Anthropometric study of craniofacial asymmetry in unilateral coronal craniosynostosis

Estudo antropométrico das assimetrias craniofaciais na craniossinostose coronal unilateral

ABSTRACT

Introduction: Unilateral coronal craniosynostosis (UCC) is the unilateral premature ossification of the coronal suture, leading to a 3-dimensional deformity that may affect the skull and orbits. Objective: The purpose of this study was to measure and compare the craniofacial asymmetry of patients with UCC in the preoperative and postoperative periods to assess the effectiveness of surgical UCC correction. Methods: Nine patients (4 girls and 5 boys) underwent surgical correction of UCC between January 2007 and December 2010. Their craniofacial dimensions were measured with a goniometer to calculate the craniofacial asymmetry index (CAI) in the preoperative and postoperative periods, and the CAI averages of the periods were compared. The CAI was calculated as the ratio of the craniofacial diagonal measurements of the unaffected side to those of the affected side. Results: The mean patient age was 2 years 1 month. The average surgical time was 2 hours 46 min. The average volume of transfused blood was 280 ml. The preoperative and postoperative CAI averages were 1.045 and 1.009, respectively (p=0.0109), indicating a strong tendency for symmetry after the surgery. Conclusion: Surgical UCC correction is effective for achieving craniofacial symmetry. Overcorrection of the bony structures is required to obtain bone symmetry postoperatively.

Keywords: Craniosynostoses. Plagiocephaly. Craniofacial abnormalities.

RESUMO

Introdução: A craniossinostose coronal unilateral (CCU) é a ossificação prematura da sutura coronal unilateramente e provoca uma deformidade em 3 dimensões, que pode afetar o crânio e as órbitas. Objetivo: O objetivo do presente trabalho foi aferir e comparar a assimetria do crânio e face dos pacientes portadores de CCU no período pré e pós-operatório. Método: Nove pacientes foram submetidos à correção da craniossinostose coronal unilateral, entre janeiro de 2007 a dezembro de 2010. Quatro pacientes foram do sexo feminino e cinco do sexo masculino. Foram aferidas medidas da região craniofacial para quantificar o índice de assimetria craniofacial no período pré-operatório e compará-lo com o período pós-operatório. O índice de assimetria craniofacial (IAC) foi determinado pela diferença entre as medidas craniofaciais diagonais obtidas com o goniômetro. Resultados: A idade média dos pacientes submetidos à correção de CCU foi de 2 anos e 1 mês. O tempo médio de cirurgia foi de 2 horas e 46 minutos. O volume médio de sangue transfundido foi de 280 ml. A média das diferenças das medidas diagonais obtidas com o goniômetro no período pré-operatório (IAC) foi de 1,045 e do pós-operatório de 1,009 (p=0,0109), indicando forte tendência à simetria craniofacial após o ato cirúrgico. Conclusão: O tratamento proposto para as CCU foi eficiente na obtenção de simetria craniofacial. Foi necessária a sobrecorreção das estruturas ósseas para obtenção de simetria óssea no período pós-operatório.

INTRODUCTION

Craniosynostosis is the premature fusion of one or more sutures of the cranial vault and its associated cranial deformities. The pattern of skull deformities is peculiar to the fused suture (metopic, coronal, sagittal, and lambdoid). Unilateral coronal craniosynostosis (UCC) is the premature fusion of the coronal suture. Its incidence is approximately 1 in 10,000 live births, and it has been described as the second most frequently identified synostosis in specialized centers for the treatment of craniofacial deformities.

The vast majority of patients with UCC typically present morphological changes of the cranial vault due to a compensatory mechanism of brain growth (Virchow’s law), usually toward the open sutures, which causes a 3-dimensional deformity. This situation leads to considerable deviation of the sagittal axis of the skull base, observed during physical examination through the discrepant position of the ears (the affected side is positioned higher and forward), deviation of the nasal root to the affected side, and deviation of the nasal tip to the opposite side. The frontoparietal region ipsilateral to the affected suture is depressed, leading to a compensatory contralateral hump. The eyebrow position is always higher on the affected side. The mentonian region is deviated to the contralateral side. The zygomaticomaxillary region may be more anteriorly positioned in relation to the contralateral side, following the 3-dimensional rotation of the skull base.

