Congenitally Corrected Transposition of the Great Arteries (CCTGA)

31

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Abstract

Echocardiographic evaluation of patients with CCTGA (also called LTGA) requires precise segmental approach. In 90% of patients, CCTGA is associated with other congenital heart defects, VSD, LV outflow tract obstruction, and apical displacement of the septal and inferior leaflets of tricuspid valve (Ebstein anomaly), as well as conduction system dysfunction. Prognosis is determined not only by associated anomalies, but also by systemic pressure effect on RV function. In the absence of other anomalies, diagnosis may be delayed. Systemic ventricular failure, and arrhythmias may be the presenting symptoms in isolated CCTGA.

Congenitally Corrected Transposition of the Great Arteries (CCTGA)

1. Remember that subcostal coronal view and apical four chamber view provide the most diagnostic findings. Define Morphologic right atrium (RA), left atrium (LA),right ventricle (RV) and left ventricle (LV) based on their echocardiographic features and evaluate discordant connection of atria and ventricles, atrioventricular (AV) discordance: RA to morphologic LV and LA to morphologic RV (Fig. 31.1).
2. Demonstrate discordant connection of ventricles and great arteries, ventriculoarterial (VA) discordance: LV to pulmonary artery (PA) and RV to aorta (AO) (Figs. 31.2 and 31.3).

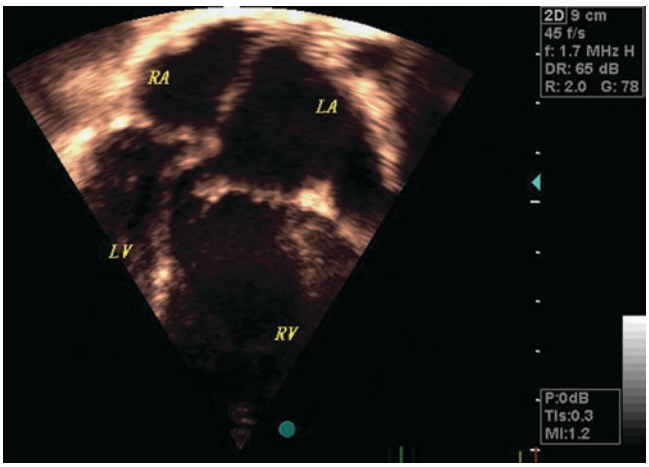

Fig. 31.1 Transthoracic apical four chamber view showing AV discordance. *RA* right atrium, *LA* left atrium, *RV* right ventricle, *LV* left ventricle

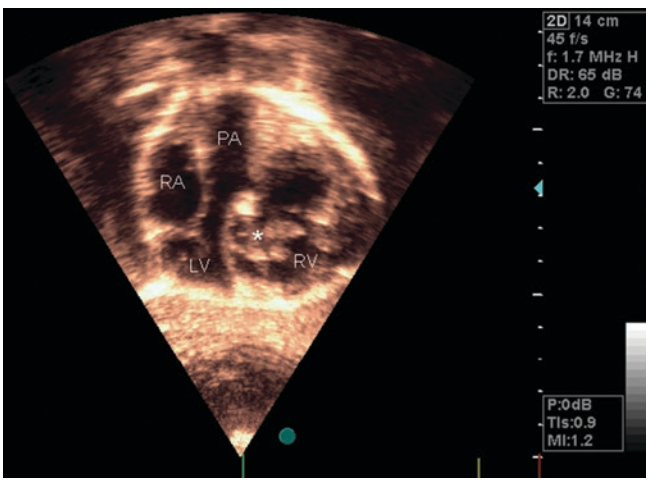

Fig. 31.2 Subcostal coronal view showing PA arising from morphologic LV. Asterisk indicates tricuspid valve in systole. *RA* right atrium, *LV* left ventricle, *PA* pulmonary artery, *RV* right ventricle

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M. Moradian, A. Alizadehasl (eds.), *Atlas of Echocardiography in Pediatrics and Congenital Heart Diseases*,
https://doi.org/10.1007/978-3-662-62341-1_31

