Try and get an accurate history regarding the FREQUENCY of the torticollis. Ask the parents to take frequent candid snapshots. If the visual gain from torticollis is not great then the frequency may be low and your office assessment when stressing the patient with small optotypes may overstate the 'real-life' defect.
4. The natural history in any one particular case is uncertain - some (?many ?most) kids improve and you should wait till age 7-8 before contemplating surgery.

##### SPECIFIC RECOMMENDATIONS

1. < 15 degrees: leave. If you get this result with surgery you and the patient are usually pleased.
2. 15-30 degrees: Classic surgery.
3. 30-45 degrees: augment Classic by 40% (18mm OU).
4. > 45 degrees: augment Classic by 60% (21mm OU).

These augmented surgeries WILL produce duction restrictions and diplopia (usually transient). Lesser surgery will produce fewer good results.

##### PATIENTS WITH TORTICOLLIS AND STRABISMUS

One common circumstance is NBS with amblyopia in one eye. Here the torticollis is determined by the fixing eye and this should be moved to the primary position. Surgery to the non-fixing eye should be performed for any residual strabismus at that or a subsequent surgery.

EXAMPLE: Dramatic face turn to the left with good vision OS when very adducted. The torticollis can be 40-50 degrees! In this common pattern the other eye is amblyopic. The face turn is 'driven' by OS, and forcing OD fixation with occlusion fixes the face turn.

One needs to really maim addiction OS, eg. a MR recess of 12mm and then do best guess surgery on the other horizontal muscles for residual strabismus. (See REINECKE in Acta Strabologica, Tel Aviv 1985 for specific and helpful guidelines).

##### IMPROVING VISION WITH SURGERY

If MLN can be reduced to LN, vision often improves. One way of doing this is to fix the strabismus (either with glasses or surgery) and by producing fusion the parameters of the nystagmus are lessened, and the acuity improves. In Zubcov's series a 57 year old patient with acquired A pattern exotropia, LMLN and oscillopsia had bilateral superior oblique tenotomies producing orthophoria and improving his binocular visual acuity from 20/80 to 20/30! (this was the best result in that study).

##### MULTIPLE RECESSION SURGERY

In 'routine' horizontal CN, Limon de Brown has had a large experience in four muscle recession (12mm each muscle) to lessen amplitude and frequency of nystagmus and improve vision in many/most of her patients. Reinecke has shown that patients with congenital PAN are the ones that are most likely to benefit from this surgery.

#### IMPROVING ACUITY WITH TORTICOLLIS SURGERY

With surgery planned for torticollis alone, some studies have suggested (eg. Taylor, Flynn and Dell'osso) that the visual acuity improves in these patients. It is not clear if this improvement is greater than that naturally seen in some patients.

#### VERY IMPORTANT REFERENCES

1. Reinecke's beautiful 3 page state of the art summary in the 1990 and 1991 Year Books Of Ophthalmology heading the Visual Physiology chapter.
2. Any recent review article by Dell'osso - an excellent one is in the Jpn Jnl Ophth 1985: 29, 351-368.
3. Mitchell Wheeler and Parks, J Ped Ophth Strab 1987: 24, 87-93, especially the Discussion by Calhoun.
4. 'The differential diagnosis of CN' Gresty et al, JD. Neurol, Neurosurg and Psych, 1984:47, 936-42. Very sensible medically orientated paper.
5. 'Compensatory mechanisms in CN' Von Noorden et al, Amer J Ophth 1987: 104, 387-397.

## CHAPTER 11

### EXODEVIATIONS IN CHILDHOOD AND IN ADULTS

Exodeviations can be considered to be of three main types:

1. 'Usual' exodeviations - childhood deviations, or those that behave that way. These begin as intermittent distance exodeviations.
2. Sensory deviations - as a result of poor vision the eye swings out.
3. Consecutive - after previous esotropia surgery.

#### WHY

When nearly everyone is anaesthetised the eyes divert. Thus the natural posture of the non-innervated orbits is exotropia. The alert state and Reticular Activating System results in relatively excessive innervation of the medial recti and the eyes straighten. It is possible that uncorrected hyperopia is a further stimulus to this. Certainly, uncorrected hyperopia in the first years of life is far more common in caucasians than in Orientals and Orientals have a far higher incidence of early onset exotropia than caucasians.

