J Neurosurg Pediatrics 9:305–315, 2012

Surgical treatment for primary spinal aneurysmal bone cysts: experience from Children's Hospital Boston

Clinical article

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Object. Spinal aneurysmal bone cysts (ABCs) constitute a rare and clinically challenging disease, primarily affecting the pediatric population. Information regarding the management of spinal ABCs remains sparse. In this study the authors review their experience with spinal ABCs at Children's Hospital Boston.

Methods. The medical records of all patients treated surgically for primary spinal ABCs between January 1998 and July 2010 were retrospectively reviewed.

Results. Fourteen cases were identified (6 males and 8 females, ages 5–19 years old). The ABCs were located throughout the spine, with an equal number in the thoracic and lumbar spine, and rarely in the cervical spine. The majority of patients presented with back pain, but neurological deficits and spinal deformity were common. A variety of radiographic techniques were used to establish the diagnosis, including needle biopsy. Preoperative selective arterial embolization was performed in 7 cases (50%), and the majority of cases required spinal instrumentation along with resection. Mean follow-up was 55.9 months (range 15–154 months) after initial intervention. Two ABCs recurred (14%), at 9 months and 8 years after incomplete initial resection, and the patients underwent reoperation. Complete resection was ultimately achieved in all cases. All patients were asymptomatic and neurologically intact at their last follow-up evaluation, and showed no evidence of deformity or recurrence on imaging.

Conclusions. Computed tomography and MR imaging are adequate for an initial evaluation of spinal ABCs, although solid variants can present a diagnostic challenge. Given the high rates of recurrence with residual disease, complete obliteration of the lesion should be the goal of treatment. Preoperative embolization is often performed, although in the authors' opinion the degree of bleeding tends not to support its routine use. Long-term follow-up is warranted as recurrences can occur years after initial intervention. However, gross-total excision in conjunction with spinal stabilization, as needed, usually provides cure of the ABC and excellent long-term spinal alignment.
(http://thejns.org/doi/abs/10.3171/2011.12.PEDS11253)

Key Words • **aneurysmal bone cyst** • **spine** • **case series** • **selective arterial embolization**

Aneurysmal bone cysts have recently been redefined by the WHO as "benign cystic lesions of bone composed of blood filled spaces separated by connective tissue septa containing fibroblasts, osteoclast-type giant cells, and reactive woven bone."[16] The WHO noted that ABCs may arise de novo (primary ABCs), or form secondary to deterioration of other benign and malignant bone tumors (secondary ABCs) that have undergone hemorrhagic cystic change.[16] Approximately 70% of ABCs are primary, with the remaining commonly associated with pathologies such as giant cell tumors, hemangiomas, osteoblastomas, chondroblastomas, and telangiectatic osteosarcomas.[37] A solid variant, which differs from the classic ABCs in that cavernous chambers and spaces are absent, accounts for 3.4%–7.5% of all cases.[3,11,29] The pathophysiology of these lesions remains elusive.[10] Although regarded as benign tumors, ABCs can be locally aggressive, causing profound destruction of their encasing bones and pathological fractures, as well as compression of adjacent vital tissues from their rapid expansion.[9,10,16,19,24,37] For this reason, ABCs of the spine, due to their close proximity to the spinal cord and nerve roots, warrant special attention, and present a unique clinical challenge.

Although ABCs have been reported to constitute

Abbreviations used in this paper: ABC = aneurysmal bone cyst; EBL = estimated blood loss; GTR = gross-total resection; SAE = selective arterial embolization.

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1%–6% of solid bone tumors, they still remain a relatively rare clinicopathological entity with an estimated annual incidence of no more than 1.4 per 100,000 people.[23] Of these tumors, 8%–30% are located in the spine, representing approximately 15% of all spinal tumors.[3,11] Reportedly, the lumbar spine is the most commonly affected area (40%–45%) followed by the cervical (30%) and the thoracic (25%–30%) spine, with the posterior elements (laminae, pedicles, and spinous and transverse processes) more commonly involved than the vertebral bodies (anterior elements).[3,11] Spinal ABCs predominantly affect the pediatric population as 75% of patients are younger than 20 years old.[10,23,24,37] Both sexes are almost equally afflicted, with many studies showing a slight female predominance.[10,23,24,37]

Evaluation of ABCs usually includes a combination of plain radiography, CT, and MR imaging, and rarely, open or imaging-guided needle biopsy.[2,3,5,6,13,17,18,22,27,28,30,32] Definitive diagnosis is established by histological analysis, although often the radiographic appearance can be classic.[2,3,5,6,13,17,18,22,27,28,30,32] If CT and MR imaging is typical, biopsy may not be necessary before definitive treatment.[6,18,22,27,28] A lytic lesion surrounded by a thin shell of cortical bone and a ballooning or blown-out bone expansion with erosion and destruction of the cortex are quite characteristic.[22,27,28] However, if there is concern for secondary ABC due to underlying pathology, a biopsy might be necessary, especially if nonsurgical modalities might be used to treat the condition. Computed tomographic imaging can better define the location and extent of bone destruction as well as the integrity of the cortical bone, and is particularly useful for evaluating instability when contemplating instrumentation.[6,18,22,28] Magnetic resonance imaging delineates well the relationship of the ABC to the spinal cord, nerve roots, and other soft-tissue components.[6,22,27,28] Septa, lobulation, and fluid-fluid levels are very suggestive of an ABC on CT and MR imaging,[6,18,22,27,28] although less commonly similar findings can be found in other pathologies such as osteoblastomas, chondroblastomas, telangiectatic angiosarcomas, eosinophilic granulomas, and giant cell tumors.[20,22,28]

Therapeutic approaches described in the literature include a combination of curettage, piecemeal excision, en bloc resection, endovascular embolization, intralesional injection of ablating agents, and radiation.[2,3,5,13,17,30,32] Because of the relative rarity of the disease, there are currently no prospective trials evaluating any treatment paradigms. Most of the available information regarding the management of spinal ABCs comes from case reports and case series.[2,3,5,13,17,26,30,32] Furthermore, the majority of these studies were conducted several decades ago, and therefore, their applicability to modern clinical practice is questionable. In this article we review our 12-year experience with primary spinal ABCs at our institution, comment on existing reports, and propose an optimal management approach based on our conclusions.

Methods

The medical records of all cases treated surgically at the Children's Hospital Boston between January 1998 and July 2010 were retrospectively reviewed. Only cases with confirmed pathology reports of primary ABC were included. Demographic characteristics, pathology reports, operative reports, imaging information, and information regarding hospital course and complications, as well as follow-up, were reviewed. The study was performed under an approved institutional review board protocol.

