Childs Nerv Syst
DOI 10.1007/s00381-010-1092-3

ORIGINAL PAPER

Factors influencing spinal canal stenosis in patients with long-term controlled hydrocephalus treated with cerebrospinal fluid shunt

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Received: 28 December 2009 / Accepted: 26 January 2010

Abstract

Purpose Spinal canal stenosis (CS) occurs in patients with hydrocephalus who are treated with cerebrospinal fluid (CSF) shunting. The pathophysiology of CS comprises CSF overdrainage. We analyzed the incidence of CS and the factors causing it.

Methods Thirty-three patients who underwent ventriculoperitoneal shunt during childhood visited the Outpatient Department in Yamaguchi University Hospital in 2006. Diameters of spinal canal at C_4 were measured. Treatment procedure, age, and type of hydrocephalus in the patients with CS were compared with those without CS.

Results Of the 33 patients, 10 (30.3%) presented CS, and two (6.1%) were symptomatic. A low-pressure valve caused CS with a significantly higher incidence than a medium- or high-pressure valve (60.0% vs. 17.4%, $P<0.05$). Although the difference was not significant, the average age of shunt insertion for a patient with CS was slightly less (0.87±0.99) than for a patient without CS (1.63±1.58). No differences in the CS incidence were observed between obstructive and communicating hydrocephalus.

Conclusion In order to prevent CS, the hydrocephalus should be appropriately controlled by using a medium- or high-pressure valve until the diameter of the spinal canal reaches the required level. Adjustment of the programmable valve with the patient's growth should be ideal.

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Keywords Hydrocephalus · CSF shunt · Overdrainage · Spinal canal stenosis · Myelopathy

Introduction

Intracranial hypotension caused by cerebrospinal fluid (CSF) overdrainage is one of the major complications in CSF shunt surgery. Orthostatic headache is a representative symptom of intracranial hypotension. When the overdrainage persists, slit ventricle with or without slit ventricle syndrome, subdural hygroma, or hematoma appears. Bony changes such as thickened cranium, microcranium, and spinal canal stenosis (CS) also present as chronic signs of intracranial hypotension. The relationship between CS and ventriculoperitoneal (VP) shunting has already been described in 1983 [4]. Previously, nonprogrammable pressure regulation valves were used. Programmable valves were introduced in the 1990s, and they have enabled the easy control of the hydrocephalus.

In the present study, we analyzed the incidence of spinal CS and factors causing CS in patients with hydrocephalus who are treated by CSF shunting. We also compared the characteristics of CS and slit ventricle, which is another overdrainage syndrome.

Materials and methods

Patients

Forty-three patients (24 men and 19 women) who underwent VP shunting for hydrocephalus during childhood visited the Outpatient Department in Yamaguchi University

Hospital in 2006. The shunting of the 43 patients was done between 1966 and 2005. Shunt insertion was performed between 1 week and 4 years after birth. The follow-up periods were 4 months to 41 years. Patients with asymptomatic shunt obstruction were excluded from the study, and those who suffered shunt malfunction and underwent immediate shunt revision were included unless the valve settings were changed. The types of hydrocephalus are listed in Table 1. We diagnosed the etiology as 11 cases of obstructive and 22 cases of communicating hydrocephalus; however, 10 cases could not be classified as either obstructive or communicating hydrocephalus.

Definition of canal stenosis

The antero-posterior (AP) diameter, which is defined as the distance from the spinolaminar line to the posterior marginal line, was measured at the narrowest point in the lateral view of the cervical X-ray film. The normal value of the AP diameter is 17±5 mm. For adults, spinal CS is diagnosed with certainty if the canal diameter is 12 mm or less, and the diagnosis is doubtful if the canal diameter is 15 mm. In the present study, CS in children was diagnosed when the diameter was below 90-percentile on the growth chart [1, 2]. Of the 43 patients, 10 were under 3 years of age and were excluded in the statistical analysis because the X-ray film evaluation is inaccurate for the infants, and there is no data available regarding the standard development curve of the canal diameter until 3 years of age.

