

# Craniosynostoses Presenting As Copper Beaten Skull Appearance

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## Abstract

Craniosynostoses is defined as a condition of premature fusion in one or more cranial sutures leading to a number of significant complications, mainly raised intracranial pressure which can itself cause other neurodevelopmental complications. Copper beaten skull appearance is a well-known presentation of craniosynostoses made by the strong impression of the brain's gyri on the inner layer of the skull. It may also represent an increase in ICP. We report a 1-month-old infant with the diagnosis of craniosynostoses, confirmed by conventional radiologic studies and a three dimensional CT scan, presenting as copper beaten skull appearance. After diagnosis, the patient underwent a successful neurosurgical intervention.

**Keywords:** Copper Beaten Skull, Craniosynostoses, Intracranial Hypertension

## 1. Introduction

About 25% of single births and 50% of twins represent with a kind of abnormality of head morphology at the time of birth (1). Some of these morphological abnormalities may happen as a result of distinct syndromes such as the tram-line appearance in Sturge-Weber syndrome, copper beaten appearance in curzons syndrome, and dolichocephaly in Marfan syndrome (1). Some others occur following different types of mechanical pressures, either in the intrauterine environment or during passage from the birth canal. These kinds of morphological abnormalities usually resolve without any intervention (2).

In contrast, some other congenital abnormalities of the skull need interventions to resolve. Craniosynostoses is a type of these aforementioned problems for which early detection and appropriate surgical intervention are essential to guarantee a normal neurodevelopmental process of the patient (2).

As cranial growth is the result of brain growth, any premature fusion in one or more sutures would result in an abnormal shape of the head due to compensatory growth in other non-fused sutures (2). Craniosynostoses usually presents itself at birth, but in mild cases it may remain undiscovered. Most patients will show cranial deformity within the first few months of life, leading to diagnosis of their disorder. The diagnosis is based on physical examination and radiographic studies, either conventional radiography or computed tomography (CT). In physical exam-

ination, head circumference measurement is essential to detect the associated microcephaly (3, 4).

If left untreated, craniosynostoses may result in a number of complications, including an increase in intracranial pressure (ICP), deformity of the face, strabismus, etc. Among these, the raised ICP is a major complication (5, 6). We present a case of craniosynostoses with elevated ICP associated with a specific radiologic finding, the copper beaten skull (CBS) pattern.

## 2. Case Presentation

A 1-month-old female infant presented with head growth retardation. The infant's past medical history was not significant for any illness. Maternal history during pregnancy was also unremarkable. In the physical examination, her head circumference was 41 centimeters (below the third percentile of standard pediatric growth charts), and the anterior and posterior fontanelles were closed. Other neurological exams were normal. Laboratory results were negative for any abnormal findings. After conventional radiographic studies (Figure 1) gave the impression of craniosynostoses, the patient underwent a three dimensional CT scan of the skull which revealed the closure of the anterior and posterior fontanelles and the sagittal, lambdoid, and coronal sutures (Figure 2). It should also be noted that severe and diffuse CBS patterns were also present in both the radiographic studies and CT scan. Thus,

the impression of craniosynostoses was confirmed and the patient was scheduled for neurosurgical intervention.

The patient's operation involved a bicoronal zigzag sagittal suture craniectomy, bilateral release of coronal and lambdoid sutures, and bilateral frontal and parietal stave craniectomy. The operation was conducted in Shiraz Namazee hospital affiliated with Shiraz University of Medical Sciences (SUMS).

### 3. Discussion

Craniosynostoses involves a premature fusion of the cranial sutures resulting in an abnormal shape of the skull and can cause significant complications (7). This condition can affect one (simple craniosynostoses), or two or more (compound craniosynostoses) cranial sutures (3). Depending on the fused sutures, different shapes of the head appear; they are categorized as six main types of clinical manifestation of craniosynostoses (2):

1. Sagittal craniosynostoses presents as scaphocephaly (dolichocephaly or "boat shaped" head) which is the most common single-suture craniosynostoses (40% - 60% of all cases)

2. Metopic craniosynostoses causes trigonocephaly (triangular head)

3. Unilateral coronal craniosynostoses results in anterior plagiocephaly (twisted head)

4. Unilateral lambdoid craniosynostoses causes posterior plagiocephaly

5. Bilateral coronal craniosynostoses presents as anterior brachycephaly (short head)

6. Multiple-suture involvement causes a more complex head morphology such as cloverleaf skull (kleeblattschädel) or oxycephaly (towering and cone-like head)

The etiology of craniosynostoses can be classified into two groups: 1) primary (syndromic or non-syndromic) and 2) secondary (3). The incidence rate of primary craniosynostoses is reported to be in the range of 0.3-1.4 per 1000 births. Among them, multiple-suture craniosynostoses accounts for 14% of all cases; about 9-15% of them are associated with craniofacial syndromes (2).

