Progressive spasticity and scoliosis in children with myelomeningocele

Radiological investigation and surgical treatment

T. S. Park, M.D., Wayne S. Call, M.D., William M. Maggio, M.D., and Diane C. Mitchell, M.D.

Department of Neurosurgery and Division of Neuroradiology, University of Virginia School of Medicine, Charlottesville, Virginia

Seventeen myelodysplastic patients with progressive extremity spasticity and scoliosis underwent radiological evaluation and surgical treatment. All but one were under 18 years of age at the time of surgical treatment. Duration of the clinical presentation ranged from 1 to 7 years. Metrizamide was instilled into the subarachnoid space in 12 patients, the lateral ventricle in two, and the hydromyelic cavity in three. Sequential computerized tomography scanning after intrathecal instillation of the contrast material clearly demonstrated hydromyelia in nine patients and compression of the brain stem in five. Posterior fossa decompression with plugging of the obex was performed in 12 patients, posterior fossa decompression alone in three, and ventriculoperitoneal (VP) shunting procedures in two. Of the 12 patients who underwent the obex plugging procedures, eight have shown partial or complete resolution of spasticity and an increase in motor strength with no significant postoperative complications. In contrast, posterior fossa decompression or VP shunting procedures alone have not led to a favorable neurological outcome. Hydromyelia may occur more commonly among myelodysplastic patients than previously recognized and may be treated most effectively by the obex plugging procedure.

Key Words • brain stem compression • hydromyelia • myelomeningocele • obex • scoliosis • spasticity • posterior fossa decompression

Spinal deformities occur in nearly 90% of survivors of myelomeningocele, causing functional decline in their later lives. Scoliosis is the most common spinal deformity, with congenital anomalies of the vertebrae, muscle paralysis, muscle malposition, and pelvic obliquity implicated as etiological factors. Of particular significance is the fact that some myelodysplastic children develop scoliosis without musculoskeletal abnormalities. The scoliosis in these children is typically located proximal to the level of the myelomeningocele, and progresses if untreated. This particular type of scoliosis occurring in myelodysplastic patients has been described as "idiopathic scoliosis" in comparison with congenital and developmental scoliosis. Since myelomeningocele is associated with diverse anomalies involving the entire neural axis, it is conceivable that neurological disorders account for idiopathic scoliosis, but these have as yet received little investigation.

Hall, et al.,12-14 demonstrated radiographically that hydromyelia noted in myelodysplastic patients communicates with the ventricle, and they therefore advocated a ventricular shunting procedure. By contrast, Hoffman, et al.,17 reporting on a group of myelodysplastic children in whom neurological deterioration was thought to be due to compression of the brain stem within the cervical canal, found that the neurological deficits of their patients resolved after decompressive laminectomy.

We have also encountered a distinct group of myelodysplastic patients whose neurological deficits are characterized by progressive spastic extremity weakness and scoliosis. Our experience with the management of these patients, however, has revealed that hydromyelia and compression of the brain stem are solely or concurrently responsible for the delayed neurological deterioration. Furthermore, hindbrain decompression combined with plugging of the obex has led to a favor-
Summary of Cases

Clinical Material

This series consists of 17 myelodysplastic patients who underwent surgical treatment for progressive scoliosis and spasticity. Sixteen of these were children under 18 years of age and one was an adult aged 32 years at the time of surgical treatment. In all patients, the myelomeningocele had previously been surgically repaired. The myelomeningocele was located in the cervical region in one patient, the thoracic region in one, the thoracolumbar region in four, the lumbar region in five, and the lumbosacral region in six.

All patients except two had ventricular shunting systems in place. Twelve patients had functioning shunts, and three had nonfunctioning shunts. The duration of shunt malfunction in the three patients was unclear, but presumed to be more than a year. Of the 12 patients with functioning shunts, 10 had normal or small lateral ventricles, one had mild dilatation of the lateral ventricles, and another had small lateral ventricles but a dilated fourth ventricle. Of the five patients with no shunt or a nonfunctioning shunt, four had moderate to marked dilatation of the lateral ventricle and one had normal ventricles.

