Case Report

Multidisciplinary Treatment Approach of Morquio Syndrome (Mucopolysaccharidosis Type IVA)

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ABSTRACT

Morquio syndrome (mucopolysaccharidosis type IVA) is an autosomal recessive disorder caused by the accumulation of mucopolysaccharides in lysosomes because of the deficiency of *N*-acetylgalactosamine-6-sulphate sulphatase. Patients with Morquio syndrome often need orthodontic treatment. This study demonstrates the correction of malocclusion by orthodontic treatment and the improvement of the masticatory function by prosthodontic treatment in a male patient affected by Morquio syndrome. Labial inclinations of mandibular and maxillary anterior teeth were corrected, and spaces necessary for prosthodontic restorations were preserved. After this procedure, the restorations were made, and Hawley appliances were used in both jaws for retention. Patient satisfaction and an acceptable occlusal relationship were achieved by following an optimal treatment plan when considering the general status of the patient with Morquio syndrome. (*Angle Orthod* 2006;76:335–340.)

KEY WORDS: Multidisciplinary treatment; Morquio Syndrome

INTRODUCTION

Morquio syndrome, mucopolysaccharidosis type IVA (MPS-IVA), was first described in 1929 by Louis Morquio,[1] a pediatrician from Uruguay. MPS-IVA is an autosomal recessive disorder caused by the accumulation of mucopolysaccharides in lysosomes because of the deficiency of *N*-acetylgalactosamine-6-sulphate-sulphatase. This enzyme hydrolyzes the sulfate ester bonds of *N*-acetylgalactosamine-6-sulfate at the nonreducing end of chondroitin-6-sulfate and galactose-6-sulfate at the nonreducing end of keratan sulfate.[2,3] This syndrome shows an autosomal recessive heredity and affects one in 40,000 live births.[4]

The syndrome is characterized by severe skeletal changes, which include hypoplasia of the odontoid process, short neck, pectus carinatum, kyphoscoliosis, and dwarfism. Hypoplastic odontoid processes causing atlantoaxial subluxation and cervical myelopathy are usual clinical findings.[5] Other characteristics are laxity of joints,[6] mineralization disturbances of tooth enamel,[7,8] and neurosensory deafness.[9] The glaucoma often observed in these patients is not considered a feature of the syndrome.[10] Other features of this disease are cardiac abnormalities, respiratory problems, acoustic deafness, and dental abnormities.[11] The heart is also affected by Morquio syndrome. Cardiac problems usually develop later in life and result from the accumulation of mucopolysaccharides in the heart valves. This can weaken the valves, leading to murmurs and heart failure.[12] The respiratory problems in Morquio syndrome may be a restrictive defect due to thoracic cage deformity or upper airway obstruction during head flexion, or both. There have been isolated reports of sleep-disordered breathing.[13] Morquio syndrome also affects hearing. Patients can have conductive or nerve deafness, and commonly have a combination of both. The use of a hearing aid can help compensate for this problem.[14]

The specific dental findings described in these patients include tapered posterior teeth with pointed cups and spaced and flared maxillary anterior teeth. The patients show an anterior open bite, despite a short vertical facial height. The temporo-mandibular joint (TMJ) is affected with severe resorption of the head of

