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Preventive and Rehabilitative Treatments in *Midline Facial Cleft* and *Atrial Septal Defect*: Case Report

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Abstract: ***Embryogenesis of the craniofacial region is extremely complex. Significant demands are placed on the coordination of cell separation, migration, and interaction during a brief 4-week period. Any mishap in this intricate program can lead to disastrous consequences and play a role in the etiopathogenesis of craniofacial clefts. Management of patients with congenital or acquired defects of the palate, enabling a connection between the oral cavity and the nose and/or the maxillary sinus, is a challenge to clinicians. Prosthetic rehabilitation strategy such as by using acrylic resin obturator has been used to improve patients' aesthetic and speech, and restore their masticatory functions including their swallowing ability. The atrial septal defect is characterized by a defect in the interatrial septum. Cardiac surgeons have to refer surgical patients to a dentist for oral prophylaxis management and to treat any oral infection before conducting a cardiac surgery to prevent infective endocarditis. Dental treatment of patients with atrial septal defect is prioritized in preventive care.***

Keywords: Atrial septal defect, craniofacial cleft, obturator, preventive care

1. Introduction

Human facial development begins at approximately four weeks post conception, development begins at approximately four weeks post conception. It consists of five processes, which surround the early oral cavity or stomodeum. Those are frontonasal process, a pair of maxillary processes of the first pharyngeal arch, and a pair of mandibular processes of the first pharyngeal arch.[1] This extremely complex process is called embryogenesis of the craniofacial region. Significant demands are placed on the coordination of cell separation, migration, and interaction during a brief 4-week period. The proper amount of tissue must be present at an exact moment in the correct three-dimensional relationship for normal craniofacial development to occur. Any mishap in this intricate program can lead to disastrous consequences. Evidence from animal and clinical studies supports a multifactorial etiology. Factors such as infection with influenza A_2 virus, infestation with toxoplasmosis protozoan, maternal metabolic disorders, and exposure to teratogenic compounds, including anticonvulsants, antimetabolic and alkylating agents, steroids, and tranquilizers, are believed to play a role in the etiopathogenesis of craniofacial clefts.[2]

The incidence of craniofacial clefts is estimated between 1.4 and 4.9/100,000 live births.[3][4] Although two genders are being affected by craniofacial clefts, girls are more commonly affected.[5] In Tessier Classification, clefts 0–14 radiate around the orbital bone rims [Figure 1].[2] Tessier number 0 and number 3 are seen to be the most commonly reported cleft types in both genders in the literature.[4] Tessier number 0 involves cleft in the median craniofacial or midline facial and is usually accompanied by hypertelorism. Hypertelorism is an abnormally widened nose with separation of the nostrils, and an occipital encephalocele.[5][6]

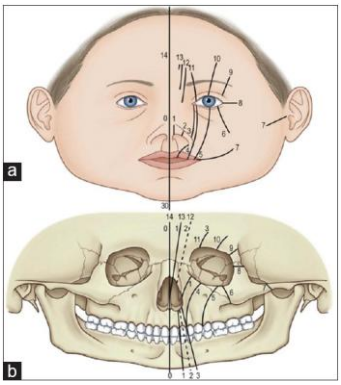

Figure 1: Tessier classification of craniofacial clefts. A. Path of various clefts on the face.
B. Location of the clefts on the facial skeleton. (Courtesy Dr. Paul Tessier)[2]

Management of patients with congenital or acquired defects of the palate, enabling a connection between the oral cavity and the nose and/or the maxillary sinus, is a challenge to clinicians.[7] Prosthetic rehabilitation strategy such as by using acrylic resin obturator has been used to improve patients' aesthetic and speech, and restore their masticatory functions including their swallowing ability. The basic design of obturator prostheses uses the available tooth and bearing tissue to achieve maximum retention and stability. The primary goals of the obturator prosthesis are to preserve the remaining teeth and tissue and provide comfort, function, and aesthetics to the patients. The goals of prosthetic rehabilitation are to allow adequate deglutition and articulation, possible support of the orbital contents to prevent enophthalmos and diplopia, soft tissues support to restore midfacial contour, and acceptable aesthetic results.[8][9]