All patients except three had developed scoliosis of between 15° and 75° proximal to the level of the myelomeningocele. Two patients had already undergone spinal fusions. Fifteen patients showed spasticity of the arms. Eleven also had progressive weakness of the arms and atrophy of the intrinsic hand muscles. Of the four patients who had spasticity without obvious arm weakness, one complained of recurrent suboccipital headaches and the remaining three patients exhibited scoliosis and hyperreflexia. Age at onset of the scoliosis or arm weakness ranged from 1 to 19 years, and duration of the clinical presentation ranged from 1½ to 7 years. The clinical course in all patients was characterized by slow but relentless deterioration.

Radiological Investigations

The neuroradiological examinations obtained on these patients consisted of cranial computerized tomography (CT) without contrast enhancement and sequential metrizamide CT myelography. The examinations were aimed at investigation of the following: 1) severity of ventriculomegaly; 2) lowermost level of the cerebellar tonsils or medulla; 3) compression of the cerebellar tonsils and brain stem; and 4) existence of hydromyelia.

For metrizamide CT myelography, contrast was instilled into the spinal subarachnoid space in 12 patients, the lateral ventricle in two, and the hydromyelic cavity in three. Sequential CT examinations were obtained immediately and 5 to 8 hours after intrathecal instillation of the contrast medium. Two patients were subjected to a third scanning, 12 and 18 hours after the injection, respectively. Axial and sagittal scanning was performed at 3- to 10-mm intervals at the levels of the posterior fossa and cervical spine. Additional scanning of the spinal cord was then performed as necessary.

The CT scans obtained immediately after intrathecal injection of metrizamide clearly visualized caudal displacement of the cerebellar tonsils and medulla in all patients. This was especially well demonstrated by sagittal reformatted images (Figs. 1 and 2). These initial scans also proved useful in demonstrating compression of the cerebellar tonsils and brain stem within the cervical canal. The compression was evidenced in some

Fig. 1. Computerized tomography scans after intrathecal injection of metrizamide in a child with recurrent suboccipital headaches and hyperreflexia. Sagittal reformatted image demonstrates caudal displacement of the cerebellar tonsils to the C-3 level. The subarachnoid space dorsal to the cerebellar tonsils is completely obliterated.
cases by complete obliteration of the subarachnoid space posterior to the displaced cerebellar tonsils. On this initial CT scan, however, metrizamide did not enter the hydromyelic cavity.

Repeat CT scanning 5 to 8 hours after metrizamide injection effectively visualized hydromyelia. Typically, the contrast medium delineated both the subarachnoid space and dilated central canal with the intervening spinal cord tissue appearing as a ring of relatively low density (Fig. 3). It is of note that hydromyelia was distinctly less prominent in the cervical spinal cord than in the thoracic and lumbar regions of the spinal cord. In cases where the hydromyelic cavity was filled by metrizamide following intrathecal instillation, the fourth ventricle was also filled by the contrast material. In three patients, there was evident continuation of metrizamide from the fourth ventricle to the hydromyelic cavity. Two patients were subjected to CT scanning 12 and 18 hours after injection because the CT scan obtained 6 hours after the injection failed to show the hydromyelic cavity. In neither case was hydromyelia demonstrated.

In three patients, contrast material was inadvertently instilled into the hydromyelic cavity (Fig. 4). Extensive hydromyelia noted in these patients involved the entire spinal cord and opened into the fourth ventricle. Of two patients who received intraventricular administration of the metrizamide through shunt reservoirs, only one demonstrated contrast material within the dilated central canal (Fig. 5).

Operative Management and Findings

Posterior fossa decompression with plugging of the obex was performed in 12 patients, posterior fossa

Fig. 2. Computerized tomography scan obtained immediately after intrathecal injection of metrizamide. The cerebellar tonsils are clearly visualized at the C-5 level and the cervical cord is flattened.
decompression alone in three, and establishment of patent ventriculoperitoneal (VP) shunts in two. Prior to plugging of the obex, we ensured that the VP shunts were functioning in all patients. Posterior fossa decompression was performed with the patients in the prone position under controlled ventilation. Care was taken not to hyperflex the neck. Through a midline incision, the occipital bone and the upper cervical laminae were exposed. The arch of the atlas was frequently bifid. A space between the posterior rim of the foramen magnum and the arch of the atlas was always prominent. A burr hole was drilled into the occipital bone close to the rim of the foramen magnum. A small portion of the occipital bone was removed, after which cervical laminectomy was performed to the lowest level of the cerebellar tonsils or the medullary kink. This required the cervical laminectomy to be carried as low as the C-5 level in our series.

