

Pitfalls in Diagnosis of Shunt Malfunction in Children

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ABSTRACT

Background: In children with a potential shunt malfunction, pediatricians and emergency physicians are often the first to assess these children prior to neurosurgical referral. Most shunts fail within the first year (67%), with 14% of children having a shunt failure within the first month of insertion, and 40% within the second year of insertion. Classical symptoms of raised ICP, such as nausea, vomiting, coma and bulging fontanelle have a high predictive value for shunt failure, but are not always present. Some patients may be asymptomatic of shunt failure and the age of the patient will also confound this symptom spectrum. Clinical signs of shunt failure are well varied and age dependent. This article presents a series of four cases of shunted patients with atypical presentations of shunt malfunction. **Materials and Methods:** Clinical, radiological and surgical data of the patients were collected to demonstrate atypical presentations of shunt malfunction. All shunt malfunctions were confirmed upon review of the shunt in the operating theatre. **Results:** Four patients are shown to have atypical presentations of shunt malfunction. The first child was completely well despite an incidental finding of papilloedema on routine follow up, despite the lack of clinical and radiological evidence for shunt malfunction. The second child was completely well with a minor intermittent CSF leak at the drain wound. The third was an expanding pseudomeningocele after tumor resection. The final child had an expanding lower back pseudomeningocele but was otherwise well. All patients had resulting successful shunt revisions. **Conclusion:** It is important for neurosurgeons, paediatricians and emergency doctors to be able to identify these atypically presenting children and to correct their shunt malfunction to prevent long-term complications of insidious raised ICP, such as blindness or worsening of pre-existing neurological deficits and more importantly to prevent the child having a rapid and devastating decompensation.

Keywords: Shunt, Hydrocephalus, Intracranial pressure, Shunt malfunction.

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INTRODUCTION

Hydrocephalus is one of the most common pathologies managed by paediatric neurosurgeons. Hence, it is not unusual to come across shunt malfunction. Pediatricians and emergency physicians are often the first to assess these children prior to neurosurgical referral. Most shunts fail within the first year (67%), with 14% of children having a shunt failure within the first month of insertion, and 40% within the second year of insertion.¹ While symptoms such as nausea, vomiting, coma and bulging fontanelle have a high predictive value for shunt failure,^{2,3} they are not always present. Some patients may be asymptomatic of shunt failure and the age of the patient will also confound this symptom spectrum. Clinical signs of shunt failure are well varied and very much age dependent. A younger child with open fontanelles may only have bulging fontanelle, distended scalp veins, sun-setting,

increasing head circumference, bradycardia and desaturations. Older children have fixed skulls so may have signs that arise rapidly. It is vital to correctly diagnose shunt malfunction that leads to increased Intracranial Pressure (ICP). This article presents a series of four cases of shunted patients with atypical presentations of shunt malfunction.

Case 1

A 13-month-old boy routinely presented with a sacral myelomeningocele, repaired at birth. A Ventriculoperitoneal (VP) shunt was inserted on day 24 of life (with no revisions) and he had known strabismus, for which he had been having regular ophthalmology review. He had been well with no signs of shunt malfunction. On a recent ophthalmology routine visit, he noted to have papilloedema. He was immediately referred to the neurosurgeons and a CT brain scan was done. The CT scan was compared to previous cranial ultrasound imaging (CRUSS) and there was no significant change in ventricle size noted (Figure 1 a-b). His mother had not noted any abnormal behaviour or any worrying symptoms. Given the definite papilloedema and despite lack of other clinical or radiological evidence of shunt



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malfunction, he had surgical exploration of his shunt at which time the proximal catheter was noted to be blocked.

Post-operatively he remained well and currently attends routine 3 monthly ophthalmology reviews.

Case 2

A 3-month-old boy presented with a history of myelomeningocele (closed day 2 of life) with simultaneous VP shunt, requiring one revision for infection. The shunt infection occurred 5 days following initial shunt placement and was treated by removing the shunt and placement of an External Ventricular Drain (EVD). After a course of intravenous antibiotics, the EVD was removed, and a new shunt was placed. The child was discharged but returned at 3 months old after his mother had noted an intermittent leak of 'clear fluid' from the site of the previous EVD (right frontal area). She commented that the baby had been otherwise well, with no vomiting, feeding issues, strabismus or irritability despite this CSF leak. On examination, he was alert and interactive, his fontanelle was soft and there had been an increase of his head circumference. CRUSS compared to previous study showed an increase in the Levene Index from 38% to 50% (Figure 2a-b). He had a revision of his shunt at which time the proximal end was noted to be blocked.

