SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES AND TECHNOLOGY
THIRUVANANTHAPURAM

DEPARTMENT OF CARDIOLOGY

CLINICAL PROFILE AND OUTCOMES OF CARDIOMYOPATHY IN CHILDREN

A THESIS SUBMITTED FOR THE DEGREE OF DM CARDIOLOGY

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DEPARTMENT OF CARDIOLOGY,

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CERTIFICATE

I hereby certify that the work in this dissertation titled "**CLINICAL PROFILE AND OUTCOMES OF CARDIOMYOPATHY IN CHILDREN** " is a certified record of original research work undertaken by Dr. Gousia Mukhtar in the Department of Cardiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology in partial fulfillment of the requirement for the award of D.M. Cardiology degree.

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I, Dr.Gousia Mukhtar, hereby declare that the project in this book, titled "Clinical profile and outcomes of cardiomyopathy in children" was undertaken by me under the supervision of the faculty, Department of Cardiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology.

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ABBREVIATIONS

DCM	Dilated Cardiomyopathy
HCM	Hypertrophic cardiomyopathy
RCM	Restrictive Cardiomyopathy
ARVD	Arrhythmogenic right ventricular dysplasia
AVC	Arrythmogenic ventricular cardiomyopathy
ICD	Intra-cardiac defibrillator
CRT	cardiac resynchronization therapy
LVNC	Left ventricular non compaction
NC:C	Non compacted to compacted ratio
LAE	Left atrial enlargement
AV block	Atrioventricular block
HF	Heart failure
CTR	Cardiothoracic ratio
IVS	Interventricular septum
SCD	Sudden cardiac death
LGE	Late gadolinium enhancement
LAD	Left axis deviation
PVH	Pulmonary veinous hypertension
LVEF	Left ventricle ejection fraction
PCMR	Pediatric cardiomyopathy registry
NACCS	National Australian Childhood Cardiomyopathy Study

ARNI	Angiotensin Receptor Neprilysin Inhibitor
LVOTO	Left ventricular outflow tract obstruction
ECG	Electrocardiography
RBBB	Right bundle branch block
LBBB	Left bundle branch block
NSVT	Non sustained Ventricular tachycardia
LVH	Left ventricular hypertrophy
NYHA	New York heart association

INTRODUCTION

Cardiomyopathies are defined as abnormalities of the ventricular myocardium unexplained by abnormal loading conditions or congenital heart disease(1).As compared to other cardiac conditions, we must admit that it is the cardiomyopathy that still remains unconquered and continues to challenge the cardiology community. Nevertheless, since long, cardiomyopathy has held the fascination of cardiologists but still has one of the worst cardiology outcomes. Though, ample data exists on adult cardiomyopathy, research on pediatric cardiomyopathy especially from this part of the world is surprisingly scarce.

Pediatric cardiomyopathies often occur in the absence of comorbidities, such as atherosclerosis, hypertension, renal dysfunction, and diabetes mellitus; as a result, they offer insights into the primary pathogenesis of myocardial dysfunction(1) A better understanding of the spectrum and outcomes of childhood cardiomyopathy would facilitate patient care and permit evaluation of newer therapies. Pediatric cardiomyopathies are genetically heterogeneous with many different causative genes and multiple mutations in each gene. Genetic variants causing cardiomyopathy in children can also have systemic features affecting noncardiac organs.The RASopathies, including Noonan syndrome, are the most well-known syndromic causes of pediatric cardiomyopathy.(1) Inborn errors of metabolism (Carnitine cycle defects in DCM) and storage disorders (Pompe disease with HCM) are associated with childhood-onset cardiomyopathy. Diagnosing such disorders especially carnitine deficiency is absolutely essential as appropriate treatment can reverse the cardiomyopathy.

Population-based studies in the United States, Finland, and Australia have estimated the incidence of primary cardiomyopathies in children. The lowest estimate, from Finland, was 0.7 cases per 100 000 person-years but includes only idiopathic cardiomyopathy(2).The highest estimate, from

Australia, was 1.24 cases per 100000 person-years,but this report included only children up to 10 years of age, the period when cardiomyopathy is most commonly diagnosed. (3). The estimate from the United States was 1.1 cases per 100000 person-years, but the incidence was 8 times higher (8.3 cases per 100000 person-years) in children diagnosed at < 1 year of age(4). All these studies have then aimed to study the specific types of cardiomyopathies separately , determine their clinical profile and outcome and identify the patients at high risk of acute cardiac events and death so that appropriate measures can be taken at the right time.

In the National Australian Childhood Cardiomyopathy Study, Nugent et al analyzed all cases of primary cardiomyopathy in children younger than 10 years of age who presented between 1987 and 1996. Dilated cardiomyopathy made up 58.6 percent of cases, hypertrophic cardiomyopathy 25.5 percent, restrictive cardiomyopathy 2.5 percent, and left ventricular noncompaction 9.2 percent of cases. 184 subjects with dilated cardiomyopathy were then studied separately. Freedom from death or transplantation was 72% 1 year after presentation and 63% at 5 years in DCM patients. Risk factors for death or transplantation were age > 5 years at presentation, familial dilated cardiomyopathy and lower initial fractional shortening z score. In the same study, 80 subjects with hypertrophic cardiomyopathy were identified. Freedom from death or transplantation was 83% 5 years after presentation and 76% 10 years after presentation. Risk factors for death or transplantation included concentric left ventricular hypertrophy, age at presentation < 1 year, lower initial fractional shortening Z score, and increasing left ventricular posterior wall thickness relative to body surface area. At the latest follow-up, 54 of 65 surviving subjects had no symptoms, and 46 were receiving no regular medication(3).

There are almost no such studies done from this part of the world on pediatric cardiomyopathies and that itself explains the need of the current study.Though this is an institution based study,it may be close to a population based study as it is from a major referral centre of the region.

REVIEW OF LITERATURE

It was as early as 1949 that Evans described a distinct heart-disease having a definite clinical, cardiographic, and pathological pattern which he called “familial cardiomegaly”(5)

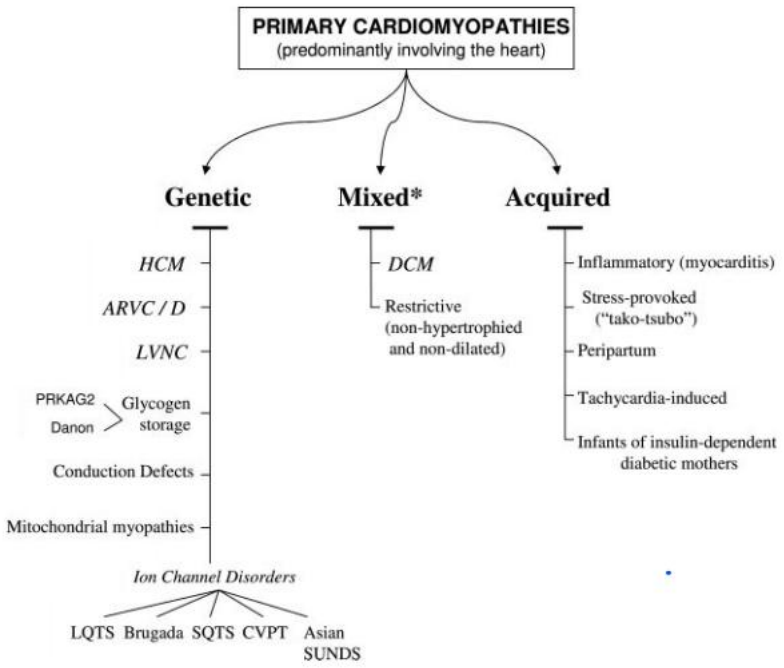
In 1951 ,Donald Teare reported “I have seen three cases of rhabdomyoma of the myocardium causing sudden death in young and healthy adults in as many months” . He collected more such cases and published his paper in the British Heart Journal in 1958 where he describes eight cases of ‘‘asymmetrical hypertrophy or muscular hamartoma’’ in young adults; seven of whom died suddenly.(6)

The term ‘cardiomyopathy ’ was first used in 1957 by Brigden, who described a group of uncommon, non-coronary myocardial diseases (7). In 1961 Goodwin defined cardiomyopathies as ‘‘myocardial diseases of unknown cause’’ (8). He described three different entities, namely ‘‘dilated, hypertrophic and restrictive’’, terms which are still in use today. With the advent of echocardiography, cardiomyopathies began to be diagnosed more frequently. In 1968, the WHO defined cardiomyopathies as “diseases of different and often unknown etiology in which the dominant feature is cardiomegaly and heart failure” (9) The first classification of cardiomyopathies was published in 1980, by the World Health Organization (WHO) and International Society and Federation of Cardiology (ISFC), and included the three subgroups proposed by Goodwin (10). The definition of ‘‘myocardial diseases of unknown cause’’ was maintained to define cardiomyopathies, which were distinguished from ‘‘specific heart muscle diseases’’, the latter comprising heart diseases with similar phenotypes, but due to an identifiable cause.

Representing a major advancement, both ‘‘arrhythmogenic right ventricular dysplasia’’ (with the inappropriate term ‘‘dysplasia’’ later changed to

‘‘cardiomyopathy’’) and a group of ‘‘unclassified cardiomyopathies’’, defined as ‘‘those that do not fit in any group’’, were added to the three original subgroups.(11)

In 2006, an expert committee of the American Heart Association proposed a new scheme in which the term “primary ” is used to describe diseases in which the heart is the sole or predominantly involved organ and “ secondary” to describe diseases in which myocardial dysfunction is part of a systemic disorder. Primary cardiomyopathies for the first time also included ‘‘ion channel diseases’’ and were differentiated in three subgroups based on their etiology as ‘‘genetic, mixed and acquired’’ (12).



AHA 2006 Classification of cardiomyopathies.The primary cardiomyopathies are further classified as genetic, mixed (genetic and nongenetic), acquired.(12)

The radical shift from a phenotypic to an etiological classification, as well as the inclusion of ion channel diseases among cardiomyopathies, although

proposed to guide future research rather than to be employed in the clinical arena, sparked a passionate transatlantic debate, culminating in a thorough reworking of the original 1995 classification by the European Society of Cardiology (ESC) Working Group on Myocardial and Pericardial diseases, in 2008.(13)

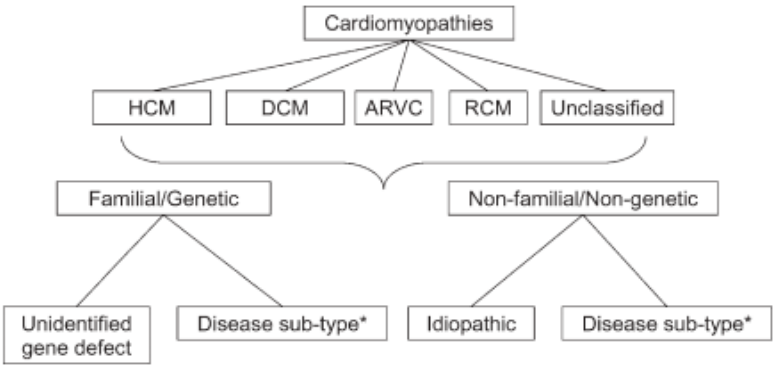
The ESC group stated that " the challenge of distinguishing primary and secondary disorders in this way is illustrated by the fact that many of the diseases classified as primary cardiomyopathies can be associated with major extra-cardiac manifestations; conversely, pathology in many of the diseases classed as secondary cardiomyopathies can predominantly (or exclusively) involve the heart" (13)

An alternative approach is to reclassify cardiomyopathies according to the causative genetic defect. Howeever, in clinical practice the pathway from diagnosis to treatment rarely begins with the identification of an underlying genetic mutation; more usually, patients present with symptoms or are incidentally found to have clinical signs or abnormal screening tests. Even when the genetic defect is known in a family, the identification of clinically relevant disease in gene-carriers still requires the demonstration of a morphological phenotype.(13)

They maintained that a clinically oriented classification system in which heart muscle disorders are grouped according to ventricular morphology and function remains the most useful method for diagnosing and managing patients and families with heart muscle diseases. ESC classification provided a simple operational framework for the medical community, which might have a direct impact in diagnosing and managing these complex diseases(13).

They defined a cardiomyopathy as: A myocardial disorder in which the heart muscle is structurally and functionally abnormal, in the absence of coronary artery disease, hypertension, valvular disease and congenital heart disease sufficient to cause the observed myocardial abnormality.(13)

Each of the time-honoured categories dilated, hypertrophic, restrictive and arrhythmogenic right ventricular were maintained, divided into familial and non-familial to replace the pre-genetic era concept of ‘‘unknown etiology’’. Familial refers to the occurrence, in more than one family member, of either the same disorder or a phenotype that is (or could be) caused by the same genetic mutation.The genetic defect can be well known or unidentified. Non-familial cardiomyopathies are clinically defined by the presence of a cardiomyopathy in the index patient and the absence of disease in other family members. They are subdivided into idiopathic (no identifiable cause) and acquired cardiomyopathies (Disease subtype) in which ventricular dysfunction is a complication of a known disorder rather than an intrinsic feature of the disease.



ESC 2008 Classification of cardiomyopathies into specific morphological and functional phenotypes which are then subdivided into familial and non familial forms.

Though considerable research has been done on adult cardiomyopathy, research on pediatric cardiomyopathies especially from this part of the world still remains limited

The earliest study on pediatric cardiomyopathy i.e.. Baltimore-Washington Infant Study (BWIS) conducted in 1992 by Ferencz et al reported a prevalence of 1 in 10,000.(14) They studied fifty-six infants who had cardiomyopathy in infancy, in the absence of a structural defect. The study included conditions like tumors, endocardial fibroelastosis, mucocutaneous lymph node syndrome, and infarction as well among the cardiomyopathies. Of particular interest is the documentation in this study of some overlap between the etiologies of Cardiomyopathies and structural cardiovascular malformations. This overlap suggests that embryonic myocardial disease might sometimes be responsible for altered cardiac structures, possibly secondary to hemodynamic changes.

Arola et al conducted a retrospective study in Finland in 1980-1991 to obtain information on the epidemiology of childhood cardiomyopathies.(2) 118 patients less than 20 yrs age were definitely identified as having idiopathic cardiomyopathy. Dilated cardiomyopathy was the most common form of cardiomyopathy, diagnosed in 52 percent, 12 of whom had endocardial fibroelastosis.37 percent had hypertrophic cardiomyopathy. Restrictive cardiomyopathy was identified in six patients and arrhythmogenic right ventricular dysplasia in three patients. Notably the proportion of infants with hypertrophic cardiomyopathy was significantly less compared to other cardiomyopathies.

In their study, ‘’The Epidemiology of Childhood Cardiomyopathy in Australia’’ published in the NEJM in 2003 , Nugent et al(3) analyzed all cases of primary cardiomyopathy in children who presented between 1987

and 1996 and who were younger than 10 years of age. 314 new cases of primary cardiomyopathy were identified, for an annual incidence of 1.24 per 100,000 children younger than 10 years of age. Dilated cardiomyopathy made up 58.6 percent of cases, hypertrophic cardiomyopathy 25.5 percent, restrictive cardiomyopathy 2.5 percent, and left ventricular noncompaction 9.2 percent of cases.The incidence of all types of cardiomyopathy except restrictive declined rapidly after infancy. Indigenous children had a higher incidence of dilated cardiomyopathy and a higher rate of death as the presenting symptom than nonindigenous children. Lymphocytic myocarditis was present in 40 percent. Authors concluded that Lymphocytic myocarditis and left ventricular noncompaction were important causes of childhood cardiomyopathy in Australia.

The Pediatric Cardiomyopathy Registry (PCMR) was established to describe the epidemiologic features and clinical course of selected cardiomyopathies in patients aged 18 years or younger and to promote the development of etiology-specific treatments. The original PCMR design consisted of 2 cohorts. The first was a retrospective cohort of children who were diagnosed between 1990, and 1995, and identified by chart review from tertiary care centers in the United States and Canada. The purpose of this cohort was to identify potential predictors of outcome as well as diagnostic approaches. The second cohort was a population-based prospective cohort of children diagnosed after 1996 at pediatric cardiac centers. The purpose of this cohort was to estimate accurately the incidence of cardiomyopathy in children.(15)

Lipshultz published their study “ The Incidence of Pediatric Cardiomyopathy in Two Regions of the United States” in NEJM in 2003 (16) which is based on the prospective component of the PCMR data base, consisting of patients who have received a diagnosis of cardiomyopathy

from 1996 to 1999. They identified 467 cases of cardiomyopathy, for an overall annual incidence of 1.13 per 100,000 children. The incidence was similar to that reported for Finland and Australia. They Showed that Dilated cardiomyopathy made up 51 percent of the cases, hypertrophic cardiomyopathy 42 percent, and restrictive or other types 3 percent; 4 percent were unspecified. The incidence was significantly higher among infants younger than 1 year old than among children and adolescents who were 1 to 18 years old.

The findings of PCMR were published in various studies.Cox et al studied the factors associated with establishing a causal diagnosis in cardiomyopathies.(17).Only one third had a know cause. For dilated cardiomyopathy, a known cause was associated with older age, lower heart rate, smaller left ventricular dimensions, and greater shortening fraction and family history of cardiomyopathy and family histories of genetic syndromes and sudden death. For hypertrophic cardiomyopathy, only blood and urine testing was associated with a causal diagnosis.

Nugent et al also reported on 80 children with Hypertrophic cardiomyopathy from the National Australian Childhood Cardiomyopathy Study in their study titled “Clinical Features and Outcomes of Childhood Hypertrophic Cardiomyopathy” published in Circulation.(18). An underlying syndromal, genetic, or metabolic condition was identified in 46 subjects (57.5%). Left ventricular outflow tract obstruction was present in 32 subjects (40%); right ventricular outflow obstruction was present in 10 (12.5%). Freedom from death or transplantation was 83% 5 years after presentation and 76% 10 years after presentation. Risk factors for death or transplantation included concentric left ventricular hypertrophy, age at presentation less than 1 year, lower initial fractional shortening Z score, and

increasing left ventricular posterior wall thickness relative to body surface area.

Colan and Lipshultz et al reported on the PCMR data on HCM in the article "Epidemiology and Cause-Specific Outcome of Hypertrophic Cardiomyopathy in Children" published in Circulation in 2007(19). Of 855 patients, 8.7% had inborn errors of metabolism, 9.0% had malformation syndromes, 7.5% had neuromuscular disorders, and 74.2% had idiopathic HCM. Children with HCM associated with inborn errors of metabolism and malformation syndromes have significantly worse survival than the other 2 groups. Patients with idiopathic HCM had a 10-year survival from the time of diagnosis of 85.3%. Patients with idiopathic HCM diagnosed before 1 year of age had worse survival from the time of diagnosis than those diagnosed after 1 year of age. Overall, patients with idiopathic HCM who are older than 1 year of age, regardless of age at diagnosis, have an annual mortality of 1%, a rate that is much lower than previously reported in children and is not different from that found in population-based studies in adult.

Maron et al assembled a multicenter international registry of ICDs implanted (1987 to 2011) in 224 unrelated children and adolescents with HCM judged at high risk for sudden death(20). Patients received ICDs for primary (n=188) or secondary (n=36) prevention after undergoing evaluation at 22 referral and nonreferral institutions in the United States, Canada, Europe, and Australia. They concluded that in a high-risk pediatric HCM cohort, ICD interventions terminating life-threatening ventricular tachyarrhythmias were frequent. Extreme left ventricular hypertrophy was most frequently associated with appropriate interventions. The rate of device complications adds a measure of complexity to ICD decisions in this age group.

Yetman et al published a study “Long-term Outcome and Prognostic Determinants in Children With Hypertrophic Cardiomyopathy” in JACC in 1998. (21). For the 99 HCM patients less than 18 years age, Median age at diagnosis was 5.0 yr with a median follow-up interval of 4.8 years. Death or resuscitated sudden death occurred in 18 patients. Sudden death rate was 2.7%/yr after age 8 yr. Increased corrected QT interval (QTc) dispersion on ECG, ventricular tachycardia (VT) on ambulatory ECG and myocardial bridging of the LAD coronary to be associated with reduced time to death or resuscitated sudden death.

Maruizi et al assessed patients with pediatric-onset hypertrophic cardiomyopathy diagnosed from 1974 to 2016 in 2 national referral centers for cardiomyopathies in Italy.(22). They published their results in the study “Long-term Outcomes of Pediatric-Onset Hypertrophic Cardiomyopathy and Age-Specific Risk Factors for Lethal Arrhythmic Events” published in JAMA in 2018. Patients with metabolic and syndromic disease were excluded. Of 1644 patients with HCM, 100 (6.1%) were 1 to 16 years old at diagnosis. 42.0% were symptomatic. During a median of 9.2 years, 24 of 100 patients (24.0%) experienced cardiac events, including 19 LAEs and 5 heart failure–related events (3 deaths and 2 heart transplants). Risk of LAE was associated with symptoms at onset and Troponin I or Troponin T gene mutations. Adult HCM risk predictors performed poorly in this population. 79.0% of the patients in this study who experienced an LAE would not have been considered high risk under adult recommendations. Annual mortality at 5 years was 5%.

Saeidi et al analysed 40 pediatric patients with HCM and published their study in Pediatric cardiology in 2017(23). In their study named “Delayed Myocardial Enhancement in Pediatric Hypertrophic Cardiomyopathy: Correlation with LV Function, Echocardiography, and Demographic

Parameters'', they have aimed to detect the presence of fibrosis by Cardiac magnetic resonance imaging (CMR) in the pediatric age group and correlate CMR findings with demographic data, LV function, and other echocardiographic parameters. The mean percentage of DE-MRI was 9.7 ± 9%. They found a significant correlation between the percentage of DE-MRI in children with HCM and the pressure gradient across LVOT, NYHA classification, and LV myocardial mass.

Initial studies on pediatric restrictive cardiomyopathy were limited to case reports and case series.

Denfield et al reported on twelve cases of restrictive cardiomyopathy by database review of patient records from 1967 to 1994 in his study "Restrictive Cardiomyopathies in Childhood - Etiologies and Natural History". (24). Four of the 12 patients had embolic events (1, recurrent pulmonary emboli; 1, saddle femoral embolus; 2, cerebrovascular accidents) and 9 of 12 died within 6.3 years despite medical therapies. The authors concluded that restrictive cardiomyopathy in childhood is commonly idiopathic, and the prognosis is poor. Embolic events occurred in 33% of our patients, and 9 of 12 patients died within 6.3 years.

Russo et al reported on the natural history of idiopathic restrictive cardiomyopathy in a paediatric population and aimed to identify any factors predictive of outcome.(25). 21 patients were identified. Probability of survival at 1, 5, and 10 years was 80.5%, 39% and 20% respectively. Median age of presentation was 3.8 years. Median survival without transplantation was 2.2 years. Right and left ventricular end diastolic pressures and ratio of left atrial to aortic root dimensions (LA:Ao) at presentation had a significantly negative correlation with survival time after diagnosis.

Weber et al analyzed outcomes of childhood RCM, with a focus on the impact of phenotype comparing pure RCM with cases that have additional features of hypertrophic cardiomyopathy (HCM) (26)..This analysis from the Pediatric Cardiomyopathy Registry identified 152 cases of restrictive cardiomyopathy among 3375 children with cardiomyopathy (4.5%). Among the 152 patients, 101 were considered to have pure disease (3.0% of total) and 51 to have mixed RCM/HCM phenotype (1.5% of total and 34% of the RCM cases). Freedom from death was comparable between groups with 1-, 2-, and 5-year survival of RCM 82%, 80%, and 68% versus RCM/HCM 77%, 74%, and 68%. Independent risk factors at diagnosis for lower transplant-free survival were heart failure, lower fractional shortening z score and higher posterior wall thickness in the RCM/HCM group. Overall, outcomes were worse than for all other forms of cardiomyopathy

Walsh et al published a retrospective study of pediatric patients with restrictive cardiomyopathy diagnosed between April 1994 and May 2011 named “Conduction Abnormalities in Pediatric Patients With Restrictive Cardiomyopathy”(27) The mechanisms of serious arrhythmic events (death or episode of acute hemodynamic compromise thought to be secondary to arrhythmia) were evaluated. Sixteen patients (1–17 years of age) were reviewed, with 5 sudden cardiac events noted, including 4 deaths. The median PR interval (222 versus 144 ms; P=0.01) and the QRS duration (111 versus 74; P=0.01) were significantly longer in those who had an acute cardiac event. Older age at presentation was associated with sudden cardiac events (P=0.01).The authors concluded that pediatric RCM patients are at risk for acute high-grade heart block, and bradycardic events represented a significant portion of all arrhythmic events. Aggressive ECG monitoring strategies looking for conduction system disease should be the rule in all

patients with restrictive cardiomyopathy. Implantation of a defibrillator/pacemaker should be considered as prophylactic management.

Rivenes et al in their study “Sudden Death and Cardiovascular Collapse in Children With Restrictive Cardiomyopathy” reviewed eighteen consecutive patients during a 31-year period to determine the clinical outcome and cause of death(28). Those who sustained sudden, unanticipated cardiac arrests were evaluated for risk factors that are predictive of sudden death. Patients who were at risk for sudden death were girls with chest pain, syncope, or both at presentation and without congestive heart failure. Histopathological evidence of acute or chronic ischemia was found in the majority of patients, with acute ischemia more common among those who sustained sudden death events.

Arola et al reported on clinical profile and course of 62 Finnish children and adolescents (median age, 13 months; range, 1 day to 20 years) with IDCM in 1980 to 1991 (29). During a mean (6SD) follow-up of 3.9 to 4.5 years (range, 1 day to 25 years), 10 patients (16%) recovered, 17 (27%) had residual disease, 4 (6.4%) underwent heart transplantation, and 31 (50%) died. Infants(less than 1 years of age) and adolescent (>15 years of age) male patients with progressing symptoms of left ventricular failure after initiation of medical therapy tended to have the poorest outcome. However, in multivariate analysis, only histologic evidence of endocardial fibroelastosis, clinical signs of right ventricular failure at presentation, and the need for anticoagulative therapy during follow-up, the last an expression of a severely impaired left ventricular systolic function, appeared to be significant predictors of long-term outcome.

Daubeney et al examined the clinical characteristics and risk factors for death and transplantation among children with dilated cardiomyopathy

enrolled in the National Australian Childhood Cardiomyopathy Study(NACCS) (30). There were 184 subjects with dilated cardiomyopathy. Freedom from death or transplantation was 72% (95% CI, 65% to 78%) 1 year after presentation and 63% (95% CI, 55% to 70%) at 5 years. Risk factors for death or transplantation comprised age > 5 years at presentation, familial dilated cardiomyopathy, lower initial fractional shortening z score and failure to increase fractional shortening z score during follow-up. At follow-up, 78 (44.6%) of 175 cases diagnosed during life had no symptoms and were not taking any cardiac medication.

Alexander et al examined the long-term outcomes for children with dilated cardiomyopathy enrolled in the NACCS registry (31). The National Australian Childhood Cardiomyopathy Study is unique in representing the longest and most complete longitudinal national cohort study of childhood dilated cardiomyopathy, with a median follow-up among surviving subjects of 15 years. Survival free from death or transplantation was 74% 1 year after diagnosis, 62% at 10 years, and 56% at 20 years. In multivariable analysis, age at diagnosis less than 4 weeks or more than 5 years, familial cardiomyopathy, and lower baseline left ventricular fractional shortening Z score were associated with increased risk of death or transplantation, as was lower left ventricular fractional shortening Z score during follow-up. At 15 years after diagnosis, echocardiographic normalization had occurred in 69% of surviving study subjects. Normalization was related to higher baseline left ventricular fractional shortening Z score, higher left ventricular fractional shortening Z score during follow-up, and greater improvement in left ventricular fractional shortening Z score. The highest-risk period for children with DCM was in the first year after diagnosis, with 26% of patients achieving the end point of death or transplantation compared with ~1% per year in subsequent years.

Few studies have been conducted on pediatric ARVC since it is a rare disease.

Deshpande et al in their study "Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVC/D): Review of 16 Pediatric Cases and a Proposal of Modified Pediatric Criteria" reviewed pathology records of pediatric patients from a large volume, academic, tertiary care center between years 2006 and 2015 to identify patients demonstrating histopathology suggestive of ARVC/D. Ultimately, 16 cases of microscopically diagnosed ARVC/D were identified. All pertinent clinical information was reviewed, including demographics, presentation, disease course, and pathology, as well as various diagnostic tests including echocardiography, magnetic resonance imaging (MRI), and genetic testing. Many pediatric patients with confirmed histopathology would not have met the current ARVC/D diagnostic criteria, resulting in delays in diagnosis and treatment. The current criteria need further revision to encompass pediatric manifestations of ARVC/D (32)

Ricardo et al reviewed 36 children with LVNC from January 1997 to December 2002(33). Five children had associated cardiac lesions. The median age at presentation was 90 days. The median duration of follow-up was 3.2 years. Twenty-seven patients (75%) had ECG abnormalities, most commonly biventricular hypertrophy (10 patients, 28%). Both ventricles were involved in 8 patients (22%). Left ventricular systolic function was depressed in 30 patients (83%). Nine patients presenting in the first year of life with depressed left ventricular contractility had a transient recovery of function; however, ejection fraction deteriorated later in life, at a median interval of 6.3years. Two patients had an "undulating" phenotype from dilated to hypertrophic cardiomyopathy. Five patients (14%) died during the study. They concluded that LVNC does not have an invariably fatal course

when diagnosed in the neonatal period. A significant number of patients have transient recovery of function followed by later deterioration, which may account for many patients presenting as adults, some manifesting an "undulating" phenotype.

Zuckerman et al identified LVNC in 50 patients, 34 of them < 1 year of age. Death or transplantation occurred in 26 patients, with a median survival of 1.17 years after presentation. Patients surviving 1 year after presentation had 75% conditional survival, and patients surviving 2 years after presentation had 92% conditional survival. Hemodynamic instability, poor ventricular function, and LV dilatation were each independent predictors of poor outcome. Of the 21 patients who presented with hemodynamic instability, 17 died or underwent transplantation at a median of 0.08 years after presentation. They concluded that LVNC is recognized more in younger patients; however age was not a predictor of outcome. Patients who present with hemodynamic instability and poor ventricular function have decreased transplant-free survival, and most poor outcomes occur within the first year after presentation(34)

Isolated right ventricular non compaction have been rarely described in literature in the form of case reports.Sert et al described it in a baby girl who presented with murmur at third day of life.(35) Echocardiographic study showed prominent trabecular meshwork with deep intertrabecular recesses in the RV apex. Color flow Doppler examination confirmed the presence of blood flow within the trabeculae. The systolic and diastolic functions of the left ventricle (LV) were normal with no evidence of noncompaction cardiomyopathy.

Burke et al reported pathological findings in 14 cases of LVNC .(36) Other cardiac anomalies were present in 8 cases (nonisolated LVNC) and noted

that RV involvement (biventricular noncompaction) is seen in 6 cases as defined by histological criteria of greater than 75% transmural thickness of the noncompacted right ventricle. Right ventricular involvement was present in 50% (4/8) of LVNC with associated anomalies versus 33% (2/6) of the isolated forms.

Yun et al. reported noncompaction cardiomyopathy in the RV in 4 of 11 cases (1 patient with RVNC and 3 patients with bilateral involvement). According to Yun et al, dilation of the RV may be a helpful supportive feature for the diagnosis of RVNC(37).

Coming to Indian studies , Data on pediatric cardiomyopathies especially hypertrophic and restrictive cardiomyopathies is limited to case series.

VK Gandhi et al reported on 20 children with cardiomyopathy , seen between January 1986 to December 1987 at pediatric and cardiology departments of V.S. General Hospital, Ahemedabad.(38) Among the 20 children , 14 had dilated cardiomyopathy, 3 had hypertrophic and 3 had restrictive cardiomyopathy. Cardiomyopathies constituted 2.1% of total cardiac cases seen under the age of 16 years over a period of two years.The prevalence was higher than reported by Menon et al from delhi.

Menon et al from the AlIMS, New Delhi reported on 69 children with cardiomyopathies upto the age of 12 years from 1973 to 1982.(39). The 69 patients constitute less than one per cent of children with heart disease seen in the clinic during this period. Of the 69 children, 64 were of the DCM. Restrictive CM was present in two cases and hypertrophic CM in three patients.

AIMS AND OBJECTIVES

- To describe the clinical profile of cardiomyopathy in children less than 18 years of age.
- To study the outcome of cardiomyopathy in children less than 18 years of age when compared to historical cohorts.

HYPOTHESIS

- The outcome of pediatric cardiomyopathy in our population is similar to historical cohorts from other populations.

MATERIALS AND METHODS

STUDY DESIGN

- It is a single centre observational study with retrospective case enrollment and cross sectional follow up.

INCLUSION CRITERIA

- All patients with cardiomyopathy who presented to us during the period from 1990 to 2020 and younger than 18 yrs of age were enrolled in the study.
- After taking an informed consent , The available medical records of each enrolled patient was reviewed.This retrospective cohort of children was prospectively followed up to study their outcome.
- The patients were eligible for inclusion if specific quantitative echocardiographic criteria were met or if the pattern of cardiomyopathy confirmed to a defined semiquantitative pattern.
- Each patient was assigned to a diagnostic category according to the phenotypic characteristics following the European Society of Cardiology Classification, after directly reviewing all available cardiac information(13).
- Cardiomyopathies were defined as abnormalities of the ventricular myocardium unexplained by abnormal loading conditions or congenital heart disease.(1)
- Dilated cardiomyopathy was defined as LV end-diastolic dimension >2 standard deviations above normal for body surface area, in conjunction with depressed systolic function (fractional shortening / ejection fraction less than 2 standard deviations below normal for age).(40)
- Hypertrophic cardiomyopathy was defined by otherwise unexplained septal hypertrophy, left ventricular free-wall hypertrophy, or both (wall thickness more than 2 SD above the normal mean for BSA.(3)
- Restrictive cardiomyopathy was defined as "normal or decreased volume of both ventricles associated with biatrial enlargement, normal left ventricular wall thickness and atrioventricular valves, impaired ventricular filling with restrictive physiology, and normal (or near normal) systolic function”(26).
- Left ventricular noncompaction was defined by

1. Presence of prominent trabeculations in the left ventricle

2. Presence of deep recesses between the trabeculations and also presence of a thin compacted layer

3. Noncompacted: compacted ratio >2 in end systole on echocardiography.(40)

- ARVC is defined by the presence of right ventricular dysfunction (global or regional), with or without left ventricular disease, in the presence of histological evidence for the disease and/or electrocardiographic abnormalities in accordance with published criteria.(13)

- Need for ICD

  Was defined as based on one or more of these major risk factors: (41)

  - Family history of HCM SCD,
  - NSVT on ambulatory monitor,
  - Massive LVH
  - Unexplained syncope.

