Craniocevical junction fusions in patients with hindbrain herniation and syringohydromyelia

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Object. Patients with hindbrain herniation or the so-called Chiari malformation Type I (CM-I) and/or syringohydromyelia are treated with dorsal decompression alone; however, a small percentage of patients with other associated abnormalities require concomitant dorsal craniocervical junction (CCJ) fusion. The authors surveyed the indications for CCJ fusions in this population.

Methods. A retrospective review of University of Iowa medical records and radiographs obtained between 1996 and 2005 was performed. Inclusion criteria encompassed patients with diagnoses of CM-I and/or syringohydromyelia requiring dorsal CCJ fusions, and others with CCJ abnormalities who had CM-I and/or syringohydromyelia.

Results. Two hundred thirty-four patients were identified, all of whom were symptomatic at presentation. Their ages ranged from 2.5 to 86 years; 33% of the patients were <16 years of age. Patients were categorized as follows, with some being assigned to >1 category: Group I, congenital or acquired CCJ abnormalities with reducible bone compression (25% of patients); Group II, previous anterior CCJ/upper brainstem decompression (44%); Group III, occipitocervical complex instability with CM-I and/or syringohydromyelia but without CCJ bone abnormalities requiring adjunctive posterior fossa decompression (26%); and Group IV, musculoligamentous instability, either from pathological states or from muscle dehiscence from repeated posterior fossa procedures (14%). Instrumentation was used in 96% of patients, with all 96% receiving semirigid fixation with titanium loop and sublaminar cables; all fusion constructs incorporated autologous bone. At last follow-up evaluation, fusion was radiographically complete in 97%, and symptom improvement was seen in 92%.

Conclusions. Dorsal CCJ fusions are required in patients with CM-I and/or syringohydromyelia who have concomitant CCJ abnormalities (Groups I and II). A definite group (CM-I and/or syringohydromyelia) without bone abnormality exists (Groups III and IV). This may be due to muscle weakness secondary to a high syrinx.

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Key Words • Chiari malformation Type I • craniocervical fusion • dorsal fusion • syringohydromyelia

Abbreviations used in this paper: BMP = bone morphogenetic protein; CCJ = craniocervical junction; CM-I = Chiari malformation Type I; CMJ = cervicomedullary junction; CSF = cerebrospinal fluid; CVJ = craniovertebral junction; FMD = foramen magnum decompression; PFD = posterior fossa decompression.
Methods

A retrospective review of University of Iowa medical records and radiographs obtained between 1996 and 2005 was performed. Inclusion criteria were patients with diagnoses of hindbrain herniation and/or the presence of a syrinx in the cervical spinal cord and who required a dorsal craniocervical fusion; this included patients with other CCJ abnormalities associated with CM-I and/or syringohydromyelia. Hindbrain herniation was defined as cerebellar tonsillar descent to below the level of the foramen magnum, and the patient needed to have referable symptomatology for inclusion.

Patient Presentation

Over this 10-year period, 234 patients were identified who met the aforementioned criteria. The patient age at presentation ranged from 2.5 to 86 years (mean 36 years); 33% of the patients were <16 years old.

The duration of symptoms ranged from 1 month to 150 months (mean 29.75 months). All patients were symptomatic on presentation, with head or neck pain (78% of patients) being the most common feature (suboccipital headache alone was present in 59% of patients). A categorization of presenting signs and symptoms is shown in Table 1.

Diagnosis of the Anomaly

Diagnosis of the anomaly was made by assessing the presenting symptoms and signs and evaluating them in the context of adequate imaging studies. This preoperative investigation was critical and was believed to be thoroughly conducted to obtain the maximum benefit of a surgical procedure. This was most effectively accomplished by complete imaging of the CVJ with both preoperative CT and MR imaging studies. When possible, these studies were performed at our institution and included a 3D reconstruction of the craniocervical CT to better define the surgical anatomy and bone anomalies. Also, dynamic MR imaging in both flexion and extension views, which were obtained to evaluate the extent of ventral and dorsal cervicomedullary compression, respectively, was found to be a key component in determining the reducibility of the abnormality at the CCJ. Such comprehensive preoperative imaging of the neck in flexion and extension was also necessary to assess the stability of the cervical spine. As reviewed previously, unstable configurations at the CCJ are outlined in Table 2. Stabilization is of paramount importance for reducible lesions to maintain the neural decompression. Irreducible lesions require decompression at the site of compression, followed by fusion. Relief of symptomatic ventral compression seen on flexion MR images obtained with the spine in the extended position was a red flag indicating the need to fuse. The position of the odontoid was taken into consideration when positioning the patient for surgery, because cervical flexion could exaggerate the ventral compression on the CMJ.

