Craniovertebral junction abnormalities with hindbrain herniation and syringomyelia: regression of syringomyelia after removal of ventral craniovertebral junction compression

Clinical article

ARNOLD H. MENEZES, M.D.

Department of Neurosurgery, University of Iowa Hospitals and Clinics, Iowa City, Iowa

Object. Hindbrain herniation syndrome, or Chiari malformation Type I (CM-I), occurs frequently with craniovertebral junction (CVJ) abnormalities when there is reduction in the posterior fossa volume. Syringomyelia is often present. Posterior fossa dorsal decompression (PFDD) is typically performed but has adverse results when ventral bone abnormality exists. This paper presents the results of a prospective study on CVJ abnormalities in patients with CM-I and syringomyelia.

Methods. Between 1984 and 2008 (the MR imaging era), 298 patients with CVJ abnormalities and CM-I underwent ventral cervicomedullary decompression. Eighty-four patients had associated syringomyelia (15 with secondary invagination and 69 with primary basilar invagination, os odontoideum, or malunion of fractures). Of these 84 patients with CVJ abnormalities, CM-I, and syringomyelia, 46 had previously undergone PFDD combined with fusion procedures or shunt placements. Of the 84 patients, a cervicothoracic syrinx was observed in 57, thoracic syrinx in 14, and holocord syrinx in 13. Studies included CT, MR imaging, and cine flow studies. All 298 patients who underwent ventral CVJ decompression had irreducible or partially reducible pathology. All 84 with syringomyelia showed brainstem dysfunction, lower cranial nerve symptoms, or myelopathy.

Results. Brainstem signs improved in 66 of the 84 patients, myelopathy improved in 58, and syringomyelia regressed in 64.

Conclusions. Neurological improvement and syringomyelia resolution can occur using only ventral cervicomedullary junction decompression in patients with basilar invagination and basilar impression. This is likely due to the relief of neural encroachment and reestablishment of CSF pathways. (DOI: 10.3171/2011.9.JNS11386)

Key Words • basilar invagination • syringomyelia • skull base • Chiari malformation • brainstem compression • craniovertebral junction

Hindbrain herniation syndrome, often referred to as CM-I, is a disorder that had traditionally been defined as downward herniation of the cerebellar tonsils through the foramen magnum of more than 4–5 mm on sagittal MR imaging.1,10,23,25 This anomaly is associated with syringomyelia in 45%–68% of patients.22 It occurs in conjunction with osseous abnormalities of the CVJ. Changes in CSF dynamics contribute to the symptoms and the clinical syndrome characterized by occipital Valsalva-type headaches, lower cranial nerve abnormalities, and spinal cord myelopathy that may be overshadowed by syringomyelia.9,18,22

Magnetic resonance imaging has revolutionized diagnosis, led to early detection, and provided a greater understanding of the pathology, genesis, and manifestations of CM-I, and has also transformed outcome studies.

In a recent comprehensive review of 364 symptomatic patients with CM-I, Milhorat et al.25 found associated syringomyelia in 65% of cases, scoliosis in 42%, abnormal retroflexed odontoid process in 26%, and basilar invagination in 12%. One of the current concepts regarding patients with CM-I and syringomyelia is that there is a reduction in posterior fossa volume compared with normal controls, with abnormalities such as shallow posterior fossa, infolding of the exoccipital bone, and CVJ abnormality (basilar invagination and secondary basilar impression) that ultimately is associated with syringomyelia.2,9,16,22,27,28

Posterior fossa decompression as treatment for symptomatic CM-I with syringomyelia has shown satisfactory results except in the presence of ventral bone abnormalities.3,4,15,17,18,25,29 In the latter circumstance, adverse outcomes have been noted either immediately or several months to years after operation.1,9 This study is a database analysis of patients with irreducible ventral CVJ abnormalities causing cervicomedullary junction compression. These patients had associated hindbrain herniation and syringomyelia and were treated with anterior cranioverte-
bral decompression. The effects of this decompression on clinical and radiographic outcome are provided, including the effect on the syrinx.

