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

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.