Results

Demographics

A total of 14 cases met the inclusion criteria of pathologically confirmed primary spinal ABCs (Table 1). These patients included 6 males and 8 females (1:1.3 ratio). Average age was 11.4 years (range 5–19 years), with no difference between males (mean 11.3 years) and females (mean 11.5 years). Two cases (14.3%) involved the cervical spine, 6 (42.9%) the thoracic spine, and 6 (42.9%) the lumbar spine. Four cases (28.6%) involved solely the posterior elements of the vertebrae, whereas 10 cases (71.4%) involved both the anterior and posterior elements. In 6 cases (42.9%) the vertebral body was destroyed by the lesion resulting in vertebral collapse, 4 of which (28.6%) were vertebrae planae. One patient underwent a needle biopsy and another underwent an incomplete resection at an outside hospital before presenting at Children's Hospital Boston. All other treatments were performed at our institution.

Clinical Presentation

Presenting symptoms included a combination of neck or back pain in almost all patients (13 of 14 [92.9%]), neurological deficits in 5 patients (35.7%), and deformity in 3 (21.4%; Table 1). In 1 patient the ABC was an incidental finding. Nine patients (64.3%) presented with chronic symptoms, which had evolved over months, while 5 (35.7%) had an acute presentation due to a pathological fracture. Six cases (42.9%) had significant cord compression at the time of presentation, and 3 (21.4%) had significant nerve root compression.

Diagnosis

Preoperative imaging was performed with a combination of plain radiography, CT, and MR imaging. In most cases initial imaging was characteristic of an ABC and appropriate surgical planning was based on these studies. One case had undergone an open biopsy at another institution, which confirmed the diagnosis of an ABC. In 3 cases (21.4%), preoperative imaging was not entirely characteristic. In 1 of these 3 cases, a CT-guided needle biopsy was performed by interventional radiology and was nondiagnostic. Final pathology confirmed the lesion to be an ABC. In the other 2 cases (14.3%), no presurgical biopsy was performed, but the pathology results confirmed the lesions to be solid variants of ABC. In 1 patient who presented with a pathological fracture, the involvement of the vertebral body preoperatively was unclear on imaging, and reactive marrow changes were the major differential. All cases were confirmed to be primary ABCs with formal histological analysis of the surgical specimens.

TABLE 1: Presentation, treatment, and outcome in the 14 patients undergoing surgical treatment for primary ABCs*

		Presentation		Treatment							
Case No.	Age (yrs), Sex	Signs/Symptoms	Imaging	Preop SAE	Surgical Procedure	Instrumented Fusion	Surgical Approach	Total EBL (ml)	Transfusion	FU (mos)	Recurrence†
1	8, M	pain/neurological deficit	T-4 (P>A) vertebra plana, severe SC compression	no	GTR	yes	pst	1600	yes	40	none
2	18, M	pain/deformity	T-6 (A>P) severe SC compression	no	GTR	yes	pst	1500	no	15	none
3	10, M	incidental finding	L-4 (A>P)	no	curettage	yes	pst	100	no	22	none
4	11, M	pain/neurological deficit	T-2 vertebra plana, 50% subluxation of T-1 on T-3, severe SC compression	yes	GTR (2-stage)	yes	pst	300	no	19	none
							pst	200			
5	5, F	pain	C-4 vertebra plana, 50% subluxation of C-3 on C-4	yes	GTR	yes	pst	150	no	67	none
6	10, F	deformity	T-8 (A=P)	aborted due to shared collaterals w/ SC	GTR	yes	pst	600	no	91	none
7	17, F	pain/deformity	L-1 (P>A) moderate SC compression	yes	GTR	yes	pst	1800	yes	73	none
8	10, F	pain/neurological deficit	L-4 (P>A) extension to L-3 & L-5, L3–5 nerve root compression	yes	GTR	yes	pst	2000	yes	25	none
9	15, F	pain/neurological deficit	C-6 vertebra plana, C5–6 nerve root compression	yes	GTR	yes	ant	300	no	154	recurrence 8 yrs after 1st op
					GTR	yes	combined ant/pst (for recurrence)	3300	yes		no recurrence 5 yrs after 2nd op
10	17, M	pain	T-11 (A=P) significant paraspinal extension, severe SC compression	yes	GTR	yes	pst	500	no	40	none
11	4, M	pain/neurological deficit	L-3 (P>A) retroperitoneal extension, cauda equina compression	yes	curettage	no	pst	50	no	37	none
12	9, F	pain	T-6 (P>A) significant SC compression, imaging not typical of an ABC	no	GTR	yes	pst	200	no	30	none
13	19, F	pain	L-5 (P>A) significant cord compression	no	GTR	yes	pst	800	yes	42	none
14	7, F	pain	L-2 (P>A) imaging not typical of an ABC	no	curettage	no	pst	100	no	127	recurrence 9 mos after 1st op
							pst (for recurrence)	100	no		no recurrence 118 mos after 2nd op

* A = anterior vertebral element involvement; ant = anterior; FU = follow-up; P = posterior vertebral element involvement; pst = posterior; SC = spinal cord.

† All outcomes intact.

Treatment

Preoperative SAE was attempted in 8 cases and was successfully performed in 7 (50%). In 1 case embolization could not be completed because the ABC and spinal cord had a common arterial supply. The indications for embolization included preoperative imaging suggestive of an ABC, and anticipation of significant bleeding. However, the decision to take a patient to the angiography suite was very much dependent on the preferences of each individual surgeon.

