Binder's syndrome: Modified approach for correction of Nasomaxillary hypoplasia.

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Abstract:

Introduction: Binder syndrome is a relatively uncommon syndrome characterized by nasomaxillary hypoplasia. Different approaches for correction of hypoplastic nasomaxillary complex has been developed and studied over years. Our study shows our experience with extra oral only technique of onlay costal cartilage graft for nasal dorsal augmentation, premaxilla augmentation and columella lengthening in three patients. **Materials and Method:** We report here three patients with nasomaxillary dysplasia whose noses were corrected with onlay costal cartilage grafts using external rhinoplasty approach for nasal dorsal augmentation, columellar lengthening, and premaxillary augmentation. L struts made for nasal augmentation, columellar lengthening, and premaxillary augmentation were fixed to one another by putting it in dissected pockets. **Results:** All the patients were operated single time. Patients were followed up with sequential photography over 6 months to 2 years. Costal cartilage maintained their volume in post operative period. **Conclusion:** Binder's Syndrome: Augmentation of the premaxilla is necessary along with nasal augmentation and columellar lengthening with autogenous costal cartilage grafts for effective treatment. Augmentation with costal cartilage is enough to give an aesthetically pleasing facial profile in mild to moderate cases.

Keywords:- Binder's Syndrome, Costal cartilage grafts, Extra oral only approach.

Introduction:

Binder's syndrome is a rare congenital anomaly characterized by nasomaxillary hypoplasia due to an abnormal development of the mid-facial skeleton.[1-3] The causative etiology of this syndrome is disturbance of the prosencephalic induction center during embryonic life.[2] Birth trauma has also been suggested as a possible etiology.[4] The essential feature of binder syndrome was initially described by Noyes in 1939,[4] and later defined it as a distinct clinical entity in 1962.

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Binder reported three cases and six peculiar features[5]:(1) arhinoid face; (2) abnormal position of the nasal bones; (3) Intermaxillary hypoplasia with consecutive malocclusion; (4) reduced or absent anterior nasal spine; (5) atrophy of the nasal mucosa, and (6) absence of the frontal sinus (not obligatory). Characteristic appearance of

individuals with Binder's syndrome makes it easily recognizable.[6] The mid-face profile is hypoplastic, the nose is flattened, the upper lip is convex with broad philtrum, nostrils are typically crescent or semi lunar-shaped giving a half-moon appearance, columella are short with deep fossa or folds between the upper lip and the nose, resulting in an acute nasolabial angle. The frontonasal angle is almost 1800, resulting in a concave mid-face profile. Till date, 250 cases have been reported with equal sex predominance and ranging from mild to severe.

By doing cephalometrical analysis, there is reduced sella nasion distance [7] and the length of the maxilla measured from the anterior surface of the maxilla to the posterior nasal spine is reduced. However, it has been suggested that there is a common concurrent induction process for both the prosencephalic area and the vertebrae, accounting for the increase in vertebral anomalies associated with this condition.[8]

It can be associated with other malformations. In the most severe forms, the syndrome is associated with true mandibular prognathism requiring both, orthodontic and surgical treatment.[10]

