

Clinical Manifestations and Surgical Treatment of Craniosynostosis in Mosul City–Iraq

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Abstract

Introduction: Craniosynostosis is a condition in which one or more of the fibrous sutures in an infant skull prematurely fuses by turning into bone.

Aim: Evaluate the clinical manifestations of craniosynostosis, imaging and surgical treatment with their complications.

Materials and Methods: The study comprised twenty one patients complaining from craniofacial deformity. The patients evaluated by recording the medical history, clinical examination and diagnosed by imaging that included a plain X-ray and computed tomography. The goal of surgery is to open the fused sutures, reshape the head and allow for normal brain and skull growth. According to the type of material that used to fixate the cranial bones, the patients divided into 3 groups, Group A: Titanium plate and stainless steel wire; Group B: Stainless wire; Group C: Absorbable suture.

Results: The clinical examination showed cranial bones deformity in all patients while 4 patients involved by papilloedema. The plain X-ray for the skull diagnosed 4 cases by showed beaten copper appearance, while the computed tomography gave excellent evaluation for all cases. The nonsyndromic craniosynostosis formed (90.4%) and coronal suture involvement formed (61.9%). The follow-up showed that the group C had more cosmetic improvement and more comfortable than group A and B, and there were 4 patients in group A and 2 patients in group B complained from extracranial prominent of plates and wire.

Discussion: The advantages of absorbable suture included lower costs, speed of application, and absence of translocation.

Conclusion: The most clinical manifestations were cranial deformity while visual deterioration was often late manifestations. The computed tomography scan represents the gold standard evaluation for craniosynostosis. The use absorbable suture in bone fixation showed more cosmetic improvement and more comfortable to the patients with low complication rate.

Keywords

Cranial deformity; Craniofacial disease; Cosmetic cranial surgery

Introduction

In the first years of life the sutures serve as the most important centers of growth in the skull [1]. The growth of the brain and the patency of the sutures depend on each other. Brain growth pushes the two sides of the patent sutures away from each other, thereby enabling growth of the neurocranium. This means that the neurocranium can only grow if the sutures remain open [2]. The brain grows rapidly in the early years of life, with growth of the neurocranium essentially ceasing at about 7 years of age. The fontanels usually close by the second year of life. Complete sutural fusion occurs after the third decade of life [3-5].

Craniosynostosis is a condition in which one or more of the fibrous sutures in an infant skull prematurely fuses by turning into bone (ossification) [6]. Normally, calvarial bones grow perpendicular to suture lines [7]. Because the skull cannot expand perpendicular to the fused suture, it compensates by growing more in the direction parallel to the closed sutures that results in an abnormal head shape and abnormal facial features [1]. Cases in which the compensation does not effectively provide enough space for the growing brain, craniosynostosis results in increased intracranial pressure leading possibly to visual impairment, sleeping impairment, eating difficulties or an impairment of mental development.

Craniosynostosis where no extracranial deformations are present is called non-syndromic or 'isolated' craniosynostosis [8]. When there are extracranial deformations present, for instance involving the limbs, heart, central nervous system or the respiratory tract it is called syndromic craniosynostosis [9]. Non syndromic type includes "simple craniosynostosis," involving only one suture, or "compound craniosynostosis," where two or more sutures are involved [6].

Article Information

DOI: 10.31021/jddm.20192128

Article Type: Research Article

Journal Type: Open Access

Volume: 2 Issue: 1

Manuscript ID: JDDM-2-128

Publisher: Boffin Access Limited

Received Date: 30 January 2019

Accepted Date: 04 March 2019

Published Date: 06 March 2019

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Citation: Abd al-Hameed IA, Taha YA, Mahmood AS. Clinical manifestations and surgical treatment of craniosynostosis in Mosul city–Iraq. J Dents Dent Med. 2019 Mar;2(1):128

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In the past, the prevalence of craniosynostosis was estimated to be one per 1,800 to 2,200 births and in a recent survey; the estimate is even higher [10]. Based on etiology, craniosynostosis may be characterized as primary (nonsyndromic and syndromic) or secondary (premature closure of normal sutures because of another medical condition such as deficient brain growth) [8]. The etiology of nonsyndromic craniosynostosis is unknown, and the condition is sporadic in most instances [11]. Fibroblast growth factor and fibroblast growth factor receptor (FGFR) regulate fetal osteogenic growth [12]. Mutations in the gene coding for FGFR1 cause Pfeiffer's disease, and mutations in FGFR2 cause Apert's syndrome and Crouzon's disease [13-15].

