

Death after late failure of third ventriculostomy in children

Report of three cases

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✓ Late failure following successful third ventriculostomy for obstructive hydrocephalus is rare, and death caused by failure of a previously successful third ventriculostomy has been reported only once. The authors present three patients who died as a result of increased intracranial pressure (ICP) after late failure of a third ventriculostomy.

Through a collaborative effort, three patients were identified who had died following third ventriculostomy at one of the authors' institutions. A 13-year-old girl with neurofibromatosis Type 1 underwent third ventriculostomy for obstructive hydrocephalus caused by a tectal lesion. Three years later her condition deteriorated rapidly over the course of 6 hours and she was found dead at home. A 4-year-old boy treated with third ventriculostomy for aqueductal stenosis presented 2 years postoperatively with symptoms of increased ICP. This patient suffered a cardiorespiratory arrest while under observation and died despite external ventricular drainage. A 10-year-old boy with previous ventriculoperitoneal (VP) shunt placement underwent conversion to a third ventriculostomy and shunt removal. Eight months after the procedure his condition deteriorated, with evidence of raised ICP, and he underwent emergency insertion of another VP shunt, but remained in a vegetative state and died of complications. Neuropathological examinations in two cases demonstrated that the third ventriculostomy was not patent, and there was also evidence of increased ICP.

Late failure of third ventriculostomy resulting in death is a rare complication. Delay in recognition of recurrent ICP symptoms and a false feeling of security on the part of family and caregivers because of the absence of a shunt and the belief that the hydrocephalus has been cured may contribute to fatal complications after third ventriculostomy. Patients with third ventriculostomies should be followed in a manner similar to patients with cerebrospinal fluid shunts.

KEY WORDS • third ventriculostomy • complication • children

COMPLICATIONS associated with the treatment of obstructive hydrocephalus with third ventriculostomy are uncommon,^{3,7,11,18} but they are often serious in nature,^{1,2,8,14} although they are rarely fatal.¹⁶ Late failure of third ventriculostomy is often heralded by the recurrence of symptoms observed during the initial presentation with hydrocephalus. Death due to increased ICP resulting from the late failure of a previously successful third ventriculostomy has been reported only once.¹⁰ We present three cases of children who died of increased ICP after the late failure of a successful third ventriculostomy.

Clinical Material and Methods

Three patients were identified, one each of whom were treated at the Hospital for Sick Children, Toronto, Canada, the British Columbia's Children's and Women's Hospital,

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computerized tomography; ICP = intracranial pressure; MR = magnetic resonance; VP = ventriculoperitoneal.

Vancouver, Canada, or the Southampton General Hospital, Southampton, United Kingdom, and who had died after undergoing a successful third ventriculostomy for obstructive hydrocephalus. Patient records were obtained and the following data were recorded: diagnosis, age at time of third ventriculostomy, previous treatment, third ventriculostomy method, clinical and radiological follow up, time from third ventriculostomy until death, and results of neuropathological examination.

Case Reports

Case 1

This 13-year-old girl with known neurofibromatosis Type 1 presented with a 6-month history of progressive headaches, poor concentration, ataxia, and evidence of papilledema on examination. Admission CT and MR studies demonstrated marked hydrocephalus involving the third and lateral ventricles, along with evidence of aqueductal stenosis (Fig. 1 *left*). The patient underwent third ventricu-

