Journal of Oral Science, Vol. 45, No. 2, 117-121, 2003

Case report

Oral findings in Noonan syndrome: report of a case

Mitsugi Okada, Naoko Sasaki, Yasutaka Kaihara, Rinzo Okada,
Hideaki Amano, Kazuo Miura and Katsuyuki Kozai

Department of Pediatric Dentistry, Hiroshima University Faculty of Dentistry, Hiroshima 734-8553

(Received 6 March 2002 and accepted 16 May 2003)

Abstract: Oral findings in a case of Noonan syndrome in an 8-year-old Japanese male are reported. Examination of the patient revealed a narrow, high-arched palate and an anterior open bite. Cephalometric measurements showed a wide gonial angle, a large mandibular plane angle, a large Y-axis and long facial height. It is suggested that the patient had a skeletal open-bite malocclusion, which included an abnormal swallowing habit. (J. Oral Sci. 45, 117-121, 2003)

Key words: Noonan syndrome; high-arched palate; open bite.

Introduction

Noonan syndrome was described by Noonan and Ehmke in 1963 as a multisystem disorder, characterized by such clinical features as short stature, hypertelorism, ptosis, and low-set ears (1). This syndrome is often likened to Turner syndrome, because they share some of the same features, including epicanthic folds, right-sided congenital heart disease, and various skeletal malformations (2). In Noonan syndrome, no consistent chromosomal abnormality has been found. Recently, by a genome-wide linkage analysis of a large Dutch kindred group with autosomal dominant Noonan syndrome, Jamieson et al. (3) localized the gene to chromosome 12 (Zmax = 4.04 at 0 = 0.0), which is located on chromosome 12q, between D12S84 and D12S366.

Noonan syndrome is relatively common, with an estimated incidence of between 1 per 1000 and 1 per 2500 live births (4). Many cases have been reported; however, few, including those with Turner syndrome, describe the characteristic oral features of a narrow, high-arched palate (5-7) and an open bite (8).

The present article describes the oral findings in a case of Noonan syndrome in an 8-year-old Japanese male.

Correspondence to Dr. Mitsugi Okada, Department of Pediatric Dentistry, Hiroshima University Faculty of Dentistry, 1-2-3 Kasumi, Minami-ku, Hiroshima 734-8553, Japan
Tel: +81-82-257-5698
Fax: +81-82-257-5699
E-mail: mitsugi@hiroshima-u.ac.jp

Case Report

Medical history

A Japanese male aged 8 years and 10 months was referred to the pediatric dental clinic of Hiroshima University Dental Hospital for orthodontic examination. He had been diagnosed as having Noonan syndrome and a heart murmur at his birth in Hiroshima, Japan. After diagnosis, the patient was followed and then underwent a catheterization procedure for pulmonary stenosis at 3 years of age. At 6 years he underwent plastic surgery for a webbed neck. The results of these two procedures were satisfactory. The patient also suffered from cervical ribs and nystagmus, but no treatment had been required.

Our physical examination revealed a very active, short-stature 8-year-old boy who was in the fifth percentile for height and weight. Rohrer's index was normal at 115.8. The patient's neck was short and webbed, and his chest was pectus excavatum. Facially, the patient exhibited hypertelorism, ptosis, a divergent squint, and a tendency for exophthalmos (Fig. 1). The family history was negative for Noonan syndrome and facial disproportion.

Oral examination

Oral examination revealed a narrow, high-arched palate and a hypoplastic mandible with a posterior cross-bite at the first primary molars (Fig. 2). In addition, we found an anterior open-bite, which had been present since the appearance of his primary dentition. His tongue was somewhat cone-shaped. The upper primary canines had exfoliated early when the upper second permanent incisors

erupted. An abnormal swallowing habit was also observed, although no nasal obstruction was present.