In an effort to further explore the indications for dorsal fusions in patients presenting with hindbrain herniation and/or syringomyelia, the patients were categorized as follows, with some in >1 category: Group I, patients with reducible CCJ bone abnormalities causing compression; Group II, patients having undergone a previous anterior decompression of the ventral brainstem; Group III, patients having occipitocervical complex instability without CCJ bone abnormalities; and Group IV, patients with musculoligamentous instability. A summary of these patient categories, definitions, and the number included in each grouping is presented in Table 3.

Sixty-five patients presented with syringohydromyelia in addition to their bone abnormalities. The bone anomalies associated with CM-I/syringohydromyelia in these patients were numerous and sometimes superimposed, as presented in Table 4.

Management of the Disease

All patients underwent dorsal fusions. Of such fusions performed, the most common concomitant decompression was that of the posterior fossa (170 patients), either alone (119 patients) or combined with a ventral decompression of the brainstem (51 patients) via the transoral approach. Dorsal fusion was the only procedure performed in 33 patients; these individuals comprised Group IV (see Table 3). In the cases in which potentially reducible bone compression was present at the CCJ (that is, in patients com-

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### Table 1

<table>
<thead>
<tr>
<th>Signs &amp; Symptoms</th>
<th>No. of Patients (%)</th>
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<tbody>
<tr>
<td>headache</td>
<td>136 (58)</td>
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<tr>
<td>neck pain</td>
<td>46 (20)</td>
</tr>
<tr>
<td>dysphagia</td>
<td>46 (20)</td>
</tr>
<tr>
<td>decreased gag response</td>
<td>91 (39)</td>
</tr>
<tr>
<td>upper-extremity weakness</td>
<td>56 (24)</td>
</tr>
<tr>
<td>upper-extremity numbness/paresthesia</td>
<td>86 (37)</td>
</tr>
<tr>
<td>lower-extremity weakness</td>
<td>5 (2)</td>
</tr>
<tr>
<td>lower-extremity numbness/paresthesia</td>
<td>16 (7)</td>
</tr>
<tr>
<td>gait instability</td>
<td>62 (27)</td>
</tr>
<tr>
<td>hyperreflexia</td>
<td>143 (61)</td>
</tr>
<tr>
<td>C-2 numbness</td>
<td>46 (20)</td>
</tr>
<tr>
<td>facial numbness</td>
<td>28 (12)</td>
</tr>
<tr>
<td>torticollis</td>
<td>34 (15)</td>
</tr>
<tr>
<td>dizziness</td>
<td>23 (10)</td>
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</tbody>
</table>

* Many patients had > 1 sign or symptom.

### Table 2

<table>
<thead>
<tr>
<th>Criteria for Unstable Configurations</th>
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<tbody>
<tr>
<td>predental space &gt;5 mm in patients &lt;8 yrs of age, &gt;3 mm in those 8–10 yrs &amp; older</td>
</tr>
<tr>
<td>separation of atlas lat masses &gt;6 mm</td>
</tr>
<tr>
<td>vertical clivus odontoid translation &gt;2 mm</td>
</tr>
<tr>
<td>gap btw occipital condyles &amp; atlas facets on lat cervical radiographs</td>
</tr>
<tr>
<td>abnormal relationship btw spinal canal &amp; foramen magnum</td>
</tr>
<tr>
<td>abnormal craniocervical motion dynamics</td>
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* Adapted from Menezes and VanGilder.

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posing Group I), the patient was placed in halo-ring traction preoperatively to obtain the optimum degree of decompression; this was maintained by instrumentation and fusion in an extended position after the PFD.

