Management of paralytic strabismus

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This paper is limited to the consideration of acquired ocular palsies attributed to involvement of the cranial nerve pathways, *i.e.* lower motor neurone lesions. It is not concerned with supranuclear problems, retraction syndromes, myasthenia, dysthyroid eye disease, or blow-out fractures. The words palsy, paresis, and paralysis tend to be used interchangeably, but usually paresis means a mild and paralysis a severe degree of palsy.

The cause of an isolated neurogenic ocular palsy is not always established but, in general terms, a IIIrd nerve palsy is often due to an aneurysm, a IVth nerve palsy to trauma, and a VIth nerve palsy to tumour or trauma.

Autopsy studies of road accident victims by Heinze (1969) have shown that avulsion of the nerve rootlets from the surface of the brain stem may cause III, IV, or VI palsies. Other IIIrd nerve lesions were due to focal softening in discrete axonal bundles in the proximal segment of that nerve, and intraneural haemorrhage at the superior orbital fissure. VIth nerve lesions were either at the brainstem surface or at its intraorbital termination in the neuro-vascular hilum of the lateral rectus muscle. Only one patient had the VIth nerve damaged by the fractured tip of the petrous temporal bone. When it comes to apportioning significance to *minor* head trauma as the possible cause of oculomotor palsy, the recent study of Eyster, Hoyt, and Wilson (1972) has shown that this may be an initial sign of a basal intracranial tumour.

In children, a posterior fossa tumour must always be considered in cases of abducent palsy. The associated vestibular upset may lead to a fall and this *injury* may be accepted as the cause of the palsy without further search. Even Gradenigo's syndrome is not always due to middle ear infection but may be caused by a tumour.

Two important causes in young adults are multiple sclerosis, and an unrecognized congenital ocular palsy that is becoming decompensated. Finally, virus infection is often invoked on slender evidence, particularly for recurrent VIth nerve palsies.

Prognosis

Hugonier and Clayette-Hugonier (1969) claimed that an isolated ocular palsy, or, one where the cause has ceased to act (*e.g.* head injury) had a good prognosis, more than 80 per cent. of such patients being completely cured within 3 to 6 months.

Bielschowsky (1939) found that only 39 per cent. of his patients recovered spontaneously, IIIrd nerve palsy having the worst outcome.

A follow-up of 45 adults (Rose, 1973) showed complete cure in one-third, residual palsy in one-third, and, surprisingly, one-third dead! He emphasized that isolated ocular palsy should be regarded seriously as a warning of significant systemic disease.

Signs and symptoms of paralysis

These are well known. Diplopia and false orientation due to erroneous projection of the visual field may provoke "ocular vertigo".

In VIth nerve paresis some abduction persists, but towards the limit of movement the motion is often jerky. This is called “paralytic nystagmus”. When both eyes are open the range of movement is usually less than when the good eye is closed and the patient compelled to use the affected eye only.

Because of the defective movement, the axes of the two eyes do not correspond in positions that necessitate the action of the paralysed muscle. The deviation of the axis of the palsied eye from parallelism with that of the sound eye, is termed “the primary deviation”. If the sound eye is covered, obliging the patient to fix the object with the affected eye, the sound eye is moved still farther in that direction. This increase in the deviation of the visual axes is called the “secondary deviation”. The occurrence of secondary deviation depends on the fact that normally two muscles which act together are equally innervated for a given moment. If one is weak and an effort is made to contract it (as in fixing with that eye), the increased innervation also influences the other muscle and causes an undue contraction. It is as if a rein acted equally on a hard-mouthed and a tender-mouthed horse yoked together; the effort to make the former deviate would cause an excessive deviation of the latter. The secondary deviation is a delicate test for weakness of an ocular muscle and may reveal its existence when the primary deviation is too slight to be observed. One result of the increased innervation that causes the secondary deviation is the erroneous projection of the visual field. The neurologists will recognize that this description of signs and symptoms, all of which including the phrase “yoked together”, is from “Diseases of the Nervous System” (Gowers, 1888).

Sometimes the patient finds that the diplopia may be counteracted by adopting an abnormal position of the head. This compensatory posture involves moving the head in the direction of action of the palsied muscle. The head position is best considered in terms of face turn, chin elevation or depression, and head tilt, which result mainly from palsies of the horizontal rectus, the vertical rectus, and the oblique muscles respectively. Although the evolution of the palsy may affect the compensatory head posture, certain generalizations are worth making. An abnormal head posture confirms the presence of a paralysis and is strongly suggestive that binocular single vision is present.

