

Sudden Death and Cardiovascular Collapse in Children With Restrictive Cardiomyopathy

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Background—Restrictive cardiomyopathy (RCM) is rare in children, and the prognosis is poor. In the present study, we evaluated all pediatric patients with RCM who were at our institution during a 31-year period to determine the clinical outcome and cause of death. Those who sustained sudden, unanticipated cardiac arrests were evaluated for risk factors that are predictive of sudden death.

Methods and Results—Eighteen consecutive patients were reviewed. Presentation, clinical course, laboratory data, and histopathological evidence of ischemia were compared between patients with and without sudden death events. The results demonstrated that patients who were at risk for sudden death were girls with chest pain, syncope, or both at presentation and without congestive heart failure. Although not statistically significant for sudden death, Holter monitor evidence of ischemia predicted death within months. 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.

Conclusions—All children with RCM are at risk for ischemia-related complications and death, and some are at risk of sudden death. In the present study, patients at risk of sudden death appeared well and had no evidence of ongoing heart failure but often had signs or symptoms of ischemia characterized by chest pain, syncope, or both. ECGs and Holter monitors may be useful screening tools. The use of β -blockade, the placement of an implantable cardioverter-defibrillator, and preferential status 1A or B listing for cardiac transplantation are proposed for pediatric patients with RCM and evidence of ongoing ischemia. (*Circulation*. 2000;102:876-882.)

Key Words: pediatrics ■ cardiomyopathy ■ death, sudden ■ ischemia ■ syncope

Restrictive cardiomyopathy (RCM) is rare in children, constituting $\approx 5\%$ of diagnosed cardiomyopathies.^{1,2} With limited numbers of affected patients, the pediatric RCM literature is limited to case reports and 4 case series.¹⁻¹⁹ Two additional studies of pediatric cardiomyopathy contain an additional 4 patients with RCM.^{20,21} The pathophysiological mechanism that characterizes this disease is a primary abnormality of diastolic ventricular function with relative preservation of systolic function and left ventricular end-diastolic dimension.²² Prognosis for the pediatric patient is poor, with 2-year actuarial survival rates of $<50\%$.¹⁻³ Syncope at presentation carries a particularly poor prognosis, with deaths occurring within 4 months in 1 study.² Other reported predictors of poor outcome include symptoms or radiographic evidence of pulmonary venous congestion at presentation.^{1,3} Sudden death has been reported, but the mechanism is unclear.^{2,5,14,19}

The purpose of the present study was to evaluate all children with RCM at our institution for clinical outcome and the cause of death. Those who sustained sudden death events, defined as

such only when the event was abrupt and unexpected,²³ were evaluated for identifiable risk factors. Although ischemia-mediated events are rare in children, we hypothesized that the patients with RCM at a significant risk for sudden death were those with ongoing myocardial ischemia.

Methods

Study Population

All patients at our institution with RCM were retrospectively reviewed. Diagnostic criteria included (1) echocardiographic features of RCM (Figure 1A), including dilated atria, normal or nearly normal left ventricular systolic function, and no significant ventricular dilatation; (2) no echocardiographic evidence of hypertrophic cardiomyopathy; (3) no evidence of constrictive pericarditis; and (4) elevated end-diastolic ventricular pressures on cardiac catheterization. Supportive evidence included ECG features of RCM (Figures 2A and 2B) and a restrictive mitral inflow pattern by pulsed-wave Doppler. (Figure 1B).

Patients were divided into 2 groups: those with unanticipated sudden cardiac death events at a time of relative well-being [SCD(+)] and those without such events [SCD(-)]. Evaluation included chart review of symptoms, physical examination, clinical course, and timing and cause

Received September 8, 1999; revision received March 27, 2000; accepted March 27, 2000.

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Guest Editor for this article was Welton M. Gersony, MD, Babies Hospital, New York, NY.