The PFD Surgical Procedure

The PFD procedure followed by dorsal occipitocervical fusion was performed in the 59 patients comprising Group I as well as in the 60 patients in Group III. The extent of PFD varied on a per-patient basis, but always involved the following: an occipital craniectomy; partial C-1 (superior two-thirds) laminectomy; intraoperative ultrasonography confirmation of dorsal decompression; intradural exploration and removal of constricting intradural bands (when present, in ~30% of patients); adequate tonsillar shrinkage and lysis of arachnoid adhesions to visualize CSF flow; watertight dural closure with a cervical fascia graft; and meticulous, layered muscle closure.

Transoral–Transpalatopharyngeal Surgical Procedure

The 103 patients comprising Group II underwent a standard transoral–transpalatopharyngeal approach for ventral decompression of the brainstem by the senior author (A.H.M.). This procedure was combined with a PFD and concomitant dorsal craniocervical fusion in 51 patients; in 52 patients a limited dorsal FMD was made to allow for the occipitocervical fusion. The transoral approach is described in detail elsewhere18,19 but is summarized as follows: induction of general anesthesia is followed by intraoral placement of the Dingman self-retaining retractor, which allows ample exposure to the soft palate. A midline incision is made here, followed by one in the posterior pharyngeal midline raphe. Magnification provided by the operating microscope is introduced where the prevertebral fascia and longus colli musculature are mobilized laterally; this allows the anterior inferior clivus as well as the anterior arches of the atlas and axis vertebrae to be visualized. The anterior arch of the atlas and the odontoid process are then removed using a high-speed drill, allowing for visualization of the retroodontoid space and ensuring that there is adequate ventral decompression of the brainstem. After ensuring that the integrity of the dura mater is preserved, the wound is then copiously irrigated and closed in multiple layers.

Dorsal Occipitocervical Fusion Procedure

Overall, 96% of patients underwent instrumented and 4% had noninstrumented fusions. Of these, 96% underwent fusion from the occiput to C-2 or C-3, with semirigid fixation via titanium loop and sublaminar titanium cables and bone. Rigid constructs included posterolateral C1–2 plating systems, C1–2 transarticular screws, and C-1 lateral mass to C-2 pedicle screw fixation constructs, and these were used in the remainder of cases. All fusion constructs incorporated autologous bone; either grafts from the patient’s right ninth rib, from posterior spinal elements, or from the calvarium. A micro-air impactor was used to harvest bone. We used BMP and/or bone graft composite in most cases. In children < 6 years of age, rib-graft strut fusion was usually performed (90%); instrumented fusion took place in the 10% who had adequate bone mass.
TABLE 4

Associated diagnoses in 234 patients with hindbrain herniation and/or syringohydromyelia treated between 1996 and 2005*

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>No. of Patients†</th>
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<tr>
<td>syringomyelia</td>
<td>65</td>
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<tr>
<td>Klippel–Feil anomaly</td>
<td>57</td>
</tr>
<tr>
<td>atlas assimilation</td>
<td>50</td>
</tr>
<tr>
<td>basilar invagination</td>
<td>56</td>
</tr>
<tr>
<td>Down syndrome</td>
<td>10</td>
</tr>
<tr>
<td>osteogenesis imperfecta</td>
<td>3</td>
</tr>
<tr>
<td>Ehlers–Danlos syndrome</td>
<td>3</td>
</tr>
<tr>
<td>VATER complex</td>
<td>2</td>
</tr>
<tr>
<td>Noonan syndrome</td>
<td>1</td>
</tr>
<tr>
<td>Goldenhar syndrome</td>
<td>1</td>
</tr>
<tr>
<td>spondylometaphyseal dysplasia</td>
<td>1</td>
</tr>
<tr>
<td>Crouzon syndrome</td>
<td>1</td>
</tr>
<tr>
<td>Aepert syndrome</td>
<td>1</td>
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</tbody>
</table>

* VATER = vertebral defects, anal atresia, tracheoesophageal fistula with esophageal atresia, and radial and renal anomalies.
† Many patients demonstrated > 1 associated diagnosis.