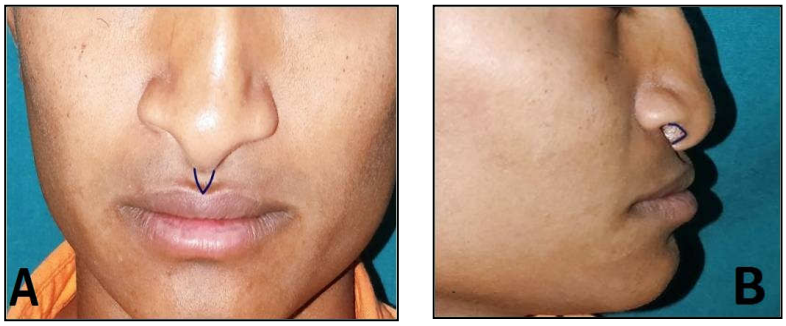
There may be pseudomandibular or true mandibular prognathism with a hypoplastic maxilla. The severity of the malocclusion correlates with the severity of the syndrome. In mild cases, orthodontic treatment may not be necessary because of compensatory effects in dental arches, while in the most severe cases, the maxillary underdevelopment is aggravated by mandibular prognathism and can only be treated by combined orthodontics and surgery. In longitudinal cephalometric studies comparing orthodontically treated children with Binder's syndrome with untreated cases, it was concluded that conventional orthodontic therapy did not produce evidence for a positive influence on craniofacial growth.[10] With increasing age, the maxilla grew forward, but not to the same extent as the mandible. Growth impediment was confined to the area around the absent anterior nasal spine in subjects with moderate forms of the syndrome. Olow-Norderam and Thilander advised postponing definitive orthodontic treatment in individuals with maxillonasal dysplasia until growth has stopped, especially in those with severe malocclusion.[10] it has been suggested that corrective surgery of the midface and nose has the potential to jeopardize acceptable occlusal results following early orthodontic correction. Olow-Norderam concluded that the severity of the malocclusion was evident at an early age. Patients who proceeded on to orthognathic surgical correction had more retrognathic maxillae, increased mandibular planes angles, large gonial angles, and markedly negative apical base angles than milder cases with Binder's who were treated orthodontically with success. The possibility of family history was put forward by Ferguson and Thompson.[11] Olow-Norderam reported positive family history in 36% of their patients.[12, 13]

Gorlin *et al.* suggest that Binder's syndrome is a nonspecific abnormality of the nasomaxillary complex. They believe that familial examples are a result of complex genetic factors, similar to those involved in producing a malocclusion.[14] in the present study, we report our experience on the correction of the nasal and premaxillary areas in 3 patients with Binder's Syndrome over a follow-up period of two years.

Materials and Methods:

In our study, we had operated 3 patients with nasomaxillary hypoplasia in 2018-19. Patients' age were ranging from 16 year to 24 years. Sex ratio male: female was 2:1. Physical examination findings include mid-facial hypoplasia, flattened nose, short columella with an acute nasolabial angle, and retrusion of the anterior nasal spine and fronto-nasal angle approaching 180°. All patients had class I dental occlusion with no malalignment of teeth. All patients were evaluated pre and postoperatively by serial photography. Our surgical treatment plan consisted of nasal augmentation, columellar lengthening, and premaxillary augmentation using costal cartilage grafts. The grafts were harvested from the right side of the chest through a small submammary incision in females and a lower oblique incision in males. To achieve an anterior projection of the nose and mid-face, usually three cartilaginous strips were implanted through a combined external rhinoplasty. L struts were made for dorsal augmentation and columellar lengthening and a separate one was made for premaxillary augmentation onto the superior alveolar process. Placement of the three cartilaginous splinters: one on the dorsum, the second into the columella, and the third onto the maxilla.

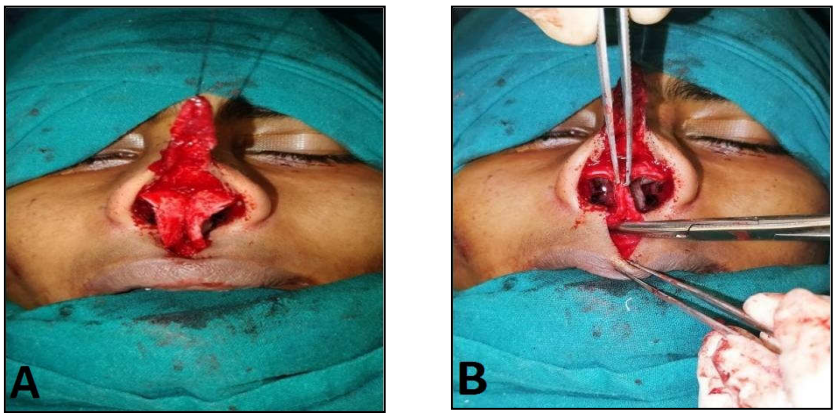
Image 1: Incision planned for elevation of columella-labial complex & anterior nasal framework



(A) Anterior view (B) Lateral view

Elevation of upper lip and columella has been done as per incision marked and anterior nasal framework is exposed. (Image-2) Dissection around piriform aperture was done through "V" incision. By this mean, we didn't require separate intra-oral incision for cartilage graft placement for premaxillary augmentation.