Methods

Study Population

This is an ongoing study of CVJ abnormalities. This database analysis of 5600 patients spans the early MR imaging era (beginning in 1984) until 2008. The studies included dynamic motion (flexion and extension) MR imaging of the CVJ, CT and 3D CT, and studies that included the effects of cervical traction. Seven hundred twenty-six patients underwent ventral CVJ decompression for irreducible pathology. Of these 726 patients, 298 had an associated CM-I. Eighty-four of the 298 patients with ventral irreducible CVJ pathology and CM-I had syringomyelia. The 84 patients ranged in age from 3 to 67 years old (mean age 20 years). All had CVJ abnormality with hindbrain herniation and syringomyelia. A cervicothoracic syrinx was observed in 57, thoracic syrinx in 14, and holocord syrinx in 13.

This group of 84 patients with syringomyelia consisted of 69 individuals who had primary basilar invagination and 15 others who had secondary invagination or basilar impression. This latter condition is due to a bone-softening state such as Paget disease, osteogenesis imperfecta, hyperparathyroidism, and varied syndromic abnormalities. Of these 84 patients with CVJ abnormalities, hindbrain herniation, and syringomyelia, 46 had previously undergone PFDD procedures. Twenty-eight of these 84 patients had an associated dorsal occipitocervical fusion and a fourth ventricle to subarachnoid shunt.

Neurological Symptoms

Neurological symptoms and signs reflected myelopathy (brainstem), brainstem dysfunction, and lower cranial nerve abnormalities. In myelopathy, motor deficits of paraparesis, hemiparesis, monoparesis, and the “central cord syndrome” picture were noted. The sensory deficits reflected loss of position, touch, and pain. Three of 84 patients suffered facial pain and another 6 experienced facial hypalgesia. Urinary frequency and incontinence were present in 14 of the 84 patients.

Brainstem dysfunction manifested with sleep apnea, dysphagia (in 40/84), regurgitation, and aspiration pneumonia (in 6 patients younger than 16 years of age). Downbeat nystagmus was present in 3 patients, and none improved. Cranial nerve abnormality presented as hearing loss in 20 of the 84 patients, most commonly in the patients with Klippel-Feil syndrome. Unilateral or bilateral palatal or pharyngeal dysfunction was present in 44, 16 of whom experienced repeated bouts of pneumonia. Hypoglossal nerve abnormalities presented with slurred speech and difficulty swallowing in 6 of the 84; all 6 had proatlas segmentation failures. Migraine headaches with intermittent loss of visual fields occurred in 6 children.

These symptoms and signs were used as outcome measures, that is, myelopathy, brainstem signs, cranial nerve abnormalities, and vascular migraines. The irreducible nature of the CVJ abnormality was clarified by obtaining MR imaging in the flexed and extended position to check for reduction of the ventral compression and with preoperative cervical traction using a halo crown for a minimum of 3 days. At the end of that time, repeat MR imaging was performed in traction, which documented either reducibility of the CVJ abnormality with relief of neural compression or an irreducible state.

Operative Procedure

All 84 patients underwent halo crown traction and transpalatopharyngeal cervicomedullary decompression. This procedure has been previously described. A posterior fossa bone decompression was made at the level of the foramen magnum followed by an occipitocervical fusion.

Prior to 1992, the ventral and dorsal procedures were performed on separate occasions with a week in between procedures. During this time the patient remained in cervical traction and MR imaging was obtained. After 1992, the transpalatopharyngeal procedure and the dorsal occipitocervical fusion were performed on the same day. Postoperative MR imaging was obtained in the 1st week, 6 months postoperatively, and subsequently at 1 year. Subsequent MR imaging was obtained only if there was no clinical or MR imaging syrinx improvement or change in neurological status. Follow-up examinations ranged from 3 to 22 years.

Results

Surgical Outcomes

The preoperative assessment included myelopathy, brainstem dysfunction, cranial nerve abnormalities, and headache. These conditions were reviewed after surgery to assess outcomes. Sixty-four of the 84 patients improved in all parameters. There was MR imaging improvement in syringomyelia and cervicomedullary junction compression in 64.

Twenty of the 84 patients experienced persistence of syringomyelia despite absence of ventral compression on MR imaging and stable fusion construct. Seven patients were operated on before 1992, and 13 cases underwent operations between 1992 and 2008. All of these 20 patients underwent posterior fossa reexploration and an intradural procedure. Twelve of these patients had undergone an intradural procedure of the posterior fossa before referral. In these 12 patients there was scarring at the foramen of Magendie. In the remaining 8 patients, an arachnoid veil overlay the foramen of Magendie or a tonsil occupied the floor of the fourth ventricle.

