An Extreme Case of Scaphocephaly From a Mound Burial Near Troy, Kansas

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Introduction

The skull which is the subject of the present paper attracted the attention of the writers when they noticed it in an exhibition case in Dyche Museum at the University of Kansas. A check of the records revealed that it was collected many years ago by Dr. R. S. Dinsmore from a burial mound near Troy, Kansas. Unfortunately, no additional archeological information is available, due to the early date at which the skull was obtained. Since it came, however, from a mound on the Missouri River bluffs, it quite possibly belongs to the Woodland Cultural Pattern, regarded by archeologists as flourishing in this particular region some time in the neighborhood of 1300 A. D. (1)

The skull had been described in the records as "artificially deformed," but since, instead, it obviously presented a marked case of scaphocephaly, it seemed well to put a description of the skull on record. In spite of many references to scaphocephaly in both anthropological and medical literature, the subject has been briefly and inadequately treated, particularly in regard to certain endocranial features of which this specimen is a beautiful example. Cranial conditions which, to the anthropometrist are distinct entities, are not clearly differentiated by many medical writers. Moreover, it is rarely that such an ancient cranium, in perfect condition, falls into the hands of both physician and anthropologist. The writers are intent upon (1) offering a description of this skull which will furnish some adequate data, both metric and morphological upon a subject which, while often mentioned, has not been very systematically explored in recent years, and, (2) discussing the specimen in relation to the various theories held, showing wherein it appears to support one or another of them. Moreover, as mentioned previously, there has been a dearth, if not an actual lack, of adequately described specimens. It is our hope that when more records of such cranial anomalies are available, their nature may be more fully understood. Thanks to the generous co-operation of Dr. Claude Hibbard of the

Transactions Kansas Academy of Science, Vol 47, No. 2, 1944.

University of Kansas Museum, this skull has been made available to us for study. We should also like to express our appreciation to Dr. W. R. Wedel of the U. S. National Museum for advice upon certain matters pertaining to its cultural provenience.

II. Description

Scaphocephaly is generally defined as that cranial condition in which, due to premature synostosis of the sagittal suture, skull growth is magnified antero-posteriorly rather than transversely. The result is an ultradolichocephalic skull, narrow and long and lacking parietal tuberosities. The skull vault slopes quickly away from a keel-like median sagittal line. Nevertheless the cranium remains bilaterally symmetrical because of the longitudinal nature of the growth process to which the other bony elements of the skull adjust.

The present specimen is that of a mature female of approximately 30 years of age. The skull is ultradolichocephalic, gracile, and completely feminine in character. Supra-orbital ridges are en-

Measurements of Skull No. 3934, University of Kansas Museum

All measurements in millimeters

*Length	194	Auricular height	121
*Breadth	121	Circumference	511
Bizygomatic width	128	Transverse arc through vertex	285
Minimum frontal width	91	Total sagittal arc	387
Interangular width	100	Frontal arc	122
Upper face height	64	*Parietal arc	135
Total face height	103	Occipital arc	130
Nasal height	45	Frontal chord	107
Nasal width	24	*Parietal chord	126
Alveolar length	53	Occipital chord	104
Alveolar width	65	Orbital height	R.33—L.35
*Biasterionic width	99	Orbital width (dacryon)	R.31—L.40
*Width at stephanion	99	Foramen magnum length	34
Basi-bregmatic height	139	Foramen magnum width	28
Basion-nasion	103	Bicanine breadth	40
Basion-prosthion	95	Bimolar breadth	63

Cranial capacity by Pearson's interracial total ht. formula 1306cc.

Indices

Cranial module	151.	Height-Length index	71.64
*Cranial index	62.3	*Breadth-Height index	114.8
Superior facial index	50.	Cranio-facial index	71.92
Maxillo-alveolar index	122.	*Transverse cranio-facial index	105.78
Nasal index	53.33	Facial module	108.6
Foramen magnum index	82.35	Gnathic index	92.3
Total facial index	80.4	Intertemporal-Interangular index	91.
Auricular height-length index	62.37		

Bicanine index 63.49

*The measurements and indices marked by an asterisk are those which reveal most clearly the influence of scaphocephalic distortion.

