



# The necessity of establishment of Craniofacial Anomalies Registry in Iran

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ARTICLE INFO	ABSTRACT
<p><i>Article Type:</i> <b>Editorial</b></p> <p><i>Received:</i> 1 Jan 2015 <i>Revised:</i> 2 Feb 2015 <i>Accepted:</i> 15 Feb 2015</p> <p><i>*Corresponding author:</i> Maedeh Bonabi Community Oral Health Department, School of Dentistry, Tehran University of Medical Sciences, Tehran, Iran.</p> <p><i>Tel:</i> +98-21-88015960 <i>Fax:</i> +98-21-88015961 <i>Email:</i> m-bonabi@razi.tums.ac.ir</p>	<p>Craniofacial anomalies include clefts, synostoses, atrophic abnormalities, neoplasias etc. among which cleft lip and/or palate caused by abnormal facial development during gestation is one of the most prevalent congenital defects. Its overall occurrence is about 1:700 ranging from 0.02 to 4.04 in 1000 live births. There are different etiologic factors considering the cleft cause. In most cases the exact cause is unknown, but it is thought to be a combination of genetic (internal) and environmental (external) factors.</p> <p>A cleft lip and palate can have a profound psychological impact both on the parents and the child. It affects the appearance of the face, also lead to problems with feeding, speech and language and hearing –due to ear infections. Speech and aesthetic concerns seem to have been important factors affecting the health-related quality of life for children with oral clefts. It is obvious that cleft lip and palate can have a substantial impact on the health economics of countries in the developing world.</p> <p>The multispecialty approach to the care of children with clefts is highly recommended. A research registry can be of invaluable assistance to physicians and researchers by providing an available panel of patient information that could assist in understanding the patients they are serving, utilization of health care services, and the design and implementation of research studies to improve patient care.</p> <p>The Cleft Lip/Palate and Craniofacial Anomalies Registry may be functioning with a mission “to promote better understanding of cleft lip/palate and craniofacial anomalies and to improve patient care and health care planning. Data collection can provide better resources for future interventional studies. As it helps to have an accurate picture of the children’s number in need of treatment in a population. Recording the data of cleft patients and treatment teams and their workload is necessary for planning about providing training centers for members of treatment teams.</p> <p><b>Keywords:</b> Craniofacial anomalies, Cleft, Registry, Healthcare</p>

## Introduction

Craniofacial anomalies include clefts, synostoses, atrophic abnormalities, neoplasias etc. among which cleft lip and/or palate is the most common congenital disease [1].

Cleft lip is defined as “congenital failure of the maxillary and median nasal process to fuse, forming a groove or fissure in the lip”. Cleft palate is defined as “congenital fissure in

the roof of the mouth, resulting from incomplete fusion of the palate during embryonic development. It may involve only the uvula or extend through the entire palate” [2].

A cleft lip and palate can have a profound psychological impact both on the parents and the child. It is common for parents to feel guilty, angry, shocked, helpless or

disappointed. On the child side it can affect the appearance of the face, also lead to problems with feeding, speech and language and hearing –due to ear infections [3]. Speech and aesthetic concerns seem to have been important factors affecting the health-related quality of life for children with oral clefts. These factors seem to be more important as children get closer to adolescence (ages 8-12 years), when acceptance by peers becomes more critical [4]. In addition, children with clefts are more likely to develop dental caries. Teeth crowding may lead to this condition due to difficulty of cleaning [3].

### Etiology

There are different etiologic factors considering the cleft cause. In most cases the exact cause is unknown, but it is thought to be a combination of genetic (internal) and environmental (external) factors [5].

**Inherited genes:** Research indicates that the genes children inherit from their parents make them more vulnerable to developing a cleft lip or palate. A number of genes have been identified that may be responsible. In some cases there is a family history of clefts, although most children of parents with clefts will not develop them.

