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Case Report

Bilateral Cleft Lip and Unilateral Cleft Palate with Unilateral Renal Agenesis: A Case Report

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Abstract

Cleft lip and palate (CLP) are one of the most frequently occurring craniofacial congenital anomalies. A syndromic association is frequently seen in such cases. A feeding obturator is a prosthetic aid that assists a patient in obturating a cleft until surgery is performed. Cleft lip surgery or cheiloplasty is often performed three months after birth, while palate surgery or palatoplasty is typically performed between the ages of six and fourteen months. A 2-day-old newborn presented to the Pediatric Emergency with a complaint of difficulty in feeding. On examination, bilateral cleft of the lip and unilateral cleft of the palate was present. The ultrasonography of the fetus before the birth had revealed non-visualized left kidney, so an ultrasonography (USG) of the abdomen and pelvis was advised which confirmed the finding i.e., left renal agenesis. Electrocardiography (ECG) revealed an arterial septal defect (ASD) of 4.5 mm. Such presentation is commonly seen in cases of isolated gonadotropin-releasing hormone (GnRH) deficiency (IGD). A custom-made feeding obturator was fabricated and delivered to help the baby overcome feeding difficulties. Parents were instructed and well-taught to feed the child with the appliance in place. Apart from giving appropriate instructions regarding feeding and maintenance of the appliance, parents were also informed about the surgical procedures that would be followed at a later date. Also, considering the exceptional psychological situation of the family and the need for a long-standing relationship between the cleft team for a successful outcome, appropriate counseling with reassurance was rendered to the family members.

Keywords: Cleft Lip; Cleft Palate; Palatal Obturators; Unilateral Renal Agenesis

Introduction

Cleft lip and palate (CLP) are opening or gap in the lip and roof of the mouth. CLP are one of the most frequent congenital anomalies of the craniofacial anatomy [1]. It affects one out of every 600 babies [2] with a probability of syndromic association found in around half of all the cases. A total of 400 syndromes have been found to be associated with CLP with the commonest being Van der Woude Syndrome, Pierre Robin sequence, Velocardiofacial syndrome and median facial dysplasia [3]. The majority of orofacial cleft cases lack additional features and are categorized as "nonsyndromic", i.e., 70% of all CL/P cases [4]. A family with only one person with clefts would appear to show no familial tendency. Hence the etiology of cleft lip and palate is multifactorial, involving both the hereditary and environmental factors [5].

Co-existence of cleft lip and palate and unilateral renal agenesis in an individual is seen in cases of isolated gonadotropin-releasing hormone (GnRH) deficiency (IGD) [6]. Low serum concentrations of the gonadotropins viz., luteinizing hormone (LH) and follicle-stimulating hormone (FSH) in the presence of low circulating sex steroid concentrations characterize IGD [6]. In about 40% of the people with IGD, their sense of smell is normal (normosmic IGD), whereas 60% have impaired smell (Kallmann syndrome) [6]. IGD can first become apparent in infancy, adolescence, or even adulthood. Low testosterone levels in the blood, incomplete sexual maturation, lack of secondary sexual traits (facial and body hair growth, deepening of the voice etc.), micropenis, and even cryptorchidism are all symptoms of Kallmann Syndrome (KS) [6,7]. Synkinesia, hearing loss, unilateral renal aplasia, brachy- or syndactyly, agenesis of the corpus callosum, cleft palate, and dental agenesis are all possible findings [7].

Cleft palate neonates have difficulty in feeding which may lead to 'failure to thrive' [8]. The ability to create negative pressure that

is required for suckling is diminished by the oro-nasal communication [9-11]. To compensate, the baby presses the nipple between the tongue and the hard palate to squeeze out the liquid and milk, but this mechanism itself is insufficient if the cleft is wide as the nipple gets trapped inside the defect [12,13]. The feeding process on top is complicated by the nasal regurgitation [9,14] and excessive air intake, which necessitates frequent burping and choking [8,11]. These all prolong the feeding time, exhausting both the baby and mother [13,14]. A feeding appliance or obturator is a prosthetic aid that is designed to obturate the cleft and restore the separation between the oral and nasal cavities [15], thereby facilitating proper and adequate feed. Hence, cleft lip and/or palate patients must receive therapy as soon as possible.

Cleft lip and palate encompass a spectrum of deformities involving the central face, upper lip, palate, alveolus, teeth, nose, and facial skeleton. The ramifications of these diagnoses have wide-ranging consequences, necessitating multidisciplinary treatment from infancy to adulthood and, in most cases, a stepwise surgical approach to correct the various components of the problem [16]. The timing and sequence of cleft repair, surgical approaches, and the use of adjunct procedures such as presurgical orthopedics are all areas of ongoing research and debate in the management of such patients [16].