Commonly, craniosynostosis is present at birth, but it is not always diagnosed when mild. Usually it is diagnosed as a cranial deformity in the first few months of life. The diagnosis relies on physical examination and radiographic studies, including plain radiography and computed tomography (CT) [2]. Optimal care of infants with craniofacial anomalies requires a multidisciplinary team approach. Infants should be evaluated within the first few weeks of life. However, referral is appropriate at any age. Once the diagnosis of craniosynostosis is confirmed, the treatment is surgical correction. The best time to intervene is when the infant is between three and nine months of age [2,6].

Because the calvarial bones are malleable and heal effectively [3]. Surgical intervention involves either strip craniectomy or cranial vault remodeling with excision of the frontal, parietal, and occipital bones, which are trimmed, reshaped, and fixed with absorbable plates [2]. Potential complications include massive blood loss and air embolism [12]. Mortality rates are low according to recent reports [2].

Aim of Study

Study and evaluate the craniosynostosis distribution, clinical manifestations, examination, imaging and surgical treatment with their complications.

Materials and Methods

The study comprised twenty one patients who were admitted to Neurosurgical Department in Mosul city-Iraq. The complaining of all patients were craniofacial deformity which may be associated with sign and symptoms of increase intracranial pressure (head ache, vomiting and deterioration of visual acuity). The ages of patients ranges between (2 months-4 years), 14 patients were male while the female were 7. All patients were evaluated by recording the medical history for the patients that included systemic disease, visual acuity and familiar congenital anomalies, also recorded the mother pregnancy history which included systemic disease, using of drug and subjected to psychological stress during pregnancy. Then the patients underwent thorough clinical examination by neurosurgeon, maxillofacial surgeon and ophthalmological surgeon, the last step of diagnosis was done by imaging that included a plain X-ray for the skull (Figure 1) and computed tomography CT scan (Figure 2). Magnetic resonance imaging (MRI) is usually reserved for the evaluation of the brain in cases of increase intracranial pressure. After surgical operation we compared the imaging results with the surgical finding to evaluate the value of each images.

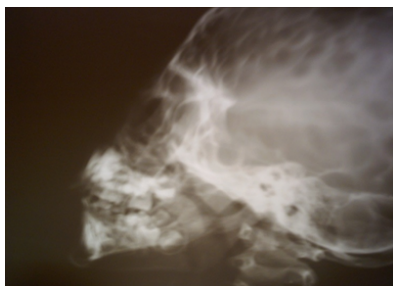


Figure 1: Plain radiograph showed beaten copper appearance

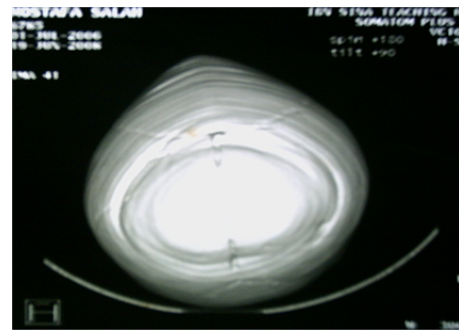


Figure 2: Computed tomography showed fused metopic suture

After diagnosed the condition as craniosynostosis, the cases were distributed in scientific classification:

- 1) Non-syndromic craniosynostosis (isolated)
 - A. Simple that involved only one suture
 - I) Trigonoccephaly (Metopic suture)
 - II) Plagiocephaly (unilateral Coronal suture)
 - III) Posterior plagiocephaly (Lambdoid suture)
 - IV) Scaphocephaly (Sagittal suture)
 - B. Compound that involved more than one sutures
- 2) Syndromic craniosynostosis in which there are extracranial deformations present, for instance involving the limbs, heart, central nervous system or the respiratory tract.

The cases that diagnosed before the age (6 months) we wait until the age (6-8 months) for doing the surgical operations while the cases that diagnosed after age (8 months) the operations were done immediately.

The goal of Surgery is to open the fused sutures, reshape the head and allow for normal brain and skull growth. In all cases the operations were done by teamwork included the maxillofacial surgeon and neurosurgeon, the operations started by neurosurgeon using coronal flap incision on the scalp to explore the cranial bone and fused sutures, then he went to craniotomy and remove the fused sutures, depending on fused sutures and cranial deformity Frontal-orbital advancement and orbital expansion may be required in which the maxillofacial surgeon removes the bones of the forehead as well as supraorbital bar and advances the forehead (Figure 3). The cranial vault remodeling by the maxillofacial surgeon through excised, trimmed, reshaped, and fixed the cranial bones to allowed the brain to grow and also to correct the abnormal shapes that occur as a result of the various types of craniosynostosis. Randomly and according to the type of material that used to fixate the cranial bones, the patients divided into 3 groups:

- a) Group A: Titanium plate and stainless steel wire (Figure 4)
- b) Group B: Stainless wire only (Figure 5)
- c) Group C: Absorbable suture (Figure 6)

After operation, follow up with clinical examination and imaging the cases give us good idea about the general condition of patients and the complication that may be associated with each type of fixation.