In all patients, there was thickening of the atlanto-occipital membrane, constricting the dural tube to a varying degree. In two cases, this peculiar variation of the normal anatomic structure had produced a visible dent on the underlying cerebellar tonsils (Fig. 6). Division of the atlanto-occipital membrane led to immediate expansion of the dural sac. A dural incision was then made in the cervical region and extended rostrally to the level of the transverse sinus, which was noted to be low in these patients. Particular caution was observed not to open the transverse sinus. The arachnoid was markedly thickened in all patients, which in some cases made it difficult to identify the underlying structures until the arachnoid was opened. It is of note that when the brain stem was free from compression, the thickened arachnoid was not adherent to the cerebellar tonsils or brain stem. All patients showed caudal displacement of the cerebellum and brain stem, but the caudal level of these structures was not necessarily identical. The cerebellar tonsils were densely adherent to the underlying brain stem and, consequently, the outlet of the fourth ventricle was invariably obliterated (Fig. 7 right). However, in some cases, CSF was found to flow through the incompletely obstructed outlet into the subarachnoid space. The choroid plexus frequently protruded into the subarachnoid space between the medulla and the displaced cerebellar tonsils. The caudal loop of the posterior inferior cerebellar artery was displaced downward and located over the cerebellar tonsils or medulla within the spinal canal. When decompression of the hindbrain alone was necessary, the dura was patched with lyophilized dura. The wound was then closed.

In cases where plugging of the obex was planned, the operating microscope was swung over the operative field as soon as the dural incision was completed. A...
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FIG. 5. Computerized tomography scans after intraventricular injection of metrizamide via a shunt outlines the hydromyelic cavity within the cervical cord.

longitudinal incision was then made in the arachnoid to facilitate careful examination of the cerebellum and brain stem. Even with the aid of the preoperative CT scans, intraoperative localization of the fourth ventricle was often difficult. We always made a longitudinal midline incision over the distal portion of the adherent cerebellar tonsils (Fig. 7). No attempt was made in any case to dissect the cerebellar tonsils of the brain stem. It was critical to carry out dissection precisely along the midline until the fourth ventricular cavity was entered. The fourth ventricle was recognized by the characteristic appearance of the floor of the midline fissure and adjacent hypoglossal trigone.

Once the floor of the fourth ventricle had been identified, the midline incision was extended caudally

FIG. 7. Left: Drawings of the obex plugging procedure showing dissection along the midline cerebellar tonsils. Right: Operative photograph showing exposure of the fourth ventricle through a midline incision over the densely adherent cerebellar tonsils.

FIG. 6. Operative photograph showing the thickened arachnoid and cerebellar tonsils below the foramen of magnum (asterisk). There is an indentation of the cerebellar tonsils and brain stem at the C-1 level (arrow).
to the level of the obex. In all cases, the caudal portion of the fourth ventricle was markedly waisted. A blunt right-angle dissector was then passed into the central canal. During the procedure, we observed in some patients that CSF was trickling from the central canal into the fourth ventricle. The central canal was often sufficiently enlarged to accommodate a 2-mm dissector without adverse effects. A small piece of muscle or fat was then pushed gently into the dilated central canal. Occasionally, transient bradycardia was associated with this procedure. The dura was closed with a graft of lyophilized dura.

Of the three children who underwent posterior fossa decompression alone without plugging of the obex, two had evidence of brain-stem compression. The remaining patient had severe ventriculomegaly that precluded the ventricular shunting procedure. She therefore underwent decompressive procedures and the hydromyelia was drained through myelotomy of the thoracic cord.

Results of Surgical Treatment

Table 1 shows the results of surgical treatment in our 17 patients. The follow-up period ranged from 2 months to 2 years. Of the 12 patients who had undergone decompression of the hindbrain and plugging of the obex, eight have shown improvement of the arm weakness and four have remained neurologically unchanged. One patient in whom plugging of the obex had led to complete resolution of arm weakness subsequently died of pneumonia 15 months after surgery. The evident improvement of motor weakness was noted as early as 5 days postoperatively. Decrease in severity of the spasticity was concomitant with improved strength. Three patients who had functionless arms prior to surgery regained motor function of the hands postoperatively. The improved motor strength has been sustained in all patients. However, none of these patients have shown reversal of the spinal deformity to a noticeable degree, despite the improvement of arm weakness and spasticity. Two of the four patients who have made no neurological recovery postoperatively underwent the surgery only recently. Another child had normal arm function preoperatively, but exhibited thoracic scoliosis (31°) and lumbar lordosis (90°). The lumbar lordosis was subsequently treated by lumbosacral spinal fusion and the thoracic scoliosis has remained unchanged and untreated.