Evolution of the paralysis if recovery does not occur is a tendency towards concomitance, explained in four stages:

(1) Overaction of the ipsilateral synergist:

– Right superior oblique → + Right inferior rectus

(2) Overaction/contracture of direct antagonist, especially if fixation is by the normal eye, and understimulation of *its* yoke muscle:

– Right superior oblique → + Right inferior oblique → – Left superior rectus

(3) True overaction of the yoke muscle, especially if the paretic eye fixes, and underaction of *its* antagonist:

– Right superior oblique → + Left inferior rectus → – Left superior rectus

(4) Underaction through secondary inhibition of the “contralateral antagonist”:

– Right superior oblique → + Right inferior oblique → – Left superior rectus
– Right superior oblique → + Left inferior rectus → – Left superior rectus

It is important to note the role of the dominant eye here. If the sound eye fixes under normal conditions, then the direct antagonist overacts, but when the paretic eye is dominant the

yoke muscle overacts. The development of contractures makes the diagnosis of the affected cyclovertical muscle more difficult, but the 3-step methods of Parks and Hamtil (1971) simplify this:

- (1) Find in which lateral gaze position the hypertropia is maximal, four possible muscles.
- (2) Determine if there is right or left hypertropia two possible muscles. *e.g.* Left hypertropia means either right superior rectus or left superior oblique.
- (3) Use the head tilt test to decide between these two possible muscles. The greater vertical deviation on tilting the head to one side while the patient maintains distance fixation gives the answer.

How may the course of a paralytic squint be assessed?

Scott (1971) has made useful suggestions from his work on lateral rectus palsies which are the simplest group to study. He has examined the *speed* of saccadic eye movements by means of electro-oculography, and by using a limbal sensing device, when horizontal saccades were generated volitionally or by optokinetic nystagmus. Robinson (1964) has shown that quick saccadic movements result from a large, briefly applied, excess force delivered by the extraocular muscles. Clearly, if the muscle is paretic, such a force cannot be generated. Following a horizontal recession-resection operation, the static position of the globe has been changed but not the mechanical damping of the orbital tissues, nor the inadequate innervation of the paretic muscle. Saccadic velocities after such surgery are not improved.

Scott (1971) made further studies of both the active force that could be generated by the lateral rectus and the development of medial rectus contracture in abducent palsies. A suction cup was attached to the eye and a silk suture connected this cup to a strain gauge. This allowed measurements of the stiffness of the muscles and the resistance of the orbital tissues to rotation. The active force produced by the lateral rectus was measured by holding the paralysed eye stable with the suction cup whilst the good eye made saccadic and following movements. A clinical estimate of lateral rectus active force was made by holding the eye still with forceps during attempted eye movements. It was interesting that electromyography did *not* correlate with the active force assessments. The factors that determine prognosis are:

- (1) How soon and how fully lateral rectus force returns,
- (2) How extensively medial rectus contracture develops in the meanwhile.

At this stage it is worth briefly reviewing the types of muscle transposing operations that have been tried. These procedures should be reserved for abducent paralysis, and in the infant this means that total occlusion of the "good" eye, doll's-head rotation, and whole-body rotation which give powerful vestibular oculo-motor drive must all fail to cause abduction. Scobee (1952) used to insist on always doing a recession-resection operation, but this is only worth while if there is some lateral rectus functional recovery.

The pioneer work of Hummelsheim (1907) in the early years of the 20th century, in which the lateral half of each vertical rectus is attached to the lateral rectus, is the basis of many "transplant" operations. The monkey experiments reported by Marina (1915), in which he transposed various ocular muscles and observed the results of cortical stimulation, led him to conclude that the central nervous system control of the eye muscles is not fixed. Several workers since then have performed experiments claiming to support this relearning thesis.

O'Connor (1921, 1935) described many different methods which followed from his belief that "it is worth while to try transplantation as the cases are otherwise hopeless and

in any event cannot be made much worse". He tried moving the whole of each vertical rectus laterally, and moving the *nasal* halves of each vertical rectus to the lateral rectus.

How these operations work was never satisfactorily explained. Re-learning processes for something as complex as co-ordinated ocular movements in adults is not an easily accepted notion and the experimental evidence for this leaves much to be desired. Woodruff (1917) felt that post-operative adhesions to the globe could account for the observed results. This view was stated much more forcibly by Verhoeff (1942), who was "perplexed that even in the dark a monkey could, within a few days interchange the functions of two ocular muscles of the same eye". He declared that each pair of transposed muscles must have crossed each other in the orbit near the globe and that each muscle must have carried with it check ligaments and parts of Tenon's fascia which together exerted some traction in the direction of the original pull of the muscle.

