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Management of Cleft Lip and Palate in 4 Years Old Saudi Girl: A Case Report

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Abstract: ***The cleft lip and palate-type deformity presents one of the most complex dento-surgical challenges in dentistry. A four years Saudi girl, born in Jizan city with bilateral cleft lip and partial cleft palate, she was referred to Dammam Medical Complex, from primary care setting for urgent treatment, and as the patient required a multi-disciplinary approach for management, the pediatric dentist, to insure the best outcome, coordinated a team. That may include a plastic surgeon, maxillofacial surgeon, prosthodontist, orthodontist, pediatrician, speech therapist, nutritionist, social worker and laboratory technician. The delayed management of the cleft lip and palate required a different path of treatment plan tailored according to the child patient needs and oral health quality of life. A complete dental rehabilitation under GA, aesthetic plastic lip surgery & palate repair were accomplished in this sequence. Conclusion: early intervention and adequate teamwork for children with cleft lip and palate provide the best care and outcome, when delay happen the challenges are higher and the outcome results will be affected.***

Keywords: Cleft lip, Cleft Palate, Multidisciplinary Management, CLP Centres

Abbreviations: Cleft lip & palate: CLP, General Anaesthesia: GA

1. Introduction /background

Craniofacial development is one of the most complex events during embryonic development, and craniofacial abnormalities are among the most common of all birth defects. Compared to many other birth anomalies, cleft lip and palate (CLP) are readily diagnosed. The primary palate is the keystone to the upper lip and the anterior portion of the definitive palate, embryological mal-development can affect breathing, sucking, swallowing, mastication, osculation, speech, and facial physiognomy. The secondary palate constitutes the floor of the nasal cavity and the roof of the mouth, which includes the anterior hard palate and posterior soft palate. It is an essential component of normal respiration, mastication, deglutition, and speech [1]. Cleft lip and/or cleft palate are structural abnormalities that occur in the embryonal period of life between the 4th and 10th weeks. Any interruption in this tightly controlled processing chain can result in a facial cleft, among which orofacial clefts and cleft lip and palate are most frequent [2]. The lip and primary palate have distinct developmental origins from the secondary palate [3]. Cleft lip defects are usually considered a single entity, and an accompanying cleft palate represents a more severe form [4]. The etiology of cleft lip and palate is still unknown, but both genetic and environmental factors may be responsible for many congenital malformations that can result from the developmental process failing to reach some developmental end point or threshold [5]. Cleft lip with cleft palate occurs twice as often as cleft lip alone. The cause of the familial aggregation of the disease may be genetic, environmental, or both [6]. A population and family-based analysis demonstrated a positive association between TGFB3 and non-syndromic cleft lip and palate. Also, a novel mutation of PAX9 may contribute to the development of cleft lip and palate [7]. Recently, evidence for the involvement of a major gene in the etiology of cleft lip and palate has been reported [8]. Poor nutrition, tobacco smoke, alcohol, viral infection, and medicinal drugs are among the lifestyle and environmental risk factors believed to be involved. Interactions between maternal and foetal genes are possible in the etiology of the disease. Foetal alcohol syndrome, in which the mother consumes too much alcohol during pregnancy, plus maternal smoking, increases the risk of orofacial clefts; the effect was observed more consistently and strongly for cleft lip with or without cleft palate than cleft lip palate [9]. Clefts, which can be syndromic or non-syndromic, are the most common of all birth defects worldwide and a serious malformation; most are non-syndromic. Cleft lip and palate affects 1 out of 1000 live born Caucasians and 1 out of 500–1000 live-born children worldwide. An epidemiological study of nearly 6 million births in 23 EUROCAT registries showed that 70.80% of clefts occurred in isolation, whereas 29.20% were associated

with other defects. Associated anomalies in multi-malformed cleft lip and palate patients in European countries, according to frequency, are musculoskeletal defects, cardiovascular defects, and defects of the central nervous system. The most frequent chromosomal anomaly was trisomy 13 [10]. Tunçbilek et al. examined 1229 cleft lip and palate patients and found that 151 had 206 additional malformations. In total, 55 were syndromic: Pierre Robin syndrome was the most common group [11].

