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Slit Ventricle Syndrome in Children : Clinical Presentation and Treatment

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= Abstract =

소아에서 틈새뇌실 증후군 : 임상 양상 및 치료

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The term slit ventricle syndrome(SVS) refers to an episodic occurrence of headache, vomiting, and possibly some degree of impaired consciousness in shunted hydrocephalic children in whom slit-like ventricles are seen on CT scan or MRI. Authors present 6 cases with SVS who were treated at our institute for last 10 years.

From 1986 to 1996, 821 patients underwent shunt surgery for hydrocephalus. The etiology of hydrocephalus included brain tumor(140 patients), post-hemorrhagic(62 patients), idiopathic normotensive hydrocephalus(64 patients), post-meningitic(58 patients), post-traumatic(54 patients), congenital(48 patients), neurocysticercosis(31 patients), and unknown etiology(364 patients). During the mean follow-up duration of 68 months, 232 shunt revisions were performed by a revision rate of 1.28 per patient. The incidence of SVS was 0.7%(6 patients).

Most of them have been operated on in infancy. Time interval from the first operation to the development of slit-ventricle syndrome ranged from 4 to 8 years, the mean was 6 years. Shuntogram showed patent shunt in all patients. Two patients with less severe clinical symptoms improved with conservative treatment. These patients were not measured ICP because of good hospital course. One patient showed high ICP and needed only revision with same pressure valve as previous shunt. Low ICP was noted in 3 patients. Pressure augmentation using an anti-siphon device(ASD) or upgrading valve system were necessary in these patients.

The authors stress that determining type of SVS is the first step in treatment planning and that the best treatment is a strategy aimed at resolving the specific type of SVS responsible for the symptoms.

KEY WORDS : Slit ventricle syndrome · ICP monitoring · Overdrainage · Shuntogram · Shunt malfunction.

Introduction

Complications of shunt due to overdrainage are poorly defined and understood and therefore, the treatment has been to subject of confusing literature. The slit ventricle syndrome(SVS) is regarded as one form of overdrainage problem in shunted children and defined as a symptom complex suggestive of increased intracranial pressure with slit-like appearance of ventricular system on a CT scan or MRI.

The incidence of SVS is not well documented because of the variable way it has been defined but it is probably 3-5%. The clinical characteristics of the SVS differ so widely as to suggest that different clinical conditions are actually being described.

Materials and methods

From 1986 to 1996, 861 patients underwent shunt sur-

gery for hydrocephalus. The etiology of hydrocephalus were brain tumor(140 patients), post-hemorrhagic(62 patients), idiopathic normotensive hydrocephalus(64 patients), post-meningitic(58 patients), post-traumatic(54 patients), congenital(48 patients) and unknown etiology(364 patients). During the mean follow-up period of 68 months, shunt revisions were performed in 232 cases. Among these 232 revisions 43(19%) were the elective lengthening of distal catheter. Proximal obstruction(42.7%) was more frequent than distal obstruction(35.3%). Slit ventricle syndrome was developed in 6 cases(0.7%) in our series. Treatment in each patient included insertion of anti-siphon device in 3 patients, conservative treatment in 2 patients and revision of same pressure valve in 1 patient. All patients were followed for over 2 years with brain CT or MRI. The medical records, the results of shuntogram and ICP monitoring, and radiographic studies of these patients were reviewed.

Results

The symptoms of SVS consisted of intermittent episodes of headache, vomiting and lethargy and occasionally some neurological signs such as upward gaze paresis and ataxia. No one progressed to unconscious state. All patients showed sluggish or absent refill of pumping device and collapsed ventricle on brain CT or MRI.

All of them have been operated on in infancy(the age at the time of initial shunt ranged from 5 to 14 months). Time interval from the initial shunt operation to the development of slit-ventricle syndrome ranged from 4 to 8 years, the mean duration was 6 years. Shuntogram showed patent shunt function in all patients. The pressure types of initially inserted shunt valves were low in 4 patients and medium in 2 patients(Table 1).

ICP monitoring was performed in 4 patients. ICP monitoring was done through the reservior or LP drainage catheter for 24 hours. One patient showed high ICP and needed only revision with same pressure valve as previous shunt system and then this patient improved. We thought that his clinical symptoms could result from partial obstruction of proximal shunt catheter in the slit ventricle even if shuntogram was interpreted as normal. Low ICP was noted in 3 patients. Pressure augmentation using an ASD or upgrading valve system were necessary in these patients. Two patients with less severe clinical symptoms improved with conservative treatment. These patients were not checked ICP because of good hospital course. We thought that the symptoms of them should be differentiated from that of migraine in these patients.