Case 3

A 5-year-old boy presented for a second resection of a posterior fossa tumour. His initial resection had been 8 months earlier with histology confirming a grade 1 Pilocytic astrocytoma and he had a shunt inserted at that stage. Five days following his second resection, he was otherwise well but noted to have a fluctuant swelling over the operative site- a pseudomeningocele. This continued to increase gradually in size. CT confirmed the presence of a pseudomeningocele with small ventricles that had not changed in size in comparison to previous imaging (Figure 3C-D). He subsequently had exploration of his shunt at which there was no CSF flowing from the proximal catheter and this was replaced. Gradually the pseudomeningocele reduced in size.

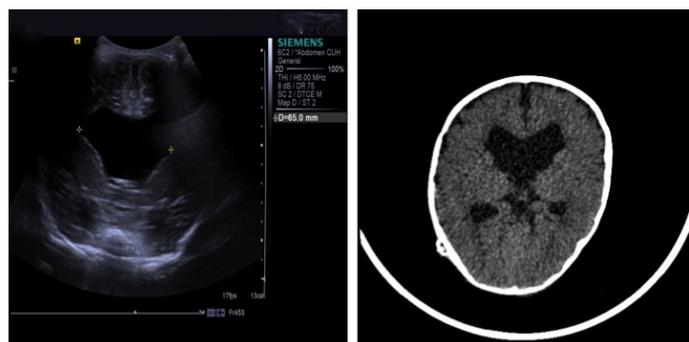


Figure 1: A. Cranial Ultrasound done after initial shunt insertion during a time the patient was well and asymptomatic for shunt failure. B. CT done at the time patient was noted to have papilloedema- there is no marked difference in ventricle size when compared to 1a.

Case 4

A 3-year-old girl with a history of repaired lumbar myelomeningocele and VP shunt, presented for routine follow-up with a long history of swelling of the lumbar area over the myelomeningocele repair site. She was otherwise well with no typical symptoms of shunt failure. Her mother noted that the swelling would intermittently deflate and recollect. MRI at the time of presentation demonstrated markedly dilated ventricles in comparison to previous imaging and a pseudomeningocele that had also increased in size (Figure 3A-B). She was admitted electively for shunt revision at which time it was noted the proximal catheter was blocked. This was replaced and subsequently over the following few days, the pseudomeningocele decompressed.

DISCUSSION

In daily practice, doctors rely initially on clinical history and examination as part of patient management. With shunted patients, the history may consist of acute symptoms in the case of complete shunt blockage or else symptoms may be prolonged or intermittent, as in the case of intermittent shunt blockage. Older children, as a result of fused sutures and closed fontanelles, have rigid craniums that are incapable of expanding to accommodate an increase in ICP that arises from shunt failure. As a result, they are more likely to present with rapid deterioration when their shunts fail whereas a child under the age of one may have more subtle symptoms of shunt failure initially, as their craniums expand with pressure.

The child's age, severe developmental delay or neurological deficit may preclude the shunted patient from giving a verbal account of their symptoms. In those cases, the parents or carers of a child are an invaluable source of information. Most parents know whether their child is deviating from his/her normal baseline, and can recount the child's previous presentation pattern with prior shunt failures, which may give the clinician a hint at the diagnosis. In children, the symptoms of shunt malfunction can often mimic those of other childhood illnesses so whenever there is doubt the safest course of action is a period of observation.

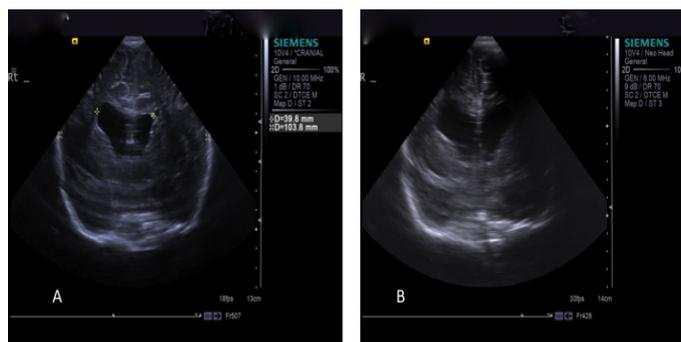


Figure 2: A. Cranial ultrasound taken after shunt insertion while patient was well. B. Cranial ultrasound at the time patient had a CSF leak.

