

Incidence of Orofacial Clefts in Croatia from 1988 to 1998

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Summary

No similar study on the incidence of orofacial clefts has been conducted in Croatia to date. This study included children born between January 1, 1988 and December 31, 1998, to mothers resident in Croatia. During the 11-year study period there were 525,298 live-births. A review of medical records on live-births from neonatology units and medical documentation from institutions providing surgical treatment for children with orofacial clefts revealed 903 children born with clefts, 24 (2.65%) of them twins, yielding a cleft incidence of 1.71 per 1000 or 1/581 for the study period. When "syndrome" clefts were excluded the incidence of "non-syndrome" clefts was 1.56 per 1000 or 1/641. During the study period the incidence of orofacial clefts varied from 1.43 per 1000 to 2.02 per 1000. Considering cleft lip with or without cleft palate (CL±P) and isolated cleft palate (CP) separately, the incidence of CL±P and CP was 1.05 per 1000 and 0.66 per 1000, respectively. According to districts of Croatia, the highest incidence of orofacial clefts was recorded in central Croatia (1.88/1000) and lowest in Lika and north Primorje (0.91/1000). According to sex, there were 56.4% male and 43.6% female children born with orofacial cleft, yielding a 1.29:1 male to female ratio. CL±P was more common in the male population and CP in the female population.

Key words: cleft lip, cleft lip and palate, cleft palate, epidemiology, incidence.

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Introduction

Orofacial clefts are the most frequent malformations in the facial area, in all populations and ethnic groups. Disassociation of the skin, muscles, bones and cartilage represents an aesthetic and functional problem, and team therapy is necessary

(including several medical and dental specialists) longitudinal and complex (1-4). Clefts may occasionally be accompanied by other congenital anomalies.

Immediately after birth as much information as possible should be offered to the parents of the child with such malformation, in order for them to be cor-

rectly oriented at the very start of the child's life, providing adequate and prompt care (5). Improved ultrasound diagnostics, and particularly more recently application of three-dimensional ultrasound, enables recognition of the anomaly during pregnancy (at the earliest from the 15th week of gestation) enabling sufficient time for the parents to acquire basic knowledge of this malformation (6). Three-dimensional ultrasound is an excellent method for detection of smaller surface malformations of the soft tissues, particularly in early pregnancy, when there is a physiologically greater amount of amniotic liquid, which enables better presentation, and also the foetus is smaller, frequently changes position and is easier to present from all sides. During routine examinations, such diagnostic methods are not applied, because the method requires more time and skill (knowledge) of the examiner. This type of examination is recommended in cases when there is a positive family history (cleft in the family) or when malformation is suspected during routine ultrasound examination.

The frequency of cleft in Europe ranges from 1.0/1000 to 2.21/1000 live-births (7). The incidence of orofacial clefts in Iran is 1.13/1000, i.e. for every 885 live-births one child is born with cleft (8,9). In North America it amounts to 1.71/1000 live-births (highest for the Caucasian population - 1.8/1000, and lowest for Afro-Americans - 0.56/1000) (10). The incidence in Canada amounts to 1.06 - 1.97/1000, while in Australia it is 1.21 - 1.73/1000 (11). The incidence of orofacial clefts in South America is 1/1000 live-births (12).

Analysed separately, the incidence of isolated cleft lip (CL) ranges from 0.29 - 0.45/1000; cleft lip and palate (CLP) from 0.71 - 1.29 per one thousand live-births. While the incidence of isolated cleft palate (CP) ranges from 0.19 - 0.83 per one thousand live-births (11).

Materials and methods

An epidemiological study was conducted during the period from 1988 to 1998. Due to the fact that in Croatia there is no integral register of malformations, the study was performed on the basis of data obtained from the neonatological departments of gynaecological-maternity departments of hospital

centres in all regions in the Republic of Croatia, 23 hospital centres and two private institutions. The study included only children born to mothers with permanent residence in Croatia.

The data obtained was supplemented with data from institutions which provide treatment for persons with orofacial clefts in the Republic of Croatia and also the Republic of Slovenia, because a small number of persons with cleft were treated in Ljubljana (13).

- University Hospital Dubrava, Clinic of Maxillofacial Surgery.
- Children's Hospital Zagreb, Department of Surgery.
- University Hospital Osijek, Department of Maxillofacial Surgery.
- Clinical Hospital Centre Rijeka, Clinic of Maxillofacial Surgery.
- Clinical Hospital Centre Ljubljana, Clinic of Maxillofacial Surgery, Republic of Slovenia.

For additional data the following patient registers were used:

- School of Dental Medicine University of Zagreb, Department of Orthodontics.
- Dental Polyclinic in Zagreb, Department of Orthodontics.
- Clinical Hospital Centre Šalata, Centre of Audiology and Phoniatrics.

During the study data were taken for patients born with orofacial cleft as a single diagnosis and for patients in whom cleft was one of the signs of a malformation syndrome.

