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Double Interatrial Septum: When Two Layers Aren't Enough

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A 61-year-old male with history of hypertension, diabetes, and atrial fibrillation presented to a cardiology consultation due to progressive dyspnea. The transthoracic echocardiogram revealed severe dilation of the right chambers, with a significant interatrial left-to-right shunt with a turbulent flow and preserved biventricular systolic function (left ventricular ejection fraction 56% and right ventricle fractional area change of 38%). Transesophageal echocardiogram was performed to better characterize the anatomy, revealing 2 parallel atrial septal structures, a septum primum (SP) and an accessory atrial septum (AAS) with an echo-free interatrial space (**Movie 1** and **2**). A left-to-right shunt was identified due to atrial septal fenestrations and septal defects in the SP and at the middle segment of AAS (**Figure 1A-D**; **Movie 3**). A cardiac computed tomography scan confirmed the echo findings (**Figure 1E and F**). The right cardiac catheterization performed before surgery estimated a Qp/Qs of 2.2 and revealed a reduced cardiac index (1.9 L/min/m^2), with no pulmonary arterial hypertension (resting mean pulmonary arterial pressure of 20 mmHg). The patient underwent cardiac surgery with complete removal of both interatrial septa, implantation of a bovine pericardial patch between the atria, tricuspid valve annuloplasty and left atrial appendage closure. At the 6 months of follow-up, he remains asymptomatic.

Double interatrial septum is a rare congenital anomaly, mostly due to abnormal development of septum secundum, resorption failure of the superior portion of SP, or persistence of the left venous valve of sinus venosus.[1)2)] This case illustrates an unusual scenario in which both septa allow a significant left-to-right shunt, being the first reported case of a double interatrial septum diagnosed due to symptoms mimicking heart failure presenting late in life.

Written informed consent for publication of clinical details and/or clinical image was obtained from the patient.

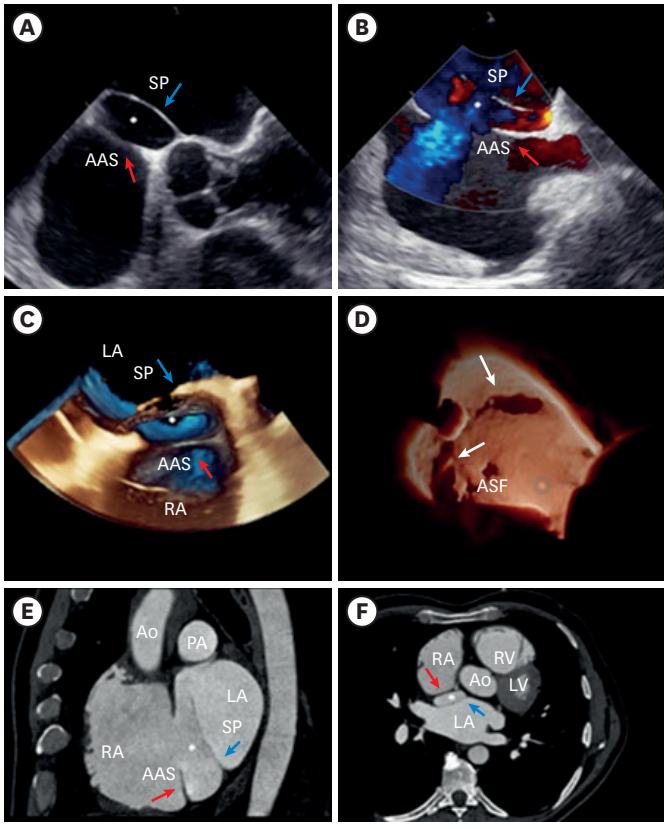

Figure 1. (A) TEE at 0º revealing two parallel septal structures creating an echo free IAS with 14 mm between them. The IAS is in a phase of expansion, as blood enters the cavity from the left atrium during ventricular systole. (B) Midesophageal bicaval view with color Doppler showing a shunt between IAS and RA through a septal defect (28 × 33 mm) at the middle segment of AAS, and with LA through the ASF and 2 main septal defects present in SP. (C) 3D TEE exhibiting the IAS between atria, with no thrombus. (D) 3D TEE showing the SP with ASF and the 2 main septal defects (the superior with major axis of 16 mm and the inferior with 19 mm) (white arrow). (E, F) Multiplanar computed tomography reconstructions in both coronal and axial planes revealing the IAS (*) with the main septal defects in both interatrial septa (E). Blue arrow: septum primum; Red arrow: accessory atrial septum.
AAS: accessory atrial septum, AO: aorta, ASF: atrial septal fenestrations, IAS: interatrial space, LA: left atrium, LV: left ventricular, PA: pulmonary artery, RA: right atrium, RV: right ventricular, SP: septum primum, TEE: transesophageal echocardiogram, 3D: three-dimensional.

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Conflict of Interest

The authors have no financial conflicts of interest.

Author Contributions

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SUPPLEMENTARY MATERIALS

Movie 1

Three-dimensional transesophageal echocardiogram revealing the 2 parallel septal structures creating an echo free interatrial space, with no thrombus.

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Movie 2

Transesophageal echocardiogram at 0º revealing 2 parallel septa, creating an echo free interatrial space with 14 mm in between. During ventricular systole, the interatrial space is in phase of expansion, as blood enters inside the cavity from the left atrium, and during ventricular diastole is in phase of partial collapse, as blood drains out of the space, into the right atrium and ventricle.

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Movie 3

Midesophageal bicaval view with color Doppler showing a shunt between interatrial space and right atrium through a septal defect (28 × 33 mm) at the middle segment of accessory atrial septum, and with left atrium through the atrial septal fenestrations and 2 main septal defects (one superior and other inferior) in septum primum.

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REFERENCES

1. Adamane Jayaram A, Uddina Kumeri AR, Rao SM, Tuli A, Nayak K. Double interatrial septum with persistent interatrial chamber: a rare but clinically significant anomaly. *Echocardiography* 2020;37:1694-7.
PUBMED | CROSSREF
2. Roberson DA, Javois AJ, Cui W, Madronero LF, Cuneo BF, Muangmingsuk S. Double atrial septum with persistent interatrial space: echocardiographic features of a rare atrial septal malformation. *J Am Soc Echocardiogr* 2006;19:1175-81.
PUBMED | CROSSREF