Macrocephaly is defined as a head circumference of greater than 2 standard deviations above the mean. There are many causes of macrocephaly, including growing, chronic subdural hematomas, familial megalcephaly, pseudotumor cerebri, benign extracerebral collections of infancy, hydrocephalus, various genetic conditions, and metabolic and dysplastic diseases.

Among these causes, untreated hydrocephalus is the most common for developing macrocephaly, leading to a condition termed hydrocephalic macrocephaly. Hydrocephalic macrocephaly is a relatively rare condition and may result in many complications such as hemorrhage due to cranial vault instability. In addition, massive cranial size could hinder neurological development.

Abbreviation used in this paper: VP = ventriculoperitoneal.
because of increased intracranial pressure, difficulties with posture and movement, and aesthetic problems.\textsuperscript{5,6,9} Because extrathecal CSF shunts can be used to treat all types of hydrocephalus, most hydrocephalic children respond to shunt surgery alone, especially ventriculoperitoneal (VP) shunt placement, and nearly normal neurological development can be ensured in most.\textsuperscript{1,12} However, there are hydrocephalic children whose conditions either could not be completely corrected through shunt surgeries or the patient did not receive effective treatment in time. Unfortunately, some of these children could develop extreme hydrocephalic macrocephaly. To date, the technical details and outcomes of using surgical methods for the treatment of hydrocephalic macrocephaly remain the focus of intensive discussions.\textsuperscript{3,4,6,7,11,15,16,18}

In this study, the clinical profile and outcome of the children with hydrocephalic macrocephaly who had undergone shunt surgeries with 1-stage total cranial vault remodeling were analyzed retrospectively. In addition, the feasibility of surgery for total cranial vault remodeling in children with hydrocephalic macrocephaly is discussed.

**Methods**

**Patient Population**

Between January 2007 and January 2011, 5 children (3 males and 2 females; age range 16–97 months, mean 54 months) with head circumferences of greater than 2 standard deviations above the mean for their respective ages were diagnosed as having hydrocephalic macrocephaly at the Department of Neurosurgery of the First Affiliated Hospital of Xiamen University. Upon detailed investigation and after obtaining properly informed consent, surgical treatment for each child was planned by the neurosurgical team. All patients underwent VP shunting or shunt revision procedures and corrective cranial vault surgery. None of the patients had craniosynostosis. This retrospective study was approved by the First Affiliated Hospital of Xiamen University Institutional Review Board.

**Operative Technique**

Prior to the single-stage total cranial vault remodeling surgery, we simulated the most probable postoperative appearance of each patient’s head and bone reconstruction using computer imaging software prepared by our group with 3D CT (Siemens) data to select the best method for remodeling and final cranial shape. A specific program was designed for each patient to remodel his or her head into the ideal shape using his or her own cranial vault bone (Fig. 1). A plaster head model with the same proportions of the patient’s head was made, and bone reconstruction was simulated on the computer (Fig. 2). The appropriate operative time was then chosen after evaluating the patient’s physical conditions and meeting with the patient’s family members.

On the day of surgery, the patient underwent induction of general anesthesia and was placed in the supine position with the head in 40°–45° ventral flexion. Adrenaline (1:200,000) (Fuzhou Neptunus Fuyao Pharma) was infused subcutaneously, and a coronal incision was made within the hair-bearing scalp halfway between the apex of the skull and the anterior hairline. Scalp flaps were undermined to the eyebrow anteriorly and to the transverse sinus posteriorly in a subperiosteal plane to expose the entire cranial vault (Fig. 3). All calvarial bones were removed according to the preoperative computer simulation plan, and a ventricular shunt catheter was introduced into the lateral ventricle to drain the CSF slowly. The length and width of the dural envelope were reduced by coagulating the dura mater with bipolar forceps in selected areas under continuous irrigation. Coagulation of the vessels of the dura and veins draining into the sagittal sinus was carefully avoided. To determine if there was any subarachnoid or subdural bleeding caused by the operation, a small incision was made in the dura, and the color of the CSF was checked to see if there were signs of subarach-

