Clinical Epidemiology and Global Health 7 (2019) 6–10

Contents lists available at ScienceDirect

Clinical Epidemiology and Global Health

journal homepage: www.elsevier.com/locate/cegh

Original research article

Epidemiology of lip and palate clefts in northwest of Iran: Prevalence, surgery complications and unrepaired patients younger than 18 years old

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ARTICLE INFO

Article history:
Received 19 June 2017
Received in revised form 4 November 2017
Accepted 10 November 2017
Available online 13 November 2017

Keywords:
Cleft lip
Cleft palate
Unrepaired cleft
Prevalence
Secondary surgery
Iran

ABSTRACT

Background: This study investigated the prevalence of lip and palate clefts, need for reoperation and primary surgery in unrepaired patients younger than 18 years old in Zanjan province, northwest of Iran.

Methods: All urban and rural healthcare centers and the education department of Zanjan province were informed about this project. After identifying all patients with cleft lip and palate, they were sent to the plastic surgery clinic of our hospital. Their data were gathered, including sex, age, clinical classification, surgical modality, complications, surgery satisfaction and un-repaired patients.

Results: A total of 87,356 people were examined by social workers. Among them 120 children and adolescents had cleft lip and palate (1.37 per 1000). 59 (49.1%) patients had cleft lip and palate, 35 (29.1%) patients had only cleft lip and 26 (21.6%) patients had only cleft palate. 25 patients (20.8%) had not done surgery (detection rate was 2.86 per 10,000). Secondary surgery was done in 39 patients (41.05%). In spite of a referral health system in Iran, 25 children with cleft lip had not been operated.

Conclusion: Although most of the identified patients had been operated before, almost half of them had to undergo surgery again. Given the lack of studies on this topic in Iran, our findings can form the basis for further studies to precisely identify the roots of cleft lip and palate and properly plan to decrease its risk factors. Further studies are needed to identify reasons of missed detections and why primary surgeries are not satisfying.

1. Introduction

Congenital anomalies are changes in the structure, performance, and metabolism at birth, and there may be one or several anomalies that lead to physical and mental disorders.[1] Prevalence of these anomalies varies in different countries.[2] Cleft lip and palate are among the common forms of these disorders.[3] In addition to facial manifestations, functional disorders such as speaking, hearing, chewing, swallowing, and breathing problems are caused by these disorders.[4] Repair is meant to regain the symmetry and alignment of anatomical landmarks and restore the child's smile.

Patients with these disorders need multidimensional surgical and nonsurgical care from birth to adulthood.[5] The rates of mortality and morbidity are also higher in them.[6] They and their families suffer severe mental and social consequences.[7] The cleft may affect the lips, nose or palate. Lip and palate clefts are primarily classified as unilateral or bilateral and also as complete or incomplete. Complete clefts affect the whole lip and extend to the nose. Incomplete clefts affect part of the lip, where there is a bridge connecting the middle and lateral lip elements. Unilateral cleft is normally associated with lip and nose anomalies. Bilateral

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https://doi.org/10.1016/j.cegh.2017.11.002
2213-3984/

cleft involves clefts on both sides of the lip and nose and the middle part of the lip (known as the prolabium). Clefts have different prevalence in different countries.

Identifying repetitive and prevalent patterns of common deformities improves surgical planning and helps the surgeon in future surgeries. Through a combination of accurate analysis, clinical patience, and technical precision, successful correction of secondary cleft lip and palate deformities can be delivered.[8] Secondary deformities are common among children born with cleft lip and palate. These children undergo numerous surgeries during the early stages of life. Thus, treatment of secondary deformities and reducing the number of interventions are crucially important for this age group.[9] Complications after cleft surgery are clinically unavoidable. More attention should be paid to the etiological factors to minimize their prevalence. For example, the overall complication rate has been 16.8% in a province of China.[10] There are more non-operated patients in developing and underdeveloped countries. In underdeveloped countries one-stage definitive repair of cleft lip and palate is considered for late-presented patients.[11]

Since patients are treated in several steps by a plastic surgeon, most patients do not undergo treatment or are treated incompletely. Moreover, because of the complexity of this condition's causes and its clinical symptoms, most patients are not clinically and accurately diagnosed. In spite of having a referral health system for identifying congenital anomalies in Iran, there is no exact estimate of the number of patients who have or have not done surgery, have complications or need reoperation. This study aimed to gain a better understanding of the prevalence of cleft lip and palate, trend changes in its incidence, surgery results, patients' satisfaction, need for secondary operation in those who had done surgery and primary surgery in un-repaired patients younger than 18 years old in Zanjan province, northwest of Iran.

