Distal cordectomies as treatment for lumbosacral myelomeningoceles

Clinical article

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Object. The purpose of this study was to evaluate the effect of distal cordectomies on motor function in the lower extremities of infants with lumbosacral myelomeningoceles.

Methods. Medical records were reviewed in 32 infants with lumbosacral myelomeningoceles who were 1 day to 7 months old, who had some lower-extremity function preoperatively, and who were treated by distal cordectomies, dividing the cord between its exit from the intact spinal canal and the neural placode. Neurological function was evaluated before and after operation by therapists who were unaware of the surgical technique.

Results. Neurological function was unchanged after operation in 29 of 32 cases. For the others, hip flexion, foot plantar flexion, and toe movement were lost in 1 case each.

Conclusions. The majority of infants treated by distal cordectomies retain preoperative motor function after operation.

(key Words: myelomeningocele, spina bifida, cordectomy, congenital)

Publications about myelomeningocele (MMC) closure techniques generally recommend preservation of the neural placode as well as nerves entering and exiting the placode, because of the postulate that the placode has neurological function. Myelomeningoceles in infants have rarely been treated with cordectomies, in which the distal spinal cord is divided proximal to the placode and the placode and its exiting nerves are resected, although cordectomies have been done in paraplegic children with recurrent MMC tethering, syringomyelia, and spasticity. Incisions were made at the junction of normal and abnormal skin around the periphery of the MMC. Subcutaneous tissues were opened down to the fascia and dissected medially to the dura mater, and then around the periphery of the dural sac (Fig. 1). Normal dura was opened in the midline at the site where the dural sac exited from the intact spinal canal, or transversely just proximal to the dura-placode junction. If normal dura was not evident at the cephalic end of the exposure, a laminotomy was performed to access normal dura. The distal normal spinal cord was followed caudally toward the placode and was transected either at its junction with the placode or distal to the lowest exiting normal-appearing nerve roots (Fig. 2). Electrophysiological stimulation/monitoring was not available at the time.

Nerves exiting the placode were divided where they entered the dura, then the placode and exiting nerves were removed. The dura was closed proximally around the transected cord with 5-0 Prolene suture, and then the skin was undermined laterally to the midaxillary line and closed in 1 or 2 layers.

Results

Thirty-two infants were identified in whom both preoperative and postoperative lower-extremity movement had been evaluated by pediatric therapists and who had been treated with distal cordectomies. Their ages ranged from 1 day to 7 months old, with a median age of 2.5 months. Preoperatively, 24 infants had some lower-extremity function, while 8 had no function. After operation, 29 of 32 infants maintained preoperative motor function. In 1 case each, hip flexion, foot plantar flexion, and toe movement were lost. These findings suggest that distal cordectomies are a viable option for treating MMCs with lower-extremity function.
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from the day of birth to 7 months, with a median of 7 days and a mean of 17 days.

Prior to the operation all 32 infants had hip flexion and 26 had hip extension; 29 had knee flexion and 28 had knee extension; 18 had ankle dorsiflexion and 4 had ankle plantar flexion; and 2 each had toe dorsiflexion and toe plantar flexion.

After the operation, 29 of 32 infants had no loss of motor function. Of note, no child who had hip flexion, hip extension, knee flexion, and knee extension but no movement distal to the knee preoperatively had any loss of motor function postoperatively. Of the 3 infants whose function was apparently worsened after operation, 1 had only hip flexion preoperatively and it was not evident postoperatively. One infant who had both ankle dorsiflexion and ankle plantar flexion preoperatively did not have ankle plantar flexion postoperatively. In 1 infant, toe flexion and extension that were observed preoperatively were not evident postoperatively.

Typical short-term complications occurred in 9 infants: 3 had wound dehiscence; 4 had wound dehiscence and infection; 1 had wound dehiscence, infection, and a CSF leak; and 1 had a CSF leak only.

Seven infants had clinically evident hydrocephalus before operation; postoperatively, 1 of the 7 did not have hydrocephalus and the hydrocephalus status of the other infants is unknown. Of the 25 infants who did not have clinically significant hydrocephalus preoperatively, postoperative data were available for 15 (10 had not returned for follow-up); 4 of the 15 infants developed hydrocephalus postoperatively.

Discussion

The development of distal cordectomy procedures in our hospital began with paraplegic infants with MMCs who were treated with a distal cordectomy, dividing the spinal cord at the level where it exited the intact spinal canal. That procedure facilitated skin closure and kyphectomies after the placode was removed, and had the potential to decrease the chance of development of a tethered spinal cord. The procedure was extended to infants with MMCs who had hip function, with or without knee extension, and we observed preservation of preoperative function. Since then, some infants with function further distally have had distal cordectomies, with the results noted above.

