Aneurysms of the interatrial septum are rare. They have been associated with complications such as embolic phenomena and atrioventricular orifice obstruction. We describe two patients, one adult and one child, with atrial septal aneurysms that were diagnosed by real-time, two-dimensional echocardiography. Atrial septal aneurysms appeared as thin, localized outpuffings of the atrial septum that protruded into the right atrium and showed marked variations in their contour and size during the cardiac cycle. They could be differentiated from other intra-atrial structures such as tumor mass, large mucouschian valve and intra-atrial haffe by their relatively low reflectance, relationship to the atrial septum, considerable alterations in their outline during the cardiac cycle and characteristic patterns during peripheral venous contrast echocardiography.

Case Reports

Case 1

Patient WL was a 68-year-old man in good health until 18 months before admission, when he noted the onset of shortness of breath while mowing his lawn. His exertional dyspnea progressively worsened and at the time of admission he could walk only 100 feet. Several weeks before admission he developed syncope while voiding. He had no other symptoms.

Physical findings included a blood pressure of 120/70 mm Hg and a regular pulse of 70 beats/min. No jugular venous distention was detected and the carotid upstroke was described as normal. The first heart sound was normal, S2 was widely split and an S3 was heard. A grade 3/6 systolic ejection murmur was audible at the base and radiated to the neck. A grade 2/6 diastolic decrescendo murmur was heard at the left sternal border. The lungs were clear. The clinical findings were consistent with the diagnosis of aortic stenosis and aortic regurgitation.

Echocardiographic Findings

M-mode echocardiography revealed a calcified aortic valve and a symmetrically hypertrophied left ventricle. The right ventricle was markedly enlarged (50 mm in end-diastole) and the interventricular septum demonstrated paradoxical anterior motion during systole. The mitral valve was normal. A band of abnormal linear echoes was also detected in the right atrium behind the prominent but structurally normal tricuspid valve and showed an undulating pattern in the cardiac cycle. Phasic differences in motion were present between the anterior and posterior limits of the echoes, and during atrial systole the anteriorly situated echoes moved toward the tricuspid orifice but did not prolapse into the right ventricular cavity (fig. 1).

Two-dimensional echocardiography was performed using a commercially available mechanical sector scanner (Picker) and confirmed the M-mode findings. In addition, parasternal and subcostal apical four-chamber views showed the presence of a thin, fragmentary linear echo in the right atrium whose convexity projected laterally to the right side. Considerable variation in the shape of the linear echo was noted during the cardiac cycle. This finding, as well as the absence of mass echoes in the right atrium, suggested the presence of a membrane-like structure rather than a solid mass. To further evaluate the nature of this lesion, an echo contrast study was performed by injecting 5 ml of 5% dextrose solution into a peripheral arm vein. Contrast echoes in the right atrium were limited by the linear echo, resulting in a filling defect (fig. 2) that showed marked variation in its contour in the cardiac cycle. It appeared to move relatively freely in the right atrium toward the region of the tricuspid valve, but did not prolapse into the right ventricle.

Cardiac catheterization revealed tight aortic stenosis, mild aortic regurgitation and evidence for an atrial septal defect. Because of the abnormal echocardiographic findings, a right atrial angiogram was performed and documented the presence of a right atrial filling defect that we initially thought represented a tumor. However, at operation no tumor was found. Instead, a thin, fenestrated, aneurysmally dilated in-
Case 2

JG was a 4-year-old white boy admitted to Strong Memorial Hospital for a Rastelli procedure and closure of a previously performed Waterston shunt (anastomosis of the ascending aorta to the right pulmonary artery). Cyanosis was noted when he was 2 days old.

Physical examination revealed an active child with a blood pressure of 90/40 mm Hg, pulse rate of 126 beats/min and regular, and respiratory rate of 32/min. He weighed 15.6 kg and was 96 cm tall. He was afebrile. Central cyanosis was noted. Neck veins were not distended. The apical impulse of the heart was diffusely palpable to the left of the sternum. The heart sounds were normal. A grade 3/6 systolic murmur was heard above the left sternal border. No gallop sounds or rubs were heard. All peripheral pulses were normal. A 12-lead ECG showed normal sinus rhythm and right ventricular hypertrophy. Cardiac catheterization demonstrated transposition of the great vessels, subpulmonic stenosis, ventricular septal defect and right-sided aortic arch.

Echocardiographic Findings

M-mode echocardiography revealed a prominent linear undulating echo behind the tricuspid valve. The great vessels were not visualized but both mitral and tricuspid valves appeared structurally normal. Real-time, two-dimensional echocardiography was performed with a wide-angle mechanical sector scanner (Advanced Technology Laboratories, Inc.). In the parasternal apical four-chamber view, a large segment of the atrial septum was seen protruding prominently into the left atrium during ventricular diastole as an undulating, curved linear echo with its convexity directed laterally. With the onset of ventricular systole, this segment of the atrial septum moved from the left atrium and bulged into the right atrial cavity, but the extent of the protrusion and the undulations were less prominent. Tricuspid and mitral valves appeared normal (fig. 3). Two transposed large vessels were identified. Multiple echoes were seen just below the posterior semilunar valve, consistent with the presence of an obstructing tissue mass in that region.