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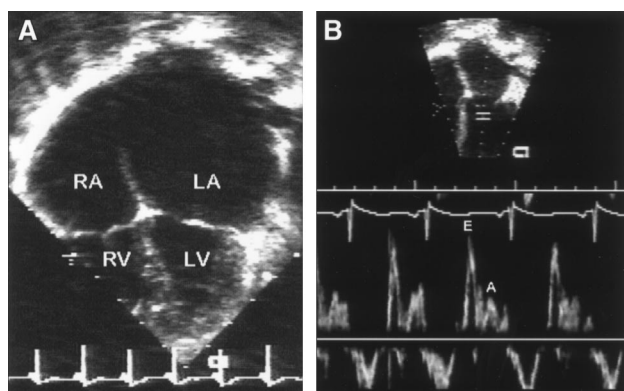


Figure 1. Echocardiographic features of RCM. A, Two-dimensional image (apical 4-chamber view) that demonstrates moderate right and severe left atrial enlargement relative to normal biventricular size and thickness. B, Pulsed Doppler mitral inflow pattern (leaflet tips) consistent with restrictive physiology that demonstrates rapid E-wave deceleration phase, high E/A-wave velocity ratio, and decreased isovolumic relaxation time. LA indicates left atrium; LV, left ventricle; RA, right atrium; and RV, right ventricle.

of death. Signs and symptoms that were attributed to ischemia included syncope and exertional chest pain. Signs and symptoms that were attributed to heart failure included tachypnea, dyspnea (with or without exertion), orthopnea, diaphoresis, hepatomegaly, jugular venous distention, and edema. Laboratory evaluation included a review of ECGs, rhythm strips, Holter monitor studies, echocardiograms, cardiac catheterization data, and autopsy or explantation findings. ECGs were reviewed for ST-segment elevation or depression, T-wave inversion, or new, deep, or wide Q waves. Holter monitor studies were assessed for ST-segment depression, with analysis performed through the Marquette 24-hour graphic display Holter monitoring system. Catheterization data were reviewed for right and left ventricular end-diastolic pressures (RVEDP and LVEDP, respectively), mean pulmonary arterial pressure (MPAP), and pulmonary vascular resistance (PVR). Finally, autopsy and explantation specimens were reviewed for evidence of acute or chronic myocardial ischemia. For all specimens, histological sections of both ventricles and all 3 major coronary arteries were examined. In 12 of 13 specimens, sections of both left ventricular papillary muscles were available for review.

Statistical Analysis

Statistical analysis was performed to identify risk factors unique to the sudden death [SCD(+)] subgroup. Clinical presentation and demographics, outcome, hemodynamic data, and laboratory and histopathological evidence of ischemia were compared. Percentages were compared with use of the Fisher exact test, and mean values were analyzed with the use of Student's *t* test. Results are presented as percentage or mean \pm SD. A *P* value of ≤ 0.05 was considered statistically significant.

Results

Patient Population

Eighteen patients ranging in age from 0.7 to 12.2 years (16 of 18 were <6 years old) with classic features of RCM were identified between the years of 1967 and 1998 (Table 1). Twelve patients were reviewed in a prior study that assessed the causes and natural history of RCM.² Two children (patients 8 and 9) were identical twins. Two other children (patients 6 and 18) were cousins. The sibling of 1 (patient 6) died of a cardiomyopathy; their parents were cousins.

Patients 1 to 5 sustained sudden death events [SCD(+)], and patients 6 to 17 did not [SCD(-)]. Patient 18 was lost to

follow-up. Of the SCD(+) patients, 1 was successfully resuscitated and underwent cardiac transplantation; the other 4 died. Of the SCD(-) patients, 5 underwent successful cardiac transplantation, and 7 died.

