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Papers must be submitted with the understanding that they have not been published elsewhere (except in the form of an abstract or as part of a published lecture, review or thesis) and are not currently under consideration by another journal published by international research journal or any published.

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Editorial

Paed. Heart J of Bangladesh (PHJB) 2021;6(1): 1

Ventricular septal defect (VSD) is one of the commonest congenital heart diseases (20-25%) in paediatric age group. It can occur in isolation or as part of more complex defects. Among different subtypes perimembranous and outlet VSD are associated with coronary cusp prolapse specially the right coronary cusp which can cause aortic regurgitation of various severity. A prospective cross-sectional study was conducted in Paediatric Cardiology Department, BSMMU to compare coronary cusp prolapse in perimembranous (PM) and outlet VSD.

Vitamin D deficiency causes oxidative stress, Immune dysfunction, secondary hyper-parathyroidism, hypocalcaemia. All these phenomenon lead to inflammation of cardiac myocyte along with thrombosis, vascular smooth muscle proliferation and vascular dysfunction. These pathological processes can be fatal in presence of congenital heart disease as patients of CHD are vulnerable to all form of cardiovascular malfunction. So a study was conducted in Paediatric Cardiology Department, BSMMU to measure Vit-D level and assess LV systolic function in children with CHD and compare that to healthy control.

The most common CHD has been ventricular septal defect, followed by atrial septal defect, patent ductus arteriosus, tetralogy of Fallot, single ventricle, atrium ventricular septal defect and double outlet right ventricle. There are very few authentic data regarding the pattern of CHD at national level in Bangladesh. A study was conducted in SCABU of Bangladesh Shishu Hospital & Institute to identify the pattern of congenital heart diseases among neonate.

These are some of the topics discussed in this issue of Paediatric Heart Journal of Bangladesh. Any comments or criticism will be highly appreciated.

Editor

Original Article

Coronary Cusp Prolapse in Perimembranous and Outlet Ventricular Septal Defect in A Tertiary Care Hospital in Bangladesh

Chaity Barua[1], Md. Tariqul Islam[2], Tahmina Karim[3], Diana Islam[4], Sunam Kumar Barua[5], Kashid Omar[6]

Abstract

Background: *Ventricular septal defect (VSD) is one of the commonest congenital heart diseases (20-25%) in paediatric age group. Among different subtypes perimembranous and outlet VSD are associated with coronary cusp prolapse specially the right coronary cusp which can cause aortic regurgitation of various severity.*

Objective*: To compare coronary cusp prolapse in perimembranous (PM) and outlet VSD*

Methods*: This prospective cross-sectional study was conducted in Paediatric Cardiology Department, BSMMU. The study included total 105 PMVSD and outlet VSD cases which were diagnosed by transthoracic echocardiography. Patients were selected according to inclusion and exclusion criteria. A total of 105 patients were studied prospectively.*

Results: *Among total 105 VSD cases- PMVSD was 80(76.2%) and outlet VSD was 25(23.8%). Prevalence of cusp prolapse was more in outlet VSD (6 out of 25 cases, 24%) than PMVSD (3 out of 80, 3.75%). Cusp prolapse occurred in older age and larger VSD shunt size in PMVSD compared to outlet variety.*

Conclusion*: Coronary cusp prolapse is more prevalent in Outlet VSD and it occures at lower age group and smaller VSD shunt size compared to PMVSD.*

Keywords: *Coronary cusp prolapse, perimembranous and outlet ventricular septal defect.*

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Introduction

Ventricular septal defect (VSD) is a developmental defect of the interventricular septum (IVS) where a communication exists between the cavities of the two ventricles. It can occur in isolation or as part of more complex defects.[1] Isolated Ventricular septal defect (VSD) is one of the most common congenital cardiac anomaly in children.Isolated VSD accounts for 20-25% of all congenital heart disease in children. The incidence of isolated VSD is about 0.3% in new born.[2] It is subdivided

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into many categories depending upon many demarcating factor. Among them one of the major subtypes are Perimembranous, muscular, outlet and inlet depending upon the anatomical position of the defect in interventricular septum.[3] VSD is associated with many complications. One distinct one is coronary cusp prolapse associated with aortic regurgitation. Combination of ventricular septal defect (VSD) and aortic regurgitation (AR) due to prolapse of right coronary or, less frequently, non-coronary cusp is known as Laubry-Pezzi syndrome. Perimembranous and outlet type VSD with coronary cusp prolapse, mainly right coronary cusp prolapses, and aortic regurgitation (AR) has been shown.[4] So this study was conducted to compare the cases of cusp prolapsed with PM and outlet VSD.

Materials and Methods

It was a prospective cross-sectional study conducted at the Department of Paediatric cardiology in Bangabandhu Sheikh Mujib Medical University (BSMMU) from August, 2020 to august 2021. All pediatric patients who underwent echocardiography in paediatric cardiology department, BSMMU having isolated perimembranous and outlet VSD aged between 6month to 18 year were included in this study. VSD associated with Complex congenital heart disease like TOF, TGA, DORV etc, patients who has aortic valve anomalies, such as bicuspid aortic valve, patients with an obvious history of infective endocarditis and patients who are not willing to take part in the study were excluded. Informed written consent was taken from the parents and ethical clearance was obtained from the Institutional Review Board (IRB) of BSMMU. Patient particulars were enlisted properly. Detailed physical examination was performed in all cases. Echocardiography was done by G&E Echo machine Model Vivid S70N. Patients were checked in supine and lateral positions. After confirming the primary diagnosis and types of VSD and cusp prolapse was identified. From these data coronary cusp prolapse in PM and outlet VSD was compared.

Results

Mean age for the study group was 35.32±50.67 month. Meanwhile mean weight and mean height was 10.68±7.24 kg and 84.1±27.3 cm respectively. Mean Body surface area was calculated as 0.48±0.24 m2. 52.3% study population came from urban area (Table-I).

Table-I

Demographic data of Study Population (N=105)

Variable	Mean±SD
Age (month)	35.32±50.67
Weight (kg)	10.68±7.24
Height (cm)	84.1±27.3
BSA (m2)	0.48±0.24
Urban area (%)	52.3%

Data was expressed as Mean±SD, BSA=Body surface area

Among total 105 patient 54 patients were male (51.5%) and 51 patients were female (48.5%). Male and female ratio was 1.05:1 (Fig.-1).

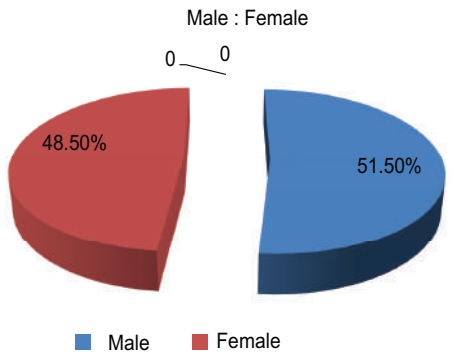

Fig-1: *Sex distribution in study population*

Majority patients presented with H/O recurrent RTI (90.5%) followed by feeding difficulty (39.4%) which was particularly evident in children belongs to lower age group. Dyspnoea was present in 26.66% children and finally cyanosis was evident in 16.19% children (Table-II).

Table-II

Distribution of clinical presentation in study population (N=105)

Clinical presentation	Percentage (n)
H/O Recurrent RTI	90.5 (95)
Feeding difficulty	39.4 (41)
Dyspnoea	26.66 (28)
Cyanosis	16.19 (17)

Data was expressed as Frequency and percentage, RTI=Respiratory tract infection

This table shows echocardiographic data obtained from Transthoracic Echocardiography of study population. Among 105 VSD cases PMVSD was 80 cases (76.2%) and Outlet VSD was 25 cases (23.8%). Mean VSD size was 6.45±3.76 mm. Mean PASP was 40.54±25.21 mmhg (Table-III).

Table-III

Echocardiographic data of study population (N=105)

VSD type	
PMVSD, n(%)	80(76.2)
Outlet VSD, n(%)	25(23.8)
Mean VSD size (mm) (Mean±SD)	6.45±3.76
PASP mmhg (Mean±SD)	40.54±25.21

Data was expressed as Mean±SD, frequency and percentage, PMVSD=Perimembranous VSD, PASP=Pulmonary artery systolic pressure

Mean age for outlet VSD is less (40.16±30.32 month) than PMVSD (128.67±48.89 month). Mean height and weight is also smaller than PMVSD in outlet variety. PMVSD required more larger VSD shunt (10.33±0.49mm) than outlet VSD (6.16±0.85mm) (Table-IV).

Table-IV

Comparison of Demographic and echocardiographic data between Cusp prolapse of Outlet VSD (N=6) and PMVSD (n=3)

Variable	Outlet VSD (n=6) (Mean±SD)	PMVSD (n=3) (Mean±SD)	p value
Mean Age (month)	40.16±30.32	128.67±48.89	0.0111
Mean Wt (kg)	11.65±3.57	25.73±6.25	0.0031
Mean Ht (cm)	90± 14.28	140.67±23.44	0.0045
Mean VSD size (mm)	6.16±0.85	10.33±0.49	0.0001
Mean PASP (mmhg)	38±15.26	38.33±1.24	0.9722

Data was expressed as Mean±SD, Unpaired t test was done as a test of significance, p value <0.05 was considered statistically significant

Discussion

Cusp prolapse and aortic regurgitation (AR) in patients with a VSD is attributed to many mechanisms that include deficient structural support for leaflets adjacent to the VSD, abnormal commissural suspension, lack of appositional forces, lack of continuity between the aortic media, annulus of AV, and the ventricular septum.[5,6]

Many studies have described more incidence rate of cusp prolapse in outlet VSD than PMVSD similar to the findings of this study. Salleb SF et al found prevalence of cusp prolapse in outlet VSD as high as 73%. But in case of PMVSD like this study they documented the prevalence of cusp prolapse as low as 14%.[7] Chiu SN et al[8] investigated 677 cases of outlet VSD and found cusp prolapse in 373 cases which was approximately 57.2%. Some other studies have reported the prevalence of cusp prolapse in outlet variety in Asian countries as high as 36%-79%.[9,10]

Outlet VSD possess more threat of coronary cusp prolapse as because these patients usually have deficiency of muscular and fibrous support below the aortic valve. This deficiency of fibroskeletal component along with venturi effect causes herniation of the right coronary leaflet through the VSD.[11]

Conclusion

Coronary cusp prolapse is more prevalent in Outlet VSD and it occurs at lower age group and smaller VSD shunt size compared to PMVSD.

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Original Article

Vitamin D Level and Left Ventricular Systolic Function in Children with Congenital Heart Disease Compared to Healthy Control

Diana Islam[1], Md. Tariqul Islam[2], Tahmina Karim[3], Chaity Barua[4], Kashid Omar[5], Rejaul Hayat[6]

Abstract

Background*: Congenital heart disease (CHD) is one of the main causes of death especially in the first year of life, and is one of the health problems that causes physical and mental disabilities in children. Vit-D is a fat soluble vitamin and vit-D defficiency is a highly prevalent condition present in all age group. Vit-D receptor is present in heart muscle and endothellium for which it is important for cardiac function by various modalities.*

Objective*: To measure Vit-D level and assess LV systolic function in children with CHD and compare to healthy control.*

Methods*: It was an observational case control study. The total sample size was 56 with 28 cases and 28 healthy controls. Patients diagnosed as congenital heart disease in paediatric cardiology department , BSMMU and aged upto 18 years were included as case group. Blood samples were sent for analysis of Vit-D in Department of Biochemistry,BSMMU. Transthoracic echocardiography was done for assessing left ventricular systolic function. Data were collected on a pre-designed questionnaire and analysed by using computer based SPSS program (version 22.0)*

Results: *The study was conducted on case (n=28) and control (n=28) group. Their mean age was 50.07±47.44 months and 80.5±54.41 months respectively. Mean Vit-D level in case and control group was 16.59±4.38 ng/ml and 27.95±6.21 mg/ml which was statistically significant. Mean Vit-D level in Acyanotic(n=15) and Cyanotic(n=13) patients was 18.69±3.25 ng/ml and 14.17±4.26 ng/ml which was also statistically significant. In both case and control group LV systolic function parameters including Ejection fraction and Fractional shortening positively correlated with serum Vitamin D level.*

Conclusion*: Vitamin D level is lower in CHD patients in comparison to healthy controls and cyanotic CHD exhibits much lower vitamin D level than acyanotic variety. LV systolic function correlates positively with vitamin D level in both CHD patients and healthy controls.*

Keywords: *Vitamin D, Left Ventricular Systolic Function, Children, Congenital Heart Disease.*

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Introduction

Congenital heart disease (CHD) represents a major health problem and most common cause of congenital anomalies. It is one of the most frequently diagnosed congenital disorders affecting approximately 0.8% to 1.2% live birth worldwide.[1] 25(OH) Vit D is a fat-soluble vitamin and a steroid hormone that is required for the proper functioning of many organs of the body.[2] 25(OH) Vit D deficiency is a highly prevalent condition, present in approximately 30% to 50% of the general population including all age groups, ethnicities and genders. In Bangladesh estimated prevalence of vit-D deficiency is approximately 75% which is much higher than other parts of Asia.[3] Vitamin D deficiency causes oxidative stress, Immune dysfunction, secondary hyperparathyroidism, hypocalcaemia. All these phenomenon lead to inflammation of cardiac myocyte along with thrombosis, vascular smooth muscle proliferation and vascular dysfunction. These pathological processes can be fatal in presence of congenital heart disease as patients of CHD are vulnerable to all form of cardiovascular malfunction.[4] So this study was conducted to measure Vit-D level and assess LV systolic function in children with CHD and compare that to healthy control

Materials and Methods

It was an observational case control study conducted at the Department of Paediatric cardiology in Bangabandhu Sheikh Mujib Medical University (BSMMU) from July 2020 to June 2021. All congenital heart disease patients of paediatric cardiology department, BSMMU aged upto 18 years will be included as case and healthy children of similar age group will be included as control in this study. Patients using drugs that affect calcium and bone metabolism, chronic liver or kidney disease, hyperparathyroidism, patients taking anticonvulsants and parents not willing to take part in the study were excluded. After obtaining written informed consent from parents patient particulars were documented properly. Patients of CHD who were diagnosed via echo-cardiography by G&E Echo machine ModelVividS70N was included as case. Then healthy children of similar age group were included as control. Blood sample was taken following standard procedure for measuring Vit-D level of both case and control and was sent to Biochemistry department of BSMMU for analysis. Then vitamin D level of case and control group was compared. Also the vitamin-D level of cyanotic and acyanotic CHD group was compared. LV systolic function of both case and control was assessed by determining ejection fraction (EF%) and fractional shortening (FS%) in M-mode echocardiography. Again these values of two groups were compared. Finally vitamin-D level and EF and FS were correlated in case and control group. Data analysis was done by statistical software, SPSS 22.