Gross-total excision was the goal of surgery in most cases. This was accomplished most often via a piecemeal excision of the ABC, as these lesions often are not very cohesive. The general strategy of surgery is to extensively dissect out the margins, with minimal entry into the lesion initially to reduce bleeding. Once the margins are defined and the neural elements are protected, the lesion can be removed relatively rapidly using a combination of rongeurs, curettes, and a high-speed drill. This strategy often allows for the surgeries to be performed without major blood loss. Eleven cases (78.6%) were treated in this manner, while 3 (21.4%) underwent simple intralesional curettage. Existing deformity or anticipated instability due to destruction of at least 2 columns of the spinal axis was treated by stabilization and fusion. Overall, 12 cases (85.7%) underwent an instrumented fusion. The fusion substrate included a combination of bone bank allografts (corticocancellous strut or bone chips) used in 8 patients and bone autografts (iliac crest in 5 patients, rib in 2 patients, and fibula in 1 patient). Iliac crest bone marrow aspirate was used in 1 patient, bone morphogenetic protein in another, and osteoinductive demineralized bone matrix gel (Grafton) in another patient. A combined anterior and posterior approach was used in 1 case involving the cervical spine, but a single posterior approach provided adequate exposure to both anterior and posterior vertebral elements in all the remaining cases. Because spinal ABCs usually arise within the posterior vertebral elements and expand the bone, posterior approaches generally allowed for wide access to the anterior disease. When the direct posterior approach was inadequate in the thoracic spine, the exposure was combined with a costotransversectomy. Transpedicular approaches were common. Intraoperative sensory and motor monitoring was used in all cases with significant spinal cord compression. Postoperative orthosis was used in 7 patients (50%), and included a halo in 3 cases and a brace in 4.

Follow-Up

Although practices varied among different surgeons, follow-up at the clinic was usually requested at 1 month, then every 6 months for the first 2 years, and subsequently at 2–3-year intervals. All patients underwent a postoperative CT scan within a few weeks after surgery to evaluate the extent of resection. Clinical follow-up was generally supplemented with plain radiography, while a CT scan or MR imaging were ordered in cases of suspected recurrences. Individual surgeons chose to follow-up with CT scans even without suspicion of recurrence.

Outcomes

Complete resection in a single procedure was achieved in most cases (10 of 14 patients). In 2 cases preoperative imaging was not diagnostic and intraoperative frozen section pathology could not exclude malignancy, so the initial approach was limited to a biopsy only. In both cases formal pathology identified the lesion as a solid variant ABC. Complete resection was subsequently achieved for both lesions, one immediately after the pathology results became available, and the other 9 months later when the lesion enlarged. Complete initial resection was not possible in 2 cases: one in which the extent of involvement of the anterior elements was initially unclear, and a second case in which significant intraoperative venous bleeding precluded complete resection. Of note, this occurred despite preoperative SAE. In the first case complete resection was achieved in a second surgery 12 days later, whereas in the latter case the ABC was found to be enlarging 8 years after the initial resection and was completely excised.

Five cases in our series (35.7%) underwent transfusion with packed red blood cells (Tables 1 and 2). Preoperative embolization did not appear to have a significant effect on blood loss (Table 2). The complications in this study were mainly associated with suboptimal positioning of hardware. A total of 4 cases were returned to the operating room for revision of instrumentation. Three of these children had lesions in the thoracic spine, and 1 in the lumbar spine. Furthermore, all 4 children were less than 11 years of age. Notably, these children were taken back for revision of their hardware, not because of hardware failure but because their instrumentation was believed to be suboptimal in a way that potential migration of the misplaced screws during the child's growth could pose a risk of injury to adjacent nerves or vessels. All revisions were performed within days of the first procedure. The first patient (Case 1) was an 8-year-old who experienced a medial broach of a screw at the pedicle of T-4 (T2–6 posterior fusion). The second patient (Case 4) was an 11-year-old who had a T-1 anteriorly protruding screw in a C5–T5 instrumented fusion. The third patient (Case 6) was a 10-year-old with 2 left screws placed somewhat more laterally than intended (T5–L1 fusion). And the last patient (Case 8) involved a 10-year-old with a misplaced screw in an L3–5 fusion. All patients recovered from the procedure uneventfully. Furthermore, all of the patients had intact instrumentation and good fusion at their last follow-up. More importantly they had no limitations in their daily activities and went on to pursue competitive sports such as tae kwon do and cheerleading.

Overall, there were 2 recurrences (14.3% of all cases), both of which were incompletely resected during the first surgery. The first case involved a 15-year-old girl (Case 9) with complete destruction of the C-6 vertebral body and significant paraspinal extension of the ABC. A GTR was performed along with instrumented fusion and grafting. Significant intraoperative bleeding precluded complete resection of a small part of the lateral capsule of the ABC's extraspinal component, although no residual was appreciated on postoperative imaging. For several years there was no evidence of recurrence clinically

TABLE 2: Average EBL per treatment group*

Treatment Group	GTR & Instrumented Fusion			Curettage w/o Instrumented Fusion		Curettage w/ Instrumented Fusion		Overall Average		
	EBL (range) in ml	Patients Transfused†	Primary Surgical Interventions	EBL (range) in ml	Primary Surgical Interventions	EBL (ml)	Primary Surgical Interventions	EBL (range) in ml	Patients Transfused†	Primary Surgical Interventions
SAE	858 (150–2000)	2	6	50	1	NA	NA	743 (50–2000)	2	7
no SAE	940 (200–1600)	2	5	100	1	100	1	700 (100–1600)	2	7
total	mean 895 (150–2000)	4	11	mean 75 (50–100)	2	100	1	mean 722 (50–2000)	4	14

* NA = not applicable.

† Interventions for recurrences/residuals are not included.

or on follow-up imaging. The patient presented again 8 years after the initial resection with neck pain, and MR imaging obtained at that time revealed a recurrence of the ABC. The second case involved a 7-year-old girl (Case 14) with an ABC involving the posterior elements of L-2. Intraoperative frozen pathology indicated a solid tumor, likely benign, but without excluding malignancy. Simple curettage was performed at the time, and the defect was packed with allograft bone chips without instrumentation. The patient was followed with serial CT scans that, 9 months after the initial procedure, showed a small area of lucency on the lamina of T-2 consistent with recurrence. Both patients underwent further operations and the lesions were completely excised (Fig. 1). There were no recurrences after complete resections. Mean follow-up was 55.9 months (range 15–154 months) after the first surgery. All patients were asymptomatic and neurologically intact at their last follow-up, and had no evidence of deformity or recurrence on imaging. Furthermore, all patients with instrumentation had adequate new bone formation and showed excellent fusion at their last follow-up evaluation.

