Oro-Facial Clefting: Experience at King Hussein Medical Center

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ABSTRACT

Objective: To evaluate and analyze patterns and factors associated with the epidemiology of cleft lip and palate in one of the most active centers for craniofacial care in Amman, Jordan.

Method: The records of 656 cleft lip and cleft palate subjects who attended the Orthodontic Department at Royal Rehabilitation Centre were registered in the Cleft Lip/Palate and Craniofacial Anomalies Registry. The sample was collected from January, 2013 to April, 2014. Variables such as cleft type and side, gender, age, family history, any associated syndromes, and prenatal diagnosis were all recorded and later analyzed.

Results: Regarding the cleft type; the most prevalent type was cleft lip and palate (43%) then isolated cleft lip (20%) followed by isolated cleft palate (8%). Bilateral cleft lip and palate was more common than bilateral cleft lip only with (21%) and (7%) respectively. Also, more clefts were found in male patients (386) than females (270), males predominated females incomplete cleft lip and palate type anomaly with 277 and 152 respectively; whereas, approximately equal numbers reported in isolated cleft lip and cleft palate among both genders. A range from 7 days to 30 years were registered in this study, with an average age (11.5 ± 5.68). A positive family history of clefting from father and mother side was seen in (3.8%) of cases. 4% of total cases had a diagnosed syndrome. Only 3% of parents had prior knowledge for their child CLP deformity via the routine prenatal ultrasound diagnosis.

Conclusion: this descriptive study for a pool of cleft lip palate patients revealed no significant differences from available national and regional epidemiological data. Further, joint hospital research and cooperation are needed to give a broader picture of the patterns and variables associated with cleft lip palate anomalies in Jordan. Prenatal diagnosis via ultrasound routine examination should be implemented and reviewed for routine screening of oral clefting.

Key words: Cleft lip and /or palate, Craniofacial registry, Patterns and variables of cleft lip/ palate.

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Introduction

Oral cleft can be defined as a furrow or disunion in the soft and/or hard tissues affecting the facial region, lips, primary and

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secondary palate. It ranges from incomplete to complete clefting and could be unilateral or bilateral.[1] Cleft lip and or palate (CLP) deformity is the most commonly seen congenital anomaly at the time of birth.[2] In Jordan, the prevalence of CLP was reported as 2.4 per 1000 new newborns.[3] This deformity is associated with major public burdens both socially and medically, updated data about patterns of CLP is needed in every craniofacial care center.

Patients with Oro-facial clefting are subject to extensive dental, surgical, medical and psychological interventions which start as early as intra-uterine screening before birth to infancy's dental and lip/palate repairs to later orthodontic preparation for bone graft and or orthognathic surgery if needed at adulthood.[4] Data covering birth defects concerning population-based studies arriving from the developing countries is insufficient.[5] Whereas, the recommendations of the Euro cleft study group; where they assessed treatment outcomes for six major centers in Europe treating cleft lip palate patients, this study group stated the value of establishing the accurate prevalence rate of the cleft lip palate population annually in order to provide the catalogue needed for carrying out and organizing the treatment for this anomaly,[6] consequently the study concludes, this will make possible dividing and planning the necessary number of treatment centers for a national future cleft program.

This holistic care provided for cleft lip palate patients and their parents aim to attain better feeding, speech, esthetics and psychosocial well-being during each stage of his/her life.[7] Variables associated with CLP, such as side and extent of cleft, gender and family history if any, also whether any associated congenital anomalies will definitely affect treatment approaches and outcomes.[8] Rather unexplored variable in the developing countries, which should be considered during the early embryonic stage of fetal life in any population, is intra-uterine screening,[9] and this study investigated its extent and availability for parents as an early diagnostic tool.

This identification of such patterns and variables associated with CLP becomes of paramount importance to obtain best intervention at best timing, consequently better treatment outcomes. In this cross sectional hospital-based survey, patient registry for CLP deformity was undertaken to report all children who attended the Royal Rehabilitation Center for consultations and treatments of orofacial clefts during a 1 year follow up period, in order to signify the patterns of CLP anomaly from the hospital-based registery at a tertiary care hospital in Amman, Jordan.

