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CASE REPORT
Cardiovascular imaging

Aortic dissection extending into the interventricular septum following redo aortic valve replacement surgery in a patient with Takayasu's arteritis: a rare case report

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Background Takayasu's arteritis is an infrequent manifestation of vasculitis affecting the aorta and its primary branches with numerous symptoms. This report details a rare case wherein a patient developed interventricular septal dissection following aortic valve replacement.

Case summary A middle-aged woman diagnosed with Takayasu's arteritis previously underwent aortic valve replacement with a mechanical valve owing to severe aortic regurgitation. Subsequently, she received a redo aortic valve replacement following an episode of prosthetic valve infective endocarditis with paravalvular leak. Heart failure symptoms emerged during follow-up, revealing aortic root dissection extending into the interventricular septum, causing significant prosthetic valve movement. A Trido Bentall operation and interventricular septum repair were performed, and the patient recovered smoothly.

Discussion Interventricular dissection, although uncommon, may be due to factors such as infection, myocardial infarction, congenital anomalies, trauma, or post-surgical shear stress. Timely diagnosis is imperative to prevent life-threatening complications; surgery remains the primary treatment. The present case report describes a rare presentation that was successfully managed through a Bentall operation and underscores the necessity of prompt intervention in treating this condition.

Keywords Aortic dissection • Takayasu's arteritis • Interventricular septal dissection • Case report

ESC curriculum 2.2 Echocardiography • 7.5 Cardiac surgery • 9.1 Aortic disease

Learning points

- Timely diagnosis of interventricular dissection, a life-threatening condition is important.
- Surgical intervention is the standard treatment for interventricular dissection in current era.

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Introduction

Takayasu's arteritis (TA) is a rare form of vasculitis primarily affecting the aorta and its major branches. Although the precise pathogenesis of TA is unclear, a cell-mediated inflammatory process within the affected vessels may be responsible for the condition. This inflammation induces vessel narrowing, occlusion, and dilation, causing numerous symptoms.

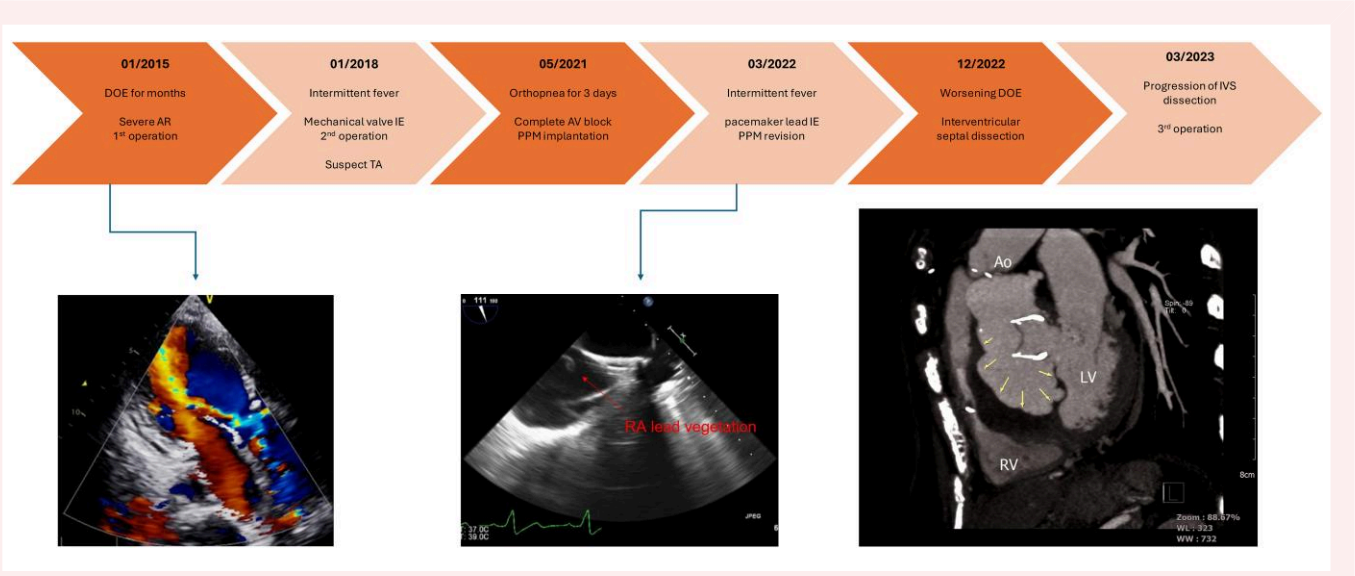
Because its underlying pathology is unknown, no laboratory diagnostic test exists for TA. Although heightened levels of inflammatory markers can aid supplementary support, normal levels cannot be used to rule out TA. Imaging studies such as magnetic resonance imaging (MRI) or computed tomography angiography (CTA) are crucial to diagnosing TA and assessing its progression.

