

An unusually-wide human bregmatic Wormian bone: anatomy, tomographic description, and possible significance

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Abstract Supernumerary ossicles (or Wormian bones) of the cranial vault are formations associated with insufficient rate of suture closure, and regarded as “epigenetic” and “hypostotic” traits. These bones rest along sutures and/or fill fontanelles of the neonatal skull. In this autoptic report of a 66-year-old Caucasian woman, a peculiar supernumerary bone is described, unusual size and shape, filling completely the bregmatic fontanelle. The skull was CT-scanned through coronal sections at 80 kV and 60 mA, with a slice thickness of 1.0 mm and a resolution of 0.35 mm/pixel. Segmentation and 3D rendering were computed using MIMICS 7.0 (digital endocast). The bone was pentagonal and remarkably large, more on the exocranial surface than on the endocranial one, involving both tables and diploe of

the vault. This feature might represent a wedge to completion of the vault architecture. Considering the functional and structural matrix of cranial morphogenesis, this case displays the possibility of discrete diversification of the ossification centres, as well as the relative stability of the structural skull matrix in response to discrete changes.

Keywords Wormian bones · Ossicle at bregma · Human skull · Gross anatomy · Digital endocast

Introduction

Cranial discrete traits (also called non-metric, discontinuous, or “epigenetic”) are “intrinsically innocuous minor skeletal variants of the human skull” [8]. Despite the term “epigenetic” was originally applied to discrete traits in order to emphasize their possible modification during ontogeny, subsequently this term has been used to stress the difference of these features versus those of the so-called “Mendelian traits”, the latter being determined by a direct gene–character relationship and completely independent on the environment [8]. These traits received attention also in the study of fossil hominids, as a source of information about growth patterns in extinct taxa [10, 12]. This evidence points to the fact that the formation and distribution of supernumerary ossicles may reflect different stress types (tension, pressure, etc.), acting on the cranial vault during late prenatal and early postnatal periods of bone growth.

Among cranial epigenetic traits, supernumerary ossicles (or Wormian bones) are irregular in shape and size, and located in the sutures or fontanelles. As other discrete traits, their formation and prevalence may be under genetic control even though the inheritance of these bones has not been perfectly assessed so far. They might be under the control

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of a number of genes with additive action (polygenic complex) and their phenotypic expression is conditioned by developmental thresholds [6]. However, external factors of stress probably act, at least on their frequency, such as artificial cranial deformation [15, 19], and all factors acting on the sutures [11]. Occurrence of craniosynostosis may also be accounted for in this way, affecting the presence of Wormian bones by altering the vectors of skull growth and development [15]. Even the mechanism responsible for the formation of Wormian bones is not fully ascertained. These ossicles could represent additional ossification centres in the fibrous tissue occurring during late fetal ages or postnatally, which separate from the primary centres of larger cranial bones [18]. Furthermore, factors altering normal tensile forces produced by the growing brain, such as artificial deformation devices, can generate localised areas of compression and tension along the sutures, resulting in their premature closure and/or can modify the postnatal growth gradients altering bone formation in sutures and fontanelles [9].

In this report, a singular Wormian bone at bregma is described. As far as we know a bregmatic bone, like this, unusual because of its large dimensions and peculiar features, has never been reported so far.

Case report

Dissection was performed on a 66-year-old Caucasian, white-skinned woman, who was a body donor for medical training and research. Only the bones of the cranial vault have been examined in this study, after routine resection of the calvaria to expose the brain surface for teaching.

