One-Week Old Treated With Shunting, Cranial Reduction For Macrocephaly

Neurosurgeons at All Children's Hospital/Johns Hopkins Medicine in St. Petersburg, Florida and the University of South Florida Morsani College of Medicine in Tampa, Florida, recently achieved excellent physical and aesthetic results in an infant born with extreme macrocephaly due to hydrocephalus. This was accomplished with implantation of a ventriculoperitoneal shunt followed by a new operation to stabilize and reduce the size of the baby's head. Both surgeries were performed during the infant's first week of life. The surgeons report that as far as they know, this is the first reported case of cranial fixation in such a young child with extreme hydrocephalus. Full details can be found in the article *Cranial reduction and fixation with a resorbable plate combined with cerebrospinal fluid shunting for difficult-to-manage macrocephaly related to hydrocephalus*, by Jotham Manwaring, M.D., and colleagues, published online, ahead of print, in the *Journal of Neurosurgery: Pediatric*s.

The baby's head measured 50 cm in circumference (more than the 99th percentile). His anterior fontanel bulged, and there was wide separation between the bones of all cranial sutures. Magnetic resonance imaging confirmed the diagnosis of severe hydrocephalus. When the infant was two days old, surgery was performed to implant a ventriculoperitoneal shunt with a programmable valve. The performance level of the valve was adjusted repeatedly over the next few days to balance drainage of cerebrospinal fluid (CSF). Despite multiple shunt valve adjustments, a satisfactory balance could not be achieved to allow sufficient CSF drainage without creating a dysmorphic skull appearance.

This often occurs in infants with congenital hydrocephalus and markedly enlarged skulls. The surgeons decided to perform a cranial reduction and stabilization procedure in an effort to improve the child's appearance, to facilitate his care, and to allow adequate CSF drainage without resulting in skull bone overlap.

Gerald Tuite, M.D., and Devon Truong, P.A.-C, the developers of the procedure,began the operation by creating an incision just behind the anterior fontanel. The surgical team drained enough CSF from the ventricles to bring both frontal and parietal bones together, resulting in a far better shape of the infant's head. The team placed a resorbable plate across the entire anterior fontanel and affixed the parietal and frontal bones firmly in place. This prevented the development of unsightly overlaps, which commonly occur when a skull is disproportionately large compared to the size of the underlying brain. The procedure took 45 minutes and the infant tolerated it well. Immediately post-operatively, the shape of the infant's head was much improved and it was much easier to care for him. There was no longer the need for frequent repositioning or shunt valve adjustments to try to regulate head shape. Not only were the cranial bones held solidly in place, but the cranial volume was reduced by approximately

49 percent.

Follow-up examinations showed that as the child became older, his cranial bones grew over the anterior fontanel to meet at the sutures and the implanted plate was absorbed by the body. At 17 months, the infant displayed near-normal development (with a slight motor delay) and required no medication or additional surgery. The shunt is still in place and functioning well. The infant's appearance remains excellent.

The importance of this paper is the timing of the cranial reduction and stabilization procedure. It was accomplished shortly after shunt implantation, within the first week of life—much earlier than reported in other papers. The authors state that this early intervention was much less technically challenging than later reconstructions, which sometimes require more than one surgical procedure, can carry more risks, and may not achieve the aesthetic outcome desired. In this case, only one 45-minute reconstructive surgery was required. The authors also suggest that further modifications of their procedure could make it simpler and even possibly include shunt implantation during the same surgery.

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