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Forum for Discussion

Myocardial bridging of the anterior interventricular coronary artery in the setting of hypertrophic cardiomyopathy in children and adolescents

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HYPERTROPHIC CARDIOMYOPATHY IS A WELL recognized cause of sudden cardiac death in children and adolescents. The incidence of sudden cardiac death related to hypertrophic cardiomyopathy in children is high, and obviously age dependent.[1] Approximately half of the children with hypertrophic cardiomyopathy in whom the diagnosis is made between the ages of 1 to 14 years die suddenly within 9 years of the establishment of the diagnosis.[2] Despite recent advances in the understanding of the genetic background of hypertrophic cardiomyopathy, such as its identification as an inherited familial disease which affects the sarcomere of the myocardium, reliable therapeutic options are still unsatisfactory for those patients known to be at risk.[3] The identification of this group of patients with hypertrophic cardiomyopathy who are at high risk is a major problem. Unfortunately, the degree of myocardial hypertrophy, excluding extreme forms, and the presence or absence of obstruction in the ventricular outflow tract, are poor prognostic predictors of sudden cardiac death.[4] Better predictors are a positive family history of sudden cardiac death, recurrent syncope, ventricular tachycardias, and abnormal response of blood pressure to exercise. Positive electrophysiological studies, with ventricular tachycardias occurring after electrical stimulation of the myocardium, have a low predictive value.[5] Furthermore, it should be noted that the mechanism which finally leads to syncope or sudden cardiac death remains speculative. Possible explanations are the occurrence of sustained ventricular tachycardia, atrial fibrillation, bradycardia, myocardial ischemia, or vasodilation during exertion.

Recently, the existence or absence of myocardial bridging of the coronary arteries, and its influence on morbidity and mortality in the young patient, has been a matter of debate in patients with hypertrophic cardiomyopathy.[6]

Myocardial bridging is far from a rare finding in healthy adults.[7] Some authorities, therefore, view myocardial bridging as part of the normal myocardial arrangement. Myocardial bridging of various degrees is found in up to half of elderly patients with left ventricular obstructive disease.[7] The incidence of such bridging in healthy children is unclear. In children and adolescents with hypertrophic cardiomyopathy, nonetheless, bridging seems to play an important role. The group from Toronto[7] found almost one third of the patients to have myocardial bridging in the setting of hypertrophic obstructive cardiomyopathy. When comparing those with and without myocardial bridging, the patients with bridging had an incidence of 80% of ventricular tachycardia as opposed to 8% in the group without bridging. It was found in half of those with a history of cardiac arrest with subsequent resuscitation, as against 4% in the group without myocardial bridging. These observations were not correlated with magnitude of the left ventricular hypertrophy or the severity of obstruction in the left ventricular outflow tract. Yetman and co-workers[7] concluded that myocardial bridging is associated with a poor outcome in children with hypertrophic cardiomyopathy, and suggested that myocardial bridging is assocciated with ischemia.

Mohiddin et al.[8] found myocardial bridging in almost half of their patients with hypertrophic

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cardiomyopathy. They showed that left ventricular septal thickness, and compression of the septal perforating arteries, but not bridging, were independent predictors of thallium perfusion abnormalities. In their investigation, bridging was not significantly associated with greater symptoms, ventricular tachycardias as revealed by Holter and electrophysiologic studies, or outcome. They argued that myocardial bridging did not result in myocardial ischemia, and may not cause arrhythmias or sudden death in children with hypertrophic cardiomyopathy.[8]

In the light of these conflicting opinions, we offer the following patient, hopefully to stimulate further discussion.

Case

A 10-year-old boy with hypertrophic obstructive cardiomyopathy had resection of the myocardium in the left ventricular outflow tract at the age of 6 years, and further resection together with replacement of the mitral valve at the age of 7 years due to significant obstruction of the left ventricular outflow tract. During follow-up, we found runs of ventricular ectopic beats in 24-hour Holter recordings. The boy was not clinically impaired, and had good systolic left ventricular function documented with echocardiography. Doppler investigations showed no evidence of residual stenosis in the outflow tract of the left ventricle. In order to rule out severe myocardial bridging, we performed selective coronary angiography. The anterior interventricular coronary artery was found to have clear systolic to early diastolic milking in its medial portion (Figs 1 and 2). We opted to surgically relieve the bridging of the coronary artery. At surgery, the artery was unroofed in its obstructed portion by complete splitting of the covering myocardial tissue. The procedure was performed on the beating heart without extracorporeal circulation. Upon completion of the unroofing, the components of the myocardium which had previously bridging separated widely over a distance of about 3 cm. This freed completely the anterior interventricular coronary artery. The post operative course was uneventful. Holter recordings did not reveal ventricular ectopy.

Comment

In the context of hypertrophic cardiomyopathy, the pattern and distribution of diastolic and systolic coronary arterial flow may be influenced by significant myocardial bridging. Findings of transmural infarction have been reported at post mortem examination.[9]

The decision to operate our patient, and to unroof the anterior interventricular artery, was made upon documentation of ventricular ectopy, not detected previously in this patient. The patient had severe myocardial hypertrophy and arrhythmias, thus fitting into both categories considered at risk by the groups of Yetman[7] and Mohiddin,[8] respectively.

