

Maxillonasal dysplasia (Binder's syndrome): Our experience.

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Abstract: Maxillonasal dysplasia or Binder's syndrome is an uncommon congenital condition characterized by a retruded mid-face with flat dorsum of nose. We report here five patients with maxillonasal dysplasia whose noses were corrected with costal cartilage graft using a combined oral vestibular and external rhinoplasty approach for nasal dorsal augmentation, columellar lengthening, and premaxillary augmentation. The cartilage graft was dipped in a solution of 100 ml 0.9% NaCl and one vial (80mg) gentamicin for 30 min to prevent warping. Right angled triangular struts were made and were fixed subperiosteally for nasal augmentation and columellar lengthening. Maxilla was augmented by two piece cartilage graft which was fixed to the pyriform aperture. This technique has been used in children, adults, and for secondary cases with promising results. All patients were of class I dental occlusion. The nasal and premaxillary augmentation which was monitored by postoperative photography was found to be stable over a follow-up period of three years.

Keywords: Binders, costochondral grafts, maxillonasal, premaxillary augmentation, rhinoplasty.

I. Introduction :

Maxillonasal dysplasia is a rare congenital malformation its etiology being unclear. It was first described by Noyes in 1932, but it was not recognized until 1962 then Binders published a comprehensive report of 3 unrelated children with Binders syndrome. He recorded 6 specific features of this malformation which are listed as follows¹ :

1. Arhinoid faces
2. Intermaxillary hypoplasia
3. Atrophy of nasal mucosa
4. Reduced or absent anterior nasal spine
5. Absence of frontal sinuses (not obligatory)

Thus, the patient presents with a typical midface deformity, underdeveloped maxilla , flat nose , acute nasolabial angle, perialar flatness, a convex upper lip². These patients show a typically Angles class 1 malocclusion.

II. Materials & Methods :

Here we report 5 patients who were treated at our institution between the period of May 2005 to July 2010. The patients presented in age gaps of 20 to 30 years .

All patients had an Angles class 1 malocclusion with no malaligned teeth. Clinical & physical examination findings included mid-facial hypoplasia, flattened nose, short columella with an acute nasolabial angle, and retrusion of the anterior nasal spine. Pre- and postoperatively Evaluation was done by serial photography; postoperative follow-up ranged from one year to three years with average follow up of one and half years.

The treatment plan we followed for the surgery was as follows :

Using costal cartilage grafts for nasal augmentation, columellar lengthening, and premaxillary augmentation. The grafts were harvested from the right side of the chest through a lower oblique incision in males & a small submammary incision in females .The cartilage graft was dipped in a solution of 100 ml 0.9% NaCl and one vial (80mg) gentamicin for 30 min to prevent warping. Right angled triangular struts were made and were fixed subperiosteally for nasal augmentation and columellar lengthening. Maxilla was augmented by two piece cartilage graft which was fixed to the pyriform aperture.

III. Results :

The technique used as well as the post operative treatment rendered was well accepted by the patient as well as the overall compliance to the treatment was positive. The costal grafts maintained its volume in all areas irrespective of the patient's age or sex. It did not result in any post operative complication such as infection or necrosis. In this technique mid face as well as the nose & nasal bone were augmented not just the tip or maxillary bone alone.

IV. Photographs :

Figures 1.1-1.4 represent pre operative and post operative pictures of 2 cases treated at our speciality & Figures 2.1-2.3 represent the cartilage grafts used for the procedure.



Fig. 1.1 Fig. 1.2



Fig. 1.3

Fig. 1.4

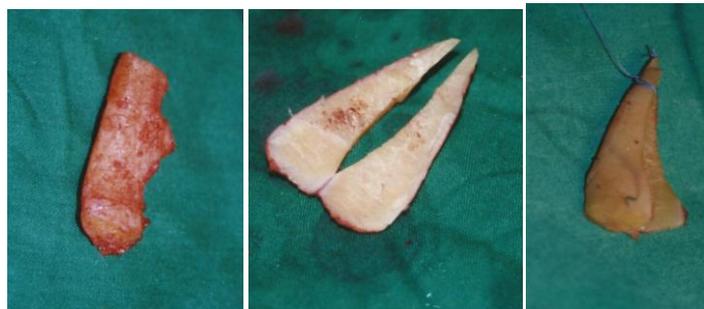


Fig. 2.1

Fig. 2.2

Fig. 2.3

V. Discussion :

Binder's syndrome is a rare disorder and the causes are unclear. It is a developmental disorder primarily affecting the anterior part of the maxilla and nasal complex (nose and jaw). Also known as Maxillo-nasal dysplasia, Maxillo-nasal dysostosis, Naso-maxillo-vertebral syndrome. Binder syndrome (Binder's syndrome)^{7,8}. The characteristics of the syndrome are typically visible. Midfacial hypoplasia with a flat nose, flattened tip and alar wings, half-moon shaped nostrils, short columella, acute nasolabial angle, frontonasal angle of almost 180° eventually resulting in a concave midfacial profile are the typical features seen in these patients. The treatment of the malformations in this syndrome was typically by the use of cartilage grafts which have been traditionally used to treat the maxillonasal hypoplasia.

Costal cartilage grafts, maintain their volume in all areas making it the ideal material for augmentation. To prevent warping of the large grafts, they must be carved from the central part of the rib then placed into 0.9% sodium chloride solution for about 30 min. Although the balance of the surface tension forces is not

maintained, the residual stress of the cartilage splinter comes to the force and even if there is some amount of warping at the end of 30 minutes, it can be taken into account when implanting.

The flat nose in Binder's syndrome has also been considered to be a problem of soft tissue deficiency in the columella. Its lengthening has been achieved by the use of a free auricular graft, small flaps from the upper lip, bilateral flaps from the nasal floor, and VY-plasty of the columella. A Le Fort two osteotomy lessens the normal glabellar depression and this may be a limiting factor as a nasal dorsum coming straight off the lower forehead is not ideal aesthetically^{8,9}. Holmström and Kahnberg recommend a two-stage surgical procedure, firstly maxillary osteotomy followed by the nasal improvement secondarily, both independent of the patient's age^{5,9}.

VI. Conclusion :

Binder's syndrome is a rare congenital malformation. Nasal reconstruction as well as orthognathic as well as orthodontic treatment is necessary. The child must be kept under observation by the orthodontist as well as plastic surgeon since birth. Surgical intervention must be done depending upon the severity of the malformation and treatment planned accordingly.

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