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Distribution of congenital heart disease in Turkey*

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Aim: To describe the epidemiology and geographical distribution of congenital heart disease. A retrospective population-based study was conducted using data collected in a large tertiary care hospital in Turkey.

Materials and methods: The medical records of 1300 patients with congenital heart disease admitted to the Gazi University Department of Pediatric Cardiology for catheter angiography from 1997-2007 were reviewed. The patients were divided into 7 groups according to geographical distribution, and each group was further divided into 2 groups, for simple (group 1) and complex (group 2) congenital heart disease.

Results: Among the 1300 cases, there was no difference between the regions regarding mean age, mean diagnostic age, and female/male (F/M) ratio. In 3 regions that have similar geographic and climatic features, a high incidence of complex cardiac malformations was observed ($P < 0.05$).

Conclusion: This is the first epidemiological study of the distribution of congenital heart disease in Turkey. Although etiological data is not sufficient to explain the findings, this study may play an important role in further prospective studies about this issue.

Key words: Congenital heart disease, epidemiology, risk factors

Türkiye'de doğumsal kalp hastalıklarının dağılımı

Amaç: Türkiye'de doğumsal kalp hastalıklarının epidemiyoloji ve bölgesel dağılımlarının belirlenmesi amacıyla üçüncü basamak bir hastanenin kayıtları geriye dönük olarak değerlendirildi.

Yöntem ve gereç: Gazi Üniversitesi Tıp Fakültesi Çocuk Kardiyoloji Bilim Dalı'na 1997-2007 yılları arasında kateter-anjiografi için gönderilen 1300 doğumsal kalp hastalığı olan hasta incelendi. Hastalar başvuru bölgelerine göre coğrafi olarak sınıflandırıldı ve her grup kendi içinde basit doğumsal kalp hastalıkları (grup 1) ve komplex doğumsal kalp hastalıkları (grup 2) olarak ikiye ayrıldı.

Bulgular: İncelenen 1300 olgu arasında coğrafi bölgeler arasında ortalama yaş, tanısal yaş ve cinsiyet yönlerinden fark izlenmedi. Benzer coğrafik ve iklim özelliklerine sahip üç bölgede komplex doğumsal kalp hastalıklarının daha sık olduğu gözlendi ($P < 0,05$).

Sonuç: Bu çalışma doğumsal kalp hastalıklarının Türkiye'deki dağılımı ile ilgili ilk epidemiyolojik çalışmadır. Etyolojik veriler bulguları açıklamak için yeterli olmasa da bu çalışmanın bu konuda yapılacak olan çalışmalar için önemli bir kaynak olacağını düşünmekteyiz

Anahtar sözcükler: Doğumsal kalp hastalığı, epidemiyoloji, risk faktörleri

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Introduction

Despite recent developments in interventional and surgical techniques, heart disease in children continues to be an important cause of morbidity and mortality. It is difficult to establish the incidence of congenital heart disease (CHD). Numerous studies show the incidence of this pattern as 0.6-1 (mean 0.8/100) per 100 live births (1). Thus, the prevalence of CHD is not uniform, but variable (2).

Much of the epidemiologic evidence on risk factors for congenital heart defects has been presented as multifactorial. In addition to genetic predisposition, environmental risk factors such as maternal diabetes, teratogens, and maternal phenylketonuria have also been implicated. Environmental physical conditions are known to be associated with many factors of human homeostasis, such as fetal development and some genetic abnormalities. Recently, an interesting report from Israel revealed that congenital heart disease in infants is directly correlated with the level of solar activity and inversely correlated with the level of cosmic ray activity during the pregnancy, predominantly in the month of conception (3).

Documentation of the relative frequencies of different cardiac defects is important for the training of physicians as well as for the management and planning of health care systems. We conducted this study to assess the prevalence and distribution of CHDs with regard to regions of Turkey, as well as age on admission.

Materials and methods

We retrospectively analyzed the echocardiographic and catheter angiography records of 1300 patients who were admitted to the Gazi University Medical Faculty Pediatric Cardiology Unit between January 1997 and December 2007. All patients were evaluated by a pediatric cardiologist with chest X-ray, standard 13-derivation ECG (including V4R derivation), and complete blood count. Echocardiographic examination was conducted using M-mode, 2-dimensional and color, pulse, and continuous wave Doppler echocardiograms with GE Vivid 7 (Wisconsin, USA). In standard parasternal long-axis, short-axis, apical 4-chamber, subcostal, and suprasternal views, 2-dimensional echocardiographic pictures were recorded. Cardiac catheterization and invasive procedures were performed with a GE Advantx LC/LP (Oklahoma, USA) when needed. Neonatal patients with patent ductus arteriosus (PDA) were not included in this study.

Geographic classification of the regions in Turkey was as follows: region 1, Marmara; region 2, Aegean; region 3, Mediterranean; region 4, Central Anatolian; region 5, Black Sea; region 6, East Anatolian; and region 7, Southeast Anatolian (Figure).

Patients were separated into 2 groups. Group 1 was the simple congenital heart disease group and included atrial septal defect (ASD), ventricular

Figure. The geographic regions of Turkey.
Region 1, Marmara; region 2, Aegean; region 3, Mediterranean; region 4, Central Anatolian; region 5, Black Sea; region 6, East Anatolian; and region 7, Southeast Anatolian.

septal defect (VSD), PDA, aortic stenosis (AS), pulmonary stenosis (PS), and coarctation of aorta (CoA). Group 2 was the complex congenital heart disease group and included atrioventricular septal defect (AVSD), tetralogy of Fallot (FT), double outlet right ventricle (DORV), and complex cardiac defects (tricuspid atresia, truncus arteriosus, pulmonary atresia, Ebstein's malformation, single ventricle, and d-transposition of great arteries).

