Correction of Progressive Severe Cervical Kyphosis in a 21-Month-Old Patient With NF1: Surgical Technique and Review of Literature

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BACKGROUND: Severe cervical kyphosis in the setting of neurofibromatosis type 1 (NF1) is a rare manifestation of the disease in the pediatric population. Dystrophic and immature bone complicate the placement of hardware necessary for surgical correction of alignment and a review of the literature yields 4 cases of pediatric patients with NF1 requiring surgical intervention in which the youngest patient was 10 yr old.

OBJECTIVE: To report the case of an 11-mo-old female with NF1 who presented with a plexiform cervical neurofibroma and focal cervical kyphosis. A comprehensive review of the literature and a detailed description of nonsurgical and surgical management for this patient population is described.

METHODS: A literature review was completed for article reviewing management of pediatric patients with cervical spine injuries and NF1. The patient’s chart was reviewed and the patient was followed for a year to provide adequate follow-up. Institutional Review Board (IRB)/ethics committee approval and patient consent were neither required nor sought for this study.

RESULTS: The literature was reviewed, summarized, and utilized for operative planning and postoperative management. Postoperative imaging and 1-yr follow-up imaging showed anterior construct and lateral mass fusion, restoration of cervical alignment, and no neurological deficits.

CONCLUSION: This is the youngest reported patient to have surgical cervical kyphosis correction in the setting of NF1. A review of the literature helped develop a long-term plan and shape a novel same-day front-back-front approach to restore alignment that will be of use to teams managing these complex patients in the future.

KEY WORDS: Dystrophic cervical kyphosis, Neurofibromatosis type 1, Spinal deformity correction

Neurofibromatosis type 1 (NF1) is an autosomal dominant disease that manifests in a variety of ways including skeletal abnormalities of the spine. Spinal involvement may be classified as dystrophic or nondystrophic changes, the former of which results in spine deformities requiring surgery. Dystrophic changes found in NF1 include defective pedicles and scalloping of the vertebral bodies that result in a dystrophic curve pattern that may occur in the cervical spine and lead to severe cervical dystrophic kyphosis with rapidly progressive paralysis. Surgical management of these patients is critical in preventing disease progression and improving symptoms; however, there are a number of challenges in the treatment of these patients related to the bone quality, anatomy, and degree of kyphosis.

Studies have found that in patients with NF1 70% of children younger than 10 yr of age and 80% in children and adolescents aged 10 to 18 yr old are symptomatic from the disease. There are patients in the infant age group who may develop dystrophic cervical kyphosis and resultant cervical myelopathy for whom surgical management has not been described in the literature. Most published reports of cervical fusion in the pediatric population are in patients older
In the context of NF1, the 2 largest case reports of surgical correction of severe cervical kyphosis total 11 cases with patients’ ages ranging from 10 to 38.5 yr old at the time of surgery.

We present a case of an 11-mo old female with NF1 who presented with a plexiform neurofibroma and focal cervical kyphosis. We carefully reviewed the literature regarding the management of patients with severe cervical kyphosis in the setting of neurofibromatosis and the management of very young (<6 yr) children with cervical instability. Given the underlying pathology and the young age of the patient, the nonoperative management considerations and the technical challenges of surgical management are reviewed in this case report.

METHODS

Presentation

At 4-mo of age, the patient was brought to clinic for evaluation of a plexiform cervical neurofibroma at C2-3-4 involving the paraspinal muscles and posterior cervical elements identified on magnetic resonance imaging (MRI; Figure 1A). The patient was asymptomatic at the

![FIGURE 1](image-url). Progressive dysplastic cervical kyphosis monitored over 1 yr. Sagittal T2-weighted MR images at 4 mo A, 10 mo B, 13 mo C, and 14 mo of age D when the patient developed neurologic deficits and cervical cord signal change.
time and returned in 6 mo with a follow-up MRI showing evidence of focal cervical kyphosis of C3-4 measuring 38° on a supine MRI (Figure 1B). The patient was followed with serial plain film radiographs and placed in a cervical hard collar. Review of the plain films showed hypoplastic vertebral bodies at C3 and C4 with anterior wedging and posterior displacement of the vertebrae with spinal canal stenosis. The progressive kyphosis appeared to be caused by the plexiform neurofibroma expanding the neural foramen and eroding the bony anatomy. Therefore, the patient underwent an anterior neck dissection and resection of the neurofibroma (Figure 1C).

