Myocardial Diseases in Thai Children

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Abstract

Myocardial diseases are among the important causes of mortality and morbidity in children. This drew the authors attention to the study of myocardial diseases in children to find out the outcome, factors affecting the outcome, and management strategies. The authors retrospectively studied children who had been diagnosed with primary myocardial diseases at six university hospitals in Thailand from January 1996 to December 2000. The total number of cases was 209 which accounted for 1.2 per cent of cardiovascular diseases in children. The patients' ages ranged from 0.1-15 years. These myocardial diseases included dilated cardiomyopathy (DCM) 45 per cent, acute myocarditis 27.3 per cent, hypertrophic cardiomyopathy (HCM) 18.2 per cent, hypertrophic obstructive cardiomyopathy (HOCM) 8.1 per cent and restrictive cardiomyopathy (RCM) 1.4 per cent. Fifty-six per cent of the patients were female. Congestive heart failure was the most common presenting symptom (75%). Median ejection fraction (EF) of acute myocarditis was 42 per cent (15-79%) which was significantly higher than DCM (33.5%, 10-57%). Serum cardiac troponin T (cTnT) was also significantly higher in acute myocarditis than in DCM (0.08 ng/ml, 0.01-0.16 *vs* 0.01 ng/ml, 0.01-0.10). Within the follow-up period of 1 year (0.1-5.5years), the mortality rates were 18.8 per cent, 17.0 per cent, 5.4 per cent and 33.3 per cent in DCM, acute myocarditis, HCM and RCM respectively. Factors associated with the mortality rate in acute myocarditis were admission to ICU and low left ventricular EF at presentation while IVIG administration and cTnT level did not.

Conclusion : Primary myocardial diseases are uncommon. Most of the patients had compromised cardiovascular reserve. Admission to ICU and low EF were factors that affected the mortality in acute myocarditis while intravenous immunoglobulin administration did not. Mortality rate in the subacute follow-up period was high in all groups.

Key word : Cardiomyopathy, Myocarditis, Myocardial Diseases

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Primary myocardial disease is not uncommon. It has been defined as a disease of the myocardium associated with myocardial dysfunction[1]. There are certain types of myocardial diseases which have specific physiological and pathological characteristics. However, there is a spectrum of diseases which could result in different clinical manifestations and outcomes. A high mortality rate of up to one-third and also morbidity rate of up to one-third with conventional anti-failure treatment has been reported[2]. For decades, many studies have focused on investigational means, supplementary medical and surgical treatment to improve outcomes in adults and a few in pediatric patients. These include serum level of biochemical myocardial enzymes[3] and endomyocardial biopsy in acute myocarditis and DCM; intravenous immunoglobulin (IVIG) administration[4,5] and immunosuppressive drugs administration[6] in acute myocarditis; coenzyme Q10 administration[7] and partial ventriculectomy in DCM; beta blocker, calcium channel blocker[8-10], disopyramide administration[11-14], dual chamber pacemaker implantation[15-17] and surgical myo-myectomy[18-21] in HCM. Better outcomes were reported with these interventions. Some standard regimens for each special type of myocardial diseases have been widely accepted. Most of the studies were done in adult patients. So far, no large study has been done in Thai children with primary myocardial diseases. Thus, a multicenter retrospective study was arranged.

PATIENTS AND METHOD

A multicenter study was designed to recruit a large number of patients with primary myocardial diseases. These included 5 university hospitals i.e. Queen Sirikit National Institute of Child Health, Chiang Mai University Hospital, Siriraj Hospital, Ramathibodi Hospital, Chulalongkorn University Hospital, and Songklanakarin University Hospital. Newly diagnosed pediatric patients with primary myocardial diseases from January 1996 to December 2000 were enrolled into the study. Secondary myocardial diseases such as HIV cardiomyopathy, myocarditis in systemic lupus erythematosus were excluded. Cardiovascular database and records were reviewed. Medical records, cardiac files and hospital files were retrospectively analyzed. Demographic data, presenting symptoms, cardiovascular investigations including serum cardiac troponin T (cTnT), creatine kinase MB fraction (CK-MB), echocardio-

graphy (ejection fraction, EF), pathological data (endomyocardial biopsy or autopsy), management, follow-up period and outcome were recorded.

