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## INTRODUCTION

Cor triatriatum (CTA) is a rare congenital heart defect in which a fibromuscular membrane forms an anomalous septum within the left atria (LA), dividing it into a posterior superior higher pressure chamber and an anterior inferior anatomically true LA. Typically all four pulmonary veins enter up stream of the anomalous septum. We present an unusual form CTA in which only the right sided pulmonary veins (RPV) were involved.

## CASE PRESENTATION

A 26 year old male with PMH of multiple lower respiratory tract infections, presented to the ER for an episode of hematemesis. Investigations revealed anemia, normal platelets, LFTs, grade 2 esophageal varices on EGD; chest CT demonstrated interlobular septal thickening of the entire right lung, along with an anomalous septum in the LA.

Cardiac MR confirmed the diagnosis of CTA by demonstrating the membrane in the right side of the LA, encompassing the right pulmonary veins (RPV) only, as well as severely impaired pulmonary venous drainage of the right lung with collateral drainage through right periesophageal varices (PEV). In order to provide relief of RPV outflow obstruction, resection of the CTA membrane was planned.

Upon arrival to the OR, anesthetic induction and endotracheal intubation were performed uneventfully. Initial intra-operative TEE revealed normal biventricular function with a LA membrane isolating the RPVs from the body of the LA. Color flow Doppler (CFD) revealed a small, highly restrictive opening through the anomalous septum. After median sternotomy and establishment of cardiopulmonary bypass, the LA was opened anterior to the RPV to enter into the high pressure chamber. The CTA membrane was visualized and seen to extend only over the RPV. The membrane was resected and the LA closed. The patient was weaned off CPB without hemodynamic support. Follow up intra operative TEE demonstrated absence of residual membrane, free flow of the RPV into the body of the LA. The patient was transported to the ICU and was extubated on POD 0 and discharged home on POD 4.

## DISCUSSION

The diagnosis of CTA is initially suspected when TTE shows a linear structure within the LA. Use of CFD and 3D TTE technology has been reported to accurately establish the diagnosis of CTA(2, 3). In the intra-operative setting, or an inadequate TTE examination, TEE provides excellent imaging of the CTA membrane, LA, membrane orifices and allows quantification of the obstruction with higher sensitivity.

We describe a variant of CTA, in which only one-half of the pulmonary venous circulation is obstructed by the CTA membrane. The unusual presentation of hematemesis was likely the result of restriction of the RPV drainage into the LA, causing venous drainage to be redirected through various collaterals, which created back pressure resulting in PEV, which eventually ruptured.

1 Trans Path Soz. 1868;19:188-190.

2 Eur J Echocardiogr 2008, 9(1):110-2.

3 J Am Soc Echocardiogr 2006;19:468