Illustrative Cases

Case 1

This 8-year-old boy presented with a several-months history of progressive midthoracic pain that became debilitating just prior to admission. The patient also had paresthesias over the left anterior thigh, and constipation and voiding difficulties, but no motor symptoms. Computed tomography and MR imaging revealed a T-4 vertebra plana (Fig. 2A–D) and mild kyphosis associated with a destructive lesion involving the T-4 left pedicle, spinous process, and vertebral body with a left anterior paraspinal component. The lesion resulted in severe cord compression and the imaging findings were consistent with an ABC, so the patient was taken directly to the operating room. Because of the extensive nature of the tumor, a gross-total removal of the T-4 vertebra—including the spinous process, lamina, pedicles, and body from a T1–6 posterior exposure—was performed from a bilateral posterior approach. A T2–6 instrumented posterior fusion was performed (Fig. 2E and F) along with placement of an anterior titanium cage. The patient's preoperative pain and neurological symptoms completely resolved. A postoperative CT scan revealed a medial broach of the pedicle at T-2 on the left side and the patient was taken back to the operating room for repositioning of the screw. At the 40-month follow-up evaluation, the patient was completely asymptomatic, and imaging showed no signs of recurrence with excellent fusion (Fig. 2G and H).

Case 3

The ABC in this case was an incidental finding in a 10-year-old boy found during the workup for appendicitis. At the time, CT and MR imaging revealed a well-circumscribed lytic lesion within the left aspect of the L-4 vertebral body, extending into the left pedicle (Fig. 3A–C). No compromise of the spinal canal was appreciated, so the patient was initially followed with serial CT

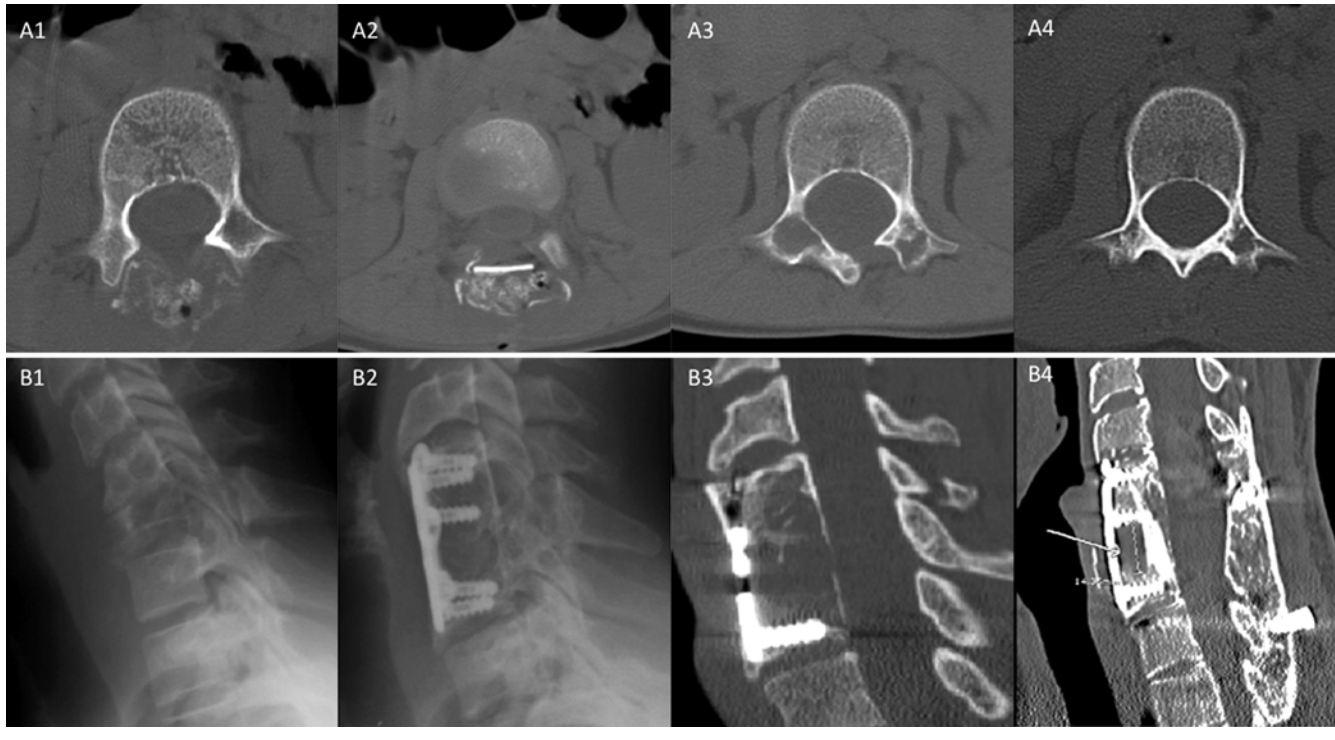

Fig. 1. Cases 9 **(A)** and 14 **(B)**. Imaging obtained in the 2 patients with lesion recurrence. **A1 and A2:** Axial CT scans obtained 2 days after curettage and bone grafting of an L-2 lesion in an 8-year-old girl involving the spinous process and lamina, extending to the inferior articulating processes. **A3:** A recurring lesion is shown again involving the lamina as well as the right pedicle of L-2 9 months after the surgery on an axial CT scan. **A4:** Follow-up axial CT scan obtained approximately 10 years after repeat surgery with recurretage and bone grafting. **B1:** Lateral radiograph showing an ABC involving the C-6 vertebra in a 15-year-old girl. The lesion had significant paraspinal extension. **B2:** The lesion was resected and an anterior C5–7 interbody fusion was performed. A small part of the capsule was left behind secondary to significant intraoperative bleeding, as shown on the lateral radiograph. **B3:** A recurrence at the C-5 and C-6 levels 8 years after surgery is shown on a sagittal CT reconstruction. This recurrence was reexcised through a combined anterior and posterior approach. The existing instrumentation was partially exchanged and supplemented with a posterior C4–T2 fusion. **B4:** Good fusion and no signs of recurrence are demonstrated 5 years after the second surgery on a sagittal CT scan reconstruction.

scans. Due to expansion of the mass, surgery was recommended. The patient underwent cyst curettage, as well as L3–4 instrumentation and arthrodesis (Fig. 3D–F). The patient recovered uneventfully. At the 22-month follow-up examination the patient was completely asymptomatic, the positioning of the instrumentation was intact, and there was no sign of recurrence or deformity on imaging (Fig. 3G and H).

