A unifying theory for the multifactorial origin of cerebellar tonsillar herniation and hydrocephalus in osteopetrosis

Clinical article

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Object. Osteopetrosis is a rare congenital metabolic bone disease. There are very few reports in the literature associating cerebellar tonsillar herniation (CTH) and hydrocephalus requiring neurosurgical attention. The authors present cases of osteopetrosis requiring neurosurgical intervention from their practice and offer a detailed account of the literature.

Methods. A retrospective review was conducted at the authors’ institution, and all children with osteopetrosis requiring neurosurgical attention were identified. Medical charts and radiographic studies were reviewed. Data including age at presentation, sex, symptoms at presentation, age at follow-up, the presence of any neurological comorbidities, and surgical procedures performed were recorded.

Results. Four patients were identified as having osteopetrosis requiring neurosurgical attention at the authors’ institution between January 1, 2005, and January 1, 2014. There were 3 females and 1 male with an average age at presentation of 11.1 years; patients were observed for a mean of 4.4 years. All of the patients were identified as harboring jugular foraminal stenosis and CTH. Seventy-five percent of these patients developed hydrocephalus, and in those cases a triventricular pattern of dilation was noted. One patient developed syringomyelia. Three of the 4 patients underwent neurosurgical procedures. Cerebrospinal fluid diversion was performed in 2 patients via a ventriculoperitoneal shunt in one case and an endoscopic third ventriculostomy (ETV) in the other. The former patient required a proximal revision at 2 years for bony overgrowth at the site of the bur hole. Two patients underwent a suboccipital decompression. In patients undergoing CSF diversion, there was improvement in ventricle size.

Conclusions. Variable degrees of hindbrain crowding and/or CTH are mentioned throughout the literature, suggesting that this entity is nearly always present in this patient population. The progressive triventricular hydrocephalus seen in these cases results from a complex combination of both communicating and noncommunicating pathophysiology, which may depend on the type of osteopetrosis, age at presentation, and the presence and degree of venous collateralization, and it appears that the hydrocephalus is more prevalent and more likely to be treated in infants and in the younger, school-aged population. The acquired hindbrain fullness in conjunction with the triventricular pattern of hydrocephalus has kept the authors enthusiastic regarding the use of ETV in these complicated cases.

Key Words • osteopetrosis • cerebellar tonsillar herniation • hydrocephalus • endoscopic third ventriculostomy • ventriculoperitoneal shunt

Osteopetrosis is a rare congenital metabolic bone disease characterized by failure of osteoclasts to resorb bone, leading to increased bone density with the potential for bone marrow failure. Although a myriad of conditions arise in this heterogeneous disease, the most debilitating consequences arise from narrowing of cranial foramina, which leads to cranial neuropathies and brainstem compression.1,14,20,56

Osteopetrosis is categorized into 3 forms with the infantile variant having the worst prognosis. Typically inherited in an autosomal-recessive fashion, the infantile variant is considered malignant and is usually fatal by 2 years of life. The intermediate and autosomal-dominant forms usually present later in life and do not typically carry a lethal course.20

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There are very few reports in the literature associating posterior fossa crowding, cerebellar tonsillar herniation (CTH), and hydrocephalus in osteopetrosis.2,12,32,63 The etiology of hydrocephalus in this scenario is incompletely understood; however, both communicating and noncommunicating pathophysiology may be present.

We present 4 patients with osteopetrosis who developed varying degrees of CTH and hydrocephalus, and we discuss the neurosurgical treatment options considered

Abbreviations used in this paper: CTH = cerebellar tonsillar herniation; ETV = endoscopic third ventriculostomy; ICP = intracranial pressure; MRV = MR venography; ONSF = optic nerve sheath fenestration; SOD = suboccipital decompression; VP = ventriculoperitoneal.
in these select cases. The pathophysiology of CTH and hydrocephalus in osteopetrosis is discussed in relation to similar processes with review of the relevant literature.

