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ORIGINAL PAPER

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Withdrawal of shunt systems – clinical use of the programmable shunt system and its effect on hydrocephalus in children

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Abstract *Objects:* The most important function of the programmable valve (PV) is to limit the shunt-dependent flow of the cerebrospinal fluid by upgrading valve pressure. This activates the regular circulation of cerebrospinal fluid, which may make successful removal of the shunt possible once sufficient cerebral development has been achieved. The purpose of this paper is to indicate the possibility of shunt removal using the programmable Medos and Sophy valves (one programmable Sophy valve was specially designed for this situation). *Methods:* Prior to regular use of the PV, removal of existing shunt systems was attempted in 57 children, since some systems malfunctioned and others had abdominal tubes that were meanwhile too short as the children had grown as they became older. Shunt removal was successfully achieved in only 18 patients (32%). However, in patients in whom PV valves were used, shunt removal was successful in 68 out of 114 patients (57%). This shows that the success rate of shunt removal becomes significantly higher when PV valves are used. The 68 cases in which PV valves were used and shunt removal was successful were divided into three groups: A, B, and C. In group A (36 cases, 53%), the Medos valve was used for the initial PV shunt implantation and the pressure was gradually increased up to 200 mmH_2O. The shunt systems were then withdrawn. Group B (29 cases, 43%) includes patients who experienced both the minor symptoms and ventricular enlargement attributable to increased valve pressure. The pressure was gradually upgraded by pumping several times and was maintained at close to 200 mmH_2O. After 6–24 months' observation shunt removal was performed, and in 21 out of 29 cases the outcome was good. However, the remaining 8 patients (12%) still had symptoms and required shunt reinsertion. The specially designed Sophy valves were then used, which allowed the pressure to be set at above 200 mmH_2O. The pressure was increased by degrees up to 400 mmH_2O and kept at the same level for 6–24 months. The shunt systems were then removed successfully. Although a high pressure setting was required over a sustained period, a total of 29 patients (43%) were able to have their shunts removed. In group C (3 cases, 4%), which included patients with aqueduct stenosis, the pressure was raised and thus allowed ventricle enlargement. Third ventriculostomy was performed under neuroendoscopy with the shunt pressure maintained at a high level. Shunt systems were removed successfully. *Conclusions:* This study showed that it is possible to remove the shunt systems in 50% or more of pediatric hydrocephalus cases in which PV valves are used. This is achieved through careful control of the valve pressure. Close observation is essential during the period when the PV pressure is maintained at a high level, as well as 6–12 months after shunt removal.

Keywords Children · Hydrocephalus · Shunt · Programmable valve · Withdrawal of shunt · Outcome

Introduction

The purpose of the clinical use of the programmable valve (PV) is to attain the intended cerebral ventricular pressure by noninvasive means. This will not only normalize the ventricular size but also prevent excessive drainage, since the high pressure limits the flow of cerebrospinal fluid (CSF) [1].

It is worth mentioning that the PV limits the shunt-dependent flow of CSF as the upgraded pressure activates the regular circulation of the CSF. This will lead to cerebral development, and shunt removal will consequently be possible [2].

In this study, we observed the practice of shunt removal in cases where PVs were used (Medos and specially designed Sophy).

Materials and methods

The total number of patients (aged 3–8 years) in whom we attempted the shunt removal operation was 176. This included 97 patients with implanted Medos valves, 14 with standard implanted Sophy valves, 8 with specially designed Sophy valves (which allow the pressure to be set at above 200 mmH_2O), and 57 with standard shunt systems.

Our basic PV pressure settings are shown in Table 1. We adjusted the pressure for each patient in accordance with the CT images and the degree of clinical improvement (Table 2). We attempted withdrawal of the ventriculoperitoneal shunt (VPS), either when the shunt had become too short as the patient grew older, or when the shunt patency became too low.

Observations

The CT images showed changes in both ventricle size and symptoms resulting from the increased pressure on the PV valves (Fig. 1). These changes can be divided into three types. In type I, the ventricle remained the same size or was slightly enlarged but no symptoms were observed. In type II, the ventricle became relatively enlarged and intermittent symptoms were seen. In type III, the ventricle was remarkably enlarged and clear symptoms appeared. In most type III cases it was impossible to remove the shunt.

