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Original Article

Binder's syndrome (maxillonasal dysplasia) different treatment modalities: Our experience

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ABSTRACT

Aim: Being an uncommon congenital condition, the treatment modalities of maxillonasal dysplasia are not clearly defined. Our aim is to discuss the availability and utility of various treatment options to achieve optimum results. In patients with Binder's syndrome, the midface appears flattened, the columella is short and the upper lip slants backwards. **Materials and Methods:** We report here 15 patients with Binder's syndrome who were operated over a period of 5 years. Different treatment options in the form of correction of the depressed nasal dorsum and maxillary hypoplasia with split cranial bone graft or synthetic materials such as high-density porous polyethylene implant were used. Two patients with Angle class III malocclusion underwent a Le Fort I osteotomy for maxillary advancement. The patients were followed over a period of 3 years. **Results:** We achieved a reasonable augmentation of the nose and the maxilla in our patients. We faced complications in two of our patients; in one patient there was fracture of the dorsal nasal bone graft and the other patient had protrusion of paranasal screws into the palate, which were removed. **Conclusion:** In this series of cases, we were able to utilise various treatment modalities appropriately to achieve satisfactory outcome with no significant complications.

KEY WORDS

Binder's syndrome, maxillonasal dysplasia, split calvarial bone graft

INTRODUCTION

Binder's syndrome or maxillonasal dysplasia is a congenital malformation and its aetiology is unclear. Noyes considered that his patient's abnormalities resulted from birth trauma, but did not comment upon absent nasal spine.[1] Hopkin later concluded it to be a result of development insult. The occurrence of this malformation is sporadic, though familial recurrence has been noted. Recurrence in siblings with unaffected parents has been noted to be seven times and it is ten times for siblings with an affected parent.[2] It was suggested by Oliver Quarrell that the phenotype be considered as an association which principally affects the maxilla and vertebra.[3] Although it was first described by Noyes,[1] the syndrome was not recognised until Binder's comprehensive report of three unrelated children was published in 1962. He recorded six specific clinical characteristics:[3-5] arhinoid face, intermaxillary hypoplasia, abnormal position of nasal bones, atrophy of nasal mucosa, reduced or absent anterior nasal spine, and absence of frontal sinus (not obligatory).

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The result is a typical appearance including 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. Maxillary hypoplasia leads to relative prognathism and Angle type III dental malocclusion.[6] Delaire *et al.* had reported on presence of small central upper incisors. There may be associated cleft palate.[7] The anatomical basis for some of the facial abnormalities can be appreciated from reviewing the lateral cephalogram.[3,5,8] Decreased intelligence is not a significant feature in this condition. Vertebral anomalies are seen in about 50% of cases with Binder's,[9] though none of our patients had vertebral anomalies. Olow-Nordenram and Radberg found no correlation between the presence of cervical spinal abnormalities and severity of malocclusion.[10] All patients with Binder's syndrome have some degree of these characteristics which may range from mild to severe. The present series of 15 cases of Binder's syndrome were diagnosed on clinical grounds and radiographs. Depending on the patient needs and demands, we have utilised various available treatment modalities in the form of onlay augmentation and orthognathic surgery. We had a follow-up period of 3 years which revealed sustained and satisfactory results.

MATERIALS AND METHODS

Our study is based on 15 patients with Binder's Syndrome treated over a period of 5 years. The age of the patients ranged from 19 to 28 years. The mean age was 22 years. The female to male ratio was 10:5. The diagnosis of maxillonasal dysplasia was based on clinical findings of typical facial features. The requirements of each patient were evaluated preoperatively with regard to occlusion, aesthetic outcome and patient expectations. Lateral cephalogram was done in all patients for preoperative analysis and for the evaluation of the results of treatment. During the pre-op consultation, it was observed that only two patients had Angle class III malocclusion while the rest had class I occlusion. The two patients who had class III dental malocclusion were treated with Le Fort I osteotomy for maxillary advancement as mentioned in Figures 1 and 2. The remaining 13 patients with Class I occlusion were given a choice of onlay augmentation with either calvarial bone grafts or with high-density porous polyethylene (HDPE) implants. Eleven patients opted for calvarial bone graft in view of advantages such as a hidden

