http://dx.doi.org/10.15218/zjms.2015.0004

Evaluation of cleft lip and palate management in Erbil

Received: 20/1/2014 Accepted: 15/5/2014

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Abstract

Background and objective: Cleft lip and palate are congenital deformities that affect the primary and secondary palates during embryogenesis. The objectives of this study was to record the pattern of presentation of cleft lip and palate in Erbil, the types of surgical procedures, the outcome and complications, hence making recommendations to ensure improved care.

Methods: This is a prospective study conducted in Erbil city, in the Department of Plastic Surgery at Rezgary Teaching Hospital and Rapareen Pediatric Hospital from March 2009 to May 2013. A total of 164 patients with cleft lip/palate were included in this study. Statistical package for the social sciences (version 18) was used for data entry and analysis.

Results: The age ranged from birth to 24 years. The ratio of male to female was 1.4:1. The combined cleft lip and palate was the commonest type (87 cases, 53%). The commonest risk factor was poor family (105 cases, 64%), followed by consanguinity (97 cases, 59%). The commonest procedure for cleft lip repair was Millard (67 cases) and that for cleft palate repair was Furlow palatoplasty (57 cases). The commonest complication of cleft lip repair was wide scar (9 cases, 7.5%) mostly below one year of age. Ninety-one percent of the parents were satisfied with the children's appearance of the lip and 86% of parents were satisfied with palatoplasty result.

Conclusion: The high degree of association of consanguinity with the cleft lip and palate emphasizes the importance of education about discouraging consanguineous marriage. Millard repair is still the commonest procedure for cleft lip repair.

Keywords: Cleft lip, Cleft palate, Palatal fistula.

Introduction

Cleft lip and palate are congenital deformities that affect the primary and secondary palates during embryogenesis.[1] Cleft lip and palate are the most common facial malformation in all populations and ethnic groups, accounting for 65% of all head and neck anomalies. Every day some 700 newborn with cleft lip and/or cleft palate are born in the world, which means that a baby with such a cleft is born every two minutes. The reported incidence varies according to geographic location, ethnicity, gender, and socioeconomic status. Cleft lip and palate is most prevalent among Asians, least in Africans, and in Caucasians its prevalence is intermediate.[2] Among the cleft lip and palate population, the most common diagnosis is cleft lip and palate at 46%, followed by isolated cleft palate at 33%, then isolated cleft lip at 21%. Unilateral clefts are nine times as common as bilateral clefts, and occur twice as frequently on the left side than on the right. Males are predominant in the cleft lip and palate population, whereas isolated cleft palate occurs more commonly in females.[3] The aetiology of cleft lip and palate is not fully understood, but the best evidence today suggests a multifactorial origin for this type of birth defect, with both genetic and environmental causal factors. Intrauterine exposure to the anticonvulsant

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http://dx.doi.org/10.15218/zjms.2015.0004

phenytoin is associated with a 10-fold increase in the incidence of cleft lip. Maternal smoking during pregnancy doubles the incidence of cleft lip. Other teratogens, such as alcohol, and retinoic acid, are associated with malformation patterns that include cleft lip and palate. Genetic abnormalities can result in syndromes that include clefts of the primary or secondary palates among the developmental fields affected.[4] Throughout the history of cleft surgery, there has been debate concerning the optimal timing of surgical repair, and, in particular, the timing of cleft palate closure. Probably the most common timing sequence adopted the world over is to perform cleft lip repair at three months, and cleft palate repair secondarily at around nine months.[5] Numerous methods have been described for repair of the cleft lip deformity. Early techniques involved a straight-line closure, and these procedures still find applicability in the repair of microform clefts. In 1955, Millard described the concept of advancing a lateral flap into the upper portion of the lip combined with downward rotation of the medial segment. The technique preserves both the Cupid's bow and the philtral dimple, and it has the additional advantage of placing the tension of closure under the alar base, thereby reducing flare and promoting better molding of the underlying alveolar processes. Since then, the two most popular types of repairs have been the Tennison and Millard techniques with many modifications.[6] The primary goal of cleft palate repair is to restore the function of the palate, one of the most important functions being the develop-ment of normal speech, in addition to an intact palate without fistula.[7] One of the first palatoplasty procedures was described by Bernhard von Langenbeck in the mid-1800s. Vou Wardill-Kilner palatoplasty, developed in 1937. The Furlow technique essentially consists of repairing palatal clefts using Z-plasties of the oral and nasal mucosa. The theoretical advantage is that the soft palate may be lengthened while preventing longitudinal Scar contracture and palatal shortening. The posteriorly based myomucosal flaps re approximate the palatal mus-culature, reconstructing the levator sling. Janusz Bardach in Poland first described the Two-flap palatoplasty in 1967.[8] This study aimed at recording the pattern of presenta-tion of cleft lip and palate in Erbil, the types of surgical procedures, the outcome and complications, and hence, making recommendations to ensure improved care.

