CLINICAL CASE

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Open Bite Deformity in Amelogenesis Imperfecta – Case Report

Zgryz otwarty w *amelogenesis imperfecta* – opis przypadku

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Abstract

Amelogenesis imperfecta hereditaria (AI) appears to be rather rare defect requiring continous observation and publishing the results. AI is frequently found in association with anterior open bite (AOB).Treatment of AOB deformities is known to be difficult and results are not always predictable. In combination with AI, surgical correction of AOB can be one of the most difficult surgical procedures in orthognathic surgery. Orthodontic options are limited, and therefore, a multisegment Le Fort I or an alveolar segmental osteotomy is necessary, followed by pro-sthetic rehabilitation. The authors present a multidisciplinary approach for correction of a severe AOB deformity in a 19-year-old female patient with AI (**Dent. Med. Probl. 2004, 41, 2, 293–298**).

Key words: amelogenesis imperfecta, dental anomalies, skeletal open bite.

Streszczenie

Wrodzony niedorozwój szkliwa (*amelogenesis imperfecta hereditaria* – AI) w połączeniu z zaburzeniami zębowymi i gnatycznym typem zgryzu otwartego jest rzadką wadą, dlatego jest celowe publikowanie własnych obserwacji i wyników leczenia. Leczenie zgryzu otwartego jest trudne, a wyniki nie zawsze przewidywalne. Chirurgiczne leczenie zgryzu otwartego w przebiegu AI jest jednym z najtrudniejszych w chirurgii ortognatycznej. Możliwości leczenia ortodontycznego są ograniczone, dlatego jest konieczna multisegmentalna osteotomia szczęki typu Le Fort I lub osteotomie segmentalne w obrębie wyrostków zębodołowych z następową rehabilitacją protetyczną. Autorzy przedstawiają przypadek leczenia wielospecjalistycznego zgryzu otwartego u 19-letniej pacjentki z wrodzonym niedorozwojem szkliwa (**Dent. Med. Probl. 2004, 41, 2, 293–298**).

Słowa kluczowe: niedorozwój szkliwa, wady zębowe, kostny zgryz otwarty.

Amelogenesis imperfecta (AI) is a group of hereditary defects of the dental enamel not associated with any other generalized or systemic anomalies. The prevalance of this condition is 1 : 12–14000 [1] to 1 : 4000 [2], depending on the population studied. Various classifications have been proposed for the different AI types [2–3]. Generally, the AI is subdivided into three types. In the hypoplastic type, the enamel matrix is imperfectly formed with deficiencies in quantity but relatively well mineralized. In the hypomineralization type, the enamel is formed in relatively normal amounts but is poorly mineralized resulting in soft enamel. In the hypomaturation type, the final stages of the mineralization process are abnormal. This type is considered to be benign manifestation of the hypomineralization type. AI, a condition primarely affecting the enamel has been associated with other dental anomalies, including taurodontism, congenitally missing teeth and delayed eruption [4–7]. AI is frequently found in association with anterior open bite (AOB) [2, 8, 10]. The incidence of AOB in patients with AI varies from 24–35% [1, 9, 10]. The treatment of AOB deformities is known to be difficult and results are not always predictable [11–13]. In combination with AI, surgical correction of AOB is even more difficult and more challenging. Orthdontic options are limited and, therefore, a multisegment Le Fort I or alveolar segmental osteotomies are necessary, followed by prosthetic rehabilitation.

This article describes a combined multidisciplinary approach for correction of a severe AOB deformity in a 19-year-old female patient with AI.

Case Report

A 19-year-old female patient was referred to the Maxillofacial Clinic, Wroclaw Medical University for surgical correction of a severe skeletal AOB deformity. The patient related that her anomaly had already started when she was a small girl and had become gradually worse. She suffered since childhood because of her facial, dental and occlusal deformity. The patient's parents did not show enamel and skeletal alterations similar to those noted in the patient. The family history excluded similar features in two ascendant generations. Clinical examination of the patient showed a convex profile, incompetent lips, a mouth breathing pattern, a severe skeletal AOB and the increased anterior face height, steep mandibular plane and retracted jaws. The mentalis and orbicularis oris muscles were hyperactive when closure was attempted (Fig. 1A, C). There was a severe AOB that extended from the first molar of one side to the first molar on the opposite side of the arch. The measured interincisal distance was 10 mm. The mandibular dental arch showed a reverse occlusal curvature. There was a high palatal vault, an omega-shaped maxillary dental arch and a transverse discrepancy between it and the mandibular dental arch. The teeth showed surface irregularities, yellow discoloration, short clinical crowns, a tapered crown form with spacing in the anterior region but tight proximal contacts between adjacent teeth in the posterior regions (Fig.1E). The teeth were not sensitive to thermal stimuli. The cephalometric X-ray showed skeletal features characteristic of AOB with a steep mandibular plane, a steep mandibular occlusal plane, increased intermaxillary plane and an inreased anterior facial height (Fig. 2A). The pantomographic X-ray revealed taurodontism in molar teeth and failure of eruption of lower wisdom teeth (Fig. 3).