---

**Fig. 1.** Computer simulation of single-stage vault remodeling surgery. The most probable appearance of each patient’s postoperative head and bone reconstruction was simulated based on preoperative computer images of the 3D CT data of the patient’s head. Preoperative calvaria (A), model of postoperative excepted calvarial shape (B), simulation in splitting the calvarial bone (C and D), and simulation in reconstructing the calvaria (E and F).
Surgical treatment of hydrocephalic macrocephaly

Fig. 2. Plaster head models before and after surgery. Left: A plaster head model in proportion with the patient’s head before surgery was built and the lines of incision were indicated. Right: Simulation of bone reconstruction after surgery for the same patient is shown on a plaster model. A sagittal bandeau was made to support the bilateral bone flap in the single-stage total cranial vault remodeling.

Fig. 3. Case 3. The calvaria of a 97-month-old boy with macrocephaly before, during, and after the single-stage total cranial vault remodeling surgery. A and B: Intraoperative photographs. Scalp flaps were undermined to the eyebrow anteriorly and to the transverse sinus posteriorly in a subperiosteal plane to expose the entire cranial vault. A sagittal bandeau was built. C and D: The head circumference of this patient decreased from 65 cm preoperatively (C) to 54 cm after a single-stage total cranioplasty (D). The patient stayed in the ICU for 22 days due to a lung infection.

Analysis of Outcome

Clinical evaluations included the comparative preoperative and postoperative examinations of head circumference, nasion-inion distance, and bitragal distance as well as postoperative complications. In addition, subjective postoperative neurological change and parent satisfaction were also evaluated. Patients’ functional outcomes, such as the ability to support the weight of the head, to sit, and to stand and walk, were assessed according to a previously published method.6

Results

Two girls and 3 boys with a median age of 35 months (range 16–97 months), at the time of surgery were included in this study. All patients had undergone at least one VP shunt operation at other institutions before coming to our institution, and the average number of shunt procedures performed was 1.4. There were no episodes of air embolism and no deaths associated with any of the procedures. Furthermore, no brain tissue was resected and no significant perioperative complications were found in this group of patients.

During surgery, preventive steps were taken to determine if there were any signs of subarachnoid or subdural bleeding caused by surgery; no subarachnoid or subdural bleeding was found. The average ICU time for these 5 patients was 10.4 days, and the average length of hospital stay was 23 days. One patient had a prolonged stay of 22 days in the ICU due to a postoperative lung infection (Fig. 3). Except for the 2 patients who underwent one-time postoperative shunt revision procedures approximately 2 weeks after the remodeling surgery, no patient underwent any other postoperative surgery for hydrocephalus or macrocephaly. In addition, no other severe postoperative complications were observed in any of the patients during an average of 22 months (range 6–32 months) of follow-up. All patients had a significant reduction in macrocephaly (Table 1) and a better appearance than before the operation (Fig. 5). The median head circumference decreased from 64 cm preoperatively to 53 cm postoperatively (Table 1 and Fig. 5). Likewise, the median nasion-inion distance decreased from 42 cm preoperatively to 34 cm postoperatively, while the median bitragal distance was reduced from 41 cm preoperatively to 34 cm postoperatively (Table 1).

The global neurological change was assessed by a score from 0 to 3: a score of 0 was assigned to permanent decrement while scores of 1, 2, and 3 were given to transient decrement, no change from baseline, and improvement from baseline, respectively. All patients received a...
score of 3 (Table 1). In addition, parent satisfaction was surveyed, and scores of 0, 1, and 2 were recorded if parents were dissatisfied, satisfied, and very satisfied, respectively. The results showed that all parents were very satisfied and were uniformly pleased with the improvement in their child’s appearance after surgery (Table 1). Moreover, motor function, including head control, sitting, and walking/standing, was assessed in patients before and after surgery using the scoring system shown in Table 2. The median total score postoperatively was 7, which was significantly higher than the preoperative score of 5 (Table 3).