Methods

This was a prospective study conducted in Erbil city, Kurdistan region-Iraq in the Department of Plastic Surgery at Rezgary Teaching Hospital and Rapareen Pediatric Hospital from March 2009 to May 2013. A total of 182 patients with cleft lip/palate managed by a plastic surgeon and a pediatric surgeon in the formerly mentioned hospitals were included in this study. The data was recorded for age, sex, type of cleft lip/palate, unilateral or bilateral, way of feeding, shortness of breathing, antenatal history, associated nasal deformity, associated anomaly, using of presurgical orthopedic appliance whether active like Latham device or passive like alveolar molding plate, time and type of lip repair, time and type of palate repair, type of anesthesia, complications, aesthetic outcome and parent satisfaction. Eighteen patients were excluded from the study because of lack of adequate follow up, therefore; only the remaining 164 patients were included in the analysis. Cleft lip closure was mostly performed with the Millard technique[9] at the third to fifth months of age. Cleft palate closure, was performed with Furlow palato-plasty (57 cases), two flap palatoplasty (38 cases), Vou Wardill-Kilner palatoplasty (12 cases), or Von Langenbeck (25 cases) between 10^{th} and 14^{th} months of age in the majority of cases. However, due to delayed referrals or long surgical lists, 22 cases (17%) were treated after the 15^{th} months of age. Follow up visits took place on

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the 4th day, first, third, sixth and twelfth post-operative weeks and sixth month by the surgeon. The follow up findings were reported and analyzed, photographs have been taken for all patients in each visits. Treatment outcomes were assessed by three raters, including two doctors who managed the cases and one patient or patient's parents, and were graded as poor (score 0), fair (score 1), good (score 2), and excellent (score 3). The average of individual scores given by each rater was taken.

Ethical considerations: The study protocol was approved by the Research Ethics Committee of the College of Medicine of Hawler Medical University. Informed was consent obtained from parents prior to participation in the study.

Statistical analysis: Data were analyzed using the statistical package for the social sciences (version 19). Chi square test of association was used to compare between proportions. Fisher exact test was used when the expected count of more than 20% of the cells of the table was less than 5. A P value ≤0.05 was considered statistically significant.

Results

The age of the patients ranged from birth to 24 years, with the mean age of twenty months. The gender distribution was 96 (58.5%) male and 68 (41.5%) female with the ratio of male to female as 1.4: 1. The distribution of type of cleft lip and palate is shown in Table 1. The combined cleft lip and palate was the commonest type. The risk factors for cleft lip and palate are shown in Table 2. Poor family was the commonest risk factor (105 cases, 64%), with P = 0.013 which is statistically significant. The procedures used for cleft lip and palate repair are shown in Table 3.

Table 1: The distribution of type of cleft lip and palate.

Type	Number	%
Combined cleft lip and palate	87	53
Isolated cleft palate	45	27.4
Isolated cleft lip	32	19.6
Total	164	100

Table 2: The risk factors for cleft lip and palate.

Risk factors	Cleft lip and palate		Cleft palate		p
	No	%	No	%	
Positive Family History	43	36	9	20	0.048
Consanguinity	78	65	19	42	0.007
Maternal age > 30 yrs	73	61.3	14	31	0.001
Paternal age > 30 yrs	76	63.8	16	35.5	0.001
Poor family	83	63	22	48.8	0.013
Smoking	9	7.5	5	11	0.46
Alcohol drinking	7	5.8	4	8.8	0.49
Drug intake	25	19	9	27	0.92

Table 3: The procedures used for cleft lip and palate repair.

