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A case of benign intracranial hypertension

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Abstract

Aim: **To describe a patient with longstanding fully accommodative esotropia who developed benign intracranial hypertension.**
Method: **A case is documented with history, orthoptic, ophthalmological and neurological findings. The diagnosis of benign intracranial hypertension is discussed in relation to the child's symptoms and when further investigation is required.**
***Results*:** **An unexplained temporary decompensation of a previously well-controlled deviation followed by several episodes of blurred vision and occasional headache 6 months later prompted the orthoptists to press for further investigations. Subsequent neurological investigation led to a diagnosis of benign intracranial hypertension.**
Conclusion: **The orthoptic team were responsible for the long-term management of this child. The onset of symptoms that could not be easily accounted for were the key elements in the orthoptists' request for further investigation.**

Key words: Benign intracranial hypertension, Blurred vision, Decompensation, Visual outcome

Introduction

Benign intracranial hypertension (BIH) is characterised by raised intracranial pressure (ICP) and normal cerebrospinal fluid (CSF) composition in the absence of an intracranial mass or ventricular dilation.[1,2] It is a rare condition in children,[2] particularly in the prepubescent child.[3]

Reports suggest that the features of BIH in adults and children are similar.[2–4] Signs and symptoms include headache, nausea, vomiting, visual loss and VI nerve palsy. Tinnitus, numbness and tingling of hands and feet, arthralgias, low back pain and difficulty in walking have also been reported.[1] BIH occurs more frequently in females and is associated with obesity. However, in prepubertal cases there is no sex predilection and obesity is not common.[3] Cinciripini *et al.*[3] reported on 10 cases of BIH in prepubertal children aged 11 years and younger in whom the main presenting signs were diplopia and strabismus, which were frequently associated with VI nerve palsy and stiff neck.

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A case is described of a 7¼-year-old girl with fully accommodative esotropia who developed BIH. The features of the case are considered in relation to the diagnosis of BIH and when an orthoptist should decide that further investigations are required.

Case report

The child had been attending the eye clinic since the age of 2½ years with fully accommodative esotropia. At the age of 6 years, the child was wearing right $+4.50/-0.75 \times 180$, left $+4.25/-0.25 \times 180$. Corrected vision with the Keeler Crowded logMAR test was right 0.100 (6/7.5), left 0.05 (6/6pt). Cover test showed a well-controlled esophoria of 12^{Δ} at ⅓ m and 6^{Δ} at 6 m. Stereoacuity was 55" using the Frisby stereo-test. AC/A was normal. Without correction there was a small right esotropia with no appreciation of diplopia or subjective awareness of the deviation. Attempts at assessing the fusion range and binocular visual acuity (BVA) had been inconclusive due to lack of patient comprehension.

At the age of almost 6½ years the child suddenly complained of horizontal diplopia. With correction logMAR vision was 0.00 (6/6) right and left, and cover test at ⅓ m and 6 m showed a slight esophoria becoming right/alternating esotropia with diplopia. Abduction of each eye was full. With correction the deviation measured 14^{Δ} E(T) at ⅓ m and 6^{Δ} E(T) at 6 m. There was no associated history of illness or trauma and no other parental concerns.

Cycloplegic refraction revealed an increase of +0.25 DS for the right eye. Examinations of the fundi and media were normal. The new prescription was given. One month later, with correction, the child showed good control of the esophoria, which measured 14^{Δ} at ⅓ m and 6^{Δ} at 6 m, and BVA to 6/6 at ⅓ m and 6 m.

Six months later

Whilst attending for orthoptic review 6 months later the mother mentioned that the child had complained of several episodes of transient blurred vision and occasional headache, which could occur at any time. Despite thorough questioning, the mother was vague about the incidence, severity and frequency of the episodes. The orthoptic findings were unchanged, with good control of the esophoria with glasses. In view of the previous temporary decompensation of the esophoria and the development of vague new symptoms the orthoptist arranged an appointment with the ophthalmic consultant.

At this appointment 2 months later, corrected vision was still logMAR 0.00 right and left. Examination of the

fundi revealed a swollen appearance of both discs. There were no haemorrhages or drusen. Goldmann visual field assessment was normal. The ophthalmic diagnosis was swollen optic discs 'probably secondary to hypermetropia' and the child was scheduled to have an ophthalmic review in 3 months.

At routine orthoptic review 3 weeks later corrected acuity had reduced to right 0.250 (6/9.5pt), left 0.125 (6/7.5pt). The control of the esophoria and measurement were unchanged. The mother mentioned two episodes of the child being distressed by transient blurred vision and pain in the eyes. The orthoptist was very concerned and arranged an urgent ophthalmological opinion. At this appointment the acuity was still reduced with no change in orthoptic status. Goldmann visual field assessment, pupil responses and colour vision using the Ishihara test were normal. The disc appearance was unchanged. Due to great orthoptic concern the consultant agreed to refer the child to a paediatrician for further investigation.