SCD(+) Group

Clinical Presentation and Demographics

Three of the 5 SCD(+) patients presented with syncope, 1 presented with chest pain, and 1 was evaluated for a murmur. All patients were female, between the ages of 1.6 and 10.5 years (mean 6.3 ± 1.6 years). The time from symptom onset to diagnosis was 0 to 0.1 years (mean 0.02 ± 0.04 years). None of the patients presented with signs or symptoms of heart failure, and none were in heart failure at the time of their arrest. Events occurred from 1 month to 3.5 years after diagnosis (mean 1.0 ± 0.6 years, but 4 of 5 occurred within 6 months) and consisted of sudden death, aborted sudden death, or rapidly progressive cardiovascular collapse. One patient died abruptly after being startled by a sprinkler (patient 1). A second patient arrested after playing outside (patient 2). A third patient vomited and then had a cardiac arrest en route to the emergency department (patient 3). A fourth patient collapsed en route to a clinic visit and died within 12 hours despite emergent cardiopulmonary bypass and ventricular assist device placement (patient 4). The 1 survivor was resuscitated from an in-hospital arrest heralded by severe chest pain (patient 5).

Hemodynamic Data

Right- and left-heart catheterizations were performed in all 5 patients. LVEDP averaged 25.2 ± 2.6 mm Hg, RVEDP averaged 13.4 ± 1.3 mm Hg, and MPAP averaged 25 ± 1.3 mm Hg. PVR index averaged 2.4 ± 0.5 U \cdot m².

Laboratory Evidence of Ischemia

Review of the laboratory data from the SCD(+) patients revealed consistent evidence of ischemia. All 5 ECGs at presentation demonstrated ST-segment depression or T-wave inversion in the inferior, lateral, or lateral precordial leads (or a combination) (Figure 2B). All 5 Holter monitors revealed ST-segment depression (5.4 to 12.7 mm), and it was most pronounced at higher heart rates (Figure 3). In the survivor with chest pain, ST-segment depression to 8.2 mm preceded the development of torsade de pointes (Figures 4A and 4B).

Pathology Specimens

Pathology specimens (3 autopsies and 1 explantation) were available for 4 of the 5 SCD(+) patients. Acute ischemia (Figure 5A) was demonstrated in 3 of 4 hearts. In 2 cases, an acute left ventricular myocardial infarction was identified; the other demonstrated biventricular subendocardial ischemic necrosis. Chronic ischemia (Figure 5B) was evident in the fourth heart, with prominent myocytolysis and vacuolization and extensive papillary muscle scarring. None of the 4 hearts had evidence of coronary artery obstruction.

SCD(-) Group

Clinical Presentation and Demographics

Nine of the 12 SCD(-) patients presented with signs and symptoms of heart failure, 1 had a murmur, 1 had an irregular

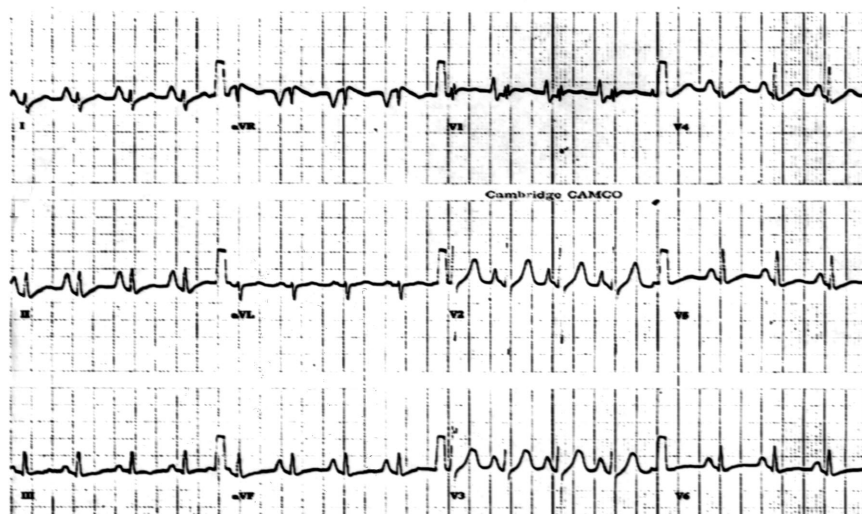
A**B**

Figure 2. ECG patterns in RCM. A, ECG without overt evidence of ischemia, demonstrating biatrial enlargement and non-specific ST-T-wave changes in inferior and lateral precordial leads. B, ECG with evidence of ischemia, demonstrating biatrial enlargement and ST-segment depression in inferior, lateral, and lateral precordial leads.