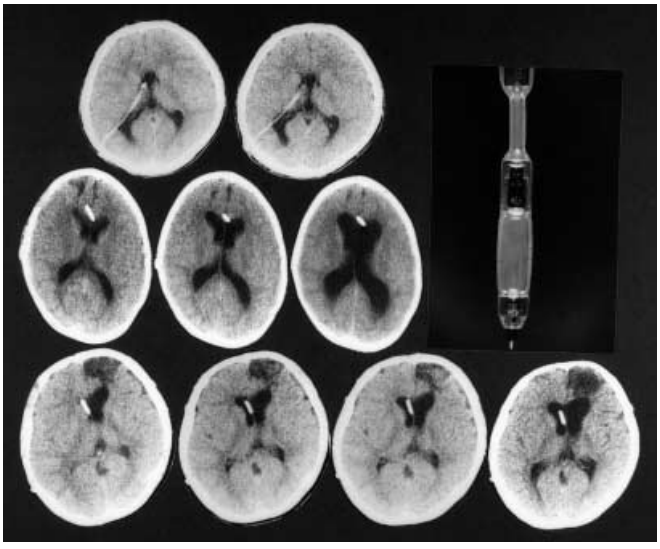

Fig. 1 CT results and symptoms that appeared after upgrading of the pressure setting of the PV. *Above*: Type I [ventricular enlargement (- ~ ±), symptoms (-)]. *Middle*: Type II [ventricular enlargement (± ~ +), symptoms (± ~ +)]. *Below*: Type III [ventricular enlargement (+ ~ ++), symptoms (+ ~ ++)]

Table 1 Basic pressure settings of the programmable valves

<1.5 years of age	30–80 mmH_2O
1.5–3 years of age	80–120 mmH_2O
>3 years of age	100–140 mmH_2O

Table 2 Procedures for step-by-step pressure setting of the programmable valve used for pediatric hydrocephalus after ventriculoperitoneal shunting

1 Start at 30 mmH_2O, then adjust the pressure based on the CT results over 1 week after the operation
2 Set the pressure at 30 mmH_2O or higher depending upon the patient's age, cerebral development and pathophysical condition over 1–12 months after the operation
3 The final result of VPS operation will be seen within 2 years of the operation
4 Increase the pressure of the shunt valve within 3 years of the operation to remove the shunt at latest

Figure 2 shows the CT images of a patient in group I. Neither clear symptoms nor ventricular enlargement can be observed. The pressure on the Medos valve was increased up to 200 mmH_2O and then shunt removal was attempted. After removal no symptoms appeared and ventricle size remained steady.

Figure 3 shows a patient in group II. Typical symptoms included headaches and inactivity. The CT images show some ventricular enlargement. Nevertheless, further progression of the symptoms was not observed and ventricular enlargement did not continue. The symptoms gradually disappeared. The pressure was then adjusted in such a way that it would reach 200 mmH_2O over a period of approximately 12 months and the shunt removal operation was performed. After removal there were no visible symptoms and ventricular enlargement was not observed.

In the cases shown in Fig. 4, the pressure was set and maintained at 200 mmH_2O for approximately 6 months and the shunt was then removed since there were no clear symptoms and ventricular enlargement was not observed. However, after the removal procedure symptoms such as headache, vomiting, and inactivity were observed, and ventricular enlargement was also found. The

