CLASSIFICATION OF CRANIOSTENOSIS

DAVID FAIRMAN, M.D.,* AND GILBERT HORRAX, M.D.

Department of Neurosurgery, The Lahey Clinic, Boston, Massachusetts

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The subject of this study is a discussion of the group of malformations in which there exists a premature synostosis of two or more membranous cranial bones. The history and pathologic anatomy of the condition will be considered and a new classification based on an anatomical, clinical and therapeutic standpoint will be suggested.

HISTORY

Homer described in the Iliad the ugliest man who came to Ilium saying that his head was peaked toward the top. Based on this Homeric reference, some French writers have used the phrase Tête à la Thersite for indicating oxycephaly.

Hippocrates was cognizant of the cranial deformities and their relation to the cranial sutures. In the following excerpt he revealed to what extent these deformities were understood about 400 B.C.

Men’s heads are by no means all like to one another, nor are the sutures of the head of all men constructed in the same form. Thus, whoever has a prominence in the anterior part of the head, ... in him the sutures of the head take the form of the Greek letter ταυ, T. ... But whoever has the prominence in the back part of the head, in him the sutures are constructed in quite the opposite form to the former. ... But whoever has a prominence of the head both before and behind, in him the sutures resemble the Greek letter δια H.

Although we were unable to find in the literature an interpretation of these statements formulated by Hippocrates, it is believed that when he spoke of the different forms of the cranial sutures he may have meant premature synostosis of the coronal or lambdoidal sutures in the first 2 cases, and closure of all the sutures including the sagittal suture in the 3rd case.

Oribasius, physician to the Emperor Julian, was aware of oxycephaly. Oribasius who, like Galen, was a native of Pergamos, wrote a digest of medicine and surgery in 70 books, of which 25 remain. He mentioned in some of these writings the cranial deformities associated with palatal defects and other abnormalities.

According to Greig the term “oxycephaides” was used in 1830 by the great naturalist H. Milne Edwards, to designate a variety of crustaceans in which the head is more or less molded like a rostrum. Sommering in 1839, stated that an association appeared to exist between certain types of cranial deformity and the closure of some sutures.

* Fellow in Neurosurgery, The Lahey Clinic, Boston, Massachusetts.
In 1851, Virchow presented a complete study of cranial deformities, giving for the first time a scientific explanation of these conditions. He established the relationship between the malformations of the skull and the premature synostosis of the cranial sutures, formulating the principle that, when synostosis of two bones occurs prematurely, normal growth is inhibited in a direction perpendicular to the obliterated suture line, and compensatory growth takes place in other directions.

Although Virchow failed to distinguish between chondrodystrophies and scaphocephaly or oxycephaly, his great merit consists in his observations that the premature synostosis of the sutures was the direct cause of the restriction of growth and, therefore, the primary cause of the cranial deformity, terming these conditions craniostenosis.

PATHOLOGIC ANATOMY

In order to understand the mechanism by which these conditions are produced, it is necessary to review the most important facts regarding the development of the skull in infancy.

From the embryologic standpoint, the cranium is primarily membranous. In the second month there appears a first and important differentiation. The inferior half is rapidly invaded by the chondrine and transformed into cartilage, while the superior part persists in the stage of simple membrane.

The first of these parts, the so-called chondrocranium, forms the base of the skull, that is, the ethmoid, sphenoid, and the inferior portions of the temporal and occipital bones. The second or membranous cranium constitutes the vault, that is, the frontal, parietal and occipital bones. The development of the base and the vault follows two different ways of ossification.

In the development of the base, there appear a number of points of ossification in the cartilaginous tissue. The ossification is not complete until the age of 6 or 7 years. Development of the vault is more complicated. According to Ford the membranous bones of the vault are still slightly separated at birth although their margins are in apposition, but no bony union exists, as may easily be determined by palpation. The anterior and posterior fontanelles are still evident. Ford stated that between the 6th month and the end of the 1st year the sutures of the vault become closed. This closure of sutures is apparent because the serrated margins begin to interlock, although x-ray examination reveals that the bones are not actually fused. The posterior fontanelle closes, under normal conditions, by the 2nd month and the anterior fontanelle closes between the 14th and the 22nd month.

