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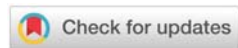
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## Introduction

Binder's Syndrome is a developmental disorder primarily affecting the anterior part of the maxilla and nasal complex [1]. The condition is variably expressed, and reconstruction must be tailored to the individual [2]. Maxillonasal dysplasia, otherwise known as Binder's syndrome, is a congenital malformation characterized by nasomaxillary hypoplasia due to underdevelopment of the midfacial skeleton. It was formerly known by other terms including "dish face," "fossae praenasalis," "facies scaphoidea," "dysostosis maxillo-nasalis," and "congenital flat nose syndrome" [2]. The first classical description of this defect by Binder in 1962 documented six characteristics: arhinoid face, abnormal position of nasal bones, intermaxillary hypoplasia with malocclusion, absent anterior nasal spine, atrophy of nasal mucosa, and hypoplastic frontal sinus [3]. Patients with Binder's syndrome usually have typical facial features characterized by midfacial hypoplasia with a flat nose, flattened tip and alar wings, crescent-shaped nostrils, a short columella, an acute nasolabial angle, an absent nasofrontal angle, a convex upper lip, and a concave midfacial

## Case Report

# Correction of maxillary hypoplasia using modification of Anterior maxillary distraction device in a patient with Binder's Syndrome- Case report

## Abstract

This is the case report of a 20year old female, affected by Binders Syndrome reported to our hospital and was operated for maxillary hypoplasia using modifications of the standard Anterior Maxillary Distraction (AMD) device. This helped in rehabilitation of the patients anterior and lateral profiles; thereby giving a satisfactory cosmetic appearance to the patient.

profile. Skeletal examination shows a hypoplastic maxilla positioned on a short anterior cranial base and an absent or hypoplastic anterior nasal spine [2,4]. Also commonly found in Binder's patients are cervical spine malformations and skeletal and dental class 3 malocclusion [1]. The etiology of Binder's syndrome is not fully understood. Most cases are sporadic, although hereditary factors may play a role because the incidence of Binder's syndrome in relatives is about 15% [2,5]. The degree of severity of the clinical features ranges from mild to severe, and the physical findings include midfacial hypoplasia, flattened nose, short columella, and retrusion of the anterior nasal spine along with class 3 malocclusion [2]. In this case report, we put forth the treatment plan we undertook for our patient specific to her complaints of functional and aesthetic needs (Figure 1).



Figure 1: Pre-operative frontal and worms view profile of patient.

## Case Report

The patient was a 20-year-old female affected by Binder's Syndrome. Her primary complaint was difficulty in chewing due to a deranged occlusion. She also had concerns regarding her missing anterior teeth and her misshapen nose. Patient also had a history of cheiloplasty done at 6 months of age, history of palatoplasty at 1 year of age. She had a flattened nasal bridge with a flattened pro-nasal and a short columella. The premaxilla was rudimentary and she had missing 11, 12, 21, 22, 23. Radiographic features showed a missing anterior nasal spine. Patient had a negative over-jet of 12mm and a class 3 malocclusion with a prognathic mandible. She also had a history of Lefort 1 advancement surgery 1 year back. However, the extensive scarring from previous surgery and the increased amount of negative overjet led to a relapse 2 months post-surgery. So, we planned her for Anterior maxillary distraction to facilitate prosthodontic rehabilitation and correction of negative over-jet as much as favorable. The absence of premaxilla made it necessary to plan the amount of correction such that, the existing anterior alveolus would be positioned in a way to facilitate prosthetic rehabilitation.

Post-distraction, the plan was to provide the patient with fixed prosthesis that would make up for the absence of premaxilla and associated missing anterior maxillary teeth (Figure 2).

After taking consent from the patient for the planned course of treatment, a hyrax orthodontic appliance was custom-prepared beforehand on the patient's maxillary dental model. The appliance was oriented so as to achieve anterior-posterior movement. It had 4 tooth-borne anchorage arms and 4 bone-borne anchorage arms on the palatal side. The four-tooth-borne anterior arms were soldered to the orthodontic bands and cemented on to the desired anchor teeth, 2 arms anterior to osteotomy line and two arms posterior to osteotomy line on either side. There are 4 bone-borne arms curve into loops that help engage the device to the palatal vault through screws.

Presurgical orthodontics includes alignment of the teeth and also to create 1mm of space between teeth through which the planned osteotomy is to be made usually in the premolar canine segment. Space was created so as to avoid any damage to the roots of the teeth while placing the vertical osteotomy cuts. The device was placed intraorally and secured with GIC cement pre-operatively (Figure 3).

Under general anaesthesia and after nasotracheal intubation, vestibular incision was placed in the maxilla intraorally from 1<sup>st</sup> molar to 1<sup>st</sup> molar. Mucoperiosteal flap was raised and buccal cuts horizontal and vertical cuts are placed as per the routine Anterior Maxillary Osteotomy (AMO) procedures.