Data on the number of children born in Croatia and other necessary data were obtained from the Institute for Statistics of the Republic of Croatia, and were supplemented with data obtained from the Institute for National Health of the Republic of Croatia.

For analysis of the incidence of orofacial clefts in Croatia according to geographical regions, we separated four regions according to the formula of the Institute for Statistics of the Republic of Croatia, as follows:

- | | |
|-------------|------------|
| 1. Slavonia | South-east |
| | North-east |
| | Podravska |
| | Posavska |

| | |
|----------------------------|--|
| 2. Central Croatia | Bilogora Međimurje and Varaždin region Croatian zagorje Upper Posavina Zagreb Banija and Pokuplje |
| 3. Lika and northern coast | Gorski kotar and Lika Croatian coastal region Istria |
| 4. Dalmatia | Dalmatian highlands Northern Dalmatia Southern Dalmatia |

During the analysis the Republic of Croatia was divided into 21 districts. The data obtained was subjected to statistical analysis (χ^2 - test).

Results

In Croatia, during the period from January 1988 to December 1998, according to the report of the Institute for Statistics, the total number of live-births born to mothers with permanent residence in Croatia was 525,298. According to available medical documents on live-births from neonatological centres in Croatia, and medical documents of centres in Croatia and Slovenia, which treat children born with orofacial clefts, there were 903 children registered with this malformation during the above mentioned period. Of the identified 903 patients with orofacial clefts in the studied 11 years, 24 patients were twins, which amounts to 2.65%.

Figure 1 represents a three-dimensional presentation of children with cleft, classified according to gender and year of birth.

In Croatia operations on children with orofacial cleft are carried out in four centres. The majority of such operations are performed in the University Hospital "Dubrava" in Zagreb (694:76.85%). Fifty patients (5.53%) were operated on in the Children's Hospital in Zagreb, 17 (1.88%) in the Clinical Hospital Osijek, 27 (3%) in the Clinical Hospital Split, and occasionally in the Clinical Hospital Centre in Rijeka and in other health centres.

Thirty-seven patients (4.1%) were treated in the Clinical Hospital Centre in Ljubljana, which gen-

erally refers to operations performed up until 1991. After which only sporadic cases were recorded.

The frequency of congenital anomalies is expressed by the incidence. The incidence is the number of new cases of malformations (orofacial clefts) during a specific period in units of time (e.g. one year), and expressed as "annually per 1000", "annually per 10000" or "annually per 1000000".

Of the total number of 525,298 live-births there were 903 children with a diagnosis of orofacial cleft, i.e. 1.717/1000 live-births. Thus, for each 581 live-births one child on average was born with a cleft. The incidence changes from year to year, and ranges from 1.43/1000 in 1989 to 2.02/1000 in 1992. If we exclude syndrome clefts (which are connected with chromosomal aberrations, monogenic syndromes and multiple congenital anomalies) then the incidence is 1.56/1000, i.e. for each 641 live-births one child on average is born with orofacial cleft, as an isolated anomaly ("non-syndrome" cleft). If we analyse separately cleft lip with or without cleft palate (CL±P) and isolated cleft palate (CP) the incidence is 1.0489/1000 for cleft lip with or without cleft palate, and 0.657/1000 for isolated cleft palate respectively.

Of the total number of children with a cleft 509 (56.4%) were male and 394 (43.6%) female. During the studied 11-year period the incidence of cleft for male children was 1.83/1000 and for female children 1.47/1000.

The incidence of orofacial clefts in Croatia is presented in Figure 2.

Croatia is divided into 21 districts. The majority of children born with a cleft came from Zagreb and the Zagreb district (29.2%), followed by the Split-Dalmatian district (10.0%) and the Osijek-Baranja district (8.1%).

Distribution of children born with a cleft, included in this study, according to districts and gender, is shown in Table 1.

If we divide Croatia into 4 geographical regions (Table 2) we can see that the incidence in certain geographical regions differs significantly, both between regions and within certain regions during the studied period. The highest incidence was recorded in central Croatia and changed over the

years from 2.3‰ (1990) to 1.49‰ (1989 and 1998). The lowest incidence was recorded in Lika and north Primorje, from 0.2‰ (1993 - war year) to 2.0‰ (1996).

The number of children born with orofacial cleft in a specific year, in each region, is shown in Table 2.