Results

Occipitocervical fusion was found to be radiographically complete in 97% of patients based on CT scans and lateral cervical spine imaging performed at last follow-up. Symptomatic improvement was seen in 92%. Not surprisingly, the symptoms of suboccipital headache and posterior cervical pain were those that were most commonly relieved by treatment. No change from the preoperative examination or persistence of symptoms was observed in 7%, with headaches again comprising the largest component (5%). Two patients reported a return of dysphagia and dysarthria, as well as lower-limb paresthesias in 1S, although these were subjective findings and were not substantiated by imaging or clinical examination.

In 4% of patients with continued symptoms, repeated tonsillar descent was seen following PFD on follow-up imaging. However, in half of these, dynamic imaging revealed CSF space around the tonsils, and thus no intervention was undertaken. Tonsillar impaction was found to be repeated in the remaining half of patients; 2 of these also had an associated residual cervical syrinx. These patients underwent repeated dorsal decompression and fusion, with placement of a fourth ventricle–subarachnoid shunt in the 2 with residual syringes. Radiographic and symptomatic improvement was confirmed at follow-up.

Sixty-five patients presented with syringohydromyelia in addition to their bone abnormality. Of these, 23 patients presented with syringohydromyelia alone, in the absence of hindbrain herniation. The most common anatomical concurrent finding in these patients was the Klippel–Feil anomaly, with > 1 cervical segmentation failure as well as dystopic os odontoideum. On evaluation of this group together at last follow-up, however, 60 of 65 patients were found to have radiographic improvement or resolution of their syrinx. The remaining 5 patients had persistent syringes, in one of whom a syringosubarachnoid shunt was placed; the other 4 patients had stable conditions and no further treatment plans were formulated.

Follow-Up Duration

The mean duration of follow-up was 20.2 months (range 1 month–10 years). Twenty-eight patients were lost to follow-up, either continuing their care at their referring institutions or failing to return after discharge. No deaths were reported in those patients who continued through to the last follow-up evaluation.

Illustrative Cases

Case 1 (Group I)

This case provides an example of atlas assimilation with basilar invagination that is reducible on extension.

History and Physical Examination. This 11-year-old boy had a history of progressive posterior headache and neck pain over several months, with an even longer history of difficulty walking. He and his family also reported worsening difficulty with gait. Physical examination revealed an absent gag reflex and a broad-based gait, but the remainder of his examination results were normal.

Neuroimaging. Sagittal dynamic MR images obtained in flexion and extension views demonstrated tonsillar herniation as well as a large syrinx extending from C-2 to the thoracic spinal cord. The ventral osseous cervicomedullary compression caused by atlas assimilation seen on flexion views was relieved in extension (Fig. 1A).

Operation. The patient underwent PFD, with intradural lysis of adhesions and tonsillar shrinkage, and concomitant dorsal Oc–C2 fusion with titanium loop and cable instrumentation, calvarial bone grafts, and BMP (Fig. 1B and C). This fusion kept the patient in moderate extension.

Postoperative Course. The patient noted complete symptom resolution after the 12-month follow-up visit and continues to do well, with complete fusion confirmed by radiographic assessment.

Another example of a CM-I with atlas assimilation and reducible atlantoaxial dislocation in extension can be seen in Fig. 1D; this patient was treated with a PFD and dorsal occipitocervical fusion.

Case 2 (Group II)

This case provides an example of a fixed ventral bone compression in which treatment was anterior decompression of the upper cervical cord followed by PFD and fusion.

History and Physical Examination. This 12-year-old girl presented initially to an orthopedist with a 7-month history of progressive claw deformity of her hands, to the extent that she was unable to play her clarinet. She was ultimately referred to our institution, where detailed questioning revealed that she sometimes aspirated liquids while drinking. Physical examination confirmed clawing of her hands, the right worse than the left, with weak interosseous muscles, as well as absent upper-extremity reflexes and hyperactive lower-extremity reflexes; the remainder of her examination, including cranial nerve and gait assessments, was normal.

