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RESEARCH ARTICLE

TREATMENT OF CHIARI TYPE I MALFORMATION IN PATIENTS WITH AND WITHOUT SYRINGOMYELIA: A CONSECUTIVE SERIES OF 66 CASES

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ABSTRACT

Background - Cleland first described the anomaly later referred to as the Arnold-Chiari malformation, in 1883.[78] Chiari and Arnold in 1891 and 1894 respectively,[79,80] reported their descriptions and students of Arnold later used a single eponymic term to describe the various abnormalities.[81] More recently, Peach[82,83,84] and Carme| [85,86,87] have reviewed these early reports and others,[88,89,90] and clearly delineate the anatomical features of the Chiari malformations. In both the Chiari I and Chiari II anomalies, the cerebellar tonsils are displaced downwards below the foramen magnum to variable levels of the upper cervical cord. The Chiari II anomaly, better known as the Arnold-Chiari malformation, usually includes caudal dislocation of the medulla, inferior vermis and/or the fourth ventricle and may involve dorsal kinking of the cervicomedullary junction. The upper cervical nerve roots course rostrally. In both Chiari I and II malformations, hydrocephalus and syringomyelia may be present.

Aim - To study the outcome of surgical treatment in patients with Chiari type I malformation with and without syringomyelia

Methods and Material - We conducted a retrospective review of the medical records of patients having Chiari type I malformation with and without syringomyelia who were treated in our hospital, Shri Sai baba hospital, shirdi, between January 2011 and December 2017. Their age, sex, initial presentation, extent of syrinx, duration of follow up, syringosubarachnoid done or not, complication, second surgery done or not and outcome were reviewed. This is a retrospectively analyzed consecutive series of 66 patients (mean patient age 15 years, range 1–53 years). The uniform posterior craniovertebral decompression consisted of a small suboccipital craniectomy, a C-1 laminectomy, microsurgical reduction of the cerebellar tonsils, and dural closure with a synthetic dural graft to increase the cerebrospinal fluid space at the craniocervical junction. The presence of a large syrinx, with significant thinning of the spinal cord tissue and obliteration of the spinal subarachnoid space, particularly when combined with syrinx-related symptoms, was an indication for the placement of a syringosubarachnoid shunt.

Results - In 32 patients Chiari I malformation alone was present, and 34 in patients it was present in combination with syringomyelia. Clinical findings included pain, neurological deficits, and spinal deformity. The presence of syringomyelia was significantly associated with the presence of scoliosis (odds ratio 74.4 [95% confidence interval 8.894–622.4]). All patients underwent a posterior craniovertebral decompression procedure. In 22 of the 34 patients with syringomyelia a syringosubarachnoid shunt was also placed. The mean follow-up period was 24 months (range 3–95 months). Excellent outcome was achieved in 54 patients (82%) and good outcome in 12 (18%). In no patient were symptoms unchanged or worse at follow-up examination, including four patients who initially required a second operation for persistent syringomyelia. Pain was more likely to resolve than sensory and motor deficits after decompressive surgery. Radiological examination revealed normalization of tonsillar position in all patients. The syrinx had disappeared in 15 cases, was decreased in size in 17, and remained unchanged in two.

Conclusion - Posterior craniovertebral decompression and selective placement of a syringosubarachnoid shunt in patients with Chiari I malformation and syringomyelia is an effective and safe treatment. Primary placement of a shunt in the presence of a sufficiently large syrinx appears to be beneficial. The question of if and when to place a shunt, however, requires further, preferably prospective, investigation.

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INTRODUCTION

As described and classified over a century ago, (Chiari H *et al*) herniation of the cerebellar tonsils through the foramen magnum into the cervical spinal canal with obliteration of the cerebellomedullary cistern is the primary feature of Chiari I malformation. It is the mildest form of the Chiari complex that includes more extensive hindbrain anomalies often combined with spina bifida and severe forms of brainstem dysfunction and structural abnormality (Arnold J: Myelocyste et al). Nevertheless, the disorder can be associated with significant symptomatology, (Adams RD, Salam-Adams M, Doherty MJ, Spence DP, Young C, et al) risk of secondary injury due to trauma, and the risk of progression and damage of the spinal cord due to associated syringomyelia. Syringomyelia (also known as hydromyelia) is found in 50 to 70% of the patients with Chiari

