This article contains **a** video clip that can be viewed with QuickTime Player. Please click here to download QuickTime software.

Toward a rational treatment of Chiari I malformation and syringomyelia

RICHARD G. ELLENBOGEN, M.D., ROCCO A. ARMONDA, M.D., DENNIS W. W. SHAW, M.D., AND H. RICHARD WINN, M.D.

Departments of Neurological Surgery and Radiology, University of Washington School of Medicine, Seattle, Washington; and Department of Surgery, Walter Reed Army Medical Center and The Uniformed Services University of the Health Sciences, Bethesda, Maryland

In patients with Chiari I malformation with and without associated syringomyelia, aberrant cerebrospinal fluid (CSF) dynamics and a spectrum of posterior fossa pathological findings are demonstrated. In this study, the authors test the validity of using prospective cardiac-gated phase-contrast cine-mode magnetic resonance (MR) imaging to define the malformation, delineate its pathophysiology, and assist in implementing a rational treatment plan.

Eighty-five cases were prospectively analyzed using cine MR imaging. Sixty-five patients, adults and children, with symptomatic Chiari malformation, with and without syringomyelia, were surgically treated from 1990 to 1999. All patients underwent pre- and postoperative cine MR evaluation. Ten patients were treated after a previous surgical procedure had failed. To establish CSF flow characteristics and normative CSF profiles, 20 healthy volunteers were examined.

Compared with normal volunteers, in Chiari I malformation patients with and without syringomyelia, uniformly abnormal craniocervical junction CSF flow profiles were revealed. After intradural exploration, nearly all patients with Chiari I malformation experienced clinical improvement and CSF flow profiles, paralleling those of normal volunteers, were shown. In all patients in whom treatment had failed, abnormal preoperative CSF flow profiles, which correlated with suspected physiological abnormalities and the pathological findings noted at reoperation, were demonstrated.

Symptomatic Chiari I malformation is a dynamic process characterized by the impaction of the hindbrain in an abnormal posterior fossa. This compression obstructs the normal venting of CSF in and out of the craniocervical subarachnoid space, throughout the cardiac cycle. Therefore, decompression or enlargement of the posterior fossa to establish normal CSF pathways should be the primary goal of surgical intervention. Aberrant CSF flow appears to be only one aspect of the pathological condition found in patients with Chiari I malformation. Arachnoid scarring in the posterior fossa and selective vulnerability of the spinal cord may also be factors in the pathogenesis and maintenance of associated syringomyelia. Phase-contrast cine MR imaging is a useful tool in defining physiological and anatomical problems in patients with Chiari I and syringomyelia, and it can help guide an appropriate primary or salvage surgical therapy.

KEY WORDS • cerebrospinal fluid • Chiari I malformation • syringomyelia • cardiac-gated phase-contrast cine magnetic resonance imaging • arachnoid scar

Chiari I malformation, as defined by tonsillar ectopia located below the foramen magnum, is being increasingly identified as a result of advances in MR imaging.[3] Consequently, more patients with poorly defined complaints are seeking treatment based on evaluation of an MR image study. The malformation appears to be complex in its presentation, cause, and natural history.[24] Furthermore, there is a wide spectrum of surgical treatments for Chiari I malformation and syringomyelia. These factors further complicate our ability to identify appropriate patients who will benefit from surgical treatment. Currently there are over 20 different practiced operations directed at improving the aberrant CSF dynamics thought to be a major coponent of this malformation.[1,6,10,12–16,18,20,23,24,28,30,32,33,40] The options include, but are not limited to, posterior fossa decompressive surgery with obex plugging, extradural decompressive surgery, syringosubarachnoid shunt placement, and reconstruction of a larger posterior fossa.

All the procedures designed to correct the suspected underlying physiological abnormalities are associated with varying degrees of success.[1,4,9,10,15,18,23,33,34] Whereas treatment failures are noted with all the operative approaches, even those that are technically well performed, success in a specific surgical therapy neither completely validates the theory it is based on nor proves it is the best or only form of treatment.

Analysis of the accumulated evidence indicates that the pathogenesis of the Chiari I malformation and associated syringomyelia may be not only complex but also multifactorial in origin.[4,24,26,40] The neurosurgeon will likely benefit from being able to used a validated tool that will help in the characterization of the Chiari I malformation and syringomyelia, in general and in each specific patient. The aim of our study was to evaluate the efficacy of one of our tools, prospective cardiac-gated phase-contrast cine MR imaging, in guiding surgical therapy and evaluating whether the treatment of the pathological and physiological features of this malformation were effective.

CLINICAL MATERIAL AND METHODS

Patient Population

Sixty-five patients and 20 healthy volunteers underwent prospective analysis using cine MR imaging. In the 65 patients with symptomatic Chiari I malformation, syringomyelia was present in some and absent in others. These 65 consecutive patients underwent surgical treatment at Walter Reed Army Medical Center or The Children's Hospital and Regional Medical Center/University of Washington School of Medicine between 1990 and 1999. Data obtained on the characteristics and the presentation of the patients are listed in Table 1. The data obtained in patients and normal volunteers were acquired prospectively and maintained in a database computer program. Of the 65 Chiari malformation patients, surgical treatments performed at an outside institution had failed in 10 patients who were referred to us for further treatment. All patients underwent a thorough examination, and baseline MR images were obtained prior to undergoing surgical evaluation. The position of the cerebellar tonsils was used

TABLE 1
Patient characteristics and presenting signs and symptoms

Characteristic/Variable	No. of Patients (%)*	
	Pediatric	Adult
sex (F:M)	23:13	16:13
mean age (yrs)	10.6	34.5
mean duration of symptoms (yrs)	2.5	4.1
precipitating event (trauma, infection, other)	9/25	4/14
mean follow up (yrs)†	6.2	4.1
headache	27 (75)	29 (100)
syringomyelia	29 (81)	22 (76)
scoliosis	19 (53)	1 (3)
weakness	17 (47)	8 (28)
sensory loss	20 (56)	13 (45)
pain	8 (22)	14 (48)
reflex change	19 (53)	7 (24)
ataxia	9 (25)	6 (21)
dizziness/disequilibrium	11 (31)	9 (31)
apnea	3 (8)	1 (3)
ocular findings	11 (31)	4 (14)
bulbar findings	8 (22)	3 (10)
hydrocephalus	4 (11)	1 (3)
basilar invagination	5 (14)	4 (14)

* There were 36 pediatric patients (age range 1.5–18 years) and 29 adult patients (age range 18–65 years).
† Value indicates the number of years since surgery.

as a general screening parameter. Asymptomatic patients in whom the cerebellar tonsils were below the opisthion were not offered surgery. Two patients with syringomyelia and tonsillar position less than 5 mm below the opisthion received surgical treatment. In the remaining surgical candidates the tonsils had descended 3 mm or more below the opisthion. Patients with Chiari malformation underwent surgery if any of the following findings were demonstrated: 1) a lifestyle-limiting Chiari malformation–related headache (one made worse on Valsalva maneuver), 2) neurological deficit attributed to their syringomyelia, or 3) progressive signs such as scoliosis, ataxia, or dysthesthetic pain. No patient, in whom the aforementioned findings were demonstrated and who sought treatment, was deferred. Surgery was not performed in patients with symptoms confined to chronic fatigue syndrome.