Discussion

1. Clinical manifestation and incidence

The 'slit ventricle syndrome' was first described by Becker and Nulsen, who in the early 1960s pointed out the possibility of inadequate drainage secondary to small ventricles in shunted hydrocephalic subjects[1]. This syndrome is still a rather poorly understood clinical entity in spite of its wide acceptance in the medical literature and the numerous reports. The syndrome consists of recurrent episodes of headache, malaise, vomiting and varying degrees of impairment of consciousness. However, the complete SVS is not based entirely on clinical symptoms and neurologic signs. It must be accompanied by a sluggish or absent refill of the pumping device and radiological evidence of a slit-like ventricle on the side of the shunt. The demonstration of slit-like lateral ventricles with CT scan or MR imaging, is a mandatory, though not sufficient finding for a diagnosis of SVS. The diminished size of the lateral ventricles is a relatively common observation in hydrocephalic children harboring shunt devices; this was seen in as many as 50 - 60% of the cases in some series reported in literature[29)30)]. Most of the patients with slit-like ventricles do not develop any kind of clinical manifestation during life. On the other hand, the symptoma-

Table 1. Summary of patients with slit ventricle syndrome

Case	Age		Valve pressure	Shuntogram	ICP monitoring	Treatment
	Initial shunt	SVS				
1	6 mon.	4 yr	Low	Patent	Low	Valve upgrade
2	8 mon.	5 yr	Low	Patent	Low	Valve upgrade
3	5 mon.	5 yr	Low	Patent	Low	Valve upgrade
4	9 mon.	7 yr	Medium	Patent	High	Revision with same pressure valve
5	12 mon.	6 yr	Low	Patent	Not checked	Conservative
6	14 mon.	8 yr	Medium	Patent	Not checked	Conservative

tology regarded as typical of SVS is also found in patients with normal or even large cerebral lateral ventricles[10)21)26)].

The time interval from the original shunt and the age of the patient are also helpful on making a diagnosis. SVS typically arises after several months or years of adequate functioning of the CSF shunt device[4)21)24)]. All patients in this paper had variable latent period, 4 to 8 years from shunt to the onset of SVS symptoms. Such a long time interval has led some authors to classify the syndrome as one of the complications of chronic CSF overdrainage[11)22)24)]. However, the headache associated with SVS differs from that accompanying intracranial hypotension : it is not related to posture and it is not relieved by recumbence. The headache due to chronic intracranial hypotension is not generally accompanied by malaise, vomiting or signs of neurological dysfunction. The intermittent character of the symptoms is also significant in distinguishing SVS from the symptoms of intracranial hypotension.

The diagnosis of SVS should be avoided and substituted by a correct diagnosis of disturbances unrelated to the CSF shunting therapy, etc., childhood migraine. Headache, vomiting and altered sensorium can be seen in childhood migraine as well as in patients with SVS. A majority of children with migraine has a family history of migraine and response to propranolol or verapamil therapy[16)].

The actual incidence of SVS is not known. In literature, the reported frequency of the syndrome varies surprisingly widely in different articles(Table 2), with values ranging from 1 to 24%, and even to 37%[2)4)6)17)21)24)].

The incidence of SVS does not seem to be related to the etiology of the hydrocephalus: in most reports it appears to be associated with all the various types of hydrocephalus, such as malformative, post-hemorrhagic, post-infective, and post-traumatic hydrocephalus[4)13)15)21)24)]. Some series show a higher risk for this kind of complication in patients with post-hemorrhagic or post-infective hydrocephalus[2)4)]. However, we could not find any etiologic factors related to the incidence of SVS.

Other factors, such as the number of previous CSF shunting surgical procedures or the characteristics of the valve system utilized for treating the hydrocephalus, do not show any demonstrable correlation with the occurrence of the syndrome(Table 3). Our series strongly suggested that low pressure valves would be responsible for the occurrence of SVS. Even though other authors have also found a higher incidence in patients originally treated with low pressure valves[2)14)15)], some have observed the complications in subjects operated on with medium pressure or even high pressure valves[4)6)30)].

