



A case of asymptomatic patient with right ventricular dilatation

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Abstract: Sinus venous defect is an uncommon type of atrial septal defect (ASD), often associated with anomalous pulmonary vein drainage and left to right shunting. If undetected, it leads to right atrial and ventricular volume overload. The current case describes an asymptomatic athlete with missed large sinus venosus defect since birth, and the different types of cardiac imaging modalities that lead to the detection of the defect and guiding the appropriate surgical intervention.

Keywords: Sinus venosus defect; anomalous pulmonary vein drainage; shunt; right ventricular dilatation

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Case presentation

This is a 28-year-old male who presented to the emergency department for traveler's diarrhea that resolved the following day. He is a super-athlete, heavy weight lifter, runner, and mountain climber and completely asymptomatic and in very good health. His physical examination however, showed regular heart beats with unexpectedly right ventricular thrill, fixed split S2 and 3/6 holosystolic murmur throughout the precordium. Electrocardiogram showed incomplete right bundle branch block and right axis deviation (*Figure 1A*). Chest X-ray showed dilated cardiac silhouette with right ventricular and right atrial enlargement, and pulmonary vasculature plethora (*Figure 1B*). Transthoracic echocardiogram showed severely dilated right ventricle (RV) and right atrium (RA) (*Figure 2A*) but with preserved systolic function and normal free wall global longitudinal strain -24% (*Figure 2B*). The interatrial septum appeared intact without secundum atrial septal defect (ASD) (*Figure 2A*). Shunt calculation revealed Qp/Qs =3.1 using the formula Eq. [1]:

$$\frac{[(RVOT \text{ diameter } 2.6 \text{ cm})^2 \times (RVOT \text{ VTI } 39 \text{ cm})]}{(LVOT \text{ diameter } 2.1 \text{ cm})^2 \times (LVOT \text{ VTI } 19 \text{ cm})} \quad [1]$$

Since there was a high suspicion of intracardiac shunt that could not be visualized by transthoracic echocardiogram (agitated saline was not performed), transesophageal

echocardiogram was performed and showed large superior sinus venosus defect (*Figure 2C*, white arrow) with anomalous right pulmonary venous drainage (*Figure 2C*). Cardiac computed tomography (CCT) was performed for better 3D images to guide the surgeon plan the surgery; it confirmed the large superior sinus venosus defect (black arrow) with communication between the superior vena cava (SVC), RA, and left atrium (LA), along with abnormal drainage and blood mixing from the right upper pulmonary vein (RUPV) and right lower pulmonary veins (RLPV) (*Figure 3*). Cardiac magnetic resonance (CMR) imaging was requested mainly to quantify the RV volumes/EF (gold standard) to establish a baseline and follow-up in 1 year for reverse remodeling. It revealed the anomalous pulmonary veins (*Figure 4A*) and quantified the shunt to be 165 mL/beat with Qp/Qs 3.2 (*Figure 4B*). Surgery was recommended and the patient underwent surgical closure of the defect (*Figure 5A*) using a patch (*Figure 5B*) along with correction of the pulmonary venous drainage. Patient recovered well and was discharged home day 4 post-op.

Sinus venosus defect

ASD is among the most common congenital abnormalities in adults. It consists of a deficiency in the atrial septum; when located in the middle of the septum (ASD secundum),

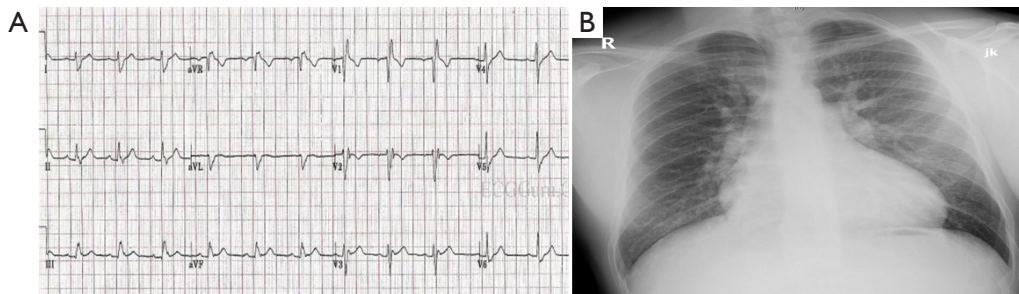


Figure 1 Twelve lead ECG on presentation showing incomplete right bundle branch block and right axis deviation (A). Chest X-ray showing dilated cardiac silhouette, particularly the RA and RV, with plethora of the pulmonary vasculature (B). ECG, electrocardiograph; RA, right atrium; RV, right ventricle.