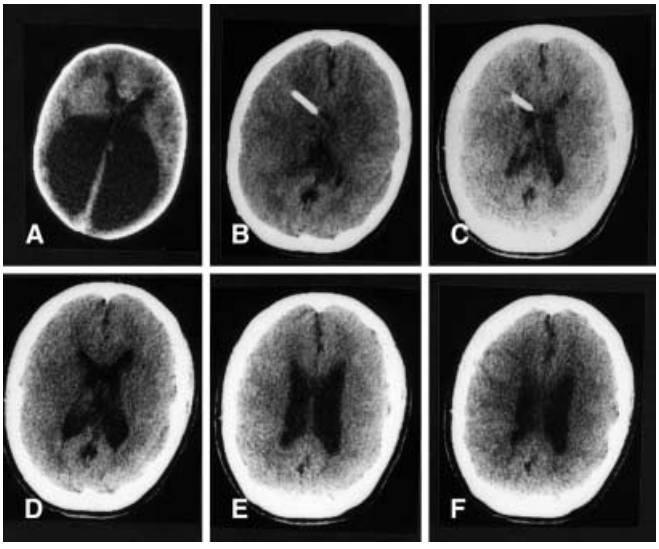

Fig. 2A–F Removal of the shunt system using the programmable valve (type I). **A** Before shunt insertion. **B** After shunt insertion (30→80 mmH_2O). **C** Valve pressure upgraded (80→120→160→200 mmH_2O). **D** One month after shunt removal. **E** Six months after shunt removal. **F** Twelve months after shunt removal

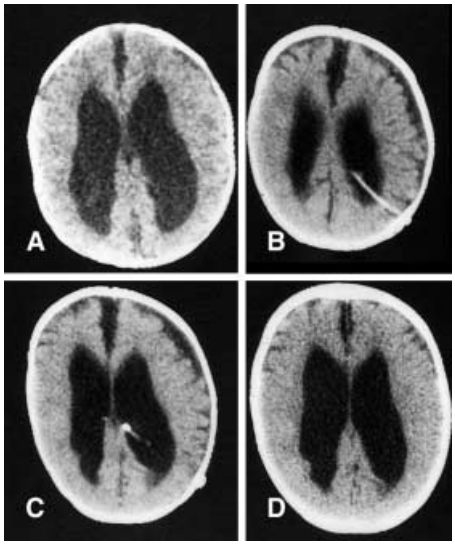

Fig. 3A–D Removal of the shunt system using the programmable valve (type II). **A** Before shunt insertion. **B** After shunt insertion (30→60 mmH_2O). **C** Increasing valve pressure (60→100→160→200 mmH_2O). **D** The shunt system was removed after 12 months with the pressure set at 200 mmH_2O

VPS was replaced with an upgraded Sophy valve, which supports pressures set at over 200 mmH_2O. The pressure was increased to 385 mmH_2O. This high pressure was maintained for 12 months, and then the shunt was removed. After shunt removal there were no symptoms or ventricular enlargement.

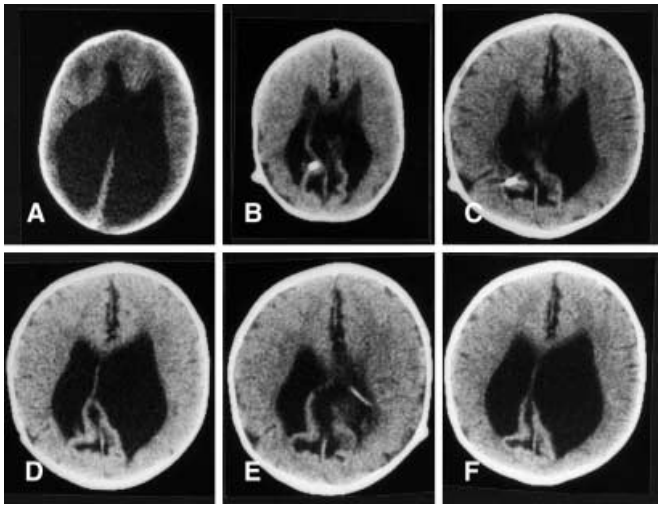

Fig. 4A–F Removal of the shunt system using the programmable valve (type III). (It was impossible to withdraw the shunt system due to persistence of the symptoms. The Medos was replaced with the specially designed Sophy. Shunt removal was achieved with the replaced Sophy.) **A** Before shunt insertion. **B** After shunt insertion (30→80 mmH_2O). **C** Before removal of the Medos (the pressure had been kept at 200 mmH_2O for 6 months). **D** After removal of the Medos (both ventricular enlargement and symptoms appeared). **E** After shunt reinsertion operation (using specially designed Sophy). **F** Shunt removal achieved using the specially designed Sophy valve with the pressure maintained at a high level for 12 months

Figure 5 shows a patient who had aqueduct stenosis and an implanted VPS. As ventricle development appeared to be normal the pressure was gradually increased to approximately 200 mmH_2O. In case the ventricle should otherwise become enlarged, a third ventriculostomy was performed using neuroendoscopy. The pressure was kept high (200 mmH_2O) for about 1 month and then the shunt was removed.

