Upper cervical spine fusion in the pediatric population

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The outcomes of 25 pediatric patients who underwent upper cervical or occipitocervical fusion at the authors' institution since 1983 were reviewed. At a mean age of 9 years, the patients presented with spinal instability that was associated with os odontoideum in 11 cases, rotatory subluxation in five cases, odontoid fracture in two cases, atlantooccipital dislocation in two cases, and congenital atlantoaxial instability in five patients, four of whom had Down's syndrome (trisomy 21). Ten children had abnormal findings on neurological examination preoperatively; however, nine experienced improvement or resolution of deficits as of their latest follow-up evaluation (mean 17 months). Fusion was achieved with the first operation in 21 of 25 patients; eventually it was attained in all but one. Four patients exhibited persistent spinal instability after an initial procedure. This was caused by erosion of a multistranded cable through the intact arch of C-2 in two cases, by pin site infection necessitating early halo removal in one case, and by slippage in a halo following a Gallie procedure, which was revised with a Brooks fusion in one case. This series, the largest yet published, shows that with appropriate surgical management, posterior upper cervical fusion in the pediatric population is highly successful. Careful attention to halo pin site care and caution in using multistranded cable in young patients may improve results.

Key Words * cervical fusion * atlantoaxial stabilization * atlantoaxial fusion * Down's syndrome * halo vest * children

Upper cervical instability may occur in children for several reasons. First, traumatic injuries to the pediatric cervical spine tend to involve the first two vertebrae, rather than the inferior cervical spine as is the case in adults.[1,10,13,18,21] Second, instability at this level may be congenital; it tends to be diagnosed early in life, especially in patients with Down's syndrome in whom screening cervical radiographs are now often required before participation in certain physical activities. Third, several rheumatological and connective tissue disorders, including juvenile rheumatoid arthritis, juvenile ankylosing spondylitis, seronegative enthesopathy and arthropathy syndrome, and Morquio's syndrome, can present accompanied by instability at this level.[5,6,11,12,14,30] Finally, os odontoideum with associated atlantoaxial instability often manifests during childhood with the onset of a neurological deficit or neck pain after minor trauma.[4,8,9,15,26] Because of the paucity of data regarding the success of upper cervical fusion in the pediatric population, we reviewed our series of 25 children treated surgically for upper cervical instability.

CLINICAL MATERIAL AND METHODS

Patient Population

The surgical case records of the Department of Neurosurgery from 1983 onward were reviewed to find all cases in which a cervical fusion procedure had been performed in a person under 18 years of age. Twenty-five patients were found to have undergone posterior upper cervical fusion (Table 1). The mean patient age at the time of presentation was 9.4 years (range 3-16 years), and both genders were represented (11 boys and 14 girls).

TABLE 1 PREOPERATIVE INDICATIONS AND TYPE OF SURGERY IN 25 PEDIATRIC PATIENTS REQUIRING UPPER CERVICAL FUSION*						
Case No.	Age (yrs), Sex	Diagnoses at Presentation	Initial Neurological Exam	Surgeon	Levels in Construct	Construct Type
1	5, F	AOD	rt hemiparesis (0-1/5), rt Horner's, rt hemidiaphragm paralysis	1	Oc-C2	sublam C-1 & C-2
2	13, M	type II odontoid fracture	intact	1	C1-2	Brooks
3	6, F	C1-2 rotatory subluxation	intact	5	C1-2	triple wire
4	3, F	os odontoideum	intact	1	C1-2	Brooks
5	12, F	os odontoideum	intact	2	C1-2	Brooks
6	3, M	trisomy 21, C1-2 instability	intact	2	C1-2	Brooks
7	10, M	os odontoideum	decreased proprioception, hyperreflexia, bilat LE	6	C1-2	Brooks
8	6, M	os odontoideum	intact	1	C1-2	Brooks
9	9, M	os odontoideum, VA dissection	rt CN VII palsy, rt nystagmus, rt dysmetria	1	C1-2	Brooks
10	5, F	trisomy 21, Oc-C1 & C1-2 instability	bilat LE hyperreflexia	1	Oc-C2	#22 ga. wire sublam C1-2; #18 ga. Oc-C2
11	4, F	os odontoideum	intact	2	C1-2	Brooks
12	16, M	os odontoideum	lt hemiparesis	2	Oc-C2	Brooks
13	15, F	trisomy 21, C1-2 instability	bilat UE hyperreflexia	2	C1-2	Gallie
14	13, F	os odontoideum	intact	2	C1-2	Brooks
15	10, F	C1-2 rotatory subluxation	intact	1	C1-2	Brooks
16	8, M	congenital C1-2 instability	dysphagia, hemiparesis	1	TOO	
17	6, M	JRA, C1-2 rotatory subluxation	intact	1	C1-2	Brooks
18	9, F	C1-2 rotatory subluxation	intact	2	C1-2	Gallie
19	12, M	os odontoideum	intact	2	C1-2	Brooks
20	9, F	type II odontoid fracture	intact	3	C1-2	Brooks w/multistranded cable
21	11, F	AOD	CN IX, X, XI, XII deficits, rt hemiplegia	2	Oc-C3 Oc-C2	pediatric CD rods multistranded cable; sublam C-2
22	14, F	trisomy 21, C1-2 instability	bilat LE hyperreflexia	4	Oc-C2	multistranded cable; sublam C-2
23	14, M	os odontoideum	intact	4	C1-2	Brooks w/multistranded cable
24	10, F	C1-2 rotatory subluxation	intact	2	C1-2	Brooks w/multistranded cable