Neuroimaging. Sagittal and coronal neutral MR imaging demonstrated hypoplastic occipital condyles resulting in basilar invagination and ventral compression of the CMJ; this, combined with cerebellar tonsillar herniation, resulted

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Illustrative Cases
in a large syrinx extending from the CMJ to the conus inferiorly (Fig. 2).

**Operation.** The patient underwent transoral–transpalato-pharyngeal resection of the odontoid and anterior arch of C-1, followed by PFD and dorsal Oc–C2 fusion with titanium loop and cable instrumentation, calvarial bone grafts, and BMP. This fusion kept the patient in mild extension and relieved compression on the spinal cord.

**Postoperative Course.** The patient has noted improvement in her claw deformity after 24 months of follow-up care, and she continues to do well; her fusion is complete according to radiographic studies.

**Case 3 (Group III)**

This case is an example of reduction of a ventral abnormality in the extended position, so that treatment was PFD and fusion.

**History and Physical Examination.** This 22-year-old woman presented to a local facility after colliding with one of her dance partners at practice and hitting her occiput, prompting evaluative imaging, which led her to our institution. She reported posterior cervical and suboccipital pain on presentation to our institution, but further investigation revealed that she had experienced lifelong trouble swallowing solids more than liquids, painful extension of the neck, posterior headaches, and occasional ringing in her ears. Physical examination confirmed a decreased gag response as well as decreased sensation in the left V1 and V2 distributions; the remainder of the results were normal.

**Neuroimaging.** Dynamic MR imaging in flexion and extension views demonstrated CVJ buckling as well as cerebellar tonsillar ectopia to C-2. At the level of the posterior arch of C-1, no CSF was seen surrounding the brainstem or spinal cord. There was no syringohydromyelia noted. On flexion of her neck, it was noted that her anterior brainstem was severely compressed, which was relieved in extension. There was no bone abnormality (Fig. 3).

**Operation.** The patient underwent PFD and dorsal Oc–C2 fusion with titanium loop and cable instrumentation, calvarial bone grafts, and BMP. This fusion kept the patient’s spine in mild extension (Fig. 3).

**Postoperative Course.** The patient has noted complete symptom resolution and at 30 months of follow-up has returned to her previous occupation, with complete fusion based on radiographic assessment.

**Case 4 (Group IV)**

This case is an example of musculoligamentous instability after repeated PFDs, which caused poor muscle approximation and fibrotic scars after repeated operation.

**History and Physical Examination.** This 13-year-old girl had a long, complex history of symptomatic CM-I with brainstem abnormalities, including 10th cranial nerve dys-

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*Fig. 1. Case 1. Sagittal dynamic T1-weighted MR images (A) demonstrating tonsillar herniation and ventral bone CMJ compression in flexion that is relieved on extension. A large syrinx extending from C-2 to the thoracic cord is noted. Intraoperative photographs show the PFD with intradural lysis of adhesions and tonsillar shrinkage (B) and dorsal Oc–C2 fusion with titanium loop and cable instrumentation, calvarial bone grafts, and BMP (C). Sagittal MR images (D) depict another example of a CM-I with atlas assimilation and reducible atlantoaxial dislocation in extension; the patient was treated with a PFD and dorsal occipitocervical fusion.*
function, which had required tracheostomy and gastrostomy when she was an infant. This was concomitant with hydrocephalus and a cervicothoracic syrinx, for which she underwent 2 PFDs between the ages of 3 and 6 months, with placement of a fourth ventricle–subarachnoid shunt. At her third PFD at the age of 7 months, a large fat graft had been placed over the dural closure to prevent scarring. She subsequently had a ventriculoperitoneal shunt placed and underwent 32 shunt revisions for headache before she first presented to us in early 2006 with severe occipital headaches, quadriparesis, ataxia, and an old tracheostomy. She had no ventriculomegaly. Physical examination findings were as follows: gross hyperreflexia, clonus, unsteady gait, and generalized weakness. Neuroophthalmological findings of upgaze paresis, diplopia, convergence–retraction nystagmus, and lid retraction localized the anomaly to the dorsal midbrain.