rhythm, and 1 was evaluated for a positive family history. None presented with syncope. Eight patients were male and 4 were female, ranging in age from 0.7 to 12.2 years (mean 3.4 ± 0.9 years). The time from symptom onset to diagnosis ranged from 0 to 2.6 years (mean 0.5 ± 0.8 years). Seven children died (patients 11 to 17), 5 underwent successful orthotopic cardiac transplantation (patients 6 to 10), and 1 was lost to follow-up. The time from diagnosis to death or transplantation ranged from 4 days to 14 years (mean 3.3 ± 1.3 years).

Hemodynamic Data

Cardiac catheterizations were performed in 12 of the 13 SCD(−) patients; the left heart was entered in 11. RVEDP, LVEDP, and PVR were uniformly elevated. LVEDP averaged 23.6 ± 2.4 mm Hg, RVEDP averaged 14.5 ± 1.3 mm Hg, and MPAP averaged 33 ± 4.1 mm Hg. PVR index averaged 6.3 ± 1.8 U · m².

Laboratory Evidence of Ischemia

Laboratory studies were not uniformly available. Among the available studies, evidence of ischemia was inconsistently

noted. Thirteen patients had ECGs available for review. Eight ECGs had evidence of ischemia that consisted of ST-segment depression, T-wave inversion, pathological Q waves in the inferior or lateral precordial leads or both, or a combination. Two ECGs had no evidence of ischemia, and 2 other ECGs had a right bundle-branch block pattern that precluded analysis.

Seven patients had Holter monitor studies available for review; however, 4 studies had a bundle-branch block, paced ventricular rhythm, or both, which precluded ST-segment analysis. The remaining 3 had evidence of ischemia, with ST-segment depression ranging from 3 to 12.7 mm. Studies were performed 1 day to 8 months before death or transplantation.

Pathology Specimens

Pathology specimens were available for review in 9 of 12 SCD(−) patients. Of the 5 patients who underwent transplantation, 3 explanted hearts had evidence of chronic ischemia. One child had extensive fibrous scarring and myocytolysis in the left ventricular subendocardium and papillary muscles. The remaining 2 explants had a pattern of myocardial

TABLE 1. Pediatric RCM: Institutional Experience (1967 to 1998)

Patient	Sex	Age at Diagnosis, y	Chief Complaint	Time From Symptoms to Diagnosis, y	SCD	CHF	Outcome	Time From Diagnosis to Death/Transplantation, y	Evidence of Ischemia			Terminal Rhythm
									ECG	Holter Monitor	Pathology	
1	F	10.8	Syncope	0	Yes	No	Death	0.1	Yes	Yes	NA	NA
2	F	4.9	Syncope	0	Yes	No	Death	0.3	Yes	Yes	Acute	NA
3	F	1.6	Murmur	0	Yes	No	Death	0.5	Yes	Yes	Chronic	NA
4	F	8.9	Syncope	0	Yes	No	Death	0.3	Yes	Yes	Acute	Bradycardia, asystole
5	F	5.3	CP	0.1	Yes	No	TX	3.5	Yes	Yes	Acute	Torsade de pointes
6	M	5.0	CHF	NA	No	Yes	TX	8.0	BBB	BBB	Chronic	NA
7	F	0.9	CHF	0.8	No	Yes	TX	3.9	Yes	BBB	Chronic	NA
8	M	2.5	Murmur	0	No	No	TX	0.2	Yes	NA	None	NA
9	M	2.5	Family history	0	No	No	TX	0.2	No	NA	None	NA
10	M	0.9	CHF	0.1	No	Yes	TX	1.5	Yes	Yes	Chronic	NA
11	M	4.0	CHF	2.6	No	Yes	Death	0.7	Yes	Yes	Chronic	VF
12	F	1.7	CHF	0.1	No	Yes	Death	1.6	Yes	Paced	Acute, chronic	VT, VF
13	M	1.6	CHF	0.9	No	Yes	Death	14.0	BBB	BBB, paced	NA	NA
14	F	4.1	CHF	0.9	No	Yes	Death	0.9	No	NA	Acute	VF
15	M	2.9	CHF	0.1	No	Yes	Death	0.6	Yes	Yes	Acute, chronic	IVR
16	F	1.1	CHF	0.1	No	Yes	Death	2.7	Yes	NA	NA	NA
17	M	12.2	Irregular rhythm	0	No	Yes	Death	5.6	Yes	NA	NA	NA
18*	M	5.0	CHF	NA	NA	NA	NA	NA	Yes	NA	NA	NA