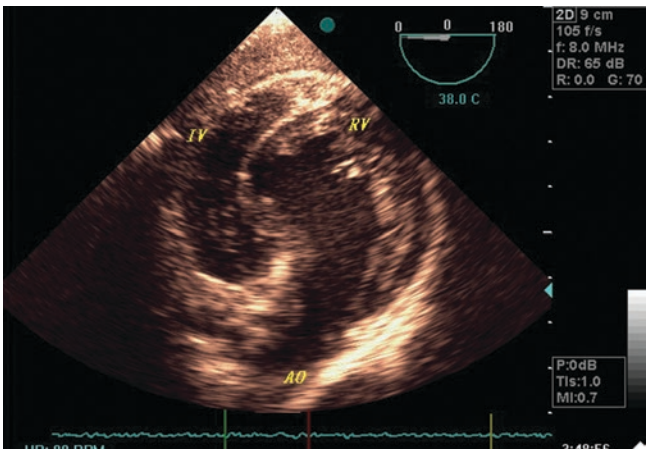

Fig. 31.3 Transesophageal echocardiography (TEE), deep trans gastric view showing origination of aorta from morphologic right ventricle. *LV* left ventricle, *RV* right ventricle, *AO* aorta

3. In some cases with superior-inferior arrangement of ventricles imaging both atrioventricular valves in a same plane are not possible and tilting the transducer is necessary.
4. Identify parallel orientation of great arteries in different views including subcostal and parasternal views also evaluate the left and anterior location of aortic valve (Fig. 31.4a, b).
5. Appreciate for common associated anomalies including ventricular septal defect (VSD), left ventricular out flow tract obstruction (LVOTO), pulmonary stenosis (PS), Ebstein malformation of tricuspid valve, or any other lesion. Also use color flow Doppler to evaluate any atrioventricular valve stenosis or regurgitation (Fig. 31.5a, b).
6. Standard parasternal long axis view in these patients is confusing because interventricular septum is vertical, ventricles are almost side by side, and great arteries are

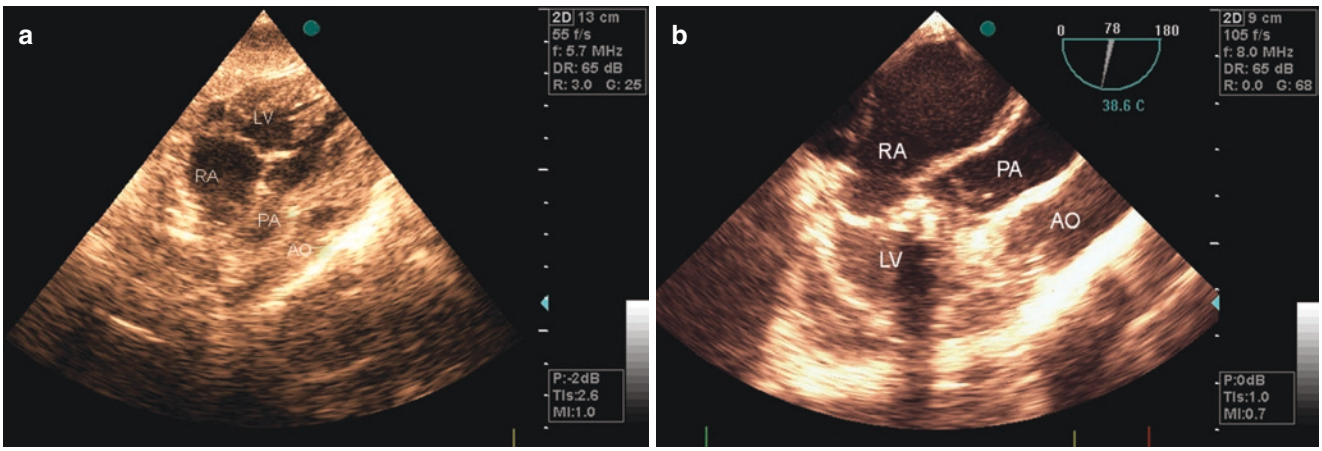

Fig. 31.4 (**a**, **b**) Parallel orientation of great arteries is shown in TTE, subcostal coronal view (**a**). Anteriorly located aorta and parallel arrangement of great arteries is shown in this transesophageal echocardiography of a patient with CCTGA at in flow-outflow view (**b**). *LV* left ventricle, *RA* right atrium, *AO* aorta, *PA* pulmonary artery