Based on characteristic skeletal and dentoalveolar features and cephalometric evaluation AI, was diagnosed.

The four-stage multidisciplinary treatment was applied:

I stage – presurgical orthodontics: an intraoral palatal splint with expansion screw was applied to expand the maxillary arch,

II stage – Schuchardt's procedure: posterior maxillary osteotomies reduced the AOB and allowed a forward and upward rotation of the mandible (Fig. 4A, B),

III stage – Köle's procedure: anterior mandibular segmental osteotomy closed the remaining AOB and reduced the excess anterior mandibular height (Fig. 4C, D),

IV stage – prosthetic rehabilitation: multiple crowns were made to stabilize the occlusion and to improve aesthetics.

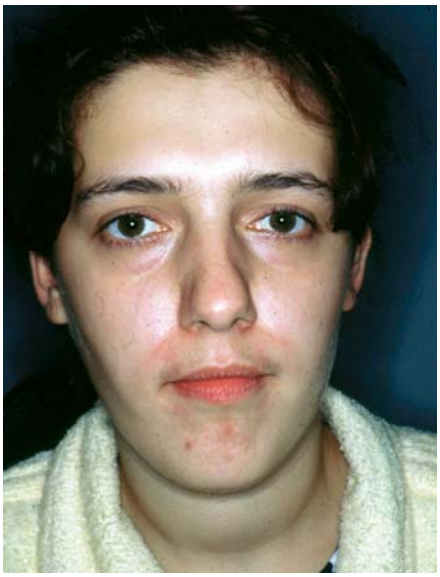
This patient who presented (with an) AOB deformity associated with AI was sucessfully treated with combined multidisciplinary approach. The photographs demonstrate the correction of the open bite and establishment of a normal Class I occlusion (Fig. 1B, D, F). Cephalometric evaluation revealed successful reduction of the anterior facial height and improvement of the soft tissue profile (Fig. 5A, B). The correction of the malocclusion was achieved with a notable improvement in the patient's self-esteem.

Discussion

Diagnosis of amelogenesis imperfecta hereditaria in the described patient was based on morphological characteristics for this disorder. The patient showed general enamel defects, as much in her decideous as in her permanent teeth, suggesting that this malformation did not occur due to agressor agents during a certain period of formation of the dental germ. Furthemore, the patient did not show any systemic disease which could cause general enamel hypoplasia, such as renal or endocrine disturbances involving calcium metabolism [14–15]. The teeth showed surface irregularities, yellow discoloration and short clinical crowns. The dental X-rays showed thin enamel and radiopacity

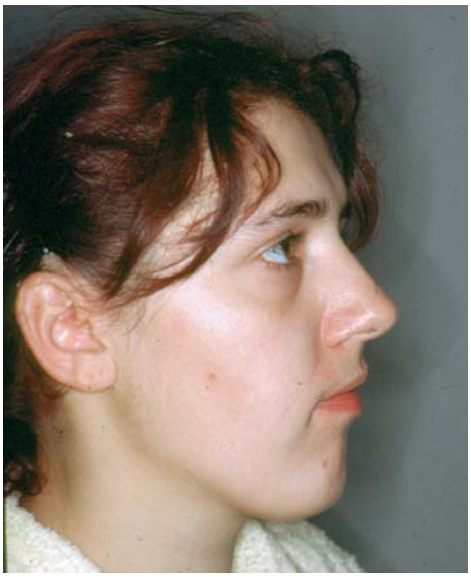
Fig. 1. A – preoperative en face view of a 19-year-old woman with AI and AOB, **B** – En face view 2 years after bimaxillary segmental osteotomies, **C** – preoperative profile view shows increased the lower anterior facial height and hyperactive the mentalis and orbicularis muscles, **D** – postoperative profile view – the lower anterior facial height is reduced, a harmonious profile is achieved, **E** – preoperative occlusal view with a severe AOB, **F** – occlusal view 2 years after surgery; multiple crowns were made to stabilize the occlusion and improve aesthetics

