

Symptomatic Myocardial Bridging in a Child Without Hypertrophic Cardiomyopathy

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ABSTRACT

Myocardial bridging (MB) is a rare coronary anomaly in children that is typically associated with hypertrophic cardiomyopathy or left ventricular hypertrophy. Several reports, mainly in adults, have suggested an association between MB and sudden death or ischemia without other cardiac abnormalities. In this report, we describe an 11-year-old girl with syncope and manifestations of cardiac ischemia associated with MB of the middle segment of the left anterior descending artery. The coronary anomaly was not associated with left ventricular hypertrophy. Surgical unroofing of the affected coronary artery segment resulted in complete recovery. MB should be included in the differential diagnosis of children presenting with syncope and signs of ischemia even in the absence of ventricular hypertrophy.

SYSTOLIC COMPRESSION OF the epicardial coronary arteries embedded by overlying myocardial tissue is known as myocardial bridging (MB). MB is often an incidental finding in adults. Childhood MB; however, is almost always associated with hypertrophic cardiomyopathy or left ventricular hypertrophy and has been strongly related to sudden death in hypertrophic cardiomyopathy patients.¹ Isolated myocardial bridges have been found at autopsy in adolescents who had cardiac arrest during strenuous physical activity.^{2,3}

Here we describe an 11-year-old girl who presented with syncope and ischemic symptoms and was subsequently diagnosed as having an isolated MB in the left anterior descending coronary artery that was not associated with hypertrophic cardiomyopathy. The MB was repaired surgically, and the child has remained symptom-free.

CASE REPORT

A previously healthy 11-year-old female of Arabic descent presented with recurrent episodes of syncope on exertion. The episodes lasted for up to 5 minutes and were associated with retrosternal chest pain, palpitations, anxiety, and dyspnea. Physical examination revealed a thin girl in no distress with normal vital signs. The cardiac examination was normal. Laboratory work-up did not reveal anemia or electrolyte abnormalities. The electrocardiogram on admission demonstrated normal sinus rhythm, a short PR interval (80 milliseconds),

and no electrocardiographic signs of hypertrophy, ischemia, or a prolonged QTc interval. There were nonspecific ST-T wave changes in the lateral leads. Echocardiography revealed normal cardiac and coronary anatomy. Ventricular size and function were within normal limits. During hospitalization, the patient had 2 witnessed episodes of syncope during ambulation. A 24-hour electrocardiographic evaluation revealed severe (8-mm) ST-segment depressions during ambulation and other strenuous activities. An exercise stress test, based on the Bruce protocol, was discontinued after 3 minutes because of septolateral ST-segment depressions with inverted T waves.

Dobutamine stress echocardiography induced ischemic symptoms, severe ST depression in V2, and hypokinesia of the septal wall and apical region. Thallium scintigraphy showed severe dipyridamole-induced ST depression and mild anterior wall hypoperfusion. Based on these findings, the patient underwent cardiac catheterization.

Key Words: myocardial bridging, coronary artery, hypertrophic cardiomyopathy, syncope

Abbreviation: MB, myocardial bridging

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terization, which revealed a 14-mm-long MB at the midsegment of the left anterior descending artery with a positive milking effect of ~80% systolic compression (Fig 1). Based on these data, the severity of obstruction was grade 5 according to Angelini's MB grading of 1 to 5.⁴ The patient continued to have ischemic manifestations and syncopal attacks despite medical treatment that included reduced physical activity and the administration of β -adrenergic and calcium channel blockers.

The child underwent surgery, during which the midsegment of the left anterior descending artery was noted to be embedded in the myocardium and was surgically unroofed (Fig. 2). There were no postoperative complications. Repeat stress test and electrocardiogram showed resolution of exercise and stress-induced ischemic changes. The girl remained symptom-free 24 months postoperatively.

DISCUSSION

MB occurs when a band of myocardial muscle fibers overlies a segment of the coronary artery, which results in mechanical stenosis secondary to systolic compression. The reported prevalence of MB at autopsies varies from 5% to 86%, the large variation being the result of differences in classification and/or methods of evaluation.⁴⁻⁶ Angiographically, the incidence of MB is <2% in the general adult population.¹ It is seen most commonly in the middle segment of the left anterior descending artery.⁷

Descriptions of bridging in children are rare, and its prevalence in the normal pediatric population is unknown. Nearly all reports of MB in children occur in association with hypertrophic cardiomyopathy, with an incidence of 28% among children with diagnosed hypertrophic cardiomyopathy.¹ There is an ongoing debate