The palatal cut was placed by tunneling through the mucoperiosteum by using finger to protect the palatal mucosa. Completion of cuts was confirmed through mobilization of the AM segment. BB arms were secured using long screws depending on the bone height usually 1.5mm×15mm screws anteriorly and using 1.5mm×10mm screws posteriorly. Device



Figure 2: A, B, C, D- orthodontic arms; E, F,G, H- loops for palatal screws.



Figure 3: Intra- op frontal view.



Figure 4: Per op cephalogram.

was activated to check the movement between the segments. Closure was done using 3-0 vicryl.

After a latency period of 3 days the distraction was started by turning the device screw at a rate of 1mm per day and a rhythm of 0.25mm by turning the device four times daily (Figure 4).

Thus, distraction of 12mm was achieved over a period of 12 days. After this the palatal screw was sealed using acrylic GIC. The consolidation period was of 3 months and during this period, monthly reviews using radiograph and clinical photographs were done.

At the end of 3 months a change in profile along with 12 mm distraction was achieved. This resulted in an edge-to-edge skeletal relation of the alveolar bones which helped to achieve a class 1 canine relation after prosthetic rehabilitation (Figures 5-8).

We have planned the next stage of her surgery wherein, we intend to augment her columella with a costochondral graft thereby giving a better nasal skeletal structure for improved aesthetics.

## Discussion

The classical features of midfacial retrusion and flat nose due to nasomaxillary hypoplasia create significant aesthetic and functional disability for the patient with Binder's Syndrome [2]. Controversy still surrounds the optimal surgical treatment plan for patients with Binder's syndrome, especially the ideal age for performing surgery and the most appropriate procedure for achieving long-term favorable aesthetic and functional results [2]. Binder's syndrome is clinically apparent when the patient is young and puts considerable psychological strain on such children as they grow up. For this reason, many authors advocate early correction [5]. There are various surgical modalities and options available for the treatment of maxillonasal dysplasia and hypoplasia seen in Binder's Syndrome.

Converse et al., [6]. described a pyramidal naso-orbito-maxillary osteotomy to correct the lack of nasomaxillary projection. A LeFort I osteotomy combined with nasal bone or cartilage grafts was advocated by Munro et al., [4], Jackson et



Figure 5: Per op cephalogram.



Figure 6: Post op cephalogram.



Figure 7: Prosthetic Rehabilitation and radiographic results at the end of 3 months. Facial profile post prosthetic rehabilitation.



Figure 8: Prosthetic Rehabilitation and radiographic results at the end of 3 months. Frontal view of occlusion post prosthetic rehabilitation.

al., [7], performed a LeFort II osteotomy in addition to nasal correction with bone grafts to the dorsum and paranasal areas and columellar lengthening with a VY-plasty. A LeFort II osteotomy and advancement, however, can create a class 2 malocclusion in a patient with previously normal occlusion, and may necessitate orthodontic treatment or a LeFort I osteotomy to restore normal occlusion [4]. Distraction osteogenesis is another treatment modality used in the treatment of maxillary hypoplasia. Although Le fort osteotomy is a commonly performed procedure for correction of maxillary hypoplasia, problems such as relapse and Velopharyngeal Insufficiency (VPI) are documented for large advancement of more than 10mm [8]. When compared, distraction osteogenesis has greater advantages such as application in growing patients and relapse free advancement in cases with more than 10mm with long-term stability, unchanged or better velopharyngeal (VP) function [8]. At this juncture, Anterior Maxillary Distraction (AMD) which was described by Dimitri Karakasis et al in 2004, with the main intention of avoiding extensive surgery and prevention of Velopharyngeal Insufficiency (VPI), has gained popularity in recent years. This procedure also helps reduce surgical time and extent of surgery.

Our patient had the primary concern of deranged occlusion along with missing anterior teeth. To address this concern of hers we sought the use of a less invasive procedure that would give us stable results. We modified the conventional tooth-

borne (TB) AMD device to a combined tooth borne- bone borne (TB-BB) device. This was necessary for this patient as the missing anterior teeth posed a threat for adequate support necessary to bring about optimum distraction forces in a conventional TB AMD device. The use of a TB device resulted in tipping of the anchor tooth due to counter forces acting on the tooth resulting in extrusion of the tooth. And finally, insufficient distraction energy generated by the device taking support from a periodontally compromised anchor tooth would have affected the optimum amount of distraction. However, the modified TB-BB device we used was able to overcome these shortcomings resulting in a stable anterior distraction length of 12mm at the end of 3 months.

Thus, the reverse over jet of the patient was corrected and rehabilitation of an aesthetically appealing dental profile was achieved.

We have planned the next stage of her surgery wherein, we intend to augment her columella with a costochondral graft thereby giving a better nasal skeletal structure for improved aesthetics.

## Conclusion

We were able to restore the dental aesthetics of a patient with Binder's Syndrome using a modified AMD device, thereby giving a good facial appearance to the patient and enhancing her personal confidence.

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