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A Review of Congenital Syngnathia In Children from a Clinical Point of View and the Effect of Treatment on Them

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Abstract

Aim: *The purpose of this review is to examine cases with maxillomandibular fusion and treatment methods and the effect on patients' recovery.*

Methods: *In order to conduct this review, original articles in English were collected from PubMed and Google Scholar databases. Articles were reviewed from 1997 to 2023. Unpublished studies and studies in non-English language were not reviewed. In this study, the gender and age of the patients were not classification criteria, and all bony and mucosal fusions of the maxilla and mandible with or without ankylosis of the jaw joint and connection to the zygoma were considered.*

Results: *The studied cases had two types of Dawson's classification. Type 1 is mucosal fusion and type 2 is bone fusion. Cases that had bone fusion and the involvement is more severe have respiratory and nutritional problems and the risk of intubation during surgery is raised for them. Tissue and bone separation and establishment of proper airway and nutrition are part of the treatment goals of this disease. Placing the distractor device in the osteotomized place and daily activation of the device is continued until the desired length of the mandible is reached, and then after the separation of the device, a long follow-up phase of the patients and physical therapy is done to stabilize the obtained result. Gender and age have no effect on the rate of disease recurrence, and patients with syndromes such as Horner, Aglossia-Adactylia, etc. show variable results.*

Conclusion: *Maxillomandibular syngnathia is a rare disease of unknown etiology. which can range from mucosal fusion to complete bony fusion of the maxilla, mandible and zygomatic arch and has different classifications that can cause many problems for patients depending on the severity of the disease. Safe airway access through endotracheal intubation or tracheostomy and alimentary access through nasal feeding tube or gastrostomy and intra- and extra-oral access to the fusion area and then isolation and osteotomy of the area and deployment of distractor devices and most importantly long follow-up and physiotherapy can save the lives of these children.*

Keywords: Syngnathia; Vertical Distraction; Maxillomandibular Fusion; Tmj Ankylosis; Osteotomy

Introduction

The term syngnathia is defined as congenital fusion or adhesion between the maxilla and mandible. This connection may be mucosa alone (synechiae) or bony connection between the maxilla and mandible or in combination with zygoma and condyle adhesion (complete synostosis) [1]. From a genetic point of view, the most reliable etiopathogenesis is the interaction of FOXC1, which is a regulator for the jaw, with FGF8, which can affect the severity of the syngnathia phenotype [2]. Syngnathia can be alone or together with syndromes such as popliteal pterygium, oromandibular limb hypogenesis, Dobrow, Horner syndrome, Vander wound, aglossia-adactylia, Nager syndrome, lateral cleft palate with alveolar mucosa adhesion [3,4]. With all the information and evidence obtained, the definitive etiology of this disease is not known. Some researchers suggest that causes such as retention of the buccopharyngeal membrane, obstruction of the amniotic cords in the developing region of the first pharyngeal arch abnormal vascularization of the stapedial artery, rapid absence or failure of migration of neural crest cells in an incomplete separation of the first arch Branchial to maxilla and mandible, lack of tongue protrusion during development and trauma, and the presence of some teratogens such as Meclozine, high doses of vitamin A are considered to interfere with the development of the mandible, although it has not been proven yet [5-7]. The first case was reported by [8]. in which he described unilateral facial atrophy with congenital TMJ ankylosis in an infant. Dawson in 1997 presented a classification that provided the first classification of congenital osseous syngnathia with functional and therapeutic outcomes. This classification includes type 1 (simple syngnathia): bony fusion of the mandible and maxilla or zygoma in the absence of other congenital anomalies of the head and neck, type 2a (complex syngnathia): bony fusion of the maxilla and mandible and absence of the tongue, type 2b (complex syngnathia). Bone fusion of maxilla and man-

dible and zygoma with agenesis and proximal hypoplasia of the mandible [9]. Laster in 2001 added alveolar ridge fusion with or without congenital deformities and accompanying cleft palate and TMJ ankylosis to this classification [6]. After them Tauro in 2012, Olusanya in 2020, and Kumar in 2021 unilateral or bilateral fusion and accurate diagnosis of micrognathia and polyhydramnios's from examinations and ultrasound and diagnosis of closed or open mouth and MRI findings added to studies in this field. although still after years, Dawson's classification is a more reliable and easier reference for this classification [10-14].