**Methods**

After obtaining institutional board review (Children’s Medical Center Dallas), we identified patients with osteopetrosis who were referred for neurosurgical consultation between January 1, 2005, and January 1, 2014, using a clinical pediatric neurosurgery database (Neurosurgeons for Children). Patients, charts, and radiographic studies were reviewed for age at presentation, sex, symptoms at presentation, age at follow-up, the presence of any neurological comorbidity, and surgical procedures performed.

**Results**

Between January 1, 2005, and January 1, 2014, 4 patients with osteopetrosis were referred for neurosurgical consultation and were evaluated by Neurosurgeons for Children in Dallas, TX. This cohort also represented the entire complement of children with this disease during this time frame at our institution. There were 3 females and 1 male in this cohort with an average age at presentation of 11.1 years (range 4 months–20 years). The mean follow-up in these cases was 4.4 years (range 6 months–7 years). All patients demonstrated jugular foraminal stenosis with resultant cranial venous outflow obstruction and CTH. Three of the 4 patients (75%) developed hydrocephalus with a triventricular dilation pattern being observed (small fourth ventricle and dilated third and lateral ventricles). One patient (25%) presented with syringomyelia and myelopathy.

Two patients with hydrocephalus underwent neurosurgical treatment; one underwent ventriculoperitoneal shunt (VP) placement, and more recently, the other underwent endoscopic third ventriculostomy (ETV). In both surgically treated cases, there was improvement in the ventricle size and periventricular edema. Two patients underwent suboccipital decompression (SOD) with decompression of the foramen magnum. There were no operative complications associated with any of the neurosurgical procedures.

**Case 1**

This 8-month-old girl, born full-term, initially presented at 1 month of age with irritability, high parathyroid levels, and low phosphate levels. She was started on calcium supplementation and presented again at 4 months with irritability, poor feeding, decreasing milestones, vision loss, and a bulging fontanelle. Further investigation revealed pancytopenia, splenomegaly, and sclerotic bone on skeletal bone survey (Fig. 1A). MRI demonstrated mild prominence of the ventricular system (Fig. 1B).

The patient was diagnosed with a TCIRG1 gene mutation and autosomal-recessive osteopetrosis. At 7 months of age she underwent chemotherapy, total lymphoid and abdominal irradiation, and subsequent bone marrow transplantation. She received immunosuppressive and prophylactic antibacterial and antiviral chemotherapy. Her medical course was complicated by respiratory insufficiency, hypertension, prolonged QT intervals, and poor feeding. During this time, she developed irritability and underwent brain MRI (Fig. 1C) and MR venography (MRV). Compared with prior imaging, the posterior fossa structures appeared more crowded, and the third and lateral ventricles had increased in size. MRV demonstrated venous outflow obstruction, characterized by markedly diminutive junctions of the sigmoid sinus and internal jugular veins bilaterally, without evidence of venous collateralization.

After correction of thrombocytopenia, at 8 months of age the child underwent an uncomplicated ETV. Postoperative MRI demonstrated a patent third ventricular stoma, improvement in the ventricle size, and decrease in the periventricular edema. She has been monitored now for an additional 6 months and has required several repeat admissions for fever, bacteremia, and apneic spells; while she continues on multiple medical therapies, her irritability and feeding have significantly improved.

**Case 2**

At the age of 12 years, this boy with known osteo-
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trosis presented with developmental delay, restrictive lung disease, worsening respiratory status, feeding issues requiring gastrostomy tube placement, chronic mandibular osteomyelitis, hypertension, visual and hearing impairments, and adrenal insufficiency. Because of recurrent infections he was admitted for sepsis. During his workup, he underwent head CT and brain MRI and MRV. The head CT demonstrated marked calvarial hyperostosis and prominence of the ventricular system (Fig. 2A). MRV demonstrated absence of the sigmoid sinuses and jugular bulbs with extensive venous collateralization, dilation of the posterior condylar emissary veins, and dilated superficial scalp veins.

