Surgical Treatment of Chiari I Malformation: Simplified Technique and Clinical Results

Michael B. Pritz, M.D., Ph.D.¹

ABSTRACT

A simplified technique for decompressing the posterior fossa and foramen magnum in symptomatic patients with a Chiari I malformation is described. The approach uses a curvilinear dural incision and an autologous pericranial graft. This procedure was used in 14 symptomatic patients, 8 of whom had a cervical or cervicothoracic syrinx. Neurologic signs and symptoms improved or were unchanged in all patients. The size of the syrinx decreased in all patients. No patient developed a new neurologic deficit, cerebrospinal fluid leak, pseudomeningocele, or infection. Postoperatively, one patient developed new, dysesthetic, unilateral arm pain that did not require medication. This technique has several advantages. The procedure is simple and requires a single skin incision. Autologous graft can be used. The occipital sinus can be avoided. Finally, the contralateral region remains pristine if a repeat procedure becomes necessary.

KEYWORDS: Chiari I malformation, surgical technique, treatment

Surgery is the treatment of choice for symptomatic patients with a Chiari I malformation either without or with a spinal cord syrinx. In symptomatic individuals without hydrocephalus or another structural lesion to explain cerebellar tonsillar herniation, various surgical procedures have been used with different degrees of success.¹⁻⁴ Because the volume of

the posterior fossa is small in Chiari I patients,⁵ decompression of the foramen magnum and posterior fossa has been the mainstay of surgical treatment. This report describes a simplified technique for posterior fossa and foramen magnum decompression that has proven successful in symptomatic patients with a Chiari I malformation.

Skull Base, volume 13, number 3, 2003. Address for correspondence and reprint requests: Michael B. Pritz, M.D., Ph.D., Indiana University School of Medicine, Section of Neurological Surgery, 545 Barnhill Drive, Emerson Hall 139, Indianapolis, IN 46202-5124. E-mail: mpritz@iupui.edu. ¹Indiana University School of Medicine, Section of Neurological Surgery, Indianapolis, Indiana. Copyright © 2003 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662. 1531-5010,p;2003,13,03,173,178,ftx,en;sbs00348x.

MATERIAL AND METHODS

Patients

Fourteen patients (5 men and 9 women) underwent the surgical procedure described below. Their ages ranged from 18 to 65 years. Eight patients (57%)

had an associated syrinx. All patients were symptomatic. No patient had arachnoiditis, hydrocephalus, or other intracranial structural pathology aside from the Chiari I malformation. Postoperative neurologic examinations and imaging studies were performed at regular intervals (range, 4 months to almost 3 years). Follow-up is ongoing in most patients.

Figure 1 Surgical technique for Chiari I decompression. (A) Midline skin incision. (B) Y-shaped fascial opening. (C) Placement of trephines. (D) Dural incision. (E) Harvest of diamond-shaped pericranial patch. (F) Pericranial patch is sewn in place. (G) Final appearance of dural closure with patch graft.

Surgical Technique

Under general anesthesia, patients are placed prone in skeletal fixation with the head elevated and all pressure points padded. Hair is shaved from the occipital area to the upper neck. Perioperative antibiotics are administered parenterally. Magnifying loupes are used for the procedure. An incision is outlined beginning about 7 cm above theinion and extending downward in the cervical area based on the inferior location of the cerebellar tonsils (Fig. 1A). The incision is carried down to the pericranium and fascia. The occipital scalp is undermined to provide adequate exposure for the donor graft. The fascia is opened in a Y-shaped fashion (Fig. 1B).⁶ Trephines are placed in the suboccipital area (Fig. 1C). A suboccipital craniectomy is performed, followed by a cervical laminectomy based on the intradural pathology and extent of tonsillar descent (Fig. 1D). The dura is incised in a curvilinear fashion avoiding the occipital sinus (Fig. 1D). The arach-

noid is opened and obvious adhesions are freed. A diamond-shaped pericranial patch is harvested from the occipital area and shaped to fit the dural defect (Fig. 1E). A piece of Gelfilm is placed over the exposed cerebellum and upper spinal cord. The pericranial patch is sewn in place using double-arm 5-0 prolene sutures in a fashion similar to that used to close a patch graft for arterial reconstruction (Fig. 1F and G). The integrity of the closure is ascertained and any areas of leakage are repaired. The suture line is covered with biological adhesive. The incision is closed in anatomical layers.⁶ No drain is used.

RESULTS

Preoperatively, patients exhibited a variety of signs and symptoms (Table 1), which are presented in a format used by others.⁷ The inferior extent of the

Table 1 Clinical Results in 14 Patients with Chiari I Malformation without or with Syringomyelia

Symptom/Sign	Preop Status (No. Patients)				Postop Status (No. Patients)			
	Absent	Mild	Moderate	Severe	Absent	Mild	Moderate	Severe
Symptom								
Headache	12	1	1	0	13	1	0	0
Dysesthetic pain	13	0	1	0	12	2	0	0
Subjective weakness	9	1	3	1	11	3	0	0
Sensory loss by Hx	8	1	2	3	8	5	1	0
Oscillopsia	11	0	1	2	12	2	0	0
Paresthesias	7	1	6	0	9	5	0	0
Dysphagia	10	0	4	0	12	2	0	0
Chronic cough	12	0	1	1	14	0	0	0
Weak voice	12	1	1	0	14	0	0	0
Sleep apnea	13	0	1	0	14	0	0	0
Impaired balance	8	0	6	0	8	6	0	0
Sign								
Weakness on examination*	10	1	2	1	11	3	0	0
Atrophy	11	2	0	1	12	2	0	0
Sensory loss on examination	7	3	2	2	8	5	1	0
Nystagmus	10	1	0	3	10	4	0	0
Vocal cord paresis	13	0	0	1	13	1	0	0
Gait impairment	9	1	4	0	10	4	0	0

*Weakness is defined as absent (5/5), mild (4/5), moderate (3/5; antigravity strength), severe (1-2/5; < antigravity strength). Hx, history; no., number; preop, preoperative; postop, postoperative.

cerebellar tonsils was 6 mm below the foramen magnum in 1 patient, at the posterior arch of C1 in 11 patients, and at the posterior arch of C2 in 2 patients. Eight patients had a syrinx, which was confined to the cervical area in 3 patients and to the cervicothoracic region in 5 patients (Figs. 2A and B).

