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IMAGES IN NEONATAL MEDICINE.....

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A cloverleaf skull associated with Crouzon syndrome

A baby born to non-consanguineous parents presented with a cloverleaf skull and Crouzon syndrome. Computed tomography scans revealed a trilobed configuration of the skull with a characteristic honeycomb appearance of increased craniolacunae (fig 1). The limbs and extremities were normal. DNA studies showed a 1040C→G mutation in the *FGFR2* gene predicting a serine 347 to cysteine amino acid substitution. The mutation was not present in either parent.

A cloverleaf skull consists of a trilobular skull with craniosynostosis. The known aetiology includes Apert syndrome, Beare-Stevenson cutis gyrata syndrome, Boston type craniosynostosis, Carpenter syndrome, Crouzon syndrome, Cumming syndrome, *FGFR2* unusual phenotypes, Muenke syndrome, osteoglophonic dysplasia, type 2 Pfeiffer syndrome, Saethre-Chotzen syndrome, type 1 and type 2 thanatophoric dysplasia, chromosomal duplications of dup(4p), dup(13q), and dup(15q), amniotic bands, and bilateral subtemporal decompression procedures for hydrocephalus.¹ Crouzon syndrome (OMIM 123500) is an autosomal dominant disorder involving craniosynostosis and *FGFR2* mutations. A frank cloverleaf skull is present in only 7% of Crouzon syndrome.¹

Figure 1 Computed tomography scans showing a trilobed configuration of the skull with the honeycomb appearance of increased craniolacunae.

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