J Neurosurg Pediatrics 2:188–193, 2008

Postnatal ascent of the cerebellar tonsils in Chiari malformation Type II following surgical repair of myelomeningocele

Nobuhito Morota, M.D., and Satoshi Ihara, M.D.

Department of Neurosurgery, National Children's Medical Center, National Center for Child Health and Development, Tokyo, Japan

Object. Postnatal improvement in Chiari malformation type II (CM-II) following surgical repair of myelomeningocele was evaluated.

Methods. The authors reviewed data obtained in 20 cases in which patients underwent postnatal myelomeningocele repair within the first 48 hours after birth between October 2002 and September 2006. In 14 patients (Group 1), myelomeningocele was diagnosed in utero and the infants were delivered by cesarean section at 35–39 weeks' gestation (mean 36.4). The 6 infants in Group 2 were born after full-term gestation (39–41 weeks), and their myelomeningoceles were diagnosed postnatally. In all 20 patients, the myelomeningoceles were surgically repaired postnatally. Dynamic change of the herniated cerebellar tonsils in CM-II before and after the myelomeningocele repair, associated hydrocephalus, and symptomatic CM-II were analyzed.

Results. In Group 1, the CM-II was confirmed before myelomeningocele repair in 13 cases (93%). The spinal level of the caudal end of the cerebellar tonsils ranged from C-2 to C-7. Ascent of the cerebellar tonsils was observed in 11 patients (range 1–4 spinal levels, mean 2 levels) and continued even after ventriculoperitoneal (VP) shunt placement in most patients. A VP shunt was required for the treatment of hydrocephalus in 12 patients (86%). Symptomatic CM-II developed in 8 of 13 patients (61%), 3 of whom required surgical decompression.

In Group 2, CM-II was confirmed in 5 infants (83%), with the cerebellar tonsils at a spinal level of C-2 to C-4 or C-5. Ascent of the cerebellar tonsils was observed in 4 patients (range 1–1.5 spinal levels, average 1.1 levels), and no patient had symptomatic CM-II. A VP shunt was placed in 5 patients (83%). No patient was lost to follow-up during the 18-month follow-up period. The only statistically significant difference between the 2 groups was the presence of symptomatic CM-II in Group 2 ($p = 0.02$).

Conclusions. Patients showed ascent of the cerebellar tonsils after postnatal myelomeningocele repair. Placement of a VP shunt helped promote the ascent. However, postnatal myelomeningocele repair in the patients in Group 1 failed to consistently prevent development of symptomatic CM-II. This limited experience suggests that postnatal repair of myelomeningocele can partially reverse the anatomical CM-II, but symptomatic CM-II cannot be prevented in some patients when the repair is performed after 36 weeks' gestation. *(DOI: 10.3171/PED/2008/2/9/188)*

Key Words • **Chiari malformation Type II** • **decompression surgery** • **hydrocephalus** • **myelomeningocele** • **neonatal surgery**

Chiari malformation Type II is a serious condition associated with myelomeningocele. Its frequency is reported to be as high as 90% in patients with myelomeningocele, and it becomes symptomatic in about 20–30%.[4,9] Affected patients experience multiple symptoms of brainstem dysfunction, including dysphagia, dysarthria, and dyspnea, which can be life-threatening in some cases. Prevention of symptomatic CM-II has long been a challenge for pediatric neurosurgeons.[6,7,9] Management of hydrocephalus has been regarded as one of the most influential factors in preventing the symptoms of CM-II. Foramen magnum and upper cervical decompression are indicated when the CM-II becomes symptomatic despite satisfactory treatment of hydrocephalus and systemic management.

Improvement of CM-II (ascent of the cerebellar tonsils) following in utero repair of myelomeningocele has been reported, although the condition had not been thought to change after birth.[2,8,10–12] A decreased frequency of developing CM-II has been attributed to early myelomeningocele repair, which can restore CSF circulation.[10,11] Based on the unified theory of the pathogenesis of myelomeningocele, fetal repair of the lesion stops the CSF leakage and improves the pressure gradient between the intracranial and spinal cavities, thus preventing CM-II development.[2–4,11]

The possibility of improvement in CM-II in the neonatal

Abbreviations used in this paper: CM-II = Chiari malformation Type II; CSF = cerebrospinal fluid; VP = ventriculoperitoneal.

