

# THE DILEMMA OF THE SLIT VENTRICLE-SYNDROME (SVS): EVALUATION AND RETROSPECTIVE STUDY OF OWN SERIES AND REVIEW OF THE LITERATURE

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**Abstract:** The purpose of this study is to review the pathophysiology and treatment of severe headache disorders in patients having a VP shunt system as CSF diversion for treatment of hydrocephalus, who developed slit ventricle syndrome after insertion of the VP shunt system.

**Materials and methods:** The literature on the management of the slit ventricle syndrome is reviewed as well as an assessment of personal experiences over a 6-year period of time in the management of severe headache disorders in shunted patients with slit ventricle syndrome. Additionally a case of pediatric patient and a case of an adult patient are demonstrated.

**Results:** If the slit ventricle syndrome is defined as severe, life-modifying headaches in patients with shunts and normal or smaller than normal ventricles with ventricular shunts for the treatment of hydrocephalus, there are five different pathophysiologies, that are involved in the process.

These pathologies are defined by intracranial pressure measurement as severe intracranial hypotension analogous to spinal headaches, intermittent obstruction of the ventricular catheter, intracranial hypertension with small ventricles and a failed shunt (normal volume hydrocephalus), intracranial hypertension with a working shunt (cephalocranial hypertension), and shunt-related migraine.

The treatment of these conditions and identifying patients with each condition are facilitated by attempting to remove the shunt.

**Conclusions:** To prevent the complication, CSF overdrainage should be avoided as much as possible. The setting of a programmable shunt should be set to high to prevent slit-like ventricles and premature suture fusion. The importance of HC monitoring in pediatric patients needs to be emphasized. Manual examination and a skull X-ray are required to assess the status of suture lines during follow-ups.

Following the analysis of attempts to remove shunts, there are three possible outcomes. In about a quarter of patients, the shunt can be removed without having to be replaced. This is most common in patients treated in infancy for post-hemorrhagic hydrocephalus or patients shunted early after or before brain tumor surgery. Another half of patients have increased intracranial pressure and enlarged ventricles. In these patients, there is an 80% success rate for endoscopic third ventriculostomy. Finally, the most severe form of the slit ventricle syndrome involves intracranial hypertension without ventriculomegaly, which is managed optimally by shunt strategies that emphasize drainage of the cortical subarachnoid space such as lumboperitoneal shunts or shunts that include cisterna magna catheters.

**Keywords:** VP-Shunt, Headaches, Hydrocephalus, Slit ventricle syndrome.

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## 1. INTRODUCTION

Headache following lumbar puncture has been generally assumed to result from leakage of CSF through the puncture-hole in the dura causing CSF hypotension and distension of the pain sensitive structures at the base of the brain. The leakage of CSF through the puncture hole in dura has been proved by isotope studies and surgical exploration. [1, 3, 7-9].

Schaltenbrand and Wolf P have reasoned that the spontaneous aliquorrhoea syndrome may result from either decreased CSF production (from an unexplained disturbance of chorioid plexus function) or enhanced CSF resorption. The latter seems improbable. Posture dependent headache following minor trauma such as a fall on the buttocks has been claimed to be due to a dural tear with subsequent leakage of CSF.

A purely spontaneous tear in the dura seems unlikely because of its strong texture. CSF hypotension has been associated with increase in brain volume<sup>7</sup> and the CT findings in our case support this view. The slit-shaped ventricles, tight basal cisterns and scant CSF over the cortex fit well with diffuse brain oedema.

Dexamethasone treatment resulted in rapid disappearance of headache, and the CT scan three months later was normal. The cause of the apparent brain oedema remains unknown but it might be secondary to dilatation of brain veins and downward brain sagging which have been observed during CSF hypotension.

The extremely tight ventricles during the headache period of spontaneous intracranial hypotension may compromise the production of CSF by the chorioid plexuses and thus lead to a vicious circle. The exact mechanisms remain unresolved but brain swelling having perhaps some role in the pathogenesis. In earlier reports the most promising therapeutic results were achieved by hypotonic saline iv infusions. In headache after lumbar puncture fludrocortisone has been advocated, probably because of its sodium retaining property.

In many cases fludrocortisone had no effect on the symptom perhaps because of increase in brain swelling. The immediate therapeutic response to dexamethasone seems logical because CT indicated the presence of diffuse brain oedema. If post-lumbar puncture headache is also associated with brain oedema, dexamethasone might be worth a therapeutic trial in that condition.

Shunt surgery is a common treatment for hydrocephalus in infancy. Shunt surgery provides immediate relief of intracranial pressure (ICP) and its symptoms in affected children. However, shunting distorts normal cerebrospinal fluid (CSF) dynamics to such an extent that many problems may develop after the procedure [1]. Overdrainage of CSF can affect the physical characteristics and growth of the brain parenchyma, ventricles, and skull [2].

SVS refers to the occurrence of headache, vomiting, and possibly some degree of impaired consciousness and visual deterioration in children with hydrocephalus and VP shunts [3-8]. The pathogenesis remains unclear. Overdrainage, periodic shunt malfunction, intracranial hypertension, and decreased brain parenchyma compliance to variations in cerebrospinal fluid volume have been proposed as SVS mechanisms. Recent research suggested that lack of ventricular dilation is related to a reduction in brain compliance, analogous to that seen in idiopathic intracranial hypertension [4-10]. Therefore, diagnosis of SVS can be delayed if systemic symptoms of increased intracranial pressure are absent and neuroimaging findings are negative. As in this case, visual symptoms may be the only clue to shunt malfunction. The anterior and posterior visual pathways and the ocular motor nerves, especially the abducens, are vulnerable to increased intracranial pressure because of their subarachnoid course [2].

Although papilloedema is not a sensitive sign of shunt dysfunction in children, it remains a pivotal finding that may suggest shunt dysfunction [5]. Nguyen et al. reported ophthalmic complications of slit-ventricle syndrome in children (Ophthalmology. 2002;109:520-524) reported ophthalmic complications of SVS in children, and two of six patients in their study developed optic atrophy later in the disease course.

As elevated intracranial pressure related to SVS can lead to permanent visual loss, visual symptoms should not be overlooked in VP shunt patients.

**Classification of Hydrocephalus:** Based on recovery of supra-vital dye there is a non-communicating hydrocephalus due to aqueduct stenosis or narrowing in the fourth ventricle.

The communicating hydrocephalus can be explained with a blockage between the SSAS and CSAS, obstruction at arachnoid villi or venous hypertension.

**Classification Based On Point Of Obstruction:** Generally, every Hydrocephalus is Obstructive!. According to Ransohoff Classification there are –Intraventricular obstructive and Extraventricular obstructive Hydrocephalus. The current imaging technology should allow the definition of the “first” point of obstruction

**The point of obstruction in post-hemorrhagic hydrocephalus:** Acutely it is in the area of the arachnoid villi; but later, in all forms of PHH the blockage is between the SSAS and CSAS, which is an ideal indication for ETV.

A decrease in ventricular size and brain compliance occasionally leads to slit ventricle syndrome, a serious complication of long-standing shunts [3, 4].

In slit ventricle syndrome, small ventricles and a stiff brain make the shunt malfunction and cause significant headache and other symptoms of increased ICP. Derangement of skull growth results in secondary craniosynostosis and small head circumference (HC). It is known that secondary craniosynostosis develops in 1–5% of patients with shunts [5, 6].

Most patients with secondary craniosynostosis are asymptomatic, but the condition is occasionally combined with slit ventricle syndrome [2].

In this situation, the 2 conditions can elicit a significant and synergistic increase in ICP and its symptoms, requiring urgent surgical intervention [2].

However, these changes in the brain and skull usually develop from long-standing shunts, usually years after shunt surgery [7]. Early-onset slit ventricle syndrome and secondary craniosynostosis are extremely rare in infants who underwent shunt surgery only months earlier. The symptoms and signs of slit ventricle syndrome and secondary craniosynostosis overlap with those of shunt obstruction, which is far more common. Therefore, differential diagnosis of these conditions in infants and young children may require clinical acumen.

Headaches are one of the most common afflictions of mankind. Based on large series of patients, about 4% of the adults in the world suffer headaches every day, with a female-to-male ratio of 2.5:1 [1]. A larger but inestimable number of individuals have occasional incapacitating headaches [2]. Not surprisingly then, patients with shunts have headache disorders.

Slit ventricles (SV) are collapsed or abnormally small ventricles apparent on computed tomographic (CT) or magnetic resonance imaging (MRI) scans after insertion of a ventricular shunt in a patient with hydrocephalus. Although many patients with SV are asymptomatic, slit ventricle syndrome (SVS) is said to exist when SV are accompanied by intermittent episodes of severe headaches, cyclic nausea and/or vomiting, and slow refill of the shunt's pumping device after compression.

These episodes are believed to reflect sudden, periodic increases in intracranial pressure (ICP), possibly related to underlying decreased intracranial compliance.

The presence of the shunt in patients with headaches always leads to the assumption that the inserted shunt system is not functioning properly. This assumption can lead to large numbers of expensive and possibly dangerous imaging studies, long waits in emergency rooms, and the expenditure of considerable money when the headaches are a chronic condition.

There is always the possibility that patients could die or develop severe neurologic dysfunction from high intracranial pressure (ICP) at the time of shunt failure. Medically, therefore, it is reasonable to ascertain that their shunt is working. Doing so, however, is not always straightforward. Since their development in the 1950s, valve-regulated shunts have probably saved more lives and more cognitive function for more years than anything else neurosurgeons have ever done [3, 4].

However, shunts have led also to a great many problems that were not easy to predict from the beginning. The most common and most chronic of these newly recognized conditions is the association of chronic headaches with the presence of a shunt.

Severe headache disorder in patients with shunts and small ventricles has been called the “slit-ventricle syndrome” (SVS) [5].

SVS is not a single condition; rather, several different pathophysiologies can underlie this constellation of findings [6]. This article defines the various causes of severe headaches in shunted patients, suggests an algorithm for the diagnosis and treatment of this common condition, and suggests a management approach to patients with shunt-related problems.