TX indicates transplantation; NA, not available; CP, chest pain; CHF, congestive heart failure; BBB, bundle-branch block; VF, ventricular fibrillation; VT, ventricular tachycardia; and IVR, idioventricular rhythm.

Patients 1 to 5 are those who sustained SCD events.

Evidence of Ischemia columns refer to ECG evidence of ST-segment depression, T-wave inversion, Q waves, or a combination; Holter monitor evidence of ST-segment depression; and histopathological evidence of acute or chronic ischemia on autopsy or explantation.

*Patient 18 was lost to follow-up.

scarring not suggestive of ischemia (Figure 5C); these were from the only SCD(−) patients without chronic heart failure. No evidence of acute ischemia was noted in the explanted hearts.

Of the 4 patients who died and underwent autopsy, 3 had evidence of acute ischemia. Two hearts had acute hypotensive

infarctions, and 1 demonstrated acute subendocardial ischemic necrosis. In both hearts with an infarction, there also were chronic ischemic changes with scarring and myocytolysis in the subendocardium, papillary muscles, or both. Chronic ischemia was not noted in the third heart, but papillary muscle sections were not available. The fourth heart had only chronic ischemic

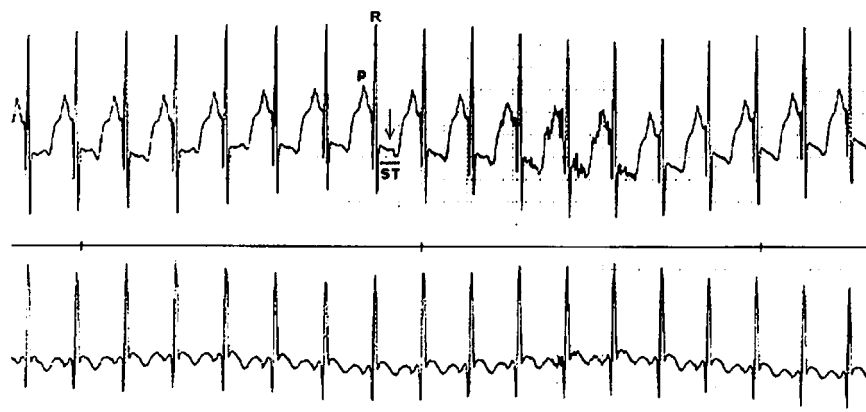


Figure 3. Holter monitor tracing from 10-year-old girl with RCM and syncope, demonstrating ST-segment depression (arrow) consistent with ongoing ischemia. She died within 5 weeks of diagnosis.

