The Cleft Palate-Craniofacial Journal 50(2) pp. 242–248 March 2013

CASE REPORT

Conservative Management of Severe Open Bite and Feeding Difficulties in Patient With Noonan Syndrome

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Noonan syndrome is a multiple malformation disorder with an autosomal dominant inheritance pattern. Congenital heart defects, short stature, thoracic deformities, short neck with webbing, hypertelorism, malocclusions, and feeding difficulties are some of the commonly observed clinical features. We report on a case of a patient with Noonan syndrome, severe open bite, associated feeding difficulties, and strong gag reflex, discussing conservative management with myofunctional therapy. Myofunctional therapy has demonstrated a successful outcome, reestablishing masticatory function and routine dental home care after an 18-month follow-up.

KEY WORDS: *Noonan syndrome, open bite, feeding difficulties*

Noonan syndrome is a multiple malformation disorder with an apparent autosomal dominant inheritance pattern (Tartaglia and Gelb, 2005) and an estimated incidence of 1:1000 to 1:2500 live births (Tartaglia et al., 2001). The first published illustration of this condition was done by Kobylinsky in 1883. Nevertheless, it was only described as a syndrome by Jacqueline Noonan in 1968. Noonan affirmed the existence of a new syndrome with associated congenital cardiac disease. Mutations in the PTPN11 gene, encoding the nonreceptor protein tyrosine phosphatase SHP2, which maps to the long arm of chromosome 12, were identified in more than 50% of studied patients (Bertola et al., 1999; Bertola et al., 2003; Tartaglia et al., 2001). It has also been affirmed that more than half the cases represent new mutations (Pierpont, 1996).

The most commonly observed clinical features associated with Noonan syndrome are congenital cardiac defects (Pierpont, 1996; Bertola et al., 1999; Saenger, 2002); thoracic deformities with superior pectus carinatum and inferior pectus excavatum (Addante and Breen, 1996); short stature (Pierpont, 1996; Bertola et al., 1999; Saenger, 2002; Bertola et al., 2003; Ogawa et al., 2004); short neck with webbing, redundancy of skin, and low posterior hairline (Addante and Breen, 1996; Bertola et al., 1999; Tartaglia and Gelb, 2005); bleeding diathesis (Bertola et al., 2003); cryptorchidism (Pierpont, 1996; Bertola et al., 1999; Saenger, 2002; Bertola et al., 2003); and ophthalmologic and orthoptic findings, including hypertelorism with downward sloping palpebral apertures, epicanthal folds, ptosis, strabismus, and amblyopia (Lee et al., 1992; Tartaglia and Gelb, 2005).

Craniofacial characteristics have been described as dental malocclusions (Tartaglia and Gelb, 2005); philtrum with deep grooves and low-set and posteriorly rotated ears (Pierpont, 1996); high arched palate (Pierpont, 1996; Ogawa et al., 2004, Tartaglia and Gelb, 2005); micrognathia (Addante and Breen, 1996; Ogawa et al., 2004); retrognathia (Pierpont, 1996); and dental abnormalities, bifid uvula, and a rarely fissured palate (Addante and Breen, 1996). These patients usually present with moderate developmental delay during infancy, affecting motor, learning, and language skills (Pierpont, 1996; Pierpont et al., 2009).

In 2010, Romano et al. published a state-of-the-art review article that summarized a meeting of health care providers coordinated by the Noonan Syndrome Support Group; this meeting gathered experts in various aspects of the disorder with the aim of developing guidelines for its diagnosis and management. The authors reported that 55% to 100% of patients with Noonan syndrome presented with high arched palate, 50% to 67% manifested dental malocclusion, 72% expressed articulation difficulties, and 33% to 43% presented with micrognathia. Development of mandibular cysts was another observed feature, characterized by multinucleated giant cells within a fibrous stroma, indistinguishable from cherubism. However, these two conditions are genetically distinct; SH3BP2 gene mutations

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Submitted September 2011; Accepted April 2012.

