Dental Management of Dysostosis Cleidocranialis-Case Report

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Abstract

Cleidocranial dysplasia (CCD) is an autosomal dominant disorder characterized by skeletal anomalies such deformities, short stature of milder degree, abnormal shoulder mobility due to hypoplastic or aplastic clavicles. Cleidocranial dysplasia is characterized by anomalies of the thorax, spine, pelvis and extremities. Of particular interest to us as dentists are disturbances of the skull and tooth development. Case presentation: A 15 year-old girl with cleidocranial dysplasia (CCD) presented in this article, had typical features of CCD: delayed tooth eruption, underdeveloped upper jaw, especially premaxilla, high arched palate and a large number of excessive teeth. According to the patient's age and clinical findings, fixed bridges prosthesis made of Ceramill CAD/CAM Sintron in both arches was the treatment of choice. Successful treatment of the orofacial problems of patient with CCD requires the combined efforts of several dental specialists. The case reported proves the efficacy of surgical, orthodontic and prosthodontic procedures in resolving of clinical dentofacial features of CCD.

Key Words: Craniofacial abnormalities, Cleidocranial dysplasia, Tooth eruption

Introduction

Cleidocranial dysplasia (CCD) is a autosomal dominant disorder characterized by skeletal anomalies such deformities, short stature of milder degree, narrow and slumped shoulders, hypoplasia or aplasia of the clavicle, one side or both sides - these people can fully merge shoulders forward, hence the name "People of rubber ".

The location of the *RUNX2* gene codes for a core-binding transcription factor protein (CBFA1), which have been shown to cause CCD, was mapped to chromosome 21. It's expression is restricted to developing osteoblasts and a subset of chondrocytes [1]. This syndrome has long been known. Sometimes people with this syndrome have performed in circuses, calling them as "a man or woman of rubber".

The skull is broad (brahicefalija brahicefalija), with a pronounced parietal and frontal protrusions (protuberances). The base of the nose is broad nasal bridge is depressed. Change the jaws and teeth, underdeveloped upper jaw, especially premaxilla, high arched palate, pseudo progeny, persistence of deciduous teeth, delayed or absence of eruption of permanent teeth, and a large number of excessive teeth. Oral manifestation in the cleidocranial dysplasia patient includes multiple impacted permanent teeth, prolonged retention of primary teeth and multiple supernumerary teeth. The orthopantomograph of one patient with syndrome dysostosis cleidocranialis shows 52 teeth. The patients are with normal intelligence.

Only several cases of Cleidocranial dysostosis and syringomyelia have been reported in the literature [2]. Syringomyelia is the development of a fluid-filled cavity of syrinx within the spinal cord. The worldwide prevalence of CCD is generally regarded as being about one per million [3]. High frequency of CCD is noticed in Cape Town [3].

Lossdorfer et al. investigated the role of periodontal ligament cells (PDL) in delayed tooth eruption in patients with cleidocranial dysostosis (CCD) and concluded that PDL cells from CCD express a less distinctive osteoblastic phenotype resulting in an impaired ability to support osteoclastogenesis which might, in part, account for the delayed tooth eruption [4].

The early diagnosis of CCD is very difficult, because a majority of the craniofacial abnormalities becomes obvious only during adolescence. As a result of late diagnosis, occlusional and psychological problem may occur. Dysostosis cleidocranialis concerns both teeth and jaw and is characterized by supernumerary teeth, dentitio tarda, tooth impaction and eventually follicular cysts [5].

The purpose of this article is to show the surgical, orthodontic and prosthodontic management of the patient with CCD and to emphasize the importance of close consultative communication and cooperation between pedodontist, orthodontist, oral surgeon and prosthodontist (multidisciplinary approach).

Case Presentation

A 15 year-old girl (AS), Albanian nationality, live in the Gostivar city (*Figures 1-3*). Family history reveals that she has sister with the same syndrome. The initial extraoral examination revealed a retrognathic and vertically deficient maxilla, resulting in a concave facial profile. Orthopanthomogram investigations revealed the presents of excessive teeth, most of which are impacted. The patient has an elongated face, parietal and frontal bossing, a pointed beard, an antimongoidally placed eyes, a bulbous nose, small lips, and a short filtrum.