The following age groups were considered: newborns (0-28 days), infants (1 month to 2 years), preschool children (2-5 years), school children (5-12 years), and adolescents (>12 years).

Statistics

SPSS 11.0 (SPSS Inc., Chicago, IL, USA) and MedCalc 7.3.0.1 for Windows were used for analysis. A 2-tailed chi-square test was used for detecting differences among the yearly prevalence rates. $P < 0.05$ was considered significant.

Results

Among the 1300 patients, there were 672 males (51.7%) and the F/M ratio was 0.9:1, with a mean age of 63.128 ± 59.743 (1-312 months). Infants formed the largest group among the age groups (39.3%) (Table 1). There were 717 patients in group 1 and 583 patients in group 2, and age and gender distribution did not differ among groups and regions.

VSD was the most frequent type of CHD (22.2%), as expected. The frequency of other cardiac defects is shown in Table 2. The largest population of patients came from region 4 (57.7%), and the majority of the patients referred to our institute came from region 4 (Table 3). Group 2 patients were most prevalent in regions 3, 6, and 7. This was found to be statistically significant ($P < 0.05$) (Table 3).

Discussion

This study aimed to document the frequency and prevalence of CHD in children referred to a pediatric cardiology tertiary care center in a country with different geographic features. There have been few epidemiological studies about CHDs in Turkey (4,5). Highlights from local data collected from different parts of the world may help to predict the influences and etiological factors of CHDs and the prevalence of the diseases in different ethnic groups. Thus, health system strategies can be developed according to the data provided.

A study from Iceland reported VSD (45.7%) as the most diagnosed heart defect, followed by ASD (12.2%) and AS (1.5%), among 338 patients (6). Similar findings from Saudi Arabia reported VSD in 32.5% of patients, PDA in 15.8%, and ASD 10.4% (7). Among 1693 children in a large tertiary care hospital located in the central Anatolian region of Turkey, the most common acyanotic anomaly

Table 1. Age at diagnosis and regional distribution.

Region	Age (months ± SD)	Neonatal		Infant		Preschool		School		Adolescent	
		n	%	n	%	n	%	n	%	n	%
1	67.875 ± 64.110	2	8.3	9	1.7	8	2.9	9	2.8	5	2.8
2	64.027 ± 54.703	-		10	1.9	15	5.6	7	2.1	5	2.8
3	59.039 ± 58.888	1	4.1	60	11.7	32	11.9	19	5.9	17	9.6
4	65.372 ± 59.684	17	70.8	281	54.9	122	45.6	222	69.1	107	60.4
5	71.626 ± 63.110	-		42	8.2	27	10.1	27	8.4	19	10.7
6	60.443 ± 67.697	2	8.3	26	5.0	16	5.9	9	2.8	8	4.5
7	50.938 ± 54.461	2	8.3	84	16.4	47	17.6	28	8.7	16	9.0
Total	63.128 ± 59.743	24	100	511	100	267	100	321	100	177	100

Table 2. Diagnostic distribution of patients.

Type of congenital heart disease	n	%
Group 1	*717*	*55.2*
VSD	288	22.2
ASD	144	11.1
PDA	120	9.2
CoA	51	3.9
AS	50	3.8
PS	57	4.4
Group 2	*583*	*44.8*
TOF	130	10.0
AVSD	51	3.9
Other	402	30.9
Total	1300	100

Table 3. Correlation of patient groups according to region.

Region	Group 1	Group 2	Total n %	P
1	16	16	32	NS
2	23	14	37	NS
3	58	70	128	0.03*
4	448	302	750	NS
5	63	52	115	NS
6	25	36	61	0.006*
7	84	93	177	0.003*

*: significant difference between groups; NS: not significant.

was isolated VSD (32.6%), and tetralogy of Fallot (5.8%) was the most frequent cyanotic anomaly. The other defects were PDA (15.9%) and ASD (13.1%) (4). Results of several studies from different countries show distributions similar to those in our study, with VSD (22.2%) being the most frequent congenital heart disease, followed by ASD (11.1%), PDA (9.2%), and PS (4.4%) (8-11).

Comparing the age at diagnosis within the regions, no significant difference was found (Table 1).

The majority of referrals to our unit were from region 4, where our hospital is located (57.7%). No significant difference between the referrals from other regions was detected. This means that our center is a referral unit for patients from all regions of Turkey (Table 3).

We observed that patients with complex cardiac defects (Group 2) were mostly from regions 3, 6, and 7 (P = 0.03, P = 0.006, and P = 0.003, respectively) (Table 3). Recently, Stoupel et al. from Israel showed that the risk of infants born with congenital heart disease is directly correlated with the level of solar activity and inversely correlated with the level of cosmic ray activity during pregnancy, predominantly in the month of conception (3). The significant differences between groups 1 and 2 in regions 3, 6, and 7 may be related to the close proximity of these regions to the Middle East and, as a result, to the solar activity in this region.

Another reason for the high incidence of complex congenital heart diseases in the regions mentioned above may be the high frequency of consanguineous marriage in Turkey (12,13). The prevalence of consanguineous marriage was reported to be 40% in regions 6 and 7 in a national study (14).

The etiological factors affecting the high prevalence of complex congenital heart disease in some parts of Turkey should be studied carefully in terms of consanguinity, genetic factors, and environmental conditions.

As this was the first such study in Turkey, we think that this report will lead to new epidemiological studies on this topic.

Limitations

Absence of data concerning consanguineous marriage and prenatal period are limitations in this study.

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