- Need for cardiac transplant was defined as (42)

  - Refractory heart failure
  - Progressive pulmonary hypertension
  - Deterioration of Functional capacity despite maximal medical treatment.
  - Refractory ventricular arrhythmias

- Need for CRT was defined as those who meet current recommendations for the implantation of a CRT as in adults with NYHA class II to ambulatory class IV HF, LVEF < 35%, and widened QRS.

- Normalization of LV Function was defined as a return to normal LV size (an LVEDD z-score less than 2) and normal LV systolic function (an LVFS or LV ejection fraction z-score less than 2) on any subsequent echocardiogram.(43)

EXCLUSION CRITERIA

- Congenital heart defects unassociated with malformation syndromes
- Uremia, acute or chronic
- Abnormal ventricular size or function ascribed to intense physical training
- Those who have undergone invasive cardiothoracic procedures or major surgery except those related to cardiomyopathy including ECMO, LVAD and AICD
- Kawasaki disease
- HIV infection or born to a HIV positive mother
- History of Rheumatic fever
- Prior exposure to anthracyclines or other cardiotoxic drugs
- Cardiomyopathy related to chronic arrythmia except if it was included prior to onset of arrythmia , except a patient with chronic arrthymia subsequently ablated

SPECIFIC OUTCOMES TO BE STUDIED

- • Death from HF
- • SCD
- • Ventricular arrhythmias
- • AV block
- • Requirement of CRT/ICD/Transplant

DATA ANALYSIS

The data analysis was performed using the SPSS Statistics software for Windows Version 21 by the principal investigator with the guidance of Prof Sankara Sarma, Department of Biostatistics, SCTIMST. Continuous variables were expressed as either mean standard deviation or median depending on the overall variable distribution. Descriptive summaries were presented as frequencies and percentages for categorical data. Continuous variables were compared using Student's t test or Mann-Whitney U test as appropriate. Group comparisons were made using $\chi 2$ tests. The data will be stored in the personal computer of the principal investigator for a period of three years, maintaining confidentiality.

RESULTS

- During the thirty year study period (1990-2020), 233 cases of pediatric cardiomyopathy were identified. 119 cases of dilated cardiomyopathy (51 percent), 63 cases of hypertrophic cardiomyopathy (27 percent), 33 cases of restrictive cardiomyopathy (14 percent), and 11 cases of left ventricular noncompaction cardiomyopathy (4.7 percent). Besides this, five cases of ARVD, one case each of RVNC and atrial cardiomyopathy were identified.

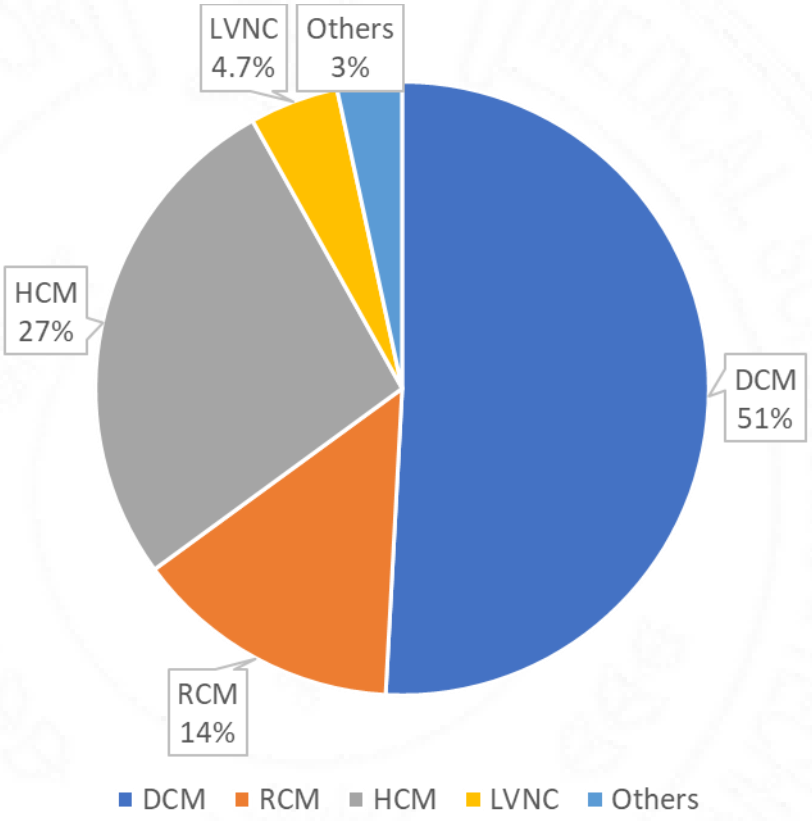


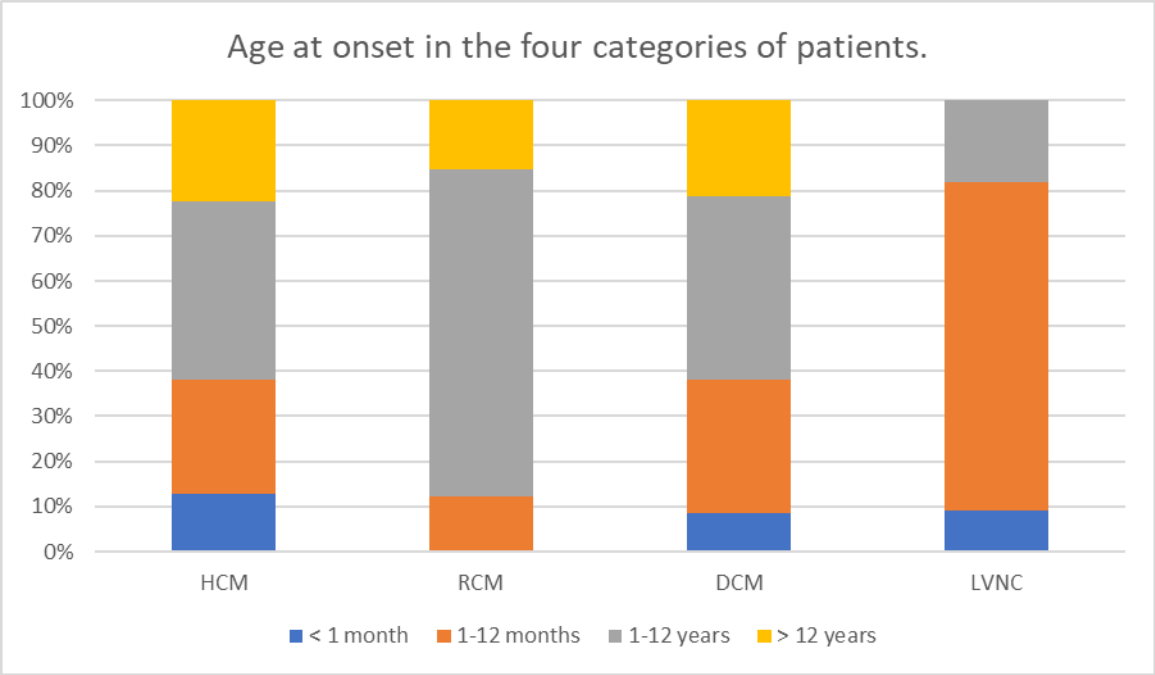
Chart showing relative percentages of the each type of cardiomyopathy

The age of presentation of each type of cardiomyopathy is described in table1.

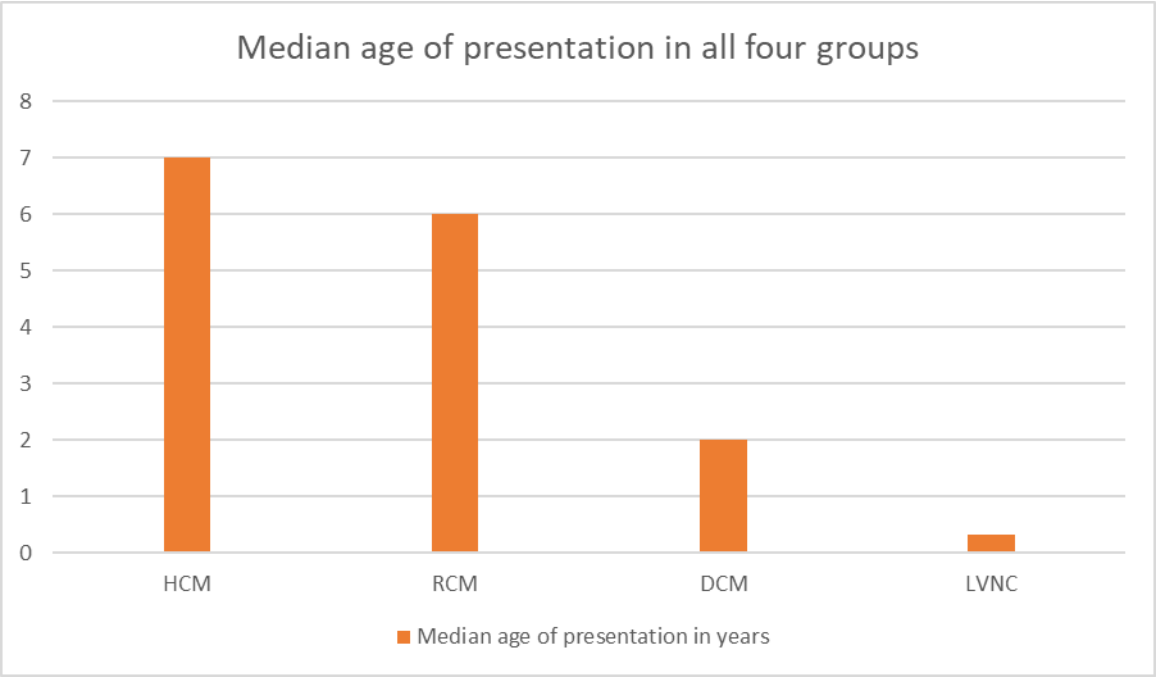
Age	HCM	RCM	DCM	LVNC	Total
< 28 days Freq (%age)	8(12.7)	0(0)	10(8.4)	1(9)	19
1-12 months	16(25)	4(12)	35(29)	8(72)	63
1-12 years	25(39)	24(72)	48(40)	2(18)	99
>12 years	14(22)	5(15)	26(21)	0(0)	45
Total	63	33	119	11	226

Table 1 showing the age distribution of the four major types of cardiomyopathy

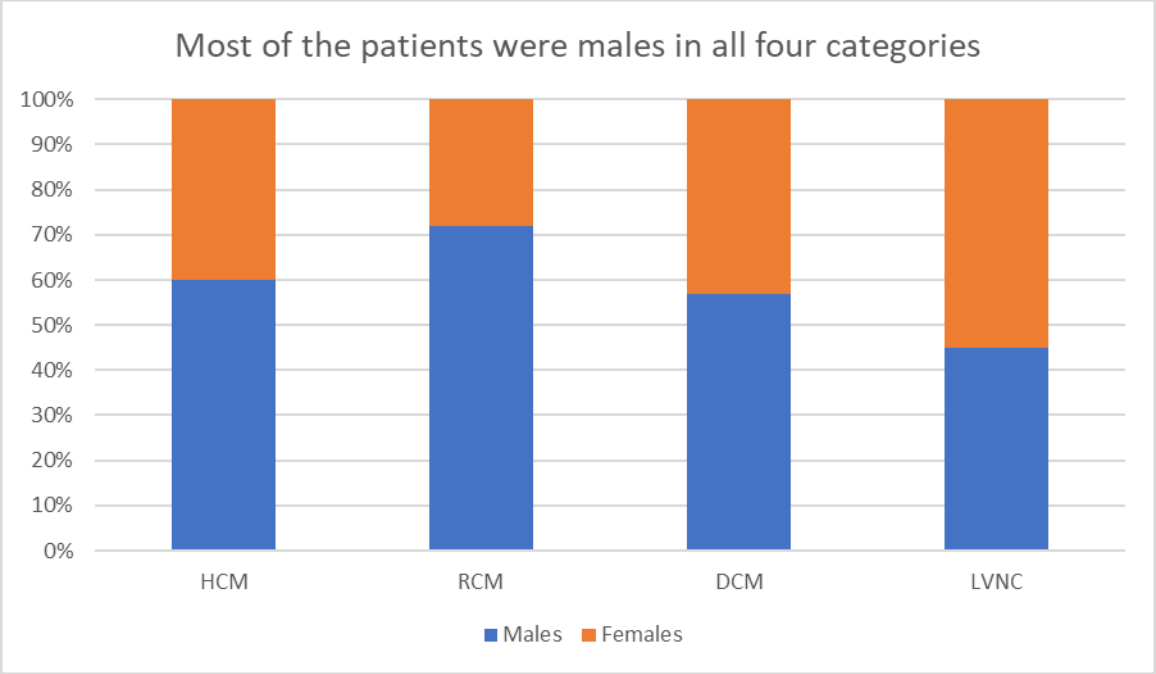
35% out of the total cases received a diagnosis in the first year of life.



Age at onset of the four categories of pediatric cardiomyopathy. None of the RCM patients presented in the neonatal period and none of the LVNC patients presented after 12 yrs. 12% of the HCM cohort presented in the neonatal period.

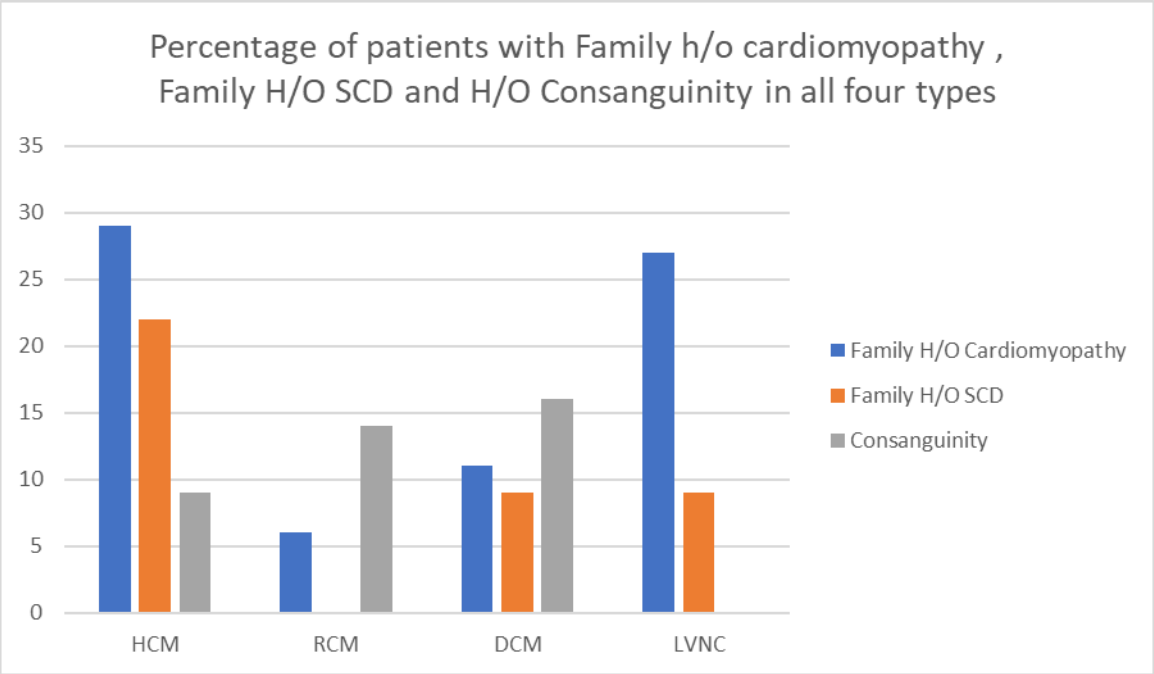


The median age of presentation of HCM patients was 7 yrs (Range 0.1-18 yrs)The median age of presentation of DCM patients was 2 yrs (Range 0.1-18 yrs) , of RCM patients was 6 yrs(Range 0.8-16) and of LVNC patients was 4 months (Range- 1 day to 10 yrs)



Among all cardiomyopathies, males were more commonly diagnosed with cardiomyopathy than females. 142 out of 232 cases were males(61%).Of the 119 children with dilated cardiomyopathy, 69 were males (57.0 percent), as were 38 of the 63 children with hypertrophic cardiomyopathy (60 percent),

24 of the 33 with restritive cardiomyopathy (72 percent), and 5 of the 11 with LV noncompaction cardiomyopathy(45%).



Family history of cardiomyopathy was most common in HCM patients(29%) and least common with RCM patients(6%). Family history of SCD was also most common in HCM patients(22%).None of the RCM patients had family history of SCD.

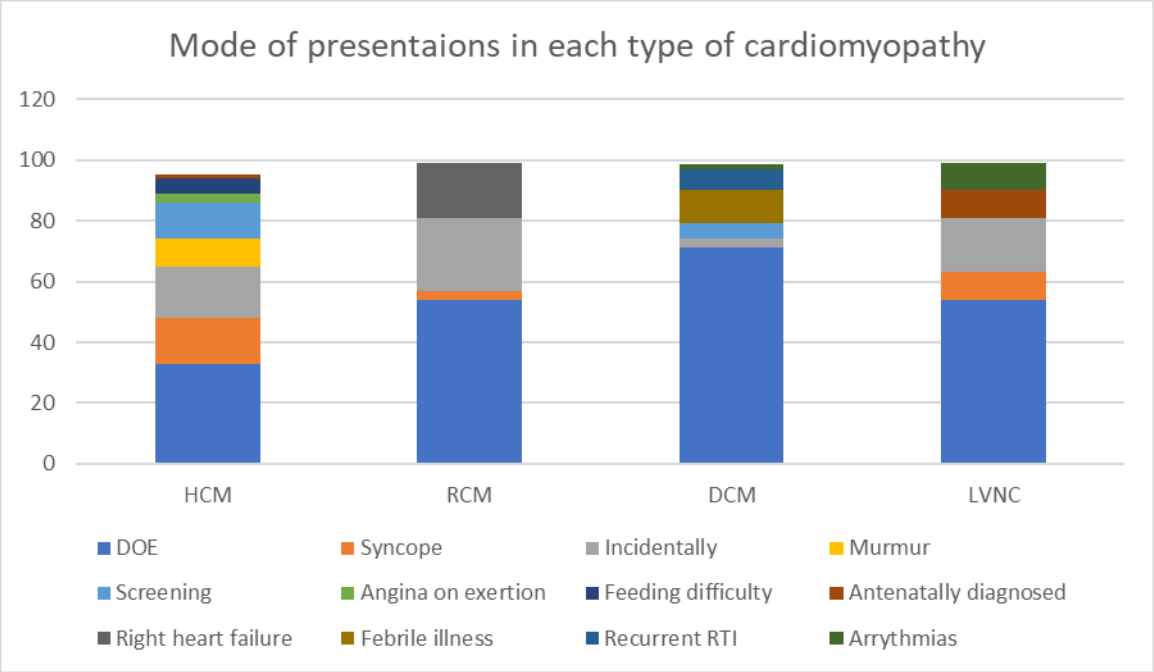


Chart depicting the mode of presentation of the four categories of cardiomyopathies. Dyspnea on exertion was the most common mode of

presentation in all four types.Syncope was most common in HCM patients.None of DCM patients presented with Syncope.

The clinical profile of HCM patients is described in Table 2

8 of the 63 HCM patients(12%) presented in the neonatal period and 24 (37%) patients presented in the Infancy. 60% were males.The most common mode of presentation was dyspnea on exertion (33%).10 patients presented with Syncope (15%). 8 patients were diagnosed as a result of family screening. Two patients presented with Angina on exertion. Out of the 63 HCM patients , H/O Consanguinity was known in 55, out of which 5 patients (9%) had history of parental consanguinity. Syncope developed at any time during the course of illness in 18 patients (28%) . Family History of cardiomyopathy was present in 18 patients (29%). Family history of sudden cardiac death was present in 14 patients (22%). 15 pateints had proven syndromic or metabolic disease (24%). Noonans Syndrome was the most common syndrome found (9 patients). One patient had Friedrichs Ataxia(GAA mutation positve). One had Pompes disease (alpha glucosidase mutation positive). One had Childhood onset myoclonus dystonia. Three had unclassified syndromes. One patient had MYBPC3 mutation positive. 44 patients (69%) were in NYHA FC II. 14 patients (25%) were in NYHA FC I . Three patients were in NYHA FC III. On examination, cardiac enlargement was present in 10 patients. LV S3 was present in 7 patients(11%). Loud P2 was present in 9 patients(14%). S4 was present in 15 patients(34%). Ejection systolic murmur was present in 47 patients(74%). Double apical impulse was present in 7 patients (11%)

Chest Xray was done in 54 patients. Mean Cardiothoracic ratio was 0.57 +/- 0.07 (Range 0.45-0.8) , Right atrial enlargement was present in 15 patients(27%).Left atrial enlargement was present in 35 patients (64%).

Pulmonary veinous hypertension was present in 24 patients (44%) on chest x ray.

ECG characteristics of HCM patients are shown in Table 3

The median PR interval was 140 msecs(Range 80-250). PR interval median z score was 1.0 (Range - 3.2 to 7.27). 10 patients (15%) had first degree AV block that is, PR interval > 2.5 z score.Median QRS duration was 100(Range 80 to 180).QRS duration median z score 2.0 (Range -1.0 to 8.3). 5 patients had QRS duration z score > 2.5. ECG was available for interpretation in 41 patients. Median QT interval in these patients was 453 ms (Range 400 ms -588 ms). QT dispersion was 52(Range 20-126). Twenty patients had left axis deviation. Two had north west axis. Two had right axis deviation. q waves were present in 37 patients(62%). ST depression was found in 16 patients (39%). Echocardiographic evidence of LVH was present in 53 patients (89%). Pre excitation was present in two patients.

Echocardiographic parameters of HCM patients are given in Table 4 and Table4.

The median Interventricular septal thickness was 16 mm (Range 7-36 mm). Weight was available for 40 patients only. So z scores could be calculated for 40 patients. The median Interventricular septal thickness z score was 4.31 (Range 2.33 to 7.3). The median posterior wall thickness was 15 mm (Range 6- 38 mm). The median posterior wall thickness z score was 2 (Range -1.49 to 8.06). Mean LA dimension was 30 +/- 12 (Range 12-80 mm). LA dimension z score(n=40) was 2.56 (-1.08 to 8.9). Mean LVEF was 71+/- 12% (Range 30%-87%).

Character(n=63)	Frequency (Percentage)
Age	
< 1 month	8(12)
1 m - 12 months	16(25)
> 1yr -12 years	25(39)
> 12 yrs	14(22)
Males	38(60)
Mode of presentation	
Dyspnea on exertion	21(33)
Murmur	6
Incidentally	11
Screening	8
Syncope	10(15)
Angina on exertion	2
Feeding difficulty	3
Antenatally diagnosed	1
Consanguinity (n=55)	5 (9)
Syncope	18(28)
Family H/O Cardiomyopathy	18(29) , Not known in 2
Family H/O SCD	14(22)
Syndromic	15(24) ; Noonan Syndrome Most common

NYHA FC	
I	16 (25%)
II	44 (69%)
III	3 (4.7%)
Cardiac enlargement	10(15)
LVS3	7(11)
Loud P2	9(14)
S4	15(34)
ESM	47(74)
Double apical impulse	7(11)
CXR (n=54)	
CTR (m+/-SD)	0.57+/- 0.07 (0.45-0.8)
RAE	15(27)
LAE	35(64)
PVH	24(44)

Table 2 showing the clinical profile of 63 HCM patients

ECG(n=59)	
PR interval, Median(Range)	140 (80-250)
PR interval z score, Median(Range)	1.0(- 3.2 – 7.27)
PR interval > 2.5 z score,n(%)	10(15)
QRS d, Median(Range)	100(80-180)
QRS d zscore, Median(Range)	2.0 (-1.0 – 8.3)
QRS d > 2.5 z score,n(%)	5(8)
QT interval, ms Median(Range) (n=41)	453(400-588)
QT dispersion (n=41)	52(20-126)
LAD,n(%)	20(32), 2 had NW axis
RAD,n(%)	2(3)
Normal axis	37(62)
q waves	37(62)
ST depression(n=41)	16(39)
LVH	53(89)
Preexcitation	2

Table 3 showing ECG features of HCM cohort patients

ECHO	Median (Range)
IVSs (mm)	16 (7-36)
IVSs z score(n=40)	4.31 (2.33 – 7.3)
PWs (mm)	15 (6-38)
PWs (z score) (n=40)	2 (-1.49 – 8.06)
LA dimension,mm (mean)	30 +/- 12 (12-80)
LA dimension z score(n=40)	2.56 (-1.08 - 8.9)
LVEF, mean	71+/- 12 (30-87)
LVOT Obstruction	32(50)
LV AO gradient,mmHg Median, (Range)	88 (15-169)
SAM	41(64)
PAH	5(7.9)
Mitral regurgitation	
No	33(52)
Mild	17(26)
Moderate	11(17)
Severe	2(3)
Biventricular	5
Apical	1
Burnt out HCM	2

Table 4 showing Echocardiographic features of HCM patients.

LVOT Obstruction is present in 32 patients(50%).Median LVOT gradient was 88mmHg (Range 15-169 mmHg).SAM was present in 41 patients (64%). Pulmonary hypertension was present in 5 patients.Thirty patients had mitral regurgitation, Two of them had severe mitral regurgitation.Five patients had Biventricular HCM.1 patient had Apical HCM.Two patients had burn out HCM.

Table 6 depicts the Holter was done in thirty patients, out of which twenty one had normal holter. Five patients had NSVT/VPC s.Two patients had Atrial arrythmias. Two patients had Conduction defects.

Cardiac MRI was done in 26 patients, out of which 16 patients had Late gadolinium enhancement. Seven had mid myocardial enhancement, three had subendocardial and six had diffuse subendocardial enhancement.

48 patients were on beta blocker,mostly on metoprolol.Eight patients were on diuretics, One was on digoxin and one was on ACE-Inhibitor.

Median NTpro BNP was 2052 (Range40-9540 pg/ml).

Cardiac catheterization was done in 17 patients.Mean PA pressure was 22+/-6. LV ed was 15+/-7.Median LV AO gradient by cath was 100 (Range 30-140).Brokenbrough braunwald sign was present in 7 patients.

Outcome parameters of HCM patients is described in Table 6.

Over a mean follow up period of 5.6 years (0.1-30 years),twenty seven were lost to follow up(42%).Out of the patients on follow up, five died(13%)and thirty one(86%) were alive.Out of the five patients who died,three died of sudden cardiac death,one from heart failure and one from ventricular arrythmias.

Sustained ventricular arrythmias were seen in two patients and atrial arrythmias in two patients. First degree AV block was seen in 10 patients(15%).Bundle branch blocks(BBB) in five patients(8%).Eight patients required ICD(12.7%).Two patients underwent septal myectomy and both are alive and asymptomatic after 24 yrs and 8 yrs of follow up respectively. Two patients underwent ICD both for primary prevention and one underwent PPI for distal AV conduction disease.

Holter (30)	
Normal	21
NSVT/ VPCs	5
Atrial tachy/AFib	2
Intermittent CHB / BBB	2
CMRI (26)	
LGE present	16
Midmyocardial	7
Subendocardial	3
Diffuse	6
LGE Absent	10
Medications	
Beta blocker	48
Metoprolol	29

Propanolol	12
Bisoprolol	3
Atenolol	4
Diuretics	8
Digoxin	1
ACE Inhibitor	1
NT pro BNP pg/ml, Median(range) (n=12)	2052 (40-9540)
Cardiac cath (n=17)	
PA mean	22+/- 6
LV ed,mean	15+/- 7
LV AO grad by cath,mmHg(median) (n=9)	100 (30-140)
Brokenbrough Braunwald sign	7(77)

Table 5 showing hemodynamic features and other clinical characteristics of HCM cohort.

Outcome (n=63)	
Years of Follow up (mean)	5.6 (0.1-30)
Alive (% of pts on follow up)	31 (86)
Death (% of pts on followup)	5 (13)
Death from HF	1
SCD	3
Death from ventrarrythmias	1
Lost to follow up(%)	27 (42)
Sustained Ventricular arrythmias	2 (3)
Atrial arrythmias	2 (3)
AV block	10 (15.8)
BBB	5(8)
Requirement of ICD/CRT Transplant	8 (12.7)
Septal myectomy	2 (Both asymptomatic,alive)
ICD/PPI	3 (2 underwent ICD for primary prevention, 1 underwent PPI for distal AV conduction disease)
EPS	4

Table 6 showing Outcome characteristics of HCM patients. SCD Sudden cardiac death, AV block Atrioventricular block , BBB Bundle Branch block

	Death or ventricular arrythmias N Y	Total
CMRI LGE N	10 0	10
CMRI LGE Y	13 3	16
	23 3	26

Table 7 Relationship between Cardiac MRI and Mortality.26 patients had CMRI done.Among the patients with LGE on MRI, three died and no patient died among the patients with no LGE on CMRI.P value was not significant.(P=0.26)

Variable	P-value
Age	0.37
Syncope	0.39
Family H/O Cardiomyopathy	0.18
Family H/O Sudden cardiac death	0.33
Consanguinity	0.11
QT Dispersion	0.34
IVS z score	0.57
LAE z score	0.23
SAM	0.68
NSVT on holter	0.45
LGE on MRI	0.26
LVOT obstruction	0.70

Table 8 depicting the relationship between various clinical, echocardiographic and radiological risk factors and death or ventricular arrhythmias in HCM cohort.None of the parameters tested had a significant relationship.

Clinical profile of 33 restrictive cardiomyopathy is depicted in table 9.

None of the 33 RCM patients(12%) presented in the neonatal period and Only 4 patients presented in the Infancy.72% were males. The most common mode of presentation was dyspnea on exertion (54%).1 patient had presented with syncope.Family History of cardiomyopathy was present in 2 patients(6%).No patient had family History of sudden cardiac death.1 patient had History of Emery Driefuss muscular dystrophy.Consanguinity was known in 28 patients, out of which 4 patients (14%) had history of parental consanguinity. Syncope developed at any time during the course of illness in 3 patients (9%) . Only three patients were in FC I(9%).17 patients (51 %) were in NYHA FC II. 12 patients (39%) were in NYHA FC III. On examination, cardiac enlargement was present in 24 patients (72%). LV S3 was present in 12 patients(36%). RVS3 was present in 2 patients(6%). Both LVS3 and RVS3 was present in 1 patient.Loud P2 was present in 18 patients(54%). Murmur on auscultation was present in 13 patients(39%).

Chest Xray was done in 32 patients. Mean Cardiothoracic ratio was 0.63 +/- 0.1 (Range 0.45-0.8) , Right atrial enlargement was present in 22 patients(68%).Left atrial enlargement was present in 18 patients (60%). Pulmonary veinous hypertension was present in 24 patients (75%) on chest x ray.

ECG characteristics of RCM patients are shown in Table 10

Two patients were in Atrial fibrillation and two were in junctional rhythm. The median PR interval was 140 (Range 100-220). PR interval median z score was 1.0 Range (- 1.5 to 4.8). 7 patients (21.8%) had first degree AV block that is, PR interval > 2.5 z score.Median QRS duration was 100(Range 80 to 140).QRS duration median z score 2.0 (Range 0 to 3.0). 2 patients had QRS duration z score > 2.5. ECG was available for

interpretation in 14 patients. So,QT interval,ST depression and q waves could be interpreted in these patients only. Median QT interval in these patients was 465 ms (Range 361 ms -508 ms). QT dispersion was 52(Range 20-130).One patient had left axis deviation.Seven had right axis deviation. q waves were present in 11 patients(93%). ST depression was found in 7 patients (50%). Echocardiographic evidence of LVH was present in 18 patients (89%). Biventricular hypertrophy was present in 7 patients.None of the patients had evidence of pre excitation. One patient had TNNI3 mutation positive , one patient had MYL3 gene mutation positive.

Character (n=33)	
Age	
< 1 month	0
1-12 months	4(12.1)
1-12 yrs	24 (72.7)
> 12 yrs	5(15.5)
Males/Female	24/9 (72)
Mode of presentation	
DOE	18 (54)
Incidentally	8(24)
Right heart failure	6 (18)
Syncope	1(3)
Syncope	3 (9)
Family H/O CMP	2 (6) , NA in 1
Family H/O SCD	None

Syndromic	1(3)
Consanguinity (n=28)	4()
FC	
I	3(9)
II	17(51)
III	12(39)
IV	1
Cardiac enlargement	24(72)
LVS3	12(36)
RVS3	2(6)
Both	1(3)
Loud P2	18(54)
ESM/PSM	13(39)
CXR(n=32)	
CTR Mean	0.63+/-0.1
LAE	18(60)
RAE	22(68.7)
PVH	24(75)

Table 9 showing the clinical profile of RCM cohort

ECG	
PR interval mm, Median (Range)	140 (100-220)
PR interval z score, Median(range)	1.0(-1.5-4.8)
PR interval > 2.5 zscore	7(21.8)
QRS d mm, Median (Range)	100 (80-140)
QRS d z score Median (Range)	2.0 (0-3.0)
QRS d z score> 2.5	2(10.7)
QT interval ms (n=14)	465(361-508)
QT dispersion (n=14)	52(20-130)
LAD	1(3.3)
RAD	7(23)
Normal axis	22(73)
Q waves (n=14)	11(93)
ST depression(n =14)	7(50)
LVH	18()
BVH	7()
Preexcitation	none

Table 10 showing ECG characteristics of RCM cohort.

ECHO	Median (Range)
LVID d	32 (22-43)
LVID d z score (n=29)	-1.36 (-5.32-2.82)
LVID s	21 (12-32)
LVID s z score (n=29)	-0.47 (-5.2-2.7)
IVS s	9 (5.0-18.0)
IVS z score (n=29)	-0.77 (-2.0 - 4.0)
PW s mm	10 (5.0-16)
PW z score (n=29)	0.51 (-2.0 – 3.0)
LA dimension mm, mean	35+/- 8.35 (20-53)
LA dimension z score (n=29)	3.82 (0-7.0)
LVEF, mean	61.9 +/- 10 (32-80)
PAH	14(42.4)
BAE	33 (100)
Pericardial effusion	4

Table 11 showing Echocardiographic characteristics of RCM patients

Echocardiographic parameters of RCM patients are given in Table 11.