Radiographic examination

Figure 3 shows frontal and lateral head radiographic views of the patient. Cranial sutures were normal, as they appeared to have closed. Digitate impressions were seen on the front of the skull, along with cervical spinal fusions at the second, third, and fourth vertebral arch. He had an intact dentition in a good state of repair with no carious lesions seen clinically or on X-ray. All permanent teeth, except for the third molars, were observed to be present

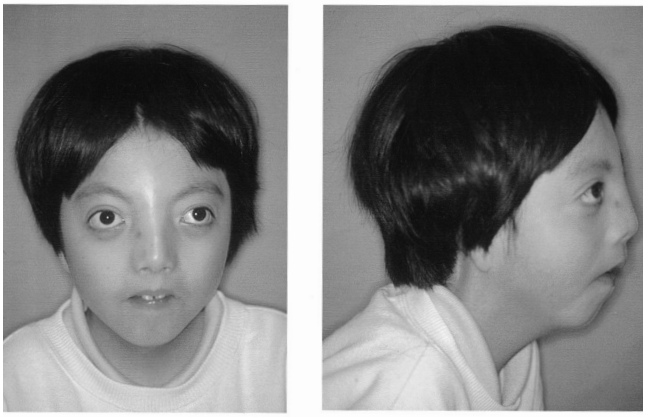

Fig. 1 Patient's full face and profile at 8 years of age.

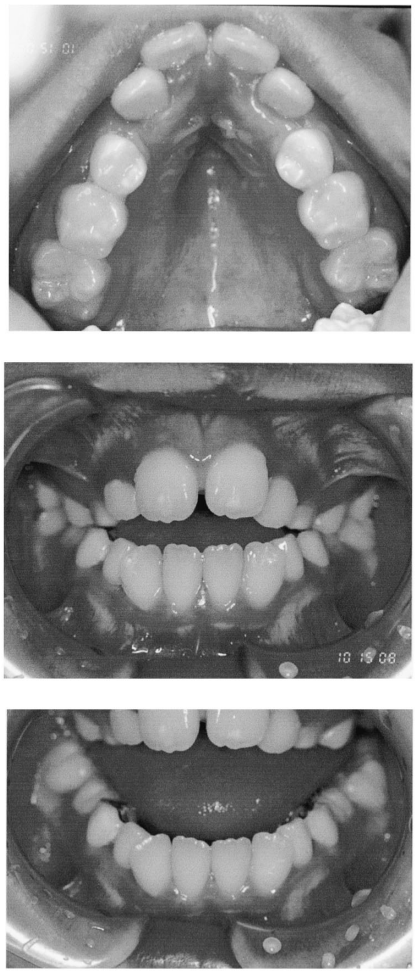

Fig. 2 Oral features of the maxilla, mandible and occlusion.

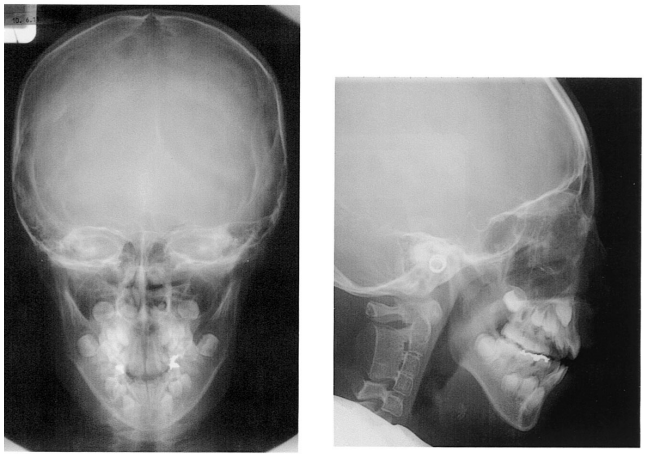

Fig. 3 Radiographs of the frontal and lateral portions of the head.

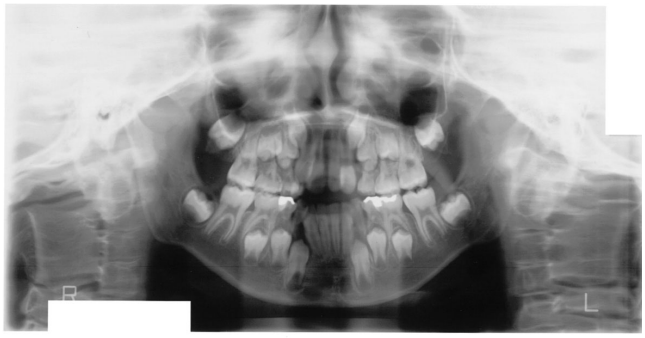

Fig. 4 Patient's orthopantomograph.

