

Improved Surgical Treatment for Craniostenosis in Children

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Volume 4 Issue 10- 2020

Received Date: 22 Sep 2020

Accepted Date: 09 Oct 2020

Published Date: 12 Oct 2020

2. Keywords

Improved surgical treatment;
Craniostenosis; Children

1. Abstract

To analyze the craniostenosis and the characteristics of development of cranial bones in children, introduce an improved surgical treatment for craniostenosis, and evaluate its efficacy and feasibility. Three children with craniostenosis were treated by our improved surgical treatment from April 2014 to April 2016, and all of them suffered scaphocephaly caused by premature closure of sagittal suture. All the children recovered well and were followed up for six months to one year during which no relevant clinical symptoms occurred again. A three-dimensional CT scan of the cranial bones showed that the sutures were not closed. The appearance of the skull was improved. The nervous system function and development were normal. Craniostenosis is more commonly seen in children. The cause is yet to be identified. Re-closure of sutures after conventional surgery is commonly found. Our improved surgical method proved to be effective in treating this disease in children because the treatment can effectively increase the volume of the cranial cavity and improve the appearance of the skull and the development of the nervous system. So this treatment can be widely applied.

The cause of craniostenosis is unknown, but it is highly related to genetic factors. It can often cause symptoms such as increased intracranial pressure, delayed development, mental retardation, abnormal mental activity, and epilepsy in children. Its clinical manifestations are different due to different types of craniosynostosis. So there are many surgical treatments for this disease. But the purpose of all treatments is to increase the volume of the cranial cavity and ensure enough space for the development of the nervous system [1]. If it is not treated in the early stage, it can cause skull deformities, abnormal neurological development, and poor prognosis. Three children with craniostenosis caused by premature closure of sagittal suture were treated by our improved surgical treatment from April 2014 to April 2016. The surgery proved to be effective in treating this disease, which can be reflected in the following aspects:

3. Case Study

3.1. General information

The 3 cases (male, aged 15 months to 37 months) in the group were all children with craniostenosis caused by premature closure of the sagittal suture, and they were admitted to our hospital for treatment from April 2014 to April 2016.

3.2. Case One

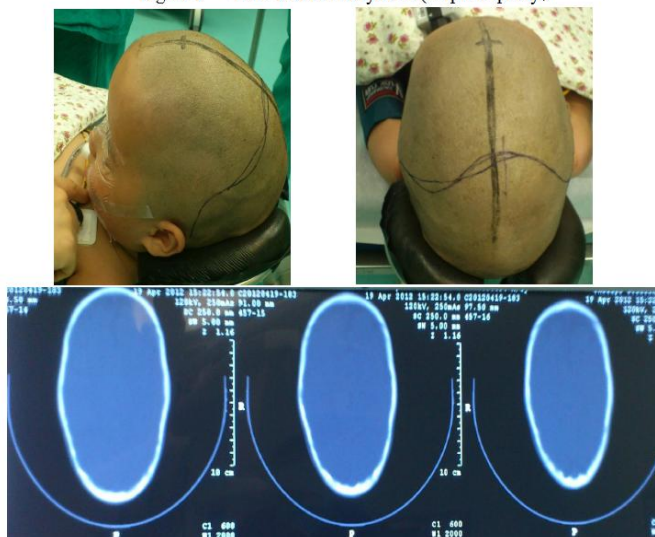
Patient: Duan (surname), male, 3-year-and 1-month-old (37 months), had a history of neonatal jaundice and no hereditary disease in the family. He was admitted to the hospital for headache and blurred vision in March. At birth, the child was found to have a scaphoid-shape head, with a long anteroposterior diameter. No special treatment was given. The headache started 3 months ago with intermittent attacks and accompanied by blurred vision. Then he was taken to our department of our hospital for treatment, and was diagnosed as craniostenosis by three-dimensional CT imaging and other examinations. Admission physical examination showed there was no significant difference in abilities of speech, memory and intelligence between them and children of their age. The gross visual acuity of both eyes was 0.5, and the visual field was normal. The head circumference was 51cm, the anteroposterior diameter was 15cm, and the transverse diameter was 10cm. The muscle strength and muscle tension of the limbs were normal. Auxiliary examination showed the pressure at lumbar puncture was 200mm-H₂O. CT scan of head showed changes in cranial bones, a sign of craniostenosis, and no obvious abnormalities in the brain parenchyma.