MANDIBLE

Bimental breadth	46	Symphyseal height	29
Minimum width ascending ramus	33	Bigonial width	97
Bicondylar width	114	Length	100

tirely lacking, and there is only a faint trace of a glabellar prominence. The sagittal suture is entirely synostosed and obliterated, although the coronal suture is still open ectocranially throughout its length except, on each side, for a very small area at stephanion.

The parietal foramina are lacking. There is a slightly flattened and depressed area in the region of the obelion and the superior border of this region carries two rather pronounced bosses arranged symmetrically, one on either side of the obliterated suture, and placed close together. The possible significance of these two bosses will be discussed at more length later.

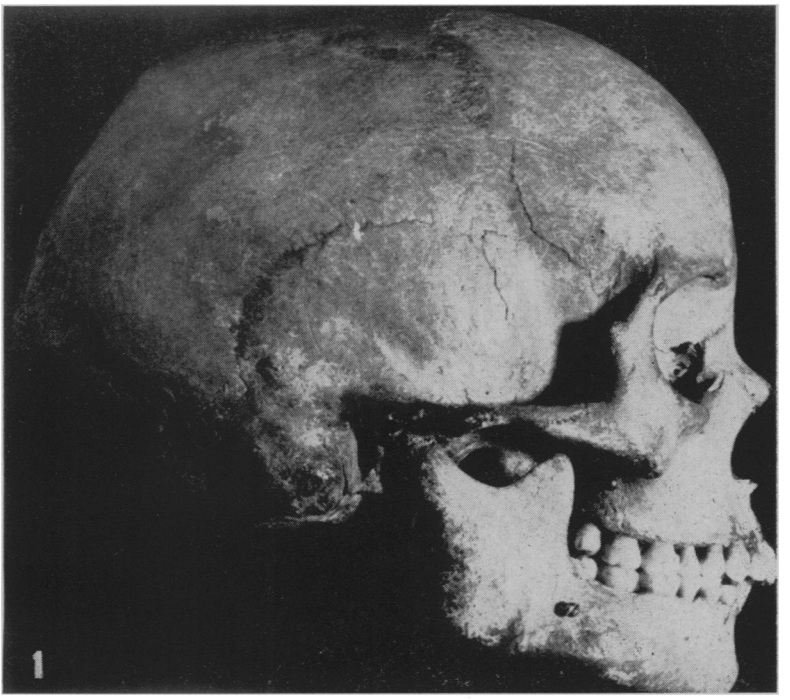

PLATE I—Norma lateralis.

The occiput is narrow and protruding. The *planum nuchale* is smooth and shows little muscular relief or rugosity. Both the superior and inferior nuchal lines are only faintly visible. The external

occipital protuberance is entirely lacking, but the *crista occipitalis externa* is faintly evident. The gracility and lack of ruggedness of the whole nuchal area is suggestive of the distinctly feminine nature of the calvarium. The zygomata are slender and delicate. In spite of the narrow skull vault, there is only a hint of phaenozygosity.

The mastoid processes are the shortest and most blunt the writers have ever seen in an adult human skull. The tips do not project beyond the *pars lateralis* of the occipital bone. The glenoid fossae are small and shallow but in no way abnormal.

The face is short, the frontal angle high, with a full and prominent forehead. The nasal root is low but not pronouncedly so. The nasal index is platyrrhine. There is a nasal spine of medium length and the nasal sill is short and well defined. The canine fossae are slightly evident. The upper incisors show the "shovel-shaped" condition so typical of Mongoloids, and there is a rather marked degree of overbite. The dentition is almost perfect. There is evidence of only moderate wear and the loss of a single tooth—a first left premolar—during life. The third molars have all erupted properly, and there is only one cavity—a large one—in the left lower second molar.

The optic foramina are more oval than round, as if compressed from side to side. The superior and inferior orbital fissures seem narrower than usual; the greater wings of the sphenoid appear to have encroached on them laterally.

