

Binders Syndrome: A Review

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Abstract

Binder's syndrome is a rare congenital condition characterized by a retruded mid-face with an extremely flat nose due to an underdevelopment of the mid-facial skeleton. It primarily affects the anterior part of the maxilla and nasal complex. Hence the condition is also known as Maxillonasal dysplasia. This paper aims to put forth the etiopathogenesis, clinical presentation & the management of Binder's syndrome.

Keywords: Binder's syndrome, congenital, Maxillonasal.

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INTRODUCTION

Maxillonasal dysplasia was first reported by Noyes in 1939 [1]. Von Binder in 1962 coined this syndrome as "maxillonasal dysostosis" [2]. He reported the six most salient features of the syndrome. They include arhinoid face, intermaxillary hypoplasia associated with malocclusion, abnormal position of the nasal bones, nasal mucosa atrophy, anteriornasal spine agenesis and a lack of frontal sinuses [3, 4].

The exact etiology of the syndrome is not clear but Binder suggested that there is disturbance of the prosencephalic induction center during embryonic development [5]. The induction process for both the prosencephalic area and vertebrae is common, accounting for increased vertebral anomalies associated with the syndrome [6].

Gorlin emphasized that Binder's syndrome is an abnormality of the nasomaxillary complex and familial examples are as a result of complex genetic factors. Although most of the cases involve only the nasomaxillary complex, a variety of other anomalies have been recorded like cervical vertebrae, skeletal defects, cardiac anomalies, orofacial clefting, strabismus and mental retardation. Other factors like birth trauma and familial history have also been reported. Gorlin suggested that familial factors involve

complex genetic factors, similar to those involved in producing a malocclusion [7].

Clinical Features

Binder syndrome is characterized by the absence of nasofrontal angle with the nose being hypoplastic having a small tip. The nostrils are usually half moon shaped and the upper lip is convex with a high arched palate. There is dental malocclusion with an anterior overbite. There will be mild hypertelorism. The frontal sinuses are usually hypoplastic / absent. There will be anomalies of the cervical spine, strabismus and there could be mild mental retardation [3, 8].

Several studies have investigated the craniofacial morphology of patients with Binder syndrome by using lateral cephalometric radiographs. Few studies compared the lateral cephalometric radiographs of 14 patients with Binder syndrome who had no prior surgical or orthodontic treatment with published age- and sexmatched norms. They found that the anteroposterior dimensions of the anterior cranial base and the maxilla were reduced, with most patients having a Class III skeletal relationship. Despite the prominence of the mandibular incisors, both overjet and overbite were within normal ranges [9].

In addition to the facial features of Binder syndrome, several authors presented evidence of cervical spine abnormalities. A study involved radiographic examinations of 43 patients with maxillonasal dysplasia and found that 44% had malformations of the cervical vertebrae. The atlas and the axis are most frequently affected. In addition, 6 of the patients in the series had dysplasia of the vertebral bodies related to persistence of the chorda dorsalis. The incidence or the severity of the malformations did not appear to be related to the degree of maxillonasal dysplasia [10, 11]. In a study of 28 cases of maxillonasal dysostosis, it was found that 54% had craniospinal and cervicospinal malformations [12].

Literature shows that most Binder's syndrome patients suffer from microdontia of the central upper incisors; a lack of lateral incisors has also been observed, but rarely [13]. The differential diagnosis of this rare disease could be Warfarin embryopathy, Down's syndrome, Apert syndrome, Stickler syndrome, Keutel syndrome and Acrodysostosis [13]. These syndromes can frequently be misdiagnosed during fetal ultra sonogram (USG) examinations. Binder's syndrome can be easily diagnosed by using 2D and 3D ultrasounds, beginning with the 21st week of pregnancy [3]. It should also be considered that Down syndrome is often associated with Binder's syndrome. It has been reported that 12% of patients with trisomy 21 lack a nasal bone [14].

Etiology

The exact etiology of the syndrome is not clear but Binder suggested that there is disturbance of the prosencephalic induction center during embryonic development [5]. Although not substantiated in literature, birth trauma has also been suggested [5]. Ferguson and Thompson have put forward the possibility of a family history [15]. Binder believed that patients with this syndrome have a mild form of arrhinencephaly, but they have no olfactory anomalies to support the hypothesis [11].

The literature survey, according to Nedev, has not disproved the possibility of a genetic etiology, although it may not be the actual cause of the syndrome. The features and degree of the abnormality depend on the time of exposure to harmful teratogenic factors [14].