The median diastolic left ventricular internal dimension(LVId d) was 32 mm (Range 22-43 mm). Weight was available for 29 patients only. So z scores could be calculated for 29 patients. The median LVId d z score was -1.36 (Range -5.32 to 2.82). The median systolic left ventricular Internal dimension(LVId s) was 21 mm (12-32 mm).Median LVid s z score -0.47 (-5.2-2.7).Interventricular septal thickness z score was 0.77(Range -2.0-4.0). The median posterior wall thickness was 10 mm (Range 5-16 mm). The median posterior wall thickness z score was 0.51 (Range -2.0-3.0). Mean LA dimension was 35 +/- 8.35 (Range 20-53 mm). LA dimension z score(n=29) was 3.82 (0.0 to 7.0). Mean LVEF was 61.9 +/-10 (Range 32-80%). PAH was present in 14 patients. Biatrial enlargement in 100% patients.Four patients had pericardial effusion.

Table 12 shows the hemodynamic features of RCM patients.

Holter was done in 9 patients.It was normal in 7 patients.1 had Atrial tachycardia/AFib and one had Frequent VPCs.12 patients had cardiac MRI done. Late gadolinium enhancement was present in 5 patients and absent in 7 patients.All had subendocardial enhancement. Out of 33 patients of RCM,28 were on furosemide, 16 were on digoxin and 13 were on Aldactone. 9 were on ACE-inhibitors.1 was on betablocker and 1 was on cordarone.NT pro BNP was done in 9 patients.Median NT proBNP was 4000(1296-10900).Cardiac cath was done in 24 patients. Mean RA pressure was 13 +/- 7 mmHg.Right ventricular systolic pressure was 47+/- 16 mmHg.Right ventricular end diastolic pressure 16+/- 6 mmHg.Mean pulmonary artery pressure was 30+/- 12 mmHg. Mean Pulmonary Capillary Wedge Pressure was 20+/-7 mmHg.Left ventricular systolic pressure was 97+/- 13 mmHg.Left ventricular end diastolic pressure was 25+/- 7

mmHg.Pulmonary vascular resistance index was 4+/-3 WU.m2 . Cardiac index was 2.4 +/- 1.1.

Holter(n=9)	
Atrial tachycardia/A Fib	1
Frequent VPC s	1
Normal	7
CMRI (n=12)	
LGE present	5
Subendocardial	5
LGE absent	7
Medications	
Digoxin	16
Furosemide	28
ACE-I	9
Aldactone	13
Beta blocker	1
Cordarone	1

NT pro BNP,pg/ml , median (range) (n=9)	4000 (1296-10900)
Cardiac cath (n=24)	
RA mean (mmHg)	13 +/- 7
RVSP (mmHg)	47 +/- 16
RVEDP (mmHg)	16 +/- 6 (6-30)
PA mean (mmHg)	30 +/- 12 (13-62)
PCWP	20 +/- 7
LVSP (mmHg)	97 +/- 13
LVEDP (mmHg)	25+/- 7 (12-42)
PVRI (W U. m2)	4 +/- 3 (1.0-16.9)
CI	2.4 +/- 1.1

Table 12 Hemodynamic features and other clinical characteristics of RCM patients.

Outcome	
Years of follow up , mean(range)	3.7 +/- 4.69 (0.1-25)
Death	6
Death from heart failure	2
SCD	4
Alive	8
Lost to follow up (% of all)	19(57%)
Ventricular arrythmias	0
Atrial arrythmias	2
AV Block	7(21)
BBB	2(3)
Requirement of CRT / ICD / Transplant	16 (48)
EPS	1
ICD/PPI	1 PPI done for atrial paralysis

Table 13 Outcome of RCM patients.

Outcome of children with restrictive cardiomyopathy has been described in Table 13

Over a mean follow up period of 3.7 years (0.1-25 years), nineteen were lost to follow up(57%).Out of the patients on follow up, six died(43%)and eight (52%) were alive.Out of the six patients who died, four died of sudden cardiac death,two from heart failure.

None of the patients had sustained ventricular arrythmias and atrial arrythmias were seen in two patients. First degree AV block was seen in 7 patients(21%).Bundle branch blocks(BBB) in two patients(3%).Sixteen patients required ICD/CRT or transplant(48%).Electrophysiological study was done in one patient showing atrial paralysis.Permanent pacemaker implantation was done in 1 patient for atrial paralysis.

Variable	P - value
Syncope	Significant (0.07)
ST depression	Not significant
AV block	Not significant
BBB	Not significant

Table 14 depicting the relationship between various clinical risk factors and death or ventricular arrhythmias in RCM cohort.The relationship with Syncope was close to be statistically significant.

Clinical profile of dilated cardiomyopathy patients presented in Table 15

10 of the 119 DCM patients(8 %) presented in the neonatal period and 45 (37%) patients presented in the Infancy. 57% were males.The most common mode of presentation was dyspnea on exertion (71%).None of the patients presented with Syncope. 7 patients were diagnosed as a result of family screening. Out of the 119 DCM patients , H/O Consanguinity was known in 106, out of which 18 patients (16%) had history of parental consanguinity. Syncope developed at any time during the course of illness in 3 patients (2.5%) . Family History of cardiomyopathy was present in 14 patients (11%). Family history of sudden cardiac death was present in 11 patients (9%). 8 patients had proven syndromic or metabolic disease (6.7%). 64 patients (53%) were in NYHA FC II. 14 patients (25%) were in NYHA FC I . 44 patients(36%) were in NYHA FC III. On examination, cardiac enlargement was present in 9 patients(75%). LV S3 was present in 64 patients(54%). Loud P2 was present in 48 patients(40%). Mean CTR was 0.65+/- 0.06.

Variable (n=119)	**Frequency (%)**
Age	
< 1 month	10 (8)
1-12 months	35 (29)
1-12 years	48 (40)
> 12 years	26 (21)
Male/Female	69/50 (57)
Mode of presentation	
DOE	84 (71)
Febrile illness	13 (11)
Screening	7 (5)
Recurrent RTI	8 (7)
Incidentally	4 (3)
Arrythmias	2 (1.6)
Syncope	3 (2.5)
Family H/O Cardiomyopathy	14 (11)
Family H/O SCD	11(9)

Consanguinity(n=106)	18
Syndromic	7 (5)
FC	
I	6(5)
II	64(53)
III	44(36)
IV	5 (4)
Cardiac enlargement	90 (75)
LVS3	64(54)
P2	48 (40)
ESM/PSM	61(51)
CXR	
CTR Mean +/- SD	0.65+/- 0.06
LAE	79(73)
PVH	94(87)

Table 15 Clinical characteristics of DCM patients

ECG characteristics of the DCM patients are given in Tab 16 . The median PR interval was 1.0 . Range -2.0 to 4.7. The median QRS duration z score was 2.0 (Range -1.0 to 7.3). LAD was seen in 13 patients(12%).ECG was available for interpretation only in 105 patients out of which ST depression was seen only in 2 patients, as compared to 50% of patients in RCM. Preexcitation was present in 4 patients with DCM.

ECG (n=119)	
PR interval ms Median (Range)	140 (80-240)
PR interval z score , Median (Range)	1.0 (-2.0 - 4.7)
QRS d , ms Median (Range)	100 (80-180)
QRS d z score Median (Range)	2.0 (-1.0 - 7.3)
LAD	13(12)
RAD	4 (3)
Normal axis	102
q wave	41 (34)
ST depression (n=105)	2 (1.6)
LVH	96(80)
Preexcitation	4

Table 16 ECG characteristics of DCM patients.

On ECHO , the median LVid z score was 4.3 (Range -0.4 to 9.06).Median LVEF was 31%. (Range 10%-67%). 28 patients had PAH.Severe Mitral regurgitation was present in only 5 patients.Holter was done in 11 patients out of which 4 had NSVT/VPCs and one had Atrial tachycardia/AFib.

Cardiac MRI was done in 24 patients.In 12 patients, LGE was present and in rest, LGE was absent.

LVId d (mm) Median (Range)	48 (20-84)
LVId d (z score) Median (Range) (n=98)	4.3 (-0.4 – 9.06)
LVId s (mm) Median (Range)	40 (15-71)
LVId s (z score) Median (Range)(n=98)	5.8 (0.12-10.2)
LVEF % Median (Range)	31 (10-67)
PAH	28(23)
Mitral regurgitation (n=104)	
Mild	46 (38)
Moderate	53 (44)
Severe	5 (4)

Table 17 Echocardiographic characteristics of DCM patients

Variable	Frequency (%)
Holter (n=11)	
NSVT/ VPC s	4
Atrial tachy/A Fib	1
Normal	5
BBB	1
C MRI (n= 24)	
LGE present	12(50)
Subendocardial	6
Transmural	3
Mid myocardial	2
Patchy	1
LGE absent	12(50)

Table 18 Holter and MRI findings of DCM patients.

Medications	
Digoxin	89(74)
Furosemide	107(89)
ACE-I	86(72)
Aldactone	48(40)
Beta blocker	57 (47)
Amiodarone	6 (5)
Warf	3 (2.5)
ARNI	4 (3)
NT pro BNP	4130 (134-35000)
Cardiac cath (n=16)	
PA mean	26 +/- 11
PCWP	18 +/- 5
LVEDP	16 +/- 9
CI	2.74 +/- 1.4
PVRI	4.26 +/- 3.9

Table 19 Hemodynamic and other clinical characteristics of DCM patients.

74% of the DCM patients were on digoxin, 89% were on furosemide, 72% were on ACE-I. Only 40% were on Aldactone and 47% were on beta blockers. Digoxin was prescribed in the early periods of the study and the practice of prescribing digoxin fell in the later periods and ACE-I and beta blockers were more commonly prescribed in the later parts.Only 6 patients were on amiodarone , 3 were on Warf.The practice of prescribing ARNI started recently and thus only 4 patients were on ARNI.The median NTpro BNP was 4130 (Range 134-35000).

Cardiac cath was done only in 16 patients in early parts of the study.The important parameters noted are described in the table.

Over a mean follow up of 2.98 +/- 3.97 years, 37% were lost to follow up.Among the remaining patients who were on follow up, 33 died(45%). 27 recovered (36%). 14 had persistent LV dysfunction (19%).

Echocardiographic and other clinical characteristics of the DCM patients are given in Tables 17, 18 and 19.

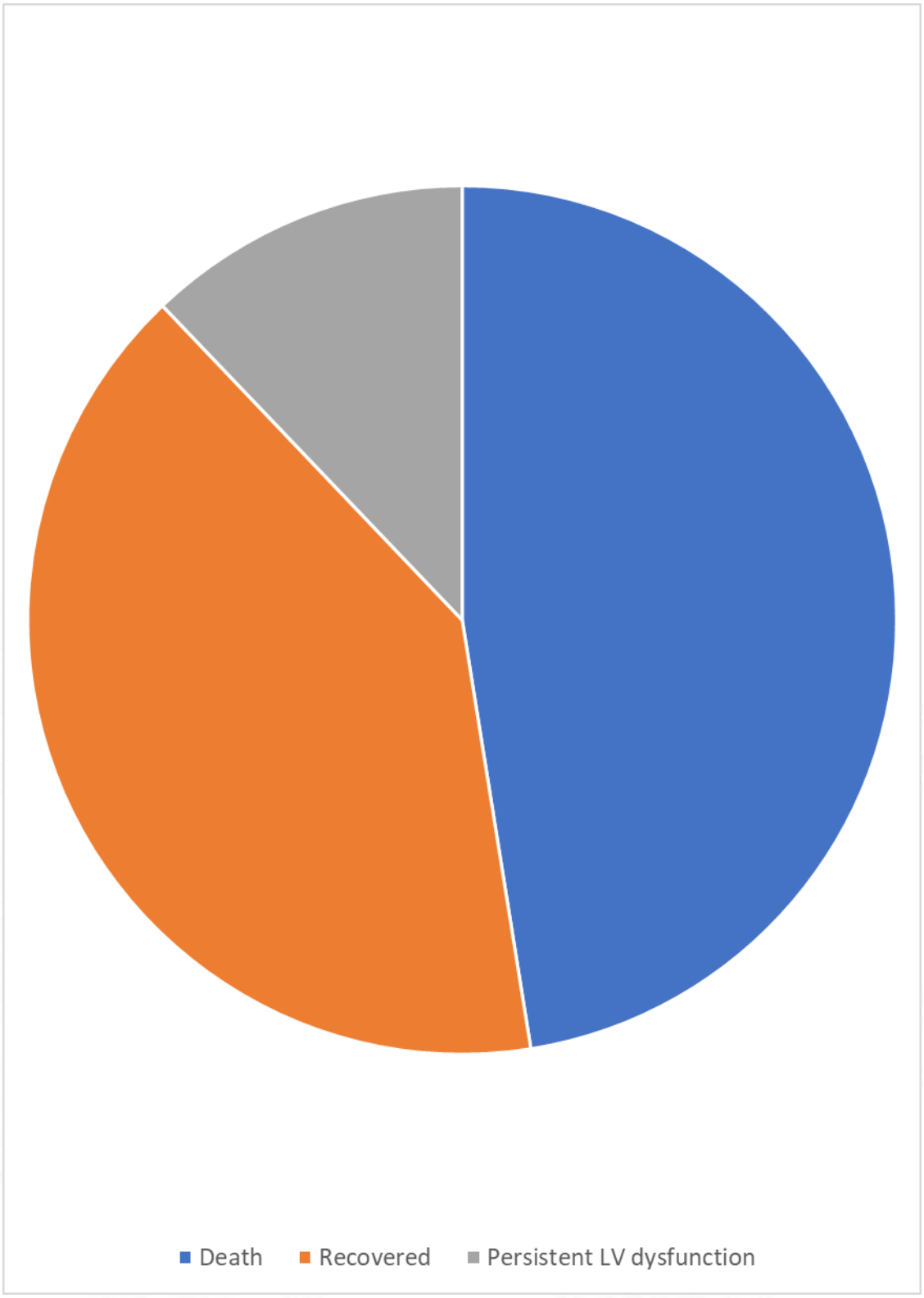
Outcome	
Years of follow up, Mean +/- SD (Range)	2.98 +/- 3.97 (0.1- 24)
Lost to Follow up, n=119 (%)	45 (37)
Death, n=74(% of patients on follow up)	33 (45)
Death from HF	25
SCD	3
Death from ventr arrythmias	5
Recovered, n=74 (%)	27(36)
Persistent LV dysfunction n=74 (%)	14 (19)

Table 20 showing outcome in DCM patients.

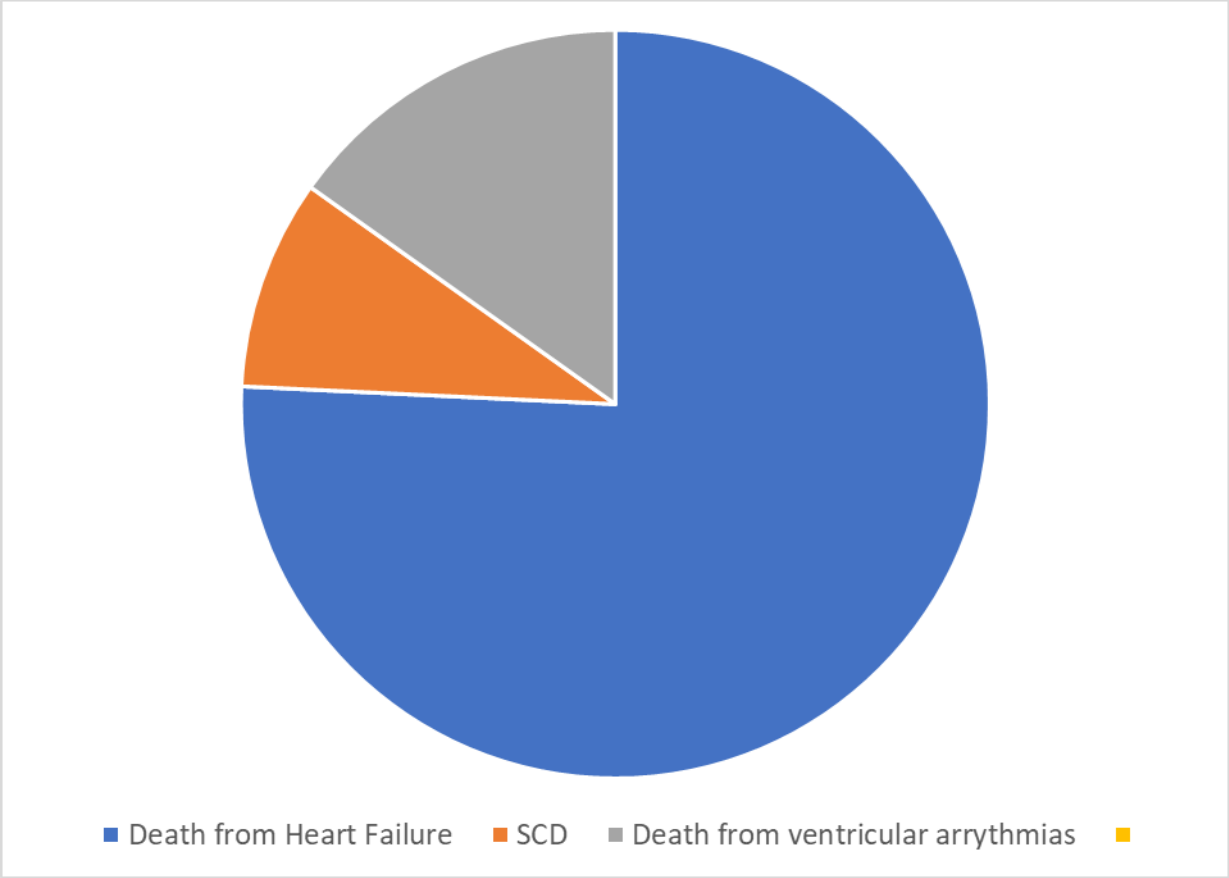
Outcome	
Sustained Ventricular arrythmias	**9 (7.7)**
Atrial / junctional arrythmias	6 (5.1)
AV block	14 (11.9)
BBB	9 (7.7)
LBBB	2
Requirement of ICD/CRT/ Transplant	41 (34)
ICD/ PPI/CRT	1 ICD , 2 CRT
EPS	1
ECMO	1

Table 20 showing outcome in DCM patients

Among the 119 DCM patients, 9 had sustained ventricular arrthmias. Atrial / Junctional arrythmias were present in 6 patients. 14 patients had First degree AV block(11.9%). 9 patients had BBB, among which only 2 had Left bundle branch block.Requirement of ICD/CRT/Cardiac transplant was considered indicated in 41 patients (34%). 1 patient underwent ICD and two underwent CRT. Electrophysiological study was done in 1 patient. 1 patient had required ECMO.



Pie diagram showing percent of patients who died , recovered or had LV dysfunction among the DCM cohort.



Most common cause of death was heart failure among the DCM patients.

Variable	P value
LVid z score > 4	Not significant (0.69)
Age,Infancy vs Non infancy	Not significant (0.25)
Family H/O Cardiomyopathy	Not significant
Syncope	Not significant

Table 21 showing **the relationship between various clinical and echocardiographic risk factors and death or ventricular arrhythmias in DCM cohort.None of the parameters tested had a significant relationship.**

The clinical profile of LVNC patients has been decribed in table 22. Among the 11 patients of LVNC, most presented in infancy. Dyspnea on exertion was the most common mode of presentation. History of syncope was present in only 2 patients Three had family history of cardiomyopathy.

Variable (n=11)	Frequency (%)
Age	
< 28 days	1
1-12 months	8
1- 12 years	2
> 12 years	0
Male	5 (45)
Mode of presentation	
DOE	6
Incidentally	2
Arrythmias	1
Antenatally diagnosed	1
Syncope	1

Variable	Frequency (%)
Syncope	2 (18)
Family H/O Cardiomyopathy	3 (27)
Family H/O SCD	1 (9)
Syndromic	1 (Unclassified)
FC	
I	0
II	9
III	2
Cardiac enlargement	4 (36)
LVS3	2 (18)
P2	4 (36)
PSM/ESM	3 (27)
CXR	
CTR	0.62 +/- 0.07 (0.5-0.7)
LAE	7 (77)
RAE	3 (33)
PVH	8 (88)

Table 22 Clinical characteristics of LVNC patients.

PR interval mm ,median (range)	140 (100-200)
PR interval z score, median (range)	1.5 (0.0-4.70)
PR interval > 2.5 z score	2
QRS d mm, median (range)	100 (80-120)
QRS d z score, median (range)	2.0 (0 -2.0)
QT interval (n=5)	437 (350-566)
QT dispersion (n=5)	45 (40-85)
LAD	3 (30) Rest normal
Q wave	4 (36)
ST depression	1 (90)
LVH	10 (90)

Table 23 ECG characteristics of LVNC patients.

LVID d mm	41 (9-55)
LVID d z score	2.8 (-3.05 – 9.01)
LVID s mm	33 (4-48)
LVID s z score	4.41 (-5.06- 9.9)
LA dimension	22 +/- 10 (7-44)
LA dimension z score	2.13 (-1.56 – 5.6)
LVEF	41 (24-80)
PAH	3 (27)
Mitral regurgitation (n=6)	
Mild	3
Moderate	2
Severe	1

Table 24 describes the ECHO parameters of LVNC

Holter (n=2)	
Normal	2
NT pro BNP median (range) pg/ml	2651 (227-35000)
Medications	
Digoxin	4
Furosemide	9
ACE-I	8
Aldactone	4
Beta blocker	8
Amiodarone	1
Warf	1
CMRI (n=6)	
LGE present	3 All subendocardial
LGE absent	3
NC : C ratio median (range)	2.8 (1.96-4.26)

Table 25 Other clinical characteristics of LVNC patients

Years of follow up mean	1.73 +/- 1.07 (0.4-3.0)
Death	4
Death from HF	3
SCD	0
Death from VT	1
Alive	3
Lost to follow up	4
Ventricular arrythmias	1
AV block	1
BBB	1
Requirement of ICD/ CRT / Transplant	4

Table 26 describes the outcome of LVNC patients. 4 were lost to follow up, 4 died and three were alive.First degree AV block was present in one patient , one had sustained ventricular arrythmias.

	Age of onset	Mode of presentation	Gender	Family H/O SCD	Family H/O CMP	Syncope	Consanguinity
Patient 1	10	DOE	M	Y	N	N	NCM
Patient 2	15	DOE	M	N	N	N	NCM
Patient 3	14	Right heart failure	M	N	N	N	CM
Patient 4	18	Clubbing	M	N	N	N	NCM
Patient 5	8	Right heart failure	F	N	N	N	NCM

Table 27 Clinical characteristics of the five pediatric AVC patients.

	ECG	ECHO	RV Angiography	Cardiac MRI
Patient 1	BAE , RBBB , T wave inversions in V1-V5 , Epsilon waves	RAE, Biventricular systolic dysfunction, Severe Tricuspid regurgitation, RV id z score 3.12	Dilated RV, RV dysfunction	Not done
Patient 2	RBBB , T wave inversions in V4-V6,II ,III , a VF , q in I , a VL,V5-V6	RAE , BV systolic dysfunction , RV Id z score 3.6	Dilated RV, RV dysfunction	Not done
Patient 3	RAD , ST depressions T wave inversions in V1-V5 II, III , a VF	RAE, RV dysfunction Severe Tricuspid regurgitation, RV id z score 2.9 , PLAX RVOT 20 mm /m2	Dilated RV	Not done
Patient 4	First degree AV block, RAD , Twave inversions V1-V4	Dilated RA/ RV, LV apex wrapped around RV	Not done	NO LGE , LV apex wrapped around RV apex, RV outpouching Dyskinetic RV
Patient 5	Junctional rhythm , RAD , I RBBB , T wave inversion in V1-V5	Mild Biventricular systolic dysfunction, Severe Tricuspid regurgitation, RV id z score4.0	Dilated RV and RVOT , Severe Tricuspid regurgitation, Small outpouching in apical region. Rved 12	Dilated and thin walled RV with fibrofatty infiltration , No LGE

Table 28 Other investigative findings in ARVC patients

	Holter	Ventricular arrhythmias	EP study	Treatment	Follow up	Outcome
Patient 1	VPC s	VT of RBBB morphology with inferior axis at 24 yrs of age	BB reentry VT Ablation done	Amiodarone Diuretics , Digoxin , Beta blockers	28	On Follow up
Patient 2	NSVT RBBB morphology	No sustained arrythmias	Not done	Diuretics Digoxin Envas	8	Lost to FU
Patient 3	Not done	No sustained arrythmias	Not done	Diuretics , digoxin	0.6	Lost to FU
Patient 4	Not done	No sustained arrythmias	Not done	No drugs	1	On Follow up
Patient 5	Junctional Tachy	None	Suprahisian and intrahisian conduction system disease. Atrial standstill	Diuretics , digoxin. PPI VVIR	3	On Folllow up

Table 29 Other pertinent findings characteristics and outcome of ARVC patients

	Age/Gender	Mode of presentation/History	Pertinent Findings	Follow up(yrs)	Outcome
Patient 1	5 m/M	Recurrent RTI No syncope No significant family history	Features of non compaction on Echo , NC: C ratio> 2 MRI- Dilated RA with preserved biventricular systolic function Subendocardial enhancement in hypertrabeculations of RV Cardiac cath- Increased RVEDP with Mild PAH	6	Alive

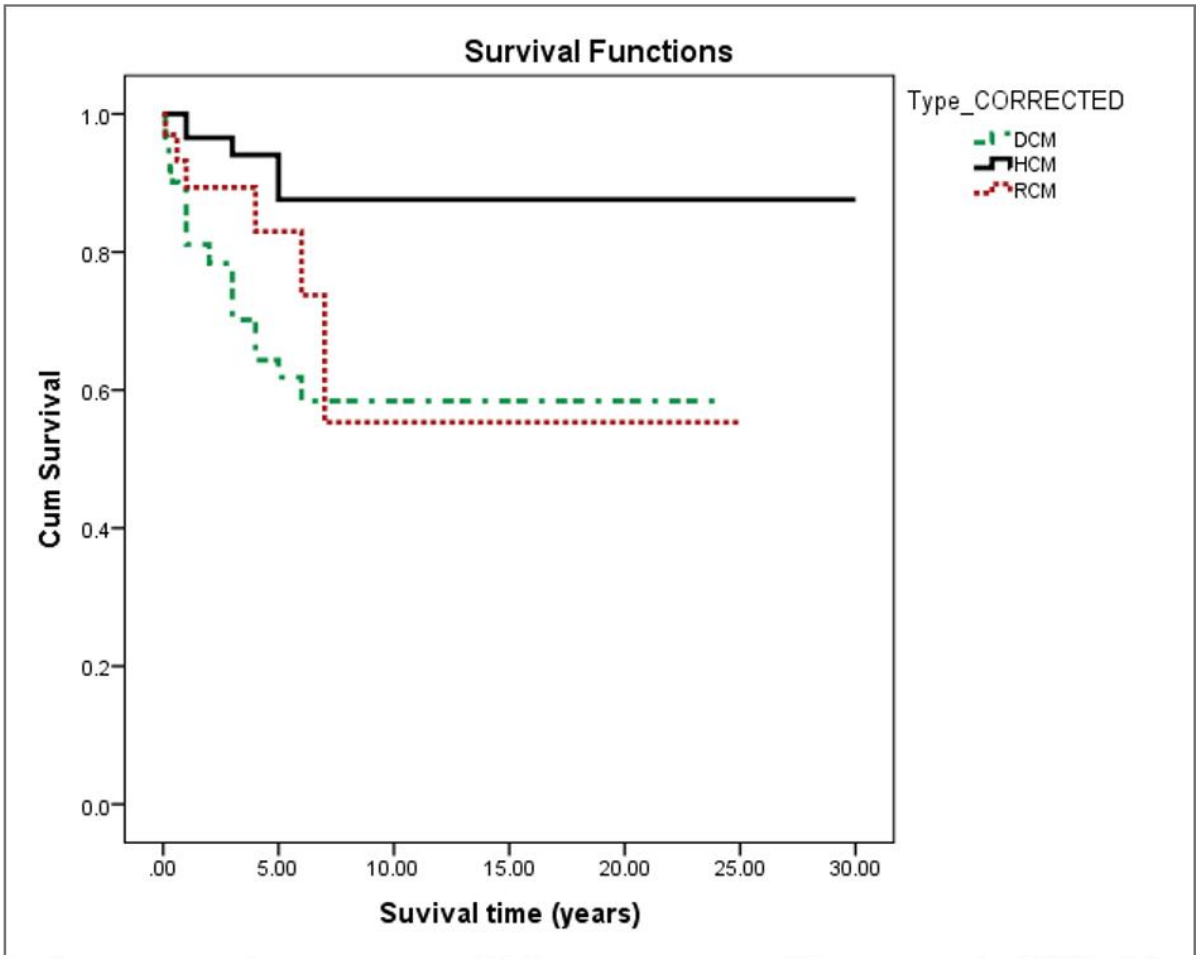
Table 30 showing findings of patient of RV Non compaction.

	Age/Gen	Mode of presentation/History	Pertinent Findings	Follow up(yrs)	Outcome
Patient 1	5y/M	Palpitation Syncope+ No significant family history	Holter- Long sinus pause 6.4 s Normal BV systolic function on echo MRI- Preserved biventricular systolic function, LGE in subendocardium of interatrial septum, RA wall,Posteromedial papillary muscle EPS- CTI dependent CCW typical atrial flutter Underwent PPI VVI	4	Alive Asymtomatic

Table 31 showing findings of patient of Possible atrial cardiomyopathy.

HCM, 16 out of 63	Frequency
Syndromic	15
Noonan Syndrome	9
Pompe disease	1
Freidrichs ataxia	1
Childhood onset myoclonus dystonia	1
Unclassified	3
Genetic	
MYBPC3	1
RCM, 3 out of 33	
Emery dreifuss muscular dystrophy	1
Genetic mutation	
TNNI3	1
MYL3	1
DCM, 8 out of 119	
Duchenne muscular dystrophy	1
Becker muscular dystrophy	1
Mitochondrial cytopathy	2
MPS Type 6	1
Kelnifelters syndrome	1
Carnitine defeciency	1
Unclassified	1
LVNC,3 out 0f 9	
Genetic mutation	
PRKAG2	1
SCN5A	1
Primary carnitine deficiency(SLC22A5 gene mutation)	1
Total	30

Table 30 showing the syndromic , metabolic or genetic diseases identified in the three subgroups.Only 30 out of 232 patients (13%) had syndromic , genetic or metabolic disease.



Kaplan Meir curve showing survival in the three major types of cardiomyopathy patients. The Kaplan meir survival curve for the three types of cardiomyopathies reveals 5 year survival rate of 89% in HCM, 82% in RCM and 61% in DCM. Overall survival was worse for RCM cohort , but 5 year survival was worse for DCM patients.

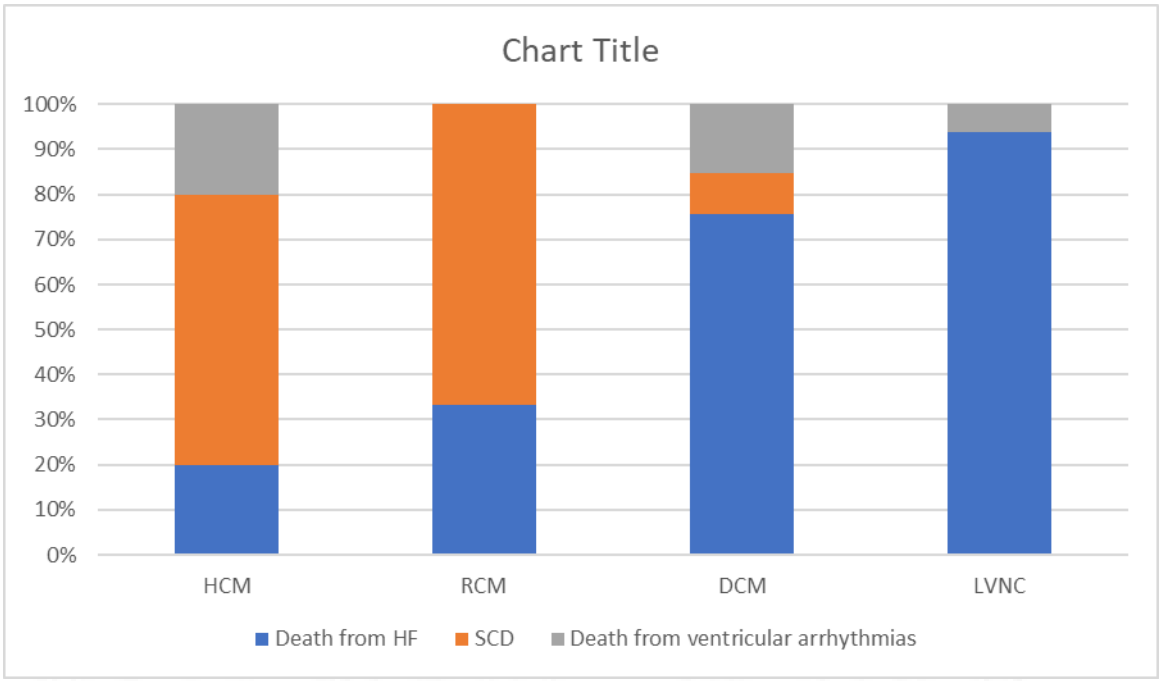


Chart depicting the cause of death in the four types of cardiomyopathies.SCD was most common in RCM.