Image 2: Elevation of upper lip & columella as per incision.

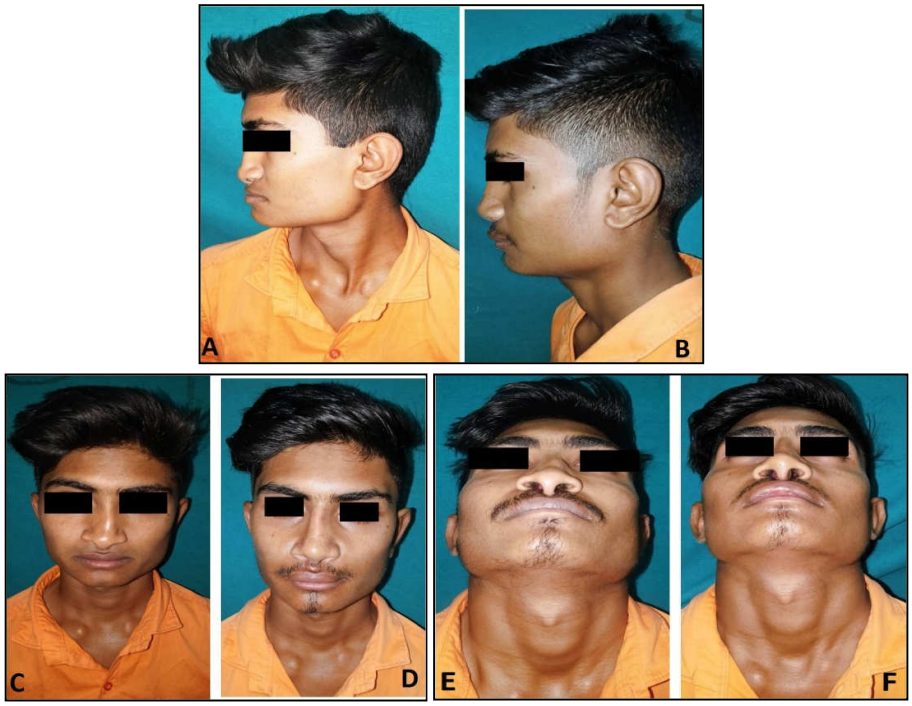


Costal cartilage graft harvested & carved to the desired shape, three separate grafts for dorsal, columellar and premaxillary portions.(Image-3)

Image 3: Costal cartilage graft.

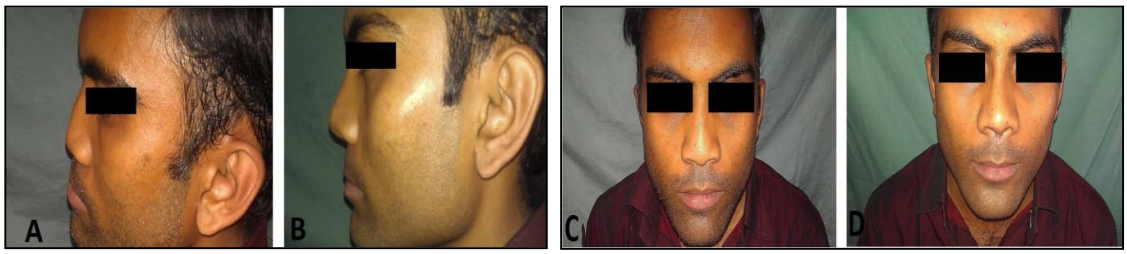
(A) Harvested cartilage graft, (B) Graft carved for desired shape

Image 4: 19 year old male with Binder syndrome.



