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REVIEW ARTICLE

Transesophageal Echocardiographic Diagnosis and Imaging of Cardiac Situs and Malposition

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ABSTRACT

Dextrocardia (DC) is a rare cardiac anomaly that can be associated with other congenital abnormalities. Transesophageal echocardiographic imaging for evaluating these patients requires modification of the omniplane angle from the standard angles used. We describe the imaging technique in this condition when it is associated with situs solitus (SS) and situs inversus.

Keywords: Dextrocardia, Echocardiography, Situs inversus, Transesophageal.

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INTRODUCTION

Dextrocardia is one of the rare cardiac anomalies with an incidence rate of <1 per 10,000 pregnancies/live birth.[1,2] It is defined as a position of the major portion of the heart in the right hemithorax with the base to apex directed to the right and caudal. It is a congenital anomaly that has to be differentiated from a similar condition called dextroposition of the heart in which the major portion of the heart is situated in the right hemithorax with base to apex directed to the left. The latter condition is mostly due to associated lung or diaphragm pathology (Fig. 1B).[3] With improvements in surgical techniques, 85% of children born with congenital heart disease reach their adulthood. Transesophageal echocardiography (TEE) is one of the definitive modalities to image the associated cardiac anomalies in DC. When these children/adults are planned for surgery, a systematic approach should be followed to delineate the anatomy and associated abnormalities. The omniplane angles need to be modified while performing TEE in these patients.

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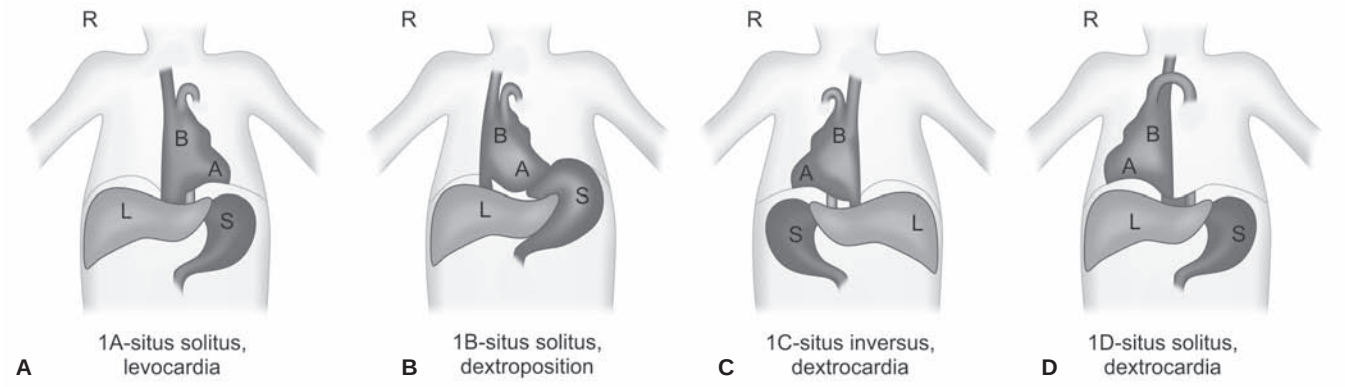
PATHOGENESIS AND ITS CLINICAL IMPORTANCE

