Raghib syndrome: Echocardiographic features

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Introduction

A 25-year-old female was referred to our hospital with a 3 year history of palpitation and cyanosis. The chest X-ray was suggestive of pulmonary artery and right heart dilatation, and the ECG showed right ventricular hypertrophy. A transthoracic echocardiogram showed absent coronary sinus (CS) on the parasternal long axis view (Fig1A). The roof of coronary sinus was absent and a secundum type atrial septal defect (ASD) was also noticed on the apical four chamber view with left-to-right shunt from left atrium (LA) to right atrium (RA) through the absent coronary sinus, on color Doppler (Figs1B-1C). The persistent left superior vena cava (PLVC) was demonstrated on the suprasternal view. A right heart contrast echocardiography showed the micro-bubbles first appearing in the LA appendage, confirming the PLVC draining into LA (Fig1D). Thus, the diagnosis was Raghib syndrome with secundum ASD, and the patient was referred for surgical repair.

The surgical procedure showed completely unroofed CS and the PLVC drained into LA between left appendage and left upper pulmonary vein. Rerouting the PLVC into RA was successfully achieved by reroofing of the CS and reconstructing the atrial septum. A post-operative transthoracic echocardiogram showed the patch between the atrial septum and CS, no residual shunt between LA and RA and PLVC draining into the RA (Figs2A-D).

Discussion

Raghib syndrome is a special type of ASD which is known as coronary sinus septal defect (CCSD), first described by Raghib 1. CCSD accounts for less than 1% of ASD and 0.1% of all congenital heart diseases (CHDs). CCSD is classified as three types according to completely or partially absent CS wall (Type I: complete; Type II: unroofed mid-portion and Type III: unroofed terminal portion) 2. Each type of CCSD is subsequently divided into 2 subtypes according to the presence of PLVC: a) with PLVC and b) without PLVC. Type Ia is also called Raghib syndrome which includes absent CS, coronary sinus ASD and PLVC draining into the LA between the appendage and left upper pulmonary vein 3. The right superior vena cava is usually thin or completely absent. In inexperienced hands the Raghib syndrome could easily be missed with simple ASD, but detailed transthoracic echocardiographic examination with all modalities, color Doppler and bubble contrast should assist in achieving the complete diagnosis. The correct diagnosis should also guide towards optimum surgical planning which involves reroofing of the CS as well as rerouting the PLVC rather than simple ASD patching 4.

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Fig1 Pre-operation echocardiogram
A No CS was found from parasternal long axis view
B The roof of CS was absent and the secundum type ASD was seen from apical four-chamber view (arrow)
C Left-to-right shunt from LA to RA through the absent CS on color Doppler imaging (arrow)
D The right heart contrast echocardiography showed the bubble contrast first appeared in the LA confirming that the PLVC drained into LA.
(CS=coronary sinus; ASD= atrial septal defect; LA=left atrium; RA=right atrium; PLVC= persistent left superior vena cava)

Fig2 Post-operation echocardiogram
A The patch between the atrial septum and the coronary sinus level was noticed.
B The left-to-right shunt between atria was disappeared on color Doppler
C and D Two-dimentional and Color Doppler echocardiography showed the PLVC flow drained into RA.
(PLVC= persistenr left superior vena cava, RA=right atrium)
References