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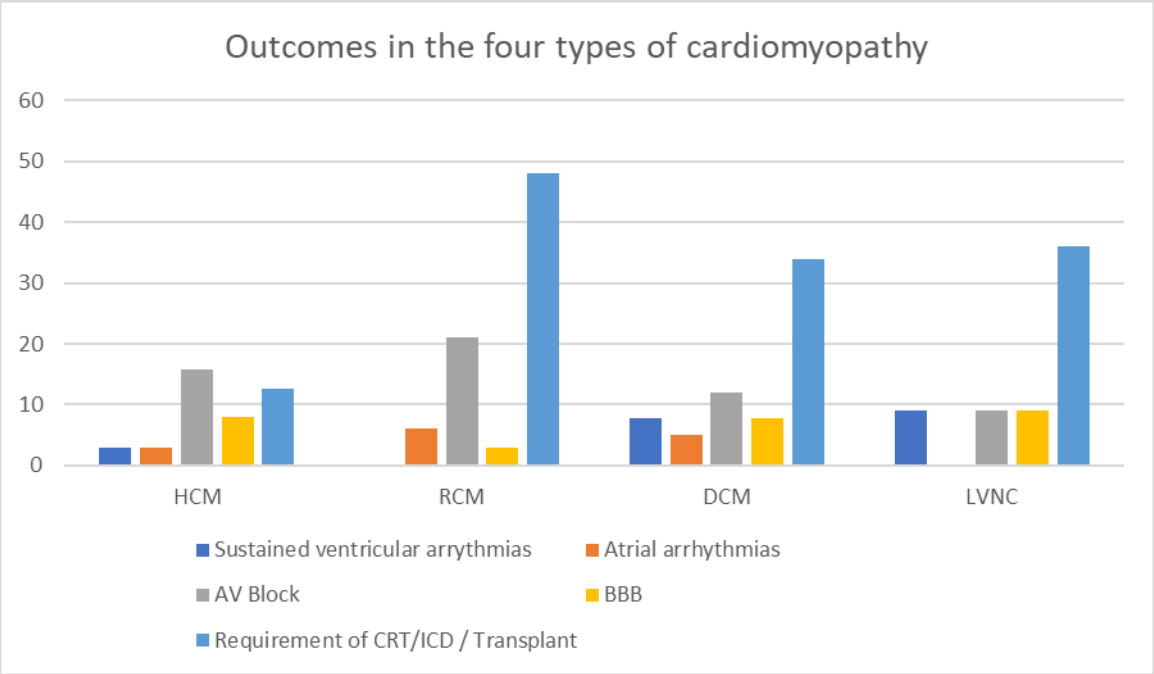
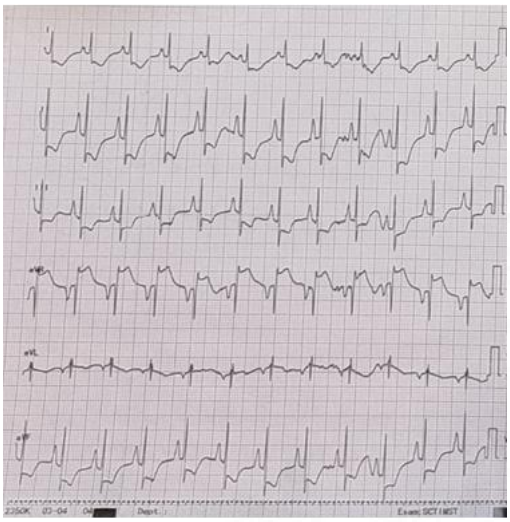


Chart depicting outcomes in different types of cardiomyopathies. The incidence of ventricular arrythmias, atrial arrhythmias, AV Block, BBB and requirement of CRT,ICD and transplant is shown. There were no ventricular arrythmias in RCM patients and no atrial arrhythmias in LVNC patients

PICTURES

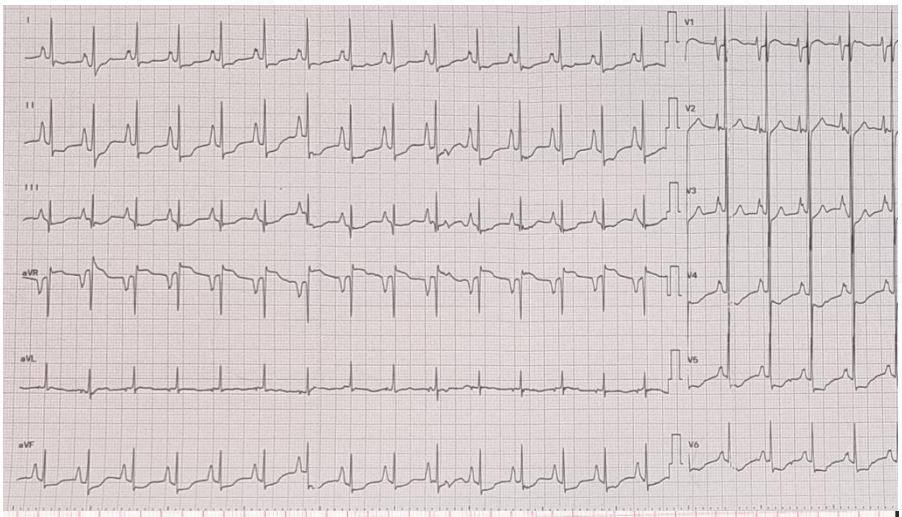


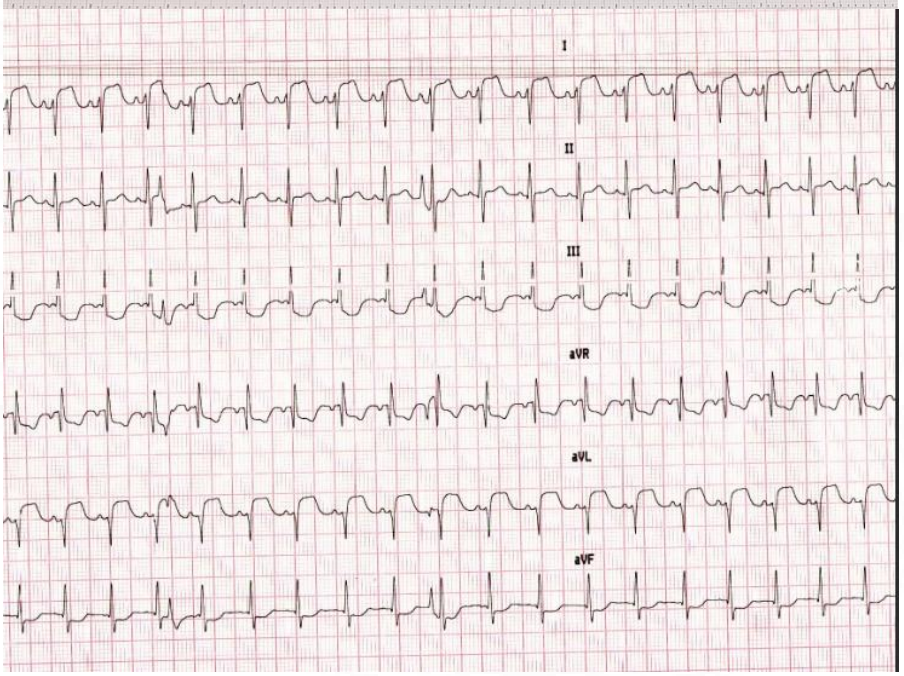
Representative ECG of three of our RCM patients

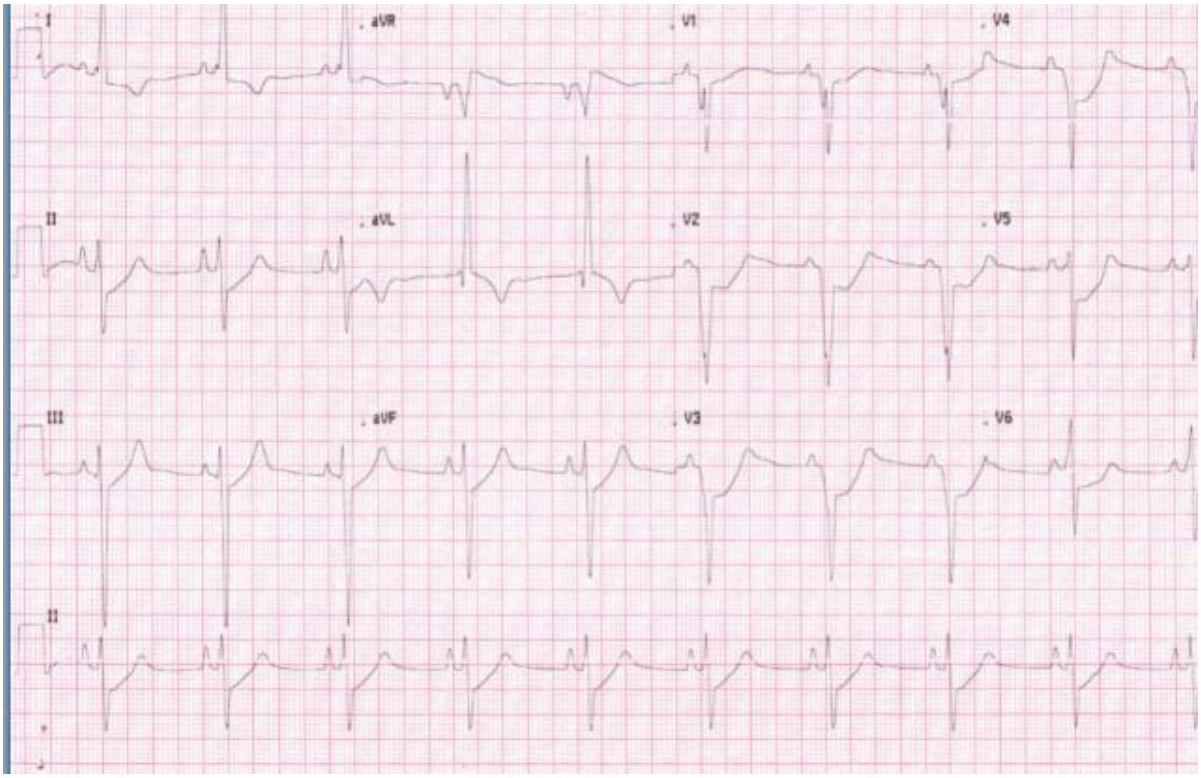
A: ST depression in I, II , III , a VF with ST elevation in a VR

B: Tall P waves signifying RAE with ST depression in II,III,a VF and ST elevation in a VR

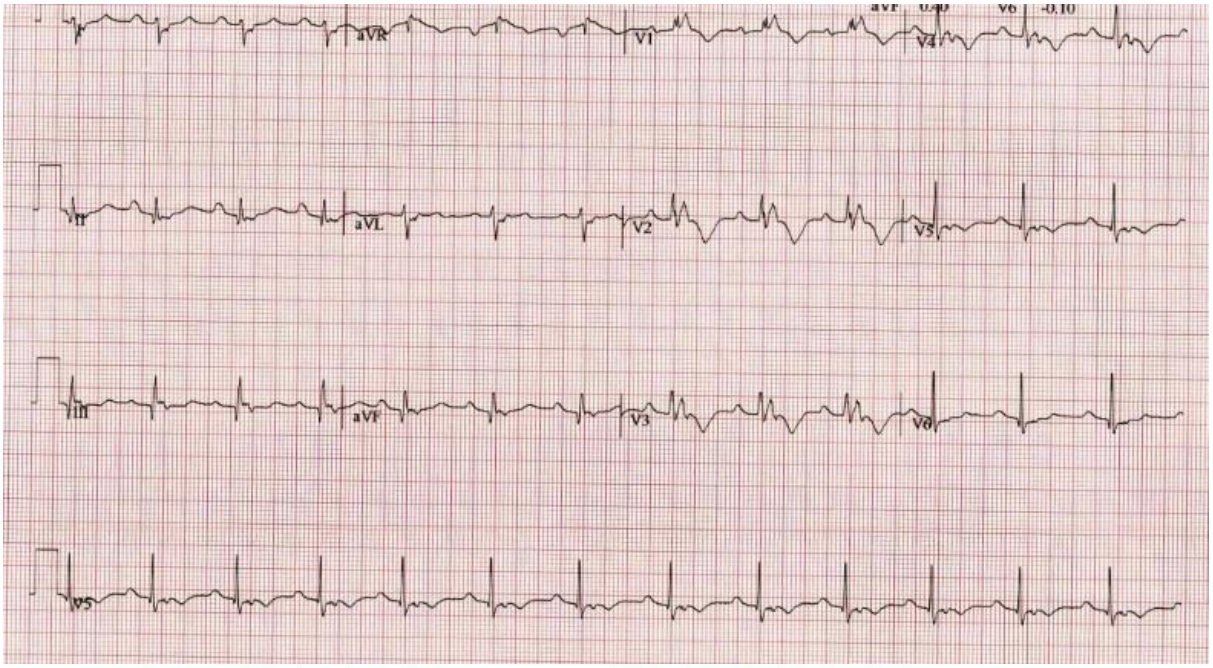
C: Another patient with ST elevation in I , Avl with ST depression in III, a VR and a VF



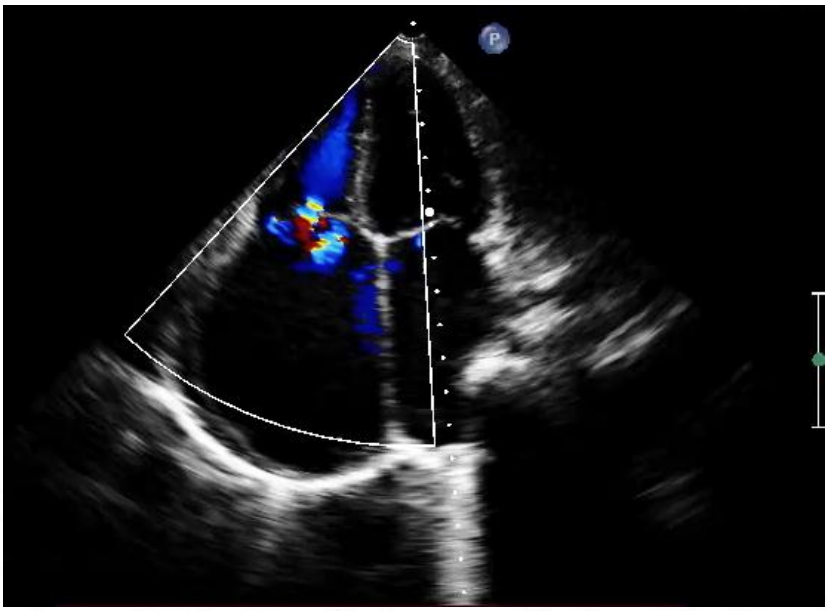




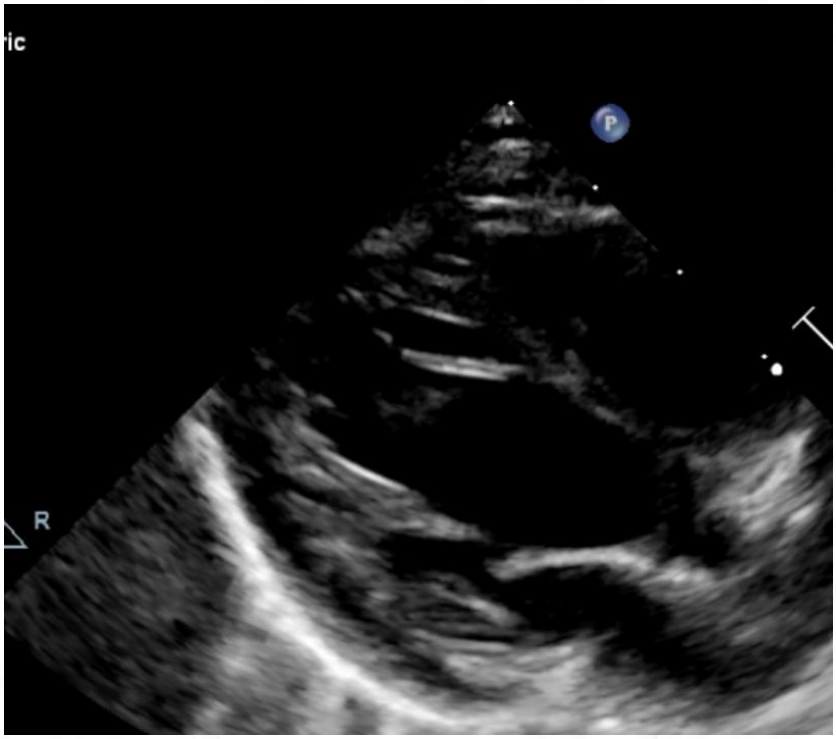
ECG of one of our HCM patients showing q in aVL and ST depression in V2-V6



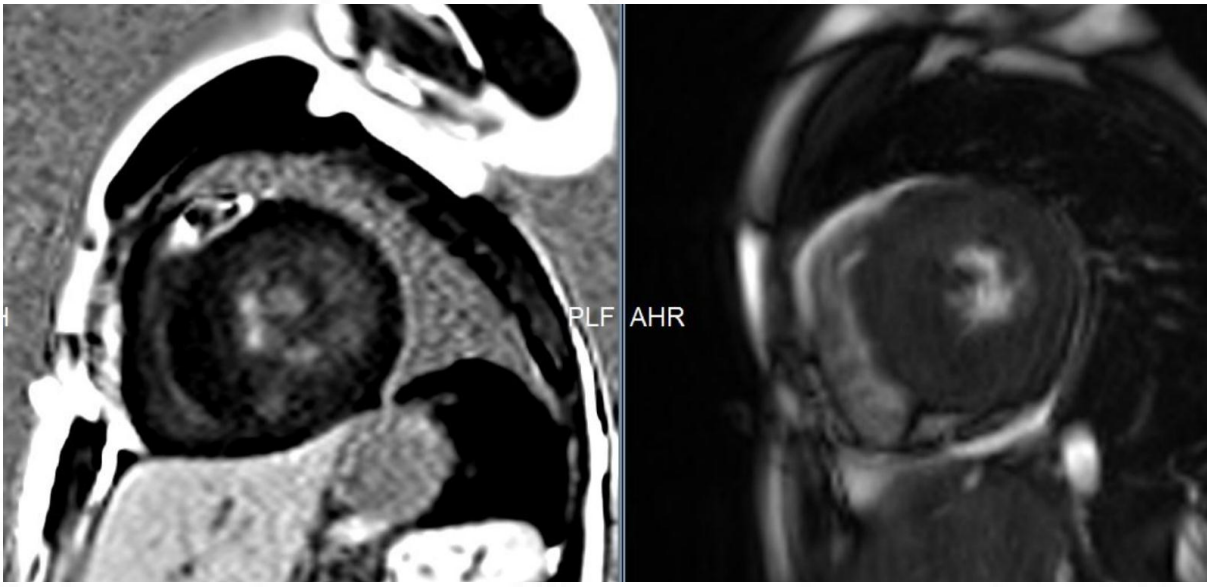
ECG of one of our ARVD patients showing repolarization abnormalities in form of T wave inversion in anterior precordial leads and prolongation of the terminal QRS indicating depolarization abnormalities.



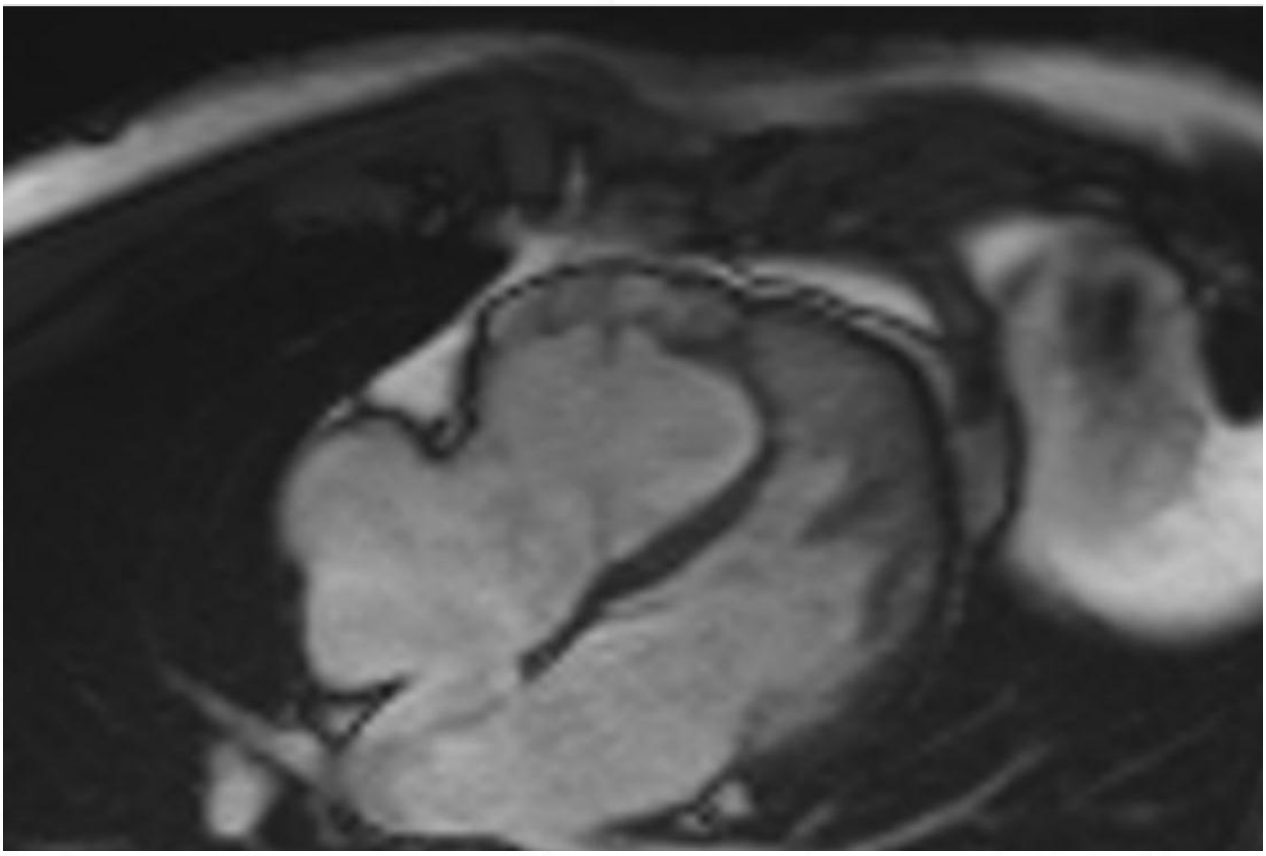
Echo image of one of our RCM patients showing biatrial enlargement and Tricuspid regurgitation



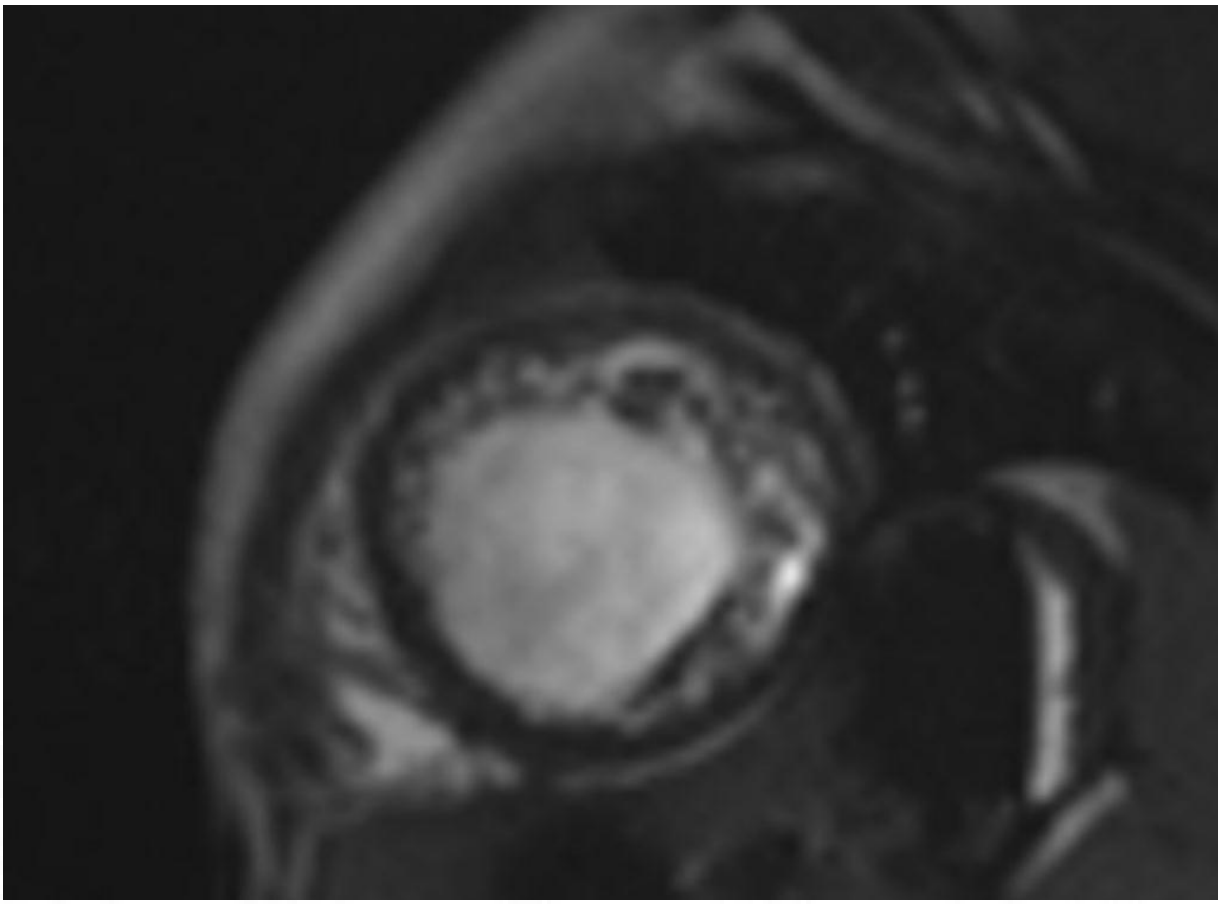
Echo image of one of our LVNC patients showing left ventricular non compaction.

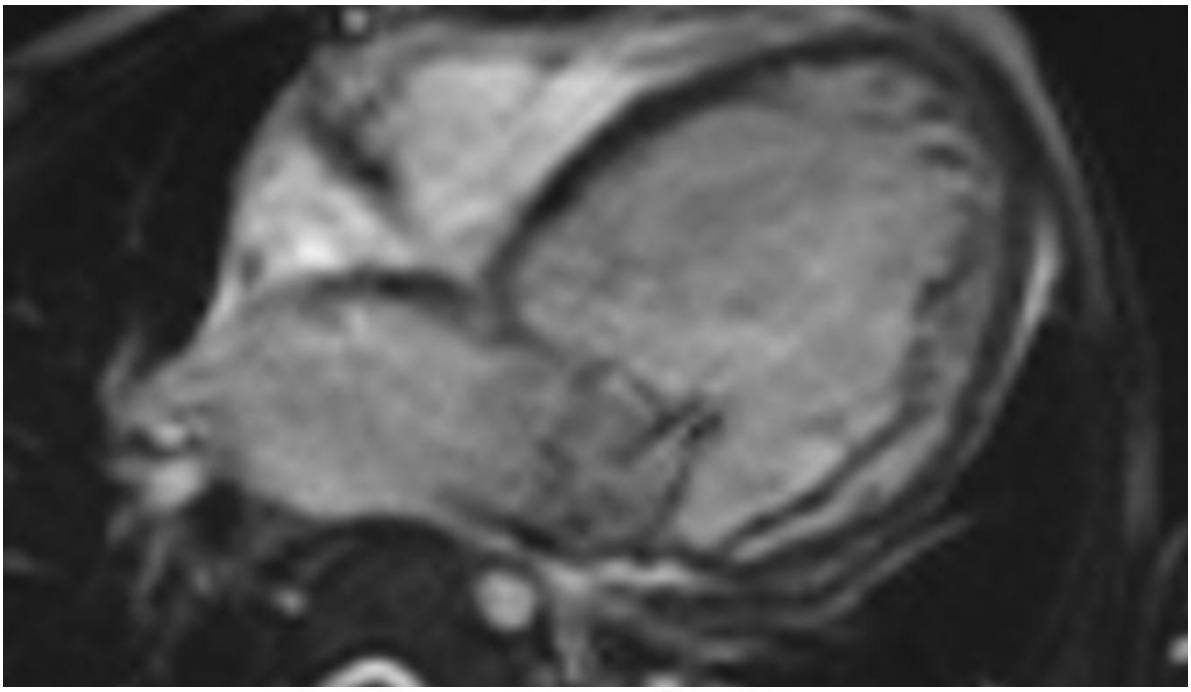


Short axis contrast enhanced images showing late gadolinium enhancement in lateral and inferior walls

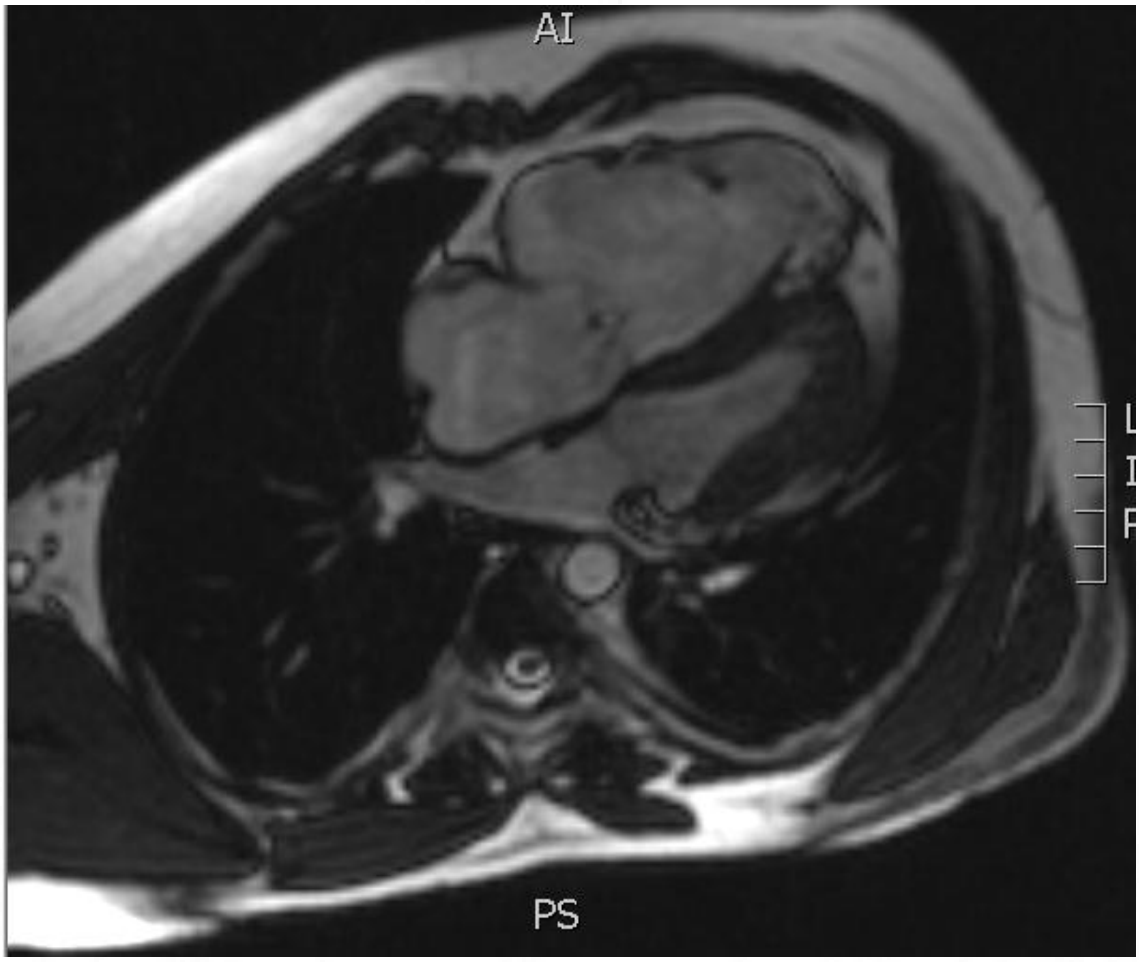


Four chamber CMR images of one of our AVC patients showing dilated RA/RV with left ventricular apex wrapping around the right ventricle.

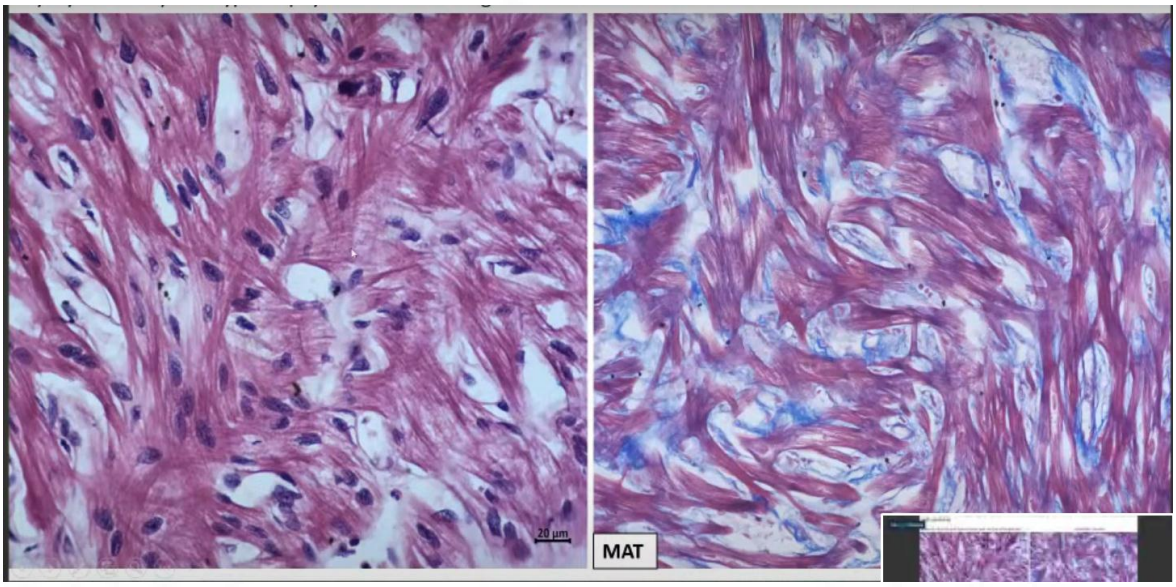




Short axis and four chamber SSFP CMR images showing Left ventricular non compacted segments.



CMR Images of one of our AVC patients showing dilated RA/RV with small outpouching seen in lateral RV wall



Histopathological findings in one of our RCM patients showing myocardial disarray and hypertrophy and nuclear enlargement with interstitial fibrosis.

SUMMARY

SUMMARY

- All patients with cardiomyopathy who presented to us between 1990 to December 2020 and were younger than 18 years of age were enrolled in the study. The available medical records of each enrolled patient was reviewed. This retrospective cohort of children was prospectively followed up to study their outcome.

- Specific outcomes to be studied were Death from Heart failure, SCD, Ventricular arrhythmias, AV block and Requirement of CRT/ICD/Transplant.

- During the thirty year study period (1990-2020), 233 cases of pediatric cardiomyopathy were identified. 119 cases of dilated cardiomyopathy (51 percent), 63 cases of hypertrophic cardiomyopathy (27 percent), 33 cases of restrictive cardiomyopathy (14 percent), and 11 cases of left ventricular noncompaction cardiomyopathy (4.7 percent).Besides this , five cases of ARVD , One case each of RVNC and Atrial cardiomyopathy were identified.

