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Non syndromic involvement of Cleft lip and palate with syndactyly: A Case Report .

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Abstract

Cleft palate and lip are the most commonly occurring craniofacial anomalies which may be syndromic or non-syndromic. The etiology of non-syndromic involvement of cleft lip and palate is multifactorial mostly resulting from the genetic and environmental factor. Patient with cleft lip and palate suffers from lots of difficulties likes difficulty in sucking milk, nasal regurgitation, esthetic and speech problem etc.In this present case of non-syndromic involvement of cleft lip and palate with syndactyly ,the primary health care was provided to the patient by the fabrication of feeding plate to overcome the problem of feeding milk and nasal regurgitation and to gain the weight for the growth and development of the child till the proper age of surgery for cleft lip and palate followed by surgery of syndactyly.

Keywords: Cleft, Non-syndromic, Syndactyly, feeding plate

Introduction

Cleft lip and palate are the most common congenital craniofacial disorder with the prevalence of 1 in every 1000 birth in India.[1] It may or may not be associated with various syndrome. In literature Thomas et al[2] reported that 30 % of the cleft cases are syndromic involving cardiovascular, musculoskeletal, genitourinary system and nasal regurgitation while 70 % of the cases of cleft are non-syndromic and isolated.[3] The etiology of non-syndromic or isolated cleft is multifactorial and complex

usually resulting from the interaction between genetic and environmental factors.[4] Its prevalence varies widely, depending on the ethnicity and geographic location of the population, ranging from 1 in 300 to 1 in 2,500.[5]

Various epidemiological studies including Grosen et al[6] reported that if one parent was affected with a cleft than there is an incidence of 3.2% more chance of having a child with cleft lip and palate and a 6.8% higher chance of having a child with isolated cleft palate. Syndactyly is one of the most common upper limb congenital anomalies with the incidence rate of 1 in 2,000–3,000 live births (Schwabe and Mundlos 2004) with prevalence rate higher in male patient[7,8] It is classified as complete (fingers are joined from the web to the tip) or incomplete (the fusion of the web spaces occurs only at a point between the web and the tip) or Simple (only skin is involved) or complex (bone, the neurovascular bundle and nail structure).[9]

In literature lots of cleft lip and palate cases involved with syndrome like Vander Woude Syndrome, Pierre Robin sequence, Velocardiofacial syndrome, Median facial dysplasia , Stickler's syndrome, Marfan syndrome etc [10,11,12]have been reported. Also various syndrome which shows the involvement of Syndactyly , cleft lip/cleft palate in association with various anamolies like Facial-Digital Syndromes (OFD), Amniotic Bands Syndrome, Ectrodactyly-Ectodermal Dysplasia-Cleft Lip/Palate Syndrome , Fraser Syndrome (Cryptophthalmos-Syndactyly Syndrome), Goltz Syndrome etc [7] have been discussed but less have been reported about the non-syndromic involvement of cleft lip and palate with syndactyly.

So, here in this case report we are presenting a case of non-syndromic involvement of cleft lip and palate with syndactyly. In the present case the first line of treatment was given to the patient by the fabrication of feeding plate and delivering it to patient to fulfil the purpose of feeding milk and nasal regurgitation occurring due to cleft palate. So that the patient should become physically fit by gaining weight for the surgery of cleft lip, palate, syndactyly and leading to proper growth and development of affected child.

Case Report

A 3 months old male child was reported to the Department of Plastic and Reconstructive, Maxillofacial Surgery, GMCH Nagpur with a chief complain of difficulty in feeding milk and nasal regurgitation. On intraoral examination left side cleft lip and Unilateral Cleft palate (Veau Type III) were seen (Fig1), while on extra oral examination Syndactyly of middle and ring finger (Fig2) of both the hands were also found. Patient was not having any other medical problems and even he was delivered through normal, full term uneventful pregnancy. The weight of patient at the time of birth was 2.5kg.