2. Materials and Method

From March 2013 to May 2016, all the urban and rural healthcare centers and the education department of Zanjan province were informed about this study. The cities of this province are mostly at near borders with five other provinces of northwest of Iran, except for Zanjan city. Afterwards, all of the children and adolescents younger than 18 years old who had cleft lip and palate were identified and sent to the plastic surgery clinic of a hospital in Zanjan city, Iran (Fig. 1).

Iran has a healthcare system network in nearly all villages and cities. This includes 'health homes' in small villages (in which there are nurses, social workers, primary drugs and vaccination), 'small clinics' in the central villages (in which there are physicians, nurses, pharmacy and ambulance), 'well-equipped clinics and hospitals' in the smaller cities (referral level 1) and the central city of a province (referral level 2) and in major cities of the country (referral level 3). Usually the congenital diseases are identified in this system from childhood. If a problem or disease cannot be managed or treated in one level of the system it will be referred to the upper levels.

We trained the all of the head nurses of clinics in the province and then they trained the nurses and social workers of villages. In most centers, social workers completed the related forms according to their trainings and sent the potential cases for further examinations. In some cases the surgery team travelled to the patients' city.

Social workers and nurses who completed the questionnaires, made the primary diagnoses. Patients were assessed on the basis of medical history, complaining and deformity examination. The final diagnoses were made by a specialist using clinical examinations and photography. Patients' contact information and their consent for doing the surgery were recorded on special forms. A photographic record of all these cases was also maintained. Afterwards, the data were analyzed by the statistical package for social sciences (SPSS) software version 20 (Chicago, IL, USA).

All data were recorded, including sex, age, clinical classification, surgical modality, complications, number of non-operated patients (late presentation), surgery satisfaction, and the need for secondary surgery in those who had done surgery before. Finally, prevalence of cleft lip and palate was found in this province, which can be generalized to the northwest of Iran because it is located exactly in the middle of northwestern part of Iran.