Ryc. 1. A – twarz en face 19-letniej kobiety z AI i zgryzem otwartym przed leczeniem, **B** – twarz en face 2 lata po dwuszczękowej segmentalnej osteotomii, **C** – profil pacjentki przed leczeniem – wydłużony dolny odcinek twarzy i hipertonia mięśnia okrężnego ust i bródkowego, **D** – profil pacjentki po leczeniu – skrócony dolny odcinek twarzy i harmonijny profil, **E** – warunki zgryzowe pacjentki przed leczeniem, **F** – warunki zgryzowe 2 lata po leczeniu chirurgicznym i rehabilitacji protetycznej



A

B



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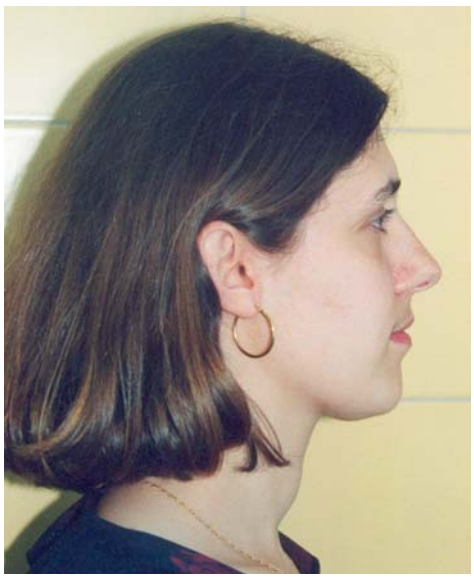

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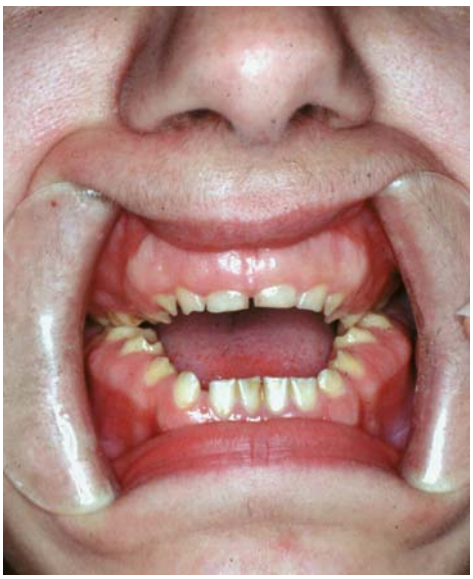
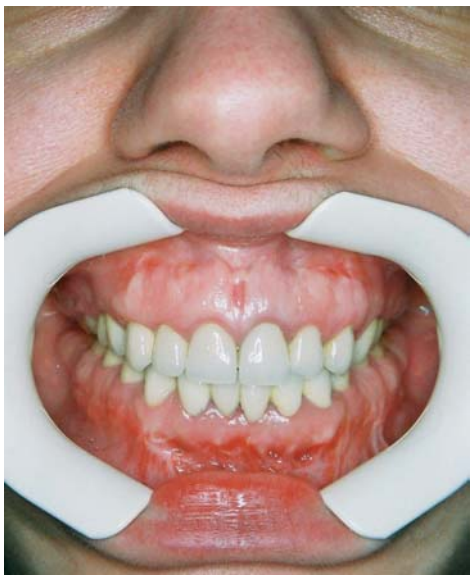