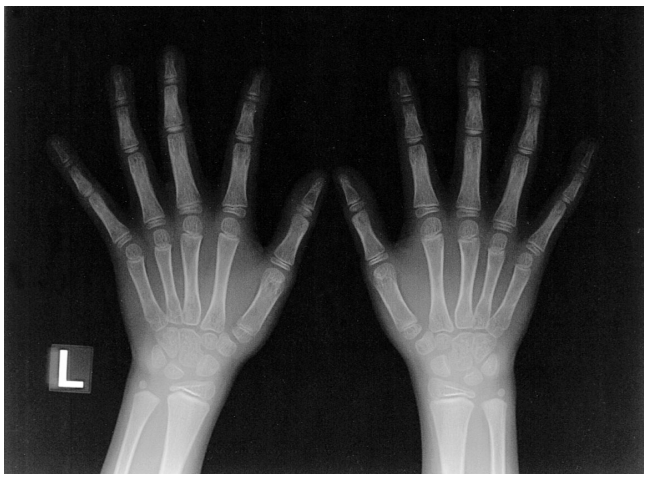

Fig. 5 Radiograph of the patient's hand-wrist.

on an orthopantomograph (Fig. 4).

Hand-wrist radiographs confirmed the bone age of an eight-year-old (Fig. 5). The landmarks and measurements used, taken according to the method of Sakamoto (9) and Japanese society of Pediatric Dentistry (10) are shown in Fig. 6. Table 1 shows a summary of the lateral cephalometric measurements of the patient. Cephalometric analysis revealed a wide gonial angle and high total and lower facial heights (N-Me, ANS-Me). In addition, the Y-axis, mandibular plane angles and ramus were over 1 SD larger than mean values, while the SNB and interincisal angle were over 1 SD smaller.

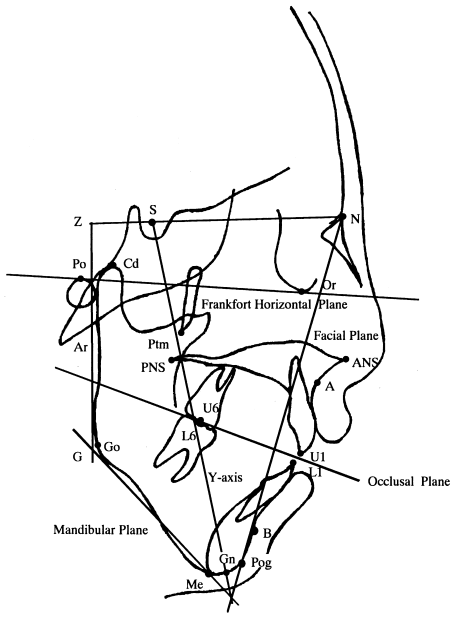

Fig. 6 Angular and linear measurements used in this study.

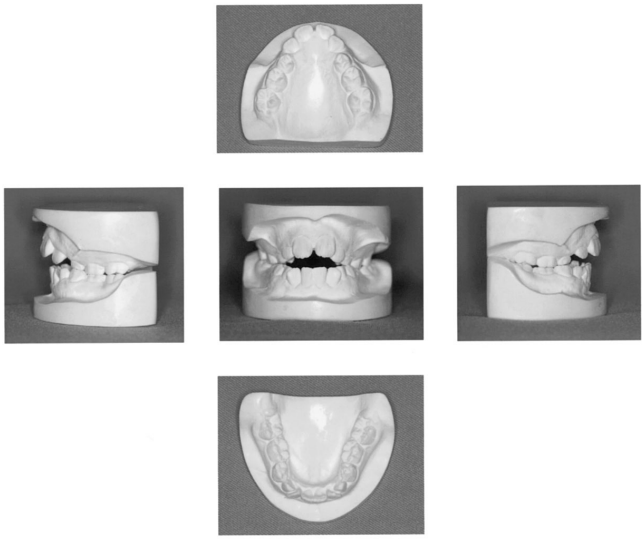

Fig. 7 Study model of the patient's maxilla.

