

Foramina parietalia permagna: the ins and outs

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[Received 25 February 2012; Accepted 20 March 2012]

Foramina parietalia permagna or enlarged parietal foramina are a rare variant estimated to be less than 1 in 25,000 cases. Out of 150 dry macerated skulls studied one skull showed 2 large parietal foramina measuring 17.38 × 27.67 mm (right) and 15.31 × 25.46 mm (left) in size. Between them, across the sagittal suture, was a transverse communicating suture interrupted by 3 very small wormian bones. There is no denial of the fact that this familial transmitted trait is caused by erratic ossification due to gene mutations. The clinical importance lies in these being markers for underlying neural or bone pathology or metabolic syndrome. The enlarged parietal foramina as expressed by the 'eyes at the back' remain a curious anatomical but a definite clinico-pathological entity. (Folia Morphol 2012; 71, 2: 78–81)

Key words: parietal bone, enlarged parietal foramina, cranium bifidum, familial

INTRODUCTION

The foramina parietalia permagna or the enlarged parietal foramina, a definite clinico-anatomical entity, is a rare variant estimated to be less than 1 in 25,000 cases [20]. Greig (1892) [7] considers it to be "sufficiently rare to make it desirable that every example be reported".

Normally the parietal foramen lies close to the sagittal suture; anterior to the lambdoid suture; 0.5–1.0 mm in size rarely enlarged to as much as 70 mm due to congenital parietal bone ossification defects; present in 60–70% cases; and transmitting an emissary vein [14]. This enlarged foramen may present without any associated abnormality [17] or along with associated congenital bony defects [3, 8, 12, 18], soft tissue pathologies [6, 16], underlying neural deficit [13, 19], or as part of metabolic syndrome [10]. It may be a variant of cranium bifidum. The condition undoubtedly has a familial inheritance due to homeobox gene *ALX4* [13].

MATERIAL AND METHODS

150 dry macerated human skulls of unknown age and sex from the departmental bone bank and from medical students were examined. One of the skulls had two thumb-sized defects (foramina) near the posterior end of the norma verticalis. This skull was examined for 150 more parameters on each half.

RESULTS

The right foramen was oval with irregular anterior margin, having diameters: sagittal 17.38 mm and coronal 27.67 mm. It was 4.6 mm from the sagittal suture, 14.71 cm from the coronal suture, and 3.86 cm from the lambdoid suture.

The left foramen was oval, having diameters: sagittal 15.31 mm and coronal 25.46 mm. It was 7.28 mm from the sagittal suture, 15.01 cm from the coronal suture, and 4.21 cm from the lambdoid suture.

There was a transverse communicating suture between two foramina across the sagittal suture interrupted by three very small wormian bones. The foramina margins were smooth except near the medial end at the transverse interconnecting suture site, well formed and sharp. There was no bevelling of margins and they were made of single compact bone without intervening diploei, ruling out any possibility of mechanical injury or trephining artefact. At the obelion the sagittal suture, as well as interiorly the superior sagittal sinus groove, were deviated towards the left foramen then distally swerved back towards the right. The sutural bones were seen at both the asterions and at the right limb of the lambdoid suture. Thorough morphometric investigation of the skull, taking into account 150 parameters for each skull half, yielded no other abnormality. A few ivory patches were scattered around the foramina as hyperostosis areas (Fig. 1).

DISCUSSION

Normally the parietal foramen is situated 1.5 cm lateral to the sagittal suture and 2.5–3.75 cm anterior to the lambdoid suture [8]. It is an extremely small foramen 0.5–1.0 mm in size transmitting an emissary vein, the Santorini's vein [14], and a minute anastomosis between the middle meningeal and occipital arteries [18]. It is present in 60% of cases; more common unilaterally (36%) than bilaterally, rarely multiple, more often on the right, accompanied by or replaced by a single median foramen, and often enclosed by a fibrous membrane of both dura mater and pericranium [1].

Foramina parietalia permagna are extremely uncommon but are a well-recognised entity. The first case was described in a dry skull [23, 24] but was later reported in the living [7], and by 1963 only 100 cases had been reported [3]. Subsequently it was reported by many anatomists [15, 17].

Morphogenesis. At the end of the second foetal month, two ossification centres at parietal tubers spread peripherally but leave an interval near the superior borders of the parietal bone called the sagittal fontanelle [21], which close by the seventh foetal month. Closure may be delayed (Broca) or rarely, due to irregularities in the ossification, a large gap of fingerbreadth size persists [9].

