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Original Research Article

Classification and Management Variants of Nasomaxillary Hypoplasia Based on the Clinical Features: A Retrospective Study

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Abstract

Objectives: To classify the variants of nasomaxillary hypoplasia based on the clinical features an advocate a logical treatment protocol for each type described.

Material & Method: This was a retrospective study of 14 cases of nasomaxillary hypoplasia (Binder's syndrome) selected from case records and photographs of 113 cases of rhinoplasties performed over a period of two years. Diagnosis of Binder's syndrome was based on the typical features.

Results: Nasal correction with loco regional autologous cartilage grafts was sufficient in mild cases. Loco-regional cartilage grafts along with costal cartilage grafts were needed for moderate and severe cases. Anterior nasal floor along with alar base augmentation was performed to achieve a proper aesthetic profile in moderate and severe cases. Post-operative results were excellent in mild and moderate cases and acceptable in severe cases.

Conclusion: Most cases of nasomaxillary hypoplasia present as mild or moderate deformity, severe hypoplasia being seen in only a few cases. During surgical reconstruction, loco regional cartilage grafts (septal and conchal) can be used extensively in mild and moderate cases without having to solely depend on costal cartilage for augmentation. 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, nasomaxillary hypoplasia

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.

Binder reported three cases and six peculiar features5:(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 [5].

There is no sexual predominance, and most cases are sporadic. [6] More than 250 cases have been reported in literature so far. [7] Surgical treatment for this deformity has been challenging to say the least. It has evolved from simple onlay bony cartilaginous grafts to Le Fort's osteotomies for maxillary advancement.

Due to the rarity of the congenital condition, literature reveals either single case reports or small series studies on Binder's syndrome. [8, 9] Hence the present study was conducted with the objectives to classify the variants of nasomaxillary hypoplasia based on the clinical features. and advocate a logical treatment protocol for each type described.

Material & Method:

This was a retrospective study of 14 cases of nasomaxillary hypoplasia (Binder's syndrome) selected from case records and photographs of 113 cases of rhinoplasties performed over a period of two years in private multi-specialty hospital of Bihar India

Diagnosis of Binder's syndrome was based on the typical features.

Age of the patients ranged from 18 to 30 years with an average age of 19.2 years. Out of 14 patients, 4 patients were males and 10 were females. All the cases underwent primary rhinoplasty. All the patients were operated by one surgeon. Ethical permission was obtained from ethical committee of the institute.

Surgical technique and operative steps:

All cases had a common surgical approach; transcolumellar incision and open rhinoplasty. V-Y advancement of columellar incision was planned in severe variety of cases. Dissection was done inferiorly towards ANS in mild cases and extended laterally up to pyriform aperture in moderate and severe cases. Bilateral sublabial incisions were made in severe cases for placement of alar base grafts. Nasal pyramid was developed in subperichondrial plane. Nasal septum was exposed from the caudal angle/border in bilateral subperichondrial planes by anterior and inferior tunneling.

Mild cases:

- ***Cartilage grafts:*** Nasal septal cartilage grafts and bilateral conchal cartilage grafts were used.
- ***Columellar projection:*** Columellar projection was done by septal cartilage graft extended from ANS below to the future tip.
- ***Dorsal projection:*** The apparent bony hump of the dorsum was resected,and stacked conchal cartilage graft was placed on the cartilage dorsum as an overlay graft.
- ***Tip projection:*** Tip projection was achieved by lateral crural steal,intra- and inter-domal suturing and a small dome graft.

Moderate cases:

- ***Columellar projection:*** A T-shaped cartilage strip assembly was formed from the rib cartilage and fixed in an inverted T manner. The horizontal limb was placed anterior to the short ANSand on the anterior nasal floor, vertical limb formed the caudal strut.
- ***Dorsal projection:*** Cartilaginous dorsum was augmented by first partially separating the upper lateral cartilages and placing two-rib cartilage strips as spreader grafts extended tothe tip.

- ***Tip projection:*** Tip projection was achieved in the same manner as in mild cases.

Severe cases:

- ***Columellar projection:*** T-shaped cartilage assembly was created as in moderate cases, but the horizontal limb was fixed to the premaxilla with 6 mm titanium screws.
- ***Dorsal projection:*** Bilateral upper laterals were detached completely, two-rib cartilage strips were placed as extended spreader grafts and raised above the existing dorsum from the keystone area (bone–cartilage junction) down to the tip. The upper laterals were reattached, and spreaders were fixed at the keystone area with a prolene stitch passed through a drill hole in the nasal bones.
- ***Tip projection:*** Tip projection was same as in mild and moderate cases, but weak alar cartilages were reinforced with pieces of conchal cartilage grafts.

The following six parameters were studied to assess the functional and aesthetic outcome:

1. Nasal breathing: Normal/reduced
2. Satisfaction scale: Very satisfied (8‑10)/satisfied (5‑7)/dissatisfied (<4)
3. Appearance improvement scale: Great improvement (8-10)/some improvement (5-7)/no improvement (<4)
4. Tip stiffness: Nil/minimal/bothersome
5. Donor site: Nil scar/hypertrophic scar/keloid
6. Columellar scar: Visible/barely visible/well visible.

Results:

Table 1: Baseline Demographics

Gender	Number	%
Males	4	28.6
Females	10	71.4
Total	14	100

Table 2: Distribution of study participants with respect to severity

Severity	Number	%
Mild	4	28.6
Moderate	8	57.1
Severe	2	14.3
Total	14	100

Discussion:

A correlation between the nature of a deformity and the treatment instituted for each case evolved, leading to the formulation of a classification system of mild, moderate and severe Binder's syndrome along with the proposed surgical correction for each group. Earliest comments on Binder's syndrome as normal length and short nose variants were made by Rintala and Ranta. [10] Although the terms mild and severe were used in many studies on Binder's syndrome, so far no single study has put forward such a classification. [11-14]

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. [15] Converse used the oral vestibular approach to insert a shell-

like segment of iliac bone. [16] Later, he proposed using an L-shaped bone graft to reconstruct the dorsum and the shortened columella. [17]

Regarding corrective rhinoplasty, this study differs from the traditional method of placing a L-block assembly of either cartilage or bone on the degloved nasal pyramid. [18,19] We have also not followed the cantilever technique of augmenting the nasal dorsum and tip. [20, 21]

Even if the septum and nasal bones are included in the advanced segment, as in aLe Fort two osteotomy, the flat nose and the depressed alar base remain and with it remain the facial characteristics of Binder's syndrome. [22] 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. [23] 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. [24]

Monasterio *et al*. [18] quote 15% of class 3 occlusion in their series of Binder's syndrome. However, if required, Le Fort II osteotomy and orthodontic treatment must be included in the management of severe cases as advised in some other studies. [25-27] At the same time, management of extreme hypoplasia where there is a requirement of nasal inlay grafting or permanent prosthesis is out of scope of this study. [28]

Conclusion:

Most cases of nasomaxillary hypoplasia present as mild or moderate deformity, severe hypoplasia being seen in only a few cases. During surgical reconstruction, loco regional cartilage grafts (septal and conchal) can be used extensively in mild and moderate cases without having to solely depend on costal cartilage for augmentation. 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.

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