The No. 3 Craniofacial Cleft

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Abstrak

Sumbing no.3 merupakan satu di antara 14 tipe kelainan sumbing kraniofasial kongenital. Lokasi sumbing ini bertepatan dengan tempat pertemuan antara processus maxillaris dan frontonasalis pada masa perkembangan embryonal. Mengetahui embryologi leher dan kepala memberi pemahaman mengenai fungsi saraf kepala, prinsip-prinsip pembentukan kepala-wajah dan kelainan-kelainannya yang merupakan akibat penyimpangan dari perkembangannya. Penanganan kelainan ini memerlukan bedah rekonstruksi yang canggih. Makalah ini melaporkan kasus seorang anak perempuan 1,5 tahun dengan sumbing kraniofasial no 3.

Kata kunci: sumbing kraniofasial, kelainan bawaan, bedah rekonstruksi.

Abstract

The No. 3 Cleft is one of 14 types of congenital craniofacial cleft anomalies. The location of no. 3 cleft coincides with the embryonic junction of the maxillary and frontonasal processes. There is no theory of the causes but some hypothetic risk factors have been proposed. Understanding of head and neck embryology gives rationale to the function of the cranial nerves, principles of craniofacial form, and anomalies that result from aberrations in their development. Treatment of this malformation needs excellent reconstructive surgery. This paper is reporting a case of the no. 3 craniofacial cleft in a 1.5-year-old female.

Key words: Cranifacial cleft, congenital anomaly, reconstructive surgery.

Introduction

"In whatever form (facial features) He wanted, He put you together". (Surah Al-Infithar, Ayah 8).[1]

"When forty-two nights have passed over the Nutfah, Alloh sends an angel to it, who shapes it (makes its face) and makes its ears, eyes, skin, flesh and skeleton. Then he says, "O, Lord! Is it male or female? And your Lord decides what He wishes and the angel records it". (Shahih Muslim: Kitab Al-Qodar).[2]

The face is the most important component of the general appearance of an individual by which he or she can be identified. The word "Shurah" in the verse above is translated to "form". In the Prophet's Hadits and general Arabic usage, it is used to indicate the facial features. Thus the Ayah can be interpreted to refer to the appearance of the individual in general and to the construction of the face in particular.[3]

Facial Development

The face is not just a single organ or structure but it is constructed from five primordia. Its development is beginning in the middle of the third week, at which time the three germ layers in the cranial part of the embryo begin their specific development. Those three primary germ layers, consisting of ectoderm, mesoderm, and endoderm, serve as a basis for differentiation of the tissues and organs within the developing embryo.

By the early of the fourth week, the cranial region of a human embryo somewhat resembles a fish embryo of a comparable stage. It forms branchial apparatus that consisting of (1) branchial arches, (2) branchial pouches, (3) branchial grooves, and (4) branchial membranes. Most congenital malformations of the head and neck originate during transformation of the branchial apparatus into adult derivatives.[3,4,5]

The branchial arches are largely responsible for the formation of the face, neck, nasal cavities, mouth, larynx, and pharynx. The first branchial arch contributes to the maxillary and mandibular prominences and the anterior portion of the auricle. The paired maxillary and mandibular prominences derived from the first arch from the lateral and caudal borders of the stomodeum (primitive mouth), respectively. The frontonasal prominence, a central process formed by the proliferation of the mesenchyme ventral to the forebrain, forms the cranial boundary of the stomodeum. Although the frontonasal prominence is not a branchial arch derivative, it merges with first arch derivatives to form an integral part of facial development. These five facial prominences (two paired and one unpaired) bordering the stomodeum are responsible for the development of adult facial features (Fig. 1).[5]

Case Report

A 1,5-year old girl was referred to surgical clinic of Sardjito Hospital for treatment of a severe facial cleft. She was born as the 7th daughter of a poor couple husband and wife from a very remote area. During pregnancy, the mother regularly took a traditional javanese remedies (*jamu*). Ante natal care was taken by traditional midwife (*dukun bayi*) as well as delivery services. The mother was 39-year old whenever she delivered the baby. There was a family history of congenital anomalies. One of patient's older brother i.e. the 3rd son was suffered from syndactily of his right hand.