Neuroimaging. Sagittal MR imaging and CT scanning as well as 3D CT reconstruction revealed an abnormal clivus–odontoid C-1 arch relationship, as follows: angulation of the clivus–odontoid articulation of \( \sim 95^\circ \) in flexion and \( \sim 120^\circ \) in extension, with indentation into the ventral medulla. The dorsal fat graft and a displaced fourth ventricle shunt catheter were visualized. Axial CT and MR imaging studies at the C-2 vertebral body level demonstrated the cervical muscles separated by fat (Fig. 4).

Operation. The patient underwent transoral–transpalato- pharyngeal resection of the odontoid process and lower clivus. The PFD procedure was followed by intradural exploration of the fourth ventricle and syrinx, with lysis of adhesions. Gross occipitocervical instability was present, and we found intradural scarring. The Oc–C2 fusion was performed with titanium loop and cable instrumentation, calvarial bone grafts, and BMP.
Postoperative Course. The patient attained complete neurological recovery within 3 weeks after the dorsal procedure and fixation. At her 1-year follow-up evaluation she remained symptom free. A follow-up postfusion MR image demonstrates marked reduction in the syrinx commensurate with symptomatic improvement (Fig. 4F).

Discussion
Osseous anomalies of the CVJ are believed to arise during the origination of the occipital bone and vertebrae from sclerotomes in the mesoderm and ensuing complex differentiation processes that occur from Week 4 to Week 12 of gestation.\textsuperscript{22–24} The CM-I is one such complication, but sometimes these bone anomalies may be superimposed,\textsuperscript{5,14} reducing the volume of the posterior fossa. Syringomyelia may develop when the CSF is also obstructed at the foramen magnum.\textsuperscript{26} The presence of such concurrent abnormalities at the CCJ makes the site subject to instability, both anteroposteriorly and on horizontal rotation of the head, which further complicates matters, especially when treatment plans must be formulated.

The simultaneous presence of CM-I and syringohydromyelia has long been recognized, but the literature has varying opinions on the appropriate treatment to be provided.\textsuperscript{1,3,4,7–11,20,22,27} The PFD procedure alone, which is the standard treatment for CM-I, is considered inadequate in a small subgroup in which the risk of postoperative instability at the CCJ requires stabilization by a fixation procedure.\textsuperscript{2,6,8,11,12,15} However, few large-scale studies have reported in detail the indications for treating such combined anomalies of the CCJ, and these entities are often the topic of isolated, smaller studies. Furthermore, the presence of concomitant CCJ bone anomalies with CM-I and/or syringohydromyelia is the subject of only a few case reports, with different treatment plans and choices for dorsal fusion.\textsuperscript{13,14}

The incidence of CM-I with concurrent bone anomalies is reported to be rare. In a series of 364 patients with symptomatic CM-I, Milhorat et al.\textsuperscript{22} reported basilar invagination in 12\%, atlas assimilation (together with condylar hypoplasia and other occipital bone atypias) in 5.2\%, and the Klippel–Feil anomaly in 4.9\%, but reported that none of these were superimposed. The incidence of these concur-
rent anomalies is far below the present series' reported instances of basilar invagination (31%), atlas assimilation (24%), and Klippel–Feil anomaly (24%). Such disparities are most probably a direct result of the fact that we focused only on those patients with CM-I and/or syringohydromyelia who required dorsal CCJ fusions, the need for fusion being largely attributed to the inherent instability or concomitant bone abnormalities causing ventral compression. Also, the referral patterns to our hospital skewed the population of patients with CM-I and/or syringohydromyelia who undergo evaluation, because the 234 patients in this series requiring dorsal fusion comprise ~20% of all patients with CM-I who were treated. Thus, a comparison to other studies of patients with CM-I is less meaningful.