Results

In cases where PVs were not used, successful shunt removal was achieved in only 18 patients out of 57 (Fig. 6). Using PVs enabled 68 out of 119 to have their shunt systems removed (Fig. 7). The rate of successful shunt removal rose significantly in comparison with cases without PV systems.

The successful 68 cases were divided into groups A, B, and C (Fig. 7). In group A (36 cases, 53% of the successful cases), the PV system used was Medos and the pressure setting was upgraded to 200 mmH_2O or near that. Group A patients had no apparent symptoms and no noticeable ventricular enlargement. The shunt removal operation was successful.

Group B (29 cases, 43%) included patients who had both minor symptoms and slight ventricular enlargement

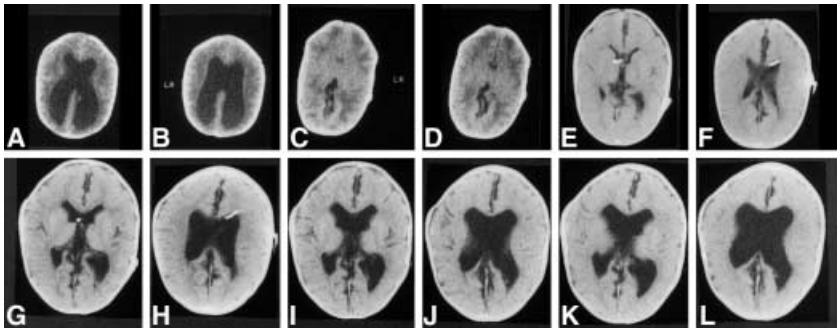

Fig. 5A–L Shunt removal by means of the programmable valve plus third ventriculostomy. **A, B** Before shunt insertion. **C, D** After shunt insertion (30→60 mmH_2O). **E, F** Upgrading of valve pressure over 6 months (60→80 →120 mmH_2O). **G, H** Further upgrading of valve pressure over 12 months (120→160→ 200 mmH_2O). **I, J** Removal of the shunt system with third ventriculostomy. **K, L** Twelve months after shunt system removal