Case 8

This case refers to a 10-year-old girl who presented with back pain and was found to have left L-4 distribution weakness. Imaging revealed a large expansile lesion centered on the left L-4 pedicle and transverse process, with extension in the vertebral body, as well as superiorly and inferiorly to L-3 and L-5. The L-3, L-4, and L-5 nerve roots could not be identified on imaging but there was extensive destruction of the L3–4 and L4–5 facet joints (Fig. 4A–F). She experienced rapid symptom progression over several days and was taken for emergency treatment with preoperative embolization followed by surgery. The patient underwent complete excision of the ABC with a left L-4 and partial L-3 vertebrectomy. Spine stabilization was achieved with L3–4 and L4–5 posterior lumbar interbody fusions and an L3–5 pedicle screw fixation. The patient recovered uneventfully with resolution of her radicular symptoms. Postoperative imaging, however, revealed a misplaced pedicle screw, and the patient was taken back to the operating room 3 days after her original surgery for repositioning of the screw. The patient recovered from this second surgery uneventfully. At the 25-month follow-up she was completely asymptomatic, had full strength, and had resumed all her favorite activities, which included tae kwon do and cheerleading. There was no evidence of recurrence of deformity, and an excellent fusion was confirmed on imaging (Fig. 4G and H).

Discussion

Demographics and Presentation

Aneurysmal bone cysts are recognized as benign lesions, which nevertheless can behave aggressively, causing significant attenuation of the affected bone structures and compression of adjacent soft tissue. For this reason, ABCs of the spine warrant special attention. Due to their proximity to the spinal cord and nerve roots, rapid expansion and/or pathological fractures can result in compressive phenomena and neurological symptoms, which can range from mild radiculopathies to overt paraparesis. Further-

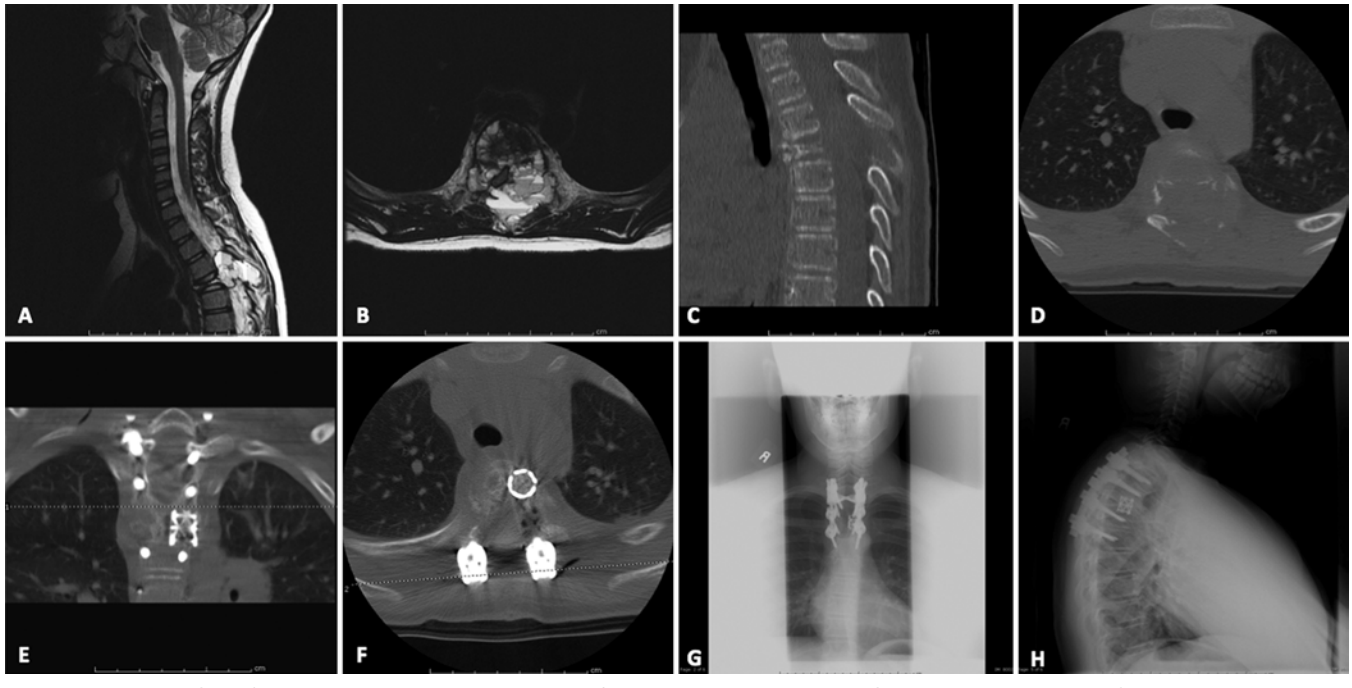

Fig. 2. Case 1. Images obtained in a patient with T-4 vertebra plana involvement. Sagittal **(A)** and axial **(B)** T2-weighted MR images demonstrating extensive involvement of the fourth thoracic vertebra. Sagittal **(C)** and axial **(D)** CT reconstructions showing profound attenuation of the bone and complete collapse of T-4. A coronal CT reconstruction **(E)** and axial CT scan **(F)** were obtained after a posterior T2–6 instrumented fusion using cancellous bone graft and a Danek titanium cage. Anteroposterior **(G)** and lateral **(H)** plain radiographs of the cervicothoracic spine obtained 5 years after resection showing intact instrumentation.

more, because these lesions commonly affect the growing pediatric spine, deformity and instability are common and need to be addressed appropriately.[2–5,13,17,25,30,32]

In our patient series, the demographic characteristics of our population were consistent with other existing reports,[2,3,5,13,17,30,32] showing slight female predominance