At 14-mo of age, the patient was brought to the emergency room with complaints of right-sided upper and lower extremity weakness and decreased weight bearing activity. A repeat supine MRI was obtained showing kyphosis to 50° (Figure 1D), narrowing of the spinal canal to 5 mm and cord compression with signal change. The degree of kyphosis would likely have been greater had it been measured with upright plain films of the cervical spine. The patient was subsequently placed in a halo with 5 pounds of traction and 5 extra pounds were added per day for 3 d with traction maintained over 5 until the patient was placed in a halo vest (Figure 2). The patient was left in the halo in order to allow time for bone maturation for rigid spinal fixation. Over the 7-mo period, the patient remained an active toddler and would fall with vest and undergo multiple adjustments by our orthotics team for pin loosening and rod motion. The implementation of a multidisciplinary team was crucial to safe and practical patient care. At 21 mo old, the patient’s bone quality had matured enough for definitive surgical management (Figure 3).

**Operation**

Elective surgery was scheduled and the patient was taken to the operating room for a same-day staged combined procedure. Monitoring of somatosensory evoked potentials (SSEPs) and motor evoked potentials (MEPs) was carried out on the upper and lower extremities through the entirety of the case, and appropriate perioperative lines were placed for fluid resuscitation and transfusion as needed during the duration of the surgery.

The patient was placed in the supine position and the halo ring was secured to the operating table with a Mayfield adapter. An incision was on made along the anterior border of the right sternocleidomastoid (contralateral to the anterior dissection carried out in the past) measuring 1.5 inches and careful neck dissection was carried out using the operative microscope to identify the prevertebral space in a standard fashion. After localization using the standard fluoroscopic technique, a C3 and C4 corpectomy was carefully carried out with attention paid to ensure the vertebral arteries were protected given the narrow operative corridor due the narrow and hypoplastic vertebral bodies. After removal of the disc material above and below, we observed that the inferior endplate of C2 and the superior endplate of C5 were at angle of close to 50°, matching our preoperative measurements of the kyphotic angle (Figure 4A). Preoperatively, the decision was made to not fuse at this point to allow for dynamic correction of the kyphotic angle via posterior cervical compression. Though the patient would be temporarily unstable, with neurophysiological monitoring in place, the decision was made to safely reposition the patient prone for the posterior cervical approach using a rigid cervical collar during transfer and positioning instead of placing the patient back in the halo due to technical difficulty with the endotracheal tube and the available support staff to help with positioning. We do acknowledge this risk and believe it would be equally suitable to place a patient back in a halo vest for repositioning. The anterior incision was then closed and the patient was placed in a rigid cervical collar. The patient was placed on a stretcher and carefully rolled and placed prone on the operating table with the halo ring secured to the Mayfield adapter.

The posterior approach was carried out in the standard manner with a midline incision from C2 to C6. The C2-5 lateral masses were exposed and the lamina freed with the use of an angled curette. The kyphosis could easily be appreciated because the spinous processes and facet joints were bulging posteriorly. Having studied the patient’s CT scan preoperatively, we knew the patient’s cervical bones were too small for effective anchoring with lateral mass or translaminar screws. We decided to use laminar hooks (VERTEX, Medtronic Corporation, Dublin, Ireland). Bilateral C2 caudal facing laminar hooks were applied along with rostral facing hooks on C5. After hooks were placed, neurophysiology motor monitoring was performed every 5 min for 20 min and continuous sensory monitoring was performed to make sure there was no mass effect on the spinal canal. A 4.5-mm precut titanium rod was then bent into a lordotic shape and secured to the hooks. The halo ring was then released from the operating table by the anesthesia staff and held with 1° of freedom to allow for extension as the hooks were then compressed while pressure was applied onto the kyphotic apex in order to reduce the kyphosis. This was done under continuous fluoroscopic guidance. Multiple neurophysiological tests were done to make sure there was no compression of the spinal cord. Kyphosis was considered reduced when the endplates of C2 and C5 were parallel to each other. Once that was achieved, we locked down the construct. The lamina, spinous process, and facet joints were then decorticated and demineralized bone matrix (DBM) (PROGENIX DBM Putty, Medtronic Corporation) was then

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applied for posterior lateral fusion and arthrodesis. Standard closure was completed and the patient was placed back into a rigid cervical collar and repositioned supine for the final portion of the surgery.

