Isolated Tricuspid Stenosis

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SUMMARY
A case is reported of isolated tricuspid stenosis with a 5-year hemodynamic follow-up after a tricuspid commissurotomy. Fusion of the commissures and chordae tendineae of the tricuspid valve was noted at surgery. There was a history of rheumatic fever, and 5 years after surgery mild mitral stenosis had developed. Seven previously reported cases of isolated tricuspid stenosis have been considered by some authors to be congenital in origin, and, therefore, isolated tricuspid stenosis of rheumatic origin is thought to be established for the first time.

Additional Indexing Words:
Cardiac catheterization Mitral valve Tricuspid valve

Seven previously reported cases of isolated tricuspid valvular stenosis1-7 were recently reviewed by Keefe and associates,7 who thought that all of these cases were congenital in origin. Isolated tricuspid stenosis caused by rheumatic fever was considered never to have been documented.

The purpose of this report is to describe the eighth case of isolated tricuspid valvular stenosis, the fourth case to have isolated tricuspid valvuloplasty and possibly the first case with convincing evidence of rheumatic etiology.

Case History
J.C., a white female who is now 43 years old, had acute rheumatic fever at the age of 9; she had migratory polyarthritis of all the large joints, a heart murmur was heard for the first time, and she was kept in bed for 6 months. In 1965, at the age of 38, she complained of marked swelling of her legs, fatigue, and slight dyspnea on exertion.

Physical examination in 1965 revealed marked edema in both legs from the knees down. There were striking a waves in the jugular venous pulse, which increased in amplitude during inspiration. There was also presystolic pulsation of the liver, which was palpable 4 cm below the right costal margin. A grade III/V, rough, presystolic murmur at the lower left sternal border was accentuated during inspiration.

The electrocardiogram in 1965 (fig. 1) showed normal sinus rhythm with evidence of right atrial enlargement. The chest roentgenogram revealed a normal heart except for right atrial enlargement.

Right and left heart catheterization, using the Seldinger technique, was performed in 1965. The right atrial tracing revealed a prominent a wave (20 mm Hg) and a small v wave (5 mm Hg) with a slow y descent. Right ventricular and pulmonary artery pressures were normal. The mean diastolic gradient across the tricuspid valve was 6 mm Hg, with a peak gradient of 15 mm Hg during atrial systole. There was no aortic valve gradient and no mitral valve gradient either at rest or exercise.

At open heart surgery in June, 1965, simultaneous pressures from the right atrium and right ventricle demonstrated a mean gradient of 4 mm Hg and a peak diastolic gradient of 12 mm Hg across the tricuspid valve. The tricuspid orifice was less than 1 cm.2 There were fused commissures and chordae tendineae. The valve leaflets were teased apart with a Gerbode dilator, and the fused chordae tendineae with Morris chordae knives. The valve then appeared competent, with an orifice of 4.5 cm.2 Pressure tracings repeated after surgery showed no tricuspid valve gradient.

In the 5 years since surgery she has been virtually asymptomatic, except for occasional slight ankle edema.

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Physical examination in 1970 at age 43 revealed a prominent $a$ wave in the jugular venous pulse (fig. 2). The first heart sound at the apex was slightly accentuated. At the lower left sternal border no opening snap was audible during expiration; with inspiration, a definite, but soft opening snap was present (fig. 2). No opening snap was audible at the apex (fig. 2). At the lower left sternal border there was a grade I/VI early-to-midsystolic murmur and a grade II/VI high-pitched early diastolic and presystolic murmur; both the systolic and diastolic murmurs were accentuated by inspiration.

The electrocardiogram in 1970 (fig. 1, bottom) showed a vertical frontal axis with P waves suggestive of left atrial enlargement. Chest roentgenograms demonstrated slight prominence of the left atrial appendage. An echocardiogram showed mild delay in movement of the anterior leaflet of the mitral valve, compatible with mild mitral stenosis.

On cardiac catheterization repeated in 1970, with simultaneous right atrial and right ventricular pressures, there was a mean diastolic gradient of 1 mm Hg across the tricuspid valve with a peak gradient of 5 mm Hg during atrial systole, but no end-diastolic gradient. There was a mean diastolic gradient of 4 mm Hg across the mitral valve and an end-diastolic gradient of 1–2 mm Hg. The pulmonary artery and right ventricular
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pressures were normal, and there was no gradient across the aortic valve.