Two patients who showed initial clinical and MR imaging improvement experienced recurrent symptoms 2 years later; both had previously undergone intradural procedures and had regrowth of posterior fossa bone.

Complications of Surgery

There were 4 pharyngeal infections in 726 cases, none of which occurred after 1990. This is attributed...
to preoperative attention to nutrition, restoration of dentition, hygiene, nystatin and chlorhexidine (Zila Pharmaceuticals) rinses 3 days preoperatively, and meticulous sharp dissection. There were no infections in the 84 patients, nor were there any deaths or neurological deterioration. The overall complications for the 726 patients are detailed elsewhere, and comprised 3 deaths and 3 neurological worsenings, which improved. The deaths included myocardial infarction in an 80-year-old patient with rheumatoid arthritis 30 days postoperatively, 1 death from a vertebral artery blowout in a previously irradiated chordoma (irradiated before referral), and 1 death from delayed brain abscess 6 months later.

Velopalatine insufficiency was noted in 2 children, both of whom had preoperative glossopharyngeal and vagus nerve dysfunction. The gag reflex recovered but nasal speech and nasal regurgitation required speech therapy for 6 months.

Illustrative Cases

Case 1

This 42-year-old woman presented in 1990 with facial numbness, nasal regurgitation, poor hand coordination, and headaches. Neurodiagnostic imaging identified atlas assimilation, fixed atlantoaxial dislocation, and odontoid invagination into the ventral medulla. A CM-I was noted with cervical syringomyelia (Fig. 1A). She underwent anterior decompression of the cervicomedullary junction. The immediate postoperative MR imaging showed medullary decompression, ascension of the cerebellar tonsils, and resolution of the syrinx (Fig. 1B). She subsequently underwent dorsal occipitocervical fusion. Her preoperative symptoms resolved by the 6-month follow-up evaluation. There had been no recurrences when she was evaluated 4 years later.

Case 2

This 12-year-old boy presented in 1991 with severe headaches and weakness in his arms. His underlying pathology was acroosteolysis. He had previously undergone ventriculoperitoneal shunt placement and a PFDD for basilar impression. Magnetic resonance imaging revealed tonsillar impaction down to the C2–3 interspace with a large syrinx originating just below it (Fig. 2 left). A high transpalatopharyngeal approach was made to resect the midline clivus and the odontoid mass. An immediate postoperative MR image showed reduction in the size of the syrinx (Fig. 2 right). His arm weakness improved significantly, and he subsequently underwent a dorsal occipitocervical fusion. This latter procedure was difficult because of the bone-softening state. Postoperatively the patient’s condition necessitated continued use of a molded occipitocervical Minerva brace.

Case 3

This 8-year-old girl presented in 1998 with difficulty swallowing, nasal regurgitation, poor tongue movement, and moderate weakness in her upper extremities, more distal than proximal. She was believed to have Klippel-Feil syndrome with segmentation failure of C-2 and C-3 vertebral bodies and atlas assimilation. There was a foreshortened clivus and the odontoid invagination indented the ventral midmedulla. A large cervicothoracic junction syrinx was identified with the tonsillar ectopia (Fig. 3A). She underwent ventral decompression of the medulla and a dorsal occipitocervical fusion on the same day. Her syrinx resolved. Twelve years after her operation (Fig. 3B) she has continued to experience complete recovery of her symptoms. An MR image obtained at the 10-year follow-up evaluation shows no syrinx.

Case 4

This 13-year-old girl presented in 2004 with a 1-year history of clawing of her left hand and subsequent weakness of the right hand. She had difficulty swallowing liquids and was a spastic quadriparetic. Neurodiagnostic imaging revealed atlas assimilation with basilar invagination and significant ventral medullary compression. A holocord syrinx was identified (Fig. 4 left). She underwent transpalatopharyngeal decompression of the ventral

---

**Fig. 1.** Case 1. **A:** Composite of midsagittal T1-weighted craniocervical (left) and axial T2-weighted (right) MR images through the plane of the invaginated odontoid process. Note the significant ventral midmedulla compression and the high cervical spinal cord syrinx. Hindbrain herniation is evident. **B:** Postoperative midsagittal T1-weighted MR image. The medulla is decompressed and the cervicomedullary syrinx has resolved.
medulla with a dorsal occipitocervical fusion. Her postoperative MR imaging showed complete resolution of the syrinx (Fig. 4 right). Seven years postoperatively she has continued resolution of the hand weakness, swallowing issues, and strength. She is currently enrolled in nursing school and is active in sports.