Statistical analysis

Variables were described as median/range or percentage as appropriate. Association between categorical variables was evaluated using the chi-square test. Comparison of difference of continuous variables between various groups of patients was performed by the Kruskal Wallis test or the Mann-Whitney U test. All significant tests were done in 2-tailed fashion. A p-value < 0.05 was considered as statistically significant.

RESULTS

From January 1996 to December 2000, there were 209 new patients with primary myocardial diseases who were cared for by 5 university hospitals in Thailand. These patients accounted for 1.2 per cent of the total number of 17,417 children with cardiovascular diseases diagnosed at the 6 hospitals during the same period. The diseases comprised of 94 cases (45%) of DCM, 57 cases (27.3%) of acute myocarditis, 56 cases (26.3%) of HCM and 3 cases (1.4%) of RCM as shown in Fig. 1. Fifty six per cent of the patients were female. Seventy two per cent of the patients presented with congestive heart failure. Demographic data, cardiovascular presentations and investigations are demonstrated in Table 1. Endomyocardial biopsy was performed in 5 acute myocarditis patients and 2 each of DCM and RCM. Pathology proved 40 per cent of acute myocarditis patients and 100 per cent for both DCM and RCM. However, within 6 months of follow-up-period the patients with negative pathology were considered cured hemodynamically by functional class and ejection fraction from echocardiogram. Managements other than conventional anti-failure drugs and outcomes are shown in Table 2. There was no difference ($p = 0.11$) in the mortality rate in acute myocarditis patients who were treated with or without IVIG. With the follow-up period of 1 year (0.1-5.5 years), the mortality rates were 18.8 per cent, 17 per cent, 5.4 per cent and 33.3 per cent in DCM, acute myocarditis, HCM and RCM respectively. Age, gender, presenting symptoms did not affect mortality in DCM, acute myocarditis and HCM. The mortality rate in DCM and acute myocarditis was not affected by intravenous inotropes administration, CK-MB level or cTnT level. Admission to ICU, and low EF at first diagnosis clearly affected the mortality in acute myocarditis ($p = 0.01$ and $p = 0.03$).

DISCUSSION

Primary myocardial disease is uncommon. In the present study, it accounted for 1.2 per cent of all the cardiovascular problems in children. Dilated cardiomyopathy has been known as the most common type of cardiomyopathy with the incidence of 8-10 per 100,000 population in the United States and Europe[22]. Hypertrophic cardiomyopathy and restrictive cardiomyopathy were reported as accounting for 20-30 per cent and 5 per cent of cardiomyopathy respectively. To the best of the authors' knowledge, the incidence of myocardial diseases in Thailand has not been reported. In this study, the numbers of various types of myocardial diseases were comparable to reports from the United States and Europe [22]. There was no sexual predominance in any type of myocardial disease.

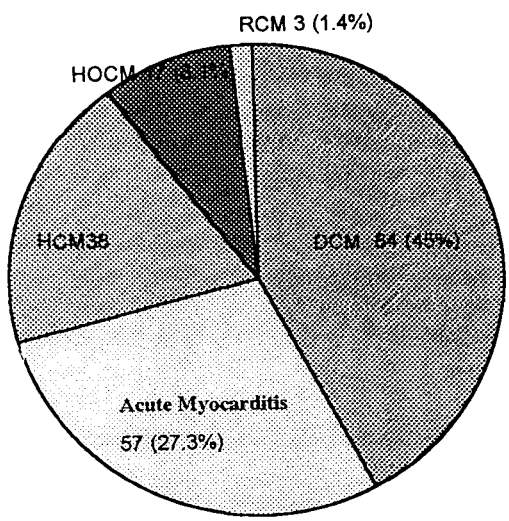

Fig. 1. The number of patients in each type of primary myocardial diseases

HCM = hypertrophic cardiomyopathy
HOCM = hypertrophic obstructive cardiomyopathy
RCM = restrictive cardiomyopathy
DCM = dilated cardiomyopathy

Table 1. Demographic data, investigations and treatment.