Paediatric assessment

At the paediatric assessment there were no problems with gait, coordination or paraesthesia, and no neurological defect other than swollen discs. The child had been seen previously by the paediatric team when aged 5¼ years with a history of headache, which was generally relieved with paracetamol. At that time the child had been reviewed over a 6 month period during which the headaches were considered to be non-specific, and as they decreased in frequency no further investigations were undertaken.

Given the history and presence of swollen discs further investigations were now undertaken. Magnetic resonance imaging (MRI) findings were normal. In view of this a lumbar puncture was arranged. Opening pressure was 23 cm H_2O,* but due to technical difficulties with the equipment it was suspected the true value was higher. The pressure was reduced to 15 cm H_2O. Samples of CSF were sent for biochemical analysis and culture and the results were normal.

Immediately after the lumbar puncture the child reported that she felt 'much better' and has been asymptomatic since the lumbar puncture 2 months ago. The current paediatric working diagnosis is BIH. The esophoria is well controlled with glasses and currently measures 12^{Δ} at ⅓ m and 6^{Δ} at 6 m, and BVA 6/6 at ⅓ m and 6 m. The disc appearance and visual acuity remain unchanged. Goldmann visual field assessment is normal.

Discussion

Following 4 years under orthoptic care the child was diagnosed as having BIH. Referral for further investigation and the subsequent diagnosis were the result of orthoptic concern over the child's symptoms. Diagnosis of BIH is one of exclusion based on the clinical symptoms and the neurological, ophthalmological, radiological and CSF findings. Prompt diagnosis of BIH and appropriate management are important to prevent loss of visual acuity and visual field.

When confronted with parental/child reports of signs and symptoms that could be accounted for by difficulties controlling a longstanding strabismus, at what point should an orthoptist become concerned that there is 'something' more serious that warrants further investigation? What were the signs that this child's symptoms could not be assumed to be due to control of the strabismus?

It is uncertain in this case what the primary presenting sign of the intracranial hypertension was. In children the reported presenting symptoms are blurred vision, headaches, diplopia due to nerve palsy and stiff neck.[3–5] In a retrospective study of paediatric cases headache was reported to be the most common symptom,[6] and headaches have been reported to be predominantly frontal.[2] The child had experienced headaches when aged 5¼ years and had been evaluated by the paediatric team. This highlights the importance of an orthoptist routinely inquiring about a child's health and any ongoing medical investigations not associated with the child's eye condition. The mother first reported occasional headaches to the orthoptist at the time she reported the blurred vision, when the child was 7¼ years old. At this time the mother considered the two to be associated.

The reason for the decompensation of the esophoria when the child was 6½ years old remains uncertain. The deviation had always been well controlled on cover test. The increase in the deviation was only 2^{Δ} at near and it seems unlikely that poor fusion reserves would account for the decompensation. Subsequent assessment has shown good fusion reserves. Ocular motility was normal with no suspicion of VI nerve palsy. Whilst there was a +0.25 DS change in prescription for the right eye, it is doubtful that this change was sufficient to restore binocularity given that the child's AC/A was normal. Any variation in the intracranial pressure at this time may have affected the general health of the child sufficiently to temporarily disrupt her control of the esophoria, even though the child was reported by her mother to be well.

At age 7¼ years the child complained of episodes of transient blurred vision and occasional headache. A variety of visual disturbances have been reported in children and adults with BIH, including transient visual loss/blurred vision.[2,7] These 'transient visual obscurations' are considered to be unilateral or bilateral episodes of visual loss, which usually last less than 1 minute and are followed by full 'recovery'. These episodes are thought to be due to transient ischaemia of the optic nerve. The orthoptist was concerned because the temporary loss of control of the esophoria could not be accounted for and, given the onset of transient blurred vision, requested an ophthalmological opinion.

The ophthalmic examination revealed bilateral swollen discs but did not cause ophthalmic concern. The child was very cooperative with Goldmann perimetry and the result was normal and considered to be reliable. The incidence of visual field loss is reported to be high in adult and child cases of BIH. Loss may be mild or marked and is most commonly an enlargement of the blind spot.[2,7,8] Three weeks later the mother described the child as being distressed by two episodes of blurred

* Most reviews on BIH in children consider 20 cm H_2O as the upper limit of normal.

vision and headache and visual acuity was reduced on testing. When the child was seen by the ophthalmologist at the urgent request of the orthoptist the ophthalmic findings were unchanged. The orthoptists expressed great concern that there was an underlying neurological defect and pressed for further investigation.