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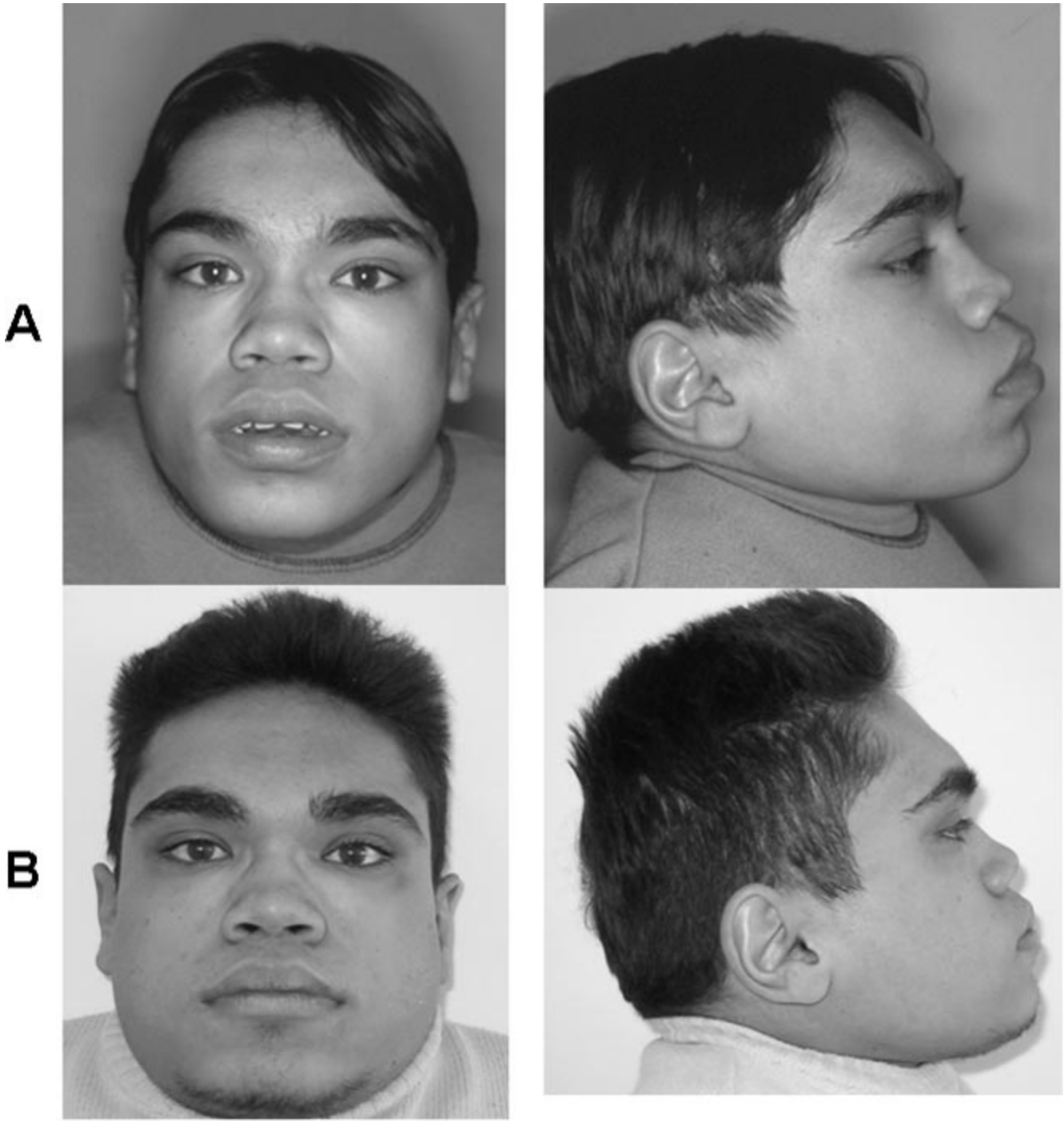

FIGURE 1. Facial photographs. (A) Pretreatment stage (21 years 4 months). (B) Posttreatment stage (24 years 9 months). Left, frontal view; right, lateral view.

the condyle. The enamel is usually less than 25% of normal thickness but of normal radiodensity on radiographs. Despite these findings, the patients exhibit a low rate of caries. The clinical and radiographic appearance of the teeth resembles hypoplastic amelogenesis imperfecta with thin enamel of normal radiodensity.[14–16] Mental retardation is not a feature of this syndrome. Life expectancy is normally less than 30 years, but isolated cases of long survival have been documented.[17]

Patients with Morquio syndrome often need orthodontic treatment.[18] This report demonstrates the correction of a malocclusion by orthodontic treatment and the improvement of the masticatory function by pros-