Large vessel biopsy and histology are rarely practical in diagnosing TA. Occasionally, aortic tissue may be procured during cardiovascular surgeries, revealing signs of active inflammation comprising lymphocyte infiltration, elastic lamina destruction, muscular media destruction, aortic scarring, and intimal proliferation.

In 1990, the American College of Rheumatology (ACR) classification criteria were formulated to aid in distinguishing TA from other forms of arteritis, which was then revised in 2022.[1] The criteria enable a diagnosis of TA with high sensitivity (93.8%) and specificity (99.2%).

This report describes an unusual case with TA who developed aortic root dissection extending into the ventricular septum following aortic valve replacement.

Summary figure



Case presentation

A 39-year-old female teacher with a history of mild aortic regurgitation (AR) experienced a worsening of dyspnoea for several months, despite receiving treatment with diuretics and ARB. She had no hypertension nor family history of cardiovascular disease. Examination revealed grade III/VI to-and-fro murmur at the left lower sternal border. Echocardiography revealed left ventricular (LV) dilation, preserved LV ejection fraction, and progression to severe AR without an apparent cause. The patient subsequently underwent aortic valve replacement with a mechanical valve due to symptomatic severe AR.

She experienced intermittent fever for 1 month at approximately 3 years after the operation, at which time echocardiography showed moderate paravalvular AR and partial dis-adhesion of the mechanical aortic valve with a fluttering mass. The patient subsequently received redo aortic valve replacement with a bioprosthetic valve (Trifecta 21 mm) due to infective endocarditis. Pathologic results revealed suppurative inflammation. However, both blood and surgical specimen cultures returned negative results. A thickened aortic root was noted during operation, and subsequent computed tomography (CT), MRI, and gallium scan results further supported the diagnosis of aortitis. Takayasu's arteritis was suspected by meeting 2022 ACR criteria: female, reduced upper extremity pulsation, involvement of two artery territories, and which got 5 points.

During the index admission, she received high-dose prednisolone for TA, which was gradually tapered over the following months while azathioprine was introduced. Recurrent episodes of intermittent fever in the subsequent years, indicative of TA flares, were effectively managed with intravenous tocilizumab.

Three years after the second operation, she underwent pacemaker implantation due to complete atrioventricular block. The pacemaker was subsequently revised 1 year after the initial implantation due to endocarditis with vegetation on the atrial lead.

Five years after the second operation, the patient developed worsening exertional dyspnoea. A follow-up CTA for TA indicated an aortic root pseudoaneurysm extending into the interventricular septum. The interventricular septal dissection had progressed incrementally over a 3-month period, approaching the insertion site of the pacemaker lead in the right ventricle, as revealed by both echocardiography (*Figures 1* and *2*; supplementary material video) and CT (*Figure 3*).

The patient subsequently underwent surgery for interventricular septal dissection and pseudoaneurysms, moderate aortic bioprosthetic valve regurgitation/stenosis, and severe mitral regurgitation due to chamber dilation. Surgery consisted of Trido-median sternotomy with the ascending aorta and direct bicaval cannulation.