At the time of operation the findings observed during cardiac catheterization were confirmed. The pulmonary valve annulus was small, the pulmonary valve was bicuspid but not stenotic and there was dense, fibrous tissue obstructing the subpulmonic region. The Waterston anastomosis was closed. The ventricular septal defect was repaired so that the left ventricle ejected into the aorta, the connection between the left ventricle and the pulmonary artery was closed and a valved external conduit was placed between the right ventricle and pulmonary artery (Rastelli procedure). The postoperative course was complicated by cardiac tamponade that required surgical closure of a bleeding point in the right atrium on the tenth day. Subsequently, the patient developed

teratrial septum was found bulging into the right atrium (fig. 1). This was excised and the interatrial septal defect was closed with a Dacron patch. The aortic valve was replaced with a #16 Lillehei-Kaster valve. The patient was weaned from the pump successfully but developed intractable heart failure and died on the sixth postoperative day.
Figure 2. Aneurysm (AN) of the interatrial septum. The apical four-chamber view shows the atrial septal aneurysm as a thin, fragmentary, linear echo in the right atrial cavity. The basal portion of the atrial septum (AS) is intact. A peripheral i.v. injection of 5 ml of 5% dextrose in water produced contrast echoes in the right heart and outlined the atrial septal aneurysm as a filling defect in the right atrial cavity, which demonstrated marked changes in its outline during the cardiac cycle. CW = chest wall; RV = right ventricle; TA = tricuspid annulus; LA = left atrium; LV = left ventricle; VS = ventricular septum; MA = mitral annulus; I = inferior; S = superior; R = right; L = left.

Figure 3. Aneurysm of the interatrial septum. The apical four-chamber view shows prominent bulging of the atrial septum (AN) into the left atrium (LA) in ventricular diastole (upper panel). During ventricular systole (lower panel), the aneurysmal segment of the atrial septum bulged into the right atrial cavity (dotted). This patient had associated transposition of the great vessels, subpulmonic stenosis and ventricular septal defect. RV = right ventricle; TV = tricuspid valve; RA = right atrium; LV = left ventricle; MV = mitral valve; VS = ventricular septum; CW = chest wall; I = inferior; S = superior; R = right; L = left.
persistent hypotension, deteriorated progressively and
died on the sixteenth postoperative day.

Postmortem examination confirmed the surgical
findings. In addition, the patient had an atrial septal
aneurysm in the region of the fossa ovalis, bicuspid
aortic valve and a double-orifice right coronary artery.

Discussion

Aneurysms of the interatrial septum usually involve
the region of the fossa ovalis.\(^8,9\) Often, they are
asymptomatic and hemodynamically insignificant.
However, serious sequelae have been noted that re-
quired surgical resection of this lesion.\(^2\) Stagnation
of blood in large atrial septal aneurysms has resulted in
clot formation, with subsequent cerebral and pul-
monary embolic episodes.\(^2,6\) An aneurysmally dilated
atrial septum may also protrude into the mitral\(^6\) or
tricuspid orifice, resulting in obstruction simulating
atrioventricular valve stenosis. During angiocardio-
graphy, an atrial septal aneurysm may present as a fill-
ing defect in the right or left atrium that is in-
distinguishable from a tumor mass.\(^2\) Therefore,
clinically, it would be valuable if a noninvasive tech-
nique such as echocardiography could be used to make
a definitive diagnosis of this entity.

M-mode echocardiographic findings have not been
found specific in the diagnosis of this entity.\(^7,9\)
Generally, patients with atrial septal aneurysms
demonstrate linear echoes in the right atrium behind
the tricuspid valve (fig. 1). In an occasional patient in
whom the aneurysm shows phasic bulging into the left
atrium, linear echoes may be observed posterior to the
aorta in the left atrial cavity.\(^8\) It is often difficult on the
M-mode to differentiate an atrial septal aneurysm
from an atrial tumor mass, although the echoes from
an aneurysm are usually less prominent than those
from a tumor and do not show significant protrusion
into the valve orifice. Furthermore, a redundant and
large eustachian valve may be mistaken for an atrial
septal aneurysm on the M-mode.

Real-time, two-dimensional echocardiography
appears to have the potential of making a definitive
diagnosis of this entity. In our limited experience with
this technique, atrial septal aneurysms present distinc-
tive and characteristic features. They appear as
localized, thin outpouchings of the atrial septum that
regularly protrude into the right atrium because the
left atrial pressure is normally higher than that in the
right atrium during the cardiac cycle. In some patients
the direction of the bulge is reversed in some phases of
the cardiac cycle, particularly ventricular diastole,
resulting in protrusion of the aneurysm into the left
atrial cavity also. This finding appears to occur in
patients with complex congenital heart diseases, such
as transposition of the great vessels (our case 2) and
tricuspid atresia\(^11\) and is presumably related to the
phasic reversal of pressure relationship in the atra.
The prominent variations in the shape and size of the
thin, irregular echo outline produced by the aneurysm
as it undulates in the atrium and its relatively low
reflectance serve to differentiate it from an atrial
tumor mass. Furthermore, the atrial septal aneurysm
usually does not protrude significantly into the ventri-
cle during diastole.