Discussion

Etiology

Keefe and associates\(^7\) concluded that all previously reported cases of isolated tricuspid valve stenosis were congenital in origin for the following reasons: (1) the murmur was detected at an early age, (2) no previous patient had a history suggestive of acute rheumatic fever, and (3) the surgical description of the tricuspid valve at surgery in two previous cases and at autopsy in one case did not suggest a rheumatic etiology. At surgery in one case the valve was described as funnel-shaped without identifiable commissures,\(^8\) and in another case, as a fibrous structure in which the left anterior commissure was identified as a thickened ridge.\(^9\) In the only case in which there was an autopsy the valve was an inflexible diaphragm-like structure, pearly white and avascular, with a rigid slitlike orifice and fused edges.\(^1\) No comments regarding the chordae tendineae were made in these three cases.

However, it should be pointed out that all of the previously reported cases were in the age group where rheumatic heart disease is common; five of the seven previously reported patients were between 32 and 37 years of age, with one patient 17 and one 43 years of age.

The present patient had a definite history of rheumatic fever at age 9. The surgical findings were consistent with a rheumatic etiology, that is, fused but identifiable commissures and fused chordae tendineae. In 1965, when she had a tricuspid commissurotomy, there was no evidence of mitral stenosis, but 5 years later she developed hemodynamic evidence for mitral stenosis. Because of the history of acute rheumatic fever, the surgical findings, and the late development of mitral stenosis, we believe the evidence for a rheumatic etiology of this patient’s isolated tricuspid stenosis is convincing.

Clinically significant tricuspid stenosis can be found in about 3% of patients with rheumatic heart diseases,\(^2,8\) but in previously reported patients the mitral stenosis was usually severe with no previous reports of hemodynamically insignificant mitral stenosis.

Clinical Findings

In tricuspid stenosis there is usually peripheral edema, which was very marked in our patient prior to surgery. Dyspnea on exertion was present in all previously reported cases of isolated tricuspid stenosis.\(^1-7\) There are prominent \(a\) waves in the jugular venous pulse, presystolic pulsation of the liver, and a diastolic (predominantly presystolic) murmur along the left sternal border—all accentuated by inspiration. There should be evidence of

Figure 2

(Left) Jugular venous tracing (JVT) and phonocardiogram from the lower left sternal border at 100 cycles/sec. Note the tall venous \(a\) waves. An opening snap is recorded with the first heart cycle during inspiration but not in the latter two cycles. Presystolic and early-to-mid-systolic murmurs are present. (Right) Carotid artery tracing (CAT), apexcardiogram (ACG), and phonocardiogram from the apex. No opening snap is present. Early diastolic and presystolic murmurs are present.
right atrial enlargement by electrocardiogram and chest roentgenogram. Although pulmonic stenosis and pulmonary hypertension would be a more common cause of prominent a waves in the jugular venous pulse, inspiratory accentuation of the a wave and of the presystolic murmur is found only in tricuspid stenosis.

Our case demonstrates the clinical difficulty at the bedside in distinguishing tricuspid from mild mitral stenosis when the combination is present in the same patient. Because the opening snap was inconstant, present only with inspiration, very soft in intensity, and localized in a small area at the lower left sternal border, it was felt to be most likely tricuspid in origin. The diastolic murmur was high-pitched and more easily heard with the diaphragm of the stethoscope than the bell. This fact, combined with the inspiratory accentuation, led us to believe the murmur in 1970 was primarily tricuspid in origin. It was only after seeing the left atrial enlargement on the electrocardiogram and chest roentgenogram that mitral stenosis was strongly suspected.

Catheterization

A right atrial angiogram is important in excluding a right atrial tumor in a patient who presents with clinical evidence for isolated tricuspid valve disease. Most important in establishing the diagnosis of tricuspid stenosis is the simultaneous recording of right atrial and right ventricular pressures, making sure that the zeros of the transducers and the baselines on the pressure recording are identical. In tricuspid stenosis there will be a large right atrial a wave of 12-20 mm Hg and a diastolic mean gradient of 4-8 mm Hg across the tricuspid valve. It should also be emphasized that a mean gradient across the tricuspid valve is more important in tricuspid stenosis, since an end-diastolic gradient may be absent with significant obstruction. This is a reflection of the lower filling pressures on the right side of the heart.

Surgery

Three previous patients have had an isolated tricuspid commissurotomy.5-7 The longest previous hemodynamic follow-up was 14 months;4 in that case a "persistent significant" gradient across the tricuspid valve was found, and there was "slight" elevation of pulmonary artery wedge pressure but a left heart catheterization was apparently not done and, therefore, it is not know if there was a mitral gradient. Another patient who had surgery5 had no left heart catheterization.

The four patients, including ours, who have had isolated surgical repair of the tricuspid valve have had subsequent clinical improvement.

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