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REVIEW ARTICLE

Unravelling Maxilla-nasal Dysplasia: A Deep Dive into Binder Syndrome

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Abstract

Binder syndrome, also referred to nasomaxillary hypoplasia or maxillonasal dysplasia, is a rare congenital craniofacial condition characterized by underdevelopment of the maxilla and nasal structures. Although the exact cause of Binder syndrome remains unclear, it is thought to result from disturbances during embryonic development. Diagnosis typically relies on both clinical examination and radiographic imaging. Treatment often involves a multidisciplinary approach, including surgical correction of facial deformities and orthodontic interventions to improve facial aesthetics and function. This article presents a comprehensive narrative review of the etiology, clinical manifestations, and management of Binder syndrome.

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Introduction

Binder-type nasomaxillary dysplasia, a rare developmental anomaly present at birth, has been documented in medical literature since the late 19th century. In 1882, Zuckerkandl[1] was one of the pioneering individuals to identify an anomaly characterized by the absence of the anterior nasal spine. Dr. Noyes further elucidated the essential features of this condition in a single patient in 1939.[2] However, it wasn't until 1962 that Dr. Von Binder formally identified and defined this condition as a distinct clinical entity in a comprehensive report detailing three affected children.[3] Binder termed the syndrome dysostosis maxilla-nasalis and outlined a constellation of symptoms, including midfacial hypoplasia marked by a flat nose, shortened columella, flattened tip and alar wings, sharp nasolabial angles, half-moon-shaped nostrils, and absence of nasofrontal (glabellar) angles. Additionally, individuals with this syndrome often exhibit midmaxillary hypoplasia and a concave midfacial profile, colloquially referred to as a "dish face".

Etiology

The precise etiology of Binder syndrome remains elusive, but it is hypothesized to result from a disruption in the prosencephalic induction center during embryonic development. This congenital anomaly, with an estimated incidence of approximately 1 in 10,000 infants, affects both genders equally but is likely underdiagnosed, particularly among specific ethnic groups.[4] Holmstrom identified a hereditary component in 16% of 50 patients diagnosed with Binder syndrome, suggesting

a potential inheritance pattern with an autosomal recessive trait displaying incomplete penetrance.[5] In addition to craniofacial dysmorphology, more than 40 percent of individuals with Binder syndrome present with vertebral anomalies that emerge during the third month of gestation, coinciding with nasal development.[6,7] The shared induction process for both the prosencephalic region and vertebrae likely contributes to the heightened prevalence of vertebral abnormalities observed in conjunction with Binder syndrome.[8]

Clinical Features

In maxillonasal dysostosis, notable findings encompass a deficiency in premaxillary development alongside deformities affecting the nasal framework and its associated soft tissues. Typically, the nasal spine is either absent or rudimentary, and the nasal floor converges anteriorly with the bony labial plate overlaying the upper incisors. This contrasts with the typical cranial anatomy, where the anterior extent of the nasal floor is distinctly delineated.[9]

Binder documented three cases and identified six characteristic features.[3] The primary facial characteristic of Binder's syndrome is the arhinoid facial appearance, which manifests as a flat and vertically oriented nose due to hypoplasia of the nasomaxillary structures.[10,11] Additionally, the syndrome presents with other distinctive facial features such as maxillary hypoplasia, resulting in a hypoplastic midface profile and severe malocclusion. Furthermore, individuals with the syndrome may exhibit diminished nasal spine, atrophy of the nasal mucosa, and abnormal positioning of the nasal bones. Occasionally, the absence of frontal sinuses may also be noted, although it is not a consistent finding.[11,12]

In Binder's syndrome, alongside the characteristic flattened nose, individuals commonly present with a concave facial profile,[13] often accompanied by a decrease in the naso-frontal angle.[14] Hypertelorism, characterized by an increased distance between the eyes, may also be evident.[12] The junction between columnella and the upper lip is typically short and retracted.[5,10] Skeletal underdevelopment in areas such as the nasal floor, prenasal fossae, and piriform aperture is typically bilateral.[13]

Individuals with Binder's syndrome often exhibit flatness around the nose and cheek area (perialar flatness)[12] and Maxillary hypoplasia contributes to a convex upper lip, leading to an acute nasolabial angle. According to Holmstroem *et al*., this angle ranges from 76–88° in Binder's syndrome, contrasting with the normal range of 103–117° reported in a cephalometric analysis by Segner and Hasund's. The upper lip's convexity, the presence of a deep fold or fossa between the nose and upper lip, and a flattened philtrum result in an acute angle.[5,10-12,15]

Skeletal Features

Skeletal alterations in Binder's syndrome significantly influence patients' facial characteristics. One notable change is the absence of the anterior crest, which typically divides the floor of the nasal cavity, alongside a hypoplastic or absent anterior nasal spine. These factors contribute to a flattened nasal profile lacking in nasal prominence.10,16 Additionally, researchers have noted the presence of a scaphoid depression within the anterior floor of the nasal cavity. While clinically observable, this depression is not typically visualized on radiographs.[10] However, the nasal bones maintain a normal length in Binder's syndrome.[13]

A prominent finding consistently highlighted in the medical literature is the prevalent hypoplasia or absence of frontal sinuses, manifesting in approximately 40–50% of cases. Additionally, there's a distinct anatomical feature where the anterior cranial base appears notably shortened, resulting in a reduced sella-nasion distance. This structural anomaly coincides with a peculiar positioning of the maxilla towards the posterior aspect. This positioning subsequently leads to specific clinical manifestations such as Class III malocclusion and relative prognathism.[10-12] Furthermore, an atypical convexity of the maxilla is evident in individuals with Binder's syndrome.[16] Additionally, a few patients of Binder's syndrome may also exhibit a cleft palate.[10] Moreover, beyond cranial skeletal alterations, approximately 44.2% of individuals with Binder's syndrome demonstrate abnormalities in the cervical spine. These anomalies emerge during the developmental phase of these structures

in the 3rd month of pregnancy, coinciding with the formation of the nose. This emphasizes the considerable influence of environmental factors in shaping this deformity. Notably, the first and second cervical vertebrae are frequently affected, with their hypoplastic arches displaying the ossification patterns are abnormal, indicating a complex interplay of genetic and environmental factors in the pathogenesis of Binder's syndrome.[10,12,17]

