Sir, Surgical management of WEBINO syndrome following pineal gland lesion removal

WEBINO (walled-eyes bilateral internuclear ophthalmoplegia) is a rare clinical syndrome of exotropia with bilateral INO associated with the loss of fusional vergence. It is usually caused by demyelination or ischaemic damage to the medial longitudinal fasciculus and medial rectus subnucleus of the oculomotor nerve,¹ hence spontaneous improvement is common. We report our experience in a persistent case following pineal tumour removal.

Case report

A 13-year-old boy was admitted for persistent headache and double vision for 6 weeks. His ocular findings were BCVA 6/5 each eye; bilateral papilloedema; minimal esophoria for near, esotropia for distance, and good stereopsis (see Table 1a). MRI scan showed a cystic

tumour arising from the pineal gland, obliterating the aqueduct of Sylvius (Figure 1). He underwent excision of the lesion (histopathology showed papillary tumour) and received supplemental radiotherapy. He was referred back to us for persistent diplopia. He still had BCVA of 6/5 in each eye, but now showed marked alternating exotropia of $>70^\Delta$ dioptres. There was limitation of adduction and variable jerk nystagmus of the abducting eye on horizontal saccades (bilateral INO). Downgaze was completely absent and upbeat nystagmus was seen in upgaze. Other orthoptic findings are detailed in Table 1b. The left pupil was semi dilated and nonreactive to light or accommodation. Hess chart showed no adduction of either eye past the midline (Figures 2a and c). His condition failed to resolve over 2 years. As he was unsuitable for adjustable suture, and to optimise the cosmetic and functional outcome a two-staged squint surgery was decided. Initially he underwent bilateral 12 mm lateral rectus muscle recession, followed by bilateral medial rectus muscle resection of 6 mm. Good ocular alignment was achieved post operatively and his diplopia was relieved (Figure 2b). Table 1c shows orthoptic findings 6 months following the surgery.

Table 1 Timeline of orthoptic findings

- A CT: minimum esophoria for near, slips to Esotropia for distance.
Motility: Normal.
4 Δ test: Bifoveal.
Frisby: 170.
Pupils: normal reaction to light and accommodation.
Convergence: to 8 cm.

Hess chart at presentation

- B CT: Marked XT $\sim 70^\Delta$ for near and distance.
Motility: Mild under action of adduction and abduction with abducting nystagmus on lateral gaze; marked limitation depression and upbeat nystagmus on elevation.
Pupils: normal right and partially dilated non-reactive left pupil, no Marcus Gunn. Convergence: Not possible.

Ocular motility diagram following pineal gland lesion removal.

- C CT: N-slight XT no diplopia, D- slight Exophoria with fair recovery
PCT: N: 25 $^\Delta$ BI, D: 10 $^\Delta$ BI.
Convergence: 20 cm. PFR: N: not possible, D: 12 $^\Delta$ BO $\rightarrow$ 2 $^\Delta$ BI
Worth 4-dot: N: diplopia response, D: Fusion response.
Frisby: -ve.

Ocular motility diagram following squint correction.

This is the second case reported in the literature of WEBINO onset following surgical intervention.² Our case is unique in having combined features of WEBINO, impaired vertical gaze, loss of accommodation, convergence, and some pupillary involvement. This condition should be differentiated from pontine exotropia in which the vertical gaze remains intact.³ It should also be differentiated from bilateral nuclear third nerve palsy, where variable degree of medial recti paresis, and bilateral ptosis are usually present, and the angle of divergence rarely exceeds 50^A dioptres.⁴

Surgical intervention is an option to achieve binocularity in WEBINO syndrome if spontaneous recovery is unlikely. Demonstrable fusion response was reported by Roper–Hall *et al*⁵ in seven out of eight patients, post operatively. In contrast to our case they achieved their results through two stages of unilateral recess–resect procedures. As the angle of squint in WEBINO is very large four-muscle operation is necessary

Figure 1 MRI head scan showing 2.5 cm lesion in the pineal gland area obstructing the Sylvian aqueduct.

and adjustable suture is advisable. In the absence of convergence, overcorrection is advisable as latent divergence could persist (shown in Hess chart Figure 2d).

Conflict of interest

The authors declare no conflict of interest.

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A Toufeeq and D Dave

Department of Eye, Chesterfield Royal Hospital NHS
Foundation Trust, Chesterfield, UK
E-mail: abbastoufeeq@yahoo.com

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Figure 2 Patient photographs showing 9-gaze positions and corresponding Hess charts before (a, c) and after corrective squint repair (b, d).