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ASSOCIATION OF HYPERTELORISM IN CHILDREN WITH CONGENITAL ACYANOTIC HEART DISEASES

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ABSTRACT

The study was done over a period of 2012-2013 in the Department of Paediatric Medicine, Medical College Hospital, Kolkata. 100 patients (Cases: 50 patients with congenital acyanotic heart disease. Control: 50 patients without it) of age group 6 months to 16 years were examined clinically. Echocardiography was done. Intercanthal distance was measured by Vernier's sliding caliper. Among 50 cases, 17(34%) male and 12(24%) female patients had hypertelorism. Male predominance is seen in both the group. Children with hypertelorism should be properly screened for congenital acyanotic heart disease to decrease the morbidity.

Keywords: Hypertelorism, Congenital acyanotic heart disease.

INTRODUCTION

Craniofacial anthropometry is a vivid tool and that can guide clinicians in description, diagnosis and surgical treatment of abnormal skeletal and facial patterns. It is principally useful in medical genetics since many of the congenital anomalies like Apert, Down, Turner etc. involve the head region and present at birth, so it can help clinicians to objectively describe what exactly they are seeing (online Medical news today article 2007). The diagnosis of many dysmorphic syndromes is based on advanced cytogenetic and molecular techniques. The recognition of subtle morphological anomalies in craniofacial region may help. Clinicians usually describe dysmorphic characters as "wide set eyes", "broad nose", "large mouth" and "posteriorly placed ear". However, this is a subjective description. The deviations from the normative values can be evaluated by comparing the values obtained from the patient with that of normal population, Naglel et al, (2005). This study was conducted to compare the intercanthal distance of cases with congenital acyanotic heart disease to controls. The normal intercanthal distance is about 2cm. to 3.6cm. Normative values of intercanthal distance could be obtained from different studies done in Nation and International level by different researchers i.e. deviation from that can be indicative of congenital anomalies.

MATERIALS AND METHOD

The study was conducted over a period of one year from July 2012- June 2013 on 100 patients of aged 6 months to 16 years attending the outpatient department (OPD) and or, admitted in the department of Paediatric Medicine, Medical College, Kolkata. Thorough clinical examination of cardiovascular system by inspection, palpation, percussion and auscultation was done. Echocardiography was performed to confirm the diagnosis. Clinical examinations were done in presence of paediatricians. The intercanthal distance (ICD) was measured by vernier's sliding caliper (Fig. 1).

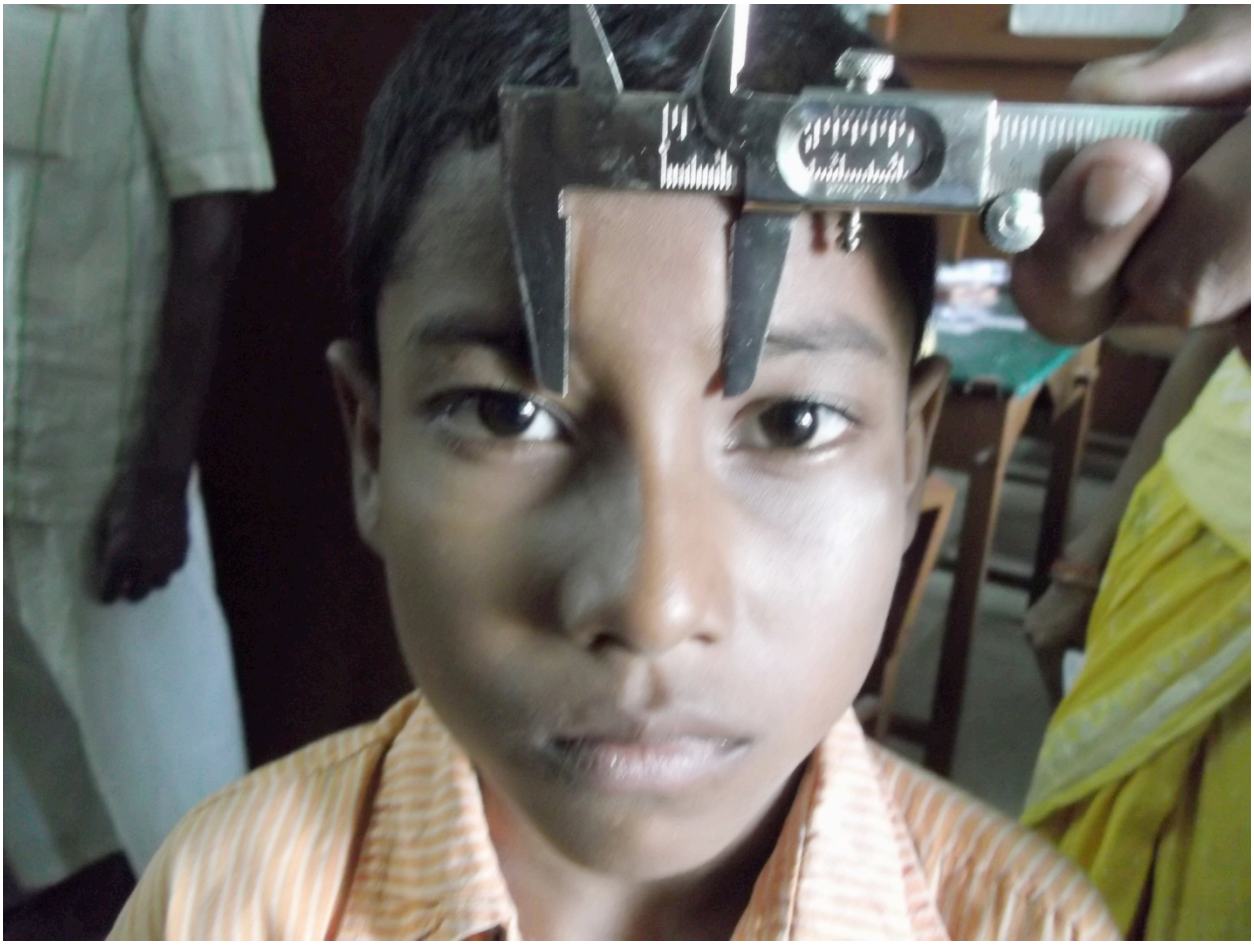

Fig.1 showing measurement of intercanthal distance by sliding caliper.