The body of the mandible is, transversely, rather thick and stocky, particularly in the molar region, but is in no way remarkable. Both mental foramina, however, are very small, almost abnormally so. They lie beneath the second premolar. The lingula overlying the mandibular foramen is unusually thick and blunt. The basilar aspect of the skull presents little in the way of unusual features except the slightly drawn out aspect of the foramen magnum, apparently in response to the unusual rearward protrusion of the occipital bone. A pharyngeal fossa replaces the tubercle of that name, but there is a rather high incidence of this anomaly among Amerinds.

Barring the premature ossification of the sagittal suture, there is little of an unusual nature to be noted about cranial closure. The coronal suture is pronouncedly complex in the region overlying stephanion and the lambdoid similarly throughout its biasterionic portion. No Wormian bones were noted. The H form of pterion characterizes both sides of the vault.

Examination of the inner anatomy of the skull is limited to information which can be gained by inspection and palpation through

the foramen magnum, since the skull is not sectioned.

The inner table of the bones of the vault is intact; an unusual series of markings is seen on all visible bony surfaces. These markings are palpable on the occipital bone. They are less marked on the calvarium than nearer the base of the skull, where they assume the form of shallow broad grooves separated by low crests of bone. The irregular pattern, breadth and depth of groove, and form of the ridged boundaries follow the pattern of cerebral convolutions and undoubtedly represent "convolutional atrophy," also known as "digital impressions." Their significance will be indicated later.

The vascular markings are of special interest. The anterior and posterior branches of the middle meningeal artery are prominently represented down to very small rami. Lying directly in line with the fused sagittal suture is a groove of varying breadth (estimated 3 to 6 mm.) and of a depth unknown in the experience of the authors. Starting as a shallow impression anterior to the coronal sulcus, it deepens rapidly until at the torcular Herophili (sinus confluens) it is so deep that the palpating finger may not be pressed against its bottom. At this latter point it turns to the right. This groove follows the course of the superior sagittal sinus. Bilaterally the transverse sulcus is easily palpated in the rim of the posterior cranial fossa, at first shallow posteriorly but deepening as the palpating finger follows it laterally and anteriorly. Parasagittally, at about vertex and just anterior to the coronal sulcus, irregular pits and depressions are seen bilaterally which suggest the Pacchionian markings.

No endocranial marking may be distinguished in the region of the pair of eminences referred to ectocranially as lying near the obelion.

The sella turcica and hypophyseal fossae do not seem enlarged or altered in shape. The anterior and posterior clinoid processes on the right are fused to form a bony bridge.

The sagittal suture is completely fused throughout its visible length. The coronal suture is open endocranially, except in its most lateral aspects; however, no light is transmitted through it. The visible portion of the squamosal suture is open.

In no place does light thrown into the skull trans-illuminate the calvarium. Neither frontal nor maxillary sinuses trans-illuminate.

A table of anthropometric data is given. It will be noted that all of the measurements which are in the least unusual pertain to the excessive longitudinal growth of the parietal, largely in a posterior

direction, and the necessary adjustments made by the other bones to the narrowed vault. Such an index as the transverse cranio-facial,