	DCM	%	Acute myocarditis	%	HCM	%	HOCM	%	RCM	%
Gender										
Female	51	54.3	38	66.7	18	47.4	8	47.1	2	66.7
Male	43	45.7	19	33.3	20	52.6	9	52.9	1	33.3
Age (yr)	2 (0.1-14.2)		3 (0.1-1.5)		0.5 (0.1-13)		4.5 (0.1-14.5)		4.0 (1.9-7.0)	
Presenting symptoms										
CHF	79	84.1	45	78.9	17	44.7	8	47.1	2	66.6
Cardiogenic shock	3	3.2	1	1.8	-	-	-	-	-	-
Heart block	3	3.2	5	8.8	-	-	-	-	-	-
Tachyarrhythmia	1	1.1	4	7.0	-	-	-	-	-	-
Stroke	1	1.1	2	3.5	-	-	-	-	1	33.3
Other	-	-	-	-	15	39.5	7	41.2	-	-
Investigations (median)										
cTnT (ng/ml)*	0.01 (0.01-0.10)		0.08 (0.0-0.16)		-		-		-	
CK-MB (ng/ml)**	19.5 (4.8-63.0)		30 (2-182)		-		-		-	
EF (%)***	33.5 (10-57)		42 (15-79)		75 (60-90)		65 (60-75)		62 (56-63)	
Treatment No. of case										
IV inotropes	38	40.4	29	50.9	2	5.3	1	5.9	2	66.7
Admit to ICU	35	37.2	23	40.4	3	7.9	1	5.9	1	33.3

* Statistical difference between DCM and acute myocarditis $p = 0.031$

** No difference between DCM and acute myocarditis $p = 0.17$

*** Statistical difference between DCM and acute myocarditis $p < 0.001$

HCM = hypertrophic cardiomyopathy, HOCM = hypertrophic obstructive cardiomyopathy

RCM = restrictive cardiomyopathy, DCM = dilated cardiomyopathy

Table 2. Treatment other than conventional anti-failure drugs.

	DCM	%	Acute myocarditis	%	HCM	%	HOCM	%	RCM	%
IVIG	10	10.6	15	26.3	-	-	-	-	-	-
Pacemaker	2	2.1	2	3.5	-	-	1	5.9	1	3.3
Steroid	5	5.3	6	10.5	-	-	-	-	-	-
Carnitine	2	2.1	-	-	-	-	-	-	-	-
Co-enzyme Q10	4	4.3	-	-	-	-	-	-	-	-
Surgery	1	1.1	-	-	-	-	2	11.8	-	-
B-blocker	-	-	-	-	-	-	1	5.9	-	-
Calcium channel blocker	-	-	-	-	-	-	1	5.9	-	-

DCM = dilated cardiomyopathy, HCM = hypertrophic cardiomyopathy,
HOCM = hypertrophic obstructive cardiomyopathy, RCM = restrictive cardiomyopathy, IVIG = intravenous immunoglobulin

Clinical presentations and diagnosis

The major presenting symptom was congestive heart failure as reported in are other study (2). Practically it is difficult to differentiate DCM from acute myocarditis. Cardiac troponin T level measured at the first presentation has been demonstrated to be significantly higher in acute myocarditis than DCM. This is helpful for early diagnosis and the appropriate line of treatment to be given. A number of studies also supported this finding(23-25). However, the authors found that the level of cTnT in the present study was low (0.08 ng/ml) in comparison to other reports (> 0.1 ng/ml). The smaller myocardial mass in children could account for this discrepancy since most of the previous studies were done in adult patients. Ejection fraction of DCM is significantly lower than acute myocarditis. This finding could assist in making the diagnosis. Endomyocardial biopsy and pathology had very low sensitivity 10-40 per cent(26-28). There was up to a 5.2 per cent risk of cardiac perforation in children with acute myocarditis who were given IV inotropes(29). This data was from 9 cases with a pathological report. There was 40 per cent sensitivity for acute myocarditis, 100 per cent for both DCM and RCM. Practically it is very important to be able to diagnose acute myocarditis considering the disease could be cured within a 6-month-period. Echocardiography is the appropriate diagnostic tool for HCM and RCM. Restrictive cardiomyopathy is very rare in children. In this study, the first 2 cases required pathological reports to confirm the diagnosis. The significant clues to the diagnosis of RCM are marked enlargement of both atria and impaired ventricular diastolic function with slightly decrease or low normal systolic function which can be demonstrated by echocardiogram.