Result

Mean age of the study subjects was 50.07±47.44 months. Mean weight and mean height was 11.8±6.99 kg and 91.9±24.4cm respectively. Mean BSA was 0.52±0.24m2. Most of the study subjects of the case (57.14%) and control (68.6%) group came from rural areas (Table-I).

Table-I

Demographic data of study population

Variable	Case (n=28) (Mean±SD)	Control (n=28) (Mean±SD)
Age (Month)	50.07±47.44	80.5±54.41
Weight (kg)	11.8±6.99	20.3±8.26
Height (cm)	91.9±24.4	116.6±30.15
BSA (m2)	0.52±0.24	0.77±0.26
Residence		
Rural	57.14%	68.6%
Urban	42.86%	31.4%

*Results were expressed as mean±SD, SD=Standard deviation

Acyanotic CHD cases were more in number 15 in total and cyanotic cases were 13. Among acyanotic CHD most of the cases were VSD 53.33% followed by PDA 40% and ASD 6.7%. Regarding acyanotic CHD majority cases were TOF 61.5% accompanied by few complex CHD including Pulmonary atresia (15.4%), Single ventricle (15.4%) and Tricuspid atresia (7.7%) (Table-II).

Table-II

Disease spectrum in Case group (n=28)

Name of Disease	Frequency (n)	Percentage (%)
Acyanotic CHD	15	53.6
VSD	8	53.3
PDA	6	40
ASD	1	6.7
Cyanotic CHD	13	46.4
TOF	8	61.5
Pulmonary Atresia	2	15.4
Single Ventricle	2	15.4
Tricuspid atresia	1	7.7

*Results were expressed as frequency and percentage; VSD=Ventricular septal defect, PDA=Patent ductus arteriosus, ASD=Atrial septal defect, TOF=Tetralogy of Fallot

Cyanosis, recurrent RTI and failure to thrive was documented as the most prevailing presenting feature with 60.7%,53.6% and 53.6% frequencies respectively. Other less common presentations were H/O cyanotic spell (32.4%), Dyspnoea(28.5%),Poor feeding(25%),chest pain (14.28%),palpitation(7.14%). Mean SPO2 was 84.6±13.7% (Table-III).

Table-III

Clinical Presentation in Case group (n=28)

Clinical presentation	Percentage (%)
Cyanosis	60.7
Recurrent RTI	53.6
Failure to thrive	53.6
H/O cyanotic spell	32.4
Dyspnoea	28.5
Poor feeding	25%
Chest pain	14.28
Palpitation	7.14
SPO2 (%) (mean±SD)	84.6±13.7

*Results were expressed as percentage; (H/O=History of, RTI=Respiratory tract infection)

Mean Vit-D level in case and control group was 16.59±4.38 ng/ml and 27.95±6.21 ng/ml respectively. As the P value of comparison of mean Vit-D level between case and control group done by unpaired t test is <.0001 the difference is extremely significant. So vit-D level was significantly lower in children with CHD than healthy control. Mean EF of case and control group was 60.71±4.73% and 63.89±5.02%. On the other hand mean FS of case and control group was 31.5±3.35% and 31.3±2.90 %. Both EF and FS was significantly lower in CHD cases (Table-IV).

Table-IV

Comparison of Vit-D and LV systolic function between case and control

Variable	Case (Mean±SD	Control (Mean±SD)	p value
Vit-D (ng/ml)	16.59±4.38	27.95±6.21	0.0001
EF (%)	62.71±3.73	63.89±5.02	0.3225
FS (%)	31.5±3.35	31.3±2.90	0.8121

*Results were expressed as (mean±SD); unpaired t test was done as a test of significance; **p value<0.05 is considered as significant; EF+Ejection Fraction, FS=Fractional shortening

Comparison of vitamin D level between case and control group in all age group shows vitamin D level is significantly lower in case group than the control group (Table-V).

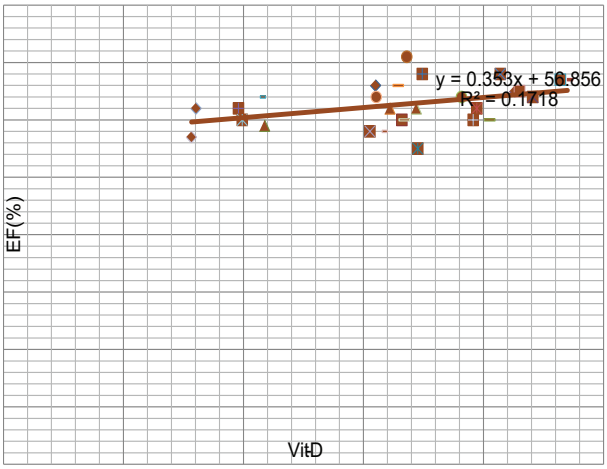
Table-V

Comparison of Vit-D level between Cyanotic and acyanotic CHD

Variable	Group	(Mean±SD)	p value
Vit-D(ng/ml)	Acyanotic (n=15)	18.69±3.25	0.0038
	Cyanotic (n=13)	14.17±4.26	

*Result was expressed as mean±SD; Unpaired t test was done as a test of significance; **P <0.05 was considered significant

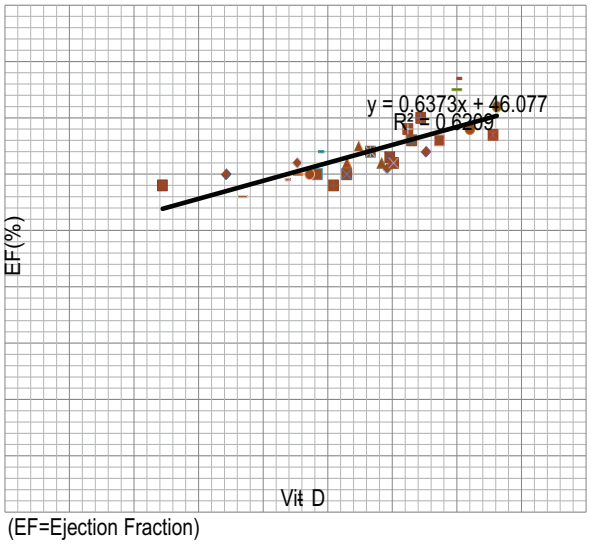
There is positive correlation between Serum Vit-D level and ejection fraction (EF%) in CHD cases (Fig.-1).



(EF=Ejection Fraction)

Fig.-1: *Correlation between Vit-D and EF(Ejection fraction) in case group*

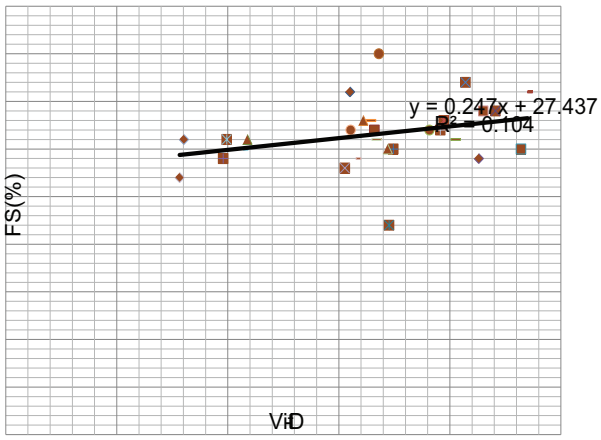
There is positive correlation between Serum Vit-D level and ejection fraction (EF%) in control group (Fig.-2).



(EF=Ejection Fraction)

Fig.-2: *Correlation between Vit-D and EF(Ejection fraction) in control group*

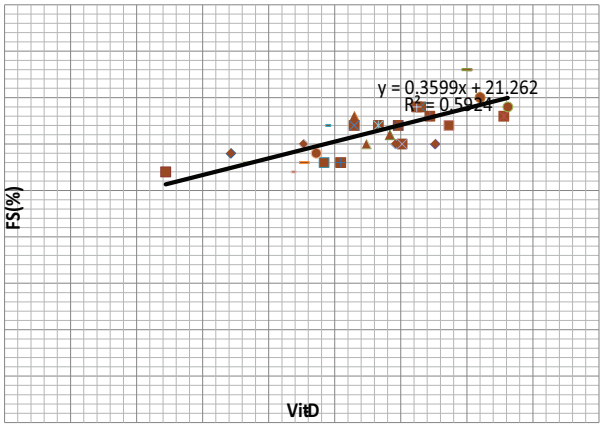
There is positive correlation between Serum Vit-D level and Fractional shortening (FS%) in CHD cases (Fig.3).



(FS=Fractional shortening)

Fig.-3: *Correlation between Vit-D and FS(Fractional shortening) in case group*

There is positive correlation between Serum Vit-D level and Fractional shortening (FS%) in control group (Fig.-4).



(FS=Fractional shortening)

Fig.-4: *Correlation between Vit-D and FS(Fractional shortening) in control group*

Discussion

Identification of 25(OH) Vit D receptors in endothelial cells, smooth muscle cells, and myocytes from the heart has led to the hypothesis that this 25(OH) Vit D has effect on cardiovascular function.[5] In this study, vit-D level was significantly lower in children with CHD than healthy control ($p<0.0001$). Noori NM et al found similar lower vit-D level in CHD cases than healthy control group in egyptian children.[6] This was in agreement with other studies also.[7]

In the present study, Cyanotic CHD has much lower vit-D level than acyanotic CHD. According to the findings of the study done by Nargesi et al[8] there was a significant lower levels of 25(OH) Vit D in the cyanotic group. Also Noori et al[6] exerted similar finding.

In the current study, LV systolic function assessed by ejection fraction and fractional shortening strongly correlate with vit-D level in both case and control indicating exclusive relation between serum vit-D level and LV systolic function.[9,10] Patange et al[11] experienced that 25(OH) Vit D inadequacy was strongly correlated with Left ventricular systolic function. The study identified that lower vit-D level was related with the expansion of left ventricular mass and subsequently systolic dysfunction.

Conclusion

Vitamin D level is lower in CHD patients in comparison to healthy controls and cyanotic CHD exhibits much lower vitamin D level than acyanotic variety. LV systolic function correlates positively with vitamin D level in both CHD patients and healthy controls.

Recommendation

- Vit-D strongly correlate with LV systolic function in both CHD and healthy control. So vit-D supplementation should be considered in children with LV systolic dysfunction
- Large sample size is required.
- Should include diastolic function along with systolic function.

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Original Article

Pattern of Heart Disease in Neonate Admitted in SCABU: Experience of Bangladesh Shishu Hospital & Institute

MA Kamal[1], Mohammad Abdullah Al Mamun[2], Md. Mahbubul Hoque[3]

Abstract

Background: *Like other developing countries, Bangladesh is facing a multitude of health problems and Congenital Heart Disease (CHD) is one of them. Without early recognition, diagnosis and treatment, a majority of infants and children with CHD die in their first month of life in developing countries.*

Objective: *This study was conducted to see the pattern of congenital heart disease in neonate admitted in SCABU.*

Methodology: *This cross sectional study was conducted in SCABU of Bangladesh Shishu Hospital & Institute from February 2020 to July 2020. Routine clinical examination was done within 24 hours of admission. All newborns recruited into the study were screened using transthoracic echocardiography by pediatric cardiologist after the initial evaluation. Neonates with CHD admitted during the study period were finally included. Data were collected and analyzed by using SPSS version 26.*

Results: *Total 47 neonates with CHD were admitted during the study period. Mean age were 9.03±7.26 days (range 1-28 days). Male were 55.32% and female were 44.68% with a male female ratio 1.2:1. Among the neonates term were 80.85% and preterm were 19.15%. A large group of neonate presented with cardiac murmur 15(31.92%), respiratory distress 20(42.55%), cyanosis 11(23.40%) and heart failure 1(2.13%). Majority of CHD were acyanotic (33, 70.21%) among them VSD 21.28%, ASD 25.53%, PDA 17.02%. Among cyanotic heart disease (8, 17.02%) TGA (4.25%), TOF (6.38%), pulmonary atresia (2.13%) and TAPVC (2.13%) were common. Among the neonates with heart disease 37(78.72%) were discharged, 5(10.64%) died, 5(10.64%) leave against medical advice.*

Conclusion: *This study shows that the most common acyanotic CHD in neonate in SCABU is ASD, VSD and PDA, and the most common cyanotic CHD is TOF and TGA.*

Keywords: *Congenital Heart Disease, pattern, neonate, SCABU.*

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Introduction

After commendable success in vaccine preventable diseases and overall reduction of communicable diseases, countries will face an epidemiological transition marked by a shift in the relative contribution of communicable and non-communicable diseases as major causes of childhood morbidity and mortality. In the next two decades, these changes are likely to occur in the 68 countries where current under 5 mortality is at least 35/1000 live births. As a result, most countries will see a steady increase in the relative importance of deaths due to congenital anomalies, non-communicable diseases, and injuries.[1]

Bangladesh is facing a multitude of health problems and congenital heart disease (CHD) is one of them. CHD occurs in 8 per 1000 live births.[2,3] CHD are serious conditions that have significant impact on morbidity, mortality. Without early recognition, diagnosis and treatment, a majority of infants and children with CHD die in their first month of life in developing countries.[4] So congenital malformations including CHD are now emerging as one of the leading cause of neonatal and under-5 mortality in Bangladesh. Without proper diagnosis and treatment, a majority of infants and children with cardiac disease both congenital and acquired die in developing countries and bear an increasing burden on health systems.

Hussain et al[5] during early nineties found only 8.3% CHD at neonatal period admitted in Dhaka Shishu (Children) Hospital. During January 1998 to December 1999 only 11.9% CHD were diagnosed during neonatal period and during January 2008 to December 2009 number increased to 27.5% in Dhaka Shishu (Children) Hospital.[6] A study at the Royal Brompton Hospital, in England, showed that most infants hospitalized with the diagnosis of CHD were neonates.[7] Recent data revealed 30% CHD was detected during neonatal period in Bangladesh.[8] That means diagnosis of CHD during neonatal period is increasing day by day. If these affected neonates left untreated causes serious morbidity and mortality, therefore early detection and proper intervention is most important.

The most common CHD has been ventricular septal defect, followed by atrial septal defect, patent ductus arteriosus, tetralogy of Fallot, single ventricle, atrium ventricular septal defect and double outlet right ventricle.[9] There are very few authentic data regarding the pattern of CHD at national level in Bangladesh. The objective of the study is to identify the pattern of congenital heart diseases in SCABU of Bangladesh Shishu Hospital & Institute.

Materials and methods

This cross sectional study was conducted in SCABU of Bangladesh Shishu Hospital & Institute from February 2020 to July 2020. Routine clinical examination was done within 24 hours of admission. This was recorded in a form that included the following parameters: Central cyanosis, murmur on chest auscultation, and respiratory distress. Furthermore, the field investigator for all newborns obtained non-invasive arterial oxygen saturation. Oximetry values were obtained from one of the feet of the baby. A persistent saturation of <95% were considered abnormal.