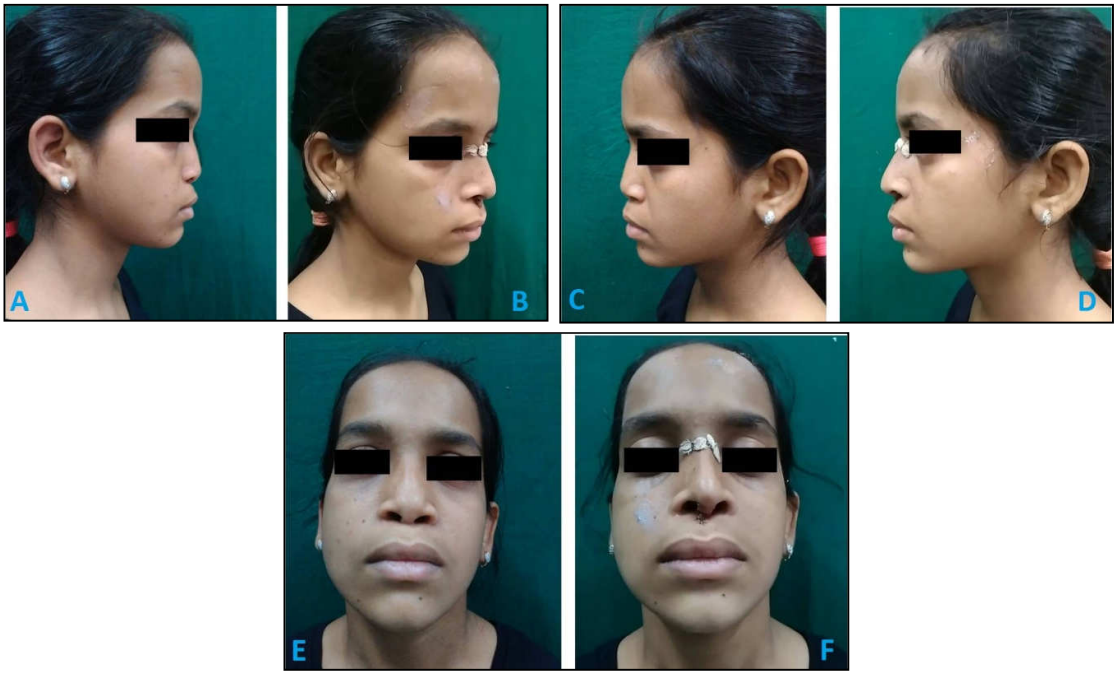
Pre operative and post operative photograph in profile view [A and B],
Front view [C and D] and basal view [E and F]

Image 5: 26 year old male with binder's syndrome corrected with costal cartilage graft.



Pre operative and post operative photograph in profile view [A and B],
Front view [C and D]

Image 6: 17 year old female with nasomaxillary hypoplasia.



Pre operative and post operative photograph in Left lateral view [A and B], Right lateral view [C and D] and Front view [E and F]

All cases were followed up for 6 months to 2 years duration showed nasal and maxillary augmentation to be permanent. Mid-face and nasal augmentations were satisfactory in all cases along with increase in length of columella and nasal opening.

Discussion:

In Binder's syndrome, the hypoplasia of the nasal floor and the adjacent part of the maxilla produces the characteristic dish-face anomaly and a flat nose mainly due to a deficient horizontal growth of the maxilla. [16, 17, 18,19,20,21, 22] Surgical correction is demanded as these deformities are evident at a very young age and often lead to severe psychological problems besides the functional restrictions.

In planning the treatment strategy, two questions have to be taken into special consideration: (1) what is the appropriate surgery, and (2) which is the optimal age for performing surgery?