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Figure 1. *Frontal facial view.*

Figure 2. *Lateral facial view.*

The planning of dental treatment varies from individual to individual and mostly depends on the patient's need, the age at diagnosis and social and economic circumstances. At the beginning of the treatment the extraction of the 33, 32, 31, 42 and 45 were made. Also dental restoration from first class were put on 17, 27 teeth and mesio-ocluso-distal (MOD) restoration were made on 16, 46. On the 45 teeth from the low arch Helio dental restoration from second class was made.

Figure 3. *Frontal intraoral view.*

The surgical and orthodontic management of unerupted teeth

Due to the excessive teeth, they must be extracted in order to provide a time and place for the emergence of impacted teeth. They often must be surgically treated to emerge in the mouth. Of course, this extraction should be done several times, removing teeth in series, in order maximum to preserve bone to become weak and deficient. They can use mobile devices to which may be added teeth to the eruption of permanent teeth, because the emergence of these patients is difficult and rather late. Alignment of teeth and the leveling of the dental arches are made with fixed appliances. Heavy upper and lower wires were placed. Often, at the end of orthodontic treatment should be made prosthetic replacements. All supernumerary teeth from the jaws were also surgically removed. After the permanent tooth eruption had been accomplished, orthodontic treatment of these teeth was required.

Orthodontic treatment was initiated with the following objectives: maxillary expansion, removal of the supernumerary teeth, guided eruption or extrusion of the impacted permanent teeth. A hyrax expander was placed for 2 months, which achieved 10 mm of expansion.

Impacted permanent teeth 13 and 23 were surgically exposed and bonded with wire ligature placed close to the cusp tips for guided eruption.

Apparatus for rapid extension of a suture mediana

The most widely used apparatus for rapid maxillary extension is Biederman's, and rarely Derishsweiler's apparatus. Their main goal is rapid spread of the maxillary dental arch in the area of the suture median. It is used in underdeveloped maxilla, difficulty breathing and lateral cross-bite. Historically, the appliances for Rapid Maxillary Extension come from the long ago time when Faushard, Baurdet and Fogg were professionally active [6]. However, more recently Derishsweiler and Timms deserve the development of this

device. Biederman's apparatus is composed of four rings (two of the first premolars and two of the first permanent molars) and a screw that is wrapped on a special wired construction (hirax screw) (*Figure 4*).

Figure 4. *RME rapid maxillary extension.*

First of all the rings are attached on the teeth in the mouth of the patient, an impression with the rings on it and after that the gypsous model is pouring out. On the working model the screw is adapted and attached on the oral side of the rings. Finally the apparatus is processed and polished. Derishsweiler's apparatus consists of four rings (two of the first premolars and two of the first permanent molars), a transverse extension screw and an acrylate plate extending from the mesial side of the first premolars to the distal side of the first permanent molars.

The rings are attached on the teeth in the mouth of the patient, an impression is taken and a working model is poured out. First, on the working model the foil is placed where the acrylic plate will be made. In the middle of the acrylic plate the screw is embedded. Than the apparatus is processed and polished. The activation of both appliances is done by opening the screw 2 to 3 times a day. In a short period of time, extension of the sutura mediana and separation between the central incisors occur. Delair's mask is an orthodontic device that is designed to act in an antero-posterior direction, which it achieves via a facebow or J hooks to the patients dental braces or palatal expander. This therapeutic method tends to correct more severe bite problems which occurred during the growth and development of the maxilla and mandible. The appliance is applied in primary and mixed dentition.

Working way of Delaire's mask

In the maxilla it emphasizes the growth of its front part. It acts on the premaxilla and the frontal-maxillary process. This mask on the mandible acts in such a way as to cause modification on the direction and development of the condylar cartilage. Delaire's mask on the dento-alveolar crest acts causing mesial displacement of the upper alveolar arch and the base of the maxilla, in the same time causing distal shift of the lower alveolar arch and the remodeling of the bone part of the chin.