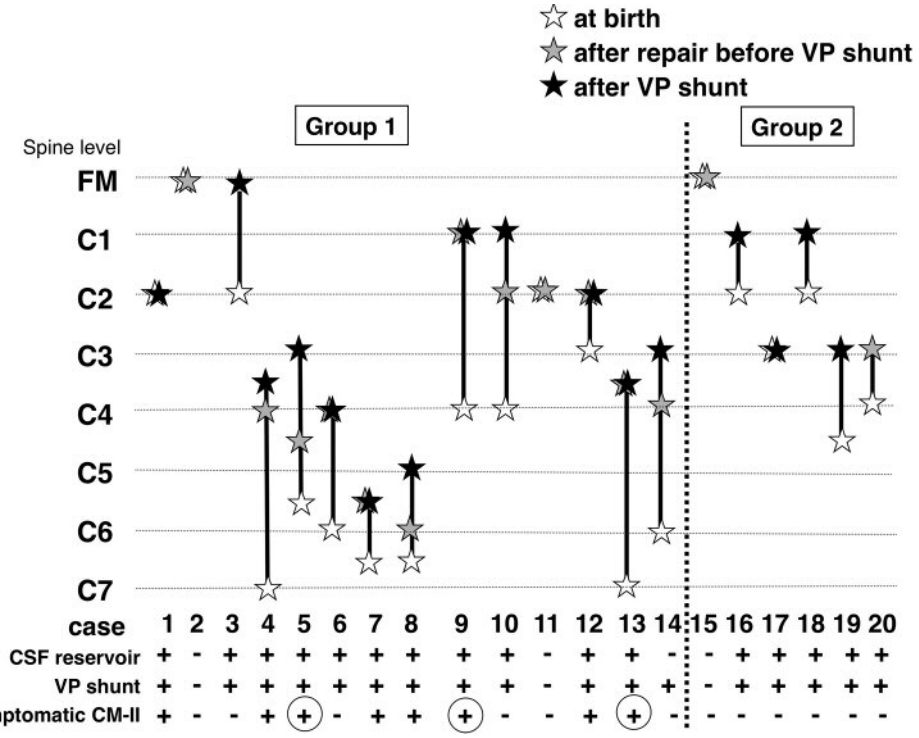

FIG. 1. Graph showing the dynamic changes seen in patients with CM-II. The spinal level of the cerebellar tonsils as well as the presence of hydrocephalus and symptomatic CM-II in each patient are shown. *Open circles* indicate patients who later required decompressive surgery. Ascent of the cerebellar tonsils was more prominent in Group 1 (fetal group) than in Group 2 (postnatal group). FM = foramen magnum.

period has not been much studied since the condition has been regarded as irreversible after birth.[10,11] Recently, authors have observed marked ascent of the hindbrain following the repair of myelomeningocele after birth, even before VP shunt placement. This observation suggests that the CM-II could be reversible even after birth. We measured the spinal level of the caudal end of the cerebellar tonsils on MR images before and after myelomeningocele repair and analyzed the postnatal dynamic changes of the CM-II.

Methods

The study population included 20 patients who underwent postnatal myelomeningocele repair within the first 48 hours after birth between October 2002 and September 2006. Myelomeningocele was diagnosed by MR imaging in utero in 14 patients (Group 1); in the other 6 patients (Group 2), it was detected postnatally. The in utero MR imaging studies in Group 1 were performed at a mean of 31 weeks' gestation (range 21–39 weeks). Based on our institutional guidelines, patients in Group 1 were delivered by elective cesarean section at a gestational age of ~ 36 weeks. The infants in Group 2 were born at full term (range 39–41 weeks, mean 39.3 weeks). All patients underwent MR imaging before myelomeningocele repair. The myelomeningocele was repaired 1 day after birth in all cases, and a CSF reservoir was placed in the lateral ventricle at the same time in those infants who showed signs of hydrocephalus before myelomeningocele repair. Intermittent CSF aspiration through the reservoir was performed, with the frequency ranging from daily to several times a week, depending on the progress of the hydrocephalus. A VP shunt was inserted when the signs of increased intracranial pressure became uncontrollable by CSF aspiration or the infant developed signs of symptomatic CM-II. Follow-up MR images were obtained before VP shunt placement, which was usually performed 1–2 weeks after myelomeningocele repair (range 3–13 days after the first MR imaging study, mean 11.1 days). When patients developed medically intractable CM-II–related symptoms, foramen magnum and upper cervical decompression was indicated. The spinal level was judged using the cervical spine level at the caudal end of the cerebellar tonsils on a mid-sagittal MR image. The final spinal level of the CM-II was measured on the MR images obtained before discharge (range 15–134 days after the first MR imaging study, mean 46.5 days).

