

ORIGINAL PAPER

Slit ventricle syndrome: clinical and diagnostic pitfalls

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ABSTRACT

Slit ventricle syndrome (SVS) consists of clinical symptoms of intracranial hypertensive syndrome (IHS), which include severe, usually intermittent headaches; vomiting and possibly some degree of decreased consciousness; and impairment in hydrocephalic children with an apparently working ventriculoperitoneal shunt (VPS), without ventricular enlargement on computed tomography (CT) or magnetic resonance imaging (MRI). Signs of IHS include increasing head circumference and papilledema. The syndrome has been typically observed in children with neonatal or infant hydrocephalus, three to six years after VPS implantation. Therapeutic decisions are difficult and often depend only on the clinical presentation of IHS. There is much controversy about the treatment of SVS. Immediate shunt revision may pose a problem in putting the shunt system in the slit ventricles or removing the old shunt, which can be attached to the ependyma.

KEY WORDS:

hydrocephalus, slit ventricle syndrome, children, intracranial hypertension, ventriculoperitoneal shunt, radiological imaging.

INTRODUCTION

Considering that the majority of children with a ventricular parietal shunt (VPS) can have mild headaches and in some cases their ventricles are smaller than normal on routine imaging studies, there are still controversies about algorithms for the treatment of patients with clinical intracranial hypertension symptoms (IHS), without any or with very slight evidence in radiological findings [1, 2].

Slit-like ventricles in CT or MRI in a shunted patient can be observed in different clinical conditions, including SVS, overshunting syndrome, and normal volume hydrocephalus, or it can be misinterpreted as a properly working shunt. Numerous opinions and recommendations in formulating an evidence-based algorithm can be found in the

literature. Subsequent classifications were based on VPS efficiency and discriminate between patients with functioning, intermittently functioning, or malfunctioning shunts [3-5]. Several authors divided patients with SVS into those having low or high intracranial pressure (ICP). Based on ICP and headache, ReKate *et al.* (1993) [4, 6-8] established a new SVS classification based on history, symptoms, and ICP monitoring results, to facilitate individual treatment.

Type 1 is caused by cerebrospinal fluid (CSF) overdrainage and is associated with low pressure. Low ICP symptoms progress during the day with the headache improving after resuming the recumbent position. It has been discussed that shunt-induced suture ossification described in chronic overdrainage of CSF via the shunt may cause slit ventricle syndrome [9].

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Type 2 is caused by intermittent proximal obstruction of a ventricle catheter. Episodic high-pressure symptoms are the mainstay, sometimes associated with activity.

Type 3 is caused by shunt failure (normal volume hydrocephalus, NVH, or shunt pseudotumour), and are associated with shunt blockage and elevated CSF pressure. The patient may have morning headaches unrelieved by analgesics or visual changes similar to the cephalocranial disproportion symptoms above.

Type 4 (hydrocephalic, BIH) is a cephalocranial disproportion that increases brain parenchymal pressure but not CSF pressure and has been attributed to shunted children with craniofacial syndromes. Headaches, vomiting, and papilledema can lead to vision loss if elevated ICP is left untreated.

Type 5 is a headache unrelated to shunt function. This group of patients may have a family history of migraines, episodic headache, or headaches relieved by rest.

PATHOPHYSIOLOGY OF SVS

There are some theories related to the pathogenesis of SVS, and it is likely that the true pathophysiology of SVS involves more than one mechanism. The first is that maximised brain growth at five or six years of age may explain the reduction of subarachnoid CSF, which can buffer the increases in ICP during times of catheter obstruction (this may explain the age range during which slit ventricle syndrome occurs). Ventricular pressure is intimately related to ICP, and when CSF pressure drops uncoupling occurs, e.g. increased venous congestion and increased brain elastance. An inability for the spinal subarachnoid space to aid in buffering high ICP (due to a smaller than normal spinal canal) may also play a role [10-12].

The second is that increased pressure with the long-standing presence of a ventricular catheter may cause sub-ependymal and periventricular gliosis, which contribute to the inability of the ventricles to dilate, increased ventricular wall stiffness, and ventricular compliance. Gliosis, however, has been found on autopsy in individuals with and without small ventricles [13, 14].