A singular bone filling the bregmatic fontanelle was noted on both the exocranial and endocranial surfaces of the vault (Figs. 1, 2). In correspondence of the bregmatic bone, the vault showed an outer regularly convex and an inner concave surface, according to and as a completion of the curvature of the calvaria. This was a single, independent bone, and appeared almost regularly pentagonal in shape, slightly rotated toward left (i.e. anticlockwise), as shown in Fig. 1. It displayed an anterior apex, two antero-lateral and two postero-lateral margins and a posterior (base) margin. The antero-lateral margins were joined with the frontal bone, the postero-lateral ones with the parietals. The right and left lateral vertices between the antero- and postero-lateral margins were joined with the coronal suture (with a subtle discrepancy on the left; see Fig. 1), and the base of the pentagon posteriorly in the middle with the sagittal suture. It was apparent that the cranial sutures were in continuity with the sutures around the bone. However, the antero-lateral margins were delimited from the frontal bone by less dentate sutures than those delimiting the postero-lateral

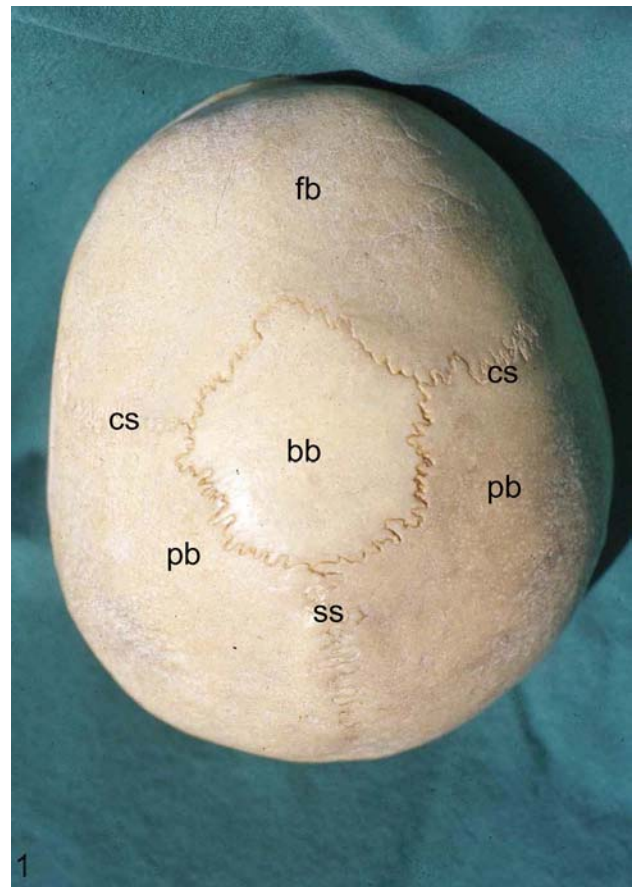


Fig. 1 External surface of the cranial vault. The unusually wide pentagonal bregmatic bone (bb) is located between the frontal bone (fb) and the parietals (pb); its lateral vertices are joined with the coronal suture (cs), and the middle point of its base with the sagittal suture (ss). The sutures delimiting this bone are moulded according to the normal pattern of the coronal and sagittal ones

margins from the parietals. These sutures were moulded according to the normal pattern of the coronal and sagittal suture respectively, so that they appeared to be the direct continuation of those.

On the external table the length of the left antero-lateral margin was 40 mm, of the right one 35 mm, of the left postero-lateral 20 mm, of the right one 30 mm, and of the base 40 mm (Fig. 1). On the internal table the sides measured 35, 30, 20, 25, and 30 mm, respectively (Fig. 2). On the whole, this bone was wider on the external aspect, than on the internal one. In fact, its sagittal axis was 57 mm and the transverse 56 mm on the exocranial aspect, and 46 and 55 mm, respectively, on the endocranial one.

When observed on the internal surface, the bone showed granular foveolae, more numerous in the posterior part, in continuity with those occurring in the parietal internal table (Fig. 2), as commonly found in recent European populations [2].