There are several reasons why syngnathia is considered an important issue in infant development, and several medical teams are working to help resolve this problem. This congenital problem can cause speech and beauty defects in mild cases and breathing and feeding problems in more severe cases [15]. The main problem of these patients will be maintaining a stable airway, although the placement of a nasal feeding tube is not an answer in the long term and is considered a temporary solution due to the risk of aspiration and pneumonia respiratory and digestive infections. The use of endotracheal intubation, respiratory aids such as oxygen masks, positive pressure ventilation, and as a last resort tracheostomy can guarantee the baby's survival to some extent. Although in milder cases, a person with tolerable respiratory and nutritional deficiency will advance to a young age. However, cases of death due to late action to establish a secure airway or infection have also been reported [16]. Treatment: Several methods are mentioned for the treatment of mucosal and bony fusion of two jaws. For a more detailed examination, all patients undergo a 3D face scan and 3D reconstruction in sagittal, axial, and coronal views. Because this information will be helpful for a detailed examination of the fusion, its type, and connection location. Surgical methods include freeing the mucous and bony connections, performing osteotomies at the connection points, and placing distractor devices at the opening of the seams, among other effective measures [17].

What is discussed in this review article is the treatment methods of patients with syngnathia problem and the investigation of the problems resolution and recovery rate from 1997 until today. And considering the importance of emergency solutions to the problem despite the rarity of the disease, this article was written.

Methods

To conduct this review, original articles in English were collected from PubMed and Google Scholar databases. Articles were reviewed from 1997 to 2023. Unpublished studies and studies in non-English language were not reviewed. In this study, the gender and age of the patients were not classification criteria, and all bony and mucosal fusions of the maxilla and mandible with or without ankylosis of the jaw joint and connection to the zygoma were examined. The keywords syngnathia, jaw synostosis, gingival synechiae, maxillomandibular union, congenital TMJ ankylosis,and maxillomandibular fusion were used to search for articles. Nutritional considerations of patients, airway preservation and surgical approach, and recurrence or non-recurrence were investigated.

Results

In the review of the articles found in the databases and after checking the entry and exit criteria in the article, all the remaining cases with syngnathia problems from 1997 to 2023 were studied. Most of the examined cases were type 1 of Dawson's classification and the rest were type 2. Although the patients who were in the syndromic field such as Dobrow syndrome, Horner syndrome, Aglossia-adactylia with different appearances such as lower ears,and tongue agenesis and tongue hypoplasia had syngnathia problems. What can be seen in this study was the lack of communication or, more correctly, less communication in the relationship between kinship and the occurrence of this problem. And only a limited number of cases had consanguineous parents. Another point to be taken from different cases was the issue of fast follow-up treatment and prevention of TMJ joint ankylosis because the importance of the jaw joint as a growth center in the face is not hidden from anyone [18]. The research also revealed that the average age and sex of the patients did not affect on their surgical results, but the issue that revealed a significant difference in the rate of recovery and progress of the treatment was the degree of involvement of the upper airway and access to the feeding route in infants. For patients who had wider bone connections, especially in the area of the zygoma arch to the tmj joint, this caused a decrease in mouth opening even after the mucosal fusion between the two jaws was removed, making the field for endotracheal intubation more complicated, and the possibility of respiratory obstruction and aspiration and occurrence It increased lung infections, including pneumonia. The lack of complete access to a safe airway and the need for breathing aids such as oxygen masks and ventilators faced anesthesiologists with a great challenge in anesthetizing these children. As a result, in some cases, the invasive way of tracheostomy was inevitably chosen [19]. In a similar case, which was performed in Iran in 2021 at Bahonar Hospital, Kerman, on a 23-days-old baby of Afghanistan origin,the complete fusion of the maxilla and mandible and the connection of the zygomatic arch to the jaw joint was also the chosen way of tracheostomy, which is a safer and more reliable way to breathe. The patient was diagnosed. The use of an NG tube was considered for feeding these patients, although this method increases the risk of aspiration, and regurgitation and pneumonia, it can be a suitable method for feeding patients, which in most cases and the case performed in Bahonar Hospital, Kerman was also used. Although some studies have proposed and used gastrostomy as a reliable and safe method, considering the need for surgery in other parts of the body, it may be opposed by families [19,20]. Another variable that was investigated in cases with syngnathia was different surgical approaches. Intraoral and extraoral access are two different access methods. The advantage of the intraoral method is fewer scars, hidden from sight, less chance of nerve damage, especially facial nerve, and loss of facial animation on the same side. But in some cases, especially complete bone fusion, especially in the area of the jaw joint and zygomatic arch, wider access and more dissection of tissue with submandibular or periauricular incision, which was also performed in a similar case in Iran by the maxillofacial surgery team. It was optional, it is required, in which case the extraoral method is considered an advantage. On the other hand, using the distractor vertically between the mandible and the maxilla, and the cheek and placing the screws in the mandible from the skin increases the accuracy of the work,

