

Coronary sinus ostial atresia (CSOA) with persistent left superior vena cava (LSVC) is a rare congenital cardiac anomaly.

A 24-year-old female with history of levo-transposition of the great arteries, left-sided atrioventricular valve atresia and non-restrictive ventricular septal defect presents with systemic oxygen desaturations during exercise (baseline 92%, peak exercise 83%). She underwent atrial septectomy as a newborn with subsequent bidirectional superior cavopulmonary anastomosis at age 2.

Cardiac catheterization revealed a left innominate vein (LIV) with unobstructed flow to right SVC. A moderate size left SVC was seen coming off the left innominate vein (LIV) and connecting to a dilated coronary sinus (CS). The ostium of CS was found to be atretic with the coronary veins draining retrogradely into the LIV via the left SVC. Two veno-venous (VV) pulmonary collaterals were also seen draining into the left pulmonary vein from the LIV and LSVC.

We deliberated between sealing the VV collaterals versus opening the coronary sinus. Opening the sinus is more invasive as it would redirect blood flow from the LIV directly into the atria via the LSVC. This would then necessitate closer of the LSVC, which is a much larger vessel than the VV collaterals. The decision was made to close the VV collaterals.

It is critical to rule out CSOA in patients with Fontan circulation. Inadvertent closure of CS decompressing vessels can lead to coronary venous hypertension and subsequent myocardial hypoperfusion and ischemia.



Injection of L SVC showing dilation of L SVC and coronary veins with lack of drainage of coronary vasculature into R atrium

Injection of LSVC showing a veno-venous (VV) collateral arising from the LSVC to the left pulmonary vein