**1.1. Severe Headaches in shunted Patients with Slit Ventricle Syndromes:**

Incidence of headaches in shunted patients with hydrocephalus: Essentially all adolescents and young adults with shunts will have headaches if asked. Most of these headaches are mild, intermittent and lead to normal function. Severe headaches in shunted patients with apparently working shunts is frequently according to so called the “Slit Ventricle Syndrome” (SVS), over drainage, subdural haematomas or hygromas.



**Figure 1: Head CT with inserted intraventricular VP shunt catheter over the ventral horn of the right lateral ventricle.**

Radiographically, slit ventricles in the head CT (Figure 1) are present in about 80% of the patients; however Slit Ventricle Syndrome is diagnosed in approximately 15% in this patient group.

Causes Of Headaches Based On ICP Monitoring: Almost five reasons were presented in the literature or “Headaches in Patients With Shunts” as according to the authors Harold L. Rekate, MD, and Dory Kranz in the publication in the Seminars of Pediatric Neurology in 2009, which can explain the etiology of this kind of headaches.

Intracranial Hypotension, intermittent proximal obstruction, intracranial hypertension with a failed shunt and small ventricles (NVH: Normal Volume Hydrocephalus), intracranial hypertension with a working shunt (Cephalocranial dysproportion) and shunt related migraine.

Headache syndromes: Intense headache can be present over the period of time 10-90 minutes and it can awaken patient from sleep. Most of the headaches was worsening in afternoon, whereas improvement was recognized by the patients after lying down.

A positive family history can increase the possibility of the headache attacks. Intermittent proximal obstruction, intracranial hypotension, NVH or CCD and shunt related migraine were discussed in previous publications as possible triggers of the shunt related headaches.

Medical Management Of Headaches: Patients with shunts can be treated similar to patients with migraine; Beta blocker (Inderal), Cyproheptadine, Sumatryptin (Imitrex) and analogs can be prescribed.

Who needs intervention: Patient should be involved in the decision when and how to intervene: Suggested threshold: The patient as pupil must leave school or as adult the work twice per month or more frequently, lying down in school nurse’s office, most of these patients are not at imminent risk and decision can be made over time and the headache diary.

The First Step: Shunt revision with incorporation of a Device for Resistive Siphon (DRS): 85% of chronic headaches are improved by this technique only. Technology of shunting has made great strides in the past decade.

What is a DRS: Device which retards siphoning. There are multiple types and designs; such as Orbis Sigma, Delta Valve, Anti-Siphon Device, Siphon Control Device, Siphonguard and gravity compensating device.

**How to Manage / The New Classification:**

As shunting of the ventricles to the jugular vein or peritoneum became standard treatment for hydrocephalus, problems related to overdrainage were recognized with increasing frequency [7]. Before valve regulation of cerebrospinal fluid (CSF) flow was developed, drainage of the lateral ventricles in children with hydrocephalus was precluded by the routine collapse of the brain with associated lethal subdural hematomas.

The development of valve regulation made it possible to treat severe hydrocephalus successfully with much less likelihood of this dreaded complication.

By the mid-1970s, problems related to the ventricles becoming too small, leading to intermittent or recurrent obstruction of the ventricular catheter, was recognized as a severe problem. Several strategies, including subtemporal decompression for the management of this condition, were introduced [8–10].

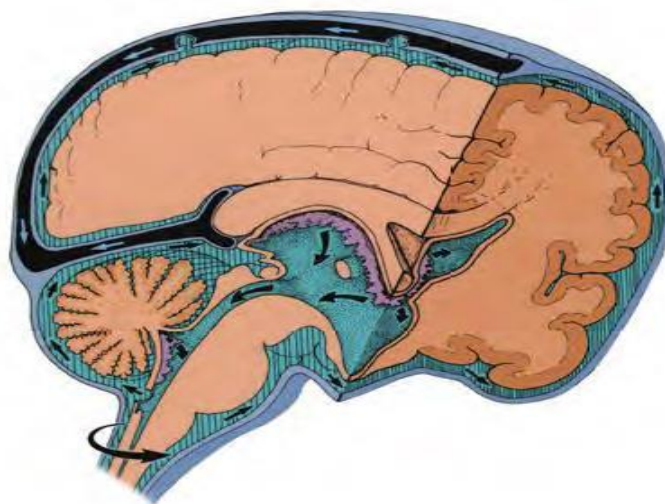
In an early report on our experiences with this condition, definition of SVS was a triad involving intermittent headaches lasting 10 to 90 min, small ventricles on imaging studies, and slow refilling of the pumping mechanism of the valve [5].

In this same article, it was demonstrated that it was possible to increase the volume of the lateral ventricles by using valve upgrading and a device that retards siphoning (DRS). It is also recommended that the use of low-pressure shunts should be abandoned unless dictated by specific indications.

As experience grew, especially with the ability to track changes in ventricular size using contemporary neuroimaging such as computed tomography (CT) and magnetic resonance imaging (MRI), it became obvious that this view of the SVS was too simplistic and that there were multiple different forms of the problem.

Reports dealing with SVS were not describing the same condition [11].

In several papers, Rekaté et al. published their experiences with ICP monitoring in patients with small ventricles and severe headaches and / or with slit ventricles. Five distinct pathophysiologies were identified for this condition (see below). There is some overlap, but each condition requires specific treatment paradigms [6]. Based on many previous studies, it has been our policy to upgrade the valve and include a DRS in all patients. All patients studied had previously undergone the procedure [5].



**Figure 2: Intracranial CSF circulation.**

All shunt operations are associated with some risk, especially the risk of infection. Therefore, it requires a significant discussion with all patients and if children, their parents, to determine whether to intervene. Surgical intervention is indicated only in patients whose headaches significantly interfere with normal life. If children have to leave school or adults need to leave work or discontinue working more than twice a month, it is generally believed, that surgical intervention is justified. There is little consensus about the definition or causes of headaches in patients cerebrospinal fluid diversion.

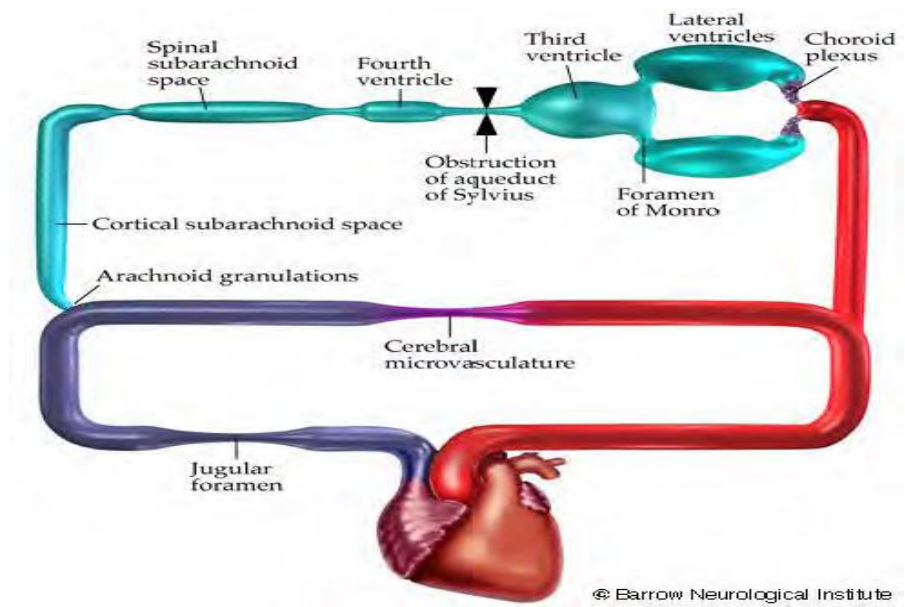
Most such patients undergo a valve pressure upgrade and incorporation of hydrostatic valves, anti-siphon devices or variable-resistance valves. About one in five of these patients, however, does not improve or improves only temporarily. Before further intervention is pursued, the causes of the patient’s headaches and the relationship of the headaches to shunt function and ICP must be understood fully. To understand the causes of the headaches, it is essential to define the relationship of ICP and shunt function to the headaches.

Based on chronic monitoring of ICP in these patients with headaches, five syndromes of shunt-related headaches were defined in the following chapters. Each syndrome leads to specific treatment strategies [6].

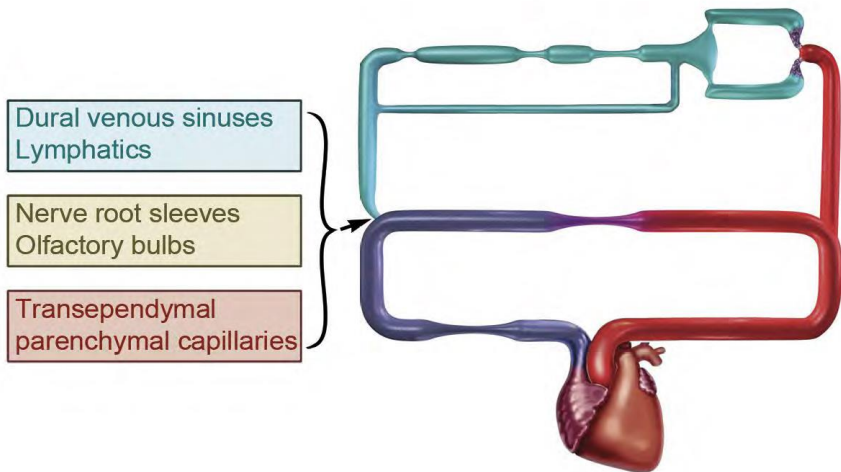
**1.2. Endoscopic third ventriculostomy (ETV):**

Candidates for ETV are ideally patients with aqueduct stenosis or stenosis in the 4<sup>th</sup> ventricle and patients with the condition SSAS to CSAS. There is no indication in patients with CSAS to SSS (arachnoid villi) and patients with venous hypertension (Pseudotumor). (Figure 3 and 4).

**Contraindications to ETV:** Patients with achondroplasia, craniofacial syndromes, history Of unresponsive ventricles, association with Spina Bifida, or multiple sites of obstruction have to be excluded from a procedure with ETV. Drake et al see ETV failure as same as shunt failure.