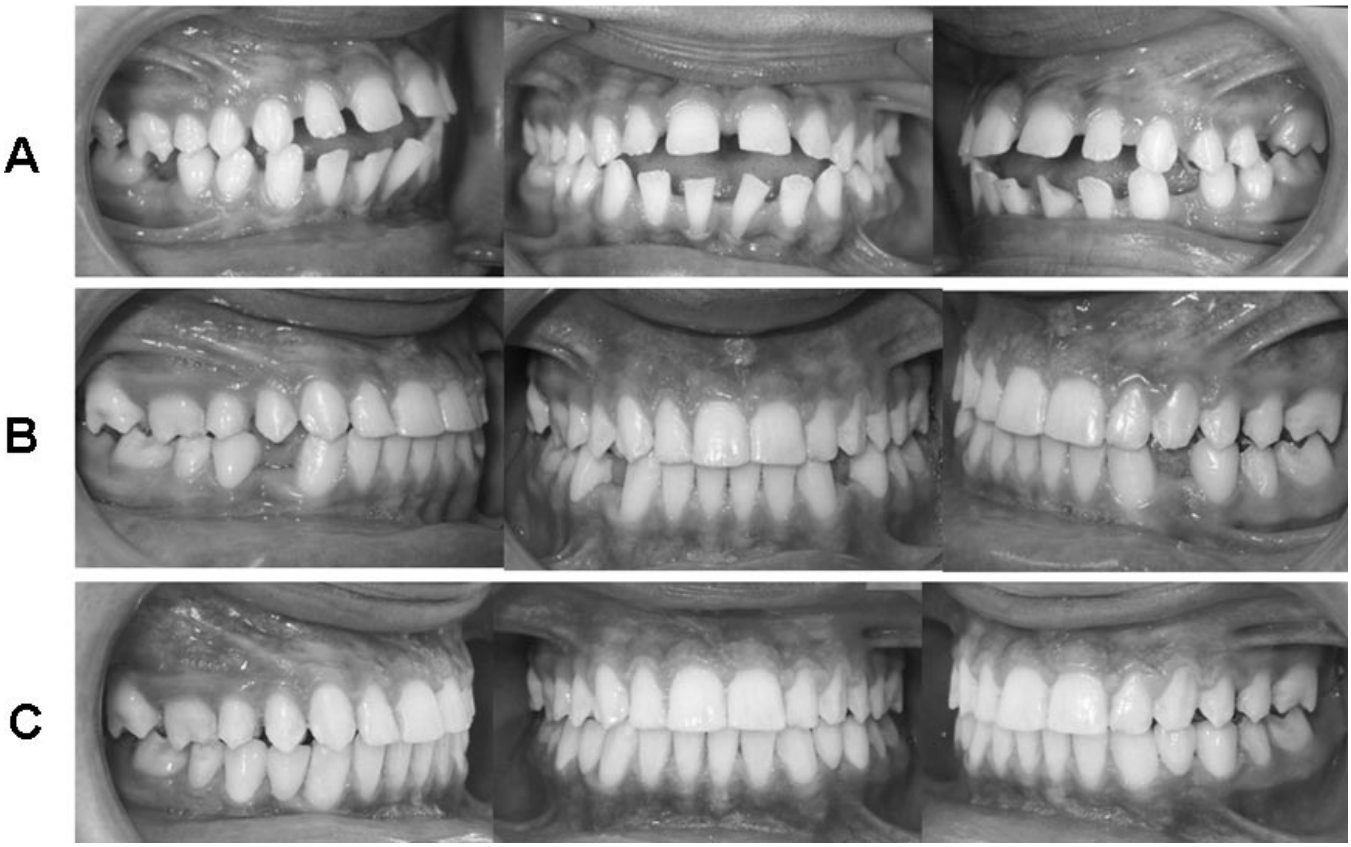

FIGURE 2. Intraoral photographs. (A) Pretreatment stage (21 years 4 months). (B) Posttreatment stage (24 years 9 months). (C) Prosthodontic treatment stage (24 years 10 months). Left, lateral view on the right side; center, frontal view; right, lateral view on the left side.

thodontic treatment in a male patient affected by Morquio syndrome.

CASE DESCRIPTION

The characteristic skeletal deformities of Morquio syndrome were discovered during the clinical inspection. The patient was 22 years 4 months old. He was 102 cm tall and weighed 21 kg. The symptoms were difficulty in walking, malformations of fingers and toes, bilateral corneal opacity, and the loss of hearing ability, 30–50 dB of 1000–8000 Hz. Respiratory problems were present under physical effort. No valvular pathology or heart murmur was found. There was no mental retardation. Cultured fibroblast from the patient showed a total absence of *N*-acethylgalactosamine-6-sulphate-sulphatase. In the patient, MRI revealed hypoplastic odontoid process, atlantoaxial subluxation, and stenotic foramen magnum. No neurological abnormalities have been observed.