The age distribution of SVS presents a peak in incidence in the first decade of life, with most authors indicating a preferential age of diagnosis between 4 and 7 years[2)4)6)21)24)]. The syndrome is definitely rare after the second decade of life. All of patients in our series have been operated on in infancy. Their ages at the onset of SVS symptoms was below 10 years.

Table 2. Incidence of slit ventricle syndrome

Authors	Period(years)	No. of shunted patients	No. of SVS	%
Benzel et al.(2)	5	92	22	24
Choux & genitori(4)	18	?	28	0.9
DiRocco(6)	17	625	6	0.9
Keucher & mealey(16)	12	228	3	1.3
Mclaurin & olivi(20)	7	?	15	3.5
Oi & matsumoto(23)	?	164	5	3
Choi & kim	10	821	6	0.7

Table 3. Opening valve pressure in slit ventricle syndrome

Authors	Opening pressure(No. of patients)				Total No. of patients
	Low	Medium	High	Unknown	
Benzel et al.(2)	20	2			22
Choux & genitori(4)	6	21		1	28
DiRocco(6)		6			6
Hyde-rowen et al.(15)	6				6
Choi & kim	4	2			6

2. Pathogenesis

The results of laboratory investigations, such as tests for evaluating shunt function and ICP recordings, appear to vary considerably. However, SVS is basically due to an overdrainage of CSF leading to collapse of the cerebral ventricles. It is recognized that in the normal individual a negative intraventricular pressure, approximately -70mm H2O, exists in the upright posture. In the shunted person this negative pressure is increased several fold as a consequence of siphon effect by the hydrostatic column of shunt catheter.

Serlo et al. stated that blockage of the ventricular catheter causes cerebral edema and subarachnoid accumulation of CSF which is usually accompanied by signs of increased intracranial pressure[29)30)]. There appears to be no evidence to support this explanation. Some authors affirm that the symptomatology of the syndrome depends on transitory phases of increased intracranial pressure[5)8)12)18)]. These phases of intracranial hypertension are thought to occur because of ventricular catheter trapping between the abutting ventricular walls, which causes transient obstructions of the catheter itself. Transitory enlargement of the cerebral ventricle during the acute phase of the syndrome is thought to be an indirect evidence of a transitory malfunction of the shunt system[10)]. However, the hypothesis of transitory occlusion of the ventricular catheter causing an intermittent shunt malfunction has never been demonstrated directly[9)12)14)19)26)30)]. When tested with radionuclide shuntogram, the CSF shunts in subjects with SVS interpreted as an intermittent CSF shunt malfunction have never been found to be patent[8)].

In facts, the reports describing the condition of the shunt device for the pathogenetic interpretation of the syndrome differ considerably : the shunt has been found to be patent[10)26)], occluded either at the proximal[11)15)] or at the distal end[7)], or only partially occluded[19)]. Such variability has led to a demand for a change in the pathogenetic interpretation of the syndrome in subsequent reports and has justified attempts to classify patients in various groups according to the functioning of the CSF shunt devices. Epstein et al. have proposed that children with SVS and intermittent shunt malfunction, who can be treated by simple revision of shunt, be differentiated from children with increased ICP and patent shunt, who would need cranial expansion[10)]. Recently Rekate[24)] classified SVS into five distinct syndrome by ICP monitoring : 1) intermittent low pressure headache, 2) intermittent proximal obstruction, 3) shunt failure with small ventricle(normal volume hydrocephalus), 4) intracranial hypertension with working shunts(hydrocephalic pseudotumor), 5) headache unrelated to shunt function. 1) - 3) can be treated with shunt revision or upgrading and 4) with cranial expansion.

We performed ICP monitoring for over 24 hours in 4 patients. One patient showed high ICP and 3 patients low ICP. We found that the proximal shunt catheter of patient with high ICP was partially obstructed in spite of normal shuntogram. He was well after only shunt revision with changing proximal catheter and the same pressure valve as that of previous shunt. The clinical symptoms of 3 patients with low ICP could be resulted from overdrainage of CSF and intermittent obstruction of proximal shunt catheter. Pressure argumentation could be a treatment of choice in these cases.