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DOI: 10.1597/11-214

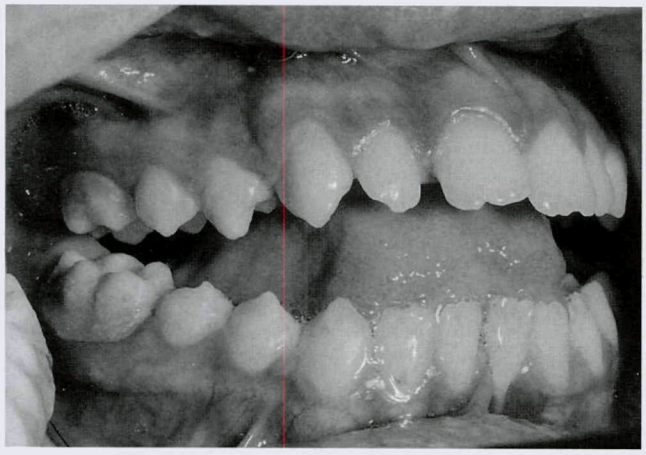

FIGURE 1 Lateral occlusal view before treatment, demonstrating lack of dental contact, visible gross plaque, and calculus accumulation.

are found in people with cherubism, whereas PTPN11126 and SOS1 mutations are found in patients with Noonan syndrome who have giant cell lesions.

Severe feeding difficulties are a common pattern in Noonan syndrome. However, it often goes unrecognized until failure to thrive and/or malnourishment becomes an obvious issue for these patients. Though the prevalence and underlying cause are usually poorly understood, delayed gastrointestinal motor development has been suggested as the plausible explanation for such condition (Shah et al., 1999). Great management difficulties may be encountered by pediatric dentists, because the association between widespread intestinal dysmotility and gastrointestinal reflux, coupled with developmental delay of the central nervous system, may lead to strong gag reflexes in these patients. Furthermore, craniofacial discrepancies may pose an added challenge, enhancing feeding difficulties because of the patient's inability to chew on solid foods. The purpose of this article is to present the case of a patient with Noonan syndrome, associated severe open bite, and a strong gag reflex and to discuss a conservative approach with myofunctional therapy.

Case Report

A boy aged 13 years and 5 months presented to the Pediatric Special Care Clinic at the Federal University of Ceará Dental School for his first dental evaluation. He was referred from Clinical Genetics. The patient's mother reported a chief complaint of the great difficulty in performing routine oral hygiene on her son, who wasn't able to chew solid foods. The son was diagnosed with Noonan syndrome soon after birth but moderate developmental delay and very limited learning and language skills had been identified at the time of presentation. The patient had reduced hearing ability in the right ear. However, other common congenital defects, such as cardiac, lymphatic, and bleeding abnormalities, were absent.

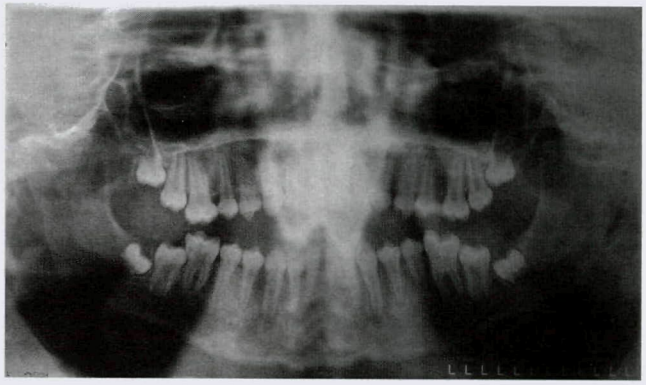

FIGURE 2 Panoramic radiograph showing erupting second permanent molars, unerupted third molars, generalized horizontal bone loss, localized vertical bone defects, symmetric condyles, and an irregular mandibular border.