Case 5

This 9-year-old girl presented in 2007 with neck pain, headaches, and balance difficulties when walking. She was of small stature and cranial nerve examination revealed diminished gag reflex with hyperreflexia in the lower extremities. She had previously undergone a dorsal occipitocervical fusion for basilar invagination. No syrinx had been identified at the time of the initial operation at age 3 (Fig. 5A). Neurodiagnostic imaging at the evaluation revealed a complete dorsal occipitocervical bone fusion. The odontoid process had invaginated into the

---

**Fig. 2. Case 2.** **Left:** Midsagittal T2-weighted MR image of the CVJ. This patient had significant basilar impression with indentation into the medulla. There is tonsillar impaction through the foramen magnum and a high cervical syrinx. **Right:** Postoperative midsagittal T2-weighted MR image reveals reduction in the size of the cervical syrinx.

**Fig. 3. Case 3.** **A:** Composite of T2-weighted (left) and T1-weighted (right) MR images in the midsagittal plane of the cranio-cervical region. Atlas assimilation with the abnormal ventral CVJ pathology indenting into the high midmedulla, tonsillar ectopia, and a cervicothoracic syrinx are also present. **B:** Composite of midsagittal T2-weighted (left) and T1-weighted (right) MR images after transpalatopharyngeal decompression of the ventral medulla. The syrinx has reduced significantly and the cerebellar tonsils have ascended.
medulla and was encircled by the vertebrobasilar arterial tree (Fig. 5B). Syringomyelia was detected. The bone fusion was complete in a posterior crescentic manner (Fig. 5C). Hence, an anterior transpalatopharyngeal approach was used with decompression of the medulla. Postoperative MR imaging a day after surgery showed reduction in the syrinx and significant neurological improvement (Fig. 5D). She was last evaluated 3 years after surgery and her swallowing function, balance, and gait had returned to normal.

Case 6

This 13-year-old girl presented with lisping speech, difficulty swallowing, and severe headaches. She was referred because of her hindbrain herniation. Diagnostic imaging showed a proatlas segmentation abnormality with atlas assimilation (Fig. 6A). Continuation of the clivus was observed into the ventral foramen magnum. This indented into the pontomedullary junction and the midmedulla (Fig. 6B). The posterior fossa volume was small. The tonsillar descent was to the level of the inferior body of C-2. A cervicothoracic syrinx was evident. The patient underwent a transpalatopharyngeal decompression of the ventral cervicomедullary junction and a dorsal occipitocervical fusion. Her postoperative MR imaging shows decompression of the medulla as well as resolution of the cervicothoracic syrinx (Fig. 6C). Her neurological examination results are now normal, and it has been 7 years since her operative procedure.

Discussion

Craniovertebral junction bone abnormalities are being recognized much more frequently now than in the past 30 years due to awareness and easy definition with neurodiagnostic imaging modalities. Patients with symptomatic CVJ abnormalities in this study underwent routine MR imaging of the brain and cervical spinal cord, dynamic flexion/extension MR imaging in the midsagittal plane to document instability, CT and 3D CT to define the bone abnormality, and traction studies to document a reducible or irreducible state.20,24

Patients with irreducible ventral bone abnormalities compressing the medulla and cervicomедullary junction underwent ventral decompression via a transoral or a transpalatopharyngeal route and a dorsal occipitocervical fusion. In reducible craniovertebral junction abnormalities, dorsal decompression was performed as needed (foramen magnum-posterior atlas arch) and a limited occipitocervical fusion (occupit to C-1 to C-2). The presence of syringomyelia and hindbrain herniation did not change this decision. An associated CM-I was observed in 298 of the 726 patients who underwent a ventral bone decompression. Eighty-four patients had an associated syrinx and hindbrain herniation had significant abnormalities. These conditions resulted in ventral medulla compression. The cerebellar tonsils extend to the midportion of the posterior arch of the axis. A cervicothoracic syrinx is evident. Hence, an anterior transpalatopharyngeal approach was used with decompression of the medulla. Postoperative MR imaging showed reduction in the syrinx and significant neurological improvement (Fig. 5D). She was last evaluated 3 years after surgery and her swallowing function, balance, and gait had returned to normal.