During childhood the margins of the cranial bones are not actually fused; therefore, it is still possible for the skull to grow. The sutures are interlocked so firmly, however, at the age of 10 years, that increased intracranial pressure causes little or no enlargement of the head.

The sutures play a very important role in the development of the skull. When the ossification invades the suture and the different bones of the
vault become actually fused, then the synostosis is physiologic, when it begins during middle age. Testut said that the physiologic synostosis begins at 40 or 45 years of age and is not complete until the age of 75 or 80. In the various conditions under consideration, the cranial bones are united prematurely, becoming fused before birth. The line of union disappears completely or is marked by a bony ridge. This process constitutes the pathologic synostosis which is of practical significance only when it takes place during intrauterine life or in the 1st year of postfetal life. During this period, growth of the skull is possible on account of the suture spaces. Later the skull develops only by periosteal apposition and resorption, just as in the basal synchondrosis.

The earlier the pathologic synostosis takes place, the more marked is the degree of skull contraction or craniostenosis. On the other hand, closure of a single suture may cause only some contraction since those sutures that remain open take on a compensatory function. It is possible to see externally this compensation, revealed in the irregular development of the skull in various directions.

PATHOGENESIS OF CRANIOSTENOSES

In 1856 Minchin, from Dublin, in a paper based on the fact that there is often a thickening in the region of the obliterated suture, conceived the idea of a single parietal bone extending bilaterally from the medially sutured center. He thought that the thickening above mentioned constituted a single center of ossification for two bones instead of the usual two different centers developing two separate bones. It is impossible to maintain this theory due to the fact that the thickening always shows, from the histologic standpoint, remnants of the early suture.

Fournier stated that syphilis was the etiologic factor in the cranial malformations under discussion, but in all cases in which Wassermann tests were done they were negative and there were no pathologic conditions indicating congenital syphilis.

Virchow, in his fundamental work already mentioned, described an adult oxycephalic skull showing an inflammatory process, involving one of the cranial bones partially. Based on this evidence only, he conceived the idea that an inflammatory process developed in the meninges, and later involved the cranial bones. Therefore, their capacity for further marginal growth was inhibited, producing the pathologic suture.

The main objection to Virchow's thesis consists in the fact that no one so far has been able to demonstrate macroscopically or microscopically an inflammatory process in the lines of the sutures in postfetal life.

Thoma, in a series of articles published in Virchow's Archives between 1907 and 1918, formulated the hypothesis that external pressure produced all pathologic closure of sutures during fetal life. He meant by external pressure the compressing mechanism of the uterine musculature.

Since the pressure exerted by the uterus under normal conditions is a fluid pressure, we must assume that Thoma believed that oligohydramnios
was always present. However, Park and Powers,⁷ in their classical work published in 1920, demonstrated that in their series of 25 cases a history of oligohydramnios was not present in any instance. On the contrary, they reported in 3 cases a history of polyhydramnios. Therefore, Thoma's theory is unacceptable.

Rieping, in 1919, expressed the theory that a defect in the blastemal stage of cranial development was responsible for the displacement of the primary ossification points of the cranial membranous bones toward the suture lines. He felt that when the primary ossification centers of the parietal bones were dislocated toward the midline, the result would be a premature synostosis of the sagittal suture. The same principle applied to the frontal and occipital bones would explain the other varieties of craniostenosis. It is interesting that the same hypothesis was formulated as early as 1875 by Morselli.