Management

About 40 per cent of DCM and myocarditis patients were admitted to the intensive care unit and 40-50 per cent required IV inotropes. These findings implied the severe hemodynamic compromise in both types of myocardial disease. Pathophysiology is different in each type of primary myocardial disease, which results in a different line of treatment. Carnitine and co-enzyme Q10 (CoQ10) have been accepted as second line drugs for adult DCM(7). It is not a standard regimen since there are only a few studies in pediatric patients. Twenty to thirty per cent of DCM including carnitine deficiency DCM is familial inheritance. In some institutions, carnitine is routinely added to the standard treatment of idiopathic DCM since it is difficult to diagnose carnitine deficiency DCM in Thailand. In adult patients with DCM, beta blocker has been added to the optimal conventional drugs with better long term outcome. The basis of this treatment is the concept that excessive neuroendocrine activation is a strong predictor of mortality(30). Partial left ventriculectomy has also been reported as an option for patients with DCM(31). High dose IVIG has been proposed to improve the recovery of left ventricular function with better survival during the first year of the disease(4,5). In this study, IVIG administration for acute myocarditis was used in some centers and there was no difference in the course of the disease. Many studies could not demonstrate beneficial effects of immunosuppression (prednisolone and IVIG) in acute myocarditis(6,32,33). Heart transplantation is the last option for patients with intractable heart failure.

In the present study, patients with HOCM were treated with standard medication i.e. beta blocker and calcium channel blocker. These 2 medi-