- 12% of HCM patients presented in the neonatal period and 37% presented in the first year of life. None of the RCM patients presented in the neonatal period and only 12% of RCM patients presented in the first year of life. Among the DCM patients, 8.7% presented in the neonatal period and 37% in infancy. Among the 11 cases of LVNC, one presented in neonatal period and nine cases in infancy (81%).

- The median age of presentation of HCM patients was 7 yrs (Range 0.1-18 yrs). The median age of presentation of DCM patients was 2 yrs (Range 0.1-18 yrs). and of RCM patients was 6 yrs (Range 0.8-16 yrs).

- Among all cardiomyopathies, males were more commonly diagnosed with cardiomyopathy than females. 142 out of 232 cases were males(61%).
- Family history of cardiomyopathy was most common in HCM patients(29%) and least common with RCM patients(6%). Family history of SCD was also most common in HCM patients(22%).None of the RCM patients had family history of SCD.
- Among the 63 HCM patients, sixteen patients had proven syndromic, metabolic or genetic disease.(25%). LV outflow obstruction was present in 32 patients(50%).
- Noonan syndrome was present in 9 of the 63 patients of HCM(14%)
- Dyspnea on exertion was the most common mode of presentation. Syncope developed at any time during the course of follow up in 28%
- Out of the 26 patients who had Cardiac MRI done,16 patients had LGE, out of which three died or had ventricular arrythmias and none of the patients with no LGE faced death or ventricular arrhythmias but the relationship was not significant (P value = 0.26)
- Out of the 16 patients with LGE, two patients had LGE > 25%.
- In the HCM subgroup, over a mean follow up period of 5.6 years (0.1-30 years), twenty seven were lost to follow up(42%). Out of the rest, five died (13%) and thirty one (86%) were alive.This gives 5 year survival rates of 89%.
- Three patients had sudden cardiac death, one died from heart failure and one died after recurrent ventricular arrythmias.

- First degree AV Block was present in 15% of HCM patients. Bundle branch blocks(BBB) in five patients(8%). Only two patients had sustained ventricular arrhythmias,two had atrial arrythmias.
- Eight patients required CRT/ICD or transplant.(12.7%)
- Two patients underwent septal myectomy and both are alive and asymptomatic after 24 yrs and 8 yrs of follow up respectively. Two patients underwent ICD both for primary prevention and one underwent PPI for distal AV conduction disease.
- Among the 33 RCM patients, Dyspnea on exertion was the most common mode of presentation. Syncope developed at any time during the course of illness in 3 patients(9%).
- ST depression was present in 7 patients(n=14)
- Over a mean follow up period of 3.7 years (0.1-25 years), nineteen were lost to follow up(57%).Out of the patients on follow up, six died(43%)and eight (52%) were alive.
- Out of the six patients who died, four died of sudden cardiac death,two from heart failure.So, the most common cause of death was SCD.
- This gives a 5 year survival rate of 82%, although RCM had the worst overall survival.
- None of the patients had sustained ventricular arrythmias and atrial arrythmias were seen in two patients, two were in junctional rhythm.
- First degree AV block was seen in 7 patients(21%) as compared to 15% in HCM patients.Bundle branch blocks(BBB) in two patients(3%).

- Sixteen patients required ICD/CRT or transplant(48%).
- Only relationship between syncope and death was found to be close to statistically significant(P=0.07).
- Electrophysiological study was done in one patient showing atrial paralysis for which permanent pacemaker implantation was done.
- Out of the 119 DCM patients , Dyspnea on exertion was the most common mode of presentation H/O Consanguinity was known in 106, out of which 18 patients (16%) had history of parental consanguinity. Syncope developed at any time during the course of illness in 3 patients (2.5%) as compared to 28% in HCM and 9% in RCM.
- 8 patients had proven syndromic, metabolic or genetic disease.
- Among 119 patients, 27 were suspected to have post viral myocarditis on the basis of association with a known viral illness. Viral identification was available only for three patients
- 47% patients were on beta blockers and 72% were on ACE-Inhibitors showing that a reasonably good percent of our population were on beta blockers. The practice of prescribing ARNI started recently and thus only 4 patients were on ARNI.
- Among the DCM subgroup, over a mean follow up of 2.98 years, 45 were lost to follow up (37%). Among the remaining 74 patients,33 died (45%), 27 recovered (36%) and 14 had persistent LV dysfunction (19%).
- Among the thirty patients who died, 25 died form Heart failure, 3 had SCD and 5 died after ventricular arrhythmias.
- The 5 year survival rate of DCM of 61%

- (11.9%) had first degree AV block which is the least among all three types. BBB was present in 9 patients (7.7%) and among them LBBB was present in only 2 patients.
- 9 patients had sustained ventricular arrythmias (7.7%) which is more than in HCM (3%) or RCM patients(none).Atrial or junctional arrythmias were seen in 6 patients.
- Requirement of ICD/CRT/Cardiac transplant was considered indicated in 41 patients (34%). 1 patient underwent ICD and two underwent CRT. Electrophysiological study was done in 1 patient. 1 patient had required ECMO.
- Among the 11 cases of left ventricular non compaction cardiomyopathy were identified, none of the patient had associated significant congenital heart disease.
- Dyspnea on exertion was the most common mode of presentation. History of syncope was present in only 2 patients.
- The median NC:C ratio was 2.8.
- Over a mean follow up of 1.73 years, 4 were lost to follow up, 4 died and 3 were alive.
- Three died from Heart failure, 1 died from Ventricular arrhythmias and none of the patients had SCD.
- One patient had sustained ventricular arrythmias and one patient had first degree AV block and one had BBB.

- Among the rarer types, five cases of pediatric ARVD were recognized. First case diagnosed as definite ARVD.Patient 2 and Patient 3 fulfilled borderline ARVD.Patient 4 and patient 5 were diagnosed as Possible ARVD.
- Only 1 patient had documented ventricular tachycardia of RBBB morphology and one patient had junctional rhythm with suprahisian and infrahisian cinduction system disease.One patient presented wirh clubbing.
- One case of Right ventricular non compaction amd one case of probabale atrial cardiomyopathy were reported.

DISCUSSION

To our knowledge , this is the largest study on pediatric cardiomyopathy from South Asia as of now. We studied the clinical profile and outcome of cardiomyopathy in children less than 18 years of age. Among the 233 cases of cardiomyopathy identified, 119 cases were dilated cardiomyopathy (51 percent), 63 cases were hypertrophic cardiomyopathy (27 percent), 33 cases were restrictive cardiomyopathy (14 percent), and 11 cases were left ventricular noncompaction cardiomyopathy (4.7 percent). Besides this, five cases of ARVD, one case each of RVNC and atrial cardiomyopathy were identified.

The frequency of different types of cardiomyopathies is similar to other studies with DCM found to be most common(51%) followed by HCM(27%) and then , RCM(14%). RCM was found to be more frequent(14%) in our study than in studies conducted in rest of the world. Lipshultz et al reported the incidence of RCM and other specified cardiomyopathies to be only three percent.(16). In the study by Nugent et al, the incidence of RCM was 2.5%. LVNC was less common in our study (4.7%) than in the study conducted by Nugent et al (9.2%)(3). The high incidence of RCM patients may reflect the high incidence of EMF seen in this part of the world. Five patients had histopathological diagnosis of EMF. Four had proven EMF on biopsy, one had EMF diagnosed after autopsy, although only 7 out of the total 33 RCM patients underwent endomyocardial biopsy.

12% of HCM patients presented in the neonatal period and 37% presented in the first year of life. None of the RCM patients presented in the neonatal period and only 12% of RCM patients presented in the first year of life. Among the DCM patients, 8.7% presented in the neonatal period and 37% in infancy. Among the 11 cases of LVNC, one presented in neonatal period and nine cases in infancy (81%).

Although classically HCM was thought not to present in infancy, in our study 12% of HCM patients presented in the neonatal period and 37% presented in the first year of life. Earlier studies did not specifically mention the percentage of cases which were diagnosed in neonatal period. The median age of presentation of HCM patients was 7 years. This is almost similar to the study by lipshultz et al who also reported a high incidence of HCM in infancy and the median age of presentation of HCM patients was reported to be 5.9 years(16). The high percentage of HCM in infancy in our study may represent metabolic causes of HCM.Even though, Nugent et al excluded patients with multisystem metabolic causes of cardiomyopathy, still they found more than 50% of HCM patients to be diagnosed in infancy and the median age of presentation of HCM patients to be unusually early, around 5.7 months.(3). This may represent the cases that have been diagnosed as a result of screening.

None of the RCM patients presented in the neonatal period and only 12% of RCM patients presented in the first year of life. Nugent et al found no cases of RCM patients in infancy in their study.Median age of presentation of RCM patients was 36 months in their study.(3)

The median age of presentation of HCM patients was 7 yrs (Range 0.1-18 yrs). The median age of presentation of DCM patients was 2 yrs (Range 0.1-18 yrs). and of RCM patients was 6 yrs (Range 0.8-16 yrs).

Among all cardiomyopathies, males were more commonly diagnosed with cardiomyopathy than females. 142 out of 232 cases were males(61%).Of the 119 children with dilated cardiomyopathy, 69% were males (57.0 percent), as were 38 of the 63 children with hypertrophic cardiomyopathy (68 percent), 24 of the 33 with restritive cardiomyopathy (72 percent), and 5 of the 11 with LV noncompaction cardiomyopathy(45%). Most of the other

studies have found cardiomyopathy to be more common in males except Nugent et al who found DCM to be more common in girls(56%).(3)

Family history of cardiomyopathy was most common in HCM patients(29%) and least common with RCM patients(6%). Family history of SCD was also most common in HCM patients(22%).None of the RCM patients had family history of SCD.

The Kaplan meir survival curve for the three types of cardiomyopathies reveals 5 year survival rate of 89% in HCM, 82% in RCM and 61% in DCM. Although the 5 year survival rate is worse for DCM, RCM has the worst overall survival of the three types followed by DCM and then, HCM. This is in accordance with other studies. Probabilities of freedom from death at 5 years after diagnosis of cardiomyopathy in the PCMR were as follows: Pure HCM: 90%, All RCM: 71%; pure DCM : 78%.(26)

Among the 63 HCM patients, sixteen patients had proven syndromic, metabolic or genetic disease(25%). LV outflow obstruction was present in 32 patients(50%).This is more than what has been reported in adult HCM patients(25%)(44) and almost similar to the pediatric studies conducted so far. In the study by Nugent et al, 40% patients had LV outflow obstruction and in the study by Yetman et al,59% had LV outflow obstruction.

Noonan syndrome was present in 9 of the 63 patients of HCM(14%). Out of the nine patients, three presented in infancy(33%). Six of the nine patients were females. Two patients had associated pulmonary stenosis and one underwent balloon pulmonary valvuloplasty for severe valvular pulmonary stenosis. Outcome was known in 5 out of the 9 patients and all are alive over a mean follow up of 7 years.One underwent septal myectomy and is doing well after 24 years of follow up.We have not compared the HCM patients with NS with those without as our numbers are small. Wilkinson et

al compared data in 74 children with NS and HCM and 792 children with idiopathic or familial isolated HCM. (45). Children with NS were diagnosed with HCM before 6 months old more often (51%) than children with HCM (28%) and were more likely to present with congestive heart failure (CHF) (24% vs 9%).Patients with NS with HCM have a worse risk profile at presentation compared with other children with HCM, resulting in significant early mortality (22% at 1 year).One patient with MYBPC3 mutation positive was a 15 year old boy a case of myoclonus dystonia with a family history of cardiomyopathy and sudden cardiac death,who had diffuse subendocardial enhancement on C MRI. Myoclonus dystonia has never been previously described in MYBPC3 positive HCM patients. Preexcitation was present in two of the HCM patients. First degree AV Block was present in 15% of HCM patients.Only two patients had sustained ventricular arrhythmias.

In the HCM subgroup, over a mean follow up period of 5.6 years (0.1-30 years), twenty seven were lost to follow up(42%). Out of the rest, five died (13%) and thirty one (86%) were alive.This gives 5 year survival rates of 89% which is comparable to other studies.Nugent et al reported a 5 year survival of 83%(3) Maruizi et al reported a 5 yr survival of 95%.(22). Three patients had sudden cardiac death, one died from heart failure and one died after recurrent ventricular arrythmias.

Although our numbers are small, we tried to analyze various postulated predictors of death and/or ventricular arrythmias. Out of the 26 patients who had Cardiac MRI done,16 patients had LGE, out of which three died or had ventricular arrythmias and none of the patients with no LGE faced death or ventricular arrhythmias but the relationship was not significant (P value = 0.26).Out of the 16 patients with LGE, two patients had LGE > 25%. None of the variables studied had a significant association with death or

ventricular arrhythmias.(Table)This may be due to the high percentage of patients lost to follow up.

Among the 33 cases of RCM, 21% had first degree AV block as compared to 15% in HCM patients. ST depression was present in 7 patients(n=14).q waves were present in 11 patients. BBB was present in 2 patients. ST depression in RCM patients is a marker of myocardial ischemia and a predictor of sudden cardiac death. Rivenes et al looked at risk factors for sudden death in a cohort of 18 patients with RCM; chest pain and syncope were identified as risk factors for sudden death. (28). Histopathologic evidence for ischemia was found in the majority of patients who died and Holter and ECG evidence of ischemia that is, ST-segment depression or T-wave inversion in the inferior, lateral, or lateral precordial leads (or a combination) predicted death within several months.Walsch et al studied the conduction abnormalities in 16 pediatric patients with RCM.(27). The median PR interval and the QRS duration were significantly longer in those who had an acute cardiac event compared to those who did not. He concluded that bradycardic events represented a significant portion of all arrhythmic events in such patients. One of the patients was a 3 year old child, product of consanguinity, had family history of cardiomyopathy in cousin and past history of atopic dermatitis. He presented with heart failure with hyperpigmentation of lips and failure to thrive. He had syndactyly of second and third left toe and had peripheral eosinophilia. He had recurrent acute cardiac events and ECG evidence of myocardial ischemia. On genetic evaluation, MYL3 gene mutation (p.A154C) was found positive. He died within 6 months of presentation of a sudden cardiac arrest. Autopsy conducted showed enlarged heart especially biatrial enlargement. Pathological findings included coagulative necrosis of myocytes with neutrophil predominant infiltrate suggesting myocardial ischemia, myocyte

hypertrophy and myocardial disarray with myocardial bridging.There was no evidence of thromboembolism.

The patient with TNNI3 gene mutation positive was a 21/2 year old male child , product of non consanguinity and no family history of cardiomyopathy who presented with right heart failure.ECHO showed glistening appearance of the myocardium, mild pericardial effusion and other features of restrictive cardiomyopathy.Cardiac MRI showed late gadolinium enhancement in interatrial septum, mitral valve and tricuspid valve but not in the ventricular myocardium. Gene analysis revealed heterozygous A170T mutation in the TNNI3 gene. He was FC III on last follow up 3 yrs after onset and was lost to follow up subsequently.

In the RCM subgroup, over a mean follow up period of 3.7 years,19 were lost to follow up (57%).Out of the rest , 6 died (43%) and 8 were alive(57% of the patients in whom outcome was known).The most common cause of death was sudden cardiac death(4 out of 6 patients).This gives a 5 year survival rate of 82%, although RCM had the worst overall survival. The high percentage of patients who have been lost to follow up has definitely overestimated the 5 year survival. PCMR reported the 5 year survival of all RCM to be 71%.The possible risk factors were studied with relation to their association with death or ventricular arrhythmias. Only relationship between syncope and death was found to be close to statistically significant.Rivenes et al also reported syncope and chest pain to be associated with sudden death.(28).

Among the 119 DCM patients, only 3 patients had syncope (2.5%) as compared to 28% in HCM and 9% in RCM.9 patients had proven syndromic or metabolic disease. Syndromic or metabolic disease must have been underestimated as all patients did not undergo screening for the same. One

patient had Duchennes muscular dystrophy gene positive and one had Beckers muscular dystrophy gene positive.Two patients had suspected mitochondrial disorder.One patient with a positive family history of DCM in other sibling presented in the neonatal period with feeding difficulty. She was evaluated and found to have carnitine deficiency.Both siblings responded to carnitine supplementation. Diagnosing such disorders especially carnitine deficiency is absolutely essential as appropriate treatment can reverse the cardiomyopathy.

Preexcitation was present in 4 patients.One of them was a 2 year old child with an accessory pathway localized to right anteroseptal location with a documented orthodomic AVRT. Electrophysiological studies were not done as they were not willing for same. First degree AV block was present in 11.9% patients.This is the least among the three types.In RCM, 21% had first degree AV block and in HCM patients, 15% had AV block.

Among 119 DCM patients, 27 were suspected to have post viral myocarditis on the basis of association with a known viral illness. Viral identification was available only for three patients. One had Herpes zoster viral myocarditis and two had Cox sackie viral analysis positive. Diagnosing viral myocarditis on the basis of a temporal history of viral illness alone can lead to overestimation of the incidence of viral myocarditis.

47% patients were on beta blockers and 72% were on ACE-Inhibitors showing that a reasonably good percent of our population were on beta blockers. Only 5% of the pediatric patients enrolled in the American Pediatric Cardiomyopathy Registry received beta blockers in the 1990s compared with 18% after 2000. (46). Beta blocker use in pediatric population has been increasing in recent times. Although Shaddy et al in their study observed that carvedilol does not significantly improve clinical

heart failure outcomes in children and adolescents with symptomatic systolic heart failure, the study may have been underpowered and the effect may have been due to the inclusion of right ventricule and single ventricle physiology patients.(47). Digoxin was prescribed in the early periods of the study and the practice of prescribing digoxin fell in the later periods and ACE-I and beta blockers were more commonly prescribed in the later parts.

Among the DCM subgroup, over a mean follow up of 2.98 years, 45 were lost to follow up (37%). Among the remaining 74 patients,33 died (45%), 27 recovered (36%) and 14 had persistent LV dysfunction (19%). This is almost similar to other studies conducted so far. In the study by Alexander et al , normalization of LV function was attained in 33% of all subjects by 15 years after diagnosis.(31). Higher LVFS Z score at diagnosis, higher LVFS Z score during follow-up, and greater improvement in LVFS Z score were all predictive of an increased likelihood of normalization. Arola et al in their study observed that during a mean follow-up of 3.9 +/- 4.5 years (range, 1 day to 25 years), 10 patients (16%) recovered, 17 (27%) had residual disease, 4 (6.4%) underwent heart transplantation, and 31 (50%) died.(29). Patients who had normalization of LV function most likely would have viral myocarditis or have undergone reverse remodelling as an effect of medications.

The 5 year survival rate of DCM of 61% is almost similar to that published by most of the other studies.Alexander et al reported the 5 year survival of DCM to be 65% at 5 years among the NACCS participants. In the PCMR registry, the 5 year survival rate of DCM was 78%

9 patients had sustained ventricular arrythmias (7.7%) which is more than in HCM (3%) or RCM patients(none).Among the patient with sustained ventricular arrythmias, six died, one is alive who received an ICD and two

were lost to follow up.14 patients (11.9%) had first degree AV block which is the least among all three types. BBB was present in 9 patients (7.7%) and among them LBBB was present in only 2 patients.This brings attention to the lower incidence of LBBB in pediatric DCM as compared to adult DCM as has been proven in previously conducted studies.The study by Pugia et al reported on the natural history of dilated cardiomyopathy in children. Out of the 927 DCM patients, 47 were pediatric DCM (5.1%). LBBB was seen in 4.4% of pediatric DCM versus 31.9% of adult DCM.(48). The reason for this has been postulated to be due to the less severe disease at baseline and earlier diagnosis due to screening.None of the risk factors studied had a significant relationship with death or ventricular arrhythmias , however this is not reliable given the percentage of patients who were lost to follow up.

11 cases of left ventricular non compaction cardiomyopathy were identified (4.7%). None of the patient had associated significant congenital heart disease since it was an exclusion criteria in this study. Over a mean follow up of 1.73 years, 4 were lost to follow up, 4 died and 3 were alive.One patient presented at 1 year of age with recurrent episodes of refractory ventricular tachycardia.Cardiac MRI showed no late gadolinium enhancement and a noncompacted to compacted ratio of 2.4.Genetic analysis revealed a positive SCN5A mutation. He later succumbed to cardiogenic shock despite being treated with multiple antiarrythmics and cervical sympathectomy. SCN5A mutation has previously been described in patients with LV non compaction cardiomyopathy. Shan et al compared the frequency of SCN5A variants in LVNC patients with or without arrhythmias.The frequency of SCN5A variants was significantly higher in the patients with arrhythmias than those without (50% vs 7%)(49). Another patient was a 2 month old infant who presented with recurrent unresponsiveness. ECG showed left lateral accessory pathway and family

screening revealed noncompaction in a 7 yr old sibling with preexcitation. Genetic analysis revealed PRKAG2 mutation positive in the child and the sibling as well as in two of the other family members. PRKAG2 cardiomyopathy is classically described to be associated with hypertrophic cardiomyopathy, preexcitation and progressive conduction system disease.Non compaction has never been reported previously. It may be possible that the child evolves into a hypertrophic cardiomyopathy in the later stages of life as has been described in literature that LVNC can follow an "undulating phenotype" (33). Recently Hisham et al reviewed the Phenotypic expression and clinical outcomes in a South Asian PRKAG2 cardiomyopathy cohort.(50). He has studied 22 patients including adults with PRKAG2 cardiomyopathy.86% had left ventricular hypertrophy.77% had a WPW pattern on ECG. None of the cohort described had LV non compaction.

Five cases of pediatric arrhythmogenic ventricular cardiomyopathy were recognized. None of the studies on pediatric cardiomyopathy conducted so far have reported on Pediatric AVC separately. First case fitted two major of the Task force criteria, 2010 and was diagnosed as definite ARVD.Patient 2 and Patient 3 fulfilled one major and one minor criteria and were diagnosed as borderline ARVD.Patient 4 and patient 5 had one major criteria and were diagnosed as Possible ARVD.(51).Four out of the five patients were males. All except one presented in adolescence. Three patients had biventricular systolic dusfunction and two had right ventricular dysfunction.Twave inversions were not taken as diagnostic criteria in patients aged less than 14 yrs.Only the patient 1 had family history of sudden cardiac death.None of the patients presented with sudden cardiac arrest. Only patient 1 had ventricular tachycardia of RBBB morphology with inferior axis.Interestingly, one of our patients was brought to medical attention when

he was noticed to have clubbing on a routine visit.Such rare presentation has been previously reported in literature and is postulated to be due to the right to left shunt across a Patent foramen ovale due to increase in right ventricular end diastolic pressures. Patient 5 had intermittent junctional rhythm and Electrophysiological study revealed suprahisian and infrahisian conduction system disease.She required a VVIR pacemaker later for recurrent syncope.Though ARVD usually presents with ventricular arrhythmias, Bradyarrythmias have also been reported as the first manifestation due to the fibrofatty infiltration of the cardiac conduction system disease. (52). Deshpande et al (32) reviewed 16 cases of pediatric ARVD that were diagnosed through histopathology, genetic testing and task force criteria.They noticed that patient who had histopathology documented fibrofatty infiltration of the right ventricular myocardium would not have fulfilled the criteria of definitive diagnosis of ARVD if the adult task force criteria would have been used for diagnosing.They therefore recommended that the current criteria need further revision to encompass pediatric manifestations of ARVC/D.

We report one case of Right ventricular non compaction in a 5 month old infant who had features of RV non compaction with preserved biventricular systolic function. Cardiac MRI showed subendocardial enhancement in hypertrabeculations of RV. RV non compaction may be missed and underdiagnosed because of the difficulty in differentiating pathological noncompaction from the normal trabeculations of RV. It has been proposed that the dilation of the RV may be a helpful supportive feature for the diagnosis of isolated right ventricular noncompaction.(37)

Among the rare unclassified cardiomyopathy , we describe one case of probable atrial cardiomyopathy in a 5 year old child with recurrent syncope. Holter showed long sinus pauses and electrophysiological study revealed

CTI dependent typical atrial flutter.Cardiac MRI showed preserved biventricular systolic function and LGE in subendocardium of interatrial septum, RA wall, posteromedial papillary muscle.This case might represent an early form of atrial cardiomyopathy which has been previously described in literature by Williams et al who described a family where three of the five siblings had some form of cardiomyopathy characterized by first degree AV block and supraventricular rhythm which progressed to persistent atrial standstill over some years.

LIMITATIONS:

The major limitation of the study is the high attrition rate in all the subgroups. That is not surprising in a retrospective study conducted over a period of three decades.The difficulty of contacting patients who visited the hospital over the past three decades is obvious.Given the natural history of rare diseases like RCM that is known from previous studies we may have overestimated survival.

Another limitation of the study is the lesser number of genetic and metabolic abnormalities identified as the testing was not available for most of the study period and even now, few parents can afford the costs involved

Much more research needs to be done on pediatric cardiomyopathies especially from this part of the world. Genetic analysis needs to be provided for all parents so that we can risk stratify the patients.

CONCLUSION

DCM is the most common type of cardiomyopathy followed by HCM , RCM and then LVNC. RCM is more common in our population than all other studies conducted so far.

Outcome of HCM and DCM in our population is similar to other population cohorts , The better survival of RCM in our population may be due to the high percent of patients lost to follow up.

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APPENDIX

श्री चित्रा तिरुनाल आयुर्विज्ञान और प्रौद्योगिकी संस्थान, त्रिवेन्द्रम
तिरुवनन्तपुरम - ६९५०११, केरल, इंडिया
SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES AND TECHNOLOGY, TRIVANDRUM
Thiruvananthapuram - 695 011, Kerala, India
(An Institute of National Importance under Govt. of India)
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Institutional Ethics Committee
(IEC Regn No. ECR/189/Inst/KL/2013/RR-16)

SCT/IEC/1485 /NOVEMBER-2019 27.11.2019

Dr. Gousia Mukhtar
Resident
Department of Cardiology
SCTIMST, Thiruvananthapuram

Dear Dr. Gousia Mukhtar,

The Institutional Ethics Committee reviewed and discussed your application to conduct the study entitled "CLINICAL PROFILE AND OUTCOMES OF CARDIOMYOPATHY IN CHILDREN (IEC/1485)" on 5th November, 2019.

The following documents were reviewed:

Original submission

1. *Covering Letter addressed to the Chairperson, IEC, SCTIMST dated 24.09.2019 with checklist*
2. *TAC Approval Letter*
3. *IEC Application Form*
4. *Project Proposal*
5. *Information Sheet and Consent Form in English and Malayalam*
6. *Child Assent Form for children under age 12 in English and Malayalam*
7. *Proforma*
8. *CV of Principal Investigator and Co-Principal Investigators*

Revised submission

1. *Covering Letter addressed to the Chairperson, IEC, SCTIMST dated 18.11.2019 with checklist*
2. *TAC Approval Letter*
3. *IEC Application Form*
4. *Project Proposal*
5. *Information Sheet and Consent Form in English and Malayalam*
6. *Child Assent Form for children under age 12 in English and Malayalam*
7. *Proforma*
8. *CV of Principal Investigator and Co-Principal Investigators*

The following members of the Ethics Committee were present at the meeting held on 5th November, 2019 at G. Parthasarathi Board Room, AMCHSS, SCTIMST

SL. No.	Member Name	Highest Degree	Gender	Scientific /Non Scientific	Affiliation with Institution(s)
1.	Dr. R V G Menon	M Tech, PhD	Male	Lay Person (Chairman)	No
2.	Dr. Kala Kesavan. P	MBBS, MD	Female	Basic Medical Scientist	No
3.	Dr. K R S Krishnan	M.E., Ph.D.	Male	Medical Technology	Yes
4.	Dr. Harikrishna Varma PR	Ph.D(Materials Science)	Male	Medical Technology	Yes
5.	Dr. S S Giri Sankar	LL.M. Ph.D.	Male	Legal Expert	No
6.	Dr. V. Raman Kutty	M D, M Phil, M P H	Male	Health Sciences Expert/Clinician	Yes
7.	Dr. Aneesh V Pillai	BA. LLB (Hons.), LLM, Ph. D, SET (Law)	Male	Legal Expert	No
8.	Smt. Sathi Nair	MA (English Literature)	Female	Lay Person	No
9.	Dr. P. Manickam	BSMS, MSc (Epid).,PhD	Male	Health Science Expert/ Social Scientist	No
10.	Dr. Harikrishnan S	MD, DM (Cardiology) DNB (Cardiology)	Male	Clinician	Yes
11.	Mr. Satheesh Chandran	MSW, PGDPM	Male	Lay person/ NGO/ Social Scientist	No
12.	Dr. Christina George	MD Psychiatry	Female	Clinician	No
13.	Dr. Mala Ramanathan	PhD	Female	Social Scientist (Member Secretary)	Yes

IEC Decision

The IEC approved the conduct of the study in the present form.

Remarks:
The Institutional Ethics Committee expects to be informed about the progress of the study, any SAE occurring in the course of the study, any changes in the protocol and patient information/informed consent and asks to be provided a copy of the final report.

There was no member of the study team who participated in voting / decision making process. The ethics committee is organized and operated according to the requirements of Good Clinical Practice and the requirements of the Indian Council of Medical Research (ICMR).

Sincerely,

Mala Ramanathan
Member Secretary, IEC

PROFORMA :

1.Name

2.Age at onset

3.Sex

4.Weight

5.Height

6.Hospital no.

7.Mode of presentation / History

8.Past History

9.Consanguinity

10.Family history of cardiomyopathy and of SCD

11.Syndromal associations

12. Clinical Examination : CVS

13. Any other relevant finding

Investigations :

14.CXR measurements (CTR , LAE , RAE, PVH)

15.ECG measurements (PR interval, QRS d , Axis, QT interval, QT dispersion, Presence of preexcitation)

16.ECHO parameters (EF , LAE, RAE , LVEDD , PWT , IVS thickness , SAM , PAH , MR , LV AO gradient , BAE , Pericardial effusion)

17.Holter monitoring results

18.NT pro BNP

19.Cardiac MRI findings (EF , LGE)

20.Cardiac cath calculations if done

21. Results of metabolic or genetic tests if done

22. Outcome parameters :

- Age at Death
- Circumstances of death
- SCD
- Ventricular arrhythmia
- AV block
- BBB
- Requirement of CRT/ICD/transplant

INFORMATION SHEET

TITLE OF THE STUDY : CLINICAL PROFILE AND OUTCOMES OF CARDIOMYOPATHY IN CHILDREN

STUDY NUMBER :
PARTICIPANT S NAME
DATE OF BIRTH /AGE IN YEARS

- Cardiomyopathy is a disease of the cardiac muscle leading to inability of the heart to meet the demands of the body. You have been informed that you / your child is suffering from a disease of the muscle of the heart for which he / she has undergone or will be undergoing some tests including ECG / Chest Xray or Echocardiography which are part of routine evaluation of such patients.

- You are being requested to participate in a study which would evaluate the clinical characteristics and outcome of such diseases. Participating in this study , in which data like your / your child s age , mode of presentation and parameters from the investigations you have already undergone for the management of the disease would be used , will in no way , influence treatment decisions.

- WHAT IS CARDIOMYOPATHY

Cardiomyopathy is a disorder of the cardiac muscle which causes inefficient pumping of the heart. So that , it is unable to fulfill the demands of the body.

- WHAT IS MEANT BY CLINICAL PROFILE AND OUTCOME

The word clinical profile would mean that we would study the age of presentation , mode of presentation , family history and other such parameters . We will also study the occurrence of adverse outcomes in such diseases and compare it to outcomes of children with such diseases from other countries .

- DOES IT INVOLVE ANY INTERVENTIONAL PROCEDURE?

It does not any interventional procedure. Only the investigations which you would already undergo in the process of evaluation of the disease woulb be utilized for the study.

- IF YOU TAKE PART WHAT WILL YOU HAVE TO DO ?

For this study , we ll be using some of the data like history and other clinical details , investigations that are already done for management of the disease. No additional cost will be incurred and no additional drugs will be given.

- CAN U WITHDRAW FROM THIS STUDY AFTER IT STARTS ?

Your participation is entirely voluntary and you are free to withdraw permission to participate from the study. If you do so , it will not affect your usual treatment at this hospital.

- WHETHER THERE IS ANY RISK RELATED TO THIS STUDY?

There is no individual risk or benefit to the patient himself as it is an observational study.

- WILL YOUR PERSONAL DETAILS BE KEPT CONFIDENTIAL ?

The results of this study will be published in a medical journal but you will not be identified by name in any publication or presentation of results. However , your medical notes may be reviewed by people associated with the study, without your additional permission should you decide to participate in the study.

- If at any time you experience any problems or you have any further questions ,

Please ask: Dr Gousia Mukhtar (principal investigator)
gousiamukhtar@gmail.com
Ph no. 7994249031
IEC Member secretary
Dr K shiva kumar
0471-2524593

Consent form : TITLE OF THE STUDY : CLINICAL PROFILE AND OUTCOMES OF CARDIOMYOPATHY IN CHILDREN

STUDY NUMBER :
PARTICIPANT S NAME
DATE OF BIRTH /AGE IN YEARS
I ----------Son /Daughter of ---------- Declare that I have read the above information provided to me regarding the study : '' Clinical profile and outcomes of cardiomyopathy in children '' and I have clarified any doubts that I had .

- I also understand that my / my child s participation in the study is entirely voluntary and that I am free to withdraw to continue to participate any time without affecting my usual treatment or my legal rights.

- I also understand that study investigators will be using some of the data like history and other clinical details. Investigation parameters like ECG , CXR , holter monitoring results , Radiological imaging (Cardiac MRI) parameters for the study if already done in the hospital during the study.

- I also understand that no additional cost will be incurred / no additional drugs would be used and there is no additional risks to me / my child while being part of the study.

- I understand that the study staff and Instituitional Ethics committee members will not need my permission to look at my health records even if I withdraw from the study. I agree to this access.

- I understand that my / my childs identity will not be revealed in any information released to third parties or published.

