Case Report

Dextrocardia, situs solitus, double–inlet, and double-outlet left ventricle – A case report

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ABSTRACT

Dextrocardia is embryologic malformation characterized by displacement of the largest axis of the heart to the right side of the chest. Dextrocardia is a rare anomaly that is associated with situs inversus and the incidence in the general population is usually 1:10,000, with situs solitus being 1:30,000 live births. It is associated with complex cardiac lesions, 5% with situs inversus, and 90% with situs solitus color. Doppler echocardiography not only provides additional information but also improves overall specificity of two-dimensional echocardiography. Few case reports of dextrocardia, double-inlet (DILV), and double-outlet left ventricle (DOLV) with situs inversus have been diagnosed and reported by echocardiography, but none are available with situs solitus. To the best of our knowledge, we are presenting the first case report of dextrocardia, situs solitus, double-inlet with DILV, DOLV, and D-malposition of great arteries with pulmonary stenosis, detected by color Doppler echocardiography.

Key words: Dextrocardia with situs solitus, Double-inlet left ventricle, Double-inlet–double-outlet left ventricle, Double-outlet left ventricle

Dextrocardia is an embryologic malformation characterized by the displacement of the largest axis (base to apex) of the heart to the right side of the chest, with reversion of the apical inclination. The malformation is caused by the anomalous rotation of the primitive heart tube, leading to bending of bulboventricular loop to the left side, around the 8th week of embryonic life [1,2].

The incidence of dextrocardia associated with situs inversus, in the general population, is usually 1:10,000, with situs solitus being 1:30,000 live births, and only 1:900,000 in the adult population [3,4]. Dextrocardia with situs solitus (Figs. 1 and 2) is intricately involved in numerous extracardiac lesions such as tracheoesophageal fistula, pulmonary hypoplasia, imperforate anus, spina bifida, and Kartegener syndrome [3-5] and cardiac abnormalities such as ventricle septal defect, atrial septal defect, transposition of great arteries, double-inlet left ventricle (DILV) (Fig. 3), double-outlet right ventricle, double-outlet left ventricle (DOLV) (Fig. 4), AV septal defects, pulmonary, mitral and tricuspid atresia.

Van Praagh *et al*. [5] described and classified in detail the different types of dextrocardia. The arrangement of the position of the abdominal viscera in dextrocardia may be normal (situs solitus), reversed (Situs inversus), and indeterminate (situs ambiguous) in 32–35%, 35–39%, and 26–28% of cases, respectively [6]. However, the atrial situs always corresponds to the visceral situs [6]. Dextrocardia with situs solitus accounts for about 75% of cases of dextrocardia [7] and is associated with complex cardiac lesions, the frequency of which varies considerably according to situs; 5% with situs inversus to 90% with situs solitus [8].

Echocardiography has substantially expanded the horizons for accurate non-invasive diagnosis of congenital cardiac defects including complex congenital heart diseases. Color Doppler echocardiography [9] not only provides additional information but also improves overall specificity of two-dimensional (2-D) echocardiography. Few case reports of dextrocardia, DILV, and DOLV in situs inversus have been diagnosed and reported by echocardiography, in the past [10,11], and despite our deep search of the literature, we could not encounter any article of such anomaly, in situs solitus. To the best of our knowledge, we are presenting the first case report of dextrocardia with situs solitus, DILV, DOLV, D-malposition of great arteries, and pulmonary stenosis, detected by color Doppler echocardiography.

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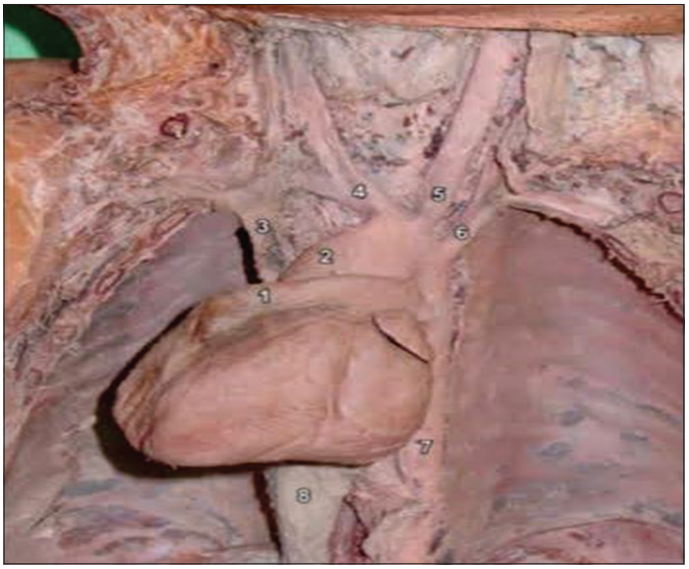