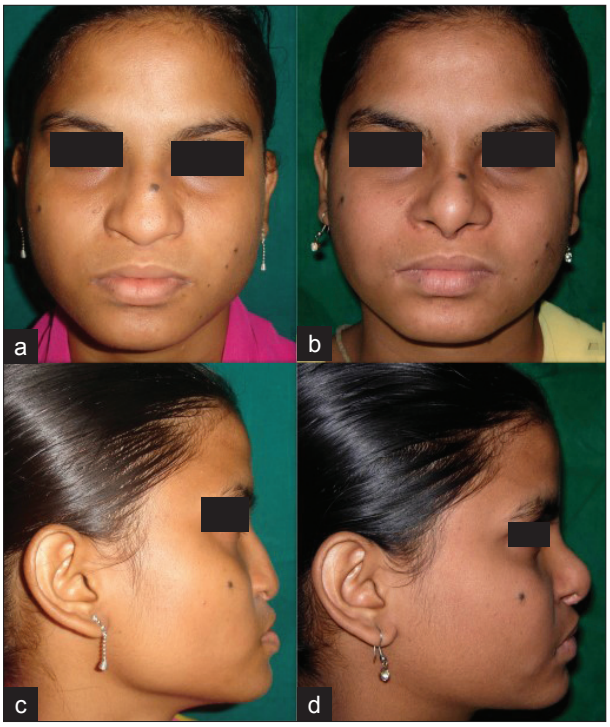

Figure 1: A 22-year-old female with Binder's syndrome who underwent Le Fort I osteotomy and nasal dorsal augmentation with calvarial bone graft. Comparison of preoperative (a, c) and postoperative (b, d) status

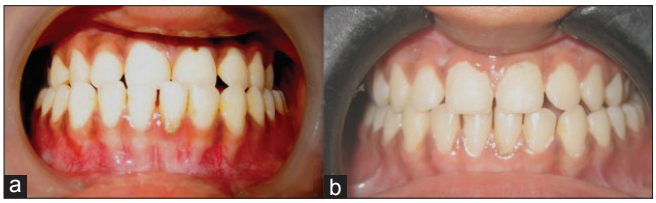

Figure 2: Comparison of pre-op dental occlusion (a) and post-op dental occlusion (b) of a patient who underwent Le Fort I osteotomy

scar, autogenous tissue, less chance of infection and exposure, while two patients opted for the HDPE implant. The choice of material was not influenced by the authors; it was purely the patient's choice, although the available options and their advantages and disadvantages were clearly explained preoperatively. A common approach was used for surgical exposure in all patients. The midface and the nasal cavities were dissected through an incision in the upper oral vestibulum and dissection was done in the subperiosteal plane to expose the maxilla. An open rhinoplasty approach with a columellar incision was used for nasal correction.[11] A coronal incision was used to harvest split calvarial bone grafts: bean-/kidney-shaped grafts for the paranasal region and boat-shaped grafts for the nasal dorsum. The nasal dorsal bone graft was placed as a cantilever graft. Various procedures which were carried out are given in Table 1. Columellar reconstruction with alar sill flaps was done in one patient and two patients underwent V-Y columellar lengthening.

In two patients, tip augmentation was also done using conchal cartilage graft.[12] One of our patients who had undergone nasal dorsum augmentation 10 years back with a costochondral graft presented with complete resorption of the graft. The patient was counselled for a revision surgery and nasal dorsal augmentation with split calvarial bone graft was done as depicted in Figure 3.

