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Transient blindness following intracranial pressure changes in a hydrocephalic child with a V–P shunt

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Abstract. A hydrocephalic child with a V-P shunt developed transient blindness following shunt revision. A year later, visual function deteriorated when shunt malfunction occurred. Following shunt revision, the child regained sight. The effects of intracranial *hypertension* and *hypotension* on the visual pathways are discussed.

Key words: Ventriculoperitoneal shunt – Shunt complication – Hydrocephalus – Blindness – Visual-evoked potential – Intracranial pressure.

It is well known that increased intracranial pressure (ICP) in patients with hydrocephalus can cause a variety of visual disturbances. Visual field defects [21] and transient or persistent blindness have been reported in shunt malfunction [2, 6, 17, 18, 22–24, 26, 42, 48, 50].

There are no data on the effect of *decreased* ICP on the visual pathways. In the case we present, intracranial hypotension following shunt revision, as well as intracranial hypertension because of shunt malfunction, caused transient blindness.

Case report

A 3.5-year-old boy was admitted to the Hadassah University Hospital with a ventriculoperitoneal (VP) shunt malfunction. He was born prematurely at 30 weeks' gestation by caesarian section. He suffered from respiratory distress and was operated on to correct patent ductus arteriosus.

Communicating hydrocephalus gradually developed and at the age of 2 months, a right ventriculoperitoneal shunt was performed (Pudenz, medium-pressure flushing valve, low-pressure peritoneal catheter).

At the age of 1 year the child underwent shunt revision and the peritoneal catheter was found to be blocked. At the age of 2.5 years he developed nausea, vomiting, ataxia and somnolence. According to the mother, there had been no change in the child's behavior in regard to visual function. The ophthalmic examination revealed that the child followed objects and light, there was a normal pupillary reaction, and fundoscopy showed a normal posterior pole. CT scan demonstrated an enlarged ventricular system. Once again, the peritoneal catheter was changed because of a distal block. Following surgery, there was an obvious improvement in the child's alertness, steadiness, and appetite. However, an obvious decrease in his visual ability was noted: the child did not follow a flashlight and did not reach for objects. Pupils reacted to light and fundoscopy revealed no change from the previous check-up. Flash visual-evoked potential (VEP) (Fig. 1) showed a bilateral nonspecific sinusoidal response interpreted as a nonfunctioning response. In view of the normal pupillary reaction, this electrophysiological finding was compatible with damage to the posterior visual pathways. Over a period of a few days, the child gradually recovered and regained sight.

One year later, at the age of 3.5 years, he again had the symptoms of somnolence, lethargy, and vomiting. The mother noticed gradual deterioration in his visual function: the child did not follow objects nor did he reach for toys. On examination, he was lethargic. Pupils were slightly dilated with a sluggish response to light. Fundoscopic examination revealed pallor of the optic disk. Flash VEP response (Fig. 2a) was extinct bilaterally. Electroretinogram (ERG) showed normal retinal activity. The child underwent a distal revision for the third time. Two-to-three days later there was an obvious improvement in visual function. He was able to recognize his parents and play with his toys. Repeated VEP (Fig. 2b) showed a remarkable change. There was normal response in the left eye and a very poor response in the right.

Discussion

The real incidence of visual loss due to shunt malfunction is not known. Several series and case reports are available [2, 6, 17, 18, 22–24, 26, 42, 48, 50]. In some large series of children with hydrocephalus, loss of sight due to shunt failure has not been mentioned [12, 25, 39, 43].

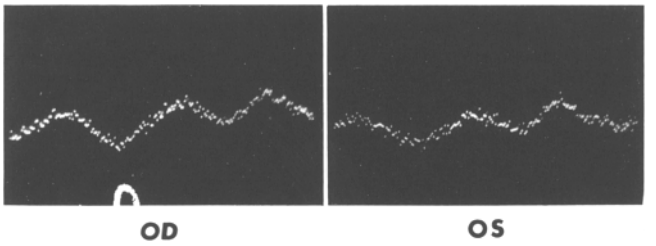

Fig. 1. Flash VEP after the second revision of the V-P shunt shows a bilateral nonspecific sinusoidal response. *OD,* Right eye; *OS,* left eye

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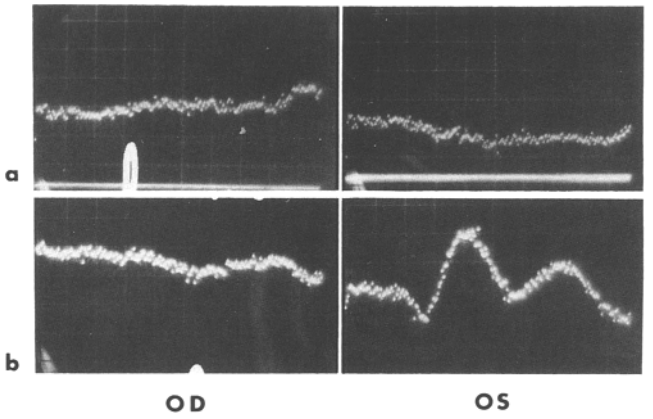