Decompressive cervical laminectomy and limited occipital craniectomy relieved suboccipital pain and decreased spasticity in one child. In another child, the decompressive procedure was combined with myelotomy for drainage of the hydromyelia. Her arm weakness improved somewhat, but the 15° thoracic scoliosis has remained unchanged. The remaining child developed entrapment of the fourth ventricle following hindbrain decompression alone, which has resulted in severe neurological deterioration. The two children whose hydromyelia was treated only by VP shunting procedures had made no neurological improvement in a postoperative period of 8 and 18 months, respectively.

The plugging of the obex performed in 12 patients was not complicated by morbidity of any significance. However, aseptic meningeal reaction (six patients), minor upper gastrointestinal bleeding due to stress ulcer (two patients), and recurrent vomiting (three patients) occurred after surgery.

Discussion

Despite the high incidence of progressive spinal deformities well recognized in the myelodysplastic population, the pathological abnormalities of the spinal cord responsible for these spinal deformities have not been studied thoroughly. This is largely due to the overwhelming complexity of the orthopedic problems suffered by all patients with myelomeningocele. In addition, neuroradiological examinations available prior to the CT era could detect only severe pathological changes involving the spinal cord. During the last 2 years, we have performed sequential metrizamide CT myelography on patients with myelomeningocele who had already developed scoliosis and marked spasticity of the upper extremities. To our amazement, the CT myelography revealed hydromyelia and/or brain-stem compression in all patients. This has led us to suspect that hydromyelia is more common than has previously been recognized as a cause of the gradual functional decline of myelodysplastic patients in later life.

The term “hydromyelia” was coined by Stilling in 1859. Association of hydromyelia with spinal bifida was described by Morgagni as early as 1761. Normally between the 4th and 5th week of embryonal life, the neural tube closes to form a single cavity that consists of the ventricle and the central canal of the spinal cord. By the 6th week, fluid secreted by the neuroepithelium and choroid plexus accumulates within the cavity and filters through the primitive roof of the fourth ventricle. The filtration pressure causes a generalized distention of the neural tube described as embryonal hydrocephalomyelia. By the 8th week, the foramina of Magendie and Luschka are formed, and the CSF flows into the subarachnoid space. As a result of this bypass, the central canal gradually narrows to become a vestigial structure. Thus, if the rhombic roof remains less
permeable than normal, an excessive amount of fluid will be retained within the neural tube. Since myelomeningocele is an intrauterine disease and is consistently associated with obliteration of the fourth ventricular outlet, infants with myelomeningocele are likely to have an enlarged central canal of the spinal cord. In fact, a high incidence of hydromyelia among this group of infants has been noted in pathological examination. Cameron found hydromyelia in 21 of 26 myelodysplastic infants. MacKenzie and Emery also reported hydromyelia in 48 of 100 myelodysplastic infants. Other investigators have demonstrated that in these infants the dilated central canal communicates with the ventricles. Furthermore, a majority of these infants are born with a patent aqueduct of Sylvius. A majority of these infants have been noted in pathological examination. Cameron found hydromyelia in 21 of 26 myelodysplastic infants. MacKenzie and Emery also reported hydromyelia in 48 of 100 myelodysplastic infants. Other investigators have demonstrated that in these infants the dilated central canal communicates with the ventricles. Furthermore, a majority of these infants are born with a patent aqueduct of Sylvius.

Multiple factors will conceivably influence the eventual outcome of the dilated central canal in their later life. Although it is not always possible to distinguish syringomyelia from hydromyelia, the distinction is particularly important when considering the etiology of the two conditions. Like others, we define "hydromyelia" as a dilated central canal filled with CSF that has been or is continuous with the ventricle. Hydromyelia is typically seen in myelodysplastic patients with an Arnold-Chiari Type II malformation. A number of hypotheses have been proposed to explain the mechanisms. To explain development and progression of syringomyelia, Gardner modified and revised the hydrodynamic theory that originates from Morgagni. According to this theory, the arterial pulsation of the choroid plexus generates a pulse wave in the ventricle that is transmitted downward into the central canal, producing a water-hammer effect and consequent enlargement of the central canal. The role of this hydrodynamic theory in the pathogenesis of syringomyelia has been seriously questioned. Nevertheless, this theory has brought to our attention the existence of persistent circulation of CSF through the central canal in patients with myelomeningocele.