The third proposed mechanism is that of temporary obstruction of the drainage system by the ependyma (which lines the ventricles and represents an interface between the CSF and the brain, and functions as a one-way membrane that allows the free flow of CSF into the ventricle), which leads to increased ventricular pressure, enlargement, and restoration of the VPS. If the membrane function of the ependymal surface is bypassed, the CSF may be forced back into the brain ECF instead of leading to the dilatation of the ventricular system. If the ventricles are drained artificially, the distending hydrostatic force of the intraventricular CSF is lost, and the ventricles become smaller than normal or collapse. Imaging studies may misinterpret this state of stable ventricular size as the absence of shunt malfunction [15-17]. Another pathophysiological

concept of SVS is that of shunt-related intracranial hypertension (IIH) due to increased sagittal sinus venous pressure, which reduces CSF absorption capacity [18].

THERAPEUTIC DECISION

Shunt malfunctions are manifested clinically by symptoms of IHS. However, shunted hydrocephalic patients may become symptomatic from shunt failure without evidence of ventricular enlargement on CT or MRI [4, 6]. One of the possibilities of intracranial hypertension syndrome without any neuroradiological findings is called slit ventricle syndrome (Fig. 1).

The patients at risk of developing slit ventricle syndrome are those who were shunted at infancy, and when a low drainage pressure shunt system was used. This does not imply inadequate treatment but means that the effect of a shunt can be unpredictable.

ICP monitoring is a good starting point in evaluating a child with small ventricles, headaches, and no evidence of a shunt malfunction because it can differentiate high from low ICP [19]. The crucial point based on extra imaging findings and ICP monitoring is to make sure that the shunt is working properly with no blockage due to malfunction of the system.

In cases of satisfactory shunt function, there are several available options. Prevention with initial placement of a programmable valve or a valve with an anti-siphon chamber has been advocated [20]. Changing to a high-pressure valve or adding an anti-siphon device at a later stage of disease can sometimes cause a fatal rise in intracranial pressure, particularly in those patients who have had a shunt or who have suffered from slit ventricle syndrome for a long time. The surgical treatment of SVS has been subdivided into surgery aimed at the restoration of the CSF circulation or correction of impaired CSF absorption, like ventriculostomy, or aimed at increasing craniocerebral compliance, like subtemporal craniotomy or calvarial expansion [21].

There is a general rule in the treatment of SVS that the first step should be to assess in which of the above-mentioned categories/types the patient belongs. Otherwise, empirical treatment, e.g. like in intracranial hypotension, is recommended. If ocular hypertension symptoms occur with double vision or visual loss or even a decline in visual acuity, operative treatment should be considered as fast as possible because the symptoms may be irreversible.

DISCUSSION

Bruce *et al.* found that 64% of children with shunts developed slit ventricles, but only 6.5% of these patients required surgery [3]. On the other hand, the clinical syndrome of debilitating headaches and small, unchanged ventricles complicated 1-37% of shunt procedures [22].

