Persistent right atrial standstill

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An asymptomatic patient with cardiomegaly caused by isolated right atrial standstill is reported. The right atrium showed no evidence of contraction on pressure records or angiographically, while the left atrium functioned normally.

Although temporary atrial standstill may result from heart disease, cardioversion, and cardiac surgery, persistent atrial standstill seems to be rare. A number of sporadic and familial cases have been reported, many associated with other heart disease or drugs. All had standstill of both atria. Persistent standstill of only one atrium in the absence of other heart disease does not appear to have been described previously.

Case report

A woman of 36 was referred for investigation of cardiomegaly associated with right bundle-branch block and a systolic murmur. She denied effort intolerance or any other symptom. The cardiomegaly had developed gradually over the previous 10 years, during which period she had had routine annual chest films taken at her place of employment. She denied any previous illness. There was no known family history of heart disease or muscular dystrophy. Clinically she appeared fit but was moderately overweight. Regular rhythm was present with a resting heart rate of 50/min. The blood pressure was 140/80 mmHg (18.6/10.6 kPa); 'a' waves were not visible in the venous pulse and were absent from a jugular venogram. No cannon waves were seen. The apical impulse was slightly displaced to the left. First and second heart sounds were normal. A soft third heart sound was audible intermittently. There was a widespread short, high pitched midsystolic murmur but no diastolic murmur. The radiological cardiothoracic ratio was 19/30.5 and the right atrium appeared to be enlarged. Over a three-month period the electrocardiogram showed a regular rhythm with very low voltage P waves, manifest only in leads III, aVL, and aVF, and right bundle-branch block. The PR interval was 0.12 s. The cycle length varied slightly as in sinus arrhythmia.

Cardiac catheterization was performed principally to exclude atrial septal defect and Ebstein's anomaly. There was no left-to-right shunt or any valvar stenosis or regurgitation. Cardiac pressures were within normal limits. The resting cardiac output was 6.6 l/min by indicator dilution. Angiographic left ventricular volume and ejection fraction, wall thickness, and left ventricular end-diastolic pressure were normal. The tricuspid valve was normally situated.

FIG. 1 Simultaneous right atrial and ventricular pressure records using side hole catheters on each side of tricuspid valve. The 'a' wave is absent. Electro-mechanical interval appears longer than in Fig. 2 since both the above pressures were filtered before recording (-3dB at 5.3 Hz).

FIG. 2 Simultaneous unfiltered right atrial pressure (RAP) and bipolar intracavitary electrogram (RAE) recorded from mid lateral right atrial wall (-3dB at 0.08 Hz and 40 Hz). The normally large atrial complex is absent. The very small wave indicated by P may represent distant left atrial activity.

FIG. 3 Simultaneous pulmonary arterial (PA) and direct left atrial (LA) pressure (filtered as in Fig. 1) recorded with electrocardiogram lead II. There is a normal left atrial 'a' wave beginning 0.04 s before the QRS complex.

The right atrium showed no evidence of mechanical activity. There was no 'a' wave in the right atrial or right ventricular pressure tracings (Fig. 1). Fluoroscopy

and right atrial angiography showed no right atrial contraction. Bipolar intracavitary electrograms from the mid lateral right atrial wall showed a single low amplitude biphasic complex synchronous with the right atrial 'c' wave instead of the high amplitude presystolic atrial complex normally recorded in this area (Fig. 2). Preceding this complex a very low amplitude oscillation of the baseline was just discernible (P in Fig. 2), possibly reflecting remote left atrial activity, corresponding to the low amplitude P waves in the surface electrocardiogram. In contrast, the direct left atrial pressure record (Fig. 3) showed both 'a' and 'v' waves, and left atrial contraction was visible angiographically. The right atrium was considerably enlarged while the left atrium was of normal size. The cardiomegaly appeared to be entirely the result of the right atrial enlargement. The patient remains well and requires no treatment.

