

Case Report

PRESURGICAL ORTHODONTIC THERAPY IN A NEWBORN WITH MEDIAN CLEFT LIP ACCOMPANIED BY HOLOPROSENCEPHALY AND AGENESIS OF THE PREMAXILLA

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Abstract. *Surgical treatment protocols for rare types of clefts do not exist, especially when they are associated with severe anomalies of the brain and face. Surgical treatment of a newborn with holoprosencephaly, premaxillary agenesis and median cleft lip is a big challenge for the surgeon. The complete lack of bone in the nasal bones and premaxilla make surgical therapy almost impossible. The role of pre-surgical orthodontic therapy plays a major role in creating the conditions for surgical repair of cleft-damaged parts of the face. Case presentation. The paper presents presurgical orthodontic therapy in a newborn with a median cleft lip using the RBJ stimulator. Conclusion. The use of stimulators of a specific construction in the therapy of rare clefts is the method of choice.*

Key words: *presurgical orthodontic therapy, newborn, holoprosencephaly, median cleft lip, complete lack of premaxilla*

Introduction

Children born with a complete cleft of the lips and palate have severe health problems. However, children born with holoprosencephaly (HCP), premaxillary agenesis, and a median cleft lip have specific and extremely difficult medical problems. In addition to the facial malformation, children with this anomaly develop mental retardation, ocular and olfactory abnormalities, endocrine and neuromuscular diseases, epileptic seizures, etc [1]. Their lifespan is not long, and depending on the severity of the anomaly, they live no longer than early childhood.

At the very beginning of their lives they struggle for survival, because feeding by natural means is almost impossible due to the extensive communication between the nasal and oral cavity, caused by the median cleft of the lip and premaxillary agenesis. Due to the inability to create the negative pressure required for milk suckling, attempts to take milk result in difficulty in swallowing. There occur nasal regurgitation, excessive air swallowing and milk aspiration, which cause very dramatic situations. The choking is followed by coughing, aspiration of milk into the lungs and cyanosis [2]. This phenomenon endangers the life of the newborn and stressfully affects the parents.

In addition to the existential problem, the newborns with this diagnosis must face serious and severe surgical interventions of the lips, palate and nose. The surgery of

the structures that form a large part of the face are particularly affected by the inborn absence of bone tissue which forms the nose, a large part of the alveolar ridge and the palate (absence of crista galli, the nasal bone, the complete premaxilla, and nasal septum), as well as by the lack of soft tissue, which plays a role in the formation of the tip of the nose, the philtrum, the tuberculum, the central red part of the lips, the frenulum, etc. Due to the defects of these tissues, the success of the surgery is questionable. The tension of the sutures often leads to dehiscence.

This paper presents the fabrication of the RBJ* stimulator and its successful application in feeding a newborn, as well as its role in presurgical period in children with HCP, premaxillary agenesis and median cleft lip.

HCP is a congenital anomaly of the prosencephaly associated with the median facial defects. It occurs between third and fourth week of gestation period and is caused by unsuccessful or incomplete division of the forebrain. According to DeMyer [3], holoprosencephaly is traditionally classified into three grades of severity: alobar, semilobar and lobar. In addition to these classic forms, a subtype of HCP, the middle interhemispheric variant or syntelencephaly has been characterized [4, 5]. Septopreopticholoprosencephaly is a mild subtype associated with midline craniofacial anomalies [6].

HCP is accompanied by a scope of distinctive craniofacial anomalies. The degree of facial deformity most

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often correlates with the degree of severity of the malformation of the brain [7].

Premaxillary agenesis with median cleft lip is a complete median lip cleft, and when associated with hypotelorism it is the fourth type of facial anomaly within HCP.

Case Report

A newborn diagnosed with HCP, premaxillary agenesis and median cleft lip was brought to the University Clinic for Dentistry in Nis, which was confirmed by the ultrasound of the brain, magnetic resonance of endocranium and three-dimensional computed tomography (3-D CT) of the craniofacial skeleton. According to the clinical examination, premaxillary agenesis was indicated by hypotelorism, flat nose, and median lip and palate cleft (Fig. 1). The complete lack of prolabium, premaxillary bone, cartilaginous nasal septum and columella was also visible, and the nasal bone was hypoplastic.

The child was primarily brought due to the inability to perform the feeding. The extensive communication between nasal and oral cavity indicated that feeding was possible only by nasogastric tubes. A RBJ obturator (Radojicic Bozidar and Julija – the authors of the device) was fabricated for these reasons.