**Figure 3: Intracranial CSF proportions and extracranial pressure relationships.**



**Figure 4: What is accomplished by ETV.**

**Benefit Assessment of ETV:** The main circumstantial rule in understanding of the difference between VP shunt insertion and ETV is that a foreign body insertion does not exist in ETV.

ETV is substantially more dangerous than a shunt procedure and significantly less dangerous than a lifetime of shunt dependency.

If ETV is planned, there is usually no reliance on implanted foreign body and most series show substantially greater longevity. On the other hand, in case of insertion of a Shunt system, it is easy to tell whether or not the hydrocephalus is being treated. The initial risks are usually more inconvenient than dangerous.

**Risk Assessment of ETV:** It is very important to find out the best possible management for the patient, whether shunt insertion is indicated or possibly patient is a candidate for ETV.

In case of Shunt systems the Rate of Infection is 8%, the rate of Failure is 20-50%. Death per year is about 1%. In case of ETV following facts to be considered: Hormonal Difficulty usually with possible diabetes insipidus (DI), loss of recent memory, diplopia, hemiparesis and death.

### 1.3. Intracranial hypotension:

These patients develop severe headaches that are not present while they are reclined in bed. The headache develops later in the day and gets worse with time as patients maintain an erect position. The headache improves rather rapidly if patients can lie down.

Monitoring shows significantly subnormal ICP. The recorded ICP was from -25 to -30 mmHg. These headaches are analogous to postlumbar puncture headaches and may be associated with enhancement of the meninges on contrast-enhanced imaging studies. This condition implies that the DRS has failed and that the patient will respond to replacement of the valve mechanism with insertion of an effective DRS and possibly a valve upgrade. Since the publication of the article [6], it is indicated to use a programmable valve. It is preferred to use the Codman Hakim Programmable Valve with Siphoguard™ (Codman Corporation, Raynham, MA). This valve does not depend on a diaphragm mechanism. Therefore, it can be placed anywhere along the course of the shunt. The skin does not have to move freely. Other programmable or adjustable valves are commercially available.

### 1.4. Intermittent proximal obstruction:

This problem is probably the most common of the five conditions and represents patients who were originally described as having SVS. Monitoring shows that patients with this condition have normal to low ICP most of the day, but their ICP increases suddenly with activity.

As their ICP increases, the headache worsens until the ventricular catheter reopens and the pressure reverts to normal again. These patients are also managed by placement of a DRS and valve upgrade. In chronically shunted individuals, proximal shunt failure is the most common form of mechanical failure of shunts. For decades, my policy has been to assume that this condition is a result of overdrainage of CSF and collapse of the ventricular walls around the ventricular catheter. Therefore, it is a sign that the back pressure or opening pressure of the valve is inadequate to maintain CSF within the ventricular system. It is believed that the valve should be upgraded and a DRS should be incorporated into the system any time a proximal obstruction occurs.

### 1.5. Shunt failure without ventricular enlargement:

Engel et al. [12] originally described intracranial hypertension with nondistending ventricles, so-called normal volume hydrocephalus (NVH). This enigmatic condition has been studied intensively. It is the most important and difficult to manage of the subtypes of SVS. In the series of Engel et al. [12], the patients were found to have signs and symptoms of increased ICP with no enlargement of the ventricular system. They recommended exploration of these shunts, which were routinely found to be nonfunctional. These patients become symptomatic with progressive symptoms of increased ICP with morning headaches progressing to all-day headaches, papilledema, visual loss, and diplopia. If the condition is not treated early, neurologic deterioration is possible and blindness is likely. This problem occurs rarely, if at all, in patients who develop hydrocephalus beyond infancy, but it is a common problem in cases of congenital hydrocephalus [13]. These patients begin life with hydrocephalus but when older have pseudo tumor cerebri. Performance of retrograde venographic measurements of sagittal sinus pressure was done in five such children.

All had elevated venous sinus pressure, as have all patients with pseudo tumor cerebri that have been tested [14]. The management of this condition is considered below.

In general, however, a shunting strategy that incorporates drainage of the subarachnoid space, such as lumboperitoneal shunts or shunts from the cisterna magna, is needed to manage these patients adequately [15–17].

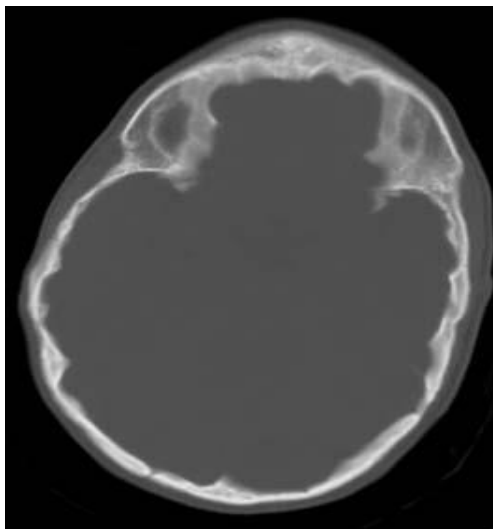
#### **1.6. Increased ICP with a working shunt: cephalocranial disproportion:**

Based on shunt flow studies and surgical exploration, these patients have working shunts but show significant signs of increased ICP. In my experience, this problem has been universally associated with hindbrain herniation (Chiari I malformation) and found exclusively in patients with craniofacial disorders such as oxycephaly, Crouzon's and Pfeiffer's syndrome (Figure 5).

Other authors have postulated that sutural closure results from decreasing ICP and from insufficient room for the growing brain [18]. With neither a significant abnormality of the shape of the skull and face nor the presence of hindbrain herniation, the problem in these patients is likely NVH and not cephalocranial disproportion. It is best managed with shunts that incorporate the subarachnoid space. Patients with true cephalocranial disproportion need a cranial expansion procedure or large subtemporal decompression. Further manipulation of the shunt is of no benefit. Hindbrain herniation in these patients can be dealt with effectively by enlarging the posterior hemicranium [19].

#### **1.7. Shunt-related migraine:**

Headaches are common in the general population. Patients with shunts can have migraines or other headache disorders that are unrelated to their shunt. Shunt-related migraine usually occurs in the context of a strong family history of migraine and is episodic. (Figure 5).



**Figure 5: CT scan of a patient with syndromic craniosynostosis and hydrocephalus showing scalloping of the inner table of the bone due to cephalocranial disproportion.**

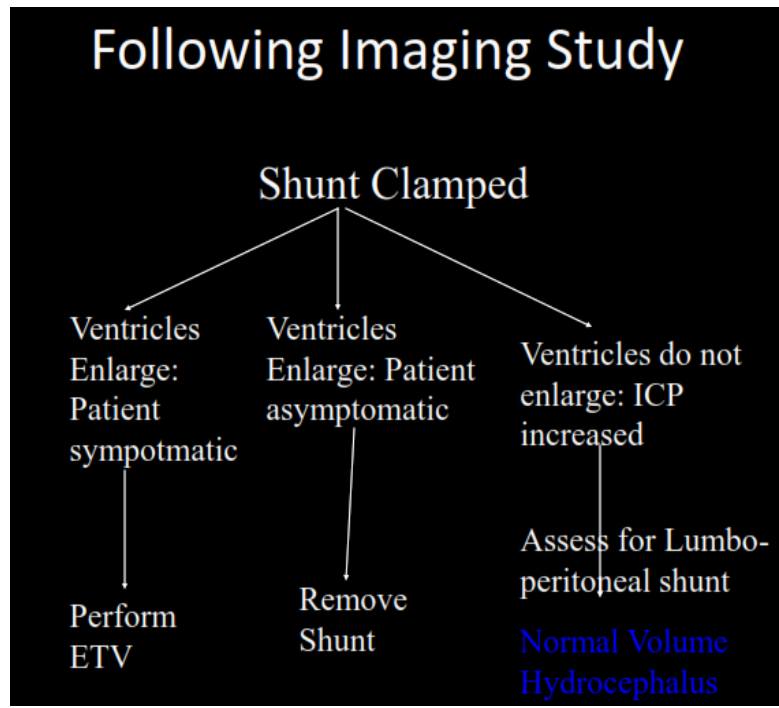
As small children, these patients usually suffer from seasonal allergies. Their descriptions of their headaches may or may not be typical of migraine. These patients often improve briefly after shunt manipulation, but the same problems return rather quickly after intervention. Because of the potential medicolegal problems associated with failure to diagnose a shunt malfunction, these patients have had many visits to the emergency room and a large number of CTscans (because these episodes are usually considered emergencies) and other diagnostic studies. Management of these patients is complicated and frustrating.

The patient, family, and neurology staff are often reluctant to believe that the headaches are unrelated to the shunt. Therefore, considerable energy and financial resources are necessary to treat these patients. Documentation by ICP monitoring is often needed to prove that the headaches are unrelated to the shunt before a commitment to medical management can be reached [6].



**1.8. Shunt-removal protocol:**

This “Shunt Removal Protocol” was developed in the following steps: Initially to exteriorize the distal end of the shunt or remove and replace it with an EVD. Then to raise drainage height or clamp ventriculostomy in ICU setting. It is necessary to obtain imaging study. (Figure 6).



**Figure 6: Shunt removal protocol.**

Fortunately, most shunted patients do well for long periods. After the first few years they may live decades with little complaints, which can lead to the final decision of the neurosurgeon, that these patients may need an operative intervention regardless of the type of shunt used or the pressure setting. Unfortunately, this statement is not true for a small percentage of shunted patients who require frequent shunt revision and who experience major interference with their function in daily life. Although most shunted patients have radiographic slit ventricles after years of shunting, only a small number have the symptoms of SVS [20]. My practice includes a large number of patients referred from outside my normal catchment area. Therefore, it is difficult for me to predict the actual incidence of symptomatic SVS. Based on my practice, however, it can be predicted that a third of infants followed for more than 5 years will have a severe chronic headache disorder that requires intervention. At least 20% will have ventricles that do not expand at the time of shunt failure (NVH).

This second percentage is supported by information from the Division of Neurosurgery of the Children’s Hospital of Los Angeles (McComb, JG, personal communication, 2000).

