A 15-year-old boy was referred for evaluation of a murmur detected during a preparticipation athletic examination (PAE) for high-school football. There was no family history of cardiopulmonary disorders. The child's birth had been a normal term delivery and he had an uneventful childhood and early adolescence. He specifically denied any exertional dyspnea, chest pain, peripheral edema, or cyanosis.

Physical examination revealed a morphologically normal male adolescent in the 90th percentiles for height and weight. He was afebrile. Respiratory rate was 14 and unlabored. Radial pulse was 80 and regular. Blood pressure was 170/80 mm Hg in the right and 170/82 mm Hg in the left arms while seated. Chest auscultation revealed clear lung fields. Precordial examination revealed a normal point of maximal impulse without lift, heave, or thrill. Heart sounds were normal with an appropriately splitting second sound. A harsh midsystolic ejection-type murmur continued into early diastole. The murmur appeared to originate at the upper sternum with radiation to the base of the neck bilaterally. There was no alteration in murmur intensity with Valsalva, squatting, or isometric maneuvers. Carotid and peripheral pulses were strong and symmetric without delays.

Transthoracic echocardiography revealed normal chamber sizes with normal left ventricular wall thickness. All cardiac valves were visualized and appeared anatomically and functionally normal. The aortic root appeared slightly dilated at 4.3 cm with evidence for turbulent flow in the proximal arch.

A chest radiograph was obtained (Fig 1).
Diagnosis: Rib notching and proximal aortic dilatation due to juxtaductal coarctation

Systolic blood pressures obtained from the lower extremities varied between 100 to 110 mm Hg, demonstrating a significant pressure drop. This finding, in combination with the rib notching on chest radiograph, prompted a thoracic MRI study that confirmed the presence of a localized juxtaductal aortic coarctation (asterisk, Fig 2). Also visible on these left anterior oblique sections are the slightly enlarged proximal aorta (arrowhead) and dilated left subclavian and inominate arteries (arrows, Fig 3).

Thoracic aorta coarctation is the fourth most common cardiac defect in infancy and childhood. It occurs at a frequency of 0.2 to 0.6 per 1,000 live births. It is frequently associated with other congenital cardiac defects, including left ventricular hypoplasia, parachute mitral valve, and, in up to 85% of cases, a bicuspid aortic valve. Despite this frequent association with other anomalies, there is no evidence for a mendelian inheritance. Current theories regarding the pathogenesis of juxtaductal aortic coarctation suggest that it evolves as an acquired vascular deformation due to abnormal hemodynamics. During fetal life, the ascending and descending aorta are divided by the narrowed aortic isthmus. Ascending aortic flow is provided primarily by the left ventricle with the bulk of distal aortic flow originating from the right ventricle via the ductus arteriosus. If a cardiac anomaly is present that impairs left ventricular output (and consequently increases ductal flow) or if the ductus joins the aorta at a right angle, the subsequent turbulent flow induces intimal hyperplasia along the opposite aortic wall. This leads to the ingrowth of a fibroelastic flap that interferes with distal aortic outflow when the ductus arteriosus closes shortly after birth.

The clinical manifestations of aortic coarctation depend on the degree of obstruction and the presence of associated anomalies. Severe outflow obstruction leads to early congestive heart failure in infancy and requires prompt surgical or dilatory intervention. More moderate degrees of stenosis allow for circulatory compensation with the development of arterial collaterals and left ventricular hypertrophy. Most children remain asymptomatic although occasionally complaints of exercise-induced claudication or cold extremities prompt medical evaluation. More often these young patients come to medical attention because of hypertension or precordial murmurs, as in this case. The origin of the murmur remains controversial but may arise from turbulent aortic flow, increased flow through tortuous collateral vessels, or from a coexistent bicuspid aortic valve.

The radiographic features of aortic coarctation result primarily from aortic dilation and the development of extensive arterial collaterals. When severe, coarctation may produce prestenotic and poststenotic aortic dilatation visible on plain chest radiographs. The ascending aortic contour may widen as seen in Figure 1. The “3” sign of aortic coarctation refers to an indented aortic knob and dilated distal vascular segment occasionally visible in the aortopulmonary window on chest radiograph (not seen in our case). A dilated left subclavian artery may form a widened upper mediastinum or left apical cap (Fig 1). Rib notching is perhaps the best recognized radiographic feature of coarctation. Rib notching appears during adolescence as collateral blood flow develops around the coarctation. The primary route
for collateral flow is between the subclavian system and the third through ninth posterior intercostal arteries. Progressive dilation of the intercostal arteries leads to erosion and remodeling of the inferior aspect of the ribs (Fig 1, ribs 4 and 5 on right and rib 5 on left). Rib notching is seen bilaterally unless the coarctation is proximal to the left subclavian takeoff (right-sided notching) or the innominate artery originates from the descending aorta (left-sided notching). Once considered pathognomonic of coarctation, rib notching is now known to accompany several disorders that produce enlargement of the intercostal neurovascular bundle, including subclavian stenosis, chronic superior vena cava obstruction, and neurofibromatosis.3

Confirmation and localization of the coarcted segment requires an aortic imaging procedure. Aortography provides the opportunity to measure pressure gradients and can be coupled with coronary angiography in older patients. However, this procedure is invasive and not without risk. Echocardiography has been reported to effectively visualize aortic coarctations and facilitates screening for other associated cardiac anomalies.1 Magnetic resonance imaging allows for noninvasive examination of the entire thoracic vascular tree and should be the initial, and perhaps only, visualization procedure prior to correction in many cases.4 The presence of a focal juxtaductal stenosis along with dilated subclavian and innominate arteries confirms the diagnosis.

If left uncorrected, 75% of patients with aortic coarctation die by age 50 years.1,5 This increased mortality is attributable to thoracic aortic rupture/dissection, aortic valvular incompetence or endocarditis, and the sequelae of chronic hypertension. Operative intervention with resection or aortoplasty remains the standard surgical approach.5 Balloon aortic angioplasty has been used successfully in infants, adolescents, and adults, but long-term studies are needed to assess the risk of restenosis and aneurysm.6

Given this child's severe resting hypertension and marked upper to lower extremity systolic pressure drop, it was recommended that he not participate in high-school football.7,8 He was referred for surgical repair, following which he should be able to return to competitive sports in about 1 year's time.9 In this era of increasing concern over health-care expenditures, the utility of large-scale PAE for high-school students has come under scrutiny.7 The PAE appears useful in documenting appropriate developmental age for competitive sports and in detecting orthopedic conditions that predispose to musculoskeletal injury.7 However, its utility in detecting life-threatening cardiovascular diseases remains unclear. Most sports medicine organizations stress that a mass screening PAE cannot and should not substitute for a primary care physician-patient relationship.7,8 We believe our case nicely illustrates the effectiveness of an astute screening primary care giver and a timely subspecialty referral.

REFERENCES
1 Bashmore T, Lieberman E. Aortic/mitral obstruction and coarctation of the aorta. Cardiol Clin 1993; 11:617-41