The physician observed that the patient suffered from bilateral cleft, cleft no 3 on the right and left side. The location coincides with the embryonic junction of the maxillary and frontonasal processes, and therefore this cleft makes embryologic sense. The right cleft is like the common cleft of the lip, alveolar process and palate (oro-nasal). There is no involvement of ocular cavity.

The left side cleft begins at the cupid's bow; undermines the nasal alar base and continues cephalad to end just medial to the inferior punctum of the lower eyelid. The nasolacrimal drainage system is disrupted and the lower canaliculus is malformed. The skeletal disruption is quite extensive. The cleft passes between the lateral incisor and the canine to involve the neighboring alveolus and the secondary palate. The lateral portion of the piriform aperture is invaded, and the medial wall of the maxillary sinus is lacking. The frontal process of the maxilla is interrupted as the cleft terminates in the lacrimal groove. Thus, a confluent cavity is formed composed of the mouth, nose, maxillary sinus, and orbit. This cleft is also called as oronasal-ocular cleft or oblique facial cleft.

The patient then undergoes the first operation to close the cleft and labioplasty. Subsequent operations are palatoplasty, velopharyngoplasty (if it is necessary), alveolar bone grafting, orthodontic operation and reconstruction of alveolar process and other parts depend on the existing deformities.

Discussion

Cleft lip is a common congenital abnormality occurring in approximately one out of 1000 life birth.[6,7] Insidency in Indonesia may be different/greater. Dr Djohansah stated it occurs once in 1083 births, while Dr. Widanto in East Nusa Tenggara stated that it occurs once in 500 births. The number of cases can be estimated $\pm$ 6640 cases per year. Most of them did not get the treatment optimally.[8] Palate cleft incidence is lower, occurs in one out of 2500 life birth, with or without lip cleft. There is no report particularly about incidence of no. 3 craniofacial cleft itself.[5]

The causes of cleft are unknown up to now, but several factors have been proposed such as: inadequate nutritional intake; teratogenic agent includes traditional javanese remedies (*jamu*) and hormonal contraception; radiation; viral and

parasit infection; hypoxia and other metabolic stress; genetic (heriditary); and mechanical factors like oligohydramnion.[9,10]

In this case, the possibility of genetic factors is supported by family history of congenital anomaly. The history of traditional javanese remedies usage also possible to be a risk factor. It is also noted that the mother was too old to get pregnant and deliver the baby (*grandimultipara*), another suspected causes of this anomaly.[10]

For treatment of the No. 3 cleft, the most two important things to be understood are principles of cleft management and time schedule of operation. There are four principles in cleft management, (1) multidiciplinary, psychologist will take care psychological stress of the parents; nutritionist to solve the diet problem; ENT doctor to handle the repeated infection; dentist to apply the obturator; physiotherapist to conduct speech therapy. Pediatrician takes the role in growth and development control. Of course, the plastic surgeon will do sequential reconstructive operations. (2) Long-term, this anomaly needs at least 5 steps of operations, with each suggested time schedule. Labioplasty should be in 3 months of age; palatoplasty in 1,5-2 year old. In this case, it is late because the patient was already 1,5 year old. Labioplasty has to be done immediately, while palatoplasty will be next 6 months later. Speech therapy has to be done during 2-4 year of age. It may require velopharyngoplasty if the speech therapy is not successful. In the 6-8 year of age, it will be done alveolar bone grafting. In the 17-18 year of age when the alveolar process does not grow anymore, it will be evaluated whether there is a hypoplastic maxilla or not. It may require a certain operation to expand maxilla with Le Fort I or II Osteotomy procedures to reach the best reconstruction. (3) Documentation, and (4) research are essential to evaluate the treatment and to develop the technical operation as well.[8,10]

What kind of the anomaly is, reconstructive operation has to be done as long as the supporting resources are available. The facial morphology, for instance, in operated children differs significantly from those of children without operation of the same age. Less than 30 years ago, medicine offered little hope to patients with severe craniofacial deformities. Today, craniofacial surgery, undertaken by highly experienced teams, has profoundly changed the outlook for these patients and their families. That which was deemed impossible in the past is now a reality through advancements in medical technology and the skill of multidiciplinary professionals.[11]

Conclussion

It has been studied a case of no 3 craniofacial cleft of 1,5 year old girl. The suspected cause may be genetic or teratogenic agent. The cleft is bilateral type, the right side is oro-nasal cleft, while the left one is oro-nasal-ocular cleft. Labioplasty and cleft closure was conducted at that time. It will be followed by palatoplasty in 6 months afterward, and other reconstructive procedures.

