

## Case Report

### Binder's syndrome: Report of two cases

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#### ABSTRACT

Binder's syndrome is an uncommon entity characterized by midfacial hypoplasia along with Class III incisal relationship. The individuals with this syndrome are easily recognizable and the syndrome is mostly associated with other malocclusions. The current article presents two cases of this rare syndrome and describes its general features.

**Key Words:** Binder's, midfacial hypoplasia, maxillofacial dysplasia

#### INTRODUCTION

Binder's Syndrome or Maxillofacial Dysplasia was first described by Noyes<sup>[1]</sup> in 1939. Although it was comprehensively described by Binder<sup>[2]</sup> in 1962 and Hopkin<sup>[3]</sup> in 1963. Binder had initially reported three cases and discussed the characteristic features.<sup>[2]</sup> The author said that the facial appearance at birth resemble darhinencephaly and in addition showed frontal sinus hypoplasia and nasal mucosal atrophy.<sup>[3-5]</sup> McWilliam and Linder Arosen<sup>[6]</sup> had further defined the craniofacial effects of the syndrome. The syndrome has also been discussed in brief by Gorlin *et al.*<sup>[7]</sup>

There has been a great deal of debate regarding the extent of deformity in patients with this syndrome. Some believe, it involves only the nasal region whereas others state that it affects the entire midface. In the present scenario, 3 dimensional scans are being used to assess the extent of the area affected.<sup>[8]</sup> Individuals with this syndrome have characteristic appearance with features such as hypoplasia of the middle third of the face, a broad flat nose, horizontal nostrils, a short

columella, and broad philtrum, a bulging upper lip and a marked groove at nasolabial junction. Palpation during the intra-oral examination reveals the absence or reduction of the bony crest at the base of the nostrils.<sup>[5]</sup> In addition to the facial features, evidence for the presence of cervico-spinal and cranio-spinal abnormalities have also been reported.<sup>[8]</sup>

This condition is thought to have an autosomal recessive inheritance with incomplete penetrance,<sup>[5]</sup> but Gross-Kieselstein *et al.*<sup>[9]</sup> have suggested a dominant mode of inheritance. There are few published reports and studies on Binder's syndrome in English literature. The present paper describes two case reports of maxilla nasal dysplasia; thus, adding a number to the shortlist of reported cases.

#### CASE REPORT

##### Case I

A 19-year-old male patient reported to dental clinics with a chief complaint of facial deformity. On extra-oral examination he presented with a mild Class III skeletal pattern. Mid-face hypoplasia was evident with a reduced fronto-nasal angle reflected in a straight profile. The lips were competent at rest with no obvious facial asymmetry [Figures 1 and 2]. No deviation was noticed on opening or closing of jaws.

Intra-oral examination revealed the absence of permanent left central and lateral incisors (21,22) along with both mandibular canines. The maxillary

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arch exhibited displaced canines and left lateral incisor whereas mild crowding was evident in the mandibular arch. In addition, reverse overjet and over bite of 3-4 mm was seen [Figures 3-5].

On radiological examination, the lateral cephalogram confirmed the clinically apparent Class III skeletal pattern, along with an increased mandibular plane angle and increased lower facial height. Pseudo-mandibular prognathism was also seen due to retrognathic maxilla [Figure 5].

### Case II

A 22-year-old patient reported to the dental clinics with a chief complaint of crowding of his teeth and wanted his teeth to be straightened. Extra-oral examination of the patient showed no facial asymmetry and a Class III skeletal relationship. Mid-face hypoplasia was seen [Figures 6 and 7] along with a collapsed nasal bridge.



Figure 1: AP view of the face (case-I)



Figure 2: Lateral view of the face showing flat nose, maxillary hypoplasia, and reduced Fronto-nasal angle (case-I)



Figure 3: Intra-oral view showing crowding and missing 21, 22 (case-I)



Figure 4: OPG (Orthopantomogram) showing crowding and missing 21, 22 (case-I)

Intra-oral examination revealed mild crowding with Class I molar relationship, edge to edge bite with respect to anteriors, missing maxillary lateral incisors and over retained maxillary right deciduous canine [Figures 8 and 9]. Oro-nasal fistula was observed in the palatal region.

The cephalometric analysis showed a Class III skeletal pattern, increased mandibular plane angle along with increased lower facial height. Since, maxilla was retrognathic pseudomandibular prognathism was seen [Figure 10].