RESULTS

The study was conducted on 100 patients, 50 cases and 50 controls. Out of those 25 were male and 25 were female in each group. The children up to 16 years of age were included in this study. The minimum age was 6 months. Out of 50 cases with congenital acyanotic heart disease, 17(34%) male and 12(24%) female children had hypertelorism (Table1).

TABLE 1: children with congenital heart disease

Sl No.	Intercanthal Distance Cm. (M)	Diagnosis	Sl No.	Intercanthal Distance Cm. (F)	Diagnosis
1	3.9	VSD	1	4	VSD
2	2.5	VSD	2	3.8	VSD
3	4	ASD	3	3.4	VSD
4	2.2	VSD	4	3.6	VSD

5	3.7	VSD	5	2.5	VSD
6	2	PDA	6	2.4	ASD
7	3.8	VSD	7	2	ASD
8	4	VSD	8	2.6	VSD
9	3.8	ASD	9	3.8	VSD
10	2.7	VSD	10	3.8	VSD
11	2.5	VSD	11	3.9	ASD
12	4	ASD	12	4	VSD
13	3.8	VSD	13	3.4	ASD
14	3.8	ASD	14	3.8	VSD
15	4	PDA	15	3.8	VSD
16	3.4	VSD	16	4.2	ASD
17	3.7	VSD	17	2	VSD
18	3.2	VSD	18	2.2	VSD
19	4	VSD	19	2.4	VSD
20	2.8	ASD	20	3.2	PDA
21	4.2	ASD	21	3.8	ASD
22	3.8	VSD	22	2.7	VSD
23	3.8	VSD	23	2.5	VSD
24	4	VSD	24	3.8	VSD
25	4.2	PDA	25	3.7	PDA

VSD – VENTRICULAR SEPTAL DEFECT, ASD – ATRIAL SEPTAL DEFECT, PDA- PATENT DUCTUS ARTERIOSUS

The mean of the cases was 3.382±0.69 with a median of 3.8 and a mode of 3.8. Whereas, 8(16%) male, 5(10%) female childrens had hypertelorism without Congenital acyanotic heart disease (table 2). The mean of the controls was was 2.872±0.709, with a median of 2.6 and mode of 2.4.

TABLE 2:CHILDREN WITHOUT HEART DISEASE

SL NO.	Intercanthal distance cm.(M)	SL NO.	Intercanthal distance cm.(F)
1	2.2	1	2.6
2	3.7	2	2.1
3	2.5	3	2.1
4	4	4	2
5	2	5	3.6
6	3.8	6	2
7	3.4	7	2.5
8	3	8	3.8
9	3.6	9	2.2
10	2.1	10	2.6
11	3.2	11	3.6
12	3.7	12	2.5
13	2.4	13	3.7

14	2.2	14	4
15	3.8	15	2.4
16	2.2	16	2.7
17	3.9	17	2.4
18	2.1	18	3.5
19	2.6	19	3.7
20	2.4	20	2.6
21	2	21	2.4
22	3.8	22	3.8
23	4	23	2.2
24	2	24	3.2
25	2.4	25	2.4

Case	**Control**
P Value- 0.191	P Value- 0.197
T Statistics- 1.46	T Statistics- 1.17

The above statistical analysis shows, p Value of cases is 0.191 and control is 0.197, both are less than 0.05. So, it can be said that values of intercanthal distance of cases are not statistically significant.

DISCUSSION

Neural crest cells are essential for formation of the craniofacial region. That also contributes to the formation of conotruncal endocardial cushions, which septate the outflow tract into pulmonary and aortic channels. So, this might be reason for infants to have craniofacial abnormalities with cardiac defects, Sadler TW (2004). Nair et al, (2012) reported one case of congenital acyanotic heart disease with Ventricular septal defect (VSD), Atrial septal defect (ASD), Patent ductus arteriosus (PDA) with pulmonary hypertension and hypertelorism, low set dysmorphic ears and other facial dysmorphism.

Ventricular septal defect (VSD) was the common congenital heart defect found in children with hypertelorism in my study. This combination of VSD as CHD is found to be most common as reported by Khalil et al., (1994) and Kapoor et al., (2008).

Noonan's syndrome is one of the multiple congenital anomalies which is related with hypertelorism, cardiac defect and many other anomalies as per study of Bertola et al., (1999). This study did show the co-relation between heart disease and hypertelorism. However it is evident that hypertelorism may be present without heart defect, but majority cases had a correlaion.

Our study revealed no statistical significance between cases and controls that may be because of less number of samples. My aim was to show if there is any co-relation between congenital acyanotic heart disease and hypertelorism as we get large number of patients with heart disease with different

ocular abnormalities and there is less number of studies in India regarding this association. But still we suggest that any child whose eyes look apart on clinical examination should be carefully investigated for coexistence of unrecognized congenital cardiac anomalies.

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