A multidisciplinary team is needed to provide comprehensive care for the patients with cleft lip and/or palate. Anesthesiology, Audiology, Genetics, Neurosurgery, Nursing, Ophthalmology, Oral and Maxillofacial Surgery, Orthodontics, Otolaryngology, Pediatrics, Pediatric Dentistry, Physical Anthropology, Plastic Surgery, Prosthodontics, Psychiatry, Psychology, Social Work, and Speech and Language specialists are all among the team members as recommended by the American Cleft Palate Association [17].

Case Presentation

A 2-day-old newborn was presented to the Pediatric Emergency by his mother with a complaint of difficulty in feeding. On examination, bilateral cleft of the lip and unilateral cleft of the palate involving alveolus, hard and soft palate were present (Figure 1). The baby was single-term, appropriate for gestation, born via emergency lower segmental caesarean section for cephalopelvic disproportion from primigravida 20-year-old female at Mechi Hospital. The newborn had an APGAR (Appearance, Pulse, Grimace, Activity, and Respiration) score of 6 and 8 in the first and fifth minutes, respectively, with a birth weight of 2.8 kilograms. The baby had hypoglycemia, and injection 10% dextrose 80 ml had been administered immediately. There was no any history of maternal illness, smoking, drug (other than the iron and folic acid tablets) or alcohol intake during pregnancy. The patient was then admitted to the neonatal ward where orogastric tube was inserted for feeding purpose.

Figure 1: Child with bilateral cleft lip and unilateral cleft palate.

Routine blood investigations were then performed which were within the normal range. The ultrasonographic image of the fetus before the birth had revealed non-visualized left kidney, so an ultrasonography of the abdomen and pelvis was advised which confirmed the non-visualization of left kidney i.e., left renal agenesis (Figure 2). Right kidney of size 4.4*1.7 cm with normal shape and echotexture with non-dilated pelvicalyceal system and well maintained corticomedullary differentiation (CMD) were observed. Gall bladder, pancreas, liver, and urinary bladder were all found to be within the normal limits. Akhiwu., *et al*. emphasized the need for routine echocardiography in all orofacial cleft patients especially children should not be overlooked [18]. Echocardiography (Figure 3) reported levocardia, normal segmental relationship, normal dimensions of the valves, and Atrial Septal Defect (ASD) of 4.5 mm with L—R shunt without Ventricular Septal Defect (VSD) and Patent Ductus Arteriosus (PDA).

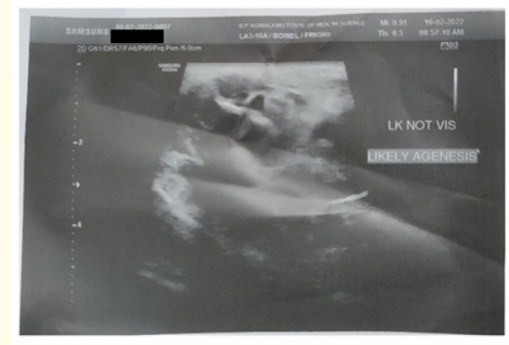

Figure 2: Ultrasonograph of the abdomen showing likely left renal agenesis.

On general physical examination; normal tone, activity and cry were present. Rooting reflex was present but suckling reflex was absent. A small vesicular ulcer (approx. 2*3 mm) was present on the mucosa of hard palate on right side for which topical application of 5% mupirocin ointment was prescribed.

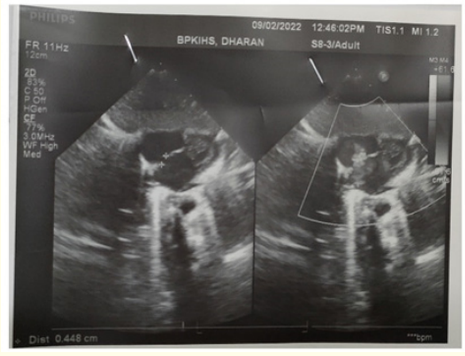

Figure 3: Echocardiograph of the infant.

After the needed counseling and reassurance of the mother along with the family members, an impression of the maxillary arch was made with an elastomeric impression material. To avoid aspiration, the newborn was held with his face to the floor. The baby cried throughout the impression making procedure ensuring a maintenance of patent airway.