Facial findings

The nasal bridge was flat, the alae of the nose were flared, the nasal tip was upturned, the mouth was broad, and the lower third of the face was prominent. The head appeared to sit squarely on the shoulders. The lips were prominent, and the head posture was slightly inclined backwards (Figure 1A).

Radiographic and dental finding

The patient showed an Angle Class I malocclusion with spacing in the maxillary and mandibular dentition. The space available was 21 mm for the maxillary teeth and 36 mm for the mandibular teeth. The maxillary and mandibular anterior teeth were spaced and flared; the posterior teeth were tapered and had pointed cusps. The patient showed an anterior open bite, despite a low vertical facial height (Figure 2A).

The lateral cephalometric radiograph showed that the anteroposterior relationship between the maxilla and the mandible was a skeletal Class II (ANB angle 5.5°). The SNA angle was 91° and SNB angle was 85.5°. Both the maxilla and the mandible were located in a more anterior position. The patient showed normal vertical facial height (SNMP = 32°). The mandibular and maxillary incisors showed a labial inclination (U1SN angle 130°, U1-NA angle 38.5°, IMPA angle 111°, L1-NB 51°). According to Ricketts E line, the pa-

TABLE 1. Cephalometric Analysis

Measurements	Pretreatment (22 y 4 mos)	Posttreatment (24 y 9 mos)
Angular (°)		
SNA	91	91
SNB	85.5	87
ANB	5.5	4
U1 to NA	38.5	37
L1 to NB	51	31.5
Intreincisal angle	85	110
SNMP	32	31
U1 to SN	130	128
IMPA	111	91
Linear (mm)		
U1 to NA	10	9
L1 to NB	12.5	7.5
Rickets E line	Upper 8 mm/ lower 9 mm	Upper 7 mm/ lower 6.5 mm

tient showed protrusion of both the upper and the lower lip (upper lip 8 mm, lower lip 9 mm) (Table 1).

The panoramic radiograph indicated that the mandibular right third molar was horizontally impacted and the other three third molar teeth were not formatted. The mandibular right and left first molar and maxillary left first molar were extracted. The tooth enamel was considered thin but normal in radiodensity. The dentin, pulp chambers, and root canal systems of all teeth were normal (Figure 3A).

Treatment plan

There was 21 mm of space present in the maxillary arch, and closure of this space was performed by mesial movement of the posterior teeth and retraction of the maxillary incisors. A total of 36 mm of space was present in the mandibular arch, and 21 mm of this space was closed with mesial movement of the posterior teeth and incisor retraction. The remaining 15 mm of space was distributed symmetrically on the right and left side between the first premolars and the canines for prosthetic restorations. Adhesive bridges were planned for prosthodontic restorations rather than implant therapy because the patient did not want to undergo the surgical procedures associated with the fixture placement.

Treatment progress

The preangulated and pretorqued 0.018-inch Edgewise appliances (Roth System, Forastadent, Pforzeim, Germany) were used. The leveling stage was completed using 0.014-inch and 0.016-inch archwires. The mesial movement of the posterior teeth was achieved using 0.016 × 0.022–inch SS archwires for both mandibular and maxillary arches, with sliding mechanics.