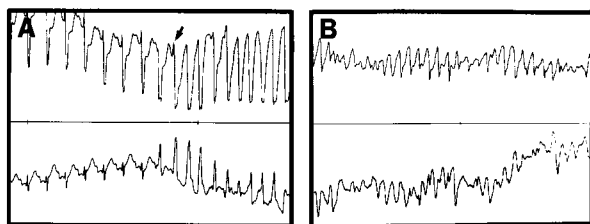


Figure 4. Holter monitor tracing from 5-year-old girl with RCM and chest pain who sustained an acute myocardial infarction during monitoring. A, Sinus tachycardia and ST-segment depression precede degeneration to torsade de pointes. Arrow indicates inciting "R-on-T" phenomenon. B, Tracing obtained several seconds later, depicting classic torsade de pointes pattern.

changes with small remote microinfarctions in the left ventricular subendocardium and papillary muscles and only modest vacuolization. None of the 9 hearts available for review had evidence of coronary artery obstruction.

Interpretation

Patient demographics and clinical presentation are compared in Table 2. Statistically significant risk factors for sudden death were (1) female sex, (2) signs and symptoms of ischemia at presentation, (3) absence of heart failure at presentation, and (4) absence of ongoing refractory heart failure.

Statistical significance was not achieved among the remaining risk factors in Table 3, including hemodynamic data, time course to diagnosis or outcome, or laboratory/pathology evidence of ischemia. There were, however, noteworthy correlations. With respect to Holter monitor evidence of ischemia, all 8 studies from both subgroups demonstrated ST-segment depression. Within 8 months of monitoring, 7 of the 8 patients sustained cardiac arrests, and the other patient underwent transplantation without event; all had histopathological evidence of ischemia. ST-segment depression on Holter monitoring, therefore, was used to accurately identify ongoing ischemia and heralded an impending death. With respect to autopsy and explantation data,

TABLE 2. Risk Factors for SCD in Pediatric RCM: Clinical Presentation and Demographics

Factor	SCD+	SCD−	P
Female, %	100	31	*0.03
(n)	(5)	(13)	
Age at diagnosis, y	6.3±1.6	3.4±0.9	0.11
(n)	(5)	(13)	
Signs/symptoms of ischemia at presentation, %	80	8	*0.01
(n)	(5)	(13)	
Signs/symptoms of heart failure at presentation, %	0	77	*0.01
(n)	(5)	(13)	
Ongoing refractory heart failure, %	0	83	*0.01
(n)	(5)	(12)	

* $P \leq 0.05$ is significant.

histopathological evidence of acute ischemia was more frequently identified in the SCD(+) group, whereas more chronic ischemic changes were noted in the SCD(−) group. Sudden death, therefore, appeared to be associated with acute ischemic events.

For all comparisons, the small sample size of the population is likely to have affected statistical significance.

Discussion

Ischemia-mediated cardiac events are common in adults but rare in children. The results of the present study suggest that pediatric patients with RCM represent a population of children who are at risk for ischemia-related complications and death. We had hypothesized that sudden deaths in children with RCM were ischemia mediated. However, it appears that many children with RCM, regardless of mechanism of death, are at risk for ischemia-related complications and death.

Sudden death occurred in 28% of our patients, with an annual mortality rate of 7% (4-year average follow-up time for all