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I malformation (Armonda RA, Citrin CM, Foley KT, et al, Dyste GN, Menezes AH, VanGilder JC et al). Numerous and thorough efforts to understand the pathogenesis of a syrinx caused by the obliteration of the craniovertebral CSF spaces have been undertaken (McLaurin RL, Bailey OT, Schurr PH, et al, Milhorat TH, Capocelli AL Jr, Anzil AP, et al, Oldfield EH, Muraszko K, Shawker TH, et al). Gardner's hydrodynamic theory was developed from the premise of an open connection from the fourth ventricle into the central canal of the cord. It suggested that the extension of the syrinx cavity is caused by a "water hammer" effect, which is a pulsatile transmission of the CSF from the fourth ventricle into the central canal in synchrony with the arterial blood pressure waves (Gardner WJ, Angel J et al). In Williams' craniospinal pressure dissociation hypothesis, (Williams B et al) he postulated an effect of venous (respiratory) pulsations resulting in a craniospinal pressure differences as the primary mechanism responsible for syrinx formation. With the advent of dynamic MR imaging the image acquisition could be related to the arterial and venous pulsations and the resulting CSF flows at the craniovertebral junction could be studied (Schroth G, Klose U et al). Based on findings of dynamic imaging technology, an explanation has been put forward for the pathogenesis of a syrinx (Oldfield EH, Muraszko K, Shawker TH, et al). This concept suggests that the cisternal obstruction at the foramen magnum results in a blockage of CSF flow during systole.

This blockage causes a systolic wave in the spinal CSF that acts on the surface of the spinal cord, which in turn results in progression of syringomyelia by repeated compression of the cord, probably by mechanical disruption of the cord tissue, and by forcing fluid into the cord from the outside in. The pulsatile pressure waves force CSF into the cord through the perivascular and interstitial spaces. In addition, the syrinx fluid is propelled longitudinally within the syrinx during each cardiac cycle.

The pathological CSF flow pattern and its restoration after decompressive surgery has been described. With the increased understanding of the pathophysiological basis for the development of syringomyelia in Chiari I malformation and with the common experience accumulated in numerous series (Armonda RA, Citrin CM, Foley KT, et al, Badie B, Mendoza D, Batzdorf U et al,), there is little controversy today that the common denominator of all treatment strategies is posterior craniovertebral decompression. Details of the procedure, its extent and invasiveness, the technique of dural closure, and whether a drainage system should be placed in the syrinx cavity, however, are subject to considerable dispute. In this report we present a series of patients with Chiari I malformation with and without syringomyelia. A standardized treatment was used: posterior craniovertebral treatment included a decompressive suboccipital craniectomy, a C-1 laminectomy, microsurgical reduction of the cerebellar tonsils, and dural closure with a synthetic graft to ensure enlargement of the craniovertebral CSF space. The spectrum of clinical presentations, radiological and surgical findings, and the postsurgical outcome is described. Details of the surgical strategy are also discussed.

Clinical material and methods

This is a retrospectively analyzed consecutive series of 66 patients, the majority of whom were treated by the senior neurosurgeon from January 2011 to December 2017. There were 40 female and 26 male patients, who ranged in age from 1 to 53 years (mean 15 years). Chiari I malformation alone was present in 32 patients, and in 34 patients it was present in combination with syringomyelia. Clinical, surgical, and radiological findings, as well as data on patient history, were reviewed from office charts and operative reports, direct review of MR images, or review of MR imaging reports. Follow-up data were obtained from office notes and from telephone interviews of patients and/or family members. The presence or absence of the following symptoms and clinical findings was specifically noted: headache, neck pain, sensory and motor disturbances, ataxia, scoliosis, and cranial nerve deficits. The time from onset of symptoms to surgery was noted. Based on their position on MR images, the cerebellar tonsils were classified into one of three categories: normal, descended to C-1, and descended to C-2 or lower. All patients underwent MR imaging within 3 to 5 days after surgery and at least one subsequent MR imaging session at the follow-up consultation, usually 3 to 6 months later. The presence or absence of syringomyelia, as well as its localization and extent in the cervical and/or the thoracic cord, were assessed preoperatively and at 3 to 6 month follow up and as needed thereafter.