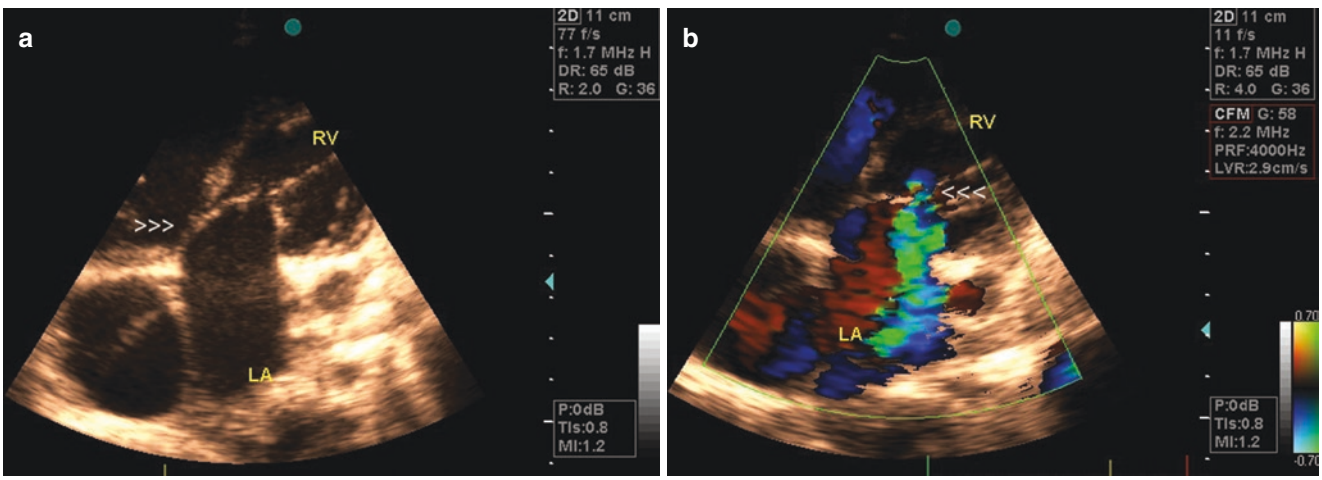

Fig. 31.5 (**a**, **b**) Apical four chamber view of a patient with CCTGA and Ebstein malformation of tricuspid valve (TV) (left AV valve). Arrow heads in figure (**a**) indicate the displacement of septal leaflet of TV. Color Doppler interrogation of the same patient shows severe tricuspid regurgitation (TR) in figure (**b**). Arrow heads point to the origin of TR jet (**b**). *LV* left ventricle, *RA* right atrium

parallel in CCTGA. So, more manipulation of the transducer is necessary

7. Because the course of ascending aorta in CCTGA is straight and leftward, aortic arch in these patients is better evaluated in high parasternal position, so called ductal view, than standard suprasternal view (Fig. 31.6).
8. Evaluate the coronary arteries in parasternal short axis view. Coronary arteries distribution in CCTGA is concordant with ventricular anatomy, the so called "coronary artery–ventricular concordance" which is the mirror-image of normal coronary distribution. The most common coronary anomaly in CCTGA is single coronary arteries.
9. During fetal life this anomaly should be ruled out in cases who are referred for evaluation of fetal bradycardia, because CCTGA may lead to complete heart block (CHB) even in fetus (Fig. 31.7)