Data in Groups 1 and 2 were compared. Differences in the presence of CM-II, the proportion of infants with symptomatic CM-II, the degree of hindbrain ascent, and the need for a VP shunt were examined using the chi-square test. Nonparametric data pertaining to the range of ascent indicated by spinal levels were analyzed using the Mann–Whitney U-test.

Results

In 13 (93%) of the 14 patients in Group 1, CM-II was confirmed on the preoperative MR images obtained before myelomeningocele repair; the caudal end of the cerebellar tonsils ranged from C-2 to C-7 (average level C-4/5, Fig.1). Ascent of the cerebellar tonsils was observed in 11 patients (range 1–4 spinal levels, mean 2 spinal levels). Signs of hy-

drocephalus were seen in 11 patients at birth, and a CSF reservoir was implanted at the time of myelomeningocele repair. These 11 patients, as well as another patient whose hydrocephalus became apparent after birth, later required a VP shunt; thus shunts were required in 12 (86%) of 14 patients in Group 1. Symptomatic CM-II developed in 8 of the 13 patients in whom CM-II was confirmed (61%). All 8 of these patients had functioning VP shunts; 3 of them had life-threatening apneic spells with bradycardia and subsequently underwent decompressive surgery.

In 5 (83%) of the 6 patients in Group 2, CM-II was confirmed on the preoperative MR images obtained before myelomeningocele repair; the caudal end of the cerebellar tonsils ranged from C-2 to C-4/5 (average level C-3). Ascent of the cerebellar tonsils was observed in 4 patients (range 1–1.5 spinal levels, mean 1.1 spinal levels). No patient in this group developed symptomatic CM-II. Signs of hydrocephalus were present in 5 patients (83%) at birth, and all received a VP shunt following placement of a CSF reservoir during the postnatal course.

There were no statistically significant differences between the 2 groups in terms of the presence of CM-II ($p = 0.52$), ascent of the cerebellar tonsils ($p = 0.65$), or hydrocephalus requiring a VP shunt ($p = 0.68$). The only statistically significant finding was the presence of symptomatic CM-II in Group 1 ($p = 0.02$). With respect to range of ascent of the cerebellar tonsils, the patients in Group 1 demonstrated a trend to greater ascent than those in Group 2 ($p = 0.07$, Table 1). Degree of ascent seemed more prominent when the caudal end of the cerebellar tonsils was located lower than C-4, but there was no relationship between degree of ascent and length of time after myelomeningocele closure.

In both groups, no complications related to surgery were observed. No CSF leakage or shunt infection was found during the initial 2 months after birth. Late complications related to VP shunts were observed in 2 infants. One required laparotomy due to ileus at the age of 3 months, and in another a shunt infection developed following a severe urinary tract infection at the age of 5 months. Another patient underwent tracheostomy for management of respiratory insufficiency due to the CM-II. All patients were alive and no patient was lost to follow-up during the follow-up period (> 18 months in all cases), but 2 of 3 patients who underwent decompressive surgery with some recovery from the preoperative condition have been admitted to the hospital for respiratory care.

TABLE 1

*Comparison of Groups 1 and 2**

Variable	Fetal Group	Postnatal Group	p Value
no. of patients	14	6	
gestational age (wks)			
range	35–39	39–41	
mean	36.4	39.3	
CM-II	13	5	0.52
symptomatic	8†	0	0.02
level of cerebellar tonsils			
range	C-2 to C-7	C-2 to C-4/5	
average	C-4/5	C-3	
ascent of cerebellar tonsils	11	4	0.65
range (no. of levels)	1–4	1–1.5	0.07
average (no. of levels)	2	1.1	
hydrocephalus Tx			
CSF reservoir	11	5	
VP shunt	12	5	0.68

* Values represent numbers of patients unless otherwise indicated.
† Three of these 8 patients required decompression surgery.