Figure 1: Position of heart in the thorax: (1) pulmonary trunk; (2) ascending aorta; (3) superior vena cava; (4) brachiocephalic trunk; (5) left common carotid artery; (6) left subclavian artery; (7) thoracic aorta; and (8) hepatic tissue covering the inferior vena cavaDextrocardia (Anatomical Specimen)

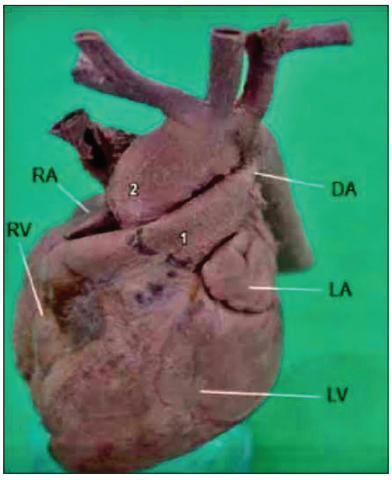

Figure 2: Anatomical specimen – dextrocardia with situs solitus (1) pulmonary artery and (2) aorta and arch vessels

CASE REPORT

A 13-year-old female child was referred to us from a private pediatric facility for a detailed color Doppler echocardiographic evaluation, for attainment of clinical diagnosis of her complex cyanotic congenital cardiac disease so that the ideal management techniques could be accordingly advised. It is worth mentioning that no cardiac intervention was provided to the ailing child from birth till date. The child was a full term normal delivery from a multipara woman of 34 years of age. She was delivered at a private hospital, and at birth was having cyanosis and low birth weight. There was no history of maternal risk factor of congenital heart disease (morbid obesity, diabetes, febrile illness, smoking alcohol intake, teratogenic drug, and radiation exposure). Presently, she was complaining of shortness of breath on minimal effort, weakness, lethargy, blue coloration of lips, tongue, and

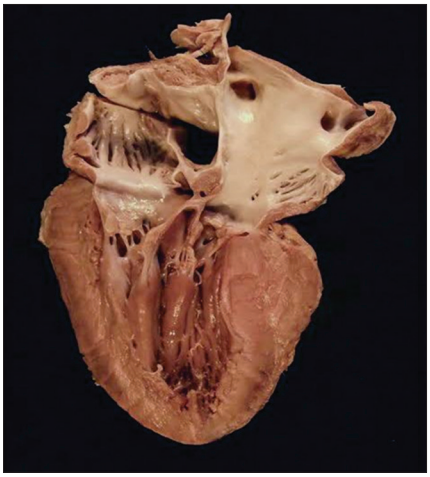

Figure 3: Anatomical specimen of double-inlet left ventricle: both the atria drain through two respective atrioventricular valves into a posterior ventricular chamber of left ventricle morphology

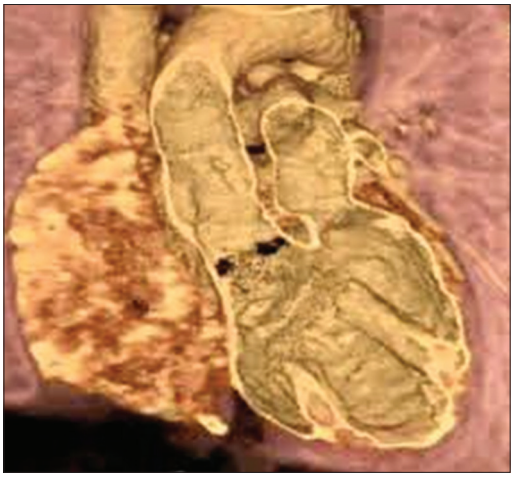

Figure 4: Multilplanar 3D reconstruction of computed tomography angiography. The hollow parts are the left ventricle chamber, with Ao on the right and posteroanterior on the left

