Pulmonary Regurgitation in Large Atrial Shunts without Pulmonary Hypertension

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SUMMARY Seven patients with pulmonary regurgitation (PR), normal pulmonary artery (PA) pressures and large left-to-right atrial shunts are reported. Six had secundum atrial septal defects (ASD) and one had anomalous pulmonary venous drainage. These comprised 4% of 180 patients with atrial shunts and normal PA pressures. Pulmonary regurgitation was diagnosed clinically by mid-frequency diastolic decrescendo murmurs beginning after the pulmonic component of the second heart sound, and diagnoses were confirmed by catheterization. In two patients who had serial preoperative catheterizations over 8 and 16 years, PR progressed in one and was present only on the second study in the other. All patients underwent shunt correction, at which time the pulmonic annulus and artery appeared dilated, but the pulmonic valves were normal and did not require revision. In all patients the PR murmur disappeared after shunt correction alone, and on chest X-ray both PA and overall heart size decreased. Although it is known that pulmonary regurgitation occurs with atrial septal defects and pulmonary hypertension, the present study demonstrates that it also occurs with high flow atrial shunts, in which setting it has different implications and is reversible with shunt correction alone.

ATRIAL SHUNTS are among the most common congenital heart lesions; therefore, it is important to be aware of even their rare associated findings, particularly those with management implications. Mid-diastolic murmurs in patients with atrial septal defect (ASD) are generally attributed to increased tricuspid flow, or to blood flow through the ASD itself. Early diastolic murmurs in patients with ASD generally are attributed to pulmonary regurgitation (PR) secondary to pulmonary artery (PA) hypertension. However, PR may also occur in patients with normal and abnormal PA pressures and resistance. It is essential to differentiate these two groups whose presentation, management and prognosis differ significantly. Although flow-related PR in patients with atrial shunts and normal PA pressures has been mentioned in the literature, we were able to find only three cases cited. The present study reports our experience with seven patients with PR secondary to atrial shunts who had normal PA pressures and pulmonary artery resistance (PAR), and discusses the incidence of these findings in ASD, the clinical presentation, management, and follow-up course.

Methods

The records of 180 patients 20 years and older with catheterization-proven atrial shunts, normal PA pressures (less than 35 mm Hg systolic) and normal PAR (less than 1 unit), who were managed at the Massachusetts General Hospital between 1950 and 1975 were reviewed. In each patient, history, physical examination, electrocardiogram, pre and postoperative chest X-rays (fig. 1), catheterization and angiographic data, operative notes, and follow-up course were reviewed.

The diagnosis of PR was established clinically by characteristic mid-frequency early diastolic murmurs, which were loudest at the upper left sternal border and radiated toward the lower right sternum. These murmurs increased on inspiration, began after the pulmonic component of the second heart sound, peaked or plateaued early, and tapered by mid diastole (fig. 2). Pulmonary regurgitation was confirmed at cardiac catheterization by a wide pulmonary pulse pressure with near equalization of the end-diastolic PA and the post A-wave right ventricular end-diastolic pressures. The diagnosis of PR was further confirmed by selective main PA angiography (fig. 3) with the catheter placed well above the pulmonic valve to minimize catheter-induced regurgitation. Shunt size was estimated by oximetry using superior vena caval blood for the mixed venous determination. In all patients, there was no clinical, catheterization or surgical evidence for aortic insufficiency. Patients' symptoms were classified according to New York Heart Association (NYHA) criteria.

Results

Seven patients (4%) of the overall group had PR. At the time of ASD closure, these patients ranged in age from 22 to 39 years and all were in NYHA class I. No patient had cyanosis or congestive heart failure, and none had findings suggestive of Marfan's syndrome. Physical examination in these patients was typical for ASD, showing right ventricular prominence, wide and fixed split second heart sounds with normal or mildly increased pulmonic components, and systolic ejection murmurs of grade II/VI in two patients, III/VI in four, and IV/VI in one. In addition, all patients had grade II/VI mid-frequency diastolic decrescendo murmurs, which increased on inspiration, were maximum at the second left intercostal space, and radiated toward the lower right sternal border (fig. 2); and in one patient there was also an associated left upper sternal border diastolic thrill. Two patients had early systolic ejection clicks. The electrocardiogram in all patients showed right axis deviation and incomplete right bundle branch block. All patients had increased pulmonary blood flow and right ventricular enlargement on chest X-ray. The PA was prominent in all patients and markedly dilated in four (fig. 1 left).