Illustrative Cases

Case 4

Ventriculomegaly was evident on fetal ultrasonography at 28 weeks' gestation in this case. Myelomeningocele was diagnosed by means of fetal MR imaging at 29 weeks' gestation, and the infant was delivered by elective cesarean section at 36 weeks' gestation. The fetal MR imaging study showed that the caudal end of the cerebellar tonsils was at C-4, but the postnatal MR images revealed that the cerebellar tonsils were at C-7. Myelomeningocele repair was carried out on Day 1, together with placement of a CSF reservoir, which was tapped 3 times before a VP shunt was placed. Follow-up MR imaging before shunt placement demonstrated elevation of the cerebellar tonsils from C-7 to C-4 and an enlarged subarachnoid space in the posterior fossa. The infant showed minor CM-II–related symptoms during the early postnatal period, but decompression surgery was not indicated. The MR images obtained before the infant was discharged from the hospital showed further ascent of the cerebellar tonsils to the C-3/4 level (Fig. 2).

Case 13

Ventriculomegaly was evident on fetal ultrasonography at 25 weeks' gestation in this case, and associated myelomeningocele was suspected. Fetal MR imaging performed at 27 weeks' gestation confirmed the presence of myelomeningocele and hydrocephalus. The infant was delivered by elective cesarean section at 37 weeks' gestation. Myelomeningocele repair and placement of a CSF reservoir were performed the next day. The initial level of the cerebellar tonsils was at C-7; they ascended to C-3/4 before the VP shunt was placed, but subsequently remained at the same level (Fig. 3). The infant later developed severe symptomatic CM-II, and a C1–5 osteoplastic laminoplasty was performed when she was 7 weeks old.

Case 14

Ventriculomegaly was evident on fetal ultrasonography at 18 weeks' gestation, and fetal MR imaging performed at 20 weeks' gestation revealed myelomeningocele with marked ventriculomegaly. A follow-up fetal MR imaging study at 32 weeks' gestation, however, demonstrated spontaneous regression of the ventriculomegaly, and the infant was born at 36 weeks' gestation by elective cesarean section. No ventriculomegaly was present at birth, and only myelomeningocele repair was performed on Day 1. Subsequently, hydrocephalus developed, and a VP shunt was placed on Day 13. Despite the fact that no CSF was removed during this period, the level of the cerebellar tonsils moved from C-6 at birth to C-4 before the VP shunt was inserted. Ascent of the cerebellar tonsils continued even after the VP shunt was inserted and reached C-3 by the time the infant was discharged from the hospital (Fig. 4). No evidence of symptomatic CM-II was observed subsequently.

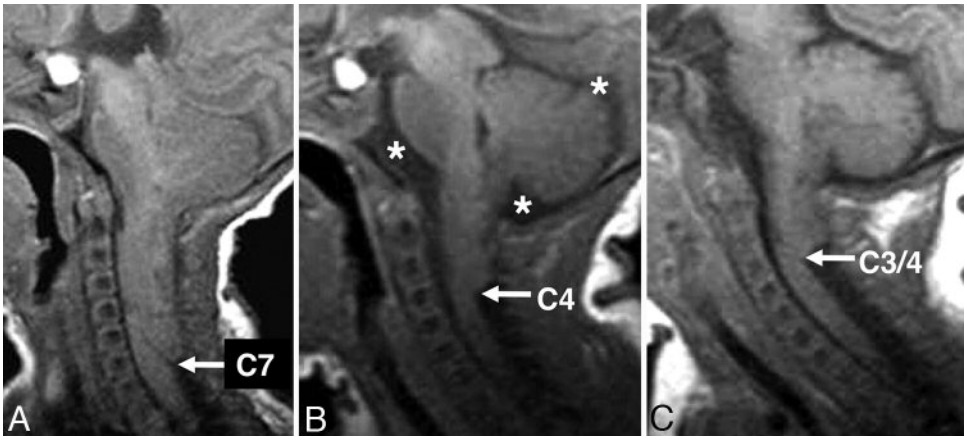

FIG. 2. Case 4. Magnetic resonance images. A: Postnatal, preoperative image obtained before myelomeningocele repair revealing the cerebellar tonsils at the C-7 level. B and C: Postoperative images obtained after myelomeningocele repair (B) and VP shunt placement (C) showing ascent to the C-4 and C-3/4 levels, respectively. Note that the subarachnoid space in the posterior fossa around the brainstem and the cerebellum is also enlarged *(asterisks)* after repair of the myelomeningocele. The patient had symptoms of CM-II, but no further intervention was required.

