Permanent Atrial Standstill: The Clinical Spectrum

JAMES WOOLLISCROFT, MD
NAIP TUNA, MD, PhD, FACC

Ann Arbor, Michigan and Minneapolis, Minnesota

Permanent atrial standstill appears to be an arrhythmia that develops because of extensive myocardial disruption. It has been noted in three clinical settings: in patients with long-standing cardiac disease, in patients with neuromuscular disease and in otherwise healthy people who present with vertigo, syncope or stroke. A patient with this arrhythmia and limb-girdle muscular dystrophy is presented.

Permanent atrial standstill is an infrequently recognized arrhythmia that was initially well described in 1946 by Chavez et al.[1] The current diagnostic criteria include: (1) absence of P waves in surface and intracavitary electrocardiograms; (2) absence of A waves in jugular venous pulse and right atrial pressure tracings; (3) supraventricular type QRS complex; (4) immobility of atria on fluoroscopy; and (5) inability to stimulate the atria electrically.[2]

Permanent atrial standstill has been diagnosed in three clinical settings: in patients with long-standing progressive cardiac disease, in patients with neuromuscular disease and in patients with absent P waves in an electrocardiogram taken during evaluation of vertigo, syncope or stroke. Recognition of the settings in which permanent atrial standstill occurs may facilitate the diagnosis and the initiation of appropriate therapy. An illustrative case is presented:

Case History

A 34 year old white man was referred to the University of Minnesota Hospitals for evaluation of bradycardia and an abnormal electrocardiogram. At age 14, he was noted to have cardiomegaly during an evaluation for a "functional" murmur. He was hospitalized for left anterior chest pain at age 21 years. Electrocardiograms showed what was interpreted as "a nodal rhythm with a rate of 38 to 52/min." At age 29, he experienced the sudden onset of slurred speech, dizziness and numbness in his left arm that resolved within 20 minutes. He was examined in an emergency room where the electrocardiogram showed absent P waves and a heart rate of 42 beats/min. He noted progressive weakness and wasting of his upper arms and pelvic musculature beginning in his late 20s.

Physical examination showed a slender man with marked upper arm and shoulder girdle atrophy. His blood pressure was 134/62 mm Hg with no orthostatic decrease; the pulse was 38/min and regular. The point of maximal impulse was in the fifth intercostal space 4 cm lateral to the mid clavicular line. Jugular venous A waves were absent. A grade 2/6 systolic ejection murmur was best heard at the left sternal border.

All laboratory data were normal except serum creatine kinase (CK), which was elevated in the range of 149 to 166 U/liter (normal 0 to 65). The CK-MB isoenzyme ranged from 2 to 4 U/liters (normal 0 to 2.0). The electrocardiogram showed bradycardia and absence of P waves (Fig. 1). An echocardiogram showed left atrial and ventricular enlargement. The aortic, mitral and tricuspid valves were normal. No evidence of "atrial kick" was present on recordings of the mitral valve indicating absence of left atrial mechanical activity (Fig. 2). A chest X-ray

From the Department of Internal Medicine, Division of Primary Care/Community Medicine, The University of Michigan, Ann Arbor, Michigan and the Department of Internal Medicine, Section of Cardiology, University of Minnesota, Minneapolis, Minnesota. Manuscript received June 16, 1981; revised manuscript received December 2, 1981, accepted December 10, 1981.

Address for reprints: James Woolliscroft, MD, Primary Care/Community Medicine, D5101 South Ambulatory Care Building, The University of Michigan Medical School, Ann Arbor, Michigan 48109.

film showed cardiomegaly. Fluoroscopy showed a large right atrium and noncontractile atria.

A treadmill exercise test was terminated because of fatigue in stage III of the Bruce protocol at 6 minutes, 5 seconds. The heart rate rapidly increased to 120 beats/min during exercise. The QRS configuration did not change and no P waves were evident.

Atrial electrical activity measured with intracardiac electrodes was absent. Atrial pacing was attempted at multiple sites at up to 15 mA with no response. The ventricles were easily paced with 0.5 mA. There was no evidence of retrograde atrial activity during ventricular pacing. Resting hemodynamic data, at a heart rate of 40 beats/min, showed a slight increase in left and right side filling pressures and a dip and plateau configuration in the right ventricular pressure tracing that was characteristic of a restrictive cardiomyopathy. Ventricular pacing produced an increase in cardiac output (Table I).