E



F

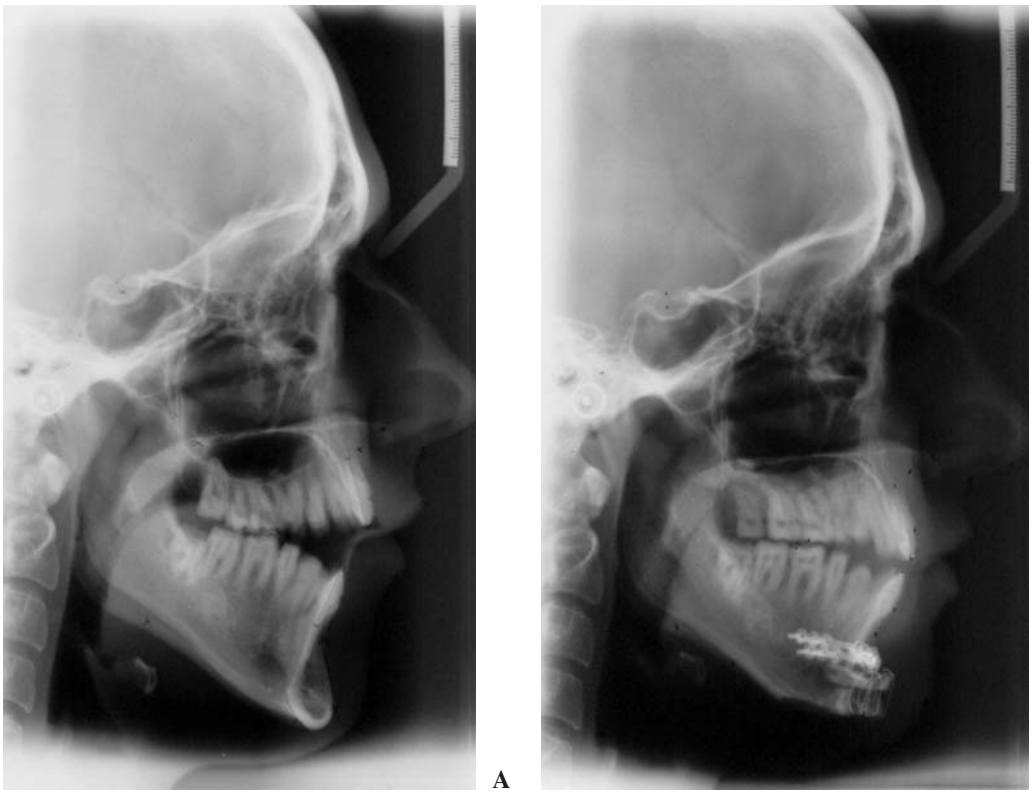


Fig. 2. A – preoperative cephalometric X-ray shows skeletal features of AOB with an increased anterior facial height, **B** – postoperative cephalometric X-ray reveals decreased intermaxillary plane and significant reduction of anterior facial height

Ryc. 2. A – zdjęcie telerentgenowskie głowy przed zabiegiem operacyjnym wykazuje cechy kostnego zgryzu otwartego i znaczne wydłużenie dolnego odcinka twarzy, **B** – zdjęcie telerentgenowskie głowy po leczeniu operacyjnym wykazuje zmniejszenie kąta podstaw i skrócenie dolnego odcinka twarzy

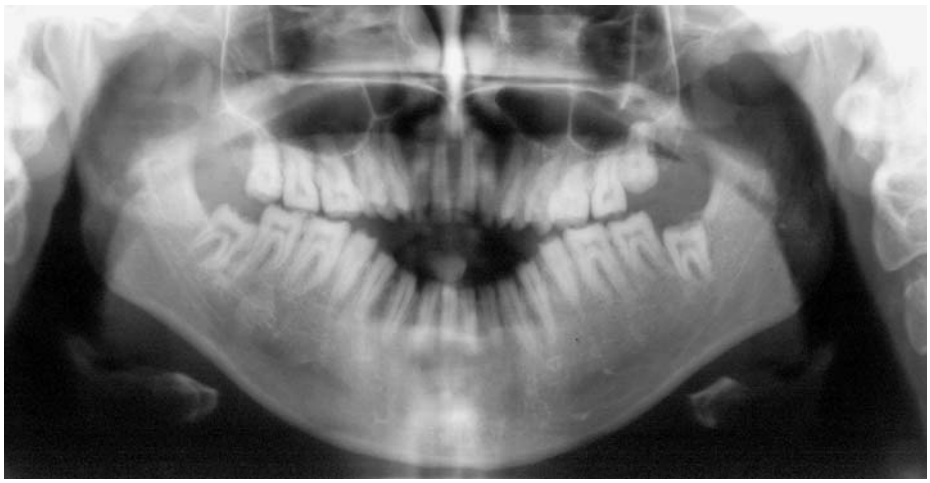


Fig. 3. Preoperative pantomographic X-ray shows taurodontism in molar teeth and failure of eruption of lower wisdom teeth

Ryc. 3. Zdjęcie pantomograficzne żuchwy przed leczeniem – widoczne poszerzenie komór miazgi zębów i kanałów korzeniowych zębów trzonowych oraz częściowo zatrzymane dolne zęby mądrości

similar to dentin. The panoramic radiograph revealed the relatively enlarged pulp chambers in molar teeth (taurodontism). Taurodontism, or elongation of the pulp chamber due to apical displacement of the root furcation, has been associated with AI in many studies [4, 14, 16].