On extraoral examination, the patient was found to have short stature, pectus carinatum, pectus excavatum, clinobrachydactyly, short neck, a low posterior hairline, low-set ears, and a flattened zygomatic bone. Intraoral exam was performed with great difficulty because of the patient's strong gag reflex and constant vomiting. The following were observed during examination: visible gross plaque and calculus accumulation in the sub- and supragingival areas, including the occlusal thirds of all permanent molars; gingival bleeding; periodontal pockets (average measurement of 4 mm); and no malpositioned teeth, aside from a rotated mandibular central incisor (Fig. 1). The patient was caries free. Also noted were the presence of hypotonic lips, lip incompetence, and tongue thrusting at rest and during deglutition, generating lack of maxillary-mandibular interdental contact. Thus, clinical evaluation of overbite and overjet was not possible. No underlying chronic respiratory condition was identified to explain the observed open-mouth posture, suggesting that the patient's mouth was kept open as a habit or because of low muscle tone. In spite of a nasolabial angle within normal range (100.9°), facial analysis showed dolicocephalic pattern and convex profile because of mandibular rotation. Also noted were an increased lower facial height with asymmetry and mandibular deviation to the right on opening. Interpupil and epicanthal distances were altered. Analysis of the models showed an intermolar mandibular arch length 7 mm greater than the intermolar distance in the maxillary arch, while the anterior length of the mandibular arch was reduced by 3 mm, with no maxillary or mandibular crowding. A flattened palate was also noted (25% Korkhaus Index).

In a panoramic view, radiographic examination showed the presence of all 32 permanent teeth, with erupting second permanent molars and unerupted third molars, generalized horizontal bone loss, localized vertical bone defects, symmetric condyles, and an irregular mandibular border (Fig. 2). Cephalometric analysis demonstrated vertical growth and a skeletal open bite (Fig. 3). Osseous bases were well interrelated but retruded relative to the cranial

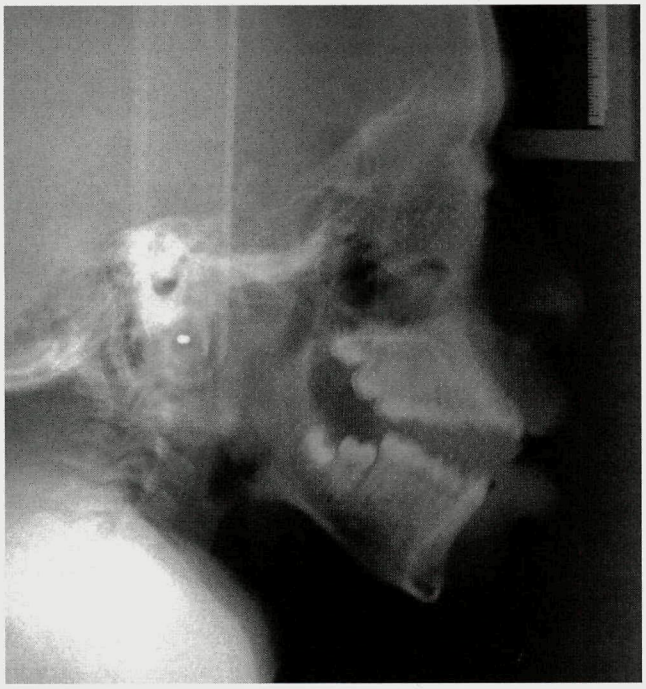

FIGURE 3 Cephalometric radiograph showing vertical growth, skeletal open bite, retruded osseous bases relative to the cranial base, anterior cranial base and mandibular lengths within normal range, inclined mandibular plane, increased lower facial height, proclined maxillary, and mandibular incisors.

base. Anterior cranial base and mandibular lengths were found to be within normal range. However, an inclined mandibular plane, rotated clockwise, resulted in mandibular retrusion, generating open bite, a lowered positioned chin, and an increased lower facial height. In spite of the predominance of vertical growth, proclined maxillary and mandibular incisors were noted. The first permanent molars were well positioned in the anterior posterior direction, and excessive eruption of mandibular incisors was present. However, despite the lack of generalized interdental contact, supraeruption of other teeth was not observed (Table 1). Soft-tissue abnormality was noted, specifically, the lips surpassed the facial esthetic plane and there was a prominent lower lip. At age 15, a hand and wrist evaluation using the Eklöf Rinjertz index identified a bone age of 9 years and 5 months, indicating a 6-year growth delay (Fig. 4).