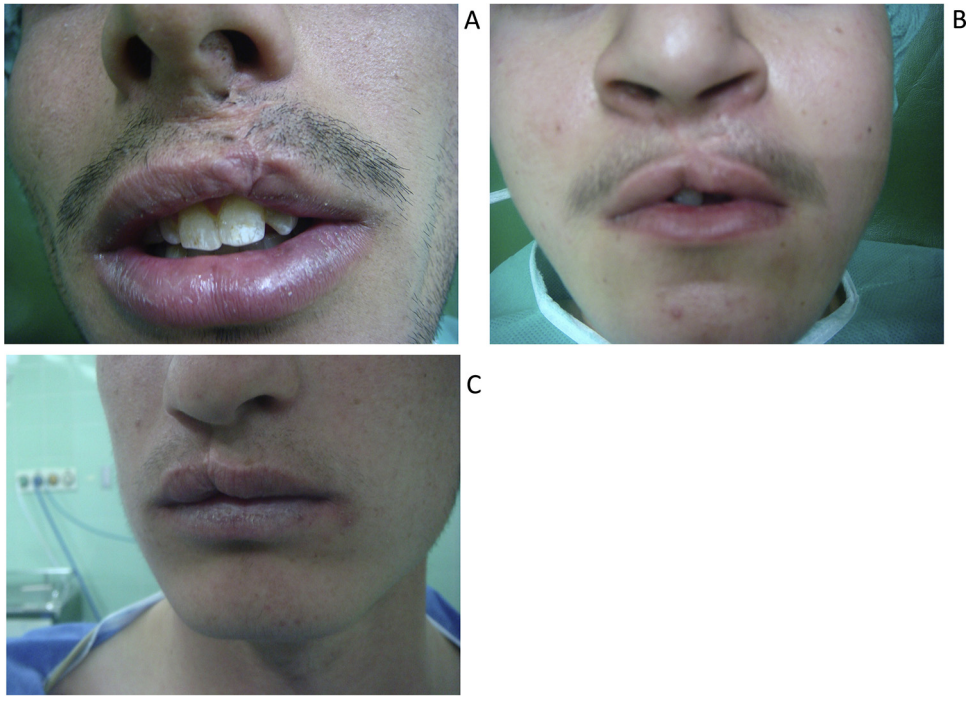

Fig. 1. Some of the adolescents with clefts that were identified in this study.

3. Results

A total of 87,356 children and adolescents (including 44,357 boys and 42,999 girls) were examined in all of the studied cities. The number of children younger than 18 years old and prevalence of lip and palate clefts in different cities of the province were estimated (Table 1). There were only 120 patients with such clefts. This means that the prevalence was 1.37 per thousand. The highest (54%) and lowest (5.8%) number of patients were in Khodabandeh and Tarom cities, respectively. 16 (13.3%) patients were two years old and one (0.8%) patient was 17 years old. People of Khodabandeh and Mahneshan cities have the least mean of revenue and people of Zanjan and Tarom cities have the highest mean of revenue in the province.

Also, 23.7% of the identified patients lived in families with three children. The three-child families had the highest prevalence of children with lip and palate clefts. 21.1% lived in two-child family, 15.8% in one-child family, 12.5% in four-child family, 15.1% in five-child family, 4.6% in six-child family, 4.6% in seven-child family, and 2% in eight-child family. Only in 21% of the families, the parents were relatives or cousins.

Around 37% of identified cases (45 patients) were younger than four years old. There were 22,783 children in this age group (26.08%). So the prevalence of lip and palate clefts in this age group was 1.97 per 1000, more than the other three age groups ($p = 0.004$, Table 2). 36 patients (30%) were girls and 84 patients (70%) were boys. The cleft lip was more observed in the left side. 59 (49.1%) patients had cleft lip and palate, 35 (29.1%) patients had only cleft lip and 26 (21.6%) patients had only cleft palate. Three children suffered rare cleft lips that were not included in the study.

Moreover, 94.2% (113 patients) had been referred by the referral healthcare system for treatment but only 79.2% (95 patients) had done the primary surgery. In this regard, 46.7% (56 patients) were operated in the first year of their life (Table 2). Among the 73 patients that had done surgery previously, cleft lips were repaired by Millard and Tennison techniques in 52 and 21 patients, respectively. 25 patients (20.8%) had not done surgery. Their deformities were cleft lip with palate in 11 cases, cleft lip in 10 cases and cleft palate in four cases. The un-repaired clefts detection rate was 2.86 per 10,000. Among them there were 21 boys and four girls.

In 56 patients from the 95 previously operated patients (58.94%), the defects were corrected with surgery (patients were satisfied), but in 39 patients (41.05%) they were not (patients were not satisfied). The most common post-surgery complication was surgical scar (Table 3). Finally, 64 (25 + 39) from among 120 identified patients underwent surgery in our center. Five patients with wide cleft palate (four needing secondary surgery and one un-repaired patient) were referred to a tertiary center (referral level 3) for treatment. Totally, 11 (7 + 4) patients underwent secondary cleft palate repair. We did only four cases of primary rhinoplasty in patients that were 15–18 years old.

Table 2
Demographic data of identified cleft lip and palate patients of Zanjan province.

Index	Frequency	Percentage
Sex		
Boy	84	70
Girl	36	30
Age		
0–4 years old	45	37.5
5–9 years old	26	21.7
10–14 years old	25	20.8
15–18 years old	24	20.0
Had been referred previously		
Yes	113	94.2
No	7	5.8
Had done surgery previously		
Yes	95	79.2
No	25	20.8
Number of surgeries (for those who had done it)		
Once	66	69.47
Twice	24	25.26
Three times or more	5	5.26

Table 3
The reason of revision in 39 patients who needed surgery from 95 patients who had done surgery before referring to us (some patients had two or complications).