Surgical Technique

All patients underwent surgery in the prone position with the head fixed in a Mayfield. A small suboccipital craniectomy and a C-1 laminectomy were performed. The craniectomy usually extended from the foramen magnum upward, including the inferior nuchal line of the occipital bone, covering the insertion area of the rectus capitis posterior minor muscle, and the medial insertion area of the rectus capitis posterior major muscle (Williams PL, Warwick R et al). The tonsils were visualized using intraoperative ultrasonography. If the tip of the tonsils could not be sufficiently exposed, a C-2 laminectomy was additionally performed. The dura was opened in a Y-shaped fashion. In the majority of cases the low-lying cerebellar tonsils were then reduced cranial ward by using bipolar coagulation of the pia (Batzdorf U et al, Won DJ, Nambiar U, Muszynski CA, et al). In cases of extensively large tonsils, subpial resection of a tonsil was occasionally required. Normalization of the position of the tonsils, visualization of the obex between the tonsils, and decompression of the brainstem and cranial nerves were the goals of this maneuver. The dura was closed with a triangular synthetic dural graft ensuring the reconstruction of open CSF space at the craniocervical junction. For placement of a syringosubarachnoid shunt, a laminotomy was placed at the level of greatest area of cord expansion.

The syrinx was identified using ultrasonography, the dura was opened (Dohrmann GJ, Rubin JM et al), and a midline myelotomy (Wisoff J: Hydromyelia et al) performed using an arachnoid knife. In three cases a dorsal root entry zone myelotomy (Iwasaki Y, Hida K, Koyanagi I, et al) was done because the cord was thinned in that area, and the least amount of damage to normal tissue was expected there. A small (0.025-inch [0.633-mm]) internal diameter; 0.047-inch [1.188-mm] external diameter) syringosubarachnoid shunt was placed (Hida K, Iwasaki Y, Koyanagi I, et al, Tator CH, Meguro K, Rowed DW et al) first into the subarachnoid space; after the myelotomy the proximal end of the tube was inserted into the syrinx cavity.

Before closure of the dura, the tube was sutured to the pia at the myelotomy by using a No. 6-0 nonabsorbable suture. The syringosubarachnoid shunt was always placed before performing the suboccipital craniectomy. The decision to place a shunt was based on two conditions: 1) radiological evidence of significant thinning of the cord tissue that surrounds the syrinx, as well as obliteration of the surrounding subarachnoid space; and 2) the presence of symptoms primarily attributable to syringomyelia rather than the Chiari I malformation alone.

The pragmatic reason for this relatively aggressive policy concerning syringosubarachnoid shunt placement at the primary procedure was the assumption that the risk of inflicting injury was judged smaller than the risk and effort of a subsequent separate operation. During all operations that involved shunt placement, intraoperative neurophysiological monitoring with motor evoked potentials and somatosensory evoked potentials was used in the same routine fashion as for operations involving spinal cord tumors (Deletis V et al, Kothbauer KF, Deletis V, Epstein FJ et al, Logue V, Edwards MR et al). Typical postoperative care involved the patient staying overnight in an intensive care unit. Patients were discharged from the hospital between 4 and 10 days postoperatively.

Procedure-related complications

The cases were analyzed for the following possible complications (Batzdorf U et al, Menezes AH et al). CSF leaks requiring either wound revision and/or lumbar drainage, wound infections requiring antibiotic treatment and/or surgical revision, infectious meningitis, intraoperative hemorrhage due to injury of the vertebral or posterior inferior cerebellar artery, visible intraoperative vasospasm in one of the latter vessels, postoperative radiological evidence of ischemia in the territories of these vessels, intraoperative changes in cardiovascular parameters, postoperative respiratory problems, and postoperative hemorrhage requiring reoperation. Postsurgical moderate headaches and the occurrence of so-called "aseptic" meningitis were regarded as adverse events rather than outright complications.

Outcome Measures

The long-term (>3 months postoperatively) surgery related result was considered excellent if symptoms resolved. The result was considered good if the patient experienced significant improvement but also residual symptoms. A poor result indicated no change in symptoms or deterioration. Specifically noted symptoms were assessed at the time of follow-up examination.

Statistical Analysis

All numerical data were stored electronically and analyzed using simple descriptive statistics.

RESULTS

Sixty-six consecutive patients with Chiari I malformation were included. In 32, Chiari I malformation alone was present, and in 34 it was present in combination with syringomyelia. Twenty-six patients were male and 40 female (Fig. 1).

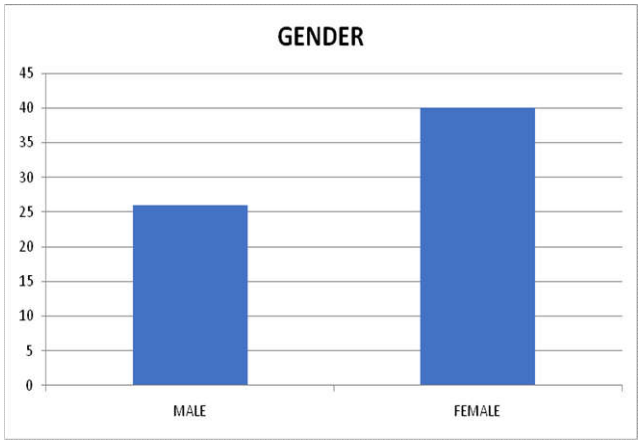

Figure 1.