As evident from the above classification of the pathophysiology underlying headaches in these patients, their management is time-consuming, frustrating, expensive, and potentially dangerous.

As stated, the first step is to use programmable shunts, that incorporate a DRS. In my opinion, the prevalence of this problem justifies the routine use of these devices. As Aschoff noted, avoiding one shunt revision would justify implanting a shunt that cost \$50,000 [21]. What are the alternatives for patients with a valve upgrade and incorporation of a hydrostatic valves, or an Anti-siphon device, or variable-resistance valve, who are still incapacitated by headaches?

Of the patients described in the discussion of the classification, a specific therapy is available only for those with cephalocranial disproportion: CRANIAL EXPANSION.

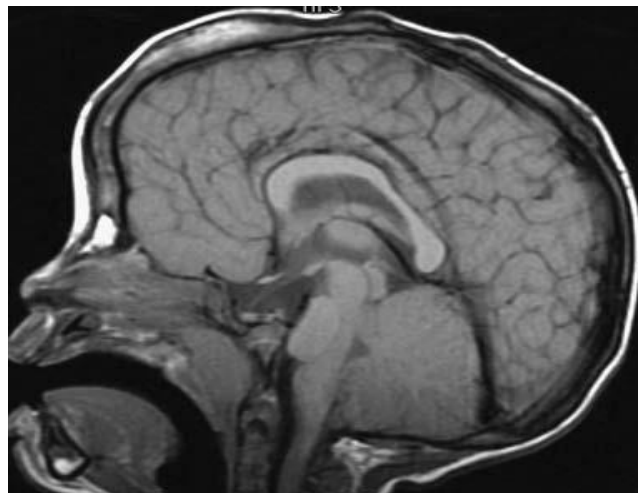
Before undertaking this procedure, it is always recommended to perform a trial of ICP monitoring to document the problem. MRI often shows a hindbrain herniation in such patients (Fig. 2). These patients require cranial expansion. The

argumentation of Dr. Di Rocco is acceptable, that occipital expansion is likely to treat both the cephalocranial disproportion and the hindbrain herniation if the bone around the foramen magnum is removed at the same time [19].

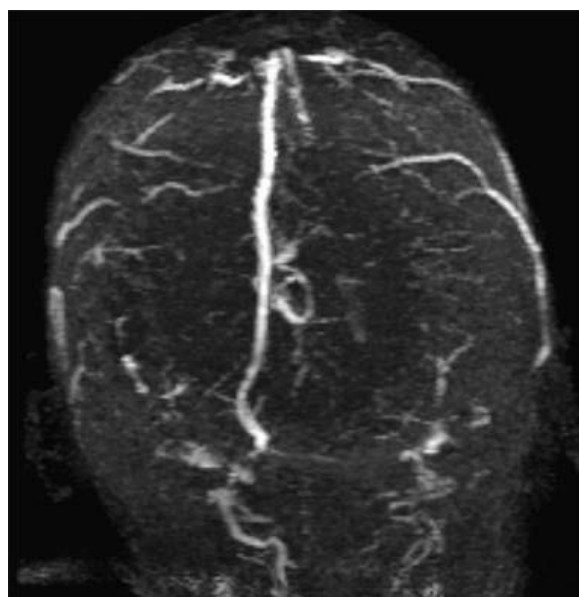
If adequate decompression can not be achieved from above, occasionally, it is necessary to explore the craniovertebral junction directly. The patients with complex craniofacial abnormalities often have an associated set of abnormalities of venous outflow from the intracranial venous sinuses and compression of the jugular foramen.

It is essential to obtain either angiographic images or MR venograms to ascertain that this flow is not interrupted (Figure 5-8). It is possible that a very large percentage of venous flow is through emissary veins and may need to be saved [22, 23]. These venous anomalies can lead to a pseudotumor-like picture, and they can coexist with NVH as discussed below. An algorithm was developed for diagnosis and treatment of all other patients with SVS who do not improve after a valve change. This strategy is an attempt to improve understanding of the pathophysiology and treatment of debilitating headaches in individual patients (Figure 7-8).

After prolonged discussions with patients and family, if appropriate, patients are offered the “shunt removal protocol [24].” This procedure has evolved over time.

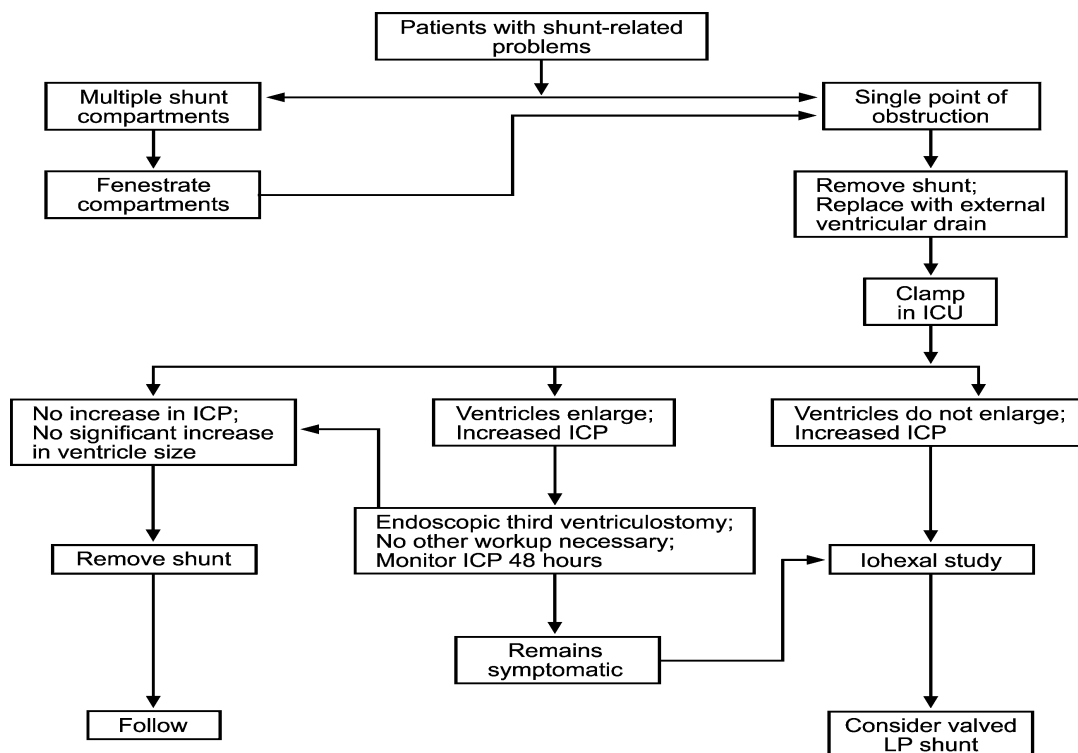


**Figure 7: MRI of brain showing hindbrain herniation**



**Figure 8: MR venography of patient with normal volume hydrocephalus showing abnormalities of venous drainage leading to late pseudotumor syndrome in NVH.**

In general, after an informed consent is obtained, the patients undergo surgery to have their entire shunt system removed and replaced with an external ventricular drain (EVD). After recovering from anesthesia, the patients are taken to the intensive care unit where the EVD is used to monitor ICP and to drain CSF if needed. Initially, the drain is left open at 25 cm H<sub>2</sub>O above the midposition of the head as long as the setting is tolerated. All participants, the patient, the nurses caring for the patient in the ICU, the family, and the neurosurgical residents are informed of the goals of the procedure and of the parameters for management of the EVD. The next morning, a scan is obtained to determine whether the ventricles have enlarged. If so, this form of treatment is continued another 24 h. If not and the patient is only mildly ill from increased ICP, the drain is closed under careful observation in an attempt to increase the size of the lateral ventricles. If the ventricles have enlarged significantly on the third hospital day, the patient returns to the operating room for an endoscopic third ventriculostomy.

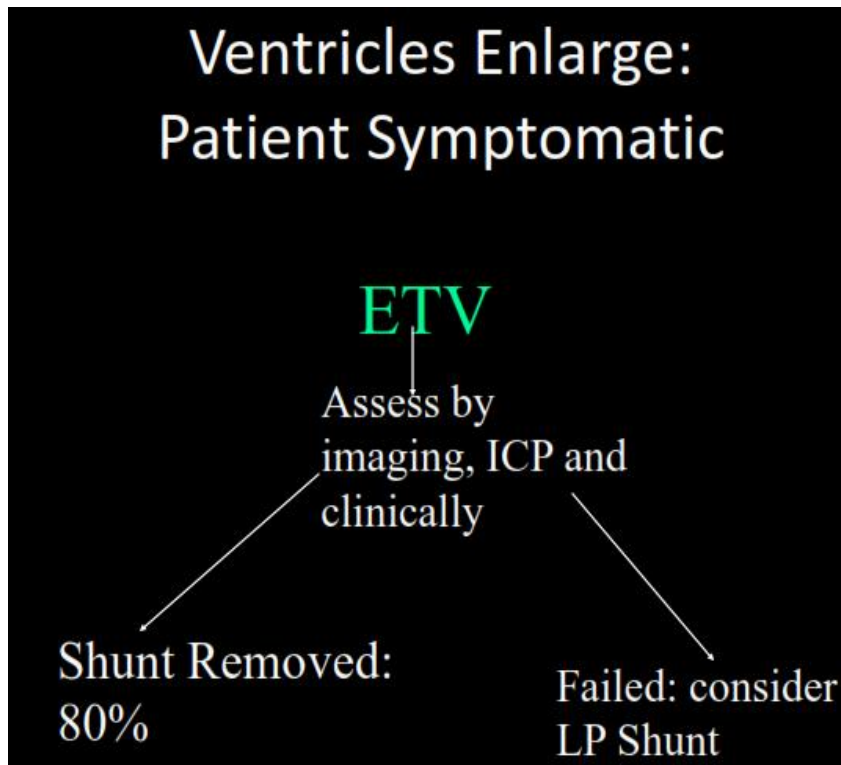


**Figure 9: Algorithm for managing shunt-related difficulties with a shunt removal protocol (with permission from Barrow Neurological Institute)**

There are three potential outcomes associated with closure of the drain. In the best scenario, the ventricles enlarge only slightly, ICP normalizes, and the patient is essentially asymptomatic. This hoped-for result has occurred in about 25% of our patients. These patients have usually undergone resection or treatment of a brain tumor or have experienced a subarachnoid or intraventricular hemorrhage. In these cases, ICP is monitored for 48 h and the drain is removed. The patient remains in close contact with our service and undergoes follow-up scans 6 weeks and 1 year after the procedure. (Figure 9).