The concept of rigid ventricular walls as the pathogenetic mechanism accounting for persistence of slit-like cerebral ventricles leading to increased intraventricular pressures has been widely accepted[23)30)]. Oi and Matsumoto noted dilatation of cortical veins and Virchow-Robin spaces, destruction and disorganization of the ependyma, gliosis in the subependymal area and decreased CSF edema in the subependymal white matter in the morphological study of a post-shunt slit ventricle in dogs[23)]. Rekate et al. detected high brain elastance in the case of SVS characterized by high intraventricular pulse pressures not associated with intracranial hypertension[28)]. They pointed out that the reduced ability of the cerebral ventricle to expand would take into account the possibility of a high brain turgor, mainly related to compression by the intracranial venous pressure.

3. Investigation and the treatment options of slit ventricle syndrome

The treatment of SVS has also been the subject of varying recommendations. Probably the first report dealing with slit-like ventricles in the literature was that of Yelin and Ehni[31)]. They described insertion of a red rubber catheter around the ventricular catheter, via a transcallosal approach, in order to prevent blockage of the ventricular catheter by the collapsed ventricle.

Portnoy et al. in 1973[25)] described the ASD which prevents flow of CSF through the shunt whenever a negative pressure of 100 - 200mm H2O is exerted, at the outlet of the device. Epstein et al.[9)] advocated a subtemporal craniectomy to decompress the intracranial contents, to permit dilatation

of the ventricles, and to provide a means of assessing intracranial pressure by palpation. This procedure was also recommended by Holness et al.[14]. It is significant, however, that 10 of the 22 patients had post-craniectomy shunt revisions and the revisions were for ventricular end obstruction in 8 of them.

The CT scan or MRI is the paramount neurodiagnostic examination. It is essential that the study should be performed during a symptomatic period and compared to a baseline scan made when asymptomatic. Any increase in previous slit ventricles may be an only objective sign of intermittent shunt obstruction. If the ventricles size unchanged, a radionuclide study(shuntogram) for the evaluation of shunt patency is performed : partially occluded catheters will usually demonstrate a prolonged clearance time. These catheters may be unable to increase the rate of CSF drainage in response to increase in intracranial volume and result in high ICP. ICP monitoring is reserved for those children with normal shunt patency and slit ventricle in radiological studies. If ICP showed low, upgrading of the CSF shunt resistance or incorporation of an ASD into the existing CSF shunt system is a choice of treatment. This option is based on the hypothesis that the cause of SVS is an intermittent obstruction of the ventricular catheter trapped between the abutting ventricular walls. The rationale of these operations is that they favor an increase in volume of the ventricle harboring the shunt catheter by augmenting its inner pressure, thus freeing the catheter and reducing the risk of further obstruction. In fact, some degree of enlargement of the cerebral ventricle following the placement of higher opening pressure valve or an ASD has been described in a few reports[12)15)]. However, such effectiveness of ASD has not been well understood in the majority of cases, because disappearance of their symptoms has not been accompanied by obvious modifications of the volume of the cerebral ventricles[2)21)30)].

ICP monitoring is necessary for at least 24 hours either through subarachnoid screw or directly into the shunt reservoir usually with a portable monitoring that permits ambulation and simultaneous pressure recording with postural alterations. In children with paroxysms of increased ICP and normal shunt function, cranial expansion is the preferred treatment. Subtemporal craniectomy was introduced with the main goal of 'freeing' the trapped ventricular catheter by allowing the ventricular wall to expand towards the area of diminished resistance. The first reported positive results obtained with this type of operation have been attributed to some degree of postoperative ventricular enlargement[15)]. However, other authors have noted a decreased volume of the ventricular system following subtemporal craniectomy, with reduction more prominent on the side of the ventricle harboring the draining catheter[3)20)]. They postulated that a decrease in the ventricular pulse pressure by this type of operation is the main factor in the disappearance of the clinical symptomatology. Other authors carried out a craniotomy with morcellation of the posterior calvarium from the coronal suture to the inion as well as laterally to the squamosal suture[10)]. They pointed out that posterior calvariotomy would provide more extensive expansion of intracranial volume than subtemporal craniectomy do. This type of operations aims at increasing craniocerebral compliance. In one of our patients who showed high ICP, shuntogram was interpreted as normal. But we found out patial obstruction of proximal catheter at time of revision. This patient with simply changing proximal catheter. In our small series we have no experience on cranial expansion. In cases of SVS with high ICP, patency of shunt system have to be investigated carefully before deciding calvarial expansion. Nowadays, the treatment protocol for SVS was Fig. 1.