Table 1 Number of the patients according to the etiology of the hydrocephalus

Obstructive hydrocephalus	11	(10)
Aqueductal stenosis	5	(4)
Brain tumor	4	(4)
Arachnoid cyst	1	(1)
Trauma	1	(1)
Communicating hydrocephalus	22	(13)
Malformation	11	(5)
Intraventricular hemorrhage	6	(4)
Arachnoid cyst	3	(3)
Meningitis	1	(1)
Trauma	1	(0)
Indistinguishable of the communication	10	(10)
Malformation	5	(5)
Arachnoid cyst	5	(5)
Total	43	(33)

The numbers in the brackets indicate the numbers of the patients over 3 years of age at follow up

Factor analysis affecting canal diameter

The shunt pressure setting, age of shunt insertion, and etiology of hydrocephalus were selected as factors that may affect canal diameter. The patients were classified into low- and medium/high-pressure groups based on the valves used. For the Hakim and Sophy programmable valve users, those with valve settings below 70 mmH_2O were included in the low-pressure group. For the Strata valve users, those with valve settings below the 1.5 level were included in the low-pressure group. The programmable valve pressures were adjusted within a month after the surgery. The patients whose settings were changed several times were excluded from the study.

These factors that may cause CS were also applied to analyze the patients with slit ventricle. Out of the 33 patients, 11 presented with slit ventricle; the width of the portion in which the ventricular catheter was placed was less than 5 mm. No patient presented the slit ventricle syndrome.

Statistical analyses were performed by the chi-square or Mann–Whitney *U* test. A *P* value of less than 0.05 was defined as a significant difference.

Results

Incidence of canal stenosis

Figure 1 illustrates the canal diameters distributed according to the patient age. The majority of the patients had

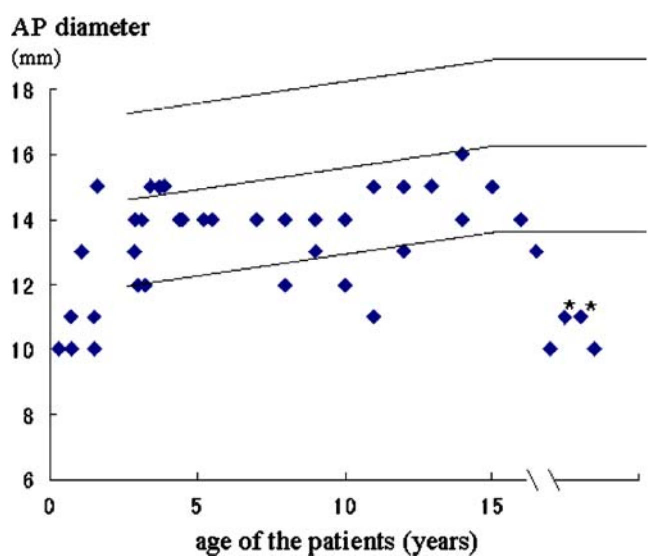

Fig. 1 Correlation between canal diameter and patient age. The X and Y axes indicate the age and the antero-posterior diameter of the cervical spinal canal at the narrowest point. The *three lines* indicate the averages and the 90-percentile mark. The majority of the patients were lower than the average. Twenty-nine percent of the patients were presented with canal stenosis, which is defined as a canal diameter less than the lower line. Two patients (denoted by *asterisks*) suffered myelopathy

