Bilateral superior oblique tendon sheath syndrome

Occurrence and spontaneous recovery in one of uniovular twins

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Brown (1950) defined the features of the superior oblique tendon sheath syndrome and reported five cases. He postulated that the restriction in elevation of the affected eye in the adducted position was caused by a congenitally short anterior tendon sheath of the superior oblique muscle. As the eye moved into adduction, this ligament-like sheath became taut and strongly opposed the action of the inferior oblique muscle. By surgery, he showed the forcedduction test for elevation in adduction was positive until the superior oblique tendon sheath was cut; the forcedduction test then became negative as the eye could be fully elevated.

Some of Brown's early cases were treated by excision of the superior oblique tendon sheath, but in his later thesis (Brown, 1957) he advised that surgery should not be performed unless the patient had a deforming head tilt. Excision of the fibrotic superior oblique tendon sheath did not usually lead to improved function of the inferior oblique muscle.

Nutt (1954, 1955) and Nutt and Mein (1963) favoured surgery, especially if a head tilt were present, but admitted that the results were not always what one would hope for, and that, to a certain extent, slow and spontaneous improvement occurred.

Costenbader and Albert (1958) were the first to report a case showing spontaneous regression of pseudoparalysis of the inferior oblique muscle. Adler (1959) added another case. At the Strabismus Symposium of the New Orleans Academy of Ophthalmology, Brown (1962) reported that, of sixty of his own patients with the superior oblique tendon sheath syndrome, only three had outgrown it and that he had seen several adults with this condition.

The present case is the first to be reported with spontaneous recovery from a bilateral superior oblique tendon sheath syndrome, and its occurrence in only one of uniovular twins is an added feature of interest.

Case report

A girl aged 2 years 11 months (Robyn) was brought for examination because almost from birth her mother had noticed some peculiarity of eye movement, but no definite cast had been seen. The family history was negative for squint. Birth was described as normal and she was one of identical twins. Her twin sister was said to have normal eyes.

Examination The eyes were parallel in the primary position and the downward movements of both eyes were good. Upward movements showed a complete paralysis of both inferior oblique muscles.
She was re-examined at 5 years of age, when her mother reported that she always held her head slightly tilted to the right (Fig. 1A). The eyes were parallel in the primary position and when looking straight down to the reading position, but on hard depression the right eye diverged (Fig. 2). With direct elevation the right eye also diverged. On side versions the adducted eye turned downwards below the horizontal, and while adducted it could not be voluntarily elevated either binocularly or uniconically.

FIG. 1 The twins—Robyn (left) and Jennifer (right). (A) aged 5 years; (B) aged 8 years; (C) aged 18\(\frac{1}{2}\) years

FIG. 2 Robyn, aged 5 years. Bilateral superior oblique tendon sheath syndrome; divergence on elevation and depression

In the primary position, fixation was steady with each eye, but in the reading position cover testing showed alternating sursumduction. Retinoscopy under atropine cycloplegia revealed no significant refractive error (\(+2.5\) D hyperopia both eyes), the ocular media were clear, and the fundi showed no abnormality. Visual acuity with the E test gave equal vision of 6/9 in each eye.
Her twin sister, Jennifer, was examined at this time (Fig. 1A). Her eyes appeared parallel and she converged evenly, but the cover test revealed a variable amount of esotropia. No abnormal action of the inferior oblique muscles could be elicited. Fig. 3 shows her normal eye positions. She had a similar amount of latent hypermetropia to Robyn. With the E test, normal and equal vision was obtained. No treatment was given for either twin.

![Figure 3] Jennifer, aged 5 years. Normal eye movements in all positions

At 6½ years of age the examinations were repeated. By now, Robyn had a more definite head tilt to the right shoulder, a slight face rotation to the left with her chin slightly raised. Ocular movements were much the same (Fig. 4), except that the right inferior oblique muscle appeared to be acting a little, whereas the left inferior oblique appeared to have no activity at all. Divergence was still present on full elevation and depression, and cover testing produced the upward drift of alternating sursumduction. Unilateral fixation produced variable latent nystagmus with slight rotation in all positions of eye movement. The uncorrected visual acuity was 5/6 each eye, and prolonged binocular fixation appeared to be steady.