24	10, F	C1-2 rotatory subluxation	intact	3	C1-2	Brooks w/multi-stranded cable
25	13, M	os odontoideum	rt hemiparesis	3	C1-2	Brooks w/multi-stranded cable

* AOD = atlantooccipital dislocation; CD = Cotrel-Dubousset; CN = cranial nerve; ga = gauge; Horner's = Horner's syndrome; JRA = juvenile rheumatoid arthritis; LE = lower extremity; Cc = occiput; sublam = sublaminar; TOO = transoral odontectomy; UE = upper extremity; VA = vertebral artery.

Patient Presentations

Of the 15 patients presenting with normal findings on examination, all but one had neck pain. Three patients presented with hyperreflexia alone. One patient presented with normal motor examination responses, but decreased proprioception in his lower extremities. Although 15 children presented without neurological deficit, only one was totally asymptomatic. A total of eight children in the study had neck pain and one reported interscapular pain. A 3-year-old boy with Down's syndrome had been noted to have a right head tilt for 1 year, but he did not have rotatory subluxation. One patient with os odontoideum had experienced whole body dysesthesias transiently after a dive into a shallow pool and another had a transient monoparesis. These deficits had fully resolved by presentation in both cases. One patient with a Type II odontoid fracture had experienced transient mild quadriparesis that fully resolved by presentation.

Ten children had abnormal neurological findings at presentation (Table 1). Three were hyperreflexic without associated motor or sensory deficits. Three had lower cranial nerve deficits, accompanied by hemiparesis in two patients. Three other children also presented with a form of myelopathy, and another presented with sensory deficits only but normal motor responses. Three children presented with deficits referable to regions of the neuroaxis above the foramen magnum. An 8-year-old boy presented with dysphagia and a mild hemiparesis. Another patient presented at the age of 9 years with repeated emesis and ataxia; on examination this child demonstrated unilateral nuclear facial weakness and dysmetria with a contralateral hemiparesis. A third patient, comatose following a severe motor vehicle accident, presented with deficits of the ninth, 10th, 11th, and 12th cranial nerves with hemiparesis. Two other patients presented with hemiparesis.

Radiological Examination

Plain cervical radiographs (anteroposterior, lateral, and open-mouth odontoid views) were obtained in all patients. Additional lateral views in flexion-extension were obtained in all cases of os odontoideum and congenital instability, but were avoided in the two patients with newly presenting odontoid fractures; the latter injuries were further evaluated using computerized tomography scanning in one case and plain tomography scanning in the other. Plain tomography scanning was also used to evaluate two cases of os odontoideum in lateral views and one case of rotatory subluxation in anteroposterior views. Computerized tomography and magnetic resonance imaging were used in all but three cases in this series. A single patient was evaluated by means of four-vessel cerebral angiography, which revealed a vertebral artery dissection and subsequent occlusion associated with os odontoideum.

In the cases of os odontoideum determined to be unstable, the atlantodental interval ranged from 6 to 11 mm, whereas movement of the atlas on the axis on flexion-extension views ranged from 4 to 12 mm.

