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Wormian Bones in Osteogenesis Imperfecta and Other Disorders

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Abstract. When are Wormian bones significant is not an easy question to answer, but its relevance is important in relation to bone dysplasias such as osteogenesis imperfecta. Recognition will differ with age of patient, radiographic objectivity, and personal subjectivity. In order to attempt an answer, the skull radiographs of 81 cases of osteogenesis imperfecta of varying ages were examined for the presence of Wormian bones. These were compared against the incidence of Wormian bones in 500 skull radiographs of normal children. Significant Wormian bones as against normal developmental variants were considered to be those more than 10 in number, measuring greater than 6 mm by 4 mm, and arranged in a general mosaic pattern. They were found in all the cases of osteogenesis imperfecta but not in the normal skulls. The occurrence of significant Wormian bones in other bone dysplasias from our material and that of the literature was recorded. Other incidental findings in the skulls of the cases of osteogenesis imperfecta were also appraised.

Key words: Osteogenesis imperfecta – Skeletal dysplasia – Wormian bones

At a recent orthopaedic meeting in Cape Town a question was asked concerning the nature and significance of Wormian bones. The difficulty of giving an exact answer and the lack of literature on the subject stimulated this communication.

Wormian bones are small, irregular bones which are found in the cranial sutures. They are named after the Danish anatomist, Olaus Wormius, who described them in a letter to Thomas Bartholin in 1643 [10, 11]. Alternative designations are Schaltknochen [7], intercalary, and intrasutural bones.

Wormian bones are detached portions of the primary ossification centres of the adjacent membrane bones. They are present in normal individuals, but they are also an important feature of osteogenesis imperfecta (OI) and other conditions. Their presence in familial studies may uncover unsuspected cases in disorders such as cleidocranial dysplasia [5]. For purposes of diagnosis in these disorders, their size, number, and appearance are of importance.

In an attempt to elucidate the pathological and diagnostic significance of Wormian bones we have examined skull radiographs of 81 patients with proven OI. In order to provide a base-line we have also studied skull radiograph from 500 normal children. Our findings are presented and discussed in this paper.

Materials and Methods

1. Normal Skulls

The skull radiographs of 500 consecutive children aged one month to ten years, who attended the Red Cross Children's Hospital for an unrelated purpose, were examined. Difficulties in interpretation were encountered due to radiographic overlap in the sutures, especially with the posterior sutures in the lateral projection. Furthermore, in the normal young child Wormian bones arranged in a linear fashion were so frequent and numerous that it was not possible to make any meaningful quantitative assessment. We therefore designated a "significant Wormian bone" (SWB) as one which had a diameter greater than 6 mm in one axis and 4 mm in another at right angles, with an arrangement tending to a mosaic rather than linear pattern. Any portion of bone which showed multiple fissuring to the extent that interpretation was impossible was excluded. Many developing skulls had three to five such bones, but double this figure was unusual (1%). Counting these bones is not always an easy matter and we took 10 SWB's as a cut off point for a possible diagnostic indicator of abnormal development. This numerical evaluation did not include isolated clusters of fragmentation. SWBs are therefore defined as sutural bones

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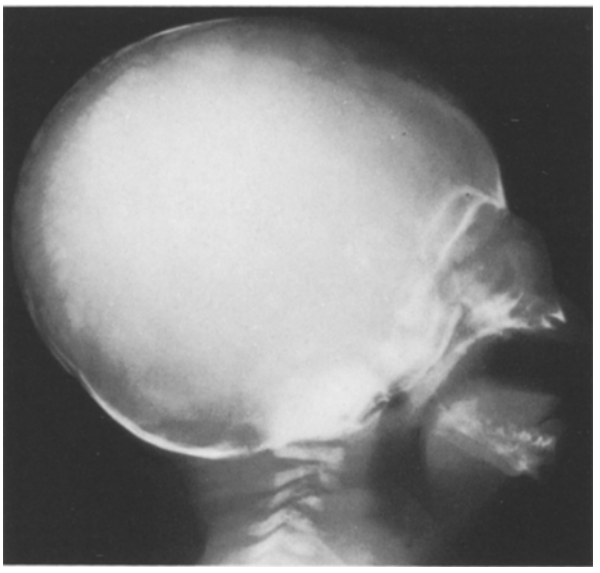