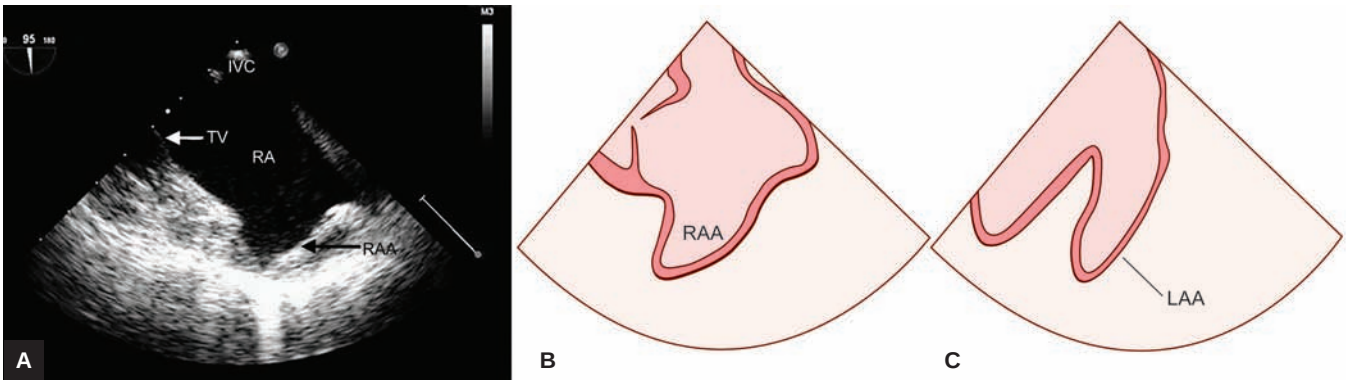
Van Praagh[4] in his study described a segmental approach in the evaluation of a child with congenital heart disease, in which the heart is divided into three segments—visceroatrial situs, ventricular loop, and position of the great vessels.

The term situs refers to the organ configuration within the body, mainly abdomen and thorax, based on which an individual can have any one of the configurations—SS, situs inversus (SI; mirror image of SS), or situs ambiguous. The position of the heart is evaluated following situs evaluation. With levocardia (LC) being the normal position of the heart (Fig. 1A), malposition can be either mesocardia or DC. The presence of DC in a patient should raise the suspicion of associated cardiac and noncardiac anomalies. Dextrocardia can be associated with any situs (Figs 1C and D) and the incidence of cardiac malformation associated with it varies with the situs, ranging from 3 to 5% (SI, DC) to 95% (SS, DC).[5]

Situs solitus with DC is associated with other lesions like anomalous pulmonary venous connections, patent ductus arteriosus, tetralogy of Fallot, coarctation of aorta, transposition of the great arteries (TGA) corrected/uncorrected, and septal defects.[6-8] Situs inversus with DC has a lower incidence of associated cardiac anomalies, which can be TGA corrected/uncorrected, ventricular septal defect, or double outlet right ventricle.[9] Patients with DC and situs ambiguous always have associated cardiac anomalies, which may be classified based on the type of atrial isomerism.[2] Patients with left atrial isomerism typically have pulmonary venous confluence returning close to the midline of an often-common atrium, atrioventricular (AV) valve with AV canal type defects, left-sided obstructive lesions like aortic stenosis and coarctation, and interrupted inferior vena cava (IVC). Patients with right atrial isomerism have bilateral SVCs, total anomalous pulmonary venous return, common AV valves with AV canal type defects, and right-sided obstructive lesions like pulmonary atresia. Both types have risk of arrhythmia and conduction abnormalities, with left atrial isomeric patients having greater risk of sinus node dysfunction.



Figs 1A to D: (A) Normal subject (SS, LC); (B) dextroposition. Note the position of the major portion of the heart in the right hemithorax. The position of the abdominal viscera and base to apex direction of the heart are still maintained (noncardiac pathology); (C) situs inversus, DC which is the mirror image of normal; and (D) SS, DC. Note the normal position of the abdominal organs. Also note the left to right—base to apex orientation of the heart. R: Right side; B: Base of the heart; A: Apex of the heart; L: Liver; S: Stomach



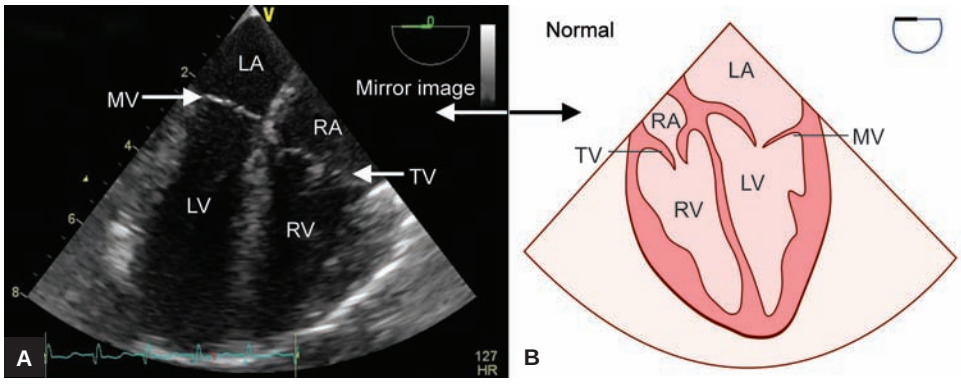
Figs 2A to C: (A) Probe in the lower esophageal position showing the junction of IVC and the right atrium. Also note the broad-based appendage of the right atrium; (B) TEE schematic of broad-based right atrial appendage; (C) TEE schematic of narrow finger-like left atrial appendage. RA: Right atrium; TV: Tricuspid valve

