Atrial Flutter Associated with Isolated Pulmonic Stenosis

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SUMMARY
Two adults with severe pulmonic valvular stenosis and atrial flutter are presented. This arrhythmia is apparently a rare complication of this particular form of congenital heart disease. In both cases surgical correction was achieved with the aid of total body perfusion. At the time of surgery, significant tricuspid insufficiency was discovered in both cases. Although the tricuspid valve was not repaired, insufficiency of this valve was not evident postoperatively. Cardioversion, applied at a different stage in each, terminated the atrial flutter in both patients. Sinus rhythm has been maintained for almost 2 years without drug therapy. The optimal time for cardioversion in such cases is discussed.

Additional Indexing Words: Valvular pulmonic stenosis Open heart surgery Cardioversion

ALTHOUGH supraventricular arrhythmias are a frequent late manifestation of many forms of congenital heart disease, patients with pulmonic stenosis and intact septum appear to be singularly free of this complication. Textbooks on congenital heart disease make no mention of atrial flutter in their descriptions of this condition,1-4 nor has it been described in the studies of large series of cases published in the recent literature.5-9

Our purpose was to present two adults with severe, isolated pulmonic valvular stenosis, in whom atrial flutter was detected at the time of diagnosis. In both patients cardioversion terminated this arrhythmia, being performed before surgery in one patient and in the postoperative period in the other. Other problems in the management of the patients are also discussed.

Report of Cases

Case 1
C.P., a 35-year-old man, was aware of the presence of a heart murmur since the age of 12.

He remained symptom free until 6 months prior to admission to hospital, when he began to suffer from palpitations, fatigue, and dyspnea on effort. On admission, physical examination revealed a slightly underdeveloped patient, cyanotic and dyspneic at rest. Pulse rate was 130/min and regular blood pressure was 120/70 mm Hg. A right ventricular uplift and a systolic thrill over the pulmonic area were palpated. A harsh systolic murmur was heard over the pulmonic area. The liver edge was palpable 2 cm below the costal margin. Hemoglobin was 18 g% and hematocrit was 55%. Chest roentgenograms showed a markedly enlarged right ventricle, prominence of the main pulmonary artery, and decreased vascularity of the peripheral lung fields. The electrocardiogram revealed atrial flutter with 2:1 or 3:1 atrioventricular conduction and severe right ventricular strain (fig. 1).

The subjective symptoms and the signs of congestive heart failure led us to the decision to instigate conversion to sinus rhythm. Under general anesthesia the flutter was terminated by the first countershock of 150 watt-sec intensity. The P waves were now notched and very prominent (fig. 2). Four days later right heart catheterization disclosed a gradient of 180 mm Hg over the pulmonic valve (table 1); no signs of shunt were disclosed. The patient was prepared for surgery by rest in bed, digitalization, and diuretics, and he lost 5 kg during 2 weeks. At operation, the right atrium, the right ventricle, and the main pulmonary artery were grossly enlarged. The interior of the right atrium was
explored and a tricuspid regurgitant jet of moderate intensity was discovered. The patient was placed on cardiopulmonary bypass, and the pulmonary artery was opened longitudinally. The pulmonary valve was very stenotic, its orifice being 3 mm in diameter, and the cusps were fibrotic and thickened. The stenosis was relieved by cutting the three commissures, and a wide opening of the order of 25 mm was achieved. The interior of the right ventricle was examined with the finger, and concentric hypertrophy of all the musculature was noted; there was no evidence of localized infundibular obstruction. The right atrium was now opened. There was no defect in the atrial septum. The tricuspid annulus was dilated, but the cusps were quite normal, and it was obvious that this valve was the seat of secondary insufficiency only. No repair was carried out.

Except for transient signs of mild cardiac failure during the second postoperative week, the patient’s recovery was smooth. Cyanosis disappeared completely, and soon after leaving the hospital, he returned to his employment as a clerk, relieved of all his symptoms. Fourteen months after surgery he was recatheterized (table 1). At this time the systolic gradient across the pulmonic valve was 20 mm Hg. The overall follow-up period after the valvotomy is now 22 months; he is not receiving drugs, and has had no recurrence of the atrial flutter or other arrhythmia.

**Case 2**

H.J., a 40-year-old mother of two children, was unaware of any heart condition until the age of 34 years. At that time she began to suffer from easy fatigability, palpitations, dyspnea,
and precordial pain. Over the years she became severely incapacitated. On admission, physical examination revealed an obese woman, slightly dyspneic and cyanotic at rest. Heart rate was 130/min and blood pressure was 110/80 mm Hg. A right ventricular uplift and systolic thrill over the pulmonic area were palpated. A very harsh systolic murmur and a faint second sound were heard over the pulmonic area. The liver
edge was just palpable. Hemoglobin was 17 g% and hematocrit 56%. Chest roentgenograms showed aneurysmal dilatation of the main pulmonary artery and extreme enlargement of the right ventricle and right atrium (fig. 3). The peripheral lung fields were poorly vascularized. The electrocardiogram showed atrial flutter with 2:1 atrioventricular conduction and severe right ventricular strain (fig. 4). Right heart catheterization revealed a systolic gradient of 150 mm Hg across the pulmonic valve (table 1). No additional lesions were detected.

