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Wormian bones in a general paediatric population

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KEYWORDS
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Abstract Wormian bones are small bones that are often found within the sutures and fontanelles of the skull. When a child presents an unexplained fracture or fracture(s), osteogenesis imperfecta is usually suggested when an "abnormally high number" of fractures are seen.

Purpose: To assess the frequency, number, and topography of wormian bones in a "normal" paediatric population.

Materials and methods: In a population aged from 0 to 3 years, we retrospectively analysed 605 CT brain scans carried out for a range of indications, excluding cases in which there was a suspicion of constitutional bone disease.

Results: In our population, wormian bones were found in 53% of children (n = 320): 43% of the children had between one and three (n = 260), 10% had four or more (n = 60), and 6% had five or more (n = 40). There was no significant relationship between the number of wormian bones and the various indications that had led to the CT scan being carried out. Wormian bones in the lambdoid suture were found in by far the greatest numbers.

Conclusion: Wormian bones are common and can sometimes be numerous without necessarily pointing to osteogenesis imperfecta, since 10% of the children in our study had at least four.

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Wormian bones are small bones that are often found within the sutures and fontanelles of the skull. They are often considered to be a simple anatomical variant. Nonetheless, they are more commonly seen in patients with certain kinds of bone dysplasia such as cleidocranial dysostosis, pycnodysostosis, congenital hypothyroidism, rickets, but above all osteogenesis imperfecta, which is the main differential diagnosis from abuse.

It is for this reason that the search for wormian bones is an important step in the radiography investigations carried out for unexplained fracture(s) in children, in order to be able to clarify: is this child subject to physical abuse or does he/she have brittle bones?

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Indeed, an “abnormally” high number of wormian bones is thought to be a strong argument in favour of osteogenesis imperfecta (types I and IV in the Silence and Glorieux classification).

This association between wormian bones and osteogenesis imperfecta has been known of for a very long time but it was established more specifically in an article by Cremin et al. [1] dating back to 1982.

There are other, less well-established associations with CNS abnormalities that have also been described, but these remain very controversial.

Presently, we have very little data on wormian bones in normal populations and no indication of the threshold beyond which they must be reported to the clinician.

The purpose of this work is therefore to bring together data about wormian bones in a “general” paediatric population, in which there is no suspicion of constitutional disorders of brittle bones.

Our analysis took in the presence, topography, and number of wormian bones by age and by the indication for the CT scan that had been carried out.

Materials and methods

Among a paediatric population aged between 0 and 36 months who had undergone investigations in the paediatric radiology department of a university hospital, we retrospectively assessed 605 CT brain scans carried out between January 2006 and May 2011 that had been indicated for a range of reasons, excluding all cases of brittle bone disorders. The various reasons for which the CT brain scan had been indicated were divided into four groups:

- group A: headaches, convulsive seizures, neurological deficits, or any other focal neurological signs (349 cases);
- group B: head injuries (132 cases);
- group C: hydrocephalus (55 cases);
- group D: abnormalities of the bones of the cranial vault (69 cases) including plagiocephaly (12 cases) and craniosynostosis.

In this way, our analysis covered the presence, topography, and number of wormian bones by age and by the reason for which the CT scan had been indicated. We were able to count wormian bones in the sagittal suture, posterior fontanelle, posterolateral or mastoid fontanelle, and the right and left lambdoid sutures as shown in Fig. 1. These analyses were carried out on a post-treatment console using 3D reconstructions with volume rendering.

Results

In our study, wormian bones were found to be very common given that they were identified in 320 patients, or 53% of the CT scans carried out, with a number per CT scan ranging from one (110 scans) to eight (five scans). The distribution in percentages is presented in Fig. 2. The distribution of numbers of wormian bones per CT scan by group is detailed in Fig. 3. It shows that 43% of children had between one and three (n=260) wormian bones, 10% had four or more (n=60), and 6% had five or more (n=40). We then assessed the mean number of wormian bones by indication:

- group A (n=349): 445 wormian bones equating to 1.28 W bones/CT;
- group B (n=132): 180 wormian bones equating to 1.36 W bones/CT;
- group C (n=55): 73 wormian bones equating to 1.32 W bones/CT;
- group D (n=69): 96 wormian bones equating to 1.39 W bones/CT.