#### COMMON ABBREVIATIONS

XT, XT' = exotropia for distance, near.

X, X' = exophoria for distance, near.

EX=O, EX'+O straight for distance, near.

( ) = intermittent deviation.

#### TYPES OF EXODEVIATIONS

1. EXOPHORIA Convergence Insufficiency (CI; X', EX=O) behaves differently to the 'usual' childhood exodeviations. It is commonly secondary to accommodative disorders, and rarely to neurological disease.
2. INTERMITTENT EXOTROPIA (XT) with EX' = O or X'

This is the commonest presentation of childhood exodeviations and usually presents around the age of 2-3. Many patients progress towards constant exotropia and definite documentation of deterioration is adequate reason to consider surgery.

Evidence of deterioration may include a history of:

- ? deviation is more frequent.
- ? episodes of deviation last longer, eg. through several blinks.
- ? deviation is less easily controlled.
- ? monocular eye closure (MEC) becomes a constant feature when the child goes out-of-doors.
- ? diplopia - children rarely mention it to parents - ASK.

MEC is probably a diplopia avoidance technique. Deterioration by history may be associated with an objective decline in stereopsis.

At the stage of (XT) we have one or more of suppression, ARC and MEC as the adaptations. These are usually reversible and I would advocate surgery at this stage and results may be better now rather than waiting for a constant XT with the development of less reversible sensory adaptations.

#### BUT BEWARE!!

Hiles and others have shown that in (XT) and especially if < 20 pd many patients (50% !) do not get worse and some even get better (25%). It would be bad to exchange equal acuities, some stereopsis and a minor cosmetic defect for an overcorrection with ET, amblyopia and little/no binocularity.

Wolff of Wilmer has pointed out that about 20% of patients in his group lose acuity or binocularity following exotropia surgery, especially if the child is younger. Finally, a significant minority of optimally corrected exodeviations will recur. The surgery in exodeviation is certainly NOT a cure-all, SO DON'T RUSH!!

### 3. CONSTANT DISTANCE EXOTROPIA XT, X' or XT, (XT')

Fusional vergence has been completely abandoned and most of the potential for binocularity has been lost. Up to this stage the angle of the XT has usually stayed fairly constant (usually  $30 \pm 5^\circ$  - WHY? - there must be some constant fascial arrangements), and the frequency of the deviation has increased. At the stage of XT, XT' we often see secondary contractures changes and LR hypertrophy which increases the angle of the XT, XT'.

### APPARENT OBLIQUE DYSFUNCTION

Oblique overaction is very common, typically BOTH SOOA AND IOOA seen together with/without the presence of an alphabet pattern. The apparent overactions are a result of the mechanical limitations of excursion of the eyeballs and not true overactions - another simple and ingenious observation made by Guyton. Ask LK to demonstrate this to you.

TRUE OBLIQUE DYSFUNCTION should only be diagnosed if there is marked asymmetry between IO OA and SO OAs and usually is seen in the presence of a significant alphabet pattern.

M.E.C. Monocular eye closure is a classical XT symptom especially in well lit outdoor situations. MEC is more common in the XT patient with NRC than one with ARC (90% c.f. 35%!!), and is thus probably a diplopia-avoidance mechanism.

### WHEN TO CONSIDER SURGERY IN EXODEVIATIONS

1. Definite documentation of deterioration.
2. Symptoms, usually poor cosmesis, M.E.C. or diplopia.

### WHEN NOT TO OPERATE

1. An exo with unexpectedly good cosmesis, eg: - exo with negative angle kappa - would be exo on cover test but straight to light reflexes - routinely measure the Krimsky angle as well as the cover test angle to pick this up, or - exo and marked telecanthus.
2. If you are not sure of your measurements - neither you nor I nor anyone else then knows what to do.
3. Avoid surgery in the amblyogenic age group. If a child meets criteria for surgery then use temporising techniques such as giving optically inappropriate and unnecessary minus lenses to stimulate accommodative convergence. Kushner has shown that patients who wear these for up to 5 years seem not to have any increased incidence of induced myopia when compared to normal controls.