Preoperative Diagnosis

Atlantoaxial instability associated with os odontoideum was the most common diagnosis following

clinical and radiographic evaluation and was made in 11 of 25 cases. Atlantoaxial rotatory subluxation was present in five patients, one of whom also had juvenile rheumatoid arthritis. Type II odontoid fractures were newly diagnosed in two patients. Congenital atlantoaxial hypermobility was found in five patients, four of whom had Down's syndrome. Two patients had traumatic atlantooccipital dislocation.

Operative Technique

After a sedative had been administered, all patients remained conscious while they were intubated using fiberoptic techniques to minimize the risk of inadvertent spinal cord injury. Most children were then placed in a halo orthosis and turned to the prone position. Cervical alignment was confirmed using intraoperative radiography. The operative technique used in 19 children consisted of a Brooks-type fusion, using an interlaminar block of bicortical bone and bilateral sublaminar wires. One case was supplemented with interspinous wires. The Gallie technique was used in two patients: one with rotatory subluxation, but no apparent translational instability, and another with a congenitally narrow canal at C-2. Four patients underwent primary occipitocervical fusion. Autologous iliac crest bone graft was used in each case.

In 19 patients, internal fixation was achieved by using 18-gauge wire. In six children treated more recently, a multistranded cable was used. Somatosensory evoked potential monitoring was used in all patients to detect dorsal column impingement during wire passage. A radiograph was obtained to confirm the alignment at the end of each procedure.

Postoperative Immobilization

Immobilization was generally continued for 3 months. Postoperatively, one child was immobilized in a hard collar and all others in a halo orthosis. The halo ring was secured using four pins in most cases, with two placed anteriorly in the forehead and two placed posteriorly. When placing the posterior pins, the patient's hair was generally not shaved but rather prepared using iodoform solution. Each pin was tightened using 0.9 J (8 in/lb) of torque and retightened 1 day later. Pin care consisted of swabbing each pin site three times daily with half-strength hydrogen peroxide solution.

RESULTS

Rate of Fusion

Fusion was achieved in 21 patients (84%) after the initial operation (Table 2) and in three more patients after a subsequent procedure. Fusion was determined to have taken place on the basis of both a lack of movement on flexion-extension lateral cervical films and the presence of intervening calcified osteoid. The mean radiographic follow-up period was 13 months (range 4-32 months).

TABLE 2
POSTOPERATIVE COURSE IN 25 PEDIATRIC PATIENTS WHO UNDERWENT UPPER CERVICAL FUSION*

Case No.	Levels Fused	Immobilization Type	Follow Up (mos)	Follow-Up Exam	Complications
1	Oc-C2	halo	76	rt UE 4/5; rt LE 5/5; lt side 5/5; rt LE hyperreflexia	mild scoliosis; rt LE 2.4 cm short
2	C1-2	halo	5	intact	halo pin site infection
3	C1-2	halo	14	intact; no pain	halo pin site infection
4	C1-2	halo	22	intact	none
5	C1-2	halo	2	intact	none
6	C1-2	halo	14	mild head tilt to rt; intact	halo pin site infection; C-2 on C-3 subluxation
7	C1-2	halo	7	intact	none
8	C1-2	halo	10	intact	none
9	C1-2	halo	14	rt dysmetria improving; rt side hyperreflexia	none
10	Oc-C2	halo	12	intact	none
11	no fusion	halo		intact	halo pin site infection; early halo removal; failed fusion
	Oc-C3	halo	28	intact	
12	no fusion	halo	32	lt UE 4/5; lt hyperreflexia	postop slippage
	C1-2	halo			
13	C1-2	halo	4	intact	none
14	C1-2	halo	20	intact	none
15	C1-2	halo	28	intact	none
16	NA	NA	14	dysphagia improving; hemiparesis resolved	none
	C1-2	halo			
17	C1-2	halo	6	intact	none
18	C1-2	halo	7	intact	halo pin site infection
19	C1-2	halo	7	intact	none
20	no fusion	halo	29	intact	graft resorption; fusion failure
	Oc-C3	halo			
21	Oc-C2	halo	9	dysarthria improving; hemiplegia resolved; hyperreflexia	none
				bilat LE hyperreflexia	
22	no fusion	halo	32		fusion failure; further surgery deferred by parents & physician
23	C1-2	collar	13	intact	none
24	C1-2	halo	4	intact	none
25	C1-2	halo	4	intact	halo pin site infection; pin moved

* LE = lower extremity; NA = not applicable; Oc = occiput; UE = upper extremity.