Author Contributions: Xing H and Jing-ping L, These authors have contributed equally to this article

Citation: Xing H. Improved Surgical Treatment for Craniostenosis in Children. *Annals of Clinical and Medical Case Reports*. 2020; 4(10): 1-7.

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Figure 1 Case 1 of craniostyosis (scaphocephaly)



Scaphoid-shape skull of the child

CT scan of the skull shows that the anteroposterior diameter is significantly longer than the transverse diameter, and the cranial cavity is narrow, which affects brain development.

3.3. Case Two

Patient: Tan(surname), male, two-year and 9-month-old, has no patient history. The disease does not run in the family. He was admitted to the hospital for abnormal head shape for more than 2 years with an intermittent headache for half a year. The family members of the child complained that the head of the child was different from the children of the same age after birth and did not receive special treatment. Half a year ago, the child began to have a headache with intermittent attacks, which became worse after activities. So the child was taken to our hospital for treatment. He was

then diagnosed as craniostenosis (caused by premature closure of sagittal suture) by the three-dimensional CT scan and other examinations. Physical examination on admission showed no obvious abnormalities in consciousness, intelligence, vision and visual field, 50cm of head circumference, closed bregma, 19cm of anteroposterior diameter, and 13cm of transverse diameter.

Movement of limbs was possible. Auxiliary examination showed the pressure at lumbar puncture pressure was 140mmH₂O. A three-dimensional CT scan of head showed changes in cranial bones, a sign of craniostenosis.

Figure 2 Case 2 of craniostenosis(cymbocephaly)



The shape of the head of the child before the operation was scaphoid, and the cranial cavity volume was small. The anteroposterior diameter was significantly longer than the transverse diameter. The results of CT imaging of the cranial bones of the child before the operation.

4. Surgical Methods

4.1 Preparation before surgery

Before the surgery, the children were given mental support, underwent routine examinations and tests to make sure the functions of the heart and lungs were normal. After the blood and scalp were prepared, the scalp was cleaned and disinfected with water and iodophor to prevent infection. Then sterile dressings were used to cover the skin of the surgical area after disinfection.

4.2. Auxiliary Examination

All the children underwent a 64-slice CT scan of the skull and three-dimensional reconstruction of the head. The results showed that the sagittal suture disappeared, and the bone accumulated and thickened at the premature closure of cranial suture. No obvious abnormalities and other diseases were found in the skull.

4.3. Surgical Methods

After general anesthesia, the child lied on the back, and the head-rest was fixed. The Steri-Strips for incision were stuck to both sides of the auricle. During the surgery, the child's body was kept warm.

Two surgeons were responsible for the disinfection of the operation area, including the areas from the nasion to the exoccipital tuberosity and lower part of the auricle. After disinfection, a large s-shaped coronal incision was made in the anterior coronal plane of the external auditory canal. The skin was incised, with the end of the incision to the upper part of the bilateral zygomatic arch. The skin, subcutaneous, and temporal muscle tissues were incised layer by layer. The scalp and muscle layers were separated forward and backward to expose the skull. After hemostasis, a hole was drilled about 1cm from the intersection of the sagittal suture, the coronal suture, and the occiput on each side. A milling cutter was used to make a slit with a width of 1.5cm parallel to the sagittal suture on each side. And 4 slits of 1.5cm wide and 8cm long were made with the sagittal suture as the midline, parallel to the coronal suture. After cauterizing the slits with an electric knife and sealing with gelatin sponge and bone wax to stop bleeding, the titanium strip was connected and fixed, and a drainage tube under the scalp was placed. Then the muscle tissues, subcutaneous and scalp incisions were sutured layer by layer (Figure 1). (See Figure 1 for details)