ROLE OF TEE IN APPROACH

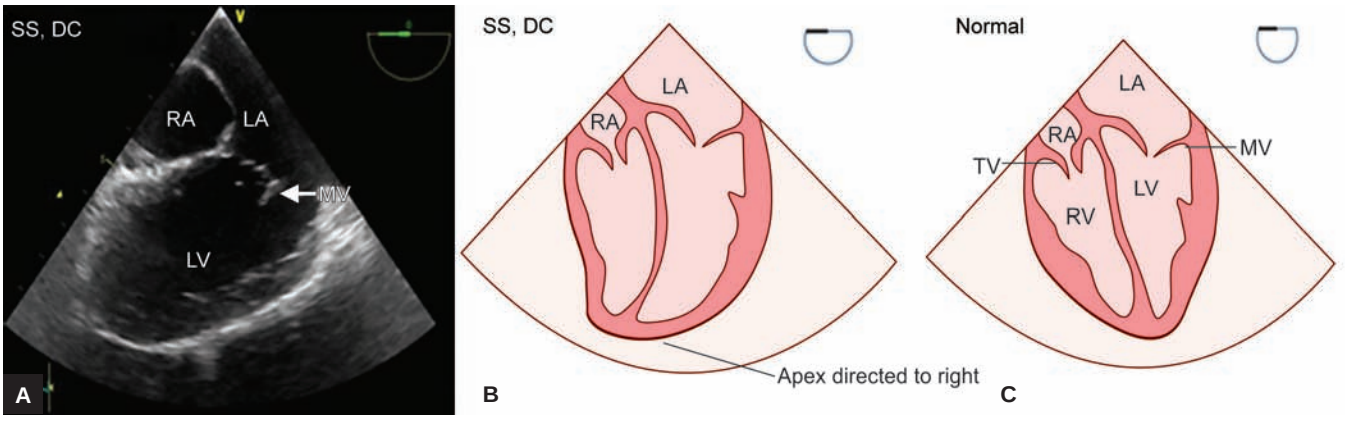
Physical examination like palpation, auscultation of the cardiothoracic system, and electrocardiography will give clues regarding the cardiac position. The common finding in SI, DC is marked right-axis deviation of the P wave (negative in aVL and lead I) and of the QRS complex, and low voltage in the precordial leads, V4 through V6. The trace of lead aVR in SI, DC is similar to lead aVL of SS, LC.[10] Echocardiography finds its role in confirming the diagnosis and evaluating the other associated cardiac anomalies in these patients.

Situs determination is fundamental in patients with congenital heart disease.[11,12] Because of the uncertain nature of the systemic and pulmonary venous drainage and subtle anatomical characteristics of the crista terminalis and AV valves in congenital heart disease patients, situs should be determined solely based on the atrial appendage. The atrial appendage maintains its characteristics even in cases of atrial enlargement due to shunt lesions.[13] Transthoracic echocardiography can be used to image the atrial morphology and the situs in children, whereas it is often difficult to do the same in adults.[14] The TEE, because of its close proximity to the atria, provides clear details of both the appendages and helps in determining situs. The right atrial appendage (RAA) is short with blunt end and smooth inner surface. The left atrial appendage (LAA) is long and narrow with crenelated cavity (Figs 2B and C). With the probe in the lower esophageal position, rotating the probe to the right brings the liver and short axis of the IVC into view in SS. Slight withdrawal, anteflexion, and increasing the omniplane angle to around 90° help visualize the entry of IVC into the right atrium (Fig. 2A). This view can also be obtained by advancing the probe from the bicaval position. The right atrial appendage can also be visualized in this view.

