

Median Cleft Face Syndrome

A 7-year-old male child with normal neuro and mental development presented with facial malformations. He had hypertelorism, exotropia, broad nasal root, midline clefting of the nose and an operated scar for frontal encephalocele (*Fig. 1*). The anterior nares were short and narrow. His visual acuity was 6/24 in both eyes and he had bilateral optic nerve atrophy. The rest of the systems were normal. A CT scan of the head revealed normal brain structure with the presence of a bone graft in the frontal region that was grafted for frontal encephalocele (*Fig. 2*).

Median cleft face syndrome (or frontonasal dysplasia) is a rare, sporadic condition. It results from embryonic failure of



Fig.1. Patient with marked hypertelorism and bifid nose.

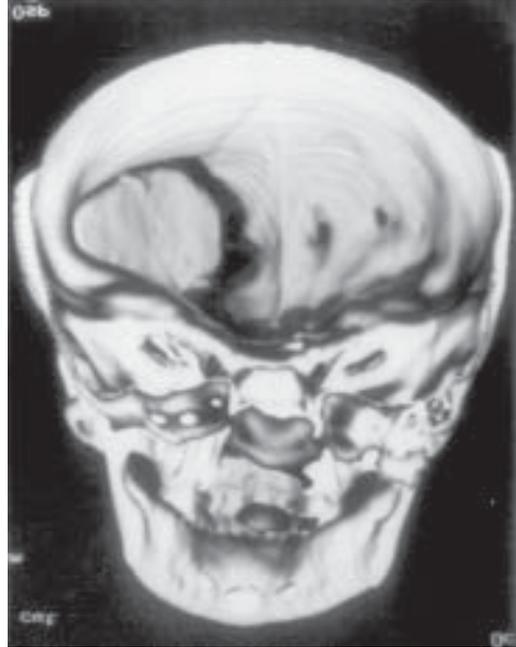


Fig. 2. Three dimensional CT scan of the head showing repaired frontal bone defect with a bone graft.

fusion of the median nasal processes. The most common findings are orbital hypertelorism, notched nasal tip to completely divided nostrils, broad nasion, low-set ears and deficit in midline frontal bone (cranium bifidum occultum). The most common associated malformations are lipoma or agenesis of corpus callosum, tibial hypoplasia, proximal hallucal polydactyly, epibulbar dermoids, ear tags and tetralogy of Fallot. Most patients have a normal intelligence.

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