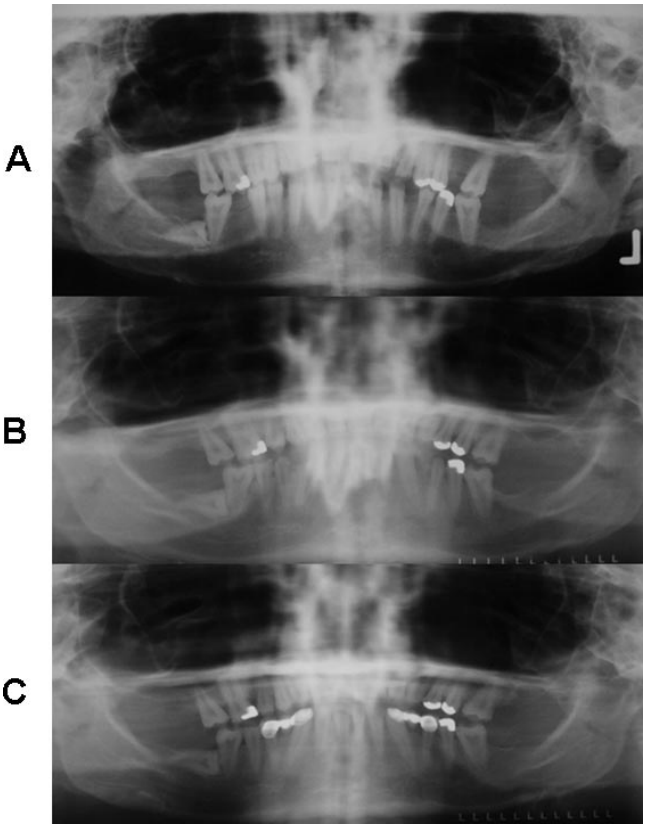

FIGURE 3. Panoramic radiographs. (A) Pretreatment stage (21 years 4 months). (B) Posttreatment stage (24 years 9 months). (C) Prosthodontic treatment stage (24 years 10 months).

Necessary spaces for prosthodontic treatment were gained bilaterally in the mandibular arch using 0.016 × 0.022–inch SS archwires. Incisor retraction was performed using 0.017 × 0.025–inch SS archwires with closing loops. For the correction of open bite, the box elastics were used. For the prosthodontic treatment, both first premolars on each side were prepared for veneer crowns, and adhesive type preparations were made for the canines.

RESULTS

Labial inclination of the mandibular and maxillary anterior teeth was corrected, and spaces necessary for prosthodontic restorations were preserved. After this procedure, the prosthodontic restorations were made as three unit bridges with one adhesive retainer on the canines (Figure 2B,C). Hawley appliances were used in both jaws for retention. As a result, optimal intercuspation of the molar has been achieved at the age of 24 years 9 months. Orthodontic treatment time was two years five months.

DISCUSSION

Morquio syndrome is characterized by short neck, deafness, and muscle weakness. In the dentofacial