Thirty-six of the Chiari I cases were pediatric patients (<18 years of age), and 29 were adult patients. In the majority of the patients (80.5% of pediatric and 76% of adult) syringomyelia was associated with their Chiari I malformation. A thorough history was obtained; neurological exam and cine MR study of the CCJ were performed preoperatively, 1 to 6 months postoperatively, and each subsequent year. Each cine loop was carefully reviewed by the treating neurosurgeon (R.G.E.) and a neuroradiologist. The mean follow-up period was 5.8 years, and no case was included in the study until the patient underwent a 1-year follow-up evaluation. The same surgeon performed the surgical treatment and uniform surgical treatment, was undertaken. The precise pathological findings observed at each operation were carefully recorded.

Volunteer Population

Twenty volunteers (eight children and 12 adults) underwent phase-control cine MR imaging to obtain baseline normative CSF profiles. Informed consent was obtained from all patients for neuroimaging studies and surgical treatment, as well as from all volunteers for neuroimaging and follow-up studies. The children ranged in age from 15 months to 15 years (mean 7.9 years) and the adults from 20 to 55 years (mean 32.5 years).

Neuroimaging Studies

A midsagittal cardiac-gated phase-contrast cine MR image of the CCJ was obtained using a 1.5 tesla Picker International (Helsinki, Finland) or a 1.5 tesla GE Medical Systems (Milwaukee, WI) scanner with a CSFflow quantitative software package. Prospective data commenced with the cardiac R wave and continued until the terminal 15 to 30% of the cardiac cycle; acquisition then ceased until the sequence was triggered by the next R wave. Approximately 190 to 200 cardiac cycles were sampled for a total of 3 to 10 minutes in each patient. A reference sequence was subtracted from a flow-sensitized sequence to construct each velocity-encoded image. In accordance with the manufacturer's specifications, scan parameters included a repetition time of 30 to 900 msec (depending on the patient's heart rate); an echo time of 18 msec; a field of view of 25 cm; a matrix of 192 × 256; a section thickness of 3 to 6 mm; an ROI of 0.04 to 0.06 cm; and a flip angle of 15 to 30°.

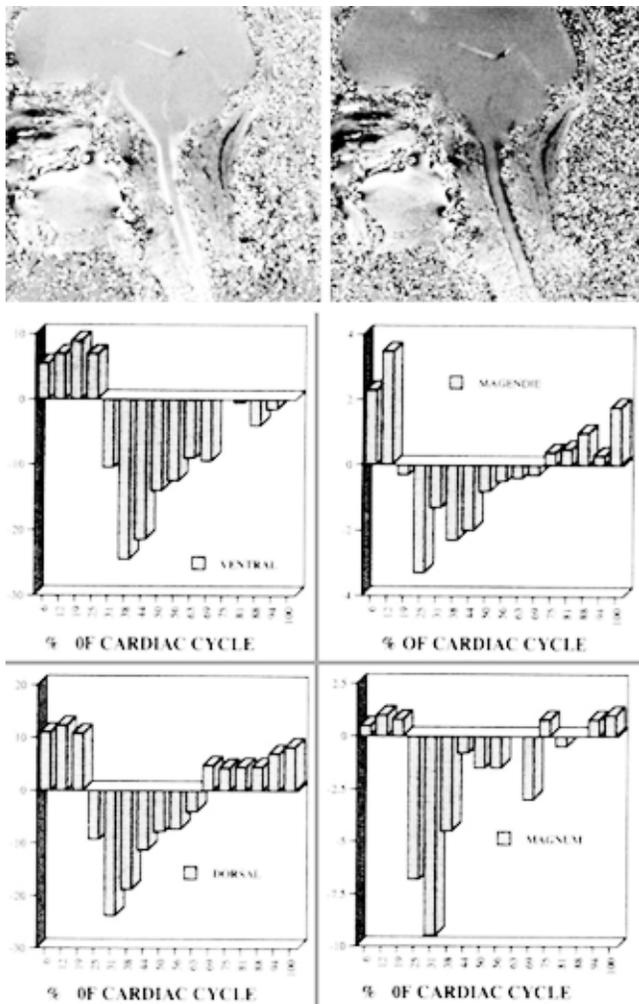The velocity-encoded images were then arranged in a cine loop. By convention, cranial fluid flow was designat-

Fig. 1. *Upper Left:* Cine MR image obtained in a healthy volunteer during midsystole, demonstrating normal caudal CSF flow as hyperintense signal in the craniocervical junction. *Upper Right:* Cine MR image showing normal diastolic cranial CSF flow as hypointense signal. *Lower:* Graphs depicting CSF velocity (in mm/second) as compared with percentage of the cardiac cycle for a normal/control patient, in all four ROIs. Cranial velocities are expressed as positive values, and caudal CSF velocities are expressed as negative values.

ed hypointense and caudal flow was designated hyperintense (Fig. 1 upper left and right). Flow velocity measurements in 55 patients (in mm/second) were obtained in four ROIs: the foramen magnum, the Magendie's foramen, and the subarachnoid spaces, dorsal and ventral to the spinal cord at the level of the midbody of C-2. The Vistar workstation (Picker Vistar, Ohio) permitted faithful localization of the ROI on all cine frames. Cerebrospinal fluid flow velocity was plotted in relation to the cardiac cycle, producing a flow velocity profile. Cranial and caudal flow velocities were expressed as positive and negative values along the y axis, respectively (Fig. 1 lower). Qualitative data only (that is without flow velocity measurements) was obtained in 10 pediatric patients at Children's Hospital.

The digitized cine-loop demonstrates this tool being applied in patient evaluation.

Video Clips

Clip A. Cine MR image obtained in a healthy volunteer. The hyperintense signal in all clips will represent caudal flow; and hypointense signal, cranial flow. Note the movement of the hindbrain. There is no obstruction to CSF flow by the very brisk but small amplitude movements in the CVJ.

Clip B. An illustrative case of a patient with Chiari I malformation. Note the pistonlike action of the hindbrain occluding the foramen magnum, which obstructs both caudal and cranial CSF flow. In contrast to the finding demonstrated in the healthy patient, the hindbrain is impacted and does not move in the cranial direction to unplug the foramen magnum and permit cranial CSF movement.

Clip C. Cine MR image of the patient represented in Fig. 4. The patient is a 3-year-old child who underwent placement of syringosubarachnoid shunt for a large thoracic syrinx. Six months later, a new high cervical syrinx formed with associated with quadriparesis, as well as apnea. Note the tonsils, which end at the opisthion. Shown here is the rostral flow of CSF in the syrinx with ventral caudal CSF flow in the cervical subarachnoid space. The patient was successfully treated with an intradural posterior fossa exploration and duroplasty.

