

The incidence of congenital anomalies associated with cleft palate/cleft lip and palate in neonates in the Konya region, Turkey

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Abstract

Additional congenital anomalies have often been found in patients with orofacial clefts. We wanted to find out the incidence and type of congenital malformations that may accompany cleft palate (CP) and cleft lip and palate (CLP) in babies born in the Konya region. A total of 121 newborn babies with CP or CLP were prospectively included in the study, and all were assessed in detail for congenital anomalies. Of 121 babies, 86 (71%) had CLP and 35 (29%) had CP. There was at least one congenital malformation in 80 (66%) of the cases. Additional congenital malformations were seen in 26 (74%) of the 35 with isolated CP, and 54 (63%) in the 86 patients with CLP ($p < 0.05$). The most common congenital malformation was congenital heart disease, followed by head and neck anomalies. The most common congenital heart disease was atrial septal defect. A serious chromosomal anomaly was found in 18/121 patients with CP or CLP (15%). Of the 80 babies in whom congenital malformations were found, 31 (39%) had dysmorphic features. While 21 (68%) of dysmorphic cases had isolated CP, 10 (32%) had CLP ($p < 0.05$). The rates of premature delivery, intrauterine growth retardation, and consanguinity between parents were higher in patients with CP or CLP. The neonatal mortality was 20% ($n = 24$). Our results indicate that at least one congenital anomaly is also present in about two-thirds of newborn babies with CP and CLP, and these anomalies significantly increase their morbidity and mortality. All newborn babies with CP and CLP should be screened for additional congenital anomalies, particularly of the cardiovascular system.

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Introduction

Orofacial clefts are among the most common congenital malformations of the craniofacial region.^{1,2} The incidence of cleft lip and palate (CLP) is 1/500–2000 livebirths.^{3–6} Despite the fact that clefts have not been completely explained, many

genetic and environmental factors have been held responsible for their aetiology and pathogenesis. Mother's smoking and alcohol consumption, the use of vitamins and anticonvulsants, and a syndrome-malformation complex, have been held responsible.^{7,8} Ethnic factors may also have a role. The incidence of CLP is highest among Asians, but it was found to be lowest among the black population.⁹ As well as genetic and environmental factors, consanguinity may play a part. Those whose parents have first degree consanguinity are subject to higher risks in terms of congenital anomalies.^{1,10,11}

There is a close relation between cleft palate and cleft lip and palate (CP and CLP) and the incidence of other congenital anomalies, and the incidence of additional congenital anomalies is between 1.5% and 63.4%.^{1,10,11} Congenital mal-

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formations and growth retardation are common, particularly among those with isolated CP.^{9,12–19}

The aim of this study was to find out the incidence of congenital malformations that occur together with CP and CLP in Konya, Turkey.

Patients and methods

We prospectively studied 121 patients who were admitted between January 2005 and April 2011 to the newborn unit of Selcuk University Meram Medical Faculty, Konya, Turkey, with the diagnosis of CP or CLP. Sex, gestational week, birth weight, age of parents, consanguinity between parents, site of CLP, accompanying anomalies, risk factors, and accompanying syndromes were recorded. A detailed physical examination was made of all babies. Those with emergency conditions such as respiratory distress or cardiovascular decompensation were immediately treated and recorded. Abdominal ultrasonography, cranial imaging, echocardiography, whole body radiography, and chromosomal analysis were done for all patients.

Babies with either CLP or CP alone were included in the study. They were admitted for difficulty in feeding, placement of a palatal prosthesis, accompanying anomalies, or an emergency condition such as respiratory distress that required immediate treatment. Babies with isolated cleft lip (CL) were excluded from the study as they are not usually admitted to hospital, and most are not examined in detail. Babies who could not be examined in detail (those who died or were transferred to another clinic) were excluded from the study.

The data collected were analysed with the help of the Statistical Package for the Social Sciences (version 17.0, SPSS Inc, Chicago, IL, USA). The results are displayed as mean (SD), or number (%). We used the Mann–Whitney *U*-test to assess the significance of differences among quantitative data, and probabilities of less than 0.05 were accepted as significant.

Results

A total of 128 babies who were admitted between January 2005 and April 2011 to the newborn unit with the diagnosis of CP or CLP were evaluated. Of the babies, 121 from whom sufficient data were collected and sufficient examinations were made were included in the study. The remaining 7 babies were excluded as insufficient data were collected, or they died before adequate examinations could be made.

Table 1 shows the details of the patients. Eighteen (15%) had major chromosomal anomalies (such as trisomy 13, 18, and 21). Thirty-four babies (28%) who had been admitted with CP or CLP had respiratory distress. Of these 34 babies, 18 (53%) had respiratory distress syndrome of prematurity, 10 (28%) had transient tachypnoea of the newborn, 3 (9%)

Table 1

Details of patients. Data are number (%) or mean (SD).

	Number (%) or mean (SD)
Sex	
Male/female	56/65 (%46/%54)
Birthweight (g)	2390 ± (87)
Gestation (weeks)	36.2 ± (3.9)
Maternal age (years)	31.2 ± (3.6)
Mode of delivery	
Caesarean section	81 (%67)
Normal	40 (%33)
Type of cleft	
Cleft lip and palate	86 (%71)
Cleft palate	35 (%29)
Type of cleft lip and palate:	
Bilateral	17 (%20)
Unilateral	69 (%80)

had persistent pulmonary hypertension, 2 (6%) had aspirated meconium, and 1 (3%) had a diaphragmatic hernia.

Accompanying malformations

At least one congenital malformation coexisted with CL or CLP in 80 (66%) of the 121 babies. Of those with malformations, 42 (52%) were boys and 38 (48%) were girls. Congenital malformations were seen in 26 (74%) of the 35 patients with isolated CP, but in 54 (63%) of the 86 patients with CLP ($p < 0.05$). Thirty-one (39%) of the 80 patients with CP or CLP and coexisting congenital malformations had dysmorphic features. Syndromes such as Smith–Lemli–Opitz, Stickler, Pierre–Robin, and Apert; and trisomy 13, 18, and 21 and achondroplasia accompanied the cases with dysmorphic features (Tables 2 and 3).

While the rate of consanguinity in the babies with CP and CLP who also had congenital malformations was 50 (62%), this rate was 14 (35%) for those without congenital malformations ($p < 0.05$). The rate of consanguinity was 64 (53%) among all babies with CP and CLP with or without congenital malformations. There was first degree consanguinity in 42 (66%) of the cases with parental consanguinity. The rate of family history of CP or CLP was 19/121 (16%) regardless of congenital malformation. More than one system or organ were involved in 38 (48%) of those with congenital anomalies.

Among our cases, there was only one mother who drank alcohol during pregnancy, and she drank only a small amount

Table 2

Distribution of accompanying malformations in 80 patients with CP or CLP.

Malformation	Number (%)
Congenital heart disease	29 (36)
Anomalies of the head and neck	28 (35)
Anomalies of the skeleton	17 (21)
Anomalies of the central nervous system	14 (17)
Urogenital anomalies	13 (16)
Anomalies of other systems	12 (15)

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