### HOW MUCH SURGERY

Tables exist based on empirical experiences. In CHILDHOOD exodeviations where you want to achieve stable orthotropia, some binocularity and no restriction of ductions then the table from Calhoun et al is a good guide:

Prism Recess Prism Resect

Diopres LR OU Diopres MR OU

15 4 18 4

20 5

25 6 27 5

30 7

35 8 36 6

45 9

55 10

70 11

### AIDES DE MEMOIRE

1. LR recess OU.  $30 \text{ pd} = 7 \text{ mm}$ .  
 $\pm 5 \text{ pd} = \pm 1 \text{ mm OU}$  over the range 15 - 40 pd.
2. MR resect OU.  $\text{Mm resect} = \text{square root of deviation in pd}$ .

A recent postal survey of American strabismologists showed that up to 25-30 pd most respondents follow the above guidelines and above 35 most did a little less surgery than the above table.

### PREOPERATIVE ASSESSMENT

1. DISTANCE TARGET is an accommodative target preferably a Snellen letter 6m away. If the acuity is 6/6 then I use a 6/12 letter for the target (to allow for blur caused by the thickness of the measuring prism). If the patient is unable to use Snellen then an interesting TV cartoon across the room may work.
2. NEAR TARGET is an interesting target that requires a significant accommodative effort, eg. a near letter or number chart, or a colourful/interesting picture object that the child is asked to describe.
3. FAR DISTANCE TARGET. This should be a target as least 100 metres away and out of the window.

4. DEVIATION MEASUREMENTS are made by cover test and prism measurement and never by eyeballing. A Krimsky measurement should always be done as well to numerically assess the cosmetic aspect of the exodeviation. You need to make the standard in office distance and near measurements in all positions of gaze. You then need to see if the distance measurement can be augmented by far distance fixation and by patching the eye for one hour. The common situation is that that patient has divergence excess exotropia. Approx half the time the distance angle is increased by one of the above augmenting procedures, and it is this augmented angle for which the surgical dose must be chosen.

More than half the time, the distance-incomitance disappears after one hour of occlusion: this breaks the tenacious proximal fusion that these patients have.

If the divergence excess persists, then you need to see if there is a true high ACA ratio and then +3 DS OU for near will abolish the distance-near incomitance. Patients who have this true high ACA ratio are more likely to need bifocals after surgery.

#### REBOUND

When you cover one eye, the other makes over-correcting movements before it fixates and it becomes impossible to define an endpoint with covertest. Sometimes this can be helped by optimising the refraction before doing the cover test measurement and/or using a more accommodative target. Sometimes these don't help and I then use a prism adaptation test.

#### DIVERGENT BLIND EYE (or nearly blind eye)

You can change the above numbers to do more MR resection. It doesn't matter if you produce a small abduction restriction, and you may get a more stable result, eg. from Parks -

#### PRISM DIOPTERS LR RECESS MR RESECT

40 8 6

50 9 7

60 10 8

70 10 9

80 10 10

#### WHAT IS THE LARGEST RESECTION/RECESSION YOU CAN DO?

In the 'usual' XT:

1. MR resect - you should produce no restriction of abduction with your MR resection. With a large XT you will have a very stretched MR and will be able to do a large MR resection without any problems. Check before you tie your final knots by intraoperative FDT that you are not causing a restriction.

2. LR recess - In order to produce an abduction deficit you probably need to recess LR 15+mm. Up to about 12mm recess should never produce a deficit and is probably always safe. If you do, WAIT; a recessed LR normally "takes up the slack" and ductions improve over a few months. If LR has already been recessed 10mm or so, a re-recession is best performed as a 5mm slingback.

In a deviated blind eye:

1. MR resect - that which restricts abduction to a cosmetically acceptable degree (typically 2/3 abduction) is fine, and may allow a resection of 10-12 or more mm in a large XT with a very stretched MR.

2. LR recess - see above.

You can augment these surgeries in very large deviations by intraoperative LR injection with Botox.

#### RECURRENT DEVIATIONS

Always do SAFE and PREDICTABLE surgery.