The patient was prepared for surgery with digitalis and diuretics, and she lost 3 kg. At operation the right atrium, the right ventricle, and the pulmonary artery were extremely enlarged. Digital exploration of the interior of the right atrium revealed moderate to severe tricuspid regurgitation. On cardiopulmonary bypass the pulmonary valve was explored by way of a longitudinal arteriotomy. The valve was very stenotic, its orifice measuring 3 mm in diameter. The three fibrotic commissures were opened all the way to the relatively small annulus and the maximum opening possible, 14 mm, was achieved. The interior of the right ventricle was explored; there was concentric hypertrophy but no localized obstruction in the infundibulum. The right atrium was opened and the absence of an atrial septal defect was confirmed. The annulus and the tricuspid valve itself showed no organic pathology; repair was considered unnecessary. The heart, which had been purposefully fibrillated at the commencement of the perfusion, was defibrillated and beat in sinus rhythm.

Following the operation, the pulse remained regular for 48 hours, at which time atrial ectopic beats appeared. These increased in frequency and on the fifth postoperative day atrial flutter recurred. The patient’s recovery was very slow.
and she complained of dyspnea and palpitations which delayed ambulation. Twenty-four days after surgery, cardioversion was performed and sinus rhythm appeared after the first countershock with 120 watt-sec (fig. 5). The patient's condition improved dramatically—she was relieved of her palpitations and her recovery was now rapid. She left the hospital 7 weeks after the operation and returned to her usual housework; no cardiac drugs were recommended. Follow-up chest x-rays have shown striking diminution in the size of the heart (fig. 6). At recatheterization 14 months after surgery, a residual gradient of 35 mm Hg was demonstrated (table 1). The follow-up period is now 20 months and sinus rhythm has been constant.

**Discussion**

Since the more advanced stages of pulmonic stenosis are often accompanied by massive right atrial enlargement, with or without tricuspid insufficiency, one would expect atrial arrhythmias to be frequent late complications
of this anomaly. Yet perusal of the literature has disclosed no reports of atrial flutter occurring during the natural course of isolated pulmonic stenosis.1-9 Both our patients were found to have atrial flutter at the time of initial presentation. In case 1, incapacitation together with the signs of congestive cardiac failure was the important factor in the decision to carry out early cardioversion, even before cardiac catheterization. In retrospect, this approach may be open to criticism in view of the severe right ventricular hypertension found at the subsequent catheterization. However, the fact remains that the patient has been in sinus rhythm from the day of cardioversion and throughout right heart catheterization, open heart surgery, and a follow-up period of almost 2 years. His immediate postoperative recovery was smooth and rapid and this may be related to the absence of atrial flutter.

The approach in case 2 was different since we considered cardioversion too dangerous for a patient with such tremendous cardiomegaly. She was catheterized, prepared for surgery, and operated on while having atrial flutter. The defibrillation performed at the termination of cardiopulmonary bypass converted her heart to sinus rhythm. This continued for 4 days only and then atrial flutter reappeared. We attributed the very slow recovery partially to the hemodynamic disadvantages of the arrhythmia. This surmise is supported by the marked improvement which followed the cardioversion.

As concentric hypertrophy of all the right ventricular muscle was found in both cases, no resection of the infundibular area was performed. This procedure would only have prolonged the period on bypass, increased the trauma of the heart, and possibly hampered efficient contraction of the right ventricle in the vital first postoperative days. The tricuspid insufficiency presented more of a problem, particularly in the second patient, in whom it was severe. However, as organic pathology of the valve cusps was obviously absent, annuloplasty did not seem to be indicated as a primary procedure. Had the heart not maintained an adequate circulation in the immediate postperfusion period, we were prepared to begin the bypass again and to repair or even replace the tricuspid valve. Tricuspid insufficiency was never a problem in the postoperative period and apparently, once the commissurotomy had relieved the right ventricle of the strain of pumping against tremendous resistance, the tricuspid valve regained competence.

Both patients were restudied 14 months after surgery. Neither had signs of pulmonary insufficiency, but a systolic murmur was audible over the pulmonary area in both patients. The roentgenograms showed that the heart had shrunk in both cases. This was particularly striking in the second patient with the gross cardiomegaly. The electrocardiograms did not show significant changes. Recatheterization of the right heart revealed a minimal systolic gradient across the pulmonic valve in the first patient and a 35-mm Hg gradient in the second. The small annulus in this patient may explain the residual stenosis.

References


"Cardiac Asthma"—Pointed Remarks by MacKenzie (1911)

Sir,—I had hoped that Sir Clifford Allbutt would have explained what he meant by the term "asthma," and would have shown how the condition I had described under the term "cardiac asthma" failed to come within the limits of his definition. I have tried to comprehend his meaning, and I gather that he assumes that the form of dyspnoea which I had called "cardiac asthma" differs from that "assemblage and procession of signs and symptoms, positive and negative, recurring with fair uniformity," which he looks on as characteristic of asthma.

I have had opportunities of studying the condition during attacks in ordinary asthma and in cardiac asthma, and I could detect no dissimilarity. Thus, the patients' sensations, their aspect and attitude, the manner of breathing, the onset and offset of the attack, the response to remedies, were identical. . . .

I have seen only a few cases during an attack, and those I saw when I was engaged in general practice, and was summoned in the middle of the night because of the patient's distress. The information derived from seeing these cases has enabled me to appreciate the condition in patients suffering from these attacks whom I have not actually seen during an attack.

Sir Clifford Allbutt puts forward the suggestion that it is possible he may never have seen such a case, and I am disposed to think that this is probably the reason why he disagrees with the term "cardiac asthma." . . .

When one reflects on the rarity of these cases, and that the attacks usually occur in the night, and the fact that the teaching physician is not likely to be called out at night to see a breathless man, it is not surprising that many physicians have no experience of this and other forms of cardiac dyspnoea.—J. MACKENZIE: Correspondence: Cardiac Asthma. Brit Med J 2: 1231, 1911.
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