Fig. 1. Typical SWBs in mosaic pattern in a 10 day old baby with OI

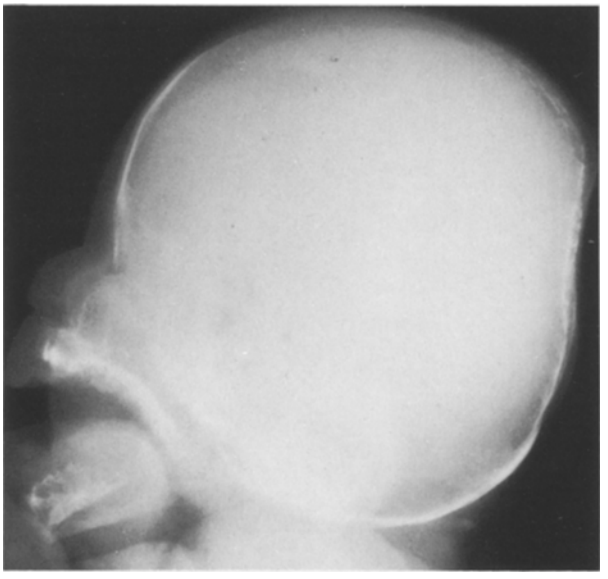

Fig. 3. Skull radiograph of a premature infant with OI, showing lack of vault calcification. Although the posterior skull shows irregular ossification, SWBs cannot be defined

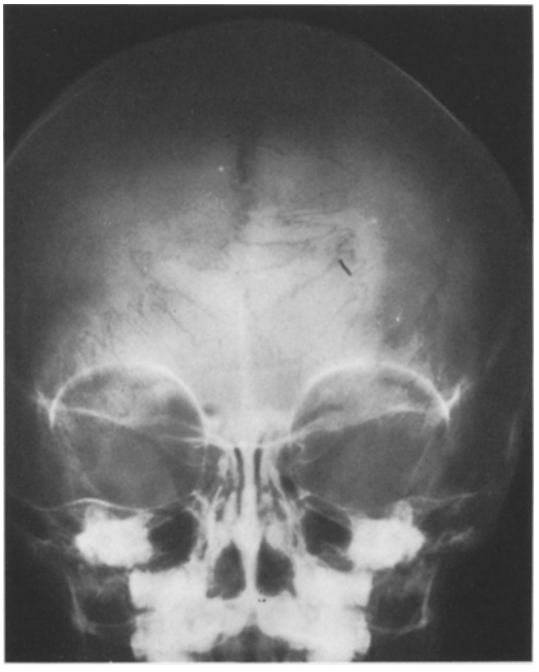

Fig. 2. SWBs shown in frontal projection of the skull of an adult with OI

which are 6 mm by 4 mm or larger, in excess of 10 in number, with a tendency to arrangement in a mosaic pattern.

2. Osteogenesis Imperfecta

The skull radiographs of 81 patients with proven OI from the University of Cape Town Skeletal Dysplasia Registry were examined for significant Wormian bones or other abnormalities. Thirty-four of the patients were under the age of 10 years, 15 being in their first year of which six were neonates. Thirty three were between 10 and 30 years of age and the rest scattered over the remaining decades up to 70 years.

Difficulties in interpretation were encountered unless lateral, frontal, and Towne's views of the skull were available. When the sutures have closed, evaluation in the anteroposterior projection with radiographs in the 60–65 kV exposure range is required.

3. Literature Survey

The literature on Wormian bones was obtained by a computerised retrieval service (MEDLARS) and the illustrations in the conditions described were perused for SWBs. The disorders noted in standard Gamut lists [12, 17] were also evaluated and the findings were correlated with those from additional material in our bone registry.

Results

1. Assessment of OI Skulls for SWBs

All but 10 of the patients with OI had more than 10 SWBs and typical appearances are shown in Figs. 1 and 2. Four of these 10 persons were adults in whom the quality or technique of the radiographs prevented accurate assessment; large Wormian bones were present but could not be evaluated quantitively. These results were deemed to be inconclusive.