Procedure		No	%
Lip repair	Millard repair	67	56.3
	Mohler repair	35	29.4
	Rose-Thompson	17	14.3
	Total	119	100
Palate repair	Furlow palatoplasty	57	43.1
	Two flap palatoplasty	38	28.8
	Von Langenbeck	25	19
	VWK palatoplasty	12	9.1
	Total	132	100

http://dx.doi.org/10.15218/zjms.2015.0004

The commonest procedure for cleft lip repair was Millard (67 cases) and that for cleft palate repair was Furlow palatoplasty (57 cases). The Complications of cleft lip repair is shown in Table 4. The commonest complication was wide scar (9 cases, 7.5%) which mostly occurred with Rose-Thompson repair for bilateral cleft lip (5/17). Among the nine cases with wide scar, four of them were associated with nasolabial fistula and one patient with whistle deformity was associated with nasolabial fistula (P = 0.49). The Complications of cleft palate repair is shown in Table 5. The commonest complication was VPI (21 cases, 15.9%) which was diagnosed clinically (the presence of hypernasality, nasal air emission, and nasal grimacing when the patient attempt to produce consonants) which mostly occurred with two flap palatoplasty (12/38).

Table 4: The Complications of cleft lip repair.

Complications	No	%	P value
Dehiscence	2	1.6	
Wound infection	2	1.6	0.49
Wide scar	9	7.5	
Whistle deformity	3	2.5	
Total	16	13.2	

Table 5: The Complications of cleft palate repair.

Complications	No	%	P value
Dehiscence	1	0.7	
Palatal fistula	6	4.5	0.23
VPI	21	15.9	
Total	28	21.1	

Discussion

Globally the sex distribution of oral clefts is roughly 60% male and 40% females.[10] This is consistent with our study where 96 patients (58.5%) were male and 68 patients (41.5%) female. In our study Cleft lip and palate, isolated cleft palate, and cleft lip, were found in 53%, 27.4%, and 19.6% respectively. This prevalence corresponds to many observations, including those of a British study from Bolivia in 2004[11] but a study in Kenya in 2007 showed a predominance of cleft lip compared with cleft lip and palate.[12] Of the total 164 patients, 52 patients (31%) had a family history of cleft lip and palate, similar to other studies done by Andrews et al that found a prevalence of cleft lip and palate in family members ranging from 20% to 42%.[13] Consanguineous marriages between first and second cousins are commonly practiced within the Iraqi culture. Similar marriage patterns are seen in the adjacent central Asian and Middle Eastern countries.[14] In the present study 97 cases (59%) were the offspring of consanguineous marriages. This result was found to be consistent with other studies done by Ravichandran K et al, Aziza A etal, and Mansoor K etal, who observed a consanguineous relationship in 56.8%, 55%, and 61.6% of cleft patients respectively.[15-17] This high degree of association of consanguinity with the cleft deformities emphasizes the importance of education about anticipated genetic consequences of consanguinity in our society of high consanguineous marriages and should be discouraged. In the present study 105 cases (64%) of the patients were from poor families who have poor antenatal care with inadequate maternal nutritional support. Findings of observational studies suggest a role for maternal nutrition in orofacial clefts. In many studies, maternal use of multivitamin supplements in early pregnancy has been linked to decreased risk of orofacial clefts. In a meta-analysis,[18] multivitamin use was associated with a 25% reduction in birth prevalence of