Park and Powers insisted on the fact that the margins of the cranial bones were kept apart by interstitial normal growth of mesenchymal tissue. Due to a defect in the germ plasm the capacity of normal growth of the mesenchyme is diminished or lost, without developing the exudate enabling it to resist ossification, thus explaining the premature synostosis.

Undoubtedly, the theory of Rieping, and of Morselli, complemented by that of Park and Powers, resists any fundamental criticism, being so far the most logical hypothesis acceptable.

CLASSIFICATION AND CLINICAL SYMPTOMS

The only formal classification of craniostenosis has been made by Greig,⁸ who considered oxycephaly as a whole and divided it into three types:

I. **True oxycephaly** which presents a general craniofacial stenosis. This is congenital and often associated with syndactyism or other deformities of the extremities.

II. **Delayed oxycephaly** which may appear at any time during childhood and never presents other deformities.

III. **False oxycephaly** which is localized synostosis, not congenital, and is often of definite origin.

Analyzing the original papers published by Greig in 1926, we find the following facts: first that his conception of true oxycephaly was based on 3 patients, aged 58, 58 and 45 years respectively. The 3 patients presented deformity of the head and symptoms of mental deficiency, with 2 of them showing deformities of the extremities. In spite of the fact that these 3 cases did not present any x-ray evidence, we know that a synostosis discovered after 45 years of age cannot be considered unusual and it is impossible to make a retrospective diagnosis to establish at what age closure of the suture occurred. Furthermore, we believe that it is impossible to make a diagnosis of oxycephaly based on the shape of the deformity of the head and the neurologic symptoms only. It is indispensable to have x-ray evidence of the premature closure of the sutures.

As far as the deformities of the extremities are concerned, we believe it
is unjustifiable to regard the combination of malformations of head and extremities as a clinical entity. The significance of their occurrence together appears to be merely an inconstant association, since there is a special tendency when malformations appear in the head to appear also in the extremities.

The second point is that Greig's conception of delayed oxycephaly is based on 2 patients aged 8 and 9 years respectively. He thought that in these cases the condition was not congenital. The deformity was not exaggerated and did not present any related concomitant somatic defect. No roentgenograms of the skull were taken of one patient. In the other patient, although there was an x-ray report of digital impressions over the cranial vault, no mention of synostosis was made. For these reasons it is doubtful whether a diagnosis of oxycephaly can be accepted.

Greig's third point is a classification of his conception of false oxycephaly based on the examination of skulls in the museum of Edinburgh (all of them middle age or older). He stated that in false oxycephaly the synostosis was limited to a few or even to one suture, that facial sutures never participated and that no association of deformities of the extremities coexisted. He stated that the base of the skull was not affected and that the condition was not congenital. Furthermore, he stated that the deformity of the head in false oxycephaly was a misleading feature and he did not recognize this group as a pathologic entity.

We believe that Greig's description of false oxycephaly fits perfectly the present classification of brachycephaly and scaphocephaly in which, as we know, partial synostosis is responsible for the clinical symptoms.

Since Greig published his article in 1926, his classification has been unanimously accepted and followed, up to the present time. In reviewing the literature no criticism of Greig's classification has been disclosed. On the contrary, his classification is followed carefully, which in our opinion is misleading and adds to confusion for the reasons previously cited.

Based on anatomical and clinical facts, the following classification is suggested.

\[
\text{Craniostenosis} \rightarrow \begin{cases} 
\text{Incomplete} \\
\text{Complete}
\end{cases}
\rightarrow \begin{cases} 
\text{Scaphocephaly} \\
\text{Brachycephaly} \\
\text{Oxycephaly}
\end{cases}
\rightarrow \begin{cases} 
\text{Compensated} \\
\text{Decompensated}
\end{cases}
\]

As may be seen in this classification, we consider the anatomical deformity of the head and the neurologic symptoms. Deformity of the skull is a result of Virchow's law: limitation of expansion in directions perpendicular
to the suture-line closure causes overexpansion to take place in the regions
where the sutures remain open.