Reason of surgery	Number	Percentage
Unfavorable scar	25	26.31%
Vermilion notch (whistle)	14	14.73%
Short lip (contracture)	21	22.10%
Tight lip	8	8.42%
Oro-nasal Fistula	7	7.36%
Total	39	41.05%

4. Discussion

Our results revealed that prevalence of cleft lip and palate is 1.37 per thousand in Zanjan province, northwest of Iran. Only 79.2% of patients had done the primary surgery, of which 46.7% were operated in the first year of their life. 39 (41.05%) patients that had done surgery were not satisfied of the result. An important finding was the number of non-operated patients or late presentations (25 cases, 2.86 per 10,000). To the authors' knowledge, this has not been reported by any study in Iran. Although 94.2% (113 patients) had been referred by the referral healthcare system for treatment, only 79.2% (95 patients) had done the primary surgery. This has been due to cultural and economic problems.

Some studies have reported different prevalence in Iran. For example one has reported that these anomalies are more prevalent in the southwest of Iran.[12] According to Golalipour et al.[13] the overall prevalence of oral cleft is 0.97 per thousand live births in Iran. Regarding parents' ethnicity, the prevalence of oral cleft is 0.86, 0.88, and 1.47 per 1000 in Fars, Turkman, and Sistani ethnicities, respectively. This prevalence varies from 0.3 to 2.4 per 1000 live birth in other Middle-Eastern countries according to

Table 1
Prevalence of identified cleft lip and palate patients in different cities of Zanjan province.

Cities	Total number of people younger than 18 years old	Number of people with lip and palate clefts	Prevalence per thousand
Khodabandeh (near Hamedan province)	36,628	65 (54.2%)	1.77
Zanjan	14,636	16 (13.3%)	1.09
Ijrud (near East Azerbaijan province)	11,255	10 (8.3%)	0.88
Tarom (near Guilan province)	8,687	7 (5.8%)	0.80
Abhar (near Qazvin province)	8,665	11 (9.2%)	1.26
Mahneshan (near Kurdistan province)	8,485	11 (9.2%)	1.29
Total	87,356	120 (100%)	1.37

Sabbagh et al.[14] In the investigation by Yassaei et al.,[15] the prevalence of cleft lip and palate in Iran was 0.85 per 1000 live births. Moreover, Jamilian and Babaiian[16] reported a prevalence of 2.14 per 1000 live births in a hospital in Tehran.

These statistics suggest that the prevalence of these disorders vary in different parts of the world and highlight the role of race and genetics in their emergence. But the important point is that many of these reports have been retrospective or in short period of time. In this study we studied the clefts in a 17-year life period and found different prevalence in different age groups. For example, the prevalence was 1.97 per thousand in the children younger than four years old but considering all age groups the prevalence was 1.37. This shows that prevalence of lip and palate clefts may differ in different years. The difference was significant between younger than four years old age group and the other three age groups.

Sah and Powar[6] stated that patients' age at referral time range from one day to 71 years with a median age of two years old. Cleft lip, cleft lip and palate, cleft palate and rare clefts were noted in 21.7%, 61.1%, 16% and 1.1% cases, respectively. Only 1.3% of cases had a family history. According to Cooper et al.[17] the prevalence rates of cleft lip were 1.30 per thousand in Chinese, 1.34 in Japanese, 1.47 in other Asians, and 1.33 in all Asia. In our study we only considered people younger than 18 years old and the prevalence was 1.37 per thousand. But the type of cleft and palate was similar to Sah and Powar[6] and Rajabian and Aghaei.[18]

In the research by Sabbagh et al.[14] the prevalence of cleft lip and palate was higher among men than women in Saudi Arabia and its neighboring countries, whereas isolated cleft lip was more common in women than men. In addition, the eight-fold difference between the highest and lowest reported prevalence of facial clefts was attributed to genetic and contextual/environmental factors. Rajabian and Aghaei[18] reported a prevalence of 0.8 per 1000 live births for cleft lip and palate in the southwest provinces of Iran with a 1.25 men to women ratio. Their findings suggest that the prevalence of cleft lip and palate in the southwest of Iran is similar to the low prevalence of cleft lip and palate in African countries and it is also lower than the prevalence of cleft lip and palate among Caucasian whites.