**Environmental risk factors:** A number of things have been identified that may increase a child's chance of being born with a cleft lip or palate: lack of folic acid during pregnancy, socio-economic status (SES), smoking, alcohol consumption, obesity, malnutrition, and medications during pregnancy.

Cleft lip and palate may be accompanied by other syndromes (for instance Pierre Robin syndrome) or birth defects [6-9].

### Incidence

Cleft lip and palate, caused by abnormal facial development during gestation, is one of the most prevalent congenital defects. Its overall occurrence is 1:700 (0.02 to 4.04 in 1000) live births [10-15]. Among those patients with cleft lip and palate, 75% are involved with the alveolar cleft and are in need of extensive treatment and rehabilitation of the defect and occlusion [16,17].

The incidence varies by ethnic group and gender. Isolated orofacial clefts, or clefts that occur with no other birth defects, are one of the most common birth defects about 70% of

**Table 1.** Incidence rates of cleft lip and palate in different races.

Race	Incidence Rate (in 1000 birth)
Japanese	0.82 to 3.36
Caucasian	1.43 to 1.86
Chinese	1.43 to 4.04
African	0.18 to 1.67
British	1.42
Norwegian*	1.9
Latin Americans	1.04

\*Highest rate in Western world.

all orofacial clefts be isolated clefts [18].

The Centers for Disease Control and Prevention (CDC) estimated that each year 2,651 babies are born with a cleft palate in the United States and 4,437 babies are born with a cleft lip with or without a cleft palate [18]. The highest prevalence rates for (CL±P) are reported for Native Americans and Asians. Africans have the lowest prevalence rates [19]. Cleft lip is more common in males while cleft palate is more common in females [20]. In general cleft lip is more common than cleft palate.

Incidence rates of cleft lip and palate in different races are shown in Table 1 [2,14,15]. According to non-official data in Iran, there are 1200 infants who are born with cleft lip and palate each year while according to the 2011 population census the live births per year was 1274000 from 2005 to 2010 [21]. This will result in at least 1 in 1000 birth incidence for this anomaly in our country. The high prevalence for this congenital anomaly speaks for providing an organization to support these in-needs of care individuals.

### The team approach

The multispecialty approach to the care of children with clefts is highly recommended. The needs of cleft children are multifactorial, and dealing with them as an independent practitioner is often to the detriment of the patient. The need for management of cleft lip and palate deformities at multiple levels is easy to appreciate when one begins to list the functional and anatomic areas affected by the deformity.

The craniofacial team is composed of nursing and physician specialists with particular interest and training in the care of children with cleft deformities [22]. Each

**Table 2.** The craniofacial team for provision of care of children with cleft deformities.

Feeding specialist	Assesses and manages feeding issues related to a cleft diagnosis
Nurse coordinator	Coordinates the multispecialty care and management of the patient
Plastic surgeon/ Oral & maxillofacial surgeon	Executes surgical procedures related to the cleft lip/palate, orthognathics, velopharyngeal insufficiency, and nose
Speech therapist	Diagnoses and treats disorders of speech
Otolaryngologist	Assesses auditory issues, tympanic membrane management
Dentist	Prevents and treats tooth and gum disorders and diseases
Orthodontist	Corrects irregularities of tooth position
Prosthodontist	Replaces teeth and makes dental and alveolar molding devices
Geneticist	Assesses and diagnoses genetically linked diseases and disorders
Social worker	Provides social services such as insurance needs

member task is mentioned in Table 2 [4,20,22]:

Team members may be less than what is mentioned. In one study the proposed team consisted of family physician, plastic or maxillofacial surgeon, orthodontist, prosthodontist, otolaryngologist, speech therapist, audiologist and social worker [23, 24].

Also a team of 4 members (Orthodontics, Plastic Surgery, Maxillofacial Surgery and Speech and Language Therapy) was latterly suggested by Mark Hammond in 1999 [25].