AI is a group of hereditary conditions. Its genetic inheritance pattern can be autosomal dominant, autosomal recessive or X-linked [2, 5, 7, 15].

Although, the patient's family history excluded AI in two ascendant generations, the genetic origin of the disease can not be ruled out.

In studies made of the prevalence of AOB in patients with AI, it was noted that it occured in 24–35% of affected group compared with only 2% in the world population at large [1, 9, 10]. AI and AOB can be than considered to be intimately related. The coexistance of the two conditions can be

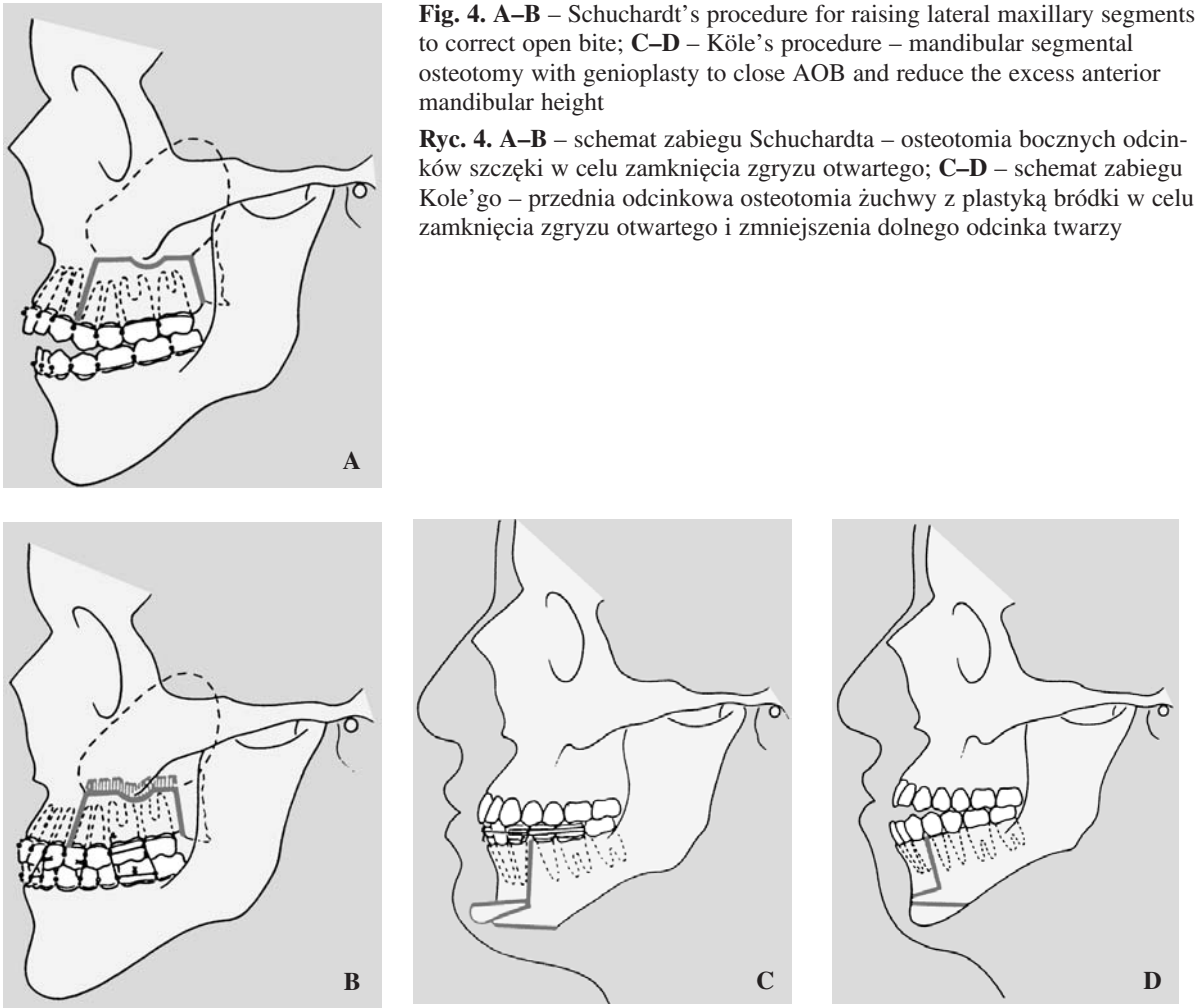
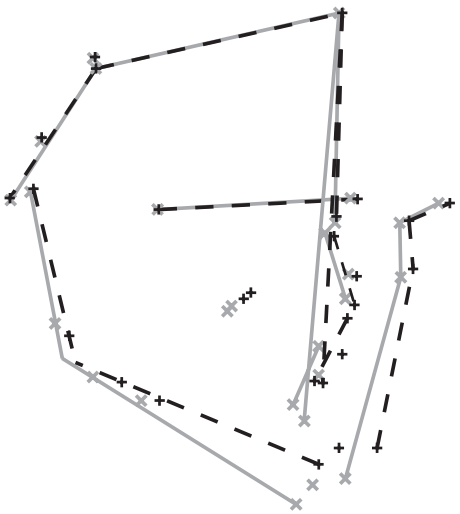
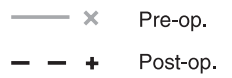