Case 6

This 13-year-old girl presented with lisping speech, difficulty swallowing, and severe headaches. She was referred because of her hindbrain herniation. Diagnostic imaging showed a proatlas segmentation abnormality with atlas assimilation (Fig. 6A). Continuation of the clivus was observed into the ventral foramen magnum. This indented into the pontomedullary junction and the midmedulla (Fig. 6B). The posterior fossa volume was small. The tonsillar descent was to the level of the inferior body of C-2. A cervicothoracic syrinx was evident. The patient underwent a transpalatopharyngeal decompression of the ventral cervicomедullary junction and a dorsal occipitocervical fusion. Her postoperative MR imaging shows decompression of the medulla as well as resolution of the cervicothoracic syrinx (Fig. 6C). Her neurological examination results are now normal, and it has been 7 years since her operative procedure.

Discussion

Craniovertebral junction bone abnormalities are being recognized much more frequently now than in the past 30 years due to awareness and easy definition with neurodiagnostic imaging modalities. Patients with symptomatic CVJ abnormalities in this study underwent routine MR imaging of the brain and cervical spinal cord, dynamic flexion/extension MR imaging in the midsagittal plane to document instability, CT and 3D CT to define the bone abnormality, and traction studies to document a reducible or irreducible state.20,24

Patients with irreducible ventral bone abnormalities compressing the medulla and cervicomедullary junction underwent ventral decompression via a transoral or a transpalatopharyngeal route and a dorsal occipitocervical fusion. In reducible craniovertebral junction abnormalities, dorsal decompression was performed as needed (foramen magnum-posterior atlas arch) and a limited occipitocervical fusion (occupit to C-1 to C-2). The presence of syringomyelia and hindbrain herniation did not change this decision. An associated CM-I was observed in 298 of the 726 patients who underwent a ventral bone decompression. Eighty-four patients had an associated syrinx and hindbrain herniation had significant abnormalities. These conditions resulted in ventral medulla compression. The cerebellar tonsils extend to the midportion of the posterior arch of the axis. A cervicothoracic syrinx is evident. Hence, an anterior transpalatopharyngeal approach was used with decompression of the medulla. Postoperative MR imaging showed reduction in the syrinx and significant neurological improvement (Fig. 5D). She was last evaluated 3 years after surgery and her swallowing function, balance, and gait had returned to normal.

Case 6

This 13-year-old girl presented with lisping speech, difficulty swallowing, and severe headaches. She was referred because of her hindbrain herniation. Diagnostic imaging showed a proatlas segmentation abnormality with atlas assimilation (Fig. 6A). Continuation of the clivus was observed into the ventral foramen magnum. This indented into the pontomedullary junction and the midmedulla (Fig. 6B). The posterior fossa volume was small. The tonsillar descent was to the level of the inferior body of C-2. A cervicothoracic syrinx was evident. The patient underwent a transpalatopharyngeal decompression of the ventral cervicomедullary junction and a dorsal occipitocervical fusion. Her postoperative MR imaging shows decompression of the medulla as well as resolution of the cervicothoracic syrinx (Fig. 6C). Her neurological examination results are now normal, and it has been 7 years since her operative procedure.

Discussion

Craniovertebral junction bone abnormalities are being recognized much more frequently now than in the past 30 years due to awareness and easy definition with neurodiagnostic imaging modalities. Patients with symptomatic CVJ abnormalities in this study underwent routine MR imaging of the brain and cervical spinal cord, dynamic flexion/extension MR imaging in the midsagittal plane to document instability, CT and 3D CT to define the bone abnormality, and traction studies to document a reducible or irreducible state.20,24

Patients with irreducible ventral bone abnormalities compressing the medulla and cervicomедullary junction underwent ventral decompression via a transoral or a transpalatopharyngeal route and a dorsal occipitocervical fusion. In reducible craniovertebral junction abnormalities, dorsal decompression was performed as needed (foramen magnum-posterior atlas arch) and a limited occipitocervical fusion (occupit to C-1 to C-2). The presence of syringomyelia and hindbrain herniation did not change this decision. An associated CM-I was observed in 298 of the 726 patients who underwent a ventral bone decompression. Eighty-four patients had an associated syrinx and hindbrain herniation had significant abnormalities. These conditions resulted in ventral medulla compression. The cerebellar tonsils extend to the midportion of the posterior arch of the axis. A cervicothoracic syrinx is evident. Hence, an anterior transpalatopharyngeal approach was used with decompression of the medulla. Postoperative MR imaging showed reduction in the syrinx and significant neurological improvement (Fig. 5D). She was last evaluated 3 years after surgery and her swallowing function, balance, and gait had returned to normal.