Inspection of the aortic root revealed a peri-annular pseudoaneurysms arising from the right coronary sinus of Valsalva resulting in an interventricular septum dissection (*Figure 4*), a rocking aortic bioprosthetic valve (Trifecta 21 mm) attaching to the intima of the dissected interventricular septum, and a previous peri-annular bovine

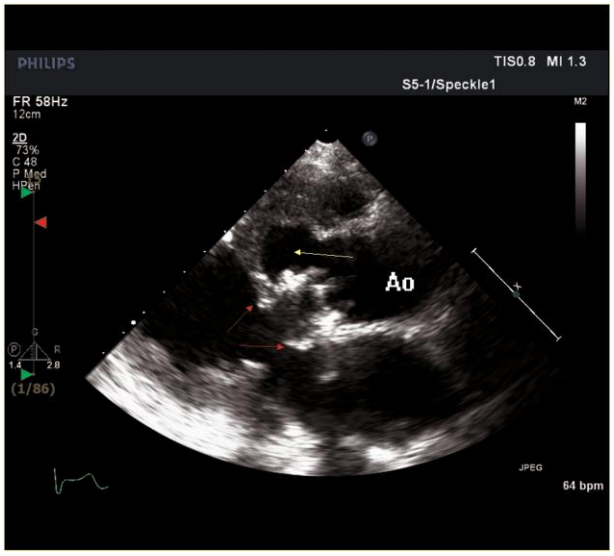


Figure 1 Parasternal long axis view of an echocardiogram performed during outpatient follow-up revealing a mispositioned bioprosthetic valve (the short arrows) and aortic root dissection extending into the interventricular septum (the long arrow). Ao, aorta.

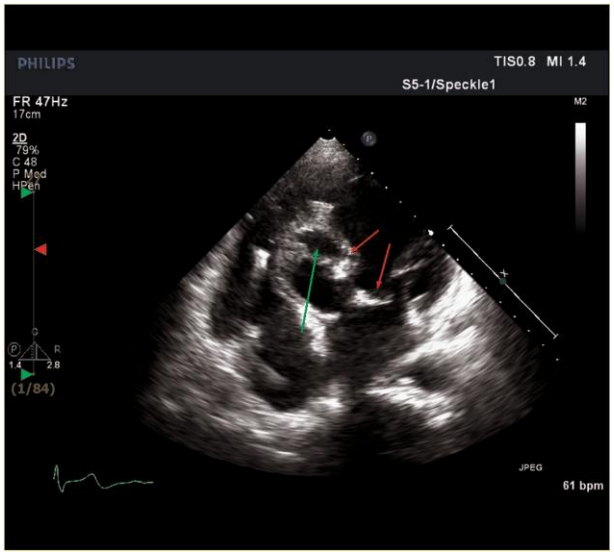


Figure 2 Apical five-chamber view of the echocardiogram depicting a dissected interventricular septum (the long arrow) and a mispositioned prosthetic aortic valve (the short arrows).

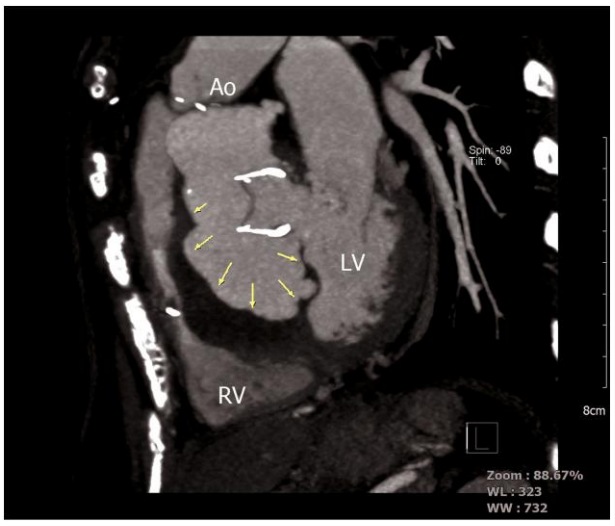


Figure 3 Chest computed tomography revealing interventricular dissection measuring 3×4 cm^2.