The halo ring was secured to operating table and the anterior incision was reopened and Caspar retractors were replaced. After Caspar retractor was placed, we proceeded with the placement of strut graft. We considered the placement of expandable cage, polyetheretherketone (PEEK) implant and fibular strut allograft. Because of the small size of the vertebral body, only the fibular strut graft was possible. The allograft was further filed down such that the coronal width of the strut graft was at 11 mm and the sagittal depth was about 10 mm. The strut allograft was filled with DBM and the patient’s autologous bone. With careful distraction on the C2 and C5 vertebral bodies, we wedged the allograft into the corpectomy space. X-Ray and neurophysiology were done at multiple points to make sure there was no dislodging of the posterior instrumentation and to make sure there was no compression of the spinal cord. A careful review of the literature preoperatively revealed the utility of using a low-profile static anterior cervical plate with a single screw at the cephalad and caudad end (Uniplate, DePuy Synthes, New Brunswick, New Jersey) for anterior cervical support in very young patients. The plate was 22 mm and it was secured with 2 10-mm screws. The screws used were self-tapping screws with a blunt end. Fluoroscopy was used at
this point to make sure that the construct was at the midline and to ensure the posterior edge of the strut was within the posterior spinal ligamentous line (Figures 4C and 4D).

**Postoperative Course**

Postoperatively, cervical immobilization was continued with the halo vest. The patient was extubated on postoperative day 2 and had a stable neurologic exam. The patient’s diet was advanced with no difficulty and tolerated solid foods. The patient was discharged home by day 4 after clearance by physical therapy and speech therapy and was followed with serial imaging studies. The patient was brought back to the operating room 6 weeks later for removal of the halo vest and placement of a cervico-thoracic-lumbar supportive orthotic brace. Four months and ten months postoperatively the patient remained in good alignment with evidence of fusion and no hardware complications (Figure 5). The plate was noted to be sitting a bit proud on postoperative imaging. This is because the longus colli was not completely dissected off the vertebral body due to the narrow nature of the body and the risk of vascular injury and Horner’s syndrome with lateral dissection. Therefore, the plate is sitting partially on the muscle and not entirely on the bone. This did not cause any swallowing difficulty and has not been an issue over a year out from surgery. The patient has had stable imaging over 1 yr without evidence of adjacent segment disease or kyphosis. The patient has had a stable neurologic exam and is meeting all her developmental milestones for a child affected by neurofibromatosis. That patient will be followed...
FIGURE 5. A, Preoperative radiograph shows a C3 and C4 anterior vertebral body collapse with resulting cervical kyphosis of 50° at C2-5. B, Six-month postoperative radiograph demonstrating kyphosis correction and early bone fusion with fibula strut graft. C, Ten-month postoperative radiograph showing complete fusion of anterior and posterior construct.

closely throughout her childhood as there is a potential for hardware failure in the future or additionally deformity as the patient grows. Fortunately, the patient’s overall disease burden is well controlled and there are no other lesions within the construct that will need to be followed.

DISCUSSION

The management severe cervical kyphosis in patients with neurofibromatosis presents a challenge to neurosurgeons,
Additionally, increased complexity is encountered in the very young pediatric population. Approximately 26% to 50% of individuals with NF1 have skeletal deformities, of which cervical kyphosis is comparatively rare, with prior series identifying 29 cases out of 146 combined reports from 3 case series. The mean age of the patients diagnosed with cervical kyphosis due to NF1 is 15 yr old and the mean age of those patients undergoing surgical treatment was 14 yr. The youngest patient to undergo cervical fusion for kyphosis correction was 1.7 yr of age and underwent an anterior posterior fusion. Prior studies have also found the angles of cervical kyphosis can range from 16° to 138° with the average of more than 50° delineating when some patients may start to develop paralysis. Careful preoperative evaluation of the patient's exam and radiographic findings ultimately guides management decisions. There are no well-defined indications for the correction of cervical kyphotic deformities in the pediatric population, though it is well documented in NF1 patients with dysplastic skeletal deformities, kyphosis results in more neurological impairment than does scoliosis due to pathological spinal flexion and spinal cord compression. Those patients that ultimately develop neurologic deficits, mechanical neck pain, or have continued progression of their deformity are generally considered candidates for surgical management having failed conservative management with brace treatment, which is not effective in most cases. However, in our very young nonneurofibromatosis patient population, we have found that conservative management with a rigid cervical collar can provide stability and time for hypoplastic vertebral bodies to develop and allow for the resolution of the kyphotic deformity over time.