### **Economic points of view**

It is obvious that cleft lip and palate can have a substantial impact on the health economics of countries in the developing world. In one study, economic modeling of cleft lip and palate in sub-Saharan Africa with retrospective demographic and economic data from 2008 was performed to assess financial profitability potential. The total number of Disability-Adjusted Life-Years (DALYs) secondary to cleft lip and palate in 2008 was calculated from accepted cleft incidence rates and disability weights taken from the Global Burden of Disease Project. DALYs were then converted to monetary terms (\$US), using both a human capital approach and Value of a Statistical Life (VSL) approach. With the human capital approach, the potential economic benefit if all incident cases of CLP (Cleft Lip and Palate) in SSA in 2008 were repaired at birth ranged from \$252 million to \$441 million. With VSL, the potential economic benefit of the same CLP repair would range from \$5.4 to \$9.7 billion [26]. These facts and figures show deep impact of consideration and launching craniofacial registry for prevention of resources waste in developing countries.

### **Necessity of data collection**

A research registry can be of invaluable assistance to physicians and researchers by providing an available panel of patient information that could assist in understanding the patients they are serving to, utilization of health care services, and the design and implementation of research studies to improve patient care [27].

The Cleft Lip/Palate and Craniofacial Anomalies Registry may be functioning with a mission “to promote better understanding of cleft lip/palate and craniofacial anomalies and to improve patient care and health care planning [2].

There are several important reasons why we should collect data.

1. **Research and audit**; there are a small number of cleft patients born in each Health District, thus pooling of data will in the future produce a larger resource for research, often making statistical significance greater. The emerging possibilities offered by clinical and molecular genetics require a clearly defined family genetic pedigree, to assess effectiveness of any new treatments.

2. **Audit**; the recording of consecutive cases of a condition helps in preparing a series of patients with similar cleft deformities and allows comparison for audit purposes, in order to assess the effectiveness of treatment and set standards for the future.

3. **Purchasing and planning cleft services**; recording the incidence of birth deformities allows for the planning and provision of services to cater for these children. It has generally been regarded that the incidence of cleft subtypes in the United Kingdom is about 1:700. It is important to have accurate figures for the number of children requiring treatment to perform a population-based needs

assessment.

4. **Care**; records cleft registrations against the hospitals and cleft teams that treat these cases. It therefore has information about cleft teams and workload that is useful in planning training centers for future cleft team members [22].

### Discussion

The FDI World Dental Association and the World Health Organization (WHO) established the first Global Oral Health Goals jointly in 1981 to be achieved by year 2000. Recently, the FDI, WHO and IADR (International Association of Dental Research) have embarked on the activity of preparing goals of the new millennium, for year 2020. This document illuminates 16 objectives which in the 8th objective, craniofacial anomalies had been mentioned in terms of: “Reducing exposure to risk factors with special reference to tobacco, alcohol, teratogenic agents and improved nutrition, increasing access to genetic screening and counseling, increasing early detection, increasing rapid referral, increasing the number of affected individuals receiving multidisciplinary specialist care, increasing early detection of seriously handicapping malocclusions and their referral” [27]. Also the objective number 16 speaks for using Healthcare information systems [28]. Increasing the proportion of population covered by satisfactory information system is one the aims of this document.

To fulfill these aims the authors suggest the ministry of health to initiate the establishment of craniofacial anomalies registry. It was suggested in cancer registry establishment in Iran to execute the registry in a limited district in the first phase of establishment [29]. As cleft is a less prevalent disease, we may register craniofacial anomalies in the whole country to be able to provide enough data for analyses in the frame of a national program. This registry may reduce the social and economic costs of lip and palate clefts for the individuals and the community.

### Conclusion

Because craniofacial anomalies are one of the affecting problems in personal and social life of a patient, it needs to be registered at birth of a person to facilitate providing needed services. Data collection can provide better

resources for future interventional studies as it helps to have an accurate picture of the children in need of treatment in a population. Recording the data of cleft patients and treatment teams and their workload is necessary for planning about providing training centers for members of treatment teams.

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