At 17 years of age, the patient developed a severe pulmonary infection and sepsis requiring additional hospitalizations. During this time he underwent brain MRI, which demonstrated worsening dilation of the lateral and third ventricles with increased periventricular edema, as well as caudal extension of the cerebellar tonsils (Fig. 2B). Due to the magnitude of his clinical illness, the patient was not clinically fit to undergo any neurosurgical interventions and died shortly thereafter in the intensive care unit.

Case 3

This 12-year-old girl with osteopetrosis and a history of hypertelorism surgery earlier in childhood presented with progressive vision loss. Workup included head CT, which demonstrated triventricular hydrocephalus. A VP shunt was placed, and decompression of the ventricular system was achieved (Fig. 3A). Brain MRI and MRV were performed shortly thereafter, demonstrating CTH, markedly abnormal bidirectional movement of neural tissue at the cervicomedullary junction, and effacement of CSF both anterior and posterior to the craniovertebral junction. Narrowing of the jugular veins bilaterally at the skull base was also noted, as were prominent suboccipital transdiploic venous collaterals. Syringomyelia was also identified (Fig. 3B).

Two years later, the patient presented with evidence of shunt malfunction. She was taken for revision, and it

![Fig. 2. Case 2. A: Head CT (left) demonstrating marked calvarial hyperostosis and mild prominence of the lateral ventricular system. Lateral skull radiograph (right) obtained at that time, demonstrating the magnitude of hyperostosis that is present in these cases. B: Noncontrast sagittal (left) and coronal (right) brain MR images obtained at 17 years, demonstrating the characteristic diminutive posterior fossa compartment and resultant CTH along with triventricular hydrocephalus.](image)

![Fig. 3. Case 3. A: Head CT demonstrating the preoperative triventricular hydrocephalus (left), and the postoperative ventricular decompression after VP shunt placement (right). B: Noncontrast sagittal brain MRI (left) and MRV (right) demonstrating CTH, evidence of a cervical syrinx, and jugular venous stenosis with evidence of marked venous collateralization. C: Noncontrast spine MRI (left) demonstrating syringomyelia. Postoperative foramen magnum decompression was performed in this case (right).](image)
was noted at time of surgery that the valve was covered in bone along with bony overgrowth at the bur hole site. Poor spontaneous flow from the proximal end of the catheter was identified, and the proximal catheter and valve were replaced. Shortly thereafter, the patient developed progressive back pain, myelopathy, and difficulty walking. Imaging demonstrated an increased size of the syrinx, and the patient subsequently underwent SOD of the foramen magnum without duraplasty (Fig. 3C). Although preferable, due to the extreme bony overgrowth and subsequent limited working area within the suboccipital space, it was not felt safe or feasible to perform duraplasty this case. Follow-up spinal imaging demonstrated a mild improvement in her syrinx, without complete resolution.

The patient has since been observed for 7 years; she has impaired but stable vision, stable hearing difficulties, continued weakness, and hyperreflexia in the lower extremities; and frequent headache and dizzy episodes. Follow-up imaging has demonstrated stability of her syringomyelia and decompressed ventricles.

Case 4

This 20-year-old woman with a history of osteopetrosis presented with increased lower-extremity reflexes, gait abnormalities, and complaints of occipital headaches. Brain MRI demonstrated normal-appearing ventricles and herniation of the cerebellar tonsils through the foramen magnum. MRV demonstrated diminutive jugular veins, occluded sigmoid sinuses, and venous collateralization through the mastoid and condylar emissary veins into a dilated suboccipital venous plexus (Fig. 4). This patient underwent SOD with duraplasty and tonsillar resection. She has been observed now for 5 years, and her headaches and myelopathy have improved; however, she continues to have a persistently wide-based gait.