All newborns recruited into the study were screened using transthoracic echocardiography after the initial evaluation. A pediatric cardiologist performed echocardiography using the ultrasound system. The technique involved performing cross-sectional echocardiography, and Doppler and color flow imaging in various views. The cardiologist was not aware of the results of the initial clinical evaluation. Data were collected and analyzed by using SPSS version 26.

Results

Total 47 neonates with CHD were admitted during the study period. Mean age were 9.03±7.26 days (range 1-28 days). Male were 55.32% and female were 44.68% with a male female ratio 1.2:1 (Fig.-1). Among the neonates term were 80.85% and preterm were 19.15% (Fig.-2).

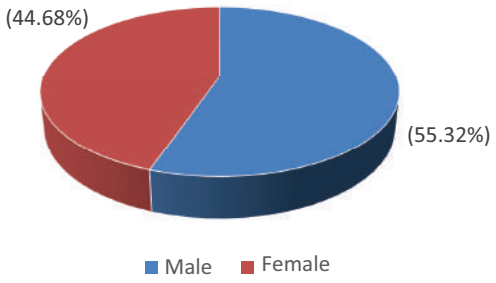

Fig.-1: *Distribution of sex of neonates in SCABU (n=47)*

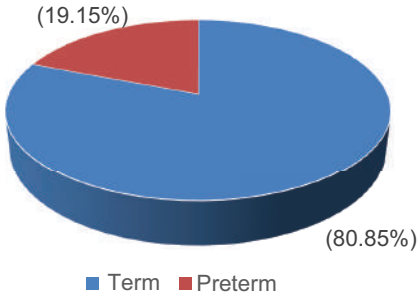

Fig.-2: *Distribution of gestational age of neonates in SCABU (n=47)*

A large group of neonate presented with cardiac murmur 15(31.92%), respiratory distress 20(42.55%), cyanosis 11(23.40%) and heart failure 1(2.13%) [Fig.-3].

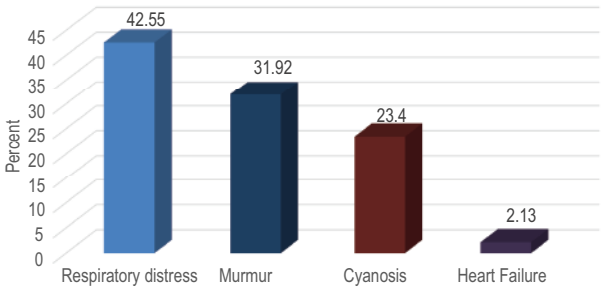

Fig.-3: *Distribution of clinical suspicion of CHD in neonate (n=47)*

Majority of CHD were acyanotic (33, 70.21%) among them VSD 21.28%, ASD 25.53%, PDA 17.02%. Among cyanotic heart disease (8, 17.02%) TGA (4.25%), TOF (6.38%), pulmonary atresia (2.13%) and TAPVC (2.13%) were common (Table-I).

Table-I

Distribution of heart disease among neonates in SCABU (n=47)

Diseases	Number (%)
Acyanotic CHD	33(70.21)
ASD	12(25.53)
VSD	10(21.28)
PDA	8(17.02)
AV Canal Defect	1(2.13)
Asymmetrical Septal Hypertrophy	2(4.25%)
Cyanotic CHD	8(17.02)
TOF	3(6.38)
TGA	2(4.25)
Pulmonary Atresia	1(2.13)
TAPVC	1(2.13)
HLHS	1(2.13)
Others	6(12.76)
PPHN	6(12.76)

*VSD: Ventricular Septal Defect, ASD: Atrial Septal Defect, PDA: Patent Ductus Arteriosus, AV canal defect: Atrioventricular canal defect, TOF: Tetralogy of Fallot, TGA: Transposition of Great Arteries, TAPVC: Total Anomalous Pulmonary Venous Return, HLHS: Hypoplastic Left heart Syndrome, PPHN: Persistent Pulmonary Hypertension of Newborn

Neonates were admitted with perinatal asphyxia (20, 42.55%), sepsis (13, 27.66%), prematurity (6, 12.77%), MAS (4, 8.51%), IDM (2, 4.25%), TTN (1, 2.13%) and RDS (1, 2.13%) (Fig.-4).

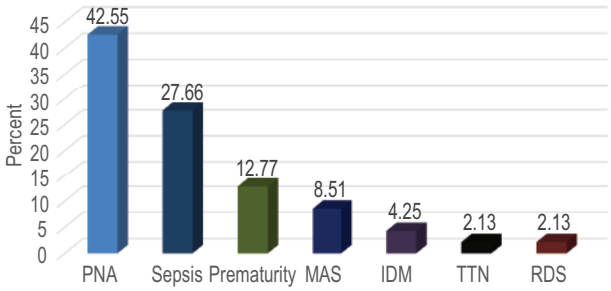

Fig.-4 *:Admission diagnosis of the neonates with HD*

Among the neonates with heart disease 37(78.72%) were discharged, 5(10.64%) died, 5(10.64%) leave against medical advice (Fig.-5). One neonate with VSD, 1 TGA, 1 PA, 1 HLHS and 1 PPHN died.

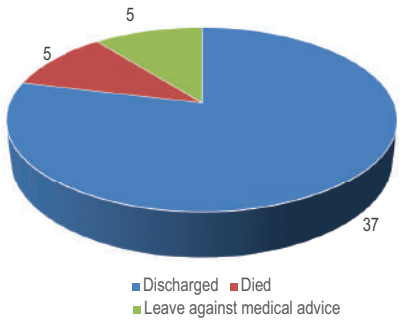

Fig.-5: *Distribution of outcome of neonates in SCABU (n=47)*

Discussion

Mean age of neonates were 9.03±7.26 days among them male were 55.32% and female were 44.68% with a male female ratio 1.2:1. Among the neonates term were 80.85% and preterm were 19.15%. A large group of neonate presented with cardiac murmur (31.92%), respiratory distress (31.92%), cyanosis (23.40%) and heart failure (2.13%). Majority of CHD were acyanotic (70.21%) among them ASD (25.53%), VSD (21.28%) and PDA (17.02%) were commonest. Among cyanotic heart disease TGA (4.25%), TOF (6.38%), pulmonary atresia (2.13%) and TAPVC (2.13%) were common. Majority (78.72%) were discharged, 10.64% died, 10.64% leave against medical advice. Majority of the neonates were admitted with perinatal asphyxia, sepsis, prematurity, MAS, IDM, TTN and RDS.

In this study, boys are outnumbered than girls with a ratio of 1.2:1. Rakkappan et al[10] in India found that CHD was more common in female births, which was very similar to the study conducted in Nigeria.[11] However, our finding is similar to that reported by Nikyar et al[12] from Gorgan, Iran where there is a male preponderance and found out the ratio of male:female is 1.35. Alabdulgader et al[13] from Saudi Arabia and Stephensen et al[14] from Iceland reported that the frequency was the same for males and females. In this study CHD was common among term neonate. Rakkappan et al[10] in India also found term neonates were affected more with CHD than preterms. In contrast to our study, Steurer et al[15] in California found out that preterm babies are at higher risk of CHD.

This study showed that majority of CHD are of acyanotic CHD. Among acyanotic CHD the pattern of CHD is ASD followed by VSD and PDA and among cyanotic CHD TOF was commonest followed by TGA and Pulmonary Atresia 2.13%. Rakkappan et al[10] in India showed that majority of CHD are acyanotic (97%) and among acyanotic CHD ASD was the commonest (52%), followed by PDA (40%) and VSD (10%). The most frequent type of CHD that this study found is ASD, which is in accordance with a recent study done in Saudi by Majeed-Saidan et al[16]. Furthermore, another study in Iran by Rahim et al[17] cited the most common CHD in the newborn as ASD, while in other studies, the most frequent type of CHD was VSD.[18-20] Uddin et al[21] in Bangladesh found that VSD was the most common type of acyanotic congenital heart disease followed by ASD and PDA. Among the cyanotic congenital heart disease, tetralogy of Fallot was the most common abnormality. Majeed-Saidan et al[16] reported the pattern as ASD, followed by VSD. In China among the live births, the top three lesions were ventricular septal defect (VSD), patent ductus arteriosus, and atrial septal defect, which accounted for 34.0%, 23.7%, and 10.8%, respectively.[22] In Bangladesh one multi-center study and one past and present situation analysis found that VSD remains at the top of the list followed by ASD, PDA, TOF and TGA. VSD was the commonest among acyanotic CHD and TOF was the commonest cyanotic CHD.[6,8]

A large group of neonate presented with cardiac murmur, respiratory distress, cyanosis and heart failure. There is a popular believe that murmur in neonatal period has no importance, it is a physiological phenomenon.[23] But it is not true at all times. Murmur in neonatal period may be the first sign of underlying serious structural cardiac defect. Uddin et al[21] have found the incidence rate of cardiac murmur is only 1.26%. These variations may be due to the examiners skills and experience,

the timing and frequency of examination. Mortality was high among neonates with complex congenital heart disease. Co morbid condition like prematurity, perinatal asphyxia and sepsis also contributed in higher mortality.

Conclusion

This study shows that the most common acyanotic CHD in neonate in SCABU is ASD, VSD and PDA, and the most common cyanotic CHD is TGA and TOF. Mortality is high among neonates with complex cyanotic heart disease.

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Review Article

Arrhythmias in Emergency in Paediatric Age Group

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Abstract

Unstable Arrhythmias are rare in Pediatric Emergency (ER)/Out Patient Department (OPD). The symptoms of palpitation and syncope are common. Sinus tachycardia & supraventricular tachycardia are more common. Emergency nurses & physicians should be prepared for diagnosis and acute management as well as further diagnostic testing and cardiology evaluation and follow up.

Keywords: *Arrhythmia, narrow complex & wide complex tachycardia, supraventricular tachycardia.*

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Introduction

True emergencies' due to unstable arrhythmias are rare in pediatric ER/OPD; symptoms of palpitation and fatigue and syncope are more common. Pediatric arrhythmias account for ~ 55/100,000 patients evaluated in ER.[1] Sinus tachycardia is the most common one- followed by supraventricular tachycardia (SVT) and bradycardia; they account for 13% & 6% respectively.[2] Primary care physicians, emergency physicians and intensive care physicians should be prepared for acute management and further evaluation and referral.

Arrhythmias in children are seen both in structurally normal and congenitally diseased hearts. They again can be broadly classified in 2 groups: Narrow QRS complex and wide QRS complex.

Tachycardia: Narrow Complex QRS

Supraventricular tachycardia (SVT) is a narrow QRS complex tachycardia, generally[2] refers to any arrhythmia originating at or above the bundle of His and QRS duration is less than 100ms (Fig.-1a). Three mechanisms are described - the re-entry tachycardia is the most common and the circuit (circus movement) can originate within atrium and may produce atrial flutter or fibrillation; it may originate at the level of AV node (AV node reentry tachycardia) or via an accessory pathway, bundle of Kent (AV re-entry tachycardia). The second mechanism is enhanced automaticity resulting in tachycardia - the focus can be sinus node (as in sinus tachycardia) or elsewhere in atria. Sinus tachycardia is commonly seen in fever, anemia, hypovolaemia and medications that increase catecholamines and is uncommonly in cardiac disease conditions. There is sinus P wave preceding every QRS complex in sinus tachycardia; rates exceed 140 bpm in children and 180bpm in infants but usually less than 200bpm (Fig.-1b). Treatment is aimed at underlying cause. Triggered tachycardia is less common mechanism of tachycardia and usually occurs in a setting of drug toxicity like digoxin overdose.[3]

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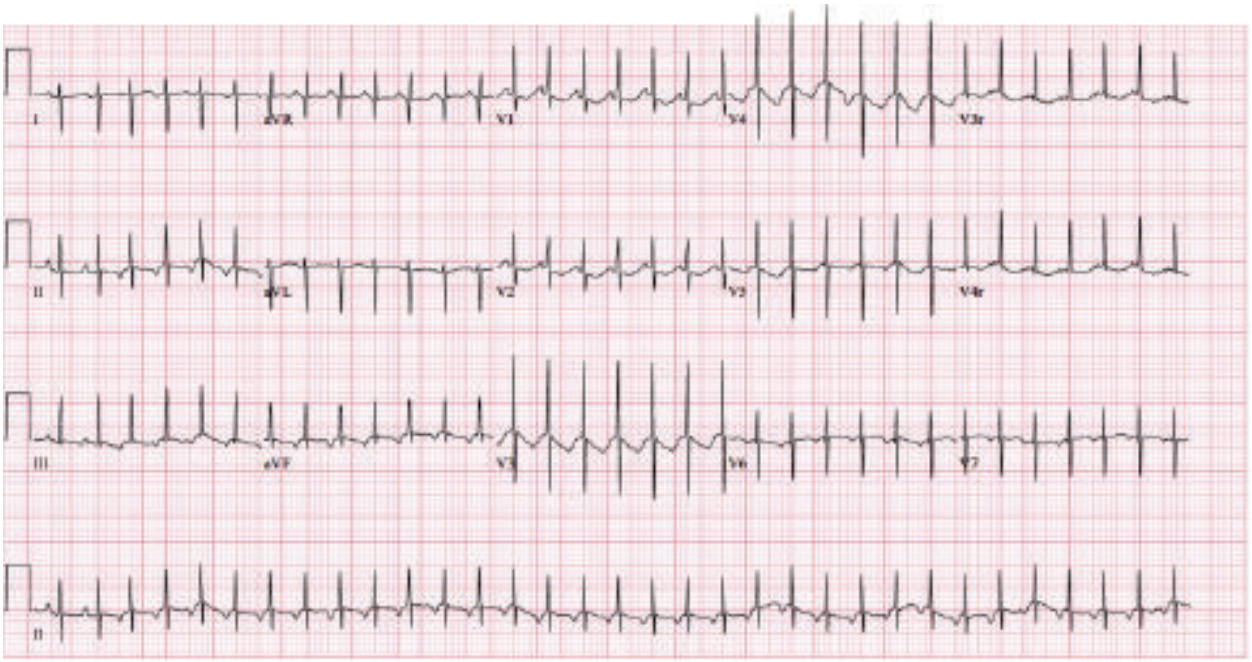
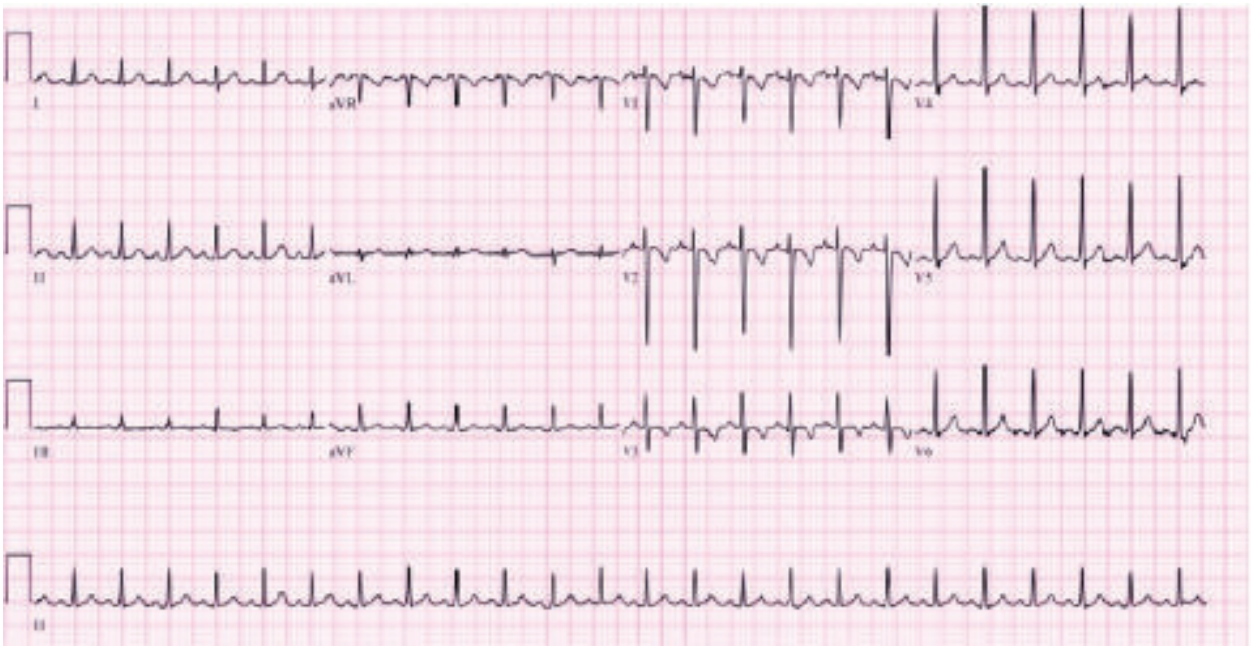