The cause of non-syndromic primary craniosynostoses is unknown and the disease is sporadic in most patients. Some probable risk factors, such as advanced maternal age, white maternal race, maternal smoking, and male infant sex are recognized for this type. In contrast, the syndromic form is associated with specific disorders, such as Pfeiffer's syndrome, Crouzon syndrome, Saethre-Chotzen syndrome, and Apert syndrome (3, 6). However, these genetic disorders are considered to be the cause of less than 15% of all cases of craniosynostoses (2). Currently, about 150 different syndromes associated with craniosynostoses are

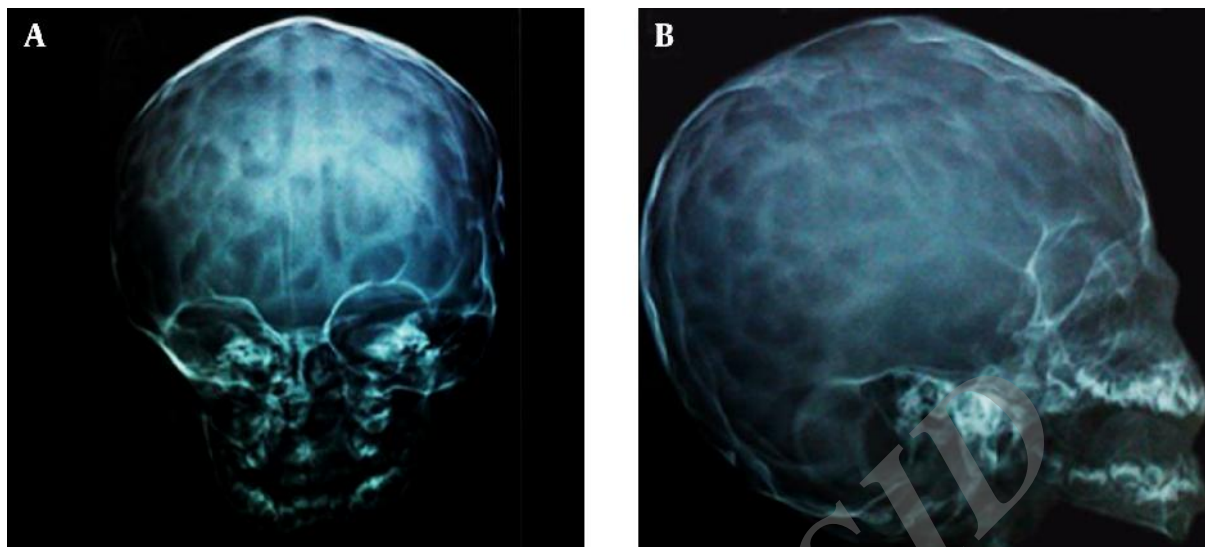
known. Some conditions, such as metabolic disorders and mucopolysaccharidosis, are recognized to be the causes of the secondary form of craniosynostoses (3, 6).

Copper beaten (also known as silver beaten or beaten brass) appearance-a well-known presentation of craniosynostoses-is similar to the hammer markings on a metal such as silver or copper. It refers to a condition in which convolutional markings are more prominent than the normal state. Convolutional markings can be normal, but they are usually restricted to the posterior part of the inner layer of the skull vault (8). They are the normal impression of the brain's gyral pattern on the inner surface of the skull and are usually seen in rapid brain growth periods during the age ranges of 2-3 and 5-7 years. The prominence of these normal markings decreases approximately after the age of 8 years (1). Whenever these markings become pronounced and more anterior, it is more likely to be a copper beaten skull, which raises the probability of an increased ICP (8). However, this appearance is neither significant nor specific enough to be an exact indicator of a raised ICP (8, 9). Accordingly, further investigations such as ICP measurement should be done for a definite diagnosis.

As mentioned above, copper beaten skull appearance is not necessarily the hallmark of an increase in the ICP. Intracranial hypertension, with about 40% prevalence in cases suffering from fusion of multiple sutures, and 14% in patients with single-suture involvement, is a well-known complication of craniosynostoses (4). A chronic increase in ICP may result in macrocrania, splitting of the skull sutures, skull demineralization and erosion, and sella turcica enlargement. These problems are more common in complex craniosynostoses (1). A raised ICP can be a life-threatening condition unless it is diagnosed and managed early (4).

It should be mentioned that, in addition to craniosynostoses, there are two other different diagnoses as the underlying cause of an increased ICP. Obstructive or non-communicating hydrocephalus may be the cause of ICP due to obstruction in the normal flow of the cerebrospinal fluid (CSF) in the ventricular system (1). Another diagnosis for increased ICP is an intracranial mass, which usually disturbs the CSF flow by its pressure effect on the ventricular system (1).

There are numerous surgical options to treat craniosynostoses, ranging from strip craniectomy to cranial vault remodeling. Current surgical interventions include resection of the involved sutures and correction of secondarily-involved bones of the face and cranium. More recently, minimally invasive endoscopic techniques have been successfully applied which resulted in a decrease in both blood loss and length of hospitalization (2, 3).



**Figure 1.** Conventional Skull Radiography Showing the Copper Beaten Skull Pattern in Anteroposterior (A) and Lateral (B) Views



**Figure 2.** Three Dimensional CT Scan of Skull Showing the Diffuse CBS Pattern in Anterior (A), Posterior (B), and Lateral Views (C)

Disregarding the cause of intracranial hypertension, the CBS pattern is the most prevalent imaging indicator of chronically increased ICP in either conventional radiographic or CT scan studies. As a late sign in the disease's course, it results from pressure remodeling of the inner calvarial surface to correspond to the underlying gyri. Among the different causes of raised ICP, the CBS pattern is more common in children with craniosynostoses, especially those less than 18 months old. The presence of this appearance may predict the incidence of some abnormal development in behavioral, writing, and reading skills. Therefore, long-term follow-up of this group of patients is recommended (10).

Copper beaten skull appearance can also diagnose the lacunar skull deformity (luckenschadel). Lacunar skull deformity is a skull anomaly of the newborn, commonly associated with conditions including spina bifida, meningocele, and sometimes encephalocele (10).

Although the CBS pattern may be normal and resolve without any intervention until puberty (1), it may instead be a sign of increased ICP. Due to the risk of intracranial hypertension for normal brain growth, further investigations should be considered to discover any underlying cause and consequently attempt early neurosurgical intervention.

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