The speech-language pathologists functional evaluation of the patient's respiration, chewing, and swallowing showed an atypical deglutition, superior respiration, hypotonic facial muscles, lip incompetence, and lack of dental occlusion. Myofunctional therapy was instituted to improve function of the stomatognathic system; muscle posture; and mobility of the lips, tongue, cheeks, and soft palate. Another aim was to prepare facial musculature for future orthognathic surgery and orthodontic treatment. To achieve these goals, awareness and training of the functions were conducted. Therefore, exercises for the lips, tongue, chin region, and chewing muscles was performed with isotonic, isometric, and isokinetic procedures for mobility, tonus, and strength, respectively.

Initially, a relaxing massage was performed on the patient's face to release the muscles for the planned exercise sequence. Subsequently, mobility exercises of the tongue were performed with rotation, vibration, and crackles. The patient was instructed to thrust the tongue against the cheeks and hard palate for strength and tonus. To correct tongue posture, the patient was coached to place the tongue on the incisal papilla. All of these exercises were alternated and sequentially followed with a specific number of repetitions. To minimize lip incompetence and exercise breathing, the patient was asked to blow into balloons. Muscle strengthening of the orbicularis oris superior, orbicularis oris inferior, buccinator, masseter, and glossal muscle groups was obtained by using a wooden tongue blade and applying contra-resistance on the muscle or on the focused group of muscles, for instance, lips, cheeks, and tongue.

Breathing exercises were performed by placing the wooden spatula on the mouth to promote nasal breathing. Finally, a rubber band was placed between the maxillary and mandibular posterior teeth, and the patient was asked to occlude and attempt chewing, resembling natural mastication cycles (Fig. 5). Two different rubber band sizes were used, and exercises were initiated from the larger to the smaller size. Once chewing ability improved, swallowing exercises were established and slowly progressed from liquids to soft and solid foods. Once this exercise sequence was concluded, speech therapy was instituted to improve language and breathing. The patient's mother was guided and asked to perform home supervision of exercises in order to collaborate with treatment. She was asked to gradually introduce solids at daily meals. Thus, in addition to a 45-minute weekly therapy session, 10-minute home daily exercises were prescribed as part of the child's routine.

Initially, preventive measures and periodontal treatment were instituted with great difficulty, using behavior guidance techniques in an attempt to desensitize patient and to achieve a better control of vomiting and gag reflex. When the patient was 15 years old, myofunctional therapy was initiated. After 4 months of therapeutic sessions and a 16-month follow-up, a significant increase in tongue, lip, and facial muscle tonus was achieved, allowing patient to chew solid foods and facilitating dental treatment, which consisted of re-examination, six sessions of scaling, root planing, dental prophylaxis, and oral hygiene instructions. (Fig. 6A and 6B). Cephalometric analysis demonstrated a decrease in lower facial height and an anticlockwise mandibular rotation, thus reducing the mandibular plane angle (Figs. 7 and 8). A decrease in facial convexity was also noted (Table 1). In addition, a reduction in the previously observed prominent lower lip led to improvement of the facial esthetic plane. The mother reported great improvement in the patient's feeding ability and gag reflex. The

TABLE 1 Ricketts-Simplified Cephalometric Analysis of Patient With Noonan Syndrome, Before and After Myofunctional Therapy