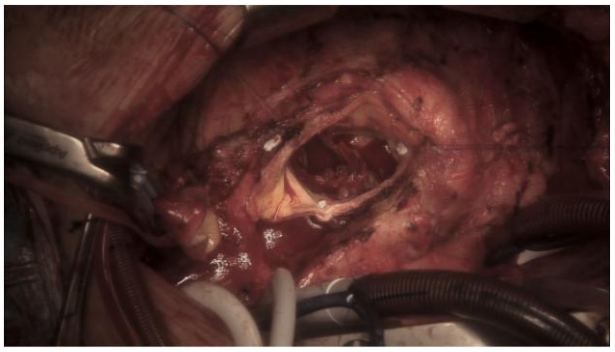


Figure 4 Aortic bioprosthetic valve was detached surgically. The surgical view with the cranial part in the left and the caudal part in the right showed left ventricular outflow tract and interventricular septal dissection.

pericardium patch over the sinus of Valsalva. Also observed was a sclerotic change of the leaflet without vegetation or abscesses.

During surgery, the coronary arteries were first separated from the sinus of Valsalva with 20 mm buttons. Second, the cavity of the pseudoaneurysm sac was obliterated using 2–0 braided polyester interrupted pledgeted mattress sutures and Tisseel, a fibrin glue (Baxter). Third, proximal anastomosis was performed by suturing the flanged composite graft (26 mm Dacron vascular graft and 23 mm St. Jude mechanical valve) to the myocardium of the LV outflow tract and the aortomitral curtain using 2–0 braided polyester interrupted pledgeted mattress sutures. Fourth, the suture line was reinforced using 4–0 polypropylene continuous sutures between the outside-folded cuff and the cut edge of the supra-annular aortic wall. Finally, the coronary arteries were anastomosed to the composite graft using 5–0 polypropylene continuous sutures and polytetrafluoroethylene felt.

Distal anastomosis was completed by suturing the flanged composite graft to the distal ascending aorta. Mitral annulus dilatation with intact leaflet was observed intraoperatively.

Downsized mitral annuloplasty was performed with a 30 mm Memo-3D ring using a superior-extended trans-septal approach. The incised left atrium roof and the right atrium atriotomy were repaired with bovine pericardium.

Three-month post-operative CTA revealed a normal position of the prosthetic valve without pseudoaneurysms or fistulae. Echocardiography

Table 1 Case reports of interventricular septal dissection

Authors	Presentation	Diagnostic modality	Diagnosis	Treatment and outcomes
Dong *et al.*[8]	57-year-old man presenting with exertional dyspnoea approximately 1 year after falling from a height with chest pain	Transthoracic echocardiography	Right sinus of Valsalva rupture with dissection into the interventricular septum and formation of a false cavity	Surgical repair
Park *et al.*[10]	50-year-old woman with a history of aortic replacement and permanent pacemaker implantation 17 years prior due to severe aortic regurgitation and complete AV block presenting with dyspnoea on exertion	Transthoracic echocardiography, magnetic resonance imaging	Peri-prosthetic leak into the interventricular septum with aneurysmal change	Discharged 18 days after surgical repair without complication
Zhao *et al.*[14]	49-year-old woman presenting with chest compression and dyspnoea	Transthoracic echocardiography, trans-oesophageal echocardiography	Dissection of the aortic root associated with perforation of the left coronary sinus and prolapse of the exfoliated endocardium into the left ventricular outflow tract	Discharged in good condition following a Cabrol procedure with aortic valve replacement plus total replacement of the aortic root and ascending aorta with a conduit and end-to-end anastomosis between the conduit and the left and right coronary ostia
Yoo *et al.*[5]	26-year-old man experiencing cardiac arrest/ventricular fibrillation while playing soccer	Transthoracic echocardiography, trans-oesophageal echocardiography, chest computed tomography	Rupture of the right sinus of Valsalva with dissection into the interventricular septum	Discharged with rehabilitation 1 month after primary closure and subsequent aortic valve replacement and permanent pacemaker implantation
Jang *et al.*[6]	58-year-old man presenting with acute ischaemic stroke	Transthoracic echocardiography, cardiac computed tomography	Perforation of the left sinus of Valsalva and a dissection involving the interventricular septum, causing communication between the left ventricle and aorta	Conservative management due to patient refusal of surgical intervention
Kumar *et al.*[3]	21-year-old man presenting with worsening dyspnoea on exertion	Transthoracic echocardiography, left ventricle root and coronary angiography	Right sinus of Valsalva aneurysm that dissected through the intraventricular septum and egressed into the left ventricle	Discharged without event 8 days following surgical repair of the right sinus of Valsalva
Fennich *et al.*[4]	23-year-old man presenting with syncope due to complete atrioventricular block	Transthoracic echocardiography, cardiac computed tomography angiography, magnetic resonance imaging	Perforation of the right sinus of Valsalva dissecting into the muscular interventricular septum	Discharged without event following surgical repair of the perforated sinus of Valsalva and conservative management of the aortic valve using a Dacron patch; transthoracic echocardiography revealed no residual flow across the repaired defect
Ghosh *et al.*[2]	52-year-old man presenting with dyspnoea, palpitation, and wide pulse pressure	Transthoracic echocardiography, computed tomography angiography	Right sinus of Valsalva aneurysm with interventricular dissection and rupture into left ventricle	Discharged without event 1 week following aortic valve replacement and repair of right sinus of Valsalva aneurysm
Ni *et al.*[12]	58-year-old man presenting with dyspnoea and dizziness	Transthoracic echocardiography, cardiac computed tomography	Left sinus of Valsalva with aneurysm and extension into ventricular septum; aortic regurgitation	Bio-Bentall procedure repairing the intimal tear with a bovine pericardial patch
Wu *et al.*[13]	36-year-old man presenting with chest distress and shortness of breath lasting than 2 months	Transthoracic echocardiography, aortic computed tomography angiography	Right sinus of Valsalva rupture with ventricular septum dissection	Successful surgical repair