The mean age at diagnosis was 15 years (range 1–53 years). Two thirds of the patients were children and adolescents (Fig. 2).

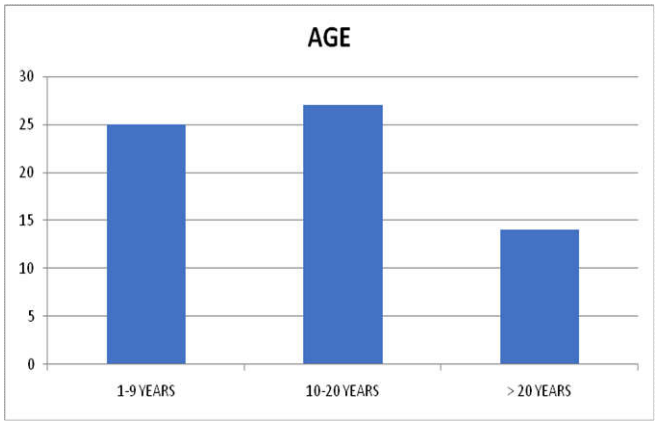

Figure 2.

Clinical Picture

The most common presenting symptoms were headache (34 patients [52%]) neck pain (22 patients [33%]), sensory deficits (20 cases [30%]), and a motor deficit (eight [12%]). Motor deficits more frequently affected the upper extremities (four patients with upper-extremity weakness only, two with lower-extremity weakness only, and one with quadriparesis) (Fig. 3).

In 11 patients (17%) scoliosis was the initial presenting symptom (Fig. 3). The average time from onset of symptoms to surgery was 11 months. The patients with syringomyelia more frequently suffered motor deficits than those without syringomyelia (ratio 7:1). Sensory symptoms were also more frequent in this group (ratio 4:1). Various cranial nerve deficits, including swallowing problems and torticollis, were found in 14 patients (21%). Ataxia was found in four patients. Gait problems, bladder dysfunction, movement disorders, respiratory problems, irritability, or motor regression were found in individual patients. There was a statistically significant relationship between the presence of some degree of scoliosis and the radiologically demonstrated presence of syringomyelia. Ten of 11 patients with scoliosis also harbored a syrinx. Thus, Chiari I malformation patients with scoliosis are significantly more likely to harbor a syrinx than those without scoliosis (odds ratio 74.4 [95% confidence interval 8.894–622.4]).

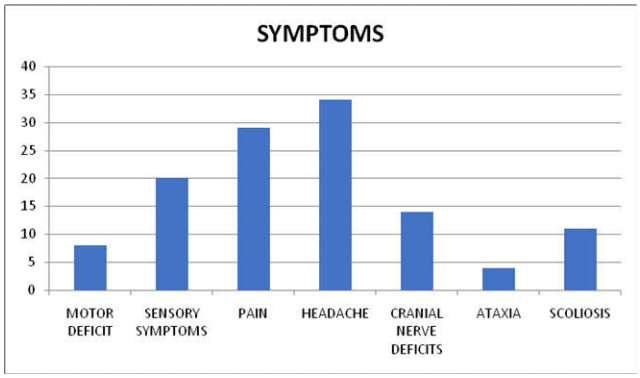

Figure 3.

Radiological Findings

Before surgery the cerebellar tonsils were extending below the foramen magnum level in all patients. The tip of the tonsils was between the foramen magnum and C-1 in 44 patients, between C-1 and C-2 in 20 patients, and below C-2 in two patients. As revealed on early postoperative MR images the position of the tonsils was normalized (that is, returned to the level of the foramen magnum) in 64 of the 66 patients in whom the tonsils had been microsurgically reduced or resected. A syrinx was found in 34 patients: in the cervical cord in 12, and with cervicothoracic extension in 22 (Fig. 4). After surgery, the syrinx had disappeared in 15 cases, was decreased in size in 17, and remained unchanged in two. Additional follow-up MR imaging findings were kyphosis in two patients, cervical stenosis in one, and progressive basilar invagination in one. In the patients with scoliosis no further progression of the deformity was seen on follow-up studies, but one patient underwent corrective surgery at another institution.