In addition to the above characteristic intra- and extra-oral features of the patient also presented polydactyly [Figure 11].

## DISCUSSION

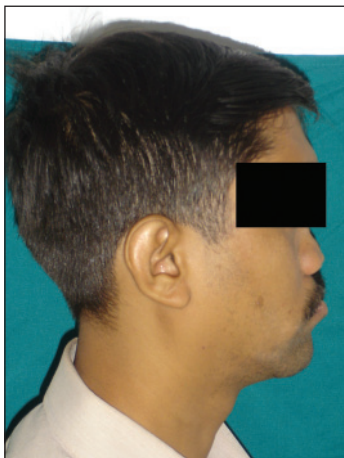
The craniofacial deformity with hypoplasia of middle third of face associated with congenital absence of anterior nasal spine and depression of nasal bones with flattened nasal alae has been



**Figure 5:** Lateral cephalogram showing maxillary hypoplasia, mandibular pseudo-prognathism and reverse overjet, and overbite (case-I)



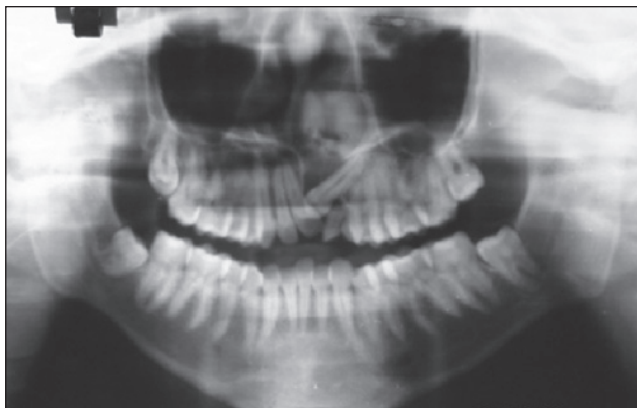
**Figure 6:** AP view of the face (case-II)



**Figure 7:** Lateral view of the face showing flat nose, maxillary hypoplasia, and reduced frontonasal angle (case-II)



**Figure 8:** Intra-oral view showing crowding and missing 21, 22 (case-II)



**Figure 9:** OPG (Orthopantomogram) showing crowding and missing 21, 22 (case-II)



**Figure 10:** Lateral cephalogram showing maxillary hypoplasia, mandibular pseudo prognathism and reverse overjet, and overbite (case-II)

described as Binder's syndrome or Maxillo-nasal dysplasia. The use of terms syndrome and dysplasia is misnomer as has been discussed by Quarrell

*et al.*<sup>[10]</sup> because of varied clinical presentation and because of absence of any reported histopathological description.



**Figure 11:** Polydactyly (case-II)

The etiology of the syndrome remains obscure. Although, Binder had suggested the defect to be of archinecephalic origin, along with disturbance of prosencephalic induction center during embryonic growth.<sup>[2,5]</sup> Noyes<sup>[1]</sup> suggested birth trauma to be one of the causative factors though in five cases reported by Hopkin<sup>[3]</sup> none had any birth trauma. He later hypothesized that the syndrome was result of developmental insult since patients had vertebral abnormalities. Ferguson and Thompson<sup>[11]</sup> had thought of genetic etiology, which was also supported to some extent by others. It was suggested that inheritance could be autosomal recessive with reduced penetrance or it could be multifactorial in heritage. Gorlin *et al.*<sup>[7]</sup> finally emphasized that Binder's syndrome is a non-specific abnormality of nasomaxillary complex and familial examples are a result of complex genetic factors. The syndrome as reported by Binder<sup>[2]</sup> had few characteristic features such as arhinoid face, abnormal position of nasal bones, inter-maxillary hypoplasia with associated malocclusion, reduced or absent anterior nasal spine, atrophy of the nasal mucosa and absence of frontal sinus.

