Cardiology in the Young (2011), **21**, 490–494
doi:10.1017/S1047951111000849

Review Article

Symptomatic myocardial bridges in children: a case report with review of literature

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Abstract Myocardial bridge is a rare congenital coronary anomaly in children, usually seen in the setting of hypertrophic cardiomyopathy and in left ventricular hypertrophy. Most myocardial bridges are believed to represent a benign anatomical variant; however, symptomatic myocardial bridge is a distinct subgroup of pathological variant, linked to myocardial ischaemia, ventricular arrhythmia, and sudden cardiac death. We present a case of a symptomatic myocardial bridge in a 5-year-old boy with mild hypertrophic cardiomyopathy who underwent supra-arterial myotomy, automatic defibrillator placement, and long-term Beta-blocker therapy. We also present 10 years of follow-up with a review of literature regarding symptomatic myocardial bridges in the paediatric age group.

Keywords: Hypertrophic cardiomyopathy; coronary ischaemia; supra-arterial myotomy; sudden cardiac death

Received: 11 February 2011; Accepted: 29 April 2011; First published online: 23 June 2011

A SEGMENT OF EPICARDIAL CORONARY ARTERY embedded by overlying myocardial tissue is known as myocardial bridge. Although we have known about myocardial bridges for more than 250 years, controversies exist regarding the clinical relevance of systolic compression of tunnelled coronary artery.[1,2] New diagnostic modalities such as quantitative coronary angiography, intravascular ultrasound, and intracoronary Doppler flow velocity measurements have become available in the last decade and they helped us to define the pathophysiology of myocardial ischaemia in a small subgroup of symptomatic myocardial bridges.[2] The management of symptomatic myocardial bridge includes mechanical decompression of tunnelled coronary segment, supplemented with automatic defibrillator and Beta-blocker therapy. The goal of this therapy is to resolve the myocardial ischaemia and to reduce the associated risk of sudden cardiac death.

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Case report

A 5-year-old boy presented for cardiac clearance before head magnetic resonance imaging for conscious sedation with history of multiple muscular ventricular septal defects and recurrent episodes of syncope with exertion, at times followed by generalised tonic-clonic seizures.

He had a previous diagnosis of multiple muscular ventricular septal defects in infancy during evaluation of heart murmur and a paediatric cardiologist followed him up to 4 years of age. He recently started with episodes of syncope, especially with running, and at times generalised tonic-clonic seizure followed such an episode. His neurological evaluation including head computed tomography and electroencephalogram was negative. He continued to have such episodes and was placed empirically on Tegretol.

His physical examination was consistent with a diagnosis of small muscular ventricular septal defect. An electrocardiogram showed normal sinus rhythm, left superior axis with left ventricular hypertrophy. A two-dimensional echo Doppler study showed evidence

of mild hypertrophic cardiomyopathy without left ventricular outflow tract obstruction and normal proximal coronaries, in addition to haemodynamically insignificant muscular septal defect. We performed a stress exercise test in the view of syncope with exertion to recreate and to assess similar scenario. During the stress test with moderate exertion and with a heart rate of more than 140 beats per minute, he suddenly screamed with anginal pain, with severe ST-T wave changes in anterolateral leads (Fig 1). Subsequently, he developed ventricular tachycardia with hypotension and required cardioversion (Fig 1). Thallium scan showed reversible perfusion defects in the distribution of left anterior descending artery. Selective coronary angiography revealed a 28-millimetre-long myocardial bridge at mid-segment of left anterior descending artery with more than 80% systolic compression and persistent narrowing in 50% of diastole (Fig 2).

He underwent supra-arterial myotomy, which revealed at least 30 millimetres of tunnelled segment of left anterior descending artery with a maximum depth up to 7 millimetres (Fig 3). We also placed an automatic defibrillator at the same time. He tolerated the procedure well. A Beta-blocker, atenolol, was started in the dose enough with adequate Beta-blockade, confirmed on holter monitor. We performed a repeat stress exercise test 3 months later, which showed resolution of exercise-induced ischaemic changes. He underwent a repeat coronary angiography after 1 year, which also showed complete resolution of myocardial bridge (Fig 4). Myocardial perfusion scan and periodic holter monitors were unremarkable in follow-up.