Table 1: Summary of cases.

	Type of hydrocephalus	Presentation of shunt failure	Image findings	Management
Case 1 13-month-old boy	Congenital- Spina bifida	Well child Papilloedema	No change in ventricle size	Shunt revised
Case 2 3month old boy	Congenital- Spina Bifida	Well child CSF leak from EVD wound	Slight increase in ventricle size on CRUSS	Shunt revised
Case 3 5year old boy	Acquired- Posterior fossa tumour	Expanding pseudomeningocele	No change in ventricle size	Shunt revised
Case 4 3year old girl	Congenital- Spina Bifida	Well child Expanding lower back pseudomeningocele	Dilated ventricles	Shunt revised

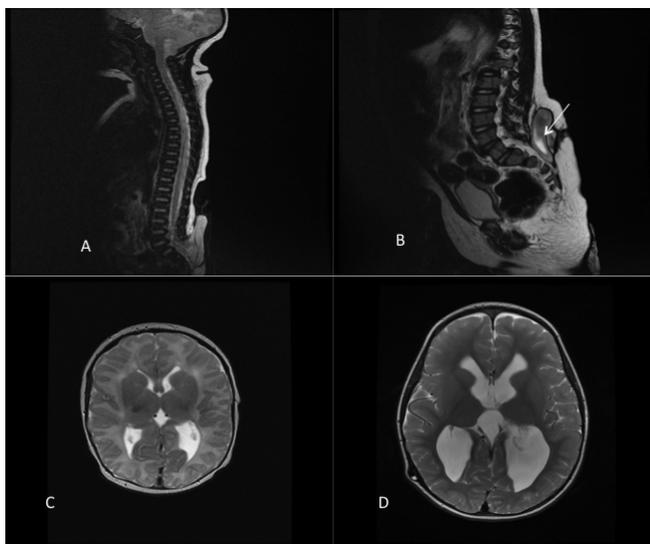


Figure 3: A-B. MRI spine before (a) and at presentation (b)- note obvious increased size of pseudomeningocele. C-D. Comparison of previous MRI Brain (c) and MRI prior to shunt revision (d).

An algorithm for management is described (Figure 4)

Pitfall 1: Papilloedema

Fundoscopy is a very useful, noninvasive tool for assessing for intracranial hypertension. Papilloedema is usually an accurate sign of raised ICP and associated visual loss is uncommon in the early stages of papilloedema.⁴ In shunted children, clinical symptoms, signs or radiological evidence of shunt failure usually accompany papilloedema. Isolated papilloedema is rarely the only sign of shunt failure as seen in case 1, but it has been reported.^{5,6} Whilst the presence of a normal optic disc does not always exclude shunt malfunction, the presence papilloedema is often correlated with intracranial hypertension as a result of shunt failure. Papilloedema usually develops after sustained exposure to raised pressure, so its sensitivity is reduced in the setting of acutely raised ICP and thus may be absent in acute shunt failure.⁷

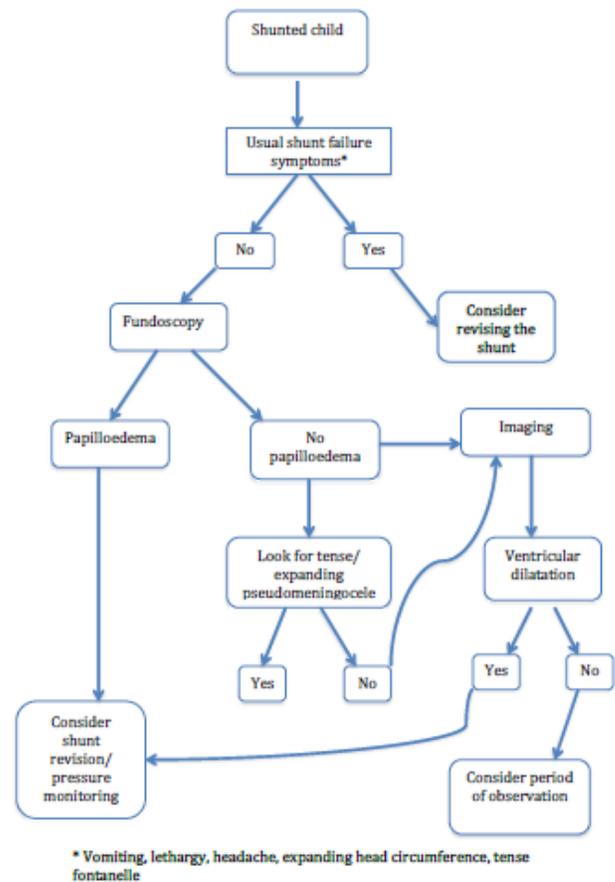


Figure 4: Proposed algorithm for managing atypical presentations of shunt failure.