1(a)

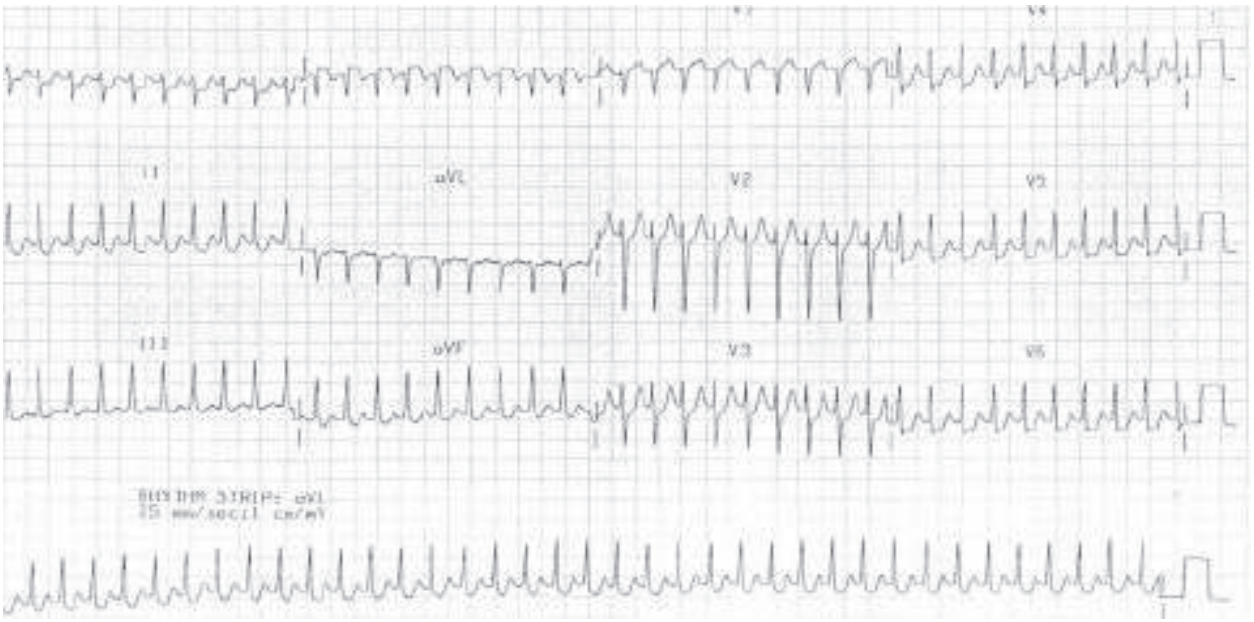


1(b)

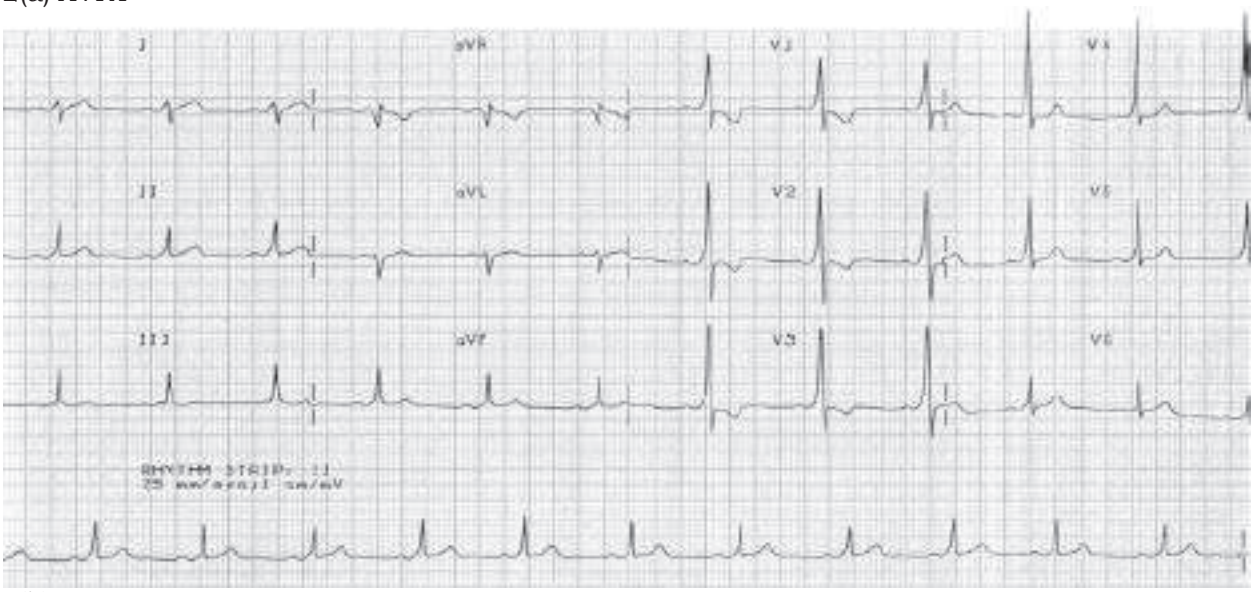
Fig.-1: (a)*Narrow QRS complex, 1:1 AV ratio, short RP interval (<"80/ ms), and stable cycle length; (b) Sinus tachycardia*

AV reentry (AVRT) and AV node reentry tachycardia (AVNRT): AVRT - this is also known as accessory pathway mediated tachycardia- an *orthodromic* reciprocating tachycardia. It is the most common type of SVT seen in children- about 82% of arrhythmias in infants. There are 2 distinct pathways between atria and ventricles-thus creating an electric circuit down the AV node and then up an accessory pathway outside AV node, thus creating a

narrow QRS complex tachycardia (Fig.-2a). The other much rarer *antidromic* tachycardia reverses the direction of conduction and creates a wide QRS coplex tachycardia. Once normal sinus rhythm is restored 50% of patients manifest pre-excitation in the form of a delta wave on electrocardiogram consistent with Wolf-Parkinson-White syndrome (WPW); the remainder of patients have a concealed pathway not evident during sinus rhythm. The only difference between the 2 forms is the manifest antegrade conduction to the ventricle in WPW; but conduction through the accessory pathway can occur before the AV node is activated and a shortened PR interval, and slurred QRS upstroke and widened QRS result (Fig.-b). This also creates the potential for rapid ventricular response during atrial fibrillation, which can result in ventricular fibrillation and sudden death. Although acute management of SVT is the same for patients with concealed and manifest pathways, definitive ablation therapy in patients with WPW is compelling.[5,6]



2(a) AVRT



2(b) WPW

Fig.-2: *(a) AVRT; (b) WPW syndrome. Note the short PR interval and slurred QRS upstroke (delta wave)*

AV node reentry tachycardia (AVNRT): This is also a reentry circuit, and the pathways involve the AV node; there are 2 discrete conduction limbs - a slow *antegrade* pathway and a fast pathway which conducts retrograde. AVNRT accounts for 15% SVT in pediatric age group, increases with age and rare in infancy. These 2 types of tachycardia are clinically indistinguishable. Infants with AVNRT may go unrecognized for prolonged periods resulting in cardiovascular deterioration. Therapy is similar in both types of SVT-briefly interrupting conduction through the AV node with adenosine terminates both types, and long-term medication and ablative therapy are similar.

Clinical features: H/O poor feeding, lethargy, irritability & pallor in infant. Palpitation dizziness chest pain syncope and shortness of breath in older children & adolescents. Signs and symptoms of congestive heart failure. H/o abrupt onset & termination.

ECG-findings: HR is >220 in infants and >180 in children with narrow QRS complex and AV ratio 1:1. In AVNRT terminal QRS notching may be visible. In AVRT inverted P waves may visible (Fig.-3).

Acute Management: Vagal maneuvers (application of ice; Valsalva maneuver, or carotid massage) help abolish acute episode; if unsuccessful - intravenous adenosine 50mcg/kg rapid bolus followed by saline flush with increasing doses as needed; effective dose ranges 100-150mcg/kg; a maximum dose is 250mcg/kg.

Synchronized DC Cardioversion if adenosine is unsuccessful; starting dose being 0.5J/kg and increased in steps upto 2J/kg. Intravenous amiodarone (verapamil) is an alternative in experienced hand and should be avoided in infants.

Further work up and disposition: Once normal sinus rhythm is established a repeat ECG is done to rule out pre-excitation and underlying cardiac disease. Serum electrolytes & thyroid function and CBC are sent. All patients under one year of age and those hemodynamically unstable should be admitted for observation. Patients with first time episode and normal hearts should be observed overnight w-out treatment. Patients with structural heart disease should be treated on individualized basis. If patient is discharged instructions on vagal maneuver should be provided to treat recurrence.

Initiation of maintenance therapy would depend on patient age and severity of symptoms and recurrence; and should be discussed with parents.

Infants w-out preexcitation should be put on oral propranolol for 12mo. Children with breakthrough SVT benefit from amiodarone orally. Verapamil should be used with caution in poor LV function and young infants. Infants & children with/-out preexcitation have long term benefit from atenolol or nadolol.

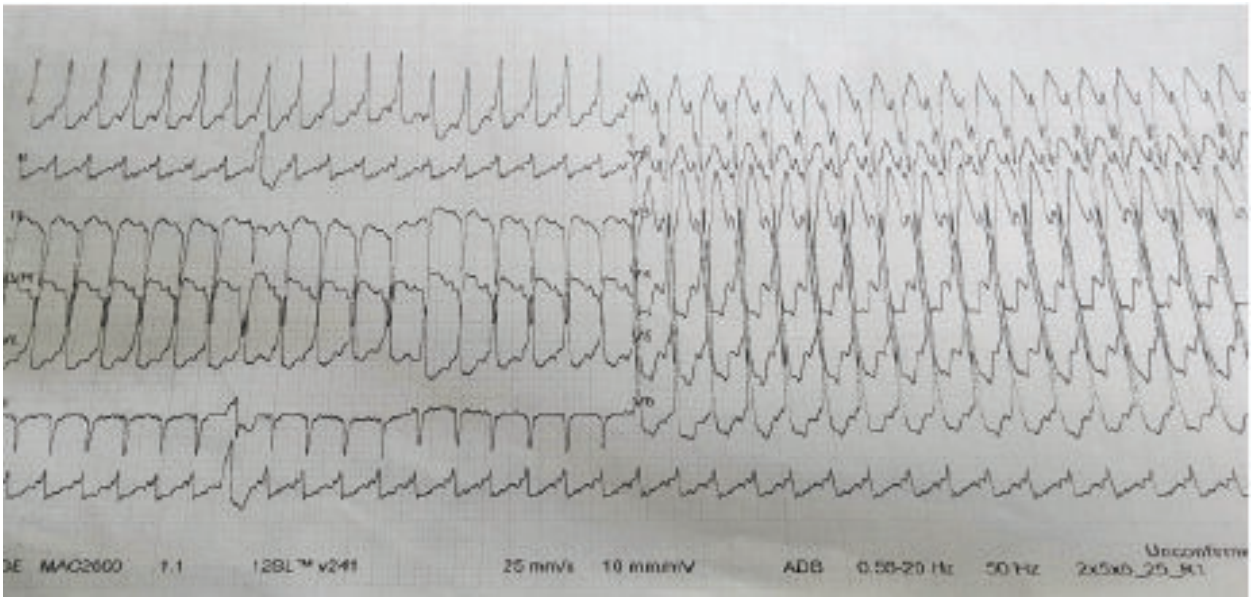


Fig.-3: *AVNRT*

Caution: Digoxin or verapamil may increase the antegrade conduction through accessory pathway and should be avoided.

Cardiology follow up is recommended for all new patients; a copy of ECG should be provided to parents to be brought during their next visit.

Infants with suspected prolonged episode should have an echocardiography to rule out thrombus and ventricular dysfunction. Otherwise, echocardiogram may be obtained during cardiology evaluation. Patients with asymptomatic preexcitation on ECG should also have routine cardiology evaluation.

Prognosis: The probability of complete resolution of SVT is dependent on age of onset. In the majority of infants diagnosed at 1 year of age or less, SVT is likely to resolve, compared to only 33% of patients diagnosed after 1 year of age .(6) Maintenance therapy includes the use of beta-blockers in cases of recurrent and/or prolonged tachycardia. Digoxin and calcium channel blockers should not be prescribed in patients with WPW prior to full cardiology evaluation (See note of caution above). Sotalol, flecainide or amiodarone may be necessary for medical control of some SVT. Catheter ablation allows for definitive therapy for SVT, at low risk in patients beyond the toddler years. The success rate for accessory pathway ablation is above 90%. Risk of heart block is 1.2-10.4% ; overall risk of complication being 3-4%

Atrial tachycardia -Atrial flutter

It is rarely seen beyond the newborn period except in the setting of underlying congenital heart disease and cardiac surgery, where it is most common in children who have undergone Fontan procedures, atrial septal closure, and tetralogy of Fallot (TOF) repair. Congenital heart disease and post-operative patients represent about 80% of older children presenting with atrial flutter.[7] It accounts for about 30% of fetal tachycardia, 18% of tachycardias in newborn and only 8% in older children.[8] Atrial flutter involves a single reentry circuit within the atrial muscle, most commonly around the borders of the tricuspid valve. Hemodynamic compromise is determined by the duration of flutter and the degree of AV block, with 1:1 AV conduction resulting in the most significant instability. Clinical presentation depends on ventricular response. In the setting of rapid ventricular conduction, patients may be hemodynamically unstable with poor cardiac output.