Discussion

Patients with myelomeningocele showed improvement in CM-II after postnatal myelomeningocele repair. The degree of ascent was larger in the patients in Group 1 than in those in Group 2. Ascent of the cerebellar tonsils continued even after placement of a VP shunt in most cases. Nevertheless, our policy of delivering patients with fetal myelomeningocele at ~ 36 weeks' gestation by elective cesarean section failed to prevent the development of symptomatic CM-II and hydrocephalus. Postnatal ascent of the cerebellar tonsils does not appear to be correlated with the development of symptomatic CM-II, which means that postnatal morphological improvement of CM-II does not imply functional improvement of the brainstem in some patients.

The clinical course of the Group 1 infants was more difficult than that of the Group 2. Patients in Group 1 tended to have more advanced, aggressive hydrocephalus, which is why the in utero diagnosis of myelomeningocele was made. Prominent CM-II was already present at the time of fetal diagnosis. In contrast, ventriculomegaly in Group 2 was not large enough to be detected on routine ultrasonographic examination during pregnancy;[1] thus, the associated CM-II was generally less severe in Group 2 than in Group 1, as shown in Fig. 1. Differences in the backgrounds of the associated pathological conditions appear to have led to the differences in the results between the 2 groups.

Reversibility

Chiari malformation Type II was thought to be irreversible until intrauterine repair of myelomeningocele was demonstrated to result in dramatic reduction of hindbrain herniation. Reversibility has not been expected following myelomeningocele repair after birth.[10,11] Tulipan et al.[10] demonstrated the impact of intrauterine myelomeningocele repair in terms of reduction of CM-II and shunt-dependent hydrocephalus, and he explained that restored hydrodynamic forces within the craniospinal axis play an important role in producing these results.[10,12] In addition, he suggested that these results could not be obtained in the full-term infant due to the different consistency of the brain and skull in these patients, together with the presence of hydrocephalus.[10,11] The present study clearly demonstrates that CM-II is revers-

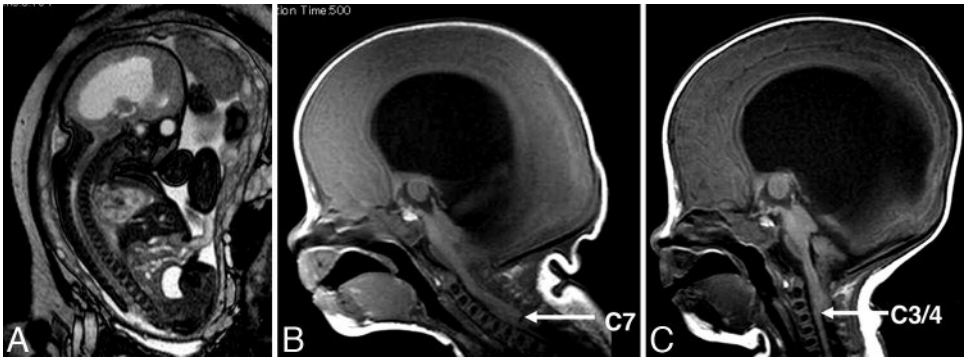

FIG. 3. Case 13. A: Fetal MR image obtained at 27 weeks' gestation showing a lumbosacral myelomeningocele with marked ventriculomegaly and CM-II. B: Postnatal, preoperative MR image obtained before myelomeningocele repair showing the cerebellar tonsils at the C-7 level. C: Postoperative MR image obtained after myelomeningocele repair showing ascent to the C-3/4 level. The cerebellar tonsils remained at the same level after VP shunt placement. Despite the improvement in hindbrain herniation, the patient required decompression surgery for severe symptomatic CM-II.