Study model analysis

A study model of the maxilla was prepared for examination of the high-arched palate (Fig. 7). The width of each tooth was within the normal range. The patient was found to have a narrow palate and a right simian crease using a scanner (PICZA model PIX-3, Roland DG Co., Japan) in which 0.5 mm interval scanning was available (Fig. 8). Table 2 shows a summary of the dental arch length measurements, using an analytical method described by Ootsubo et al (11). The width of the dental arch was over 2 SD narrower at the maxilla than the standard value, and over 1SD narrower at the mandible. Further, the patient had an over bite of -4.2 mm and an over jet of 2.8 mm.

The height of the palate was measured from the maxillary casts, using a previously established three-dimensional measurement system for dental casts (12). The standard horizontal plane was defined by 4 points; P2 and P3 showed the deepest point of the cervical portion on the

Table 1 Summary of cephalometric measurements

Measurements	Mean	SD	Case
SNA	81.4	3.3	78.9
SNB	76.2	2.9	72.1*
ANB	6.7	2.4	7.0
Y-axis (FHP to S-Gn)	63.8	3.6	74.7*
Mandibular plane angle (SN)	39.5	4.0	44.3*
Occlusal plane angle	14.5	4.0	18.1
UU to SN	96.8	8.2	99.1
LL to Mand. plane	89.5	6.5	89.6
Interincisal angle	133.9	12.1	121.9*
Gonial angle (MGZ)	130.1	1.3	134.3*
Ramus (GZN)	89.4	5.3	95.1*
N-S (mm)	64.6	2.8	62.1
N-Me (mm)	106.5	3.9	124.6*
N-ANS (mm)	47.8	2.7	48.5
ANS-Me (mm)	62.4	3.3	79.5*

*: 1 SD more or less than standard

Table 2 Summary of dental arch lengh measurements

	Mean	SD	Case
Maxilla			
1-E	28.1	1.9	29.3
C-C	34.2	2.2	-
D-D	33.7	2.0	19.2**
E-E	37.9	2.1	26.2**
6-6	41.8	2.2	33.0**
Mandible			
1-E	24.9	1.5	26.1
C-C	27.1	2.3	20.6**
D-D	29.1	2.1	25.2*
E-E	32.5	1.9	24.1**
6-6	36.1	2.6	30.1**

*: 1 SD less than standard, **: 2 SD less than standard (mm)

palatal side of the upper first primary molars and the upper second primary molars on both sides, respectively (Fig. 9). Four lines were drawn (3 parallel and 1 perpendicular); from the highest vertex of the papilla of the incisor (P1) to the midpoint between the deepest points of the cervical portions of the upper first and second primary molars, and the first permanent molars on each side of the palate (L1, L2, L3 and L4 in Fig. 9). The height of the palate was defined as the distance between the intersections and the standard plane.

Figure 10 shows the patient's palatal height compared to data from twenty normative subjects at Hellman's dental stage IIIA. P1, P2, P3 and P4 show the upper palatal point on the first permanent incisor, first primary molar, second primary molar, and first permanent molar, respectively. The palatal depth was over 1 SD higher at P2, P3 and P4 than standard values. The palatal depth at P3 was the highest of all measured points, with a value of 14.3 mm.

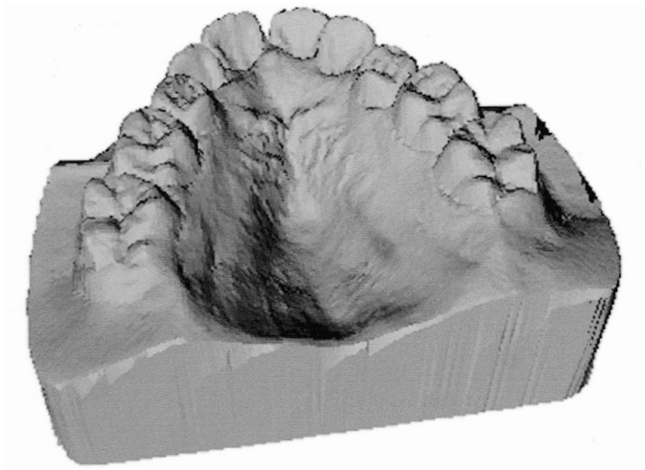

Fig. 8 Scanning image of maxilla using PICZA.