So, if reoperating for a blind divergent eye, redo the blind eye and nauseam and never operate on the good eye (=SAFE surgery). If there is no large sensory difference between the eyes choose virgin muscles (=PREDICTABLE surgery).

#### NEUROLOGICAL EXOTROPIA

##### 1. HEAD INJURY PATIENTS

XT is not uncommon and represents a partial third or a posterior fossa massage. Some characteristic features in these patients include:

i. CONVERGENCE INSUFFICIENCY - is common. Most recover spontaneously. Those that do not can be difficult to manage well. Prisms tend not to be effective because the loss of vergence gives little flexibility where the reading material can be held. Standard MR resect does not give good results - the distance/near incomitance persists. Nemet has published 3 cases of 'Biased MR resect' where he resects the lower half of the MR more than the upper half, and claims good results which I have confirmed.

ii. Some of these patients have lost their fixation reflex and no matter how straight you get them with surgery they drift out again within weeks.

iii. Diplopia is sometimes not a problem because of concurrent field defect, optic atrophy or general cognitive depression.

## 2. BRAIN DAMAGED KIDS

XT in these patients is often variable, surgery difficult to plan and the result unpredictable. A reasonable result is small angle ESOTROPIA that is probably more stable and attractive than small angle exotropia.

### ALPHABET PATTERNS IN EXODEVIATIONS

These are common and are treated in the usual way. A special case is A exo with marked SO OA and IO UA. SO weakening is important to prevent recurrence of the XT. Vertical shift of horizontal recti will be inadequate.

### SURGICAL EXPECTATIONS

Using the approaches indicated here, one typically expects an early esotropia. By 6-8 weeks the esotropia should have resolved. If it has not resolved by 3 months, consideration should be given to medial rectus Botox if the problem is troublesome.

At 12 months 18% of patients are orthotropic.

There is an approx 5% deterioration rate per annum after that.

### VARIABLE EXOTROPIA

How do you treat a patient with variable XT eg. varying from 0-45 pd? -- with/without diplopia?

If the patient has symptoms warranting treatment, then the options may include:

#### 1. PLUS LENSES:

If an XT patient has optically significant plus (eg. > +4) then this should be worn no matter what the deviation. If someone has a variable and lesser plus, then giving full plus may stabilise the XT. If it does do this, it should be worn and surgery performed for the stable (and often larger) XT.

#### 2. PRISMS

In someone with variable XT I will often do a PRISM ADAPTATION TEST. I prescribe the 'best guess' prism as a Fresnel to be worn on the day of the next consult. Any residual or new XT is then corrected with a larger Fresnel worn on the day of the next consult, etc. For most patients, only one or two visits are necessary. This is continued until the largest CT is 'released', and appropriate surgery is performed for the deviation. Very few patients require this.

3. LESSER SURGERY Variable XT indicates some fusional vergence with a 'capture range' that is not quite good enough. Less than the usual amount of surgery (and often single muscle surgery) may allow the patient to 'capture' the XT.

## **OPHTHALMIC MANIFESTATIONS OF HEAD INJURY**

### **25TH MEETING OF EUROPEAN STRABISMOLOGICAL ASSOCIATION JERUSALEM 1999**

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The information in this talk is based on an examination of several hundred patients with head injury over a fifteen year period. The core group is 290 consecutive patients who were first seen during the period 1984-1994. Nearly all of these patients were referred from Neurology Rehabilitation Units.

#### Epidemiology of Head Injury

In the state of Victoria, Australia (population greater than four million), there are approximately 5,000 hospital admissions per year for head injury. Two thirds of these victims are male. Two thirds of the total are under twenty-five.

## This Study

Two hundred and ninety patients were seen over the period 1984-1994 and all were referred because the non-ophthalmic medical staff in a Rehabilitation Unit detected symptoms or signs.

Eighty percent of the patients were seen in the first year after head injury. Follow-up was achieved in 43% of patients. The other 57% were not seen for follow-up. The reasons for this were varied and included geography (having been discharged from the Neurology Rehab Unit, they no longer found it convenient to attend a metropolitan ophthalmologist), being cured or disinterested. There is some anecdotal evidence to suggest that most of the patients who do not return for follow-up are in fact cured, as there is often significant medicolegal and fiscal advantage to them returning for care if they have persisting problems.