The first phase in the construction of the RBJ obturator is taking the anatomical impression of the upper jaw (Fig. 2A). Impression taking in a newborn with HCP is very specific. Bearing in mind that the newborn completely lacks the primary palate, it is necessary to take the impression of the palatal extensions which are widely separated and form the secondary palate. Impression taking is largely based on the skills and experience of the orthodontist and the process excludes general anesthesia for the newborns. The premedication given to reduce stress in infants could provoke prolonged sedation in the post-anesthetic period and some functional disorders such as eating and sleeping problems even 15 days after the intervention. In addition, endotracheal intubation has

proved to be difficult in a certain number of such cases (4–7% infants), and occupies a large part of the space within the oral cavity, which makes operating difficult during the impression taking, thus affecting its precision.

Irreversible impression materials are used for taking a preliminary impression. The preliminary impression is anatomical and still imprecise, and it only gives rough outlines of the palatal extensions. Its only role is to be the basis for creating an individual spoon, which will be helpful in taking the precise impression. The secondary (corrective or functional) impression is a crucial step in the process of constructing a RBJ stimulator. It must include the finest details of a child's alveolar ridges (creases, frenulum). Additional silicones from Group A (Fig. 2B) are used for this purpose. They are distinguished by high accuracy, long lasting dimensional stability, hydro-compatibility and liquidity under pressure.

There are certain differences in the procedure in comparison to the standard impression taking, with the goal to reduce the time necessary for impression taking to minimum. Reducing the binding time of the impression material is achieved by doubling the amount of activator from the one used in standard procedures. However, one should be careful, because if the amount of the activator is excessive, it may have the opposite effect and result in not binding of the impression material at all, risking the aspiration of the impression material. Therefore, special attention should be paid to this phase of impression taking. The prepared materials are inserted into the mouth of the newborn via the anatomical impression. It is essential that the process of binding of the prepared material with addition silicones has begun immediately before the insertion (Figs. 2B, 2C). The binding process lasts 40 seconds, as the child's body temperature accelerates the binding process (Fig. 2D). After inserting the impression material, this semi-liquid material seeks its way to the free areas, and as a big part of a bone is missing due to premaxillary agenesis, there is therefore no bone support that would stop the flow of the material in the upper part of the nasal cavity. At one moment it cannot be controlled



Fig. 1 A: View of the entire face immediately after birth. B: Intraoral details.



Fig. 2 A: Taking the anatomical impression of the upper jaw. B: The preliminary impression and additional silicones from Group A. C, D: Taking the corrective impression of the upper jaw. E: The corrective impression.

any more, and we rely only on the process of material binding and its extraction in one piece after the binding.

It is important to continually pull down the child's cheeks with the left hand in order for the movable immobile mucous membrane, which is the vestibular boundary of the stimulator, to be impressed precisely. Any extension of the acrylate of the future obturator above or below this boundary destabilizes the apparatus. If it goes beyond this limit, any movement in the child's mouth will disrupt the apparatus, because the movable mucous membrane will tense, and if it is below the boundary of the

mucous membrane, it will not adhere in the mouth even while steady. Thus, the fabrication is similar to the principles of making of a total dental prosthesis in elderly people with edentate alveolar ridge.

Precise impressions, regardless of their discontinuity, clearly show all other anatomic details (surfaces) typical for this stage of the child's development, and can serve as orthodontic retainers, or quite the opposite, destabilize the stimulator (Figs. 2E, 4A).

After the corrective impression has been precisely taken, a plastic model is developed in the laboratory. The



Fig. 3 A: Plastic model of the upper jaw and RBJ obturator. B, C: A skillfully taken impression enables the retention of the appliance within the mouth of the newborn, without any extraoral fixation, which is one of the advantages of this RBJ obturator. D: Feeding of the newborn with the RBJ obturator.

plastic model must contain the smallest details typical for this period of the newborn's upper jaw development (Fig. 3A). Alveolar ridges are not fully developed and we call them alveolar margins. The maxillary alveolar ridge is horseshoe-shaped and has constant configuration. It develops into two parts: the external or labio-buccal and the interior or lingual part. The labio-buccal portion is diffused first and grows more rapidly. This fact should be kept in mind when constructing the obturator in order to allow for the growth of the alveolar ridge. For this reason, this side of the plaster model is covered with a thin layer of wax before filling the cold polymerizing acrylate. The thickness of the applied wax will correspond to the size of the tissue growth for this part of the alveolar edge. In the next phase, the gap that is clearly visible on the plastic model is filled with wax. This gap results from discontinuity in the fusion between the two palatal shelves. Using the working cast prepared in this manner, the RBJ obturator is made of polymerizing acrylates (Figs. 3A, 3B, 3C). The obturator must be free of sharp edges, well-polished, thin and glossy.