2. Case Presentation /Case Study

A 4yrs old Saudi girl, born and lived in Jizan city, she comes from low social and economic class with three siblings.

Living in a remote rural area, the family low social economic status was an obstacle as they couldn't afford treatment travel expenses and transportation difficulties.

Later on, when the patient`s family was transferred to Dammam city, she was seen in Primary Health Care and referred to Dammam Medical Complex for urgent management.

Patient general health was within normal limit, with no known allergy, never been hospitalized, and with up to date vaccination record.

Extra-oral examination was within normal limit, Non-palpable lymph nodes, and below average in height and weight. She had bilateral cleft lip with incomplete right side, incompetent lip, and straight profile. Fig (1)

Figure 1

Intra-oral examination revealed incomplete cleft palate, with rotated teeth in the cleft line, primary dentition with multiple carious teeth /badly decayed teeth, poor oral hygiene & plaque accumulation. Fig (2, 3)

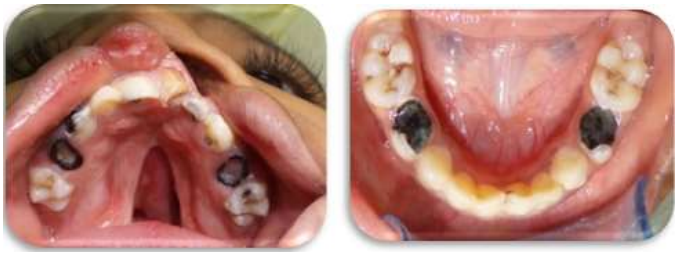

Figure 2 and 3

Full mouth radiograph taken showed multiple carious teeth with multiple badly decayed teeth and missing permanent teeth in the cleft line. Fig (4)

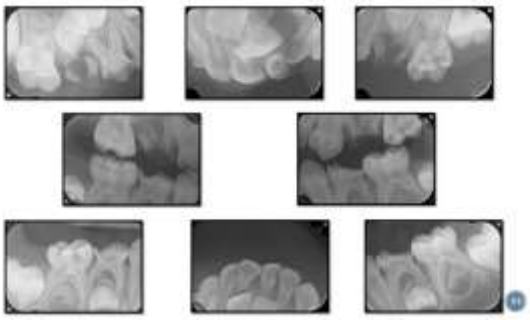

Figure 4

She was categorized as a high-risk patient with severe early child hood caries, definitely negative behavior according to Frankl scale, and requiring extensive complete dental rehabilitation under GA.

Patient was cleared by Pediatrician to rule out any underlying syndrome, disease and as part of the pre-anesthetic assessment.

As part of a Multidisciplinary Team Approach Consultation was done with various specialties: (prosthodontist, orthodontist, maxillofacial, etc.)

An obturator was made using alginate impression to give the patient a better oro-palatal seal so she can eat and drink without risk of aspiration. During the impression, the large defect was blocked out with moist 4x4 cotton gauze (using petroleum jelly) for easier insertion.

The obturator was constructed with bilateral acrylic teeth to act as space maintainer, restore extracted badly decayed teeth and improve esthetic. Fig (5)

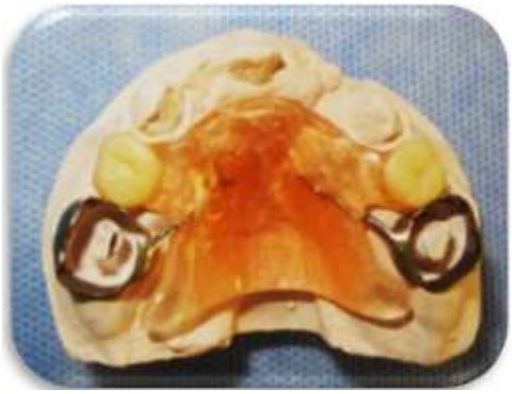

Figure 5

Complete dental rehabilitation under GA was performed treating all carious and badly decayed teeth according to the AAPD (American Association of Pediatric Dentistry) Guidelines. Fig (6, 7, 8)