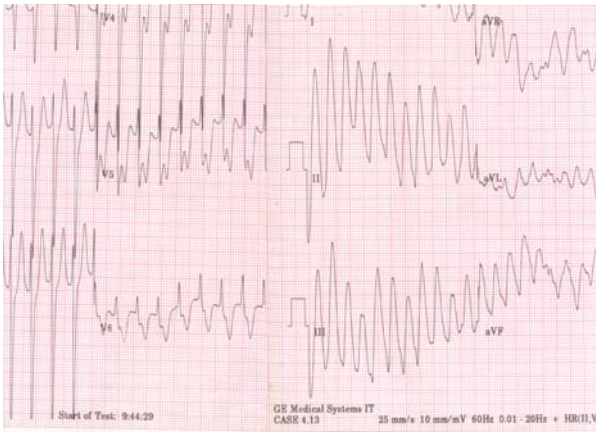

Figure 1.
Stress exercise electrocardiogram showing severe ST-T wave changes of ischaemia induced by exertion, followed by ventricular tachycardia.

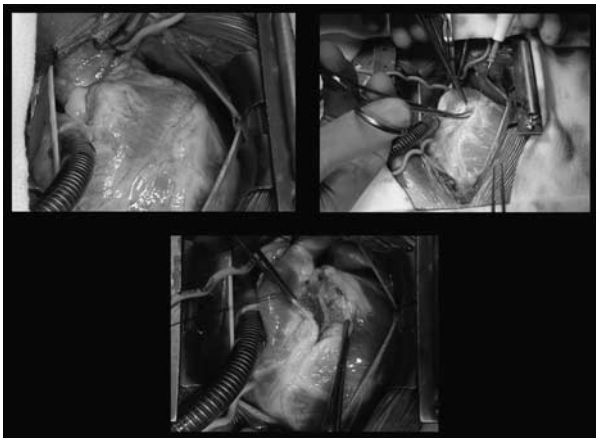

Figure 3.
Supra-arterial myotomy of myocardial bridge utilising cardio-pulmonary bypass.

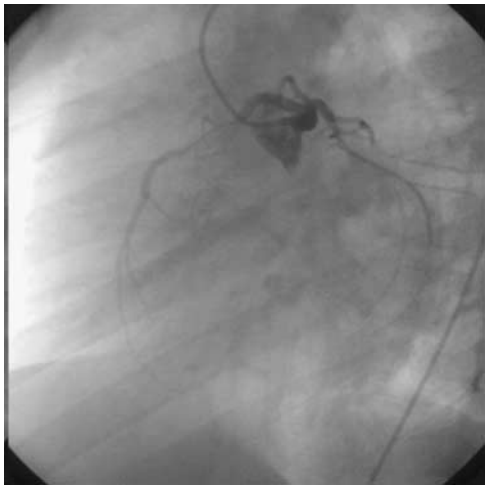

Figure 2.
Selective coronary angiography showing more than 80% systolic compression of mid-left anterior descending artery.

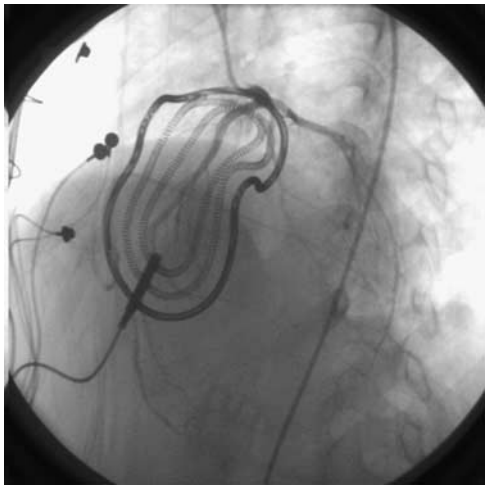

Figure 4.
Follow-up coronary angiography after supra-arterial myotomy showing normal filling of left anterior descending artery.

https://doi.org/10.1017/S1047951111000849 Published online by Cambridge University Press

The patient is doing well 10 years after his surgery. He had two appropriate defibrillator discharges with brief episodes of ventricular tachycardia at the rate of 140–160 beats per minute during the first 2 years. The changes of hypertrophic cardiomyopathy have remained stable on echocardiogram. He continues to participate in physical activities with mild to moderate exertion, such as regular gym in school and recreational sports without any problems.

