

What is the role of dentists in managing craniofacial ciliopathies?

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A syndrome can be defined as "a collection of traits, health problems, and/or birth defects in an individual which usually has a single underlying cause". Among various syndromes, ciliopathy is classified as a disorder that results from aberrant form or function of primary cilia, manifesting as a broad range of clinical manifestations that ranges from organ specific to a broad pleiotropic syndrome.1

A large number of craniofacial conditions were identified as possibly ciliopathic.2 The term has been proposed by Brugmann et al.3 and classified by Raja et al.4 The problems of this syndrome might include a combination of physical problems, health problems, learning difficulties and/or challenging behavior. After initial diagnosis, some relevant evaluations are recommended.

Dental evaluation to assess hygiene, dental crowding, and hypodontia. Ophthalmologic assessment, cardiac evaluation, neurologic examination, endocrinology testing, renal function studies, and nutritional assessment can aid in identifying medically significant complications. Then, the patient can be considered for referral to a clinical geneticist.

Dental primary complications can be prevented by dental extractions in the case of dental crowding. Secondary complications can be prevented by antibiotic prophylaxis for surgical and dental procedures, in the case of individuals with structural cardiac anomalies.⁴

Dentists, as health promoters, must be aware of the existence and treatment options of these ciliopathies.4 It is important to deepen the knowledge of these ciliopathies to recognize the type of disability afflicting the patient and offer comprehensive and inter-disciplinary treatment to promote the dental health of these patients.

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