Figure 5. *Delaire-mask.*

Delaire's mask simultaneously acts on the maxilla, mandible, masticatory musculature and tongue. The basic mechanism of action of this mask is: extracting the maxilla as a "drawer" in the "front-naso-maxillary joint", indentation of the mandible (stopping the anterior growth of the mandible), the use of the force of the masticatory muscles (focusing on the action of the masticatory muscles in the direction of the action of the mask's forces) and the use of the tongue's strength, i.e. placing the tongue up and forth. Such a placement of the tongue forwardly pulling forces on upper frontal teeth and providing a normal vertical and horizontal overlap. The Delaire's mask is constructed from an extraoral part (mask), an intraoral part (dual intraoral arch) and elastic strips that connect these two parts (*Figure 5*).

The external part of the mask is custom-made and is consist of frontal (forehead) part, chin part and wire structure. The frontal part (forehead) is placed in the forehead region, and the forehead (cap) of the chin. These two parts are made of plastic mass, and the inner part is coated with a skin or canvas that does not cause allergy in the patient.

The wiring structure is composed of two vertical metal lateral rods with a thickness of 2 mm. The lateral woven elements are merged with each other with a horizontal "pre-labial wire" which is located 1 to 1.5 cm below the labial commissure and in which two hooks are wrapped, with a wire thickness of 0.7 mm, for each side, which serve to lay the elastic tires for traction.

The external part of the mask, forehead and chin part, is easily adapted to the patient's face by warming in hot water.

There are metal openings on the frontal and chin plastic part on each side for placing rigid wires that are modified in width and length according to the face of the patient. When fully adapted to the forehead, the chin and the wired part of the mask are fixed with an anchor key.

The intraoral double wired construction consists of two wires (vestibular and palatinal) that are merged in the first permanent molars or the second molar molars.

The process of making an intraoral double bow

At first four rings are adapted on the first permanent molar or second molar molars. An impression from the upper jaw is taken with the alginate mass, leaving rings in it. The rings remaining in the alginate mass are isolated from the inside with a wax, which will later enable us to extract the rings from the model more easily.

Initially on the molded model the palatinal part of the double-wired construction is manually made, which accurately adapts to the necks on the oral side of all teeth (never above the cingulum), which gives stability to the apparatus. The palatinal wire must not be twisted in a zigzag, with the exception only in those cases where we have tooth dystopia; the wire is adapted to the palatinal necks of teeth with dystopia.

The vestibular wire of the double-wired construction must not touch the vestibular surfaces of the teeth. The wire follows the tooth-line in the neck region of the teeth in a weak arc and needs to stand 1.5 to 2 mm from the incisive surface. This deviation of the arc, avoid close contact between the teeth from the arc, on that way allows the natural extension of the tooth to the front.

On the vestibular arc between the lateral incisions and canines' teeth on both sides there is a hook that is distally directed, or the wire itself is twisted in the form of a loop, which are used for placing elastic tricks.

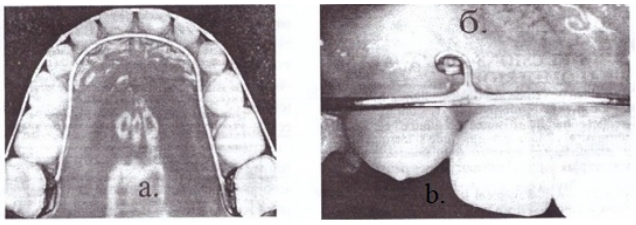

Figure 6. *(a) Double-wired construction in the patient's mouth, (b) Vestibular wire as part of the double-wired construction.*

The double-wired construction (arc) is made from a wire with a thickness of 1 to 1.1 mm, while the hooks are made from the wire with the thickness of 0.7 mm. The two wires, each on its own side, which are merged with rings on the teeth and polished. After that the double-wired construction is tested and cemented in the patient's mouth (*Figure 6*).

The orthodontic elastics for traction represent "motor force" for the antero-posterior traction system. The orthodontic elastics are placed on the hooks of the vestibular wire and the hooks of the pre-labial extra-oral wire from the face mask. At the beginning, weaker forces are used (from 600 to 800 g), and after 3-4 weeks greater forces from 1500 g are used (*Figure 7*).