Clinical features: Commonly detected *in-utero* during fetal ultrasound evaluation and results in fetal hydrops if prolonged. Most newborns are asymptomatic unless tachycardic for >48hours. Infants with prolonged tachycardia may present with history of poor feeding, irritability, lethargy, diaphoresis, and pallor and older children may complain of palpitations, chest pain and/or dizziness.

Electrocardiographic findings: Regular atrial rates of 200 bpm; AV ratio >1:1, variable but generally regular, ventricular rate ~100; typical saw tooth appearance may be seen in lead II, III and aVF and normal appearing QRS complex (Fig.-4):

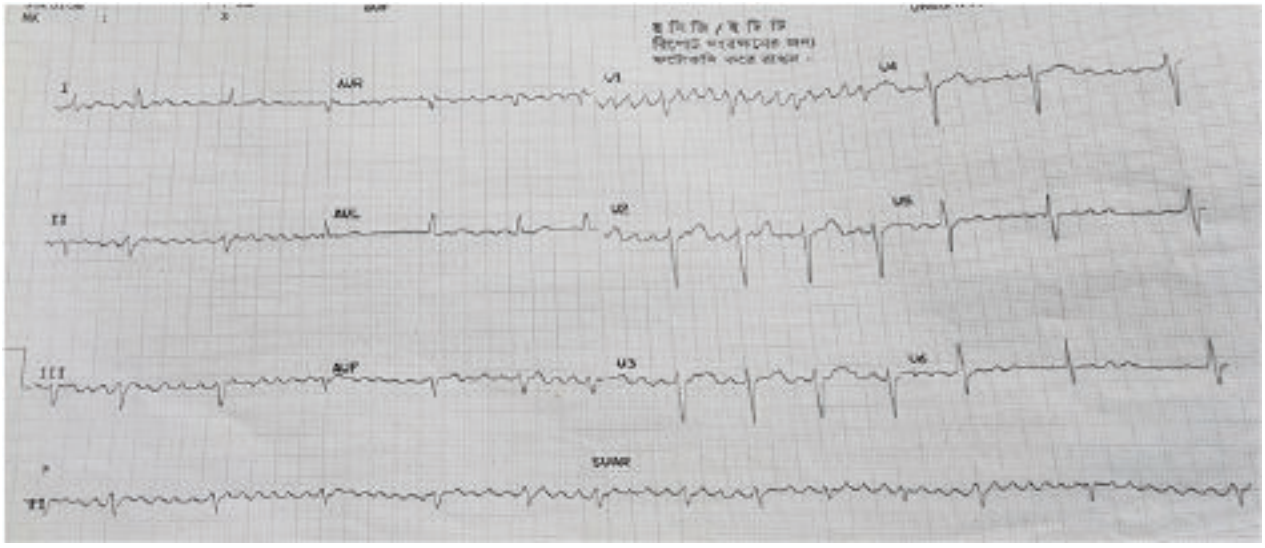


Fig.-4: *Atrial flutter with atrial rate more than 200 and ventricular rate of 100 bpm. Note the typical "sawtooth" appearance and AV conduction >1:1*

Acute management: Vagal maneuvers or adenosine do not convert this rhythm but may increase the degree of AV block, unmasking underlying flutter waves. If patient has rapid ventricular response with reduced cardiac output, or for elective cardioversion, use DC cardioversion starting at 0.5J/kg, and increasing to 1-2J/kg if needed is done. If stable, pharmacotherapy can be aimed at rhythm control with IV amiodarone or ibutilide, or at rate control with beta-blockers or calcium-channel blockers.

In patients with pacemaker, pace termination can be attempted by the electrophysiologist cardiologist.

Further work-up and disposition: Serum electrolytes and thyroid function tests.

Cardiology consultation and echocardiogram to rule out/in underlying structural heart disease, tachycardia-mediated cardiomyopathy and thrombus. Admission for observation and Anticoagulation therapy for episodes >48 hours and in all Fontan patients.

Prognosis: In the presence of a structurally normal heart, the risk of recurrence of neonatal atrial flutter is low and no long-term therapy is needed. In cases of structurally abnormal heart and/or recurrent atrial flutter, long-term therapy may be required. Management is directed at either rhythm control with amiodarone and flecainide, or control of ventricular response with digoxin, calcium channel blockers or beta-blockers in selected patients. Catheter ablation can be highly successful in the older child.

Ectopic atrial tachycardia (EAT)

Ectopic atrial tachycardia is uncommon, accounting for about 10% of SVT in children.[9] It is due to enhanced automacity of single or multiple foci outside the sinus node and is often refractory to medical therapy and cardioversion. EAT is the most common cause of tachycardia-induced cardiomyopathy due to its persistent and chronic nature; improvement in cardiac function has been observed following successful treatment. The precise etiology is unknown; however, viral illness, atrial tumors and genetic predisposition have been implicated. Most patients present with minor symptoms in the setting of preserved function.

Clinical features: EAT is predominantly observed in infants and children with structurally normal hearts. Patients present with symptoms of chest pain, palpitations, pre-syncopal or syncopal events. Older children may present with exercise intolerance or be asymptomatic. Infants may present with feeding difficulties, diaphoresis with feeds or distress secondary to chronic tachycardia.

Electrocardiogram findings: Atrial rate that is inappropriately rapid for age. Sometimes difficult to distinguish from sinus tachycardia. Abnormal P wave morphology and axis. Atrial rate variable, ranging from 120 to 300 bpm. May exhibit a "warm up" period at initiation with progressive P-P interval shortening and a "cooling down" period prior to termination and >1:1 AV conduction can be seen inpatients at rest or while asleep without termination of the tachycardia (Fig.-5).

Acute management: EAT responds poorly to adenosine and DC cardioversion. First line treatment for symptomatic patients includes IV amiodarone loading with 5mg/kg bolus over 20–60 minutes; can be followed by a maintenance drip at 10-15mg/kg/day. Minimally symptomatic patients do not require acute treatment

Laboratory work-up: serum electrolytes, complete blood count, toxicology screen. If cardiac function is poor, additional laboratory evaluation should include viral panel, blood culture and cardiac enzymes to help differentiate infection versus tachycardia-induced cardiomyopathy. Cardiology consultation and echocardiographic evaluation is recommended. Obtain 24-hour cardiac monitor to determine duration of tachycardia and ventricular rates. Treatment is dependent on patient age, symptoms, clinical status, tachycardia rate and duration, and ventricular function. Asymptomatic patients can be treated conservatively. Children younger than 1 year

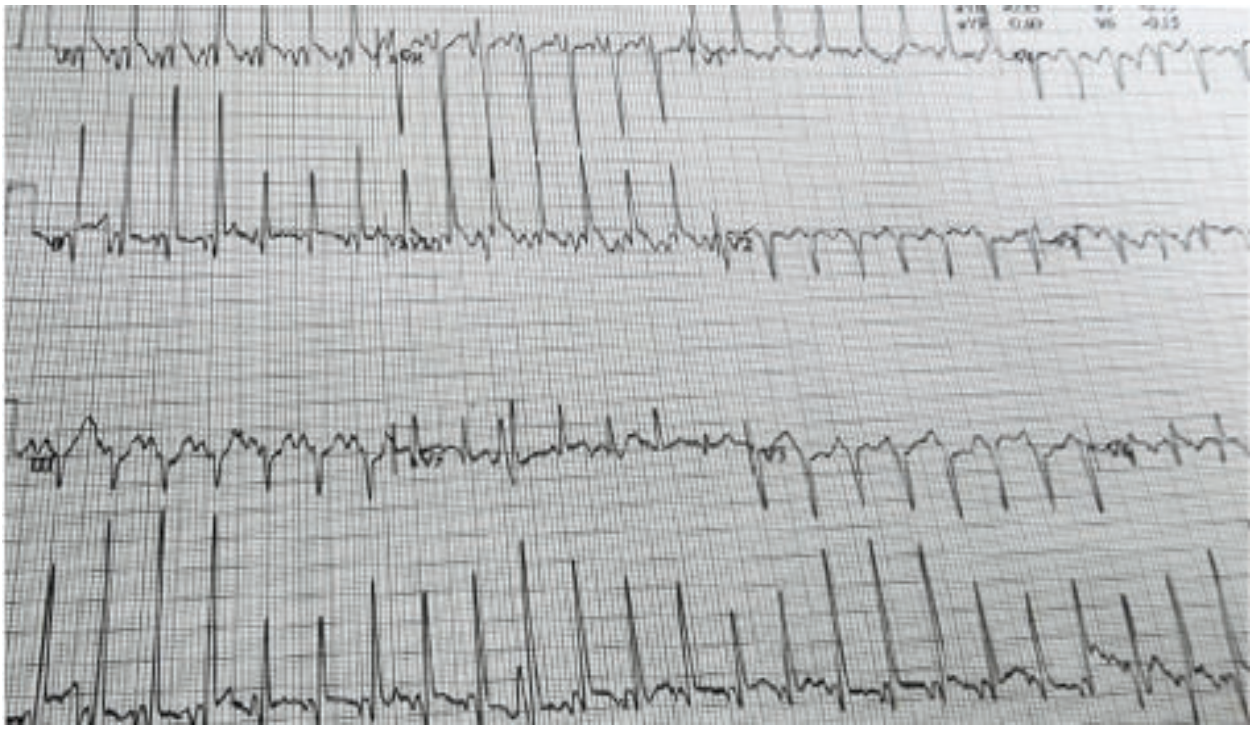


Fig.-5: *EAT in an asymptomatic infant with abnormal P wave axis. Note merging of T and P waves, HR 176*

and symptomatic patients should be admitted for management of arrhythmia and congestive heart failure. In the setting of normal cardiac function, beta-blockers can be useful by slowing AV node conduction, lowering the ventricular rate and improving symptoms. More aggressive treatment consists of flecainide, amiodarone or sotalol which have moderate success rates. Patients who fail medical therapy can undergo catheter ablation.

Prognosis: Spontaneous resolution may be observed in 30–50% of affected children. Children younger than 3 years have higher incidence of AET resolution with treatment versus older children who are likely to require RF ablation because of persistent tachycardia despite optimal medical management.[10,11]

Atrial fibrillation

Atrial fibrillation is mostly seen in children with underlying structural heart disease and in those who have undergone cardiac surgery. In infants and children with structurally normal hearts, atrial fibrillation is often associated with an AV accessory connection such as WPW. These patients can be at increased risk of sudden death if there is rapid conduction to the ventricle via a manifest accessory pathway. Atrial fibrillation can also be associated with cardiomyopathies, myocarditis, pericarditis and hyperthyroidism, and in rare instances has a genetic predisposition. The mechanism involves multiple reentry circuits predominantly within the left atrium. Patients are usually symptomatic at presentation and in the setting of rapid ventricular rates, hypotension and syncope may ensue. As in adults, the possibility of atrial thrombus with the risk of embolic stroke is of great concern.

Clinical features: Children and adolescents present with complaints of palpitations. Weakness and signs of congestive heart failure may be seen. In patients presenting with syncope, WPW should be highly suspected.

• Electrocardiogram findings (Fig.-6). They are as follows.

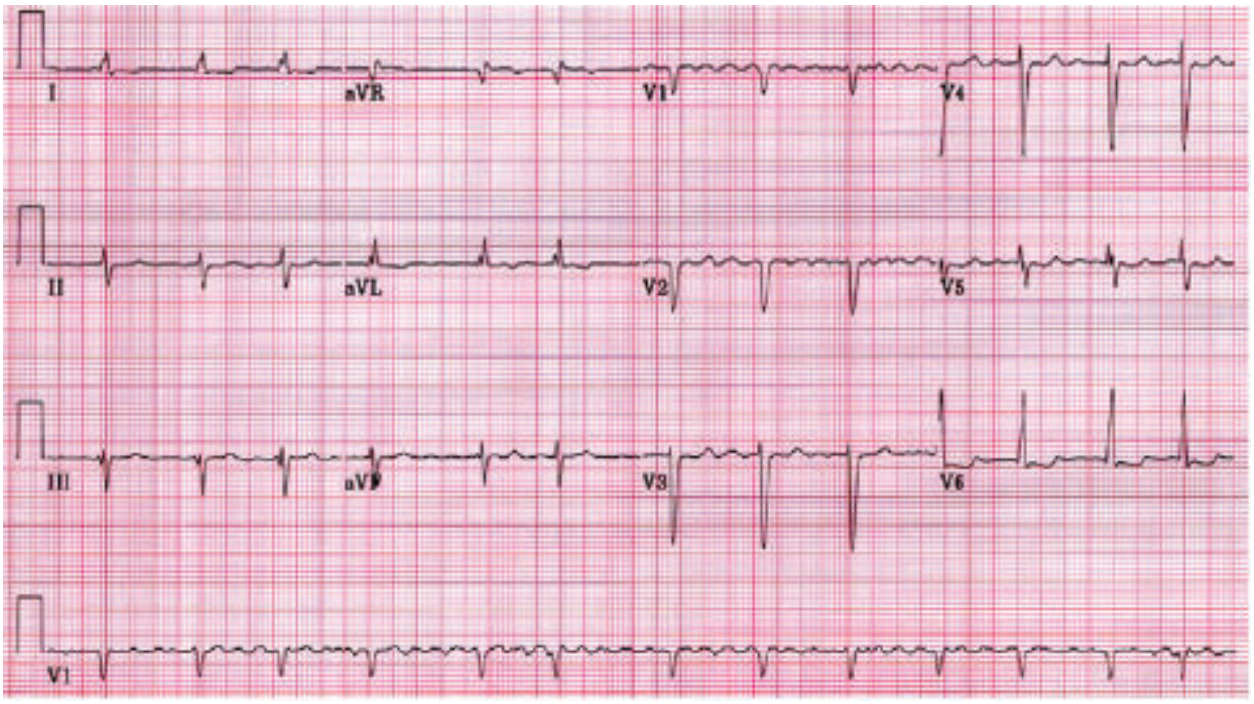


Fig.-6: *Atrial fibrillation in a 2½ year old asymptomatic patient. Note chaotic atrial waves with irregular ventricular response*

Chaotic/irregular atrial waves best seen in lead V Atrial rates of 350-600 bpm; no discrete, uniform P waves; AV ratio >1:1; variably changing ventricular response rate (irregularly irregular), ranging from 110 to 200 bpm and norrmal appearing QRS complex, except in WPW.

Acute management: If the patient has rapid ventricular response with reduced cardiac output, DC cardioversion starting at 2J/kg is used. For stable patients, pharmacotherapy aimed at ventricular rhythm or rate control can be used. IV rhythm control agents include amiodarone and ibutilide. Less effective are sotalol, digoxin and procainimide. Cardioversion in stable patients with unknown or prolonged (>48 hours) duration of tachycardia should be delayed until assessment for atrial thrombus is made with echocardiography.

