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CASE REPORT
Cardiac imaging

A case report showing unusual atrial communication with severe regurgitation of multiple valves and pulmonary aneurysm: double atrial septum with persistent interatrial space

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Background We discuss an unusual association: double atrial septum, pulmonary artery aneurysm, and severe regurgitation of multiple valves.

Case summary A 70-year-old man was admitted into the hospital because of progressive dyspnoea. Physical examination showed a blood pressure of 132/70 mmHg, a systolic murmur on the right upper sternal border, another systolic murmur at the apex, and a diastolic murmur at the lower left sternal border. Electrocardiogram revealed atrial fibrillation and complete left bundle branch block. Transthoracic echocardiography showed mitral prolapse, severe mitral, aortic, and pulmonary regurgitation, a 60 mm diameter pulmonary artery aneurysm, mild to moderate tricuspid regurgitation, and moderate pulmonary hypertension. Transoesophageal echocardiography also showed an unusual atrial communication consisting of a double atrial septum with a mid-line chamber between both atria. A cardiac magnetic resonance scan was performed and confirmed echocardiography findings and Q_P:Q_S ratio = 1.3.

Discussion In our knowledge, this is the first case report with this association. We present the main clinical features of the double atrial septum with persistent interatrial space, its echocardiography anatomy, differential diagnosis, and embryology.

Keywords Case report • Double atrial septum • Persistent interatrial space • Severe regurgitation • Multiple valves and pulmonary aneurysm

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Learning points

- We discuss a rare association: this is the first ever case report consisting of double atrial septum, severe regurgitation of multiple valves, and pulmonary artery aneurysm. Transoesophageal echocardiography was essential in the diagnosis workup. Cardiac magnetic resonance was also useful to quantify the interatrial shunt.
- The right heart catheterization (RHC) was essential to diagnose a severe group 2 pulmonary artery hypertension and to decide surgical therapy.
- Even though our patient did not suffer any embolism, it is compulsory to be aware of this cardiac anomaly, because it is associated with an increased thrombo-embolic risk. It is for that reason that we must rule it out in the cryptogenic ictus diagnosis workup.

Introduction

Double atrial septum is a very unusual cardiac anomaly that features a double atrium septum (septum primum and another septal structure) and a persistent space between them.[1,2] Both fenestrated structures run parallel and, as a result, it can be seen an interatrial septal defect between them. The accessory septal structure is a persistent left venous valve, which is a remnant of the embryological sinus venosus.[2] To the best of our knowledge, this is the first case showing this unusual kind of interatrial communication, mitral valve prolapse, severe mitral, aortic and pulmonary regurgitation, and pulmonary artery aneurysm.

Timeline

Investigation	Findings
Clinical presentation	The patient underwent pulmonary valvulotomy 50 years ago. He complains of exertional dyspnoea
Electrocardiogram	It shown new atrial fibrillation at 120 beats/min and left bundle branch block
Echocardiogram	It shown severe multiple valve regurgitation (mitral, aortic, and pulmonary), pulmonary artery aneurysm double atrial septum, and interatrial communication
Cardiac magnetic resonance	It shown ulmonary artery aneurysm, double atrial septum, and interatrial communication
Right heart catheterization and coronariography	It shown severe second group pulmonary hypertension and normal coronary arteries
Cardiac surgery	The patient underwent triple valve replacement, closure of double septum, and resection of pulmonary aneurysm

Case presentation

A 70-year-old man with a past medical history of pulmonary valve stenosis for which he underwent surgical valvulotomy when he was 20-year-old. He came into the clinic complaining of worsening exertional dyspnoea in the last 6 months. There was no fever. We were unaware of any co-existing cardiac conditions at that time. The previous surgery took place in another hospital, and we did not have access to surgical data or any previous echocardiograms. We could look into a report showing an 82 mmHg mean pressure gradient through the pulmonary valve, before the valvulotomy. Other cardiac conditions were unknown at that moment. Physical exam revealed blood pressure 130/70 mmHg, irregular heartbeats, a systolic murmur on the right upper sternal border, and both diastolic murmurs at the lower left sternal border and systolic murmur at the apex. The respiratory examination revealed diffuse bilateral crackles. Electrocardiogram showed atrial fibrillation and complete left bundle branch block. The chest X-ray showed pulmonary congestion. Blood tests were normal, only pro-blood natriuretic peptide (BNP) was increased (3420 ng/dL) (normal range 0–900).