Neurological Examination

No patient experienced neurological decline after surgery. Of the 10 patients with abnormal neurological findings, three displayed hyperreflexia without a deficit, which resolved in two patients and remained unchanged in the third. One patient, who presented with normal motor function but decreased proprioception in his lower extremities, had normal responses when examined 7 months after surgery. Three children presented with deficits referable to regions of the neuroaxis above the foramen magnum. An 8-year-old boy with congenital atlantoaxial instability presented with dysphagia and mild hemiparesis. Fourteen months after odontoidectomy and atlantoaxial fusion, his hemiparesis had resolved and his dysphagia was improving. Another patient presented with cerebellar infarctions due to atlantoaxial instability, an association previously reported by others.[2] At the age of 9 years, this patient developed unilateral vertebral artery dissection (which led to occlusion) associated with os odontoides

and instability. He presented with unilateral nuclear facial weakness, dysmetria, hemiparesis, and repeated emesis. After atlantoaxial fusion, these deficits resolved and his only limitation was moderate dysmetria. A third patient who had survived atlantooccipital dislocation caused by a motor vehicle accident presented with deficits of the ninth to 12th cranial nerves and with hemiparesis. Nine months after an occiput-C2 fusion, she showed improvement of her hemiparesis and had only a moderately severe dysarthria. Two other patients presented with hemiparesis. One experienced resolution of his deficits and the other had improved at the latest follow-up review. Overall, of the 10 patients presenting with abnormalities on initial neurological examination, five experienced resolution and four improvement of the original findings, whereas one patient presenting with hyperreflexia only showed no improvement at a mean clinical follow-up period of 17 months (range 2-32 months).

Postoperative Complications

Repeated Operation. Repeated operation was required in three patients. A 16-year-old boy with os odontoideum sustained an early loss of reduction in the 1st week after undergoing a Gallie procedure; he underwent a Brooks procedure with subsequent fusion. No additional autologous bone was harvested for the second procedure. A 4-year-old girl with os odontoideum developed multiple halo pin site infections, leading to early removal of the hardware at 4 weeks. Her fusion failed, being associated with a recurrence of 12 mm of movement of C-1 on to C-2. She then underwent an occiput-C2 fusion procedure that required additional autologous as well as allograft bone 10 weeks after the first operation. Radiographic follow-up evaluation at 16 months showed fusion extending to include C-3 as well. This was the only case in which fusion extended beyond the construct. A 9-year-old girl with a Type II fracture showed evidence of resorption of her autologous bone graft, severe thinning of C-1 and complete erosion of the posterior ring of C-2 by both multistranded cables 3 months after her initial surgery. After her pathological hypermobility did not respond to further halo immobilization, an occiput-C3 procedure with pediatric Cotrel-Dubousset rods and additional autologous bone resulted in successful fusion.

Of the four children who underwent primary occiput-C2 fusion, fusion was successful in three. Two months after surgery, the fourth, a 14-year-old girl with instability associated with trisomy 21, experienced complete erosion of the ring of C-2 where a multistranded cable had been positioned. This appeared to be caused by a sawlike effect of the cable on the bone caused by micromotion at the level of fusion despite postoperative halo immobilization. Thirteen months after surgery, flexion-extension radiographs showed 5 mm of movement between C-1 and C-2, a reduction from 11 mm; however, the patient's parents declined further operation. This complication associated with multistranded cable occurred in two of the six cases in which it was used, despite the avoidance of overtightening. In both cases there was a clear indication for repeated operation, although surgery was declined in one case.

Orthosis Complications. Complications related to the use of a halo device occurred in seven of the 24 patients in whom it was used, two of whom required repeated operation. Slippage in the halo with resultant loss of reduction occurred in one patient, a 16-year-old boy with os odontoideum who required two surgeries. Of the six patients in whom a pin site infection occurred, the device was removed early in two after multiple attempts at infection control had failed. In one of the two, a 4-year-old girl with os odontoideum, the halo was removed after 4 weeks, and as a result she required a second operation for fusion.