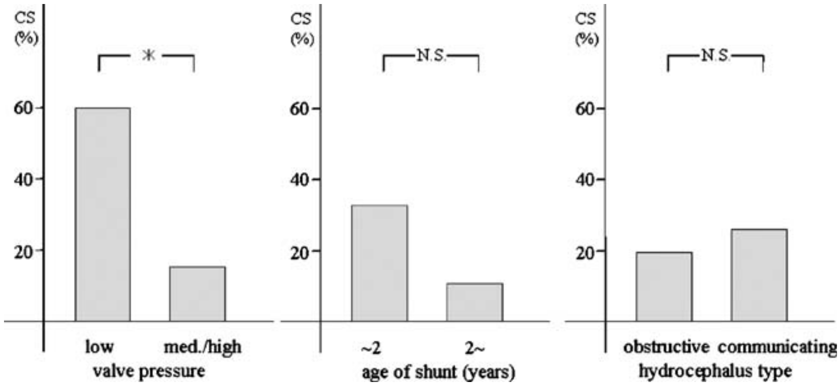

Fig. 2 Effects of low-pressure valve, age of shunt insertion, and types of hydrocephalus. *Left* incidence of canal stenosis (CS) in the group treated with low-pressure valves was significantly higher than that treated with medium/high-pressure valves; *center* the incidence of CS decreased when the age of first shunt insertion was greater than 2 years; however, the difference was not significant; *right* no differences in the incidence of CS were detected between the obstructive and communicating hydrocephalus (*$p<0.05$)

smaller diameter than the average. Out of the 33 patients, 10 (30.3%) were included in the CS category. Two patients (6.1%) presented myelopathy.

Effect of low-pressure valve, age of shunt insertion, and types of hydrocephalus

Of the 10 low-pressure valve users, six (60.0%) presented CS and four (17.4%) of the 23 medium/high-pressure valve users presented CS. Low-pressure valves caused a significantly high incidence of CS ($P<0.05$, Fig. 2, left). The average age of shunt insertion in a CS patient was 0.87±0.99 years and that without CS was 1.69±1.56 years. The CS incidence decreased when the age of the first shunt insertion was greater than 2 years. However, this difference was not significant (Fig. 2, center). Of the 10 patients with obstructive hydrocephalus, two (20.0%) presented CS and of the 14 communicating hydrocephalus patients, four (28.6%) presented CS. Thus, no differences in the incidence of CS were observed between the two types of hydrocephalus (Fig. 2, right).

Correlation between canal stenosis and slit ventricle

The average canal diameter of the slit ventricle group was 13.2±1.2 mm, which was same as the group without slit ventricle (12.1±1.8 mm). Among the 10 low-pressure valve users, only two patients presented slit ventricle (20.0%). In contrast, among the 23 medium/high-pressure valve users, nine patients (39.1%) presented slit ventricle. The average age of shunt insertion in patients with slit ventricle was 0.39±0.61 years, which was significantly lower than that of patients without slit ventricle (1.95±1.47 years, $P<0.05$). Slit ventricle was observed in six patients (42.9%) of the 14 with communicating hydrocephalus; this rate of incidence was higher than that among obstructive hydrocephalus patients (10.0%), although the difference was not significant. Slit ventricle occurred independent of CS in the patients who underwent shunt insertion in infancy.

Case presentation

A boy presented with macrocrania underwent cyst-peritoneal (CP) shunting with a low-pressure shunt valve for the treatment of the posterior fossa arachnoid cyst at the age of 3 years. When he was 32 years old, he suffered hypesthesia of both the hands and spastic lower paraparesis. His spinal cord was tightly compressed in the narrow spinal canal of an AP diameter of 11 mm, and the center of the cord revealed high signal intensity in a T_2-weighted image (Fig. 3, left). Although laminectomy was considered for the myelopathy, we opted for conservative observation. He suffered from intracranial hypertension 2 years later

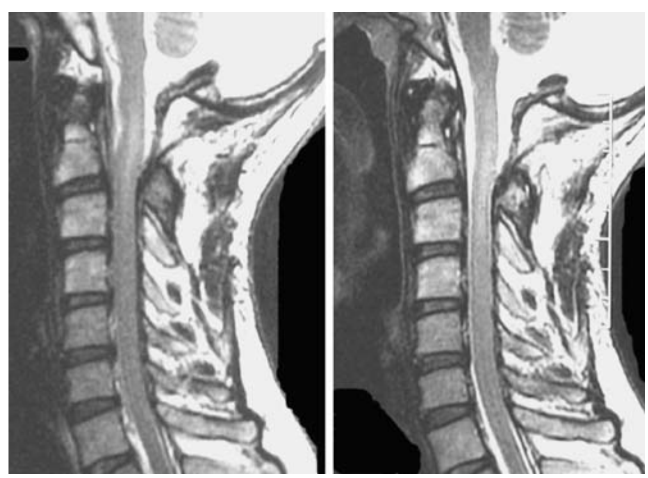

Fig. 3 Cervical MRI of the presentation case before the CP shunt malfunction (*left*) and after the CP shunt revision (*right*). *Left* the spinal cord was tightly compressed in the narrow spinal canal, and the center of the cord revealed high signal intensity. *Right* The cord compression is released, and the subarachnoid CSF space appeared around the cord

