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Surgical timing of craniosynostosis: What to do and when

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ABSTRACT

Craniosynostosis, both isolated and syndromic, are challenging malformations for the craniofacial team. They present the team with an articulated cascade of choices, which need to be addressed early in life and in the growing age to intercept, remove, or correct the direct and indirect consequences of the malformation.

Timing of treatment is thus critical and it stands on the experience of a multi-specialty trained craniofacial team.

In this paper the authors discuss the timing of treatment of the major craniosynostosis, isolated and syndromic, reviewing the options for treatment and their experience in this complex field.

1. Introduction

The timing of surgery for craniosynostosis is still controversial. The aim of this study is to find a common and practical protocol to treat this kind of patients over their lives. Craniosynostosis, the premature fusion of one or more cranial sutures, was first described by Otto in 1830 (Otto, 1830). Studies by Virchow in the 1850s led to the proposal of calvarial growth in a plane parallel to that of the fused suture, with sagittal synostosis resulting in a scaphocephalic, or boat-shaped, skull secondary to compensatory growth in the anteroposterior axis (Virchow, 1851). Virchow, however, also attributed craniosynostosis to either cretinism or an inflammation from the meninges (Virchow, 1851). Subsequent work by Park and Powers (1920) led to a conceptual revision in the 1920s, as congenital abnormalities in the suture mesenchyme became thought of as responsible for early suture fusion. In the 1950s, aetiological explanations for craniosynostosis once again changed, with studies by Moss (1959) purporting aberrations in the basicranium altering transmission of force, via the dura mater, to the overlying cranial sutures ultimately affecting premature fusion. While recent genetic and mechanical studies have now supplanted this notion, investigations by Moss, nonetheless, engendered a radical shift in the surgical approach to craniosynostosis. Given the observation that suturectomy alone did not restore normal calvarial development, complex craniofacial procedures were also deemed necessary to allow for proper growth and cranial expansion (Moss, 1959). Such realizations resulted in the pioneering work of Paul Tessier (1967).

Epidemiologically these kinds of pathologies may be divided into isolated, further divided into single or multiple synostosis, and syndromic.

2. Isolated craniosynostosis

Premature fusion of one or more cranial suture results in restriction of the growing brain, with subsequent morphologic bony deformities due to specific patterns of compensatory growth. The most common type of craniosynostosis, whether isolated or part of a larger syndromic pattern, is sagittal synostosis, resulting in a scaphocephalic deformity (Posnick, 2000) (Figs. 1 and 2). Other forms include metopic synostosis, resulting in a trigonocephaly deformity, unilateral coronal synostosis, resulting in a plagiocephaly deformity (Figs. 3 and 4), and bilateral coronal synostosis, resulting in a turribrachycephalic deformity. While lambdoid synostosis may also result in a plagiocephalic skull, this clinical entity has been infrequently encountered (Posnick, 2000). In addition to this calvarial dysmorphologies, premature pathologic suture fusion, as demonstrated by Moss, may be associated with multiple craniofacial deformities (Moss, 1957). Hypertelorism, downward slanting of the lateral canthi, palpebral fissure widening, displacement of the orbital rim and/or ear, and deviation of the nasal bone

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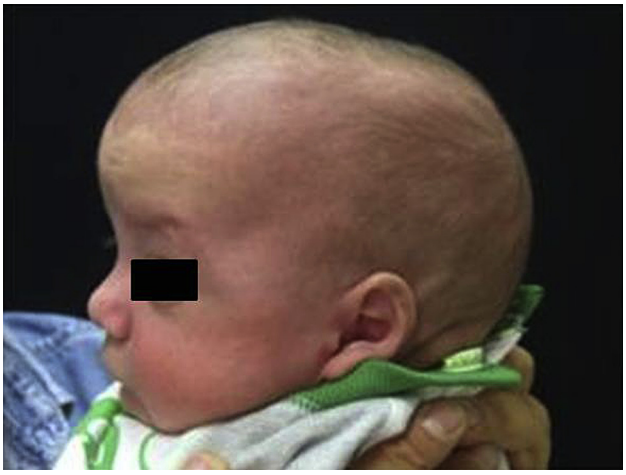

Fig. 1. Patient with scaphocephaly pre.

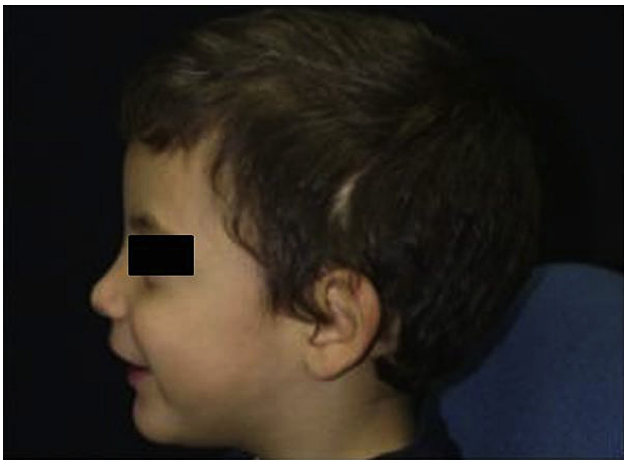

Fig. 2. Patient with scaphocephaly post.