Figure 3 Improved reconstruction of cranial bones

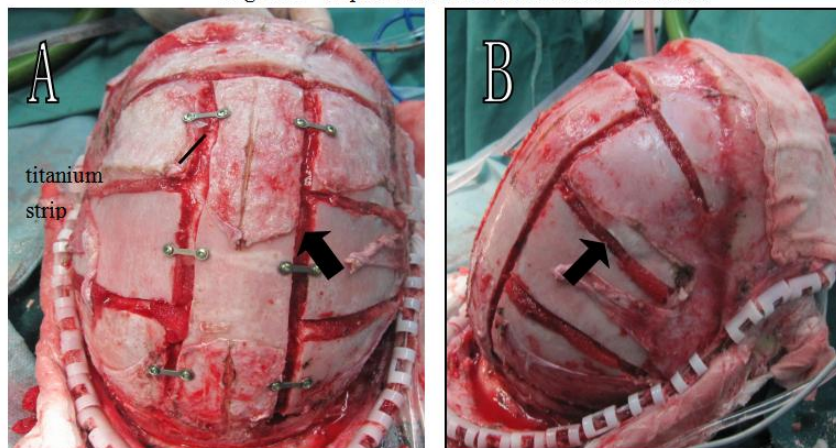


Figure A. Horizontal view. The black arrow points to a slit of 1.5 cm wide, 1 cm from and parallel to sagittal suture on each side.

Figure B. Sagittal view. Four slits of 1.5cm wide parallel to the coronal suture on each side.

4.4. Postoperative Treatment

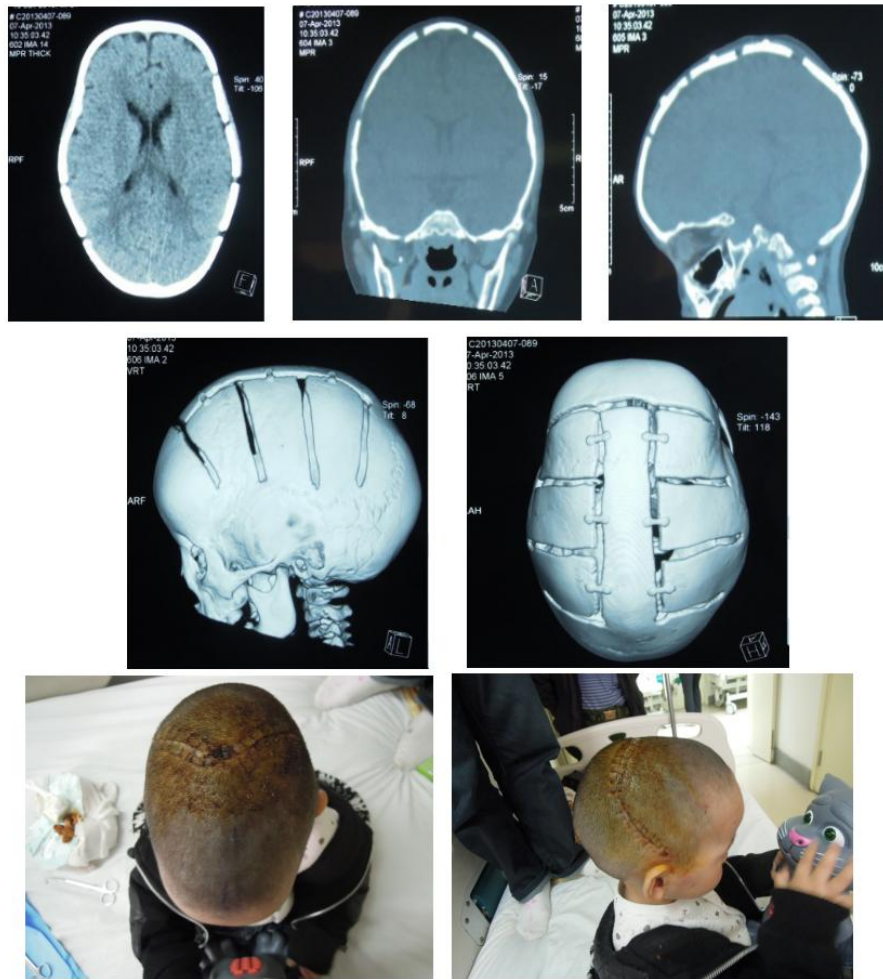
For younger children, electrolyte disturbances and anemia are likely to occur after surgery, and the changes in vital signs should be monitored dynamically after surgery. The hemogram, liver and kidney functions and changes in electrolytes should be reexamined at the same time. Anemia and electrolyte disturbances were treated promptly, and treatments such as anti-infection, hemostasis, and nutritional support were also given. Attention was paid to changes in the subcutaneous drainage of the surgical area.

