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Paediatrics Section

Prevalence of Arrhythmic Events in Paediatric Patients with Congenital Heart Disease-A Retrospective Study

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ABSTRACT

Introduction: Various types of arrhythmias occur in patients with Congenital Heart Disease (CHD) and are the leading cause of morbidity and mortality. Knowledge regarding arrhythmias in CHD is important for safe and effective management of arrhythmia and in reduction of sudden cardiac deaths.

Aim: To assess the profile of various arrhythmic events in children with CHD.

Materials and Methods: A retrospective study was conducted from January 2018 to December 2020. Data were collected from the Department of Paediatrics of a tertiary care hospital in Durgapur, West Bengal, India. A total of 232 children of the age group 0-18 years, diagnosed with CHD were included in this study. Children who underwent cardiac intervention were excluded from the study. Age, sex, age at diagnosis, nature of CHD, clinical findings, Electrocardiogram (ECG) findings were recorded. Data analysis was performed using Fisher's-Exact test on Statistical Package for the Social Sciences (SPSS), version 27.0. A p-value <0.05 was considered as statistically significant.

Results: A 20.3% children of 0-18 years of age with CHD had conduction abnormalities. Statistically significant association was found between atrial septal defect with Atrial Fibrillation (AF) (p-value <0.001) and atrial flutter (p-value=0.008), Ventricular Septal Defect (VSD) with Premature Ventricular Contractions (PVC) (p-value=0.0001), Atrioventricular (AV) canal defect with first degree AV block with (p-value=0.0004), Tetralogy Of Fallot (TOF) with Ventricular Tachycardia (VT) (p-value=0.021), L-Transposition of Great Vessels (L-TGA) with complete AV block (p-value=0.012), Ebstein's anomaly with Supraventricular Tachycardia (SVT) (p-value <0.001).

Conclusion: Specific conduction abnormalities are significantly associated with specific CHD. These results demand attention for effective management of arrhythmia and reduction of sudden cardiac death in children with CHD.

Keywords: Conduction abnormalities, Congenital cardiac defect, Sudden cardiac death

INTRODUCTION

The incidence of haemodynamically significant CHD is 8 per 1000 live births [1-4]. Nearly 1,80,000 children per year are born with CHD in India [5,6]. Incidence of paediatric arrhythmias is approximately 55.1 per 100,000 patients evaluated in paediatric emergency departments [7]. Sinus tachycardia is by far the most commonly reported arrhythmia, followed by SVT which represents about 13%, and bradycardia accounting for about 6% of all cases [8].

Arrhythmias are seen in children with structurally normal hearts as well as those associated with CHD and cardiomyopathies. Cardiac arrhythmia is a common cause of acute deterioration, emergency hospital admission and sudden cardiac death [9]. The incidence generally increases with age. There are multifactorial predisposing features for development of arrhythmias in CHD that may include congenitally malformed or displaced conduction systems, altered haemodynamics and mechanical or hypoxic stress [10,11].

The care of the patient with CHD and arrhythmias may involve pharmacological therapy, catheter ablation, implantable cardiac devices, and surgical interventions [11]. In developing countries like India, majority of CHD remain undiagnosed and uncorrected due to lack of accessible heathcare facilities. Prompt diagnosis and management of arrhythmias comprises of a significant role in management of congenital heart defect [12]. Electrocardiography should be included in routine care of children with CHD.

In most of the previous studies regarding arrhythmic events in CHD, study participants were selected from post cardiac intervention patients in Intensive Care Unit (ICU) set-up. There is higher chance of detection of arrythmia in post cardiac intervention patients because most of the patients stay in ICU under cardiac monitoring in the immediate postoperative period and are under regular follow-up. Moreover, multiple confounding factors like surgery technique, postoperative medications and electrolyte imbalance may affect the results significantly [10,11,12].

On the other hand, in children with uncorrected CHD arrythmias often remain undiagnosed as ECG is not advised routinely by treating physician. Undetected arrythmias in uncorrected CHD may be fatal due to risk of sudden cardiac death. In India, as there is huge burden on uncorrected CHD [5], prevalence and profile of arrythmia in uncorrected CHD and risk of sudden cardiac death should be extensively studied. In the present study, patients with a history of cardiac intervention were excluded because the present study was conducted with the aim to emphasise upon the true prevalence and profile of arrhythmia in uncorrected CHD for early detection and effective management of arrhythmia and reduction of sudden cardiac death.