Cephalometric Measurements	*Before 13 y 5 mo*	*After 16 y 8 mo*	*Norm*	*SD*
Dental				
Interincisal angle°	115.10	104.19**	130	±10
Maxillary-mandibular relation				
Convexity (mm)	5.25	5	2	±2
Inferior facial height°	58.28**	52.63	47	±4
Teeth/osseous bases				
Maxillary molar position (mm)	15.23	19.56	Age + 3	±3
Mandibular incisor protrusion (mm)	6.11**	6.88**	1	±2
Inclination of mandibular incisor	24.78	32.48**	22	±4
Esthetics				
Labial protrusion (mm)	8.38**	4.05**	−2	±2
Craniofacial relation				
Facial depth (°)	83.48	86.96	87	±3
Facial axis (°)	87.01	92.23	90	±3
Facial cone (°)	66.02	66.69	68	±3.5
Mandibular plain angle (°)	30.50	26.34	26	±4
Maxillary depth (°)	88.92	92.36	90	±3
Internal structures				
Cranial deflection (°)	21.35	22.42	27	±3
Anterior cranial length (mm)	54.78	56.61	55	±2.5
Mandibular arch (°)	31.74	23.83	26	±4
Mandibular body length (mm)	65.00	68.00	65	±2.7

* Patient's age in years + 3 mm. ** Values over 2 standard deviations (SD) above or below the norm.

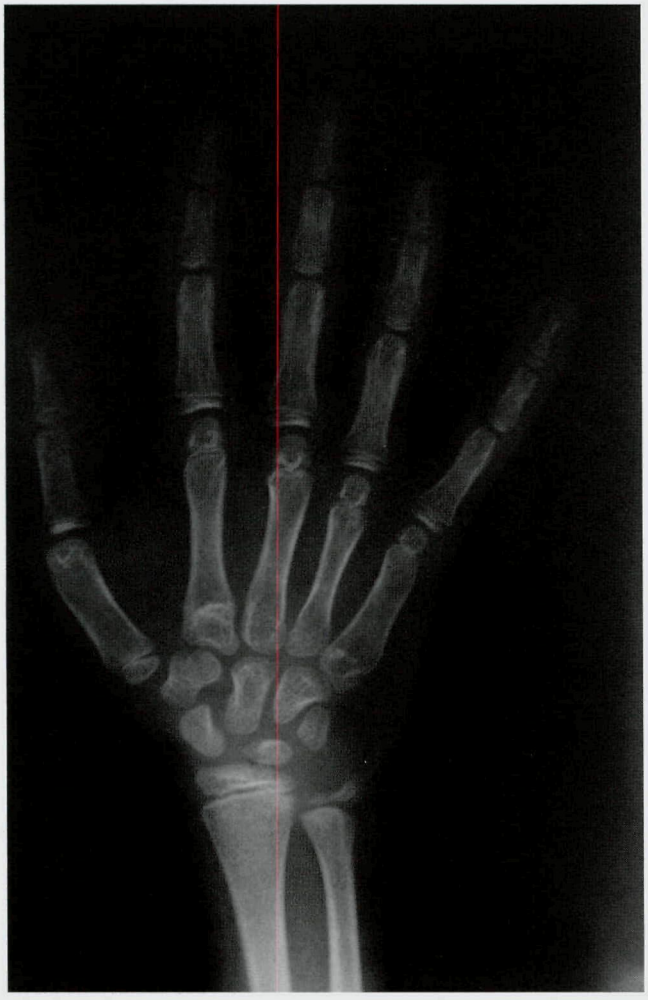

FIGURE 4 Hand and wrist radiograph showing growth delay at 15 years of age.

patient has been on monthly follow-up visits for maintenance of periodontal health and daily home oral care.

Discussion

In 2007, Shaw et al. reported a long-term follow-up of 112 patients with Noonan syndrome. The authors found that dental caries was a common cause of dental morbidity, with 15% of patients requiring extractions due to caries, whereas periodontal disease with significant bone loss and dental mobility was previously reported by Torres-Carmona et al. (1991). We have found no carious lesions associated with this patient, whereas periodontal disease with generalized bone loss was a significant finding. Most descriptions of dental abnormalities related to Noonan syndrome refer to dental malocclusions, high arched palate,