Hopkin, in 1963, proposed that Binder's syndrome was the result of a developmental abnormality [11]. In 1987, Narcy *et al.*, reported a case of Binder's syndrome in association with esophageal achalasia and abnormal autonomic reflexes, suggesting that the phenotype could be the result of an abnormal migration of neural crest cells [16]. There was evidence that vitamin K-deficiency, during human pregnancy, caused by some chemicals such as lithium and ethanol or the therapeutic use of warfarin or phenytoin, could be

a causative factor. Pregnancy histories of three cases of Binder's syndrome were reported earlier, associated with warfarin and phenytoin exposure and alcohol abuse [17].

Several authors in the past have attempted to determine the etiology of Binders syndrome. Olow-Nordenram and Valentin studied 60 families to see whether there was an autosomal recessive inheritance pattern. They concluded that the syndrome either is inherited as an autosomal recessive trait with incomplete penetrance or a threshold characteristic with a genetically multi-factorial background [18]. In contrast, Gross-Kieselstein *et al.*, suggested a dominant mode of inheritance after examining an affected mother and daughter [19].

Management

The management of these patients depends on the level of complexity because of variations in the midface discrepancy and the occlusal relationship. Some lesser-affected patients can be managed by using orthodontic camouflage alone, whereas others require multi-disciplinary input.

Surgical treatment for Binder's syndrome is primarily aimed at nasal dorsum reconstruction – elevation of the tip of the nose and lengthening of the nasal dorsum [4]. The convexity of face at the tip of the nose (the glabella-pronasale-pogonion angle) improves after surgery. The type of surgical procedure depends on the facial malformation – in case of a depressed nasal dorsum, an L-shaped bone graft is the favored procedure, while repositioning the septum is indicated if the upper section of the nose is found normal. A cartilage graft requires secondary correction more rarely than a bone graft. As much as 28% of the volume of a transplant is resorbed [4].

Osteotomy and grafts in the form of bone or cartilage are frequently used in the management of Binder syndrome. Grafts, when limited to the nasal sill, do not address the “dish face” deformity characteristic of the syndrome. LeFort II osteotomy, in addition to correcting the midface deficiency, can overcorrect the occlusal relationship if good dental camouflage is achieved, making a LeFort I osteotomy or orthodontic therapy necessary. It has been suggested that mild cases of Binder syndrome, without severe malocclusion, can be managed by using a cartilage graft around the nostril sill, dorsum, and the pyriform aperture, correcting the paranasal and infraorbital deformity without osteotomy [20]. In more severe cases, however, a LeFort II osteotomy can correct the midface deficiency and the Class III occlusion seen in patients with Binder syndrome, but it does not always fully address the nasal flatness.

The overall goals were to augment the skeletal deficiencies of the midface and begin the soft-tissue

expansion process as early as possible. In a young patient, sequential lengthening procedures of the dorsum and the columella are beneficial. Paranasal and midfacial augmentation is reserved until midfacial growth is near complete, when patients are in their midteen years [21].

A case of Binder syndrome was diagnosed at 21 weeks of gestation using two-dimensional and three-dimensional ultrasound. The first sign of any abnormality was a flattened fetal nose demonstrated in the mid-sagittal plane. Further ultrasound imaging showed the virtual absence of naso-frontal angle giving the impression of a flat forehead and small fetal nose. Suspected mild hypertelorism is also seen in the transverse and coronal planes [22].

Due to the graft's resorption or a lack of bone reorganization after surgery (mainly in the nose tip projection) relapse is observed. Therefore revision of the surgery and a secondary bone or cartilage graft is necessary in one fourth of the patients. Bone and cartilage grafts are usually carried out from 14 years of age, and precede osteotomy of the maxilla and/or nose (performed at the age of 18 or later). If the appearance of the nose after surgery is not sufficiently satisfactory, nasolabial flaps may be used to resurface the lining [23, 24].

CONCLUSION

Binder's syndrome is a rare congenital malformation that mainly affects facial features. There has been much debate about the etiopathology and management options pertaining to Binder deformity. Many authors believe that it is restricted to the nasal area, whereas others state that it affects the entire midface. Three-dimensional scans can give a precise detail regarding the extent of the area affected in a patient with Binder syndrome. The information from the scan will enable enhanced planning of any future surgical intervention to create an ideal facial balance and allow the patient to be better informed about his options. It serves as a useful 3-dimensional soft-tissue template from which a treatment plan can be made.

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