Discussion

Muscle overlying the intramyocardial segment of an epicardial coronary artery, first described by Reyman in 1737, is termed as a myocardial bridge and artery coursing in the myocardium is called as a tunnelled artery.[1] Portmann and Iwig[3] in 1960 reported the first radiological description of a transient occlusion of segment of left anterior descending artery. Baurassa et al[2] in 1976 described the angiographic appearance of myocardial bridge, termed a "milking effect", as at least 50% systolic compression of coronary segment on selective coronary angiography. Grondin et al[4] described the first successful cases of supra-arterial myotomy for mid-left anterior descending artery myocardial bridge in three adults with pectoral angina in 1977. The clinical relevance of myocardial bridge has remained controversial and reports of only eight cases of symptomatic myocardial bridges with surgical management and limited follow-up are available in the paediatric age group.[5–8]

Reports of the incidence of myocardial bridge in general population are between 15% and 85% in pathological series and only between 0.5% and 2.5% in angiographic series. Such wide discrepancy suggests that only a minority of patients with myocardial bridges are in fact at risk of clinical symptoms and cardiac events.[2,9] Myocardial bridging is usually seen in the setting of hypertrophic cardiomyopathy/left ventricular hypertrophy and is rarely reported in a structurally normal heart.[8] Myocardial bridges occur in 30–50% of adults with hypertrophic cardiomyopathy, and angiographic evidence of myocardial bridging is reported between 28% and 40% in a selected group of children with hypertrophic cardiomyopathy in two studies.[10,11] Selective coronary angiography is not routinely performed in children and prevalence of myocardial bridges in general, as well as in an unselected group of children with hypertrophic cardiomyopathy, is unknown. Symptomatic myocardial bridges constitute a small subgroup of children, in whom, ischaemic symptoms can be linked directly to their presence.

Myocardial bridges are most commonly located over the middle segment of left anterior descending coronary artery and occasionally can involve diagonal branches, posterior descending right coronary artery, and marginal branch of circumflex artery.[2] They are located at a depth of 1–10 millimetres with a typical length of 10–30 millimetres. Ferreira et al[12] distinguished two types of myocardial bridges: superficial bridges with a prevalence of 75% crossing perpendicularly or at an acute angle towards the apex and deep bridges with a prevalence of 25% with muscle bundle arising from the right ventricular trabeculae that cross left anterior descending artery transversely, obliquely, or helically before terminating in the interventricular septum. Symptomatic myocardial bridge is usually deep, involving a long segment with muscle bundle wrapping around obliquely or helically. In our case, we noted the greatest depth to be 7 millimetres with a length of 30 millimetres. Greater than 90% of systolic compression of tunnelled coronary segment with ongoing compression of at least 50% of diastole characterises a significant myocardial bridge, as seen in our case.[9]

The contribution of systolic compression of tunnelled segment of coronary in myocardial bridge for ischaemic symptoms has been controversial as coronary perfusion predominantly occurs in diastole. The new investigational modalities such as quantitative coronary angiography, intravascular ultrasound, and intracoronary Doppler flow velocity have elucidated haemodynamic disturbances occurring in the diastolic phase of cardiac cycle in patients with symptomatic myocardial bridge. There are two mechanisms believed to be responsible for reduced coronary flow reserve in distal vessels and clinical symptoms of myocardial ischaemia:[2,9] phasic systolic compression of tunnelled segment of coronary with persistent compression well in diastole (up to 30–75%) when the largest proportion of coronary flow normally occurs and increased intracoronary Doppler flow velocities with abnormal qualitative flow profiles. Intracoronary Doppler flow measurements in cases of symptomatic myocardial bridge shows early diastolic flow acceleration as a result of delayed vessel relaxation, followed by rapid mid-diastolic deceleration and by mid-late diastolic plateau. There is also local retrograde systolic flow seen proximal to the myocardial bridge due to systolic compression of coronary segment. Additional factors such as coronary atherosclerosis, platelet aggregation, and left ventricular outflow tract obstruction in hypertrophic cardiomyopathy also contribute in ischaemic symptoms. Atherosclerotic changes in coronaries are frequently seen in adults as systolic compression increases trauma sustained by intima proximal to the myocardial bridge. Platelet aggregation secondary to endothelial dysfunction can also precipitate symptoms of myocardial ischaemia. The haemodynamic burden of left ventricle in the setting of hypertrophic obstructive

https://doi.org/10.1017/S1047951111000849 Published online by Cambridge University Press

Table 1. Cases of symptomatic myocardial bridges in children.