RESULTS

The patients were followed over a period of 3–4 years, with a mean follow-up of 3 years. During the follow-up period, assessment of results was carried out by comparing preoperative and postoperative clinical photographs and lateral cephalograms as seen in Figures 4-5. There was minimal bone resorption and nasal and maxillary augmentation was persistent as clearly seen in Figures 6-8. In the patients who underwent Le Fort I osteotomy, no relapse was noticed and maxillary advancement was maintained as seen in the lateral cephalogram in Figures 1a and b. The columellar lengthening achieved was satisfactory. We had complications in two patients. One patient had fracture of the dorsal bone graft in the nose, for which corrective surgery in the form of nasal augmentation with an HDPE implant was carried out after 6 months. It was patient's choice to use the HDPE implant as she had previously undergone surgery using a calvarial bone graft. Another patient had protrusion of the paranasal screws into the palate which needed removal. None of the patients had infection, skin necrosis or any other complications. All patients were asked a few basic questions to evaluate their perspective and feelings towards their general outcome and the patient feedback was quite positive.

Table 1: Various phenotypes and appropriate surgical procedures for Binder's syndrome

Procedure	*Phenotype*	*Number of patients*
Nasal dorsal/ paranasal augmentation with split calvarial bone graft	Maxillary hypoplasia with class I dental occlusion	7
Nasal dorsal/ paranasal augmentation with HDPE implant	Maxillary hypoplasia with class I dental occlusion	2
Only nasal dorsal augmentation with calvarial bone graft	Minimal maxillary hypoplasia with severe nasal deformity	4
Le Fort I osteotomy with nasal dorsal augmentation with calvarial bone graft	Maxillary hypoplasia with class III dental occlusion	2
Columellar lengthening		3

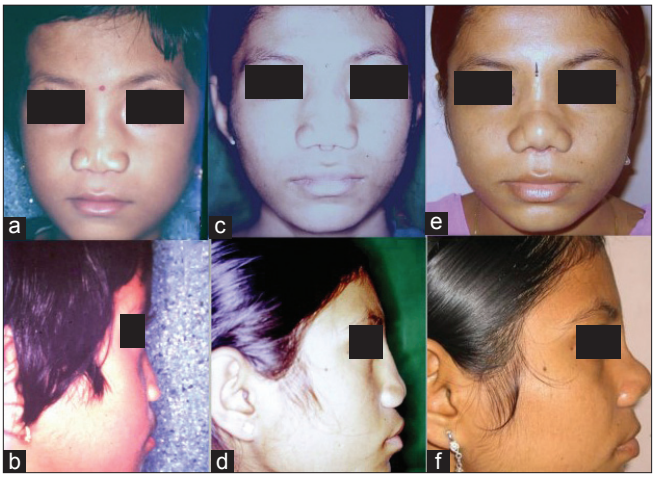

Figure 3: A 20-year-old female patient was first operated at the age of 5 years (a, b) second surgery at the age of 15 years where nasal dorsal augmentation with costal cartilage graft (CCG) was done followed by complete resorption of CCG (c, d) postoperative status after third surgery with calvarial bone grafting (e, f)

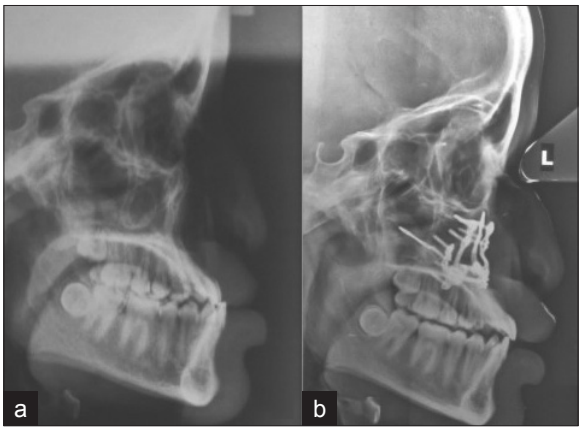

Figure 4: (a) Preoperative lateral cephalogram and (b) postoperative lateral cephalogram of a patient with Le Fort I osteotomy