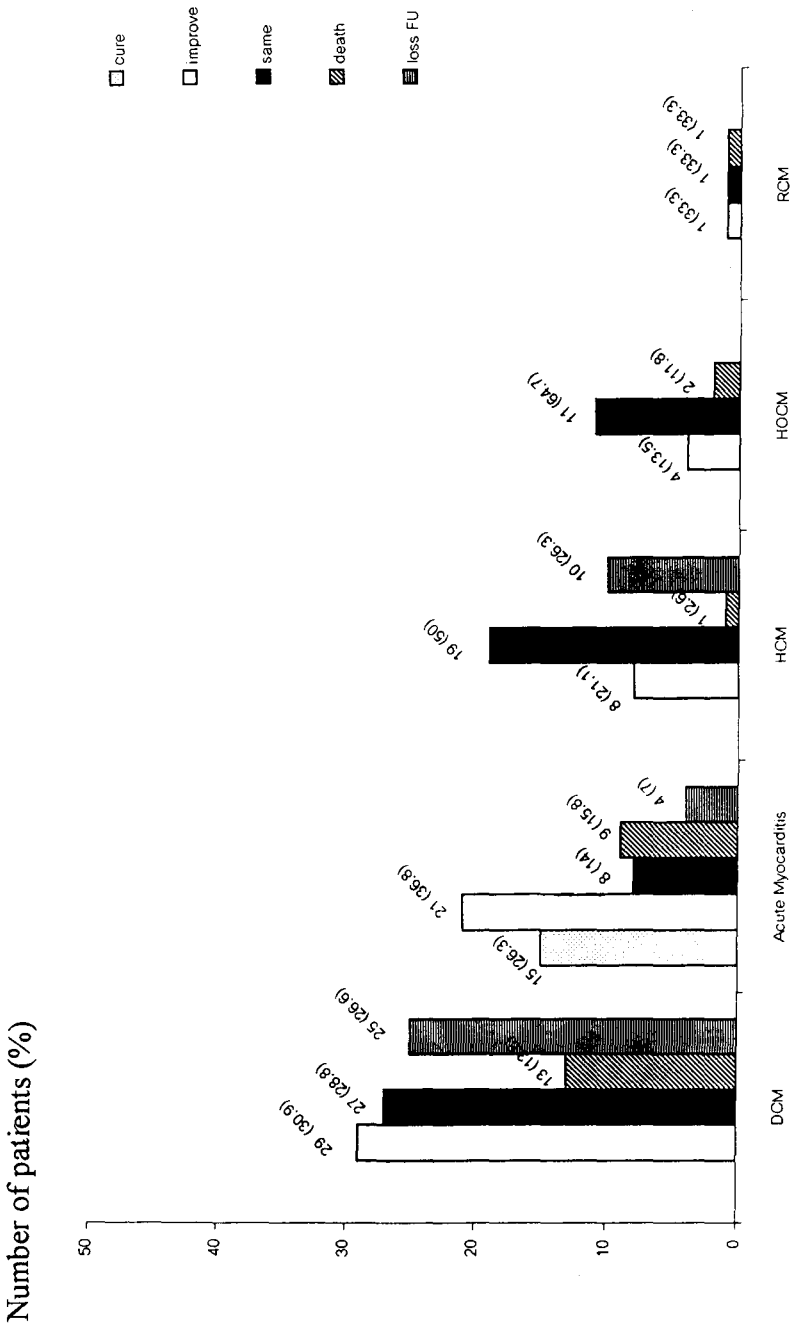

Fig. 2. Outcome of treatment at 1 year (0.1-5.5 years) follow-up.

cations and disopyramide have been proposed as the option for symptomatic HOCM. A study has demonstrated the mechanism of action of disopyramide by decreasing ejection acceleration and the hydrodynamic force on the protruding mitral leaflet resulting in a lower final pressure gradient[10]. However, there was no evidence that this medication could protect a patient with HCM from sudden death. Antiarrhythmic drugs especially amiodarone has been recommended in patients with frequent or prolonged episodes of nonsustained ventricular tachycardia on a single Holter recording. Amiodarone and implantable defibrillators have been suggested as an option to prevent sudden death[9]. Surgical myo-myectomy has been shown to have good results (especially in outflow pressure gradient > 50 mmHg). Low operative mortality (< 2%) with 93 per cent and 79 per cent survival at 5 and 10 years post-operation were reported[18-20]. Two patients in the present study underwent myo-myectomy with improving outflow pressure gradient. One patient developed high pressure gradient 6 months later. Maron et al reported that the dual-chamber pacemaker did not objectively improve cardiovascular performance in patients with HOCM[15]. Implantable cardiovertors and heart transplantation are the last options for patients with cardiomyopathy.

Outcome

Patients with DCM have demonstrated a high incidence of ventricular arrhythmia and increased risk of sudden death[34]. Lewis reported survival rates of 70 per cent, 64 per cent, and 52 per cent at 2, 5 and 11.5 years in infants and children with DCM[35]. The present study showed a 1-year survival rate of 82 per cent and found that age, gender, presenting symptoms, admission to ICU, CK-MB level, cTnT and EF at presentation did not affect the mortality in DCM. Mortality rate in acute myocarditis was 17 per cent at 1 year after presentation which was comparable to other studies[4, 6]. Low left ventricular EF and admission to ICU affected the mortality in acute myocarditis. A study showed that fulminant myocarditis; severe hemodynamic compromise and rapid onset were independent factors of survival[36]. Annual mortality rate in children with HCM has been reported to be up to 6 per cent[9]. The present study showed a 5.4 per cent mortality at 1-year-follow-up. The available data suggests that ventricular arrhythmia is the cause of sudden death in most patients, either as primary or secondary(9). No single test can reliably predict the risk of sudden death in HCM[9]. Rivens et al reported children with RCM were at a high risk for ischemia related complications and death with an annual mortality rate of 7 per cent[37]. The mortality rate in this study was much higher at 33 per cent, which might be due to the small number of patients.