Methods

The data for 670 cleft lip and/or cleft palate subjects -14 were excluded due to insufficient data records- who attended the Orthodontic Department of the Royal Jordanian Rehabilitation Center were registered in the Cleft Lip/Palate and Craniofacial Anomalies Registry created for this study purposes. The sample was collected from January, 2013 to April, 2014. All patients presenting with oro-facial clefting whether for consultation, new comers, under treatment or referral cases were recorded, all attempts were done to avoid repetition or inaccurate cleft description, a simple descriptive classification system was used to record oro-facial clefting, and data were collected via an experienced clinician in this field (fourth author).Variables such as cleft type and side, patient's gender, age, family history, and whether any associated syndromes or anomalies with oro-facial clefting were collected , finally intra-uterine screening was evaluated as a tool to detect CLP deformity via ultrasound examination by parents interview.

All cleft types were classified by simple descriptive classification as follows: cleft lip and alveolus (right, left, or bilateral); cleft lip and palate (right/left, unilateral/bilateral); cleft palate and facial cleft.[10] Information about the associated major anomalies or recognizable syndromes was recorded as well. Data were collected by fourth author via directly filling a written questioner prepared for this purpose. Later, data were statistically

analyzed using SPSS software (Statistical Package for Social Sciences, release 10.0.5, SPSS Inc., Chicago, Illinois, USA).

Results

The distribution of the cleft type in relation to gender-during the period from January 2013 to April 2014- is showed in Table I. Of the total sample of this one year follow-up study; 43% had a unilateral cleft lip and palate either right or left, this was considered as the major representative pattern for this sample, followed by cleft lip only then bilateral cleft lip and palate with 27% and 21% respectively. Other patterns reported with lesser values e.g. isolated cleft palate with 8% and facial cleft with only 1%. The side and location of unilateral clefts showed numerical differences; left side was more predominate than the right side in affected cleft lip and palate patients with (32%) and (11%) respectively, as well as isolated cleft Lip (15%) for left side and (5%) for right side. Variable presentations of this study are shown in Fig. 1, 2, 3 and 4.

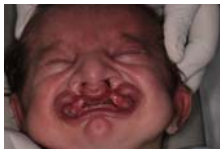

Fig. 1: A bilateral complete cleft lip and palate presented in this 25days old male neonate

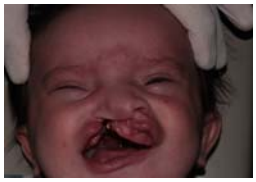

Fig. 2: A right sided unilateral complete cleft lip and palate is shown in this 33days old female neonate

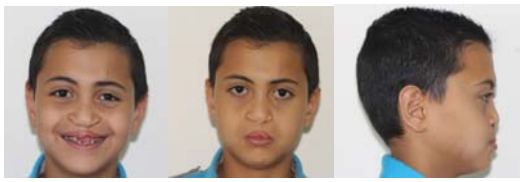

Fig.3: A 14 years old male with bilateral cleft lip only.

Fig. 4: shows a 21 years old non syndromic adult with an operated Bilateral Cleft Lip and Palate

Of the 656 subjects identified, there was a predominance of males in overall prevalence of cleft types with 386 in comparison with only 270 for girls. More males than females were affected by cleft lip and palate with (42%) and (23%) respectively. Whereas, isolated cleft lip and cleft palate reported with approximately equal numbers among both genders. See Table II.

The age range of subjects was from as young as 7 days to 30 years, with an average of 11.5 years ± 5.68. Upon evaluating the relevant family history with medical records, only 3.8% stated positive history in their family tree. Cleft lip palate part of a diagnosed syndrome was present in only 28 subjects, giving us a percentage of 4% for syndromic CLP. See Table III

As for intra-uterine ultrasound routine examination sensitivity to detect cleft lip palate deformity, only less than 20 (3%) families had prior knowledge of this deformity preconception, and none of the cleft palate only anomaly was diagnosed.

Discussion

Oro-facial clefting is considered a substantial long-term disability in children born with CLP deformity, as well as an overwhelming psychosocial and financial stress for the affected families and individuals. The CLP anomaly treatment is a long-term process starting soon after birth and may continue into the mid adulthood of life with orthodontic and pediatric care, multiple surgeries and simultaneous ear, nose, and throat clinics care as well as audio logical and speech therapy sessions.[11] Therefore, continuous epidemiologic updates are needed for each craniofacial national program and for any cleft care tertiary center.

The precise prevalence of orofacial clefts in the Middle East is still unknown.