- I voluntarily agree to take part in this study

- I received a copy of this consent form Name Signature Date Name of witness Relation to participant

Principal investigator : Dr Gousia Mukhtar

Ph no. 7994249031
IEC Member Secretary : Dr K Shivakumar 0471-2524593

DATE (Person obtaining consent)

I attest that the requirements for informed consent for the medical research project described in this form have been satisfied. I have discussed the research project with the participant/participant s parent and explained to him or her in non technical terms all of the information contained in this informed consent form , including any risks and adverse reactions that may be expected to occur. I further certify that I encouraged the participant / parent of the participant to ask questions and that all questions were answered ----------
NAME AND SIGNATURE OF PERSON OBTAINING CONSENT

സമ്മതപത്രം

പഠനശീർഷകം: കുട്ടികളിലെ കാർഡിയോമയോപ്പതിയുടെ ചികിത്സാലയ സംബന്ധിയായ രൂപരേഖയും ഫലങ്ങളും

പങ്കെടുക്കുന്നയാളുടെ പേര്.....
ജനനതിയത്/വയസ്സ് (മാസത്തിൽ/വർഷത്തിൽ)_____ പുത്രൻ/പുത്രി

..... (ദയവായി കോളങ്ങളിൽ ശരിയടയാളപ്പെടുത്തുക)

- മുകളിൽ പറഞ്ഞ കുട്ടികളിലെ കാർഡിയോമയോപ്പതിയുടെ ചികിത്സാലയ സംബന്ധിയായ രൂപരേഖയും ഫലങ്ങളും എന്ന പഠന സംബന്ധിയായി എനിക്കു നൽകിയ വിവരങ്ങൾ വായിച്ചു എന്നു പ്രസ്താവിക്കുന്നു.[ ]
- എന്റെ എല്ലാ സംശയങ്ങളും പരിഹരിച്ചു.[ ]
- എന്റെ കുട്ടിയുടെ ഈ പഠനത്തിലുള്ള പങ്കാളിത്തം പൂർണ്ണമായും സ്വമേധയാ ആണെന്നും അനുവാദം എനിക്ക് ഏതുസമയത്തും എന്റെ കുട്ടിയുടെ ചികിത്സയെയോ നിയമപരമായ അവകാശങ്ങളെയോ ബാധിക്കാതെ പിൻവലിക്കാൻ അവകാശമുണ്ടെന്നും ഞാൻ മനസ്സിലാക്കുന്നു. [ ]
- ആശുപത്രിയിൽ രോഗംകൈകാര്യം ചെയ്യുന്നതിനായി നടത്തിയ പരിശോധനകൾ, രോഗചരിത്രം, മറ്റ് വൈദ്യാലയസംബന്ധമായ വിശദാശംങ്ങൾ. ഇസിജി, സിഎക്സ്റേ , ഹോൾസ്റ്റർ നിരീക്ഷണ ഫലങ്ങൾ, റേഡിയോളജിക്കൽ ചിത്രീകരണങ്ങൾ (കാർഡിയാക് എംആർഐ) എന്നിവയുടെ പരിശോധനാമാനദണ്ഡങ്ങൾ എന്നിവയുടെ ചില വിവരങ്ങൾ ഈ പഠനത്തിനായി ഉപയോഗിക്കുമെന്നും ഞാൻ മനസ്സിലാക്കുന്നു.
- പഠനത്തിൽ പങ്കെടുക്കുന്നതുകൊണ്ട് അധികച്ചിലവ് ഉണ്ടാകില്ലെന്നും അധികം മരുന്ന് നൽകില്ലെന്നും അധികം അപായം എനിക്ക്/എന്റെ കുട്ടിക്ക് ഉണ്ടാകില്ലെന്നും ഞാൻ മനസ്സിലാക്കുന്നു.
- എന്റെ കുട്ടി ഈ പഠനത്തിൽ നിന്നും പിൻമാറിയാലും പഠനം നടത്തുന്നവർക്കും സ്ഥാപനത്തിലെ നൈതിക കമ്മിറ്റി അംഗങ്ങൾക്കും എന്റെ കുട്ടിയുടെ ആരോഗ്യരേഖകൾ പരിശോധിക്കുന്നതിന് എന്റെ അനുവാദം ആവശ്യമില്ലെന്ന്ഞാൻ മനസ്സിലാക്കുന്നു. അതിനോട് ഞാൻ യോജിക്കുന്നു. [ ]
- എന്റെ കുട്ടിയെ തിരിച്ചറിയാനുതകുന്ന വിവരങ്ങൾ ഒന്നും മൂന്നാം കക്ഷികൾക്കു നൽകുകയോ പ്രസിദ്ധീകരിക്കുകയോ ചെയ്യില്ലെന്ന് ഞാൻ മനസ്സിലാക്കുന്നു. [ ]
- എന്റെ കുട്ടിയെ സ്വമേധയാ പഠനത്തിൽ പങ്കെടുക്കാൻ സമ്മതിക്കുന്നു. [ ]
- സമ്മതപത്രത്തിന്റെ ഒപ്പിട്ട ഒരു കോപ്പി എനിക്കു കിട്ടി [ ]

പങ്കെടുക്കുന്നയാളുടെ പേര്

ഒപ്പ്

തീയതി

സാക്ഷിയുടെ പേര്

ഒപ്പ്

പങ്കെടുക്കുന്ന ആളുമായുള്ള ബന്ധം

തീയതി

(സമ്മതം വാങ്ങുന്നയാൾ)

മെഡിക്കൽ റിസർച്ച് പ്രോജക്ടിനാവശ്യമായ സമ്മതപത്രത്തിനു വേണ്ടുന്ന എല്ലാ ഘടകങ്ങളും തൃപ്തികരമായി നിർവഹിച്ചിരിക്കുന്നുവെന്ന് ഞാൻ ബോധ്യപ്പെടുത്തുന്നു. പഠനപങ്കാളിയുമായി ഗവേഷണപദ്ധതിയെപ്പറ്റി സാങ്കേതികേതര പദങ്ങളുപയോഗിച്ച് എല്ലാ വിവരങ്ങളെപ്പറ്റിയും ചർച്ച നടത്തുകയും പ്രതീക്ഷിക്കാവുന്ന അപകടസാധ്യതകളും പാർശ്വഫലങ്ങളും വിശദീകരിക്കുകയും ചെയ്തു. പങ്കാളിയെ ചോദ്യങ്ങൾ ചോദിക്കാൻ പ്രേരിപ്പിക്കുകയും എല്ലാ ചോദ്യങ്ങൾക്കും ഉത്തരം നൽകുകയും ചെയ്തു എന്നും ഞാൻ സാക്ഷ്യപ്പെടുത്തുന്നു.

സമ്മതപത്രം വാങ്ങുന്ന ആളുടെ പേര്

ഒപ്പ്

പ്രധാന ഗവേഷക

Child Assent for Children under Age 12 (To be read aloud to the child)

My name is Dr Gousia Mukhtar. I work with parents and children but I am also a student. Right now, I am trying to learn more about disease of the muscles of the heart in children like you.

We want to learn how such children like you present to the doctor , what is the age of presentation , wether your family members also have this disease. What are the findings when we examine your body.We will use the results of some tests which you will be undergoing during your management plan .

If you agree, we will use some of the information like the age at which it started , how you presented to the doctor , history of your family members and results of some tests which you already have to undergo for the routine evaluation of the disease.

You may be helping us understand the normal age and mode of presentation of such diseases. And the normal range of values of the tests which you already have to undergo during routine management of such diseases.

There will be no additional discomfort caused to you because of your participation in the study.

You should also know that if you decide to help us or if you decide to say “no,” your choice will not affect treatment plan or management of the disease.

Please talk this over with your parents before you decide if you want to be in my study or not. I will also ask your parents to give their permission for you to be in this study, but even if your parents say “yes,” you can still say “no” and decide not to be in the study.

If you don’t want to be in my study, you don’t have to be in it. Remember, being in the study is up to you and no one will be upset if you don’t want to be in the study or if you decide to stop after we begin, that’s okay, too

You can ask any questions that you have about the study. If you have a question later that you didn’t think of now, you can call me or ask your parents to call me at 7994249031

Would you like to share the history , examinatrion findings and results of your tests with me Answer as Yes or No

Name and signature of participant :

Principal Investigator : Dr gousia Mukhtar Ph no. 7994249031

IEC Member secretary, Dr K shiva kumar, Ph : 0471-2524593

DM Thesis

by Dr Gousia Mukhtar

General metrics

31,354	4,878	283	19 min 30 sec	37 min 31 sec
characters	words	sentences	reading time	speaking time

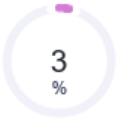
Score

This text scores better than 41% of all texts checked by Grammarly

Writing Issues

581	269	312
Issues left	Critical	Advanced

Plagiarism



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AGE in yrs	Age in months	Age in days	Years on FU	SEX	MODE OF PRESENTATION / HISTORY	syncope	CONSANGUINITY	FAMILY HISTORY OF CMP	FAMILY H/O SCD	SYNDROMES	NYHA FC	CVS	S3	P2	PSM/ESM	Double apical impulse	CXR	LAE	RAE	PVH	Axis LAD/ RAD/N	Rhythm	PR interval	PR interval z score	q wave	QRS d	QRS d z score	QT interval	QT dispersion	ST depression	LVH	LAH	RAH	Low voltage contents	Preexcitation	ECHO 1 , LVIDd	Z SCORE LVIDd	LVIDs	Z score LVIDs	IVS s	IVS , Z SCORE	PW s (mm)	PW , Z SCORE	LAE , Left atrial diameter mean , mm	LAE, Z score	LVEF
16				M	Palpitatior	N	NCM	Y	N	N	II	CE-	N	Y	Y	S4+	0.5	N	N	N	LAD	SR	120	-1.05	N	80	-1			NA	Y	N	N	N	N					27	5.6	16	2.1	34	2.56	71
5.5			1991-1998	F	Screening	N	NCM	N	N	Noonan sy	II	CE-	N	N	Y	S4+	0.6	Y	N	Y	N	SR	120	0	Y	120	2.64	400	60	Y	Y	Y	N	N	N					20		17		33		78
13			1987-1993	F	Palpitatior	Y	NOT KNOV	N	N	Noonan sy	II	CE-	N	N	Y	N	0.6	N	N	N	NW	SR	240	5.7	Y	100	1	400	50	NA	Y	Y	N	N	N					13		15		28		60
11			1987-2020	F	AOE	N	NCM	N	N	N	II	CE-	N	N	Y	N					N	SR	200	3.35	Y	100	1	453	50	NA	Y	Y	Y	N	N					35	7.3	13	1.1	34	2.7	75
17			1987-1991	M	DOE	N	NCM	N	N	N	II	CE-	N	N	Y	N	0.7	N	Y	Y	LAD	SR	220	4.15		150	4.9				Y	Y	N	N	N	45	0.37	39	2.93							20
17			1989-1996	M	DOE	N	NCM	N	N	N	II	CE-	N	N	Y	S4+	0.5	Y	N	N	NW	SR	120	-1	N					NA	Y	Y	N	N	N					14		20				85
5			1989	M	DOE	N	NCM	N	N	N	III	CE+	Y	Y	N	N	0.7	N	Y	Y	N	SR	120	0		120	2				Y	N	N	Y	N	45	2.14	41	4.75					30	2.84	21
9			09/89-11/8	M	DOE	N	NCM	N	N	N	II	CE-	N	Y	N	N	0.5	Y	Y	N	N	SR	140	1		140	2			NA	Y	Y	N	N	N	39	0.33	22	-0.47					41	4.83	65
7			1991-2001	M	Recurrent	N	NCM	N	N	N	III	CE+	Y	N	N	N	0.5	Y	N	Y	N	AFIB	140	1		120	2			NA	N	Y	Y	N	N	34	-0.34	24	0.78					38	4.69	57
10			1991-2019	M	DOE	N	NOT KNOV	N	Y	N	II	CE-	N	N	Y	N					N	SR	180	2.35		150	4.69	N			N	Y	Y	N	N	37	-0.46	29	1.5					27	1.72	45
13			1991-1998	M	DOE	N	NOT KNOV	Y	N	N	II	CE-	N	N	Y	N	0.5	N	N	N	N	SR	120	-1	N	120	2			NA	Y	N	N	N	N					25	5.7	8	1.46	25	3.03	86
	8		1992-2011	M	DOE	N	NCM	N	N	N	II	CE+	Y	Y	Y	Y						SR	120	1		120	2				N	N	N	N	N	39	4.45	29	4.98					27	4.14	50
5			03/03/199	F	DOE	N	NCM	N	N	N	III	CE+	Y	Y	Y	N	0.8	Y	Y	Y	RAD	SR	140	0.94						NA	N	Y	Y	N	N	22		16		10		11				58
7			1992-1997	F	DOE	N	NCM	N	N	N	II	CE+	Y	N	Y	N	0.7	N	Y	Y	RAD	SR	220	4.8		110	2			NA	N	N	Y	Y	N	27	-3.26	12	-5.2					28	2.18	60
12			1992-1995	M	DOE	Y	NCM	N	N	N	III	CE-	Y	Y	Y	S4+	0.55	N	N	N	N	SR	120	-1	Y	120	2	420	60	NA	Y	N	N	N	N					19		24		38		50
5			06/1992-0	M	DOE	N	NOT KNOV	N	N	N	III	CE+	Y	N	N	N	0.8	N	N	Y	LAD	SR	120	0	Y	120	2.17				Y	N	Y	N	N	40	0.87	35	3.43					27	2.04	29
16			10/92-02/9	M	DOE	N	NOT KNOV	N	N	N	IV	CE+	Y	Y	Y	N	0.65	Y	Y	Y	LAD	SR	110	-1		100	1				Y	N	N	N	N	75	5.74	67	7.42					41	4.09	10
11			6/11/92-1	M	DOE	N	NCM	N	N	N	IV	CE+	Y	N	Y	N	0.65	N	N	Y	N	SR	160	1.42		120	2.58				Y	N	N	N	N	60	4.46	53	6.34					32	2.97	32
		24	29/11/92-	F	Respirator	N	NCM	N	N	N	III	CE-	N	N	N	N															NA	NA	NA	NA	N											
2			02/93-02/9	F	DOE	N	NCM	N	N	N	III	CE+	Y	Y	Y	N	0.6	Y	N	Y	RAD	SR	120	0		120	2.3				Y	N	Y	N	N	53	4.93	44	6.23					22	1.29	22
2			03/93-04/9	M	Right hear	N	NCM	N	N	N	II	CE+	RV S3+	N	N	N	0.6	Y	N	N	N	SR	140	1.16		120	2			Y	N	Y	Y	N	N	28	-3.47	18	-2.45	17	3.85	16	3.07	48	5.65	65
12			04/93-08/9	M	DOE	N	NCM	N	N	N	II	CE-	N	Y	Y	N	0.7	N	N	Y	N	SR	100	-2		100	1				Y	N	N	N	N	48	-0.08	34	1.01					32		50
1.2			18/08/93-	M	Screening	N	NCM	N	N	N	II	CE-	N	N	N	N	0.6	N	N	N	N	SR			Y	100	2			NA	Y	N	N	N	N					10		7		22		65
1			1987-1993	M	Recurrent	N	NCM	N	N	N	II	CE-	N	Y	Y	N	0.55	N	N	N	RAD	SR	120	1	Y	110	2	361	50	Y	N	N	Y	N	N	NA										
2			11/94-12/9	F	Screening	N	NCM	N	N	N	II	CE-	Y	N	Y	N	0.6	N	N	N	N	SR	150	1		120	2.3				Y	N	N	N	N	39	3.93	29	4.57					17	0.59	52
	3		03/02/94-	M	Screening	N	NCM	N	N	N	II	CE-	Y	N	Y	N	0.6	Y	N	N	N	SR	120	1	N	110	2			NA	Y	N	N	N	N					14		15		16		75
1.6			02/94-03/9	F	Febrile Illn	N	NCM	N	N	N	III	CE-	N	Y	Y	N	0.6	N	N	Y	N	SR	120	0		110	2				Y	N	N	N	N	40	4.15	34	5.8					21	2.07	30
		7	02/94-03/9	M	DOE	N	NCM	N	N	N	II	CE+	Y	Y	Y	N	0.8	Y	Y	Y	LAD	SR	120	1	Y	100	1			NA	N	N	Y	N	N					12	5.1	6	0.86	14	1.4	64
7			05/94-96	M	DOE	Y	NCM	N	N	N	III	CE-	Y	Y	Y	y	0.6	Y	N	Y	RAD	SR	120	0	Y	120	2			NA	Y	Y	N	N	N					17	3.9	12	1.23	37	3.85	70
		3	22/07/94-	M	Feeding di	N	NCM	N	N	N	II	CE-	N	N	N	N					LAD	SR	120	1	N	100	2			NA	Y	Y	Y	N	N					8		7		14		82
12			1994-2000	M	Syncope	Y	NCM	N	N	N	II	CE+	Y	Y	Y	N	0.6	N	Y	N		JR	100	-1.5		100				NA	Y	N	Y	N	N	37	-1.36	15	-4.45	8	-0.77	9	-1.43	35	2.94	80
	9		11/94-03/9	F	Febrile Illn	N	NCM	N	N	N	III	CE+	Y	Y	Y	N	0.6	Y	N	Y	N	SR			Y	100	2				Y	N	N	N	N	54	8.52	44	8.92					29	5.09	32
	4		95-96	M	DOE	N	NCM	N	N	N	III	CE+	N	N	Y	N	0.6	Y	N	Y	N	SR	160	2		100	2				NA	N	Y	NA	N	32	3.87	26	5.22							30
2			1995-2004	M	DOE	Y	NCM	N	N	N	II	CE-	N	N	N	N	0.6	N	N	N	N	SR	130	1	Y	100	2				Y	Y	Y	N	N	48	3.73	33	3.79					57	7.9	67
13			995-2019	M	DOE	N	NCM	Y	N	N	II	CE-	Y	N	Y	N	0.5	Y	Y	Y	N	SR	120	-0.88		120	3.5			NA	Y	N	Y	N	N	32	-2.61	19	-2.41	9	0.06	9	-1.18	37	3.49	72
1.5			6/6/95-8/6	M	DOE	N	CM	N	N	N	II	CE-	Y	N	Y	N	0.6	Y	N	Y	N	SR	120	0		100	2				Y	Y	N	N	N	46	4.33	40	6.12					26	2.91	30
2			95-97	M	DOE	N	NOT KNOV	N	N	N	III	CE+	Y	Y	N	N	0.6	N	Y	N	RAD	SR	120	0		100	1.5			NA	Y	Y	Y	N	N	25	-3.77	16	-2.71					20	-0.05	
1			95-95	M	DOE	N	NCM	N	N	N	II	CE-	N	N	N	N	0.7	Y	Y	Y	N	SR	140	1		100	1.5			NA	Y	N	Y	N	N	34	0.65	21	0.51	9	1.41	8	-0.15	37	5.05	
6			27/02/96-	M	DOE	N	NCM	N	N	N	II	CE-	RV S3+	N	N	N	0.6	Y	Y	Y	N	SR	120	0	Y	100	1.5			NA	Y	Y	Y	N	N	27	-3.12	17	-2.33					35	3.84	67
		2	95	M	DOE	N	CM	N	N	N	II	CE-	Y	Y	Y	N	0.7	Y	Y	Y	N	SR	120	1		120	2.3				Y	N	Y	N	N	41	8.05	39	9.7					13	0.64	31
1			96-2008	M	DOE	N	NCM	Y	Y	N	II	CE+	Y	Y	Y	N	0.6	Y	Y	Y	N	SR	140	1.44		100	2				Y	N	Y	N	N	45	1.39	31	1.94					25	1.11	58
1			97-2021	F	DOE	N	NCM	Y	Y	Noonan Sy	II	CE+	N	N	Y	N	0.55	N	N	Y	N	SR	160	2.55	N	100	2			NA	Y	Y	N	N	N					16		12		51		68
	6		1997	F	DOE	N	NCM	N	N	N	II	CE+	N	N	Y	N	0.6	N	N	Y	N	SR	120	1		100	2				Y	N	N	N	N	44	6.09	39	7.6					20	2.25	28
13			1998-2005	M	DOE	N	CM	N	N	Emery Dre	II	CE+	Y	N	Y	N	0.6	N	N	N	N	SR	120	1		100	2			NA	NA	N	Y	Y	N	34	-2.1	24	-0.65					27	1.18	73
	3		1998-2021	M	DOE	N	NCM	N	N	N	II	CE+	Y	Y	Y	N	0.7	Y	N	Y	N	SR	120	1		100	2				Y	N	N	N	N	45	5.49	30	4.94					23	2.8	50
9			1998-1999	M	DOE	N	NCM	N	N	N	III	CE+	Y	Y	Y	N	0.7	Y	Y	Y	N	SR	120	-0.5		100	1				Y	N	N	N	N	53	2.48	49	5.11					40	4.07	18
2			2001-2021	F	DOE	N	NCM	N	N	N	III	CE+	Y	Y	Y	N	0.7	N	N	Y	N	SR	120	0		100	2				Y	N	N	N	N	51	8.39	45	4.55					29	3.93	24
15			1977-1984	F	DOE	N	NOT KNOV	NOT KNOV	NOT KNOV	N	III	CE+	N	N	Y	N	0.85		Y	Y	N	AFIB	120	-1	Y					NA	Y	N	Y	N	N	NA										
1			1992-2019	F	MURMUR	N	NCM	N	N	Noonan Sy	I	CE-	N	N	Y	N	0.5	N	N	N	N	SR			N	90	1			NA	Y	NA	NA	NA	N					10		10		24		78
13			1985-1986	F	MURMUR	N	NCM	N	N	N	I	CE-	Y	N	N	N	0.5	Y	Y	Y	N	SR	220	4.6						NA	Y	Y	Y	N	N	43	0.05	22	-1.49	11	0.85	10	-0.86	40	3.8	79
18			1994-2000	M	DOE	N	NCM	N	Y	N	III	CE+	N	N	N	S4+	0.6	N	N	Y	LAD	SR	160	1		150	6				NA	Y	Y	N	N	57	2.42	52	4.96					30	1.38	27
17			2000-2001	M	DOE	N	NCM	N	N	N	I	CE+	N	N	N	N	0.55	Y	N	N	N	SR	200	3.1							Y	N	Y	N	N	78	5.59	69	7.22					41	3.61	23
6			2001-2021	M	MURMUR	Y	NCM	Y	N	N	I	CE-	N	N	Y	S4+						SR			N					NA	Y	Y	N	N	N					27		15		35		90
9			1995-2001	M	Arrythmias	Y	NCM	N	N	N	III	CE-	N	N	Y	N	0.55	N	N	Y	N	SR				180	6.64				NA	N	N	N	N	44	0.28	39	3.03					20	-1.1	28
8			2002-2005	M	DOE	N	NCM	N	N	N	III	CE+	Y	N	N	N	0.65	N	Y	Y	N	SR	180	2.76		100	1				Y	NA	NA	N	N	63	4.33	57	6.41					31	2.36	25
9			2002-2002	F	DOE	N	NCM	N	N	N	III	CE+	Y	N	N	N	0.6	N	N	Y	N	SR									Y	Y	Y	N	N	56		50						36		23
15			2003-2003	M	DOE	N	NCM	N	N	N	III	CE+	Y	Y	Y	N	0.7	Y	N	Y	N		160	1		100	1				N	Y	N	N	N	68	4.85	63	7					42	4.33	21
8			2003-2012	F	Incidentall	N	NCM	Y	Y	N	I	CE-	N	N	Y	S4+	0.65	Y	N	Y	N	SR	210	4.8	Y	140	3.78			Y	Y	Y	N	N	N					24	5.17	38	8.06	22	0.56	68
	6		2003-2020	F	DOE	N	NCM	N	N	N	III	CE+	N	Y	Y	N	0.8	Y	N	Y	N	SR	120	0							Y	NA	NA	N	N	48	5.72	41	7.08					23	2.57	37
15			2003	M	DOE	N	NOT KNOV	NOT KNOV	NOT KNOV	N	III	CE+	RV S3+	N	Y	N					N	SR	140	0							N	NA	NA	N	N	64	4.11	60	6.5					40	3.87	18
18			2004-2012	M	DOE	N	NCM	N	N	Noonan Sy	II	CE+	N	N	Y	Y	0.55	Y	Y	Y	N	SR			Y	100	1	420	44	Y	Y	Y	N	N	N					29	5.36	26	4.55	35	2.13	72
13			2005-2006	M	DOE	N	NCM	N	N	N	II	CE+	N	Y	N	N	0.55	y	Y	Y	N	SR	120	-1	N	120	2.8	465	52	N	Y	Y	Y	N	N	41	-0.81	32	1.17	11	0.47	13	0.41	50	5.04	51
14			2004	M	Right hear	N	CM	N	N	N	II	CE+	RV S3+	Y	Y	N	0.6	N	Y	N	N	SR	120	-1		100	0	412		Y	N	N	Y	N	N	36	-1.56	18	-2.94					25	0.62	73
11			2005-2011	F	DOE	N	NCM	N	N	N	II	CE+	N	Y	N	N	0.6	Y	Y	Y	N	SR	140	0						NA	Y	Y	Y	N	N	39	-0.75	29	0.83	8	-0.69	8	-2.09	32	2.37	63
3			2005	F	DOE	N	NCM	N	N	N	III	CE+	Y	N	N	N	0.75	Y	Y	Y	N	SR	160	2							Y	Y	Y	N	N	49	5.2	46	7.41					40	6.09	18
13			2005	M	DOE	N	NOT KNOV	N	N	N	II	CE+	Y	Y	N	N	0.65	N	Y	Y	N	SR , Atrail f	240	5.7		150	4.83				N	Y	N	N	N	55	2.51	46	4.34					46	4.7	35
15			2006-2012	M	DOE	N	NCM	N	N	N	II	CE+	N	N	N	N	0.6	N	Y	Y	N	SR	120	-1	Y	100	1				N	N	N	N	N	60	3.47	47	4.58	9	-0.03	9	-1.29	22	-0.35	45
8			2006-2007	M	Right hear	N	NCM	N	N	N	I	CE-	N	Y	Y	N	0.5	N	N	Y	N	SR	140	1		100	1	406	50	N	N	N	Y	N	N	40	0.19	27	0.83	8	-0.18	11	0.67	45	5.25	67
13			2006	F	DOE	N	NCM	N	N	N	III	CE+	N	N	N	N	0.7	Y	Y	Y	N	SR			Y						Y	Y	N	N	N	52	2.48	47	4.94					43	4.72	20
4			2007	F	DOE	N	NCM	N	N	N	II	CE+	N	N	N	N	0.52	Y	N	Y	N	SR	140	1	Y	80	0				NA	Y	N	N	N	44	0.26	34	1.93					30	1.75	45
1.5			2007-2013	M	DOE	N	CM	N	N	N	II	CE+	N	Y	Y	N	0.72	Y	N	Y	LAD	SR	250	7.27	Y	120	2	476	57	Y	Y	Y	N	N	N					20	4.35	16	2.56	80	8.9	68
11			2007-2008	M	Syncope	Y	NOT KNOV	NOT KNOV	NOT KNOV	N	II	CE-	N	N	Y	N	0.5	Y	N	N	N	SR	160	1	N					NA	Y	Y	N	N	N					23		12		26		71
2.5			2007	F	DOE	N	NCM	N	N	N	II	CE+	N	Y	Y	N						SR	140	1							Y	Y	N	N	Y	55	7.09	49	8.49					28	3.97	25
12			2004-2008	M	DOE	N	NOT KNOV	N	NOT KNOV	N	III	CE+	N	Y	N	N	0.5	Y	Y	Y	RAD	SR	220	4.6		120	2			NA	Y	N	Y	N	N	NA										
2			2008	F	DOE	N	NCM	N	N	N	II	CE+	Y	Y	Y	N	0.6	N	N	Y	N	SR	160	2	Y	100	2				Y	N	Y	N	N	42	5.83	26	4.62					35	6.32	67
8			2008-2010	M	Right hear	N	NCM	N	N	N	I	CE+	LV S3+ RV	N	N	N	0.6	N	Y	Y		SR	130	0		120	2.35			NA	N	Y	N	N	N	38	0.41	23	0.15	8	0.20	8	-0.93	53	6.84	68
	7		2008-2013	F	Recurrent	N	NCM	N	N	N	I	CE-	N	N	Y	S4+					N	SR	120	1	N	120	2	413	80	N	Y	Y	Y	N	N					15	3.91	11	1.57	34	4.08	82
2			2009	M	DOE	N	NCM	N	N	N	II	CE+	N	N	Y	N	0.65	N	N	Y	N	SR	140	1		120	2				Y	Y	N	N	N	45	5.07	35	5.82					24	2.87	43
3			2009-2010	M	DOE	N	NCM	N	N	N	II	CE-	N	N	Y	N	0.75	Y	N	Y	N	SR	140	1	Y						Y	Y	N	N	N	57	4.67	47	5.97					39	4.79	21
	6		2009	M	DOE	N	NCM	N	N	N	II	CE+	Y	N	Y	N	0.6	Y	N	Y	N	SR	80	-2	Y	140	5.38				Y	Y	N	N	Y	42	6.19	39	8.11					25	4.13	50
	2		2009	F	DOE	N	NCM	N	N	N	II	CE+	Y	N	Y	N	0.7	Y	N	Y	N	SR			Y						Y	N	N	N	N	42	6.89	32	7.1					26	4.79	40
2.5			2010	F	Hyperdyna	N	NCM	N	N	N	II	CE+	Y	N	N	Y	0.6	N	N	Y	N	SR	200	4.5	Y	120	2.5	443		Y	N	Y	Y	N	N	32	0.5	22	1.24					25	2.53	60
13			2012	M	DOE	N	NCM	Y	N	Mytochon	III	CE+	N	N	N	N	0.67	N	N	Y		SR	210	4.1		128	2				Y	Y	N	N	N	60	3.47	54	5.68					37	3.33	23
8			2010-2021	M	DOE	N	NCM	N	N	N	II	CE+	N	Y	N	N	0.55	N	N	N	LAD	SR	180	2	N	120	2	424	70	N	Y	Y	Y	N	N					10	0.33	14	1.36	35	3.35	58
9			2010-2011	F	Incidentall	N	NCM	Y	Y	Noonan Sy	I	CE-	N	N	Y	N					LAD	SR	160	2	Y	80	-1	588	126	N	Y	Y	N	N	N					15	3.17	15	2.64	35	3.41	79
13			2008-2012	M	DOE	N	NCM	Y	N	N	III	CE+	Y	N	N	N	0.6	Y	Y	Y		SR	240	5.7							Y	Y	N	Y	N											
11			2004-2011	F	Syncope	Y	NCM	N	N	N	II	CE-	N	N	N	S4+	0.52	Y	N	N	LAD	SR	120	-0.65	Y	140	3	N		Y	N	N	Y	N	N					14		14		34		45
0.6			2012-2021	M	Recurrent	N	NCM	N	N	N	I	CE-	N	N	Y	N	0.5	Y	N	N	N	SR	150	2.1	N					NA	NA	Y	N	N	N					10		9		22		57
5			2011	M	Febrile Illn	N	NCM	N	N	N	III	CE+	Y	N	Y	N	0.73	Y	N	N	N	SR	120	0		120	2.3				Y	Y	N	N	N	55		50						28		20
11			2011-2021	M	Syncope	Y	NCM	N	N	Friedrich	II	CE-	N	N	N	N	0.5	N	N	N	N	SR	120	-1	Y	120	2			N	Y	Y	N	N	N					14		10		26		85
13			2012-2021	F	Syncope	Y	NCM	Y	Y	N	I	CE-	N	N	Y	N					LAD	SR	160	1	N	80	-1	432	50	Y	Y	Y	N	N	N					19	3.5	16	1.9	42	3.9	67
12			2012-2013	M	DOE	N	NCM	N	N	N	III	CE+	N	Y	N	S4+	0.73	N	N	Y	N	SR	200	3.2	Y	100	2			NA	NA	Y	Y	N	N	28	-3.02	20	-1.26	2.21		13	2.03			62
17			2012	M	DOE	N	NCM	N	N	Kelnifelter	II	CE+	Y	N	Y	N						SR	240	5.2	Y	120	2				Y	Y	N	N	N	76	5.53	64	6.78					50	5.18	31
4			2012- 201	F	DOE	Y	NCM	N	N	N	II	CE+	Y	N	N	N					N	SR	160	2	Y	80	0	462	75	N	Y	Y	Y	N	N	22	-3.8	12	-3.99	12	2.94	10	1.27	34	4.4	80
	2		2013-2020	M	DOE	N	NCM	N	N	N	II	CE+	N	N	Y	N	0.56	Y	N	Y	LAD	SR	160	2.6		120	3.4				Y	Y	N	N	N	42	4.43	35	5.87					33	5.15	37
8			2012-2017	F	Syncope	Y	NCM	N	N	N	III	CE-	N	N	Y	N	0.5	N	N	N	N	SR	140	1	Y	80	-1	410	60	N	Y	N	Y	N	N					10		12		26		45
2			2012	M	DOE	N	NCM	N	N	N	II	CE+	Y	N	Y	N	0.7	Y	N	Y	N	SR	120	0		130	3.46				Y	Y	N	N	N	50	5.14	45	7.03					22	1.72	19
	7		2013-2020	F	DOE	N	NCM	N	N	N	II	CE+	Y	N	N	N	0.7	Y	N	Y	N	SR	120	1		120	2.3				Y	Y	N	N	N	43	4.35	35	5.62					24	2.73	40
	6		2013-2016	M	DOE	N	NCM	N	N	N	II	CE+	Y	N	N	N	0.7	Y	N	Y		SR	120	1		130	2.4				Y	Y	N	N	N	46	3.95	39	5.6					28	3.23	33
16			2013	F	DOE	N	NCM	N	N	N	IV	CE+	Y	N	Y	N	0.7	Y	N	Y		SR	160	1							Y	Y	Y	N	N	63	3.87	57	6.03					32	2.22	22
15			2013	M	DOE	N	NCM	N	N	N	IV	CE+	Y	N	Y	N	0.68	Y	N	Y	LAD	SR	200	3.26							Y	Y	N	N	N	72	4.15	65	6.26					51	4.67	20
2.5			2013-2021	M	DOE	N	NCM	Y	N	N	II	CE+	Y	Y	N	N	0.66	N	N	Y	N	SR	120	0		80	1				N	Y	N	N	N	43	4.1	37	5.86					24	2.59	28
	5		2013-2019	M	Recurrent	N	NCM	N	Y	N	II	CE+	Y	N	Y	N	0.55	N	Y	Y	N	SR	100	0	N	80	1				Y	Y	N	N	N	26	2.32	15	1.28	7	01-Jan	5	-1.12	16		60
3			2013-2016	M	Family Scr	N	NCM	Y	Y	N	II	CE-	N	N	N	Y	0.6	Y	N	N	LAD	SR	140	1	N	120		540	70	N	Y	Y	N	N	N					16	3.5	13	1.7	25	1.04	85
	6		2013-2014	M	Murmur	N	NCM	N	N	N	I	CE-	N	N	Y	Y					N	SR	140	1.5	N	100				NA	Y	Y	N	N	N					14		15		18		90
13			2013-2021	F	DOE	Y	NCM	N	N	N	II	CE-	N	N	Y	N	0.57	Y	N	Y	LAD	SR	160	1	Y	180	7.33			N	Y	N	N	N	N					21		18		34		92
16			2014-2018	M	Syncope	Y	NCM	N	N	N	II	CE-	N	N	N	N	0.5	Y	N	N	N	SR	80	-3.2	N	120	2	416	45	N	Y	Y	N	N	Y					18		23		34		73
	7		2015-2021	M	Recurrent	N	CM	N	N	N	III	CE+	Y	Y	Y	N	0.7	Y	N	Y			120	0	Y	110	2.4				Y	Y	N	N	N	47	7.31	42	8.69					20	2.55	20
3			2014-2020	F	DOE	N	NCM	N	N	N	III	CE+	Y	N	N	N	0.7	Y	N	Y	N	SR	130	1		100	2				Y	Y	Y	N	N	35	0.82	32	3.75					27	2.76	19