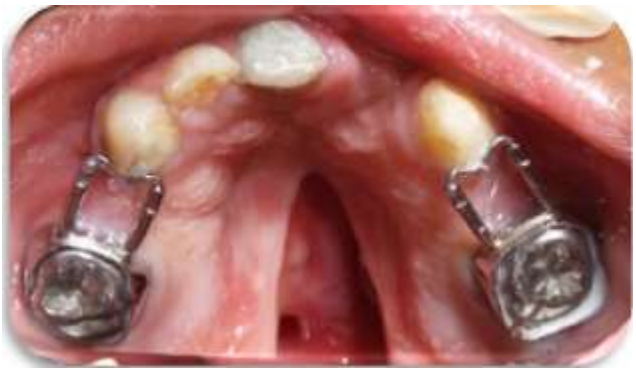

Figure 6

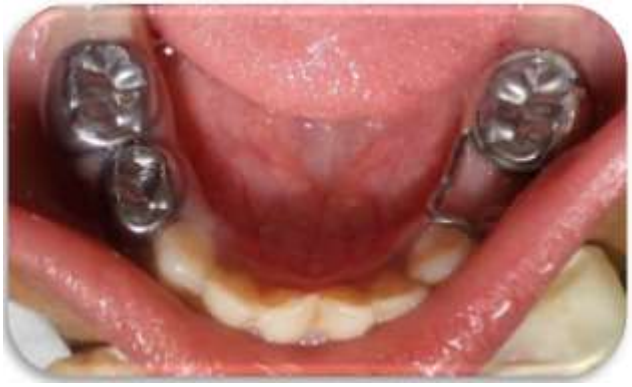

Figure 7

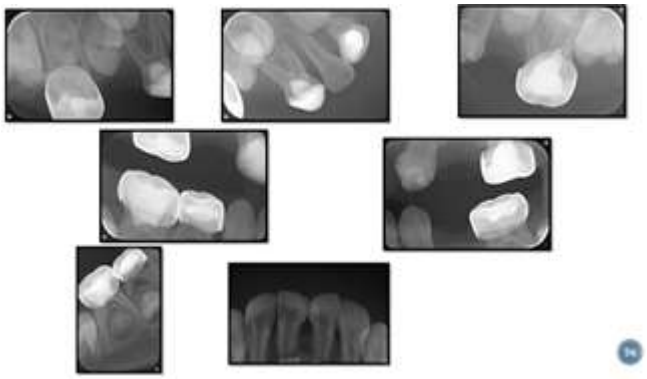

Figure 8

Patient was seen for follow up after one week, full postoperative photographs and radiographs were made. Patient compliance was very poor in following the oral hygiene instructions and in wearing the obturator, therefore an orthodontist consultation suggested to replace the obturator with bilateral band and loop to maintain the space for the full eruption of the successor teeth.

After a couple of months, Primary lip repair was performed by the Plastic Surgeon as follow: Incision with straight-line procedure repair closed. Mucosa, muscle and skin Nasal floor were repaired. Fig (9, 10, 11, 12)