Spinal fusion procedures in patients with neurofibromatosis and in very young pediatric patients provide their own individual challenges and when taken together can pose a technical challenge. The young age of the patient is advantageous in that the patient tolerates halo fixation relatively well and there is a potential for good bone fusion after surgery. Previous works have shown that circumferential cervical fusion is required to restore normal alignment in patients with severe cervical kyphotic deformities. Posterior fusion alone has a reported incidence of failure as high as 72% with dystrophic kyphotic curves of 50° or more, while those patients who underwent sequential anterior and posterior fusions in 1 study found the failure rate to decrease from 53% to 23%. Children with neurofibromatosis- and dystrophic-type spinal deformities are at high risk for pseudoarthrosis, pseudoarthrotic defects in the fusion mass, spondylolisthesis and hardware failure.

Surgical management and instrumentation in the very young patient population for the cervical spine instability has been discussed in a few studies. Due to the small nature of the cervical bones in very young patients, compromises must be made with the instrumentation. Additionally, there is variation in the surgical management of these patients. In 8 patients managed by Ma, 6 patients underwent a 3-part 540° combined procedure spread over weeks. Those patients undergoing the 540° combined approach underwent a dorsal wedge osteotomy for fixed cervical kyphosis to release dorsal elements followed by a ventral corpectomy and final posterior instrumentation and fusion. This sequence mandates that the anterior corpectomy and graft placement with fusion provide most the correction due to “superior surgical leverage” that may be only attainable in older patients.

Given the bone quality in very young pediatric patients, there would be significant risk to end plate damage if the corrective burden and leveraging were to be placed on the anterior construct initially. Therefore, we started with the anterior procedure to release the structural elements to allow for more range of motion with extension. The use of sublaminar hooks and compression allowed for safer manipulation with dynamic compression of the construct and patient extension to correct the kyphotic angle by monitoring the alignment of the anterior vertebral body endplate under live fluoroscopy. Therefore, the final placement of the fibula strut graft could be completed without concern for possible violation of the endplates after removal of the stress from the kyphotic curve. All stages of the 540° combined approach were completed during 1 surgery that has not been described before. Surgical correction of the kyphotic deformity using the combined approach and the careful pre- and postoperative management of this patient highlights the variety of tools that may be employed in treating this very rare pathology in a unique patient population.

CONCLUSION

The management of very young pediatric patients with NF1 who develop cervical dystrophic kyphosis poses a variety of management, surgical, and technical difficulties. Our case, to our knowledge, is the first to describe the complete management of a patient presenting at 4 mo of age to undergoing definitive surgical treatment at 21 mo of age. Using cervical orthotics, halo traction, halo vest, and cervical fusion, we were able to develop a management schema that offered low morbidity and improved functional outcome. Careful review of the literature on patients with NF1 and cervical deformities undergoing surgical correction and the literature focused on young pediatric patients requiring subaxial cervical fusion allowed us to develop a novel surgical technique to provide for cervical alignment and stability in a 21-mo old patient. We agree that a combined anterior and posterior internal fixation and fusion is a good option for kyphosis correction and a 540° combined approach during a single surgery is feasible in a very young patient by using sublaminar hooks for posterior fixation and dynamic realignment and a static single screw uniplate system for anterior stabilization and fusion.

Disclosure

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.
REFERENCES


COMMENTS

The authors should be commended for the thoughtful management of a complex problem. Management of progressive cervical kyphosis in an NF1 patient is very challenging at this young age and the authors have reported a successful short-term outcome. They summarize available literature to support their surgical approach. Ultimately, given the infrequent presentation of such difficult patients, larger multi-center studies may provide additional insight into the optimal management of these patients.

Steven Hwang
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The authors are to be congratulated for the management of a very difficult case of progressive kyphosis of the cervical spine in a toddler with NF1. Their surgical approach, while aggressive, seems to have achieved satisfactory clinical and radiographic outcomes at 14 months (short-term follow-up) after surgery. Long-term follow-up for children entails monitoring them for decades. This patient will require life-long follow-up to survey for iatrogenic consequences of spine surgery at such a young age, such as short neck, crankshaft deformity, and adjacent level disease. It is hoped that the authors will publish a continuation of their patient’s story 5, 10, 15, and even 20 years down the line. From a technical standpoint, as a specialty, we have developed ways to place adult-sized spinal instrumentation into the smallest of spines, but what we have not answered as a specialty is if we are truly improving our patients’ quality of life and natural history of disease, or have we “kicked the can down the road” and simply created new problems for these patients to deal with as adolescents and young adults.

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