The RBJ obturator closes the communication between the two cavities and enables the creation of the negative pressure necessary for milk suckling and undisturbed feeding (Fig. 3D).

The fabrication of the RBJ stimulator begins after a week. It has an active impact and is the start of the pre-surgical preparation of the newborn. The stimulator is made on the same principle as the obturator. The accuracy of the impression is now at a higher level, since the impression is taken via the obturator, which now functions as the individual spoon (Fig. 4A). The borders of the future RBJ stimulator are clearly visible on the impression: movable immobile mucous membrane (the height of the stimulant), posterior border determined by the most distal line of palatal extensions, lateral sulci and interdental furrows (Fig. 4A).

The primal role of the RBJ stimulator is to close the communication between the nasal and oral cavity, to bring the split palatal segments closer and direct their growth in the direction of reducing the width of the cleft, which would ultimately lead to the improvement of surgical reconstruction (Figs. 4B, 4C).

The RBJ stimulator owes its effects to an open orthodontic screw positioned in its framework. The open screw is placed on the sagittally cut acrylic plate of the stimulator. Before each activation of the screw, it is necessary to remove a part of the acrylate on the sagittal plane for the exact size of the space which will be closed again by turning the screw. In this way, there is always the continuity of the acry-

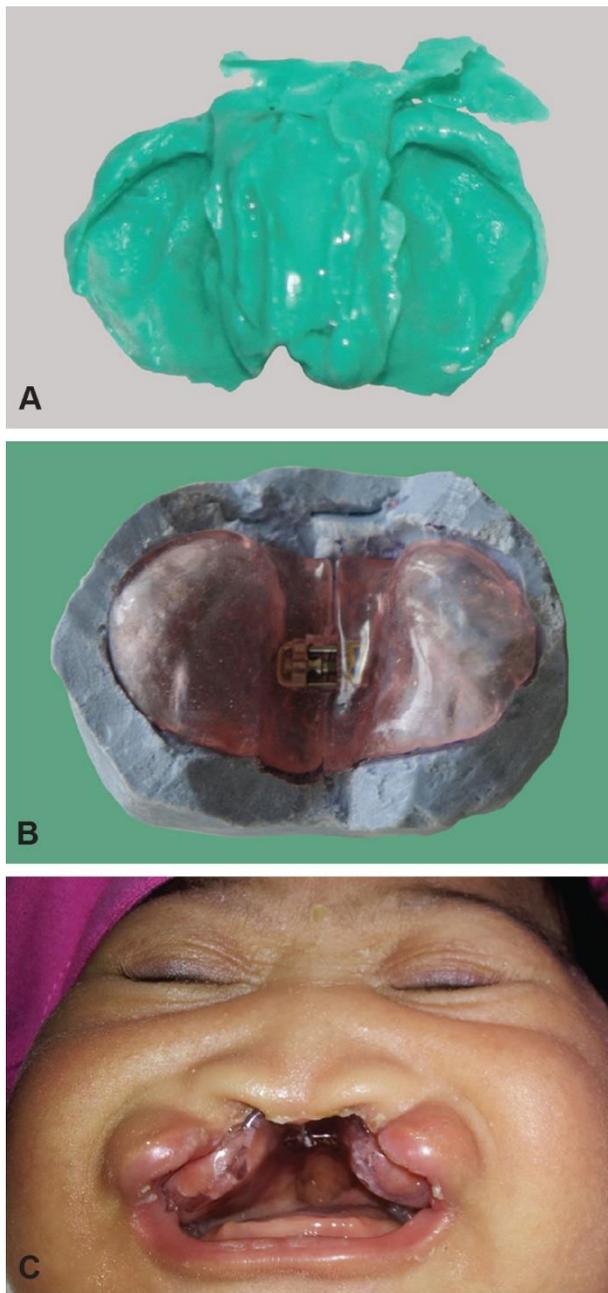


Fig. 4 A: Corrective impression. B: Plastic model of the upper jaw and RBJ stimulator. C: RBJ stimulator into the mouth.

late plate, which is necessary for the stimulator to create the negative pressure required for feeding. The palatal segments move toward each other by activating the screw, thus reducing the cleft (Figs. 5A, 5B).