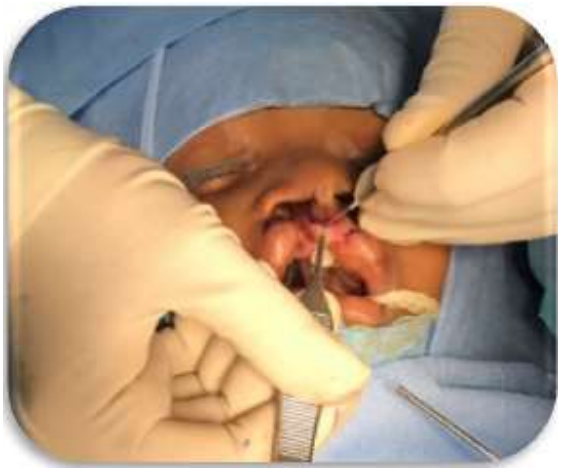

Figure 9

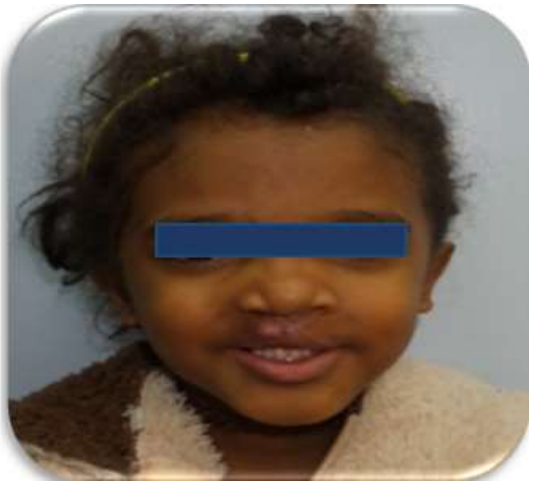

Figure 10

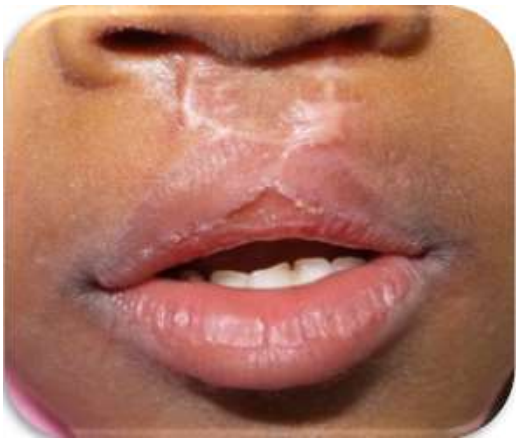

Figure 11

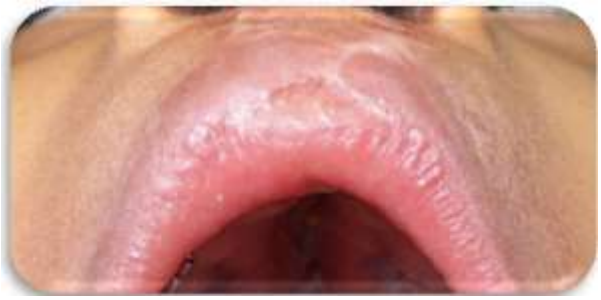

Figure 12

After complete healing of the lip repair surgery, and under GA with oral intubation, hard and soft palate repair was

done using superiorly based vomer flap, to reconstruct the nasal layer, bilateral Von Langenbeck technique was used to repair the hard palate and intervelar veloplasty for the soft palate and uveula after identifying and repairing the muscles of the soft palate. Closure of the flap was done with tension free after bilateral Hamulus fracturing by using 4.0 resorbable sutures. Fig (13, 14)

Figure 13

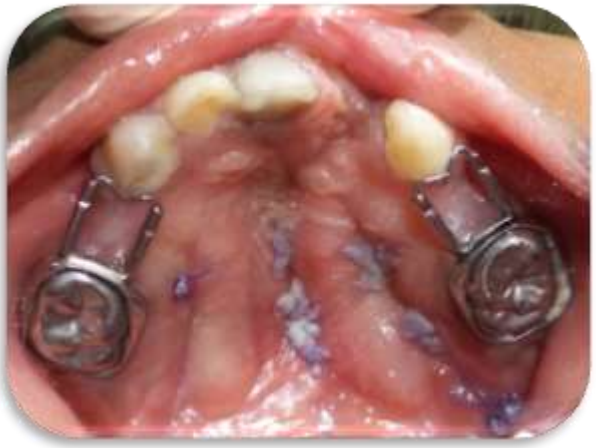

Figure 14

After complete healing of the palatal repair, Patient was referred to a speech therapist for speech education. Patient was booked on a recall schedule for follow up and monitoring.

