

Boil over VP Shunt site as a gateway to CNS Infection: A rare case in a child with Dandy-Walker variant and Situs Inversus Totalis

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Abstract

Background:

Hydrocephalus in a clinical syndrome of raised intracranial pressure with ventriculomegaly. It usually occurs from defective flow of cerebrospinal fluid or rarely from excessive secretion. Management typically involves ventriculoperitoneal (VP) shunt insertion, which carries a risk of infection, particularly in young and immunocompromised children. The coexistence of rare anomalies such as Dandy-Walker variant and situs inversus totalis further complicates clinical management and may reflect a shared embryological basis.

Case Summary:

We present a case of a 15-month-old female with communicating hydrocephalus and VP shunt in situ (placed at age of 9 months) who presented with vomiting, seizures, and an infected, exposed shunt tract. Neuroimaging revealed features of the Dandy-Walker variant, while chest and abdominal imaging were suggestive of situs inversus totalis. CSF analysis and shunt tip culture confirmed *Pseudomonas aeruginosa* meningitis. The patient underwent emergency VP shunt removal and insertion of a ventricular reservoir and was treated with intravenous meropenem and linezolid. Clinical improvement was noted, and VP shunt reinsertion is planned.

Keywords: Congenital Hydrocephalus, Situs Inversus Totalis association, Exposed VP Shunt, Shunt Meningitis

Introduction

Hydrocephalus is a significant cause of neurological morbidity in children and is frequently managed by cerebrospinal fluid (CSF) diversion through ventriculoperitoneal (VP) shunt insertion. While generally effective, VP shunting is associated with a notable risk of complications, particularly infection, with rates ranging from 5–15% in pediatric populations. These infections, often caused by skin flora, can also be due to aggressive gram-negative organisms like *Pseudomonas aeruginosa*, especially in hospital settings.

Structural anomalies of the central nervous system often coexist with hydrocephalus. One such anomaly is the Dandy-Walker variant, which features inferior vermian hypoplasia, cystic dilatation of the fourth ventricle, and communication with the cisterna magna.

This variant may impair CSF flow and contribute to developmental delays and hydrocephalus.

Situs inversus totalis is a rare congenital condition involving mirror-image reversal of the thoracic and abdominal organs. Though typically asymptomatic, its recognition is important in diagnostic imaging and surgical procedures. Emerging evidence suggests a possible link between laterality defects, such as situs inversus, and disorders of neural tube development. Both anomalies may reflect underlying ciliopathies or disruptions in early embryonic left-right axis determination.

Here, we report a case of VP shunt infection due to *Pseudomonas aeruginosa* in a child with communicating hydrocephalus, who was incidentally found to have both a Dandy-Walker variant and situs inversus totalis. This case highlights the intersection of

complex congenital anomalies with infectious complications and emphasizes the need for multidisciplinary management and awareness of potential syndromic associations.

Case Summary:

A 15-month-old female, diagnosed with communicating hydrocephalus and VP shunt insitu placed at 9 months of age, presented with vomiting for three days, followed by a generalized tonic-clonic seizure lasting 15–20 minutes. A month ago, the child developed a boil on the scalp directly over the shunt insertion site. Only topical antibiotic cream was applied, without any systemic antibiotics. Over time, the lesion began to slough and discharge pus, eventually leading to exposure of the cranial end of the shunt through the overlying skin (Figure 1). On examination, she was drowsy, HR-60-70/min, RR- 20-22/min, hypotonic in all four limbs, and had a low Glasgow Coma Scale (GCS) score. The shunt tubing was visibly exposed with signs of local infection. On detailed history taking, there was delay in gross motor and language milestones and moderate acute malnutrition was present.

Neuroimaging (CT Brain) revealed moderate ventriculomegaly, bilateral parieto-occipital white matter hypodensities, thinning of the corpus callosum, a choroidal fissure cyst, and features consistent with Dandy-Walker variant, including inferior vermian hypoplasia and communication between the fourth ventricle and cisterna magna. Chest X-ray and abdominal ultrasound confirmed dextrocardia and complete situs inversus totalis (Figure 2). Echocardiography revealed a structurally normal heart. CSF analysis showed total WBC count (1350), neutrophilic pleocytosis (755), elevated protein (187), and low glucose (55); culture from both the CSF and shunt tip grew *Pseudomonas aeruginosa*.

Emergency shunt removal was performed, and a ventricular reservoir was placed. Intravenous meropenem and linezolid were started, along with IV levetiracetam and phenytoin. Over the next few days, the child showed neurological improvement, resumed oral feeding, and no convulsion episodes. Hydrocephalus is currently being managed by repeated cerebrospinal fluid (CSF) drainage through the reservoir. Repeat CSF analysis shows improved CSF picture, with planned reinsertion of a VP shunt upon normalization.