This defect presents mostly bilaterally, sometimes unilaterally [1], or at times as a large single midline posterior parietal defect known as cranium bifidum. With the growth of the child these gaps gradually obliterate [5, 15, 17] to close at a later date [5] or

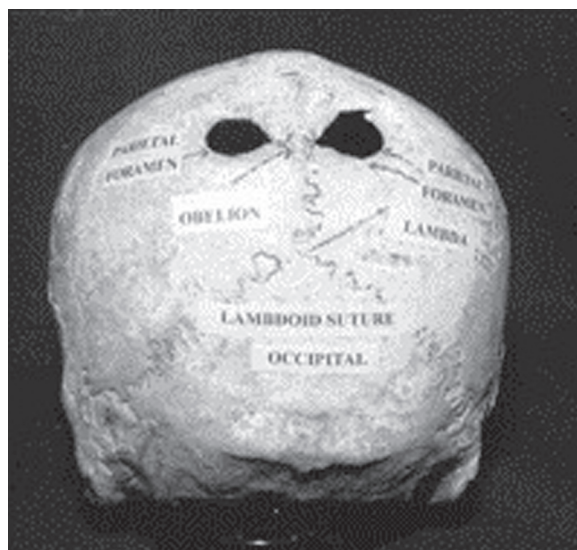


Figure 1. Norma occipitalis, showing enlarged parietal foramina.

may persist throughout life [18]. It may be summed up that the cranium bifidum and the enlarged parietal foramina are age dependant variable expressions of the same trait [11]. These defects sometimes appear as bone-filled large demarcated areas in conjunction with normally sized and positioned parietal foramina [5, 12, 22] thereby dismissing earlier hypotheses that rather than merely being enlarged parietal foramina these are separate entities.

The shape of this foramen is well formed often symmetrical, but not always [10]; if asymmetrical then sagittally smaller, and many times a remnant of the transverse suture extends from the medial margin of the defect to the mid-sagittal suture [8], as in the current case. In rare cases it is like a unilateral cleft.

The size varies from a few mm to 70 mm [8] but most frequently between 30–40 mm [15, 16]. Usually the reduction in the size of the foramen during childhood ends at three years [20] and further reduction is minimal [8].

Although so much enlarged but does not transmit an enlarged vein evident by the absence of any vascular groove marking around, but recently some studies have correlated a falcine venous sinus with these defects [4, 19]. They are in fact defects of ossification, as supported by the overall thinness of rest of the parietal bone [1]. The term 'enlarged foramina' is a misnomer as no significant structure passes through it; rather 'congenital parietal defects' would be a more appropriate nomenclature [3].

These foramina are reported concurrently associated with other bony defects like defective lateral clavicular end [3], craniosynostosis with temporal skewing [12], cleft lip and cleft palate [8], Crouzon variant with brachycephaly [10], abnormal facial features [6], and with aplasia cutis congenita [16] or without any other congenital abnormalities and any hereditary or familial transmission [2]. The spectrum of intelligence in this defect is wide from the bright, intelligent children of Goldsmith's Catlin family to mental retardation that can be attributed to the association of this anomaly with aberrant vascular evolution, which can also affect the skull, cerebrovascular system, and brain resulting in associated occipital cortical infolding [19], focal encephalomalacia, and mentally compromised children, which is now confirmed by the appearance of delineation of mental retardation loci on the chromosomes of these patients [13].

Apart from a few isolated cases [10] familial transmission seems the rule, as it was first observed in 16 out of 56 members of the Catlin family (spanning over five generations) by Goldsmith, (1922) [5]; hence, the defect acquired the name 'Catlin marks' and was also reported in 14 out of 32 members (spanning four generations) [8]. The variant was thought to be due to a gene probably an autosomal dominant but with an irregular penetration and expressiveness. Recently more advanced studies have found heterozygous mutations of the homeobox genes *ALX4* on chromosome 11 and *MSX2* on chromosome 5 in such cases [13, 24].

Enlarged parietal foramina or "eyes at the back" serve no useful purpose but represent the inheritance of an entirely useless or even injurious character. It is not a serious handicap to the possessor except for some headaches, nausea, and vomiting [18] or vulnerability to injury or infection at the time of birth or later on during active life. The presence of the enlarged parietal foramina is to be differentially diagnosed from cranial vault defects, meningocele, meningoencephalocele, dermoid cyst, multiple myeloma, secondary neoplasms, syphilis, xanthomatosis, trauma, and trephine artefacts. This condition being present in association with many diseases can act as a marker for diseases like osteoporosis, cleidocranial dysostosis, aplasia cutis congenita, cranio-lacunia, renal or celiac rickets and Crouzon syndrome [18].

CONCLUSIONS

Although a rare occurrence, foramina parietalia permagna occurs in conjunction with or without the normally sized and positioned parietal foramina thereby dismissing earlier hypotheses that rather than merely being enlarged parietal foramina, these are separate entities. These foramina have variable gross features like size, site, and number. It has a familial transmission due to heterozygous mutations of the homeobox of a totally useless rather an injurious character genes. This transmission is not only useless but also injurious in nature. Being associated with other pathologies these may act as marker to envisage them. Leaving aside the varied views of the enlarged parietal foramina as expressed by Goldsmith, the 'eyes at the back' remain a curious anatomical but definite clinico-pathological entity.

ACKNOWLEDGEMENTS

I am grateful to my teachers and colleagues.

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