Metz and Scott (1970) and Metz, Scott, O'Meara, and Stewart (1970) have recently settled the question by electromyographic recordings from the transposed superior rectus and the untouched superior rectus in the contralateral eye. The EMG firing patterns showed that both muscles still recruited together on upward gaze, and inhibited together on downward gaze. Similar EMG tracings from a transposed right lateral rectus showed that this muscle still recruited in right gaze and was inhibited in left gaze. Furthermore, orbital dissection of these monkeys *post mortem* showed extensive adhesions between the muscles at their crossing point and to the adjacent globe which became the functional insertion.

The clue to how these repositioned muscles work was given by a demonstration in a human patient (Hildreth, 1953). He operated under local anaesthetic on a man with esotropia (but *not* due to VIth nerve palsy) and first detached both horizontal recti (Fig. 1). A silk loop was passed laterally around the muscle bellies of the superior rectus and inferior rectus and tightened so that the pull of the vertical rectus muscles was displaced lateral to the vertical axis of rotation of the eye. On command the patient could abduct this eye

immediately, and the tighter the loop was pulled, the greater the abduction power. Hildreth rightly concluded that, since the abduction was immediate, no new nerve pathways had developed and that re-learning did not have to be invoked. The delay in re-establishment of "normal" ocular movements in various animal experiments which was claimed to be the central re-education time was merely allowing for adequate adhesions to develop. To my knowledge Hildreth's experimental demonstration was never employed as a routine surgical procedure, but Jensen (1964) reported a new operation to restore abduction which was clearly derived from Hildreth. He split each vertical rectus, but did not disinsert it, and sutured each lateral half vertical rectus to the adjacent half of the similarly split lateral rectus.

'The 4-0 cotton suture is then threaded once more around the muscle bundles and tied with sufficient tension to hold the muscles together but not so tightly as to shut off the blood supply'.

I rather doubt if any blood still gets through if the suture is pulled tight enough to approximate the muscle halves at the equator, but normally the vertical rectus muscles have two or more anterior ciliary arteries running forwards over the muscle insertion and the nasal vessels of each vertical rectus are left untouched (Fells, 1972).

Results with Jensen's operation are still unpredictable. Jensen (1964) reported an average of 30 Δ of abduction in eighteen cases, but even he produced exotropia in five.

Another group of patients who have not been easy to help in the past are those with *bilateral* superior oblique palsy, usually from closed-head injury. If the accident was sufficiently severe to cause loss of consciousness then I always assume that the trochlear palsy is bilateral unless examination proves otherwise. The torsional diplopia, particular on downward gaze, is peculiarly disabling to the patient and may be helped only slightly by chin depression. The bilaterality of the lesion may be missed from a perusal of the Hess charts when one side is more severely affected than the other, but the switch-over in torsional diplopia on latero-depression leaves no doubt. Lyle (1964) described three types of operation that could help cyclotropia: bilateral inferior oblique myectomies; superior oblique plications; and re-alignment of the inferior rectus attachments so as to reduce the obliquity of their insertion, making the medial end further from the limbus than the lateral end of its insertion.

Harada and Ito (1964) described anterior partial advancement of the superior oblique muscle to correct extorsion.

The assessment of cyclotropia before surgery leaves much to be desired and until recently we have used specially devised slides in the synoptophore. Unfortunately the results are variable, but in the past few months Dr. Harden and Mrs. Dulley in my Department have adapted an old method using the Hess-Lees screen to give accurate quantitative results. The patient's wand ends in a T-piece with a universal joint allowing it to be placed easily in various directions in relation to the arm. The orthoptist indicates the main points on the screen in turn, and the patient has to place the bar as accurately as possible along the horizontal line of the screen passing through that point. The bar is made equal in length to two divisions on the screen. Both ends of the bar have their position accurately marked on the screen. After all the relevant points have been plotted, the examiner uses a modified protractor to measure the angle made between the bar and the horizontal line passing through that particular point. This test usually reveals more torsion than the synoptophore slides, but we believe it to be more accurate because the patient finds the test easier and hence quicker to do and is less fatigued by it. Finally, the results are more readily repeatable.

The operation that I do for correction of excyclotorsion is bilateral advancement of the anterior half of each superior oblique, moving the superior oblique forwards and laterally (Fig. 2).