Fig 1- Cranial end of VP shunt seen through gaped scalp wound



Fig 2- Chest X-Ray with abdomen showing Situs Inversus Totalis

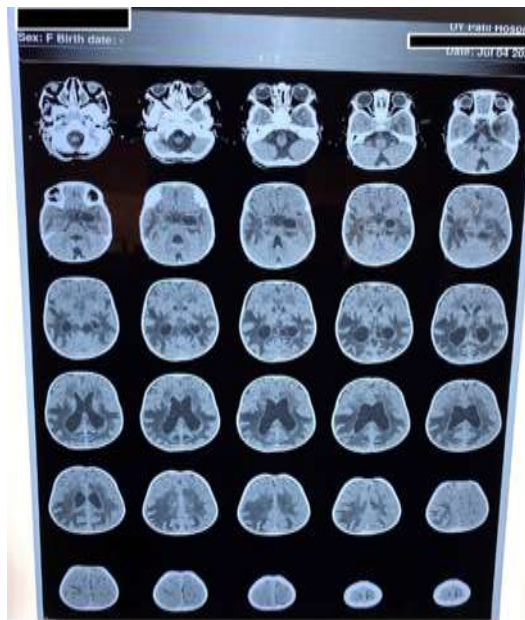


Fig 3: CT Brain: Immediately after shunt removal; CSF leak seen in brain parenchyma and ventricular dilatation is seen



Fig 4: CT Brain Bone window: Arrow showing the previous VP Shunt insertion site

Discussion

Hydrocephalus is a common neurological disorder in children, often secondary to structural brain malformations. The Dandy-Walker variant, a milder form within the Dandy-Walker complex, is characterized by partial agenesis or hypoplasia of the inferior cerebellar vermis, cystic dilatation of the fourth ventricle, and preserved posterior fossa size. These structural abnormalities can disrupt normal CSF flow, particularly at the level of the fourth ventricle, leading to progressive communicating hydrocephalus. In our patient, neuroimaging confirmed inferior vermian hypoplasia and fourth ventricular communication with the cisterna magna—hallmarks of the Dandy-Walker variant—along with corpus callosum thinning and choroidal fissure cysts. These features likely contributed to both the hydrocephalus and her developmental delay.

VP shunting is the standard of care for hydrocephalus, but shunt-related complications occur in 30–40% of cases, with infection being among the most serious. The risk is particularly high in infants and malnourished children due to immature immune responses and poor skin barrier function. Shunt infections often occur within six months of insertion and may present with subtle or delayed signs, such as fever, vomiting, seizures, or drowsiness. In this child, the presence of exposed and infected shunt tubing with purulent discharge and positive cultures for

Pseudomonas aeruginosa from both the CSF and shunt tip indicated a high-grade infection. Unlike the more common staphylococcal infections, *Pseudomonas* infections are highly resistant to antibiotics, and capable of forming robust biofilms on foreign materials. These characteristics necessitate shunt removal and targeted antimicrobial therapy. In our case, the child responded well to intravenous meropenem (sensitive on culture) and linezolid, and CSF parameters are in improving trend.

The incidental finding of situs inversus totalis—confirmed through imaging—adds further clinical and embryological interest. Situs inversus totalis, where thoracoabdominal organs are a mirror image of normal anatomy, is typically asymptomatic but clinically significant in surgical and diagnostic contexts. It is found in approximately 1 in 10,000 live births and may occur in isolation or as part of syndromes such as primary ciliary dyskinesia (PCD), Kartagener syndrome, or other laterality disorders. Embryologically, laterality is established early during gastrulation through the function of motile cilia in the embryonic node, which generate leftward flow of signaling molecules. Dysfunction of these cilia can disrupt left-right axis formation, potentially leading to both situs anomalies and neural tube defects such as Dandy-Walker malformations. This suggests a possible shared pathogenesis rooted in defective

ciliary motility or gene expression during early embryogenesis.

The co-occurrence of Dandy-Walker variant and situs inversus in this child supports the hypothesis of a broader ciliopathy or disturbance in embryonic patterning. Some studies have linked such associations to mutations in genes regulating ciliogenesis, such as DNAH11 and LRRC6, or to disruptions in planar cell polarity. Though this child did not have overt features of primary ciliary dyskinesia (such as chronic respiratory symptoms), her combination of CNS and visceral laterality defects warrants further genetic and syndromic evaluation.

This case exemplifies the diagnostic and management challenges in children with multiple congenital anomalies. It emphasizes the need for a high index of suspicion for shunt complications in children presenting with neurological symptoms, especially when structural brain anomalies and syndromic features coexist. A multidisciplinary approach—including paediatric surgery, infectious diseases, pediatrics, and nutrition—is essential to ensure timely intervention and improved outcomes.

Conclusion:

This case emphasises the critical vulnerability to shunt infections within the first six months post-insertion, particularly in pediatric patients with compromised immunity and malnutrition. The progression from a seemingly benign scalp boil to a full-blown *Pseudomonas* meningitis highlights a rare but significant extraluminal route of infection—direct skin breach over the shunt tract. The concurrent presence of Dandy-Walker variant and situs inversus totalis adds a unique dimension to this case. Such an unusual presentation emphasizes the importance of vigilant follow-up and early recognition of skin lesions over shunt sites.

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