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## The use of an adjustable valve to treat over-drainage of a cyst-peritoneal shunt in a child with a large sylvian fissure arachnoid cyst

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**Abstract** *Introduction:* The cyst-peritoneal shunt is a recognised surgical alternative in the management of sylvian fissure arachnoid cysts. Shunt overdrainage is well described in literature on ventriculo-peritoneal shunts, but not often appreciated as a complication of cysto-peritoneal shunts. *Case report:* A 5-year-old boy presented with a symptomatic left sylvian fissure arachnoid cyst. This was initially treated by craniotomy and membrane fenestration in the carotid cistern. Recurrence led to insertion of a valveless cyst-peritoneal shunt 5 months later. Initial progress was followed by persistent headaches 18 months after shunt insertion. CT scan revealed a significant reduction in the cyst size, enlargement of the ipsilateral lateral ventricle, collapse of the contra-lateral ventricle and midline shift towards the side of the shunt. These findings were interpreted

as over-drainage of the cyst-peritoneal shunt. *Result:* A Codman Medos adjustable valve was inserted, with the intention of gradually increasing the pressure until the midline shift was restored and the contra-lateral ventricle was reconstituted. This was achieved with the valve set at 90 mm H<sub>2</sub>O, verified by CT scan. Radiological improvement was associated with dramatic symptomatic improvement. *Conclusion:* Over-drainage of cyst-peritoneal shunts is often not appreciated, especially when the main manifestation is headaches. As it is difficult to predict the required valve pressure setting, it may be advisable to consider the use of an adjustable valve.

**Keywords** Hydrocephalus · Arachnoid cyst · Adjustable valve · CSF shunt

### Introduction

Arachnoid cysts are benign congenital space-occupying lesions containing cerebrospinal fluid. Surgical treatment is indicated when they become symptomatic by causing intracranial hypertension, focal neurological deficits, intractable epileptic seizures or haemorrhage following minor head trauma [1]. The pathogenesis and the natural history of arachnoid cysts are not fully understood, and this has led to the development of a variety of surgical approaches, including cyst-peritoneal shunting, ventriculo-cisternostomy, cyst fenestration, and recently neuroendoscopic cyst fenestration [1, 3, 6, 7, 9]. In a study in which shunting

was compared with fenestration procedures, it was concluded that for middle fossa cysts, shunting is the procedure of choice [3]. However, another similar comparative study found membranectomy and opening of the basal cisterns to be the procedure of choice [9]. The success of cyst-peritoneal shunting is usually defined as obliteration of the arachnoid cyst. Shunt over-drainage is a well-described complication of ventriculo-peritoneal shunts [5, 8], but is not often recognised as a complication of cyst-peritoneal shunts. We present a patient who developed symptomatic over-drainage following initially successful shunting of a large sylvian fissure arachnoid cyst.

## Case report

A 5-year-old boy presented with a symptomatic large left sylvian fissure arachnoid cyst (Fig. 1). A craniotomy was performed, and a substantial part of the cyst was excised and membrane fenestration was performed in the region of the carotid cistern. After an initial symptomatic and radiological improvement, symptoms recurred and on imaging the cyst was enlarged after 3 months. It was decided to treat the patient with a cyst-peritoneal shunt. A valveless cyst-peritoneal shunt was inserted 5 months after the craniotomy. Initial progress was followed by persistent headaches for 18 months after shunt insertion. The headaches became progressively worse, and interfered with the child's performance at school. A CT scan revealed a marked reduction in the cyst size, dilatation of the ipsilateral left lateral ventricle, obliteration of the contralateral right lateral ventricle and midline shift towards the shunted side (Fig. 2). In conjunction with the symptoms, these findings were interpreted as over-drainage of the cyst-peritoneal shunt. It was decided to employ an adjustable pressure valve, to try to partially "re-enlarge" the cyst and counteract the symptoms of over-drainage.

A Codman Medos adjustable valve was inserted in the circuit of the cysto-peritoneal shunt, 22 months after initial insertion. The pressure was initially set at 50 mm H<sub>2</sub>O. The initial pressure setting was deliberately chosen to be low, to avoid a sudden change in intracranial pressure. The patient was followed up closely and the pressure was gradually increased by 10 mm H<sub>2</sub>O every month, titrated against the symptoms. At a pressure setting of 90 mm H<sub>2</sub>O, the cyst had moderately enlarged, the midline shift was restored and the contralateral ventricle enlarged (Fig. 3). Radiological improvement was associated with a dramatic symptomatic improvement.

## Discussion

Careful observation of the images of this patient (Fig. 1) will reveal that the left side of the skull, which houses the