In Sah and Powar's study[6] on clefts, 55.7% were men and 44.3% were women. In Kling and colleagues' study[19] 63.5% of cases were men. In our research, 70% of identified patients were boys. In a study by Yassaei et al.[15] prevalence of cleft lip and palate was 0.86 in a thousand live births. In their study 30.4%, 23.2%, and 46.4% of patients suffered from cleft palate, cleft lip, and cleft lip and palate, respectively. Isolated cleft lip was also more common among women. Blanco-Davila[20] found 1.1 per thousand incidences of cleft lip and palate in Mexico. Clefts of the left side occurred more often. The highest incidence was found in patients of one to six months old. This was similar to our study in that the highest incidence was seen in children younger than four years old.

Sitzman et al.[21] studied 130 children with cleft lip and palate in North America. The children had been operated in different centers and were younger than 18 years old. According to their results, there was a considerable difference between rates of secondary lip operation in different centers. In their research, 53%, 33%, and 19% of patients were exposed to secondary rhinoplasty, secondary lip, and secondary palate surgery, respectively. However, in our study only 41.05% of patients needed secondary lip surgery. The most common deformity was unfavorable scar. Zhang et al.[10] reported an overall complication rate of 16.8% for their previously operated cleft lip and palate patients. Cheema and Asim[22] stated that secondary correction in 189 cases was the most common in the second decade of life with 82 cases. The most common deformity was unfavorable scar in 150 cases followed by notch at the vermilion border in 124 cases. Short lip was found in 119 cases. Complete revision of the repair was required in 158 cases and 25 cases required partial redo. In our study 39 patients did secondary surgery which is less than Cheema and Asim[22] and more than Zhang and colleagues' study[10].

Although cleft lip surgery is most effective during childhood, 32.2% of children had not been treated in a study in India.[23] Also, in Kianifar and colleagues' study[24] 22.9% of patients had been operated more than once, whereas in developed countries almost all patients undergo repeated operations. They found 1.9 per thousand live births prevalence for cleft deformities in Northeastern Iran which is close to the findings in East Asian countries. In another study, revision surgery accounted for 24.2% of cleft lip surgeries.[25]. Shu et al.[26] found 1.84 per 10,000 people prevalence for un-repaired cleft lip or palate in Gansu province of China. This had been a disease burden of this province, especially in the rural areas. In our study 25 patients had not been operated even once which is less than Murthy's study[23] and more than Shu and colleagues' study[26]. Because of the high rate of complications of lip and palate clefts in many studies, some surgeons have tried to overcome the problems by suggesting better approaches[27].

Again, an important finding of our study was identification of non-repaired (late presented) cleft lip and palate patients in a country which is said to have a good referral health system for such cases. Also, many of the patients who undergo treatment are not satisfied of the result. This shows that the quality of revision surgeries should be improved. Some of the reasons that 20% of the identified cases did not refer for surgery are related to cultural and economic problems.

5. Conclusion

Although most of the identified patients had been operated before, almost half of them had to undergo surgery again. Given the lack of studies on this topic in Iran, our findings can form the basis for further studies to precisely identify the roots of cleft lip and palate and properly plan to decrease its risk factors. This study implies that Iran's current health system network is not doing its best to find congenital diseases. So, training courses or workshops should be held regarding congenital diseases periodically for the nurses and social workers of small cities and villages to avoid new cases which are not treated until adolescence.

There is a crucial need for several surgeries in cleft lip patients and some patients have not been operated or have not sought treatment yet. Because of the long-term problems of cleft lip and side effects of surgeries on patients' life quality, well-timed and secondary correction of these defects seems necessary. So, their identification and assistance and providing cultural and healthcare trainings are vital for controlling the complications and improving patients' satisfaction and life quality. Further studies are needed to identify reasons of missed detections and why primary surgeries are not satisfying.

Funding

This study was funded by Zanjan University of Medical Sciences (Grant No: A-11-152-1).

Conflict of Interest

None.

Acknowledgment

The authors thank Seyed Muhammed Hussein Mousavinasab for his sincere cooperation in editing this text.

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