Congenital Anomalies of Orbit

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The orbit is subject to a wide variety of congenital and acquired diseases. Emphasis will be given to the rare but very characteristic craniofacial syndromes which give rise to ocular complications. These will include craniosenotic syndromes and facial clefting syndromes. Crouzon's disease and Apert's syndrome are the most commonly encountered. The ophthalmological signs of these conditions include proptosis due to shallow orbits, strabismus, hypertelorism (which is a separation of the bony orbit), visual loss secondary to raised intracranial pressure, optic nerve stretching or compression and exposure keratopathy. Eighty percent of patients with Crouzon's have optic atrophy. Exposure keratopathy occurs as a result of the shallow orbits which may present with spontaneous subluxation of the globes.

The management of these cases require a multi-disciplinary approach. The ophthalmological assessment should include visual acuity, visual field, extraocular movement, exophthalmometer measurement, inner and outer canthal measurements, length and width of palpebral fissure, inter papillary distance, refraction and fundus examination. The basic approach in correction of these conditions, as pioneered by Tessier will also be mentioned. Cosmetic corrective surgery does not fully correct the ophthalmological complications.

Several of the more common conditions like orbital dermoid, orbital haemangioma will also be discussed with regard to clinical presentation and management. Both these conditions make up 47% to 61% of all orbital tumours in children.