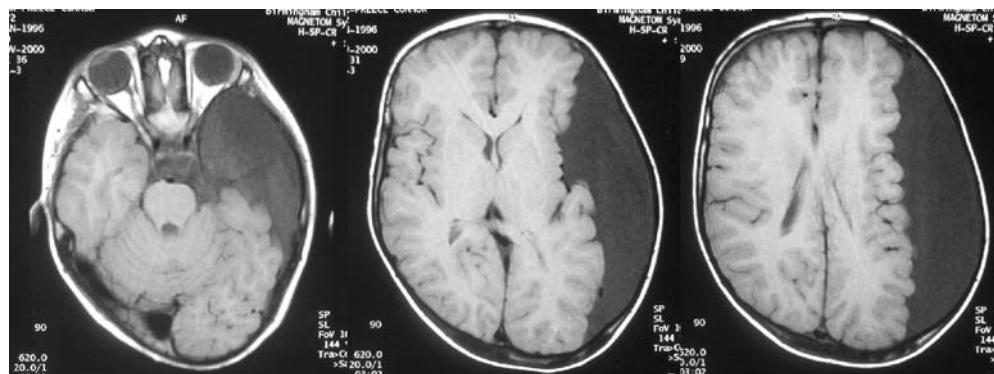
**Fig. 2** Computed tomography scan obtained 18 months after insertion of a valveless cyst-peritoneal shunt, when the patient had persistent headaches that interfered with school performance. The sylvian fissure arachnoid cyst has been reduced in size dramatically, the left lateral ventricle has enlarged to a size bigger than normal, the right lateral ventricle has been effaced and there is a midline shift to the left.

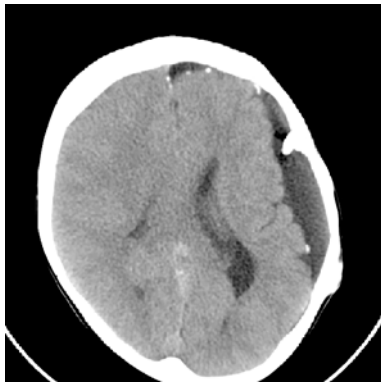
large sylvian fissure arachnoid cyst, is larger than the normal right side of the skull. This is a common finding in patients with large sylvian fissure arachnoid cysts and is believed to be due to the chronic pressure effect of the cyst on the adjacent temporal bone. It is reasonable to speculate that complete obliteration of the cyst in response to shunting will force the brain to expand abnormally in order to occupy a much larger volume than it was originally meant to.

The appearance of Fig. 2 corresponds to this possible explanation. The collapse of the arachnoid cyst due to shunting has "drawn" the left hemisphere to the left, forcing it to expand by enlarging the left lateral ventricle, and resulting in a midline shift to the left when this ventricular expansion is exceeded. This represents an "over-treatment" of shunting due to over-drainage.

In a recent report in which SPECT scanning was used to define the perfusion defects caused by arachnoid cysts, it was found that the defects disappeared even though the middle fossa cysts were not completely collapsed. This implied that the drive to collapse the cyst entirely with a

**Fig. 1** Axial T1-weighted MR scan of a 5-year-old boy who presented with persistent headaches and papilloedema. There is a large left sylvian fissure arachnoid cyst, which extends over most of the convexity of the left hemisphere. The underlying hemisphere appears compressed.





**Fig. 3** Computed tomography scan obtained 3 months after insertion of a Codman Medos adjustable valve in the cyst-peritoneal shunt, when the patient was asymptomatic. In comparison to Fig. 2, the sylvian fissure arachnoid cyst has increased in size, the left lateral ventricle is smaller and the midline shift is not present.

cyst-peritoneal shunt may represent an unnecessary over-engineering of treatment [10]. This case report renders further support to this observation.

Symptomatic shunt over-drainage is seen in a small proportion of patients with ventriculo-peritoneal shunts [5]. It presents usually with chronic or recurring headaches, and is associated with slit ventricle appearance on neuro-imaging. Over-drainage has not been described in association with cyst-peritoneal shunts. The only related conditions that have been described are shunt dependency [4] and development of slit ventricles [11]. Over-drainage as a complication should be suspected in patients with cyst-peritoneal shunt who present with persistent headaches, after an apparent reduction in the size of the sylvian fissure cyst. In this patient, a gradual increase in the pressure in the cyst led to symptomatic improvement and restoration of a more normal configuration of the brain. A good radiological result and symptomatic control was achieved by setting the pressure to 90 mm H<sub>2</sub>O. This pres-

sure setting was achieved by titrating against the patient's symptoms and was guided by periodic CT scans as there were no clear guidelines in the literature. A report has been published recently in which programmable shunts were used in seven patients with arachnoid cysts [6]. The authors achieved satisfactory results at a pressure setting of 100 mm H<sub>2</sub>O, which is similar to our setting. They postulated, probably rightly, that it is not necessary to aim for total collapse of the cyst with shunting and that "partial" drainage with an adjustable valve may not lead to bowing of the overlying temporal bone. More studies will be needed to define the best pressure setting, as well as the long-term effect of "partial" shunting on the underlying brain and the overlying temporal bone.

The programmable Codman Medos valve has been shown to be very helpful in managing difficult conditions like normal pressure hydrocephalus, multi-loculated hydrocephalus, and arachnoid and porencephalic cysts [2]. Zemack and Romner [12], describing their experience with the programmable Codman Hakim valve in 583 patients with different conditions, conclude that a programmable valve should be used in all cases requiring CSF drainage. It may be difficult to generalise indications to all cases of hydrocephalus, but the use of programmable shunts in arachnoid cysts, especially the very large sylvian fissure arachnoid cysts extending over the surface of the hemisphere, seems to be a reasonable option.

There is increasing evidence that complete obliteration of a sylvian fissure arachnoid cyst probably should not be the goal of treatment. Open fenestration through craniotomy or endoscopic fenestration certainly do not lead to total collapse of the arachnoid cyst, and are rarely associated with symptoms of over-treatment. When shunting is chosen as the main treatment, it may be advisable to employ an adjustable pressure valve, in order to gradually reduce the size of the cyst, without collapsing it completely, guided by symptomatic improvement.

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