Case 6

This 13-year-old girl presented with lisping speech, difficulty swallowing, and severe headaches. She was referred because of her hindbrain herniation. Diagnostic imaging showed a proatlas segmentation abnormality with atlas assimilation (Fig. 6A). Continuation of the clivus was observed into the ventral foramen magnum. This indented into the pontomedullary junction and the midmedulla (Fig. 6B). The posterior fossa volume was small. The tonsillar descent was to the level of the inferior body of C-2. A cervicothoracic syrinx was evident. The patient underwent a transpalatopharyngeal decompression of the ventral cervicomедullary junction and a dorsal occipitocervical fusion. Her postoperative MR imaging shows decompression of the medulla as well as resolution of the cervicothoracic syrinx (Fig. 6C). Her neurological examination results are now normal, and it has been 7 years since her operative procedure.
sociation and syringomyelia. This is observed in patients with osteogenesis imperfecta who develop a progressive reduction in the size of the posterior fossa volume due to skull base invagination, leading to secondary invagination and hindbrain abnormalities with the later development of syringomyelia.

An important question that arises is the cause of deterioration in patients with ventral CVJ abnormalities who undergo a primary PFDD. It is possible that the early deterioration is due to angulation that takes place at the cervicomedullary junction onto the offending ventral pathology during the operation in a prone position, and the presence of instability that may not be detected. Delayed worsening may be due to a gradual appearance of instability and cranial settling. This latter condition is caused by the cantilever effect of the cranium leading to further instability over a period of time. The possibility of cerebellar reimpaction despite the PFDD further aggravates the ventral angulation that may have occurred.

The issue that is paramount to this presentation is the significance of the syrinx disappearing after ventral CVJ decompression in the face of hindbrain herniation (Fig. 7). It is more than likely that this results from removal of the CSF block at the level of the foramen magnum and also from restoration of the posterior fossa volume. It is possible that there is a reversal of the CSF craniospinal pressure dissociation. Postoperative cine studies of CSF flow have not been convincing in proving a reversal of this latter phenomenon. However, a combination of events most likely takes place to provide for the significant clinical as well as radiographic improvement.

In this series, prior to 1992, the ventral decompression procedure was followed a week later by the dorsal fusion procedure. During this time interval postoperative MR imaging was performed and showed early improvement in the syrinx. Thus, one cannot attribute reduction in the syrinx to a posterior procedure as was performed after 1992, when the ventral decompression and dorsal occipitocervical arthrodesis were performed on the same day.

Fig. 5. Case 5. A: Midsagittal T1-weighted MR image of the brain and cervical spinal cord at age 3 years showing odontoid basilar invagination into the midmedulla oblongata, and segmentation failure of C-2 and C-3 vertebral bodies without syringomyelia. B: Midsagittal T1-weighted MR image of the brain and cervical spinal cord obtained in the patient at 9 years old. Note the severe odontoid invagination into the midmedulla. Holocord syringomyelia is present. The patient underwent a bone dorsal occipitocervical fusion at age 3. C: Midline sagittal reconstructed craniocervical CT scan (corresponding to B). Note the dorsal occipitocervical fusion to the C-3 level. The bone invagination into the medulla has an acute clivus-odontoid angle. D: Midsagittal T2-weighted MR image of brain and cervical spinal cord obtained a day after the ventral transpalatopharyngeal resection of the odontoid process. The medullary compression is significantly reduced and there is CSF present dorsal to the cervicomedullary junction. The syrinx has already deflated.
Case 5 developed syringomyelia over several years with documented progressive decrease in the posterior fossa size due to the forward bending of the skull and upward invagination of the odontoid process and anterior skull base. This is observed in children where the downward growth of the clivus proceeds until 16–18 years of age. This phenomenon has been observed in several other pediatric patients with similar underlying pathology and operations.6,20 The syrinx was deflated on MR imaging obtained 24 hours after ventral decompression and did not return on the last follow-up evaluation 3 years later.