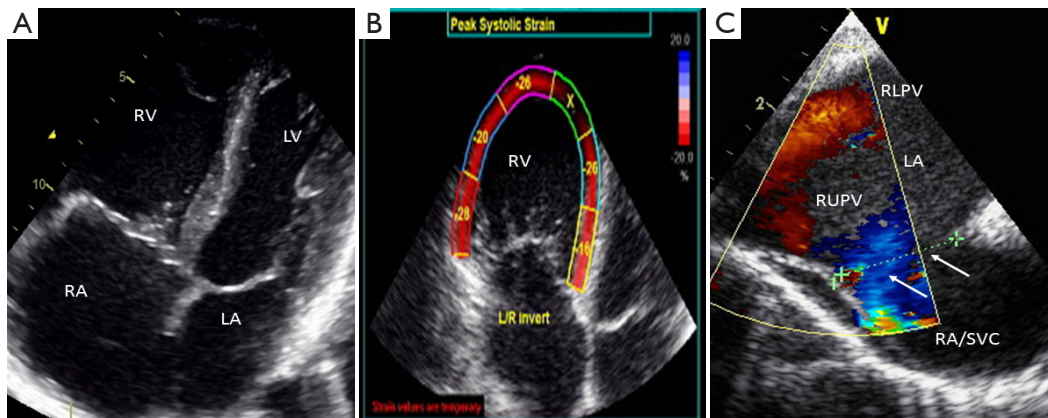


Figure 2 Transthoracic echocardiogram showing dilated RV and RA (A) with normal right ventricular longitudinal strain (B). Transesophageal echocardiogram showed a superior sinus venosus defect (white arrow, C). LA, left atrium; LV, left ventricle; RA, right atrium; RLPV, right lower pulmonary vein; RUPV, right upper pulmonary vein; RV, right ventricle; SVC, superior vena cava.

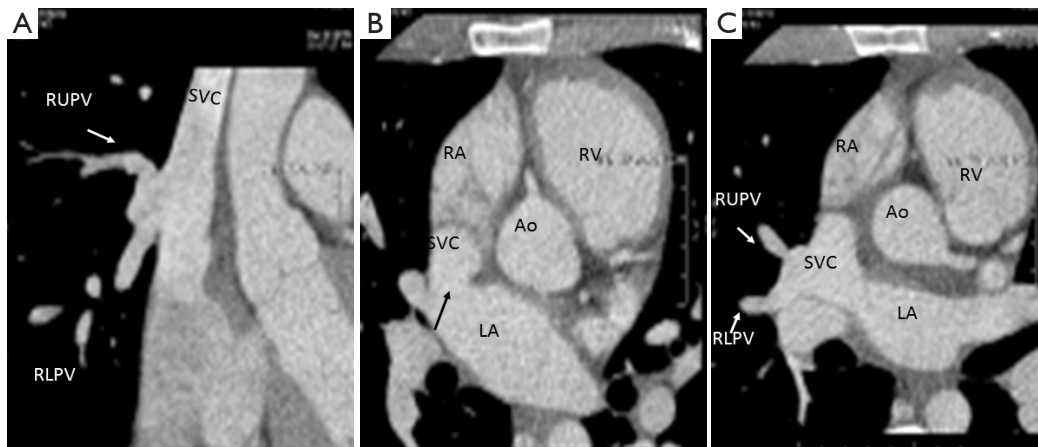


Figure 3 CCT demonstrating the sinus venosus defect (black arrow), with anomalous pulmonary venous drainage. SVC, superior vena cava; CCT, cardiac computed tomography; LA, left atrium; RA, right atrium; RLPV, right lower pulmonary vein; RUPV, right upper pulmonary vein; RV, right ventricle; SVC, superior vena cava.