because the CP shunt was disconnected. Surprisingly, the myelopathy improved. To prevent overdrainage, we revised the CP shunt using a programmable valve set at 150 mmH_2O. The symptoms of intracranial hypertension and myelopathy were alleviated. The follow-up MRI demonstrated that the cord compression was released, and the subarachnoid CSF space appeared around the cord (Fig. 3, right).

Discussion

Inhibition of the developmental widening of the spinal canal may be caused by long-term CSF overdrainage since infancy. However, the CS alone cannot explain the reason for the paradoxical improvement in myelopathy following the increase in shunt pressure settings. Cases of compressive myelopathy caused by CSF overdrainage through VP shunting have been reported [5, 6, 8, 10, 11]. The myelopathies occurred 15 months to 25 years following the VP shunting. Of the five patients, three were adults; therefore, bony changes in the spine did not occur in them. In these reports, dilation of the epidural venous plexus or meningeal thickening due to intracranial hypotension was the cause for cord compression. Venous dilation should have been the main cause for myelopathy in our patient as well, and the presence of the narrowed spinal canal suggested an effective association. To prevent symptomatic CS, the canal diameter should be maintained within normal limits.

The incidence of CS in hydrocephalus patients who underwent VP shunting at infancy has already been reported by Kobayashi et al. [4] in 1983. We investigated the factors influencing the canal widening, namely, the shunt pressure settings, the age of shunt placement, and etiology of the hydrocephalus; these factors have not been described in the previous report. The use of low-pressure valves caused a significantly higher incidence of CS than the use of the medium- or high-pressure valves. Since we have treated children with hydrocephalus using low-pressure valves, these patients present with myelopathy over the next few years. Recently, we changed the programmable valve pressure settings to relatively high, particularly for pediatric hydrocephalus patients who are capable of sitting or standing without assistance. Moreover, the pressure setting will be adjusted with the increase in height and weight; this is similar to the determination of the optimal shunt setting based on the hydrostatic and intra-abdominal pressures [3, 7]. We estimate that these children will not suffer from myelopathy.

Once the spinal canal has been sufficiently widened, it is rare that it will narrow again even with prolonged intracranial hypotension. It is important to know the lower limit of the inhibition of canal widening that does not impact cord compression. Although statistically insignificant, patients who undergo shunting after 2 years of age seldom present with CS. We should closely observe the cervical canal diameter of patients who underwent shunt placement before the age of 2 years.

Despite having the same cause for intracranial hypotension, there was no correlation between the characteristics of CS and slit ventricle patients. Rekate et al. [9] indicated that severe slit ventricle syndrome is due to the disturbed CSF absorption due to intracranial venous hypertension. Unlike CS which is purely caused by CSF overdrainage, CSF and cerebral blood circulations may play roles in the formation of slit ventricle.

Conclusions

The pathophysiology of compressive myelopathy observed in hydrocephalus patients treated with CSF shunt comprises CS and dilation of the spinal epidural venous plexus. Both are caused by chronic CSF overdrainage. The placement of a low-pressure shunt in infancy causes a significantly higher incidence of CS than a medium- or high-pressure shunt. To prevent CS, proper control of hydrocephalus by using programmable valves and close observation of the cervical canal diameter are required.

Acknowledgment This work was supported in part by a Grant-in-Aid for Specially Promoted Research (Project No. 20001008) granted in 2008 to Kyushu Institute of Technology, Yamaguchi University, and Shizuoka University by Japan Ministry of Education, Culture, Sports, Science, and Technology.

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