Transthoracic echocardiogram (TTE) revealed a non-dilated left ventricle with normal ejection fraction at 65%. The ventricular walls thickness was normal, and there were no regional wall motion abnormalities apart from paradoxical septal motion, probably related to the left bundle branch block. The right cardiac cavities were slightly dilated and the right ventricle contractility was good (TAPSE 18 mm). Mitral valve prolapse (P2 and P3 segments of the posterior leaflet) and severe mitral regurgitation (PISA diameter 10 mm, vena contracta 8 mm, regurgitant volume 62 cc) were also noted on the echocardiogram. The aortic valve was trileaflet in terms of its structure with severe regurgitation (vena contracta (VC) 8.5 mm, regurgitant volume 70 cc) but no vegetation or mass was found. The regurgitation was central and its jet broad. The right coronary leaflet was thickened and hypoplasic. The aortic size was normal (sinus aorta 34 mm, ascending aorta 35 mm, descending aorta 26 mm). We also found a 60 mm diameter pulmonary artery aneurysm and severe pulmonary regurgitation (regurgitation jet area /RAA 53% and vena contracta 7.2 mm) (*Figures 1* and *2*). The pulmonary transvalvular gradient was 20 mmHg. The pulmonary valve was dysplastic. Finally, we found mild to moderate tricuspid regurgitation and a 40 mmHg systolic gradient between the right ventricle and right atrium was calculated. We explored the interatrial septum in standard parasternal and subcostal views, finding a thickened interatrial septum but without any anatomical or haemodynamic abnormality.

We continued the patient's workup with transoesophageal echocardiogram that confirmed the presence of severe regurgitation of multiple valves and the pulmonary artery aneurysm. Moreover, we found a double atrial septum with a midline chamber between both atria (*Figure 3*) We did not calculate the pulmonic to systemic shunt ratio because the presence of severe pulmonary and aortic regurgitations could artefact the measure. Moreover, we did not perform a

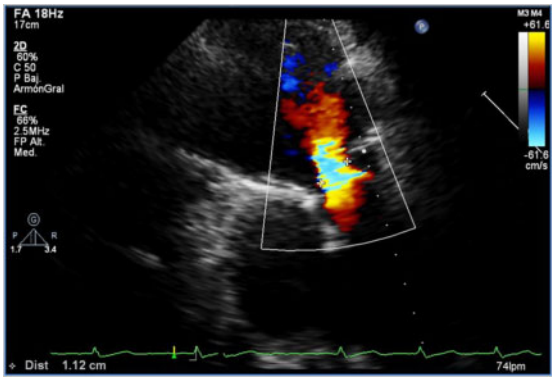


Figure 1 Transthoracic echocardiogram severe pulmonary regurgitation.

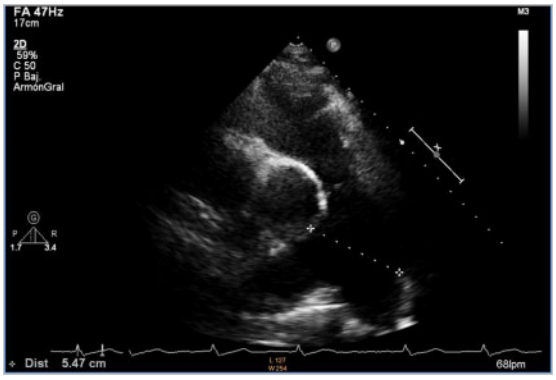


Figure 2 Transthoracic echocardiogram: pulmonary artery aneurysm.

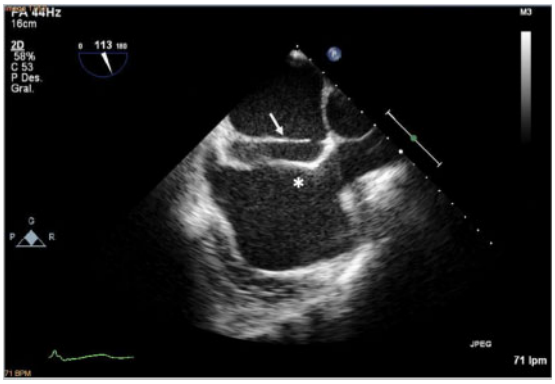


Figure 3 Transoesophageal echocardiogram double atrial septum and interatrial communication: Arrow: septum and ostium primum. Asterisk: accessory atrial septum and its ostium.

