Acquired Chiari I malformation following baclofen pump placement in a child

Case report

R. SHANE TUBBS, PA-C, PH.D., CHARLES LAW, M.D., W. JERRY OAKES, M.D., AND PAUL A. GRABB, M.D.

Departments of Pediatrics, Rehabilitation Medicine, Pediatric Neurosurgery, and Cell Biology, University of Alabama at Birmingham, Children’s Hospital, Birmingham, Alabama

The authors present a case of a child suffering from shunt-treated hydrocephalus and spastic quadriplegia who underwent surgery for placement of a baclofen pump. Magnetic resonance (MR) imaging performed prior to pump placement demonstrated no hindbrain herniation. Afterward, however, the patient exhibited symptoms of brainstem compression, and MR imaging revealed a significant Chiari I malformation along with a fully functioning ventriculoperitoneal shunt. Posterior fossa decompression was performed, and the patient’s symptoms abated. The authors believe this to be the first report of an acquired Chiari I malformation in a patient with a baclofen pump. Clinicians should consider Chiari I malformation as a rare but severe complication of baclofen pump placement.

KEY WORDS • spasticity • cerebrospinal fluid • complication • pediatric neurosurgery

BACLOFEN pumps are frequently used in the treatment of spasticity. Although many complications have been reported, an acquired Chiari I malformation resulting from a baclofen pump system has not been reported in the literature. We review the case of baclofen pump–induced Chiari I malformation and review the pertinent literature.

Case Report

History and Treatment. We report the case of a 13-year-old boy, born at 28 weeks’ gestation, who weighed approximately 900 g and suffered from an intraventricular hemorrhage. Hydrocephalus was soon diagnosed. At age 7 days a VP shunt was placed. Three shunt malfunctions occurred, each preceded by severe emesis and irritability. A diagnosis of spastic quadriplegia was made when the patient was a toddler. A seizure disorder was also diagnosed, for which the patient receives Tegretol (100, 50, and 150 mg morning, noon, and night, respectively), and scoliosis was present with a thoracic convexity to the right. At 6 years of age the patient underwent MR imaging of the brain and craniocervical junction to ascertain the causes of his seizures (Fig. 1 left). At 8 years of age the patient underwent placement of a subfascial baclofen pump with the intrathecal catheter positioned near the T-6 vertebra. Three years later the alarm in the pump signaled a low battery; a new pump was placed at that time and the original intrathecal catheter was left in place. Both pumps delivered 190 μg of baclofen every 6 hours. After placement of the second pump the patient experienced decreased drooling, increased range of motion in the extremities, and enhanced clarity of speech. The patient has significant contractures at both the elbows and knees, however, and 6 months ago he developed what was thought to be a right-sided Bell palsy. In response to the patient’s continued facial weakness, including right nasolabial fold flattening with good bilateral orbicularis oculi function for a period of 4 weeks, an MR image (Fig. 1 center) was obtained, which demonstrated well-decompressed ventricles and new tonsillar ectopia. Four months later, the patient’s facial weakness persisted, his speech had slurred, and he had developed excessive drooling. His muscle tone during this period remained well controlled as a result of the intrathecal administration of baclofen. Magnetic resonance imaging this time revealed a Chiari I malformation (Fig. 1 right) with no syringomyelia.

Operation. The patient underwent surgery for exploration of his baclofen pump and VP shunt (Fig. 2). The catheter site showed no evidence of cerebrospinal fluid leakage and the pump was functional. The VP shunt, hampered by an adherent intracranial catheter with marginal flow, was found to be working suboptimally and, conse-
quently, its intracranial catheter, reservoir, and valve were replaced. After revision of the shunt the patient’s symptoms did not resolve. A decompression of the Chiari malformation was performed 4 months following this procedure.

Postoperative Course. The last clinical follow-up examination was performed 6 months after decompression of the malformation; the patient’s symptoms of brainstem compression have now improved.

Discussion

Complications following placement of baclofen pumps have been widely reported. Gooch, et al., described complications in 100 consecutive children and young adults in whom pumps were placed for the treatment of severe spasticity. Disconnection of the catheter at its connection to the pump, which occurred in 9% of implanted pumps, was the most common complication. Chiari I malformation was not reported in this large series or in other series or case reports found in a search of the literature spanning from 1966 to the present.

We believe that the baclofen pump resulted in progressive, acquired Chiari I malformation in this patient who suffered from chronic shunt-treated hydrocephalus. Chronic leakage from around the catheter or through an unrecognized fracture may be the underlying cause of the Chiari I malformation. Chiari I malformation was not radiologically evident in this patient during the period in which the first pump system was being used. It was only after undergoing surgery for pump revision that his symptoms began. Interestingly, Akman, et al., have reported retrograde leakage of cerebrospinal fluid into the infusion pump reservoir that had been caused by hyperbaric oxygen therapy. Catheter leakage and obvious fistula formation have also been reported. We do not believe that the suboptimally working VP shunt, although clearly a risk factor, was the cause of this patient’s symptomatic tonsillar ectopia. Historically, shunt malfunction in this patient was always accompanied by severe emesis and irritability, neither of which was observed after the third shunt revision was performed in 1998. We propose that lumbar punctures accompanied by normal intracranial pressure led to an acquired Chiari I malformation. Other situations in which the Chiari I malformation has been induced include spinal subarachnoid shunts, lumbar drains, epidural anesthesia with subarachnoid space penetration, and lumboperitoneal shunts. In fact, Chumas, et al., have reported a death due to chronic tonsillar herniation in a child with Crumon disease who received a lumboperitoneal shunt.

Conclusions

In summary, symptoms of Chiari I malformation should be investigated if noted in patients in whom baclofen pumps have been placed; clinicians should recognize that Chiari I malformation is a rare but possible complication of this treatment modality.
Acquired Chiari I malformation

References


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