In the six neonates, four had deficient ossification of the calvaria, while in two others, who were very premature, the skull size was too small to permit effective measurements (Fig. 3). In these cases the findings also were regarded as inconclusive. No relationship was established between the number of SWBs and the ages of the OI patients.

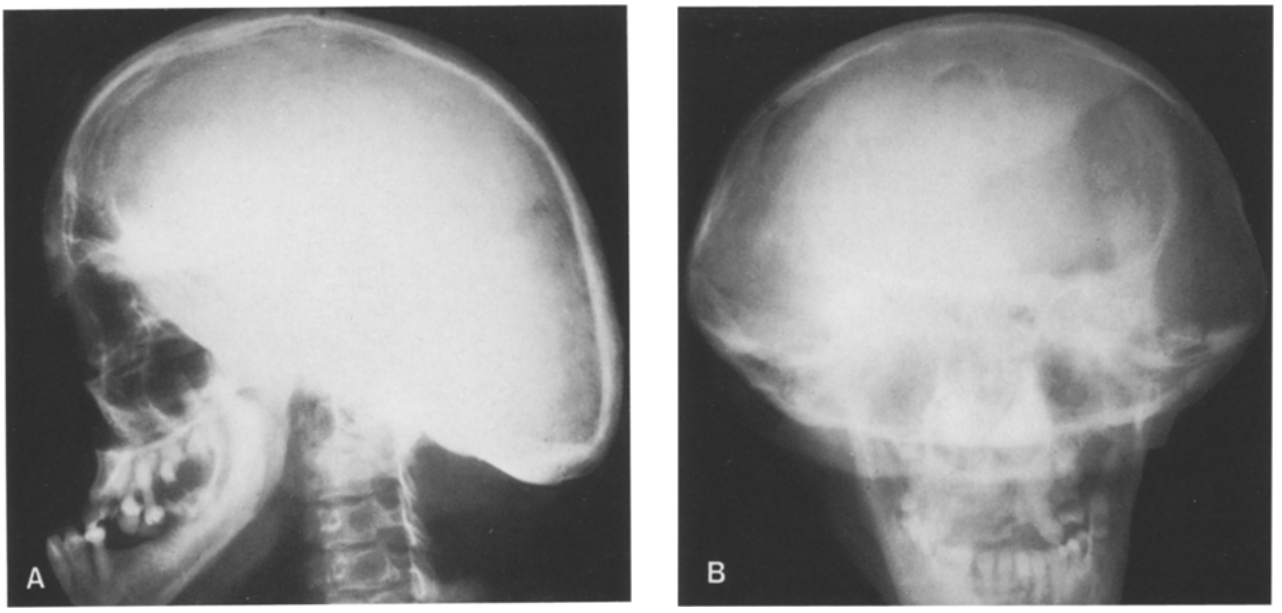

Fig. 4A and B. Anteroposterior and lateral projections of the skull of a 30-year-old patient with OI. Gross basilar invagination is evident

2. *SWBs in Other Conditions*

a) Consistent SWBs

i) From Our Own Cases:

Cleidocranial dysostosis
Cretinism
Pachydermoperiostosis (familial osteo-arthropathy) [2]
Metaphyseal dysplasia (Jansen type)

ii) From the Literature:

Kinky-Hair syndrome (Menkes syndrome) [18]
Acro-osteolysis (Hajdu-Cheney syndrome) [1]
Prader-Willi syndrome (Hypotonia – hypomentia – hypogonadism – obesity) [9]
Trisomy F Translocation [16]

b) Inconsistent SWBs

i) From Our Own Cases:

Pyknodysostosis
Sclerosteosis
Osteopetrosis – infantile type
Hydrocephalus
Down syndrome

ii) From the Literature:

Aminopterin-induced fetal changes [14]
Hallerman-Strieff syndrome (oculomandibulodyscephaly) [6, 15]
Progeria [3, 8]
Rickets

3. *Incidental Abnormalities in OI Skulls*

Platybasia or basilar invagination was present in six of the adults with OI and in two it was particularly severe (Figs. 4A and B). One of the patients was aged 15 years but none of the 34 children under 10 years of age showed this feature.

Excessive pneumatisation of the frontal sinuses and/or mastoids was present in 17 of the OI patients who were above the age of 10 years. This is a difficult feature to assess but it was considered to be gross in four cases (Fig. 5).