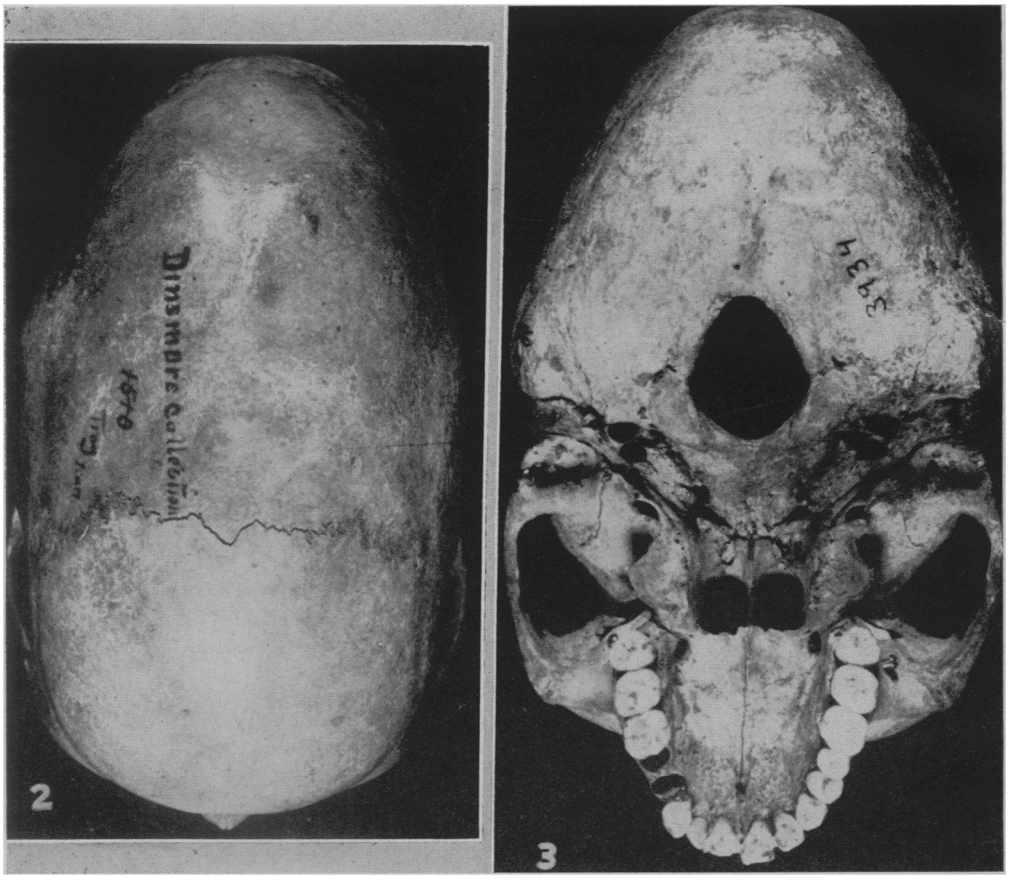

PLATE II—Norma verticalis. PLATE III—Norma basalis.

for example (105.78) is of a noticeably primitive character because of the narrowness of the vault in comparison with the zygomata. We have starred in the above tables certain indices which reflect the nature of the pathology. The basilar region of the skull anterior to the foramen magnum seems less affected, in terms of width, than the drawn-out occiput. This difference can be noticed in the photographs and is also reflected in the low biasterionic width of 99 mm. The skull gives an appearance of complete bilateral symmetry and adjustment despite its obviously pathologic nature.

III. Etiology

Dye and Kinder have commented that: "Of the factors which have been considered important in determining the shape of the skull, the following may be mentioned: 1) intelligence, 2) brain development, 3) growth of sinuses, 4) heredity, 5) growth of sutures, and 6) muscle development and muscle pull or pressure. No two investigators will give quite the same importance to each of these factors."([2]) From the information available on examination of the Troy skull, it is possible to comment primarily only on the fifth and sixth of these, and incidentally on the second and third.

One of the most conspicuous, and perhaps fundamental, deviations from normal in this cranium is the complete fusion of the sagittal suture. Bonnet has classified suture closure according to the time at which it occurs with reference to brain growth. As quoted([3]) he lists:

1. Premature synostosis, where closure occurs before full brain growth is reached.
2. Synostosis praecox: suture closure after complete brain growth but before normal obliteration.
3. Senile suture obliteration; a normal phenomenon of middle age and later.

With respect to age groups in the human being, it may be remarked that brain volume at birth is about 330 c.c.; at one year of age, 950 c.c.; average adult brains vary around approximately 1450 cc. as a norm, over 1300 cc. of this increase being achieved by the age of ten years.([4]) The age-group twenty-six to thirty is the elective period for suture obliteration, both ecto- and endo-cranial.([5])

Increase in brain volume is seldom inhibited by the occurrence of premature synostosis. Adequate cranial capacity is provided by continued bony growth, but the shape of the cranium and its contents may be altered. If two cranial bones undergo premature bony union, the normal lateral growth of bone will be inhibited in a direction which is perpendicular to the obliterated suture line (since it is at the non-fused bony margins that growth normally occurs) and the compensatory growth will occur in other directions. The nature and amount of deformity will be a function of three factors: 1) the sutures which fuse; 2) the completeness of the fusion; and 3) the amount of brain growth which follows the fusion.