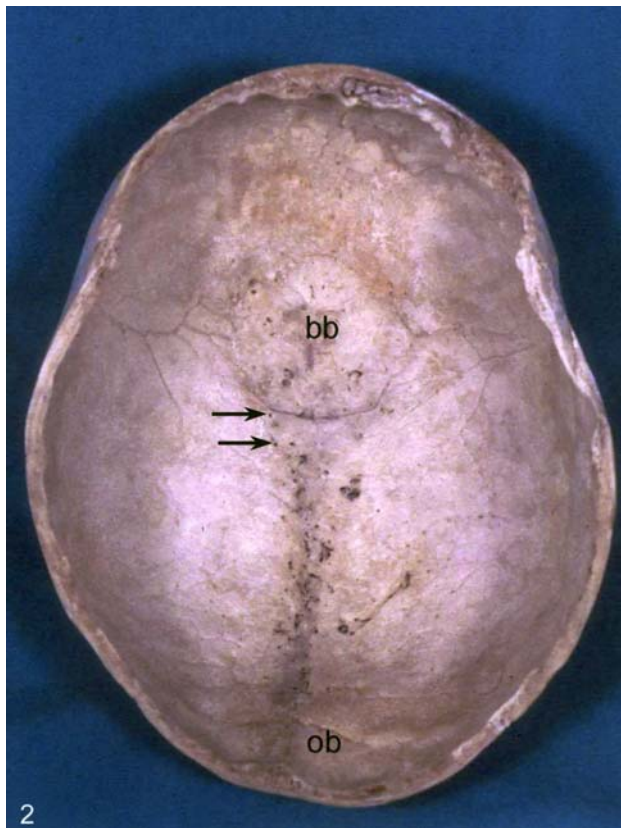
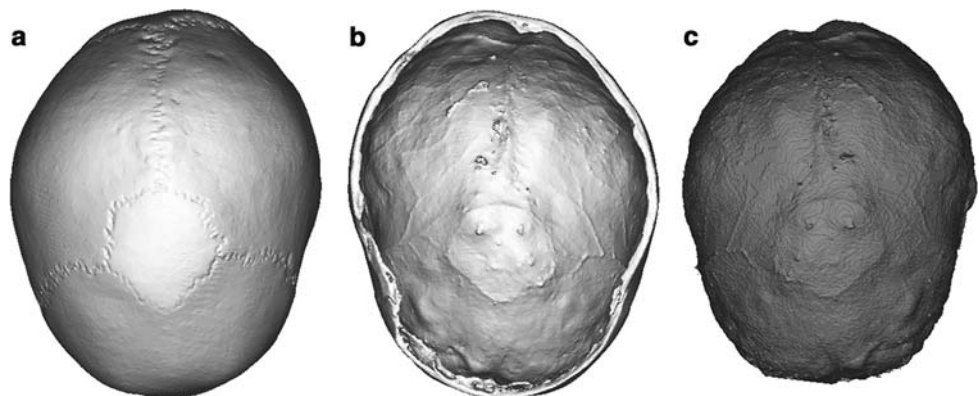


Fig. 2 Internal surface of the cranial vault (*ob* occipital bone). The bregmatic bone (*bb*) shows a pentagonal shape also on the endocranial aspect, although its dimensions on this side are inferior in comparison with the external surface. Granular foveolae (*arrows*) are seen more numerous in the posterior part of the bregmatic bone. These are in continuity with the foveolae on the parietal internal table, as it is common for aged skulls

The vault was CT-scanned through coronal sections at 80 kV and 60 mA, with a slice thickness of 1.0 mm and a resolution of 0.35 mm/pixel. Segmentation and 3D rendering of the exocranial and endocranial volumes were computed using MIMICS 7.0 (Materialise, Leuven). The digital replica (Fig. 3a, b) and endocast (Fig. 3c) of the specimen

Fig. 3 Digital replica of the specimen after CT segmentation and 3D rendering of the cranial vault. The exocranial (**a**) and endocranial (**b**) surfaces, and the endocast (**c**) are shown



are shown. Internally, the pentagonal shape of the bregmatic bone was clearly recognisable and the main sutures were still visible but entirely closed. There was a certain folding of the inner table at the lambdoid suture (Fig. 3b).

