What is normal in an abnormality? Central venous cannulation in a patient with Situs inversus totalis with dextrocardia and polyCystic kidney disease

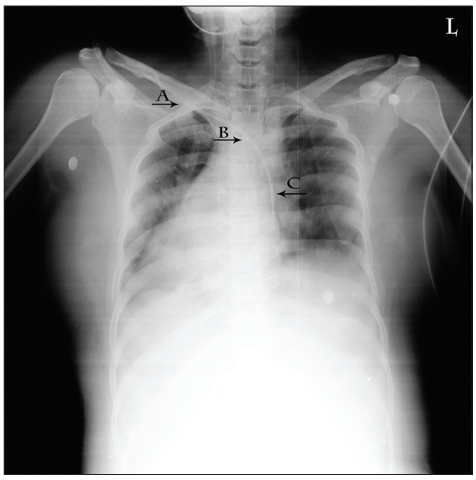

Figure 1: What is normal in an abnormality? Central venous cannulation in a patient with situs inversus totalis with dextrocardia and poly cystic kidney disease

Sir,

Situs inversus totalis (SIT) with dextrocardia and poly cystic kidney disease (PCKD) is a rare condition, which can be associated with total anomalous pulmonary venous connection (TAPVC) and persistent right-superior vena cava (SVC).[1,2] Central venous catheterization (CVC) in patients with anatomical abnormalities are complex and challenging, especially in emergency situations.

Our patient was a 38-year-old lady diagnosed with SIT coupled with dextrocardia and PCKD, who underwent ureteroscopic lithotripsy under spinal anesthesia. Postoperatively, she developed urosepsis, hemodynamic instability, requiring CVC. Although preoperative echocardiography had revealed dextrocardia, normal cardiac chambers, right aortic arch, and left SVC, relative position of internal jugular vein (IJV) and carotid artery in the neck was doubtful. Because of unavailability of ultrasonography during emergent cannulation, right subclavian vein (A) was cannulated and correct positioning confirmed with pressure waveform. Post CVC chest X-ray [Figure 1] revealed the CVC crossing the midline through the right-brachiocephalic vein (B) entering the left SVC (C) toward the morphologic right atrium. In normal patients, correctly positioned right CVC remains on the right side without crossing the midline. If it crosses the midline, it could be because of the malposition into the left internal jugular, left subclavian vein, or left persistent SVC. There are few exceptions for the correctly positioned right CVC to cross the midline, and SIT is one of them.[3]

In patients with SIT, if right CVC has not crossed the midline and below the level of carina, its entry into persistent right SVC, TAPVC, and malposition into the aorta should be suspected.

In our patient, we later confirmed using ultrasound the normal great vessel relationship in the neck. In SIT, the relationship of carotid artery and internal jugular veins remains normal. Such anatomical relationships are as equally important during CVC as during the cardiothoracic surgery. Preferable site of CVC in a patient with SIT would be left IJV, as it has a straighter course using ultrasonography.

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Quick Response Code:

Website: www.ijccm.org

DOI: 10.4103/0972-5229.118406