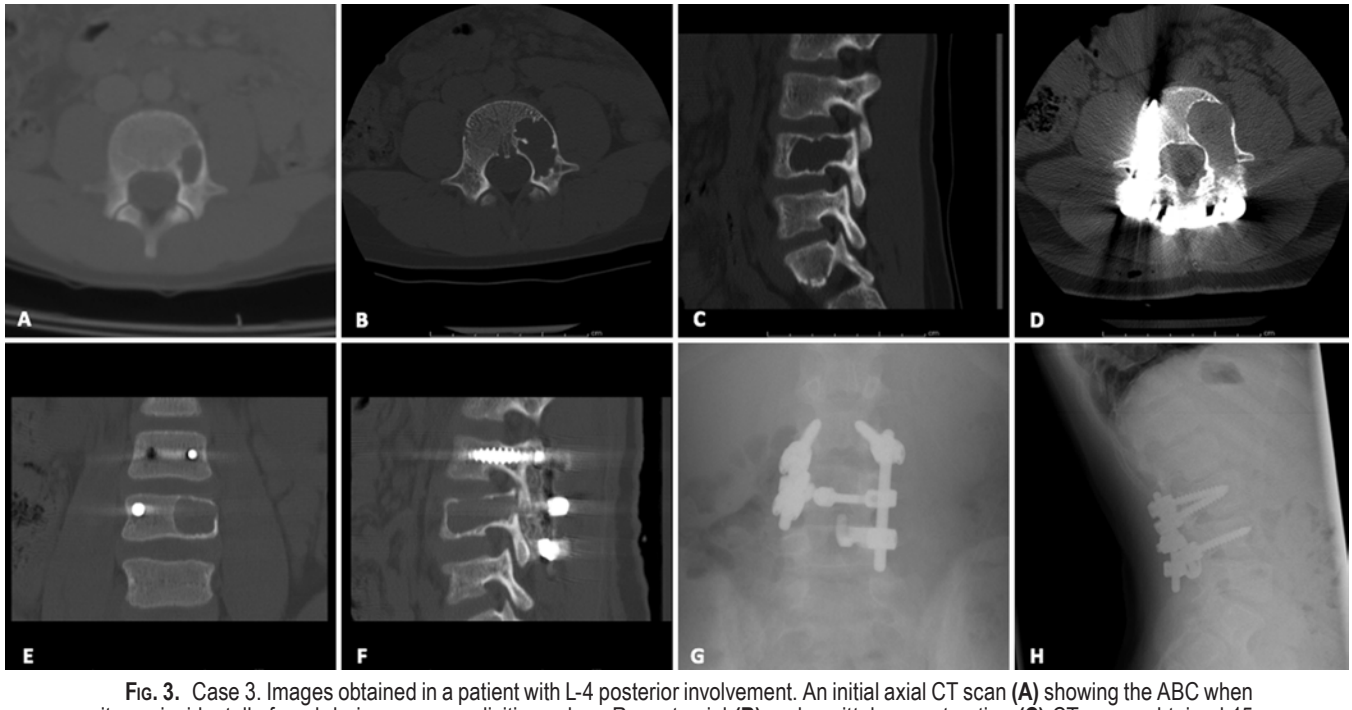

Fig. 3. Case 3. Images obtained in a patient with L-4 posterior involvement. An initial axial CT scan **(A)** showing the ABC when it was incidentally found during an appendicitis workup. Repeat axial **(B)** and sagittal reconstruction **(C)** CT scans obtained 15 months after the initial scan original demonstrating progression of the disease. Postoperative axial **(D)**, coronal reconstruction **(E)**, and sagittal reconstruction **(F)** CT scans were obtained after curettage and L3–4 instrumented fusion. Anteroposterior **(G)** and lateral **(H)** plain radiographs of the lumbar spine obtained 2 years after surgery showing intact instrumentation.

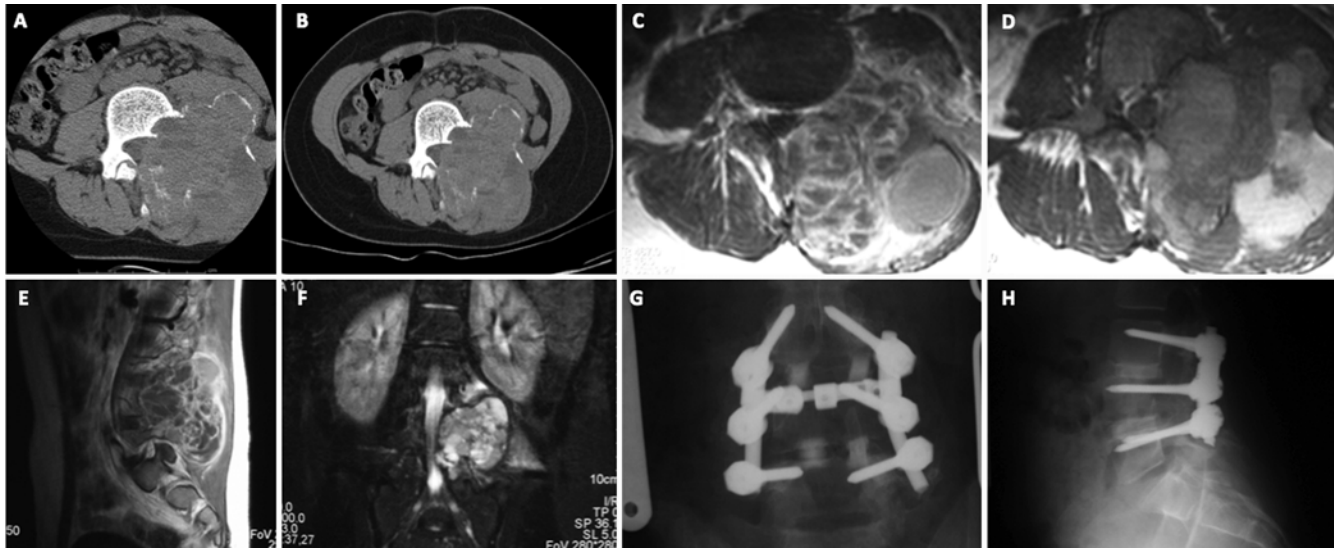

Fig. 4. Case 8. Images obtained in a patient with L-4 anterior involvement. Axial CT scans **(A and B)** showing extensive involvement of the left posterior elements of L-4 with significant paraspinal extension and distortion of the anatomy. Axial **(C and D)**, sagittal **(E)** and coronal **(F)** MR images delineating the extensive soft-tissue involvement. Anteroposterior **(G)** and lateral **(H)** plain radiographs of the lumbar spine were obtained 9 years after surgery, which involved an L-4 and partial L-3 vertebrectomy, an L3–5 fusion with L3–4 and L4–5 posterior lumbar interbody fusion, iliac crest bone grafting, and pedicle screw fixation using the Synthes system.

(male to female ratio 1:1.33), and the ABCs usually arising within the posterior elements of the vertebrae. The thoracic and lumbar spine, however, was more commonly affected than the cervical spine, contrary to previous reports.[2,3,5,13,17,30,32] The mean age was 11 years, but the fact that our institution exclusively specializes in the treatment of pediatric diseases must be taken into consideration. Although complete vertebral collapse and vertebrae planae have rarely been reported in the literature,[8,31] 28.5% of our patients presented in this manner.