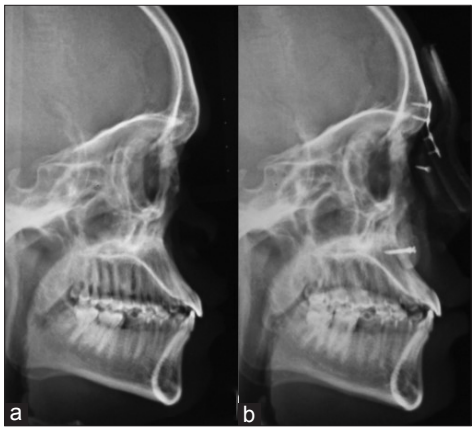

Figure 5: (a) Preoperative lateral cephalogram and (b) postoperative lateral cephalogram of a patient with nasal dorsal and paranasal augmentation with calvarial bone graft

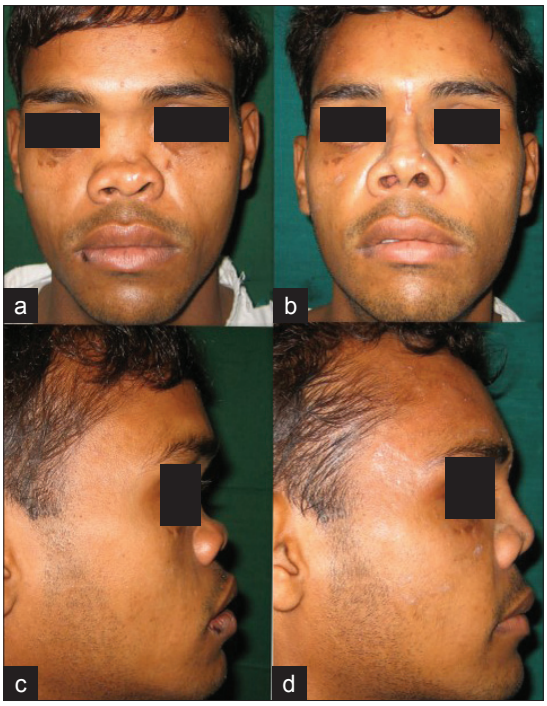

Figure 6: A 20-year-old male with nasal dorsum and paranasal calvarial bone grafting; (a, c) shows preoperative status and (b, d) shows postoperative results

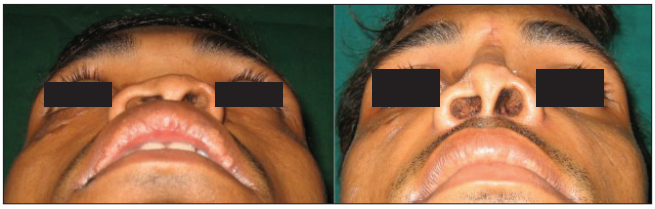

Figure 7: (e) Showing preoperative status and (f) showing postoperative results in worm's eye view of patient in case 2

DISCUSSION

The facial features of maxillonasal dysplasia are well described. The midface appears flattened, the columella is short and there is backward slanting of the upper lip. In most reports on Binder's syndrome, the local maxillary hypoplasia of the floor and walls of piriform aperture have been corrected with bone grafts. Various methods of correcting the deformity associated with the Binder's syndrome have been mentioned in the literature, although no rigid protocols for treatment are followed. The nasal deformity can be corrected with bone grafts, cartilage grafts or the alloplastic materials. Similarly, paranasal onlay grafting or a Le Fort I or II osteotomy has been described for the correction of the midface hypoplasia and malocclusion. Converse used the oral vestibular approach to insert a shell-like segment of iliac bone,[13,14] and Ragnell applied cancellous bone chips to the anterior surface of the maxilla through a