Discussion

The occurrence of hydrocephalic macrocephaly is uncommon. As hydrocephalus is often neglected and/or treated ineffectively, defects in neurological function and physical development together with a cosmetic deformity of the calvaria are frequently seen in these patients. Reduction cranioplasty is required in certain cases of hydrocephalic macrocephaly to decrease the size, weight, and contour of the significantly enlarged head.

In our study, 5 patients with an average age of 54 months (range 16–97 months) underwent reduction cranioplasty and all had improvement in motor function, including head control, sitting, and walking/standing. In addition, all parents were very satisfied and were uniformly pleased with the improvement in their child’s appearance after surgery. Thus, reduction cranioplasty is a very effective treatment to correct hydrocephalic macrocephaly.

Various surgical methods of reduction cranioplasty for the treatment of hydrocephalic macrocephaly have been described in the literature. Takahashi et al. considered that aesthetics were important but not as important as neurological function; they performed multistage reduction cranioplasty to treat 2 cases of hydrocephalic macrocephaly to decrease the associated risks.

In addition, Gage et al. described a limited cranioplasty technique that was shown to be associated with a shorter hospitalization and lower shunt revision rate. This technique focused on the posterior turricephaly but did not address any anterior frontal bossing. Other surgeons reported that a single-stage total cranial vault reduction cranioplasty is safe and effective for the treatment of mac-

**TABLE 1: Postoperative measurements and subjective outcome assessment**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (mos), Sex</th>
<th>Head Circumference (cm)</th>
<th>Nasion-Inion Distance (cm)</th>
<th>Bitalag Distance (cm)</th>
<th>Neurological Change*</th>
<th>Parent Satisfaction†</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Preop</td>
<td>Postop</td>
<td>Preop</td>
<td>Postop</td>
<td>Preop</td>
</tr>
<tr>
<td>1</td>
<td>93, M</td>
<td>64</td>
<td>53</td>
<td>46</td>
<td>35</td>
<td>45</td>
</tr>
<tr>
<td>2</td>
<td>16, F</td>
<td>64</td>
<td>54</td>
<td>42</td>
<td>34</td>
<td>41</td>
</tr>
<tr>
<td>3</td>
<td>97, M</td>
<td>65</td>
<td>54</td>
<td>43</td>
<td>35</td>
<td>42</td>
</tr>
<tr>
<td>4</td>
<td>35, F</td>
<td>63</td>
<td>53</td>
<td>42</td>
<td>34</td>
<td>40</td>
</tr>
<tr>
<td>5</td>
<td>29, M</td>
<td>62</td>
<td>52</td>
<td>41</td>
<td>34</td>
<td>40</td>
</tr>
</tbody>
</table>

* A score of 3 indicates improvement from baseline.
† A score of 2 indicates very satisfied.
Surgical treatment of hydrocephalic macrocephaly

We reasoned that a simulated computer program to reconstruct the head prior to surgery should be used and, together with the aforementioned surgical techniques, could provide good aesthetic outcomes post-operatively. We also used a sagittal bandeau as described by Sundine et al. in all of our total cranial vault remodeling procedures for the correction of hydrocephalic macrocephaly (Figs. 3 and 4). All patients survived and had good outcomes in terms of neurological function and aesthetics. We conclude that a sagittal bandeau is important in achieving the stability and bilateral symmetry of the remodeled cranial vault.

Hydrocephalic macrocephaly is rare. In general, each patient with this condition has a different head deformity. Therefore, each total cranial vault reduction cranioplasty is unique. From our experience, we recommend the following steps for a successful reduction cranioplasty. First,
A computer imaging system should be used to simulate the patient's postoperative head appearance and bone reconstruction followed by selection of the best method for remodeling and cranial shape in every case. Second, an operative plan should be designed around remodeling the patient's head to an ideal shape using the patient's cranial vault bones (Fig. 1). Third, the techniques of bone reconstruction should be practiced according to the computer-simulated procedures using a plaster head model made in proportion to the patient's head before performing the total cranial vault reduction remodeling surgery (Fig. 2).