The clinical implication of the rotation of the skull base in UCC has been intensely debated, and this rotation is believed to be responsible for all the clinical findings of UCC. As described by Silva-Freitas et al., the orbit in Harlequin (elevation of the superolateral corner of the orbit) is a typical sign of UCC, because of the anomalous position of the greater sphenoid wing on the affected side. The premature fusion of the coronal suture may be associated with craniofrontonasal dysplasia, Saethre-Chotzen syndrome, and the Pro250-to-Arg mutation in the FGFR3 gene, also known as the Muenke mutation. The clinical features vary according to the identified genetic mutation and may involve the upper limbs.

The purpose of the present study was to measure and compare the craniofacial asymmetry of patients with UCC in the preoperative and postoperative periods to assess the effectiveness of surgical UCC correction.

METHODS

Nine patients (4 girls and 5 boys) underwent surgical correction of UCC between January 2007 and December 2010. Their craniofacial dimensions were measured with a goniometer to calculate the craniofacial asymmetry index (CAI) in the preoperative and postoperative periods and the CAI averages of the periods were compared.

The CAI was calculated as the ratio of the craniofacial measurements of the unaffected side to those of the affected side. For example, a preoperative CAI average greater than 1 indicates a larger unaffected side before surgery (Figure 1). All patients included in the analysis were photographed in a standardized photography studio while lying down to obtain photographs in the full frontal, lateral, oblique, and frontobasal views. Patients suffering from syndromic craniofrontonasal dysplasia associated with UCC were excluded.

Operative Technique

With the patient in the supine position, under general anesthesia, UCC correction was begun with a coronal incision and subgaleal undermining of the frontal flap. Subsequently, a pericranial flap was elevated from the coronal suture level to the orbital roof. The supraorbital neurovascular bundle was carefully identified, elevated, and released from the frontal bone together with the periorbit. With access to the inner orbit, periorbital undermining was performed along the subperiosteal plane.

Craniotomy was performed through the fontanel, providing safe access to the intracranial area. The cranial bone was easily removed and cut with a piecemeal instrument, Jackson scissors, or a reciprocating saw. Cranioplasty was performed on the surgical table. The anterior cranial base was detached at the central and lateral portions, with extensive detachment
of the greater sphenoid wing. Osteotomy was performed from
the region of the greater sphenoid wing toward the lateral
wall of the orbit. The basilar region was decompressed by
using a malleable retractor, with appropriate protection of
the notable structures.

Fronto­orbital advancement was not performed because
the rapid and intense brain growth in the first year of life allows
the orbital structures to become symmetrical naturally; this
surgery was reserved for older patients with UCC. Children
younger than 1 year of age present a favorable combination
of flexibility and possibility of large brain expansion during
growth of the craniofacial skeleton. This combination enables
surgical correction without the use of molding helmets and the
need to move the orbital structures.

Suction drains were placed and maintained for 24-48 h.
Dressings were used until 12 h after drain removal. The scalp
was sutured with 4-0 nylon sutures3.

Statistical Evaluation
Student’s t-test was used to compare the preoperative and
postoperative CAI averages. P values less than 0.05 were
considered significant.

RESULTS
The patients underwent the same surgical technique,
with completion of cranioplasty and decompression of the
skull base. Decompression of the skull base usually provides
enough room for brain growth.

The average patient age was 2 years 1 month. The average
surgical time was 2 h 46 min. The average volume of blood
transfused was 280 ml.

The preoperative and postoperative CAI averages were
1.045 and 1.009, respectively, showing a significant diffe-
rence (p=0.0109; Figures 2-6).