Further work-up and disposition: Once in normal sinus rhythm, repeat EKG.

Laboratory work-up: serum electrolytes, thyroid function, complete blood count, toxicology screen. If cardiomyopathy is suspected, additional laboratory evaluation should include viral panel, blood culture and cardiac enzymes. Echocardiography is indicated in all cases. Admit for observation and treatment. Initiate anticoagulation therapy in most cases.

Prognosis: Despite medical therapy, atrial fibrillation has a high recurrence rate and often requires catheter or surgical intervention. Chronic anticoagulation therapy is indicated in patients with persistent or recurrent atrial fibrillation. Ablation of the AV node with implantation of a pacemaker may be necessary in some refractory cases.

Wide QRS complex tachycardias

Although less common in children than in adults, wide complex tachycardia still occurs with some frequency. Not all wide complex tachycardias are due to ventricular tachycardia (VT). The differential diagnosis of wide complex tachycardia includes VT, bundle branch block during SVT and SVT with pre-excitation in patients with WPW. If available, a previous 12-lead electrocardiogram may show underlying bundle branch block and can be helpful in distinguishing VT from SVT with aberrancy.

Due to the potential life-threatening nature of VT, all wide complex tachycardia should be treated as VT until proven otherwise.

A) Ventricular tachycardia

VT is a potentially life-threatening arrhythmia recognized as a cause of sudden death in both adults and pediatrics. It is rare in children and accounts for about 6% of patients followed for tachycardia.[12,13] Defined as a tachycardia originating below the bundle of His, rates can range from just over the sinus rate to well over 200 bpm. Episodes lasting less than 30 seconds are termed as non-sustained VT and those more than 30 seconds as sustained VT. VT can further be classified as monomorphic, with a regular rate and a single QRS morphology, versus polymorphic, with variability in rate and QRS morphology. The same basic mechanisms of automaticity, reentry and triggered tachycardia exist in VT as for other arrhythmias. Etiology is widely variable and includes idiopathic, drug toxicity, cardiomyopathy, myocarditis, cardiac tumors and metabolic abnormalities, to name a few.

Clinical features: Variable may present with symptoms ranging from dizziness and palpitations to syncope and cardiac arrest.

Electrocardiogram findings: Figure 7 shows the findings which are as follows.

Hallmarks are prolonged QRS duration for age and VA dissociation with ventricular rates exceeding atrial rates (AV ratio < 1:1). Rates range from ~110 to >200 bpm.

When VA conduction is 1:1, VT cannot be excluded. Features consistent with the diagnosis of VT rather than SVT with a wide QRS include variation in RR interval and presence of fusion complexes. Complexes may appear uniform or vary from beat to beat as in polymorphic VT.

Acute management: If unstable, synchronized cardioversionis started at 2J/kg and repeated, increasing the dose if needed. If stable, may attempt amiodarone at 5mg/kg IV over 30–60 minutes or procainimide at 15mg/kg IV over 30–60 minutes.

Further work-up and disposition: History should focus on prior symptoms, symptoms suggestive of myocarditis or long-standing cardiomyopathy, and the possibility of drug toxicity, as well as a thorough family history for known arrhythmias or history of sudden death.

Once in normal sinus rhythm, repeat EKG to rule out underlying abnormalities including long QT, Brugada, arrhythmogenic right ventricular cardiomyopathy, structural heart disease, electrolyte abnormalities and ischemia.

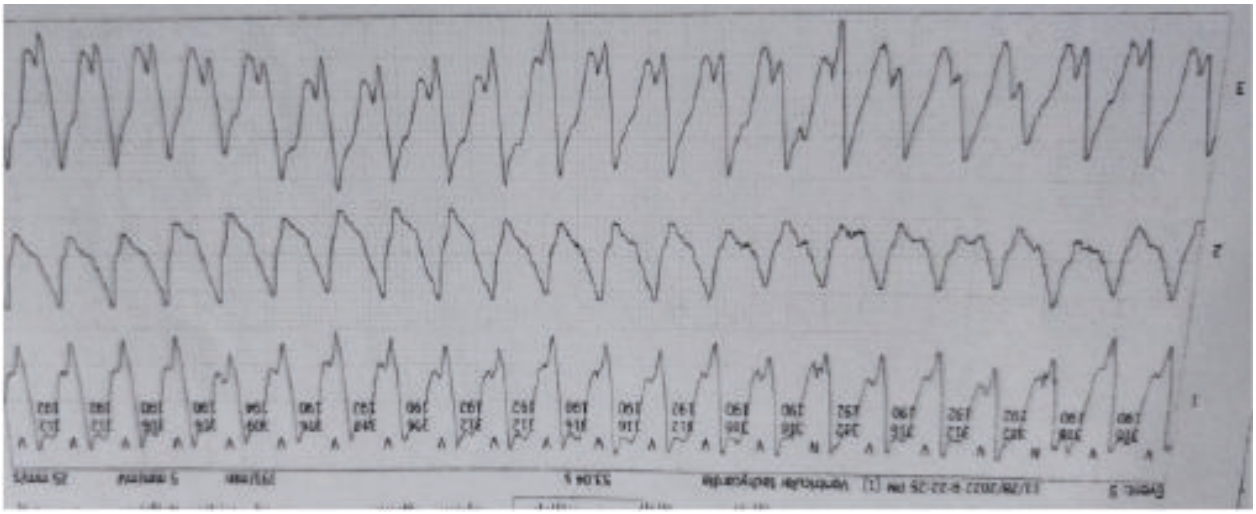


Fig.-7: *VT in a sick sinus child*

Laboratory work-up should include toxicology screen, serum electrolytes, complete blood count, viral panel, blood culture and cardiac enzymes.

Cardiac consultation and echocardiographic evaluation are done to rule underlying structural heart disease, cardiomyopathy, cardiac tumors.

Admission for observation: After cardioversion, return to sinus rhythm may be transient and continual infusion of amiodarone may be required.

Prognosis: In the setting of a structurally normal heart and stable, well-tolerated monomorphic VT, many minimally symptomatic patients can be followed closely without therapy. Such patients may benefit from B-blockers to reduce ectopy. Chronic treatment otherwise depends on rate, duration, symptoms, type of VT and the presence of genetic channelopathy such as long QT. Aggressive antiarrhythmic therapy usually in addition to implantable cardioverter-defribrillator (ICD) placement is required for life-threatening VT.

B) Long QT syndrome

The congenital long QT syndrome is a genetic disorder of prolonged cardiac repolarization that may cause cardiac arrest and sudden death. Abnormalities in cardiac ion channels predispose patients to a characteristic polymorphic VT called "torsades de pointes". Events are often precipitated by adrenergic stimuli. Acquired long QT may also occur and can be caused by drugs, underlying medical conditions or electrolyte imbalances.

Clinical features: Patients may present with presyncope, syncope, seizures, or cardiac arrest. Precipitating factors may include exercise, especially swimming, emotional stress, exposure to loud noises or even sleep. Although rare, infants can present with poor feeding, or with episodes of lethargy, cyanosis or poor perfusion.

Electrocardiogram findings: Sinus rhythm ECG, QTc of >460 in post-pubertal females and 450 in others, best obtained from lead II (Bazett Formula QTc= QT Interval/"-RR). Borderline QTc>440 ms in the setting of clinical symptoms and/or family history should be investigated. Abnormal T wave morphology including notching and low amplitude. *Torsade de pointes* seen during events.

Acute management: For torsades de pointes, emergent defibrillation followed by administration of magnesium sulfate and possibly lidocaine. Correction of underlying problem if acquired long QT. Intravenous beta-blockade may calm an adrenergic storm.

Further work-up and management in suspected congenital long QT syndrome: To obtain thorough family history of rhythm abnormalities, sudden death, deafness.

To ascertain all medications: To review history of event that may have triggered arrhythmia. To obtain electrolytes and treat underlying abnormalities. If presented with symptoms or documented VT, admision for observation, cardiology consultation and treatment. For patients presenting with non-cardiac issues and noted to have abnormal QTc interval, out-patient cardiology follow-up may be arranged. Restrict all strenuous activity pending cardiology follow-up. Provide list to the patients of medications known to prolong QT that should be avoided. Immediate family members should also be screened with 12-lead EKGs.

Prognosis: Prognosis is poor in untreated symptomatic patients, with an annual mortality of 20%. B-blockers are the mainstream therapy and reduce risk of sudden death to about 6% annually but do not eliminate it completely.[14] High risk patients may benefit from ICD placement which has been shown to reduce mortality risk. Factors known to increase risk include history of previous syncope, deafness, previous torsade, female gender and genotype.[13]

Congenital Heart Disease

Patients with congenital heart disease are at lifelong risk for the development of arrhythmias. Types of arrhythmias depend on the underlying cardiac anomaly and more importantly on the surgical repair and can range from atrial flutter to ventricular fibrillation (Table I).[10,12-18]

Table I

Congenital heart disease and associated arrhythmias

Congenital Heart Disease	Associated arrhythmia
Tetralogy of Fallot	Atrial tachycardia
Double outlet right ventricle	Ventricular tachycardia Sinus node dysfunction
Transposition of the great arteries	Ventricular arrhythmias Atrioventricular block
Ebstein's anomaly	Supraventricular tachycardia
Ventricular septal defect repair	Heart block Ventricular arrhythmias
Atrialseptal defect	Atrial tachycardia
Atrial septal defect repair	Sinus node dysfunction

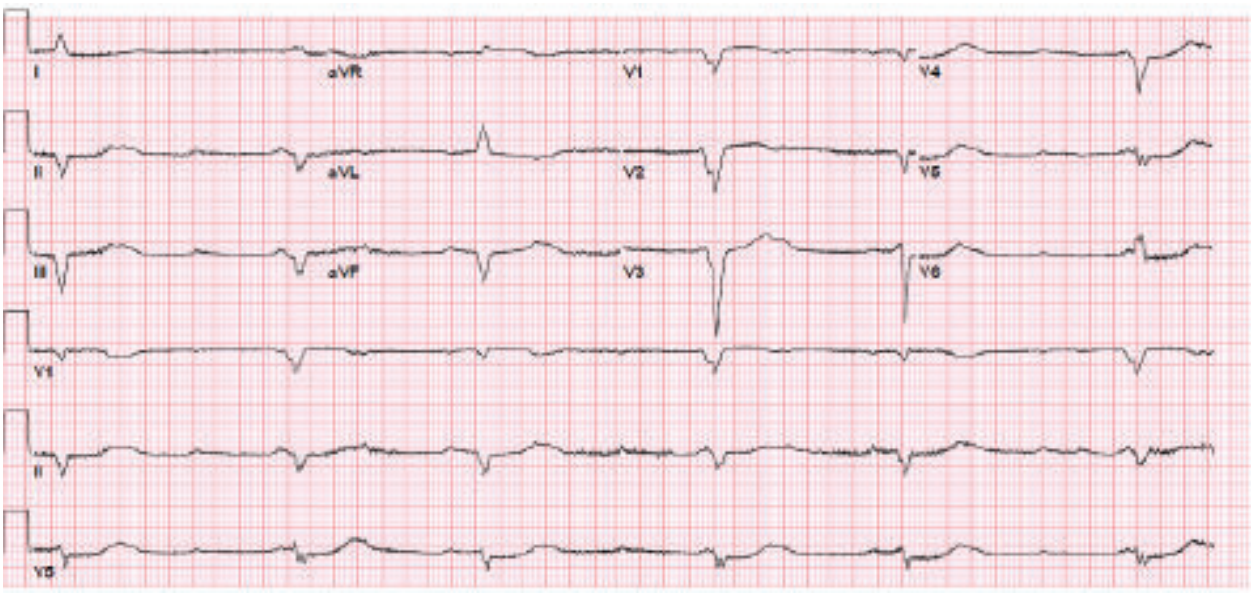


Fig.-8: *Atrioventricular block in an infant with myocarditis*

Bradycardias

Bradycardias are a group of disorders that include sinus node dysfunction and abnormal conduction through the AV node. AV conduction abnormalities include first, second or third degree heart block. First-degree heart block, prolongation of PR interval without loss of conduction, does not cause bradycardia or hemodynamic instability and is primarily of interest as a marker for an underlying cause. Causes of first-degree block are numerous and include enhanced vagal tone, previous cardiac surgeries, myopathies, and infections including Lyme disease, myocarditis, endocarditis, and hypothyroidism.

1. Sinus bradycardia and sinus node dysfunction

Sinus bradycardia can result from multiple disease states, most of which are not primary cardiac. The exception is sinus node dysfunction, an abnormality of impulse generation and propagation of the sinus node usually caused in pediatrics by direct injury or disruption of blood supply to the node from previous cardiac surgery. Associated atrial reentry tachycardias are common and when they occur, the term "brady-tachy syndrome" is applied. Etiology of sinus bradycardia other than congenital heart disease is diverse and includes increased intracranial pressure,

electrolyte abnormalities, respiratory compromise, hypothyroidism and certain medications. The need for acute treatment for sinus bradycardia itself is rare. The presence of sinus bradycardia in an otherwise healthy appearing child is generally of no concern, although anorexia nervosa should be considered. In the moribund patient with sinus bradycardia, urgent determination of the underlying cause is of paramount importance. Atropine and epinephrine increase sinus rates in most patients. Symptomatic patients with true sinus node dysfunction and/or those with coexistent tachycardias are likely to require elective pacemaker implantation.

Although primarily a disease of the elderly, Sick Sinus Syndrome (SSS) is an important clinical problem in pediatric patients, because treatment of associated exercise intolerance, presyncope or syncope, usually requires lifelong cardiac pacing. In children a previous history of cardiac surgery for congenital heart malformation is noted in approximately 80% of SSS cases.