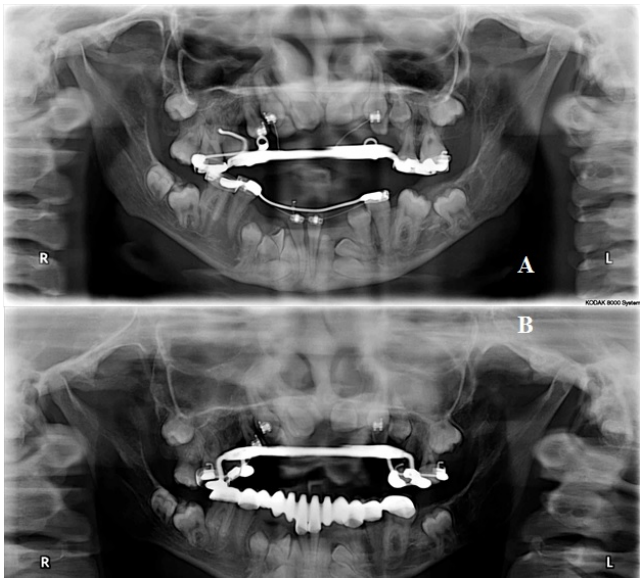

Figure 7. *(A) Orthopantomogram made after initial orthodontic therapy, (B) Orthopantomogram made after placing the first lower bridge.*

The mask is worn 12 hours a day, at least 6 months. After wearing Delaire's mask, a functional retention device is sets in the patient mouth.

Prosthodontic treatment

Standard impressions of the oral situation of the patient were taken with light body polyvinyl siloxane impression material (Low viscosity C - Silicone Oranwash L, Zhermack, Italy). Preparation of the patient for the impression in the upper jaw was shown on *Figure 8.*

Figure 8. *Preparation of the patient for the impression in the upper jaw.*

In the dental technique lab PROMEDIKA, the preparations and scans of the upper and lower models were done. The working models were digitized along with the abutments, the models of the lower and upper jaw and the bite (*Figure 9*).

Figure 9. *Working gypsum model.*

Finally, the abutments were scanned individually, one by one because the shoulders of the abutments were located subgingivally and could not be accurately covered only by the model scans.

CAD construction

First, the position of the digitized model was defined by design software according to the insertion direction of the abutments. In the second step, the abutment's shoulder lines were marked and the thickness of the interspaced was defined. The marginal line is the "margin of the preparation" of the restoration. In our case we set the interspaced of 0.2 mm and the space for cement of 0.4 mm. The thickness of the marginal border was set at 0.1 mm. According to our experience, these settings result in excellent accuracy of the restoration fitting on the model and eliminating the need for subsequent corrections and adjustments in the patient's mouth. In the end, the design of the structure was once again tested against the individually designed parameters. If the wall thickness is less than the acceptable minimum, the software sends a warning, allowing automatic correction. The full contours of the teeth from 13 to 23 were reduced by 0.9 mm from the vestibular side to make space for the veneers.

Milling

The complete CAD design divides the basic structures of crowns on 18.000 to 20.000 coordinates and generates harmonious surface texture and excellent marginal sealing. The complete design was transferred to the CAM device. CAM software uses the information of the specified interspaced and margins of restoration to calculate milling's parameters that make difference between different areas of restoration. For example, when the process of milling the margins of restoration id going on, the device reduces the speed, as well as the filling rate, thus contributing for prevention of the crown's margins from breaking or gaining cracks. As a result, even cervical margins that are very thin and have a 0.1 mm thickness may be safely cut and require only a little processing after the sintering process. In less sensitive areas, the device uses a higher cutting speed. Once the output format is entered, a milling strategy is selected. In this case we used a milling strategy using 25 mm, 10 mm and 0.7 mm cutters for bridge construction. A structure that supports sintering has been designed and which allows the restoration to be sintered in an upright position with a sintering furnace. The sintering frame minimizes the distortion which may occur during the process of sintering and is an instrument for achieving high precision in the passage of large restorations.

