Development of syringohydromyelia associated with Dandy-Walker malformation: treatment with cystoperitoneal shunt placement

Case report

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Dandy-Walker malformation (DWM) is a well-described clinical entity, which includes vermian agenesis, posterior fossa cysts, and frequently, hydrocephalus. The authors report the clinical course and present the radiographic findings pertaining to a 1-month-old girl with DWM who was treated initially with a ventriculoperitoneal shunt and endoscopic fenestration of a posterior fossa cyst. After decompression for hydrocephalus, an increased mass effect at the foramen magnum from her posterior fossa cyst was demonstrated, as well as subsequent development of syringohydromyelia from C-4 to T-7. She was treated with a cystoperitoneal shunt. At the 6-month follow-up examination, the child (15 months of age) had achieved gains in developmental milestones, and complete resolution of the syrinx was established through MR imaging. This is the fourth nonautopsy pediatric case of DWM-associated syringohydromyelia reported in the literature, and the third in a child to demonstrate impaction of the posterior fossa cyst at the foramen magnum leading to syrinx formation with subsequent treatment and resolution. Spinal imaging may be useful in the evaluation of patients with DWM who do not experience expected improvement after shunt procedures.

Key Words • syringomyelia • hydromyelia • syrinx • Dandy-Walker malformation • shunt • pediatric neurosurgery

Abbreviations used in this paper: CSF = cerebrospinal fluid; DWM = Dandy–Walker malformation; MR = magnetic resonance; VP = ventriculoperitoneal.
Postoperative Course. On postoperative imaging performed when the patient was 5 months of age, displacement of the cyst into the foramen magnum and an increased mass effect on the brainstem could be discerned (Fig. 2A). Cystoperitoneal shunt placement was recommended, but because there was no apparent symptomatology, this procedure was deferred. Follow-up MR imaging at 9 months demonstrated increased mass effect at the cervicomedullary junction, as well as interim development of a noncommunicating syrinx from C-4 to T-7 (Fig. 3A). The syrinx measured 8 mm at its greatest diameter.

Second Operation. On the hypothesis that the syringohydromyelia was caused by abnormal CSF flow dynamics at the foramen magnum resulting from the mass effect of the cyst, we placed an independent cystoperitoneal shunt.

Second Postoperative Course. Postoperatively, the cyst was demonstrated to be well decompressed and the syrinx had collapsed. The patient experienced a gain in developmental milestones after placement of the second shunt. At the 6-month follow-up examination when the patient was age 15 months, MR imaging findings were normal, with no recurrence of the syrinx (Fig. 3B).

Discussion

There have been 20 reports of syringohydromyelia associated with DWM, including Gardner’s original description in 1957.1,3-10,16,17,21,23-25 Reported symptoms include upper-extremity pain and weakness,1 gait disturbance,1 paralysis,24 and headache.7 Surgical strategies that have been used for...
the treatment of DWM-associated syringohydromyelia include posterior fossa decompression,\textsuperscript{3,5,6,21} plugging the obex with muscle,\textsuperscript{1} VP shunt revision,\textsuperscript{23} cystoperitoneal shunt revision,\textsuperscript{7} and placement of syringoperitoneal shunts.\textsuperscript{4}

The role of posterior fossa cyst impaction into the foramen magnum as the mechanism of syrinx formation in DWM (which is analogous to the development of a Chiari malformation Type I) was first proposed in 1997 by Cinalli, et al.,\textsuperscript{3} and Tekkok and Ventureyra.\textsuperscript{23} One patient had persistent syringohydromyelia despite a functioning cystoperitoneal shunt and was successfully treated with posterior fossa decompression and duraplasty.\textsuperscript{3} The other child had a functioning VP shunt but had syrinx resolution after placement of a low-pressure valve.\textsuperscript{23}

Both communicating\textsuperscript{16,17} and noncommunicating\textsuperscript{3,4,7,21,23} syringohydromyelia have been documented in association with DWM, which fuels the debate over the pathophysiological mechanism of syrinx formation. In communicating cases, some have argued that the syringohydromyelia results from hydrocephalus, with the syrinx acting as a “fifth ventricle.”\textsuperscript{16} In the present case, the syrinx was not in communication with the posterior fossa cyst. The association with impaction of the foramen magnum caused by posterior fossa cyst herniation can be used to argue against such an interpretation and for a mechanism of altered CSF flow at the foramen magnum, as seen in Chiari malformation Type I.\textsuperscript{6,19,26}

Children with DWM are at risk for syrinx formation as a presumed rare complication. Because spinal imaging is not part of routine management for these disorders, the incidence is unknown. A high index of suspicion for syrinx formation may be important in caring for these patients, and spinal imaging may be helpful in the evaluation. Cystoperitoneal shunt placement alone appears to be effective as an initial treatment. If syrinx collapse is not achieved with a functioning shunt, however, posterior fossa decompression is the reasonable next step.

**Conclusions**

There is a clinical association of DWM with syringohydromyelia, with five previous case reports involving children. This is the third report of an increase in foramen magnum impaction of the posterior fossa cyst and resultant formation of syringohydromyelia. The syrinx resolved with successful decompression of the foramen magnum through insertion of a cystoperitoneal shunt. This unusual case demonstrates that alterations in CSF dynamics can occur with cyst progression and can result in the formation of syringohydromyelia. Spinal imaging may be useful in the evaluation of patients who fail to show the expected improvement after CSF shunt procedures.

**References**

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**Fig. 3.** A: Sagittal T1-weighted MR image obtained at 9 months of age demonstrating formation of a cervicothoracic syrinx. B: Sagittal MR image obtained after placement of a cystoperitoneal shunt, showing complete resolution of syrinx.
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