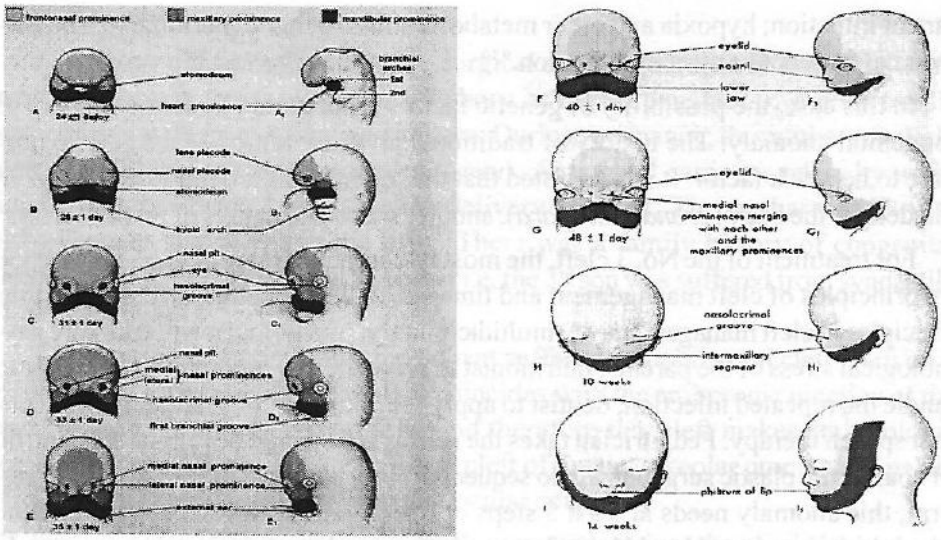

Figure 1. Facial development)[3]

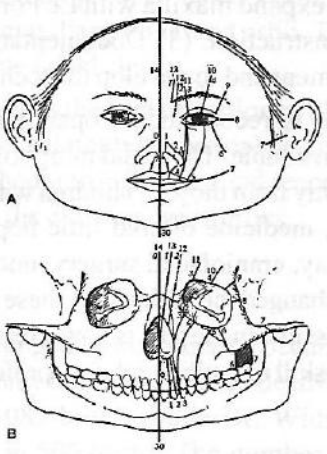

Figure 2. Tessier classification of cleft)[5]

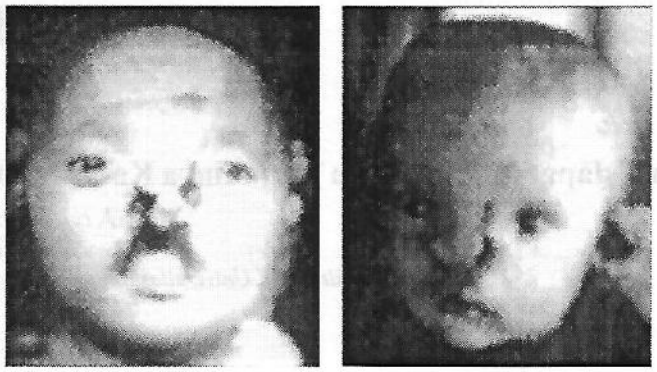

Figure 3. The existing case of no.3 facial cleft (pre and post operative)

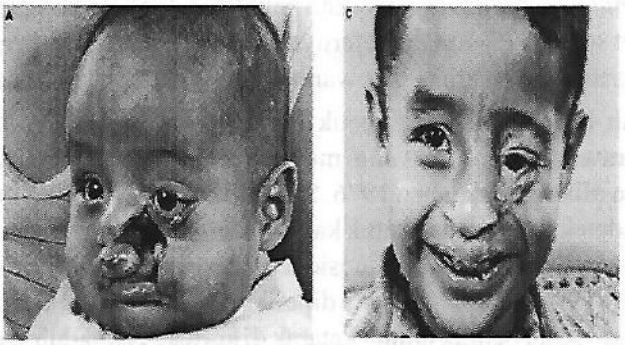

Figure 4. Pre and postoperative photograph of other patient)[5]

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