Discussion

Osteopetrosis is a rare and potentially severe congenital metabolic bone disease that is characterized by the failure of osteoclasts to resorb bone, which leads to increased bone density and the potential for bone marrow failure. Although a myriad of conditions can arise in this disease, such as failure to thrive, susceptibility to infections, anemia, thrombocytopenia, and hepatospleno-megaly, the most debilitating consequences are those that affect the nervous system via narrowing of cranial nerve foramina.

First described in 1904 by Albers-Schonberg, human osteopetrosis has been categorized into 3 types: infantile or autosomal recessive, intermediate, and autosomal dominant, with the infantile type having the worst prognosis. This type is typically considered “malignant,” or “lethal,” and is thought to affect roughly 1 in every 200,000 children, carries a nearly 100% mortality at 10 years, and is usually fatal by 2 years of life. The mutation in over 50% of cases involves the TCIRG1 gene, which encodes the a3 subunit of the vacuolar proton pump responsible for acidifying the bone/osteoclast interface. The only known effective treatment for this disease is matched bone marrow transplantation. The intermediate and autosomal-dominant forms are more benign, usually present later in life, and affected individuals typically have a full life expectancy.

Involvement of Pediatric Neurosurgery

The role of pediatric neurosurgery in cases of osteopetrosis typically involves treatment of 1 or more progressive entities, including increased intracranial pressure (ICP), cranial neuropathies, CTH, or hydrocephalus. The decrease in cranial capacity related to thickening of the calvaria is thought to contribute to elevated ICP in these cases, and narrowing of the cranial nerve foramina has led to deafness, facial paresis, blindness, and sequelae of brainstem compression. Cervical and foramen magnum stenosis in these cases has also caused spinal cord compression, resulting in myelopathy and syringomyelia requiring surgical decompression. Variable degrees of posterior fossa crowding and CTH in osteopetrosis are also frequently mentioned in available literature and appear to be common findings in these rare cases, with the majority of cases associated with concomitant triventricular hydrocephalus.

Pediatric neurosurgical intervention in these rare cases has been described since the late 1960s, when Baird et al. first described VP shunting for a 3-month-old boy with osteopetrosis and hydrocephalus. While mostly outside of the United States, several reports have surfaced since...
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that time, describing children with this disease coming to neurosurgical attention.2,5,9,12,20,25,32,35,63

Increased ICP

In osteopetrosis, the decrease in cranial capacity resulting from thickening of the calvaria contributes to the elevated ICP seen in these cases.2,4,6,20,39 In 1963, Klintworth31 reported on high CSF pressure evidenced on ventriculography in a patient with osteopetrosis. In that report, the ventricles were small, suggesting a possible pseudotumor-like phenomenon. Similar findings in a case of infantile osteopetrosis were again reported in 1977 by Lehman et al.35 More recently, Mirsadeghi et al.39 reported on a 19-year-old woman presenting with headache, papilledema, elevated CSF pressure, and small ventricles. The posterior fossa was not mentioned in this report; however, MRV demonstrated jugular venous stenosis. This patient was treated with a lumbo-peritoneal shunt, which reportedly ameliorated the headache and corrected the papilledema.

Visual loss and papilledema in osteopetrosis may be the result of increased ICP, or as a result of optic canal compression. Thus, while some patients require optic nerve sheath fenestration (ONSF), others may benefit from optic canal decompression. The presence of papilledema necessitating ONSF and/or optic canal decompression in this patient population has been reported in the ophthalmology literature since the late 1970s.1,4,23,54,64 More recently in 2006, Allen et al.4 described the case of a 33-year-old man with osteopetrosis who presented with decreasing visual acuity, papilledema, and an elevated opening CSF pressure. The facial CT did not demonstrate severe optic canal stenosis. Neither the ventricular size nor the posterior fossa anatomy in this case was mentioned. Vision loss in this case was believed to be secondary to increased ICP as opposed to a compressive neuropathy, and ONSF was performed, resulting in improvement in both visual acuity and papilledema. This case underlines the importance of determining the precise cause for vision loss in these cases. Although limited to a single report,28 it bears mentioning that reversal of optic canal stenosis has been identified after bone marrow transplant in an 8-month-old child with malignant osteopetrosis.