In summary, primary myocardial diseases are uncommon. They affect the quality of life in most of the patients. Making a diagnosis is not too complicated. Mortality with standard treatment is still high. Intravenous immunoglobulin administration in acute myocarditis did not affect the mortality. Age, gender, presenting symptoms and inotropes administration had no effect on mortality in all groups. Admission to the ICU and low left ventricular EF at presentation in acute myocarditis did affect mortality rate, whereas CK-MB level and cTnT level did not. A randomized, double blind prospective study is required to find a new option of treatment to improve the quality of life and outcome of these patients.

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ภาวะกล้ามเนื้อหัวใจผิดปกติในเด็ก

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ภาวะกล้ามเนื้อหัวใจผิดปกติ เป็นหนึ่งในสาเหตุการตายที่สำคัญในเด็กจึงเป็นเหตุนำให้มีการศึกษาในผู้ป่วยเด็กในครั้งนี้ เพื่อหาแนวทางการรักษาที่เหมาะสม ผลของการรักษาและปัจจัยที่มีผลต่อการรักษาในผู้ป่วยเด็กที่มีปัญหาภาวะกล้ามเนื้อหัวใจผิดปกติ โดยได้ทำการศึกษาย้อนหลังในเด็กทุกคนที่มารับการรักษา และได้รับการวินิจฉัยเป็นภาวะกล้ามเนื้อหัวใจผิดปกติที่โรงพยาบาลระดับมหาวิทยาลัย 5 แห่งของประเทศไทยในช่วงเวลาตั้งแต่ มกราคม 2539-ธันวาคม 2543 พบว่า ผู้ป่วยทั้งหมด 209 ราย ซึ่งคิดเป็น 1.2% ของผู้ป่วยโรคหัวใจเด็ก พบว่าเป็น dilated cardiomyopathy (DCM) 45%, acute myocarditis 27.3%, hypertrophic cardiomyopathy (HCM) 18.2%, hypertrophic obstructive cardiomyopathy (HOCM) 8.1% และ restrictive cardiomyopathy (RCM) 1.4% เป็นเพศหญิง 56% อายุอยู่ในช่วง 0.1-15 ปี อาการนำของผู้ป่วยพบ 75% เป็นภาวะหัวใจวาย ค่ามัธยฐานของ ejection fraction (EF) ของ acute myocarditis 42% (15-79%) ซึ่งจะมากกว่าของ DCM 33% (10-57%) อย่างมีนัยสำคัญทางสถิติ เช่นเดียวกับระดับ Cardiac Troponin T (cTnT) ใน acute myocarditis จะสูงอย่างมีนัยสำคัญทางสถิติกว่าใน DCM (0.08 ng/ml range 0.01-0.16 *vs* 0.01 ng/ml range 0.01-0.10) จากการติดตามเป็นระยะเวลานานเฉลี่ย 1 ปี (0.1-5.5 ปี) พบอัตราตาย 18.8%, 17%, 2.6%, 11.8% และ 33.3% ในกลุ่ม DCM, acute myocarditis, HCM, HOCH และ RCM ตามลำดับ การรักษาตัวในหออภิบาลผู้ป่วยหนักและ EF ของเวนตริเคิลซ้ายต่ำ มีผลต่ออัตราตายในผู้ป่วย acute myocarditis ในขณะที่การให้ IVIG และระดับ cTnT ไม่มีผล

สรุป : ภาวะกล้ามเนื้อหัวใจผิดปกติพบได้ไม่บ่อย ผู้ป่วยส่วนใหญ่จะมีภาวะหัวใจล้มเหลว อัตราตายที่ 1 ปีพบว่าค่อนข้างสูง ปัจจัยที่มีผลต่ออัตราตายใน acute myocarditis คือ การรักษาตัวในหออภิบาลผู้ป่วยหนัก และ EF ของเวนตริเคิลซ้ายต่ำ ในขณะที่การให้ IVIG ไม่มีผลทำให้อัตราตายลดลง

คำสำคัญ : กล้ามเนื้อหัวใจทำงานผิดปกติ, กล้ามเนื้อหัวใจอักเสบ, โรคของกล้ามเนื้อหัวใจ

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