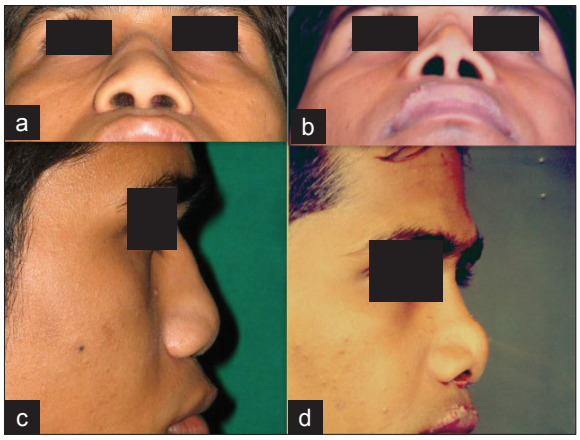

Figure 8: A 26-year-old male; (a, c) showing preoperative status and (b, d) showing postoperative results after 15 months, with satisfactory result

median incision at columellar base.[15] In our cases, we have used onlay bone grafts as well as alloplastic implants to correct the hypoplasia in patients with class I occlusion. An upper vestibular approach was utilised to place the graft in the paranasal region and columellar incision was used to place nasal dorsal graft. The split calvarial bone graft was harvested in the shape required for the onlay graft. The split calvarial bone graft has the advantage of concealed scar site and has comparatively lesser pain as compared to other bone harvest sites. The HDPE implant was used in two patients who refused bone grafting. The use of cartilage from the costochondral junction as a cantilever was preferred by Munro, Sinclair, and Rudd because this gave a softer nasal tip.[5,12] It does have the disadvantage of cartilage warping, and when the deformity is severe, bone grafting allows greater correction of the sunken nose.[15,16] Cartilage grafts, however, cannot be used as stress-bearing structures. Bhatt mentioned the use of costal cartilaginous grafts for nasal dorsum augmentation and the premaxillary area with satisfactory results.[17] In two of our patients, only tip augmentation was done using conchal cartilage grafts.. We have also placed our bone grafts as cantilever. Remodelling is always to be expected in grafts. Tessier had stated that nasal skin can be lengthened or stretched to almost any extent in short noses without the need for skin grafts or flaps.[18] We also observed as was mentioned by Hans Holmstrom that a thorough undermining of the skin at the lip–columellar junction made it possible to advance the skin around the columellar base up into columella with the help of the nasal bone graft. In most cases, there was no real shortage of skin in the columella.[19,20,21] The soft tissue deficiency in the columella is identified as one of the reasons for the flat nose in Binder's. The lengthening of the columella in maxillonasal dysplasia has been achieved by the use of a free auricular graft, small flaps from the upper

lip and V-Y plasty.[22] We had two patients who required columellar lengthening which was achieved with V-Y plasty and one patient underwent columellar reconstruction with alar sill flaps. It is important to give a good projection to the tip without endangering the circulation of the overlying skin. This can be accomplished by thorough subperiosteal dissection of the soft tissues of the midface.[23] In cases with malalignment of teeth, particularly type III occlusion, orthodontic treatment followed by orthognathic surgery is required such as a Le Fort I or II maxillary or nasomaxillary advancement.[14] However, even if the septum and the nasal bones are included in the advanced segment, as in the Le Fort I I osteotomy, the flat nose and the depressed alae persists.[24-26] In this series, two patients had class III occlusion for which Le Fort I osteotomy with nasal dorsal augmentation with calvarial bone graft was done. In all of our patients, good augmentation of the maxilla and the nasal dorsum was achieved and persisted. More importantly, all patients were satisfied with the outcome.

CONCLUSION

Binder's syndrome or maxillonasal dysplasia is an uncommon clinical entity, but the exact birth prevalence remains unknown. It is important to understand that Binder's syndrome has a variable presentation, and therefore needs to be tackled with various treatment options. Of utmost value is a careful preoperative assessment and evaluation to determine which treatment modality will give maximum possible benefit both aesthetically and functionally. Depending on the facial correction needed and patients' demands, the treatment strategy may include orthognathic surgery or onlay augmentation with autogenous graft or alloplastic implant. In this series of cases, we were able to utilise various treatment modalities appropriately to achieve satisfactory outcome with no significant complications.

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