Bone and cartilage grafts have been traditionally used to treat the maxillonasal hypoplasia. Ragnell described the application of iliac cancellous onlay bone chips to the anterior surface of the maxilla through a median incision at the columellar base.[23] Converse used the oral vestibular approach to insert a shell-like segment of iliac bone.[24] Later, he proposed using an L-shaped bone graft to reconstruct the dorsum and the shortened columella.[25] To raise the nasal contour, Holmstr Öm[26] as well as Losken[27] and later Rune,[28] used L-shaped bone grafts taken from the iliac bone and the skull, respectively. They also augmented the premaxillary region with bone chips[26] or a U-shaped bone segment[27] through an oral vestibular approach[26] or a perialar crease incision or one just below the columella.[27] However, the results of bone grafts remain unpredictable. Resorption

often occurs especially if the soft tissue cover is very tight and displacement of the bone strut has been described to lead to disappointing long-term results. [28] The patients are very often disturbed by the stiffness of the tip of the nose and the rigidity of the bone implant leads to easier fractures.[16, 28] The pain in the bone graft donor site lasts longer and delays ambulation.

Costal cartilage grafts, on the other hand, maintain their volume in all areas and produce a more natural feeling to the nose, making it the ideal material for augmentation. Ortiz Monasterio *et al*.[30] also described convincing results in augmenting mid-facial deficiencies by using cartilaginous onlay grafts to the piriform area, such as L-strut grafts for dorsal and columellar areas.

Some authors have proposed the use of alloplastic implants but the risk of increased extrusion rates and infection are more as they are not an autogenous material, and it is not cost-effective considering the Indian scenario.

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.[26]

Our concept is to lengthen the columella by VY-plasty if there is a real shortage of skin, but if there is just a retraction into the hypoplastic nasal floor; skin advancement can be achieved by undermining the skin at the lip-columellar junction and with the help of nasal cartilage grafts. If necessary, the cartilaginous septum is rotated forward to additionally support the nasal dorsum. A limitation to the achievement of an optimal result is presented by the constriction of the soft tissue covering the nose and of the lining of the nasal cavities which were not expanded progressively, as it occurs in normal patients. According to Ortiz Monasterio *et al.*,[30] this problem can be prevented if surgical treatment starts early because the corrected facial conditions follow a pattern similar to normal growth. At least equally important is the advantage of improving the self-image of the patients during their growth period when surgery is performed early in life. Therefore, we cannot agree with Tessier *et al.* [31] that the ideal age is 16 years for surgery in Binder's syndrome when growth of the maxilla is completed; one should at least use an onlay graft technique without osteotomies. In our series, all patients had Class one dental occlusion and no malalignment of the teeth, so no orthodontic treatment was required. In cases with severe malocclusion, particularly Type three, maxillary retrognathia should be corrected by a Le Fort one maxillary advancement. However, even if the septum and nasal bones are included in the advanced segment, as in a Le Fort two osteotomy, the flat nose and the depressed alar base remain and with it remain the facial characteristics of Binder's syndrome.[32] This is mainly due to the absent septal support of the nasal dorsum and the relative retrusion of the septum with respect to the nasal base.[25,33]

Furthermore, 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.[34] These facts point to the major importance of nasal correction in patients with Binder's syndrome.

In severe cases of the syndrome, Holmstr Öm and Kahnberg[35] recommend a two-stage surgical procedure, firstly maxillary osteotomy followed by the nasal improvement secondarily, and both independent of the patient's age.[36]

As the degree of malformation in Binder's syndrome varies significantly, surgical correction needs to be individually tailored based on the demonstrated principles.[37] The onlay grafting technique seems to positively influence facial growth with minor secondary corrections being an option at any time.

Conclusion:

Binder's Syndrome: Augmentation of the premaxilla is necessary along with nasal augmentation and columellar lengthening with autogenous costal cartilage grafts for effective treatment. Extraoral only approach makes deformity correction easier. Augmentation with cartilage graft gives an aesthetically pleasant result in mild to moderate cases.

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