Cerebellar Tonsillar Herniation

While various theories exist, the etiology of CTH in osteopetrosis has been incompletely understood. We believe the cause this condition results from not only venous hypertension (producing an abnormal back pressure on CSF drainage inducing cerebellar engorgement), but also the diminutive size of the posterior fossa from calvarial hyperostosis. Together, these phenomena produce an acquired CTH, which generates a mechanical CSF outflow obstruction at the level of the cisterna magna and the cranio-cervical junction.

Variable degrees of posterior fossa crowding and CTH in osteopetrosis have frequent mention in available literature and thus appear to be common findings in these rare cases, with the majority of these cases associated with concomitant triventricular hydrocephalus.2,5,9,12,20,25,32,35,63 In 2007, Kulkarni et al.32 reported on a 15-year-old Indian boy presenting with headache, bulbar dysfunction, extremity motor loss, and hyperreflexia. Imaging in this case identified CTH and a cervical syrinx. There was no mention of the ventricular configuration, treatment, or outcome in this case. In the report by Al-Tamimi et al.,27 all of the 3 children described presented with various degrees of CTH, and while 1 child died in infancy, the other 2 children continued to complain of occipital and neck pain thought to be related to hindbrain herniation. In 2009, Jamjoom et al.25 reported on a 4-year-old British boy with autosomal-recessive osteopetrosis, oxycephaly, and CTH. Unfortunately, this child died intraoperatively during cranial expansion surgery. In 2011, Dlouhy et al.22 reported on a 25-year-old woman presenting with headache and loss of her gag reflex. Brain MRI demonstrated severe posterior fossa calvarial thickening, a small posterior fossa, and CTH with compression and deformation of the lower brainstem. She underwent suboccipital craniectomy, C-1 laminectomy, and duraplasty, which reportedly resolved her preoperative symptoms. Most recently, Mahmoud Adel et al.25 reported on a 9-month-old Saudi Arabian child who was found to have CTH in the presence of hydrocephalus and clinical myelopathy.

Hydrocephalus and Its Treatment

The presence of postmortem ventriculomegaly in osteopetrosis has been identified since the 1920s; however, the etiology of hydrocephalus in these cases has also been incompletely understood. Our contention for this phenomenon concludes that both a communicating, somewhat partially obstructive component from venous hypertension, in concert with an obstructive component from the acquired hindbrain craniocerebral disproportion, creates the progressive triventricular hydrocephalus pattern that is seen in these rare cases.

Stated above, the neurosurgical treatment of hydrocephalus in osteopetrosis was first described in 1968 by Baird et al.,6 who reported on a 3-month-old boy with osteopetrosis and hydrocephalus who was treated with a ventriculocaval shunt. The patient was subsequently found dead at home at 10 months. In 1977, Amacher7 reported on twin infants with osteopetrosis presenting with hydrocephalus, sagittal sinus thrombosis, skull base foraminal stenosis, and paraparesis. While the etiology of this spectrum of clinical findings was left unknown, it was suggested that the hydrocephalus may have been the result of an outflow obstruction in the posterior fossa. In 1986, el Khazen et al.14 reported on 2 successive cases of “lethal infantile osteopetrosis,” where the fetuses were diagnosed with skeletal hyperplasia and hydrocephalus in utero.