The Neurology Rehabilitation source of these patients means there is under referral of patients who would normally not be admitted to a Neurology Rehabilitation Unit and therefore there is under referral of the mildest cases and under referral of the severest cases of the head injury. Even though this is a different population to that in other studies, the similarity of the findings when compared to other head injury studies is striking.

## The Types of Problems

Following a closed head injury (CHI) there are problems that one can anticipate and can expect to find, eg. cranial nerve palsy or marked optic atrophy. There are also findings that effect many of these patients that are peculiar to the head injury population and are somewhat unexpected. These findings are not well acknowledged in the literature, are not well known by Ophthalmologists, are sometimes vague, often ignored by Ophthalmologists and if not ignored are misunderstood or undiagnosed.

The groups of symptoms and signs peculiar to head injury are:

1. Problems with accommodation and convergence (35%)
2. Non paretic motility disturbances (15%)
3. Subtle optic atrophy (20%)
4. Other non-specific visual disturbances

The groups of symptoms and signs that are expected are:

1. Cranial nerve palsies (50%)
2. Marked OA (5%)

## PROBLEMS WITH ACCOMMODATION AND CONVERGENCE

These are seen in 1/3 of patients. The three types of problems that are seen singly or in combination are:

1. Convergence insufficiency (19%)

This is defined as crossed diplopia for near in someone who is straight for distance.

2. Accommodative insufficiency (14%)

This is defined as someone under the age of thirty-five who develops symptomatic "presbyopia" whilst wearing the full manifest hyperopic correction for distance.

3. Pseudomyopia (13%)

This is someone who has never been myopic whom suddenly becomes myopic, or someone who had a lesser degree of myopia who suddenly needs a much stronger myopic correction. The acquired or change in myopia is abolished with cyclopentolate.

The treatment of convergence insufficiency can be:

1. No treatment. Monocular eye closure when required.
2. Prisms for reading.
3. Surgery. Bimedial resection and slanted bimedial resection have been tried.

The early results of surgery are variable and the medium-long term results are worse. Today I would only offer this to a patient who was not coping with conservative techniques and found a less than 50% expectation of improvement to be an attractive proposition.

The treatment of accommodative insufficiency is to give the appropriate plus that allows the patient to function for near. I tell the patient that they are likely to improve with time.

The treatment of pseudomyopia is to give the appropriate script as a crutch. The natural history of pseudomyopia is somewhat variable. There is the odd patient who gets worse from year to year. There is the odd patient who ends up needing different pairs of glasses at different times of the day. Early on in my experience, I would give cyclopentolate and bifocals as this seems to be a theoretically purer approach, but this was abandoned after a small number of trials because of patient complaints.

If we assume that all those patients who are lost to follow-up are OK, then convergence and accommodative insufficiency persists in 1/3 of patients and pseudomyopia persists in a little over half of the patients in whom it is first an issue.

## CRANIAL NERVE PALSIES

The data on cranial nerve palsies is offered largely so you can compare this series with your own experience to get a “feel” for the overall severity of this patient population.

Fifty percent of the patients had a cranial nerve palsy.

#### Fourth Nerve Palsy

One in three patients had a fourth nerve palsy and of these one in three were bilateral. Most get better. Forty percent end up requiring surgery. Of those that require surgery most end up requiring a superior oblique tuck for a Knapp class 2 palsy.

#### Sixth Nerve Palsy

One in eight patients had a sixth nerve palsy and one in eight of these were bilateral. Of the unilateral ones most get better and only one in four have surgery. Of the bilaterals only one in four get better and most have surgery.

#### Third Nerve Palsy

One in eight patients had a third nerve palsy. Of these nearly all were unilateral. It is probable that most patients with injury severe enough to cause a bilateral third nerve palsy don't survive. One in three have surgery to make their appearance normal or to lessen diplopia.

How long does it take for cranial nerve palsies to recover?

The “old” rules were that it takes a year for cranial nerve palsies to recover and that it is reasonable to offer surgery beyond twelve months.

We had a small number of patients with fourth and sixth nerve palsies who continue to improve beyond twelve months and continue to improve through to eighteen months. We had a small number of patients with third nerve palsies who continue to improve beyond two years. Miller has reported a similar experience in a small number of patients whose third nerve palsies had not improved at all by twelve months, but then did improve in the second year.