In intramembranous osteogenesis (the type of bone formation in the skull vault) the advancing bone edge is preceded by osteoblastic activity in a fibrous connective tissue membrane of the collagen-

ous variety. When bone from two adjacent centers of ossification approaches a juncture, synostosis is prevented for a time by continued new formation of this "membrane," thus maintaining the classic articulation known in arthrologic terminology as "suture": a union of skeletal elements by a continuous intervening layer of fibrous connective tissue. When synostosis occurs, it is not so much a positive phenomenon on the part of bone as it is a failure of this fibrous connective tissue to maintain its interposition by continued replacement of itself.

The etiologic theories which have been based on premature synostosis revolve about three main ideas:

1) Virchow's concept of some inflammatory process in the fetal cranium which deprives the bone margins of their normal growth potential and possibly, due to the fact that calcification is not an unusual sequela of inflammation, promotes the suture closure.

2) A defect in the blastemal skeleton, perhaps as a result of a congenital abnormality in the interstitial mesenchyme, or from injury in very early embryonic life. On the basis of such considerations as the occasional association of syndactylism with skull deformity, it is believed that the mesenchymal abnormality must date from earlier than the seventh week of embryonic life.

3) Displacement of primary ossification centers of the synostotic bones toward each other. Cases have been reported in which the ossification centers of separate bones were merged into one common center in the fused suture.

Another set of theories of the origin of these skull deformities is founded not on premature synostosis as primary, but on localized changes in the skull. Two such concepts are:

1) Defects in the sphenoid bone. Scaphocephaly has been noted very frequently in connection with underdevelopment of the greater wings of the sphenoid; the oxycephalic (or "steeple-skull") deformity, concersely, is often associated with abnormally large greater wings. Ogilvie and Posel([6]) describe cases in support of the concept that the primary defect is basilar, with consequent alterations in shape of the brain case to accommodate the broader or narrower sphenoid.

2) Generalized basilar hypoplasia. Gunther is quoted([3]) as stressing the importance of such developmental

failure, the mechanism of production of deformity possibly being related to disturbances in blood supply to the developing bones of the calvarium, resulting from narrowing of the basal vascular foramina. The hypothesis, attributed to Broca, that an abnormally shaped brain is the primary cause of deformity is receiving very slight attention at present.

Very few studies on pathology are available. Of cases diagnosed while living, roentgenographic studies furnish the only information to supplement that gained from inspection, palpation, and craniometry. There are obvious difficulties involved in following such cases to an autopsy so complete that the skull is examined freely. On the other hand, the occasional diagnosis made on skulls from osteologic collections is very infrequently accompanied by any data regarding soft-part anatomy or symptomatology and functional defect during life. Of the case here presented, no such data are available save by implication.

Classifications of this group of skull deformities reveal the difference in emphasis from the standpoint of the physician or the physical anthropologist. When described at all in the medical textbook, seldom is any term except oxycephaly applied. Although there is general agreement as to the appearance of the deformity, such emphasis seems to result from the clinical impression that this skull type of all the group, is the most productive of spectacular symptomatology. Greig's([11]) classification may be representative: when synostosis involves all the sutures of the head and face, we have true oxycephaly; with one or a few synostosed sutures, the term pseudo-oxycephaly may be applied. Greene and Brown ([7]) observed synostotic deformities in the rabbit which, although they are not maintained to be identical with those in man, have a great deal in common with them (including, probably, mode of origin). Greene's specific definitions ([8]) of the deformities should be of value in clarifying classification systems; they are repeated here.