5. Results

5.1 Postoperative Outcomes

Symptomatic treatments such as hemostasis, neuro-nutrition, and infection prevention were given to all children after surgery; at the same time, anemia was treated as appropriate according to the mechanism of coagulation and intraoperative bleeding. Pressure bandaging was given and the healing of the incision and the drainage tube were paid attention to ensure smooth drainage. During and after the operation, no incision infection was found. The subcutaneous hemorrhage/fluid and the symptoms of intracranial hypertension were significantly relieved. No obvious complications occurred, so the patients were recovered and discharged.

Figure 4 Postoperative skull shape and imaging results



The results of CT scan of the head and 3D reconstruction after surgery. The postoperative head shape of the child.

5.2 Results of Follow-up

Postoperative follow-up was conducted by telephone and visits in outpatient. The follow-up was 12 months, 8 months and 15 months for case 1, case 2 and case 3, respectively. None of the children re-occurred with symptoms of intracranial hypertension, like headache, vomiting and blurred vision. Significant improvements in head shape were seen. No obvious neurological dysfunction was found. The development of nervous system function was the same as that of children of the same age. Results are as follows:

A three-dimensional CT scan of the head was performed 12 months and 8 months after surgery. The reconstructed bone sutures were not closed and the symptoms before surgery did not occur. The results of the Wechsler Adult Intelligence Scale (WAIS) showed the children’s intelligence development was not affected.

Table 1: Postoperative outcomes(cm)

		Clinical Symptoms	Head Circumference	Anteroposterior Diameter	Transverse diameter
Case 1	3 months after surgery	(-)	51	15	10
	6 months after surgery	(-)	51	15.5	11
	12 months after surgery	(-)	52	16	15
Case 2	3 months after surgery	(-)	50	19	13
	8 months after surgery	(-)	51	19.5	16
Case 3	6 months after surgery	(-)	47	18	13
	12 months after surgery	(-)	50	19	14.5

6. Discussion

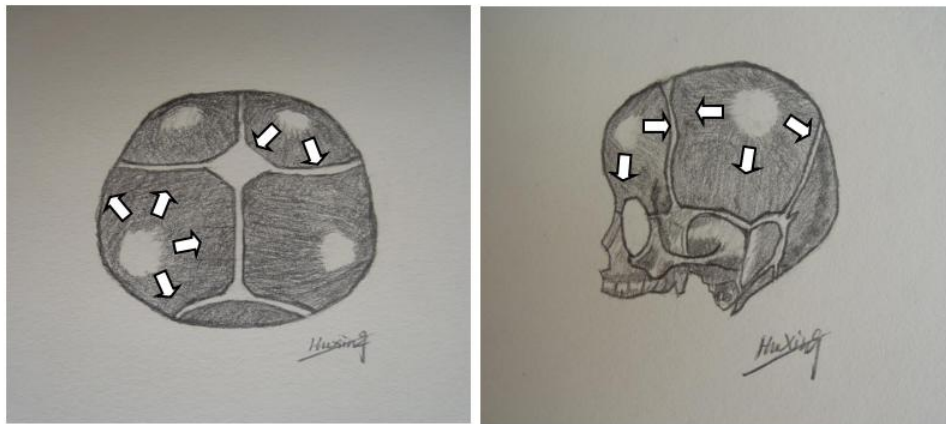
Craniosynostosis is more commonly seen in children. The male/female ratio among the patients who suffer this disease is 3:1. This disease may run in the family [2]. The cause is yet to be identified. Some scholars believe that abnormal growth factors secreted by the endocranium may affect the closure of the cranial bones [3]. Scholars at home and abroad believe that the disease should be treated with surgery as soon as possible [4-5] because the brains of infants and young children develop rapidly. According to reports, the weight of the brain can increase by 135% at 1 year old, and it can develop into 80% of the adult brain at 3 years old. Delayed treatment may lead to a missed window of opportunity for surgery. If the surgery is performed after the brain tissues have developed, the surgery will only serve as improving the appearance of the head.