The eustachian valve may present as a prominent
linear echocardiography in the lower portion of the right atrium,
but usually does not show prominent undulations
when viewed from the apex. Also, when the atrial sep-
tum is studied in multiple planes, it would appear to be
intact, with no evidence of outpouching. Peripheral
venous contrast echocardiography may also help in
the differentiation, because the atrial septal aneurysm
(fig. 2), unlike the eustachian valve (fig. 4), will appear
as a filling defect in the right atrium. An intra-atrial
baffle inserted during the Mustard procedure for dex-
trotransposition of the great vessels may mimic an
atrial septal aneurysm (fig. 5). However, a complete
echoangiographic examination would demonstrate
other segments of the baffle in other planes and dextro-
transposed great vessels.

Aneurysmal bulging of the atrial septum is a well-
known finding in patients with hypoplastic right-heart
syndrome with tricuspid atresia\(^6,9\) (fig. 6) and can be
suspected by M-mode echocardiography.\(^8,9\) In the
only such case studied by real-time, two-dimensional
echocardiography, the authors noticed abnormal
echoes "in the position of the atrial septum" moving
toward the right atrium during systole and toward the
left atrium during diastole.\(^9\) However, their published
illustration seems to demonstrate only a localized
thickening of the atrial septum that does not show ob-
vious bulging or bowing, and hence, no convincing
evidence for the presence of an aneurysm. In our ex-
erience, localized thickening or abnormal echoes in
the region of the atrial septum have been noted in
patients with cardiomyopathy who have no evidence of
associated atrial septal aneurysms.

Localized bulging of the atrial septum is indicative
of an aneurysm formation because it points to
localized weakening in the septal wall that results in
the outpouching. Bulging of the entire atrial septum
does not necessarily imply a totally aneurysmal atrial
septal wall. Tei et al.\(^12\) described prominent, gen-
eralized bulging of the atrial septum into the right
atrium in patients with atrial pressure overload, as in
mitral regurgitation, and the septal bulge has been
noted to disappear after mitral valve replacement,
suggesting absence of aneurysmal involvement.
Furthermore, to our knowledge, aneurysmal involve-
ment of the entire atrial septum has not been doc-
umented pathologically.

Our patients had interesting associated cardiac
lesions. Case 1 had calcific aortic stenosis and a left-
to-right shunt through the fenestrated atrial septal
aneurysm. Case 2 had transposition of the great
great vessels, ventricular septal defect, pulmonary valve
stenosis, bicuspid aortic valve and a double-orifice
FIGURE 4. Eustachian valve (EV). The apical four-chamber plane was tilted slightly inferiorly to image the tricuspid annulus (TA), the eustachian valve and the inferior vena cava (IVC). The eustachian valve appears as a prominent linear echo that does not show undulations when viewed in this plane. A peripheral arm vein injection of 5% dextrose in water produced contrast echoes that completely filled the right atrial cavity and the eustachian valve-inferior vena cava complex and no filling defects were observed. In this patient with mitral stenosis, prominent filling of the vena cava from an arm vein injection probably resulted from tricuspid regurgitation. In patients without tricuspid regurgitation, it is usually necessary to inject through a leg vein to demonstrate adequate filling of the eustachian valve-inferior vena cava complex. CW = chest wall; RV = right ventricle; RA = right atrium; LA = left atrium; MV = mitral valve; TA = tricuspid annulus; LV = left ventricle; VS = ventricular septum; I = inferior; S = superior; R = right; L = left.

FIGURE 5. Intra-atrial baffle. In this patient with dextrotransposition of the great vessels after the Mustard procedure, the intra-atrial baffle resembles an aneurysmal atrial septum when it is viewed in the apical four-chamber plane (arrows). CW = chest wall; RV = right ventricle; TA = tricuspid annulus; RA = right atrium; LA = left atrium; MV = mitral valve; LV = left ventricle; VS = ventricular septum; I = inferior; S = superior; R = right; L = left.

right coronary artery. In both cases, confirmation of the presence of atrial septal aneurysms and the associated cardiac lesions was obtained at surgery and/or at autopsy. Neither patient had tricuspid atresia or right-heart hypoplastic syndrome.

Our preliminary experience indicates that real-time, two-dimensional echocardiography is useful in identifying patients with atrial septal aneurysms.

Addendum

Since the submission of the manuscript, we have made prospective diagnosis of an atrial septal aneurysm in a patient with mitral stenosis using real-time, two-dimensional echocardiography. The echocardiographic features of the aneurysm were similar to those of case 1 and the findings were confirmed at surgery.
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