

# Arrhythmia Profile in Children Presenting at a Tertiary Care Centre in India - From Fetal Life to Fifteen Years of Age

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## Abstract

Many of the arrhythmias tend to be asymptomatic and detected incidentally, while some others may be picked up too late after a catastrophic presentation. The goal of this study was to document the arrhythmia profile in children presenting to our tertiary care hospital in India. We also reviewed the early and late outcomes of arrhythmias in these patients (including in isolated and post cardiac surgical patients). Amongst 100 patients with arrhythmias, 35% were detected incidentally for presentations like breathlessness (29%) and refusal to feed. A majority of patients (70%) did not have any underlying structural defect and had no cardiomegaly on chest X-Ray. Of those with structural defects, 90% were congenital with the majority being acyanotic (58%) and would have gone clinically undetected. 19 children (20%) presented with heart failure. Tachyarrhythmias (61%) were more common, and all the immediate deaths were from this group. Bradyarrhythmias were mostly complete heart blocks. Almost half the patients required emergency antiarrhythmics. Holter study was done to check for antiarrhythmic efficacy and to decide regarding pacing. In 48% cases, clinical management was changed based on the Holter report. Almost half the patients required emergency antiarrhythmics (adenosine, isoprenaline, amiodarone). The long-term outcomes were favorable with 50% of the patients not requiring long term antiarrhythmics and most remained asymptomatic. The need for interventional therapy like pacemaker insertion and radiofrequency ablation was less than 2%. Arrhythmias in children have varied symptoms and pediatricians need to be aware about this. With optimal management, long term outcomes for these children are usually favorable.

**Keywords:** Pediatric arrhythmia, arrhythmia in children, pediatric Holter, tachyarrhythmias in children, bradycardia in children.

## Introduction

A cardiac arrhythmia simply defined is a variation from the normal heart rate and/or rhythm that is not physiologically justified (1). Although just 5% of the emergency hospital admissions in the pediatric population is attributed to symptomatic arrhythmias (2), they represent upto 15-20% of sudden cardiac death in children (3). These remain unexplained after necropsy and leave no trace after death. Some causes include polymorphic ventricular tachycardia in congenital long QT syndrome, atrial fibrillation in Wolff-Parkinson-White syndrome and congenital complete atrioventricular block (4). Of note other channelopathies like CPVT, Brugada syndrome, short QT syndrome or syndromes related to myopathies and SCN4A channel dysfunctions are associated with sudden cardiac death in children (5-8). The symptoms of arrhythmias in children depend on the underlying rhythm disorder, its effect on haemodynamics and the age of the patient. Majority of patients will be asymptomatic, and hence warrant a high index of suspicion on the part of the clinician (9). Typically, neonates and infants with arrhythmias tend to present with congestive cardiac failure secondary to tachyarrhythmias (particularly following incessant supraventricular tachycardias like Permanent junctional reciprocating tachycardia (PJRT) and Ventricular tachyarrhythmias (VT)). Tachyarrhythmias also occur as complications after cardiac surgery and represent a major source of morbidity and mortality.

Arrhythmias that may be well tolerated in a younger patient can be a major cause of morbidity and mortality after cardiac surgery for congenital heart disease (10,11).

Although the diagnostic yield is low in the pediatric age group, Holter monitoring enables cumulative evaluation of heart rhythm and rhythm variability, which is important in diagnosing silent arrhythmias in high risk groups (abnormal heart in cardiomyopathies or postoperative heart) (12,13). Early detection and management play a vital key to survival (14).

## Methods

The study was a 5-year single centre retro-prospective observational study conducted in a tertiary centre multispeciality referral hospital for children. This centre had a specialized pediatric cardiology unit catering to the outpatient department, general ward, pediatric and neonatal intensive care unit (ICU), medical cardiac ICU and post-operative cardiac ICU. The study was a 5-year single centre observational study. All fetuses and children aged less than 15 years diagnosed and referred with arrhythmias and treated during the study period were selected. The study was approved by the Institutional Ethics Committee.