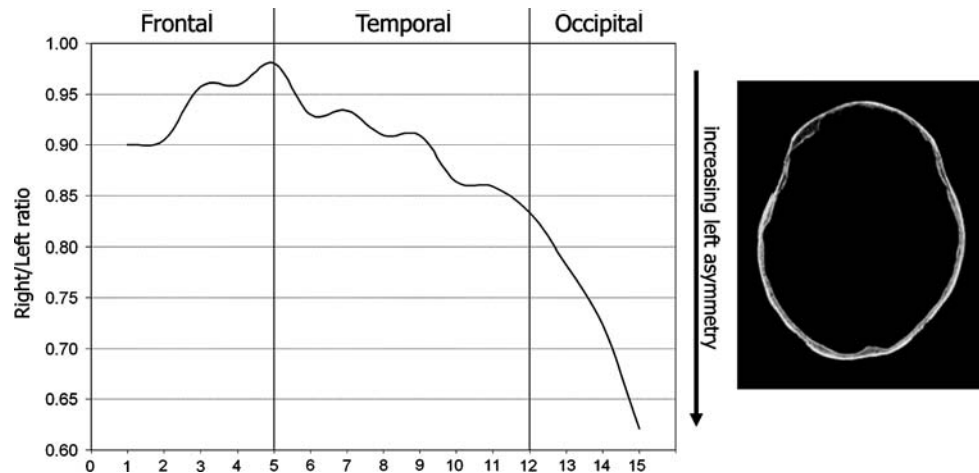
On the endocast (Fig. 3c) the frontal cortical sulci were visible, showing some additional ridges of bone deposition at the left prefrontal cortex (middle tract of the first and second frontal circumvolutions). The temporal gyri were particularly recognisable, with a large boss at the middle portion of the first temporal circumvolution.

The left and right hemispheres were 161 and 159 mm long, respectively. The endocranial maximum width was 131 mm at the temporo-parietal areas, and 98 mm at the frontal lobes (Broca's cap).

The right frontal pole was just slightly longer than the left one, while posteriorly the left hemisphere clearly exceeded the right one backwards. The endocranium was characterised by an evident asymmetry of the posterior region, and general dominance in size of the left hemisphere (Fig. 3c). Along a transect approaching the midsagittal plane from the internal frontal to the internal occipital protuberance, the right and left widths were sampled on 15 equally spaced intervals using the lower transversal CT profile, and the right/left ratio is reported accordingly (Fig. 4). The right frontal lobe was slightly narrower than the left one. The left dominance increased backwards, with the right lateral diameters being less than 65% of the counterlateral part at the occipital poles. Therefore, this specimen showed only partially the common right-frontal left-occipital petalias (torque), the left hemisphere always being wider and the degree of asymmetry strongly increasing backwards. However, such pattern does not depart from the normal range of human variation. Also when the parietal lobes were considered, the left hemisphere was larger. From a posterior view, the parietal areas were gently curved, but steeply inclined from the midline to the supramarginal gyrus, at which level the maximum endocranial width lay.

Pathological alterations of the diploic thickness were not recognised using the present CT resolution.

Fig. 4 Ratio between right and left hemispheric width. This is reported along the midsagittal profile, along a transect at 15 equally-spaced locations (from the frontal to the occipital areas), using the lowest available CT section (on the right)



Discussion

The ossicle at bregma is certainly a rare occurrence. In a recent survey on a sample of 127 deformed and undeformed crania, no bregmatic bones were detected [15]. Hauser and De Stefano [8] reported that in a series of 100 male adult middle Europeans there was none of these ossicles. The same Authors summarised their incidence in several cranial samples from different geographical regions. Some samples showed no ossicle at bregma, whereas in the others the incidence was low, from a minimum of 0.2% for a sample of 657 Bohemians (historical period) to a maximum of 2.5% for a sample of 260 Sardinians (unknown age). In an ongoing analysis at the Museum of Anthropology “G. Sergi” (University “La Sapienza”, Rome), in a sample of 400 recent Romans, ossicles at bregma have been found in four cases only. The prevalence of ossicles at bregma would be similar in both sexes [8]. According to Hauser and Bergman [7] ossicle at bregma is significantly correlated to the sagittal ossicle and the ossicle at lambda.