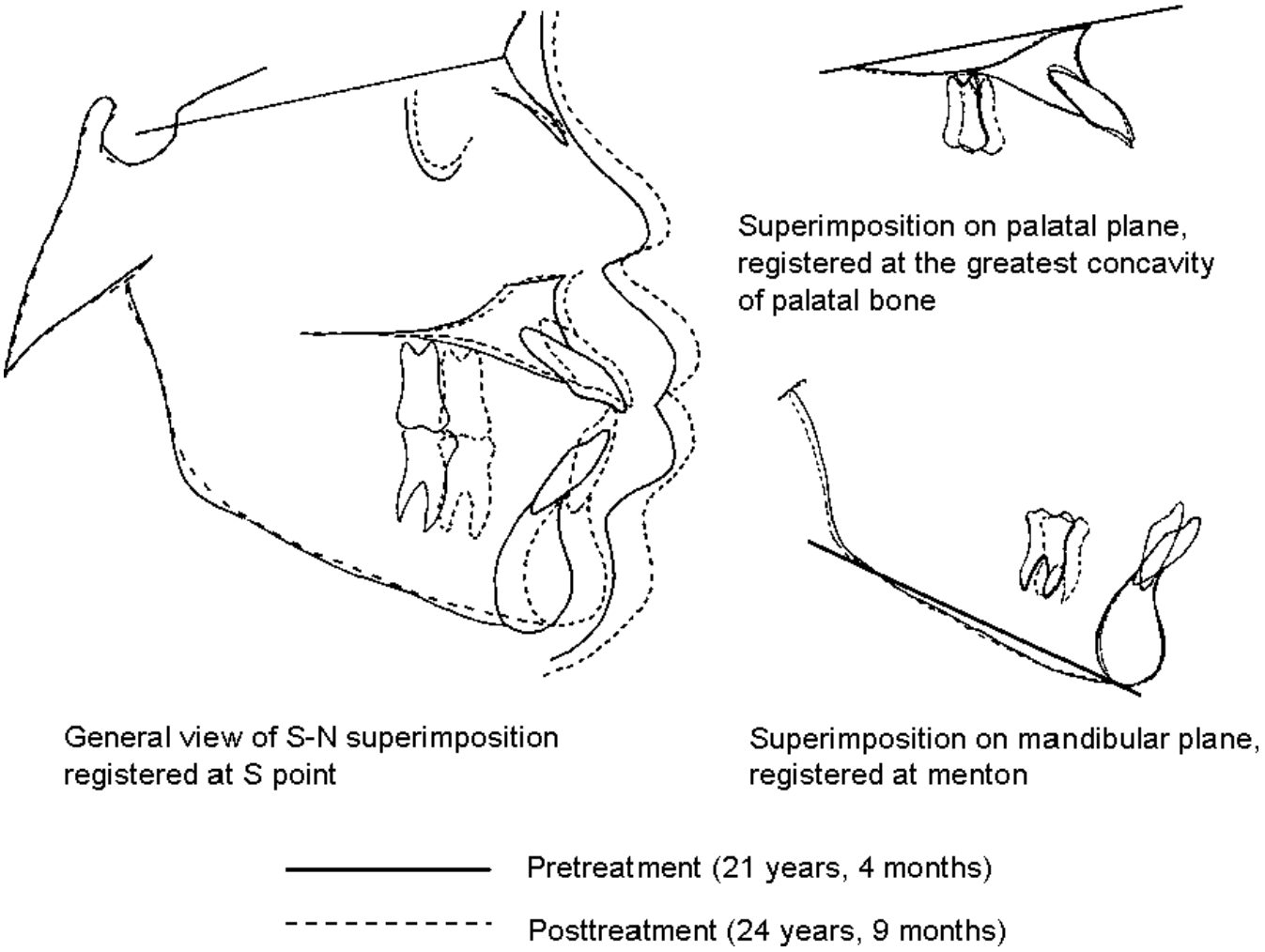

FIGURE 4. Superimposed tracings of lateral cephalograms.

area, this syndrome is characterized by a flat nasal bridge, flared alae nasae, broad mouth, labially inclined maxillary and mandibular incisors, spaced arch, thin enamel, and pits of the enamel. This case showed these characteristics.

In the patient, MRI revealed a hypoplastic odontoid process, atlantoaxial subluxation, and a stenotic foramen magnum. In Morquio syndrome, both mortality and morbidity are related primarily to atlantoaxial subluxation resulting from the instability of the odontoid process.[5] A minor fall or extension of the neck can result in cord transection and subsequent quadriparesis or death,[19] and it can be avoided by surgical craniocervical fusion.[5] Regarding the magnetic resonance imaging (MRI) findings, the patient is under neurological follow-up and surgical intervention is planned.

In the literature, it is suggested that strict oral hygiene instruction and use of nonbonded orthodontic appliances are desirable in the orthodontic treatment of a patient with Morquio syndrome.[18] In this case, the oral hygiene status of the patient was evaluated, and the use of fixed appliances was deemed appropriate. With the high level of cooperation of the patient, the oral hygiene was good throughout the treatment procedure. There was no enamel breakage in debonding.

The labially inclined teeth were retracted, and box elastics were used to correct the anterior open bite (Figure 4). The open bite was corrected not only with incisor extrusion but also with a decrease of the mandibular plane angle. Hawley appliances were used for retention, and long-term retention was planned. Adhesive bridges were used for prosthodontic restorations.

The right mandibular third molar was impacted (Figure 3B,C). After a surgical consultation, it was decided to follow the third molar with periodic radiological evaluations, especially considering the lack of odontogenic symptoms and the systemic risks of a surgical procedure.

As a result, patient satisfaction and an acceptable occlusal relationship were achieved by following an optimal treatment plan when considering the general status of the patient with Morquio syndrome.

CONCLUSIONS

- Impaired facial appearance is a most common finding in Morquio syndrome.
- In this case, orthodontic and prosthodontic treatment created an esthetically satisfactory occlusion and emphasized the importance of dental care for patients with Morquio syndrome type IVA.

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