FIG. 2 Left eye seen from above, indicating how the anterior half of the superior oblique is moved forwards and laterally, moving the axis around which the superior oblique rotates the globe closer to the antero-posterior axis and thus improving intorsion

Because I believe that the superior oblique rotates the eye around a fixed axis within

FIXING RIGHT EYE		FIXING LEFT EYE	
To Patient's Right	To Patient's Left	To Patient's Right	To Patient's Left
$-1^{\circ}L/1\frac{1}{2}^{\Delta}$ R Ex 7°	ϕ R/ 1^{Δ} R Ex 7° L Ex 2°	ϕ L/ 2^{Δ} R Ex 5° L Ex 2°	ϕ R/ $1\frac{1}{2}^{\Delta}$ R Ex 4° L Ex 2°
	$\circ$ R Ex 7°		$+1^{\circ}R/2^{\Delta}$ R Ex 10°
$+3^{\circ}L/2^{\Delta}$ R Ex 17° L Ex 2°	$+5^{\circ}R/3^{\Delta}$ R Ex 17° L Ex 2°	$+3L/2\frac{1}{2}^{\Delta}$ R Ex 17° L Ex 2°	$+3^{\circ}R/5^{\Delta}$ R Ex 15° L Ex 2°

Case 1 Preoperative synoptophore readings (December 11, 1969)

FIG. 3 Case 1. Bilateral IVth nerve palsies. Preoperative synoptophore readings and Hess chart

the orbit, I regard this operation as altering the plane of action of the superior oblique, making it more antero-posterior and therefore producing the greater intorsion that is required. If there is significant extorsion on upward gaze, then both inferior obliques are recessed and I normally deal with all four obliques at one operation. An example is given in Case 1.

Case 1, a married woman aged 26 years, fell from a horse and was unconscious for the next 2 weeks. On recovery she complained of intolerable diplopia, mainly on downward gaze. A total of 19° of excyclotorsion was measured on downward gaze, with 7° of excyclotorsion on upward gaze (Fig. 3i). The Hess chart (Fig. 3ii) shows the bilateral superior oblique weakness.

After surgery, at which both inferior obliques were recessed and both superior obliques had their anterior halves advanced and moved laterally, her head position immediately improved. In fact her friends commented on her upright posture with the chin now slightly raised because of some incyclotorsion on up gaze (Fig. 4i). An appropriate improvement is seen in the Hess chart (Fig. 4ii).

FIXING RIGHT EYE		FIXING LEFT EYE	
To Patient's Right	To Patient's Left	To Patient's Right	To Patient's Left
$+2^\circ R/4^\Delta$ R In 2° L In 5°	$\phi R/2^\Delta$ R In 5° L In 5°	$+1^\circ R/5^\Delta$ R In 3° L In 4°	$\phi R/2^\Delta$ R In 3° L In 4°
	$+1^\circ R/1\frac{1}{2}^\Delta$ R In 2°		$\phi R/2^\Delta$ R In 3° L In 2°
$\phi R/1\frac{1}{2}^\Delta$ R Ex 1°	$+1^\circ R/1\frac{1}{2}^\Delta$ R Ex 1° L In 5°	$+1^\circ \ominus$ R Ex 1°	$+1^\circ R/1^\Delta$ R Ex 1°

Case 1 Post operative synoptophore readings (June 10, 1970)

FIG. 4 Case 1. Postoperative synoptophore readings and Hess chart

After 3 years the field of binocular single vision was central but reduced in extent (Fig. 5, overleaf). A course of orthoptic exercises soon restored this to a satisfactory area (Fig. 6, overleaf).

It is vital to remember that any person with diplopia after head injury must be carefully tested for fusion. Trauma may also destroy fusion and in these circumstances any surgery merely worsens the diplopia.

IIIrd nerve palsy is the most difficult of all because of the number of muscles involved.

FIG. 5

FIG. 5 Case 1. *Field of binocular fixation 3 years later.*

FIG. 6

FIG. 6 Case 1. *Improvement in field of binocular fixation after orthoptic exercises.*

oblique just above the medial rectus insertion (Metz and Yee, 1973). Using this approach An additional complication is that the regenerating nerves have too many routes to choose between, with the resultant intriguing misdirection—regeneration syndrome. Two common signs of this misdirection are lid retraction on looking down and away from the affected side, and pupillary constriction on downward gaze or on adduction. Treatment is disappointing. Some surgeons confine their attention to the good eye and do maximal recessions of the superior and lateral rectus muscles. Helveston (1973) described a ‘super-maximal’ recession-resection operation when the vision was poor in the affected eye. A 10-mm. lateral rectus recession was followed by a double 80 per cent. marginal myotomy of the same muscle, plus a 12-mm. resection of the medial rectus. If necessary, vertical rectus recession-resection procedures could be done 4 months later for hypotropia.