### Primary Objectives

- To study the demographic variables, etio-pathogenesis and clinical presentation of children diagnosed with arrhythmias.
- To study the electrocardiographic patterns and treatment modalities required.
- To correlate clinical variables, electrocardiographic variables and echocardiograms with outcome.
- To study the effect of Holter monitoring on management of children with arrhythmias.

### Study duration and sample size

The selection of 100 children for the study was based on referral statistics and records in the cardiology division with arrhythmias which is approximately 20 per year.

Recruitment for the retrospective cohort was over a period of 4 years from August 2014 to July 2018 and the nature and course of their disease was studied for a maximum of 5 years. Recruitment of prospective cohort from the study was from August 2018 to July 2019 (12 months) and follow-up was for a minimum of 3 months.

Of these 100, 9 were excluded as 4 were lost to follow-up (insufficient follow-up data) and 5 had incomplete records in the retrospective cohort.

Finally 91 children with diagnosis of arrhythmia were included in the analysis..

### INCLUSION CRITERIA

- All children from fetal life to 15 years of age, newly diagnosed with arrhythmia or a known case coming for follow up during the study period.
- Children admitted in the ICU settings including post cardiac surgery children, who suffered from cardiac arrhythmias were also included.

### EXCLUSION CRITERIA

- Those evaluated and detected to have sinus tachycardia or sinus bradycardia at presentation.

### Outcome Parameters

This is an observational, analytical and retrospective plus prospective study. Basic demographic data consisted of the child's age, sex and age at presentation. Symptomatology included chest pain, palpitations, syncope, refusal to feed, increased respiratory activity/breathlessness, lethargy, asymptomatic/ incidental detection and other complaints if any. Concurrent medical/surgical illness along with duration of symptomatology (if present) were noted.

Past history of similar episodes, previous admissions for similar complaints and past history of use of anti-arrhythmics was noted. In children with underlying heart diseases, details of the primary defect (both congenital and acquired) were entered along with details of cardiac surgery and duration between surgery and arrhythmia in operated cases. Perioperative arrhythmias as well as those sustained during cardiac catheterization were also included and recorded.

Previous management history included emergency anti-arrhythmic received and its response and any need for cardioversion and inotropic support.

Family history of arrhythmias or unexplained sudden death if present was noted. Family history also included presence of connective tissue disorders in parents or siblings and positive antibodies in mothers of children with congenital heart block.

Examination findings noted were heart rate, respiratory rate, blood pressure according to centiles, peripheral pulses, capillary refill time and interpreted as normal or abnormal, pulse oximetry, presence/absence of cardiac murmur, crepitations and hepatomegaly at initial presentation with arrhythmia.

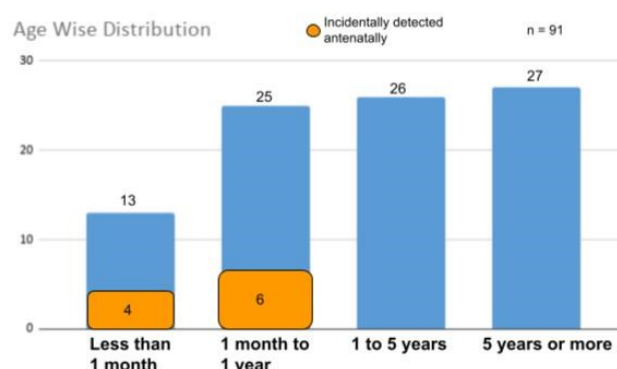
Investigations included serum electrolytes, chest X-ray, standard 12-lead Electrocardiogram (ECG), 2D Echocardiography/colour Doppler and Holter. Indications for Holter monitoring included for diagnosis, monitoring and assessment of antiarrhythmic efficacy. 'Philips Zymed 1810 Holter System' was used. The machine was attached to the patient for 24 hours and patients were allowed to do all daily activities. Each Holter was reported by an independent certified pediatric cardiologist with training in electrophysiology.