revealed a properly functioning aortic prosthetic valve (supplementary material video). The patient had an uneventful, symptom-free daily living 1 year after the operation.

Discussion

Interventricular septal dissection is a rare clinical condition accompanied by several symptoms, such as dyspnoea,[2] chest pain,[3] acute heart failure, conduction disturbance,[4,5] stroke,[6] and cardiac arrest.[5] Transthoracic echocardiography frequently reveal a cyst-like mass within the interventricular septum, with potential shunts from rupture sites and communication with the cardiac chambers.[7] In the present case, CT scans may have underestimated the severity owing to the static nature of CT images, which cannot capture the dynamic interactions and shear forces between the valves and the septum; these interactions are only discernible through motion echocardiography.

Interventricular septal dissection is frequently caused by the Valsalva sinus rupture.[7] Additional causes comprise congenital abnormalities, infective endocarditis, deceleration trauma, and cardiac surgery.[2,3,7,8] Few reports have described interventricular septal dissection following aortic valve repair, which have been linked to post-operative shear stress[9,10] and may partially explain the presentation in our case.

Vascular inflammation caused by TA may have partially contributed to the vascular complications in the patient. Several case reports have documented instances of aortic dissection and even myocardial dissection in individuals with TA, suggesting a potential causative relationship.[11,12] However, the English-language literature on TA has not described interventricular septal dissection following aortic valve replacement. Similarly, previous reports have presented several cases of interventricular septal dissection in patients with Behcet's disease, a condition also characterized by chronic systemic vascular inflammation.[6,13,14] In many of these cases, surgical interventions revealed a pathology characterized by inflammatory cell infiltration and fibrinoid necrosis, mirroring the findings in the present case.

Several case reports (Table 1) have documented treatment of a ruptured Valsalva sinus using a transcatheter approach,[15] although none has involved interventricular complications, as in the present case. Surgical repair is the standard treatment for interventricular septal dissection,[10] although conservative management was utilized in deference to patient preference in at least one case.[6] In the present case, the patient underwent a Trido Bentall operation and recovered smoothly.

Conclusion

Interventricular septal dissection is a rare and potentially life-threatening condition, rendering timely diagnosis from clinical suspicion and image examinations critical. Interventricular septal dissection may be associated with inflammatory arteritis, but any suggestion of causation requires further investigation. Surgical interventions such as those in the present case are the standard treatment for this condition.

Lead author biography

Chan-Han Hu is a medical doctor, graduated from National Cheng Kung University in Taiwan. He is currently a cardiology resident and is passionate about echocardiography and cardiac imaging.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Consent: The authors confirm that written consent for submission and publication of this case report including the images and associated text has been obtained from the patient in line with the COPE guidelines.

Conflict of interest: None declared.

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Data availability

Data will be made available on request due to privacy/ethical restrictions.

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