CLASSIFICATION OF CRANIOSTENOSIS

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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 tau, 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 ἀτα Ὠ.

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 “oxycephalides” 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.

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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