The atrial septal defect is characterized by a defect in the interatrial septum allowing pulmonary venous return from the left atrium to pass directly to the right atrium. Dental treatment of patients with an atrial septal defect is prioritized in preventive care. Depending on the size of the defect, size of the shunt, and associated anomalies, this can result in a spectrum of disease ranging from no significant cardiac

sequelae to right-sided volume overload, pulmonary arterial hypertension, and even atrial arrhythmias. Atrial septal defect (ASD) is a congenital cardiac disorder caused by the spontaneous malformation of the interatrial septum. Note the following types of ASD: 1. Ostium secundum ASD: The normal septum primum does not close this type of abnormal foramen ovale at birth. A combination of excessive resorption of the septum primum and a large foramen ovale produces a large ostium secundum ASD; 2. Ostium primum ASD: These defects are caused by incomplete fusion of septum primum with the endocardial cushion; 3. Sinus venosus ASD: An abnormal fusion between the embryologic sinus venosus and the atrium causes these defects. Anomalous drainage can be into the right atrium, the superior vena cava, or the inferior vena cava; 4. Coronary sinus ASD: The coronary sinus defect is characterized by unroofed coronary sinus and persistent left superior vena cava that drains into the left atrium. This can result is desaturation due to right-to-left shunt into the left atrium.[10]

The three major types of atrial septal defect (ASD) account for 10% of all congenital heart disease and as much as 20-40% of congenital heart disease presenting in adulthood. Ostium secundum is the most common type of ASD accounting for 75% of all ASD cases. ASD occurs with a female-to-male ratio of approximately 2:1. ASD malformation can go undiagnosed for decades due to subtle physical examination findings and a lack of symptoms. Even isolated defects of moderate-to-large size may not cause symptoms in childhood. However, some may have symptoms of easy fatigability, recurrent respiratory infections, or exertional dyspnea. In childhood, the diagnosis is often considered after a heart murmur is detected on routine physical examination or after an abnormal finding is observed on chest radiographs or electrocardiogram (ECG).[10]

The management of children with ASD is prioritized in preventive care. It consists of the prevention of dental and oral diseases by systemic and local. Fluor prescriptions, deep fissure closure, and dental and oral health maintenance at home. This procedure can prevent bacterial endocarditis, which is an infection of the heart valve or endocardium caused by streptococci such as *Streptococcus sanguis* and *S. mitis*. In the field of dentistry, bacterial endocarditis is closely related to dental infection. Preventive management of bacterial endocarditis includes the administration of prophylactic antibiotics prior to invasive dental treatment which involves manipulation of the gingiva, periapical area or perforation of the oral mucosa, such as dental extraction, periodontal procedures (scaling, root planning, probing), apical surgery, biopsy, installation of orthodontic or matrix bands and rubber dam clamp (subgingival), and placement and avulsion of dental implants. Dental procedures that do not require the administration of prophylactic antibiotics are dental restoration, radiological photographs, dental printing, anesthesia injection in non-infectious tissues, fissure sealants, prosthodontics or orthodontic removable devices, orthodontic brackets in the supragingival area, and loose deciduous teeth naturally.[11][12]

2. Case Report

A 13-years-old girl with a disorder appeared on her face came to the Pediatric Dentistry Department of Padjadjaran University Dental Hospital. Her mother complained that her child was born with a cleft in the face and without a nose. This patient is a twin, most likely dizygotic. The pregnancy was unexpected, and her mother took an herbal medication in order to terminate the pregnancy. The medication was taken at an early stage of pregnancy and her mother feels that this was cause of her child's problems. There is a history of premature rupture of membranes and premature labour. The twins were delivered 8 days later. Her twin sister is said to have normal development. This child was undergoing cleft repair surgery from the age of 1 year and 3 months, and the other was at the age of 8 years. The patient's mother said there was still a small hole in the palate that caused her child having difficulty in feeding and talking, and she also said that her ECG result shows a hole in the heart. Based on on the ECG result, this patient had a large secundum atrial septal defect (21mm) that needed immediate medical intervention.