Management of each child was as per standard treatment protocols. Emergency management was done as per Pediatric Advanced Life Support (PALS) guidelines which included both pharmacological treatment and cardioversion. Details of the emergency anti-arrhythmic received and response along with need for cardioversion and inotropic support were recorded. Immediate outcome was assessed as upto discharge or upto 3 months in the cardiology clinic. Long term outcomes were considered as between 3 months upto a maximum of 5 years for the retrospective cohort. Outcomes were categorized as alive or dead on immediate and long term follow up. Long term antiarrhythmic drugs received, their dose, frequency and duration, whether the child underwent surgical /catheter intervention including radiofrequency ablation and pacemaker insertion, were recorded. Long term outcome was assessed by following up each patient for a minimum of three months to maximum of 5 years. Outcome was classified on the basis of last follow-up.

## Results

### History and Symptomatology

Neonatal cases made a minority (13/91), while most of the arrhythmias presented after the 1st month of life (51% between 1 month to 5 yrs, while 29% being over 5 years). 10 cases were incidentally detected antenatally, of which 4 presented in the neonatal period with symptoms (Fig. 1) requiring an NICU admission, while the rest 6 presented after the 1st month. All the antenatally detected cases presented before completing infancy with one presenting in the 11th month of life. There was no particular sex predilection, with males (50.5%) and females (49.5%) being almost equal.



**Fig. 1:** Age Wise Distribution.

Most of the patients were asymptomatic (35%) (Table 1). These children presented with an intercurrent illness and were incidentally detected. There were no other cardiac symptoms. The most common presenting symptom was breathlessness (28.6%).

17 patients had a history of cardiac surgery. Cardiomegaly was found in 21% children on Chest X-Rays. Interestingly, 11% had significant family history in the form of unexplained sibling deaths or autoimmune disorder in mothers of children with congenital heart block. Of the mothers of children with congenital complete heart block, Anti Ro/Anti La antibodies were positive in 63.6%, with five (71%) of these children being detected antenatally.

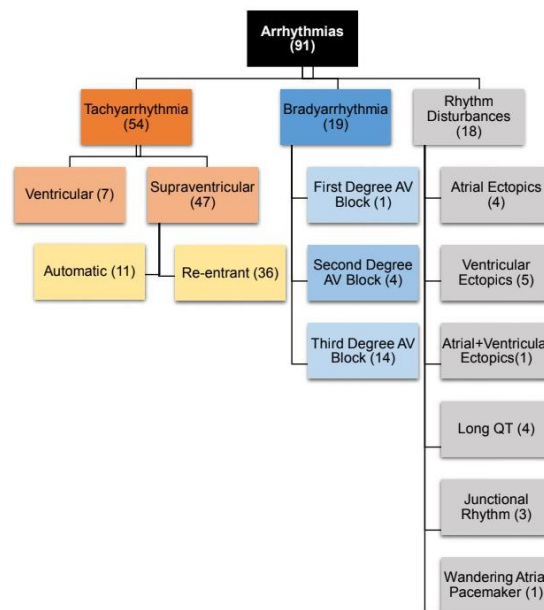
Structural heart changes were present in 30% of the children, with 90% being congenital. 52% of the cases were acyanotic heart defects with VSD accounting for the majority (57%).