Williams has introduced a concept of craniospinal pressure dissociation. He found that, in patients with Arnold-Chiari malformation, the cisternal pressure in the posterior fossa was elevated after coughing, while the lumbar pressure fell. This pressure dissociation disappeared after posterior fossa decompression or ventricular shunting. Williams attributed this pressure dissociation to a valve-like action of the Arnold-Chiari malformation which allows upward flow of CSF from the spinal canal into the basal cisterns of the posterior fossa and subsequent intracranial entrapment of the fluid. He contended that this craniospinal pressure dissociation is initially responsible for displacement of CSF into the central canal through the fourth ventricle.

Du Boulay, et al., agreed with Williams that craniospinal pressure dissociation does occur in Arnold-Chiari malformation; however, they held a different view in regard to underlying mechanisms, based on two important observations. First, in patients with Arnold-Chiari malformation, Pantopaque entered the fourth ventricle during myelographic examination. Second, during systole in normal individuals, CSF was displaced into the spinal canal from the basal cisterns of the posterior fossa volumetrically 10 times greater than that displaced from the ventricle. They speculated that narrowing at the foramen magnum in cases of Arnold-Chiari malformation interferes with the egress of CSF during systole from the cranium into the spinal canal and, as a result, pressure within the posterior fossa rises to a greater degree than that within the spinal canal. Irrespective of the underlying mechanism, entrapment of CSF and the resulting preferential increase in pressure within the posterior fossa is significant. Ball and Dayan contended that syringomyelia is produced by CSF forced into the spinal cord from the subarachnoid space along the Virchow-Robin space. Similarly, Aboulker believed that stenosis at the foramen magnum causes CSF to filter through the spinal cord parenchyma or via a pathway along the posterior roots. Results of CT studies in our patients support the contention that the CSF filling the dilated central canal arises from the lateral ventricle or the spinal subarachnoid space and flows through the fourth ventricle. However, we are unable to comment on the possibility of CSF tracking into the central canal through the spinal cord.

Performing CT immediately after metrizamide injection effectively demonstrates the caudal level of the cerebellar tonsils and medulla. It also visualizes the compression of the brain stem. Therefore, the initial CT examination is safely limited to the cervical and upper thoracic regions. In our experience, conventional myelography provides little additional information in this regard. Delayed CT scans performed 5 to 8 hours after the injection has proved most useful in the diagnosis of hydromyelia. Sagittal reformation of this delayed CT scan will delineate the contrast material within the fourth ventricle and the central canal. In agreement with Gardner's description, the hydromyelia noted in our patients was most extensive in the thoracic and lumbar regions. However, expansion of the spinal cord was not a striking feature of hydromyelia. In fact, the spinal cords of all patients except one were smaller than normal. Presumably the small spinal cord reflects diminished intramedullary fiber tracts secondary to myelomeningocele and a relatively minimal distending force on the central canal.

Figure 8 depicts our proposed approach for management of myelodysplastic patients who have shown clinical evidence of hydromyelia. This approach requires establishment of a functioning shunt. The rationale for the shunting procedure as the initial step of management is that hydromyelia represents a diversion for ventricular CSF and plays an important role in the compensation of hydrocephalus. Therefore, abrupt occlusion of the hydromyelic cavity by Pantopaque or surgical ligation in the absence of a functioning shunt can cause acute decompression of hydrocephalus resulting in death.
oblation of the central canal. The shunting procedure alone, however, may be performed in patients with minimal neurological deficits. Close postoperative follow-up observation of the patients will avoid delay in detection of neurological deterioration.

Conclusions

Hydromyelia and/or compression of the brain stem caused progressive spastic extremity weakness and scoliosis in this series of 17 myelodysplastic patients. These pathological abnormalities may occur solely or concurrently, to account for the delayed neurological deterioration. The clinical course of the disorders was characterized by slow but relentless neurological deterioration. Metrizamide CT myelography accurately visualized hydromyelia and compression of the brain stem. In the present series of myelodysplastic patients, decompression of the hindbrain with plugging of the obex has led to a favorable outcome without causing significant postoperative complications.

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References

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