

Noonan syndrome: A case report

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Abstract

Noonan syndrome is a developmental disorder which is characterized by ocular, facial, cardiac, and dental anomalies. The etiology of Noonan syndrome is unknown. This may result due to either complete or partial absence of an X-chromosome or could be genetic effect. Dental features of the Noonan syndrome that have been described includes: malocclusion, greater risk of caries, in a greater proportion of subjects, while odontogenic keratocyst or giant cells lesions in a smaller proportion. The aim of the current article is to present the oro-facial findings in a case with Noonan syndrome in a 7 year-old male. Multidisciplinary treatment is the key to success in managing children with Noonan syndrome. Pediatric dentists play a significant role in leading the health team.

Keywords: Noonan syndrome, Dental, High arch palate

Introduction

Noonan syndrome was first described as a multisystem disorder by Noonan and Ehmke in 1963. It is characterized by clinical features such as short stature, hypertelorism, ptosis, and low-set ears. Noonan syndrome was often correlated to Turner syndrome which is characterized by epicanthic folds, right-sided congenital heart disease, and skeletal malformation.^(1,2) Noonan syndrome is a rare genetic alteration and the gene responsible is located on the long arm of chromosome 12.⁽³⁾ It can affect both males and females and the severity of the features varies in patients. The estimated incidence is of 1 per 1,000-2,500 live births.⁽⁴⁾ Teeth anomalies are seen with late eruption and increased risk of dental caries.⁽⁵⁾ Numerous cases of Noonan syndrome have been reported in the dental literature, however very few of them described some details of the oral features.⁽⁶⁻¹⁰⁾ The aim of this paper is to describe a case of Noonan syndrome, highlighting oro-facial findings in subjects affected by this genetic disease.

Case Report

This is a case report of a 7 year-old boy who complained of pain in his tooth. The patient was referred to a pediatric dentist for management. A detailed medical and dental history was obtained from the patient and his parents.

Medical History

The patient had been diagnosed as having Noonan syndrome at 2 months. He presented mild pulmonary stenosis, gastro esophageal reflux disease, and bronchial asthma. Physical examination revealed that the child had a relatively short stature, facial asymmetry, hypertelorism, depressed nasal bridge, drooping eyelids, broad philtrum, low set ears, short neck, and clubbed fingers.

Facial findings

The patient presented a facial dimorphism and facial shape was of an inverted triangle. He had a tendency of lip incompetence and reported the habit of mouth breathing (Fig. 1, 2). The family history was negative for Noonan syndrome and facial disproportion.



Fig. 1: Frontal View



Fig. 2: Side View



Fig. 3: Oral view of occlusion



Fig. 4: Oral view of maxilla



Fig. 4: Oral view of mandible

Intra-oral findings

Intraoral examination revealed primary dentition with multiple caries lesions and defects in teeth mineralization. The patient presented a narrow high-arch palate with prominent rugae, retrognathic maxilla, prognathic mandible, and hypoplastic jaws. Patient had anterior open-bite since the appearance of his primary dentition (Fig. 3,4,5). The patient was uncooperative to take a panoramic radiograph for further investigation on the status of the developing dentition.

Treatment plan

A multidisciplinary approach, involving pedodontic, orthodontic, and endocrinologic treatment was planned which was approved by the ethics committee of the institution.

Discussion

Noonan syndrome is an autosomal dominant anomaly affecting both males and females.⁽¹¹⁾ It is reported that over 50% of cases of Noonan syndrome are accounted for Missense mutations in PTPN11.⁽¹²⁾ Alike to most patients with Noonan syndrome, this patient had systemic problems. Ptosis, a relatively large nose, full lips, a high arch palate, and anterior skeletal open bite were the oro-facial features frequently reported and observed in this patient.^(5,6,8) Patients with Noonan syndrome have marked mandibular prognathism with an anterior open bite.⁽¹³⁾ This patient showed a high arch palate in the posterior region and a small maxillary arch. Growth of the dental arch may be influenced by the premature closure of the cranial sutures.⁽¹⁴⁾ Noonan syndrome is diagnosed mainly based on clinical features.⁽⁷⁾ Dental treatment should be carried out under antibiotic prophylaxis as these patients present with congenital heart disease which may cause bacteremia.⁽¹⁵⁾

Conclusion

Noonan syndrome is an autosomal dominant multisystem disorder. The children with Noonan syndrome usually have a wide range of health problems. Hence, it is essential for all practitioners to be aware of the child's special care needs. Multidisciplinary treatment is the key to success in the management of children with Noonan syndrome and the pediatric dentists play a significant role.

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