Fig. 2. **a** Flash VEP before the third shunt revision; **b** flash VEP 4 days after shunt revision. *OD,* Right eye; *OS,* left eye

Various mechanisms have been suggested to explain loss of vision in shunt failure and increased ICP. The lesion is usually classified on a clinical basis as pregeniculate (anterior visual pathways) if optic atrophy or decreased light reflex exist, or postgeniculate (cortical blindness) if these findings are absent. It should, however, be remembered that optic atrophy can result from trans-synaptic degeneration after posterior lesions [20, 28, 46]. Thus, anatomical localization is not always possible on the basis of physical examination alone.

Ischemia of the optic nerve head can follow papilledema [18]. The anterior optic tracts are supplied by a network of small arteries that lie over bone. Pressure on these arteries can produce circulatory stasis and ischemia [2, 48].

Lesions to the lateral geniculate body (LGB) from downward herniation of the parahippocampal gyrus in the tentorial notch can occur [18]. Disturbances in cerebral circulation can happen secondary to compression of the posterior cerebral artery or one of its branches along the tentorial notch [8, 19, 23, 30, 44]. This mechanism is more likely to occur with sudden rise in ICP, whereas with slow rise collateral circulation is usually established.

Suggested mechanisms specific for hydrocephalus stress the close anatomical relationship between the ventricular system and the visual pathways. Engel [7] hypothesized that ventricular dilatation may stretch projection fibers of the optic tracts and change various electrophysiological parameters. There is evidence of periventricular edema, followed by gliosis and damage to white matter fibers in acute hydrocephalus [49].

Cushing and Walker were among the first to realise the importance of the dilated III ventricle acting as a mechanical compressing and distorting force on the visual pathways [5]. An enlarged III ventricle can bulge the infundibulum downwards, pressing the posterior angle of the chiasm [4, 21, 47, 48]. However, there is no direct positive correlation between increase in ventricular size and abnormalities in VEP [51]. Some authors report that persistent ventriculomegaly after shunting was compatible with normal VEP [14, 51].

VEP serves as an important tool for investigation and monitoring of visual disturbances in hydrocephalus [1, 7, 11, 14–16, 27, 33, 34, 41, 51]. Latency delays, fatigability, and asymmetries occur. The best correlation was found between increased ICP and latency delays of N2 [51] and P2 [14] components of the VEP.

The case we present demonstrates that the visual pathways are sensitive not only to intracranial hypertension but also to a sudden decrease in pressure. To our knowledge, this kind of shunt revision complication has not previously been reported [13, 32, 38, 39]. Intracranial hypotension may develop after trauma with cerebrospinal fluid (CSF) rhinorrhea or otorrhea. It can be fatal when iatrogenically caused by an external drainage system [45].

One must differentiate between the syndrome of acute decompression in marked hydrocephalus and the syndrome of chronic slit ventricles. In the syndrome of collapsed or slit ventricles, caused by long-standing excessive CSF drainage through a shunt system, loss of appetite, lethargy, headache, nausea and vomiting occur [9, 29, 35, 36]. Signs of acute intracranial hypertension occur upon shunt obstruction due to the collapsed ventricles [36]. These children who have undergone excessive drainage can also develop changes in cranial volume and shape (microcephaly, scaphocephaly) and subdural hematomas [29, 31, 37]. It has been shown [10] that these patients are intolerant of minimal pressure elevations, even in the normal range, probably the result of evacuation of a spatial buffer [10]. In the acute decompression syndrome, severe neurological symptoms may occur [10]. It has been speculated that it is a result of upper shifting of the brain stem. Sometimes symptoms are easily reversible by placing the patient in a prone position with the head lower than the body [11]. Currently there are no experimental data on the effect of intracranial *hypotension* on visual function [3, 40].

In the case we present, the change in pressure was acute and probably happened during the operation itself. In this case, we do not know what the exact mechanism was of the reversible damage to the visual pathways. It seems likely that after the second revision, when transient blindness occurred, it was due to damage to the posterior visual pathways since pupillary function was preserved. At the third revision, optic atrophy had already developed. This atrophy was most probably due to sustained increased intracranial pressure. However, posterior damage with trans-synaptic degeneration cannot be ruled out [46]. As no other neurological symptoms were present, it is unlikely that brain-stem movement was the cause of the deterioration of visual activity.

In any case it is advised, when inserting a new shunt or when revising one, that uncontrolled loss of the CSF be avoided and thus acute change in pressure.

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