1. Scaphocephaly is used in reference to the deformity resulting from fusion of the sagittal suture.
2. Plagiocephaly refers to that deformity resulting from fusion of a single segment of the coronal, right or left.
3. Trigonocephaly refers to fusion of both coronals.
4. Oxycephaly refers to fusion of the sagittal and both coronal sutures.

In oxycephaly the synotosis may or may not involve the entire suture, where elevated bony ridges mark the line of fusion. Diploic tissue is scanty, and the inner table of bone may be absent through pressure atrophy. Characteristic is the "convolutional atrophy" of the endocranium, in which shallow bony grooves afford a map of the gyri and sulci of the cerebral cortex. The cranial base is hypoplastic and depressed, with middle and posterior fossae especially deep and foreshortened. Sella turcica and hypophyseal fossae are usually unaltered. The sphenoidal greater wings are often unusually large. The face may be broadened; orbital foramina are occasionally narrowed. Radiographically the paranasal sinuses are often small or absent.

Scaphocephaly differs from oxycephaly only in two essential respects: the shape of the head and the less extensive resultant deformity. The sagittal suture is primarily involved, with a narrow skull devoid of parietal eminences. The convolutional atrophy and thinning of the skull is present as in oxycephaly; orbital change is inconstant. A change rather constantly remarked is the hypoplastic state of the greater wings of the sphenoid bone.

Defects most constantly associated with premature synostosis include syndactylism, polydactylism, joint structure ankylosis and deformity (most commonly of the elbow), hemolytic icterus, and vascular anomalies. The relationship of all of these conditions to the mesenchyme has been pointed out as evidence in support of an interstitial mesenchymal etiology.

Deviations from normal range of skull anatomy in the present specimen requiring comment in view of the foregoing etiologic and pathologic generalities follow.

The fusion of the sagittal suture is complete, involving both inner and outer table, throughout the length of the suture. That this suture closure is intimately related to the classic scaphoid form of the cranium is demonstrated by examination of indices $\frac{\text{chord} \times 100}{\text{length}}$, where the chord is measured antero-posteriorly over the surface of the bone concerned and the length is nasion-basion. For the sagittal chord, this index is 64.4, a distinct elevation over normal range. The frontal, with its index similarly computed, is low with 55.1, while the occipital bone is within normal range for dolichocephalic skulls with 53.6. The bony disproportion is, therefore, especially marked in terms of parietal bone elongation and (compensatory ?) frontal bone foreshortening.

The two prominences near obelion, close to the sagittal suture

and lying almost midway between lambda and bregma, are of special interest in relation to the theory that premature synostosis is a result of displaced ossification centers. Martin says of scaphocephaly: "The ridge or keel-shaped elevation of the cranium (culmen cuneiforme) is brought about through a smoothing-out of the parietal bones, and has origin in the synostosis of the sagittal suture, usually beginning in the fetal period in the region of obelion and completed within the first decade of life."([9]) In the absence of the usual parietal eminences the masses indicated may represent the displaced center; in this skull they attain a prominence not described specifically to our knowledge. Their presence does not invalidate the theory based on abnormality of the interstitial mesenchyme; a defect in the germinal tissue separating the two ossification centers would account for their displacement toward each other. The flattened, depressed area between them may be of interest from this aspect.

In view of the prominence attached to the sphenoid bone in etiological hypotheses, it is surprising that the sphenoid of this specimen does not appear to deviate markedly from normal. In the absence of metric criteria of sphenoid form, this observation, as also those in the literature, must remain only an impression.

The appearance of constriction and hypoplasia of the basilar foramina does not necessarily establish evidence for the basilar hypoplasia theory of etiology, since the anomaly could be secondary to vault changes. In Greene's ([7]) craniostotic rabbits, the base of the skull (including the sphenoid) was still cartilaginous while suture union of the calvarium was being established (as early as the third week of embryonic development). The narrowing of the foramina does, however, have special interest in attempting to account for the anomaly remarked in the next paragraph.