The impression was then poured to make a cast model with dental stone. Major undercuts in the areas of cleft were blocked with plaster of paris. A clear, square sheet of low-density polyethylene material (ethylene vinyl acetate) of 1.5 mm thickness was used in a vacuum former machine (Erkodent Erkopress 300tp) to fabricate the feeding obturator. It was then trimmed with a scissor to the correct fit of the arch and secured with a dental floss to prevent any accidental swallowing while feeding (Figure 4).

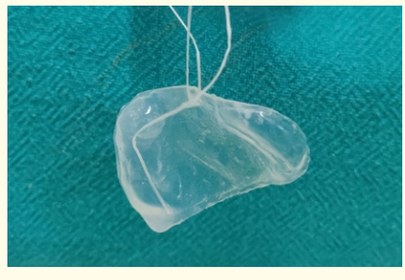

Figure 4: Custom-made Feeding Obturator.

The parents were then instructed about the methods of insertion and removal of the feeding obturator, feeding methods, and maintenance of oral hygiene along with the hygiene of feeding obturator. The newborn was able to feed adequately through bottle on the day of appliance delivery. Breastfeeding was encouraged and soon the baby was able to feed through breast, comfortably and adequately. Finally, the baby was discharged after two days receiving a BCG vaccine and was kept under regular follow up.

Discussion

Patients with cleft lip and palate require coordinated care involving multiple disciplines from birth throughout adolescence. Primary care plays a vital role in these patients who often have numerous health care needs, including feeding difficulties, speech disorders, chronic ear infections and dental and orthodontic problems. Cleft lip and palate can be diagnosed via Ultrasonography (USG) in the intra-uterine life [19]. The majority of orofacial cleft cases are not found to have a syndromic association [4]. Workup for these syndromes should have been done during intrauterine life and parents should have been counselled. But in our setup, due to lack of adequate facilities, cleft lip and palate were diagnosed later at the time of the birth. In this case, cleft lip and palate were associated with left renal agenesis which might correlate with isolated gonadotropin-releasing hormone (GnRH) deficiency (IGD). So later on, the patient might develop other symptoms as well for which the parents must be counselled well enough.

The early repair of the cleft lip and palate is associated with good esthetics, better feeding, adequate velo-pharyngeal competence and good speech and hearing development [20], thereby, improving the quality of life significantly. The evaluation of oral mucosa, which is very delicate and can easily be damaged by the obturator, necessitates a regular follow-up. Every three months, a new obturator should be fabricated to accommodate the enlarged craniofacial sutures that occur rapidly throughout this early growth period[15]. Similarly, surgical intervention should be carried out at the appropriate time for a better prognosis. When the child is around three months old, lip repair surgery is usually performed [21], whereas, palate repair surgery is normally performed when the child is between the age of six and 14 months [22], though the timing of repair is still the subject for discussion [23]. Bilateral CLP with a severely protruded premaxillary segment can present as a challenge during the surgical lip repairing. Pre-surgical alveolar molding (PNAM) is one of the infant orthopedics techniques which aids in the approximation of the cleft segments thereby improving the esthetic result of the surgical repair [24]. A condition known as glue ear, also known as adhesive otitis, is a disorder that arises when the middle part of the ear fills with fluid. Children with a cleft palate are more likely to develop this condition [21]. Although repairing a cleft palate reduces the risk of speech issues, some children with repaired cleft palate nevertheless require the speech therapy [21]. Also, Wikstad., *et al*. concluded in their study that in patients with a single kidney since childhood, the long-term prognosis is good but the late decrease in Glomerular filtration rate (GFR) and increase in albumin excretions may indicate a moderate risk for premature renal damage [25].

Conclusion

Cleft lip and palate are one of the most frequently seen congenital anomalies. Such cases, if diagnosed earlier in the intra-uterine life, would help the parents as well as the healthcare providers manage the patient in a much better way. However, in the present case, the parents were adequately counselled for the need of long-term follow ups for adequate monitoring of their child. The feeding obturator immensely helped the infant in coping with the feeding difficulties. Later on, the surgical closure of lips and palate is in turn expected to help with the functional as well as the esthetic concerns of this child. Regarding other signs and symptoms, the patient would be monitored in regular follow-ups to look for any syndromic association.

Consent

Consent Form was signed by the patient's mother, as the patient is a minor and the original article is attached with the patient's chart.

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None.

Conflict of Interest

None.

Author Contribution

- AK and SKS: Data collection and manuscript write-up.
- SK: Patient management, manuscript editing and proofreading.
- GB, LS and BK: Manuscript editing and proofreading.

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