SAM	PAH	MR	LV AO GRADIENT	OBS/ NON OBS	HOLTER	NT pro BNP	C MRI LGE Y/N		PERCENTAGE	MAX THICKNESS	MYOCARDIAL MASS	NC:C RATIO	Area of NC	Medications , Digoxin	Lasix	ACE I	Aldactone	Beta blocker	CARDIAC CATH, RA mean	RV Peak	RV ed	PA mean	RPAWedge	LVSP	LV ed	CO	CI	PVR	PVRI	LV AO grad	BB phen	Type_CORRECTED	years of FU	OUTCOMES : AGE AT DEATH	Age at death date of daeth	DEATH FROM HF	SCD	Death from VT	VENTRICULAR ARRYTHMIAS	Atrial/Junctional Arrythmias	AV BLOCK	BBBB	REQUIREMENT OF CRT / ICD / TRANSPLANT	Septal myectomy
Y	N	2		NON OBS	not done		ND								n	N	N	Atenolol				18			35							HCM	2	Not known					N	N	N	N	N	
Y	N	2	90	OBS	not done		ND							N	N	N	N	Ppnl				15			24					120		HCM	8	Not known					N	N	N	Y	N	
Y	N	2	87	OBS	not done		ND							N	N	N	N	Ppnl				17			13					30	Y	HCM	6	Not known					N	N	Y	N	N	
N	N	1		NON OBS	NSVT		ND							N	Y	Y	N	Metoprolol				14			12							HCM	34	Doing well					N	N	Y	N	N	
	N	2			not done		not done							Y	Y	N	N	N							21							DCM	4	Not known					N	N	Y	Y	Y	
Y	N	0		NON OBS	not done		ND							N	n	N	N	Atenolol				20			13							HCM	9	Not known					N	N	N	N	N	
	N	2			not done		not done							N	Y	Y	N	N														DCM	2	Not known					N	N	N	N	Y	
	N	0			not done		NA							Y	Y	N	N	N	7	36	14	20	17	112	32	1.5	1.82	2	1.64			RCM	0.2	NOT KNOWN					N	N	N	N	N	
	Y	2			not done		NA							Y	N	N	N	N	10	39	15	22		92	28	1.74	2.48	3.44	2.41			RCM	10	NOT KNOWN					N	N	N	N	N	
		0			VPC s		Done							y	y	n	n	Y	11	22	12	16				5.76						ARVD	20	Not known					Y	N	N	Y	Y	
Y	N	0	60	OBS	NORMAL		ND							N	N	N	N	Ppnl				24			8					50 Provok	N	HCM	7	Not known					N	N	N	N	N	
	N	2			Atrial tachycardia / Af		not done							Y	Y	Y	N	N														DCM	20	Recovered					N	Y	N	N	N	
	Y	1			Not done		NA							Y	Y	N	N	N	14	75	24	50	32	65	40							RCM	0.1	Death	8, 03/1992	Y	N	N	N	N	N	N	N	
	N	0			not done		NA							Y	Y	N	Y	N	18	30	20	18	14				2.4	3.96	3.16			RCM	5	Not known					N	N	Y	N	Y	
Y	N	2		NON OBS	not done		ND							N	N	N	N	N				20			12							HCM	3	Not known					N	N	N	N	N	
	N	2			not done		not done							Y	Y	Y	Y	N														DCM	0.1	Death	5, 07/1992	Y	N	N	N	N	N	N	Y	
	Y	3			not done		not done							Y	Y	Y	N	N														DCM	0.5	Death	16.5, 1993	Y	N	N	N	Y	N	N	Y	
	Y	3			not done		not done							Y	Y	Y	N	N				34	25			3.16	3.09	2.84	2.9			DCM	0.1	Not known					N	N	N	N	N	
	N	3			not done		not done							NA																		DCM	0.1	Death	24 days, 1	Y	N	N	N	N	N	N	Y	
	Y	1			not done		not done							Y	Y	Y	N	N														DCM	1	Not known					N	N	N	N	Y	
	N	1			not done		NA							N	Y	N	Y	N														RCM	0.1	Not known					N	N	N	N	N	
	N	0			not done		not done							Y	Y	Y	N	N														DCM	3	Recovered					N	Y	N	N	N	
N	N	0		NON OBS	not done		ND							N	N	N	N	Ppnl														HCM	0.1	Not known					N	N	N	N	N	
	N	2			not done		NA							N	N	N	N	N	10	62	13	44	28	95	28			6.5	4.6			RCM	6	Not known					N	N	N	N	Y	
	N	2			not done		not done							Y	N	N	N	N				16			10	1.43	3.41	4.19	1.75			DCM	1	Not known					N	N	N	N	N	
N	Y	2		NON OBS	not done		ND							Y	Y	N	N	Ppnl														HCM	0.1	Not known					N	N	N	N	N	
	N	3			not done		not done							Y	Y	N	N	Y				17	14		12	0.93	2.46	3.22	1.22			DCM	0.1	Not known					N	N	N	N	N	
Y	N	0	120	OBS	not done		ND							N	y	N	N	N				22			10					120		HCM	0.1	Not known					N	N	N	N	N	
Y	Y	2	103	OBS	not done		ND							N	n	N	N	Ppnl				22			22					60		HCM	2	Not known					N	N	N	N	N	
Y	N	0		NON OBS	not done		ND							N	n	N	N	N														HCM	0.1	Not known					N	N	N	N	N	
	N	1			not done		NA							N	Y	N	N	N	12	36	12	20	16	116	15	3.22	2.8	1.25	1.55			RCM	6	Not known					N	N	N	N	Y	
	N	3			not done		not done							Y	Y	Y	N	N				15	12		8	0.85	2.44	3.52	1.23			DCM	0.4	Not known					N	N	N	N	N	
	N	2			not done		not done							y	y	Y	N	N							6							DCM	1	Not known					N	N	N	N	N	
	Y	4			not done		not done							y	y	N	N	N				28	18		18	1.29	2.08	7.75	4.8			DCM	9	Not known					N	N	N	N	N	
	N	0			not done	2810	NA							N	N	N	N	N	3	50	10	15	5	125	15	1.76	1.6	5.68	6.36			RCM	25	Doing well					N	N	N	Y	Y	
	N	2			not done		not done							Y	Y	Y	N	N														DCM	0.3	Not known					N	N	N	N	N	
	Y	1			not done		NA							Y	Y	Y	N	N	11	50	18	34	18	93	24	1.15	1.53	13.9	10.42			RCM	2	Not known					N	N	N	N	Y	
	Y	2			not done		NA							y	y	Y	N	N	7	60	11	30	19	85	25	1.2	2.03	9.16	5.3			RCM	0.1	Not known					N	N	N	N	Y	
	N	1			not done		NA							Y	Y	N	Y	N	14	36	18	23	15	97	27	2.4	3.07	3.33	2.6			RCM	0.1	Not known					N	N	N	N	N	
	N	2			not done		not done							y	y	N	N	N														DCM	0.1	Not known					N	N	N	N	N	
	Y	3			not done		not done							Y	Y	N	N	N				18			6	4.2	4.68	2.85	2.57			DCM	12	Not known					N	N	N	N	N	
Y	Y	3	100	OBS	not done		ND							N	n	N	N	Ppnl				36			18					140	Y	HCM	24	Doing well					N	N	N	N	N	Y
	N	3			not done		not done							y	y	Y	N	N														DCM	0.1	Not known					N	N	N	N	N	
	N	2			not done		NA							N	N	N	N	N	14	30	16	19	12	100	20	2.24	1.94	3.12	3.43			RCM	8	Not known					N	N	N	N	Y	
	N	1			not done		not done							y	y	N	N	N				14			8	3	6.9	2	0.86			DCM	24	Recovered					N	N	N	N	N	
	N	3			not done		not done							y	y	Y	n	N														DCM	1	Not known					N	N	N	N	Y	
	N	3			not done		not done							y	y	N	n	Y														DCM	19	Recovered					N	N	N	N	N	
	N	2			not done		NA							N	Y	N	N	N	26	35	26	28		115	26							RCM	7	Death	21, 1984	Y	N	N	N	N	N	N	N	
N	N	0		NON OBS	NSVT		ND											N				15			12							HCM	30	Doing well					N	N	N	N	N	
	N	2			not done		NA							N	Y	N	N	N	15	40	17	29		80	35							RCM	1	Not known					N	N	Y	N	N	
	Y	2			not done		not done							y	y	Y	N	N														DCM	6	Death	23 , 2000	Y	N	N	N	N	N	Y	Y	
	N	2			VPC s		not done							NA																		DCM	1	not known					N	N	Y	N	Y	
Y	N	1	40	OBS	NSVT		ND											Metoprolol														HCM	21	Doing well					Y	N	N	N	N	
	N	0			Recurrent VT		not done							N	N	Y	N	N														DCM	6	not known					Y	N	N	Y	N	
	N	3			not done		not done							Y	Y	Y	N	Y														DCM	5	Death	13, 2005	Y	N	N	N	N	N	N	Y	
	Y	3			not done		not done							Y	Y	N	Y	Y														DCM	0.3	Death	9.5, 2002	Y	N	N	N	N	N	N	Y	
	Y	2			not done		not done							Y	Y	N	N	N														DCM	0.4	Death	15.5, 2003	N	N	Y	Y	N	N	N	Y	
Y	N	3	108	OBS	Atrial tachycardia / Af		Y											Metoprolol														HCM	9	Not known					Y	N	Y	Y	Y	
	N	3			not done		not done							Y	Y	Y	N	Y														DCM	17	Recovered					N	N	N	N	N	
	N	3			not done		not done							Y	N	Y	N	N														DCM	0.3	Death	15.3, 2003	Y	N	N	N	N	N	N	Y	
Y	N	2	64	OBS	Normal		Y			43	499					y		Atenolol				22			12					40 provok	N	HCM	8	Not known					N	N	N	N	N	
	Y	2			not done		NA							N	Y	N	N	N	14	70	20	50	30	100	24	3.2	2.14	6.25	9.5			RCM	1	Not known					N	N	N	Y	Y	
		0			Not done		ND							Y	Y	N	N	N	12	25	14	17	6		18	3.13	2.72	3.51				ARVD	0.3	Not known										
	N	2			not done		NA							Y	N	N	N	N	6	40		34	24	100	24	4.5	3.8	2.22	2.48			RCM	6	Not known					N	N	N	N	N	
	Y	3			not done		not done							y	Y	Y	N	N														DCM	3	Not known					N	N	N	N	Y	
	N	2			not done		not done							Y	Y	Y	Y	Y				30	22		28	3.44	2.64	2.32	3.02			DCM	0.4	Not known							Y			
		1			NSVT									Digoxin	Y	Y	N	N	5	27	6	11	6			4/3.55		1.25	1.46			ARVD	8	Not known										
	N	2-3+			not done	1296	N							N	Y	N	N	N	30	64	30	46	26	106	36	2.11	2.31	9.47	8.62			RCM	1	Not known					N	N	N	N	Y	
	Y	2			not done		not done							y	Y	Y	Y	Y														DCM	0.1	Not known					N	N	N	N	Y	
	N	0			not done		not done							y	Y	Y	N	N				14	10		10	4.59	3.47	0.88	1.17			DCM	1	not known					N	N	N	N	N	
Y	N	4	124	OBS	not done		ND											Metoprolol														HCM	6	Not known					N	N	Y	N	N	
N	N	1		NON OBS	NSVT		ND											Metoprolol														HCM	1	Not known					N	N	N	N	N	
	Y	3			not done		not done							Y	Y	N	Y	Y														DCM	2	Not known										
	N	1			Atrial tachycardia / Af		NA							Y	Y	Y	Y	N	15	90	12	62	36	110	42	1.9	1.52	13.6	16.9			RCM	4	Not known					Y	Y	Y	N	Y	
	Y	3			not done		not done							Y	Y	Y	N	N														DCM	1	Death	3, 2008	Y	N	N	N	N	N	N	Y	
	Y	3			not done		NA							N	Y	Y	N	N														RCM	2	Not known					N	N	N	N	N	
Y	N		107	OBS	Normal		Y											Metoprolol														HCM	5	not known					N	N	N	N	N	
	N	2			not done		not done							Y	Y	Y	N	N														DCM	0.1	Not known					N	N	N	N	N	
	N	3			not done		not done							Y	Y	Y	N	N														DCM	1	Not known					N	N	N	N	Y	
	N	3			not done		N							Y	Y	N	N	N														DCM	1	Not known					N	N	N	N	N	
	N	3			not done		not done							Y	Y	N	N	N														DCM	0.4	Not known					N	N	N	N	N	
	N	2			not done		N							Y	Y	N	N	N	4	28	9	24	20	101	25	2.14	3.89	1.86	1			RCM	0.1	Not known					N	N	Y	N	N	
	Y	3			not done		N							Y	Y	N	Y	Y														DCM	1	Not known					Y	N	N	N	Y	
N	N	2		NON OBS	VPC s		Y								Y			Metoprolol				25			25							HCM	10	Doing well					N	N	N	N	N	
Y	Y	3	90	OBS	not done		ND											Metoprolol				18			14					140	Y	HCM	1	Not known					N	N	N	N	N	
	N	2			not done		not done							Y	Y	N	N	N														DCM	4	Death	17, 2012	Y	N	N	N	Y	N	N	Y	
N	N	1		NON OBS	INTERMIT	3000	Y			21								N				28										HCM	6	Not known					N	N	N	Y	Y	
N	N	0		NON OBS	not done		Y											Metoprolol							9							HCM	10	Doing well					N	N	N	N	N	
	N	3			not done		N							Y	Y	N	N	Y														DCM	0.3	Death	5.3 , 2011	Y	N	N	N	N	N	N	Y	
Y	N	0		NON OBS	Atrial tachycardia / Af		ND											Metoprolol														HCM	10	Doing well					N	N	N	N	N	
Y	N	1	16	OBS	not done		N			31	171							Metoprolol														HCM	9	Doing well					N	N	N	N	Y	
	Y	1			not done		NA							Y	Y	Y	Y	N														RCM	1	Not known					N	N	Y	N	N	
	Y	4			not done		not done							Y	Y	Y	Y	N														DCM	1	Not known					N	N	Y	N	N	
	Y	1			not done		Y		LGE along LV lateral wall					Y	Y	N	N	N														RCM	6	Death	12, 2019	N	Y	N	N	N	N	N	N	
	N	3			LBBB		not done							Y	Y	Y	N	N														DCM	6	Persistent LV dysfunction					N	N	Y	Y	N	
N	N	2		NON OBS	Normal	1704	Y			13					y			Ppnl				32										HCM	5	Death		N	N	Y	Y	N	N	N	Y	
	N	2			not done	10000	not done							Y	Y	N	N	N														DCM	9	Recovered					N	N	N	Y	N	
	N	3			not done		Y		Subendocardial enahncement at AS and Inferior regions					Y	Y	Y	N	N														DCM	7	Persistent LV dysfunction					N	N	N	N	N	
	Y	3			not done		N							Y	Y	Y	N	N				58	23			1.34	2.43	26	14.3			DCM	3	Death	3.6.2016	Y	N	N	N	N	N	N	N	
	N	2			not done	10000	not done							Y	Y	Y	N	N														DCM	0.8	Death	16.9.2013	Y	N	N	N	N	N	N	N	
	Y	2			not done	9000	not done							Y	Y	N	N	N														DCM	1	Death	16, 2013	Y	N	N	Y	N	Y	N	N	
	N	2			not done		Done							y	y	y	N	y														DCM	8	Recovered					N	N	N	N	N	
8 mm ASD BD shunt re		1			not done	167	Y	Y	Subendocardial in RV										8	29	12	18			9	1.36	2.46	6.53	1.69			RVNC	6	Doing well					N	N	N	N	N	
Y	N	2	15	OBS	Normal		N											Ppnl														HCM	3	Not known					N	N	N	N	N	
Y	N	2	30	OBS	not done		N											Atenolol														HCM	1	Not known					N	N	N	N	N	
Y	N	3	102	OBS	not done	2375	N								y			Metoprolol				28								100	Y	HCM	7	Doing well					N	N	N	Y	N	Y
Y	N			NON OBS	Normal	50	N			27								Metoprolol														HCM	6	Not known					N	N	N	N	N	
	N	3			not done		not done							Y	Y	Y	N	Y														DCM	7	Recovered					N	N	N	N	N	
	Y	2			not done		N							Y	Y	N	N	Y														DCM	6	Recovered		N		N	N	N	N	N	Y	

AGE in yrs	Age in months	Age in days	Years on FU	SEX	MODE OF PRESENTATION / HISTORY	syncope	CONSANGUINITY	FAMILY HISTORY OF CMP	FAMILY H/O SCD	SYNDROMES	NYHA FC	CVS	S3	P2	PSM/ESM	Double apical impulse	CXR	LAE	RAE	PVH	Axis LAD/ RAD/N	Rhythm	PR interval	PR interval z score	q wave	QRS d	QRS d z score	QT interval	QT dispersion	ST depression	LVH	LAH	RAH	Low voltage contents	Preexcitation	ECHO 1 , LVIDd	Z SCORE LVIDd	LVIDs	Z score LVIDs	IVS s	IVS, Z SCORE	PWs (mm)	PW, Z SCORE	LAE , Left atrial diameter mean, mm	LAE, Z score	LVEF
	4		2014-2019	M	DOE	N	NCM	N	N	N	II	CE+	N	N	Y	N	0.6	N	N	Y	N	SR	150			80					Y	Y	N	N	N	48		37						13		45
3.5			2014	F	Right hear	N	NCM	N	N	N	III	CE+	N	Y	N	N	0.76	N	N	Y	N	SR	140	1	Y	90	0			NA	NA	Y	Y	N	N	29	-2.18	22	-0.11	10	1.46	10	0.61	40	4.91	44
	3		2015	M	DOE	N	NCM	N	N	N	III	CE+	Y	Y	N	N	0.7	Y	N	Y	N	SR	170	3.26		90	2				Y	Y	N	N	N	58	9.06	53	10.2					27	4.49	16
10			2015-2021	M	DOE	N	NCM	N	N	N	III	CE+	Y	N	Y	N	0.7	N	N	Y	N	SR	200	3.52		90	0	389		N	Y	Y	Y	N	N	24	-5.32	15	-4.14	18	3.96	15	2.32	38	3.8	67
18			2015	M	DOE	N	NCM	Y	Y	N	III	CE+	Y	N	Y	N	0.65	Y	N	Y	N	SR	150	1		100	1.5				Y	Y	Y	N	N	70	4.4	59	5.9					32	1.78	31
18			2014-2021	F	Murmur	Y	NCM	N	N	N	II	CE-	N	N	Y	Y	0.6	N	N	N	N	SR	110	-1.73	N	90	1	540	100	Y	Y	Y	N	N	Y					35		26		28		55
9			2014	M	DOE	N	NCM	N	N	N	III	CE+	Y	N	N	N	0.65	Y	Y	Y	N	SR	180	2.88	Y	100	1				N	N	N	N	N	52	2.26	36	2.65					35	3.11	58
3			2014-2018	M	DOE	N	NCM	N	N	N	II	CE-	N	N	Y	N	0.56	Y	N	Y	LAD	SR	120	0	N	110	2			NA	Y	Y	N	N	N					25		19		28		76
11			2011-2014	F	DOE	N	NCM	N	N	N	II	CE+	Y	Y	N	N	0.69	Y	N	Y	RAD	SR	160	1		110	1				Y	Y	N	N	N	62		56						48		21
15			2015-2019	M	DOE	N	NCM	N	N	N	II	CE+	Y	Y	N	N	0.7	Y	N	Y	N	SR	200	3.26		130	2.921				Y	Y	N	N	N	84		71								30
	2		2015	M	DOE	N	NCM	N	N	N	III	CE+	N	N	N	N					N	SR	120	1	N	90	2				Y	Y	N	N	N	40	7.65	35	8.7					18	2.86	27
	1.5		2015	M	Feeding di	N	NCM	Y	N	N	II	CE+	Y	N	Y	N	0.66	Y	Y	Y	LAD	SR	140	1	Y	90	2				Y	N	N	N	N	39	5.75	30	6.27					21	3.07	25
7			2015-2021	M	Syncope	Y	NOT KNOV	N	N	N	II	CE-	N	N	N	N	0.5	N	N	N	N	SR	100	-1.33	Y	90	1	503	20	Y	Y	N	N	N	N					19	1.5	17	-0.01	43	1.6	72
1			2015-2018	F	Syncope	Y	CM	N	N	N	II	CE+	N	N	Y	Y	0.63	Y	N	Y	LAD	SR	120	0	N	110	2	400	40	NA	Y	Y	Y	N	N					16	3.32	17	3.2	45	5.0	80
		2	2015	M	Feeding di	N	NCM	Y	Y	N	II	CE-	N	N	Y	N	0.6	Y	Y	N	N	SR	100	-1	Y	90	2	497	62	Y	N	N	N	N	N					8	2.85	9	3.39	14	1.27	74
	6		2015-	F	Recurrent	N	NCM	N	N	N	II	CE-	N	N	N	N					LAD	SR	120	-1	Y	100	2	400	40	NA	N	N	N	N	N					12	4.26	6	-0.32	12	-1.03	56
	3		2015	M	Feeding di	N	CM	N	N	N	II	CE+	N	Y	N	N	0.65	Y	N	Y	N	SR	100	0	Y	90	2	350	45	N	Y	N	N	N	N	37	5.22	31	6.53					19	2.36	36
		10	2015-2021	F	Feeding di	N	CM	N	N	N	II	CE+	N	N	N	N	0.6	Y	Y	Y	N	SR	120	1	Y	80	1				Y	N	N	N	N	20	0.36	15	1.8					12	-0.2	46
1			2015-2021	F	DOE	N	NCM	N	N	N	II	CE+	N	Y	Y	N	0.64	Y	Y	Y	N	SR , 2:1 AV	200	4.6		110	2.3				Y	Y	N	N	N	60	4.48	49	5.7					39	4.38	37
16			11/2015-1	M	DOE	N	NOT KNOV	N	N	N	III	CE+	Y	N	Y	N	0.6	Y	Y	Y		SR	170	1.47		120	2				Y	Y	N	Y	N	68		54						42		40
16			2015	F	DOE	N	NOT KNOV	N	N	N	III	CE+	N	N	N	N	0.9		N	Y	N	JR				110	2.18			NA	N	N	N	N	N	33	-2.13	28	0.79	10	0.77	12	0.90	36	3.42	32
	4		2016	F	DOE	N	CM	N	N	N	II	CE+	N	N	N	N					LAD	SR	140	1.68	Y	90	2				Y	Y	N	N	N	55	7.09	49	8.4					26	3.44	25
8			2016-2019	M	Screening	N	CM	N	N	BMD	II	CE-	N	N	N	N					N	SR	110	-1.05	Y	90	0				Y	Y	N	N	N	46	0.37	35	1.9					30	1.45	46
3			2016	M	Febrile Illn	N	NOT KNOV	N	N	N	II	CE+	Y	N	Y	N	0.6	N	N	Y	N	SR	150	1.66	Y	90	1.5				Y	Y	N	N	N	35	0.67	22	0.66					28	2.93	52
8			2016-2021	M	Family Scr	N	NCM	Y	Y	N	II	CE-	N	N	Y	N	0.58	Y	N	Y	N	SR	160	1	Y	100	1	491	70	N	N	Y	Y	N	N					25	5.75	18	3.59	49	5.61	64
	1.5		2016-2018	M	MURMUR	N	NCM	N	N	Pompe dis	I	CE-	N	N	Y	N	0.7	Y	Y	Y	LAD	SR	80	-1	Y	90	2	481	80	Y	Y	Y	N	N	N					14	4.85	15	5.39	17	1.06	60
3			2016	F	Febrile Illn	N	NCM	N	N	N	II	CE+	Y	Y	Y	N	0.65	Y	Y	Y	N	SR	150	1.5	Y	90	1.5				Y	N	Y	N	N	49	5.17	42	6.67					25	2.75	31
6			2018	M	DOE	N	NCM	N	N	N	III	CE+	N	Y	N	N	0.8	Y	Y	Y	N	SR	200	4.29		100	1.5				Y	Y	Y	N	N	64	4.89	57	6.74					31	2.62	23
	3		2016	M	Family Scr	N	CM	Y	Y	N	III	CE+	Y	N	Y	N	0.6	Y	Y	Y	N	SR	140	1.68		140	4.73				Y	Y	N	N	N	32	3.87	26	5.22					20	2.78	40
1.5			2016	M	DOE	N	NCM	N	N	N	II	CE+	Y	Y	Y	N	0.7	Y	Y	Y	N	SR	140	1.33		120	2				Y	Y	Y	N	N											25
	6		2016-2019	M	Incidentall	N	NCM	N	N	N	II	CE-	N	N	Y	N	0.5	N	N	N	N	SR	120	1	Y	100	2	413	49	NA	Y	N	Y	N	N					15	2.88	11	0.21	25	0.76	80
		2	2016	M	Respirator	N	NOT KNOV	Y	Y	N	I	CE-	N	Y	N	N						NA									NA	NA	NA	NA	N	22	2.39	18	4.1					9	-1.65	39
15			2016-2021	F	DOE	N	NCM	Y	N	N	II	CE+	Y	Y	N	N	0.6	Y	Y	Y	N	SR	120	-1		120	2				Y	Y	N	N	N	67	4.27	63	6.66					46	4.6	12
13			2016-2017	M	DOE	N	NCM	N	N	N	III	CE+	Y	N	N	S4+	0.74	Y	Y	Y	LAD	SR	208	4	Y	180	7.33				Y	N	N	Y	N	73	5.13	60	6.27					55	5.86	24
1			2016-2017	F	DOE	N	NCM	N	N	N	II	CE+		Y	Y	N	0.57	Y	Y	Y	N	SR			Y	120	2.3				Y	Y	N	N	N	26	-0.41	17	0.12					21	1.93	34
		1	2016-2019	F	Antenatall	N	NCM	N	N	Unclassifie	II	CE-	N	Y	Y	N	0.7	Y	N	Y	LAD	SR	180	4	Y	100	2	400	50	N	Y	Y	N	N	N	9	-3.05	4	-5.06	8	4.16	7	3.47	7	-1.56	80
13			2011-2016	F	NA	N	NOT KNOV	N	N	N	II	CE+	N	N	N	N	0.6	Y	N	Y	LAD	SR	180	2	Y	90	1				Y	N	N	N	N	53		42						40		43
6			2016-2018	F	Screening	N	NCM	Y	Y	N	III	CE-	N	Y	N	N	0.65	Y	N	Y	N	SR	220	5.47	Y	90	1				Y	Y	N	N	N	58	4.26	48	5.66					41	4.81	33
1.5		0.6	2016-2018	M	Recurrent	N	NCM	N	N	N	II	CE-	N	N	N	N	0.55	Y	Y	Y	N	SR	100	0		90	1				Y	N	N	N	N	36	1.75	30	3.75					16	-0.59	38
	3		2016-2021	M	MURMUR	N	NCM	N	N	Noonan S	I	CE+	N	N	N	Y	0.65	Y	N	N	LAD	SR	130	1	Y	90	2	416	56	N	Y	Y	N	N	N					18	4.40	19	4.51	25	1.29	80
9			2016-2019	M	DOE	N	CM	N	N	N	II	CE+	N	Y	Y	N	0.63	Y	N	Y	N	SR	110	-1	Y	120	2				Y	Y	N	N	N	52	2.26	36	2.65					30	2.02	59
14			2016	F	DOE	N	NCM	N	N	N	III	CE+	N	Y	Y	N	0.58	Y	N	N	N	SR	180	2.2		120	2.4				Y	N	Y	N	N	59	3.16	48	4.63					24	0.14	37
	8		2016-2021	M	DOE	N	CM	N	N	N	II	CE+	N	N	N	N	0.7	Y	N	N	N	SR	120	1	Y	110	2.4				Y	Y	N	N	N	44		35						30		45
4.5			2016	F	Febrile Illn	N	NOT KNOV	N	N	N	IV	CE+	Y	Y	N	N	0.7	N	Y	Y	N	SR	110	0		100	2				Y	Y	N	N	N	52		43						34		33
14			2017	F	DOE	N	CM	N	N	N	III	CE-	N	N	N	N	0.7	Y	Y	Y	RAD	SR	180	2	Y	140	3.75				N	Y	N	Y	N	55	2.51	50	5					29	1.52	20
2.5			2017-2020	M	DOE	N	NCM	N	N	N	III	CE+	N	Y	N	N	0.7	Y	Y	Y		SR	120	0	Y	120	2.5	495	130	N	Y	Y	Y	N	N	23	-3.37	16	-1.73	5	-1.82	7	-0.22	20	0.64	59
13			2017-2018	M	Syncope	Y	NCM	N	N	N	I	CE-	N	Y	Y	N	0.5	N	N	N	N	SR	140	0	Y	110	1	432	45	Y	Y	Y	N	N	N					11		9				66
4			2017	F	DOE	N	NCM	N	N	N	III	CE+	Y	N	Y	N	0.65	Y	N	N	N	SR	160	2		90	1				Y	Y	Y	N	N	61	4.65	50	5.89					38	4.2	26
1			2017	M	DOE	N	NCM	N	N	N	II	CE+	Y	Y	Y	N	0.7	Y	Y	Y	N	SR	140	1	Y	90	0				Y	y	y	N	N	45	6.3	39	7.6					17	1.1	30
	5		2017	F	DOE	N	NCM	N	N	N	III	CE+	N	N	Y	N	0.65	N	N	N	LAD	SR	80	-2		120	2.4				Y	Y	N	N	Y	38		34						14		26
3			2017	M	Febrile Illn	N	NCM	N	N	N	II	CE-	Y	N	N	N	0.5	N	Y	Y	N	SR	200	4.4	Y	90	1.5				Y	Y	Y	N	N	45		42								13
1			2017-2021	M	Feeding di	N	NCM	N	N	N	I	CE-	N	N	N	N	0.5	N	N	Y											NA	NA	NA	NA	N	41	2.58	32	3.88					24	2.02	45
1.5			2017	F	Feeding di	N	CM	N	N	N	II	CE+	Y	Y	N	N	0.66	Y	Y	Y	N	SR	100	0	Y	90	1.5				Y	N	N	N	N	37		33						22		25
2.5			2017-2021	F		N	NCM	Y	Y	N	II	CE-	N	N	Y	N	0.6	N	N	N	N	SR	120	0	Y	140	4.13	432	60	N	Y	Y	N	N	N					14		11		33		72
	8		2017-2021	m	DOE	N	NCM	N	N	N	III	CE+	N	Y	N	N	0.62	Y	Y	Y	N	SR	120	1	Y	90	2	506	80	Y	Y	N	Y	N	N	32	2.82	21	2.7	5	-0.73	5	-1.86	21	2.55	
2			2017-2021	F	Recurrent	N	NCM	N	N	N	I	CE+	N	Y	N	N	0.6	N	N	N	N	SR	120	0	Y	90	1.6				Y	N	N	N	N	31	0.83	21	1.38					19	1.08	60
16			2017	M	Incidentall	Y	NOT KNOV	N	N	N	II	CE+	N	N	Y	S4+	0.55	N	Y	N	N	SR	100	-2	N	100	1	486	55	Y	Y	N	N	N	N					16	2.59	17	2.29	35	2.60	78
1			2017-2019	M	Hyperdyna	N	NCM	NOT KNOV	NOT KNOV	N	I	CE-	N	Y	Y	S4+	0.65	Y	N	Y	N	SR	140	1	Y	90	2	479	60	Y	Y	N	N	N	N					18	4.24	17	3.56	29	2.16	68
10		2	2017-2019	M	DOE	Y	NCM	N	Y	N	II	CE-	N	N	Y	N	0.47	N	N	Y	N	SR	100	-1	Y	90	1	508	70	N	Y	N	N	N	N					29	5.75	15	1.38	39	3.31	84
11			2017-2014	F	DOE	N	NOT KNOV	N	N	N	II	CE-	N	N	Y	S4+	0.58	Y	Y	N	LAD	SR	180	2	Y	90	1	490	50	Y	N	N	N	N	N					36	6.97	20	3.33	42	3.87	88
	6		2017-2019	M	NA	N	NCM	N	N	N	II	CE-	Y	N	Y	N	0.64	Y	N	Y	N	SR	200	4.7	N	120	2	490	40	N	Y	Y	Y	N	N	55	4.69	48	6.43					26	2.13	24
		7	2017-2021	M	Feeding di	N	NCM	N	N	N	II	CE-	N	N	N	N	0.6	Y	Y	Y	N	SR	200	4.6		90	1.5				Y	N	N	N	N	34		26						18		52
	5		2017-2021	M		N	NCM	N	N	N	II	CE-	Y	N	N	N	0.58	Y	Y	N	N	SR	120	1	Y	120	2.4				Y	N	N	N	N	37	4.63	33	6.5					15	0.37	23
5			2017-2021	F	Family Scr	N	NCM	Y	N	N	II	CE-	N	N	Y	S4+	0.55	Y	N	Y	N	SR	180	3.35	Y	90	0	485	46	Y	Y	Y	N	N	N					18	2.95	26	4.76	25	0.26	86
	6		2018	F	Recurrent	N	NCM	N	N	N	II	CE+	N	N	N	N	0.72	Y	Y	N	N	SR	120	1		100	2				Y	N	Y	N	N	46		43						24		17
1.5			2018-2019	M	Pre excitat	N	NOT KNOV	NOT KNOV	NOT KNOV	N	II	CE+	N	N	N	N	0.7	Y	N	Y	N	SR	100	-2		120	2.4				Y	N	N	N	Y	38	3.52	33	5.47					22	2.33	24
	2		2018-2021	F	DOE	N	NCM	N	N	N	III	CE+	N	N	Y	N	0.65	Y	N	N	N	SR	100	0		100	2				N	N	N	N	N	29	4.64	26	6.6					16	2.14	25
5			2018-2021	M	Palpitation	Y	NCM	N	N	N	II	CE-	N	N	N	N	0.6	Y	Y	N	N	SR	160	1		120	2				Y	N	N	N	N	54	1.76	31	0.78					24	-0.29	71
5			2018-2019	M	Right hear	N	CM	N	N	N	III	CE-	N	Y	N	N	0.8	Y	Y	Y	LAD	SR	200	4.27	Y	120	2	441	20	Y	N	N	Y	Y	N	25	-3.21	19	-0.91	8	0.45	10	0.88	38	4.81	45
	4		2018	M	DOE	N	NCM	N	N	N	III	CE+	N	N	Y	N					N	SR	120	1	N	100	2				N	Y	N	N	N	53	9.01	47	9.9					30	5.6	25
18			2018	M	DOE	N	NOT KNOV	N	N	N	II	CE+	Y	Y	Y	N	0.66	Y	Y	Y	N	SR	180	1	Y	100	2.36			N	Y	N	Y	N	N	73		70						36		11
3			2018	M	Febrile Illn	N	CM	N	N	N	III	CE+	Y	Y	Y	N	0.71	Y	Y	Y	N	SR	120	0	Y	120	2.4				Y	N	N	N	N	43	4.97	39	6.97					35	5.73	15
		15	2018-2021	F	Respirator	N	NOT KNOV	N	N	N	II	CE-	N	N	N	N	0.7	Y	Y	Y											NA	NA	NA	NA	N	48	5.22	40	6.49					26	3.17	35
2			2018	M	DOE	N	NCM	N	N	N	II	CE+	Y	Y	Y	N	0.8	Y	Y	Y	N	SR	180	0		100	1				N	Y	Y	N	N	55	6.32	50	8.05					30	4.04	21
		10	2018	F	Respirator	N	CM	N	N	MPS Type	II	CE+	Y	Y	Y	N	0.6	Y	N	Y	N	SR	100	0	Y	90	2			Y	Y	N	N	N	N	34	6.1	26	6.49					23	4.6	46
17			2018-2021	M	DOE	N	NCM	Y	Y	N	II	CE-	N	N	Y	N	0.6	Y	Y	Y	LAD	SR	200	3.05	Y	90	0	432	65	Y	Y	Y	Y	N	N					26	5.16	17	2.21	29	1.21	54
		20	2018	M	DOE	N	CM	N	N	Unclassifie	II	CE-	N	N	Y	N	0.7	Y	Y	Y	LAD	SR	120	1	N	100	2			NA	Y	N	N	N	N	NA										
15			2018	M	DOE	N	NCM	N	N	N	III	CE+	Y	N	Y	N	0.7	Y	N	Y	N	SR	160	1		90	-1				NA	N	N	N	N	57	1.64	52	4.36					47	3.96	20
1			2018-2019	F	Febrile Illn	N	NCM	N	N	N	II	CE+	Y	N	N	N	0.65	Y	N	Y	N	SR	100	-1		90	1			Y	Y	N	N	N	N	37	2.66	30	4.25					21	1.68	40
	9		2018-2021	F	DOE	N	NCM	N	N	N	II	CE+	Y	N	N	N	0.7	Y	N	Y	N	SR	140	1.5	Y	100	2				Y	N	N	N	N	36	4.51	28	5.38							42
1			2018	F	Arrythmias	Y	NOT KNOV	N	N	N	III	CE-	N	N	N	N	0.7	Y	Y	Y		SR	120	0	N	80	1	437	40	N	NA	Y	N	N	N	27	0.26	21	2.02							47
3			2018-2020	M	Incidentall	N	CM	N	N	N	II	CE+	N	N	Y	N	0.6	Y	Y	N	RAD	SR	120	0		100	2	479	50	N	N	Y	Y	N	N	33	-0.01	21	0.22	9	1.25	8	-0.37	36	4.64	60
1			2018-2020	F	Febrile Illn	N	NCM	N	N	N	III	CE+	Y	N	Y	N	0.68	Y	Y	Y	N	SR	120	0		80	1				Y	N	N	N	N	47		40						22		31
4			2018-2021	F	Less active	Y	NCM	N	N	N	II	CE-	N	N	Y	N	0.7	Y	Y	Y	N	As Vp	120	0		160					Y	N	N	N	N	80	5.97	70	7.43					37	2.99	26
		10	2018-2021	M	ANTENATA	N	NCM	N	N	N	II	CE-	N	N	Y	N	0.8	Y	Y	Y		SR	100	0	N	90	2	474	30	N	Y	N	N	N	N					16	4.79	15	4.33	NA		75
		15	2018-2021	M	Family Scr	N	CM	Y	Y	Unclassifie	II	CE-	Y	N	N	N	0.6	Y	Y	N	N	SR	100	0	Y	90	2	453	57	N	N	Y	N	N	N					7	2.26	7	0.79	12	-0.92	66
7			2019	F	DOE	N	NCM	N	N	Congenita	II	CE-	Y	Y	N	N	0.6	N	Y	Y	LAD	SR	200	4.29		80	-1				N	Y	Y	Y	N	42	2.33	37	4.64							27
15			2019	M	DOE	N	CM	Y	Y	Childhood	II	CE-	N	N	Y	S4+	0.45	Y	N	Y	N	SR	160	1	N	100	1	500	45	N	Y	Y	N	N	N					28	4.84	13	-0.4	43	3.19	78
10			2019	M	Presyncop	N	NOT KNOV	Y	Y	N	I	CE-	N	N	N	S4+	0.6	Y	Y	N	N	SR	200	3.52	N	120	2	432	40	N	Y	Y	N	N	N					27	5.33	16	1.77	38	3.09	66
3			2019-2020	F	Febrile Illn	N	CM	N	N	N	II	CE-	N	N	Y	N	0.65	Y	N	Y	N	SR	180	3.33		100	2				N	Y	Y	Y	N	42	4.12	37	6.06					23	2.43	25
12			2019-2021	F	Incidentall	N	NCM	N	N	Noonan S	I	CE-	N	N	Y	N	0.5	N	N	N	N	SR	160	1	N	100	2	465	30	N	Y	N	N	N	N					22	4.97	12	0.81	NA		87
1			2019	F	Incidentall	N	NCM	N	N	N	III	CE-	Y	N	N	N	0.65	Y	N	Y	LAD	SR	160	2		100	2				Y	Y	N	N	N	46	5.94	41	7.59					19	1.57	24
1			2019	M	DOE	N	NOT KNOV	NOT KNOV	NOT KNOV	N	II	CE-	N	Y	N	N					N	SR	160	2.58	N	120	2			N	Y	Y	N	N	N	53	1.99	45	4.05	10	0.17	10	-1.05	44	4.34	24
1			2019-2021	F	DOE	N	NCM	N	N	Noonan S	I	CE+	N	N	N	N	0.47	N	N	N	LAD	SR	160	2.5	Y	100	2.46	452	35	N	N	N	N	N	N					16	2.93	9	-1.49	27	1.06	66
	3		2019	M	Feeding di	N	NCM	Y	Y	N	II	CE-	N	N	N	N	0.55	Y	N	Y	LAD	SR	160	2	Y	100	2			N	Y	N	Y	Y	N	25	2.5	22	4.76					18	2.62	30
1.8			2019	F	DOE	N	CM	N	N	Unclassifie	III	CE+	Y	N	Y	N	0.73	Y	Y	Y	N	SR	160	2		80	1				Y	N	N	N	N	41	3.72	30	4.28					24	2.64	35
8			2019-2021	M	Less active	N	NOT KNOV	N	N	N	II	CE+	N	Y	Y	S4+	0.7	Y	Y	Y	N	SR	180	2		120	2.2	508	70	Y	Y	Y	Y	N	N	30	-1.62	19	-1.1	11	2.07	12	1.95	35	4.09	57
1			2019-2021	M	Family Scr	N	CM	N	Y	N	I	CE+	N	N	N	N	0.7	Y	Y	Y	N	SR	160	2.3	Y	100	2				Y	N	N	N	N	32	2.71	27	4.61					18	1.41	37
		10	2019-2021	F	Feeding di	N	NCM	N	N	N	II	CE-	Y	Y	Y	N	0.69	Y	Y	N	N	SR	140	1	Y	100	2.3	465	52	N	Y	N	N	N	N					11	4.54	8	2.56	NA		70
2			2019-2020	F	DOE	N	NCM	N	N	N	II	CE+	Y	N	N	N	0.65	N	N	Y	N	SR	140	1	Y	90	0			N	Y	Y	N	N	N	45	5.79	35	6.4	6	0.1	6	-0.87	19	1.61	42
3.5			2019-2021	M	Hyperdyna	N	NCM	N	N	N	II	CE+	Y	Y	Y	N	0.62	N	Y	Y	RAD	SR	140	1	Y	100	2	476	45	N	Y	Y	Y	N	N	32	-0.34	22	0.57	10	1.82	10	1.10	28	2.87	60
	8		2019-2020	F	DOE	N	NCM	Y	N	N	II	CE-	N	N	Y	S4+	0.65	Y	Y	Y	N	SR	180	3.82	Y	100	2.3			NA	Y	Y	N	N	N					21	4.69	18	3.43	44	4.74	76
		6	2019-2021	F	DOE	N	NCM	N	N	N	II	CE+	Y	N	N	N	0.77	Y	Y	Y	RAD	SR	110	0	Y	100	2				NA	N	Y	N	N	54	6.53	47	7.87					20	1.38	28
	3		2019-2021	F	DOE	N	NCM	Y	N	N	II	CE-	N	Y	N	N	0.5	N	N	N	LAD	SR	120	0	N	100	2			N	Y	N	N	N	N	49	3.16	37	4.07	6	-0.91	7	-1.21	24	1.35	50
8			2019-2021	F	Right hear	N	NCM	N	N	N	III	CE+	RV S3+	N	Y	N	0.6	Y	Y	Y	RAD	JR				120					N	N	N	N	N	34/17mm										80
		28	2019-	F	Family Scr	N	NCM	N	N	N	II	CE-	N	N	N	N	0.8	Y	Y	Y											NA	NA	NA	NA	NA											20
	6		2019-2021	M	Family Scr	N	NCM	Y	Y	N	I	CE-	N	N	N	N	0.7	Y	Y	Y	N	SR	120	1	Y	100	2				Y	Y	N	N	N	37	4.78	34	6.91					18	1.74	21
9			2020-2021	M	DOE	N	NCM	Y	Y	N	II	CE+	N	Y	Y	N					N	SR	180	2.5	Y	100	1				Y	Y	Y	N	N	53	3.06	46	5.09							22
	6		2020	F	DOE	N	NCM	N	N	Unclassifie	II	CE-	N	N	Y	N	0.6	Y	N	Y	LAD	SR			Y	100	2.3	405	20	N	Y	Y	Y	N	N					9	2.35	9	1.88	17	0.91	79