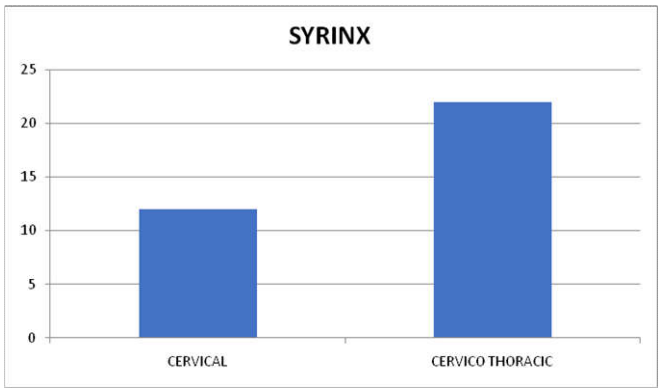

Figure 4.

Surgery

The standard posterior craniovertebral decompression was performed in all patients. An additional C-2 laminectomy was conducted in 11 patients. A resection of the tonsils, in place of just microsurgical reduction, was performed in three patients. Of 34 patients with a syrinx, a syringosubarachnoid shunt was placed in 22 in addition to the craniovertebral decompression; in all 22 cases the syrinx resolved or was reduced in size. One patient in this group required a second surgery to treat recurrent symptoms associated with the migration of the tube and increased syrinx size. In eight of the remaining 12 patients in whom no shunt was placed the syrinx resolved or significantly decreased in size. In two cases the syrinx persisted without change and in two it enlarged. These four patients required a second operation, which included insertion of a syringosubarachnoid shunt and resulted in significant decrease in syrinx size. In all patients in whom the syrinx was radiologically demonstrated to be improved, there was also a clinical improvement. In the patients who required repeat surgery, persistent symptoms or clinical progression were present. In three patients with syringomyelia some form of posterior fossa decompression had been performed at an outside facility prior to surgery at our institution. In all three we performed a syringosubarachnoid shunt procedure at our institution. One of these patients in whom posterior fossa decompression and tonsillar resection had previously been performed also required a shunt.

Procedure-Related Complications

Cerebrospinal fluid leakage occurred in four cases and was successfully managed using lumbar drainage and secondary suture. A superficial wound infection, treated with antibiotic agents, occurred in one patient. At later follow up, a pseudomeningocele was either clinically or radiologically demonstrated in three patients. No treatment was required. Five patients developed significant postsurgical neck pain and fever. Analysis of a CSF sample did not reveal evidence of infection. The patients were considered to have “aseptic meningitis,” or reaction to the graft, and were successfully treated with steroid and analgesic medications. The symptoms resolved within a postoperative period no longer than 3 weeks and had no influence on the final treatment result. One patient without previous neurological deficit developed a new distal hypesthesia in the foot as a complication of syringosubarachnoid shunt placement. This had improved at the time of last follow up. In one case intraoperative changes in motor evoked potentials were detected on one side, and the patient subsequently experienced a transient ipsilateral monoparesis of the leg. This deficit resolved within several days postoperatively and no further dysfunction was observed.

Treatment-Related Outcome

The mean follow-up time was 24 months, with a range of 3 to 95 months. An excellent outcome was achieved in 54 patients (82%) and a good result in 12 (18%). In no patient were symptoms unchanged or worsened postoperatively. There was no mortality. Improvement of specific symptoms was achieved in the large majority of cases (Fig. 5). Postoperative MR imaging assessment of the operative site in all patients demonstrates that the level of the cerebellar tonsils was normal and that the cisternal space at the level of the foramen magnum was indeed enlarged (Fig. 3). Postoperative spinal MR images obtained to assess of the syringomyelia revealed the disappearance of the syrinx in 15 cases, a significant decrease in syrinx size in 17, and no change in the syrinx size in two patients (Fig. 6).

DISCUSSION

The majority of our patients were children and adolescents. The sex distribution showed a female preponderance, which corresponds to some previous reports (Hida K, Iwasaki Y, Koyanagi I, et al, Krieger MD, McComb JG et al, Munshi I, Frim D, Stine-Reyes R, et al) but not to others in which even distribution was found. The profile of clinical presentations was comparable with what is known about the clinical entity. There were no clear differences in the incidence of major symptoms between this series and other published reports (Adams RD, Schatzki R, Scoville WB et al, Badie B, Mendoza D, Batzdorf U et al, Dohrmann GJ, Rubin JM et al).