Discussion

The incidence of orofacial clefts in Croatia during the period from 1988 to 1998 amounts to 1.71 per 1000 live-births. If we separate "syndrome" clefts from "non-syndrome" the incidence for "non-syndrome" clefts amounts to 1.56/1000. In studies performed in European countries incidence was recorded from 1.30 to 1.94 per 1000 live-births (14; 7; 15; 11). In an epidemiological study carried out in Slovenia during the period from 1973 to 1993 the incidence of clefts recorded was 1.64/1000 live-births (16,17), while the incidence in northern Italy for the period from 1978 to 1986 amounted to 1.33/1000 (18). Both data are slightly lower than the values in our study. A lower incidence was recorded in epidemiological studies in France 1.17/1000 (19), Belgium 1.30/1000 (7), south-east Scotland 1.4/1000 (20,21) and 1.53/1000 west Scotland (22); in Great Britain 1.40/1000 (when syndromes and congenital anomalies are excluded) i.e. 1.67/1000 (including syndromes and isolated orofacial clefts) (23), in Hungary 1.45/1000 (15) and in East Berlin 1.48/1000 (15). For Finland Rintala (24) reports the incidence of cleft 1.74/1000, while for Sweden Millerad et al (25) found 1.7/1000, which agrees with the findings in our study. A higher incidence was recorded in Denmark 1.89/1000 (26), i.e. 2.1/1000 (27), in Norway 1.92 - 2.08/1000 (28) and in Czechoslovakia 1.81/1000 ("non-syndrome") i.e. 2.03/1000 for all types of clefts (29), and the former Democratic Republic of Germany 1.85/1000 (15).

Analysed separately, the incidence of cleft lip and cleft lip and palate (CL±P) ranges from 0.6 - 1.6 per 1000 live-births, while the incidence of isolated cleft palate (CP) ranges from 0.27 - 1.21 per 1000 live-births. According to our findings the incidence was 1.0489/1000 for cleft lip with or without cleft palate (CL±P), and 0.657/1000 for isolated cleft

palate (CP), which does not differ from the values of other authors: Long et al (19) 0.67/1000 for CL±P and 0.58/1000 for CP, Fitzpatrick et al (22) 0.74/1000 for CL±P and 0.79/1000 for CP, Calzolari et al (18) 0.75/1000 for CL±P, and 0.58/1000 for CP, Owens et al (23) 0.93/1000 for CL±P and 0.47/1000 for CP, Croen et al (10) 1.05/1000 for CL±P and 0.66/1000 for CP, Tolarova (29) 1.22/1000 for CL±P and 0.6/1000 for CP, Christensen (27) 1.4-1.5/1000 for CL±P and 0.7-0.9/1000 for CP.

The incidence of isolated cleft palate (CP) appears to vary more within the same primary population groups, apart from black populations, and significantly less among population groups. This finding, and also the correlation of CP with the age of the mother and also multiple births (number of births), suggests that external factors more than genetic are included in the occurrence of isolated cleft palate (CP) compared to cleft lip (CL) (30).

Differences in the data obtained, presented here only for the Caucasian population, can to a certain extent be explained by the different sources of data (neonatal register, register of operations), and also the selection of data. Obviously, differences in the incidence depend on the inclusion of live-births only, the inclusion of the stillborn, i.e. the gestation age of the stillborn. In this study only data for live-births were used, and during classification submucosal cleft palate was also included as a true cleft. When calculating incidence some epidemiological studies only used data for orofacial clefts, which occur as isolated anomalies, which in that case is emphasised.

Of the identified 903 patients with orofacial cleft during the period from 1988 to 1998, 24 patients were twins, which amounts to 2.65%, and is identical with the results of Menegotta et al (12) of 2.6%, and with Van den Akker (7) 2.7%. Jensen et al (26) in Denmark and Rintala (24) in Finland found a slightly lower percentage - 1.6% of twins in a group of children born with orofacial cleft (from 1950 to 1982).

The data from this study show that the largest number of children born with a cleft were from Zagreb and the Zagreb district (29.2%), Split-Dalmatian district (10.0%) and the Osijek-Baranja dis-

trict (8.1%). It can be seen that there was a greater concentration of children born with this malformation in the region of large towns and agronomic areas of Croatia, which agrees with the results of Koželj (17) for Slovenia. The lowest incidence was recorded in industrially and agriculturally undeveloped regions (Ličko-Senjska district), which also agrees with the results obtained for Slovenia (17).

When Croatia is divided into regions the largest number of children with orofacial cleft were born in the region of central Croatia; as many as 54.37%, where the highest incidence was recorded (1.88/1000) and the lowest incidence in Lika and north Primorje (6.53%) with incidence of only 0.91/1000.

During the years of the war (1991-1995) increased incidence of cleft was observed compared to pre-war and post-war years.

Conclusions

1. This epidemiological study included all children born with orofacial cleft during the period from 1 January 1988 to 31 December 1998, i.e. a period of 11 years.
2. The incidence for the aforementioned period was 1/581, i.e. 1.717/1000, which differed from year to year and changed from 1.43‰ (1989) to 2.02‰ (1992).
3. If we exclude "syndrome" clefts, the incidence was 1.56‰, or for each 641 live-births one child on average was born with orofacial cleft, as an isolated anomaly ("non-syndrome" cleft).
4. The incidence according to geographical areas for the same period ranged from 0.91‰ in Lika and north Primorje to 1.88‰ in central Croatia.