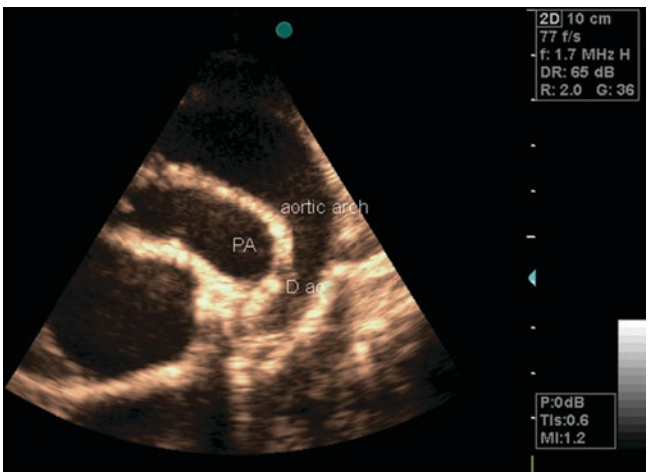

Fig. 31.6 Left high parasternal view showing left sided aortic arch, left anterior aorta in this case of CCTGA. *PA* pulmonary artery, *DAO* descending aorta

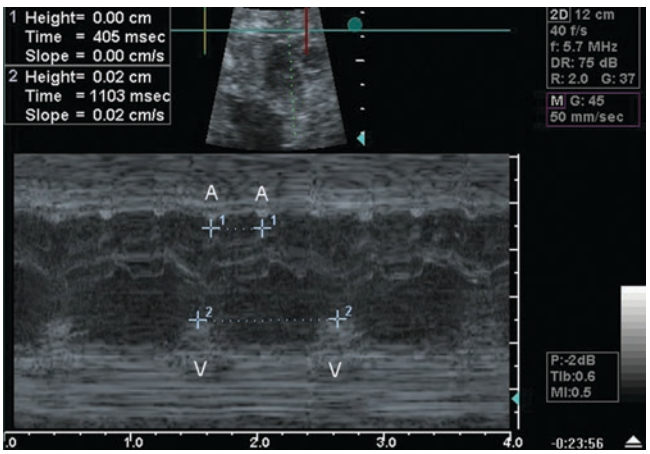

Fig. 31.7 Fetal echocardiogram using M mode to evaluate atrial and ventricular wall motion in a fetus with complete heart block (CHB). The mechanical A to A interval corresponding to electrical PP interval, is 405 milliseconds (ms) so atrial rate is 148 beat per minute (bpm), and V to V interval corresponding electrical RR interval is 1103 ms, predicting ventricular rate 54 bpm, and there is no electrical association between atria and ventricles

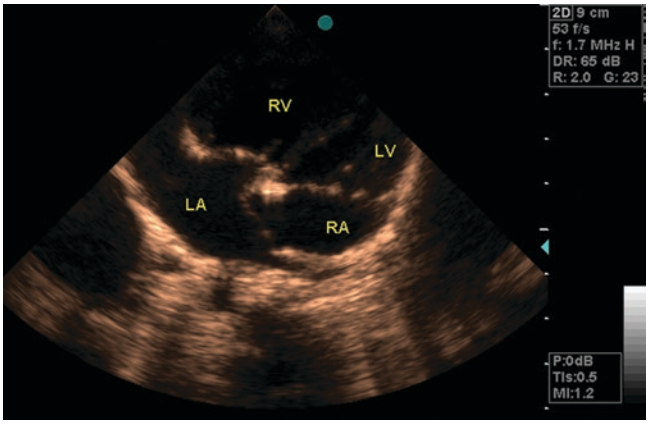

Fig. 31.8 Apical four chamber view in a patient with situs inversus, AV and VA discordance. *RA* right atrium, *LA* left atrium, *RV* right ventricle, *LV* left ventricle

10. Congenitally corrected transposition of the great arteries may be seen in with situs inversus, dextrocardia and/or mesocardia (Fig. 31.8).
11. Very important in these patients is the evaluation of RV function as systemic ventricle. Serial evaluation RV function using fractional area change, tricuspid annular excursion (TAPSE), tissue Doppler and strain imaging are useful.

Suggested Reading

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