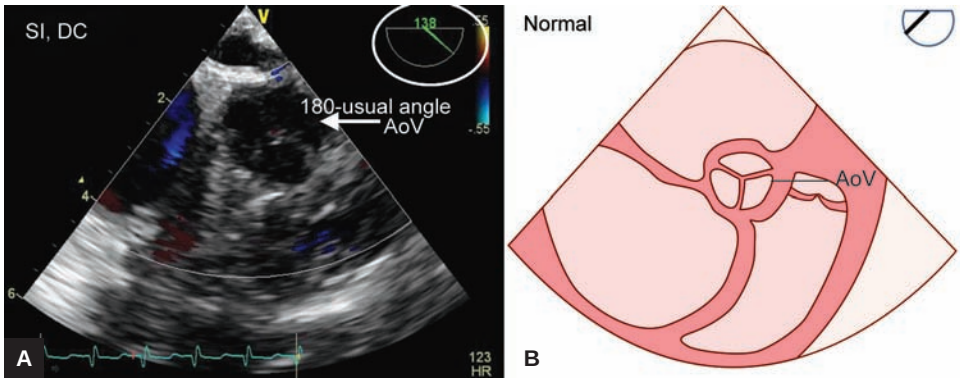
Apart from determining the situs, TEE helps in finding the associated anomalies. However, use of standard multiplane angles for TEE recommended in SS, LC patients will not image the desired views in cardiac malposition. The midesophageal four-chamber view of SI, DC is a mirror image of the view conventionally obtained in a normally positioned heart, with the right atrium on the right side of the screen (Fig. 3 and Video 1).



Figs 3A and B: (A) Midesophageal four-chamber view of SI, DC, tetralogy of Fallot (postrepair) subject, which is a mirror image of that obtained in a normal subject. (B) Midesophageal four-chamber view of a normal subject. LA: Left atrium; RA: Right atrium; MV: Mitral valve; TV: Tricuspid valve; LV: Left ventricle; RV: Right ventricle



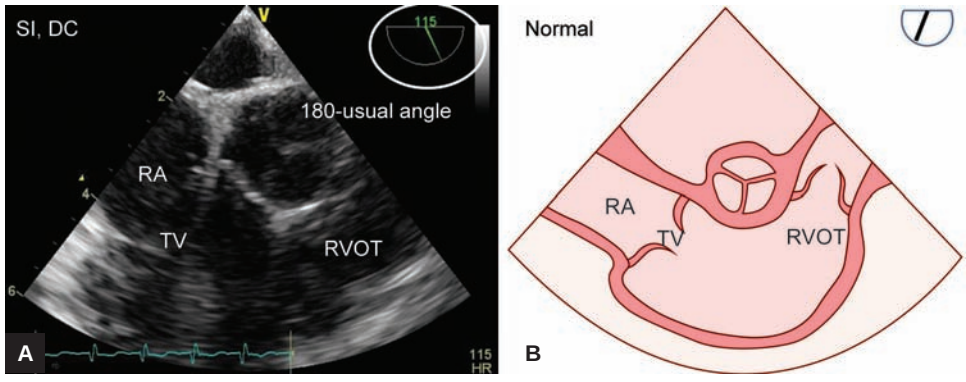
Figs 4A to C: (A) Midesophageal view of a patient with SS, DC, tricuspid atresia. Note that the right atrium is on the left side of the screen as in a normal subject. Right ventricle is hypoplastic; (B) TEE schematic of four-chamber view in SS, DC. Note the base to apex direction. The ventricles may be concordant or discordant; (C) TEE schematic of a midesophageal four-chamber view in a normal subject. RA: Right atrium; LA: Left atrium; MV: Mitral valve; LV: Left ventricle; RV: Right ventricle