Nevertheless, a few published reports gave an idea about the incidence of cleft in this part of the world. Suleiman et al from Sudan found an overall incidence rate of oral clefts per every 1000 live births to be at 0.9.[12] Whereas in Oman researchers found that the prevalence of CLP in Oman is 1.5 per 1000 live births. The prevalence of clefts in Iran is 1.03 per 1000 births.[13]

Although this study is again using hospital-based data, it investigates a pool of cleft lip palate patients for more than one year follow up while assessing new variables of interest such as intra-uterine screening. We believe that this study, with its fairly large number of subjects, may fill a vital information gap. There was no attempt to investigate the relevant prevalence or incidence of CLP.

Table I: Distribution of orofacial cleft patterns for Boys and Girls.

Total	Girls	Boys	Cleft Type
50 (8%)	26 (52%)	24 (48%)	Isolated Cleft Palate
48 (7%)	22 (46%)	26 (54%)	Bilateral Cleft Lip
96 (15%)	50 (52%)	46 (48%)	CL Left
30 (5%)	16 (53%)	14 (47%)	CL Right
212 (32%)	76 (36%)	136 (64%)	UCL P Left
74 (11%)	24 (32%)	50 (68%)	UCL P Right
140 (21%)	52 (37%)	88 (63%)	Bilateral CL P
6 (1%)	4 (67%)	2 (33%)	Facial Cleft
656 (100%)	270 (41%)	386 (59%)	Total

Table II: Sex distribution of cleft lip and/or palate.

Total	Facial Cleft	Bilateral CL P	UCL P RT	UCL P LT	CL RT	CL LT	Bilateral C L	Isolated C P	Sex
386	2	88	50	136	14	46	26	24	Boys
270	4	52	24	76	16	50	22	26	Girls
656	6	140	74	212	30	96	48	50	Total

Table III: Number and Percentages of Patients with oro-facial Clefts Associatedwith Major Anomalies and/or Syndromes

Total	Girls	Boys	
28 (4%)	10	18	Syndromic cleft
628 (96%)	262	366	Non-syndromic cleft

In Jordan, the exact number of subjects with CLP deformity is unknown exactly, due to a lack of a birth-defect registering system and an absence of national surveys on that issue. However, there was an eleven years retrospective report investigating birth prevalence rate to be around 1.39 per 1000 live births.[14] In the present study, CLP was observed more often than was cleft lip or cleft palate only. And this was consistent with the oral cleft literature and studies worldwide. In general, CLP epidemiology and pattern in this descriptive evaluation was no different than other major population studies,[15-17] a rather inconsistent variable with the CLP literature epidemiological studies was that we had about equal numbers of males and females affected with cleft palate only, in contrast to higher prevalence in female patients[17] but again this descriptive study is not indicative of a population incidence or prevalence as explained above.

Prenatal ultrasonography screening for fetus anomalies has become a routine practice of prenatal routine care; it is considered a diagnostic tool that is non-invasive, fairly cheap and surely accepted by pregnant women with different backgrounds. With no doubts, continual improvement in technology, equipment and skill of ultrasonography examiners has led to increased detection rates of craniofacial anomalies. [18-20] However, the investigation of this last variable yielded a rather unexpected finding; in our study which

was the first study to evaluate the sensitivity of this diagnostic tool in this part of the world, the findings should sound the alarm bells, less than 20 parents (3%) identified this CLP deformity via the routine intrauterine ultrasound screening, we think the benefits of this diagnostic tool is not fully appreciated; parents counseling could provide better neonatal care at a specialized center, psychological trauma could be hugely lessened, also early detection of such malformation allows discussing the possibility of termination of pregnancy for those anomalies that are lethal or significantly handicapping.[21] In some centers intrauterine treatment or planned delivery at a tertiary center may be also well thought-out.[22,23]

Conclusions

Craniofacial anomalies registry database is needed for any referral tertiary center, this registry aids for continuous update on incidence, etiological and genetic studies on a local and later a national level. Future studies should focus on adopting protocols for treatment and management of the cleft lip palate population.

Intrauterine screening as a diagnostic, non-invasive, cheap and routinely available tool-can lead to higher detection rates of oro-facial malformations, so forth; screening those during the antenatal period shouldn't overlook a cleft lip palate anomaly. Further studies should assess and investigate this simple but important screening tool in Jordan. Medical staff especially Gynecologist and radiologist should take more advantage of this rather sensitive detection tool.

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