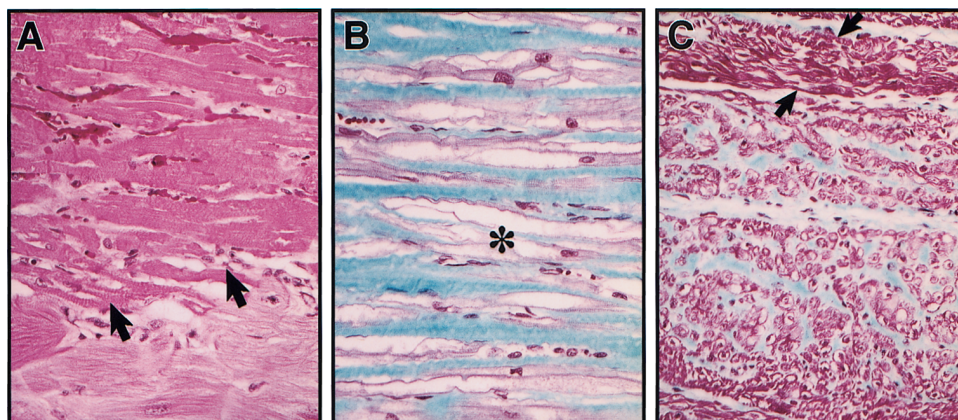


Figure 5. A, Acute ischemia. In contrast to viable myofibers below arrows, remainder of myocardium shows acute ischemic necrosis, with hypereosinophilic cytoplasm, absent myocardial nuclei, and vascular congestion (magnification $\times 66$, hematoxylin and eosin stain). B, Chronic ischemia. Asterisk overlies 1 of multiple clusters of pale, vacuolated myofibers with increased green-staining interstitial fibrous tissue. These findings, which localized to subendocardium and papillary muscles, are indicative of chronic ischemia (magnification $\times 66$, Gomori's trichrome stain). C, No ischemia. Myocardium in center demonstrates randomly distributed perimycocyte fibrosis and modest vacuolization, consistent with findings seen in restrictive cardiomyopathy. For comparison, arrows identify an adjacent myocardial bundle with normal interstitial fibrous tissue and no vacuolization (magnification $\times 66$, Gomori's trichrome stain).

TABLE 3. Risk Factors for SCD in Pediatric RCM: Hemodynamics, Clinical Course, and Evidence of Ischemia

Factor	SCD +	SCD –	P
LVEDP _{max} , mm Hg	25.2 ± 2.6	23.6 ± 2.4	0.70
(n)	(5)	(11)	
RVEDP _{max} , mm Hg	13.4 ± 1.3	14.5 ± 1.3	0.63
(n)	(5)	(12)	
MPAP, mm Hg	25 ± 1.3	33 ± 4.1	0.24
(n)	(5)	(12)	
PVR index, U · m ²	2.4 ± 0.5	6.3 ± 1.8	0.20
(n)	(5)	(12)	
Time from symptoms to diagnosis, y	0.02 ± 0.04	0.5 ± 0.8	0.07
(n)	(5)	(11)	
Time from diagnosis to death/transplantation, y	1.0 ± 0.6	3.1 ± 1.3	0.31
(n)	(5)	(12)	
ECG evidence of ischemia, %	100	82	0.9
(available n)	(5)	(11)	
Holter monitor evidence of ischemia, %	100	100	0.9
(available n)	(5)	(3)	
Acute ischemia on autopsy/explant, %	75	33	0.27
(available n)	(4)	(9)	
Chronic ischemia on autopsy/explant, %	25	67	0.27
(available n)	(4)	(9)	
No ischemia on autopsy/explant, %	0	22	0.9
(available n)	(4)	(9)	

P ≤ 0.05 is significant.

patients). There is a comparable 31% incidence of sudden death in children with hypertrophic cardiomyopathy and a lower incidence of 11% in those with dilated cardiomyopathy.^{24,25} A lower annual mortality of 4% from sudden death is reported for pediatric hypertrophic cardiomyopathy.²⁵ Among the available case reports and series on pediatric RCM, 10% (4 of 40 patients) had sudden death events. A 7-year-old boy with exertional chest pain died suddenly the day after catheterization, within 3 months of presentation.⁵ A 6-year-old boy sustained an abrupt syncopal spell associated with ventricular fibrillation and died within 2 months of presentation.¹⁹ Their symptoms of ischemia and rapid time to death were similar to those of our study population, but their sex and symptoms of heart failure at presentation were dissimilar. The 2 other patients described were sisters, ages 6 and 16 years, who also died suddenly (no other information); they were members of a 5-generation family with autosomal dominant RCM, atrioventricular block, and skeletal myopathy.¹⁴

Patients with RCM, atrioventricular block, skeletal myopathies, and arrhythmias have demonstrated both autosomal dominant and autosomal recessive inheritance patterns; in these patients, desmin inclusions have been isolated by electron microscopy, and in some, missense mutations in the desmin gene have been identified.²⁶ Four patients in this study had a positive family history, although the pattern of transmission cannot be determined. Two patients were identical twins, and 2 were cousins. One of the cousins had a sibling who died of a cardiomyopathy; their parents were cousins. Desmin inclusions were not detected.