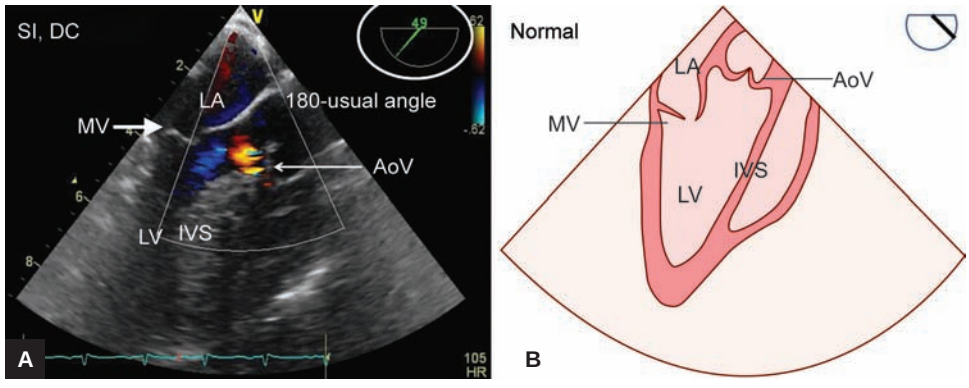
Figs 5A and B: (A) Midesophageal aortic valve short-axis view of SI, DC, tetralogy of Fallot (postrepair) subject obtained at an omniplane angle between 135 and 155°. Image is similar to that obtained in a normal subject; and (B) midesophageal aortic valve short axis view of normal subject—note the difference in omniplane angle. AoV: Aortic valve

However, in SS, DC, it is only the base to apex direction that is changed and the right atrium is visualized on the left side of the screen (Fig. 4 and Video 2).

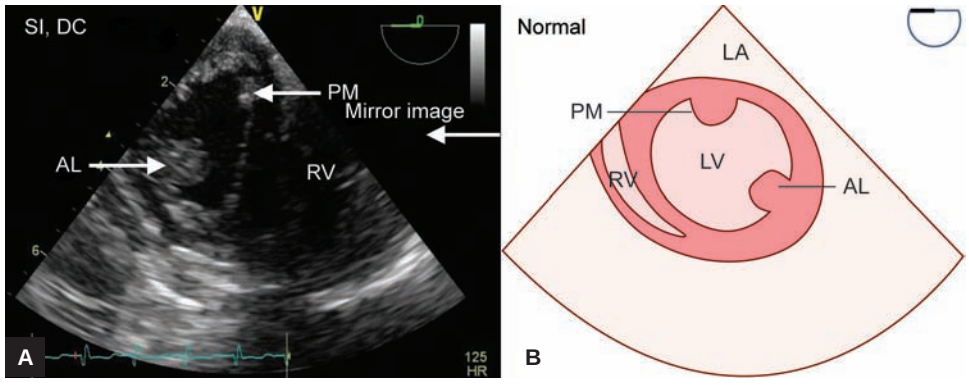
Other midesophageal views in SI, DC yield similar images to those obtained in normal cardiac position when the omniplane angles are at 180 minus the prescribed angle for the standard view (Figs 5 to 7 and Videos 3 to 5). This may not be possible in SS, DC because the axes of the TEE planes and the axis of the heart are no longer parallel to each other, as depicted in Figure 4. Hence, the other standard midesophageal views cannot be obtained.



Figs 6A and B: (A) Midesophageal right ventricle inflow–outflow view in SI, DC, tetralogy of Fallot (postrepair) subject obtained at an omniplane angle between 110 and 130°. Image is similar to that obtained in a normal subject; and (B) midesophageal right ventricle inflow–outflow view of normal subject—note the difference in omiplane angle. RA: Right atrium; TV: Tricuspid valve; RVOT: Right ventricle outflow tract