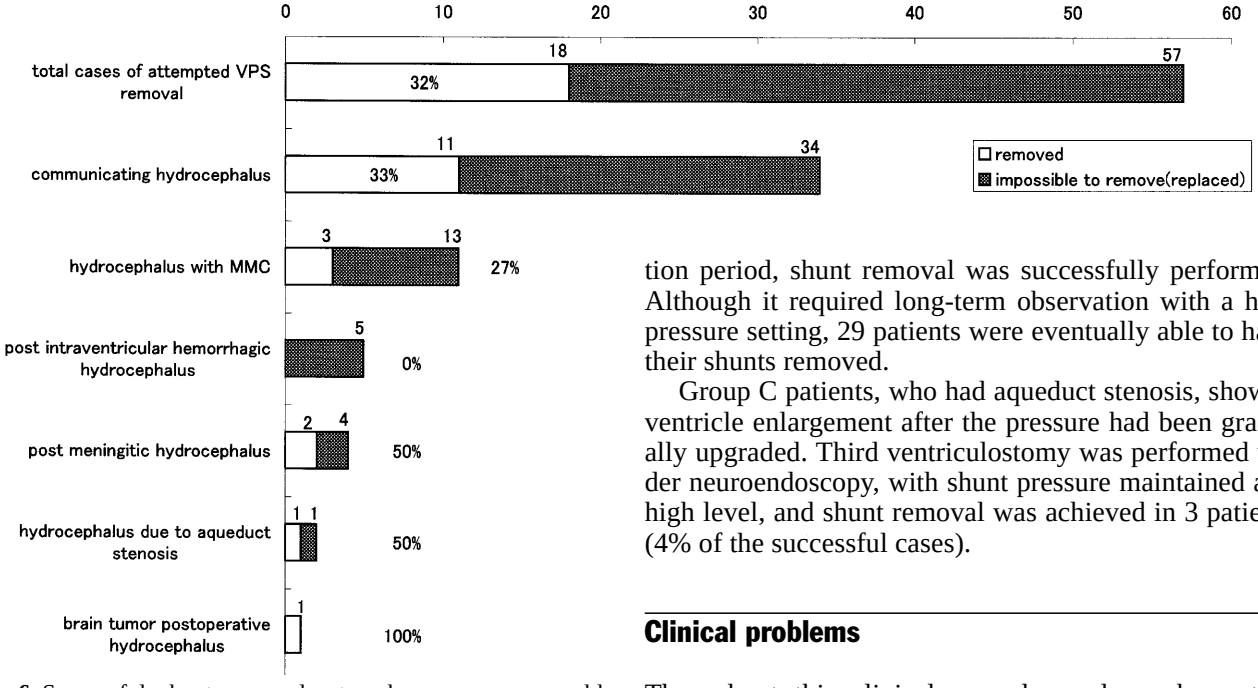

Fig. 6 Successful shunt removal rate when nonprogrammable valves were used (*VPS* ventriculoperitoneal shunt, *PV* programmable valve, *MMC* meningomyelocele)

after the valve pressure was increased. The pressure setting of the Medos valve was upgraded by degrees by pumping several times and was kept close to 200 mmH_2O. It required 6–24 months' observation before the shunt removal operation was attempted, and it was only successful in 21 cases. Although the remaining 8 patients were subjected to the same procedure, they still had symptoms, so that shunt reinsertion was performed. For these reinserted shunts, the specially designed Sophy valves that allow the pressure to be set at above 200 mmH_2O were used. The pressure setting was increased by degrees to almost 400 mmH_2O and maintained at that level for 6–24 months. After the observation period, shunt removal was successfully performed. Although it required long-term observation with a high pressure setting, 29 patients were eventually able to have their shunts removed.

Group C patients, who had aqueduct stenosis, showed ventricle enlargement after the pressure had been gradually upgraded. Third ventriculostomy was performed under neuroendoscopy, with shunt pressure maintained at a high level, and shunt removal was achieved in 3 patients (4% of the successful cases).

Clinical problems

Throughout this clinical procedure, close observation was necessary as any patient's condition could become critical. As the PV forced the shunt-dependent CSF flow to shift to the regular physiological circulation, clinical problems appeared, especially in group B.

In cases where the valve pressure was kept high over a long period, such symptoms persisted even after removal of the shunt. Intracranial pressure was measured in the lateral position in patients who displayed minor symptoms such as mild headaches. Five patients in group B had intracranial pressure as high as 320–480 mmH_2O, and this persisted for 2–6 months after shunt removal. The intracranial pressure was determined by means of lumbar puncture (Fig. 8). Figure 9 shows the cases where the cranial sutures split. It should be noted that no clear symptoms were observed in many of the patients, in spite of persistently high intracranial