The endocranial groove for the superior sagittal and portions of the transverse sinuses is markedly deepened. This deepening is of more interest from the standpoint of bone formation and mechanics than etiology. If the narrowing of foramina around emergent channels should have impeded the normally free outflow of blood, it may be hypothecated that the pressure of blood within the dural sinuses rose. No great pressure differential is needed to account for bone erosion and resorption; Policard has emphasized to the contrary that "very weak pressures are capable of provoking absorption"([10]). The continuous application of the pressure is a necessary condition. It would be possible to lay less emphasis on the venous sinus pressure here if the sagittal sinus alone were involved, in which case the

groove might be more directly related to synostosis of the overlying suture. With the involvement of the lateral aspects of the transverse

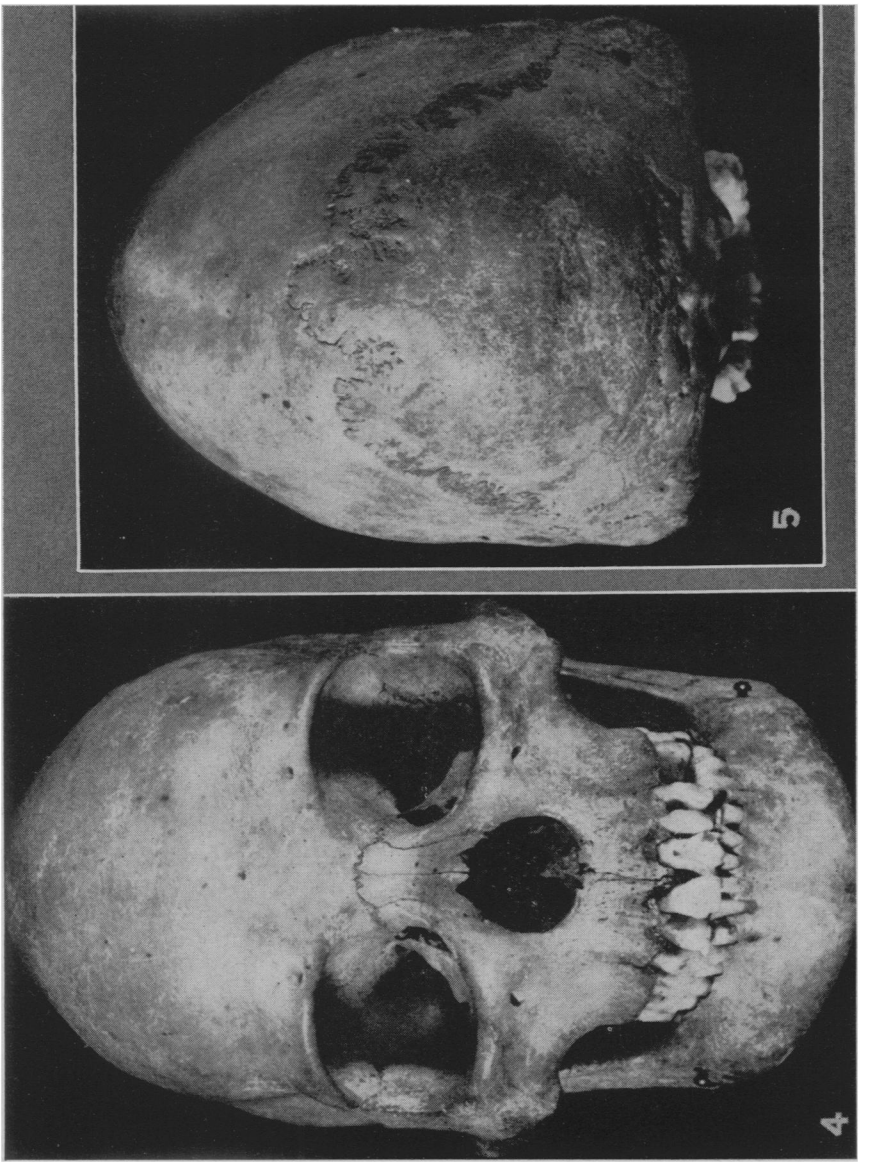

PLATE V—Norma occipitalis.

PLATE IV—Norma frontalis.

sinus, however, and the great depth of the torcular Herophili, a more generalized explanation is needed.