Clip D. Preoperative and postoperative images providing a comparison of the patient shown in Clip C and Fig.4.

Clip E. A 16-year-old pediatric patient with a holocord syrinx and no posterior CSF flow. After surgical therapy, the patient's syrinx collapsed and the dorsal foramen magnum flow improved with the patulous graft.

Clip F. Axial cine MR image of the patient shown in Clip E, demonstrating a unique perspective of the anatomy.

Clip G. Cine MR image obtained in a 57-year-old man had with ventral and dorsal compression. After undergoing dorsal decompression and fusion, restored dorsal flow and improved ventral flow were demonstrated, in part because the brainstem did not move down any further. The patient's outcome was good.

Clip H. An axial image obtained in the patient shown in clip G, revealing both dorsal and ventral compression.

Clip I. Cine MR image acquired in a female patient whose only symptoms were tonsillar ectopia of 5 mm and headache. Her CSF flow was shown to be relatively normal. The patient received no surgical therapy. She responded well to conservative medical therapy for her headaches. No progression of her disease was demonstrated.

Surgical Technique

All patients underwent a similar procedure. The goals of surgery were threefold: 1) reconstruction of a more capacious posterior fossa, 2) normalization of CSF flow from the fourth ventricle into and out of the craniocervical subarachnoid space, and 3) identification and correction of any chronic pathological entity that would prevent the success of former and latter goals. We implemented an operative procedure that was effective, safe, and simple but flexible enough to address the three aforementioned issues we viewed as important. We were not dogmatic about its application; we merely desired to adopt a uniform approach to test our objective in this prospective study.

The operation consisted of a suboccipital craniectomy; C-1 (and, when necessary, C-2) laminectomy; intradural lysis of arachnoid adhesions between the cerebellar tonsils and floor of the fourth ventricle; shrinkage of the prolapsed cerebellar tonsils with bipolar electrocautery; and duroplasty in which we used lyophilized cadaveric dura, pericranium, or bovine pericardium. The bone removal was sufficient to create an enlarged foramen magnum. Specifically, a moderate posterior fossa decompressive procedure was performed. A 3 by 3–cm craniectomy with

TABLE 2
Pathological findings observed intraoperatively

Pathological Finding	Chiari I and no Syrinx (14 patients)	Chiari I With Syringomyelia (51 patients)
extradural compression*	14 (100)	51 (100)
tonsillar impaction	14 (100)	51 (100)
arachnoid scar, dense†	6 (43)	19 (37)
gliosis‡	1 (7)	5 (10)
vascular distortion§	2 (14)	4 (6)

* Based on MR imaging revealing distortion of cerebellum and presence of extradural bands at foramen magnum and/or C-1.

† Arachnoid scar adherent to impacted tonsils requiring microsurgical technique to remove.

‡ Gliotic tonsil tips; longstanding changes noted.

§ Tonsils displaced inferiorly with loop(s) of PICA(s) embedded in arachnoid membrane or aberrant position from longstanding compression.

laminectomy in a child and a 4 by 5–cm craniectomy with laminectomy in an adult were deemed adequate. Care was taken not to remove too much bone and thus cause cerebellar slump, a rare but difficult to treat complication of an overly aggressive craniectomy.[23]

Thickened, constricting extradural bands were found and always removed in the foramen magnum and/or C-1 lamina. Microscopic dissection was used. Ultrasonography was used to guide the procedure when indicated. Gentle bipolar coagulation of the tonsils permitted increased CSF egress from the fourth ventricle. It was performed on all elongated, irregularly shaped or gliotic tonsils that obstructed CSF flow. No cerebellar tissue was resected. Meticulous technique was used to avoid spilling blood products in the CSF. All patients received perioperative antibiotics and steroid therapy. A direct approach to the syrinx, placement of a fourth ventricular stent, or placement of a muscle plug in the obex was not undertaken in any patient. In all patients undergoing surgery, there was evidence of compression of the CCJ. Pathological findings are summarized in Table 2.

Three patients with associated basilar impression required posterior fusions, and one required a transoral odontoid resection. The 10 patients referred because of persistence of their syrinx, recurrence of their syrinx, or persistence of preoperative symptoms underwent procedures similar to the other 55 patients.

RESULTS

Healthy Volunteers

Eight healthy pediatric and 12 normal adult volunteers were studied using the same cardiac-gated phase-contrast cine MR imaging technique described earlier. The goal was to obtain normative values produced on our specific imaging machines so that a comparison could be made.

The time periods for cranial and caudal CSF flow were comparable in these two groups. The caudal brain motion of the cerebellum was followed by that of the brainstem. Cranial flow velocities were measured in the first 75 to 100 msec of the cardiac cycle. Initial caudal flow velocities were noted at 100 to 115 msec after the R wave, with

dorsal C-2 and foramen magnum caudal CSF flow velocities preceding ventral C-2 caudal velocities. Peak caudal CSF velocities occurred at 130 to 175 msec following caudal movement of the brain in the posterior fossa. A second, smaller caudal CSF velocity peak occurred at 450 to 500 msec, corresponding to the peak of the dicrotic notch. The resumption of cranial flow velocities was also similar in these two control groups. Cranial CSF velocities began at 350 msec after the R wave at dorsal C-2, then within the foramen magnum, and last at 375 msec after the R wave at ventral C-2 and lasting up to 50 msec after the R wave. A major difference between the pediatric and adult groups is the observation of higher velocities in the pediatric patients. Cerebrospinal fluid dynamics have shown age-dependent differences. Differences between healthy pediatric and adult volunteers were observed. In children a higher mean maximum caudal velocity in all four ROIs were demonstrated. These differences were most dramatic in the foramen magnum and Magendie's foramen, where the CSF velocities were at least 50% higher than those measured in adults.[1] Ohara, et al.,[29] using cardiac-gated spin-echo MR imaging, noted increased pulsatile CSF flow in the posterior fossa of children as compared with adults. They attributed this difference to greater intracranial compliance in children.

In summary, there was a pulsatile caudal movement of CSF following the downward movement of the brain in the posterior fossa. This occurred during 50% of the cardiac cycle followed by a shorter rebound of CSF flow from the cervical subarachnoid space into the cranium during diastole. Figure 1 lower provides a graphic interpretation of the CSF flow in a normal subject.

Chiari I Malformation

Sixty-five patients with Chiari I malformation were studied. In patients with herniation of the cerebellar tonsils located more than 3 mm below the opisthion of the foramen magnum, obstructed CSF flow was demonstrated. This was manifest by decreased flow velocities and a shorter period of caudal CSF flow in Magendie's foramen and foramen magnum. In two patients tonsillar ectopia was of 3 mm or less, but both had holocord syringes. Both patients with borderline tonsillar ectopia had significantly abnormal CSF flow profiles. In addition, a progression of their tonsillar descent was observed in two patients while being followed prior to surgical intervention.