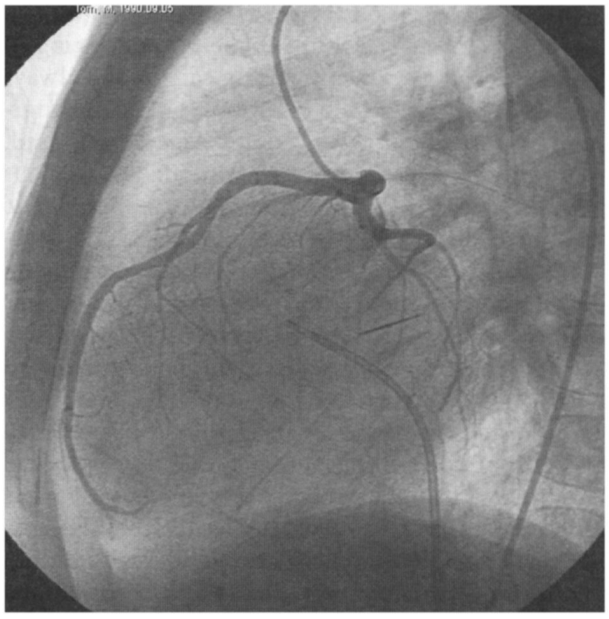

Figure 1.
Lateral view of an angiogram of the left coronary artery in a 10-year-old boy with hypertrophic cardiomyopathy after left ventricular myectomy and replacement of the mitral valve. Note the normal unobstructed lumen of the anterior interventricular coronary artery.

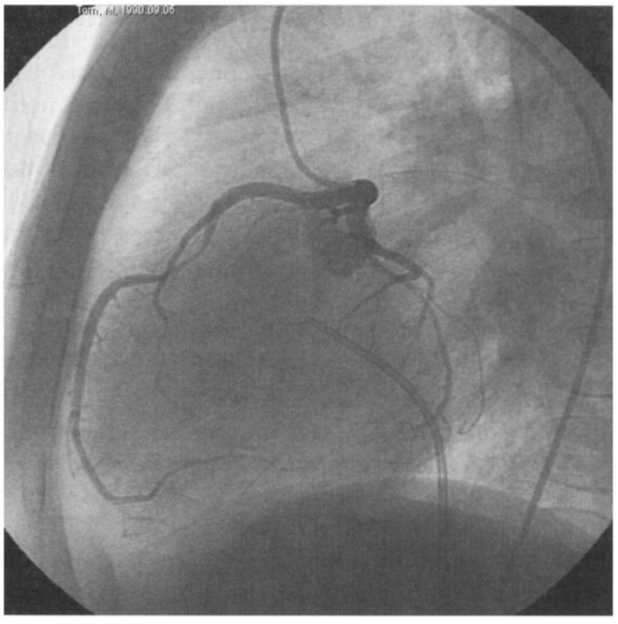

Figure 2.
Same angiography, with milking of the coronary artery in its medial part. In order to visualize the dynamic changes, a video clip of this angiogram is available on the Journals' website: http://www.greenwich-medical.co.uk.

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We tried to find out whether or not the bridging was pre-existent by looking at the former angiographies. We discovered that bridging cannot always be distinguished on the basis of left ventricular angiography. As several other groups have emphazised, selective coronary angiography is needed to rule out myocardial bridging in the setting of hypertrophic cardiomyopathy. Selective angiography of the coronary arteries is clearly not as often performed in children as in adults, and there are no indications for coronary angiography in healhy children. The true incidence of myocardial bridging in children therefore, is unknown.

We would request guidance from the readers of the Journal concerning the fact that hypertrophic cardiomyopathy too often leads to sudden death. Does significant myocardial bridging increase the individual risk for sudden death? Is there a sound argument to support the surgical unroofing procedure? If so, under which circumstances, and when?

Website

We would welcome comments, which can be posted on the website of the Journal (http://www.greenwich-medical.co.uk). The cineloops from our patient are also available for study on the website.

References

1. Maron BJ, Gardin JM, Flack JM, Gidding SS, Kurosaki TT, Bild DE. Prevalence of hypertrophic cardiomyopathy in a general population of young adults. Echocardiographic analysis of 4111 subjects in the CARDIA study. Coronary artery risk development in (young) adults. Circulation 1995; 92: 785–789.
2. Dilsizian V, Bonow RO, Epstein SE, Fananapazir L. Myocardial ischemia detected by thallium scintigraphy is frequently related to cardiac arrest and syncope in young patients with hypertrophic cardiomyopathy. J Am Coll Cardiol 1993; 22: 796–804.
3. Watkins H: Sudden death in hypertrophic cardiomyopathy. N Engl J Med 2000; 342: 422–424.
4. Spirito P, Seidman CE, McKenna WJ, Maron BJ. The management of hypertrophic cardiomyopathy. N Engl J Med 1997; 336: 775–785.
5. Maron BJ, Roberts WC, Epstein SE. Sudden death in hypertrophic cardiomyopathy: a profile of 78 patients. Circulation 1982; 65: 1388–1394.
6. McCrindle BW, Yetman AT. Myocardial bridging of the left anterior descending coronary artery in children with hypertrophic cardiomyopathy. J Am Coll Cardiol 2001; 38: 921–922.
7. Yetman AT, McCrindle BW, MacDonald C, Freedom RM, Gow R. Myocardial bridging in children with hypertrophic cardiomyopathy–a risk factor for sudden death. N Engl J Med 1998; 339: 1201–1209.
8. Mohiddin SA, Begley D, Shih J, Fananapazir L. Myocardial bridging does not predict sudden death in children with hypertrophic cardiomyopathy but is associated with more severe cardiac disease. J Am Coll Cardiol 2000; 36: 2270–2278.
9. Maron BJ, Epstein SE, Roberts WC, Hypertrophic cardiomyopathy and transmural myocardial infarction without significant atherosclerosis of the extramural coronary arteries. Am J Cardiol 1979; 43: 1086–1102.