Spinal ABCs not associated with a pathological fracture usually present with a several-week or -months history of progressive back pain, reduced range of motion, or a palpable mass on the spine. Pathological fractures causing spinal cord or nerve root compression are reported to occur in as many as 20% of patients.[2,3,5,13,17,26,30,32] Kyphosis and scoliosis caused by pathological fractures or pain and muscle spasm are also quite common, and occur in approximately 10%–15% of patients.[2,3,5,13,17,26,30,32] In our series, almost all patients had pain (92.9%), and a large fraction had neurological deficits and deformity (35.7% and 28.6%, respectively). Paresis is only rarely reported in the literature.[6,14,38] In our series 1 patient presented with paraparesis. In this patient, as well as in 4 other patients, a pathological fracture caused an acute exacerbation of their symptoms leading to significant neurological deficits (35.7% of all patients). Fortunately, all of our patients regained normal neurological function after prompt intervention. Others, however, have reported incomplete recovery and permanent deficits in such patients.[2,3,32] These examples emphasize the fact that, although considered benign lesions, spinal ABCs can behave quite aggressively with possible devastating outcomes. It is therefore prudent that intervention be provided even in minimally symptomatic patients.

Diagnosis

Preoperative CT and MR imaging were very suggestive of the diagnosis of ABC in most cases. A CT-guided needle biopsy was used in 1 case in which preoperative imaging was unclear, but did not yield useful results. Two cases in our series (14.3%) proved to be solid variants of ABCs, an incidence slightly higher than in other series.[3,11,29] Interestingly, intraoperative frozen pathology from both of these cases could not exclude the possibility of a malignant neoplasm. In light of the fact that nearly 30% of spinal ABCs are secondary to another pathology including malignant neoplasms,[37] a more conservative approach that preserves mechanical stability is appropriate at the initial intervention in these patients, as the ultimate treatment might be nonsurgical. Other groups have also reported that the solid variant ABC presents a diagnostic challenge.[3,11,29]

Management

Surgery. Incomplete excision of spinal ABCs is associated with a high incidence of recurrence.[2,3,5,13,17,26,30,32] Therefore, the general aim of treatment is complete obliteration of the lesion. A number of available options exist and are discussed below.

Although simple curettage is successfully used in the management of ABCs of the long bones,[35] it has been associated with high rates of recurrence in ABCs of the vertebral column.[2,3,5,13,17,26,30,32] One possible explanation is that the use of adjunctive strategies commonly used in long bones, such as cryotherapy or methylmethacrylate, which can expand the margins of ablation beyond the surgical margins,[10,35] are avoided in spinal ABCs because of the close proximity to the spinal cord.[4,25] Furthermore, the higher complexity of the vertebral anatomy may in-

crease the possibility of incomplete resection. Simple intralesional curettage was used in 3 of our cases with good results when complete obliteration was achieved. This technique resulted in less intraoperative blood loss (Table 2), and negated the need for an instrumented fusion in 2 of 3 cases (Table 1). We believe that intralesional curettage is a viable option for lesions where there is confidence that complete obliteration can be achieved. These include relatively smaller lesions, with fairly regular margins, without significant distortion of the anatomy or paraspinal extensions. Furthermore, this is also a valid option when the preoperative diagnosis is unclear.

En bloc resection virtually eliminates the possibility of recurrence, although it is associated with higher morbidity,[2,3,5,13,17,26,30,32] and is far more difficult to achieve in the spine. In our patient series, as a compromise between intralesional curettage and en bloc resection, most of the cases were treated with a wide piecemeal excision. Gross-total resection was overall associated with more intraoperative blood loss, as well as an increased transfusion rate (Table 2), and made the need for instrumentation more likely (Table 1). However, after complete excisions, we did not see any recurrences.

Combined anterior and posterior exposures have been described for complete resection of ABCs significantly affecting both the anterior and posterior elements of the vertebrae.[3,5,13,32] However, in our experience, a posterior approach provided an adequate exposure in all but 1 of our cases due to the large corridor access provided by the large expansile lesion.

Role of SAE. Selective arterial embolization of spinal ABCs has been proposed as both a preoperative adjunct to surgery[2,5,13,30,32] as well as definitive treatment.[3,34] In our practice SAE was used in selected cases preoperatively to decrease intraoperative bleeding. A number of groups expressed concerns and were generally conservative in attempting SAE.[2,3,30,32] Shared collaterals between the spinal cord and the ABC are concerns, particularly in the thoracolumbar spine with involvement of the artery of Adamkiewicz or in the cervical spine where the vertebral arteries may also be involved.[2,3,30,32] Furthermore, preoperative SAE was generally not recommended by other groups in the presence of pathological fractures with spinal cord compression.[2,30,32] In our series, successful and safe preoperative SAE was performed in 50% of our patients, 60% of whom had significant cord compression, and included cases from both the cervical and thoracolumbar spine. In 1 patient SAE was aborted because of shared collaterals with the spinal cord. We believe that modern digital subtraction angiography can better delineate collaterals than the technology used in most of the published series,[2,3,5,13,17,26,30,32] and thus inadvertent spinal infarction is much less likely.

However, with the advancements in modern surgical techniques, SAE may be less necessary than it had previously been. Given the fact that this was an uncontrolled retrospective study, we did not observe a substantial reduction in blood loss in our group after embolization (Table 2). Other groups have argued that significant intraoperative bleeding often hinders complete excision of the ABC, and may lead to increased rates of recurrence.[2–5,10,13,17,24–26,30,32] However, significant bleeding can occur with very vascular lesions even after SAE.[2,13,17,30] The 1 patient in our series who had an incomplete resection and recurrence because of significant bleeding during surgery had undergone preoperative SAE. Furthermore, the transfusion rates were not much different in the SAE group versus the group not treated with SAE (25% vs 33% of all procedures, respectively (Table 2). The only case with very high blood loss was a reoperation with prior fusion, who had an EBL of 3300 ml at the second surgery (Case 9, Table 1). This patient had undergone embolization prior to the first surgery, but not prior to reoperation. Although the strength of our data are somewhat limited secondary to the study's retrospective nature, it must be noted that the decision to perform SAE was very dependent on the preferences of each individual surgeon and not entirely on the objective propensity of each lesion to bleed. Furthermore, not all the patients underwent initial angiography and therefore the degree of vascularity was not known; that is, the lesions that were not embolized were not chosen because they did not have significant vascularity. In our review of the literature, we were unable to find data proving that SAE in ABCs of the spine either leads to decreased intraoperative blood loss or that it leads to overall better outcomes. The available data are even sparser in the pediatric population (< 20 years old). A number of potential factors may change the role of SAE for ABCs in the pediatric spine. First, the overall anatomy and vessels are smaller, with less well-defined feeders amenable to embolization. In our experience, many of the patients who initially underwent embolization did not have a dramatic drop in the vascularity of the lesion, as often there were only a few well-defined feeders that could be embolized, or even when these existed, shared collaterals with the spinal cord limited the number of vessels that could be safely sacrificed. Even with contemporary neuroendovascular techniques, these factors may continue to limit the usefulness of SAE. Furthermore, children are less likely to be receiving any anticoagulants or have any other impairment in coagulability that would contribute to increased intraoperative blood loss. In addition, besides the obvious risks associated with SAE, other factors may have to be considered when contemplating embolization, such as whether taking a rich collateral blood supply hinders the ability of a compressed and already ischemic spinal cord to regain function, even though initial injection of sodium amytal did not show changes as it was already ischemic; whether local ischemia limits/delays the ability to heal and form new bone; and can this lead to asymmetrical growth? These are legitimate questions to which we have no answers.