The numbers (mean, maximum, and minimum) of wormian bones per CT scan in terms of indication and patient age are summarised in Table 1.
Table 1  Distribution of numbers of wormian bones per CT scan (mean [max–min]) by age and group.

<table>
<thead>
<tr>
<th>Group/age</th>
<th>&lt; 3 months</th>
<th>3–6 months</th>
<th>6–12 months</th>
<th>1–2 years</th>
<th>2–3 years</th>
<th>Total wormian bones</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>0.69 [6–0]</td>
<td>1.36 [8–0]</td>
<td>1.75 [7–0]</td>
<td>1.17 [7–0]</td>
<td>1.25 [6–0]</td>
<td>445</td>
</tr>
<tr>
<td>B</td>
<td>1.16 [6–0]</td>
<td>1.48 [7–0]</td>
<td>1.48 [8–0]</td>
<td>1.68 [7–0]</td>
<td>0.95 [6–0]</td>
<td>180</td>
</tr>
<tr>
<td>D</td>
<td>3.5 [5–2]</td>
<td>2 [8–0]</td>
<td>1.32 [4–0]</td>
<td>0.77 [2–0]</td>
<td>0.25 [2–0]</td>
<td>96</td>
</tr>
<tr>
<td>Total wormian bones</td>
<td>92</td>
<td>152</td>
<td>230</td>
<td>182</td>
<td>138</td>
<td>794</td>
</tr>
</tbody>
</table>

Finally, we assessed the topography of these wormian bones. We counted a total of 794 wormian bones of which 22 were in the sagittal suture, 252 in the right lambdoid suture, 256 in the left lambdoid suture, 45 in the right squamosal suture, 41 in the left squamosal suture, and finally 178 in the posterior fontanelle. The percentages distributed in each area are shown in Fig. 4. Figs. 5 and 6 illustrate the presence of wormian bones in the sagittal and lambdoid sutures as well as in the posterior fontanelle.

Figure 3. Distribution of the numbers of wormian bones per CT scan by group.

Figure 4. Percentage of wormian bones in each location.

Figure 5. Example of a single wormian bone in the posterior fontanelle (inca bone).

Figure 6. Example of wormian bones in the sagittal suture and the right lambdoid suture.
Discussion

We have known of wormian bones for a very long time, with the name deriving from the description of intrasutural bones made by Ole Worm (1588–1654), a Danish anatomist, in a letter addressed to Thomas Bartholin in 1643. However, the first description is attributed to Paracelsus (1460–1541) who named a bone located in the posterior fontanelle the "ossiculum antiepilepticum". Over the centuries that followed, numerous studies described various associations between the presence of wormian bones and congenital diseases and attempted to explain the mechanisms by which they formed by rather questionable hypotheses. The result was an abundant, descriptive literature that was highly divided on several points.

Frequency of wormian bones

In our study, wormian bones were very common since they were identified in 53% of children. The incidence of wormian bones varies in the literature and this could in part be down to the method used to look for them. It is worth repeating that our study is the first to use computed tomography, which is today considered to be the gold standard method for studying the cranial vault. This is one of the strong points of our study. The archaeologist Brothwell [2] studied the incidence of wormian bones in different populations basing investigations on anthropological data and found an incidence of 55% in an Anglo-Saxon population and 80% in a Chinese population. The main radiographic study was carried out by Pryles and Khan [3], who analysed 515 skull radiographs from children aged between 0 and 14 years. For these authors, the incidence of wormian bones decreased with patient age, falling from 68% at 0 to 4 years old to 24% at 5 to 9 years old, and 8% between 10 and 14 years old. Our study was focused on a much smaller age range (0–3 years old) and found a slightly lower frequency than was identified by Pryles and Khan.

Mean number of wormian bones

No studies reporting on numbers of wormian bones in a general population have, to our knowledge, been published. There have been only assessments of target populations like that of Cremin et al. [1] into osteogenesis imperfecta, which proposed a threshold of ten wormian bones to be pathological. In our study, the maximum number of wormian bones was eight, seen in five children equating to 1.5% of our "normal" population.

Topography

In our study, wormian bones located in the lambdoid sutures were significantly more numerous (around 64% of cases). This is also seen in other authors’ findings, and especially in those of Bergman et al. [4]. There are other less common topographies for wormian bones, particularly in the coronal and sagittal sutures as the Khan study showed [5].