Other complaints were scoliosis, with a structurally normal spine on X-ray, and deformity of her right hand fingers (fixed deflection deformities). Other abnormalities found in this patient were midline/bilateral facial cleft, developmental delay, mild plagiocephaly, hypertelorism, frontal encephalocele, periventricular nodular heterotopia, hypotonia. There is no apparent significant family history. This patient has three older brothers claimed to be fit and well. No parent admits any significant health problem.

3. Case Management, Figures, and Tables

A. B. C.

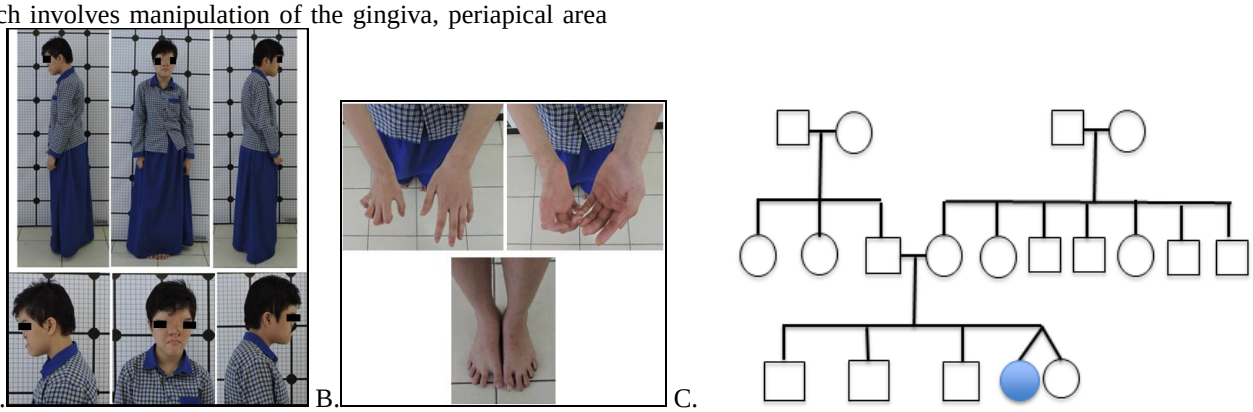

Figure 1: A. Body posture (scoliosis); B. Extremities (fixed deflection deformities on the right hand); C. Pedigree.

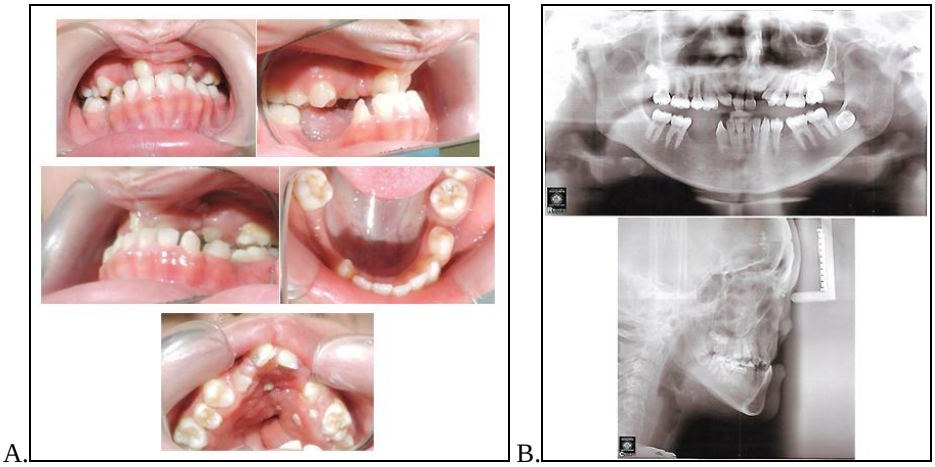

Figure 2: A. Intraoral photography; B. Panoramic and cephalometry radiography.

