Unroofed Coronary Sinus – A Rare Type of ASD
Ali Karaosmanoglu, MD, Mannudeep Kalra, MD; Moussa Mansour, MD; Wilfred Mamuya, MD, PhD; Suhny Abbara, MD

Clinical History
A 53-year old man presented to the cardiology clinic with a shortness of breath and atrial fibrillation with a rapid ventricular response. His past medical history was remarkable for hypertension, hyperlipidemia and a non ST - segment elevation myocardial ischemia. A computed tomography angiogram (CTA) evaluation of the pulmonary veins was requested as a part of pre-procedural work - up for endovascular pulmonary vein isolation.

Findings
Contrast enhanced CT examination revealed normal pulmonary vein anatomy. The coronary sinus was found to be mildly dilated in the left atroventricular groove (figure 1, 2) and there was also a 2.2 cm segment of abnormal communication between the base of the left atrium and the roof of the coronary sinus, consistent with an unroofed coronary sinus (figure 3). There was no evidence of persistent left sided superior vena cava (LSVC). Contrast material was seen entering the coronary sinus at the site of unroofing and it shunted into the right atrium via the normal coronary sinus ostium. As there was no clinically significant amount of shunting no shunt related intervention was made and the patient was continued to be medically managed.

Discussion
Unroofed coronary sinus is a rare congenital cardiac anomaly which might be difficult to diagnose (1). It is classified as an atrial septal defect and constitutes the rarest form of this group of congenital heart disease (2). The anatomic abnormality is variable and classified into four groups: type 1, completely unroofed with persistent LSVC; type 2, completely unroofed without persistent LSVC; type 3, partially unroofed mid portion; and type 4, partially unroofed terminal portion (2). The presented case appears to be consistent with type 4 subgroup of this anomaly.

The development of symptoms appears to be related to the size of the defect, and the severity of the inter-atrial shunt, which may lead to the development of right heart failure. The diagnosis should be suspected in a patient with LSVC and associated brain abscess or cerebral embol; or in a patient with unexplained arterial oxygen desaturation (1). Management depends on the clinical symptoms and surgical intervention should be considered when the symptoms cannot be managed medically. Imaging plays a crucial role in the diagnosis. Transthoracic echocardiography is, limited in its ability to evaluate the posterior structures. Cross sectional imaging with computed tomography (CT) and magnetic resonance imaging Are well suited to identify this abnormality.

REFERENCES