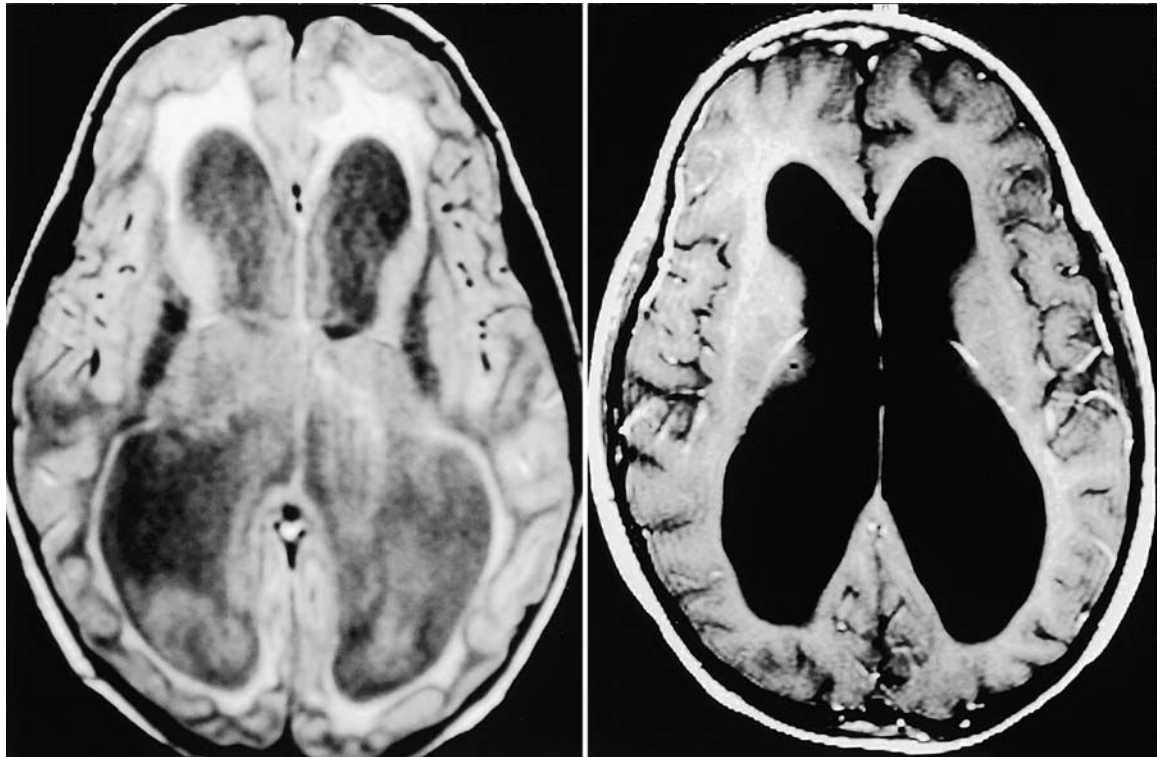


FIG. 1. Case 1. *Left:* Axial fluid-attenuated inversion-recovery MR image demonstrating enlargement of lateral ventricles with surrounding periventricular edema. *Right:* Axial T₁-weighted MR image obtained after third ventriculostomy demonstrating resolution of periventricular edema and an increase in convexity CSF.

lostomy involving blunt perforation of the thinned third ventricle floor and enlargement with a Fogarty balloon catheter. At the end of the procedure, radioactive tracer was injected into the lateral ventricle for postoperative examination of the patency of the ventriculostomy. A CT scan obtained 24 hours after the procedure demonstrated normal passage of the tracer through the patent ventriculostomy and circulation around the cerebral convexities and spinal subarachnoid space, in addition to a slight decrease in ventricular size.

All symptoms were resolved at the initial follow-up review, and CT scanning demonstrated a slight decrease in ventricular size, with resolution of periventricular edema and an increase in pericerebral spaces. On MR images obtained 2 years after presentation (Fig. 1 *right*), a stable decrease in ventricular size and a new 5-mm nonenhancing lesion were observed in the midbrain, obstructing the cerebral aqueduct. No flow void could be identified; however, the floor of the third ventricle was slack and therefore the ventriculostomy was believed to be patent. At her 3-year follow-up review, the patient was well and no neuroimaging was performed.

Six months after her last follow-up visit, she awoke, was well, and went to school, only to return home suffering from headache and vomiting, which she reported to her mother by phone. Several hours later she was found dead at home. Postmortem examination demonstrated marked ventricular enlargement with evidence of cerebral edema, as well as uncus and cerebellar tonsillar herniation. The floor of the third ventricle was noted to be covered by a thin gliotic membrane with discontinuous segments of arachnoid

membrane. On histological examination the tectal lesion was confirmed to be a pilocytic astrocytoma.