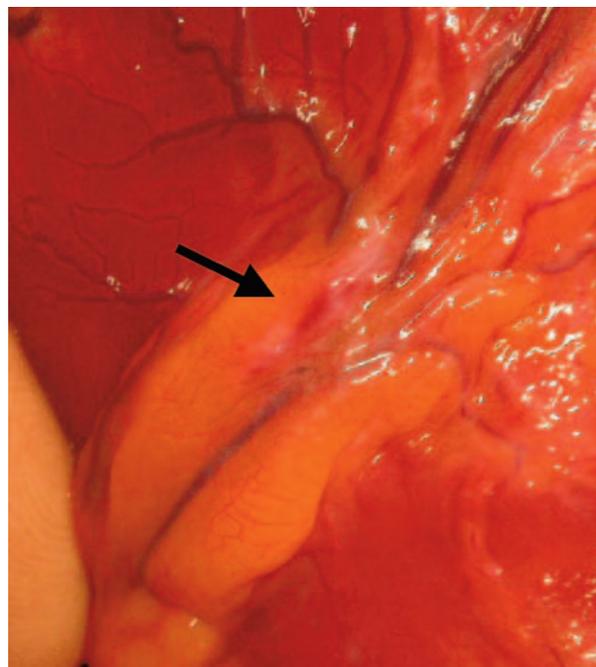


FIGURE 2
Myocardial bridge. Mid-left anterior descending artery embedded by muscle (arrow) before surgical unroofing.

whether hypertrophic cardiomyopathy-associated MB is an independent risk factor for ischemia and sudden cardiac death in children or simply an indicator of the severity of left ventricular hypertrophy. Yetman et al^{1,8} showed a strong independent relationship between MB and either chest pain, history of cardiac arrest, or reduced exercise capacity. Children with combined hypertrophic cardiomyopathy and MB had a worse prognosis as compared with those with hypertrophic cardiomyopathy alone.¹ Surgical unroofing of the MB often results in clinical improvement.⁹ On the other hand, Mohiddin et al¹⁰ demonstrated that MB does not result in myocardial ischemia, arrhythmias, and/or sudden death in children with hypertrophic cardiomyopathy. They suggested that cardiac surgery is not indicated for hypertrophic cardiomyopathy-associated MB, and medical therapy may provide adequate antiischemic therapy. Still, small series and case reports have offered compelling evidence for an association of MB and sudden death or myocardial infarction in children and young adults.¹¹⁻¹³ For example, Hillman et al¹⁴ reported a 10-year-old boy with MB of the left anterior descending artery who presented with a history of exertional chest pain, syncope, and evidence of left ventricular hypertrophy on electrocardiogram. The patient underwent successful supraarterial decompression myotomy. Downar et al⁹ reported 5 patients with hypertrophic cardiomyopathy and MB in whom symptoms or evidence of myocardial ischemia resolved or improved after supraarterial myotomy.

This case is unique because the patient presented at a

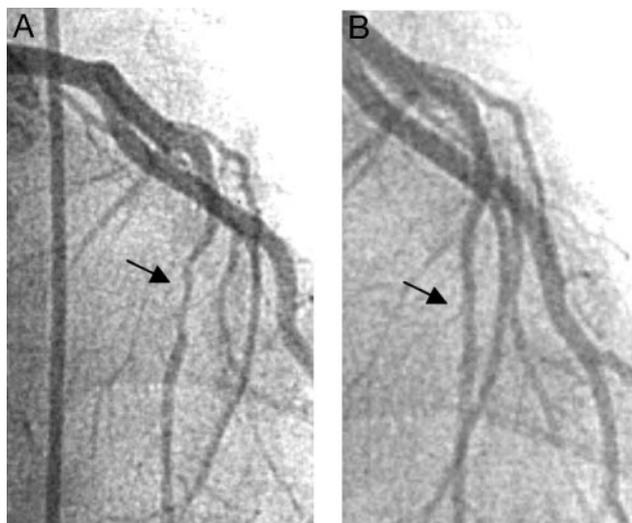


FIGURE 1
Cardiac angiography. A, Compression of the mid-left anterior descending artery during systole (arrow). B, Same segment during diastole (arrow).

relative young age with symptomatic MB without associated left ventricular hypertrophy or hypertrophic cardiomyopathy. We conclude that MB can be a risk factor for serious cardiac events in the form of anginal symptoms, myocardial ischemia, recurrent syncopal attacks, and reduced capabilities to exercise in children. Despite MB being rare in children, this entity should be considered in any child who presents with clinical or electrophysiologic manifestations that are consistent with myocardial ischemia. Because the defect is primarily mechanical, decompression surgery may be necessary to relieve the pressure on the compressed segment of the coronary artery, especially when the response to medical therapy is suboptimal.

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