Cerebrospinal fluid diversion in osteopetrosis has yielded interesting findings. In 2004, Grossman and Feldman20 reported on a 7-year-old boy from Israel who underwent VP shunt placement for hydrocephalus in the setting of progressive visual impairment. Unfortunately, this procedure did not improve vision; however, this case emphasized the magnitude of bone growth that can be seen in these cases and highlighted a possible postoperative shunt complication, as this child required a proximal
shunt revision at 19 months postprocedure due to excessive bone overgrowth at the cranial bur hole site. We also experienced this complication, as described in our Case 3. In 2008, Al-Tamimi et al. reported on 3 Asian children, all of whom had CTH and triventricular hydrocephalus. Two of these patients had papilledema and a decline in vision, and ONSF had been performed; however, both patients ultimately required CSF diversion. ETV was performed in both cases, which reportedly ameliorated the visual deterioration. The third case in this series involved a 13-month-old boy who underwent VP shunt placement for hydrocephalus and worsening vision, as well as a posterior fossa decompression for worsening bulbar function; however, despite all efforts, this child died 3 months later due to sepsis. This report was the first to comment on the success of ETV in osteopetrosis, suggesting its success implies that the fourth ventricular outflow obstruction may be a significant contributing factor for the hydrocephalus in these cases. In 2010, Turgut et al. reported on a 6-year-old Turkish girl who had triventricular hydrocephalus in addition to extensive calcification of the falx cerebri and tentorium cerebelli. The posterior fossa anatomy was not mentioned; however, there was MRI evidence of “brainstem compression.” This child underwent placement of a VP shunt, which was complicated by persistent subdural fluid collections that did not correct with valve revision. This child died shortly thereafter due to a severe pulmonary infection. This case suggested overdrainage as a potential complication of shunt placement in this population and highlighted that dural calcification in this patient population is occasionally identified. In 2011, Dhamija et al. reported on a 9-month-old British girl with hydrocephalus who was reported to be successfully treated with ETV. Although the etiology of the hydrocephalus was uncertain in this case, the authors suggested that venous outflow obstruction and reduced intracranial space for normal CSF flow were contributing factors. Most recently, Mahmoud Adel et al. reported on a 9-month-old Arabian infant with autosomal-recessive osteopetrosis who presented with visual impairments, facial palsy, hypertonia, CTH, and hydrocephalus. This case was complicated by a congenital CNS cytomegalovirus infection earlier in life, which reportedly resulted in “brain atrophy on head CT.” The child was treated with a VP shunt. The clinical outcome in this case was not reported.

While implantable shunt systems have been used in select cases of osteopetrosis, our contention is that due to the higher susceptibility for infection in these cases, the well-known mechanical and infective complications that accompany ventricular shunt systems, and the aggravation of intracranial physiology associated with shunts in children with already impaired bone growth, ETV seems a potentially safer option, with the opportunity to provide a more physiological approach toward CSF diversion in these complicated cases. Accounting for all reported cases mentioned in this paper, including our own experience (13 children with hydrocephalus), 12 children (92%) underwent CSF diversion procedures. The average age of these 13 children was 4.3 years (range 3 months–12 years) with no identifiable sex predilection (8 males and 5 females). In this surgical cohort, 8 children (67%) underwent a VP shunt procedure, while 4 children (33%) underwent ETV. No complications from or revisions for ETV procedures have thus far been reported in these few cases; however, in the shunt-treated cases, 2 children required revision, representing a 25% shunt revision rate. In both of these cases proximal shunt failure was encountered, related to bony overgrowth at the site of the valve and/or proximal portion of the ventricular catheter. After revision, it appears that these children quickly return to their preoperative neurological state. A review of the aforementioned cases of osteopetrosis associated with hydrocephalus is depicted in Table 1.

Correlative Experiences

The spectrum of posterior fossa crowding, variable degrees of CTH, abnormal skull growth affecting both the posterior fossa volume as well as the cranial foramina, intracranial venous hypertension, and triventricular hydrocephalus is not a dissimilar clinical picture to select patient populations we have encountered with achondroplasia and complex craniosynostosis. The association between craniosynostosis and CTH has been well recognized, and most believe that these hindbrain malformations are an acquired defect occurring sometime during the postnatal period as a result of premature fusion of the cranial sutures. In cases of craniosynostosis associated with CTH, the frequent coexistence of hydrocephalus and venous hypertension complicates the overall management, which differentiates this population from those with Chiari malformation Type I in the general population. Numerous cases of complex craniosynostosis have been associated with stenosis of the jugular foramen and various dural venous sinuses. This stenosis results in venous hypertension and is one of the proposed mechanisms for the elevated ICP that is seen in these patients.