Not surprisingly, there was a major difference in the time from onset of symptoms to surgery (11 months in this series), compared with pre–MR imaging series (Paul KS, Lye RH, Strang FA, et al, Rhoton AL Jr et al, Schlesinger EB, Antunes JL, Michelsen WJ, et al). During the last decade this interval between onset of symptoms and treatment has been similar in other reports (Nagib M et al). The severity of patient's symptoms is another major difference when older series are compared with this and other recent series. Magnetic resonance imaging documentation leads to an earlier diagnosis than would have been the case 35 years ago and, thus, symptoms are less severe. Obviously, MR imaging identifies asymptomatic cases as well (Meadows J, Kraut M, Guarnieri M, et al). Headache was the principal symptom in the majority of patients in this series. Some patients may have been treated for complex migraine like headache before the correct diagnosis was made. Based on the results of this series, headaches respond very well to decompressive surgery, as 33 of 36 patients with preoperative headaches experienced complete relief immediately after surgery. In the remaining three patients a longer period was required before the headaches resolved. Another commonly reported symptom of the Chiari malformation complex is pain other than headache (Heiss JD, Patronas N, DeVroom HL, et al, Milhorat TH, Kotzen RM, Capocelli AL Jr, et al). Most often this is neck, shoulder, arm pain, and frequently it is exacerbated with Valsalva-like maneuvers such as coughing, sneezing, or playing a brass instrument (Oldfield EH, Muraszko K, Shawker TH, et al, Schlesinger EB, Antunes JL, Michelsen WJ, et al,). At follow-up examination in our series, only one patient reported persistent neck pain, which was stronger with coughing; three patients reported significantly improved pain; all others were pain free. This is similar to results reported by other authors who, however, rarely distinguished between simple headache and more complex and extensive pain syndromes. In our series of the 20 patients who presented with sensory deficits, 16 had syringomyelia. Hypesthesia and moderate dysesthesia in the upper extremities were present. In 10 of these 20 patients complete normalization of sensory symptoms was achieved (seven syrinx, and three without), in nine improvement occurred, and in one patient no change was demonstrated. In previous series and studies the authors have reported resolution or significant improvement of sensory symptoms in 70 to over 90% (Munshi I, Frim D, Stine-Reyes R, et al, Krieger MD, McComb JG et al).

Preoperative motor dysfunction was only present in cases of Chiari I malformation associated with syringomyelia. Motor deficits were present in eight patients, which is 12% of the entire patient group. This incidence is lower than that in other reports (Blagodatsky MD, Larionov SN, Alexandrov YA, et al, Paul KS, Lye RH, Strang FA, et al), perhaps because of the improved neurodiagnostic modalities widely available in the last decade. In our series, these motor deficits resolved in two of eight cases, improved in five, and remained unchanged in one. Deterioration of function occurred in no patient. Thus, it appears that motor dysfunction improves somewhat less significantly than sensory dysfunction and pain, which, makes a strong case for early diagnosis and treatment. These findings differ from reports in which there was no postoperative improvement of a once established motor deficit.[39] Again these reports are almost 30 years old, and the patient population then was likely different from those studied during the recent decade. Other symptoms, such as ataxia and cranial nerve deficits, improved postoperatively in our series, which corresponds to outcomes reported by other authors. Scoliosis is an important finding in patients with Chiari I malformation in whom syringomyelia is also present. This was particularly notable in a previous series in which 20 of 26 patients with syringomyelia were found to have a significant degree of scoliosis (Krieger MD, McComb JG, Levy ML et al). In the present report, 11 of the 66 patients had scoliosis, and all but one of the 11 harbored a syrinx. There is a statistically significant association between scoliosis and syringomyelia in this patient group. None of these patients suffered further progression of their spinal deformity after surgery. This rate is similar to that cited in the aforementioned report, in which only one of 20 patients eventually underwent surgery for correction of the scoliotic deformity (Krieger MD, McComb JG, Levy ML et al, Milhorat TH, Chou MW, Trinidad EM, et al). Moreover, the treatment of Chiari I malformation has been shown to improve the spinal curvature when scoliosis was present. Magnetic resonance imaging is the standard preoperative neurodiagnostic tool for assessing patients with Chiari Imalformation. The defining feature in this entity is the presence of cerebellar tonsils that hang below the foramen magnum, and this was documented in our entire series; in approximately one third of them the tonsils have even become displaced caudal to the arch of C-1. A general impression that the posterior fossa is smaller in these patients has been reported previously, but it was not further corroborated in the present series. Early postoperative MR images were obtained, however, and the craniocaudal position of the cerebellar tonsils had normalized in all patients. Only in the two early cases, in which microsurgical reduction of the tonsils was not completed, was this normalization was not obtained (Fig. 3). Only three of our patients were found to have associated craniocervical anomalies, such as basilar invagination or atlantoaxial fusion. The incidence of such anomalies was low compared with other series in which these abnormalities appear on average to be present in every fourth patient (Menezes AH et al, Milhorat TH, Chou MW, Trinidad EM, et al). The reason for this discrepancy is most likely referral bias.