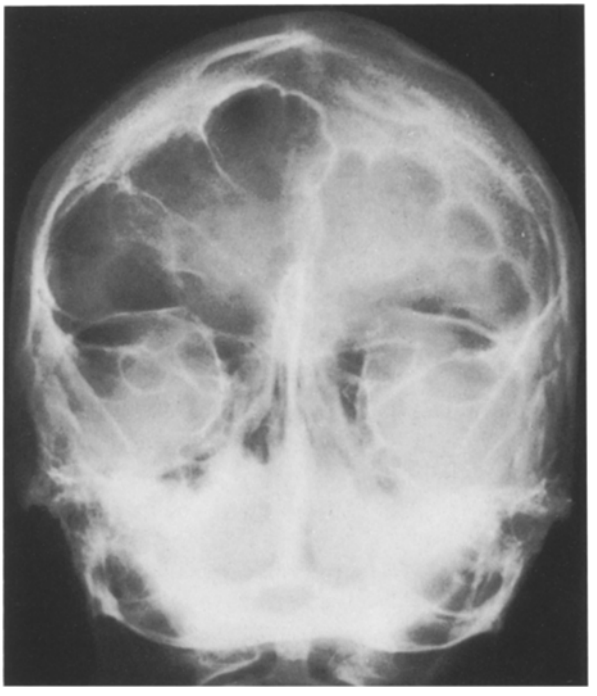

Fig. 5. Skull radiograph of a 25-year-old patient with OI, showing gross overaeration of frontal sinuses

Discussion

Multiple Wormian bones are a normal feature in the growing skull and the concept of SWBs as defined in this paper is only of value if it permits differentia-

tion between normal and abnormal situations. For this, recognition of a mosaic pattern is important. The term "Wormian bone" implies a normal developmental structure and the designation "dysplastic sutural bones" would be more appropriate when pathological changes are implied. However, as the term "Wormian bones" is hallowed by time and widely used, the retention of this eponymous designation is warranted.

The multiple fragmentation of a single isolated large Wormian bone, such as the Inca bone was not considered as SWBs as this is a common normal variant [13]. Furthermore it must be emphasized that the enumeration of Wormian bones in pathological states is difficult (172 were counted in an anatomical specimen of OI by Hektoen in 1903 [10]).

Apart from OI it is evident that SWBs occur in a few other disorders, as indicated in Section 2a of the results, while large sutural bones which are not arranged in a mosaic pattern or in significant numbers, may occur in the conditions listed in Section 2b of the results. However, many of the disorders given in Gamut lists [12] do not merit inclusion. For instance, in our and other's experience[1] SWBs are not a feature of dietary rickets.

It has been suggested that Wormian bones may serve as markers of underlying epilepsy or mental retardation [7, 11]. The study of this suggestion in depth was not part of our project, but it is significant that in the 60 children referred with problems of mental retardation during this period we did not note the presence of SWBs. In addition, we examined the skulls of 12 children suffering from mental retardation associated with the fetal alcohol syndrome and found no SWBs.

Skull growth is affected by dural attachments and is related to brain development. The development of Wormian bones occurs at an early membranous stage of skull growth and it is difficult to correlate this with heterogeneous brain disorders or relate it specifically to any one type. The development of SWB points to a bone growth disorder rather than an underlying brain disorder. A final comment on Wormian bones and skull growth is that it is not affected by the mechanical distortion of the skull, which is a traditional practice in certain tribal communities [4].

Our observation of a high frequency of platybasia in OI is in accordance with previous observations. Excessive pneumatisation of the frontal and mastoid sinuses was a surprising finding, which does not appear to have been recorded previously. This feature is probably an inconsistent and non-specific reflection of abnormal skull development in OI.

1 Taybi H (1981) Personal communication

In conclusion, we consider that the concept of "significant Wormian bones" will be of value in the radiographic confirmation of a clinical diagnosis of OI. They are not pathognomonic of OI, as they occur in other conditions, but the presence of more than ten SWBs is sufficiently unusual to warrant further investigations to identify an underlying cause or heritable disorder that has affected skull growth in an early stage of development. We have also tried to draw attention to the difficulties of assessing Wormian bones and, from the material available, have analysed the conditions in which their recognition may be of importance.

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