Case 2

This 4-year-old boy presented with a 2-month history of headache, nausea, and vomiting, and there was evidence of a sixth cranial nerve paresis. Admission MR imaging demonstrated triventricular hydrocephalus caused by aqueductal stenosis (Fig. 2 *upper left and center*). A third ventriculostomy was performed using blunt perforation with water dissection, followed by external ventricular drainage for 1 day. At the 2-month follow-up visit, all symptoms were resolved, and a CT scan performed at the 3-month follow up demonstrated that the ventricles were significantly reduced in size (Fig. 2 *upper right*). The patient was well at 16 months of follow up, at which time repeated imaging was not performed.

Two years after his initial presentation, the patient awoke with a headache and vomited after having been well the night before. He vomited four more times that day and was admitted to an emergency department, distant from the location of his initial treatment, with complaints of persistent headache and malaise. Results of his neurological examination were normal. A CT scan was obtained that demonstrated significant enlargement of the third and lateral ventricles, although no earlier films were available for comparison (Fig. 2 *lower left*). The patient was admitted for observation. He complained of severe headache in the middle of the night and was noted to be lethargic on assessment, but he was easily rousable and acting appropriately. Later

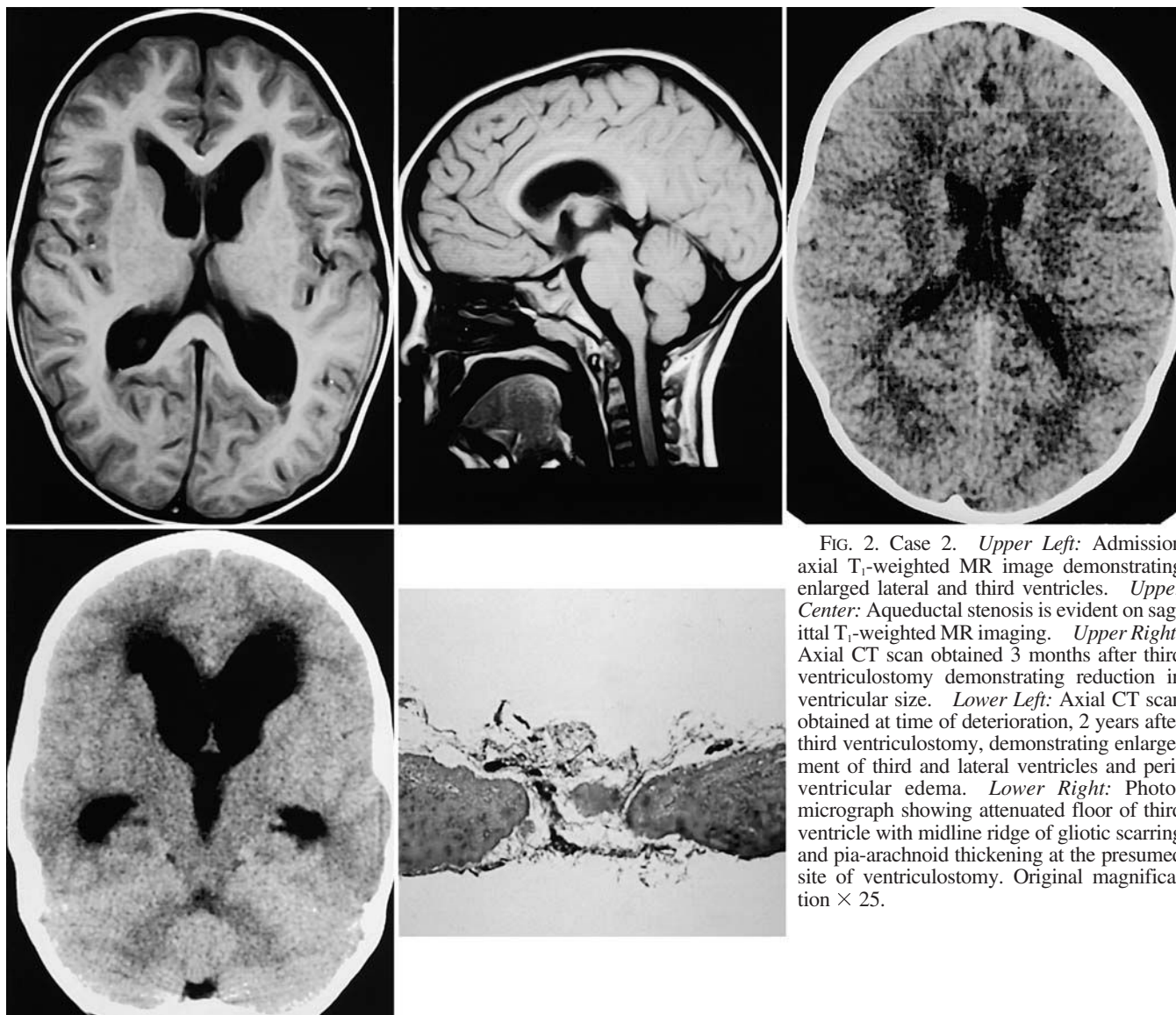


FIG. 2. Case 2. *Upper Left:* Admission axial T₁-weighted MR image demonstrating enlarged lateral and third ventricles. *Upper Center:* Aqueductal stenosis is evident on sagittal T₁-weighted MR imaging. *Upper Right:* Axial CT scan obtained 3 months after third ventriculostomy demonstrating reduction in ventricular size. *Lower Left:* Axial CT scan obtained at time of deterioration, 2 years after third ventriculostomy, demonstrating enlargement of third and lateral ventricles and periventricular edema. *Lower Right:* Photomicrograph showing attenuated floor of third ventricle with midline ridge of gliotic scarring and pia-arachnoid thickening at the presumed site of ventriculostomy. Original magnification $\times 25$.