These results support findings from our previous study on the possibility of shunt-independent arrest of hydrocephalus [25]. The second possibility is that the ventricles expand and the patient becomes ill. If the patients with a coexistent myelomeningocele (Chiari II malformation) are excluded, these patients are excellent candidates for endoscopic third ventriculostomy. The success rate for shunt-independent arrest of the hydrocephalus is 80%. It is essential to ascertain that patients who have had chronic headaches for a long time are safe and that they have no recurrent increase in ICP. Consequently, over time it was tended to leave a ventricular access device or tapping reservoir in place after the ETV has been inserted. Affixing of a butterfly needle into the reservoir for ICP monitoring for 48 h is acceptable. If patients return with headaches, the needle remains in place for later assessment of ICP and to inject contrast to ensure that the stoma is open. Using this protocol, only one late failure was observed, which occurred 1 year after the procedure. Re-exploration

of that patient revealed that the basilar artery had herniated through and sealed the stoma. A second hole was made anterior and lateral to the artery, and the patient’s symptoms resolved. The third possibility is that the ventricles do not expand, ICP increases, and patients seem ill. Such patients have NVH and are not candidates for ETV. There are two reasons why ETV is inappropriate in this condition. The first reason is practical. Even with the use of frameless stereotaxy, it is difficult to manipulate an endoscope within such small ventricles without potential damaging important structures. The second reason relates to the underlying origin of the hydrocephalus. In our experience, all patients in this series obviously had hydrocephalus during early infancy, when their first shunts were placed. Based on retrograde venous manometry performed in several of these patients, their sagittal sinus pressure is higher than normal. (Figure 10).



**Figure10: When to perform ETV.**

The ventricles have failed to expand in more than 30% of patients undergoing the protocol. It also has occurred in at least 20% of patients with hydrocephalus related to a Chiari II malformation. (Figure9). Consequently, no longer consideration was in these patients as candidates for the shunt-removal protocol. Several scenarios frequently result in NVH and respond in this way to attempts at shunt removal. The first group begins life as premature infants and is in the neonatal ICU for prolonged periods. Their hydrocephalus has been attributed to intraventricular hemorrhage, but venous studies reflect abnormal venous drainage either from congenital anomalies or chronic central lines associated with stenosis of the jugular veins or superior vena cava. The other group is usually diagnosed with congenital aqueductal stenosis. Triventricular hydrocephalus is diagnosed when their heads are discovered to be enlarging and crossing percentile lines.

Developmental assessments in these babies are normal, except for some delays in gross motor behavior related to the relatively large head. Later in life at the time of shunt failure, these children develop the signs and symptoms of increased ICP but without ventriculomegaly. At this point, MRI shows open flow through the aqueduct of Sylvius. In such cases, the hydrocephalus has actually caused the closure of the aqueduct [26].

In all of these patients and particularly in the two classes of patients discussed here, ETV offers no advantage because communication between the third ventricle and intrapeduncular cistern enables CSF to flow briefly. Intraventricular injection of Iohexal confirms free communication among the ventricles, cortical subarachnoid spaces, and cisterns except in children who experienced severe ventriculitis after their original shunt procedure [17, 24]. For these patients, it is essential to develop treatment strategies that access the cortical subarachnoid spaces [16].

**1.9. NVH: pathophysiology and treatment:**

Increased ICP associated with small ventricles that can be managed by draining CSF is called pseudotumor cerebri. Older children and adults whose ventricles do not expand at shunt failure no longer have hydrocephalus. Rather, they have a form of pseudotumor cerebri that may be more difficult to treat and more dangerous than pseudotumor cerebri that begins in adulthood. Increased pressure in the dural venous sinuses is likely to be a universal mechanism in the pathogenesis of pseudotumor cerebri in adults [14].

As can be seen in the context of a radical bilateral neck dissection, bilateral ligation of the jugular veins increases ICP and is associated with high venous pressure, increased turgor of the brain, and increased CSF in the subarachnoid spaces (i.e., pseudotumor cerebri). If bilateral radical neck dissection is performed in an infant, CSF in the cortical subarachnoid space, ventricular size, ventricular volume, or hydrocephalus would increase with increases in ICP. After shunting, the sutures close and the skull can no longer expand. The result is a shunted patient with pseudotumor cerebri. The clinical situation is the same as would have occurred at the time of shunt failure. Clinically, these patients are at great risk because the level of recognition of this common condition among general neurosurgeons, radiologists, and emergency physicians is relatively low. Almost all of these patients have been in emergency rooms where imaging studies (almost always CT scans) have been interpreted as “no evidence of hydrocephalus” or “no evidence of shunt failure.” The patients with complete and irrevocable shunt failure often have prolonged periods of suffering and occasional blindness with no evidence of ventricular dilatation. Such patients have almost universally been shunted during infancy, and some measurement of ICP may be the only way to determine if their shunt is working. In one patient with NVH in the context of spina bifida, ICP was recorded at 70 mmHg with no increase in ventricular size [15, 16]. Usually, ventricular collapse around the catheter leads to intermittent failure of the shunt and to intermittent severe intracranial hypertension and symptoms. That these patients actually have NVH and nonresponsive ventricles may not be obvious. Although patients undergo many imaging studies (usually CT scans with the attendant risks of radiation), the diagnosis is elusive. Only when papilledema or constant agony leads to a shunt tap, ICP monitoring, or shunt exploration is the condition finally diagnosed. (Figure 11).

“Normal Volume Hydrocephalus” must involve usually the cortical subarachnoid spaces. Approximately one in 5 children will be found to have non-responding ventricles at the time of shunt failure.

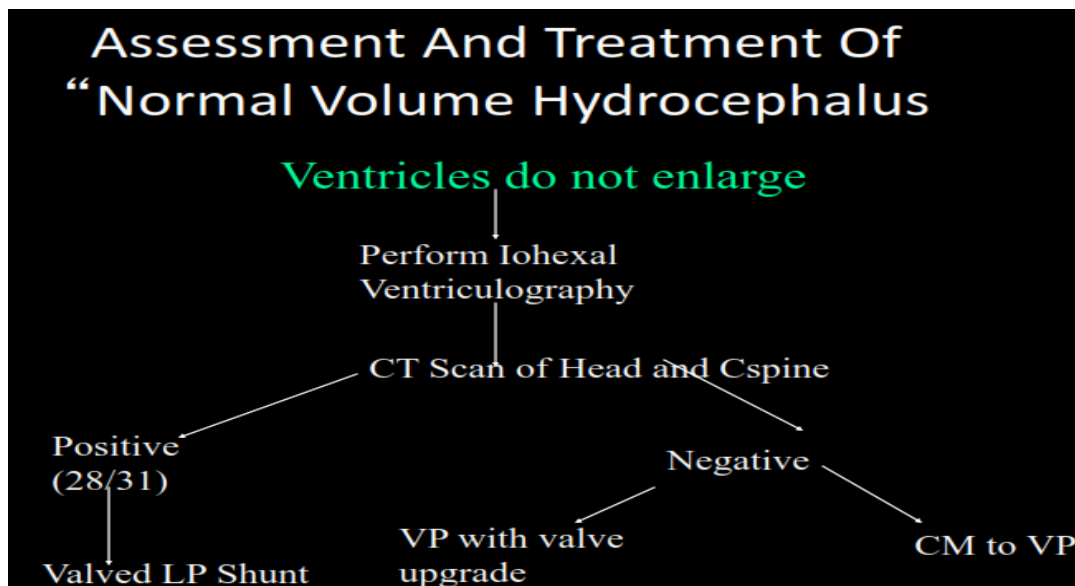


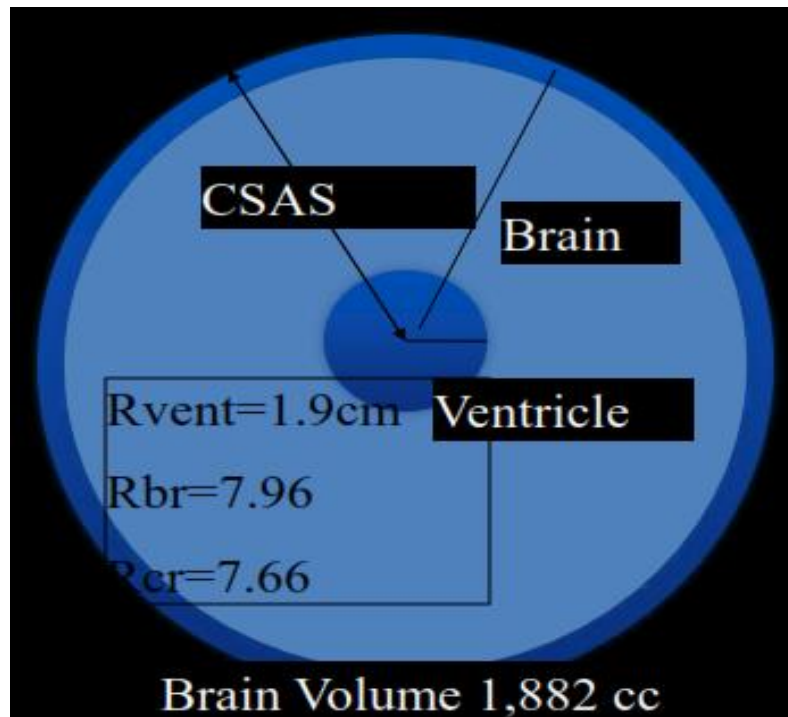
Figure 11: Assessment And Treatment Of “Normal Volume Hydrocephalus

**1.10. Lumbo-peritoneal Shunt:**

It does not require a valve except in NPH. It is usually only in patients with communicating hydrocephalus. It should be performed after Iohexal ventriculogram followed by CT of C-spine. The value of programmable valve for fine-tuning should be considered as well as to be performed with ventricular reservoir left behind.