Figure 10. *The circular bridge with its metal construction on a gypsum model.*

Figure 11. *Testing of the metal part of the upper bridge in the mouth of the patient.*

The absolute precision with which the milling machine works is apparent through the excellent cutting results obtained in the occlusal and palatinal surfaces as well as in the incisal edge of the restoration. The circular bridge with its metal construction on a gypsum model was shown on *Figure 10*, while testing of the metal part of the upper bridge in the mouth of the patient was shown on the *Figure 11*. The completed bridge with harmonic color effect and homogeneous surface texture was shown on *Figure 12*.

Figure 12. *The completed bridge with harmonic color effect and homogeneous surface texture.*

Figure 13. *The cemented bridges meet their beautiful natural appearance and fulfill the patient's functional and aesthetic expectations.*

The cemented bridges which meet their beautiful natural appearance and fulfill the patient's functional and aesthetic expectations are shown on the *Figure 13*.

Discussion

Surgical, orthodontic and prosthodontic management of the orofacial manifestations of CCD is lengthy and challenging. Careful planning and execution by an interdisciplinary team is required in the dental treatment of patients with CCD. The length of treatment ranges widely, in our cases it last five years, while in the literature cases that lasted 7 years and more are described [7]. Generally the possibility of treatment exists until growth has ended. In the treatment of these patients it is very important to maintain the patients' motivation and keep the treatment within the patient’s tolerance.

Hardy reports a case of bilateral posterior glenohumeral dislocation in male 31 year patient with cleidocranial dysplasia [8]. Feldman stated that abnormal dentition which occurs in cleidocranial dysplasia patient may result in dental caries. Although this disorder is benign, if dental caries is left untreated, it may progress to osteomyelitis of the maxilla or mandible [9].

The four main therapeutic approaches published in the literature are the Toronto-Melbourne, Belfast-Hamburg, Jerusalem and Bronx methods. Toronto-Melbourne approach involves serial extraction of deciduous teeth and depends on the development stage of the roots of the permanent teeth. Procedure is performed under general anesthesia and supernumerary teeth are also removed with the aim to facilitate the eruption of the unerupted permanent teeth [3].

Belfast-Humburg approach involves surgical procedure under general anesthesia to extract all retained deciduous and supernumerary teeth. After healing of the surgical sites, orthodontic appliances are placed on fully erupted teeth with traction bands attached to partially erupted teeth to facilitate their further eruption. The advantage of this procedure is single surgical operation under general anesthesia [10].

Jerusalem approach is based on 2 surgical interventions. During the first procedure anterior deciduous teeth and supernumerary teeth are extracted and orthodontic brackets and traction elastics are applied on the permanent anterior exposed teeth. After that surgical flaps are closed. Second surgical intervention take place at approximately 13 years of age in which the residual deciduous teeth are extracted, unerupted canines and premolars are exposed and obvious orthodontic and surgical processes are completed [3].

The Bronx approach involved extraction of all the retained primary and supernumerary teeth followed by an observation period and a second surgical phase of exposing and bonding impacted permanent teeth. In our case after orthodontic alignment and coordination, the treatment was completed with the restorative phase using fixed metal ceramics prostheses.

Recent new introduce technologies in dentistry have yielded better therapeutic options and outcomes for individuals with CCD. Dental implants can be successfully used for replacing teeth that cannot be guided into the arch orthodontically. Also, orthodontic tooth movement with reduced side effects can be made by micro-implants.

The cone-beam computed tomography (CBCT) as a new technology is now routinely used to provide a three-dimensional image of the dentition and ensuring better anatomical localization of supernumerary and impacted teeth. CBCT can provide pertinent precise information about localization of a supernumerary tooth in relation to important structures such as the cortex of the nasal floor, labial cortex of the nasal ridge, nasopalatine duct and adjacent root apices [11].

Conclusion

The treatment of dental anomalies in the dysostosis cleidocranialis patient should start at the time of normally expected deciduous tooth exfoliation.

Management of the patient must be adapted to the patients' orthopedic and dental needs. With the further developments of new technologies the management of complex dental cases will continue to be improved.

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