In this series, 20 patients with syringomyelia and CVJ abnormalities did not experience reversal of the syrinx after ventral decompression (64 of 84 reversed). It became evident on reexploration of the posterior fossa with an intradural procedure that a cerebellar tonsil occupied the floor of the fourth ventricle, or there was scarring from arachnoiditis from previous intradural surgery or the formation of new bone. Eleven percent of patients in our CM-I series22 and also that of Tubbs et al.34 showed a veil over the foramen of Magendie. Thus, all patients who have had previous posterior fossa intradural procedures should be carefully studied prior to performing the dorsal fixation. The predictors of failure of the ventral craniovertebral decompression procedure are previous intradural posterior fossa dissection and presence of the cerebellar tonsil occupying the floor of the fourth ventricle on preoperative imaging.22,34

In a recent clinical study Aghakhani et al.1 analyzed 157 cases of Chiari-related syringomyelia in adults. All patients underwent a posterior fossa decompression with intradural dissection and a duraplasty. Eleven patients (7%) had basilar invagination or basilar impression. Poor results were noted in this group of patients with basilar invagination regarding the syrinx and CM-I. In 1990 Kohno et al.13 presented a case of successful treatment of “adult Arnold-Chiari malformation with associated basilar impression and syringomyelia” using the anterior transoral route. This patient underwent an anterior fusion together with the transoral decompression. Postoperative MR imaging showed that the cerebellar tonsils had ascended and the syringomyelia had regressed; no dorsal decompression was conducted.

In the author’s review of large series of patients with CVJ abnormalities who underwent transoral decompression of the cervicomedullary junction, the following studies were examined. Kale et al.12 from the All India Institute of Medical Sciences described a 1-stage surgery combining anterior and posterior procedures for CVJ abnormalities. Four hundred ninety-nine patients underwent this procedure. The authors excluded patients with CM-I and syringomyelia from the study. Goel et al.7 presented 190 surgically treated patients with basilar invagination. They were divided into 88 patients in Group 1 with no CM-I and 102 patients in Group 2 that had a CM-I. Of this latter group, 51 had a syrinx and 81 underwent a posterior fossa decompression. A transoral procedure was performed in 11 of these individuals with a syrinx and CM-I. There was no focus on the result of the syrinx. In a subsequent publication, Goel and Sharma8 presented CVJ “realignment for the treatment of basilar invagin-
tion with syringomyelia.” However, there was no postoperative MR imaging reported because stainless steel was used in the fusion construct. Thus, the absence of syrinx outcome leaves the reader uncertain. Mummaneni and Haid26 reviewed their experience with 70 patients who underwent transoral odontoidectomy; there was no mention of hindbrain herniation or a syrinx and its resolution. Recently, Perrini et al.30 discussed the transoral approach and its superior extension to craniocervical junction malformations. Thirty-four patients underwent a transoral procedure and fusion. Thirteen of these individuals had a CM-I but no mention is made of the syrinx. In a more recent publication, Jian et al.11 discussed their treatment of posterior reduction and fixation for the management of basilar invagination with atlantoaxial dislocation. These patients had a reducible abnormality. Of the 29 individuals, 7 had syringomyelia that showed improvement in postoperative studies. This improvement is likely due to restoration of CSF dynamics and improved posterior fossa volume, and provides evidence for the possibility of reducible abnormality.

Conclusions

Patients with symptomatic CVJ abnormalities have a 33%–38% incidence of hindbrain abnormalities. A syrinx is associated in half of these individuals. A ventral decompression of the cervicomedullary junction in irreducible pathology has been shown to allow for regression of the syrinx and patient improvement.

Disclosure

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

References

10. Haroun RI, Guarneri M, Meadow JJ, Kraut M, Carson BS: Current opinions for the treatment of syringomyelia and chiari malformations: survey of the Pediatric Section of the
Syrinx regression after transoral surgery


Manuscript submitted March 4, 2011. Accepted September 27, 2011. Please include this information when citing this paper: published online November 18, 2011; DOI: 10.3171/2011.9.JNS11386.
Address correspondence to: Arnold H. Menezes, M.D., Department of Neurosurgery, University of Iowa Hospitals and Clinics, 200 Hawkins Drive, 1824 JPP, Iowa City, Iowa 52242. email: arnold-menezes@uiowa.edu.