Patients with and without syringomyelia had similarly aberrant CSF flow profiles observed at the CCJ. Specifically, the initial 100 msec of the cardiac cycle showed cranial CSF velocities that were diminished in magnitude in the foramen magnum compared with those found in normal volunteers. The most dramatic difference was the duration of caudal CSF flow both dorsal and ventral to the brainstem in the foramen magnum as compared with that in normal subjects. The terminal 50% of the measured cardiac cycle showed dampened CSF velocities and a trend toward preferential cranial flow compared with those in healthy volunteers in whom caudal CSF flow was shown during that period of their cardiac cycle. The period of caudal CSF flow in the foramen magnum was substantially decreased in patients with Chiari I malformation, lasting 20 to 35% of the measured cardiac cycle, with maxi-

Fig. 2. Midsagittal MR image obtained in a pediatric patient with Chiari I malformation and an isolated midthoracic syrinx. The patient's spinal cord ended at T12–L1 and was not tethered.

mal velocities of 1.5 to 4.5 mm/sec. Caudal CSF flow velocity was limited to 15 to 40% of the measured cardiac cycle, with maximal velocities of 1.5 to 10.7 mm/sec.

The cerebellar tonsils were uniformly observed to move downward as they impacted the CCJ during systole. During diastole, the tonsils moved upward; however, the degree of amplitude varied in each patient. As a result, the foramen magnum remained physiologically occluded during most of the cardiac cycle. In those with longstanding symptoms and dense scarring observed intraoperatively, the least upward movement during diastole, most impaction, and poorest dorsal cranial CSF flow were demonstrated. In three children and one adult, with the most exuberant subarachnoid scar observed, only slight upward movement of the hindbrain structures was seen during diastole.

Cerebrospinal fluid flow in the syringes was difficult to assess in patients with a small syrinx. In patients with a holocord syrinx, pulsatile flow of fluid in the syrinx was revealed in both the cranial and caudal direction during the cardiac cycle. It did not appear to be turbulent flow but was most often found to flow in the opposite direction as that in the adjacent subarachnoid space.

Postoperative Care

Postoperative studies in patients managed surgically demonstrated marked changes as compared with preoperative studies. In all patients there was an increase in the absolute value and duration of caudal CSF velocities, which was most notable within the foramen magnum. A less dramatic increase in CSF velocities was observed ventrally at C-2 except in those patients with basilar impression. The two- to threefold increase in CSF velocities dorsal to the spinal cord at C-2 reflects the expanded foramen magnum with less resistance to outflow. An increase in the magnitude of caudal CSF flow velocities was associated with an increased period of caudal CSF flow, a dramatic change from that measured preoperatively. The ven-

TABLE 3
Long-term outcome in 65 patients with Chiari I malformation

Parameter	No. of Patients (%)	
	Pediatric	Adult
good	23 (64)	20 (69)
improved	12 (33)	8 (28)
poor	1 (3)	1 (3)

* Refer to *Long-Term Outcome* section for description of parameters.

tral CSF caudal velocities were markedly improved in the nine patients with basilar impression who underwent surgery via a posterior approach. The only exception was one patient who was referred to us after primary surgery failed to resolve his condition. He required a transoral odontoid resection.

Syrinx size in the cervical and thoracic regions was measured as a percentage of the spinal cord by dividing the greatest axial diameter of the syrinx by the diameter of the spinal cord at the corresponding level. The postoperative size of the syrinx was compared with its preoperative size at the same axial level. The syringes were considered improved if the cavity had been collapsed by 50% or
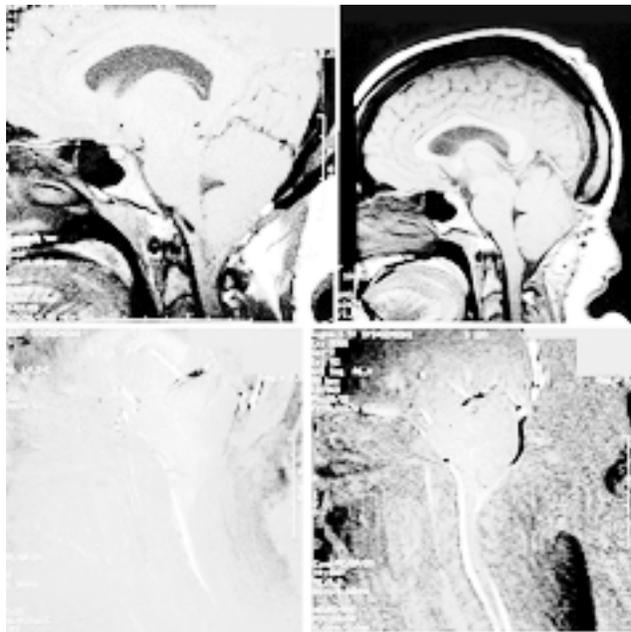

Fig. 3. Imaging studies in an adult patient who underwent previous extradural procedures for treatment of her Chiari I malformation and cervical syrinx. The patient remained symptomatic (with headaches), and the syrinx did not collapse. *Upper Left:* Preoperative MR image. *Upper Right:* Postoperative image demonstrating results after intradural decompression, lysis of significant scar, and duroplasty. The patient remains asymptomatic after collapse of her syrinx. Note the new cisterna magna and pathway established from the fourth ventricle. *Lower Left:* Cine MR image demonstrating results after extradural decompression. Note the lack of dorsal CSF flow during systole. This study directed us to perform an intradural exploration. *Lower Right:* Cine MR image obtained after the intradural procedure. Note the dorsal CSF (caudal) flow now apparent. There was improved ventral flow, as well.
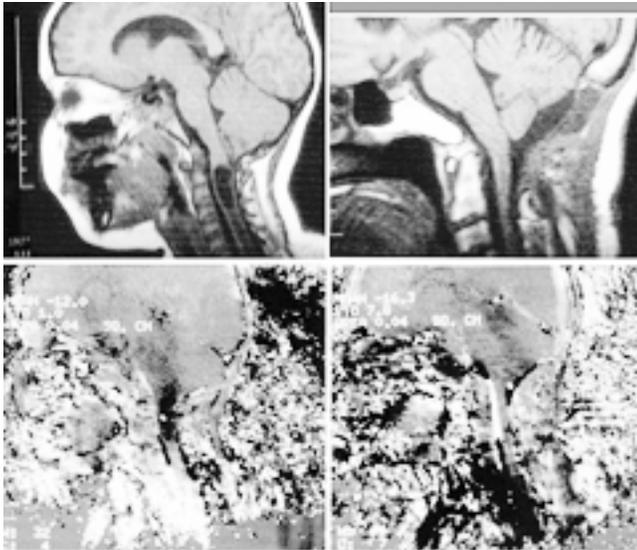
Fig. 4. Imaging studies obtained in a 3-year-old child after placement of syringosubarachnoid shunt for a large thoracic syrinx. The patient was referred to us 6 months later for treatment of a new high cervical syrinx associated with quadriparesis, as well as apnea. *Upper Left:* Preoperative MR image. Note the tonsils that end at the opisthion. *Upper Right:* Postoperative MR image obtained after an intradural exploration. The syrinx has collapsed, the patient could ambulate, and she experienced no further apnea spells. *Lower:* Preoperative and postoperative cine MR images revealing no dorsal CSF flow at the CCJ before surgery (left) and improved flow postsurgery (right). The white dot represents the marker at which site velocities were measured. The direction of flow in the syrinx was opposite that measured in the cervical subarachnoid space before syrinx collapse.

more. In the patients with a collapsed syrinx it was difficult to assess either the direction of flow or velocity of flow within the collapsed cavity. An interesting finding was the presence of a primarily thoracic-level syrinx in three patients without spinal cord tethering (Fig. 2). All three were treated successfully.