Figure 3: Forehead advancement



Figure 4: Fixation of cranial bone by plates and wire (Group A)

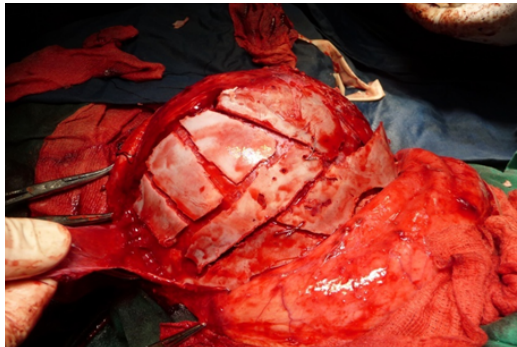


Figure 5: Fixation of cranial bone by stainless steel wire (Group B)

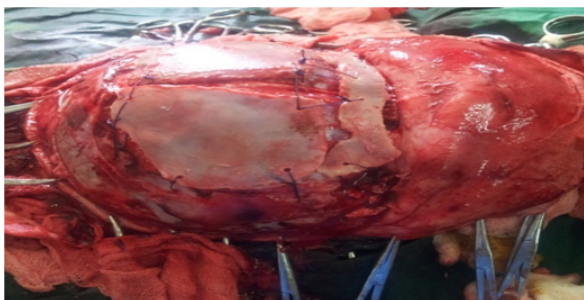


Figure 6: Fixation of cranial bone by absorbable suture (Group C)

Statistical analysis

Data were collected and managed by SPSS System (Statistical Package for Social Science) on Pentium Four computer. The Percentage Statistical test was done for study groups.

Results

Patient's age, sex and chief complain

The patient's ages at the time of diagnosis showed a wide range (3 months-4 years) with mean of (13.5 months). 14 Patients (66.6%) from all patients underwent operations during first year of life and the age at operation not less than 6 months in all cases. The males represented 66.5% (14 patients) from all patients, while females constituted only 33.3% (7 patients) from all patients. The age range and sex distribution of patients enrolled in the present research is shown in Table 1.

The complaining in all patients was cranial bones deformity while only 4 patients (19%) were complaining from visual deterioration in addition to cranial deformity.

History, clinical examination and imaging

The study showed that there were no systemic disease or familiar congenital anomalies for the patients and their mothers, only 3

mothers received drug during pregnancy (amoxicillin, paracetamol and antihistamine) for 4 days during first trimester. Also the study recorded that 7 patient's mother complained from sudden stress during first trimester of pregnancy.

In addition to cranial deformity, the clinical examination showed there were 4 patients involved by papilloedema which were the same patients that complaining from deterioration of visual acuity and 2 of them complained from exophthalmus.

Then compares between the results of imaging and the finding during surgery showed that the plain X-ray for the skull had role in diagnosed 4 cases (19%) complaining from papilloedema and deterioration in vision which gave finger printing and a beaten copper appearance of the calvarium (Figure 1) while The computed tomography (CT) scan represents the excellent evaluation for all cases (100%).

Classification of craniosynostosis

The nonsyndromic craniosynostosis (isolated) involved 19 patients that formed (90.4%) while only 2 cases (males) were involved by syndromic craniosynostosis that formed (9.6%). The study recorded that single suture involvement (simple craniosynostosis) presented in 10 patients (52.6%) which included 8 male and 2 female, the most common suture (simple craniosynostosis) was metopic suture that involved 5 patients. The multiple sutures involvement (compound craniosynostosis) presented in 9 patients (47.4%) which included 4 male and 5 female, the most common combined sutures involvement were bicoronal sutures that presented in 8 cases.

Therefore the most common suture involved in this study that include syndromic and non syndromic craniosynostosis was coronal suture that involved 13 cases (61.9%) in which 3 cases simple and 10 cases associated with other sutures (Table 1).

Surgical operation and complications

Immediately after operation there have been high degrees of improvement in craniofacial shape in group A that used titanium plates in bones fixation, and less in group B that used stainless steel wire and much less improved in group C that used absorbable suture.

The follow-up for all cases for 1 to 2 years showed that the group C had more cosmetic improvement results and more comfortable than group A and B, there were 4 patients (47%) in group A and 2 patients (28.5%) in group B complained from extracranial prominent of plates and wire that caused discomfort to children and cosmetic problem with possibility of intracranial interference with brain tissues. In addition that all cases in group A and B that treated with stainless steel wire cannot be tolerate with MRI in future to evaluate brain condition or other body parts (Table 1).