The clinical features, muscle biopsy, electromyogram and nerve conduction studies were consistent with the diagnosis of limb-girdle muscular dystrophy. A right ventricular demand pacemaker was inserted and treatment with oral anticoagulant agents was begun. Muscular weakness has progressed, but no other problems have developed in 3 years of follow-up study.

Family history: Evaluation of the patient's son showed a normal electrocardiogram and no muscular abnormalities. The patient's brother had a history of bradycardia and a murmur noted at age 14 years. At age 23 years, cardiac catheterization showed multichamber cardiac enlargement without valve disease. The electrocardiogram showed absent P waves and a supraventricular type QRS complex at a heart rate of 38 beats/min. He died suddenly at age 32 years. The heart weighed 500 g at autopsy. Autolysis prevented microscopic examination. There was no history of muscle weakness.

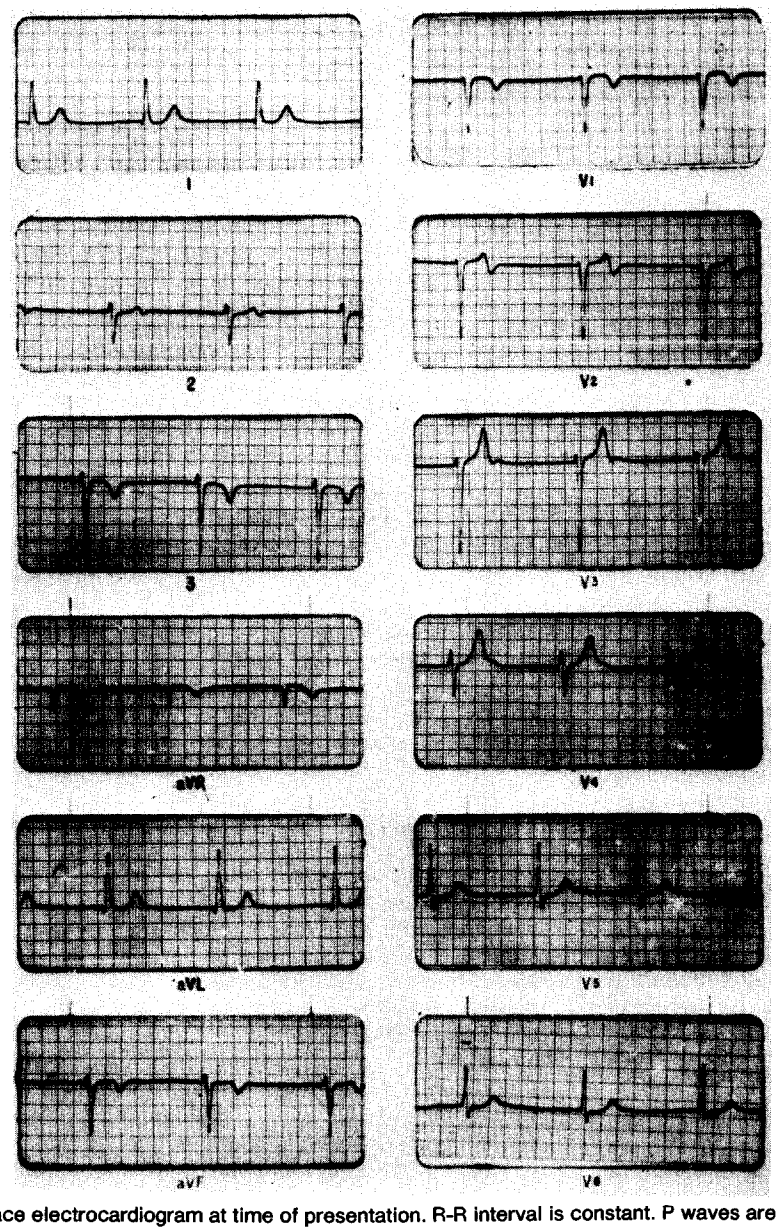

FIGURE 1. Surface electrocardiogram at time of presentation. R-R interval is constant. P waves are absent throughout.