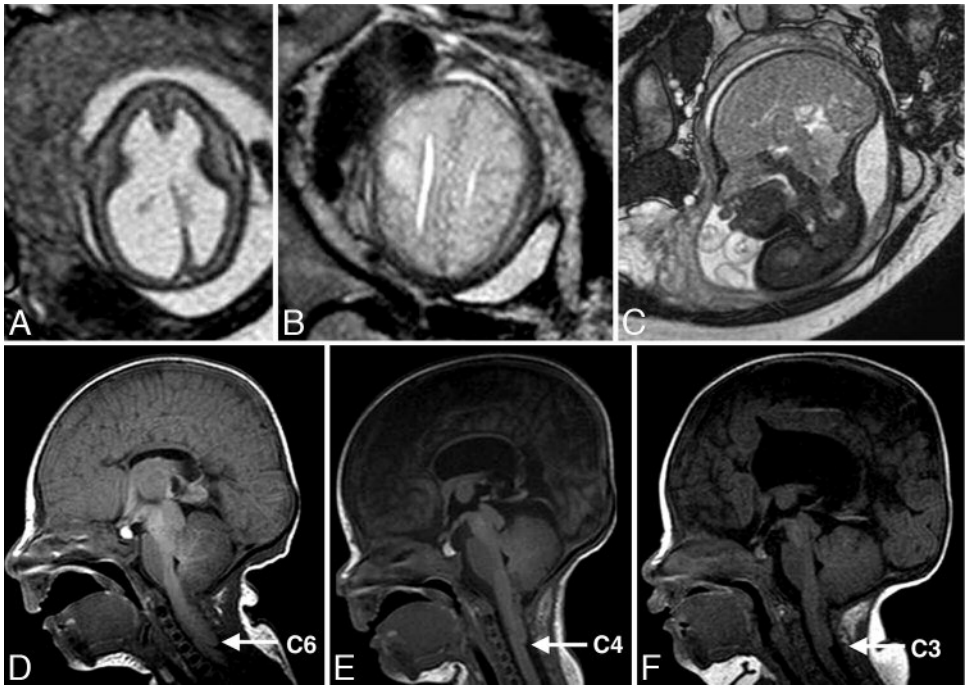

FIG. 4. Case 14. A–C: Fetal MR images. Ventriculomegaly was evident at 20 weeks (A) but had spontaneously disappeared by 32 weeks (B) when the CM-II was confirmed (C). D–F: Preoperative (D) and postoperative (E and F) postnatal MR images. The image obtained before myelomeningocele repair (D) shows the cerebellar tonsils at C-6. Postoperatively (E), they ascended to the C-4 level without any CSF aspiration. After VP shunt placement (F), they ascended to the C-3 level.

ible even with postnatal myelomeningocele repair. Based on McLone's unified theory, restored hydrodynamic forces after postnatal myelomeningocele repair push the herniated hindbrain structure back in the cranial direction, thus improving the anatomical level of the CM-II.[3,4]

Resolution of CM-II Symptoms

Despite the ascent of the cerebellar tonsils following postnatal myelomeningocele repair, resolution of symptoms related to CM-II does not always occur. The mechanism of the development of symptomatic CM-II has been attributed to 2 factors: direct mechanical compression of the brainstem by the herniated hindbrain structure and secondary degenerative change of the brainstem following chronic compression.[4] The secondary degenerative change may remain despite elimination of the direct mechanical compression. Unlike the situation following intrauterine myelomeningocele repair before 25 weeks' gestation, secondary degenerative change would play a more important role in the development of symptomatic CM-II in babies delivered after 36 weeks' gestation.

Influence of CSF aspiration

Aspiration of CSF appeared to have no significant effect on the ascent of the cerebellar tonsils. In Case 14, described above, ascent of the cerebellar tonsils was confirmed without CSF aspiration before the VP shunt was placed. It has been widely acknowledged that CSF drainage by means of a VP shunt improves CM-II–related symptoms.[7,9] Although intermittent aspiration of the CSF could temporarily delay the development of hydrocephalus in our case series, all patients in whom a CSF reservoir was placed eventually needed a VP shunt. In addition, the degree of tonsil ascent was greater after myelomeningocele repair than after VP shunt placement. The widespread effects of restored CSF circulation are also shown on the MR images obtained before the VP shunt was placed (Figs. 2B, 3C, and 4E), which demonstrate reexpansion of the subarachnoid space in the posterior fossa. These findings support the notion that the main factor responsible for CM-II ascent after postnatal myelomeningocele repair is the restored hydrodynamic forces following direct closure of the myelomeningocele.