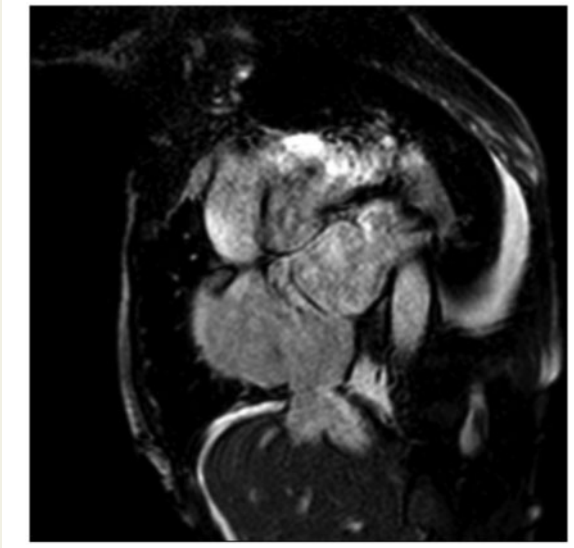
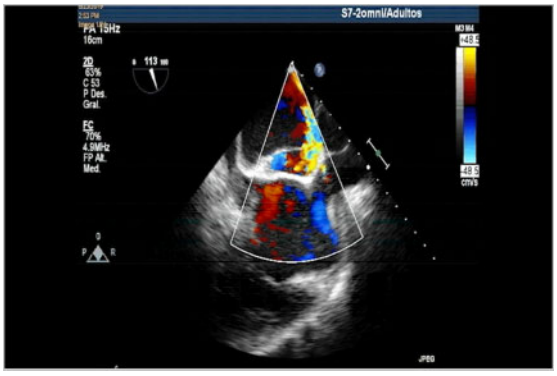


Figure 4 Cardiac magnetic resonance: double septum and pulmonary aneurysm.

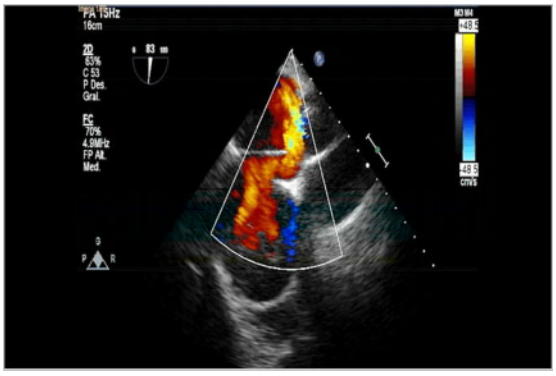
contrast study because we considered that Colour Doppler clearly defined the interatrial walls or double septum, the cavity between them and the small interatrial communication. A cardiovascular magnetic resonance scan confirmed the presence of a double atrial septum and interatrial communication. It showed a non-dilated left ventricle with slightly depressed contractility (58 cc/m^2 and ejection fraction 56%). The administration of gadolinium did not show any global or segmental enhancements. The right ventricle and right atrium were slightly dilated, and the right ventricular ejection fraction was 40%. It also showed a 60 mm artery pulmonary aneurysm (*Figure 4*) and a shunt Q_P/Q_S ratio = 1.3. The severity of the regurgitations was not analysed and the drainage of the four pulmonary veins was normal. The right heart catheterization found a severe second group pulmonary artery hypertension [PAP 72/21 mmHg (medium 38 mmHg), pulmonary capillary wedge pressure (PCWP) 25 mmHg, and pulmonary vascular resistance (PVR) 8 WU]. The coronariography showed normal coronary arteries.

Finally, a comprehensive aetiologic workup was performed and syphilis, endocarditis, carcinoid syndrome, and collagen vascular diseases were ruled out. It was considered that the most likely aetiology was congenital.