The most noticeable features of this entity are maxillary hypoplasia and flat vertical nose.<sup>[10]</sup> The major skeletal abnormality is small maxilla positioned posteriorly on a short anterior cranial base.<sup>[3,5]</sup> Maxillary hypoplasia leads to relative mandibular prognathism and Angle type III malocclusion.<sup>[10]</sup> Cephalometric studies by various authors show smaller anterior cranial base(n-s), more retrusive nasal bones (s-n-r), maxillary retrognathism (smallers-n-ss angle), smaller maxillary length (sp-pm), and reduced pharyngeal airway dimension (pm-ad3).<sup>[3,5,6]</sup> Malocclusions such as proclination of upper incisors, mandibular prognathism (pseudoprogathism), open bite and crowding are also common.<sup>[5]</sup> Few dental abnormalities such as small central incisors, congenitally absent maxillary incisor, and molars have also been

reported.<sup>[1,3]</sup> Most of the aforementioned characteristic features were observed in our cases of this rare syndrome. In addition to these features, lateral skull radiographs may reveal that the normal crest dividing the floor of the nasal cavity from the anterior aspect of the maxilla is missing and anterior nasal spine is either absent or hypoplastic. Palpation of upper lip and gingival can help in detection of absence of the nasal spine.<sup>[10]</sup> Several authors have also described cervical spine abnormalities. Malformations frequently affected C1 or C2 and included hypoplastic arches or abnormal ossification patterns.<sup>[4,8]</sup>

The treatment of this complex maxilla nasal deformity would mainly require orthodontic and surgical intervention after assessing the degree of this complex maxilla-nasal deformity. LeFort I and II osteotomy with nasal grafting can be carried out for severe cases. Olow-Nordenram and Thilander<sup>[5]</sup> advised postponing definitive orthodontic treatment in individuals with maxilla-nasal dysplasia until growth has stopped, especially in those with severe malocclusion. The treatment should be aimed at improving facial esthetics, relieve crowding, co-ordinate arches and obtain a Class I molar relationship.<sup>[5,12,13]</sup>

## CONCLUSION

Despite the fact that malocclusion is closely related to maxillo-nasal dysplasia traditionally patients with this condition have been treated by plastic surgeons alone. Nevertheless, the presence of a variety of dental and facial deformities necessitates an inter-disciplinary approach for management of these cases, along with a proper treatment planning. This is very essential since milder cases of Binder's syndrome can be treated by orthodontic treatment alone unlike the more complex cases requiring surgery.

## REFERENCES

1. Noyes FB. Case report. *Angle Orthod* 1939;9:160-5.
2. Binder KH. Dysostosis maxilla-nasalis, einarhinen cephaler Missbildungs komplex. *Deutsche Zahnartzliche Zeitschrift* 1962;6:438. Cited in: Dyer FM, Willmot DR. Maxillo-nasal dysplasia, Binder's syndrome: Review of the literature and case report. *J Orthod* 2002;29:15-21.
3. Hopkin GB. Hypoplasia of the middle third of the face associated with congenital absence of the anterior nasal spine, depression of the nasal bones, and angle class III malocclusion. *Br J Plast Surg* 1963;16:146-53.
4. Resche F, Tessier P, Delaire J, Tulasne JF. Craniospinal and cervicospinal malformations associated with maxillonasal dysostosis (Bindersyndrome). *Head Neck Surg* 1980;3:123-31.

5. Olow-Nordenram M, Thilander B. The craniofacial morphology in persons with maxillonasal dysplasia (Bindersyndrome). A longitudinal cephalometric study of orthodontically treated children. *Am J Orthod Dentofacial Orthop* 1989;95:148-58.
6. McWilliam J. Hypoplasia of the middle third of the face-a morphological study. *Angle Orthod* 1976;46:260-7.
7. GorlinR, PindborgJJ, CohenMJr. Maxillonasal dysplasia (Bindersyndrome). *Syndromes of the Head and Neck*. 2<sup>nd</sup>ed . NewYork: McGrawHill;1976.
8. Kau CH, Hunter LM, Hingston EJ. A different look: 3-dimensional facial imaging of a child with Binder syndrome. *Am J Orthod Dentofacial Orthop* 2007;132:704-9.
9. Gross-Kieselstein E, Har-Even Y, Navon P, Branski D. Familial variant of maxillonasal dysplasia? *J Craniofac Genet Dev Biol* 1986;6:331-4.
10. Quarrell OW, Koch M, Hughes HE. Maxillonasal dysplasia (Binder'ssyndrome) *J Med Genet* 1990;27:384-7.
11. Ferguson JW, ThompsonRP. Maxillonasal dysostosis (Bindersyndrome); are view of the literature and case reports. *Eur J Orthod* 1985;7:145-8.
12. Jackson IT, Moos KF, Sharpe DT. Total surgical management of Binder's syndrome. *Ann Plast Surg* 1981;7:25-34.
13. Holmström H. Surgical correction of the nose and midface in maxillonasal dysplasia (Binder's syndrome). *Plast Reconstr Surg* 1986;78:568-80.

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