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orofacial clefts. Data suggest a possible interaction between maternal hyperthermia during pregnancy and use of vitamin supplements, such that supplementation diminishes the increased risk for orofacial clefts associated with hyperthermia.[19] Maternal nutritional support with multivitamins in early pregnancy is strongly encouraged to decrease the birth prevalence of cleft lip and palate. Maternal age is a contributing factor for many congenital anomalies. In the present study in 87 cases (53%) the maternal age was above 30 years and in 92 cases (56%) the paternal age was above 30 years. Our result is consistent with that of Habib[20] who stated that the incidence of CL±P probably increases with increasing maternal age, and that of Harville et al[21] who found that the risk of CP with other defects rose with paternal age. In our study the majority of mothers did not remember history of drug consumption during the first trimester of pregnancy; however, 34 cases (20.7%) of those who remembered this event reported drug consumption other than iron and vitamins. In the present series Presurgical nasoalveolar molding (PNAM) was not done for any patient and consequently primary gingivoperiosteoplasty (GPP) was not possible at time of cleft lip repair simply because this facility is not available in our country. This is in contrast with Santiago et al[22] using PNAM who also perform GPP and reported reduction in the need for secondary bone grafting and minimal growth inhibition. Additional benefits of PNAM are improved feeding efficiency and growth.[23] Cleft lip repair represents the initial surgical endeavor in the care of an individual with cleft lip and palate. In the present series 93% of the lip repair was done between 3-5 months of age. This is consistent with studies done by Slade et al, Mchiek et al, and Goodacre et al who concluded that cleft lip repair between 3-6 months provides improved esthetic results, because the lip musculature is more developed and allows for proper reconstruction, decreased risk of anesthesia- related complications, and allows time for the parents to accept the malformation. Earlier cleft lip repair has not been shown to improve maternal bonding or have other psychosocial benefits. Neonatal repair has not seen improvement in esthetic outcomes over repair at three months.[24-26] In the present series, 83% (109/132) of the palate repair was done between 10-14 months of age. This is consistent with Chapman et al study who suggest that cleft palate repair performed before age 14 months is associated with better speech when compared with repairs performed later.[27] In the present series, 86% (102/119) of the lip repair was done with Millard, Figure 1 and Mohler technique which is a modified Millard rotation advancement repair. This is consistent with recent surveys of active North American cleft surgeons which indicate that the Millard rotation advancement or a modification of the technique is used by 84% of respondents.[28]

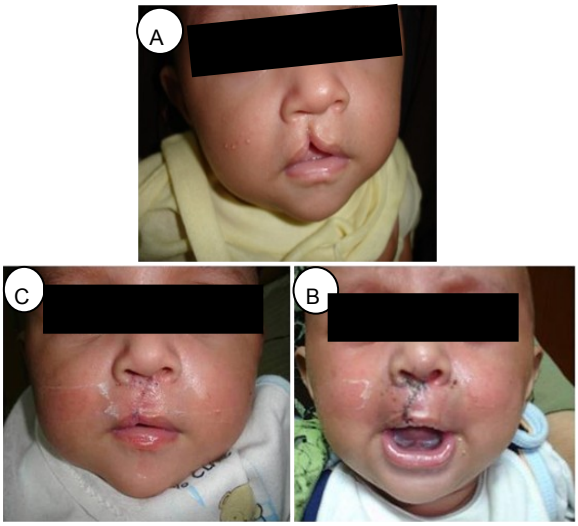

Figure 1: a- Right side incomplete cleft lip scheduled for Millard lip repair, b- The same patient seven days after the operation, c- Immediately after suture removal.

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In the present series, the commonest technique used for palate repair was Furlow palatoplasty (40.8%, 51/132), Figure 2, followed by two flap palatoplasty (30.4%, 38/132), Figure 3. This is consistent with many centers that have adopted the Furlow and two flap palatoplasty and have reported better outcomes.[28,29] Successful cleft palate repair requires adequate muscular reconstruction of the velum to create a dynamic and functional soft palate. The two flaps and Furlow palatoplasties reconstruct the velar musculature into a dynamic sling but do so in different ways. In the present series, the commonest complication of cleft lip repair was wide scar which happened in 9 cases(7.5%), Figure 4.

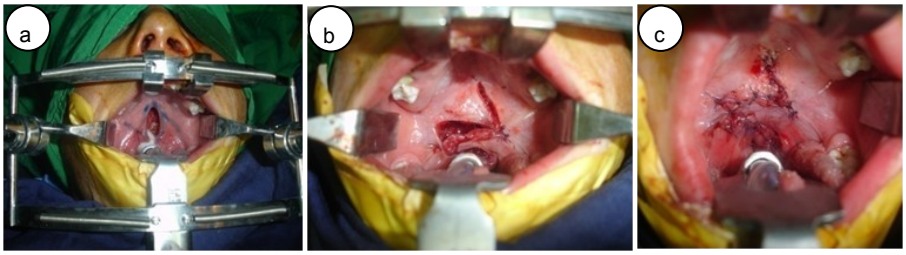

Figure 2: a- Isolated cleft palate, marking done for Furlow palatoplasty, b- Nasal layer closure completed with Z-plasty, beginning of oral layer closure with reverse z-plasty, c- Complete closure, there was no need for lateral release incisions.