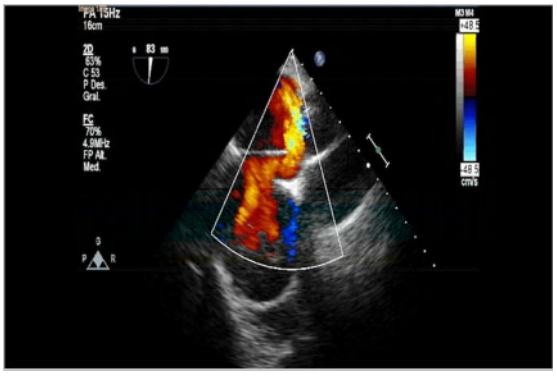
The pharmacological therapy was lisinopril 20 mg o.d. and furosemide b.i.d. The latter was needed because of fluid overload. Taking into account both clinical and cardiac image findings, it was considered that the patient eligible to surgical repair. The decision was made by the multidisciplinary team (cardiologists and cardiac surgeons). It was decided triple valve replacement because of the patient's heart failure and pulmonary hypertension, as well as the closure of the interatrial communication due to the thromboembolic risk, as it is considered below. Finally, it was also decided that the patient needed resection of the pulmonary aneurysm because of



Video 1 Severe mitral and aortic regurgitations.



Video 3 Double septum.



Video 2 Double septum and interatrial communication.

its size (60 mm) and the presence of pulmonary hypertension. The patient has successfully undergone surgery last month (triple valve replacement, tricuspid annuloplasty, pulmonary aneurysm resection, and double septum closure). The patient currently remains asymptomatic and a post-surgical TTE showed left ventricular normal function (left ejection fraction 65%) and normal prosthesis function.

Discussion

To the best of our knowledge, this is the first case report describing double atrium septum, multiple valve regurgitations, and pulmonary aneurysm. The aetiology of the aortic regurgitation was congenital and the mitral regurgitation was secondary to a valve prolapse. The pulmonary regurgitation was related to a congenital pulmonary stenosis for which the patient underwent valvulotomy 50 years ago. The pulmonary aneurysm was secondary to the pulmonary stenosis and a long-standing pulmonary artery hypertension. We also found slight biventricular dysfunction. The right dysfunction was related to the pulmonary regurgitation, whilst the left cardiac dysfunction has been attributed to both mitral and aortic regurgitation. However, it remains unclear why the left heart was undilated given the volume overload state precipitated by both aortic and mitral valve regurgitation severity. We believe that the most likely mechanism is the acuteness of the development of regurgitation of both left-sided heart valves.

The double interatrial septum is a rare congenital condition. The first case was described by Thilenius,[1] who published a case report featuring a double interatrial septum, mitral atresia, double outlet right ventricle, ventricular septal defect, and aortic coarctation. It was found that the cavity between both interatrial walls did not receive any pulmonary veins and was bordered by the fossa ovalis.

This anomaly most likely represents the persistence of the left venous valve derived from the embryogenic sinus venosus.[2] Other more unlikely possibilities are that the additional atrial septum structure represents an abnormal duplication or persistence of either the primum or secundum atrial septal tissue. It is estimated that half of the few reported cases have left-sided obstructive anomalies (left ventricle, left atrium, mitral valve, pulmonary venous structures, and left heart hypoplasia) requiring surgical intervention during childhood. In contrast, older patients, usually only show a small and non-significant left to right shunt, as demonstrated in this case.

It is possible that the cavity between septum primum and accessory septum primum may increase the risk of thrombus formation regardless of the presence or not of atrial fibrillation. Seyfert[3] in 2008 reported a patient with a double atrial septum, persistent interatrial space and a transient ischaemic attack. Breihardt[4] described another case of coronary embolism. Xiao[5] reported a case of a double atrial septum with an interatrial chamber and stenosis of the inferior vena cava orifice, which led to recurrent paradoxical embolism. Harding[6] and Deegan[7] published incidental findings of double interatrial septum in two patients undergoing pulmonary vein isolation because of atrial fibrillation. The former author postulated that, during the transseptal puncture, it was necessary to target a confluent portion of the interatrial septum, avoiding the double septum. The presumed mechanism of this thrombus structures. There is limited experience about surgery in these patients, but the closure of the double septum was performed in two published cases.[5,8] Therefore, it is possible that surgical closure of the cavity may be necessary, despite of the shunt

being small, which is the normal situation in older patients, in order to remove the risk of systemic thromboembolisms.

Lead author biography

José Antonio Ortiz de Murua working as cardiologist for 30 years. I have written more than 50 papers in national and international Cardiology journals. I work as echocardiographist, and I am involved in three lines of investigation at the moment (cardiac amyloidosis, anti-aggregation, and acute coronary syndrome and new oral anticoagulants).

Supplementary material

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

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

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

Conflict of interest: none declared.

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