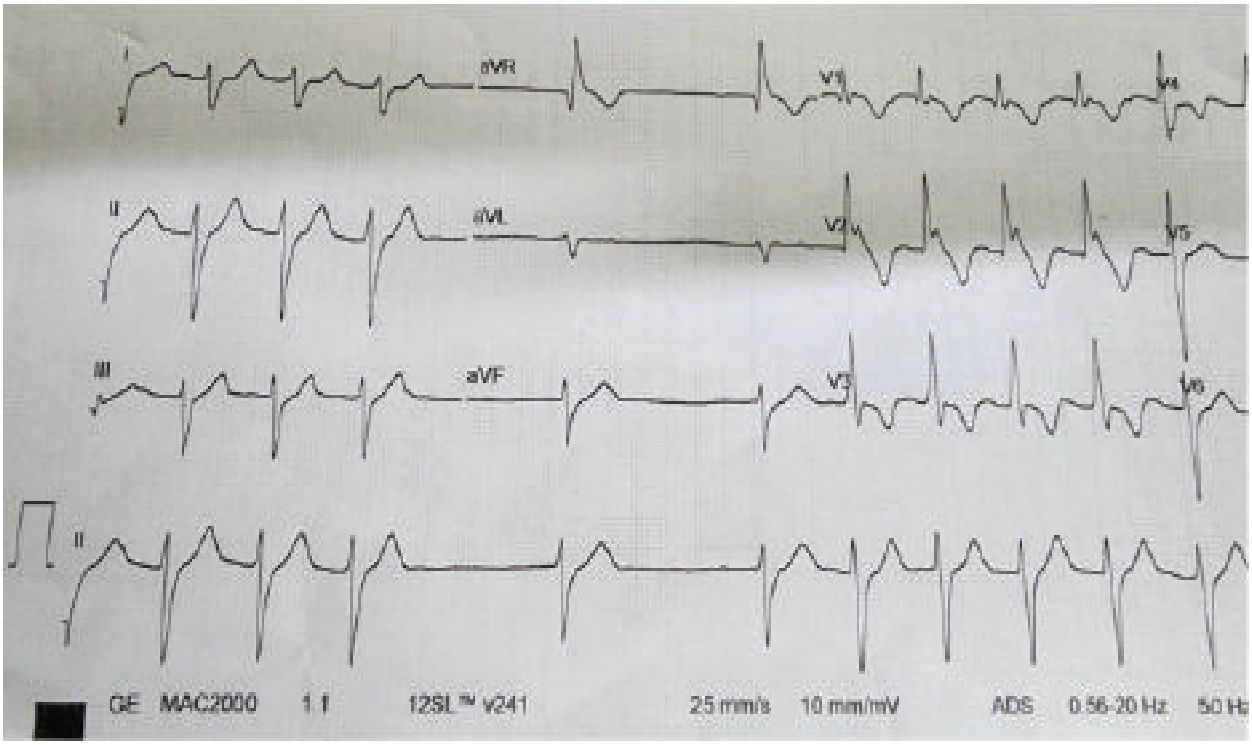


Fig.-9: *Sick sinus syndrome of long pause and absence of P-waves*

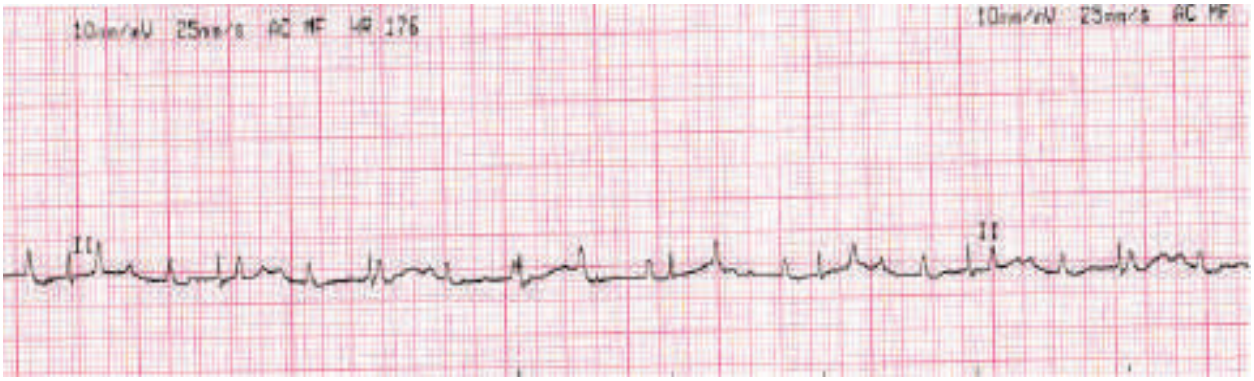


Fig.-10: *Second-degree heart block (Mobitz type II)*

2. Second-degree heart block

Second-degree heart block refers to intermittent failure of conduction through the AV node. It is further subdivided into Mobitz type I (Wenkebach), a gradual PR interval prolongation followed by a nonconducted beat, and Mobitz type II, an abrupt loss of conduction without previous change in PR interval duration (Fig.-10). Mobitz type I is generally more benign and may be a normal finding in adolescents during sleep. Other causes are similar to those described for first-degree heart block. Treatment is aimed at underlying reversible causes. Progression to higher degree block has been reported and symptomatic patients may benefit from treatment with atropine or isoproterenol. Mobitz type II is thought to be more likely to progress to complete heart block. Elective pacemaker implantation has been advised for symptomatic patients in this group.[13,14,19]

3. Third-degree AV block

Third degree AV block is the absence of conduction from the atria to the ventricle, manifested by AV dissociation. QRS duration of the escape rhythm may be normal or prolonged (Fig.-11). Third-degree block can be congenital, mainly associated with maternal collagen vascular disease or congenital heart disease, or acquired, due to AV node injury from cardiac surgery, viral myocarditis, Lyme disease, or metabolic or neuromuscular disorders. Many children are asymptomatic when diagnosed. In infancy, symptomatic patients may appear severely ill at presentation. Older children and adolescents may present with symptoms of exercise intolerance, fatigue, dizziness, syncope or signs of congestive heart failure. Sudden death may occur. Most patients eventually require pacemaker implantation, unless the cause is reversible such as in Lyme disease, where temporary pacing may suffice.

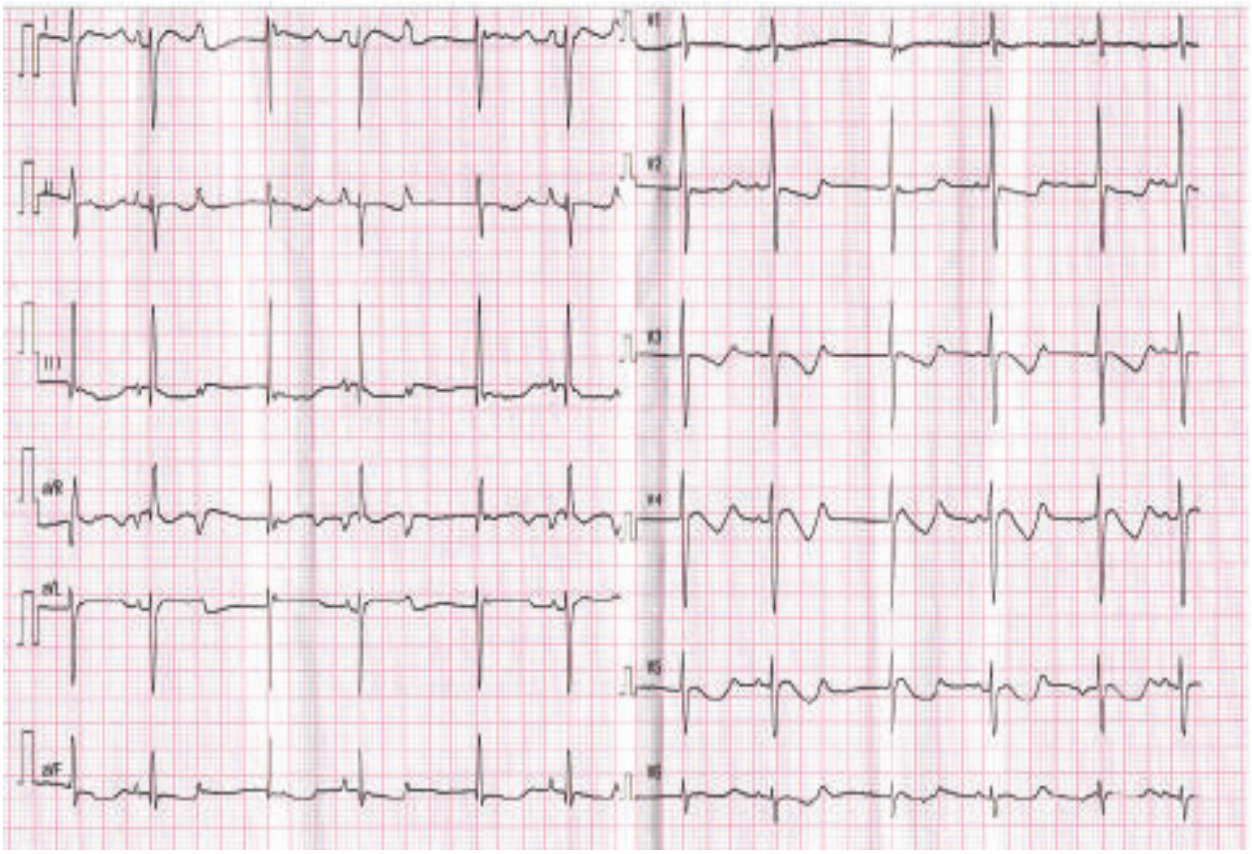


Fig.-11: *Complete heart block*

Conclusion

Proper diagnosis and management of the diverse rhythm disturbances in children is challenging. Emergency healthcare providers must be comfortable in performing a systematic evaluation and interpretation of ECG findings. An understanding of the variety of diseases that can predispose children with normal hearts or with structural heart disease to arrhythmias is essential for appropriate treatment.

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Case Report

Anomalous Origin of Left Coronary Artery from Pulmonary Artery (ALCAPA) in an Infant: A Case Report

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Abstract

Anomalous left coronary artery from pulmonary artery (ALCAPA) is a rare congenital heart disease that affects one in every 300000 live births and accounts for 0.24–0.46% of cases of congenital heart disease. In infancy, usually presents with congestive cardiac failure secondary to myocardial infarction or ischemia. It is associated with a mortality rate of 90% within the first year of life. Surgical correction to re-establish a two-coronary artery perfusion system is the treatment of choice, once patients are medically stable. Here we have reported a rare case of ALCAPA which was diagnosed by echocardiography & confirmed by selective coronary angiogram.

Keywords: *Anomalous Origin of left coronary artery, Pulmonary artery, echocardiography, coronary angiogram.*

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Introduction

Anomalous origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA) or Blade-White-Garland syndrome is a rare, but serious congenital cardiac malformation. ALCAPA represents one of the most common causes of myocardial ischaemia and infarction in children. If left untreated, the mortality rate is up to 90% within the first year of life. ALCAPA accounts for 0.25-0.50% of all congenital heart diseases.[1] Although the anomalous origin of the coronary arteries that arise from the pulmonary artery was first described in 1886.[2] It was not until 1933 when Bland *et al*[3] described the first clinical features with an autopsy finding of anomalous left coronary artery arise from the pulmonary artery (ALCAPA). The anomaly has thus been called the Bland-White-Garland Syndrome.[3,4] A coronary artery anomaly may involve an abnormal number, origin and/or course, termination or structure of the coronary arteries. [5]

The left main coronary artery normally arises from the left coronary sinus of valsalva. Instead of arising from the left aortic sinus, it may arises from the right sinus of valsalva or the proximal right coronary artery, from the non-coronary (posterior) aortic sinus, from the proximal part of the ascending aorta as well as from the pulmonary trunk or artery.[6] There are mainly two types of ALCAPA i.e. the infant type

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and the adult type, each of which has different manifestations and outcomes. Approximately 90% infant dies within the 1st year of life, due to myocardial infarction and congestive heart failure (CHF). Restoration of a dual-coronary-artery system is the ideal surgical treatment for ALCAPA syndrome.[7] Here we report a case that presented in early infancy with respiratory distress and diagnose as ALCAPA on echocardiography.

Case Report

A 8 months old male infant, 1st issue born to a non-consanguineous parents from Chittagong was admitted in Bangladesh Shishu Hospital & Institute with the complaints of cough & cold for 7 days & respiratory distress for 3 days. He had history of intermittent episodes of breathing difficulty associated with feeding difficulty and forehead sweating since 3 months of age. He had 2 episodes of respiratory tract infection. He had no history of cyanosis. With these complaints the baby was treated initially as a case of Viral Myocarditis & got IVIG, injectable antibiotics & anti failure medication at a peripheral medical college hospital. Color doppler Echocardiography was done by adult cardiologist & was diagnosed as a case of Dilated Cardiomyopathy (DCM) with severe LV systolic dysfunction. As the child was not improving satisfactorily he was referred to Bangladesh Shishu Hospital & Institute for further evaluation and better management.

On examination the baby was afebrile, acyanotic, Sp02 was 96% in room air, mildly pale, dyspneic (RR- 68/min), normotensive and having tachycardia (HR-180/min), all the peripheral pulses were palpable having no bracheo-femoral delay but pulse volume was low and capillary refilling time was 3 second. Precordium was normal in shape. There was cardiomegaly. There was no left parasternal heave, thrill or palpable P_2. S1 & S2 were audible in all area but S1 was soft. There was a pansystolic murmur best heard over apical area, grade 3/6 and radiating towards the left axilla. Breath sound was vesicular without any added sound. There was hepatomegaly. Our provisional diagnosis was Dilated cardiomegaly (DCM) with mitral regurgitation (MR) with Heart failure. Differentially we thought of viral myocarditis. Chest X-ray showed huge cardiomegaly (Fig.-2). There was pathological Q wave in lead-I and T-wave inversion in lead I & V1 - V6 on electro-cardiogram (Fig.-3).