nails. There was no history of syncope, chest retractions, or congestive heart failure (CHF). On clinical examination, the child was having a weight of 18 kg, was very thin with clubbing, involving all fingers of the upper and lower extremities, along with the presence of deep central cyanosis. Her vital signs were HR 93/min, BP 120/86 mmHg, RR: 30/min, and SpO_2 77% at room air. No overt clinical signs of CHF were detected. On cardiovascular examination, there was Grade 2/6 ejection systolic murmur in pulmonary area with normal second heart sound. The pulses were symmetric and palpable. Chest X-Ray showed dextrocardia, situs solitus, mild cardiomegaly with the presence of wiry and slender main, and branch pulmonary arteries (Fig. 5).

Color echocardiography was technically challenging and was done by the author himself, with patient lying in the unorthodox, right lateral decubitus position, and the transducer placed on the right chest and maneuvered gently by the left hand of the operator, to obtain optimum echocardiographic images. There was dextrocardia, situs solitus (Fig. 6), left aortic arch (Fig. 7), intact atrial septum with superior vena cava, and inferior vena cava draining into the right atrium and four pulmonary veins

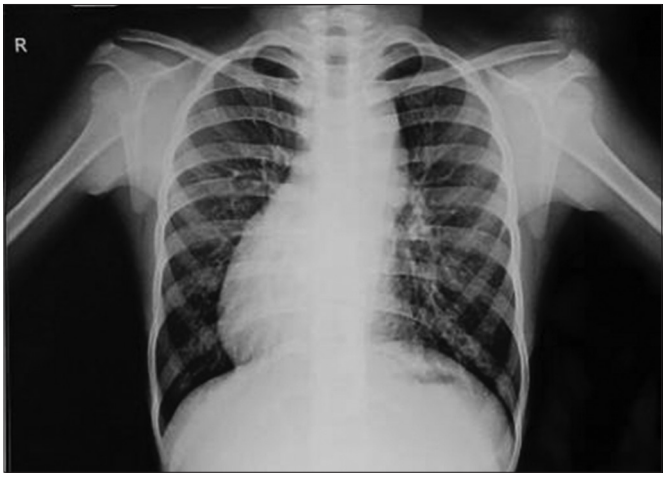

Figure 5: X-ray chest posteroanterior: Dextrocardia, situs solitus, cardiomegaly, thinned out main, and branch pulmonary arteries

