Reduction Cranioplasty for Macrocephaly
—Two Case Reports—

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Abstract
Multi-stage reduction cranioplasty was performed on two children with severe macrocephaly secondary to hydrocephalus. One patient underwent a four-stage operation, and the other underwent a two-stage operation. The postoperative course of both patients was uneventful. Reduction cranioplasty improved quality of life for both patients, and good cosmetic results were achieved. Reduction cranioplasty is effective for the treatment of macrocephaly, and multi-stage surgery can reduce the associated risks.

Key words: reduction cranioplasty, hydrocephalus, ventriculoperitoneal shunt, macrocephaly, head circumference

Introduction
Reduction cranioplasty procedures for macrocephaly have been reported sporadically since a two-stage reduction cranioplasty was performed in 1964.\(^1\) Various surgical procedures have subsequently been reported to improve the patient's quality of life. However, reduction cranioplasty is only appropriate in a limited number of patients, as it carries major risks of surgical complications.\(^2\) The major aim of reduction cranioplasty is to improve the quality of life, and cosmetic results are important but secondary. Here we describe two cases of multi-stage operations for macrocephaly which markedly improved the quality of life and achieved good cosmetic results.

Case Reports
Case 1: An 8-year-old boy presented with severe macrocephaly. His head circumference at birth was 39.5 cm. The prognosis was poor, so his parents rejected treatment for hydrocephalus. He was not treated for nearly 2 years. Eventually, a ventriculoperitoneal shunt was placed at age 3 years because of greatly enlarged ventricles. Intraventricular pressure was 160 mmH\(_2\)O when the ventricular catheter was inserted. The diagnosis was severe hydrocephalus with normal intracranial pressure. However, his head circumference continued to increase gradually. The shunt function was never satisfactory. The patient could not support his head without assistance. Physical examination confirmed severe macrocephalus (head circumference 85 cm) with a massive frontal bossing (Fig. 1). Computed tomography (CT) demonstrated enlarged ventricles and a thin cerebral mantle (Fig. 2 left).

Reduction cranioplasty was accomplished in four stages. The first stage was performed at 8 years of age. The patient was placed in a lateral position. The left temporal and parietal bony calvariae were removed. Each bone plate was trimmed. The pieces of bone plate were fastened with a wire, forming a patchwork and the forehead reconstructed. The second stage was performed one year later at age 9 years. The bifrontal bone flaps were removed and the forehead was again reconstructed with trimmed bone flaps. The dura was not opened during the first and second operations. The third stage was done at age 11 years. The bilateral frontoparietal bones were removed followed by reduction of the vault which required fragmentation of the shell. The parietal dura mater was sutured in a parasagittal curvilinear line, taking great care to avoid kinking of the superior sagittal sinus. The third operation resulted in a lowering of the cranial vertex by 5 cm. The fourth...
stage, done one year after the third, dealt with the huge occipital prominence. The patient was placed in the prone position. Three free plates of bone from the occipital region were divided into several irregular fragments, then reassembled and secured to the edges of the calvariae. Following the third and fourth operations, cerebrospinal fluid (CSF) was removed via ventricular drainage. The brain was slack, so the reduction cranioplasty was easy to perform. The shunt system (ventricular catheter, valve) was exchanged for a new one during each operation. The estimated total blood loss was 2000 ml.

The patient's head circumference was 68 cm after the last operation (Fig. 3) and has remained the same till now. Shunt function was satisfactory. At present, the patient can walk independently. Neuropsychological testing showed an intelligence quotient of 49 on the Tanaka-Binet intelligence scale. CT revealed thickening of the cerebral mantle, and dilated ventricles (Fig. 2 right).

Case 2: A 3-year-old boy was born with an abnormally large head. Hydrocephalus was diagnosed and a ventriculoperitoneal shunt was inserted at age 1 month. CT demonstrated a Dandy-Walker malformation with agenesis of the corpus callosum. Three-dimensional CT revealed scaphocephaly (Fig. 4 left). His head circumference was 54 cm. Follow-up CT after the shunt operation revealed the ventricle size was unchanged and his head stopped en-
The diagnosis was non-progressive hydrocephalus. Neuropsychological testing at 3 years old demonstrated a developmental quantity (DQ) of 60.