**Table 1:** Clinical Symptoms.

Clinical Symptoms	n=91	Percentage
<b>Asymptomatic (Incidental Detection)</b>	32	35.1
<b>Palpitations</b>	10	11.0
<b>Chest Pain</b>	10	11.0
<b>Breathlessness</b>	26	28.6
<b>Lethargy</b>	4	4.4
<b>Refusal to Feed</b>	5	5.5
<b>Syncope</b>	4	4.4

**Arrhythmia morphology**

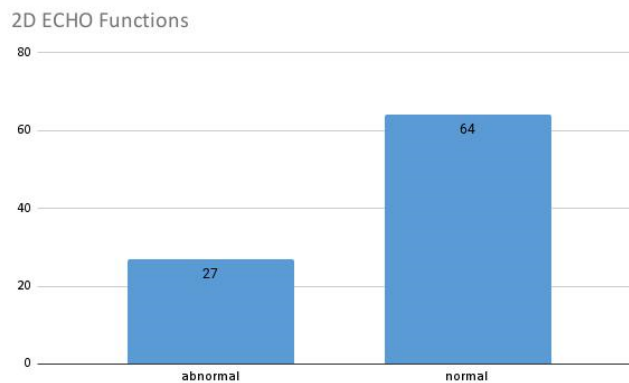
Of the 10 arrhythmias detected antenatally, 8 were bradyarrhythmias (6 with Complete heart block [CHB] and 2 with Mobitz type 2 heart block) and 2 were Supraventricular tachycardias (SVT). 61% of the arrhythmias were tachyarrhythmias (Fig. 2), and 75.8% were hemodynamically stable. 50% of the tachyarrhythmias were supraventricular (76% of these being re-entrant). 73% of the bradyarrhythmias constituted CHB.



**Fig. 2:** Arrhythmia morphology in study subjects.

18% had features of left ventricular hypertrophy on ECG. Of these, 5 had SVT, 3 had CHB, 2 had atrial ectopics and 1 patient had a long QTc interval (Fig. 3).

47% of these abnormal ECGs had significant 2-D Echocardiography and colour Doppler findings. Ejection Fraction was impaired in 29.7% of cases (Fig. 3) indicating cardiomyopathy secondary to arrhythmia (Ejection fraction <55%).



**Fig 3:** Cardiac Function on 2D-Echocardiogram/Color Doppler.

### Holter Findings

Holter study was done in 26/91 (30%) children. Holter analysis helped to change management for antiarrhythmic efficacy, decision to pace or for diagnosis of the arrhythmia.

Of the arrhythmias detected using Holter, 9 were diagnosed as SVT, 10 were AV blocks, 3 were ectopics, 2 were junctional rhythm, and one case each had a long QTc interval and wandering atrial pacemaker (Table 2).

Management was changed in 42% of cases (Table 3).

**Table 2:** Arrhythmias on Holter.

1) SUPRAVENTRICULAR TACHYCARDIA	9
a) PJRT	1
b) Ectopic atrial tachycardia	1
c) Atrial fibrillation	1
d) Others	6
2) AV BLOCK	10
a) Complete heart block	8
b) 2 <sup>nd</sup> degree AV block	1
c) 1 <sup>st</sup> degree AV block	1
3) ATRIAL ECTOPICS	3
4) JUNCTIONAL RHYTHM	2
5) LONG QT	1
6) WANDERING ATRIAL PACEMAKER	1

**Table 3:** Management changes based on Holter.

WHETHER HOLTER CHANGED THE MANAGEMENT	Frequency	Percent
NO	15	57.7
YES	11	42.3
<ul style="list-style-type: none"> <li>• Amiodarone stopped due to QT prolongation</li> <li>• Pacemaker inserted</li> <li>• Pacemaker not inserted</li> <li>• RFA done</li> <li>• Beta blocker added for sinus tachycardia +palpitation</li> <li>• Dose of flecainide increased</li> </ul>	<p style="text-align: center;">1</p> <p style="text-align: center;">2</p> <p style="text-align: center;">5</p> <p style="text-align: center;">1</p> <p style="text-align: center;">1</p> <p style="text-align: center;">1</p>	<p style="text-align: center;">3.8</p> <p style="text-align: center;">7.6</p> <p style="text-align: center;">19.1</p> <p style="text-align: center;">3.8</p> <p style="text-align: center;">3.8</p> <p style="text-align: center;">3.8</p>
Total	26	100.0