With the limitations of this retrospective review, and the data from the available literature, we conclude that SAE may be beneficial in selected cases in which it can be safely performed, and where significant intraoperative bleeding is anticipated because of the vascularity, extent, and location of the lesion. However, we believe that, at least in the pediatric population, it should not be considered a requirement or the standard of care, because the effects of embolization may be mitigated by the advances in

modern surgical techniques. Furthermore, although complications associated with SAE are rare, given the benign nature and excellent long-term prognosis associated with these lesions, a catastrophic inadvertent embolization of the spinal cord would be tragic.

Nonsurgical Options. Other options for treatment of spinal ABCs are available. Radiosurgery or radiation therapy has been considered by some investigative groups, but it is associated with a published incidence of malignant transformation.[2,3,13,17,26,32] Some interesting options are available for large sacral lesions, including both extensive embolization and direct sclerosis. While these may be reasonable for nonmotion segments, they do not afford any stability to the spine in the short term, with unclear ability to form bone over time.[1,7,36] Groups have reported using SAE in multiple sessions as a successful definitive alternative for spinal ABCs.[3,34] Given the unpredictable course of these lesions and the variable amount of time after embolization until involution of the lesion is achieved,[12,15,21] this might not be appropriate in cases of spinal cord compression or instability. However, SAE does carry risks.[33] We believe that SAE may be a viable option as monotherapy in large ABCs of the sacrum in which instability is not so much of a concern and when surgical approaches are associated with higher morbidity.[34]

Prognosis

The main complications in this study were related to initial suboptimal positioning of hardware requiring revision. The thoracic location of the lesions in 3 of 4 patients, as well as their young age (all younger than 11 years old) added to the technical difficulty of instrumentation. Of note, all 4 children were taken back for revisions of their hardware, not because of hardware failure, but because their instrumentation was believed to be suboptimal in a way that potential migration of the misplaced screws during the child's growth could pose a risk of injury to adjacent nerves or vessels. All revisions were performed within days of the first procedure. Given the young age and the absence of significant comorbidities, the aforementioned risk was believed to outweigh the risk of another procedure to revise the instrumentation. All patients recovered uneventfully. In addition, all of the patients experienced an excellent long-term outcome, with intact instrumentation and good fusion at their last follow-up. More importantly they had no limitations in their daily activities and they went on to pursue competitive sports such as tae kwon do and cheerleading. Based on our experience, we believe that the long life expectancy after the procedure in children, the plasticity of the growing pediatric spine, and the physical activities children engage in after the surgery mandate striving for the perfect construct, even if this translates into revision of hardware. A durable and safe construct is generally associated with an excellent long-term outcome.

Recurrences of spinal ABCs have been reported to occur within the first 1 or 2 years after the initial intervention. For this reason, most groups advocated a 3–5-year follow-up period.[2,3,5,13,17,26,30,32] In our series, 1 of the 2 recurrences occurred 8 years after the initial treatment, something that to our knowledge has not been reported to date. Therefore, a longer follow-up of patients may be prudent. None of our patients experienced any recurrences after complete excision was achieved, but all of them led completely normal lives with no limitations in their activities at their last follow-up. Of note, the solid variant ABCs, despite the initial diagnostic challenge they presented, shared the same excellent prognosis after complete resection with the other cases in our series.

With the limitation of this retrospective review and the data from the available literature we make the following recommendations regarding follow-up. First, all patients should undergo at least a postoperative CT scan or MR imaging to assess the extent of resection within a few weeks or months after the procedure. Follow-up imaging will then depend on the extent of resection. Second, for complete resections with instrumentation and fusion, obtaining plain radiographs yearly for the next 4 years followed by 3-year interval plain radiographs for an additional 6 years would be a reasonable approach. More sophisticated imaging such as CT or MR imaging should be sought if there are clinical signs of recurrence. And third, for incomplete resections, follow-up with CT or MR imaging is likely indicated. The time intervals between each of these studies should be individualized according to the extent of resection and the condition of instrumentation, and should be tailored to the social situation of each patient.

Conclusions

Primary ABCs of the spine constitute a unique pathology due to their proximity to the spinal cord and nerve roots, and their frequent association with deformity. Several radiographic presentations of cysts are possible, including the typical lesion consisting of multiple fluid-filled cystic spaces, a solid variant, and a vertebra plana. Computed tomography and MR imaging are usually adequate for the initial evaluation of spinal ABCs. Given the high incidence of recurrence with residual disease, complete obliteration of the lesion should be the goal of treatment. Although recurrences usually occur during the first few years after treatment, they can present as late as 8 years after incomplete resection, thus a long-term follow-up evaluation is warranted. Complete excision can destabilize the spine, and instrumentation may be necessary in the majority of cases after complete excision. Preoperative SAE should not be considered a requirement for surgery, but might be helpful in selected patients. After complete resection and stabilization spinal ABCs are associated with an excellent long-term prognosis.

Disclosure

Dr. Zenonos was financially supported by an Alexander S. Onassis Foundation Scholarship.

Author contributions to the study and manuscript preparation include the following. Conception and design: Zenonos. Acquisition of data: Zenonos, Jamil, Jernigan. Analysis and interpretation of data: Zenonos. Drafting the article: Zenonos. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Proctor. Study supervision: Proctor.

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Manuscript submitted June 20, 2011.
Accepted December 2, 2011.
Parts of this paper were presented as an oral presentation at the 39th Annual Meeting of the AANS/CNS Section on Pediatric Neurological Surgery, November 30 to December 3, 2010, in Cleveland, Ohio.
Please include this information when citing this paper: DOI: 10.3171/2011.12.PEDS11253.

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