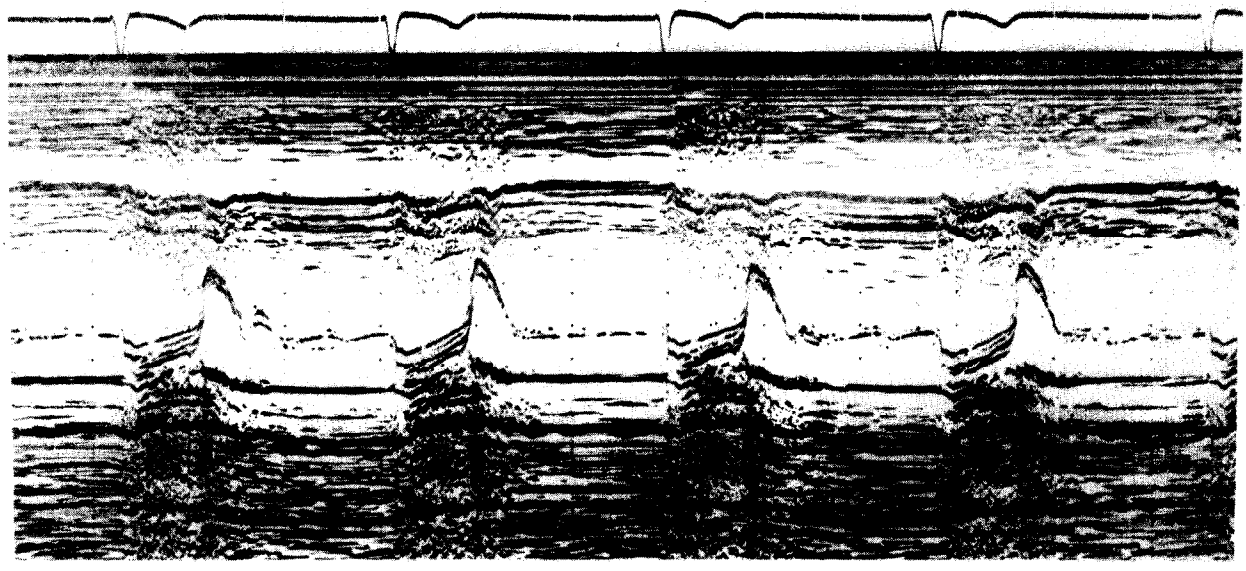

FIGURE 2. Echocardiogram of mitral valve. End-diastolic "atrial kick" or A wave of anterior mitral leaflet is absent.

Discussion

We have identified 55 patients with well-documented permanent atrial standstill. They fall into three clinical groups (Table II).

Patients with long-standing cardiac disease: In the first group permanent atrial standstill develops progressively as a sequela to long-standing cardiac disease with a prior history of supraventricular arrhythmias. There appears to be a continuum of myocardial disruption and fibrosis that may result in permanent atrial standstill. The first step is temporary atrial standstill that spontaneously converts to normal sinus rhythm. Patients with this condition respond to atrial pacing during standstill.[41] Further impairment is evidenced by absent atrial activity in surface electrocardiograms, fluoroscopy and pressure tracings. However, intracardiac electrocardiograms show low voltage atrial potential and a response to atrial pacing at high voltage remains.[3] More limited right atrial activity localized to the septum and responsive to atrial pacing only in that area[4] progresses to similar findings of low voltage atrial activity recorded in the tricuspid area[5-8] or high lateral atrial wall,[9] but without response to atrial pacing.

Perhaps the final progression to permanent atrial standstill is documented in a patient with two surface electrocardiographic patterns; neither showed atrial electrical activity.[10] Intracavitary electrocardiograms showed no electrical activity in the right atrium and coronary sinus during the first pattern. Atrial pacing was unsuccessful. Intracavitary electrocardiograms during the second pattern showed regular electrical activity in the lower portion of the right atrium and coronary sinus. Epicardial mapping showed low amplitude atrial activity in the tricuspid area, with outward propagation from a center near the tricuspid area. Bi-

TABLE I

Hemodynamic and Ventricular Pacing Study

	Intrinsic Rhythm	Pacing		
		60/min	80/min	100/min
Wedge pressure (mm Hg)	(18)	(16)	(16)	(17)
Aortic pressure (mm Hg)	151/81(109)	151/87(120)	153/94(131)	194/100(12)
Pulmonary arteriolar pressure (mm Hg)	40/18(25)	39/16(27)	40/16(27)	40/20(26)
Right atrial pressure	(11)	—	—	(7)
Cardiac output (liters/min)	5.5	7.8	9.2	9.9
Cardiac index (liters/min per m^2)	3.1	4.5	5.3	5.7
Heart rate (beats/min)	40	60	81	100
Stroke volume (ml)	138	130	114	99
Systemic resistance (dynes/cm^{-5})	1,585	1,231	1,139	994
Total pulmonary resistance (dynes/cm^{-5})	364	277	235	210
Pulmonary arteriolar resistance (dynes/cm^{-5})	102	113	91	72

Values in parentheses indicate mean of systolic and diastolic pressure.