### Management and Outcome

Of 91 cases, 51(56%) required emergency antiarrhythmics. Of these 55% reverted with the emergency drug, 45% did not. Of these 51, 36% did not require further antiarrhythmics, while 10% required a second and another 9 percent needed more than 2 antiarrhythmics. 14% cases each of SVT and VT had to be cardioverted. Almost 50% did not require long term antiarrhythmics. Most (91%) didn't require surgical or transcatheter interventions. Of those that did, pacemakers were inserted in 3%, and RFA was done in 2%. Only 1% required temporary pacing. 40% had a hospital stay of between 3-7 days.

All the 8 deaths in the immediate period were tachyarrhythmias, of which 4 were ventricular and 4 were SVT. Out of these 8, 5 had a clinical diagnosis of myocarditis.

Over long term follow-up, 6 were symptomatic and required an escalation of treatment. There were 4 further deaths; all of them had complete heart block. Of these 2 had pacemakers inserted, while a 3rd was advised. Of the 4 deaths, 2 had complex structural heart defects and developed postoperative heart block. The other two had AntiRo/La antibodies positive in mothers.

### Discussion

Pediatric arrhythmology which studies arrhythmias (dysrhythmias) in children, is still an area ripe for further exploration in the field of pediatric cardiology. Development of this niche of medicine would primarily lead to a reduction in childhood mortality. The incidence of significant arrhythmias (pre-excitation, sinus pauses, AV blocks) in children is 2%, and if all insignificant rhythm disorders are included then it increases up to 25%(15).

Arrhythmias are life threatening in children and may present as sudden cardiac death. Analysis of all children presenting or detected with arrhythmias is hence essential for optimal outcome.

Our study showed that 35% of arrhythmias were asymptomatic and detected incidentally; either on a routine screen, a fetal echo, or while being evaluated for another condition. This is coherent with the work by Nagashima et al, where they found between 15-50% of healthy, asymptomatic children to have arrhythmias on 24-hour ambulatory ECG monitoring (16). A similar study by von Bernuth et al, using ambulatory ECG monitoring found between 13%-35% of healthy children to have supraventricular extrasystoles (17). Some arrhythmias causing hemodynamic compromise to require early interventions to revert and could lead to mortality if not controlled. This would confirm the suspicion that arrhythmias remain a silent killer contributing to Sudden Cardiac Deaths (3).

Our study also shows that the majority of these children remain asymptomatic after primary treatment and can lead healthy lives even though more than half of them require an emergency drug intervention.

Any suspicion of an arrhythmia warrants a complete evaluation to rule out life threatening arrhythmias, with at least an ECG and a 2D-Echocardiogram to rule out underlying structural heart defects. Arrhythmias can be detected in fetal, neonatal and adolescent life, and with timely treatment have good outcomes.



Antenatally detected arrhythmias allowed prompt and early referral to our tertiary care center, preventing further deterioration. This study has shown that a high percentage of ECG detected arrhythmias have significant echocardiography and Doppler findings, similar to a study by Mene-Afejuku et al (12). Thus, also warranting that all detected arrhythmias be advised for an echocardiography and colour Doppler study. While the majority of the structural cardiac defects would present with murmurs (18), most of the arrhythmias have no giveaways. We found that the majority did not have any cardiomegaly on X-rays or any clinically obvious structural cardiac defects. Of those that did have structural defects, most would be acyanotic congenital heart diseases, and miss clinical detection. These findings are also reflective of another study done in the pediatric age group by Premkumar et al (9). Arrhythmias were equally likely to affect male and female children. Breathlessness has been one of the most common presentations in symptomatic cases, with chest pain, palpitations and refusal to feed in infants summing the next majority. We hence believe the clinician be wary of these seemingly docile presentations in young children.