The typical symptoms of craniosynostosis include: (1) cranial deformity which varies according to the different time of premature closure of sutures; and (2) intracranial hypertension, such as headache, vomiting, and blurred vision; and (3) abnormal intelligence and mental development. The disease was diagnosed mainly by X-ray and three-dimensional CT scan as well as clinical symptoms, like craniosynostosis and intracranial hypertension. It is important

to differentiate it from the microcephaly in which there is no symptom of craniosynostosis by X-ray.

Surgery is the first choice for the treatment of craniosynostosis, and there are different kinds of surgical treatments. The initial surgical treatment was not effective until Marchac and Tessier improved it. They argue that the cranial cavity should be enlarged through surgery rather than the growth of the brain [6]. The commonly used surgical treatments include (1) cranial suture reconstruction; and (2) fronto-orbital advancement (FOA); and (3) fronto-orbital advancement and andorbital reconstruction; and (4) fronto-facial advancement; and (5) large-scale craniotomy and reconstruction of skull flap. These surgeries are more effective than before, but there are defects such as poor appearance and coloboma of cranial bones which lead to reduced brain protection. In the conventional surgery, linear incisions are usually performed at the closed sutures, with the main purpose to keep the cranial sutures unclosed, promote the growth of the cranial bones, and provide the brain with space for normal growth and development. Some scholars believe that the desired effects of surgery can be achieved by keeping the incision sutures unclosed for 2 to 3 years [7]. But infants and children are in the stage of growth and development, so the closure is commonly seen after surgery.

Figure 5 Children's cranial bones and their growth and development characteristic



The continued proliferation of parietal tubercle, the frontal process, and bones and cells around external occipital protuberance, plus the fact that the intramembranous ossification and sutures of coronal sutures, sagittal sutures and lambdoidal suture are not closed, makes the volume of the cranial cavity to enlarge to provide space for brain growth and development.

The growth and development of children's skulls have characteristics. As shown in Figure 5, a child's skull is composed of 8 cranial bones after birth, including paired parietal bones, temporal bones, and unpaired frontal bones, occipital bones, ethmoid bones, and sphenoid bones. There are sutures, such as coronal sutures, sagittal sutures, and lambdoidal suture, between the bones. The cranial bones thicken at the parietal tubercle of parietal bones, the frontal process of the frontal bones, and the occipital process of the occipital bones, which are germinal center for the growth and