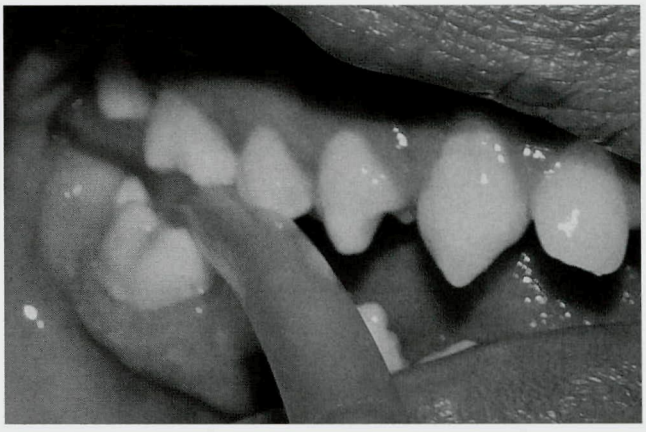

FIGURE 5 Rubber band between the maxillary and mandibular posterior teeth while patient exercised masticatory movements.

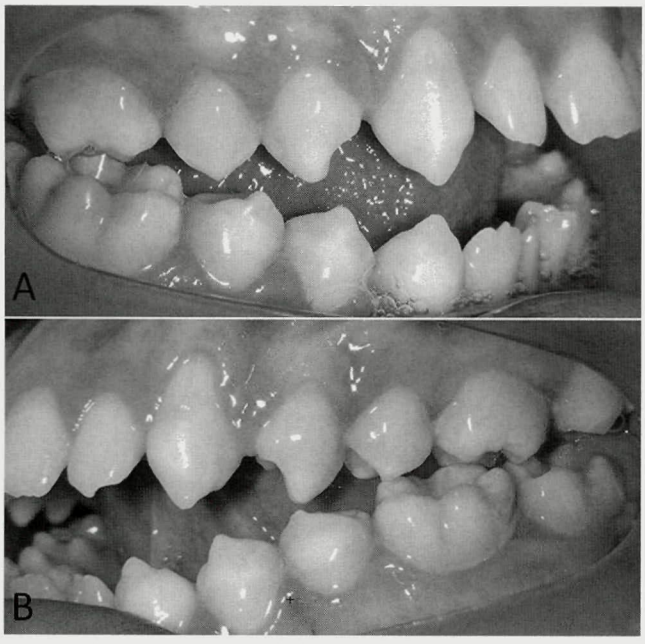

FIGURE 6 A: Right and B: Left occlusal view after myofunctional therapy (16-month follow-up), showing open bite improvement.

micrognathia, and anterior open bite as common characteristics (Torres-Carmona et al., 1991; Bertola et al., 1999; Tartaglia and Gelb, 2005; van der Burgt, 2007). In contrast to previous case descriptions (Pierpont, 1996; Ogawa et al., 2004; Tartaglia and Gelb, 2005), a flattened palate was observed. We have reported on a patient with Noonan syndrome in whom cephalometric analysis demonstrated vertical growth, mandibular retrusion, increased lower facial third, proclined incisors, and skeletal open bite. These findings concur with previous reports (Okada et al., 2003). Apparently, hypo-developed muscles, a current finding, has

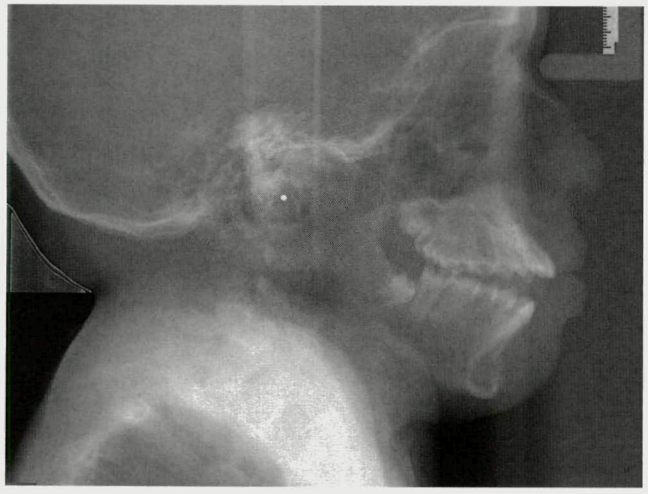

FIGURE 7 Cephalometric radiograph showing a decrease in lower facial height, an anti-clockwise mandibular rotation, and reduced mandibular plane angle.