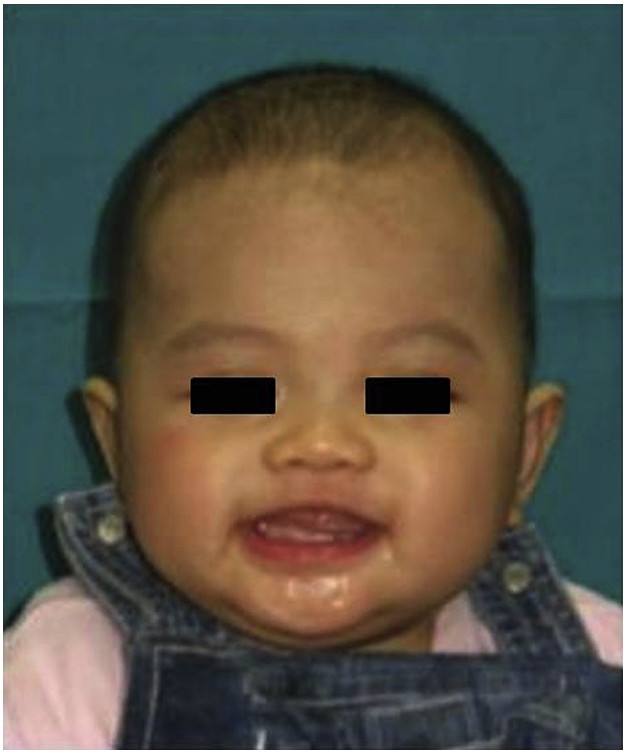

Fig. 3. Patient with plagiocephaly.

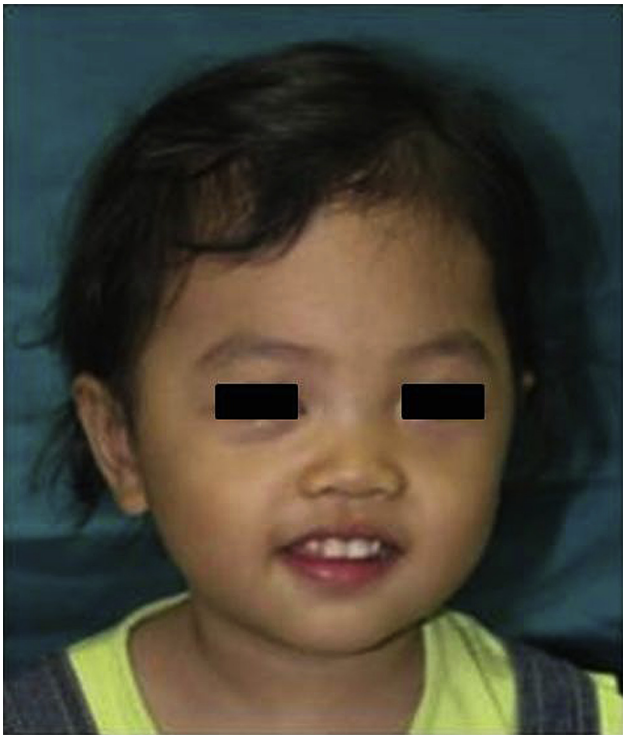

Fig. 4. Patient with plagiocephaly.

have all been commonly reported (Grabb et al., 1991). To allow for proper physical and psychological development as the child the clinician must address each of these dysmorphisms, like the deformities in the skull. In addition to these morphologic abnormalities of the calvarial vault and craniofacial skeleton, several functional aspects of premature suture fusion also merit significant consideration. Studies by Renier et al. (1982) have suggested the risk for the increase in intracranial pressure to be associated with multiple suture involvement, and that decreases in pressure measurements may follow surgical remodeling of the skull. Gault et al. (1992), likewise, demonstrated high intracranial pressure to occur most frequently in children with multiple premature suture fusions. Considering the dramatic growth by the brain during the first two years of life, it would be reasonable to expect a mismatch in cranial volume to result in elevated pressures and possible mental retardation. Though this is not often the case, continued concern has eventually driven early surgical intervention. Other functional considerations, in addition to intracranial pressure, have also been well described in association with craniosynostosis. With continued development in radiographic imaging, hydrocephalus, responsible for raised intracranial pressure, has become an entity frequently observed in patients with both syndromic and non-syndromic craniosynostosis (Kreiborg and Cohen, 1991). Visual disturbances are also commonly reported in those patients with premature pathologic suture fusion. Exorbitism and optic nerve atrophy can be readily attributed to abnormal craniofacial development (Newman, 1991). In addition, stretching of the nerve, compression by carotid vessels, or secondary effects of increased intracranial pressure have also been associated with optic nerve dysfunction (Grabb et al., 1991). Finally, the risk for mental