Continuous ECG Holter monitoring has become an irreplaceable tool in management of arrhythmias in children. Holter monitoring has been established as one of the most effective noninvasive clinical tools in the diagnosis, assessment and risk stratification of cardiac patients. However, studies in the pediatric age group are limited.

Tachyarrhythmias were the most common type of arrhythmia; most being Supraventricular reentrant, similar to what was observed by McCammond et al (19). They are mostly hemodynamically stable; however, they were also the major cause of immediate death in our cohort. These immediate deaths were also associated with cardiomyopathy (in 5 of the 8 children), which was poorly tolerated and would lead to early death. Fulminant myocarditis gives no time for nature to compensate in these young ones.

Bradyarrhythmias, on the other hand, lead to late cardiac death. Majority were Complete Heart Blocks (CHB). Most of the cases of congenital CHBs are secondary to autoimmune defects like Systemic Lupus Erythematosus (SLE) in mothers with positive AntiRo/Anti La antibodies (20). CHBs can contribute upto 15% mortality, which increases to 42.4% if associated with a structural heart defect (21).

Pediatric cardiology over the years has significantly advanced and is now on the verge of artificial intelligence breakthroughs. We believe every high volume cardiology center should have a Holter being prescribed for children presenting or detected with arrhythmias. Being a resource limited country, Holter monitoring isn't available in most of the centers in India. In our study, Holter was indicated for evaluating antiarrhythmic efficacy, to decide regarding pacemaker insertion and for diagnosis of arrhythmia. This 24-hour investigation was useful to detect AV blocks and SVTs, including one case of long QTc and resulted in change in management in a significant number of cases. It was most beneficial in children with complete heart blocks, with regards to decision regarding pacing. While the low heart rate on an ECG may indicate the need for pacing, the average heart rate on Holter gave a clearer view regarding underlying cardiac electrical activity. Holter monitoring positively contributed to the diagnosis and management of cases in our cohort similar to other studies (15,22,23).

Management wise, all patients were assessed hemodynamically and were managed as per PALS Guidelines (24). Children with tachyarrhythmias received adenosine, amiodarone or magnesium sulfate depending on ECG rhythm. Our study showed that although most responded to the emergency drug, a few required further doses for reversal of the rhythm. 56% of SVTs receiving adenosine reverted with the first dose. Of those that didn't, two were WPW syndrome, two were SVT with aberrancy, two had dyselectrolytemia, three were congenital heart defects, two dilated cardiomyopathy and one had myocarditis. We believe these factors could have led to their refractoriness to adenosine, although refractoriness has been observed in infants and young children (21).

As per our study, half of these children will not need long term antiarrhythmics, and 90% require no surgical intervention. Holter was indicated in 9 patients to determine requirement for pacing, of which a pacemaker was inserted in 2. This shows that the clinical outcome for these patients is quite favorable. With a high clinical suspicion, and prompt early treatment a significant amount of morbidity and mortality can be prevented.

## Conclusion

Our study has shown that in a tertiary referral center, a significant number of arrhythmias are asymptomatic, warranting a high degree of suspicion. Most of these arrhythmias would be hemodynamically stable on presentation and responsive to treatment, while if left untreated, they could lead to deaths due to tachycardiomyopathy. Fetal 2D-Echo would be indicated strongly in mothers with Anti Ro/La positivity to detect CHBs and Holter also plays a significant part, especially in determining need for pacemaker insertion.

## Strengths and Limitations

### Strengths

- 1) Follow up for 3 months to 5 years, allowing a study of immediate as well as long term outcome.
- 2) One of few studies from resource limited countries on childhood arrhythmias including Holter monitoring.

### Limitations

- 1) Referral bias- being a tertiary care referral center, the cohort may be skewed.
- 2) A larger cohort would be warranted, as a multicentric study, to track the natural history of arrhythmias over a longer follow up period.

### Conflict of Interest

The authors declare no conflict of interest.

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