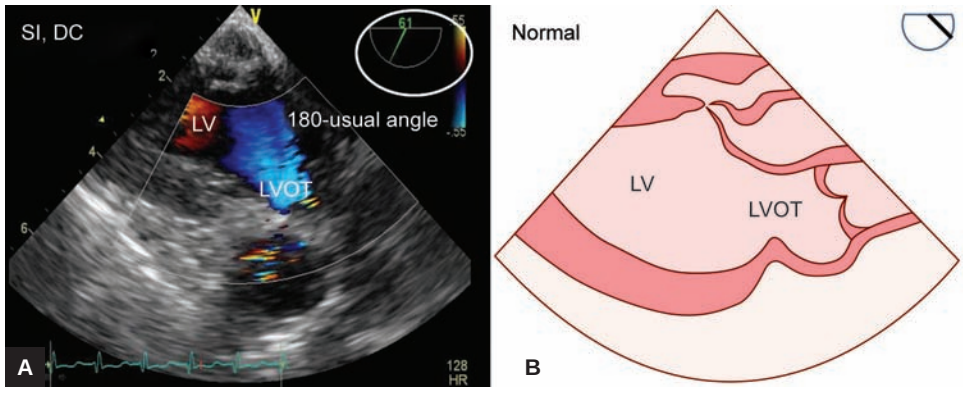


Figs 7A and B: (A) Midesophageal long-axis view of a SI, DC, tetralogy of Fallot subject obtained at an omniplane angle between 40 and 60°. Image is similar to that of a normal subject; and (B) Midesophageal long-axis view in a normal subject—note the difference in omiplane angle. LA: Left atrium; MV: Mitral valve; AoV: Aortic valve; LV: Left ventricle; IVS: Interventricular septum

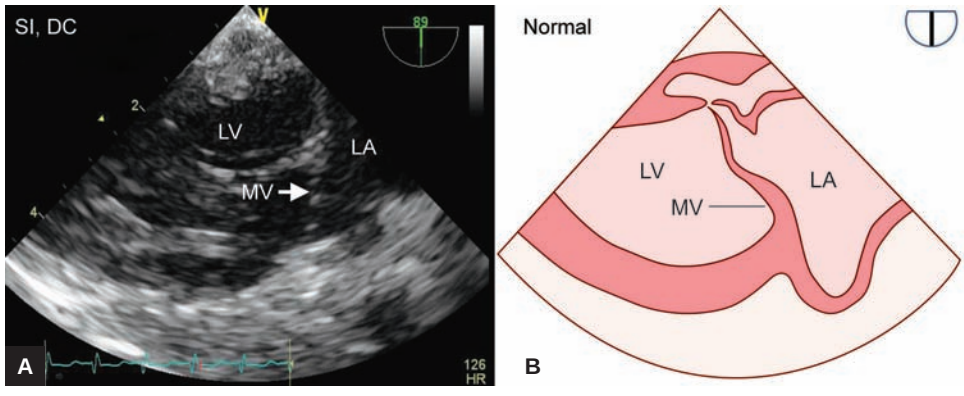


Figs 8A and B: (A) Transgastric midpapillary short-axis view of a SI, DC, tetralogy of Fallot patient, which is a mirror image of that obtained in a normal subject; and (B) Transgastric midpapillary short-axis view in a normal subject. LV: Left ventricle; PM: Posteromedial papillary muscle; AL: Anterolateral papillary muscle; RV: Right ventricle

Similar principles apply to imaging in the transgastric view in a SI, DC patient, with mirror image of the normal obtained at 0° (Fig. 8 and Video 6) and other standard images at 180 minus the standard angle (Figs 9 and 10, Videos 7 and 8). The presence of stomach and heart on opposite sides of the midline precludes much of the transgastric imaging in SS, DC. In SI, DC, three-dimensional image can also be obtained from the two-dimensional image, the only difference being that the left–right orientation on the screen will be reversed.



Figs 9A and B: (A) Transgastric long-axis view of a SI, DC, tetralogy of Fallot (postrepair) patient obtained at an angle between 40 and 60°. Image is similar to that obtained in a normal subject; and (B) Transgastric long-axis view in a normal subject—note the difference in angle. LV: Left ventricle; LVOT: Left ventricle outflow tract



Figs 10A and B: (A) Transgastric two-chamber view of a SI, DC, tetralogy of Fallot (postrepair) patient. Note that the image and angle are similar to that of a normal subject.(B) Transgastric two-chamber view in a normal subject. LA: Left atrium; LV: Left ventricle; MV: Mitral valve

CONCLUSION

Dextrocardia is a rare cardiac anomaly that can be associated with other cardiac and noncardiac abnormalities. It is pertinent upon the perioperative echocardiographer to be aware of the anomalies associated with DC, the maneuvers by which the views are obtained, and the situations when the images cannot be obtained.

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