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Apert's Syndrome: Lesser Known Aspects

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Apert's syndrome is a syndrome which is characterized by craniosynostosis, midface hypoplasia, and symmetric syndactyly of both hands and feet with an incidence of 1/160,000 live births. Ocular and dental examinations are mandatory for such patients but are often missed as attention is mainly focused on the disease triad as mentioned above.

Proptosis, strabismus, amblyopia and optic atrophy have been reported with this syndrome. Craniofacial surgery, while alleviating proptosis, may cause an alteration in strabismus, visual loss, and tearing problems in these patients. Therefore assessment and follow of these patients by an ophthalmologist is also required. [2]

Oral involvement in the form of tooth crowding, anterior open bite of the maxilla, impactions, delayed eruption, thick gingiva, supernumerary teeth or congenitally missing teeth are the hall marks of maxillary dental development in Apert

patients and needs a special mention. Secondly, failure in the anteroposterior and downward growth of the maxilla causes maxillary hypoplasia resulting in contraction nasopharyngeal airway and may obstructive sleep apnea and premature death. And finally, Apert syndrome has been found to be in parallel with Glucose 6 Phosphatase Dehydrogenase deficiency(G6PD deficiency) and hence drugs which are contraindicated in G6PD deficiency should also be avoided in Apert syndrome so that haemolytic anemia is not precipitated.[3]

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