that morning he suffered a cardiorespiratory arrest and died, despite full resuscitation efforts, including the insertion of an external ventricular drain. Autopsy results showed chronic hydrocephalus due to an acquired aqueductal stenosis caused by gliotic ependymal scarring (granular ependymitis) extending into the fourth ventricle. At the presumed site of the ventriculostomy, there was a focal zone of gliotic thickening and broadening of the pia-arachnoid within the apex of the ballooned (dilated) third ventricular floor, which was indicative of sealing by reactive gliosis and pia-arachnoid proliferation (Fig. 2 lower right). The cause of death was acute transforaminal herniation of the cerebellar tonsils.

Case 3

This 9-year-old boy, in whom a VP shunt had been placed when he was 2 days old, which had only been revised once previously (shortly after its initial insertion), presented with a 2-week history of headache and bilateral sixth

cranial nerve palsies. A CT scan demonstrated evidence of mild ventriculomegaly and an MR study confirmed the diagnosis of aqueductal stenosis; therefore, a third ventriculostomy was considered. While he was on the ward, the boy became unresponsive, with abnormal respirations necessitating emergency third ventriculostomy. The ventriculostomy was performed using blunt perforation, and it was widened by passage of the scope into the prepontine cistern and avulsion of some thinned fragments of the floppy floor. The patient awoke immediately postoperatively.

Follow-up MR images obtained 3 months after the third ventriculostomy revealed a slight reduction in ventricular size but no obvious flow void. The patient was well at his 8-month follow-up visit. One week later, however, he suffered a severe occipital headache in the early evening, and was found unconscious shortly after his parents were advised to go to the local hospital's emergency department for assessment. There the patient was initially irritable and disoriented, and he suffered a respiratory arrest that required intubation before transfer. A CT scan performed on arrival

at the neurosurgical center, within 2 hours of his presentation, showed severely enlarged ventricles. A VP shunt was inserted by which time his pupils were fixed and dilated despite mannitol therapy. His pupils became small and reactive again on return to the recovery room. Follow-up CT scans demonstrated decompressed ventricles and signs of prior transtentorial herniation, with left posterior cerebral artery infarction. The patient remained in a vegetative state and died 2 months later after withdrawal of life support.