The surface of the initial gypsum impression taken with the newborn immediately after birth, as well as the gypsum impression of the baby after the completed early orthodontic treatment through the RBJ stimulator were digitized, reconstructed and computerized by CEREC and LAB 3D (System Scanner BLUE CAM, Sirona

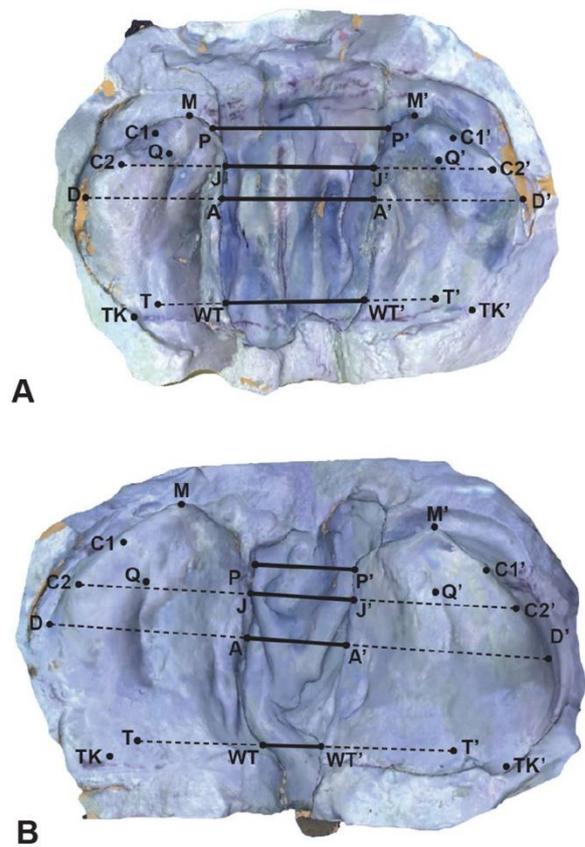


Fig. 5 The reduction of the cleft width which is achieved after carrying the RBJ stimulator from day 7th to the end of the 6th month of the newborn's upper jaw and HCP. A, B: 3D digitized models, landmarks and measured distances.

Dental Systems Germany). The analysis of the model of the damaged upper jaw of the baby with HCP, the premaxillary agenesis and the median cleft lip were performed with the help of the determined anatomic and constructed points and dimensions (Table 1, Figs. 5A, 5B).

In addition to reducing the cleft, the 3D analysis of digitized upper jaw models by using the software (CEREC in LAB 3D Scanner of the system BLUE CAM, Sirona Dental Systems Germany) has shown that the stimulator exhibits other advantages, such as correcting palatal extensions from a steep to a more horizontal position, and growth of the damaged jaw in transversal and vertical directions (Table 1, Figs. 5A, 5B).

In order to avoid the discontinuity in the transverse development of the maxilla, acrylates are simultaneously removed from the side of the stimulator which is in contact with the vestibular side of the alveolar ridge.

Ethical approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Table 1 Landmarks and measured distances on maxillary cast measured in the three-dimensional cast analysis of a newborn with holoprosencephaly, premaxillary agenesis and median cleft lip

Landmark	Point	Location		
<i>Maxillary cast</i>				
M/M'	The most anterior point	The most anterior point of the crest of the alveolar ridge [8].		
P/P'	Cleft edge point	Cleft edges of the ridges [9].		
C1/C1'	First canine point	Point at which the anterolateral sulcus crosses the crest of the ridge [9].		
C2/C2'	Second canine point	Point at which the lateral sulcus crosses the crest of the ridge [9].		
Q/Q'	Gingival groove point	Intersection of the gingival groove and lateral sulcus [10].		
TK/TK'	Tuberosity point	Junction of the crest of the alveolar ridge with the outline of the tuberosity		
T/T'	Base point	Posterior shelf pit (foveola palatina) [9, 11].		
WT/WT'	Cleft edge point	Perpendicular to the course of the alveolar crest on the level of T and T', respectively [9].		
<i>Constructed landmarks</i>				
J/J'	Author's landmark	C2-C2' cross-section point and edge of the palatine process		
A/A'	Author's landmark	D-D' cross-section point and edge of the palatine process		
D/D'		A point on the alveolar shelf on which the parallel of the line connecting the points M and T becomes the tangent		
Distances measured	Reference point		Before therapy	After therapy
P-P' (mm)	Anterior cleft width		18.30	10.51
J-J' (mm)	Middle cleft width on the level of C2 and C2'		15.51	10.88
A-A' (mm)	Middle cleft width on the level of D and D'		15.84	10.42
WT-WT' (mm)	Posterior cleft width on the level of T and T'		14.56	6.47

Discussion

HPE, premaxillary agenesis and median cleft lip are rare anomalies occurring in 1 among 8000 liveborn, healthy children [12]. Therefore, there is no standardized protocol for the management of varying degrees of severity of craniofacial malformations. The treatment is multidisciplinary, and the need to improve the quality of life is more than necessary [13]. The surgery of facial malformations is one of the most prevalent problems, largely due to the lack of bone and soft tissue of a large part of the face.