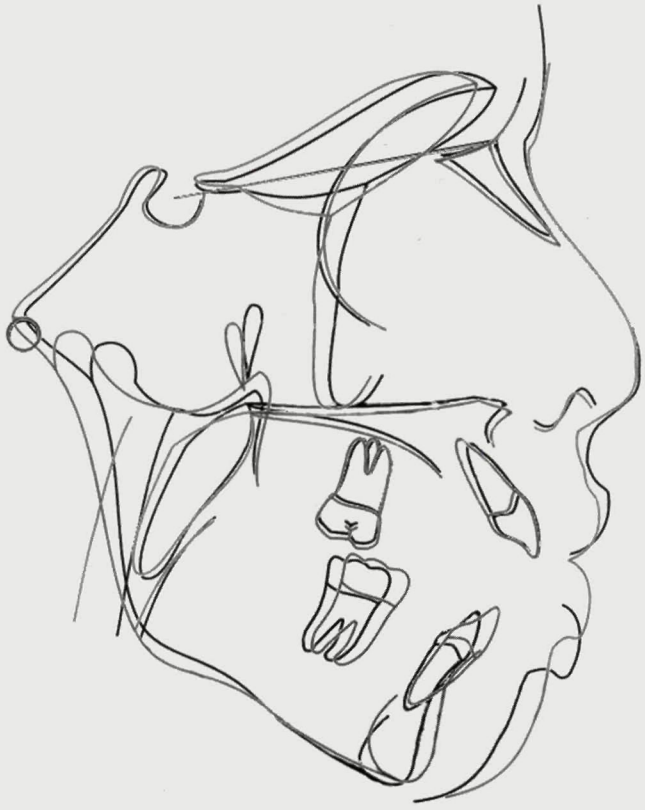

FIGURE 8 Superimposition of cephalometric tracings, before (black) and after (gray) myofunctional therapy, showing decreased lower facial height, anticlockwise mandibular rotation, and reduced mandibular plane angle.

contributed to the severe skeletal open bite of our patient. No other clinical reports have mentioned such a finding.

Masticatory function is of the utmost importance for an acceptable dentofacial development; for instance, its absence has been proposed to lead to dental crowding; disrupted dental eruption, which would generate an altered anterior-inferior facial height; and abnormal vertical growth (Profitt and Fields, 2000). Feeding difficulties in Noonan syndrome usually manifest during infancy, but there is remarkable improvement after the age of 3 or 4 years (Shah et al., 1999). It has been suggested that these abnormalities are probably an additional feature of developmental delay. Thus, when present, they could be considered an early marker of delayed language development and long-term educational achievement (Shaw et al., 2007). In this patient, reported feeding difficulties have persisted into adolescence, but there have been no descriptions of symptoms other than extreme sensitivity in the oral cavity with strong gag reflex and inability to chew solid foods, which frequently leads to immediate vomiting. To our knowledge, there are no reports in the literature of feeding problems associated with Noonan syndrome in adolescent patients. We believe that severe open bite, when associated with feeding difficulties, could possibly exacerbate such a condition. Hence, masticatory

and speech problems have been attributed to this skeletal and/or dental discrepancy (Ngan, 1997).