Discussion

The high frequency of cardiac, ophthalmic, growth, and orthopedic symptoms, associated with Noonan syndrome, emphasizes the need for early diagnosis. Early diagnosis of this relatively common condition would result in better use of diagnostic and therapeutic approaches (13). Moreover, it is important to recognize the marked phenotypic changes with age from newborn to adult.

The present case was diagnosed at birth, and the results of procedures undertaken for pulmonary stenosis and webbed neck have been satisfactory.

The present report describes a narrow high-arched palate and an open bite with accompanying Noonan syndrome. There have been several other reports of oral findings with this syndrome (5-8), however, few have reported a narrow high-arched palate with an open bite (7). Sugar et al. (8) described a Noonan syndrome patient who had marked mandibular prognathism with an anterior open bite.

In the present case, the patient's palatal height was markedly higher at the measured points of the upper first primary molar, second primary molar, and first permanent molar than those of standard values, while the values for dental arch length of the maxillar and mandibular were

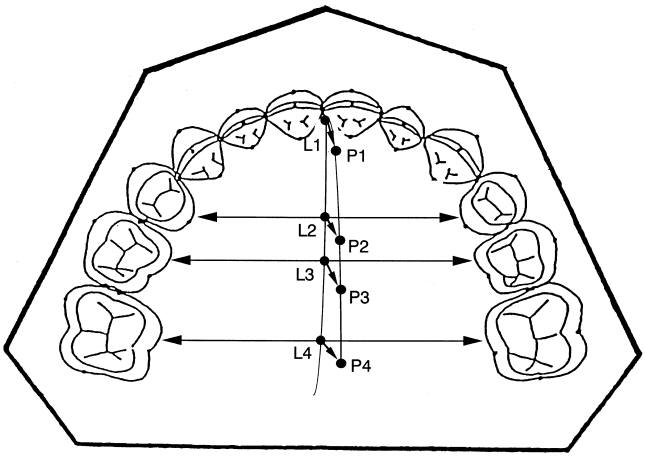

Fig. 9 Diagram of palatal height measurements used in this study: P1: palatal point on the first permanent incisor; P2: palatal point on the first premolar; P3: palatal point on the second premolar; P4: palatal point on the first permanent molar; L1: midpoint on the first permanent incisor; L2: midpoint on the first primary molar; L3: midpoint on the second primary molar; L4: midpoint on the first permanent molar.

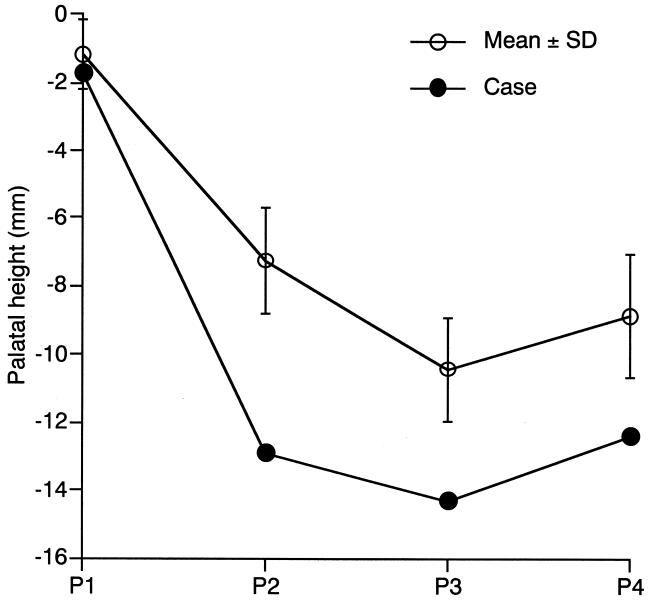

Fig. 10 Palatal height of the patient, compared with data from twenty normative subjects at Hellman's dental stage IIIA.

normal. In contrast, the width of the dental arch was markedly narrow at both the maxilla and mandibular landmarks. Early closure of cranial sutures may influence the growth of the dental arch. The present case exhibited hypertelorism, ptosis, a divergent squint and a tendency for exophthalmos. Noonan (14) reported that deformity of the sternum with an early closure of sutures was a frequently-seen feature. Unremarkable cranial sutures, along with digitate impressions were noted in this patient. Kreiborg and Pruzansky (15) suggested that, in premature craniosynostosis, malocclusion is best explained by a lack of maxillary sutural growth in all three planes of space. In addition, a corn-shaped tongue might also contribute to a narrow dental arch. Longitudinal data on individual patients are required in future studies.