Rationale for Intrauterine Surgery

Early delivery and early treatment of myelomeningocele, instead of controversial intrauterine myelomeningocele repair, showed no advantage in terms of reduction of symptomatic CM-II. Our policy is to deliver fetuses with myelomeningocele at 36 weeks' gestation, which is considered to be the earliest safe time for delivery based on fetal lung maturation. The message derived from this limited study is that postnatal myelomeningocele repair appears to be unable to change the current clinical course of CM-II. Management of fetal myelomeningocele is controversial and differs among countries.[5] If pediatric neurosurgeons wish to prevent serious, symptomatic CM-II, intrauterine myelomeningocele repair should be reconsidered.

Conclusions

Chiari malformation Type II may be partially reversed through postnatal repair of myelomeningocele. Early ascent

of the cerebellar tonsils suggests that the improvement was the result of restoration of the CSF circulation after myelomeningocele repair. Placement of a VP shunt also helped promote tonsillar ascent. Unlike intrauterine repair of myelomeningocele, however, postnatal repair of myelomeningocele did not prevent the development of symptomatic CM-II in the majority of the patients in whom myelomeningocele was diagnosed prenatally. Our limited experience implies that even though postnatal repair of myelomeningocele can improve the anatomical degree of CM-II, the preexisting brainstem dysfunction caused by long-standing brainstem compression can be irreversible in some patients when the baby is born after 36 weeks' gestation.

Disclaimer

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

References

1. Biggio JR Jr, Wenstrom KD, Owen J: Fetal open spina bifida: a natural history of disease progression in utero. **Prenat Diagn 24:** 287–289, 2004
2. Johnson MP, Sutton LN, Rintoul N, Crombleholme TM, Flake AW, Howell LJ, et al: Fetal myelomeningocele repair: short-term clinical outcomes. **Am J Obstet Gynecol 189:**482–487, 2003
3. McLone DG, Dias MS: The Chiari II malformation: cause and impact. **Childs Nerv Syst 19:**540–550, 2003
4. McLone DG, Knepper PA: The cause of the Chiari II malformation: a unified theory. **Pediatr Neurosci 15:**1–12, 1989
5. Oi S: Current status of prenatal management of fetal spina bifida in the world: worldwide cooperative survey on the medico-ethical issue. **Childs Nerv Syst 19:**596–599, 2003
6. Park TS, Hoffman HJ, Hendrick EB, Humphreys RP: Experience with surgical decompression of the Arnold-Chiari malformation in young infants with myelomeningocele. **Neurosurgery 13:** 147–152, 1983
7. Pollack IF, Pang D, Albright AL, Krieger D: Outcome following hindbrain decompression of symptomatic Chiari malformations in children previously treated with myelomeningocele closure and shunts. **J Neurosurg 77:**881–888, 1992
8. Sutton LN, Adzick NS, Johnson MP: Fetal surgery for myelomeningocele. **Childs Nerv Syst 19:**587–591, 2003
9. Tubbs RS, Oakes WJ: Treatment and management of the Chiari II malformation: an evidence-based review of the literature. **Childs Nerv Syst 20:**375–381, 2004
10. Tulipan N, Bruner JP, Hernanz-Schulman M, Lowe LH, Walsh WF, Nickolaus D, et al: Effect of intrauterine myelomeningocele repair on central nervous system structure and function. **Pediatr Neurosurg 31:**183–188, 1999
11. Tulipan N, Hernanz-Schulman M, Lowe LH, Bruner JP: Intrauterine myelomeningocele repair reverses preexisting hindbrain herniation. **Pediatr Neurosurg 31:**137–142, 1999
12. Tulipan N, Sutton LN, Bruner JP, Cohen BM, Johnson M, Adzick NS: The effect of intrauterine myelomeningocele repair on the incidence of shunt-dependent hydrocephalus. **Pediatr Neurosurg 38:**27–33, 2003

Manuscript submitted March 13, 2008.
Accepted May 23, 2008.

Address correspondence to: Nobuhito Morota, M.D., Department of Neurosurgery, National Children's Medical Center, National Center for Child Health and Development, 2-10-1 Okura, Setagaya, Tokyo 157-8535, Japan. email: morota-n@ncchd.go.jp.