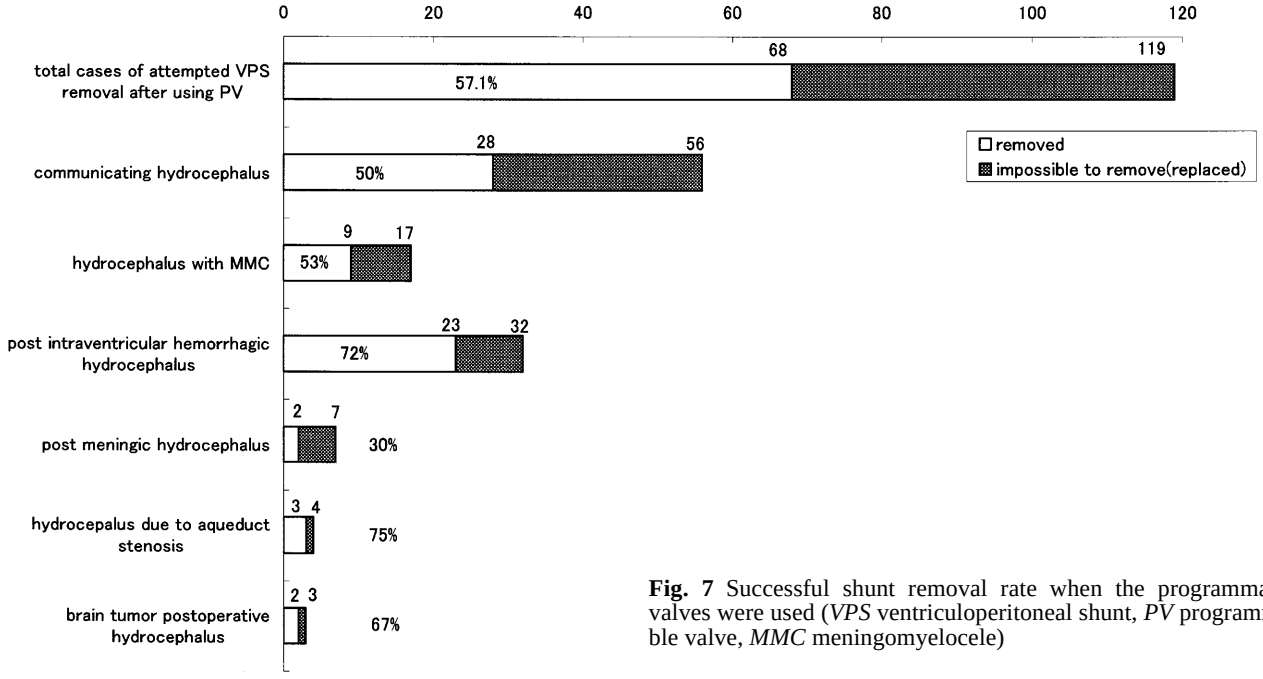

Fig. 7 Successful shunt removal rate when the programmable valves were used (*VPS* ventriculoperitoneal shunt, *PV* programmable valve, *MMC* meningomyelocele)

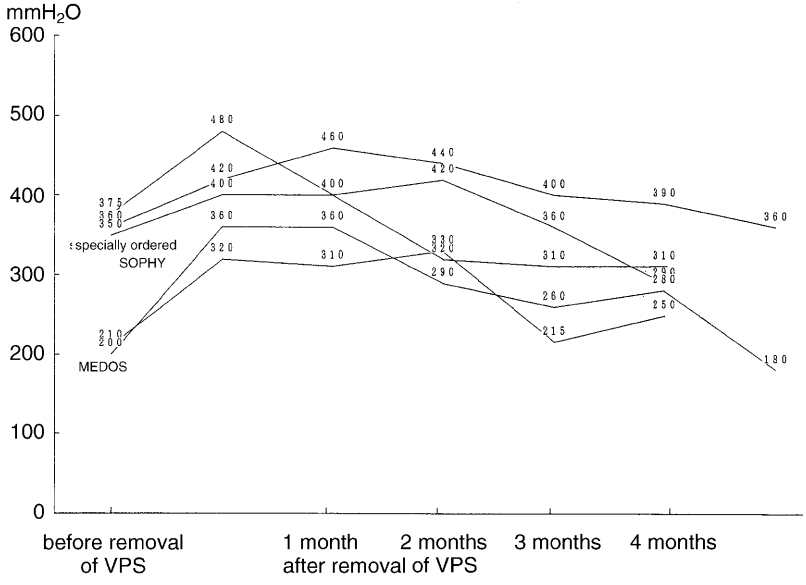

Fig. 8 Transition of the intracranial pressure in the patient whose shunt system was removed after long-term observation with a high pressure setting

pressure. Ophthalmic examinations also revealed no cases of noticeable choked disk. Close observation was required during the period of high valve pressure and also for 6–12 months after its removal. If the symptoms such as vomiting, headache, inactivity persist, shunt reinsertion should be considered.