Cases	Age/sex	Cardiac status	Presentation	Pre-operative ischaemic data	Management	Post-operative resolution data	Follow-up
Hillman et al (1999)	10 years/M	BVH	Chest pain with exertion	Dobutamine stress echo	SAM	Exercise EKG	1 year
		Apical myocardial infarction	Syncope	Perfusion scan		Perfusion scan	
Sebening et al (2002)	11 years/M	HOCM	Asymptomatic VT on Holter	Holter	SAM	Resolution of VT on Holter	–
McCrindle et al (2004)							
1.	5 years/M	HCM	Cardiac arrest	Holter Exercise EKG	SAM Preexisting AICD	–	4.5 years Late SCD
2.	9 years/M	HCM	Cardiac arrest	Exercise EKG Perfusion scan	SAM AICD	Exercise EKG	5.5 years
3.	9 years/M	HCM	Palpitations	Perfusion scan	SAM AICD	–	9 months
4.	10 years/M	HCM	Cardiac arrest	Holter	SAM AICD	Holter Exercise EKG Perfusion scan	8 years
5.	14 years/M	HCM	Recurrent syncope	Holter Exercise EKG Perfusion scan	SAM	–	4 months
Danna et al (2006)	11 years/F	Normal heart	Syncope with exertion	Holter Exercise EKG Perfusion scan	SAM Beta/calcium channel blockers	Exercise EKG	2 years
Sharma et al (2010)	5 years/M	HCM	Syncope with exertion	Holter Exercise EKG Perfusion scan	SAM AICD Beta-blocker	Holter Exercise EKG Perfusion scan	10 years

AICD = automatic defibrillator; BVH = biventricular cardiac hypertrophy; EKG = exercise electrocardiogram; HCM = hypertrophic cardiomyopathy; HOCM = hypertrophic cardiomyopathy; M = male; SAM = supra-arterial myotomy; SCD = sudden cardiac death; VT = ventricular tachycardia

cardiomyopathy secondary to significant left ventricular outflow tract obstruction and increased wall tension can also influence overall symptomatology.

Chest pain and syncope in symptomatic myocardial bridges are predominantly associated with exertion. An increase in sympathetic drive with exercise induces ischaemia as tachycardia increases systole–diastole ratio at the expense of diastolic flow, increased contractility aggravates systolic and diastolic compression, and coronary spasms contribute to constriction of tunnelled coronary artery.[9] Criteria for symptomatic myocardial bridging in cases of non-obstructive hypertrophic cardiomyopathy include typical symptoms of chest pain or syncope with exertion, supported by ischaemic data, such as stress exercise electrocardiogram and perfusion scans.

There has been a debate whether myocardial bridge is an independent risk factor for ischaemia and sudden cardiac death in children with hypertrophic cardiomyopathy or is simply an indicator of the severity of disease.[10,11,13] Reports of symptomatic myocardial bridges are available in cases with mild hypertrophic cardiomyopathy such as in the present case, as well as in a child with structurally normal heart.[5,8] On the basis of reported cases in literature, a very small subgroup of children with myocardial bridges suffer from myocardial ischaemia and sudden death.[5–8] Therefore, myocardial bridging need not be an indicator of severity of disease in hypertrophic cardiomyopathy or always be a risk factor for ischaemia or sudden cardiac death.

A review of literature concerning symptomatic myocardial bridges in paediatric age group is reported in Table 1. All patients had significant symptoms of chest pain or syncope with exertion or cardiac arrest, supported by evidence of coronary ischaemia linked to myocardial bridge, which ultimately resolved after mechanical decompression in short-term follow-up. Our case is unique as symptomatic myocardial bridge is associated with mild hypertrophic cardiomyopathy. In addition to supra-arterial myotomy, he also received automatic implanted cardioverter defibrillator and long-term Beta-blocker therapy. The patient has resumed a normal life style and is doing well 10 years after intervention.

Management strategies for symptomatic myocardial bridges in the paediatric age group include surgical intervention in the form of supra-arterial myotomy, Beta-blocker therapy, automatic implanted cardioverter defibrillator, and coronary stenting. Coronary stenting is not a favored approach due to limited data and high incidence of restenosis.[14] The use of an automatic implanted cardioverter defibrillator and long-term Beta-blocker therapy may provide an extra layer of protection and may further reduce the risk of sudden cardiac death.

In summary, myocardial bridges are usually found in the context of hypertrophic cardiomyopathy/left ventricular hypertrophy and rarely with structurally normal heart. The differential diagnosis of syncope with exertion should consider myocardial bridging, supported by ischaemic data. Myocardial bridge in general is neither a reflection of severity of disease nor an independent risk factor for sudden cardiac death in majority of the cases of non-obstructive hypertrophic cardiomyopathy in children. Symptomatic myocardial bridges constitute a very small subgroup and may represent a pathologic variant, which is linked directly to myocardial ischaemia, ventricular arrhythmia, and sudden cardiac death and requires an aggressive approach.

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