

Brief Report

A patient with aneurysms of both aortic coronary sinuses of Valsalva obstructing both ventricular outflow tracts

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Abstract Congenital unruptured aneurysms affecting both the right and left sinuses of Valsalva are extremely rare. To the best of our knowledge, there has been only one previously documented case. We report here a patient presenting with congestive cardiac failure who also had unruptured aneurysms of both the right and left sinuses of Valsalva. The aneurysms produced obstruction of the outflow tracts of both ventricles, burrowing as they did into the muscular ventricular septum.

Keywords: Intracardiac cystic mass; echocardiography; congestive cardiac failure

ANEURYSMS OF THE SINUS OF VALSALVA ACCOUNT for only 1% of congenital cardiac anomalies. Of these aneurysms, seven-tenths arise from the right coronary aortic sinus of Valsalva, one-quarter from the noncoronary aortic sinus, and less than one-twentieth from the left coronary aortic sinus¹. An unruptured aneurysm of the left sinus of Valsalva, which extends into the left ventricular outflow tract, is very rare. When present, the aneurysms usually present subsequent to their rupture into a cardiac chamber. Symptomatic patients with unruptured aneurysms of both left and right aortic sinuses are extremely rare, with to the best of our knowledge only 1 such case being previously reported in the absence of obstruction of the ventricular outflow tracts.² Another case has been reported in the setting of an obstructed left ventricular outflow tract.³ We report here a patient with congestive cardiac failure in whom unruptured sinuses involving both right and left aortic coronary sinuses burrowed into the muscular ventricular

septum, producing obstruction of the outflow tracts from both ventricles.

Case report

A boy of 8 years presented with a history of cough and exertional breathlessness of two months duration. The breathlessness, over the preceding 2 weeks, had progressed to orthopnoea and on admission he was in class IV in the system of grading produced by the New York Heart Association. On examination, he was very sick, with tachycardia, tachypnoea, pallor, and pitting oedema of both feet. His pulse, at 120 beats per minute, was regular and bounding. Blood pressure was measured at 120 over 50 millimeters of mercury. The jugular venous pressure was elevated. His precordium was hyperdynamic, with the apical impulse felt in the left sixth intercostal space just lateral to the midclavicular line. Both parasternal and suprasternal pulsations were seen, with a parasternal heave graded at 1. The first heart sound was normal, the second heart sound was normally split, and the pulmonary component was loud. A third heart sound was present, with a pansystolic murmur graded at III from VI heard in the mitral area radiating to axilla. An early diastolic murmur, also graded at III from VI, was heard in the left third

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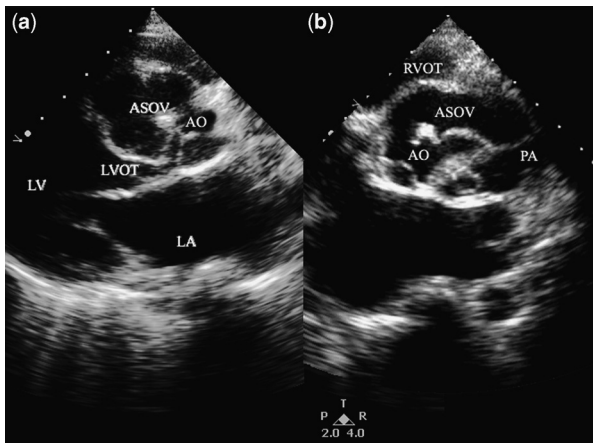


Figure 1.

The parasternal long axis view (a) shows aneurysms of both aortic coronary sinuses of Valsalva which obstruct the left ventricular outflow tract. The parasternal short axis view (b) shows the aneurysm of the right sinus protruding into the right ventricular outflow tract. Abbreviations : LA- Left atrium; LV- Left ventricle; AO- Aorta; LVOT- Left ventricular outflow tract; RVOT- Right ventricular outflow tract; ASOV- Aneurysm of sinus of Valsalva; IVS- muscular ventricular septum; MR- Mitral regurgitation; AR- Aortic regurgitation

intercostal space. Abdominal examination revealed tender hepatomegaly.

Investigations showed microcytic hypochromic anemia, with counts of leukocytes within normal limits, and sterile blood cultures. The chest radiograph showed cardiomegaly, with left ventricular apical and biatrial enlargement. The electrocardiogram showed sinus tachycardia, with left axis deviation with volume overload of left ventricle, left atrial enlargement, and right ventricular hypertrophy.

Transthoracic echocardiography in the parasternal long axis view showed thin-walled saccular lesions extending from the right and left coronary aortic sinuses and prolapsing into the left ventricular outflow tract, with obstruction of flow (Fig. 1a). The parasternal short axis view showed the aneurysms also to protrude into the right ventricular outflow tract (Fig. 1b), while the apical four chamber view showed the aneurysm from the right sinus to burrow into the muscular ventricular septum (Fig. 2a). The apical five chamber view showed the aneurysm from the left coronary aortic sinus of Valsalva to be saccular, resembling cystic masses of hydatid cysts (Fig. 2b). The leaflets arising from both the right and left coronary aortic sinuses were prolapsing, producing moderate-to-severe aortic regurgitation. The leaflets of the mitral valve were thickened and non-coapting, with colour Doppler showing moderately severe mitral regurgitation. Left ventricular function, however, was normal.