Before operation After successful operation

Fig-1: *Ayat, a 8 months old infant (Before & after operation)*

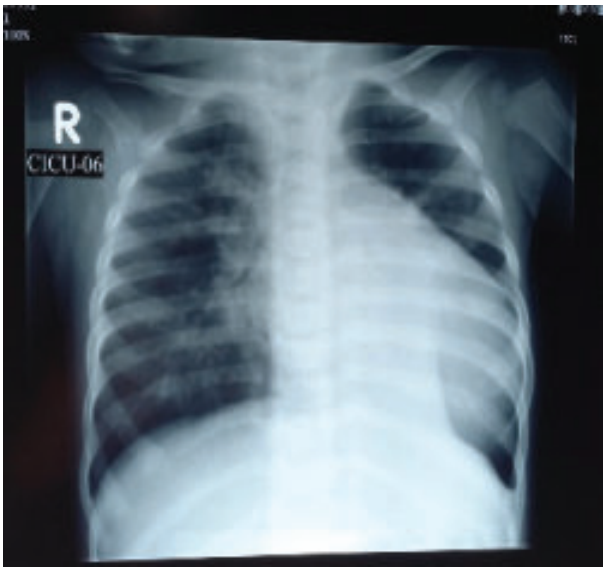

Fig-2: *Huge Cardiomegaly on Chest X-ray*

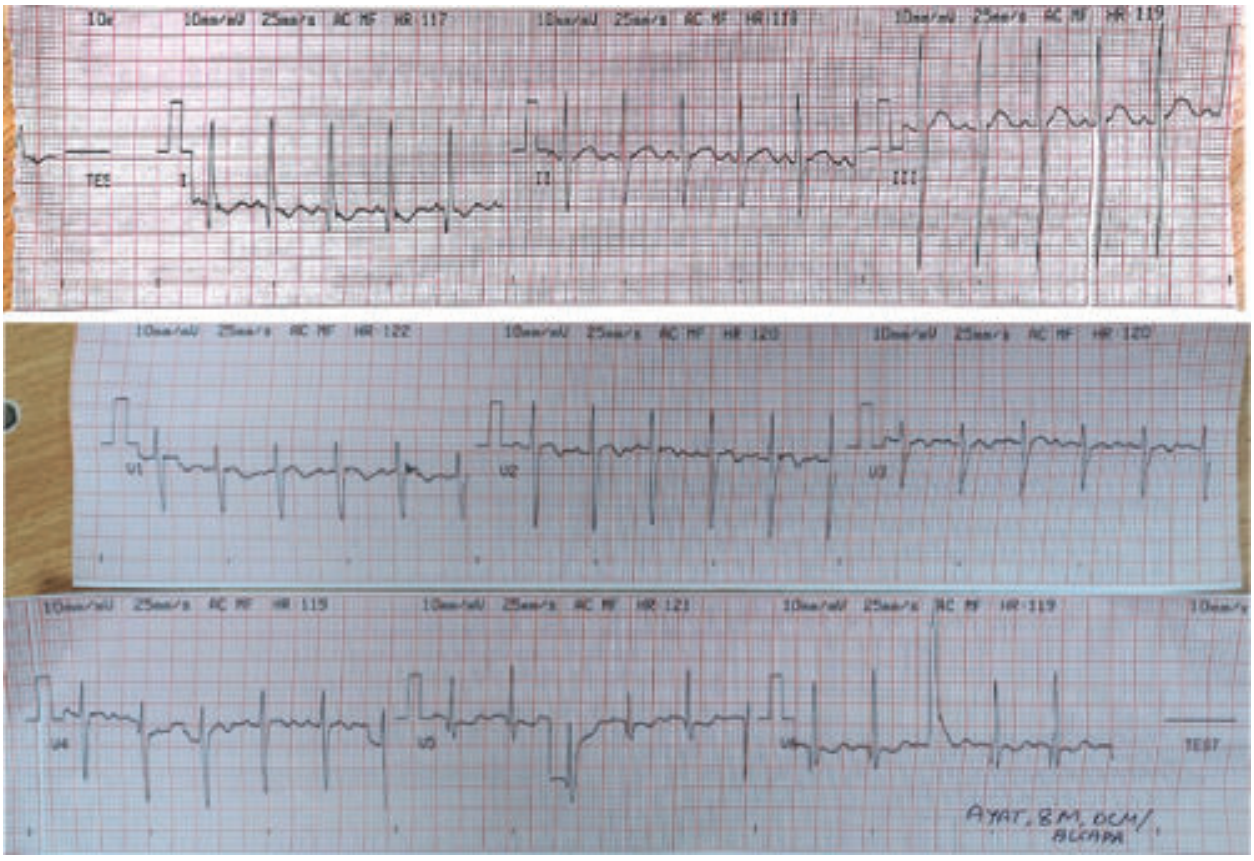

Fig-3: *ECG- Pathological Q wave in Lead I and T inversion in Lead I & V1-V6*

Color Doppler echocardiogram revealed that the left coronary artery arises from the left posterior sinus of pulmonary artery and diastolic flow seen in pulmonary artery from left coronary artery (coronary steal), moderate MR, hugely dilated LV with global hypokinesia and severe LV systolic dysfunction (Fig.-4). We confirmed the case by Selective Aortic Root

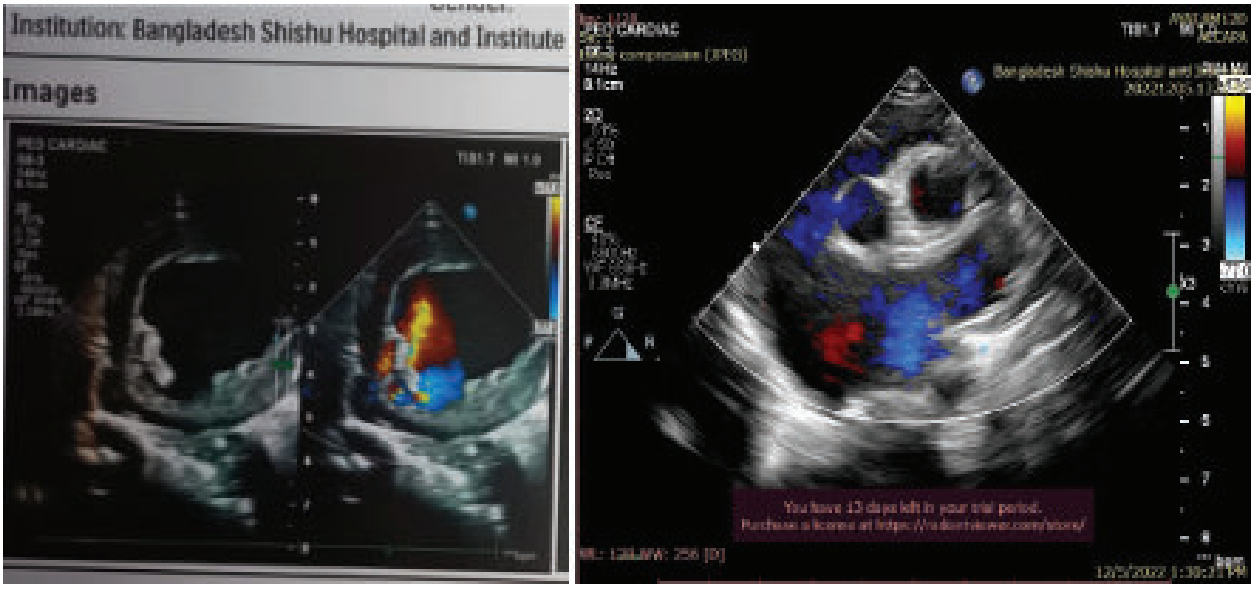

Fig-4: *Short axis view showing ALCAPA, Dilated LV with Echogenic chordae & papillary muscle*

Angiogram at Cath Lab which showed dominant right coronary originated from right coronary sinus & anastomatic collateral from RCA supplying left system which is connected to MPA (ALCAPA) (Fig.-5).

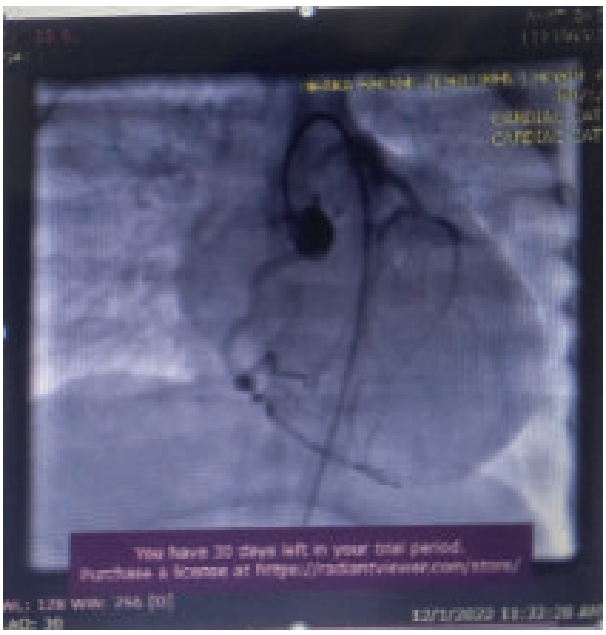

Fig-5: *Selective coronary angiogram showing ALCAPA with collateral from RCA to LCA*

The baby was treated with O_2 inhalation, Inj. Frusemide, Tab. Spironolactone, Tab. Enalapril and Tab. Digoxin. After having clinical improvement the baby was discharged & referred to an advanced cardiac center at India for immediate surgical correction and Left Coronary Artery translocation with Aortic re-implantation was done successfully.

Discussion

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital heart defect with an incidence of 1 in 300,00 live births and is the most common cause of myocardial infarction in children.[8] ALCAPA anomaly may result from abnormal separation of the cono-truncus into the aorta and pulmonary artery or from persistence of the pulmonary buds together with involution of the aortic buds that eventually form the coronary arteries.[9] There are four types of ALCA defined by the path the left coronary artery takes after arising from the right coronary sinus. The path of the first type is anterior to right ventricular outflow tract before reaching the anterior sulcus, the usual area of bifurcation. The second type courses behind the right ventricular outflow. The third one courses dorsal to the ascending aorta. These three types in absence of atherosclerotic plaque obstruction are benign. The fourth type arises from the right sinus of Valsalva and passes obliquely between the aorta and pulmonary trunk. This latter type is the only one predisposing to sudden death.[10]

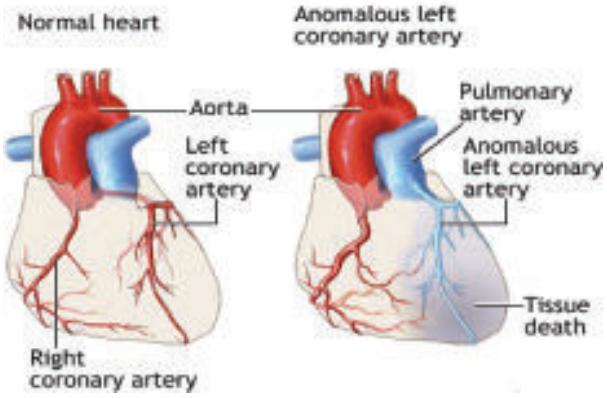

Fig-6: *Normal coronaries & ALCAPA*

ALCAPA usually manifests as an isolated defect, but in 5% of cases it may be associated with other cardiac anomalies such as Atrial septal defect, Ventricular septal defect, and Coarctation[11], it also may be associated with Tertralogy of Fallot (TOF), Transposition of great arteries(TGA) and Patent ductus arteriosus (PDA).[5]

In the neonatal period, the baby is asymptomatic as there is anterograde flow of de-saturated blood from the pulmonary artery to the left coronary artery. As pulmonary arterial pressure drops, the combination of low flow and de-saturated blood causes myocardial ischemia, especially during exertion. Collateral vessels develop between the right and left coronary arteries. Further decreases in pulmonary arterial pressure result in reversal of flow, as the left coronary artery drains from the right coronary artery, through collaterals, into the pulmonary artery. This is known as myocardial steal or coronary steal.[12,13]

In the infants, the chief symptom was irritability elicited by only slight effort, such as feeding, with signs of poor peripheral perfusion. These features started within 2 months of birth, coinciding with substantial reduction in pulmonary vascular resistance that resulted in coronary steal from the anterolateral aspect of the LV. The ECG signs of ischemia accompanied the deteriorating ventricular function. This progression created strong suspicion of ALCAPA.[14] Almost all of these features were present in our case.

The older children were asymptomatic and might have remained so until adolescence or young adulthood, presumably because of effective collateral blood supply to the LV from the RCA. Such patients can have angina upon exertion and are at risk of sudden death. Alternatively, they can present with signs of CHF caused by decompensation of already-borderline cardiac function, secondary to acute infectious illness. Their baseline ECG usually shows ischemic changes only in cases of decompensation or during a stress test.[14]

Clinical findings in late presenters (adults) with ALCAPA were investigated by Yau et al in a large review including 151 patients. They found a predominance of females 2:1, 66% presented with angina, dyspnea, palpitations or fatigue. A further 17% presented with ventricular arrhythmia, syncope or sudden death and 62% of these had no antecedent symptoms. The average age for sudden death in untreated adult ALCAPA was 35 years, and the recommendation for all newly diagnosed adult ALCAPA is therefore to undergo surgical treatment as early as possible.[15]

The diagnosis requires a high index of suspicion during history and physical examination.[9] Objective findings include Cardiomegaly on chest X-ray[14], this finding was perfectly present in our patient. In the study by Yau et al[15], the ECG demonstrated Q-waves in 50%, left ventricular hypertrophy in 28% and left axis deviation in 15%. ECG was normal in 4%. In contrast, symptomatic infants with isolated ALCAPA all had ECG abnormalities with ST-de-pres-sion, (pathological) Q-wave in V5-V6, and negative T-waves in V5.[16.] 2-Dechocardiography may identify ALCAPA and color doppler shows an abnormal jet in the pulmonary arteryand of retrograde bloodstream in the system of LCA. Another finding is abnormal dilatation of the proximal RCA, an abnormal "brightness" (echogenicity) of left ventricular papillary muscles and sharply delimited sectors of the left ventricular endocardial. Mitral valve regurgitation, left ventricular dysfunction, and wall motion abnormalities may be present.[17] Color doppler echocardiography of our patient done in our Echo lab which revealed anomalous origin of left coronary artery from the left posterior sinus of the pulmonary artery with diastolic flow in the pulmonary artery from the left coronary artery (coronary steal), Hugely dilated LV and global hypokinesia, mild to moderate MR and severe LV dysfunction (LVEF - 25.0%).Cardiac catheterization and angiography, the classical gold standard for confirming the diagnosis of ALCAPA.[18] Based on clinical suspicion supported by X-ray, ECG & Echocardiography we did selective coronary angiogram which confirmed the diagnosis of ALCAPA.

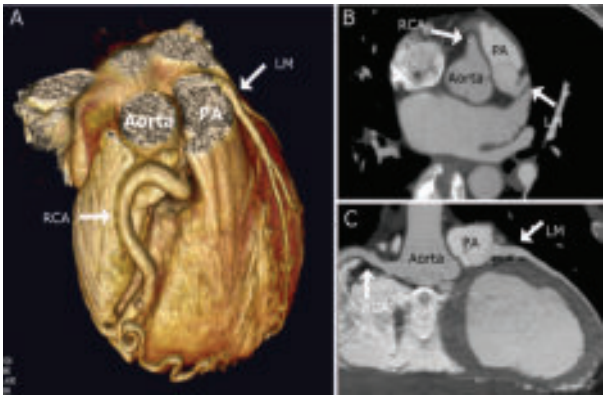

Fig-7: *CT angiogram of ALCAPA*

Myocardial perfusion studies are indicated selectively in patients with severe global hypokinesia where heart transplantation is a possibility. The current recommendation of management is to operate at any age when the diagnosis is made, due to the risk for ventricular arrhythmias and sudden death. Current surgical procedures are directed at establishing revascularization by creating a two-coronary artery system via either (i) Intra pulmonary Tunnel Operation i.e Takeuchi

procedure (creation of an aortopulmonary window and an intrapulmonary tunnel extending from the anomalous ostium to the window) (ii) Left coronary artery implantation, (iii) Subclavian artery to left coronary artery anastomosis, or (iv) Tashiro repair. By establishing a patent two-coronary artery system, most patients experience normalization of LV systolic function and improving long-term survival.[19,20] We referred the baby to advanced cardiac centre in India & Left Coronary Artery translocation with Aortic re-implantation is done successfully.

Conclusion

ALCAPA is a rare but fatal congenital heart malformation. A combination of a high index of suspicion, typical ECG and echocardiographic findings in a young infant presenting with LV dysfunction could lead to an earlier diagnosis of ALCAPA. Re-implantation of the anomalous LCA in the aortic root is the treatment of choice. Early diagnosis and surgical intervention at optimum time generally results in an excellent prognosis.

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