As most clinicians do not routinely examine the optic nerve, subtle papilloedema can be easily missed. As a matter of course, children with shunts who present with ambiguous symptoms should have formal ophthalmological examination where feasible. Regular routine ophthalmologic assessment of shunted children may be useful in identifying those children in whom shunt failure presents only as papilloedema.

Pitfall 2: Imaging

Neuroimaging is an integral part of the management of shunted patients. In young children with an open anterior fontanelle, CRUSS is sufficient to assess ventricle size and any change in ventricle size if previous CRUSS are available for comparison.⁸ CT is a fast and readily available modality to investigate older children along with X-ray (Shunt Series). The utility of a shunt series has been called into question with a few authors finding that in many cases, the test rarely yields information that is not evident on CT or on clinical examination of the patient.⁹⁻¹¹ Cases of children with a shunt failure, despite lack of radiological evidence of the same, are not uncommon. The sensitivity of CT in detecting shunt malfunction can be low, with the majority of children having a shunt revision despite scans being reported as normal or no signs of raised ICP.⁹ Furthermore, children with slit ventricle syndrome, as a result of poor ventricular compliance, will classically have unaltered imaging while being symptomatic for raised ICP, and so gaining a detailed medical history is key.

Neuroimaging in shunt failure is only as good as the previous imaging being used as a comparison. It is not sufficient to use images taken at the episode of a previous shunt failure as a comparison. This may result in misplaced reassurance as the ventricles may not have dilated to the same extent as the previous episode and hence may look smaller in comparison. Only imaging taken when the child is asymptomatic should be used as a comparison to imaging when the child is symptomatic.^{2,11,12} It is also important to compare like modalities where possible.

Shunted children should ideally have a “baseline” scan done at least 3 months following shunt revision as a reference point. In case 1, the child had no prior CT scan to compare with and comparison with a CRUSS was made. It can be difficult to identify subtle changes in this case.

Pitfall 3: Child looks well

Various situations exist where a child may be well but have an underlying shunt problem. A blocked shunt, in a child under 12 months, may present as a full fontanelle or rapidly expanding head circumference. The open fontanelles and un-fused sutures in this case allow the cranium to expand to accommodate the increasing intracranial pressure. An expanding pseudomeningocele even in a well child is usually a sign of intracranial hypertension and should arouse the suspicion of a blocked shunt. Small fluid collections directly over the shunt hardware are commonly seen in some children with malfunctioning shunts. Another variant of accommodating this pressure is seen in our second and third cases where a skull and dural defect, no matter how small, acts as a release valve for the increasing pressure. Case 4 had a congenital bone defect and dural defect from her myelomeningocele, which also offered the CSF a path of least resistance to relieve rising ICP. Once the CSF is distributed through these defects the pressure

reduces, the clinical symptoms of shunt failure may not develop and child remains well.

The presence of tumours often predisposes to shunt blockage as some tumours produce highly proteinaceous material.¹³ Another risk factor for shunt blockage is the presence of surgical debris following a procedure, such as a child post tumour resection. Table 1 summarises the cases discussed.

In all these cases it is important to be able to identify these children and to correct their shunt malfunction (Figure 4) to prevent long-term complications of insidious raised ICP, such as blindness or worsening of pre-existing neurological deficits and more importantly to prevent the child having a rapid and devastating decompensation.¹⁴

CONCLUSION

In all these cases, it is vital that the treating Paediatricians, Emergency physicians, Neurologists and Neurosurgeons should be able to identify these children and to correct their shunt malfunction as elucidated in the algorithm-Figure 4. This is aimed to prevent long-term complications of insidious raised ICP, such as blindness or worsening of preexisting neurological deficits and more importantly to prevent the child having a rapid and devastating decompensation.

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