Loss of visual function is considered to be the only serious permanent implication of BIH and management of the condition is directed at the relief of symptoms and preservation of vision. Visual field loss or reduced visual acuity has been reported in 13–27%[2] and 13–38%[3] of children at presentation or during the course of the disorder. Rush[9] reported that the visual outcome is not related to the duration of the symptoms, the degree of papilloedema, the presence of visual obscurations or the incidence of recurrent increased ICP.

The child reported feeling much better immediately following the lumbar puncture and 2 months later she continues to feel 'well'. Such resolution of symptoms on lowering of the ICP by lumbar puncture has been reported for 13 of 38 children[4] and 4 of 22 children.[2] This has also been reported in adults and suggests that symptoms are directly caused by the increased ICP.[1]

Treatment regimes for BIH have been described[2,4] and it is proposed that treatment is considered on an individual basis.[2] Patients should have regular ophthalmic surveillance from the time of diagnosis to monitor visual acuity and visual fields. Wall and George[7] considered visual field testing to be the most sensitive indicator of 'incipient' vision loss. The length of follow-up is undetermined, as the natural history of the condition and the risk factors for visual outcome are not known.[2] To date this child's orthoptic and ophthalmic status are unchanged and regular review is planned.

The orthoptic team were responsible for the long-term management of this child. The onsets of symptoms that could not be easily accounted for, with the subsequent ophthalmic finding of swollen optic discs, were the reasons why the orthoptists pressed for urgent investigation.

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There are no competing interests.

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Acute-onset esotropia (horizontal skew deviation) following fasting for Ramadan, whilst neglecting to adjust epilepsy medication accordingly: a case report

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Aim: To report a case of acute-onset esotropia (horizontal skew deviation) that was probably the result of the toxic effects of two epilepsy-mediating drugs, sodium valproate (epilim) and lamotrogine (lamictal). Sodium valproate and lamotrogine are both anticonvulsants whose mode of action is to reduce electrical discharges within the brain. Acute-onset esotropia is a relatively rare neurological disorder in which a concomitant horizontal deviation occurs as a result of lesions within the posterior fossa. This results in a disturbance of the otolithic input pathways to the horizontal vergence centre.

Methods: Details are reported of an 18-year-old epileptic taking sodium valproate and lamotrogine who suffered acute-onset esotropia within 2 weeks of commencing fasting for Ramadan. Orthoptic, ophthalmological and radiological findings are presented and considered in relation to the patient's medication and fasting at the time of onset.

Results: The patient presented with a sudden-onset esotropia with diplopia. At this time the patient was in the process of fasting for Ramadan. Orthoptic assessment revealed a concomitant esotropia with no limitation of ocular motility. Following full orthoptic, ophthalmological and radiological investigations the patient was diagnosed with acute-onset esotropia (horizontal skew deviation). The horizontal skew deviation resolved completely within 6 weeks of ceasing fasting for Ramadan. The patient regained binocular single vision and there was no need for further management.

Conclusion: High doses of epilepsy medication may cause significant brainstem damage, and in this case are the most probable cause of the acute-onset esotropia. Sodium valproate and lamotrogine are known to have significant dose-related side-effects, including diplopia. The dosage of these drugs is calculated on an individual basis and is dependent on age and weight. The patient should be advised to take anticonvulsants after food. This advice was not adhered to whilst fasting for Ramadan. A case of acute-onset esotropia has also been reported in a young boy after taking carbamazepine, another anticonvulsant.[1] Patients and clinicians should consider adjusting the dose of highly toxic drugs such as anticonvulsants prior to patients undertaking a significant change in diet.

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Familial horizontal gaze palsy and skew deviation highlighting the need for full ocular motility assessment

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Aim: To report a case of familial bilateral horizontal gaze palsy and skew deviation. A literature review did not reveal any articles associating these two conditions.

Methods: Details are reported of two female siblings. Orthoptic and ophthalmological findings are presented.

Results: Orthoptic assessment revealed bilateral horizontal gaze palsy, skew deviation, global developmental delay and a similar physical appearance. As a result of the gaze palsy, both infants utilised head movements to follow targets and view objects in their peripheral field. This was misinterpreted as head thrusting and consequently a diagnosis of ocular motor apraxia (OMA) was made. By the age of 4 years more accurate assessment of the sisters was possible and the diagnosis was revised. Both siblings were referred to Great Ormond Street Hospital, London. Following extensive genetic testing, their condition was deemed to be autosomal recessive.

Conclusion: Bilateral horizontal gaze palsies result in complete loss of both horizontal smooth pursuits and saccades. Conversely, OMA is a form of saccadic paralysis whereby the patient is unable to execute voluntary saccades but their smooth pursuits are often unaffected. These two patients highlight the difficulties associated with testing young children and how an accurate differential diagnosis can be hindered by these limitations. In addition they reinforce the need for thorough investigations to be undertaken, which should include an assessment of all ocular movement systems wherever possible, in order for an accurate diagnosis to be obtained.