In an anthropological study of 1500 skulls, Tewari et al. [6] did not find any wormian bones in the coronal or sagittal sutures. These results seem surprising in view of our series (22 wormian bones in the sagittal suture, equating to around 3%). The coronal suture was not investigated in our study.

Aetiology and pathogenesis

Over the course of several centuries, numerous causes have been suggested in an attempt to explain the development of wormian bones but none of these has been universally accepted. A “mechanical” origin is the cause most often cited in the literature. For example, in the 1851 book “Peruvian Antiquities” by Riveiro and Von Tschudi [7], wormian bones were described as being a characteristic of the Inca population. The authors suggested that wormian bones developed in response to artificial deformities induced in the skull; the indigenous Incas had a custom of practising cranial deformation in children using mechanical methods. The term “Inca bone” is still found in the literature. This hypothesis was taken up again in 1897 by Dorsey [8] and in 1965 by Bennett [9] who specified that although these skull deformities could be have been of artificial origin, they may also have been pathological, indicating, for example, hydrocephalus.

Later, Ell-Najjar and Dawson [10] refuted this theory in 1977 by comparing the incidence of wormian bones in an Indian population divided into two groups (one group with skull deformities and the other without), and concluded that there was no significant difference between these two groups.

Today, we could relate this to plagiocephaly, which is asymmetry due to pressure exerted on the infant skull. In this context, it is interesting to note that among our small sample of 12 plagiocephalies, the mean number of wormian bones (2.33 W bones/CT) was notably higher than that in the other groups of children. Parker [11] suggests that the number of wormian bones increases in proportion to the volume of the skull and that this is applicable irrespective of the cause. Bergman et al. [4] suggested that wormian bones may develop due to rapid cerebral expansion, and this would explain why they are found in higher numbers in patients with hydrocephalus.

In our study, the hydrocephalus group did not present greater numbers of wormian bones than other groups.

Significance of the presence of wormian bones

This point is much debated in the literature. Most of the time they are considered to be a simple anatomical variant and our study tends to point to this theory. Nonetheless, some authors attribute wormian bones with a particular significance. Thus, for Pryles and Khan [3], the prevalence of central nervous system abnormalities (micro- and macrocephaly, hydrocephalus, craniosynostosis, cerebral palsy, epilepsy, and learning difficulties) in a population of children with wormian bones varied from 93 to 100% in a randomised group, leading this author to state that wormian bones were a true marker for developmental abnormalities of the central nervous system. The work of some other authors does not support such high percentages. On the contrary, Jeanty et al. [12] reported that wormian bones were identified in several foetuses on sonography with no corresponding abnormality being found.
The specific disorder of osteogenesis imperfecta

Cremin et al. [1] were for their part interested in the association between wormian bones and osteogenesis imperfecta and they introduced the idea of "significance". For this team, wormian bones were considered to be "significant" when there were more than ten of them and especially when they were arranged in a mosaic pattern. Based on this, they suggested that the presence of significant wormian bones could be a strong argument in favour of osteogenesis imperfecta. In their study, 88% of 81 patients with a diagnosis of osteogenesis imperfecta had a significant number of wormian bones (more than ten) on radiography investigations. The authors pointed that significant wormian bones were not only found in osteogenesis imperfecta but also in other bone dysplasias.

Nonetheless, picking up on these criteria, Patterson et al. [13] pointed out that an absence of significant wormian bones did not exclude a diagnosis of osteogenesis imperfecta. Indeed, in their study of 11 patients with proven type IV A osteogenesis imperfecta, only one of these presented wormian bones in excess of the threshold of significance on skull radiography.

Conclusion

Wormian bones are very common and sometimes occur in high numbers in children even when there is no background of osteogenesis imperfecta, and they must usually be considered to be a simple anatomical variant whose mechanism of development is not entirely understood. When identified in radiography investigations carried out due to a suspicion of physical abuse, the presence and number of wormian bones should always be reported but it does seem to be important, in view of our results, to bear in mind that they are common in a normal population before drawing any diagnostic conclusions from such findings.

Indeed, in our study, 6% of children had five or more wormian bones and 1.5% had at least eight.

In view of our study, the threshold of ten wormian bones established by Cremin et al. [1] for considering a diagnosis of osteogenesis imperfecta still seems to us to be valid.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References