MATERIALS AND METHODS

A retrospective study was conducted over a period of three and half years in the Department of Paediatrics of IQ City medical college and hospital, Durgapur, West Bengal, India. Data were collected from January 2018 to December 2020 and data were analysed from January 2021 to June 2021. Ethical clearance was obtained from Institutional Ethical Committee (IEC) (Ethical committee approval number IQMC/IEC/LTR/17/02/29). Informed consent was obtained from all the parents of children included in the study.

Inclusion criteria: Children of 0-18 years of age diagnosed with CHD by 2D echocardiography and colour doppler examination were included in the present study.

Exclusion criteria: Patient with previous history of cardiac intervention were excluded from the present study.

Sample size calculation: A total of 232 children, who presented to the department within the study period were enrolled in the present study.

Study Procedure

Data were collected from the data record of outpatient and inpatient Department of Paediatrics of IQ City Medical College and Hospital, Durgapur, West Bengal, India. Age, sex, age at diagnosis, nature of CHD (acyanotic or cyanotic, diagnosis), clinical examination findings (presence of cyanosis, presence of murmur), ECG findings (any conduction anomaly) were recorded.

All the children were broadly categorised into two groups- cyanotic heart disease and acyanotic heart disease. Among 232 children with CHD, 66 children had cyanotic heart disease and 166 children had acyanotic heart disease. Congenital cyanotic heart disease is a heterogeneous group of abnormalities of cardiac development that result in deoxygenated blood being pumped to the body without first passing through the lungs. Acyanotic heart defects are congenital cardiac malformations that affect the atrial or ventricular walls, heart valves, or large blood vessels pathophysiologically characterised by a left-to-right shunt [12].

STATISTICAL ANALYSIS

Data was collected in a semi-structured data tool and finally entered into and analysed using SPSS 27.0. Descriptive statistics were generated as frequencies and distributions. Associations were derived between the independent and the outcome variables (arrhythmias). Fisher's-Exact test was applied for statistical analysis of the data. A p-value <0.05 was considered as statistically significant.

RESULTS

The study population consisted of 232 children of 0-18 years of age diagnosed with CHD (mean age 6±4.7 years) of which 151 (65.1%) of the study population were males, while 81 (34.9%) were females. Among 232 children, 66 (28.4%) of the children had cyanotic heart disease and 166 (71.6%) had acyanotic heart disease. [Table/Fig-1] shows detailed distribution of CHD in the study population. About 47 (20.3%) children of 0-18 years of age with congenital heart disease had conduction abnormality. [Table/Fig-2] shows number of different types of conduction abnormality present in the present study population. Among different conduction abnormalities AF was most common in the present study population (8.6%).

[Table/Fig-3] shows distribution of different types of conduction abnormalities in different CHD. Statistically significant association was found between Atrial Septal Defect (ASD) with AF (p-value=0.0012) and Atrial Flutter (p-value=0.0087), VSD with PVC (p-value=0.0001), AV canal defect with first degree AV block with (p-value=0.0004), TOF with VT (p-value=0.0218), L-TGA with complete AV block (p-value=0.0129), Ebstein's anomaly with SVT (p-value=0.0011).

Congenital acyanotic heart defect	N (%)	Congenital cyanotic heart defect	N (%)
ASD	91 (39.2)	TOF	36 (15.5)
VSD	42 (18.1)	DTGA	5 (2.1)
PDA	13 (5.6)	LTGA	3 (1.3)
VSD+PDA	9 (3.9)	TAPVC	3 (1.3)
Congenital AS	4 (1.7)	TA	2 (0.9)
COA	2 (0.9)	Single ventricle with PS	3 (1.3)
AV Canal defect	5 (2.2)	Ebsteins anomaly	5 (2.1)
		DORV with PS	9 (3.9)
Total	166 (71.6%)		66 (28.4)

[Table/Fig-1]: Distribution of congenital heart disease in the study population.
ASD: Atrial septal defect; VSD: Ventricular septal defect; PDA: Patent ductus arteriosus; AS: Aortic stenosis; COA: Coarctation of aorta; AV: Atrio ventricular; TOF: Tetralogy of fallot; TGA: Transposition of great arteries; TAPVC: Total anomalous pulmonary venous connection; TA: Tricuspid atresia; PS: Pulmonary stenosis; DORV: Double outlet right ventricle