and the problem of the patient's skin irritation was solved by placing a silicone sheet or a plastic protector [21,22].

What is always taken as a successful result of surgery is the establishment or return of the patient to the correct function. In this rare case, the ability to be separated from the ventilator after the treatment period is over, the possibility of oral feeding without problems, and the prevention of recurrence due to the inherent adhesive property of the tissues and their genetic program are discussed. The correct activation of the distractor device after the latency time has passed and the activation of the device 0.5 mm twice a day and then the consolidation time and finally the lengthening of the mandible up to 11 mm or less and more and the complete separation of the connections and then placing the guard or splint in the period of the patient's inability to function until a certain time is a way to reduce recurrence and accelerate recovery. But what is important as a key point in the treatment is the long follow-up of the patients and complete physiotherapy, otherwise, all the completed steps will return and the patient will return to reduced function, like the case in Italy due to migration and return of the patient. Afghanistan was left out of the follow-up cycle [8]. Recurrence occurred mostly in cases that were type 2 of Dawson's classification

Discussion

Surgical teams and researchers are trying to forcefully control syngnathia because it can cause respiratory and nutritional problems. TMJ ankylosis should also be prevented as much as possible. Treatment protocols should be based on three important issues, i.e. preparing CT scan images and 3D reconstruction to check the severity of the problem, checking the safe airway for surgery, and long follow-up after treatment. The goal of surgery for children with double jaw fusion problems is to create a safe path for breathing and feeding and to help them grow normally, although this goal may not be fully realized in patients who are in a syndromic field [17,5]. It is better that the performed CT scan has an interpretation part of the facial vessels, because the presence of vascular anomalies, especially arteries in the involved area in the face and neck, can cause a big problem during surgery. Although different classifications were presented over the years, Dawson's classification is still a simple and understandable solution to categorize these patients. What can be seen from the examination of the cases is that type 1 is associated with mucosal involvement and type 2 is associated with bone involvement. And the premolar canine area in type 1 and the zygoma ramus area in type 2 are among the most commonly affected areas. Although the exact pathogenesis of syngnathia is not yet known, the remaining buccopharyngeal membrane, failure of migration of neural crests, incomplete separation of the branchial arch, which can be caused by the use of drugs, vitamin A in large amounts by the mother during pregnancy, are the causes of this problem. Although the socio-economic conditions, the cultural status of families ,and access to medical facilities do not determine the length of life of these children, they can be effective factors [6-8].

Conclusion

Maxillomandibular syngnathia is a rare disease of unknown etiology. which can range from mucosal fusion to complete bony fusion of the maxilla, mandible, and zygomatic arch and has different classifications that can cause many problems for patients depending on the severity of the disease. Safe airway access through endotracheal intubation or tracheostomy and alimentary access through a nasal feeding tube or gastrostomy and intra- and extra-oral access to the fusion area and then isolation and osteotomy of the area and deployment of distractor devices and most importantly long follow-up and physiotherapy can save the lives of these children.

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