The convolutional atrophy, as well as meningeal vascular markings, on the inner table of the skull agrees with previous descriptions, and has been accounted for by the suggestions of increased intracranial pressure. The mechanism of this atrophy is not clear; the accuracy of reproduction of gyri and sulci would seem to require a reduction in the amount of fluid in subarachnoid spaces, such that brain tissue might press directly against bone. It is even more remarkable to note that convolutional atrophy is a frequent finding, while clinically patients seldom show convulsions or epilepsy. The brain cortex, while highly sensitive to pressure, seems yet not so sensitive to continuous minute pressure elevations as is the bone covering it; here again, evidence is afforded for the general concept of the great plasticity of bone.

The bridging of the clinoid processes has not been recorded as a part of the syndrome. Although it is probably an incidental finding here, it should be noted that clinoid abnormalities are not unknown. The oxycephalic and plagiocephalic rabbits studied by Greene showed the posterior clinoids to be slightly longer and directed more laterally than normal. No attempt to relate these observations can be made at present.

The mastoid hypoplasia is, like the clinoids, not described as a concomitant deformity in scaphocephaly. If re-examination and future experience should establish it as a part of the characteristic syndrome, it would appear preferable not to interpret it in terms of primary or etiologic significance. Although no data for an analysis of musculo-skeletal mechanics can be had for the present specimen, the following line of speculation is of interest. Traction has a predominant effect on the determination of number and arrangement of osteoid fibrils.([10]) The mechanism is physico-chemical, possibly by orienting the colloidal micellae of the collagenous substance in the direction of line of traction (similar to orientation in India rubber and in photo-plastic substances). The mastoid process is a traction apophysis, and its development and size are dependent on the activity of the sterno-cleido-mastoid muscle, much of which attaches to it. In man, the sterno-cleido-mastoids, acting bilaterally, cause the head to tilt forward; with hyperextension, they may increase the upward and backward tilt. (This potentially double activity is, of course, a result of their attachment on to the mastoid processes at a site almost directly in line with the axis about which the nodding movement

occurs.) If, through the distortion of the scaphocephalic head, normal distribution of forces should be shifted on the anterior and posterior arms of the lever whose fulcrum is at the occipital condyles, it is conceivable that less power might be required of the sterno-cleido-mastoids in moving or fixing the head, with resultant diminution of the traction force on the mastoid site of attachment. In support of this relationship of sterno-cleido-mastoid muscle to mastoid process, three types of mastoid hypoplasia not associated with synostosis may be cited:

1. Cleido-cranial dysostosis, in which (with clavicular aplasia) there is a lack of formation of the cleido-mastoid portion of the sterno-cleido-mastoid muscle. Greig's case shows an absence of the mastoid processes.([11])

2. Purves-Stewart's case showed an association of unilateral absence of the sterno-cleido-mastoid with absence of the corresponding mastoid process.([12])

3. In torticollis, or "wry-neck," the mastoid processes are asymmetrical; the larger process is that to which the contracted muscle attaches.

The anomalous lingula of the mandible is difficult to place. Although possibly an incidental observation, it should be remarked that this process is the site of attachment of the spheno-mandibular ligament, running from the angular spine of the greater wing of the sphenoid. It may be a related and secondary phenomenon resulting from some disturbance of skeletal mechanics.

IV. Conclusion

The anthropometric and medical data presented in this paper are intended to place on record a thorough description of a scaphocephalic cranium. Details, which are sadly lacking in most of the literature upon the subject have been presented at some length, and attention has been drawn to several features not previously noted in the literature dealing with this type of cranial disorder. Notable in this respect are the deeply channeled cranial sinuses, the peculiar form of the mastoids, and the two curious bosses near obelion, which may represent displaced ossification centers. Theories as to the cause of the defect are summarized and discussed. No other pathology is suggested, and there is no evidence to indicate, in this particular specimen, that the scaphoid condition was responsible for any marked physical disorganization or failure of the bodily processes. The skull is a beautifully symmetrical specimen of its kind, perfectly preserved, and worthy of archeological and medical attention.

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