Dental Abnormalities

Delaire et al. noted a prevalent occurrence of microdontia affecting the central upper incisors among individuals with Binder's syndrome, and an absence of lateral incisors has been reported but less frequently.10,16 Dental anomalies are frequently encountered, predominantly arising as a consequence of malocclusion, and often manifest as dental crowding and alterations in tooth positioning.10 Additionally, a case of amelogenesis imperfecta was documented in one patient, further underscoring the spectrum of dental abnormalities associated with this syndrome.5

Management

Treatment modalities for patients with Binder's syndrome vary significantly based on the severity of the deformity and the time at which it is diagnosed.

Orthopaedic Correction

Traditional cephalometric methods may not accurately analyze facial morphology in Binder's syndrome due to the absence or displacement of key measurement points such as the anterior nasal spine, nasion, and subspinale (point A). Delaire and colleagues utilized the "architectural and structural craniofacial analysis (lateral view)" developed by Delaire to characterize abnormalities in thirty-four patients with Binder's syndrome.[6] Their observations emphasized the necessity of initiating orthodontic therapy promptly, focusing primarily on maxillary advancement achieved through heavy extra-oral postero-anterior traction applied via an orthopaedic face mask. This proactive approach aims to prevent the need for later surgical maxillary advancement, with surgical interventions limited for nasal corrections.[18] Following the completion of orthodontic-orthopedic treatment, patients typically attained favorable functional and aesthetic outcomes, often complemented by straightforward plastic surgery interventions targeting nasal aesthetics.[19]

Surgical Management

Surgery of Binder's syndrome is typically conducted by Plastic surgeons and primarily focuses on reconstructing the nasal dorsum, including elevating the nasal tip and elongating the nasal dorsum.[12,20] Following the surgery, there is an increase in the nasolabial angle to the desired 100–106° from 76–88°, although a slight tendency for relapse may occur.[5,13] Additionally, the convexity of the face at the nasal tip, known as the glabella-pronasale-pogonion angle, typically improves post-surgery.

The choice of surgical approach is determined by the specific facial malformation observed. In cases of a depressed nasal dorsum, the favored procedure involves an L-shaped bone graft, whereas septum repositioning is recommended when the upper portion of the nose is unaffected. Cartilage grafts typically necessitate secondary correction less frequently compared to bone grafts. Approximately 28% of the transplant volume is resorbed over time. Rune *et al*. reported an average decrease of 13 mm in nose length from the immediate post-surgery measurement. The majority of resorption occurs within the initial two years following the procedure. It is plausible that the short arm of an L-shaped bone graft is substituted with fibrous tissue. Consequently, relapse may occur due to graft resorption or inadequate bone reorganization, particularly in the nasal tip projection. As a result, approximately one-fourth of patients may require revision surgery and a grafting of secondary bone or cartilage.[5,13,20] Bone and cartilage graft procedures are typically initiated from the age of 14, preceding maxillary and/or nasal osteotomy, typically performed at 18 years or later.[12] In cases where the postoperative appearance of the nose is unsatisfactory, nasolabial flaps may be employed to resurface the lining.[21] The most severe cases of malocclusion may necessitate Le Fort I or Le Fort II

osteotomy, typically performed in adult patients in conjunction with grafting of nose.

Orthodontic Treatment

Following surgical correction of nasal and maxillary abnormalities in Binder's syndrome, an essential step towards achieving optimal aesthetics and function involves orthodontic treatment. The approach to orthodontic treatment is contingent upon the severity of the malocclusion. In cases with mild malocclusion and compensatory effects within the dental arches, orthodontic intervention may not be deemed necessary.[11,12,22] However, the majority of Binder's syndrome patients exhibit Class III malocclusion, characterized by prominent mandibular incisors.[23] Proclination of upper incisors often serves as a compensatory mechanism for a retrognathic maxilla, masking the malocclusion. In mild cases, orthodontic therapy may involve the use of fixed appliances with Class III elastics facilitating proper alignment, overjet, and overbite correction.[11]

For more severe malocclusions, such as those requiring Le Fort I or Le Fort II osteotomy, a comprehensive orthodontic approach is essential. Fixed orthodontic treatment is initiated to decompensate the malocclusion, thereby preparing the patient for surgical intervention and ensuring optimal outcomes.[10,12] It is important to initiate orthodontic treatment once growth has been completed, ensuring a stable foundation for long-term results.[11] Through a collaborative effort between surgeons and orthodontists, individuals with Binder's syndrome can achieve functional improvements with enhanced facial aesthetics and overall quality of life.

Conclusion

Binder's syndrome presents a complex array of craniofacial abnormalities that necessitate a multidisciplinary approach for effective management. Early diagnosis is indeed critical, particularly for planning growth interventions such as the protraction of the maxilla. Surgical correction of nasal and maxillary deformities with subsequent orthodontic treatment enhances aesthetic outcomes and functional outcomes. Collaborative efforts between surgeons and orthodontists are paramount in delivering effective treatment strategies that enhance facial aesthetics and promote functional occlusion with long-term stability. Further research and advancements in treatment modalities are essential for continually improving the medical management of Binder's syndrome and enhancing the quality of life for affected individuals.

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