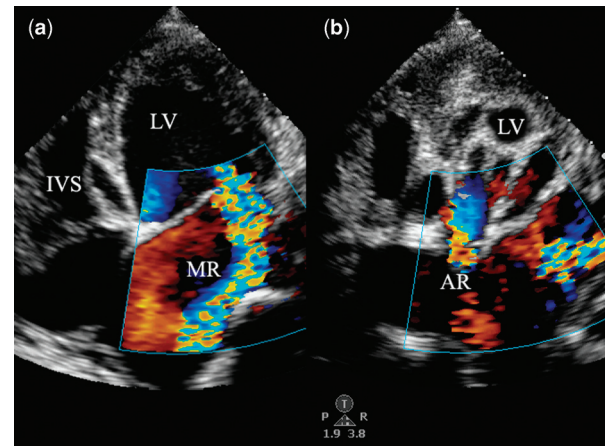


Figure 2.

The apical 4-chamber view (a) shows dilation of the left ventricle and left atrium, with the aneurysm of the right sinus of Valsalva burrowing into the ventricular septum. Note the moderately severe mitral regurgitation. The apical five chamber view (b) shows the aneurysmally dilated left sinus of Valsalva prolapsing into left ventricle, producing saccular protrusions and aortic regurgitation. Abbreviations as for Figure 1.

At emergent surgery, the pulmonary trunk, left atrium, and left ventricle, were all found to be enlarged. The large aneurysm of the left coronary aortic sinus of Valsalva extended into the left ventricle, while the unruptured aneurysm of the right sinus of Valsalva extended into the muscular septum. The leaflets of the aortic valve arising from both these sinuses were thickened, retracted, and not coapting. Surgery was carried out using cardiopulmonary bypass with moderate systemic hypothermia and cold blood cardioplegia. The left ventricular aspect of the aneurysm of the left sinus of Valsalva was closed using pledgetted sutures. The aortic opening of the aneurysm of the right sinus of Valsalva was closed with a patch. Both aortic leaflets were augmented with tanned pericardium, and an annuloplasty was performed on the mitral valve. The patient was weaned easily from cardiopulmonary bypass, and the postoperative course was uneventful. On follow-up after six months, the child is doing well, with moderate aortic regurgitation, mild mitral regurgitation, but normal left ventricular function.

Discussion

Aneurysms of the sinus of Valsalva are thought to be due to discontinuity between the aortic tunica media and the hinges of the aortic valvar leaflets.⁴ Most aneurysms arise from the right and non-coronary aortic sinuses, with less than one-twentieth arising from the left coronary aortic sinus.⁵ They typically occur above the hinge of the valvar leaflets,

and may extend in any direction. The aneurysms involving the right or noncoronary sinuses commonly involve the right atrium and the right ventricular outflow tract. Several cases have been reported where an aneurysm arising from the right coronary aortic sinus has extended into the muscular ventricular septum.^{6,7} Saccular extension into the left ventricular outflow tract, in contrast, is very uncommon. To our knowledge, only two such cases have been reported, both arising from the right coronary aortic sinus.^{8,9} Another case has been reported causing obstruction of the left ventricular outflow tract. Again to the best of our knowledge, however, ours is the first case to be reported with aneurysms of both coronary aortic sinuses extending into the outflow tracts of both ventricles, causing obstruction and also burrowing into ventricular septum. A recent report has collected together over 50 cases of aneurysmal sinuses of Valsalva, but has used a cumbersome alphanumeric system for classification.¹⁰ It is much better, as in our patient, and in view of the rarity of these lesions, simply to describe the findings.

References

1. Wells T, Byrd B, Neirste D, Fleurelus C. Sinus of Valsalva aneurysm with rupture into the interventricular septum and left ventricular cavity. *Circulation* 1999; 100: 1843–1844.
2. Zannis K, Tzvetkov B, Deux JF, Kirsch EW. Unruptured congenital aneurysms of the right and left sinuses of Valsalva. *Eur Heart J* 2007; 28: 1565.
3. Pepper C, Munsch C, Sivananthan UM, Pye M. Unruptured aneurysm of the left sinus of Valsalva extending into the left ventricular outflow tract: presentation and imaging. *Heart* 1998; 80: 190–193.
4. Edwards JE, Burchell HB. The pathologic anatomy of deficiencies between the aortic root and the heart including aortic sinus aneurysms. *Thorax* 1957; 12: 125–139.
5. Dev V, Goswami KC, Shrivastava S, Bahl VK, Saxena A. Echocardiographic diagnosis of aneurysm of the sinus of Valsalva. *Am Heart J* 1993; 126: 930–936.
6. Ahmad RA, Sturman S, Watson RD. Unruptured aneurysm of the sinus of Valsalva with isolated heart block: Echocardiographic diagnosis and successful surgical repair. *Br Heart J* 1989; 61: 375–377.
7. Dev V, Shrivastava S. Echocardiographic diagnosis of unruptured aneurysm of the sinus of Valsalva dissecting into the ventricular septum. *Am J Cardiol* 1990; 66: 502–503.
8. Mok CK, Cheung KL, Wang RY. Unruptured right coronary sinus to left ventricle aneurysm diagnosed by cross sectional echocardiography. *Br Heart J* 1985; 53: 226–229.
9. Heydorn WH, Nelson WP, Fitterer JD, Floyd GD, Strevey TE. Congenital aneurysm of the sinus of Valsalva protruding into the left ventricle. Review of diagnosis and treatment of the unruptured aneurysm. *J Thorac Cardiovasc Surg* 1976; 71: 839–845.
10. Vural KM, Sener E, Tasdemir O, Bayazit K. Approach to sinus of Valsalva aneurysms: a review of 53 cases. *Eur J Cardiothorac Surgery* 2001; 20: 71–76.