Irregular Rhythm in a 25-Year-Old Man With Three Prior Cardiac Operations

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The patient underwent closure of an atrial septal defect at age 3, had a leaking “mitral” valve repaired at age 9, and at age 13 had a “mitral” valve replacement. He began taking warfarin sodium at that time and remained symptom-free until 10 days before his initial visit here when he presented to another hospital with dyspnea and palpitations. Treatment there consisted of lisinopril 10 mg qd, carvedilol 6.25 mg bid, aldactone 25 mg qd, furosemide 40 mg qd, digoxin 0.25 mg qd, and a continuation of warfarin sodium 7.5 mg qd. An echocardiogram showed a left ventricular ejection fraction of 20%. After diuresis, he was referred to our cardiology clinic.

On his initial visit here, his heart rate was an irregular 120 beats/min, his blood pressure was 106/77 mmHg, and closing and opening snaps of a normally functioning mechanical mitral valvular prosthesis were heard. He was obese (height, 5' 9’’; weight, 272 lbs). An electrocardiogram was recorded (Figure 1).

Figure 1

What is your diagnosis?

Explication is on p. 41
ECG of the Month
Presentation is on p. 40

DIAGNOSIS: Coarse atrial fibrillation with a rapid ventricular response, left anterior fascicular block, left ventricular hypertrophy with repolarization abnormality.

Coarse atrial fibrillation is characterized by fibrillatory waves > 0.1 mV (here up to 0.3 mV in lead V₁), which have been considered a sign of left atrial enlargement and have been associated with rheumatic heart disease and congenital heart disease, whereas fine fibrillatory waves have been associated with atherosclerotic disease. Surawicz, however, thinks that separating atrial fibrillation into coarse and fine forms is not justified clinically in the current era. Coarse atrial fibrillation is sometimes mistaken for atrial flutter. Atrial flutter waves, however, all have the same morphology, including the same height and width, and the rate is usually 250 - 350 flutter waves/minute. None of these criteria is met in this electrocardiogram, and the rate is nearly 450/minute.

Criteria for left anterior fascicular block as proposed by the World Health Organization/International Society and Federation of Cardiology (WHO/ISFC) task force are left axis deviation of the QRS in the frontal plane of -45° to -90°, qR pattern in aVL, R peak time in aVL ≥45 ms, and QRS duration <0.12s. The ECG in Figure 1 meets all four of those criteria and, in addition, shows two other features usually present in left anterior fascicular block: a rS pattern in the inferior leads and, because of the broad counterclockwise loop inscribed by the QRS in the frontal plane, the R wave peaks in aVL before it peaks in aVR.

The QRS voltage is huge and meets most of the standard criteria for left ventricular hypertrophy (LVH). In addition, it meets two criteria for LVH that have been validated in patients with left anterior fascicular block: S wave in lead III + largest R+S in any single precordial lead >3.0 mV6 and SV₁ + RV₅ + SV₅ > 2.5 mV7. Morphologic and pathophysiological diagnoses in this patient can be made from the history and the electrocardiogram. The atroventricular septum in the normal heart separates the left atrium from the left ventricle. Absence of the atroventricular septum results in a common atroventricular junction with a common atroventricular valve that may have left and right orifices or a single orifice. Depending on the attachments, or lack thereof, of the common atroventricular valve to the atrial and ventricular septums, there may be a defect in the lowermost portion of the atrial septum, in the ventricular septum, or both. On the left side of the common valve, there is usually a space or cleft between the superior and inferior bridging leaflets such that during systole, part of the left ventricular ejectate regurgitates into the left atrium and often across the atrial septal defect into the right atrium. There may also be incompetence of the right side of the common atroventricular valve. A disproportionately long outlet dimension of the ventricular septum and a narrow subaortic outflow tract result in the characteristic gooseneck deformity seen on left ventriculography.

In patients with atroventricular septal defect, the left anterior fascicle is hypoplastic and longer than usual, features that probably account for the pattern of left anterior fascicular block so commonly seen. Other electrocardiographic findings may include varying degrees of atroventricular block and right ventricular hypertrophy if a large left-to-right shunt and/or pulmonary arterial hypertension are present.

The cleft in the left side of the common atroventricular valve, often erroneously called a cleft in the anterior leaflet of the mitral valve, is not easily repaired. That despite two prior open-heart operations our patient required a third to replace the left side of the common atroventricular valve to prevent ventricularialtrial regurgitation is not surprising. During his six months in our clinic, his ventricular rate has slowed; his symptoms have disappeared; and his left ventricular ejection fraction, as determined by echocardiogram, has increased from 20% to 40%. Left atrial enlargement and left ventricular hypertrophy and dilatation persist. The mean diastolic pressure gradient across the prosthetic valve is 6 mmHg, and pulmonary arterial systolic pressure is <35 mmHg.

REFERENCES


Drs. Glancy, Ahmed, LeLorier, and Helmcke are Faculty, and Drs. Ayalloore and Diwan are Fellows at Louisiana State University Health Sciences Center and the Louisiana State University Interim Public Hospital in New Orleans.