Introduction

Studies on the Noonan syndrome had been first conducted in 1963 by Jaqueline Noonan (paediatric cardiologist of the University of Kentucky) who noticed a heart defect (stenosis of the pulmonary valve) associated in the majority of the cases to common physical characteristics such as short height, hypertelorism, low junction of the ears, and deformed thorax and face in some of her patients [Noonan, 2006]. The first description of the syndrome, however, was probably made in 1883 by Kobylnski.

Of great importance in the identification of this syndrome was caries susceptibility, which in these patients is normal [Barberia-Leache et al., 2003]. In fact, initially the Noonan syndrome was mistaken for the Turner syndrome for the similarity in the physical characteristics. But, the normal caries susceptibility and the manifestation of the Noonan syndrome only in women allowed to differentiate between the two syndromes.

Subsequently, studies conducted by Tartaglia [2001] allowed to identify the gene responsible for the Noonan syndrome, which is located on the long arm of chromosome 12 (12q24.1). Later, other studies highlighted the genetic heterogeneity of the condition [Addante and Breen ,1996; Betts et al. 1993; Cancino et al.,2007, Lange et al.,2007; Lee and Cooper, 2005]. The gene PTPN11 situated on chromosome 12q24 could be responsible for such syndrome. In fact, it codifies the protein SHP-2, crucial fro transferring the intracellular signal that regulates some developmental processes, amongst which the formation of the semilunar valves of the heart. In order to test this hypothesis they have screened this gene, searching for possible mutations, first in familiar cases and subsequently in sporadic cases [Dunlap et al., 1989; Levine et al.,1991].

Although currently the diagnosis is exclusively based on clinical criteria, the discovery of the mutation of gene PTPN11 in about 50% of the patients affected by the syndrome might lead to a better understanding of the pathogenetic mechanisms that determine some of the phenotypes of the syndrome and, probably, will enable in the future a laboratory test for confirming the diagnosis or the prebirth diagnosis. The syndrome affects various ethnic groups.

General characteristics

The diagnosis, still based on the phenotype, can be evaluated at different ages, although the average age at which a diagnosis is carried out is nine years. In infants clinical characteristics are often vague; therefore, if a congenital cardiopathy or a positive family history is not present, the diagnosis is postponed until childhood. The general characteristics are summarised in Table 1. There is not an aethiologic therapy for the Noonan syndrome, but a certain number of complications of the syndrome can be dealt with medical and/or surgical therapy.

Dental features

In scientific literature dental aspects of the Noonan syndrome that have been described are: malocclusion, greater risk of caries processes, in a greater percentage of subjects, while odontogenic keratocyst or giant cells lesions (though these may not be correlated with the syndrome) in a smaller percentage (Table 2).

Patients affected by Noonan syndrome must be followed up by a multidisciplinary team, in which an important role is played by the paediatric dentist. In fact, pathologies
more common such as caries, periodontal problems, frequent malocclusions, in patients affected by the syndrome should be identified in their initial phase in order to put into effect primary or secondary prevention. It will be fundamental to identify the degree of compliance to treatments and choose the more effective operating method to establish a trusting relationship.

For those who have a light or moderate mental delay an outpatient treatment is preferable, avoiding all the disadvantages of the hospital. For those who, instead, have a serious mental delay it is advisable, in agreement with the parents, to carry out the treatment in the hospital, performed in a single session and under general anaesthesia.

Given their mental delay, children with Noonan syndrome are bottle-fed for a longer time, and sometimes the diet is inadequate for the age of the child. Besides, this extended consumption of liquids and sugars is also the primary cause of their caries. An early diagnosis and prevention of baby bottle caries are important in order to avoid subsequent problems during the eruption of permanent teeth [Nelson et al., 1978; Nirmal et al., 2001; Okada et al., 2003].

Among the most important otorhinolaryngology pathologies that can be found in patients with Noonan syndrome, there is oral breathing (bad habit), which consists in the exclusive or partial passage of air through the mouth instead of the nose.

A good percentage of patients with Noonan syndrome have, as mentioned, secondary malocclusion due to micrognathia and a higher probability of having to undergo orthodontic treatment because of dental crowding. Therefore, the role of the paediatric dentist is monitoring all those situations that hinder the corrected eruption of permanent teeth and intervene in order to resolve crowding.