Family history revealed that mother was having cleft lip and cleft palate, for which she was operated in her childhood. Parents are not having a history of consanguineous marriage. As told by the parents, the patient was having the difficulty in sucking milk and having nasal regurgitation while feeding. It was observed that the weight of the child was not increasing after the birth. So, the first line of treatment i.e. feeding plate was planned for the patient and after gaining proper weight by the patient, surgery will be planned for cleft lip followed by cleft palate and in last will be plan for operating syndactyly.

So, for fabricating the feeding plate, the preliminary impression was made with impression compound (Fig3) with due care without any anesthesia or premedication. During the procedure the child was made to cry by tapping on his foot to ensure the airway and all the care was taken to avoid aspiration by the child.

A cast (Fig4) was obtained by pouring the impression with dental stone, on which feeding plate was fabricated with auto polymerised acrylic resin. Feeding plate was properly finished, polished and checked in the patient mouth. A floss was then attached on it for easy removal of plate by parents to avoid an accidental swallowing and in case of gagging. It was then delivered to patient(Fig5). Mother was asked to feed the child with feeding plate to check for nasal regurgitation and difficulty in feeding. All the instruction regarding the use and maintenance of feeding plate along with maintenance of oral hygiene was given to parents.

Patient was recalled after 24 hours to check for any irritation and redness in the mucosa. Patient was recalled after everyone month for the reduction of the border of the feeding plate to accommodate growing arches. The weight of the patient was checked which was found to be in normal range.

Then after three months, a second feeding plate was fabricated to accommodate the enlarged craniofacial sutures at growth. For which the same procedure of impression making was carried out. The weight of the patient was periodically checked which was gradually increasing and was found to be in normal range.

Discussion

Cleft lip and palate whether it may be syndromic or non-syndromic needs to be surgically treated at right time and at right age. But the lack of knowledge, awareness and resources results in delay in medical care of such patient which even results in life threatening congenital abnormality due to malnutrition or infection.[5]

To prevent this such patient should need the primary health care as soon as possible like feeding plate which aid them in feeding and prevents the nasal regurgitation and infection. It helps the cleft patient to gain the weight and make them fit for surgery.

Conclusion

Cleft lip and palate can be successfully treated surgically but in India there is a need to bring awareness and to educate the people about the cleft and to provide information to the poor persons about the government health care centers, so that they should report their child suffering from cleft to the medical centers as soon as possible for their treatment to prevent them from life threatening abnormality. Also proper evaluation of the patient with cleft can be carried out by the physicians because sometime the cleft is isolated or nonsyndromic and can be associated with other anamolies involving other body parts which should also be required to be noticed and treated.