Treatment of Patients in Whom Earlier Surgery had Failed

Ten patients referred to us required reoperation due to the failure of earler intervention (Table 3). In all 10 cases cine MR studies revealed persistent aberrant CSF flow profiles and demonstrated at which anatomical location the obstruction was greatest. Four were children and six were adults.

Six patients had a syrinx that either failed to collapse or increased in size after a posterior fossa operation that was purely extradural. Two of the syringes slowly recurred after a year in which improvement had been shown. All six patients had dense intradural adhesions that once lysed permitted improved CSF flow with subsequent collapse of the syrinx and improvement in symptoms (Fig. 3). Two of those patients had significant past history of trauma. In two other patients syringosubarachnoid shunts had been placed, which ultimately resulted in a new syrinx formation and a recurrence of symptoms. One child presented with quadriparesis and apnea caused by the development of a new cervicomedullary syrinx that developed after placement of a syringosubarachnoid shunt caused the cer-

vicothoracic syrinx to collapse. She had borderline tonsillar ectopia. On a cine MR study significant dorsal obstruction to CSF flow was noted. This finding directed us to perform a posterior fossa decompression with intradural lysis of dense arachnoid adhesions and gliotic tonsils. Collapse of her high cord syrinx and resolution of apnea and weakness occurred within 6 weeks of surgery (Fig. 4).

There were two patients with Chiari I malformation and syringomyelia whose initial surgeries failed because more bone needed to be removed. Both patients improved after we performed surgery, in part because the cine MR studies provided guidance. One adult, previously described, required a transoral resection of his odontoid followed by an occipitocervical fusion. Cine MR imaging demonstrated the aberrant ventral flow with downward pulsation with his brainstem onto his odontoid process, which correlated to his clinical findings. He underwent successful surgery, and his symptoms slowly resolved. The second patient was a child who presented with caudal descent of her tonsils to C-2 was demonstrated. Despite undergoing posterior fossa decompression and laminectomy, her tonsils did not change configuration or position. An intradural operation was performed after C-2. Gentle bipolar coagulation of her tonsils created a more rounded shape, which ultimately permitted improved caudal CSF flow.

Clinical Picture

There are several important and interesting observations with regard to the clinical presentation. The first concerns the headaches experienced by patients with Chiari I malformation. One hundred percent of the adults and 75% of the children experienced headache. These are very specific headaches characterized by severe, pounding head pain usually in the vertex or posterior occiput, and they are almost always worsened by bending, straining, or performing any Valsalva-related activity. These headaches are relatively easy to distinguish from other headache syndromes. Approximately 75% of the symptomatic patients were involved in regular sports-related activities that were discontinued because of the exercise-limiting headaches. Two patients were involved in competitive tennis activities at the national level. They were unable to compete because of severe, debilitating headaches experienced during their serve. Both have returned to competitive sports 6 months after surgery.

Two female patients with Chiari I malformation without syringomyelia presented with significant spinal cord dysfunction. Both women had developed a hemiparesis and had obstructed CSF flow at the foramen magnum. In both there was marked compression of the tonsils, and an ectatic PICA was noted at surgery. Both improved within weeks of operation.

Thirty-one percent of the children and adults described an unusual neurootological syndrome consisting of dizziness or disequilibrium. Milhorat and associates[25] describe this finding in detail, in a higher percentage (74%) of their patients. The adults often correlated the symptoms with fatigue, but all had an associated Chiari I malformation. The findings were almost always observed in conjunction with nystagmus, oscillopsia, or simple vertigo. Most patients underwent a thorough otological or ophthalmological exam prior to seeking a neurosurgical opinion. No dis-

TABLE 3
Long-term outcome in 65 patients with Chiari I malformation

Parameter	No. of Patients (%)	
	Pediatric	Adult
good	23 (64)	20 (69)
improved	12 (33)	8 (28)
poor	1 (3)	1 (3)

* Refer to *Long-Term Outcome* section for description of parameters.

crete otological or ocular deficit was ever uncovered. Like trigeminal neuralgia, vascular displacement and compression of a PICA or anterior interior cerebellar antery may contribute to this difficult-to-define symptom in Chiari I malformation.

Long-Term Outcome

The long-term outcome in all 65 patients with Chiari I malformation recorded after their last surgical procedure is summarized in Table 3. The categories have been simplified to take into account the patient's perspective of their outcome in each case. Outcome is grouped into three categories: 1) good, which is defined as an asymptomatic Chiari I malformation patient, a Chiari I malformation patient with a syrinx that has collapsed greater than 75% of its original diameter, or a patient with one small residual component of the disease such as sensory loss or nystagmus; all patients return to normal lifestyles without restriction; 2) improved, which is defined as marked overall symptomatic improvement, syrinx collapse of 75% or greater, and only one major residual deficit such as mild spinal cord–related pain or scoliosis progression; all patients return to a normal lifestyle but individually restrict their sports activities; and 3) poor, which is defined as no change or syrinx enlargement, or the presence of residual pain, especially dysesthetic pain that is recalcitrant to therapy; the patient leads a restricted lifestyle.

A residual syrinx was seen in the cervical spine of two patients, representing the two "poor" outcomes. In one case (an adult patient) a demyelinating disease as well as a Chiari I malformation with syringomyelia were diagnosed. Although the patient's headaches and CSF profile improved postoperatively, the small cervical syrinx had not changed in size and was associated with severe, dysesthetic pain. The other patient, a girl, remained neurologically stable but had a holocord syrinx, which did not changed in size. This patient had the most severe arachnoid adhesions in the series. The initial microsurgical attempt at establishing CSF flow from the floor of the fourth ventricle was not successful. A large aberrant PICA embedded in thick arachnoid adhesion lay over both cerebellar tonsils. Her postoperative cine MR study demonstrated a CSF profile in which there was obstruction to dorsal CSF flow despite the posterior fossa craniectomy, indicating that her scar was a significant physiological issue. This child's parents described and documented a history of severe birth-related trauma with a neonatal subdural and intraventricular hemorrhage. There were no cases in which neurological worsening occurred.

The syrinx collapsed 75 to 90% or more of its original diameter in 31 and approximately 50 to 75% in eight of the 41 patients with syringomyelia. The majority of pediatric and adult patients have experienced long-term sustained improvement of their symptoms. Postoperatively, the Chiari malformation–related headache either resolved or improved such that it was experienced fewer than several times a year. All patients were able to engage in sports-related activities with the exception of three patients: two were body builders and competitive weight lifters and one was a marathon runner.