**1.11. Implication Of Non-expanding Ventricles:**

Cortical Subarachnoid Spaces (CSAS) must be involved in the hydrocephalic process. Usually implies venous hypertension; it is the initial cause of the hydrocephalus and it does not occur when process begins as adults. Point of Obstruction is distal to interpeduncular cistern serviced by the 3<sup>rd</sup> ventriculostomy. (Figure 12).



**Figure 12: Effect of Blocked Shunt.**

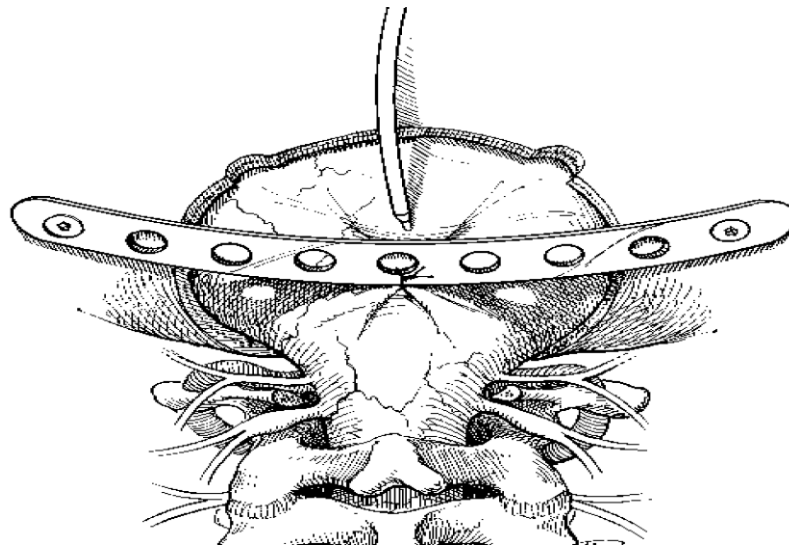
**1.12. Strategies To Overcome Severe Slit Ventricle Syndrome (NVH):**

The opening pressure has to be >5mmHg. In this case a lumbo-peritoneal shunt (must have valve) can be inserted.

The shunt system to cisterna magna , to ventricle to peritoneal shunt with LP shunt tubing in cisterna magna is the ideal treatment (Figure 13 a and 13b). The pressure of valve must be higher than the sagittal sinus.

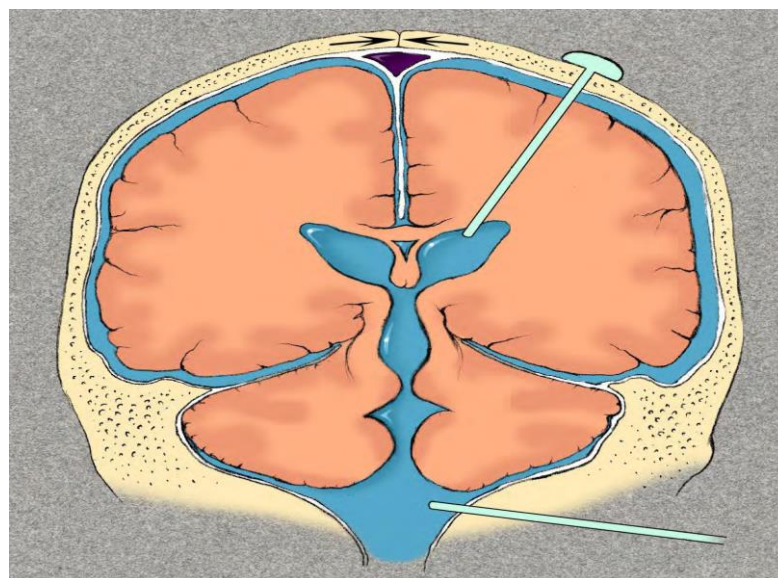


**Figure 13a: Cisterna Magna To Ventricle To Peritoneal Shunt**



**Figure 13b: Cisterna Magna To Ventricle To Peritoneal Shunt**

It is recommended to avoid shunting if at all possible. Otherwise a cortical subarachnoid shunt to peritoneum is advisable with ventricular shunts with high resistance; if possible a cisterna magna shunt to peritoneum. (CALEB procedure; Figure 14).



**Figure 14: The CALEB procedure.**

## 2. DEMONSTRATIO OF OWN CASES

### 2.1. Spontaneous intracranial hypotension with slit ventricles of a 38 years old woman:

After standing up in the morning, a previously healthy 38-year-old woman felt severe headache which was psychosis with paranoid features and the treatment was relieved by lying down. On the previous day she had felt quite fit and participated in a 10 kilometer cross-country ski-tour. Three days after onset of the headache she visited the out-patient department, and on the 4th day she was admitted to the Department of Neurology. The posture dependent headache which continued unchanged was associated with dizziness, nausea and malaise. Except for slight tenderness of the neck muscles the clinical examination was normal. A lumbar puncture on the 3rd day after onset resulted in a dry tap. This was the result also on the 4<sup>th</sup> day but aspiration with a syringe gave a colourless and clear sample of CSF with normal cell and protein content. Radiographs of the skull and chest were normal, as was the brain scan.

The dominant activity in the EEG was 9-5 Hz alpha mixed with episodic, symmetrical and synchronous theta activity. ECG was normal, as were the blood tests (ESR, complete blood cell count, blood glucose, creatinine, alkaline phosphatase, SGOT, serum Na, K, Cl, Ca, P and Mg). The osmolalities of serum and urine were normal, as were the thyroid function test and serum cortisol.

CT examination of the head was performed. The lateral, 3rd and 4th ventricles were extremely narrow, slit-shaped, and the basal cisterns were virtually non discernible. On the cortex only minimal amounts of CSF were present. The headache did not improve and then later on, fluorocortisone was started 0.3 mg/day by mouth. After five days' unsuccessful treatment dexamethasone i.m. 20 mg/day was instituted. In one day the patient became practically asymptomatic.

The dose was gradually tapered and from 5 May the corticosteroid treatment was continued with prednisolone 10 mg/day by mouth. After one week, however, the patient displayed symptoms of a manic psychosis with paranoid features and the treatment was stopped.

The psychiatric symptoms disappeared in a few days, the patient was remaining free from headache. A followup CT on 9 July showed ventricles and basal cisterns of normal size. At one-year follow-up the patient was symptom-free. CSF hypotension has been associated with increase in brain volume and the CT findings in this presented case support this view. The slit-shaped ventricles, tight basal cisterns and scant CSF over the cortex fit well with diffuse brain oedema. Dexamethasone treatment resulted in rapid disappearance of headache, and the CT scan three months later was normal. The cause of the apparent brain oedema remains unknown but it might be secondary to dilatation of brain veins and downward brain sagging which have been observed during CSF hypotension. The extremely tight ventricles during the headache period of spontaneous intracranial hypotension may compromise the production of CSF by the chorioid plexuses and thus lead to a vicious circle. The exact mechanisms remain unresolved but brain swelling having perhaps some role in the pathogenesis. (See Figure 15 with the CT scan images below)

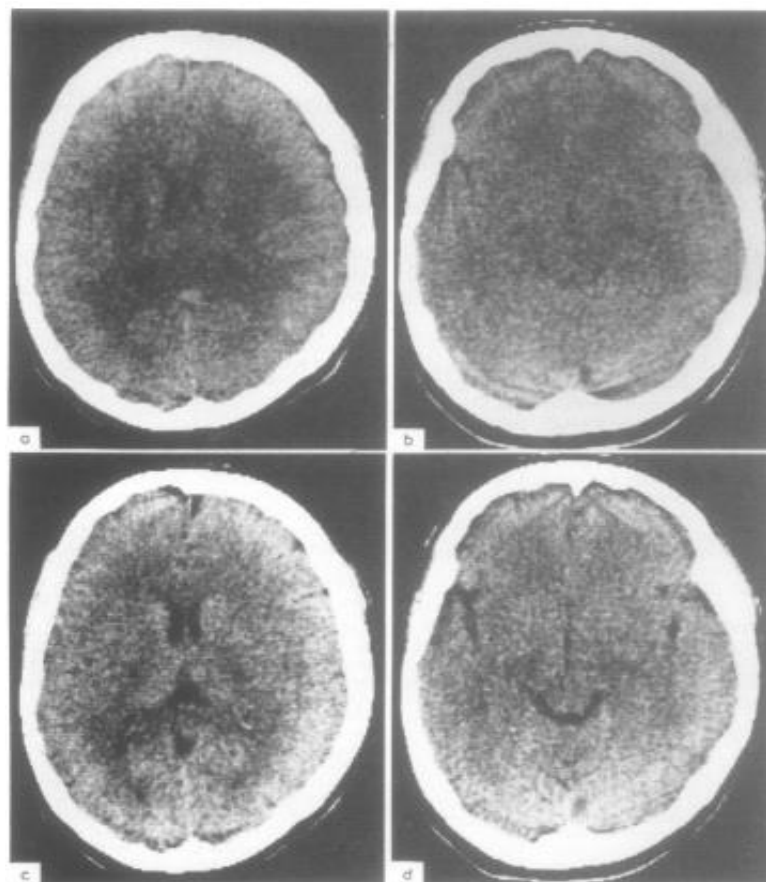


Fig: Computed tomography during the symptomatic period, showing slit ventricles and tight basal cisterns (a, b). Three months later CT showed ventricles and cisterns of normal size (c, d).