Since these patients are considered at risk of bacterial endocarditis (they have a more or less serious congenital cardiopathy), appropriate therapeutic protocols must be followed. All manoeuvres executed in the oral cavity that involve bleeding should be carried out under prophylaxis [Sharma et al., 2007; Waldman and Pearlman, 2007; Wolvius et al., 2006].

Case report

A caucasian girl of 8 years with Noonan syndrome born from healthy parents, with no family history, came to observation because her parents were concerned for the anomalous position of her teeth during shedding.

The child was born at the end of the 42nd week after a normal pregnancy. At the time of delivery, however, an emergency occurred because the baby was not in the right position (emergency dystocic delivery).

At birth the baby weight was 3,300 g, her length was 50 cm, with an apgar index of 8/9. Subsequently she was supported because of the foetal suffering (meconium-stained amniotic fluid) and control tests were carried out. Since the thorax X-ray showed cardiomegaly, the baby echocardiography assessments were immediately carried out. On her 10th day of life the patient suffered cardiac decompensation. The echocardiography showed an interatrial ostium secundum defect of approximately 10 mm and wide patency of Botallo arterial duct of approximately 8 mm, with pulmonary hypertension. On the same day a surgical intervention to close the Botallo's duct was carried out. There were no postoperative complications and the baby was released without the need for cardiac therapy.
At the first dental visit, it was assessed that the child was in good general health conditions, calm, collaborating and she was used to periodic medical visits. At the time, the girl weighted 23 kg and was 123 cm tall.

Similarly to most patients with Noonan syndrome, also our patient had systemic problems, such as otitis, heart trouble, language disturbances and asymmetry of the lower limbs. Light mental delay was also found. She had the bad habit of sucking the lower lip.

At the intraoral examination, she exhibited bilateral molar and canine I Class relationship, deep bite, ectopic eruption of tooth 24 and 25, and the OPG showed agenesis of tooth 29 and 31. The cephalometric study highlighted a neutral growth, skeletal I Class, normal facial length (fig. 1-6; Table 3).

The treatment plan included extraction of teeth F, P, O, N, E under sedation and with antibiotic prophylaxis for bacterial endocarditis, as well as sealing of teeth n. 3, 14, 19, 30.

From the orthodontic point of view, the limited collaboration of the patient did not allowed taking of the impression. Thus, it had to be used a pre-shaped orthodontic device (Occlus-o-Guide) for the treatment of the deep bite and in order to guide the incisors in their

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<th>Table 3: Initial cephalometric data.</th>
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FIG. 6 - Initial teleradiography.

FIG. 7 - The “Occlus-o-Guide” device.

FIG. 7 - The “Occlus-o-Guide” device in situ.

FIG. 11, 12 - Radiographs after treatment.
correct eruption (Fig. 7-10). The device was well accepted and helped achieving the expected therapeutic results (Fig. 11-13; Table 4).

Conclusion

Patients with Noonan syndrome often exhibit alteration of the stomatognathic apparatus. For these subjects dental care is necessary starting from the first years of age, in order to prevent problems that could become irreversible and difficult to manage in day hospital. Therefore, either a primary or a secondary prevention are recommended, with customised protocols that should also envisage a rehabilitation process aimed at the normalisation of the musculature of the lower third of the face, which is compromised by the syndrome.

References


Table 4 - Cephalometric data after treatment.

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Book review

Early Childhood Oral Health

Editors: Joel H. Berg and Rebecca L. Slayton.

Paediatricians and dentists have been waiting for a long time for a resource to collect, organise and explain information on the dental and oral health of very young children. This comprehensive text answers to them. Chapters about caries risk, managing caries, and use of fluoride provide well-known concepts on techniques and dosage in early childhood. But the real importance of this book are the chapters on anticipatory guidance and family education, very useful in order to draw the child’s awareness to oral healthcare. The book provides practical protocols and step-by-step procedures for treating young children and allowing them access to dental care. “Early Childhood Oral Health” brings together contributions from many well-known researchers and clinicians in this field. It will provide dentists, dental professionals, and non-dental healthcare providers with an interest in promoting childhood oral health for the best quality of life. All chapters are illustrated with clinical photographs.