We were unable to find articles about distal cordectomies in infants with MMCs who were not paraplegic, although cord transections have been performed as a component of kyphoscoliosis treatment in those who are paraplegic.6

In a series of 10 patients who underwent cordectomies for syringomyelia, spasticity, and pain, 2 children had cordectomies in conjunction with treatment of thoracic kyphosis.5 The authors described the pathological findings in one of the MMC specimens as follows: “The resected neural placode was available for pathological examination in only one patient…. The specimen was a closed neural placode, 5 cm long, 3.5 cm wide, and 1 cm in anteroposterior diameter. The center of the specimen contained a large cyst lined with a glistening membrane consisting of dense leptomeningeal fibrosis, presumably infolded when the myelomeningocele was originally repaired. Other features of this specimen included a dermoid cyst, gliosis, and the fact that only an occasional neuron remained.” The observation that “only an occasional neuron remained” correlates with the general preservation of motor function after removal of the placode.

Blount et al.7 reported the effectiveness of spinal cord transection for definitive untethering in 14 patients with recurrent tethered cord, 13 of whom had experienced multiple episodes of retethering. These practitioners divided the cord at the most distal normal segment above the placode in children with extremely impaired motor and sphincter function. They noted that neurosurgical reservations about the procedure included the aesthetic aspect of dividing the spinal cord and the hope that stem cell research or other technologies might be developed in the future.
McLaughlin et al. performed distal cordectomies in conjunction with intraspinal rhizotomies in 7 paraplegic children with MMCs to treat spasticity of the lower extremities effectively, dividing the cord between the scarred conus medullaris and the normal-appearing spinal cord.

The effects of cordectomies on bladder function in older children with MMCs who were undergoing kyphoscoliosis treatment have been reported. In the urological literature, Linthorst et al. compared the post-operative bladder function of 7 children with MMCs to that in 13 controls and found improved bladder compliance and capacity, with no deleterious effects on bladder function. Lalonde and Jarvis performed urodynamic studies before and after cordotomy in conjunction with correction of kyphosis in 9 children with MMCs. They found improved bladder capacity and compliance in 8 of 9 patients, and an increase in urethral pressure in 5 of 9. Bladder function deteriorated after operation in 1 patient.

Although our results indicate loss of function in 3 patients, those data reflect the evaluation by therapists, and differed from our findings on examination of 2 of those 3 infants: Case 3, who was thought by the therapist to have hip flexion (only), was thought by us to have no lower-extremity function, and the patient who was thought by the therapist to have lost ankle plantar function postoperatively was thought by us to have had no ankle dorsiflexion or ankle plantar flexion preoperatively. Data from the pediatric therapists were used as the basis for this manuscript because their data lacked the bias about motor outcomes that operating neurosurgeons would potentially have. If our evaluations had been used, only 1 of 32 infants would have been considered to have any decrement in postoperative function.

Distal cordectomies may be particularly indicated in developing countries, where MMC closures are frequently performed by non-neurosurgeons. Distal cordectomies facilitate wound closure by removing the bulk of the neural placode and may facilitate dural closure. They may also lessen the development of tethered spinal cords, which are far harder to diagnose in developing countries.

Neurosurgeons have preserved the neural placode for multiple reasons, primarily because it has been considered to have, or potentially have, some neurological function. Stark and Drummond stimulated the neural placode in 75 infants with MMCs shortly after birth, before closure of the MMC. They stimulated the neural placode with bipolar electrodes, 0.5 cm in diameter and 1 cm apart, and evaluated the responses of 11–13 lower-extremity muscles visually and by palpation. Placode stimulation resulted in contraction of approximately 80% of lower-extremity muscles and confirmed neural continuity between the placode and these muscles.

Reigel et al. evaluated somatosensory evoked potentials in 7 newborn infants with MMCs. When the peroneal nerve was stimulated, potentials were recorded from the neural placode in 5 of 7 cases and from the scalp in 2 of 7, with monophasic, long-duration responses. When the neural placode was stimulated, scalp responses were recorded in 5 of 7 cases. The authors concluded that sensory nerves to and from the neural placode are intact and they recommended meticulous preservation of the neural placode during repair of MMCs.

More recently, in 2 neonates with MMCs, Pugh et al. stimulated nerves exiting the neural placode and evoked electromyographic responses in lower-extremity muscle groups. They also stimulated the placode and observed “an alternating pattern of activity” in the legs.

It seems clear that nerves exiting the neural placode innervate lower-extremity muscles. It seems there is some neural connection between the neural placode itself and lower-extremity muscles. But what seems to be unknown is whether there is a connection of motor pathways within the spinal cord proximal to the placode and nerves exiting from the placode to innervate lower-extremity muscles. The results presented above suggest there is little such connection. Neurophysiological studies are underway to evaluate the question.

Conclusions

In infants with MMCs and some lower-extremity motor function, distal cordectomies—dividing the spinal cord proximal to the neural placode and excising the placode and its exiting nerve roots—are usually associated with no loss of motor function in the lower extremities. The procedure facilitates wound closure, may be associated with a reduced incidence of tethered spinal cord, and seems to be particularly appropriate in developing countries.

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Disclosure

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