Cardiac catheterization findings are shown in table 1, and confirm large left-to-right atrial shunts, normal PA pressures and PAR, and near equalization of PA and right ventricular end-diastolic pressures. Patients 6 and 7 had serial cardiac catheterizations 8 and 16 years apart, respectively;
the PA end-diastolic pressures decreased from 10 to 3 mm Hg in patient 6 and from 20 to 8 mm Hg in patient 7. In three patients, selective main PA angiography demonstrated the dilated PA and PA anulus and the presence of PR evidenced by opacification of the right ventricle (fig. 3).

At the time of surgery, six patients had large secundum ASDs which were closed by patches in five and by sutures in one. The seventh patient had partial anomalous pulmonary venous return from the right lung to the right atrium, and a patent foramen ovale, and had patch redirection of the anomalous pulmonary veins to the left atrium and closure of the foramen ovale with sutures. In all patients, the PA and pulmonic anulus were dilated, but the pulmonic valves were anatomically normal and surgical revision was not warranted. At discharge, the murmur of PR was no longer audible in any patients, and continued to be absent over a follow-up interval ranging from 1 to 13 years in the six patients in whom follow-up was available. Repeat chest X-rays in all patients showed decrease in both overall heart size and the size of the PA segment, although no PA returned entirely to normal size (fig. 1 right).

Discussion

With the large number of ASD patients presently undergoing cardiac catheterization and surgery, it is important to be aware of any unusual associated findings, their significance, and their appropriate management. Pulmonary regurgitation in the absence of elevated PA pressures and PAR is rare, being present in seven (4%) of 180 patients with atrial shunts and normal PA pressures, and in only three additional patients reported in the literature.\(^6\)

While it is clear from the loss of PR following shunt correction that PR was in part flow related, it is unlikely that pulmonary blood flow alone entirely explains PR, because most ASDs with comparable shunts do not develop PR. It is also unlikely that PA dilatation alone causes PR, because most ASD patients with comparable PA dilatation also do not usually have PR.\(^6\) Furthermore, not all patients in the present study had the marked degree of PA dilatation illustrated in figure 1. It is possible that dilatation of the PA anulus alone accounts for PR in some of these patients. Such dilatation is not well seen on chest X-ray but can be detected by selective PA angiography.

It is possible that coincident connective tissue disorder of the main PA or anulus permits exaggerated dilatation and PR in response to high pulmonary blood flow. Pulmonary regurgitation has been described in patients with idiopathic dilatation of the PA presumably on this basis.\(^6\) Autopsy examination or PA biopsy would be helpful in determining the etiology for PR in these patients. In our study, as well as in those patients mentioned in the literature, histologic data...
are not available. Flow-related PR is an unusual finding in children. As demonstrated in our patients who had serial cardiac catheterization, PR is acquired and progressive if the underlying shunt lesion is not repaired.

The clinical significance of PR in patients with atrial shunts concerns differentiating those with normal PA pressures and resistance from those with elevated pressures. Patients with PA hypertension and elevated PAR typically present with signs and symptoms related to obstructive pulmonary arteriolar disease, in contrast to those with flow-related PR who are minimally symptomatic. Furthermore, in the latter patients, cardiac catheterization and surgery are indicated, in contrast to the former group in whom both offer little and involve significant risk.

In patients with flow-related PR, selective PA angiography should be performed to rule out intrinsic pulmonary valve disease which might also cause PR. Because it may be difficult to clinically differentiate aortic and pulmonary regurgitation, it is of obvious importance to rule out the former; when the diagnosis is uncertain an aortogram is also warranted. In the present study, ASD correction was performed without complications related to PR. No patient required revision of the pulmonary valve, and in all, PR disappeared following shunt correction alone. Early shunt correction in patients with normal PA pressures and PAR is indicated. We have shown that persisting atrial shunts may cause progressive PR which, although well tolerated in the presence of good myocardial reserve, may be poorly tolerated with increasing ventricular dysfunction or with loss of normal sinus rhythm.

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References

Pulmonary regurgitation in large atrial shunts without pulmonary hypertension.
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