In nine of the 19 pediatric patients with scoliosis, the deformity was stabilized. In 10 deterioration in alignment has been observed despite marked improvement in all their other symptoms.

Five patients had concomitant hydrocephalus. Two children had hydrocephalus requiring permanent CSF diversion prior to the posterior fossa decompressive procedure. One girl required ICP monitoring for headaches associated with opisthotonic posturing. Preoperatively the ICP monitor demonstrated elevated ICP with plateau waves. The elevated ICP and posturing were completely alleviated after posterior fossa decompressive surgery was performed. One adult with hydrocephalus and syringomyelia underwent a successful endoscopic third ventriculostomy prior to posterior fossa decompressive surgery. One adult required concomitant placement of a temporary external ventricular drainage tube for 1 week. After undergoing posterior fossa decompression, the patient's elevated ICP levels normalized, and the ventriculostomy was removed without the need for placement of a shunt. That patient returned to work as a physician's assistant.

There were no surgery-related deaths in our series, and the morbidity rate was limited to four patients (6.2%) with postoperative pseudomeningoceles with CSF leak who were treated early in the series. The pseudomeningoceles appeared to be the result of CSF leaking through the suture holes in the dural graft material. We switched to using bovine pericardium, and no further complications occurred. Three of the pseudomeningoceles were successfully surgically repaired, and one resolved after tapping and wrapping of the scalp.

DISCUSSION

Data analysis from this study and several previous studies demonstrates that aberrant CSF flow is a major component of the pathological state in Chiari I malformation patients with and without syringomyelia.[1,4,8,13,17,18,24,35,39]

DuBoulay, et al., and Lane and Kritcheff in the 1970's, used invasive techniques such as myelography, ventriculography, and pneumoencephalography to show that CSF flow is more likely to be linked to brain expansion and contraction during the cardiac cycle. Myelography revealed that, in normal patients, caudal CSF pulsations were greatest in systole and cranial pulsations were greatest in diastole.[8,21] These were important observations on the normal movement of CSF. We used these studies as a historical reference with which to compare to our results in our normal volunteers obtained using cine MR imaging. However, invasive techniques, such as those used by DuBoulay, et al., often contaminate the closed system of CSF circulation that they were meant to study and, thus, have been supplanted by modalities involving MR imaging.[11,17,22,31]

Chiari I malformation and syringomyelia have been studied using each new technology to define better the precise dynamics of aberrant CSF movement. Since the report on cardiac-gated MR imaging of CSF flow published in 1985 by Bergstand and colleagues,[5] there has been much excitement about the application of this technique to study the normal and abnormal brain. Cine MR imaging has shown that pulsatile CSF motion in the brain occurs in response to a pulse propagation from cardiac systole.[17] It is this pulsatile velocity of CSF, not the bulk CSF movement (bulk flow), that is measured by cine MR imaging, as used in our study.[7]

The cardiac-gated phase-contrast cine MR technique used in our study has the advantage of being noninvasive, reproducible, and comparable in a longitudinal fashion. It can be used to obtain flow direction and flow velocity values at many sites simultaneously. Furthermore, normative CSF values can be obtained for use in a direct comparison. However, data supporting the validity of its use in directing surgical therapy have only recently been published.[1,18]

We have gained many insights into the normal movement of CSF through the application of cine MR imaging. Arterial outflow and venous inflow are not equal during the cardiac cycle. There appears to be a short period of brain expansion during systole. Because the Monro–Kellie doctrine, as modified by Weed,[39] assumes that the intracranial contents (blood, brain, vessels, and CSF) must remain constant through out the cardiac cycle, the CSF must be vented.[19,25,39] Enzmann[11] has shown that approximately 60% of the subarachnoid CSF in the brain is displaced across the foramen magnum into the cervical subarachnoid space during systole; this flow arises from the expansion of posterior fossa structures. Greitz, et al.,[17] have described the normal brain motion as movement that occurs in a funnel-shaped direction; thus, the resultant pulsatile cervical CSF motion is caused by the brain acting as an expanding and retracting piston. The subarachnoid space around the spinal cord exhibits a recoil reaction during diastole causing cranial CSF flow, due to the elasticity of the spinal dura mater.[1] This venting of the brain and CSF at the CCJ and cervical subarachnoid space is the site at which we noted most of the Chiari I malformation–associated disease.

Morphometric, anatomical studies have demonstrated that the cranial abnormalities in Chiari I malformation cause overcrowding of the cerebellum within a too small posterior fossa.[26,37,38] Milhorat, et al.,[24] have used radiological studies to provide a meticulous description of the abnormal anatomical findings in 364 symptomatic Chiari I patients. Their study and the morphometric study undertaken by Nishikawa, et al.,[27] provide convincing evidence that Chiari I malformation is a disorder of the paraaxial mesoderm that causes the underdevelopment of the posterior fossa cranium as well as compression of the normally developed hindbrain.

We have shown in our study of 65 patients with Chiari I malformation that the downward movement of the tonsils and subsequent obstruction of the foramen magnum during systole and diastole prevent the normal venting of CSF at the CCJ. Thus, a series of related observations in conjunction with our observations is pertinent.

In most patients with Chiari I malformation, the hindbrain sits in a small or abnormally configured posterior fossa cranium.[24,27] Pulsatile downward movement of the cerebellar tonsils into the funnel-shaped foramen magnum occurs after the cardiac pulse propagation (5–6 m/second peak velocity) begins. It is subsequently followed by the caudally directed hindbrain and associated pulsatile CSF flow which occurs 100 to 400 msec later (after the R wave). The subarachnoid space in the spinal canal does not have adequate compliance to accept the large pulses of CSF being vented into it by the impacted cerebellar tonsils. Furthermore, the spinal recoil, which normally produces cranial CSF flow during diastole, does not occur to its fullest extent. We noted that the cerebellar tonsils do not elevate sufficiently to vent the cervical subarachnoid spaces, seen in normal cases. Cerebrospinal fluid is trapped by the impacted tonsils. In this way the tonsils act more like a cork in a bottle than as a piston in an engine.

Over time, the incessant impaction of the tonsils contributes to arachnoid scarring, gliosis, and chronic compression and further displacement of the hindbrain structures. We agree with those who speculate that CSF trapped in the cervical subarachnoid space could act and compress structures within that space, thus propagating syringomyelia.[18] However, current theories cannot satisfactorily explain the existence of an isolated thoracic syrinx, as seen in three of our patients. The trapped CSF pulse pressure should be diminished by the time it reaches the thoracic subarachnoid space and should not act exclusively in the thoracic subarachnoid space. We remain perplexed that in some Chiari I malformation patients without syringomyelia CSF flow profiles are similar to those found in a patients with syringomyelia. It is attractive to speculate that given time all patients with tonsillar impaction and abnormal cervical subarachnoid CSF flow will develop a syrinx. However, we have not found that to be the case in our patient population. Several of our adult patients with known, long-standing symptoms and significant neurological deficits (such as hemiparesis or ataxia) did not develop a syrinx. Some of our patients, including an 18- and a 36-month-old child, developed large syringes and exuberant subarachnoid scarring at a very young age.

