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Background

- The incidence of congenital heart disease (CHD) in the parturient is ~1% of U.S. births per year [1].
- Situs or heterotaxy anomalies are among the least common maternal congenital lesions in pregnancy. Heterotaxy syndrome is estimated at 380 births per year in U.S. [2].
- We describe the anesthetic considerations in a rare case of a parturient with situs ambiguus undergoing cesarean delivery.

Figures

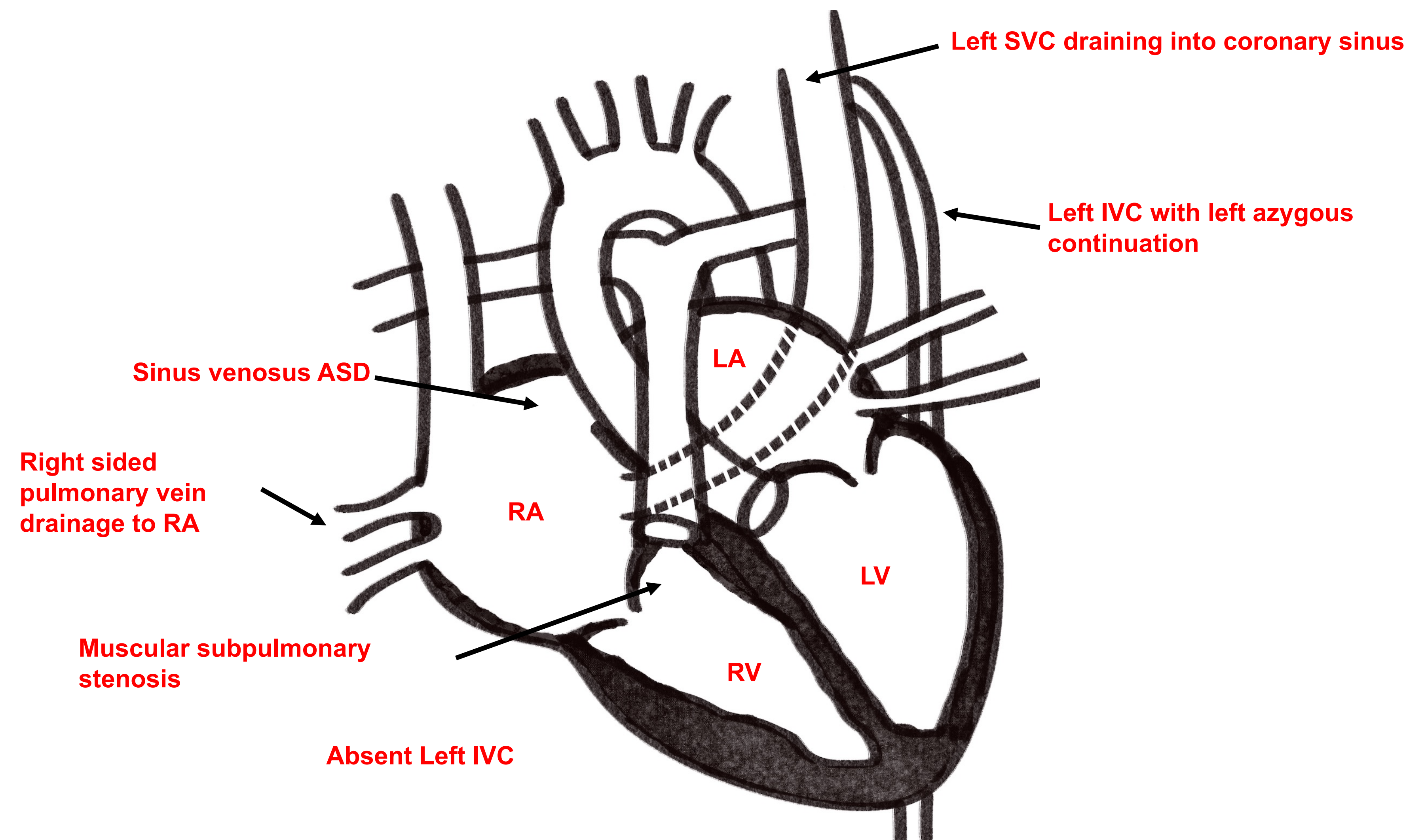


Figure 1. Congenital heart findings prior to correction. SVC = superior vena cava, IVC = inferior vena cava, RA = right atrium, LA = left atrium, RV = right ventricle, LV = left ventricle, ASD = atrial septal defect

Case Description

A 35-year-old G2P1 at 38 weeks gestation presented in latent labor with fetus in breech presentation. Due to a recent transfer of care to our ACOG Level IV hospital, the patient had no electronic medical records available on admission. Past medical history included cardiac surgery at 3-years-old and uneventful cesarean delivery 8 years ago. Patient was unsure about the specifics of her CHD and was not currently followed by a cardiologist. In the past, she was told that she would require a pacemaker as she gets older.

We ordered a congenital TTE, EKG, and a congenital cardiology consult. Significant findings (Figure 1) included situs ambiguus, interrupted left inferior vena cava with left azygous continuation, bilateral superior vena cava with left superior vena cava draining into coronary sinus, superior sinus venosus defect with PAPVR of both right-sided pulmonary veins into right atrium and muscular subpulmonary stenosis, and a perforated cor triatriatum sinister status post intracardiac baffle and rerouting of right pulmonary veins and RVOT myectomy. EKG revealed accelerated junctional rhythm with frequent PVCs at 80bpm suggesting 2 sinus nodes with the implication that we may not be able to easily increase her heart rate if needed, e.g. in the setting of hypotension.

Patient underwent a repeat cesarean delivery under combined spinal-epidural analgesia with a narcotic-only spinal (morphine 0.15 mcg/fentanyl 20 mcg intrathecal) and slow loading of lidocaine 2% + epinephrine 5 mcg/ml via epidural to minimize chance of hypotension. Due to her CHD and situs ambiguus, we used right uterine displacement and had multiple classes of vasopressors on standby. The cesarean was uncomplicated.

Discussion

Parturients with history of CHD, even after repair, require thorough evaluation and multidisciplinary care to understand their physiology and anatomy. Transfer to an appropriate higher level of care hospital and multidisciplinary care is needed. Heterotaxy syndrome is rare, complex, and has many different subtypes and presentations which makes every case unique. Anesthetic considerations will ultimately depend on the type of heterotaxy syndrome, and specialized/rare lesions benefit from a physiologic discussion with cardiology.

References

1. *Data and Statistics on Congenital Heart Defects*. 2022: Centers for Disease Control and Prevention.
2. Lambert, T.E., et al., *Abnormalities of Fetal Situs: An Overview and Literature Review*. Obstetrical & Gynecological Survey, 2016. **71**(1): p. 33-38.