The success of surgery of the median cleft lip is mostly influenced by two factors: the degree of soft tissue hypoplasia and the width of the cleft. Presurgical orthopedics plays an important role in reducing the width of the cleft.

The use of RBJ stimulators in presurgical preparation has shown its positive effects. In addition to the significant reduction in the width of the cleft, which is the ultimate goal of using this apparatus, it should be emphasized that it is an apparatus without extraoral fixation. Extraoral fixation is not necessary because the retention of the apparatus is excellent. Only the apparatus which is perfectly positioned in the mouth and intimately attached to the mucosa of the palatal extensions can have an active effect, in this case directed to cleft reduction. Any clucking or moving in the mouth indicates poor retention and an absence of an active effect. If the firm acrylic connection of the apparatus does not include the

vestibular sides of the palatal extensions to the movable immobile mucous membrane, there will be no reduction in the cleft. Excellent retention of the apparatus is achieved only by immaculate impression taking, which is described in this paper. We consider that the retention of the RBJ apparatus is more comfortable for the patient, as opposed to the different ways of apparatus retaining, which are described in the literature as: by placing two bone screws in the mid-facial area (titanium 2.0x8 mm through the palatal holes) [14].

Orthodontic presurgical therapy with RBJ apparatus is based on biological principles and the aspect of individuality. In the course of its construction, we were guided by the fact that every person with a cleft is bearing in themselves an inherent growth [15]. The knowledge of growth and development in people with clefts is a key factor for successful orthodontic therapy.

The therapy starts immediately after birth. Not only is the undisturbed feeding enabled and the serious existential problem with the newborn solved, but it also has advantages from an orthodontic perspective. The early onset of therapy is a guarantee of its success, because the palatal segments, as well as all sutures (junctures), become more rigid after months 8-10 [16]. Unlike the previously described apparatuses that are inserted at about three months of age when the reparation of the lips is performed and when they serve only to prevent the damage to the surgical site by premature removal of the sutures [17], the RBJ stimulator significantly reduces the width of the cleft to the very

moment of surgery. In addition, when inserted during the operation, it also participates in the prevention of violation of the suture integrity, as it prevents the interposition of the tongue, as stated by Acharia et al. [14] Furthermore, the early appliance of the RBJ apparatus as well as its constant carrying until the surgery has affected the tongue position and has brought the tongue position to the level of the lower jaw to a certain extent.

In addition to anomalies, the newborn also had many other health problems (epileptic seizures) that involved daily use of anti-epileptic drugs. In order for the organism of the feeble infant to be able to tolerate the shocking doses of drugs, it was necessary to create conditions for the adequate intake of the necessary nutrients and thus eliminate the possibility of getting the newborn into a state of exhaustion. For these reasons, it was necessary to solve the functional problem (swallowing in the neonatal period), and establish the proper mastication along with the further development of the child and the eruption of the first teeth. The intact palate and closing the communication with the nasal cavity were the imperative for both orofacial functions. By using the RBJ stimulator after several months, the reduction of the split was brought to the level that with the help of the available tissue it was possible to perform a palate operation, and the communication between the lips and the nasal cavity was closed. The newborn then developed well until the early child-

hood. We used a 3D analysis of gypsum impressions to verify the results of the efficiency of orthodontic treatment we performed [9]. In addition to 3D analysis, 3D scanning in cleft care provides numerous benefits in diagnosis and treatment plan [18, 19]. By intraoral scanning, 3D scanning, and the development of CAD CAM stimulators the process of taking an impression in the baby could be avoided, as well as the complications that can occur in its course [20]. However, having in mind that 3D cleft care is very expensive and that it is still not available to the vast majority of patients, especially in poorly developed countries, such as Serbia, making RBJ stimulators in a conventional way is a method of choice.

Conclusion

The RBJ stimulator has an active effect in the treatment of children with HCP, premaxillary agenesis and median cleft lip. It significantly reduces the width of the cleft, and thus greatly contributes to providing the conditions for the palate surgery to become possible. Moreover, it ensures the survival of the newborn by enabling the feeding. Since there is no single protocol in the treatment of children with this anomaly, the use of RBJ stimulators is recommended for its positive therapeutic effects.

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