The 4 patients that complained from visual deterioration showed dramatically improvement after days from operations. The mortality of this study reported one case that dead after five hours from operation due to unknown significant cause.

Discussion

In the results reported by this study, it is seen that there was a clear male preponderance among patients with craniosynostosis (66.5%), and this result compatible with Gerald M et al. [16] who reported that male formed (62.8%) from study group.

The patient's ages at the time of diagnosis showed a wide range (3 months-4 years) with cranial deformity as a main chief complain and this point was explained by Panchal J et al. [2] who mention that craniosynostosis is present at birth Commonly, but it is not always diagnosed when mild and usually it is diagnosed as a cranial deformity in the first few months of life, therefore the late diagnosis usually associated with sing and symptoms of increase intracranial pressure in addition to cranial deformity [17]. Also Panchal J et al. [2] reported that the diagnosis relies on history of pregnancy, physical

Group	Age	Sex	C.C	Type of disease	Suture involvement	Type of bone fixation	Complication of surgery
A	10 M	Male	Cd	Compound	Bic	Plate/wire	Prominent
	18 M	Male	Cd	Simple	Me	Plate/wire	Non
	4 M	Female	Cd	Simple	Me	Plate/wire	Dead
	14 M	Male	Cd/Vd	Syndromic	Bic/S	Plate/wire	Prominent
	4 Y	Male	Cd/Vd	Compound	Bic/S	Plate/wire	Non
	3Y	Male	Cd/Vd	Syndromic	Bic/S	Plate/wire	Prominent
	14 M	Female	Cd/Vd	Compound	Bic/Me/S	Plate/wire	Prominent
	10 M	Male	Cd	Compound	Bic/S	Wire	Non
B	1 Y	Male	Cd	Simple	Unic	Wire	Non
	3 M	Female	Cd	Compound	Me/S	Wire	prominent
	12 M	Male	Cd	Simple	Me	Wire	Non
	12 M	Male	Cd	Compound	Bic/S	Wire	prominent
	12 M	Male	Cd	Simple	Unic	Wire	Non
	5 M	Male	Cd	Simple	Unic	Wire	Non
	18 M	Female	Cd	Compound	Bic	Suture	Non
C	4 M	Female	Cd	Simple	Me	Suture	Non
	3 M	Female	Cd	Compound	Bic/M	Suture	Non
	10 M	Male	Cd	Simple	S	Suture	Non
	10 M	Male	Cd	Simple	Me	Suture	Non
	12 M	Male	Cd	Simple	S	Suture	Non
	18 M	Female	Cd	Compound	Bic	Suture	Non

M: Month; Y: Year; Cd: Craniofacial deformity; Vd: Visual deterioration; Bic: Bicoronal; Unic: Unicoronal; S: Sagittal; Me: Metopic

Table 1: Clinical and surgical manifestations of craniosynostosis

and clinical examination with radiographic studies, including plain radiography and computed tomography (CT), which agree with this study that mention that CT scan represents the gold standard evaluation for cranial bones and fused sutures while plain X-ray for the skull had limited utility in diagnosing of craniosynostosis although finger printing and a beaten copper appearance of the calvarium may point to increased intracranial pressure in late stage.

The study reported that the coronal suture was the most common suture involvement in craniosynostosis which formed (61.9%), but this result not compatible with Sun PP, Persing JA [12], who showed that the sagittal suture was involved in 40 to 60 percent of cases while the coronal suture formed 20 to 30 percent of cases which may be due to different genes and races. Cohen MM Jr. [18] was reported that syndromic craniosynostosis is less common (20%) than non-syndromic, which agree with this study that showed only (9.6%) of cases were involved by syndromic.

The study reported the use absorbable suture in bone fixation more comfortable than other material which compatible with Fearon JA [19], that reported the advantages of absorbable suture which included lower costs, speed of application, and the absence of observed intracranial translocation. Therefore the use of absorbable suture fixation was associated with both acceptable aesthetic outcomes and low complication rates.

Conclusion

The male were more affected by craniosynostosis than female. The most clinical manifestations in all patients were cranial bones deformity while visual deterioration and increase intracranial pressure in addition to cranial deformity are often a late manifestation of the disease process. The computed tomography (CT) scan represents the gold standard evaluation for cranial bones and fused sutures while the plain X-ray for the skull had limited utility in diagnosing of craniosynostosis. The most common type was the nonsyndromic craniosynostosis (isolated) while the most

common suture involvement was coronal suture. The follow-up for cases concluded that surgical operation that used absorbable suture in bone fixation showed more cosmetic improvement and more comfortable to the patients with low complication rate.

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