TABLE II

Clinical Characteristics in 55 Reported Cases

	Age (yr) (mean & range)	Sex	History of Syncope†	History of TIA or Stroke†	Cardiomegaly†
Group I (n = 18)[1,3–19] (PAS in patients with long-standing cardiac disease)	57 (25–78)	9M; 9F	3/15	0/15	15/15
Group II (n = 12)[2,20–26] (PAS in patients with neuromuscular disorders)	31 (15–46)	9M, 3F	1/12	4/12	5/6
Group III (n = 24)[8,14,27–40] (PAS in apparently healthy persons)	36* (21–61)	12M, 6F	12/21	4/21	16/18

* Age and sex data available in only 18 patients. † Data not available in all reports; the denominator indicates the number of patients for which information is available.
PAS = permanent atrial standstill; TIA - transitory cerebral ischemic attack.

opsy of the right atrium showed diffuse atrial fibrosis.

All available pathologic data from patients with permanent atrial standstill have confirmed severe and widespread atrial fibrosis and degeneration.[1,6,9–13,27–30] The arrhythmia appears to be a nonspecific result of advanced structural changes.

Patients with neuromuscular disease: The second clinical group in which permanent atrial standstill occurs is in association with various neuromuscular diseases. Originally described in patients with fasciioscapulohumeral muscular dystrophy,[2,20–21] atrial standstill has now been recognized with Charcot-Marie muscular dystrophy,[22] an X-linked humeroperoneal neuromuscular disease,[23] Emery-Dreifuss muscular dystrophy,[24] and limb-girdle muscular dystrophy.[25,26] Our patient is the first with limb-girdle muscular dystrophy in whom the onset of permanent atrial standstill preceded the clinical muscular disease. The finding of permanent atrial standstill in this group of patients is not surprising because Perloff et al.[12] emphasized the frequency of myocardial disease as a feature of the muscular dystrophies; they described the spectrum of cardiac dysfunction ranging from asymptomatic supraventricular arrhythmias to severe failure and death. Autopsy findings showed diffuse myocardial fibrosis and scarring.

Patients with syncope, vertigo or stroke: The third group of patients in whom permanent atrial standstill occurs, generally in the 3rd to 5th decade, are without known cardiac or neuromuscular disease. The diagnosis is made when the patient experiences vertigo,[27,31,32] syncope[14,30,33–35] or stroke[14,36,37] or during a routine physical examination.[38] These patients deny previous cardiac symptoms except for awareness of their slow heart rate. If the observations in other patient groups hold true, one would expect these apparently healthy persons to have extensive myocardial disease which is otherwise silent. In one series, biopsy of the right atrium or ventricle, or both, showed diffuse degeneration, an increase in interstitial tissues and thickening of the right atrial endocardium.[29]

Familial occurrence: As in our case, the familial occurrence of permanent atrial standstill has been noted with muscular dystrophy[22,23] as well as in apparently healthy persons.[7,12,35,36,39] Further longitudinal family studies as well as pathologic study are needed to clarify the disease process in these persons.

Diagnosis: The diagnosis of permanent atrial standstill is important because the morbidity is high. It should be suspected in any patient with absent P waves in the surface electrocardiogram and a regular R-R interval. The echocardiogram provides another noninvasive means to demonstrate atrial standstill. Under normal conditions the mitral valve is ballooned out by the force of the atrial contraction at the end of diastole. In atrial standstill, as in atrial fibrillation, the atrial kick is not present on mitral valve recordings (Fig. 2). If the noninvasive studies are consistent with permanent atrial standstill, invasive studies are needed to confirm the diagnosis.

Treatment of patients with permanent atrial standstill should be directed toward the major complications of this condition which consist of syncope, stroke, congestive heart failure and sudden death. Permanent ventricular pacing and long-term anticoagulation appear to be the most promising treatments for these patients.

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