Details of the Surgical Procedure

Decompression of the cerebellum and cervical spinal cord has been suggested as a treatment for Chiari malformation and syringomyelia since the 1930s (Gardner WJ, Goodall RJ et al, Gustafson WA, Oldberg E et al). Plugging of the obex in an attempt to block presumed CSF flow into the central canal has not worked well (Gardner WJ, Angel J et al), carries a manipulation-related complication risk (Williams B et al), and has therefore been largely abandoned. In the present series the area of the obex was carefully avoided, and there were no complications associated with lower brainstem function. The correct way to treat Chiari malformations is one of the most disputed issues in neurosurgery. The most common form of treatment is the posterior fossa decompression. Some authors suggest that this craniectomy must be of considerable size (Feldstein NA, Choudhri TF et al, Milhorat TH, Chou MW, Trinidad EM, et al), and some even combine this with some form of cranioplasty. Others, like us, prefer small craniectomies; the recently described cerebellar ptosis that occurs after larger craniectomy may support this more conservative approach. Some authors have advocated that the craniectomy alone may be sufficient, whereas others have argued that the dura must be opened. Some have opening of the dura only, but not the arachnoid (Feldstein NA, Choudhri TF et al, Hida K, Iwasaki Y, Koyanagi I, et al). Interestingly, whereas there appears to be a large number of centers where it is customary to use microsurgical procedures to reduce the size

of the cerebellar tonsils, there are comparatively few reports in which this practice is described (Depreitere B, Van Calenbergh F, Van Loon J, et al, Fischer EG et al). Mostly the results are favorable and differ little among series. This was confirmed by the present study as the outcomes were good, the number complications were small, and those complications that did occur were not severe. The issue of dural closure is disputed. Some have advocated no dural closure at all, whereas others use an autograft obtained from the fascia lata; however, this gives the patient an additional incision that causes significant discomfort. In some series pericranium has been used for the dural graft (Feldstein NA, Choudhri TF et al, Sakamoto H, Nishikawa M, Hakuba A, et al). In most cases the dural closure is achieved using an artificial dural substitute (Oldfield EH, Muraszko K, Shawker TH, et al, Weinberg JS, Freed DL, Sadock J, et al).

Many of these technical questions at this point are decided according to the experience of the surgical team and reflect the choice of the individual surgeon rather than a scientific consensus. Our preference for the dural substitute is based on the experience gained from spinal reoperations performed following surgeries in which the dura was closed using this graft type compared with autografts such as fascia lata. We have found little scar tissue after placement of the artificial substitute but a lot when using autografts. What is the best way to treat syringomyelia in the context of Chiari I malformation? There is considerable evidence that posterior craniovertebral decompression leads to reduction in syrinx size in the majority of cases (Batzdorf U et al, Menezes AH, Smoker WRK, Dyste GN et al). We have adopted a pragmatic approach to this problem. As a primary treatment we placed a syringosubarachnoid shunt when the symptomatology was believed to result from the syrinx rather than the Chiari malformation–induced compression, and when the syrinx was large with thinned spinal cord tissue and obliterated spinal subarachnoid space. Even though there are clinical and a radiological aspects upon which to base a decision to use a syringosubarachnoid shunt, some patients with smaller syringes still experienced progression despite the craniovertebral decompression and subsequently required a second surgery for the syrinx shunt. Shunt tubes from a syrinx to the subarachnoid space provide an outflow route for CSF during external cord compression, which occurs during cardiac systole (Heiss JD, Patronas N, De Vroom HL, et al).

Other authors conducted similar comparisons in larger series and found a better outcome after placement of the shunt alone (Hida K, Iwasaki Y, Koyanagi I, et al, Iwasaki Y, Hida K, Koyanagi I, et al). Obviously these results cannot be compared with those in the present series because all patients in whom shunts were placed also underwent craniovertebral decompressive surgery. In a large recent report, (Goel and Desai et al) concluded that posterior craniovertebral decompression and placement of a syringosubarachnoid shunt are superior to placing a shunt alone, which certainly agrees with the present results. They also stated that craniovertebral decompression alone is an option for treatment, which is corroborated by other reports as well (Sgouros S, Williams B et al). A syrinx extending over a long stretch of spinal cord (>four to five spinal segments) may likely require a shunt, the axial diameter of the syrinx and the thickness of spinal cord tissue, however, are important factors. Currently there are no accepted imaging parameters on which to base the decision of whether to place a shunt.