Type of arrhythmia	N (%)
Atrial fibrillation	20 (8.6%)
Atrial flutter	5 (2.1%)
Premature Ventricular Contraction (PVC)	8 (3.5%)
Ventricular Tachycardia (VT)	8 (3.5%)
First degree AV block	2 (0.9%)
Complete AV block	1 (0.4%)
Supraventricular Tachycardia (SVT)	3 (1.3%)
Total	47 (20.3%)

[Table/Fig-2]: Types of conduction abnormality in the present study population.

Parameters			
Distribution of Atrial Fibrillation (AF) in ASD	**AF detected**	**AF not detected**	**p-value**
ASD	15	76	0.0012
Other CHD	5	136	
Distribution of atrial flutter in ASD	**Atrial flutter detected**	**Atrial flutter not detected**	
ASD	5	86	0.0087
Other CHD	0	141	
Distribution of PVC in VSD	**PVC detected**	**PVC not detected**	
VSD	7	35	0.0001
Other CHD	1	189	
Distribution of VT in VSD	**VT detected**	**VT not detected**	
VSD	3	39	0.1596
Other CHD	5	185	
Distribution of atrial fibrillation in PDA	**AF detected**	**AF not detected**	
PDA	1	12	1.000
Other CHD	19	200	
Distribution of atrial fibrillation in VSD+PDA	**AF detected**	**AF not detected**	
VSD+ PDA	2	7	0.1762
Other CHD	18	205	
Distribution of PVC in VSD+PDA	**PVC detected**	**PVC not detected**	
VSD+PDA	1	8	0.2749
Other CHD	7	216	
Distribution of VT in congenital AS	**VT detected**	**VT not detected**	
Congenital AS	1	3	0.1318
Other CHD	7	221	
Distribution of first degree AV block in AV canal defect	**First degree AV block detected**	**First degree AV block not detected**	
AV Canal defect	2	3	0.0004
Other CHD	0	227	
Distribution of VT in TOF	**VT detected**	**VT not detected**	
TOF	4	32	0.0218
Other CHD	4	192	
Distribution of atrial fibrillation in TOF	**AF detected**	**AF not detected**	
TOF	2	34	0.7469
Other CHD	18	178	
Distribution of complete heart block in L-TGA	**Complete heart block detected**	**Complete heart block not detected**	
L-TGA	1	2	0.0129
Other CHD	0	229	
Distribution of SVT in Ebsteins anomaly	**SVT detected**	**SVT not detected**	
Ebsteins anomaly	2	3	0.0011
Other CHD	1	226	

Distribution of SVT in DORV with PS	SVT detected	SVT not detected	
DORV with PS	1	8	0.1124
Other CHD	2	221	

[Table/Fig-3]: Distribution of specific conduction abnormality in specific CHD. Fisher exact test; significant p-value <0.05 is statistically significant

DISCUSSION

The number of individuals with CHD is continuously on the rise. Increased number of patients are reaching adulthood. A significant portion of this population will suffer from arrhythmias due to the underlying CHD itself or as a sequalae of interventional or surgical treatment [12,13]. After thorough search of existing literature, it is clear that, even though majority of children with arrhythmias have structurally normal heart, they are frequently found in children with underlying heart disease [14].

Prevalence of arrhythmias among children with CHD in other studies ranges from 6-27% [15,16]. Prevalence was slightly lower in present study when compared to the study by Batte A et al., [16], probably because patients with cardiac intervention were excluded from the study population. However, the prevalence in the present study was lower than the prevalence of arrhythmias detected by Holter ECGs, which showed a prevalence of upto 41.2% in infants and children with uncorrected heart defects [17]. Holter ECGs are not widely available in developing countries and thus, standard 12 lead ECG was used in present study.

Prevalence of atrial fibrillation was 8.6% and atrial flutter was 2.1% in present study. Similar studies among children with CHD detected comparable prevalence of these arrhythmias [27]. AF and atrial flutter significantly associated with ASD due to dilated atria with volume overload. AF has also been reported in children with TOF at a prevalence of 6.7% [30,31]. In the present study, prevalence of AF in TOF was 5.5% but, no statistically significant association found between TOF and AF. Early detection and management of atrial arrythmias are significant to prevent the thromboembolic events.