**Figure 15: CT scan during the symptomatic period, showing slit ventricles and tight basal cisterns (a,b). Three months later, CT scan showed ventricles and cisterns of normal size (c,d).**



**2.2. Case of a 3 years old girl with SVS, who developed brain edema after removal of the VP-Shunt system:**

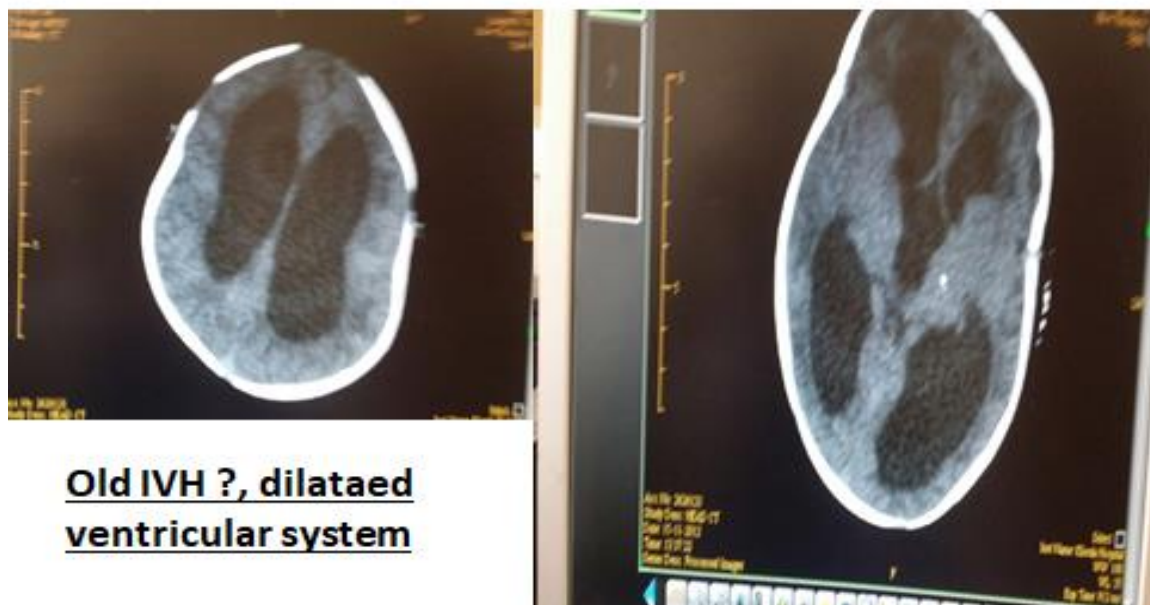
The 3 years old female patient had a history increasing size of head, for what the patient was admitted with the age of 2 months. Patient underwent CT brain, which revealed hydrocephalus.

CSF diversion device in form of a ventriculoperitoneal shunt system with a medium pressure valve was inserted. Patient was admitted again after and he underwent readjustment of the ventricular catheter. The F/U postoperative imaging showed features of slit ventricle syndrome. Patient was admitted multiple times because of recurrent headache and vomiting. Her condition used to improve with hydration and analgesics. MRI brain with MRA and MRV was performed and it was reported normal. Patient was re-admitted again with the age of three years with the symptoms of severe headache and repeated vomiting. O/E there was no CNS deficit. CT scan of brain showed features of slit ventricle syndrome with correct localization of the right parietal intraventricular VP shunt catheter. The peripheral VP shunt catheter was tied at the neck for 7 days and the patient’s clinical and neurological condition was remaining stable. MRI of brain was done and it revealed no hydrocephalus and no periventricular lucency. VP shunt was removed totally. The perioperative course was uneventful. Postoperative CT scan of brain was satisfactory. Patient was kept under close observation. His clinical condition deteriorated in the night after the last operation and then she was shifted to ICU.

Patient was intubated and ventilated, pupils were 4 mm and bilaterally non reactive to light. Immediately, EVD was inserted right parietal and the CT scan of brain showed severe global brain edema. After there was no clinical improvement, patient was taken immediately to Operation Room and decompressive craniectomy was done left fronto-temporo-parietal. After the last operation, patient’s condition remained critical. The last CT scan of brain was showing severe intracranial hypodensities with compressed brain stem in foramen magnum and midline shift to left side with severe bulging of brain through the trepanation defect.

Neurosurgically, maximal ICU care was provided to stabilize the vital functions of the patient. She was for longer period of time on inotropic support and died 2 months after the last operation. (See the images below).

**Prae-operative CT scan**



**Figure 16: Preoperative CT scan before insertion of the VP shunt system**

## Slit Ventricle Syndrome (I)

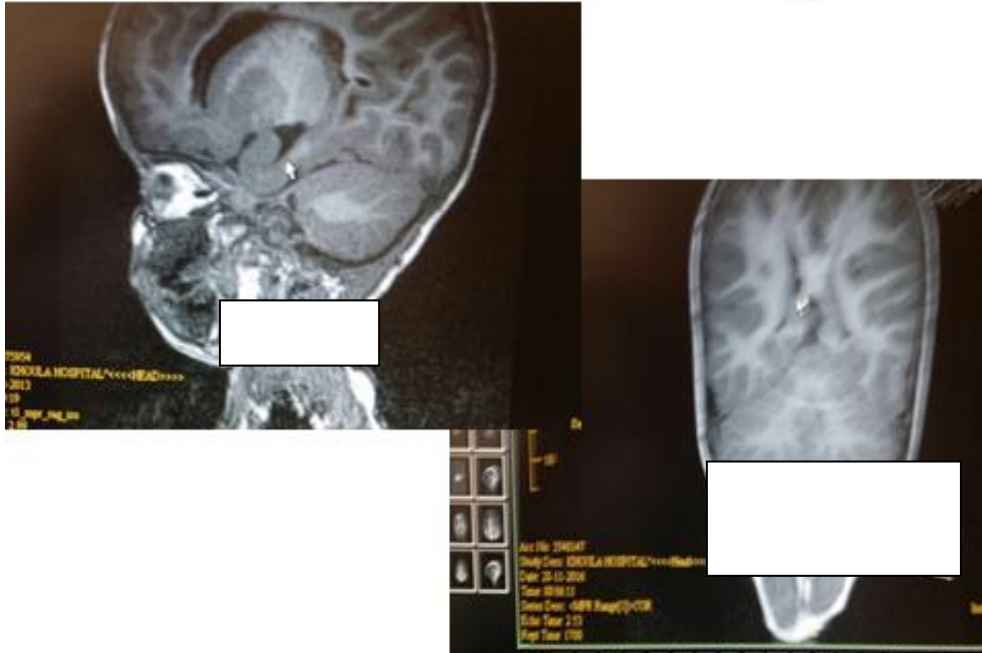


Figure 17: SVS after 2 years with VP shunt.

## **CT before last operation (Left sided Decompressive Hemicraniectomy)**

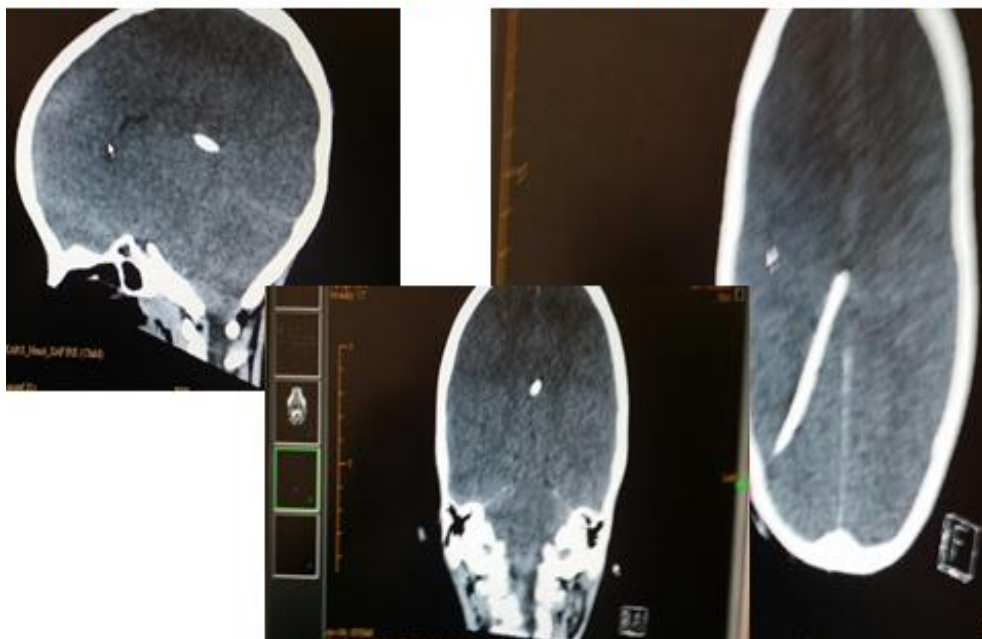


Figure 18: Severe brain edema after removal of the VP shunt system. Right parietal EVD was then inserted after deterioration of the clinical condition of the patient.

# Decompressive Craniectomy

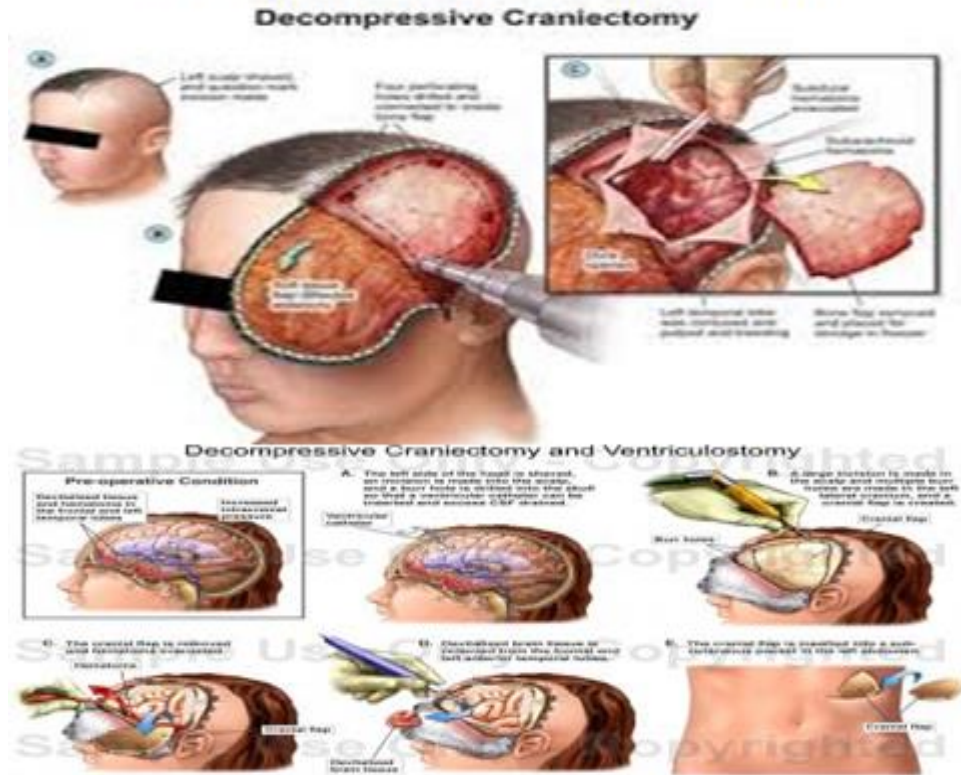


Figure 19: Decompressive craniectomy left fronto-temporo-parietal was performed as last operative procedure.