The issue of whether a syringosubarachnoid shunt is necessary regardless of the syrinx size is also not sufficiently resolved. Because many authors consider the risk of the myelotomy unacceptable in a patient with otherwise little dysfunction, they have argued that posterior fossa decompression alone is adequate. On the other hand, other authors favor the shunt, particularly in cases of rapid neurological deterioration, because they claim that it facilitates and accelerates neurological recovery (Barbaro NM, Wilson CB, Guting PH, et al, Hida K, Iwasaki Y, Koyanagi I, et al). Intraoperative monitoring and pre- and postoperative assessment with somatosensory evoked potentials have shown measurable improvement in the neurophysiological properties of the spinal cord around a syrinx after placement of a shunt as compared with before (Wagner W, Perneczky A, Maurer JC, et al). This may be due to the relieved mechanical stretching of the axons (Milhorat TH, Kotzen RM, Capocelli AL Jr, et al). In any event, the resolution or improvement of the syrinx demonstrated on follow-up imaging has reportedly been achieved in a very high percentage of patients without recurrences (Fischer EG et al, Morota N, Deletis V, Constantini S, et al). Thus, there is evidence to support the therapeutic effect of a syrinx shunt placed at the time of posterior craniovertebral decompression.

In the present series, syringomyelia recurred in three patients who harbored a preoperative syrinx but who underwent decompressive surgery alone. Of the patients who underwent decompressive surgery and placement of a syringosubarachnoid shunt, one patient suffered a recurrence due to shunt migration. The recurrence of the syrinx may be a question of follow-up time; thus, when patients who have undergone decompressive surgery alone are followed long enough, a syrinx may recur in a significant number (Logue V, Edwards MR et al). In addition to numerous preoperative symptoms and deficits found in patients with Chiari I malformation, numerous possible surgery-related complications have been described. In the early days of neurosurgery, decompressive treatment of this disorder appears to have been a dangerous operation, with significant mortality (Penfield W, Coburn DF et al, Williams B et al).

More recently the standard Chiari decompression is a procedure of comparatively low risk. In our series, there were no cases of mortality, postoperative respiratory dysfunction, or severe infections requiring wound revision. The 7.5% CSF leakage rate is similar to or smaller than that reported in other series. Some authors have reported that posterior fossa decompression without duraplasty prevents this complication (Williams B et al). Existing evidence, however, supports the rationale of enlarging the craniocervical CSF spaces (Milhorat TH, Chou MW, Trinidad EM, et al, Oldfield EH, Muraszko K, Shawker TH, et al). It appears that this makes the essential difference in surgical treatment of this condition. We believe therefore that the risk of CSF leakage has to be accepted as a downside of the essential procedure, which is the opening the dura and widening the cisternal space with a graft. This series includes five cases of irritative (“aseptic”) meningitis with neck pain, low-grade fever, malaise, discomfort, but negative CSF cultures (Munshi I, Frim D, Stine-Reyes R, et al, Van Calenbergh F, Van Loon J, et al). These patients were successfully treated with of steroid and analgesic medications. In three cases a pseudomeningocele was seen on the 3-month follow-up MR images. Only minor neck pain was associated with the pseudomeningoceles, and none of these patients

required further treatment. Overall the significant and sustained postoperative improvement demonstrated in these 66 patients suggests a favorable therapeutic result in itself as well as when compared with other results obtained in previously reported studies.

Conclusion

Treatment of Chiari I malformation with posterior craniovertebral decompression was undertaken in a large series on standardized basis. The surgical approach included microsurgical reduction of the cerebellar tonsils and placement of an artificial dural graft, which ensured enlargement of the cisternal space at the craniocervical junction. Associated syringomyelia is treated with placement of a syringosubarachnoid shunt if the syrinx is of significant size and causes symptoms. This treatment strategy is safe and effective for the related symptomatology. The presence of syringomyelia is a sign of advanced structural abnormality, as it is associated with the presence of sensory and motor deficits, as well as with the presence of spinal deformity. The preoperative presence of deficit is a predictor of poorer neurological outcome, making a strong case for early surgical intervention.

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