DISCUSSION
In the present study, the postoperative CAI average was
close to 1, indicating that important cranial symmetry was
achieved surgically. Surgery for UCC correction also has
direct implications on facial symmetry and positioning of
the orbits, and therefore, it should always be performed
before 1 year of age. The brain normally doubles in size in
the first year of life4. This rapid brain growth acts directly
on the repositioning of the craniofacial structures in patients
surgically treated for UCC3. The main goal of early surgical
intervention is to interrupt the deleterious effects of abnormal
suture fusion on the craniofacial skeleton. In addition, cranio-
synostosis may be associated with increased intracranial pres-
sure, even when only 1 suture is compromised5, and can lead
to deficits in the neurocognitive development of the affected
child6. The immediate relief of clinical signs of intracranial
hypertension by surgery usually allows normal craniofacial
development.

The craniofacial skeleton of a child aged 3-5 months is
malleable and easy to reshape. The postoperative bone defects

Figure 2 - Preoperative photograph of a patient with unilateral
coronal craniosynostosis (right). Photograph of the patient 2 years
after surgery to correct the craniosynostosis (left). Considerable
improvement in the contour of the frontal region and orbital
dystopia were observed; the improvement was achieved because
of rapid brain expansion, usually occurring in patients
undergoing surgery within 6 months of birth.

Figure 3 - Preoperative photograph of a patient with unilateral
coronal craniosynostosis, strabismus associated with congenital
muscular torticollis, and vertical orbital dystopia who underwent
surgery to correct the craniosynostosis at 3 months of age (right).
Postoperative photograph of the patient (left). Note the
considerable improvement in the contour of the
frontal region and orbital dystopia.
undergo reossification due to the osteogenic potential of the dura mater and periosteum in patients under 1 year of age. Therefore, early surgery has considerable benefits. The mean age of the patients who underwent surgical UCC correction in this study was 25 months; 2 patients aged 3 years accounted for the increase in mean age. The remaining 7 patients were operated on before 1 year of age. Preferably, patients should be operated on at around 4 months of age. However, many patients are referred to the hospital when they are older, mainly because of the lack of early diagnosis. The unmanageable skull bones of older patients make this surgery more difficult, with more intense blood loss. Often, it is prudent to delay the start of the operation until blood products are physically present in the operating room. Anticipation of the need for blood replacement and careful intraoperative monitoring of the volume of blood loss are essential for successful surgery.

Although several agents for bleeding control are available, such as bone wax, Surgicel, and fibrin glue, blood loss continues to occur in the immediate postoperative period. Therefore, blood transfusion is frequently required in the first 2 postoperative days. In the present study, all the patients required blood transfusion (~280 ml blood), and this volume may correspond to 40% of the total blood volume in a 3-month-old child. New technologies such as preoperative injection of erythropoietin and autologous transfusion are used by large North-American centers. However, because of their high costs, these modalities have not yet been introduced in the Brazilian National Health System (SUS). The current economic growth in Brazil and an increase in the resources for public health will facilitate access to new technologies in hospitals, ultimately reducing the potential risks inherent to a major surgery.

Measurement of the craniofacial structures is essential for diagnosis and monitoring of changes in growth; moreover, it may indicate the effectiveness of surgical intervention. Several methods for these measurements are available, such as X-ray, 3-dimensional computed tomography (CT), and magnetic resonance imaging. CT has the disadvantage of exposing children to a radioactive load; moreover, it requires the presence of an anesthesiologist during the surgery, which may not be possible on short notice. Alternative measurement methods, such as 3-dimensional photography and plagiocephalometry, can be used; however, they are not always applicable because of technical difficulties and scarcity of the required materials. In our department, the measurements were carried out by the same evaluator using a metric-scale tape and goniometer.

The CAI was used to verify, through measurements in the preoperative and postoperative periods, whether the surgical results visible upon clinical examination were significant. The obtained p value (p=0.0109) demonstrated that the postoperative CAI average was significantly lower than the...
preoperative CAI average, indicating a strong tendency for symmetry after the surgery. Clinically, the skull of all the patients had advanced toward symmetry, demonstrating the efficiency of the surgical technique for correction of UCC.

**CONCLUSION**

Surgical correction is effective for achieving craniofacial symmetry in patients with UCC. Overcorrection of the bony structures is required to obtain craniofacial symmetry postoperatively.

**REFERENCES**


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