SAM	PAH	MR	LV AO GRADIENT	OBS/ NON OBS	HOLTER	NT pro BNP	C MRI LGE Y/N		PERCENTAGE	MAX THCKNESS	MYOCARDIAL MASS	NC:C RATIO	Area of NC	Medications , Digoxin	Lasix	ACE I	Aldactone	Beta blocker	CARDIAC CATH, RA mean	RV Peak	RV ed	PA mean	RPAWedge	LVSP	LV ed	CO	CI	PVR	PVRI	LV AO grad	BB phen	Type_CORRECTED	years of FU	OUTCOMES : AGE AT DEATH	Age at death date of daeth	DEATH FROM HF	SCD	Death from VT	VENTRICULAR ARRYTHMIAS	Atrial/Junctional Arrythmias	AV BLOCK	BBBB	REQUIREMENT OF CRT / ICD / TRANSPLANT	Septal myectomy
		3			not done		Y		Mid myocardial from base to apex thin linear					Y	Y	Y																DCM	6	Recovered		N		N	N	N	N	N	N	
	N	2			not done		NA							N	Y	N	Y	N														RCM	1	Death	4.5, 2015	N	Y	N	N	N	N	N	N	
	N	2			not done		not done							Y	Y	Y	Y	N														DCM	1	Death	4, 2016	Y	N	N	N	N	Y	N	Y	
	Y	0			not done		N		NO LGE Diastolic dvs conc LVH					Y	Y	N	N	N	18	42	20	28	23	96	30	1.8	1.73	2.7	2.88			RCM	6	Doing well					N	N	Y	N	N	
	N	2			not done	2217	N		NO LGE					Y	Y	Y	Y	Y	7	40	10	29	23			2.57	1.99	2.33	3.16			DCM	1	Death	19, 2015	Y	N	N	N	N	N	N	N	
Y	N	3	37	OBS	Normal	6847	Y			36/42				N	N	N	N	Metoprolol														HCM	7	Doing well					N	N	N	N	Y	
	N	2			Normal		not done							Y	Y	Y	N	Y														DCM	1	Not known					N	N	N	N	N	
Y	N	0	110	OBS	Normal		ND											Metoprolol														HCM	4	Not known					N	N	N	N	N	
	Y	3			not done		Y							y	Y	Y	y	N														DCM	0.1	Not known					N	N	N	N	Y	
	N	2			not done	4528	N							Y	Y	Y	Y	Y														DCM	3	Death	17, 2019	N	Y	N	N	N	Y	Y	N	
	N	2			not done		not done							Y	Y	N	N	N														DCM	0.1	Death	1.1, 2015	N	N	Y	Y	N	N	N	Y	
	N	3			not done		not done							Y	Y	N	N	N														DCM	5	Recovered										
Y	N	2		NON OBS	Normal		Y			23				N	N	N	N	Metoprolol														HCM	6	Doing well					N	N	N	N	N	
Y	N	3	110	OBS	not done		N											Ppnl														HCM	3	Not known					N	N	N	N	N	
N	N	0		NON OBS	not done		ND							N	N	N	N	N														HCM	5	Death	5 , Dec 202	N	Y	N	N	N	N	N	N	
N	N	1		NON OBS	not done		ND							N	N	N	N	N														HCM	1	Not known					N	N	N	N	N	
	N	1			not done	2651	not done							Y	Y	N	N	N														LVNC	1	Not known					N	N	N	N	N	
	N	0			not done	2605	not done							Y	Y	Y	N	N														DCM	6	Recovered					N	N	N	N	N	
	N	3			not done	528	not done							y	y	y	y	Y														DCM	6	Persistent LV dysfunction					N	N	Y	N	N	
	N	3			not done		N		NO LGE					Y	Y	Y	Y	Y														DCM	0.2	Death	16.1 , 201	Y	N	N	N	N	N	N	Y	
	N	1			not done		NA							Y	Y	N	N	N														RCM	0.2	Not known					N	Y	N	N	N	
	N	2			not done		not done							y	y	y	y	N														DCM	2	Death	3, 2018	Y	N	N	N	N	N	N	N	
	N	1			NORMAL		not done							N	N	y		Y														DCM	3	Persistent LV dysfunction					N	N	N	N	N	
	N	0			not done		not done							N	Y	Y	N	N														DCM	6	Recovered					N	N	N	N	N	
Y	N	3+	64	OBS	Normal	1730	Y		30	21	298							Bisoprolol														HCM	6	Doing well					N	N	N	N	N	
N	N	NA		NON OBS	not done		ND							N	N	N	N	Ppnl														HCM	2	Not known					N	N	N	N	N	
	N	3			not done	9000	not done							Y	Y	Y	Y	Y														DCM	0.1	Not known					N	N	N	N	N	
	N	3			not done		not done							Y	Y	Y	Y	Y														DCM	1	Not known					N	N	Y	N	Y	
	N	3			not done		N							y	y	N	N	N														DCM	3	Death	3, 2019	Y	N	N	N	N	N	N	N	
	Y	4			not done		not done							Y	Y	Y	N	Y														DCM	1	Death	2, 2016	Y	N	N	N	N	N	N	N	
N	N			NON OBS	not done		ND							n	n	n	n	Metoprolol														HCM	3	Doing well					N	N	N	N	N	
	N	1			not done		not done							NA																		DCM	1	Not known					N	N	N	N	N	
LV Dp/Dt 4	N	2			not done	3950	N		NO LGE					Y	Y	Y	Y	Y	8	40	8	29	11		15	2.15	1.5	8.3	11.9			DCM	5	Recovered					N	N	N	N	N	
	N	3			recurrent V	4130	not done							Y	Y	Y	Y	Y	15	68	15	40			30	2.62	1.54	4.19	5.69			DCM	1	Death	14, 2017	N	N	Y	Y	N	Y	Y	Y	
	N	3			not done		not done							N	Y	Y	Y	Y														DCM	1	Not known					N	N	N	N	N	
	Y	1			NORMAL		Y							n	n	n	n	N														LVNC	3	Not known					N	N	N	N	N	
	N	2			not done		not done							N	Y	N	Y	Y														DCM	5	Not known					N	N	N	N	N	
	Y	2			NORMAL		Y		yes , Sub endocardial in IVS mid and basal					y	y	y	N	Y														DCM	2	Not known					N	N	N	N	N	
	N	1			not done		not done							Y	Y	N	N	N														DCM	2	Not known					N	N	N	N	N	
Y	N	3	126	OBS	Normal	5830	ND											Metoprolol														HCM	5	Doing well					N	N	N	N	N	
	N	3			not done	7990	Y		LGE > 50% Transmural enhancement IW/IS/IL					Y	Y	Y	Y	Y														DCM	3	Death	12, 2019	N	Y	N	N	N	N	N	N	
	N	2			not done	1220	N		NO LGE					Y	Y	Y	Y	Y														DCM	5	Recovered					Y	N	N	N	Y	
	N	3			not done	4100	N		NO LGE					Y	Y	Y	Y	N														DCM	5	Recovered					N	N	N	N	N	
	N	2			not done	10100	not done							N	Y	Y	Y	Y														DCM	3	Death	7.5, 2019	Y	N	N	N	N	N	N	N	
	N	2			NORMAL	4440	Y		LGE Transmural enhancement RV , LV free wall IVS					Y	Y	Y	N	Y	10	30 ed 18		18				2.42	1.72	0.82	1.06			DCM	1	Not known					N	N	N	Y	N	
	Y	0			normal	7890	Y		Subtle LGE in IAS ,MV, TV Not in ventricular myocardium					Y	Y	Y	Y	N														RCM	3	Doing well					N	N	N	N	N	
Y	N	1		NON OBS	Normal	4250	Y			22								Bisoprolol														HCM	1	Death	14, Dec 20	N	Y	N	N	N	N	N	N	
	N	3			not done	9320	not done							Y	Y	Y	Y	Y	10	42 ed 13		32	22		28	1.75	1.92	6.64	5.64			DCM	0.3	Death	9.5, 2018	N	N	Y	Y	N	N	N	Y	
	Y	2			not done		not done							N	Y	Y	Y	Y														DCM	4	Death	4.5, 2019	Y	N	N	N	N	N	N	N	
	N	2			not done		not done							N	Y	Y	Y	Y														DCM	1	Death	1, Jan 201	Y	N	N	N	N	N	N	Y	
	N	2			not done		not done							N	Y	Y	Y	Y														DCM	1	Not known					N	N	Y	N	Y	
	N	3			not done		not done							Y	N	Y	N	Y														DCM	4	Recovered					N	N	N	N	N	
	N	3			not done		not done							Y	Y	Y	Y	N														DCM	1	not known					N	N	N	N	Y	
Y	N	3	96	OBS	Normal	768	N											Bisoprolol														HCM	5	Doing well					N	Y	Y	Y	N	
	Y	1			normal	1960	Y		Subendocardial enhancement IVS in basal, mid and apic					n	y	y	y	N														RCM	5	Doing well					N	N	N	N	N	
	N	1			not done		not done							N	Y	N	N	N														DCM	5	Persistent LV dysfunction					N	N	N	N	N	
Y	N	0	84	OBS	Normal	960	ND											Metoprolol														HCM	10	Not known					N	N	N	N	N	
N	N	1		NON OBS	Normal		ND							N	N	N	N	Metoprolol														HCM	2	Doing well					N	N	N	N	N	
y	N	1	62	OBS	Normal		Y			29	147			N	N	N	N	Metoprolol														HCM	2	Doing well					N	N	N	N	Y	
Y	N	3	70	OBS	Normal		Y			32	216			N	N	N	N	Metoprolol														HCM	2	Not known					N	N	N	N	N	
	N	2			NORMAL		Y					1.96		N	Y	Y	Y	Y														LVNC	3	Death	2.5, 2019	Y			N	N	N	N	N	
	N	1			not done	212	not done							N	N	Y	Y	N														DCM	2	Recovered					N	N	N	N	N	
	N	0			not done	587	not done							Y	Y	Y	N	Y														DCM	5	Recovered					N	N	N	N	N	
Y	N	2	136	OBS	not done		ND							n	n	n	n	Metoprolol														HCM	5	Doing well					N	N	Y	N	N	
	N	3			not done		not done							N	Y	Y	N	Y														DCM	4	Recovered					N	N	N	N	N	
	N	2			not done		not done							y	y	y	y	Y														DCM	2	Not known					N	Y	N	N	Y	
	Y	3			not done	35000	not done							Y	Y	Y	N	N														DCM	4	Persistent LV dysfunction					N	N	N	N	Y	
		1			Long sinus pause		Normal LV function		LGE in subendocardium of interatrial septum, RA wall,Po					N	N	N	N	N														Atrial Myopathy	4	Doing well No prob					N	N	N	N	Y	
RV Dp/Dt :	Y	1			normal		N		NO LGE					Y	Y	N	Y	N	22	53	25	33		96	25			3	2.01			RCM	4	Death	6, 2019	N	Y	N	N	N	Y	N	N	
	N	3			not done	3350	not done							Y	Y	Y	Y	Y														LVNC	1	Death	10 months	Y			N	N	N	N	N	
	Y	3			not done	4080	Y		Patchy LGE in basal and mid levels of LC in AS/AL segmen					Y	Y	Y	Y	Y														DCM	0.1	Not known					N	N	N	Y	N	
	N	3			not done	3870	not done							N	Y	Y	N	N														DCM	0.1	Not known					N	N	N	N	N	
	N	1			not done		not done							N	Y	Y	Y	Y														DCM	4	Recovered					N	N	N	N	N	
	Y	2			not done		not done							N	y	y	Y	N														DCM	1	Not known					N	N	N	N	Y	
	N	4			not done		not done							N	Y	Y	N	Y														DCM	4	Recovered					N	N	N	N	N	
N	N	1	30	OBS	LBBB		Y		25	28	96				N	N	N	Metoprolol														HCM	4	Doing well					N	N	Y	N	N	
Y	N		169	OBS	not done		ND											N														HCM	1	Death	2 months ,	Y	N	N	N	N	N	N	N	
	N	3			not done	16600	Y		Homogenous myometrial hyperenhancement					Y	Y	Y	Y	N	15			32			28	2.82	1.61	4	7			DCM	1	Death	15.5, 2018	N	N	Y	Y	N	N	N	Y	
	N	2			not done	1810	not done							y	y	y	n	N														DCM	3	Recovered					N	N	N	N	Y	
	N	3			not done		not done							N	Y	Y	Y	Y														DCM	3	Recovered					N	N	N	N	N	
	N				not done		N					2.8		N	Y	Y	N	Y														LVNC	1	Death	2, 2019	Y			Y	N	Y	Y	N	
	Y	0			normal	4000	Y		LGE in IAS , Basal IVS Poor suppression of myocardium					n	y	y	Y	N	22	54	18	35	23	100	18	3.33	5.37	3.60	2.27			RCM	4	Doing well					N	N	N	N	N	
LVdp/Dt 9	N	3			not done	7940	not done							Y	Y	Y	Y	Y														DCM	4	Death	3, 11/2020	N	Y	N	N	N	N	N	N	
	N	3			not done	1670	not done							Y	Y	Y	Y	Y														DCM	4	Persistent LV dysfunction					N	N	N	N	Y	
Y	N	3	100	OBS	not done		ND							N	N	N	N	Metoprolol														HCM	3	Doing well					N	N	N	N	N	
N	N	0		NON OBS	not done		ND							N	N	N	N	Metoprolol														HCM	3	Death	3, 2021	N	Y	N	N	N	N	N	N	
	Y	2			not done		not done							N	Y	Y	N	Y														DCM	1	Persistent LV dysfunction					N	N	Y	N	N	
Y	N	2	30	OBS	Normal	40	Y			28	225			N	N	N	N	Metoprolol														HCM	1	Doing well					N	N	N	N	Y	
N	N	0		NON OBS	not done	3270	N							N	N	N	N	Metoprolol														HCM	3	Doing well					N	N	Y	N	N	
	N	2			not done		not done							N	Y	Y	Y	N														DCM	2	Recovered					N	N	Y	N	N	
Y	N	0		NON OBS	Normal	not done	N			18				N	N	N	N	Ppnl														HCM	2	Doing well					N	N	N	N	N	
	N	2			not done		not done							N	Y	Y	Y	Y														DCM	2	Death	2, 2020	Y	N	N	N	N	N	N	Y	
	Y	4			not done		not done							N	Y	N	N	Y														LVNC	1	Not known					N	N	N	N	N	
Y	Y	0		NON OBS	not done	148	ND							no drugs				N														HCM	2	Doing well					N	N	N	N	N	
	Y	2			not done	35000	not done							Y	Y	Y	N	Y														LVNC	2	Death	1, 2020	Y			N	N	N	N	N	
	N	2			not done	19900	not done							Y	Y	Y	Y	Y														DCM	0.3	Not known					N	N	N	N	N	
	N	1			normal	8410	N							N	Y	N	Y	N	7	28	6	13	10	84	12	1.6	1.2	1.98	1.34			RCM	3	Doing well					N	N	N	N	N	
	N	0			not done	691	Y		subendocardial					y	y	y	n	N														DCM	2	Recovered					N	N	N	N	N	
N	N	0		NON OBS	not done		ND							n	n	n	n	N														HCM	3	Doing well					N	N	N	N	N	
	N	3			not done	1262	Y							y	y	y	y	N														LVNC	3	Not known					N	N	N	N	N	
	Y	1			normal	2740	NA							N	y	y	N	y	6	54	16	34	22	90	16	2.3	3.70	5.2	3.23			RCM	3	Doing well					N	N	N	N	Y	
Y	N	4	59	OBS	Normal		Y		<10	29				N	N	N	N	Metoprolol														HCM	2	Doing well					N	N	Y	N	N	
	N	3			not done		not done							Y	Y	Y	Y	Y														DCM	3	Persistent LV dysfunction					N	N	N	N	N	
	N	1			not done	227	not done							N	N	Y	N	Y														LVNC	3	Doing well					N	N	N	N	N	
		2			Junctional tachycardi		N							n	n	n	Y															ARVD	3	Doing well					N	N	N	N	Y	
	N	2			not done		not done							NA																		DCM	2	Not known					N	N	N	N	Y	
	Y	3			not done		not done							N	Y	Y	N	Y														DCM	3	Persistent LV dysfunction					N	N	N	N	N	
	N	4			not done		not done							N	Y	Y	Y	Y														DCM	1	Persistent LV dysfunction					N	N	N	N	N	
N	N	0		NON OBS	not done		ND							n	n	n	n	N														HCM	1	Doing well					N	N	N	N	N	

AGE in yrs	Age in months	Age in days	Years on FU	SEX	MODE OF PRESENTATION / HISTORY	syncope	CONSANGUINITY	FAMILY HISTORY OF CMP	FAMILY H/O SCD	SYNDROMES	NYHA FC	CVS	S3	P2	PSM/ESM	Double apical impulse	CXR	LAE	RAE	PVH	Axis LAD/ RAD/N	Rhythm
17			2020	M	DOE	N	NCM	N	N	DMD	III	CE+	N	Y	N	N	0.63	Y	N	Y	N	SR
12			2020	F	Febrile Illn	N	CM	Y	Y	N	II	CE-	N	N	N	N	0.7	Y	Y	Y	N	SR
8			2020	M	Family Scr	N	NCM	Y	Y	N	I	CE-	N	N	N	N					N	SR
17			2020	M	DOE	N	NOT KNOV	NOT KNOV	NOT KNOV	N	II	CE+	Y	Y	Y	N					N	SR
3			2020-2021	M	Right hear	N	NCM	N	N	N	III	CE+	N	Y	N	N	0.7	Y	Y	N	N	SR
	2		23-11-202(	F	Syncope	Y	NCM	Y	N	N	II	CE-	N	N	N	N	0.68	Y	N	Y	N	SR
15			2020	F	AOE	N	NOT KNOV	N	N	N	II	CE-	N	N	Y	N	0.5	Y	Y	Y	N	SR
1			2020	M	Febrile Illn	N	NCM	N	N	N	III	CE+	N	Y	Y	N					N	SR
1			2021	M	DOE	N	NCM	N	N	N	II	CE+	Y	N	N	N	0.7	Y	Y	Y	RAD	SR
10			2021	F	Incidentall	N	NOT KNOV	NOT KNOV	N	N	II	CE+	N	N	N	N	0.57	Y	Y	Y	N	SR
11			2021	M	Palpitatior	N	NCM	N	N	N	II	CE-	N	N	N	N	0.5	N	N	Y	N	SR
8			2021	F	DOE	Y	NCM	N	N	N	II	CE-	N	N	N	N	0.57	Y	N	Y	N	SR
4			2021	M	Incidentall	Y	CM	Y	N	N	II	CE-	N	Y	Y	N	0.6	Y	Y	Y	N	SR
15			2021	F	DOE	N	CM	Y	Y	N	II	CE-	N	Y	N	N	0.5	N	N	N	N	SR
18			2021	M	Clubbing	N	NCM	N	N	N	II	CE-	N	N	N	N	0.5	N	Y	Y	N	SR

AGE in yrs	PR interval	PR interval z score	q wave	QRS d	QRS d z score	QT interval	QT dispersion	ST depression	LVH	LAH	RAH	Low voltage contents	Preexcitation	ECHO 1 , LVIDd	Z SCORE LVIDd	LVIDs	Z score LVIDs	IVS s	IVS , Z SCORE	PW s (mm)	PW , Z SCORE	LAE , Left atrial diameter mean , mm	LAE, Z score	LVEF
17	200	2.3	Y	100	0				Y	Y	Y	N	N	60	3.43	55	5.79					48	5.13	21
12	240	4.5		120	2				N	N	Y	N	N	54		48						32		26
8	160	2	Y	120	2	438	70	N	N	Y	N	N	N					17	3.85	11	0.59	25	1.04	74
17	160	1	Y	100	1				Y	Y	N	N	N	67	4.01	63	6.46					40	3.39	20
3	140	1.5		80	1			NA	NA	Y	Y	Y	N	31	-0.39	22	0.78	15	4.34	13	2.84	32	3.96	58
	140	2	N	80	1	566	85	Y	Y	Y	Y	N	N	24	0.5	18	1.92					15	0.47	52
15	180	2	Y	80	-1			N	Y	Y	Y	N	N					12	1.54	17	2.91	24	0.36	84
1	100	-1	Y	90	1.8				Y	N	N	N	N	61	8.07	54	9.22					30	4.42	25
1	200	4.94	Y	180	8.3	465	52	N	N	Y	Y	N	N					13	1.76	12	0.34	48	5.09	30
10	140	1	N	120	0			N	Y	Y	N	N	N	57	3.31	50	5.36							27
11	180	2		100	1				Y	N	N	N	N	45	0.75	37	2.81							38
8	160	2	N	100	1	460	40		Y	Y	Y	N	N					21	4.5	16	2.42	archived on 05/02 se		65
4	160	2		100	1	475	85	Y	Y	Y	Y	N	N	33	0.11	23	1.03	6	-0.88	6	-2.18	31	3.67	64
15	200	3.26		120	2				Y	N	N	N	N	62	3.02	47	3.98					35	2.28	47
18	200	2.78		100	0	372	43	N	N	N	N	N	N	40	-0.89	28	0.25							57

SAM	PAH	MR	LV AO GRADIENT	OBS/ NON OBS	HOLTER	NT pro BNP	C MRI LGE Y/N		PERCENTAGE	MAX THICKNESS	MYOCARDIAL MASS	NC:C RATIO	Area of NC	Medications , Digoxin	Lasix	ACE I	Aldactone	Beta blocker	CARDIAC CATH, RA mean	RV Peak	RV ed	PA mean	RPAWedge	LVSP	LV ed	CO	CI	PVR	PVRI	LV AO grad	BB phen	Type_CORRECTED	years of FU	OUTCOMES : AGE AT DEATH	Age at death date of daeth	DEATH FROM HF	SCD	Death from VT	VENTRICULAR ARRYTHMIAS	Atrial/ Junctional Arrythmias	AV BLOCK	BBBB	REQUIREMENT OF CRT / ICD / TRANSPLANT	Septal myectomy
	Y	3			not done		not done							Y	Y	Y	Y	Y														DCM	0.1	Death	17, Apr 20	Y	N	N	N	N	N	N	Y	
	N	2			VPC s NSV	4210	Y		transmural					Y	Y	N	Y	Y														DCM	0.3	Recovered					N	N	N	N	N	
N	N	0		NON OBS	Normal		ND											N														HCM	1	Doing well					N	N	N	N	Y	
	Y	3			not done	6650	not done							N	y	n	eplerenon	Y														DCM	1	Death	18, 2021	Y	N	N	N	N	N	N	N	
	N	3			normal	10900	N							N	Y	N	y	n	7	40	8	25	21	90	28	1.72	1.07	2.33	1.49			RCM	0.6	Doing well					N	N	N	N	Y	
	N	0			Not done		N							N	Y	Y	N	Y														LVNC	0.7	Doing well					N	N	N	N	N	
Y	N	2	30	OBS	normal		N											N														HCM	1	Doing well					N	N	N	N	N	
	N	3			not done		not done							N	Y	Y	Y	Y														DCM	1	Persistent LV dysfunction					N	N	N	N	N	
N	N	2		NON OBS	not done	9540	Y		15					N	Y	Y	Y	N														HCM	15	Doing well					N	N	Y	Y	N	
	N	2			not done		N					4.26		N	Y	Y	Y	Y														LVNC	0.4	Doing well					N	N	N	N	N	
	N	2			not done	203	not done							Y	N	N	N	Y														DCM	0.3	Recovered					N	Y	N	N	N	
N	N	2		NON OBS	not done		ND							N	N	N	N	N														HCM	0.3	Doing well					N	N	N	N	N	
	N	1			normal	5368	Y							N	Y	N	Y	N														RCM	0.6	Death	4.5, 2021	N	Y	N	N	N	N	N	Y	
n	N	2			Normal	134	Y		subendocardial					N	N	N	Y	Y														DCM	0.6	Persistent LV dysfunction					N	N	N	N	N	
					NOT DONE	50	N							N	N	N	N	N														ARVD	0.3	Doing well					N	N	N	N	N	