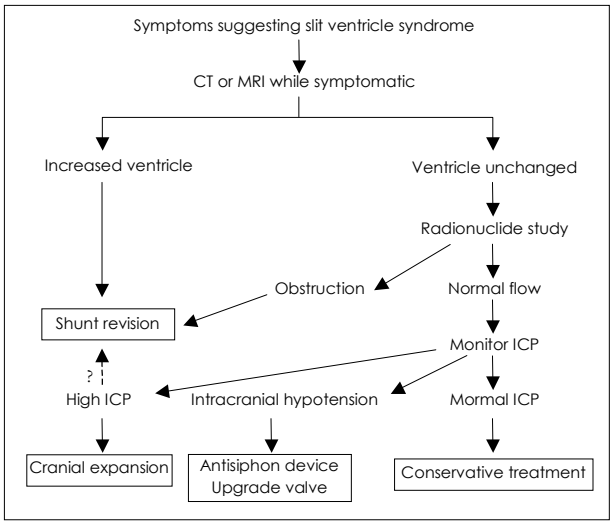

Fig. 1. Algorithm for treatment of slit ventricle syndrome.

Other important point in SVS is that initial shunt operation should be designed for the restoration of the CSF circulation or correction of the impaired CSF absorption as well as trying to decrease the risk of shunt malfunction by favoring the diversion of CSF to an extra-ventricular level (lumboperitoneal shunt, endoscopic third ventriculostomy[27)]).

All these operations have been so rarely utilized in patients with SVS that reliable evaluation of their efficacy is not yet possible. We hesitate to perform lumboperitoneal shunt in infant with unstable and growing spine. However, we believe that endoscopic third ventriculostomy or combination of endoscopic procedure and shunt is simple and effective method to avoid the shunt dependency and the overdrainage of CSF in hydrocephalus of infant with the presence of a dilated the third ventricle and to decrease the risk of shunt malfunction.

Conclusion

Slit ventricle syndrome is not a pathological entity, and is distinguishable from low-pressure headaches and from irreversible shunt obstruction. The pathogenesis of SVS is not entirely clear but various suggestions have been proposed such as an intermittent shunt obstruction due to collapsed ventricles etc. Determining the type of SVS by investigation is important to select the treatment. The placement of ASD or upgrading the valve resistance was effective in relieving the symptoms of SVS in most patients of our series.

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소아에서 틈새뇌실증후군 : 임상양상 및 치료

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= 국문초록 =

틈새뇌실증후군은 수두증으로 션트술을 시행받은 환아에서 CT나 MRI소견상 틈새처럼 좁은 뇌실을 가지고 있으면서 간헐적으로 두통, 구토 또는 의식장애등이 동반되는 질환이다.

1986년부터 1996년까지 총 821명의 환자가 수두증으로 션트술을 시행받았다. 이 환자들의 수두증의 원인은 뇌종양(140명), 출혈(62명), 정상압수두증(64명), 뇌수막염(58명), 외상(54명), 선천성(48명), 뇌낭미충증(31명), 그리고 이유를 알 수 없었던 경우가 364명으로 나타났다. 평균 추적 관찰 기간은 68개월이었으며 총 232명에서 재수술을 시행하였으며 1인당 1.28회의 재수술율을 보였다. 이들중 틈새뇌실 증후군은 6예로 0.7%였다.

틈새뇌실증후군 대부분의 환자는 영아기에 수술을 시행받았다. 처음 수술로부터 틈새뇌실증후군이 발생하기까지의 기간은 4~8년으로 평균 6년후 틈새뇌실증후군이 발생하였다. 동위원소 검사상 6예에서 션트의 기능은 모두 정상이었다. 증상이 경미한 2명의 환자에서는 보존적 치료를 시행하였는데 치료후 증상의 호전을 보였다. 이 환자들은 뇌압 측정을 시행하지 않았다. 6명중 1명은 뇌압 측정시 높게 나타나 기존과 같은 압력밸브를 이용하여 재수술을 시행하였다. 3명의 환자는 낮은 뇌압을 보여 안티사이펀밸브나 기존보다 높은 압력의 밸브를 이용하여 재수술을 시행하였다.

틈새뇌실증후군의 치료는 첫째, 증후군의 양상을 정확하게 판단하여, 둘째, 증상을 완화시킬 수 있는 방향으로 치료방침을 설정해야 할 것으로 생각되었다.

중심 단어 : 틈새뇌실증후군 · 뇌압 · 션트 · 션트기능부전.