Slit ventricle syndrome

see also Slit ventricle.

Slit ventricle may be asymptomatic.Slit ventricles may be seen on CT in 3-80 % of patients after shunting [1) 2)] ,most are asymptomatic.

These patients may occasionally present with symtoms unrelated to the shunt e.g true migraine

see Slit ventricle syndrome.

Intracranial hypotension: symptoms often relieved by recumbency.

Subtypes

(1) Intermittent shunt occlusion: Overshunting leads to slit ventricles which causes the ependimal lining to occlude the inlet ports of the ventricular catheter. producing shunt obstruction.

With time , many develop low ventricular compliance [3)] , where even minimal dilatation results in high pressure which produces symptoms. Expansion then eventually reopens the inlet ports allowing resuption of drainage.

Symptoms may resemble shunt malfunction: intermittent headaches unrelated to posture often with N/V, drowsiness, irritability and impaired mentation.

Signs may include abducens nerve palsy.

Incidence in shunted patients 2-5 % [4) 5)]. CT or MRI scans may also show evidence of transependymal absortion of CSF.

2. Total shunt malfunction ('normal volume hydrocephalus') may occur and yet ventricles remain slit-like if the ventricles cannot expand because of subependymal gliosis due to Law of Laplace.

3. Venous hypertension with normal shunt function: may result from partial venous occlusion that occurs in some conditions (e.g. at the level of the jugular foramen in Crouzon´s syndrome). Usually subsides by adulthood.

Rekate, prefer to limit the use of the term slit-ventricle syndrome to the triad of intermittent headaches lasting 10-30 min, smaller than normal ventricles on imaging studies, and slow refill of shunt-pumping devices. In other situations, a description relating to the presumed pathogenesis should be used [6)].

Of the 5 different types classified by Rekate:

Type 1 is caused by shunt overdrainage and is associated with low pressures

Types 2 and 3 are associated with shunt blockage and elevated CSF pressures

Type 4 is craniocerebral disproportion that increases brain parenchymal pressure but not CSF pressure

Type 5 is headache unrelated to shunt function. The low and normal CSF pressure types are relatively well understood, but the high-pressure forms are more problematic. In the high-pressure forms of SVS it is said that the lack of ventricular dilation is related to a reduction in brain compliance analogous to idiopathic intracranial hypertension or pseudotumor cerebri. Despite this, there is little evidence in the literature to support this conjecture.

The MR venography findings and hemodynamics of 3 cases are shown to be identical to those of pseudotumor cerebri. A literature review indicates that an underlying venous impairment may be functioning in the patients who re-present with small ventricles following shunt malfunction [7].

Treatment

see Slit ventricle treatment.

Case series

2006

Ten patients treated with cisterna magna-ventricle-peritoneum (CMVP) shunts for complex problems of shunt function were reviewed retrospectively. All patients had documented increases in ICP and ventricles that did not expand despite life-threatening increases (> 80 mm Hg in one case) in ICP. Between 1995 and 2003, 10 patients (four males and six females, age range 4-32 years) were identified as having life-threatening increases in ICP despite small or slit-like ventricles on imaging studies. Each episode was documented with intraparenchymal pressure monitoring. All patients had documented ventricular catheter failures at the time of the intervention, and all had undergone at least one previous attempt to treat the condition with a valve upgrade and replacement of the ventricular catheter. Three patients had achondroplasia, four had spina bifida, and three had a preexisting Chiari malformation Type I. All patients improved after the procedure, and none suffered permanent complications. For at least 48 hours after surgery, all patients underwent intraparenchymal monitoring of ICP (an intraparenchymal monitor was used that documented normal ICP).

The CMVP shunts are an excellent option for patients who are not candidates for LP shunts but who have high ICP and ventricles that do not enlarge at shunt failure. The ability to access the spinal fluid in the cortical subarachnoid space presumably accounts for this success [8].

1987

McLaurin and Olivi reviewed 15 cases treated by a fairly uniform technique during the past 5 years. The syndrome consists of: (1) intermittent, but self-limiting episodes resembling shunt malfunction, usually lasting a few days, (2) nonfilling of the pumping device after compression, and (3) a slit-like ventricular system on CT scan. In all but 2 patients the initial shunt was performed in infancy. The mean interval from the initial shunt to treatment of SVS was 6 years. The age range at onset of SVS varied from 2 to 17 years with a mean of 7 years. All patients in this series were relieved of symptoms by placement of an antisiphon device and, in most patients, upgrading the valve resistance. Analysis of this series has led to the following conclusions: (1) SVS is a characteristic clinical entity, usually

distinguishable from persistent shunt malfunction and from low-pressure headache, (2) the pathogenesis is intermittent obstruction of the ventricular catheter, (3) there is no good evidence that changes of brain compliance or La Place principles apply, and (4) placement of antisiphon device and upgrading valve resistance are effective treatments [9].

Case reports

A 5-year-old female with a premedical history of neonatal IVH, secondary hydrocephalus, multiple VP shunt revisions, and slit ventricle syndrome was evaluated using a noninvasive intracranial pressure monitoring device at the early stages of the clinical symptoms, evidencing increased intracranial pressure and poor brain compliance. Serial MRI images demonstrated a slight ventricular enlargement, leading to the use of a gravitational VP shunt, promoting progressive improvement. On the follow-up visits, they used the noninvasive ICP monitoring device to guide the shunt adjustments until symptom resolution. Furthermore, the patient has been asymptomatic for the past 3 years without requiring new shunt revisions.

Slit ventricle syndrome and VP shunt dysfunctions are challenging diagnoses for the neurosurgeon. The noninvasive intracranial monitoring has allowed a closer follow-up assisting early assessment of brain compliance changes related to a patient's symptomatology. Furthermore, this technique has high sensitivity and specificity in detecting alterations in the intracranial pressure, serving as a guide for the adjustments of programmable VP shunts, which may improve the patient's quality of life.

Noninvasive ICP monitoring may lead to a less invasive assessment of patients with slit ventricle syndrome and could be used as a guide for adjustments of programmable shunts [10].

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