Concerning the possible origin of the bone described in this report, unusual because of shape and size, it should be pointed out, according to Oostra et al. [17], that supernumerary bones resulting from normal, non-fused ossification centres, are different from bones deriving from additional centres within the sutures (sutural bones) and fontanelles (fonticular bones). In fact, supernumerary bones cause abnormal partition of the cranial vault (e.g. the so-called Inca bone, corresponding to the position of the lambdoid fontanelle), whereas sutural and fonticular ones are independent bony formations, probably associated with a genetic predisposition. It should also be noted that in mice the frontal bone is neural in origin whereas the parietal bones are mesodermal [13]; therefore, the coronal suture is the boundary between the neural crest and the mesoderm. This suture is an important regulation centre of the cranial vault growth, because the boundary between the two tissues

of different embryonic origin is essential in mediating their interaction, on which the vault development is dependent. If this boundary is lost (for instance, as a consequence of a gene mutation), so that mixing of the two tissues occurs, a craniosynostosis may result [13]. At this point, it must also be added that the development of the vault bones is strictly coordinated with that of the brain [20]. Signals from the dura mater such as fibroblast growth factors (FGFs) might regulate suture patency [16].

Concerning the role of mechanical stress in affecting Wormian bone formation, no significant difference as to incidence of these ossicles in artificially deformed and undeformed skulls of Southwestern Native Americans has been reported. This might suggest that the artificial cranial deformation does not affect the incidence, but rather the number of the ossicles, once they are present [3]. On the other side, it has been reported that sagittal synostosis redirects the growth vectors of the neurocranium in an antero-posterior fashion [14], possibly leading to an increased number of lambdoid ossicles, with a slightly decreased frequency of coronal Wormian bones [15]. In general, the relationship between mechanical factors (such as cranial deformations), skull morphogenesis, and Wormian ossicles, is rather supported, and quite relevant for the studies on functional craniology [15]. Finally, these ossicles must be considered the result of a lack of complete balance between cranial changes in size (growth) and shape (development) during ontogenesis [4].

Nevertheless, it is worth noting that the unpaired structure described in this paper could not represent a supernumerary ossicle, at least according to the current definition associated with the structural processes related to the Wormian bones. Considering the unusual size of this structure, the regular shape, and also the lack of evidence of peculiar morphological patterns (other epigenetic traits, large asymmetries, synostoses), it should be regarded as a supernumerary bone more than a supernumerary ossicle. According

to the peculiar position (the bregmatic fontanelle, and the neural crest-mesoderm boundary) it should represent a true additional ossification centre, more than a compensatory response of the neurocranial system. In fact, the anterior fontanelle is generally the largest and most persistent one, even if true Wormian bones have rarely been found inside [1, 5, 21]. The anterior fontanelle is usually rhomboid or quadrangular in shape, and therefore not consistent with the shape of the ossicle at bregma described in this report, that is pentagonal. Such discordance might provide further evidence about the etiology of the bone, as deriving from a proper additional ossification centre and representing a true independent entity, enclosed by the flat bones of the skull.

Finally, the difference in size between the axes of the exocranial and endocranial surfaces means that its sutural facets were obliquely-arranged, converging toward the endocranial surface. Therefore, this bone might represent a wedge to completion of the vault architecture.

Conclusions

Taking into account the information available, two hypotheses must be considered. First, this supplementary bone is the result of a biomechanically-induced morphogenesis that on the whole does not involve the general pattern of the upper vault bones. Soft structures like the meningeal introgression of the falx cerebri have a major role in the neurocranial organisation, and may deeply influence the bone morphology. Alternatively, this bone can be the result of a definite non-adaptive genetic change, producing a singular unpaired structure. Often discrete traits are said to be genetic in the sense that they are heritable but, as discussed, they are the ultimate product of a biomechanical system. On the other hand, this bone may be the result of a direct genetic influence. In both cases, what is really worth noting is the stability of the vault morphology to such marked and discrete variation. In fact, although this individual presented an additional vault bone, the bone layers showed no visible alteration, the vault morphology did not depart from the common human variability, and the endocranial traits did not display peculiar features. This must be interpreted in terms of stability of the vault morphology, organized through levels of integration and modularity that are able to support large discrete changes without major rearrangement. All these aspects are of great importance when considering the craniofacial pattern of variation within human evolution.

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