

Case Reports

Ventriculo-sagittal sinus shunt malfunction. *Causes of failure, avoidance, and alternatives*

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ABSTRACT

توضح هذه الحالة أسباب فشل عملية تحويل السائل النخاعي إلى الجيب الوريدي السهمي العلوي وكيفية تفاديها. تبلغ المريضة 14 عاماً، وتعاني من استسقاء دماغي منذ الولادة ناتج عن نزيف ببطينات المخ. تعرضت المريضة لأكثر من عملية لتسليك الأنبوب ونقله من تجويف البطن إلى الأوردة الوريدية بالرقبة، ثم إلى التجويف البلوري بالصدر، وفي النهاية تم تركيب تحويله من بطين المخ إلى الجيب الوريدي السهمي العلوي. أصيبت المريضة بفشل في عملية التحويل نتج عنه التهاب ببطينات المخ أدي إلى وفاتها. هناك عدة ملاحظات يجب مراعاتها لتفادي فشل تحويل السائل النخاعي إلى الجيب الوريدي السهمي العلوي وهي: استخدام قسطرة أذينية (ذات شقوق جانبية) في الجيب الوريدي لمنع ارتجاع الدم إلى الصمام، تصوير أوردة المخ بأشعة الرنين المغنطيسي للتأكد من عدم انسدادها، كما يجب قياس ضغط الدم بالجيب الوريدي السهمي قبل وضع القسطرة.

This case report highlights causes of failure of the ventriculo-sagittal sinus (V-S) shunt and precautions to avoid them. We present, a 14-year-old girl, a case of post-hemorrhagic hydrocephalus with multiple revisions of ventriculo-peritoneal (V-P) and ventriculo-atrial (V-A) shunts. She developed malfunctioned V-S shunt, and ventriculitis that was complicated with massive cerebellar and brain stem infarction and the patient died. To avoid malfunction, a cardiac catheter with side slits should be used, magnetic resonance angiography is recommended before shunt placement to check the patency of the sinus, and the pressure in the superior sagittal sinus should be measured at the time of surgery. In patients with problematic distal catheters, direct placement of the catheter into the right atrium using thoracoscope could be an alternative to gall bladder or ureter shunts.

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Ventriculo-sagittal sinus (V-S) shunt has been described for CSF diversion in patients with inaccessible traditional distal drainage sites for almost 40 years, during this period reports with modifications of its techniques were described.¹⁻³ The V-S shunt placement is technically easy and theoretically appears more physiological because it avoids the dynamics of CSF siphoning, which can be quite problematic for some patients with shunts. This report gives an example of a complicated course of V-S shunt and discusses the causes of shunt malfunction and technical tips to avoid such complications as well as alternatives of CSF diversion in patients with a problematic distal catheter.

Case Report. A 14-year-old girl who had ventriculo-peritoneal (V-P) shunt inserted since her infancy to treat post-hemorrhagic hydrocephalus, developed shunt infection and had multiple shunt revisions that ended with insertion of a right side ventriculo-atrial (V-A) shunt at the age of 10. She did well for 2 years then developed V-A shunt malfunction. An attempt at endoscopic third ventriculostomy failed and a V-P shunt was tried again. It failed after 3 months due to abdominal adhesions and accumulation of CSF in the abdomen and so a ventriculo-pleural shunt was inserted for a short time then it had to be removed because of development of hydrothorax, and the V-A shunt was reinserted on the opposite side. At the age of 14 she was readmitted because of malfunctioning left V-A shunt; an external ventricular drain (EVD) was inserted, duplex scan and MRV showed thrombosis of both internal jugular veins. Ventriculo-sagittal sinus shunt was then inserted using the El-Shafei (retrograde) technique,⁴ together with insertion of Ommaya reservoir on the left side (Figure 1). Following the V-S shunt she became lethargic and had repeated vomiting; she developed subgaleal CSF collection and CSF leak from the wound. Conservative treatment in the form of CSF drainage from the Ommaya reservoir and acetazolamide was tried for a few days, but did not stop the CSF leak, and she developed ventriculitis (mixed growth of gram negative bacilli) despite antibiotic prophylaxis. All implants were removed and the EVD was reinserted. After 4 weeks of treatment she suddenly became unconscious, the pupils



Figure 1 - A CT scan, showing the ventriculo-sagittal sinus shunt with the distal catheter inside the superior sagittal sinus (long arrow) and Ommaya reservoir is inserted in the left occipital horn (short arrow).