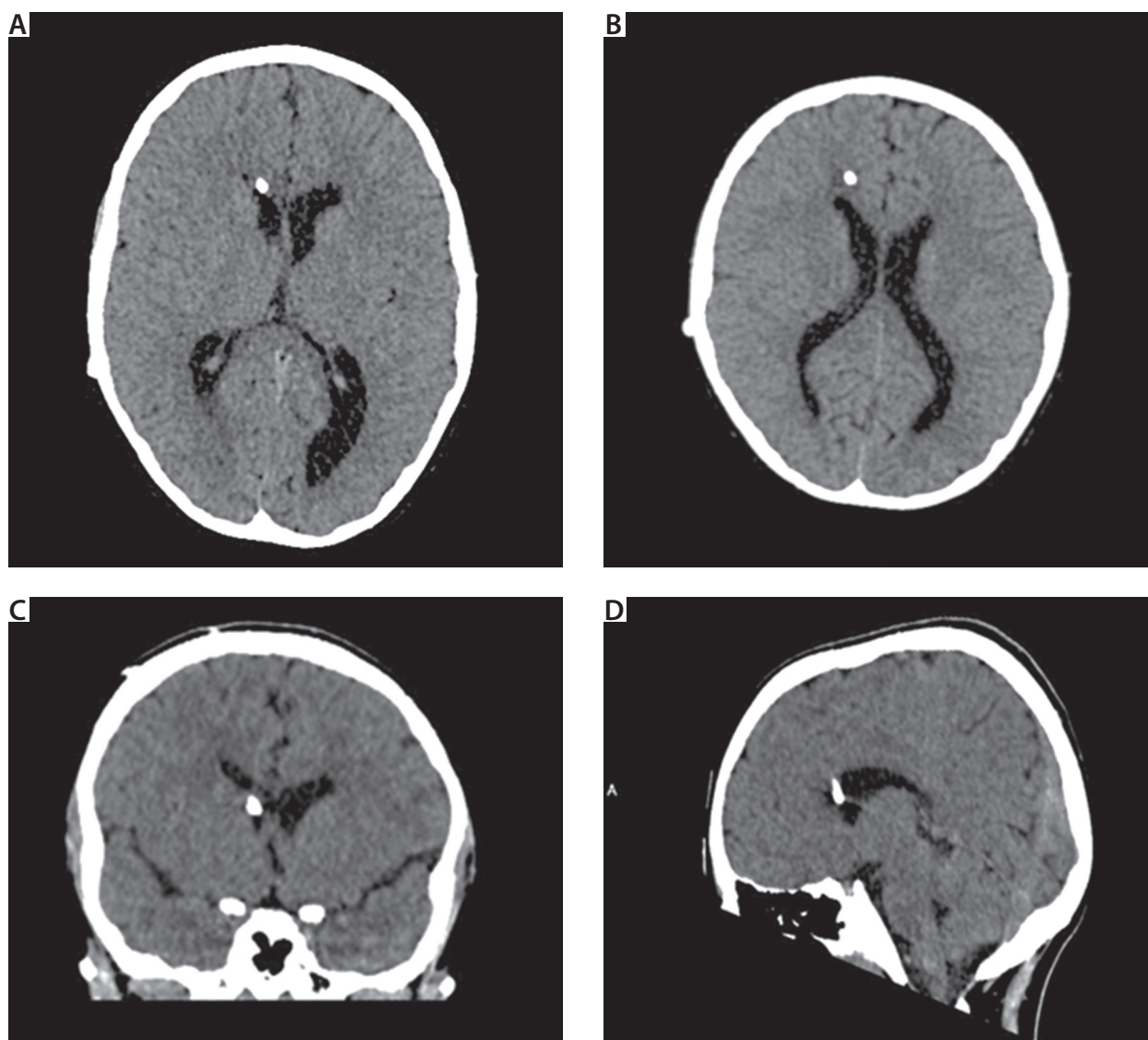


FIGURE 1. Slit ventricles – CT. Proper shunt position

The optimal therapeutic strategy for slit ventricle syndrome remains controversial. The goal of surgical intervention for this syndrome is the resolution of symptoms. Secondary benefit is obtained if shunt independence is achieved. Conservative management of SVS symptoms via excessive intravenous hydration and reclined head position was demonstrated to be successful in the early stages. Short-term steroids have been reported to be useful in transiently improving symptomatic complaints and delaying the need for surgery [23, 24].

The role of endoscopy in slit ventricle syndrome is controversial, such as endoscopic third ventriculostomy (ETV), which has been proposed to treat slit ventricle syndrome in patients shunted for hydrocephalus due to aqueductal stenosis with shunt failure. ETV, compared with shunt revision, has substantially greater longevity and no reliance on the implanted foreign body. The important condition is that slit ventricle syndrome does not preclude endoscopic treatment, but the ventricles must be large enough to allow therapeutic access [25].

In symptomatic elevated ICP (acute or semi-acute episodes of headache, severe papilledema, nausea, vomiting associated with varying degrees of impairment of consciousness, or lethargy) emergent shunt revision or ETV is required.

The authors would like to point out the significance of ocular hypertension symptoms (headaches, papilledema, and a decline in visual acuity: double vision, vision loss, temporary episodes of blindness) in making the therapeutic decision about hydrocephalic patients with shunts. Brain imaging, such as CT or MRI scans, performed to look for a brain tumour, injury, or other potential cause of symptoms usually show normal findings, but lumbar puncture demonstrates raised CSF pressure. The decision to perform urgent intervention depends on the clinical condition of the patient with IHS. Close monitoring of vital signs, checking for bradycardia, hypertension, or respiratory compromise is essential (Cushing's triad). Patients with normal ICP should undergo shunt removal without ETV. The shunt can be removed without the need of revi-

sion or ETV if the patient does not complain of headaches after 2-3 days of shunt exteriorisation and blockage when an ICP monitoring facility is not available [3, 18, 26, 27].

Baskin *et al.* [28] reintroduced the concept of using neuroendoscopic techniques and proposed a treatment algorithm for all patients presenting with slit ventricle syndrome. Kulkarni *et al.* [29] reported a higher risk of initial failure in ETV than shunt in children, which progressively decreased about three months after the procedure. Patients can experience a long-term treatment survival advantage after an early high-risk period of ETV failure in comparison with shunt removal.

CONCLUSIONS

There are still many important questions concerning algorithms and clinical scenarios in the context of children with clinical intracranial hypertension symptoms. Therapeutic decisions in symptomatic patients with IHS are based mainly on the clinical condition.

DISCLOSURE

The authors declare no conflict of interest.

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