In cases of achondroplasia, hydrocephalus is known to occur and is thought of as a communicating phenomenon, resulting from jugular foramen stenosis and impaired CSF absorption due to increased dural sinus pressure. Interestingly, with advances in MRI and reports of venous anatomy in these cases, the severity of hydrocephalus has been found to correlate with the degree of jugular foramen stenosis and collateral venous drainage patterns, with those demonstrating collaterals through the condylar emissary veins having a lower likelihood of experiencing progressive hydrocephalus. Some investigators have suggested that the hydrocephalus in achondroplasia “arrests” or becomes “compensated” following the development of sufficient venous collaterals after early childhood, a condition that is often viewed with some skepticism in pediatric neurosurgery. In this patient population, we have noticed intracranial anatomy that is similar to several of the aforementioned cases of osteopetrosis. Specifically, there are frequent findings of jugular foramen stenosis, the fourth ventricle appears small, the posterior fossa appears “crowded,” and a triventricular pattern of ventricular dilation is encountered. This ventricular pattern alone has been found to relate to the success of ETV in the
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TABLE 1: Cases of osteopetrosis undergoing intervention for hydrocephalus, including the present experience*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>CTH</th>
<th>HCP</th>
<th>CSF Diversion†</th>
<th>Surgical Complications</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baird et al., 1968</td>
<td>3 mos, M</td>
<td>NM</td>
<td>yes</td>
<td>shunt</td>
<td>NA</td>
<td>death at 10 mos</td>
</tr>
<tr>
<td>Amacher, 1977</td>
<td>4 yrs, F</td>
<td>NM</td>
<td>yes</td>
<td>shunt</td>
<td>NA</td>
<td>control of HCP</td>
</tr>
<tr>
<td>Mahmood Adel et al., 2013</td>
<td>9 mos, F</td>
<td>yes</td>
<td>ETV</td>
<td>NM</td>
<td>control of HCP</td>
<td>NM</td>
</tr>
<tr>
<td>current series</td>
<td>8 mos, F</td>
<td>yes</td>
<td>ETV</td>
<td>shunt‡</td>
<td>proximal revision</td>
<td>death at 6 yrs</td>
</tr>
<tr>
<td>Turgut et al., 2010</td>
<td>6 yrs, F</td>
<td>NM</td>
<td>yes</td>
<td>shunt‡</td>
<td>persistent subdural fluid collection</td>
<td>death at 19 mos</td>
</tr>
<tr>
<td>Grossman &amp; Feldman, 2004</td>
<td>7 yrs, M</td>
<td>NM</td>
<td>yes</td>
<td>shunt</td>
<td>proximal revision</td>
<td>control of HCP</td>
</tr>
<tr>
<td>Al-Tamimi et al., 2008</td>
<td>5 yrs, F</td>
<td>yes</td>
<td>yes</td>
<td>ETV</td>
<td>NA</td>
<td>control of HCP</td>
</tr>
<tr>
<td></td>
<td>13 mos, M</td>
<td>yes</td>
<td>yes</td>
<td>shunt‡</td>
<td>NA</td>
<td>death at 17 yrs</td>
</tr>
</tbody>
</table>

* HCP = hydrocephalus; NA = not applicable; NM = not mentioned.
† Mode of CSF diversion (e.g., shunt, ETV).
‡ This patient also underwent SOD.
§ No procedures were performed due to severity of the medical condition.

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Disclosures

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Author contributions to the study and manuscript preparation include the following. Conception and design: Scott. Acquisition of data: Scott, Swift. Analysis and interpretation of data: all authors. Critically revising the article: Scott, Swift. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Scott. Administrative/technical/material support: Weprin. Study supervision: Swift.

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