Fig. 4. A–B – Schuchardt's procedure for raising lateral maxillary segments to correct open bite; **C–D** – Köle's procedure – mandibular segmental osteotomy with genioplasty to close AOB and reduce the excess anterior mandibular height

Ryc. 4. A–B – schemat zabiegu Schuchardta – osteotomia bocznych odcinków szczęki w celu zamknięcia zgryzu otwartego; **C–D** – schemat zabiegu Kole'go – przednia odcinkowa osteotomia żuchwy z plastyką bródki w celu zamknięcia zgryzu otwartego i zmniejszenia dolnego odcinka twarzy





		Pre-op.	Post-op.
GntgoAr	122.0±7.0	132.7	126.4
ML-NSL	28.0±5.0	44.6	35.3
ML-NL	20.0±7.0	35.3	25.5
index	80.0±7.0	61.3	71.5

Fig. 5. Superimposition of pre- and postoperative cephalometric tracings. Pre- and postoperative cephalometric measurements used to evaluate skeletal and dental changes

Ryc. 5. Nałożone na siebie obrysy zdjęć telerentgenowskich z przed i po zabiegu z wybranymi pomiarami analizy cefalometrycznej

due to a pleiotropic action of the AI genes, influencing the growth of the craniofacial skeleton [9]. Witkop and Sauk [cit. according to 11] suggested that AOB was of a dentoalveolar nature, due to the patient inserting his tongue in a reaction to protect sensitive teeth, resulting in local interference that would prevent alveolar growth.

In treated patient, no indications were seen that the tongue was interposed to protect sensitive teeth. She and her mother did not mention any sucking habit or complaints of pain. This almost rules out the possibility of the AOB resulting from a local machanical interference.

This is in general agreement with the observations of Rowley [9] who has reported that in patients with AOB suffering from AI, tongue iner-

positioning is the result of vertical dysgnathia rather than its cause.

Treatment of AOB in patients with AI is known to be difficult and results are not always predictable [11–13]. Orhodontic treatment options are limited because of lack of crown height and the condition of the enamel. The unfavourable morphology of the affected teeth makes them innapropriate for removable and fixed orthodontic appliances. In the described case, we used an intraoral splint with activated screw to coordinate and expand the omega shaped maxillary arch before surgery. Correction of a severe AOB was accomplished in a two-stage surgical procedure. Posterior maxillary segmental osteotomies and anterior mandibular segmental osteotomy closed the open bite and significantly reduced the anterior facial height. Multisegment maxillary Le Fort I intrusion osteotomy or alveolar segmental osteotomies are recommended by many authors in treatment of AOB in AI patients [11–13, 15].

In our patient, a retention occlusal splint was used in the postoperative period, until final occlusion with crowns and bridges was performed one year after surgery. The multidisciplinary approach used to treat a severe AOB in the presented case gave the excellent aesthetic and functional results.

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