In patients with associated syringomyelia, we noted cranial and caudal movement of the CSF within the syrinx. This movement seemed to occur in a direction opposite that of the pulsatile flow in the cervical subarachnoid space. This may explain why syringes can expand in both cranial and caudal directions.

The precise source of the syrinx fluid remains unclear. However, there is mounting evidence that the CSF tracks through the spinal cord from the subarachnoid space. Ball and Dayan[2] were the first to hypothesize that this fluid makes its way from the spinal subarachnoid space through the perivascular channels. Stoodley, et al.,[36] have used Milhorat's animal model to show that a pulse pressure rostral to the subarachnoid block seen in Chiari malformations causes an increase of perivascular flow into the spinal cord and thereby causes a syrinx formation. And finally, Heiss, et al.,[18] have hypothesized that the CSF acts from outside the spinal cord and not from within. Their study, in which they used both cine MR and invasive monitoring procedures, provides evidence that CSF acting from the subarachnoid space causes fluid to flow into the cervical spinal cord. Our findings that CSF flow occurs in both directions in the syrinx but in an opposite direction to

that of the pulsatile flow in the adjacent subarachnoid space provide additional indirect support of this mechanism. The last bit of convincing data to be obtained would be the reproducible visualization on MR imaging of CSF percolating through the perivascular spaces of the human spinal cord propagating the syrinx. Further technological advances will clarify this issue.

There is an increasing number of reports in which the authors advocate a purely extradural approach for the treatment of this disease.[12,18] We agree that in the majority of cases, simply achieving relief of the anatomical and physiological block by an extradural approach is appropriate and sufficient. In the majority of our patients (62%), this most likely would have worked well. However, in 38% of our patients including eight of ten in whom previous surgery had failed, the degree of arachnoid scarring obviated that option. Unfortunately, cine MR imaging did not provide the detail with which to discern which patient would require an intradural lysis of scar and, thus, benefit from an intradural as compared with extradural procedure. Specifically, decompression of an ectatic, displaced, scarred PICA, dissecting free exuberant scar over the fourth ventricle, or making a gliotic scarred cerebellar tonsil assume a more round shape to permit CSF egress may be important in correction of a CSF block. Thus, we are cautious about undertaking an extradural approach without the addition of ultrasonography. Furthermore, long-term follow-up data should be obtained in the initial cohort of patients who undergo on extradural procedure. In addition, before such techniques are applied in a uniform fashion, they must be used in a diversity of patients, especially in those patients whose history points to an etiological factor that may cause arachnoid scarring.

We are not in agreement with the authors who claim that a carefully performed intradural operation creates more scarring and subsequent failure than an extradural procedure. That appears not to be the case in our series or others in which long-term follow-up data have been obtained.[4,9,10,20,40] Clearly, a poorly performed surgical procedure, intradural or extradural, can lead to additional problems, and we emphasize that the surgeons choose not only the approach most appropriate for the patient's suspected etiological or physiological findings but also that with which the patient is most comfortable.

Our craniectomies are appropriately moderate based on our observation that the impaction of the cerebellar tonsils at the foramen magnum is the site most in need of being expanded. By concentrating on creating an enlarged cisterna magnum, the physiological block can be removed. The diminished caudal (systolic) and cranial (diastolic) flow velocities can be reversed in the foramen magnum with moderate bone removal (Fig. 5). Removing bone near the lateral sinus or over the lateral cerebellum may have added very little to the goals of our surgical plan. However, we recognize that creating an enlarged posterior fossa is beneficial and can be addressed in many creative ways. We are circumspect about removing too much bone in a patient with basilar impression, and we have treated one such individual for the consequences of cerebellar sag, as described by Menezes and Batzdorf.[23] We are less enthusiastic about placing a muscle plug in the obex because of the procedure's associated morbidity and because it does not benefit most patients. In the overwhelming
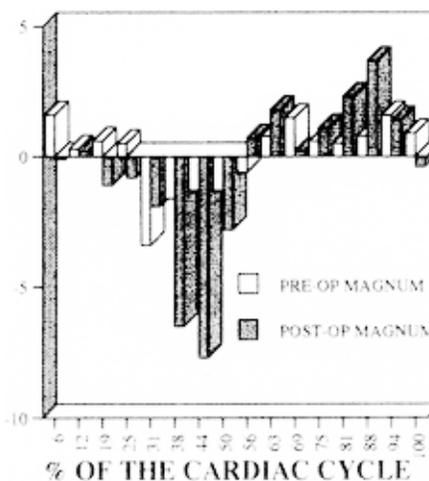

Fig. 5. Graph showing a summary of flow velocities measured pre- and postoperatively in the foramen magnum as compared with the percentage of the cardiac cycle measured. Positive values represent cranial flow, and negative values represents caudal flow. Note the sustained postoperative caudal flow demonstrated during systole and improved cranial flow observed during diastole.

majority of patients with Chiari I malformation, no connection can be visualized between the fourth ventricle and the syrinx. The placement of a shunt to drain the syrinx should be used as a last-resort procedure. Although it is a procedure that works, its longevity is not long lived.[35]

CONCLUSIONS

We present data that support the primary finding in Chiari I to be a pathological and thus physiological block to caudal and cranial CSF flow in the CCJ during systole and diastole, respectively. Whereas tonsillar position provides a general screening parameter, the CSF profile dynamics in conjunction with the clinical examination findings provide an equally useful parameter in the diagnosis of Chiari I malformation. Successful treatment requires reestablishing CSF pathways by a surgical procedure appropriate for the presumed origin and nature of the obstruction. It is reasonable, based on our data, to hypothesize that selective vulnerability of the spinal cord to the following factors also play a significant role in the differential development and propagation of syringomyelia: 1) pulsatile CSF pressure, 2) trauma and scarring, and 3) anatomical distortion.

In patients in whom surgery has successfully resolved symptoms of Chiari malformation CSF profiles will parallel those obtained in normal volunteer. In patients in whom previous surgery failed abnormal CSF profiles will be demonstrated. Cardiac-gated phase-contrast cine MR imaging is a useful tool in defining the pathophysiology in a case in which treatment has previously failed and, thus, in isolating a solution. Phase-contrast cine MR imaging can play an important role in defining the pathophysiology in a patient with Chiari I malformation and syringomyelia, by directing rational surgical therapy and by testing new treatment paradigms based on these physiological and anatomical data.