Changes in functional patterns may cause deviations in craniofacial development, resulting in facial skeletal discrepancies and dental malocclusions (Proffit and Fields, 2000). Craniofacial muscles perform a series of functions involving head movement, posture, chewing, swallowing, speech, and facial expression. For instance, a reduction in muscle tone, a feature of many syndromes, may allow excessive displacement of the jaws, rendering a significant vertical growth, excessive eruption of the posterior dentition, and severe open bite (Kiliaridis et al., 1989). Therefore, alterations in these functions have the potential to establish changes in the facial skeleton and the development of occlusion. Open bite has been defined as an open vertical dimension between the incisal edges of the upper and lower dentition (Subtelney and Sakuda, 1964). Although vertical malocclusion is an abnormality of a multifactorial nature, variations in growth intensity, function of the soft tissues and jaw musculature, and individual dentoalveolar development seem to influence the evolution of open bite problems (Ngan and Fields, 1997). Orofacial myofunctional disorders are specific conditions or behaviors that can negatively affect oral postures and functions, changing the vertical rest posture dimension of the interdental arch (Mason, 2005). Myofunctional therapy is a treatment procedure designed to establish new neuromuscular patterns, promoting correction of functional and resting postures; improving chewing, swallowing, and feeding patterns; and eliminating deleterious behaviors (Benkert, 1997). We have chosen a conservative approach with myofunctional therapy as a means for increasing muscle tonus in order to regain occlusion, with improvement in masticatory ability as the final outcome. The effectiveness of treatment was measured through cephalometric analysis, as has been previously described (Haruki et al., 1999).

In the present case report, myofunctional therapy led to a change in the mandibular rotation pattern, reducing mandibular plane angle and lower facial height and favoring reduction of the clinically observed open bite. These favorable skeletal changes allowed establishment of masticatory function and resulted in great improvement in feeding disorder associated with the syndrome. Furthermore, therapeutic measures favored lip competence, allowing closure. In agreement with the observed outcome, it has been suggested that orofacial myofunctional therapy can lead to improvement of open bite and overjet in the absence of prior or concomitant orthodontic treatment (Benkert, 1997). There is an understanding that orthodontic treatment and orthognathic surgery are necessary measures for full correction of severe open bite. However, because of the multifactorial nature of open bites, the benefits of noninvasive therapy do not preclude the need for an invasive treatment strategy (Ngan and Fields, 1997; Ng et al., 2008).

Hypersensitive gag reflex is a somatic natural response in which a body reaction leads to muscle contraction at the base of the tongue and the pharyngeal wall in an attempt to remove instruments or agents from the oral cavity (Bassi et al., 2004). Routine dental care in patients with strong gag reflex is usually a challenge for both the patient and the dental professional, generating increased anxiety that must be overcome. Interestingly, in the case described here, myofunctional therapy also led to a significant reduction in gag reflex, allowing routine oral hygiene procedures to be easily performed. Apparently, improvement of the stomatognathic system function and muscle posture helped the patient deprogram a previously acquired behavior pattern, which is the main goal in such cases (Wilks and Marks, 1983; Ramsay et al., 1987). A pharmacologic approach using nitrous oxide (Kaufman et al., 1988) or intravenous sedation with propofol (Yoshida et al., 2007) has been described as successfully depressing strong gag reflex in anxious young adult patients. However, in the current case report, gagging was not solely associated with dental treatment, but rather a limiting factor for routine tooth brushing and feeding, so we believed a slower, more conservative approach, with behavior guidance as the main focus, seemed more appropriate. This treatment strategy showed marked improvement in quality of life and concomitantly facilitated dental treatment. Orthodontic treatment was not considered during this stage for the reasons stated previously. Clearly, the patient would benefit from full orthodontic appliances and orthognathic surgery for complete correction of skeletal and dental malocclusions, including the remaining skeletal open bite. The extent of treatment will depend on the family's decision and the patient's ability to cooperate and maintain good oral hygiene. Muscle hypotonia is a factor that increases relapse in anterior open bite cases, and caution should be taken if orthognathic surgery is not considered. Also, the patient was identified as having skeletal growth delay, and any treatment, as well as timing, should take this factor into account.

Conclusion

Myofunctional therapy has demonstrated acceptable efficacy in the management of a patient with Noonan syndrome with feeding difficulties, hypersensitive gag reflex, and severe open bite associated with muscle hypotony, successfully rehabilitating masticatory function after a 16-month follow-up.

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