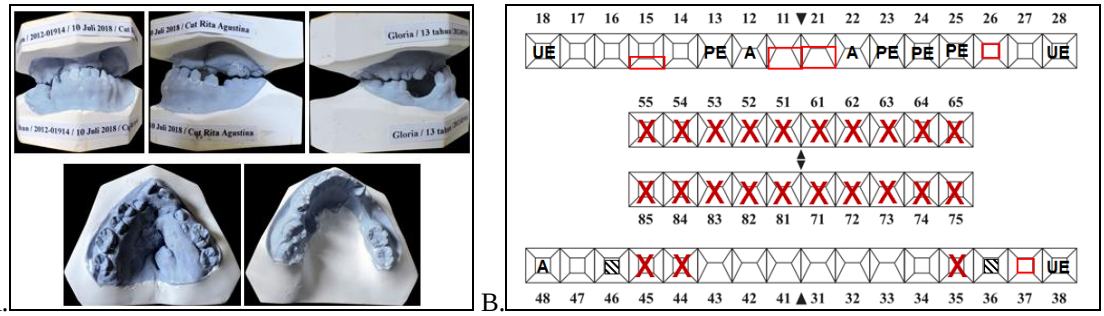

Figure 3: A. Study model cast; B. Odontogram

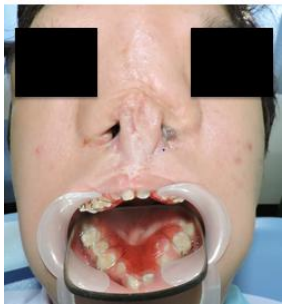

Figure 4. Obturator insertion.

4. Discussion

A genetic examination of chromosome 22q11.2 sub telomere MLPA to observe the patient's frontonasodysplasia declared to be normal and periventricular nodular heterotopia was examined through mutation DNA testing in filamin A, but the results were also insignificant. Therefore, it was taken into consideration that abnormalities occured in the patient might strongly caused by the herbal medication, and as no other family member experienced the same disorder as the patient. The examination results also revealed that the tendency of the patient's parents to have children with similar disorder was as low as 5%, because it tends to be resulted from teratogenic factors exposures, and not hereditary.

The management of the patient was handled by a multidisciplinary team, consisted of craniofacial specialists, cardiac pediatricians, surgeons, anesthesiologists, dentists, ophthalmologists, lung specialists, geneticists, radiologists, speech therapists, social workers, and psychologists. This multidisciplinary team provides comprehensive approach in diagnosing, treatment planning and delivering, and further management of the disease.

Craniofacial disorders experienced by this patient include midline facial cleft, mild plagiocephaly, hypertelorism, bilateral lip, and palate clefts, and frontal encephalocele. All of these abnormalities refer to the Tessier 0 classification, accompanied by bilateral cleft of the lip and palate. The patient underwent slit repair surgery at the age of 1 year and 3 months old and another surgery to close the skyline was done when she was 8 years old.

The obturator prosthesis has been used to restore masticatory function and improve speech for maxillary defect patients. The basic design of obturator prostheses uses the available tooth and bearing tissue to achieve maximum retention and stability. Additional retention is using Adams clasp on 14, 16, and 26. The primary goals of the obturator prosthesis are to preserve the remaining teeth and tissue and provide comfort and function to the patient.

Dental treatment in patients with atrial septal defects is prioritized in preventive care to prevent dental and oral diseases by systemic and local fluor prescriptions, deep fissure closure, dental and oral health maintenance at home. First dental visit for this patient were examination, dental hygiene education, plaque score, panoramic and cephalometry radiograf, and study model cast. Next visit were fissure sealant on 16 and 27; preventive resin restoration on 15, 26, and 37; restoration class III glass ionomer on 11 and 21; and topical flour varnish application. Cardiac surgeons have to refer surgical patients to a dentist for oral prophylaxis management and to treat any oral

infections before conducting a cardiac surgery to prevent infective endocarditis. The oral preventive care given were fissure sealant, preventive resin restoration, and topical fluor applications. These preventive care are not invasive, so the use of prophylactic antibiotic was not carried out.

5. Conclusion

The oral rehabilitative treatment aims to restore aesthetic, function, comfort, phonetic, and health of the patient. In dentistry, it aims to prevent tooth decay further deterioration or severity of the disease by maintaining a healthy oral cavity, in an effort to improve health in general. The management of craniofacial and cardiac disorders requires a team work of experienced professionals in various medical fields. This multidisciplinary approach will provide comprehensive efforts in diagnosing, treatment planning and delivering, and further management of craniofacial and cardiac disorders.

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