References

1. Armonda RA, Citrin CM, Foley KT, et al: Quantitative cine-mode magnetic resonance imaging of Chiari I malformations: an analysis of cerebrospinal fluid dynamics. **Neurosurgery 35:** 214–224, 1994
2. Ball MJ, Dayan AD: Pathogenesis of syringomyelia. **Lancet 2:** 799–801, 1972
3. Barkovich AJ, Wippold FJ, Sherman JL, et al: Significance of cerebellar tonsillar position on MR. **AJNR 7:**795–799, 1986
4. Batzdorf U: Syringomyelia related to abnormalities at the level of the craniocervical junction, in Batzdorf U (ed): **Syringomyelia. Current Concepts in Diagnosis and Treatment.** Baltimore: Williams & Wilkins, 1991, pp 163–182
5. Bergstrand G, Bergstrom M, Nordell B, et al: Cardiac gated MR imaging of cerebrospinal fluid flow. **J Comput Assist Tomogr 9:**1003–1006, 1985
6. Blagodatsky MD, Larionov SN, Alexandrov YA, et al: Surgical treatment of Chiari I malformation with or without syringomyelia. **Acta Neurochir 141:**963–968, 1999
7. Citrin CM, Sherman JL, Gangarosa RE, et al: Physiology of the CSF flow-void sign: modification by cardiac gating. **AJR 148:** 205–208, 1986
8. DuBoulay G, Shah SH, Currie JC, et al: The mechanism of hydromyelia in Chiari I malformations. **Br J Radiol 47:**579–587, 1974
9. Dyste GN, Menezes AH: Presentation and management of pediatric Chiari malformations without myelodysplasia. **Neurosurgery 23:**589–597, 1988
10. Dyste GN, Menezes AH, VanGilder JC: Symptomatic Chiari malformations. An analysis of presentation, management, and long-term outcome. **J Neurosurg 71:**159–168, 1989
11. Enzmann DR, Pelc NJ: Cerebrospinal fluid flow measured by phase-contrast cine MR. **AJNR 14:**1301–1310, 1993
12. Gambardella G, Caruso G, Caffo M, et al: Transverse microincisions of the outer layer of the dura mater combined with foramen magnum decompression as treatment for syringomyelia with Chiari I malformation. **Acta Neurochir 140:**134–139, 1998
13. Gardner WJ, Angel J: The mechanism of syringomyelia and its surgical correction. **Clin Neurosurg 6:**131–140, 1959
14. Goel A, Achawal S: The surgical treatment of Chiari malformation association with atlantoaxial dislocation. **Br J Neurosurg 9:**67–72, 1995
15. Goel A, Bhatjiwale M, Desai K: Basilar invagination: a study based on 190 surgically treated patients. **J Neurosurg 88:** 962–968, 1998
16. Grabb PA, Mapstone TB, Oakes WJ: Ventral brain stem compression in pediatric and young adult patients with Chiari I malformations. **Neurosurgery 44:**520–528, 1999
17. Greitz D, Wirestam R, Franck A, et al: Pulsatile brain movement and associated hydrodynamics studied by magnetic resonance phase imaging. The Monroe-Kellie doctrine revisited. **Neuroradiology 34:**370–380, 1992
18. Heiss JD, Patronas N, DeVroom HL, et al: Elucidating the pathophysiology of syringomyelia. **J Neurosurg 91:**553–562, 1999
19. Kellie G: Appearances observed in the dissection of two individuals; death from cold and congestion of the brain. **Trans Med Chir Edinb 1:** 84, 1824
20. Krieger MD, McComb JG, Levy ML: Toward a simpler surgical management of Chiari I malformation in a pediatric population. **Pediatr Neurosurg 30:**113–121, 1999
21. Lane B, Kricheff II: Cerebrospinal fluid pulsations at myelography: a videodensitometric study. **Radiology 110:**579–587, 1974
22. Levy LM, Di Chiro G: MR phase imaging and cerebrospinal fluid flow in the head and spine. **Neuroradiology 32:**399–406, 1990
23. Menezes AH: Chiari I malformations and hydromyelia—complications. **Pediatr Neurosurg 17:**146–154, 1991
24. Milhorat TH, Chou MW, Trinidad EM, et al: Chiari I malformation redefined: clinical, radiographic, and genetic features in 364 symptomatic patients. **Neurosurgery 44:**1005–1017, 1999
25. Monro A: **Observations on the Structure and Functions of the Nervous System.** Edinburgh: Creech & Johnson, 1783
26. Nishikawa M, Sakamoto H, Hakuba A, et al: Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. **J Neurosurg 86:**40–47, 1997
27. Nishikawa M, Sakamoto H, Kitano S, et al: [A clinical study on long-term outcome in patients with syringomyelia associated with Chiari malformation.] **No To Shinkei 51:**318–323, 1999
28. Ogilvy CS, Borges LF: Treatment of symptomatic syringomyelia with a ventriculoperitoneal shunt: a case report with magnetic resonance scan correlation, **Neurosurgery 22:**748–750, 1988
29. Ohara S, Nagai H, Matsumoto T, et al: MR imaging of CSF pulsatory flow and its relationship to intracranial pressure. **J Neurosurg 69:**675–682, 1988
30. Pollack IF, Pang D, Albright AL, et al: Outcome following hindbrain decompression of symptomatic Chiari malformations in children previously treated with myelomeningocele closure and shunts. **J Neurosurg 77:**881–888, 1992
31. Quencer RM, Post MJ, Hinks RS: Cine MR in the evaluation of normal and abnormal CSF flow: intracranial and intraspinal studies. **Neuroradiology 32:**371–391, 1990
32. Royo-Salvador MB: [A new surgical treatment for syringomyelia, scoliosis, Arnold-Chiari malformation, kinking of the brainstem, odontoid recess, idiopathic basilar impression and platybasia.] **Rev Neurol 25:**523–530, 1997
33. Sahuquillo J, Rubio E, Poca MA, et al: Posterior fossa reconstruction: a surgical technique for the treatment of Chiari I malformation and Chiari I/syringomyelia complex—preliminary results and magnetic resonance imaging quantitative assessment of hindbrain migration. **Neurosurgery 35:**874–885, 1994
34. Sakamoto H, Nishikawa M, Hakuba A, et al: Expansive suboccipital cranioplasty for the treatment of syringomyelia associated with Chiari malformation. **Acta Neurochir 141:**949–961, 1999
35. Sgouros S, Williams B: Management and outcome of posttraumatic syringomyelia. **J Neurosurg 85:**197–205, 1996
36. Stoodley MA, Gutschmidt B, Jones NR: Cerebrospinal fluid flow in an animal model of noncommunicating syringomyelia. **Neurosurgery 44:**1065–1076, 1999
37. Stovner LJ, Bergan U, Nilsen G, et al: Posterior cranial fossa dimensions in the Chiari I malformation: relation to pathogenesis and clinical presentation. **Neuroradiology 35:**113–118, 1993
38. Vega A, Quintana F, Berciano J: Basichondrocranium anomalies in adult Chiari I malformation: a morphometric study. **J Neurol Sci 99:**137–145, 1990
39. Weed L: Some limitations of the Monro-Kellie hypothesis. **Arch Surg 18:**1049–1068, 1929
40. Williams B: Syringomyelia. **Neurosurg Clin North Am 1:** 653–685, 1991