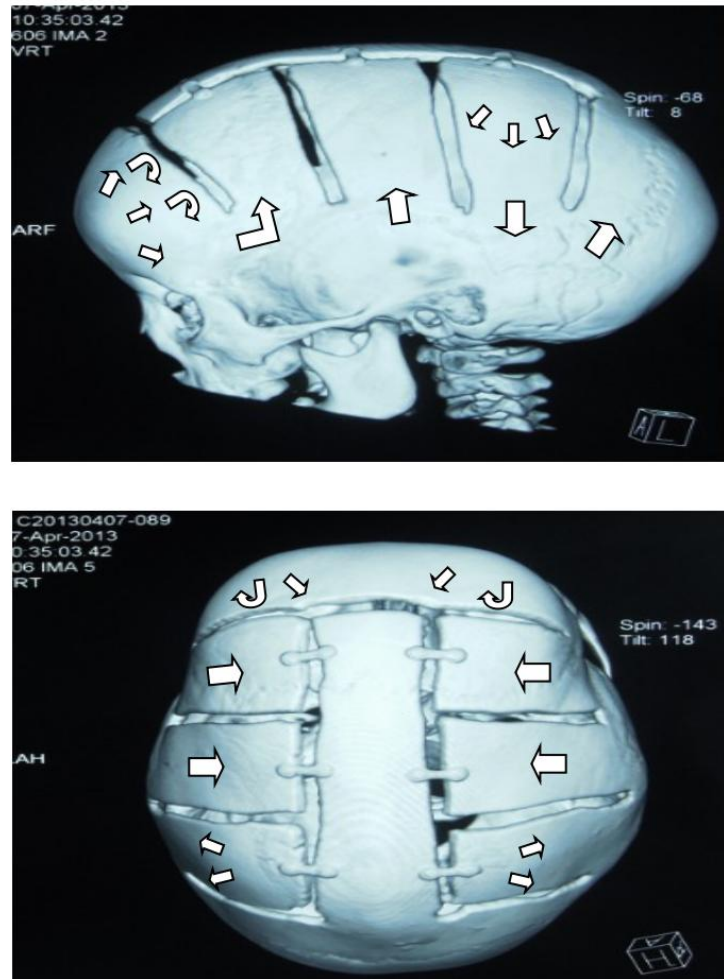
development of the infant's cranial bones. The proliferation of osteoblasts in these places and the sutures of cranial bones makes the bones grow and expand to the periphery to increase the volume of cranial cavity and provide the space needed for brain development. The unclosed sutures during the growth and development of the skull cause cranial bones to expand from the germinal center to the surroundings, expanding the volume of the brain and allowing the cranial bones to keep growing. Yet the premature closure of the cranial sutures will cause the bones to grow abnormally, resulting

in scaphocephaly, trigonocephaly and brachycephaly.

The traditional skull reconstruction has some limitations, including poor appearance, defect on the skull, unstable bone structure and less brain protective. We have improved the traditional surgical method by having regard to the growth and development characteristics of each suture after craniosynostosis in children [8] and applied it to the treatment for children with craniostenosis. According to the characteristics of craniosynostosis, as shown in the

figure, one slit was cut on both sides of the sagittal suture, and four slits parallel to the coronal suture. The slits were cauterized with an electric knife and sealed with gelatin sponge and bone wax to stop bleeding. The 4 artificial slits on the coronal plane stop the bones in areas of the forehead, parietal tubercle on both sides and occipital germinal center from growing anterior and posterior along the sagittal plane. Skull growth and development after surgery is shown in Figure 6.

Figure 6 Improved reconstruction of cranial bones and characteristics of their growth



The improved reconstruction of sutures of cranial bones prevents the bones from continuing to grow deformed along the anteroposterior diameter. Due to the blocking of the sutures, the bones have to bypass the sutures to grow laterally in the direction of the transverse diameter. The purpose is to effectively stop the longitudinal growth of the bones along the anteroposterior diameter and promote the normal growth along the transverse diameter.

The improved surgical treatment provides enough space for the normal growth and development of the cranial bones, and wearing a custom-made helmet after surgery can prevent the bones from growing deformed, to increase the volume of the cranial cavity, improve the skull shape and ensure the nervous system development. Compared with the traditional cranioplasty [9, 10], the improved treatment saves the trouble of large-scale cutting and disorderly rearrangement of the cranial bones to reduce the injuries and improve safety during and after the surgery significantly [11]. The

results of the follow-up confirm that improved surgical treatment is effective.

7. Conflicts of Interest

We declare that we have no financial and personal relationships with other people or organizations that can inappropriately influence our work, there is no professional or other personal interest that could be construed as influencing the position presented in, or the review of, the manuscript entitled.

8. Ethical Statements

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors. Informed consent was obtained from all individual participants included in the study.

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