In their study of 130 pediatric and young adult patients with CM-I, Tubbs et al. reported on 7 patients with Klippel–Feil anomaly, and basilar invagination was present in 5 patients; a syrinx was reported in 58%, and preoperative cervical instability was found in 1 patient. Treatment was with the PFD procedure, and 83% of patients attained alleviation of symptoms postoperatively. No patient initially underwent a dorsal fusion; 1 patient with subsequent ventral brainstem compression later underwent a transoral decompression followed by occipitocervical fusion. Other past studies have recognized that concomitant bone abnormalities with CM-I and/or syringohydromyelia may require adjunctive procedures to the dorsal decompression to address the ensuing instability. Goel and colleagues reported the importance of performing a posterior decompression followed by occipitocervical fusion via plate and screw fixation in patients with CM-I and atlantoaxial dislocation. Nishikawa et al. reported on 2 patients with CM-I as well as concomitant basilar invagination and atlas assimilation who required simultaneous PFD and posterior fixation for their fixed anomalies. We have pointed out before that atlas assimilation and segmentation failures of the upper cervical spine may contribute to ligamentous laxity and proliferation of granulation tissue at the CCJ. Such instability may eventually lead to basilar invagination and irreducibility, and thus a stabilization procedure is indicated.

In their study of 40 pediatric and young adult patients with CM-I, Grabb et al. sought to determine the amount of ventral brainstem compression that can be successfully treated with a PFD alone. They reported that 75% of patients had some degree of ventral brainstem compression, with 55% having syringohydromyelia. Four of the patients with severe ventral compression underwent preoperative halo-ring traction in an attempt to reduce the compression; in 3 of these patients a subsequent dorsal CCJ fusion was performed after PFD, whereas 1 patient had a transoral decompression and then fusion. One of the 3 patients receiving a PFD and fusion had residual compression and declined neurologically, however, necessitating a second operation and a ventral decompression.

Such a sequela of clinical worsening because of ventral compression after PFD for CM-I is not unknown; indeed we have reported it at our institution. It is thus imperative to be able to quantify the amount of ventral compression at the time of initial evaluation so as to provide the appropriate initial operative approach, either ventral or dorsal decompression, as well as during long-term follow-up. This can be most effectively performed through comprehensive preoperative neuroimaging studies, including dynamic MR imaging of the patient with the spine in cervical flexion and extension. Our long experience with CCI abnormalities has taught us that, when presented with such combined problems of ventral and dorsal compression, the safest and most effective procedure of choice is to address the vector causing maximal cervicomедullary compression.

The patients presenting with musculoligamentous instability (Group IV) were a relatively small component of the overall patient population studied (14%). However, the source of such instability is important to elucidate so that a course of treatment may be fashioned to address the underlying problem most correctly. Although musculoligamentous laxity at the CCI can be found in patients with congenital syndromes (that is, Ehlers–Danlos and Down syndromes), often muscular weakness is an acquired finding and can possibly be attributed to one of several states: 1) repeated muscle retraction with disturbed neurovascular supply to muscle; 2) fibrotic changes occurring in muscle due to repeated operation; 3) inadequate closure at surgery; and 4) possible loss of innervation of cervical musculature secondary to high cervical cord syrinx. Fibrosis due to repeated operation or poor muscular reapproximation may be addressed at the time of surgery by lysis of adhesions or excellent muscular–fascial closure, respectively, whereas a high cervical syrinx causing denervation atrophy of cervical musculature may ultimately require decompression of the syrinx before musculoligamentous stability returns. These possible causes should be entertained when a patient presents with recurrent symptomatology of brainstem compression despite previous posterior decompression surgery, so that the best treatment may be provided.

Conclusions

In addition to PFD/FMD, dorsal CCI fusions are required in those patients with CM-I and/or syringohydromyelia who have concomitant CCI abnormalities that cause compression of the brainstem and upper cervical spinal cord. Such bone abnormalities causing ventral compression can be further categorized into those that are reducible on cervical extension (Group I) and those that require an additional ventral surgical decompression (Group II). A definite group (CM-I and/or syringohydromyelia) without bone abnormalities exists, requiring dorsal CCI fusion to maintain relative decompression in cervical extension (Groups III and IV). This may be due to muscle weakness secondary to a high syrinx, ligamentous laxity, or from repeated posterior procedures. In any of these cases, it is imperative that the treating physician obtains a comprehensive preoperative imaging survey of the CCI, including dynamic MR cervical extension and flexion images, to be able to formulate the best treatment plan.

References

Fusion of the CCJ for hindbrain herniation and syringohydromyelia


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