

E-CHALLENGES AND CLINICAL DECISIONS

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Adult Congenital Heart Defects: How Many Is Too Many?

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CASE HISTORY

A 47-YEAR-OLD MAN presented to the authors' institution for an elective surgical closure of an atrial septal defect (ASD) after a failed attempt at percutaneous closure (Fig 1, Panel A). He was known to have a peri-membranous ventricular septal defect (VSD) and a quadricuspid aortic valve (Fig 1, Panels B and C; Video 1). On the day of surgery, after uneventful induction of general anesthesia, an intraoperative transesophageal echocardiography (TEE) examination was performed, which confirmed the presence of an ASD, a non-restrictive peri-membranous VSD, and a quadricuspid aortic valve. These findings were confirmed with real-time, 3-dimensional (3D) TEE imaging (Fig 2; Video 2).

ECHOCARDIOGRAPHIC FINDINGS

1. Two-dimensional (2D) imaging revealed that the patient had a prominent and mobile filamentous, echodense membrane extending from the lateral wall of the right atrium to the base of the interatrial septum (Fig 3; Video 3).
2. Additionally, an echo density was visualized in the right atrial appendage (RAA), which appeared to be sessile and immobile on 2D imaging (Fig 4; Video 4).

CLINICAL CHALLENGES

1. What was the extent of the filamentous, membranous structure in the right atrium?
2. Was the other immobile echo density in the RAA a clot or a tumor?

CLINICAL COURSE

The authors performed real-time 3D TEE imaging of the 2 unanticipated findings. Considering the mobility and extent of the filamentous membrane, its appearance was consistent with a cor triatriatum dextrum. Examination of the RAA using the "live" (narrow and wide angle) and R-wave-gated 3D imaging modes demonstrated it to be bifid in nature. The echo density seen on 2D imaging was part of a longitudinal ridge dividing the RAA into 2 lobes. Therefore, a clot and RAA tumor were excluded.

Functional examination of the quadricuspid aortic valve excluded the presence of stenosis or regurgitation. The surgical team decided that the cor triatriatum dextrum was non-obstructive and did not require any intervention. The patient underwent surgical closure of the ASD and VSD and was weaned successfully off cardiopulmonary bypass. The postoperative course was uneventful, and the patient was discharged a few days later.

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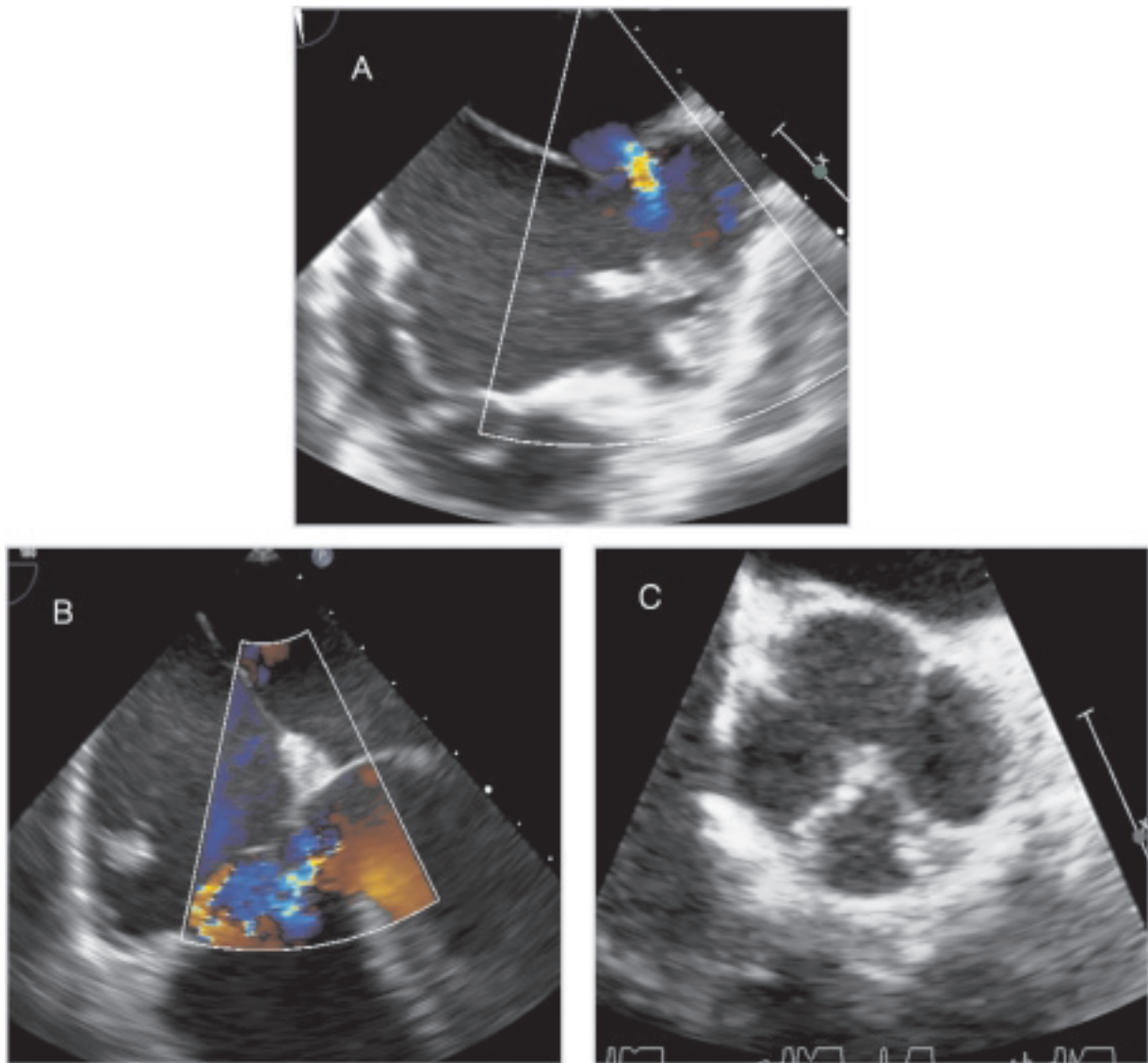


Fig 1. Multiple congenital abnormalities. Panel A, Midesophageal bi-caval view with color-flow Doppler demonstrating the secundum-type atrial septal defect with a left-to-right shunt. Panel B, Midesophageal 4-chamber view with color-flow Doppler showing a perimembranous ventricular septal defect with a left-to-right shunt. Panel C, Quadricuspid aortic valve.

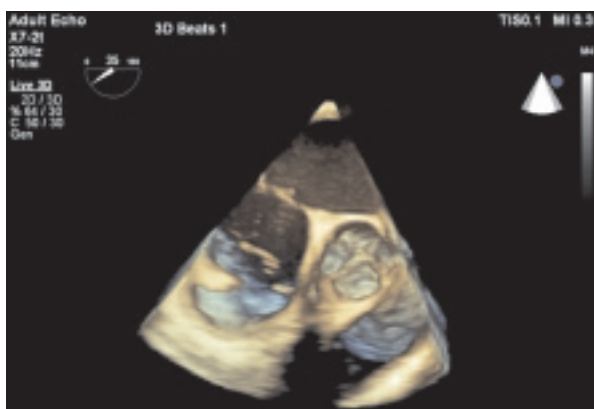


Fig 2. Three-dimensional view of the aortic valve. A 3-dimensional single-beat, full-volume en-face view of the quadricuspid aortic valve. An echo density in the right atrium also is visualized.

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