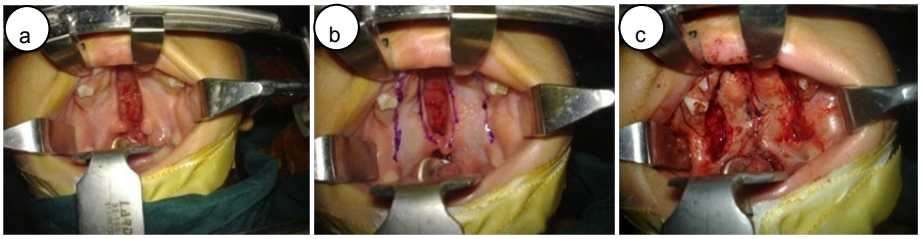

Figure 3: a- Left side complete cleft lip and palate, b- Marking done for two flap palatoplasty, c- Complete three layer closure.

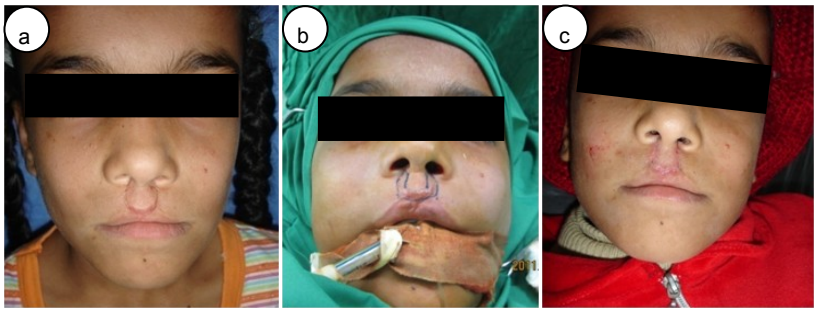

Figure 4: a- Wide scar after Rose-Thompson repair for bilateral cleft lip for whom scar revision was decided, b- marking for scar revision under general anesthesia, c- Immediately after suture removal there is improvement in the upper lip appearance.

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It was acceptable by five of them and in the remaining four cases scheduled for revision. There were two instances of wound dehiscence, the first one was due to falling on ground on third post operative day while the second one was due to wound infection which not responded to antibiotic and local wound care. Both of them scheduled for revision. The other case of wound infection responded to local wound care and systemic antibiotic for few days. There were three cases of whistle deformity which scheduled for revision. In the present series the commonest complications of cleft palate repair was Velopharyngeal insufficiency (VPI) (21 cases, 15.9%). This is consistent with other study done by Williams et al who reported VPI rate of 13% with the Furlow and 25% with Von Langenbeck palatoplasties.[30] Palatal fistula occurred in 6 cases (4.5%); four of them were located at the junction between hard and soft palate, one in the hard palate and the last one in the soft palate. The soft palatal fistula was less than two mm and closed spontaneously after six months, but the other palatal fistulas scheduled for repair after minimum of one year from the cleft palatal repair. This is consistent with other study done by Helling et al[31] who reported a fistula rate of 3.2% with the Furlow technique, and Noorchashm et al[32] who reported 3.4% fistula formation after two flap palatoplasty. In the present series, 91% (109/119) of the parents were satisfied with the children's appearance of the lip and 86% (114/132) of parents were satisfied with palatoplasty result (i.e. reported good to excellent outcome). The parents of those who developed complications and some with unrealistic expectations were unsatisfied. This is consistent with the other study done by Anke et al[33] who reported 93% parental satisfaction with lip appearance and Munz et al[34] who showed that patients and parents were rather satisfied with the treatment outcome of cleft lip and palate.

Conclusion

This high degree of association of consanguinity with the cleft deformities emphasizes the importance of education about anticipated genetic consequences of consanguinity in our society of high consanguineous marriages and should be discouraged. Maternal nutritional support with multivitamins in early pregnancy is strongly encouraged to decrease the birth prevalence of cleft lip and palate.

Conflicts of interest

The authors report no conflicts of interest.

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