The first is scaphocephaly involving closure of the sagittal suture with
compensatory lengthening of the skull in its anteroposterior dimension and
often widening and elevation of the frontal region.

The second is brachycephaly involving synostosis of the coronal sutures,
presenting normal growth in the anteroposterior direction. The skull be-
comes broad and short; the root of the nose is also widened so that the eyes
are placed very far apart. This condition also has been termed acrobrachy-
cephaly, but the prefix "acro" has been applied to oxycephaly as well,
creating a misnomer.

The third, oxycephaly, involves the coronal and the sagittal sutures and
prevents normal expansion both in the transverse and anteroposterior direc-
tions. Other sutures may be involved as well; the skull can expand only in
the vertical direction. This condition has been termed turricephaly, and
acrocephaly also.

In our opinion, therefore, the above terms, namely, oxycephaly, brachy-
cephaly and scaphocephaly, represent the only logical nomenclature for the
various types of craniostenosis.

The neurologic symptoms are dependent upon the degree of contraction
of the skull. The craniostenosis is decompensated when the patient develops
chronic increased intracranial pressure as evidenced by headache, vomiting,
papilledema, failing vision and convulsive seizures. Optic atrophy with blind-
ness and psychic disturbances may also occur.

An extremely striking feature of craniostenosis is bilateral exophthalmos,
especially in oxycephaly and scaphocephaly. In well developed cases the
protrusion of the eyeball may be so severe that it is impossible for the pa-
tient to close the lids and is frequently associated with strabismus. The ex-
ophthalmos may be due to the fact that the orbits are very shallow and
oblique with the anterior and middle fossae deeper than usual and shortened
in the anteroposterior diameter.

It is impossible from the neurologic symptoms to make a differential diag-
nosis between the varieties of craniostenosis, the difference being based only
on the degree of deformity of the head correlated with the roentgenographic
findings.

Craniostenosis is compensated when a compensatory growth of the skull
occurs at the open sutures so that symptoms of increased intracranial pres-
sure do not develop.

The x-ray examination enables us to determine with accuracy the variety
of craniostenosis and the degree of contraction of the skull. In oxycephaly
the coronal and sagittal sutures are seen to be synostosed. It is essential in
this condition that the premature synostosis of the sutures above mentioned
be present, although the lambdoidal suture may be involved also.

In brachycephaly the coronal suture is synostosed more or less completely
but the sagittal and lambdoidal sutures remain open in many cases long after
they should be closed.
Finally in scaphocephaly the characteristic feature from the x-ray standpoint is the premature closure of the sagittal suture while the coronal suture may be widened.

DIFFERENTIAL DIAGNOSIS

We have excluded from this study the craniofacial dysostosis of Crouzon, included by some authors, because it is very rare. In addition, the description of this condition given by Crouzon in 1929 is not clear enough to consider it as a form of craniostenosis.

As far as chondrodystrophy is concerned, included by Virchow in the group of craniostenosis, it is now definitely identified as a different pathologic entity. This condition is due to a premature union of the bones which form the base of the skull. The defective development involves not only the bones of the base of the skull, which are, as we know, of cartilaginous origin but also affects practically all the other cartilaginous bones, producing among other symptoms shortening of the arms and legs. The enlarged head seen in chondrodystrophy is produced, in some cases at least, by hydrocephalus. Dandy, who demonstrated by air injection in 191 the existence of hydrocephalus in this condition, suggested that it might be due to the shortened base of the skull which causes bending of the aqueduct or obstruction of the cisterna magna, at least during early life and later corrected.

SUMMARY

Deformities of the head which are now designated as craniostenosis were known to the ancient Greeks and were described by Hippocrates in 400 B.C. The various explanations of the pathogenesis of this condition since Virchow’s original study have been discussed and analyzed. Based on anatomical and clinical grounds a new classification of the forms of craniostenosis is offered.

REFERENCES