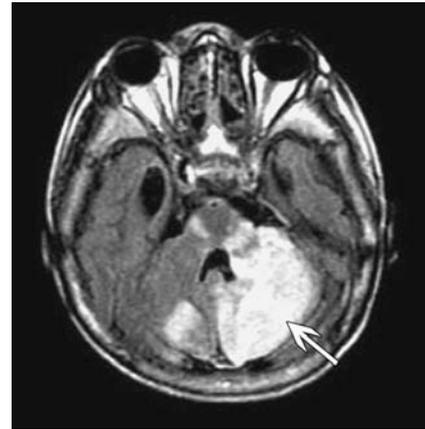


Figure 2 - An MRI scan (FLAIR-sequence) showing infarction of the cerebellum and brain stem (white arrow).

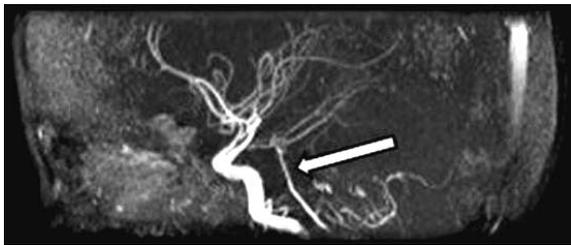


Figure 3 - An MRA showing severe spasm of the vertebro-basilar arterial system (arrow).

became dilated and she developed respiratory arrest. After resuscitation, MRI scan showed massive cerebellar infarction involving both cerebellar hemispheres and brain stem. An MRA revealed severe spasm of all cerebral arteries, which was more apparent in the posterior circulation (Figures 2 & 3), she remained on ventilation for a few days and subsequently died.

Discussion. The main problems of CSF shunting are infection and shunt obstruction. Once infection occurs, the course of the disease becomes complicated and a series of problems arise leading to multiple surgical procedures.^{3,5,6} Our patient developed shunt infection that triggered a series of complications including peritonitis, abdominal adhesions, and failure of absorption of CSF from the peritoneal cavity leading to massive ascites, poor feeding, and impairment of the child's growth in addition to multiple surgical operations. In such situations where distal shunt placement is problematic, CSF diversion to superior sagittal sinus (SSS) becomes an alternative. The patient presented in this report is a tragic case and illustrates the complicated course of V-P shunt, which was fatal in her case. Failure of her V-P and V-A shunts as well as the ventriculo-pleural shunt left us with few treatment options; choroids plexus coagulation, shunting

the CSF to SSS (V-S shunt) or uncommon shunts to the ureter and gall bladder. The V-S shunt was preferred for 2 reasons; it is technically easier, and theoretically has less chances of shunt infection than the other types of shunts. There are different techniques for insertion of the V-S sinus shunt.^{1,2,4,7,8} The retrograde technique for insertion of the catheter into the SSS described by El-Shafei,² was used for this patient because the incidence of sinus thrombosis is less. Another technical point for placement of the SSS shunt is the use of cardiac catheter (with side slits) instead of the peritoneal catheter, which has an open lower end that allows retrograde flow of blood into the shunt reservoir, causing malfunction. It is also important to measure the pressure in the SSS (using a 24G butterfly catheter connected to a manometer) before placement of the shunt into the sinus. In our patient, the opening pressure in the SSS was high (150 mm water), which was attributed to thrombosis of the neck veins. The high pressure in the SSS lead to shunt malfunction and CSF leak from the wound that resulted in severe gram-negative bacilli ventriculitis and the patient's death.

In conclusion, V-S sinus shunt can be used, as a last option, in patients with a problematic distal catheter. To avoid malfunction, it is recommended to use MR-venography to check the patency of the SSS and to measure the pressure in the sinus before shunt placement. Another important technical point is the use of a cardiac catheter with side slits instead of the peritoneal catheter, which is open at its lower end. In patients with a problematic distal catheter, direct insertion of the catheter into the right atrium using minimally invasive techniques might be an alternative to gall bladder and ureter shunts.

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