PVC was detected in 8 patients (3.5%) in the present study. PVCs have been described in children with VSD and Complete AV canal defect [32]. Though occasional PVC is benign in nature, complex PVC (frequent PVC, multiform PVC, ventricular couplets) should always be treated with β blocker and other antiarrhythmic drugs.

In present study, 1st degree AV block was detected in 2 patients (0.9%) and was significantly associated with complete AV canal defect. First degree heart block is present in the majority and prolongation of the QRS complex in over half of patients with complete AV canal defect [29]. There is also an increased risk of complete heart block due to displacement of the atrioventricular node [33]. In the present study, it was found that overall prevalence of conduction abnormality in patient with CHD was 20.3% which is comparable with the other studies [Table/Fig-4] [16,17,34,35].

Study	Place and publication year	Sample size	Prevalance of conduction abnormalities in CHD	Reason for difference of prevalance	Most common arrhythmias detected
Batte A et al., [16]	Uganda 2016	194	27%	Slightly higher prevalence due to inclusion of cases with cardiac intervention	First degree heart block
Grosse-Wortmann L et al., [17]	Canada 2010	494	41.2%	Holter ECG used for detection of post cardiac operative arrythmias	Junctional ectopic tachycardia
Hoffman TM et al., [34]	Philadelphia 2002	629	29%	Slightly higher prevalence due to inclusion of postoperative cardiac cases	Non sustained VT
Öztürk E et al., [35]	Istanbul 2021	670	12.4%	Lower prevalence because of short postoperative follow-up period	Junctional ectopic tachycardia
Present study, Banerjee A and Mitra M	West Bengal, India 2022	232	20.3%	Lower prevalence because of exclusion of cardiac interventions	Atrial fibrillation

[Table/Fig-4]: Comparison with other studies [16,17,34,35].

Among all the arrhythmias detected in the present study population, two most life-threatening arrhythmias were complete AV block and VT. Complete AV block was detected in 1 child (0.4%) with L-TGA. This arrhythmia in association with structural heart disease has a case fatality rate of 29% in infancy and 10% in childhood [15]. Complete AV dissociation has been described in literature in children with L-TGA (up to 22% cases), complete AV canal defect and TOF [16,18-21].

In present study, VT another life-threatening arrhythmia was detected in 8 patients (3.5%). Statistically significant relationship was found between TOF and VT in the present study. VT occurs in approximately 10% of patients with TOF [22] and a major cause of sudden cardiac death in uncorrected TOF [20]. Implantable Cardioverter-Defibrillators (ICD) are increasingly utilised in the primary and secondary prevention of sudden death in patients with TOF [23].

The SVT was detected in 3 patients (1.3%) in the present study. Statistically significant relationship was found between SVT with Ebstein's anomaly. Right-sided accessory pathways, classically associated with Ebstein's anomaly, have been reported in 25% cases. They may be multiple, which is the major predisposing factor for the development of SVT [24-27]. For the patients with recurrent SVT related to an AV re-entrant mechanism, radiofrequency catheter ablation technique has been successful. Success rate was 95% in those with isolated right sided accessory pathways and 76% in those with multiple pathyways [28,29].

As reflected by the results of the present study, arrhythmias occur in approximately one in five children with CHD. In a developing country like India, these arrythmias often remain undetected due to limited knowledge amongst paediatricians about the relatively high incidence of arrythmias and lack of access to medical facilities. It is suggested that Electrocardiography should be included in routine care of all children diagnosed with CHD. Early detection and effective management of arrythmias should be the mainstream strategy for reduction of sudden cardiac death in uncorrected CHD.

Limitation(s)

The present study was carried out using a standard 12 lead ECG and not a Holter ECG thus the prevalence of arrhythmias reported may be lower than the actual prevalence. The present study was unable to assess other factors which could predispose these children to arrhythmias such as genetic factors, infections, electrolyte imbalance, effect of cardiac medication like digoxin.

CONCLUSION(S)

According to the present study, arrhythmias occur in approximately one in five children with CHD and specific conduction abnormalities are significantly associated with specific CHD. Electrocardiography should be included in routine care of children with CHD. Further large scale studies are required to find significant association

between specific conduction abnormalities with specific CHD in uncorrected CHD.

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