DISCUSSION

As noted by Smith and colleagues,[25] fusion of the upper cervical spine in the pediatric population has

rarely been addressed in previous communications. We present 25 patients who underwent such a procedure at our institution. Fusion was achieved in all but one patient; however, a second operation was required in three patients. The four patients in whom the first procedure failed varied in age and diagnosis, and their procedures were performed by different surgeons using different techniques. The mean of their ages (4, 9, 14, and 16 years) exceeds that of the whole series by only 18 months. The four patients were surgically treated by three different surgeons. The fusion methods also varied: one patient underwent a Gallie fusion, one an occiput-C2 fusion, and two underwent Brooks fusions. Autologous iliac crest bone grafts and postoperative halo immobilization were used in all four patients. No trend toward greater blood loss or longer operative time, which may suggest a technically more difficult case, could be found.

Each case of fusion failure was associated with a complication of either postoperative immobilization or internal fixation. In two of the six cases in which multistranded cables were used, the cable appeared to have sawed through the lamina of C-2. It should be emphasized that the cables in these patients were not overtightened and both patients were also immobilized in a halo vest. Based on the intraoperative appearance of the rings of C-1 and C-2 in the one patient who underwent repeated operation, it was clear that the braided wires had eroded through the posterior arches at these levels. The bone medial to the wires at C-2 had largely been resorbed. The mechanism for this effect may be either avascular necrosis of the intervening bone or micromotion-induced sawing of the cable through the bone, which is inherently thinner and more fragile than that of adults. Accordingly, we have stopped using this type of cable in young children and have returned to using 18-gauge wire.

The other two fusion failures were associated with halo complications. In one case, severe, multiple pin site infections lead to early removal of the halo after 4 weeks. Despite subsequent immobilization in a rigid cervical collar, this patient later required occiput-C2 fusion. Another patient immobilized in a halo suffered an early loss of reduction that could not be regained nonsurgically, leading to a second operation 9 days after the first. Fusion was ultimately achieved in both patients.

Postoperative Immobilization

This series raises an additional issue concerning the optimum mode of cervical immobilization after a fusion procedure. Although some surgeons have favored using a hard collar after a Brooks-type operation,[25] our results suggest that this may not provide optimum immobilization. While placed in a halo device, two of 24 children had early loss of alignment, as did one of the two children in whom the halo was removed early because of pin site complications. It is our belief that in a potentially active pediatric patient who may not obey the same activity restrictions as an adult, rigid external immobilization is needed to minimize intersegmental micromotion that can interfere with a successful fusion. Although a Minerva cast may provide a comparable degree of immobilization,[17] this type of brace is often tolerated less well than a halo.

A hard collar might provide adequate immobilization in appropriately selected older children who may be candidates for highly rigid internal constructs such as C1-2 transarticular screws or an odontoid screw (for an acute odontoid fracture).[3,7,16,28] These techniques have only been in widespread use for the last few years and are often not applicable in preadolescent children, who may lack adequate bone thickness for good screw purchase. None of our recently treated patients has been a good candidate for such procedures.

Down's Syndrome

Four of the patients in our series had trisomy 21 (Down's syndrome), which can be associated with either atlantoaxial or atlantooccipital hypermobility.[19,20,22-24,27,29] Although most of the literature concerning this association deals primarily with instability of C1-2, instability at occiput-C1 must also be ruled out in each patient prior to atlantoaxial fusion or greater instability may result. This point is well illustrated in a study by Menezes and Ryken[20] in 1992, in which they described two adolescents with instability at both segments who progressed to develop basilar invagination following fusion only at the atlantoaxial level. Of the four patients in our series with trisomy 21, two had hypermobility at both levels and underwent occiput-C2 fusion. Fusion failure associated with the use of multistranded cable occurred in one of these patients. Fusion was successful in both patients who underwent atlantoaxial fixation. Although neither patient developed postoperative atlantooccipital hypermobility, one does have a fixed 3-mm C-2 on-C-3 anterior spondylolisthesis that was not present preoperatively; however it has not required further treatment. The presence of hypermobility at two levels in two of the four children with trisomy 21 in this series further illustrates the importance of determining whether craniocervical instability is present prior to atlantoaxial fusion in these patients.

CONCLUSIONS

We conclude that posterior upper cervical fusion in the pediatric population, when coupled with rigid external immobilization, has a high rate of success if properly performed. Because fusion failures may result from inadequate postoperative immobilization with resultant intersegmental micromotion, we favor continuing rigid immobilization until a solid fusion mass is visible radiographically. We also conclude that multistranded cable should be used for such patients with caution because the cable may erode through the underlying lamina before a solid fusion can occur.

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