Shunt overdrainage syndrome

Shunt overdrainage is a condition with various clinical presentations, ranging from mild to very severe symptoms, often requiring multiple evaluations and admission in some cases.

The condition typically presents with postural headache as well as nausea, emesis, and irritability, exacerbated in an upright position and alleviated in a supine position.

Symptomatology can only be explained by sudden venous entrapment following chronic venous distention as a result of chronic overdrainage. Subsequent therapeutic management with an overdrainage preventing shunt and satisfying clinical outcome with complete ceasing of headache attacks adds insight into the pathophysiology of chronic overdrainage syndrome [1].

Diagnosis

The diagnosis is primarily clinical, but clinical information obtainable from patients with cognitive impairment and developmental delays may be limited. Imaging studies may reveal slit ventricles in some patients, but others may have normal or enlarged ventricles.

The lack of sensitive and specific neuroimaging findings complicates the diagnosis. Intermittent symptoms are considered to be a result of intermittent shunt occlusion caused by collapsed ventricle obstructing inlet port of the ventricular tube [2].

Complications

Intracranial hypotension.

Intracranial epidural hematoma after ventriculoperitoneal shunt overdrainage.

Overshunting associated myelopathy (OSAM) is a very rare complication of ventricular shunt therapy, and only 12 previous cases have been reported in the literature [3].

Treatment

Overdrainage of CSF is a chronic complication that remains an unsolved problem in shunt therapy and is a well-documented in hydrocephalic patients as well in shunted pediatric patients with hydrocephalus.

Modern adjustable and gravity-assisted valves enable surgeons to set the opening pressure relatively low to avoid underdrainage without significantly raising the incidence of overdrainage and to treat overdrainage-related clinical and radiological complications without surgical intervention [4] [5].

Antisiphon device (ASD).

The prophylactic implantation of an ASD in every pediatric hydrocephalus shunt is recommended [6].

Rare clinics

Cervical myelopathy produced by an engorged suboccipital epidural venous plexus due to chronic cerebrospinal fluid (CSF) overdrainage [7].

Case series

2017

Nagahama et al., treated 24 patients with symptomatic CSF overdrainage by inserting a Strata valve. The severity of symptoms was graded, and the frequency of hospital visits and shunt operations was recorded before and after insertion of the valve. Additionally, results of brain imaging and intracranial pressure monitoring were reviewed. Nineteen patients (79.2%) had severe symptoms at the time of the insertion; 1 year after Strata valve insertion only one patient (4.17%) still suffered severe symptoms. The number of hospital admissions was 3.38/patient/year before placement and 1.21 for the 1st year, 1 for the 2nd, and 0.4 for the 3rd postoperative year. The number of operations was 3.42/patient/year during the year before placement of the valve, and then 0.71 for the 1st, 0.56 for the 2nd, and 0.25 for the 3rd postoperative years. During the 1st year after placement of the Strata valve, the settings were changed 2.79 times/patient/year, 1.29 for the 2nd, and 1.33 times/patient/year for the 3rd year.

The Strata valve was effective in improving the symptoms of overdrainage in the majority of patients in this series. The number of hospital admissions and operations for valve malfunction was reduced [8].

2013

Khan et al. reviewed the charts of 164 consecutive patients with iNPH who underwent shunt surgery at their institution from 2005 to 2011. They noted age, sex, presenting symptoms, symptom duration, hypertension, body mass index (BMI), imaging findings of atrophy, white matter changes, entrapped sulci, LPOP, valve opening pressure (VOP) setting, number of valve adjustments, serious overdrainage (subdural hematoma requiring surgery), radiological overdrainage (subdural hematomas or hygroma seen on postoperative imaging), clinical overdrainage (sustained or postural headache), other complications, and improvements in gait, urine control, and memory.

Eight patients (5%) developed subdural hematomas requiring surgery. All had an LPOP of greater than 160 mm H2O and an LPOP-VOP of greater than 40 mm H2O. Radiological overdrainage was more common in those with an LPOP of greater than 160 mm H2O than in those with an LPOP of less than 160 mm H2O (38% vs. 21%, respectively; $p = 0.024$). The BMI was also significantly higher in those with an LPOP of greater than 160 mm H2O (median 30.2 vs. 27.0, respectively; $p = 0.005$).

Serious overdrainage that caused subdural hematomas and also required surgery after shunting was related to LPOP and LPOP-VOP, which in turn were related to BMI. If this can be replicated, individuals with a high LPOP should have their VOP set close to the LPOP, or even higher. In doing this, perhaps overdrainage complications can be reduced [9].

Case reports

2017

A 29-year-old woman with a shunt since the postnatal period suffering from chronic but the most severe intermittent headache attacks, despite an open shunt and with unchanged ventricular width during attacks. Intracranial pressure (ICP) recordings were performed during headache attacks and thereafter.

Massively increased ICPs, a continuous B wave “storm,” and severely compromised intracranial compliance despite an open shunt were found, a scenario that was always self-limiting with the resolution of symptoms after several hours. When mobilized to the upright position, her ICPs dropped to - 17 mm Hg, proving shunt overdrainage.

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A 3-year-old boy with drug-resistant focal onset seizures due to hemimegalencephaly who previously underwent functional hemispherotomy followed by ventriculoperitoneal shunt placement for postoperative hydrocephalus. The subsequent clinical course was complicated by delayed diagnosis of shunt overdrainage in the absence of significant image findings. Maintaining a high index of suspicion for the possibility of shunt overdrainage is critical even in the face of unremarkable imaging findings [11].

2006

In two patients with over-drainage due to shunted occlusive hydrocephalus ventricles were dilated by integrating an antisiphon device (Miethke ShuntAssistant, Aesculap AG, Tuttlingen, Germany) into their shunt systems. The resistance of the antisiphon device, which is available in configurations from 10 to 35 cm H2O was chosen 10 cm H2O higher than necessary to prevent siphoning in the individual patient.

Both patients gradually recovered from their over-drainage symptoms and the ventricles enlarged enough to allow access with an endoscope. Using a standard procedure, ETV was performed 7 days and 1 month later, respectively. In the same operative session the shunts were occluded. Shunts were removed within 6 weeks after ETV. During follow-up of more than 3 years, both patients remained free of symptoms.

The incorporation of an antisiphon device with resistance level selected 10 cm H2O higher than needed to prevent anti-siphoning into a pre-existing shunt system in patients suffering from shunt-related over-drainage is a safe and effective technique to render ventricles large enough to allow endoscopic access for ETV [12].

[1], [10]
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