## Last CT scan



Figure 20: Postoperative CT after the decompressive craniectomy.

### 3. DISCUSSION

The clinical picture of spontaneous intracranial hypotension or spontaneous aliquorrhoea syndrome has been well known for more than 40 years but the cause of this syndrome, unlike the clinically identical post-lumbar puncture headache, has been speculative. Headache following lumbar puncture has been generally assumed to result from leakage of CSF through the puncture-hole in the dura causing CSF hypotension and distension of the pain sensitive structures at the base of the brain. The leakage of CSF through the puncture hole in dura has been proved by isotope studies and surgical exploration.

Schaltenbrand and Wolf P have reasoned that the spontaneous aliquorrhoea syndrome may result from either decreased CSF production (from an unexplained disturbance of chorioid plexus function) or enhanced CSF resorption. The latter seems improbable. Posture dependent headache following minor trauma such as a fall on the buttocks has been claimed to be due to a dural tear with subsequent leakage of CSF.

A purely spontaneous tear in the dura seems unlikely because of its strong texture. In earlier reports the most promising therapeutic results were achieved by hypotonic saline iv infusions. In headache after lumbar puncture fludrocortisone has been advocated, probably because of its sodium retaining property. But fludrocortisone had no effect on the symptoms of SVS, perhaps because of increase in brain swelling. The immediate therapeutic response to dexamethasone seems logical, if in the CT findings indicated the presence of diffuse brain oedema.

If post-lumbar puncture headache is also associated with brain oedema, dexamethasone might be worth a therapeutic trial in that condition.

Shunt obstruction (failure) is the usual presumptive diagnosis when a child with a shunt develops symptoms of increased ICP. Imaging studies and shunt tapping may be the next step for the diagnosis of shunt obstruction. However, the background etiology of recurrent shunt failures is frequently unnoticed in clinical practice; slit ventricle syndrome is an example of this. Symptoms of shunt obstruction, slit-like ventricles in imaging studies, and slow filling of the shunt reservoir are the classical triad of slit ventricle syndrome [4]. Nonetheless, the intermittent nature of symptoms, subtle changes in ventricle size, and inexperience of physicians often lead to the misdiagnosis of simple shunt obstruction. Weinzwieg et al. [2] reported that an average of 4.9 shunt revisions are performed in children with slit ventricle syndrome and secondary craniosynostosis until they receive cranial vault expansion.

The utmost importance of serial HC measurement in pediatric neurosurgical practices is very important for monitoring of patients with hydrocephalus, especially if a VP shunt system is inserted. If the patient's HC decreased far more than expected after shunt surgery and remained extremely small, then a proper neurosurgical assessment might be indicated.

Looking carefully at the HC curve may lead to questions about and appropriate evaluation of possibility of occurrence of secondary craniosynostosis in some patients.

Postoperative cerebral edema can occur in children with slit ventricles without a logic explanation. The authors Elizabeth A. Eldredge, Mark A. Rockoff, Michael D. Medlock, R. Michael Scott, Michael B. Millis, Pediatrics, April 1997, VOLUME 99 / ISSUE 4 described postoperative Cerebral Edema Occurring in Children With Slit Ventricles, who developed symptomatic cerebral edema after uncomplicated orthopedic surgery. One had a fatal outcome. Hyponatremia occurring in the postoperative period was the likely precipitating factor. Recommendations for recognizing, treating, and potentially preventing this catastrophic complication are discussed.

#### 3.1. Goals of management:

As stated, 4% of humans have chronic daily headaches. Neurosurgeons need to do what ever is possible to "normalize" the dynamics of ICP. Medical management is complicated and frustrating and requires the cooperation of neurologists, internists, and mental health specialists to improve the quality of life for patients with normal ICP dynamics. A review of the management of such headaches, which must be individualized, is beyond the scope of this discussion. Based on reviews of management strategies that treat such headaches, however, narcotic medication must be avoided if possible. Narcotics are widely recognized as leading to the well-defined entity classified as "medication overuse headaches." How does the physician decide that all has been done to "normalize" ICP dynamics? In normal patients, all CSF compartments, including the ventricles, spinal subarachnoid, cortical subarachnoid spaces and basal cisterns, freely communicate with each other. When multiple CSF compartments are isolated from each other, rapid changes in the dynamics of these

compartments occur. In response, the brain, which is a viscoelastic substance, shifts. Even if all the compartments are shunted, this rapid shifting can occur unless all CSF catheters are spliced together proximal to the valve mechanism. Such rapid shifts at the time of Valsalva maneuvers or sudden positional changes cause distortion of the pain sensitive intracranial structures such as the basal dura. Ventricular shunting in the context of very large ventricles is also problematic. It is difficult to drain the cortical subarachnoid spaces because retrograde flow through the foramen of Monro is restricted [27].

### 3.2. General aspects of management:

The role of neurosurgeons in the management of shunt related headaches or SVS must be to ensure that ICP dynamics are normalized as much as possible. Doing so requires ascertaining that all CSF compartments communicate without resistance or obstruction.

Neuroendoscopic procedures can be used to fenestrate membranes that lead to compartmentalization or by splicing multiple ventricular or subarachnoid space catheters proximal to a single programmable valve containing a DRS. The final step is to monitor ICP over time to ascertain that ICP remains normal during all positions and during sleep.

What should be done if patients return complaining of re-exacerbation of their headaches? First, the above shunt system must be identified as working as planned. If the ventricles do not expand at the time of shunt failure, which is the likely scenario in these cases, performing CT scans is futile. If it is essential to image the brain, rapid sequence MRIs are preferred. Typically, these patients have undergone many CT scans. At best, patients' risk of cataracts increases. At worst, there is a theoretical possibility that the risk of developing induced malignancies is increased over the ensuing decades. In NVH patients with worsening symptoms, a reservoir tap is placed. ICP is measured while the patient is recumbent. The patient is asked to sit up to ensure that their ICP falls to a maximum of 5 cm H<sub>2</sub>O or lower. At this point about 5 cc of Iohexal is injected. The patient is scanned within an hour. The cortical subarachnoid and spinal subarachnoid spaces of the upper cervical spine are analyzed carefully. If there are no obstructions, the system is working as hoped, and the headaches are unrelated to the ICP.

These patients can then be managed medically with the full understanding that all pressure manipulations that can be done have been done. These systems would greatly benefit by the incorporation of a telemetered monitoring device that allows the accurate noninvasive measurement of ICP in all positions.

## 4. CONCLUSION

Hydrocephalus is a lifelong condition, and decisions made for the patients, especially in infancy can affect the quality of patients' lives as they age. Headaches in patients with shunts do not necessarily mean that the shunt has failed. They may mean that patients are experiencing extremely abnormal cerebrospinal fluid pressure dynamics. However, new technological advances may have the ability to normalize intracranial dynamics to help more of these patients to live more normal lives. Full understanding of the cerebrospinal fluid dynamics will lead to a more rational approach to the management of headaches in this challenging patient population.

A ventriculoperitoneal (VP) shunt is effective for hydrocephalus. However, the shunt may malfunction due to obstruction, breakage, migration, or infection. Slit ventricle syndrome (SVS) is a rare symptomatic condition presenting with neuroimaging findings of small ventricles in a patient with a VP shunt. Accurate diagnosis can be difficult and adequate treatment may be delayed because neuroimaging of small ventricles can be misinterpreted as a properly working shunt.

SVS can present with visual symptoms occurring in the absence of other neurologic symptoms in pediatric patient. Development of visual symptoms in patients with VP shunts necessitates close follow-up and neurologic work-up to evaluate intracranial pressure and to proceed possibly with operative management.

General clinical, neurological and radiological aspects of the patients with SVS have to be investigated, if the complaints are increasing or becoming persistent; such as blood investigations in the laboratory for electrolytes and other parameters and chest / abdominal problems.

The most frequently used imaging study in the immediate postoperative period is CT, due to fast acquisition and availability in the urgency. On the majority of occasions, CT provides enough information for a correct handling.

As ventricular shunting is the most widely accepted treatment of hydrocephalus, it is important for radiologists to be familiar with the normal imaging findings, the signs of intraextracranial complications and the role of the different imaging modalities to assess them. Although patient history and physical examination provide an initial suspicion of a shunt failure, medical imaging often confirms the diagnosis and reveals the underlying cause. Understanding of the specific types of obstructive shunt malfunction, potential causes of failure, and management options is important for neurologists and pediatricians who often are the first providers to evaluate and triage these often markedly ill patients. Radiologists have a variety of procedures they can use to discover the cause of the malfunction or complication. Communication with the treating clinicians is essential for tailoring the diagnostic imaging work-up to the particular problem.

Every shunt failure is a chance to test shunt dependency, whereas most patients with communicating hydrocephalus are candidates for ETV.

At least 70% of shunted patients may be candidates for shunt removal. Communicating hydrocephalus is a misnomer and previously shunted patients are excellent candidates for ETV.

Rekate's Rules Of Problematic Shunt Management are to be considered:

- Make certain shunt is really needed.
- Attempt shunt removal.
- If remains shunt dependent make certain that all CSF compartments see the same pressure either internally or externally.
- Make certain ICPs 5-15 mmHg recumbant and -5 to +5 mmHg standing.

"If it aint broke don't fix it" but the neurosurgeons and pediatricians have to make certain that they have a plan for the next step.

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