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Legend Figure

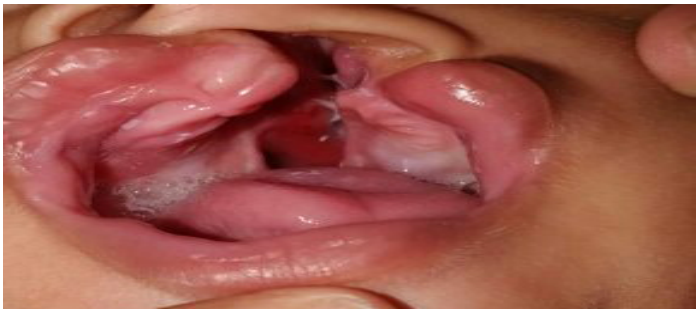

Fig 1. Intraoral photograph showing Veau's Class III Cleft palate

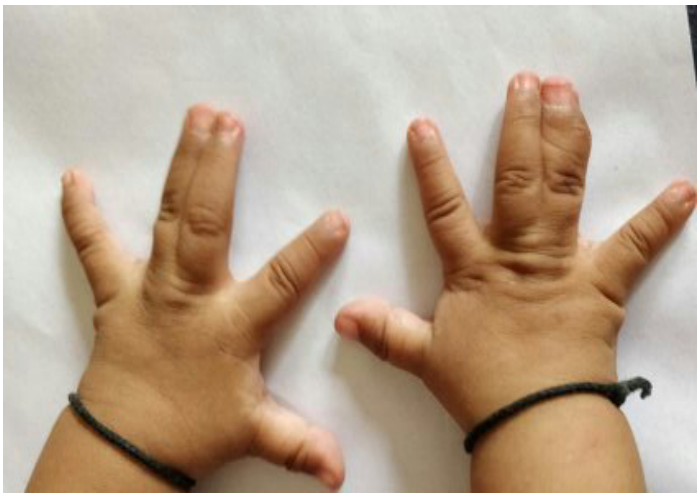

Fig 2. Syndactyly of middle finger

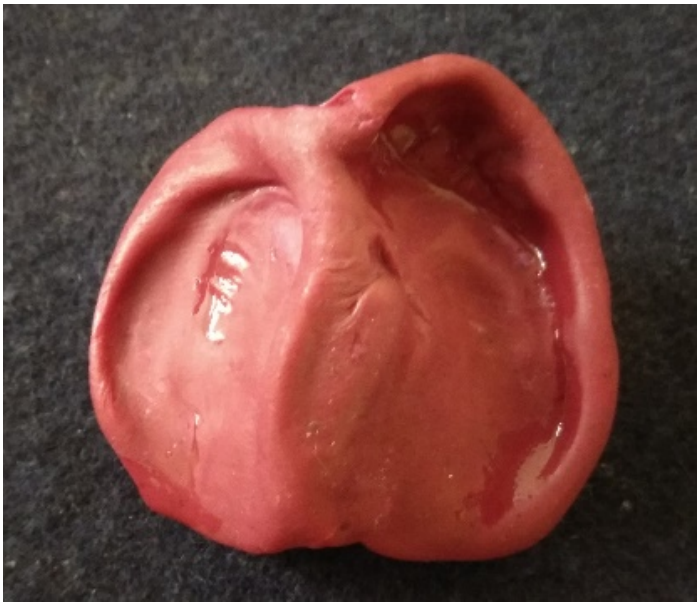

Fig 3. Impression made with Impression compound

Fig 4. Dental Cast

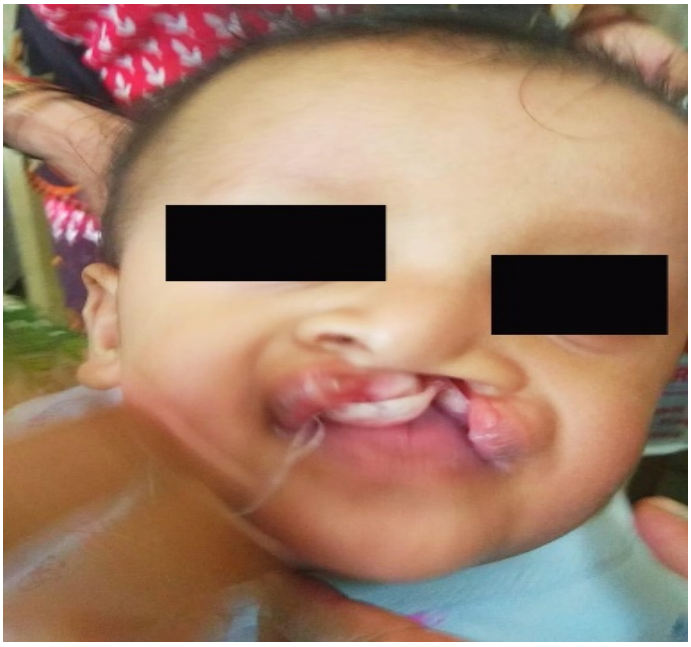

Fig 5. Feeding plate attached with floss placed in mouth