3. Discussion

Management of children with clefts shortly after birth are more favorable and less invasive than when they are older [12].

The typical management start shortly after delivery when the patient is fitting the **Rule of 10s** criteria, which are: 10 weeks of age, 10 pounds of weight and 10g of hemoglobin [13].

Treatment usually begin with pre-surgical orthopedics at 1-2weeks , then lip repair at 3-4 months , followed by palate repair at 9-10 months, and later in adolescence lip or nasal revision surgery as needed[12].

In our case the decision to start with the full dental rehabilitation was to improve the oral health, reduce the source of infection from the multiple carious teeth and the plaque accumulation.

Patient dental health was considered very poor with multiple carious and badly decayed teeth in addition to rotated teeth that increase plaque accumulation and poor oral hygiene. Patient compliance in following the oral hygiene instructions was a challenge even with the high emphasis on the importance of good oral hygiene that were explained to the patient`s parent.

Patient occlusion fortunately was growing in favorable pattern with mesial step molar relation and no maxillary constriction, and that is something considered rare with cleft palate patients. The only explanation was that, the maxilla was growing normally and without the tension effect of the palatal repair, as usually the case if it was done in the recommended age. Orthodontic follow up is still required to monitor the eruption of the permanent teeth and to intervene earlier for achieving optimal outcome.

Lip repair surgery was done as soon as possible after 2 months of the dental rehabilitation to improve the patient`s esthetic and self-esteem.

Children born with a cleft involving the palate are at risk of developing abnormal speech patterns. Repairing the palate helps to optimize speech development, and decreases the likelihood of compensatory speech errors developing or prevents such issues altogether [12]. In our case, patient articulation was poor and unsatisfactory prior to the operation.

Four weeks after palate-repair surgery, patient was referred to speech therapist for evaluation and management. About 20 to 30 percent of children who have cleft palates will have velopharyngeal incompetence or hyper nasal speech after surgery and might require a pharyngeal-flap procedure at age 4 or 5 years [12, 14].

Ear infection and hear loss during infancy and childhood are very common finding with cleft palate due the poor functionality of the muscles responsible for the opening of the Eustachian tube. That can result in fluid building up frequently in the middle ear and can cause otitis media or ear infections, all of which can lead to fluctuating hearing loss and may affect language development [14]. Fortunately, our case never had any ear infection or complaints.

Feeding with clefts lip and palate is very challenging task for the patient`s parent during infancy. Which need education and practicing to master it. In our case patient mother did not have the chance to learn the proper way of feeding, so she used to drip the milk and fluid using dropper clip, later on, the patient start to grow and she start to accommodate her own way of feeding and eating.

Finally, due to the delay of the patient accessibility for seeking treatment, the patient and her family were accommodated and adapted to their child condition; hence, there was no need to refer the patient from our behalf to the rest of the multidisciplinary team: social worker, nutritionist, psychologist and ENT.

4. Conclusion

Children born with cleft lip and palate may have severe difficulties in many aspects of their social lives. The management of these developmental malformations is multidisciplinary and involves multiple specialties throughout childhood. The cleft lip and palate-type deformity presents one of the most complex surgical challenges.

Adding to that, the delay in seeking the treatment make it more challenging and set different priority task, which will affect the treatment plan, the prognosis and the end result. The Lead pediatric dentist can act as a coordinator for the cleft team to insure the best communication and outcome.
Teamwork and communication between multi-disciplinary specialties is a corner stone in the management of such cases, hence the presence of trained CLP team in multiple governmental centers covering all the territory of the Kingdom is primordial in detecting and providing the best treatment in such specialized centers, that will have an open access and easy reach for any CLP patient.

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6. Ethical Consent

The protocol of the present case report was approved by The Ethics Committee at D.M.C., Eastern Province Directorate, Ministry of Health. The informed consent was agreed about by the patient carer (father). All dealing performed in this case was made in accordance with the ethical standards of the 1964 Helsinki declaration and its later amendments in all comparable ethical standards.

7. Conflict of Interest

Authors declare no potential conflict of interest

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