Syncope has been an ominous sign in our population, with 3 of 3 patients dying abruptly within months of presentation. Lewis,¹ however, reported a female patient with recurrent syncope who survived 11.8 years. Her spells were not arrhythmia related, and ST-segment depression was not noted on a treadmill test, suggesting that her syncope was not ischemia mediated. Gewillig et al⁴ also reported a female patient with syncopal episodes who survived 11.5 years. Her episodes were secondary to cerebral ischemic attacks due to thromboembolism. These cases and our patients demonstrate that a thorough evaluation of syncope in all patients with RCM is necessary. Ischemia, arrhythmias, and thromboembolism must all be ruled out given the association of these entities with RCM.^{1–4,14}

In the adult population, ischemic myocardium is an established substrate for lethal ventricular arrhythmias.²⁷ In our study population, this same mechanism is documented for pediatric patients. This is demonstrated by the Holter monitor recording of the resuscitated sudden death of patient 5 (Figures 4A and 4B). In these tracings, ST-segment depression is pronounced at faster heart rates, followed by degeneration to torsade de pointes. The association between torsade de pointes and ischemic myocardium is well documented in the adult literature,²⁸ suggesting a mechanism of death in some pediatric patients with RCM who die suddenly and unexpectedly. Children with a more chronic course may also sustain terminal ventricular dysrhythmias, given the presence of ischemic myocardium in these patients as well. Documentation is provided in 3 of the patients with chronic heart failure and ischemic changes (patients 11, 12, and 14). Rhythm

strips demonstrated ventricular tachycardia, ventricular fibrillation, or both during resuscitation attempts.

The results of this study suggest that there are identifiable risk factors for sudden death in this population. Patients who appear to be at a greater risk include female patients and those who present with signs and symptoms of ischemia, such as chest pain and syncope. Patients who appear to be at a lower risk for sudden death include those with signs and symptoms of heart failure at presentation and those with ongoing refractory heart failure. Nevertheless, although this is the largest number of patients analyzed to date, the patient number remains small, potentially affecting statistical interpretation.

Optimal medical management is difficult to determine due to small numbers and variable management approaches during the past 30 years. Based on the findings of this study, routine screening for ischemia in all pediatric patients with RCM should include a baseline ECG and Holter monitoring with digital ST-segment analysis. Follow-up testing should be performed approximately every 6 months, with increased frequency dictated by clinical status. Patients with symptoms attributable to acute myocardial ischemia, such as chest pain or syncope, warrant immediate evaluation. With respect to medication, captopril was deleterious in 1 small study, with 4 pediatric patients with RCM undergoing cardiac catheterization that demonstrated systemic hypotension without an improvement in cardiac output.²⁹ β -Blocking agents may be beneficial in the subset of patients with RCM with clinical evidence of ischemia. Their anti-ischemic properties and ability to suppress catecholamine-provoked arrhythmias have been well documented in the adult with ischemic heart disease.³⁰ The placement of an implantable cardioverter-defibrillator has similarly improved morbidity and mortality rates in the adult with myocardial ischemia³¹ and should be considered in patients with RCM who have clinical evidence of ischemia, particularly with documented ventricular arrhythmias. Last, immediate cardiac transplantation work-up and listing are warranted. Preferential status IA or B listing is proposed for pediatric patients with RCM and is justifiable by several factors. First, these patients are at a high risk of sudden death; second, survival time is short; and last, relative preservation of systolic function and potential negative effects of intravenous inotropic agents limit the potential of meeting current criteria for status IA or B listing.

In summary, RCM is a rare but often rapidly lethal disease in childhood. Clinical evidence of myocardial ischemia may herald an impending demise. Medical management and preferential status IA or B listing for cardiac transplantation are recommended.

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