If the affected eye has some adduction Metz advises medial rectus recession and lateral rectus resection of the affected eye. For a complete IIIrd nerve palsy he balances vertically-acting forces by fracturing the trochlear and attaching the resected tendon of superior

FIG. 7 Case 2. *Left partial IIIrd nerve palsy, preoperative Hess chart*

to the affected eye, Reinecke (1972) has corrected over 60 Δ of exotropia in congenital IIIrd nerve palsies. Metz and Yee (1973) state clearly that the goal is not to normalize ocular rotation in a total IIIrd nerve palsy since this is impossible, but to straighten the eyes in the primary position. No one seems keen to invoke re-learning here, claiming that the transposed superior oblique acts as an adductor. As a last resort, Knapp recesses all tissues on the temporal aspect of the globe to the level of the lateral orbital rim and anchors the eye in a slightly overcorrected position with a suture. If the affected eye has good vision, Hugonier and Clayette-Hugonniere (1969) refers to 'providential ptosis' in

FIG. 8 Case 2. After right inferior oblique recession

FIG. 9 Case 2. Right hypertropia 8 months later

FIG. 10 Case 2. After right superior rectus recession

total IIIrd nerve paralysis and, although defeatist, this realistic attitude may be best for the patient in the end. An example is given in Case 2.

Case 2, a married woman aged 62 years, suffered a left partial III nerve palsy in a road accident (Fig. 7) for which a right inferior oblique recession was performed (Fig. 8) but with only a temporary effect. Right hypertropia soon became apparent again (Fig. 9) and the right superior rectus was recessed (Fig. 10). This gave an almost full field of binocular single vision (Fig. 11), yet the patient still complained! She was particularly distressed when trying to look out of the window of a moving vehicle, and also when walking along the street. Checking the traffic before crossing the road always had to be done with her left eye closed because the quick glances to right and left caused diplopia. The surgical 'cure', as judged by static measurements, was not effective under everyday conditions of viewing because the *velocities* of the eyes could not be matched.

FIG. 11 Case 2. Final field of binocular fixation

Summary

Patients with ocular palsies need full investigation to try and establish the cause. Whilst waiting for spontaneous improvement, the affected eye may be occluded if adequate binocular single vision cannot be attained by adopting a compensatory head posture. When the patient is in familiar surroundings, the good eye should be occluded as this may reduce the development of contractures. Prisms have a very limited role, if any. After 6 months, if the Hess charts show a static situation, appropriate corrective surgery may be performed. The problems of contracture and of reduced speed of movement of the affected eye, even after good static alignment, should make us all cautious in our prognoses.

A cine film demonstrated the simplicity of the operation of advancement of the anterior half of the superior oblique, particularly when the speculum is replaced by lid clips to the upper lid only to give the maximum exposure. It is never necessary temporarily to disinsert the superior rectus for full access to the superior oblique insertion.

A second film illustrated the reduced speed of abduction in an adult who had had full abduction restored by horizontal recession/resection for a left partial VIth nerve palsy. As this patient followed an object moving across his field of vision from right to left the eyes alone pursued the target until only a few degrees to the left of the mid-position and the following was then continued by face turning which still allowed binocular fixation. This was his compensation for slow left abduction.

I wish to thank the Audio-Visual Department of the Institute of Ophthalmology for the films and slides. I am indebted to Mrs. Barbara Dulley for her invaluable orthoptic assistance.

Discussion

VON NOORDEN I would question the value of moving the tendon of the superior oblique for exocyclotropia rather than weakening the inferior oblique muscle or tucking the superior oblique.

FELLS What evidence have you that the inferior oblique produced exocyclotropia on downward gaze? I limit this procedure to correcting extorsion on downward gaze.

VON NOORDEN I agree that the correct procedure for this specific circumstance is to tuck the superior oblique or to do the procedure that Mr. Fells has described. However, if the exocyclotropia was greatest in the primary position or on upward gaze then I should still do the inferior oblique myectomy.

FELLS The only time an exocyclotropia is troublesome is when it occurs on downward gaze and interferes with reading, and this is the reason why I prefer my procedure. Superior oblique plication produces very little improvement in the cyclotropia but always limits elevation in adduction. The procedure which I have shown is very easy.

LYLE I agree that this is an extremely good procedure which I have performed for many years. As far as I know it originated in Japan. It is, however, quite possible to overdo its incyclotropic effects.