

Persistent Left SVC Syndrome in a Patient with Turner's Syndrome Undergoing Aortic Valve Repair

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Introduction

- Turner Syndrome (TS) is a chromosomal disorder in females characterized by the partial or complete loss of the second X-chromosome.
- PLSVC is an extremely rare anatomical anomaly with an incidence of less than <0.5% and typically drain into the coronary sinus and the right atrium.
- Our patient with Turner’s Syndrome had a persistent left superior vena cava (PLSVC) that directly affected our management as she was undergoing aortic surgery and would be undergoing cardiopulmonary bypass (CPB).

Case Description

- A 25-year-old female with Turner syndrome and developmental delay was evaluated for worsening breathing difficulties.
- A transesophageal echocardiogram (TEE) showed severe aortic insufficiency with a bicuspid aortic valve, and she was brought in for surgical aortic valve repair.
- The patient’s airway proved difficult to manage, requiring multiple attempts with video laryngoscopy before intubation was successfully achieved.
- The patient’s neck also caused difficulty in placing a central venous line on the right side, thus a central venous line was accessed via left internal jugular vein.
- TEE showed that the patient had a persistent left sided superior vena cava (PLSVC) and that the central line was in the right atrium instead of the SVC.
- Throughout the case, the patient had elevated coronary sinus pressures and had multiple episodes of new onset atrial fibrillation.
- Careful monitoring and treatment allowed the surgeons to repair the aortic valve and safely finish surgery.

Discussion

- Anesthetic concerns in TS involve physical abnormalities like short neck and micrognathia, and cardiovascular issues such as a bicuspid aortic valve and coarctation of the aorta, which complicate airway management, intubation, and hemodynamic monitoring.
- Thus, thorough pre-operative evaluation of cardiac status and airway anatomy is essential.
- While often asymptomatic, PLSVC can present in conjunction with other cardiac abnormalities, necessitating close monitoring and management during surgery.
- Central venous access and pulmonary artery catheters can also prove to be difficult and should be ideally confirmed via TEE.
- In patients undergoing CPB it is crucial to carefully monitor the patient’s hemodynamics as they are at much higher risk for suddenly developing an unstable arrhythmia.

Conclusion

- Turner syndrome has an incidence of 1 in 3000 live births and a persistent left SVC syndrome is a rare occurrence that can be associated with TS.
- Due to physical and cardiovascular concerns, thorough pre-operative evaluation of cardiac status and airway anatomy is essential.
- As with our patient, uncomplicated surgery also requires careful monitoring for arrhythmias and elevated pressures.

Imaging



CT scan demonstrating the persistent left superior vena cava (PLSVC), a rare congenital venous anomaly. The PLSVC is seen as an additional vessel on the left side of the mediastinum (indicated by the arrow), which typically drains into the coronary sinus.

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References

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