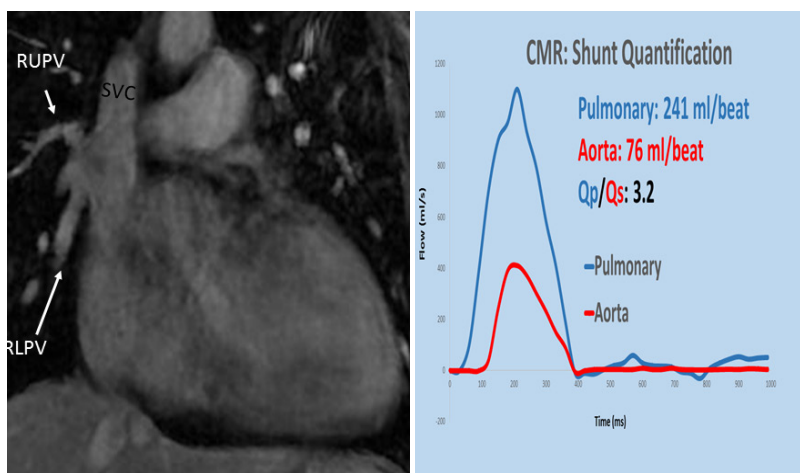


Figure 4 CMR imaging confirming the anomalous pulmonary veins and quantifying the intracardiac shunt using flow measurement. RLPV, right lower pulmonary vein; RUPV, right upper pulmonary vein; SVC, superior vena cava; CMR, cardiac magnetic resonance.

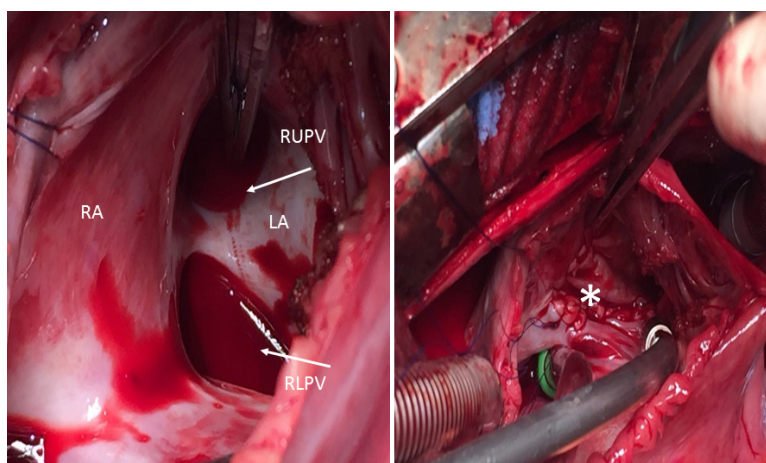


Figure 5 Intraoperative visualization of the defect followed by patch closure and redirection of the pulmonary veins. *, patch repair; RLPV, right lower pulmonary vein; RUPV, right upper pulmonary vein; LV, left ventricle; RA, right atrium.

it is easily identified on transthoracic echocardiogram, and accounts for more than 70% of cases (1). However, when the defect is located superiorly close to the SVC (sinus venosus defect), it is not well seen on transthoracic echocardiogram and is often missed (2). Sinus venosus defect accounts for 10% of ASD and is associated with left to right shunting; if significant, it results in right sided volume overload pattern with right ventricular and right atrial dilatation. Patients are often asymptomatic, particularly when they are still young. Typical electrocardiogram shows incomplete right bundle branch block and right axis deviation (3). A fixed split S2 with signs of right ventricular overload on physical exam

and on chest X-ray should raise suspicion for an ASD (2). In addition, sinus venosus defect is much more likely to be associated with anomalous pulmonary vein drainage as compared to ASD secundum (70% *vs.* 10%). Dilated RV and RA in the absence of known cause such as tricuspid or pulmonary regurgitation, and with an apparent “intact” inter-atrial septum on transthoracic echocardiogram, should raise the suspicion of a sinus venosus defect and/or anomalous pulmonary vein drainage (2). The next step involves performing transesophageal echocardiogram (4), CCT (5) or CMR (6). CCT is easy to perform, non-invasive, and provides great anatomical definition in three-

dimension with reconstruction that is fundamental to guide the surgeon pre-operatively (5). CMR allows the accurate and reproducible quantification of the intracardiac shunt by calculating pulmonary and systemic flow; a shunt ratio less than 1.5 is small and clinically non-significant (6). Unlike ASD secundum, sinus venosus defect cannot be close percutaneously (7). Moreover, identification of concomitant anomalous pulmonary veins is essential to avoid leaving behind a residual left to right shunt.

In our case, a detailed physical exam with an unusual electrocardiogram and chest X-ray raised the suspicion of an intracardiac shunt; appropriate investigation resulted in the correct identification of a missed congenital defect for 28 years that was surgically corrected to save the patient future complication of irreversible right ventricular damage and heart failure. While there are several published similar cases in the literature, the late age at presentation, the fact that he was a super athlete and completely asymptomatic, multi-modality imaging including intra-operative surgical images make this case original.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at <http://dx.doi.org/10.21037/jxym.2016.12.09>). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent

was obtained from the patient.

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