Discussion

The diagnosis of right atrial standstill in this case was based on the absence of right atrial mechanical activity, absence of the normal atrial complex in the right electrogram, and isolated right atrial enlargement. There was a striking contrast between the large, motionless right atrium and the small, normally contracting left atrium when each was visualized angiographically. There was nothing to suggest other heart disease, muscular dystrophy, or any systemic disorder.

At first sight the single complex shown in the mid right electrogram in Fig. 2 suggests ventricular activation. It would, however, be difficult to distinguish this from abnormally reduced right atrial activity occurring at the same time. For this to be so, right atrial activation would have to occur about 0.1 s later than left atrial activation to explain the timing of left atrial contraction. Either right atrial electrical activity is absent, as in the case described by Patton *et al.* (1970), or there is an intra-atrial conduction defect, in which right atrial activation is delayed. This might occur if the underlying rhythm originated near the AV node or in the left atrium. However, the heart rate and phasic variations in cycle length suggested sinus rhythm rather than an ectopic atrial or AV nodal rhythm. If so, this would imply that conduction from sinus node to left atrium remained intact despite absence of right atrial activation and raises the possibility of sinoven-tricular conduction along the specialized conducting pathways in the right atrium, unless the AV node was activated via left atrial pathways. Interatrial block or dissociation, though uncommon, is well described (Deitz *et al.*, 1957; Cohen and Sherf, 1965). However, no mention has been found of any case with documented persistent unia-trial asystole or right atrial fibrillation in the presence of left

atrial rhythm. Two of the patients reported by Rowlands, Logan, and Howitt (1967) did not develop left atrial activity after cardioversion for atrial fibrillation, when right atrial activity returned. In one case this situation was confirmed at thoracotomy. The reverse situation did not occur in their series. Rubino (1936) reported left atrial standstill with right atrial sinus rhythm in one case in response to ocular pressure.

The cause of the abnormality in this case is obscure. Right atrial standstill has been reported in patients with facioscapulohumeral dystrophy (Bloomfield and Sinclair-Smith, 1965; Caponnetto, Pastorini, and Tirelli, 1968; Baldwin *et al.*, 1973), suspected cardiac amyloid disease (Allensworth, Rice, and Lowe, 1969; Harrison and Derrick, 1969), and familial heart disease (Allensworth *et al.*, 1969; Harrison and Derrick 1969; Williams *et al.*, 1972).

Many reported cases had features of other heart disease (Rosenbaum and Levine, 1939; Magnusson, 1946; Nasi and Legnani, 1966; Jouve *et al.*, 1967; Raynaud *et al.*, 1968; Patton *et al.*, 1970), but cases without evidence of other heart disease have been reported (Wada, Takada, and Mise, 1966; Jouve *et al.*, 1967).

Of interest is the similarity between the present case and cases of idiopathic right atrial enlargement first described by Pastor and Forte (1961) and more recently reviewed by Eshaghpour, Olley, and Collins (1969). The three cases described by Pastor and Forte were not catheterized, and all had associated medical problems. In two catheterized cases (Sumner *et al.*, 1965) the right atrial electrogram, pressure, and angiogram were stated to be normal, though the specific presence of P and 'a' waves was not mentioned. The authors stated, however, that there was 'no evidence to support the existence of an abnormal sequence of activation, or contraction in the involved chamber'. Eshaghpour *et al.* (1969) illustrate a right ventricular and atrial withdrawal pressure tracing from a 6-year-old boy with idiopathic right atrial enlargement in which it is not clear whether 'a' waves are present or absent.

Possibly cases of persistent atrial standstill and idiopathic right atrial enlargement share a common pathology.

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Addendum

After this case report went to press, the patient was noted to have developed complete atrioventricular dissociation with a ventricular rate of 42/min and a left atrial rate of 56/min. The P waves were visible in the surface electrocardiogram, and in bipolar recordings from the coronary sinus.

Dissociated atrial waves were present in an apex cardiogram. In contrast, there was again no detectable mechanical or electrical activity in the right atrium. The left atrium was easily paced via the coronary sinus while there was no response to direct bipolar stimuli of up to 7 volts to the right

atrial endocardium. The patient remains well with a permanent pacemaker.

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