![Figure 4] Robyn, aged 6½ years. Slight improvement of right inferior oblique muscle action

Robyn was brought again at the age of 8 years and her mother reported that the deviation was becoming less noticeable. Examination (Fig. 5) showed a little activity of both inferior oblique muscles, divergence with direct elevation was slightly less, alternating sursumduction was still present. The head tilt had not improved (Fig. 1B).
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Her next visit was at the age of 18\(\frac{1}{2}\) years, when she was working as a secretary and having no eye symptoms. She had no head tilt (Fig. 1C) and eye movements appeared to be full (Fig. 6). Although she found difficulty in looking upwards there was no divergence on elevation and inferior oblique activity appeared to be normal. Her right eye still diverged with extended depression.

Orthoptic examination (conducted by Mrs. J. Craig (Miss Diana Mann)). The report was as follows:

"23 Oct. 1968, age 18\(\frac{1}{2}\) years; unaided vision: right eye 6/5; left eye 6/4; binocularly 6/4 and N.5; far cover test: alternating sursumduction with quick recovery; Worth's lights: 2 red or 3 green with position changing; near cover test: alternating sursumduction with fine right latent nystagmus and quick recovery; synoptophore: angle +10°, fusion from \(-2°\) to \(+25°\) and accurate stereopsis. Cover test showed alternating sursumduction in all positions with esophoria in elevation and exophoria in depression".

Her twin sister Jennifer was also examined (Fig. 7, overleaf). The orthoptic report was as follows:

"Unaided vision: right eye 6/4; left eye 6/4; N.5; far and near cover tests: no deviation; Maddox rod test at 6 metres: 3 eso, no height; synoptophore: angle +3°, fusion \(-2°\) to \(+30°\), normal retinal correspondence, stereopsis to maximum on Wirt circles; ocular movements: normal".
Dr. Ronald Rome kindly supplied the following obstetrical history:

"Pregnancy was normal except for a slight haemorrhage at 14 weeks which settled at home uneventfully. The twins were born prematurely at 36 weeks on 25 March, 1959. The twins were posterior positioned; the first weighed 4\(\frac{1}{2}\) lb. and the second 5\(\frac{1}{2}\) lb. There was a single placenta and the vessels on the surface of the placenta passed from one section to the other. Although this is not absolute proof that they are uniovular twins it is generally accepted that they are similar twins'.

There seems virtually no doubt that the twins were uniovular. Their likeness in general appearance, skin texture, and freckling, and the equal small refractive errors of hypermetropia are all in strong accord with the obstetrician's report.

**Discussion**

Forced duction test was not performed but the diagnosis of superior oblique tendon sheath syndrome can be substantiated by the divergence in elevation. This is regarded by Brown (1957) as a characteristic feature of the tendon sheath syndrome, whereas convergence in elevation is expected in uncomplicated paralysis of the inferior oblique muscles.

For spontaneous recovery to occur, not only must the tight superior oblique tendon sheath become more pliable, but the apparently paralysed inferior oblique muscles must develop functionally. The inferior oblique muscle weakness appears to be an important cause of disappointments after surgery when the tight superior oblique tendon sheath is cut, yet curiously, the inferior oblique muscles can apparently strengthen in late childhood.

Although the divergence in elevation disappeared, the divergence in extended depression persisted, perhaps indicating mild overaction of the superior oblique muscles. On a modified Hess screen, charting of the diplopia fields was a little difficult even at age 18\(\frac{1}{2}\) years, but a definite A phenomenon was elicited.

From the age of 3 to 6 years the head tilt increased, and by age 8 it was no better, but between then and age 18\(\frac{1}{2}\) it disappeared. This case therefore shows that, even though the head tilt may increase, this is not necessarily an indication for early surgery.

Children with weak inferior oblique muscles are at a big disadvantage while they are small, and their vertical squints are bad because they always have to look up at adults. As they grow taller their appearance seems to improve and finally, nowadays, most grown children look down at their parents so that, quite apart from actual spontaneous improvement, their appearance becomes much better, especially if they are binocular in the primary and lower positions of gaze.
Although at the age of 18½ years Robyn had lost her head tilt and her inferior oblique muscles seemed to act normally, she nevertheless had some residual weakness of binocular vision shown by alternating sursumduction on cover testing and suppression with Worth's lights. Even so, she is symptom-free in a secretarial position requiring much close work.

Two families showing possible inheritance of the superior oblique tendon sheath syndrome were reported by Gowan and Levy (1968). The syndrome is not excessively rare and this aspect appears worthy of further investigation.

Summary

Bilateral superior oblique tendon sheath syndrome presenting in only one of uniovular twins is described.

Despite apparent complete paralysis of the inferior oblique muscles, and an increasing head tilt for some years, functional and cosmetic recovery occurred without treatment.

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References

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