The results of cephalometric measurements showed a wide gonial angle and a large mandibular plane angle. In addition, the facial height (N-Me and ANS-Me) was longer than that of the mean values. It was also noted that the Y-axis was larger than that of the mean value. Nahoum (16) suggested that the severity of an open-bite malocclusion is determined by several craniofacial measurements (the gonial angle, mandibular plane and facial height) that are large when compared with corresponding statistical norms. Kagami (17) also showed that in Japanese open-bite cases, the gonial angle is large and the height of front lower portion of the face (ANS-Me) is long. It is suggested that this case had a skeletal open-bite malocclusion, which included an abnormal swallowing habit. Orthodontic treatment for the narrow dental arch and open-bite are planned for the future, since obvious discrepancies have been revealed in the permanent dentition.

Acknowledgments

We wish to thank the Research Center for Molecular Medicine, Faculty of Medicine, Hiroshima University for the use of their facilities.

References

1. Noonan JA, Ehmke DA (1963) Associated noncardiac malformation in children with congenital heart disease. J Pediatr 63, 468-470
2. Tuner HH (1938) A syndrome of infantilism, congenital webbed neck and cubitus valgus. Endocrinology 23, 566-574
3. Jamieson CR, van der Burgt I, Brady AF, van Reen M, Elsawi MM, Hol F, Jeffery S, Patton MA, Mariman E (1994) Mapping a gene for Noonan syndrome to the long arm of chromosome 12. Nature Genet 8, 357-360
4. Nora JJ, Nora AH, Sinha AK, Spangler RD, Lubs HA (1974) The Ullrich-Noonan syndrome (Turner phenotype). Am J Dis Child 127, 48-55
5. Nelson JF, Tsaknis PJ, Konzelman JL (1978) Noonan's syndrome: report of a case with oral findings. J Oral Med 33, 94-96
6. Bernier-Buzzanga J, Su WP (1990) Noonan's syndrome with extensive verrucae. Cutis 46, 242-246
7. Horowitz SL, Morishima A (1974) Palatal abnormalities in the syndrome of gonadal dysgenesis and its variants and in Noonan's syndrome. Oral Surg Oral Med Oral Pathol 38, 839-844
8. Sugar AW, Ezsias A, Bloom AL, Morcos WE (1994) Orthognathic surgery in a patient with Noonan's syndrome. J Oral Maxillofac Surg 52, 421-425
9. Sakamoto T (1959) A study on the developmental changes of dentofacial complex of Japanese with special reference to sella turcica. Nippon kyosei shika gakkai zasshi 18, 1-17 (in Japanese)
10. Japanese society of Pediatric Dentistry (1995) A study on the cephalometric standards of Japanese children. Shouni shika gaku zasshi 33, 659-696 (in Japanese)
11. Ootsubo J, Ishikawa F, Kuwahara Y (1964) A longitudinal study of dental development between 6 to 13 years of age. -Growth changes of dentition-. Nippon kyosei shika gakkai zasshi 23, 182-190 (in Japanese)
12. Kaihara Y, Amano H, Miura K, Nagasaka N (1998) A study of a new method for diagnosis of the dentition and occlusion in children with a three-dimensional measurement system of dental casts. Shoni Shikagaku Zasshi 36, 839-847 (in Japanese)
13. Lucker GP, Steijlen PM (1994) Widespread leucokeratosis in Noonan's syndrome. Clin Exp Dermatol 19, 414-417
14. Noonan JA (1968) Hypertelorism with Turner phenotype. A new syndrome with associated congenital heart disease. Am J Dis Child 116, 373-380
15. Kreiborg S, Pruzansky S (1981) Craniofacial growth premature craniofacial synostosis. Scand J Plast Reconstr Surg 15, 171-186
16. Nahoum HI (1977) Vertical proportions: a guide for prognosis and treatment in anterior open-bite. Am J Orthod 72, 128-146
17. Kagami H (1972) A morphological consideration of the open bites and over bites with reference to analytical values of the cephalograms and casts. Nippon kyosei shika gakkai zasshi 31, 45-60 (in Japanese)