Postoperatively, signs and symptoms either improved or were unchanged in all 14 patients. One patient developed new, unilateral dysesthetic arm pain but required no medication (Table 1). The size of the syrinx decreased in all patients (Fig. 2C) based on studies performed 9 months to nearly 3 years after surgery. No patient experienced a new neurologic deficit, wound infection, cerebrospinal fluid leak, pseudomeningocele, or recurrent syrinx after an initial decrease in syrinx size. No patient declined neurologically during the interval of postoperative follow-up.

DISCUSSION

Various surgical approaches have been described to treat Chiari I malformations without and with a spinal cord syrinx.¹⁻⁴ The present technique has proven successful in terms of treatment of both clinical signs and symptoms. Radiologic evidence has documented a decrease in syrinx size. Although somewhat controversial, opening the arachnoid allows direct inspection of structures at the cranio-cervical junction. In certain instances, inspection has led to additional bone removal and to the separation of arachnoidal adhesions. No patient developed a cerebrospinal fluid leak, pseudomeningocele, or infection. No patient incurred a new neurologic deficit after surgery, although a single patient developed mild, dysesthetic, unilateral arm pain that did not require medication. Follow-up has been rel-

Figure 2 Pre- and postoperative images of a symptomatic patient with a Chiari I malformation and cervicothoracic syrinx. T2-weighted sagittal magnetic resonance images show (A) tonsillar herniation extending to the posterior arch of C1 and (B) a large syrinx extending from C2 (A) to T9. (C) After surgery the size of the syrinx had decreased markedly.

actively brief. In patients followed for several years, however, their postoperative neurologic improvement has been maintained and the size of their syrinx has remained diminished.

The increased incision length needed to enable adequate exposure of the pericranial graft is a disadvantage of this technique. The procedure, however, has several advantages. First, the operation is simple. Second, it requires only a single incision. Third, it uses autologous tissue. Fourth, it avoids the occipital sinus. Fifth, if repeat surgery requiring further intradural expansion or exploration is needed, the contralateral side is intact, minimizing difficulties related to a prior operation. In our experience, this technique has been associated with satisfactory clinical and radiologic outcomes and complications have been minimal.

ACKNOWLEDGMENTS

I am grateful to J. Corbitt for manuscript preparation, S. Teal for the illustration shown in Figure 1, and J. Murphy for help in preparation of Figure 2. I thank the following physicians for patient referral: H. DeLeeuw, M. Farlow, J. Helfrich, F. Khairy, G.J. Loomis, B.G. Maybury, J. Munshower, T. Poe, E. Pretila, J. Vivio, J. Wojcieszek, and R. Yee.

REFERENCES

1. Batzdorf U. Syringomyelia, Chiari malformation, and hydromyelia. In: Youmans JR, ed. *Neurological Surgery: A Comprehensive Reference Guide to the Diagnosis and Management of Neurosurgical Problems*. 4th ed. Philadelphia, PA: WB Saunders; 1996:1090-1109
2. Ellenbogen RG, Zeidman SM. Craniovertebral decompression for Chiari malformation. In: Kaye AH, Black PMcL, eds. *Operative Neurosurgery*. New York, NY: Churchill Livingstone; 2000:1725-1741
3. Batzdorf U. Microsurgery of syringomyelia and syringomyelia cord syndrome. In: Schmidek HH, ed. *Schmidek & Sweet Operative Neurosurgical Techniques: Indications, Methods, and Results*. 4th ed. Philadelphia, PA: WB Saunders; 2000:1946-1954
4. Rhoton AL Jr. Microsurgery of syringomyelia and the syringomyelia—Chiari complex. In: Schmidek HH, ed. *Schmidek & Sweet Operative Neurosurgical Techniques: Indications, Methods, and Results*. 4th ed. Philadelphia, PA: WB Saunders; 2000:1955-1969
5. Milhorat TH, Chou, MW, Trinidad EM, et al. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. *Neurosurgery* 1999;44: 1005-1017
6. Kempe LG. *Operative Neurosurgery*. Vol 2: Posterior Fossa, Spinal Cord, and Peripheral Nerve Disease. New York, NY: Springer-Verlag; 1970:14-23
7. Heiss, JD, Patronas N, DeVroom HL, et al. Elucidating the pathophysiology of syringomyelia. *J Neurosurg* 1999;91: 553-562

Commentary

This is another variation on procedures for decompression of the posterior fossa for the treatment of Chiari I malformations. The bony decompression is the same as in other procedures. The authors, however, limit the dural opening and arachnoidal dissection and then make a dural graft from autologous pericranium. After treatment most of their patients improved, and all syringes resolved. The procedure requires a larger incision than usual but avoids using foreign materials for dural grafts and limits subarachnoid scarring. It is important to remember that the decompression must include the entire region that appears to be restricted on preoperative magnetic resonance imaging.

Chandranath Sen, M.D.¹