Discussion

Delayed failure after successful treatment of obstructive hydrocephalus by third ventriculostomy is thought to be uncommon in the modern era and is reported to occur in only 2 to 15% of treated patients,^{3,5,6,9,11,18,19} although definitions of delayed failure, indications for retreatment, and series of patients with long-term follow up are lacking. Failures that resulted in a second CSF diversionary procedure or death more than 6 months after the initial ventriculostomy occurred in only three of 51 patients in a series reported from the Hospital for Sick Children.¹⁹ Late failures have been reported as long as 6 years after a successful procedure.³ These are thought to occur as a result of obstruction of the ventriculostomy caused by arachnoid and glial scarring, based on radiological observations and on those noted during repeated endoscopy.^{3,6} Death resulting from increased ICP after the failure of a previously successful third ventriculostomy has been reported only once.¹⁰ Neuropathological examinations in two of our patients showed that scarring was present, which confirmed that the loss of patency of the third ventriculostomy was responsible for the fatal failure.

Predicting which patients may be at risk for later failure after successful third ventriculostomy is difficult. Imaging correlates of successful third ventriculostomy include a reduction in ventricular size and the presence of a flow void on T₂-weighted MR imaging,^{4,5,12,13} although a resolution of periventricular edema and an increase in subarachnoid space over the convexities with no change in ventricular size^{3,12} may be seen in clinical successes. Ventricular volume changes after successful third ventriculostomy have been described as varying from a mean of only 11%¹³ to as much as 36%.⁶ Flow voids on T₂-weighted MR imaging or cine phase-contrast imaging are evidence of a patent third ventriculostomy and correlate highly with a good clinical picture.^{6,12,19} The absence of a flow void, although occasionally present in patients with a good clinical outcome, was found in 12 of 13 patients presenting with delayed treatment failure in a series reported by Cinalli, et al.³

Late failure of the third ventriculostomy in our patients occurred a mean of 25 months after the initial procedure, and each died after a short prodrome of symptoms lasting less than 24 hours. The reasons for the rapid progression of the symptoms and subsequent deaths is unclear; however, the complete blockage of flow at the orifice, which was confirmed on neuropathological examination in two patients, and the presence of shunt-dependent noncompliant brain in the other patient may have contributed. Postoperative MR imaging in two patients revealed no obvious flow void and a minimal change in ventricular size, although an improvement in periventricular edema and sulcal patterns was evident, along with a stable clinical picture. In one pa-

tient no postoperative MR study was performed, and in two patients no follow-up imaging was performed in the year before they suffered fatal deterioration. Cinalli, et al.,³ reported on three of 114 patients with successful third ventriculostomy in whom an asymptomatic enlargement of the ventricles developed on follow-up review and who underwent a second diversionary procedure. Routine imaging may be important to identify patients with asymptomatic increases in ventricular size and those with an absent flow void who may be at risk for delayed failure. The duration of such follow up is unclear although in the series by Cinalli, et al., all failures occurred in the first 5 years posttreatment.

Death as a result of increased ICP is an uncommon complication of shunt malfunction and is reported to occur in 1 to 2.8% of children who receive VP shunts.^{15,17} Sgouros, et al., described two patients who died of raised ICP after a sustained period of years of apparent stability without shunt revisions. Both were admitted to primary hospitals for observation, with new symptoms that were not appreciated. An important factor believed to be responsible for death from shunt malfunction was the underappreciation of related symptoms and the delay in taking appropriate action. Symptoms of increased ICP in one of our patients may have been underappreciated in part because of the absence of a shunt. Delay in the recognition of recurrent ICP symptoms, a false feeling of security on the part of family and caregivers because of the absence of a shunt, and the belief that the hydrocephalus has been cured may contribute to fatal complications after third ventriculostomy.

Conclusions

Late failure of third ventriculostomy that results in death from increased ICP is a rare, seldom-reported complication. Surveillance imaging performed on a regular basis after third ventriculostomy to detect asymptomatic changes in ventricular size or loss of patency of the ventriculostomy may allow us to identify patients at risk for late deterioration. Education of families and caregivers about the possibility that late deterioration may occur after third ventriculostomy is important to allow prompt recognition of new symptoms related to increased ICP and to permit timely intervention. Also, patients with third ventriculostomies should probably be followed in a similar manner to patients with CSF shunts.

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