Hydrocephalic macrocephaly is usually only found in infants and young children. Kohan et al. performed total cranial vault remodeling in 8 children with a mean age of 18 months (range 9–26 months) at the time of surgery, and a high satisfaction rate with appearance and ease of caring for the child were noted after surgery.

Likewise, Gage et al. performed reduction cranioplasty for macrocephaly in 10 patients with an average age of 17.9 months (range 6–53 months), and all patients showed good outcomes at an average of 41.5 months of follow-up. However, Mathews et al. suggested that the reduction cranioplasty should be avoided in patients younger than 3 years. Obviously, the minimum age at the time of reduction cranioplasty for hydrocephalic macrocephaly is still under debate. In our study, the mean age of patients undergoing total vault reduction remodeling surgery was 54 months (range 16–97 months). It should be noted that patients with hydrocephalic macrocephaly usually have significant growth retardation in the body and limbs, and some of them have very weak physical conditions that may not be suitable for a lengthy reduction cranioplasty. For these reasons, we offered this surgery to appropriate patients regardless of their age.

There are no reports about long-term follow-up after cranial vault remodeling in patients with hydrocephalic macrocephaly. Therefore, it would be difficult to predict the outcome of our patients 5 or 10 years after surgery. More cases and a longer follow-up time are needed to evaluate this surgical treatment.

**Conclusions**

From our experience, single-stage total cranial vault remodeling with shunt surgery for patients with hydrocephalic macrocephaly who are older than 1 year is safe and effective. The strategic goal of total cranial vault reduction cranioplasty is to improve the patient's quality of life and to correct the deformity, which requires meticulous preoperative evaluation and planning. We suggest using a computer 3D imaging system to simulate the patient's bone reconstruction before surgery for better aesthetic outcome.

**Acknowledgment**

We would like to thank John Jane Jr., M.D., (Department of Neurosurgery, University of Virginia, Charlottesville, VA) for his valuable advice throughout the study and for his critical reading of the manuscript.

**Disclosure**

This work was supported by the Natural Science Foundation of China (81271332). The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**References**

1. Di Rocco C, Massimi L, Tamburrini G: Shunts vs endoscopic

---

**TABLE 3: Comparison of functional outcome before and after surgery***

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (mos)</th>
<th>Head Control</th>
<th>Sitting</th>
<th>Standing &amp; Walking</th>
<th>Total Score</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>preop</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>93</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>2</td>
<td>16</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>3</td>
<td>97</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>4</td>
<td>35</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>5</td>
<td>29</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>median</td>
<td>35</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>postop</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>94</td>
<td>3</td>
<td>2</td>
<td>3</td>
<td>8</td>
</tr>
<tr>
<td>2</td>
<td>17</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>3</td>
<td>98</td>
<td>3</td>
<td>2</td>
<td>3</td>
<td>8</td>
</tr>
<tr>
<td>4</td>
<td>36</td>
<td>3</td>
<td>2</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>5</td>
<td>30</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>median</td>
<td>36</td>
<td>3</td>
<td>2</td>
<td>2†</td>
<td>7†</td>
</tr>
</tbody>
</table>

* Refer to Table 2 for definitions of functional assessment scores.
† p < 0.05 versus the preoperative value.
Surgical treatment of hydrocephalic macrocephaly

third ventriculostomy in infants: are there different types and/or rates of complications? A review. *Childs Nerv Syst* **22**:1573–1589, 2006


Please include this information when citing this paper: published online November 29, 2013; DOI: 10.3171/2013.10.PEDS12573.

Address correspondence to: Zhan-Xiang Wang, M.D., Department of Neurosurgery, The First Affiliated Hospital of Xiamen University, 55 Zhenhai Rd., Xiamen 361003, Fujian, People’s Republic of China. email: sjwk123@yahoo.com.cn.