We thus recommend careful longitudinal assessment of these patients and intervention only when it is quite clear that they have stopped spontaneously improving, or have begun to get worse on measurements.

#### OTHER (NON-PARETIC) MOTILITY DISTURBANCES

These were seen in 15% of all the patients. None of these findings were anticipated and they are all probably peculiar to closed head injury population.

The commonest was exotropia (8%). This behaved just like a comitant childhood exotropia, but childhood exotropia was ruled out by history and there was no evidence of any third or partial third nerve palsy or INO. Some of these patients had surgery and in general the result was not as good as the results in “standard” childhood exotropia surgery.

Other motility disturbances that were seen and seem to be peculiar to the closed head injury population include:

- a. Binocular diplopia without any tropia. A small number of patients were seen who complained of binocular diplopia for which no cause was ever found.
- b. Esotropia. A small number of patients have comitant esotropia and another similar small number of patients have divergence insufficiency. These may be partly resolved sixth nerve palsies.
- c. Tropia, no diplopia. A small number of patients quite clearly have an acquired tropia, but never experience diplopia. This may be due to poor cognitive function. It may be due to acquired suppression. Diplopia may become symptomatic as cognition improves in, say, the second year after head injury.
- d. Skew deviation. Apparent IO palsy.
- e. Supranuclear gaze palsy.

#### OPTIC ATROPHY

Severe optic atrophy is an expected situation, and is seen in approx 5% of the total group of patients. It is easily diagnosed. The vision is 6/60 or worse and the disc is pale. There may be a chiasmal injury underlying it.

Subtle optic atrophy is a condition that is peculiar to head injury. Many of the patients complain of very non-specific symptoms, eg. blur which the examiner may not understand.

The condition of subtle optic atrophy is typically a constellation of several of the following:

1. Snellen acuity of 6/12 or better
2. Low contrast acuity (especially 10% acuities) are particularly bad.
3. Colour vision is often non-specifically abnormal. Desaturated colour vision tests may be more abnormal.
4. Non-specific field constriction is seen.
5. Unilateral mild pallor is easier to detect than bilateral mild pallor. NFL drop out or thinning can be appreciated especially with the direct ophthalmoscope.

Approximately 20% of patients have this mild optic atrophy and their symptoms can be misinterpreted if elegant psychophysical tests are not carried out. Standard Snellen acuity is quite inadequate in understanding the symptoms that these patients have.

## NON-SPECIFIC VISUAL DISTURBANCES

There are many other non-specific (sensory) visual disturbances that seem to be peculiar to head injury patients. Some of these symptoms may be a manifestation of the excessive introspection that patients may experience following a near death experience.

These include:

- a. Optically insignificant spectacle prescription is required for clear vision.
- b. Acquired reading disorder
- c. Loss of stereo
- d. Typical blepharospasm
- e. Acquired colour vision defect
- f. Palinopsia

## IS IT ORGANIC?

Are these “peculiar” symptoms functional or due to malingering or are they “real”?

By the label “functional” one means there is no organic basis to these symptoms. By the label “malingering” one means the patient is trying to deceive the examiner in some way. Ophthalmologists are very lucky in that we can examine “our organ” well and can do lots of different tests. It is thus easier for us to recognise “malingering” than, say, the neurologist who has to evaluate the patient who comes in with a weak arm.

Too many patients described the same symptoms for them to be imagined. The same symptom complexes are described in different patient populations! This strongly suggests that these peculiar problems are indeed “real”.

Most commonly, standard neuro-radiological investigations are either normal or non-specifically abnormal. SPECT (a nuclear medicine investigation) demonstrates diffuse multi-focal areas of damage following the acute head injury and these changes may persist. This test may be valuable for some patients in order to convince sceptical observers that there is indeed an organic problem.

## CONCLUSION

Visual problems following head injury are common. The commonest symptom seen in this patient group is diplopia, which can be due to a variety of causes. Many of the visual symptoms and signs that are found are peculiar to this group of patients and if the clinical patterns are not appreciated they may be inappropriately ignored or mislabelled.