Discussion and conclusion

By controlling PV pressure, it was possible to produce a clinically significant change in both intracranial pressure and ventricular size. When excessive drainage of CSF from the slit ventricle or subdural fluid collection occurred, use of the PV was seen to be the optimal choice for limiting the shunt-dependent CSF flow by controlling the pressure [1].

The PV system will normalize intracranial pressure and stimulate cerebral development. Moreover, it is possible to limit the shunt dependency of CSF flow by controlling the valve pressure and stimulating the regular circulation of CSF, which will eventually lead to the patient's becoming independent of the system. Children who suffer from hydrocephalus and their families dream

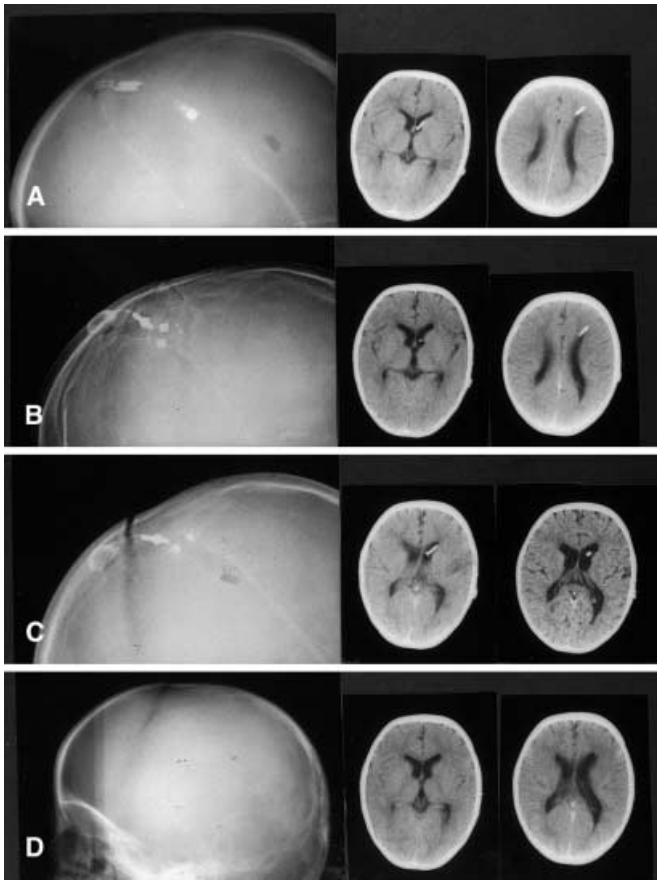

Fig. 9A–D Transition of skull X-P and CT results after long-term observation with a high pressure setting. **A** 200 mmH_2O (Medos). **B** 200 →245 mmH_2O (Medos replaced by specially designed Sophy valve). **C** 375 mmH_2O (specially designed Sophy). **D** 400 mmH_2O (1 month after shunt removal)

of having their shunts removed. In this sense, PVs are the most efficient shunt systems currently available.

Gradually upgrading the valve pressure activates the physiological circulation of CSF, and approximately 60% of the patients in our study had their systems removed. The PVs most widely used are the Medos and Sophy valves. It should be noted that there are also some disadvantages to these two types of PV, as well as their advantages: with Sophy valves it is not easy to set the pressure accurately, although this should theoretically be possible, whilst Medos valves do not support pressure settings above 200 mmH_2O. Therefore, in cases where a Medos valve is not adequate to the task of shifting the shunt-dependent flow, the specially designed Sophy valve will need to be used.

Use of PVs may make it possible to solve the clinical problems associated with hydrocephalus and achieve the withdrawal of shunt systems. This would mean that the patients would be able to lead a normal life in the future. Precise understanding of each patient's clinical condition, plus a detailed knowledge of the PV shunt systems available for use, largely determines the effectiveness of PVs. In addition, a great deal of consideration must be given to each patient's future prospects.

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