Reduction cranioplasty was performed in two stages. The initial operation was done at age 3 years. The patient was placed in the supine position. The bifrontal bone flaps were removed to maintain the midline bony bridge overlying the superior sagittal sinus (Fig. 5). The anterior sagittal bar was shortened by 2 cm and connected to the frontal bone. The calvarial bone was reshaped and secured in place with a wire. The procedure resulted in a 2 cm reduction in the head circumference. One year later, the posterior half of the calvaria, including the portion below the transverse sinus, was removed. The posterior sagittal bar was shortened by approximately 2 cm and connected to the occipital bone (Fig. 6). The calvarial bone was reshaped using bone-molding forceps. CSF was removed during both operations via a ventricular tap. The total blood loss was approximately 520 ml.

Postoperative CT demonstrated that the ventricular size was decreased (Fig. 4 right). The patient’s head circumference was reduced from 54 to 50 cm (Fig. 7). Developmental testing revealed a DQ of 68 at 5 years old.

**Fig. 5 Case 2.** Photographs showing the first operation. The design of the bilateral frontal bone (left). Construction of the midline bony bridge overlying the superior sagittal sinus (sagittal bar method) (right).

**Fig. 6 Case 2.** Schematic drawing showing the reduction cranioplasty.

**Fig. 7 Case 2.** Pre- (left column) and postoperative (right column) three-dimensional computed tomography scans (upper row: anteroposterior view, lower row: lateral view).

**Discussion**

Reduction cranioplasty is intended to reduce the size of the cranium and improve the patient’s quality of life. The procedure should, of course, be considered for all patients with macrocrania. Patients who have good neurological function, but whose heads are so large that they cannot walk because of the weight, as in Case 1, are good candidates. Reduction cranioplasty is suitable for the treatment of uncontrolled or neglected hydrocephalus, craniocerebral disproportion, and grotesque macrocrania. However, not all patients with macrocrania

*Neurol Med Chir (Tokyo) 39, June, 1999*
are appropriate candidates for reduction cranioplasty. The important point is that the procedure should provide a better quality of life. This procedure should never be undertaken by surgeons with only a casual interest in craniofacial surgery or neurosurgery. We think that there are relatively few suitable candidates for reduction cranioplasty. Our two patients described were both appropriate candidates for this complex procedure. In both cases, the parents hoped that reduction cranioplasty would improve the quality of life for their child. Parental considerations are an important factor in making the decision to carry out reduction cranioplasty.

Reduction cranioplasty methods are tailored to the specific requirements of individual cases. Cross bar craniotomy allows reduction of the cranium, and the quadrantal, picket fence, and cross bar techniques are also available. Three cases of reduction cranioplasty used total calvarial reconstruction using contoured skull fragments as in our cases. Drainage of CSF was necessary in all cases, as was imbrication of the dura due to the laxity of the brain tissue. However, excessive CSF drainage increases the risk of bleeding into the epidural subgaleal space. Other risks include massive operative blood loss due to laceration of the sinus, air embolism, and infection. Thus, we opted for multi-stage operations for decrease these risks.

We recommend that staged reduction cranioplasty be considered for cases similar to ours. The time interval between procedure stages and the initial operative site of the cranial vault are important considerations in multi-stage surgical procedures. An interval of one year between operations appears to be necessary to achieve re-osseification of bone defects. Follow-up CT at one year revealed good ossification of bone defects in our patients. Craniotomy of the anterior part of the calvariae seemed less risky than the posterior half of the calvariae including the venous confluence. The sagittal bar technique reduced the intraoperative blood loss. Thus, we recommend that the first cranioplasty be performed at the safest possible cranial vault site in young patients.

References


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