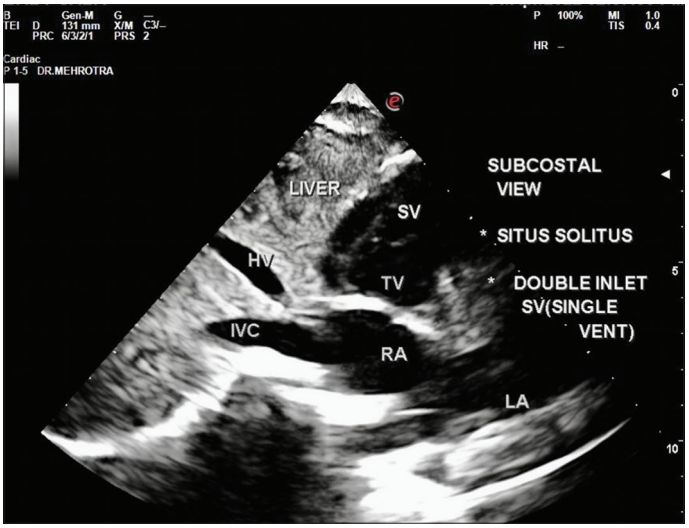

Figure 6: Situs solitus, double-inlet left ventricle

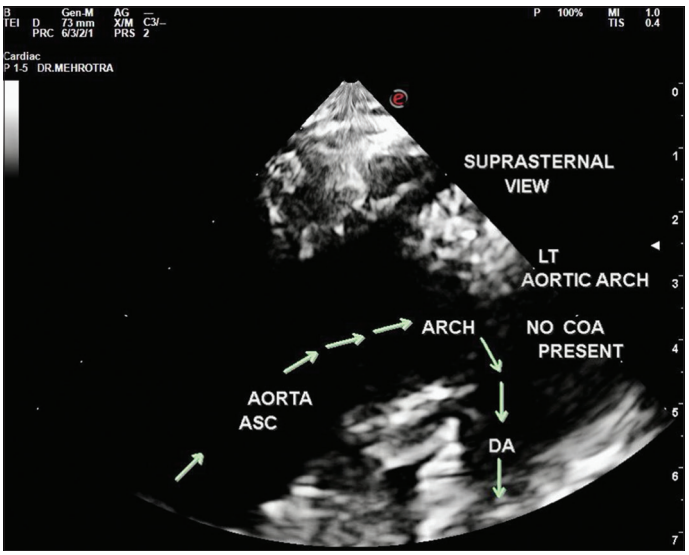

Figure 7: Left Aortic Arch (No COA/PDA)

draining into the left atrium. Both mitral and tricuspid valve were at the same level and were opening into single ventricle (SV) of the left ventricle morphology (DILV) (Fig. 8). Their respective

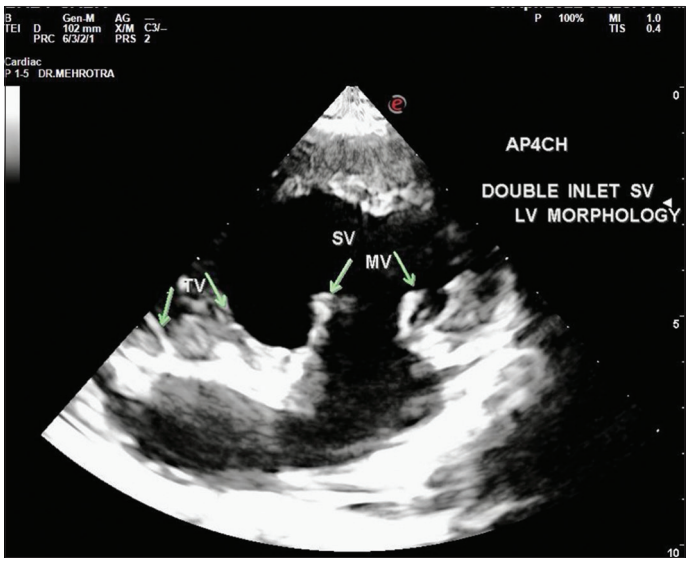

Figure 8: Subcostal View: Situs solitus, double-inlet single ventricle of left ventricle morphology

chordae and papillary muscles were attached to the lateral walls of the SV. The ventricular septum was absent and a small rudimentary RV was identified as a blind pouch, positioned on the anterior superior shoulder of SV (Figs. 9 and 10). The SV walls were smooth and finely trabeculated. Both the great arteries were arising from SV (DOLV) (Fig. 11), in a D-malposition arrangement, with aorta being anterior and to the right and pulmonary artery being posterior and to the left. The aorta is dilated (annulus diameter 19.1 mm) and the infundibulum was recognized with its attachment to confluent pulmonary arteries. The pulmonary valve was domed and there was severe pulmonary valvular stenosis with marked overgrowth of muscles in the infundibular region causing moderate infundibular obstruction (peak and mean gradient was 43.8/20.3 mmHg and 93.3/64.9 mmHg across infundibular region and pulmonary valve, respectively) (Fig. 12). The main and branch pulmonary arteries are hypoplastic (pulmonary valve annulus [D] 5.00 mm, main pulmonary artery [D] 4.70 mm, left pulmonary artery [D] 6.40 mm, and right pulmonary artery [D] 5.60 mm). The SV was dilated with normal systolic function – EF 61%.

Once the comprehensive echocardiographic diagnosis was accomplished, the patient was transferred to a tertiary care facility for a probable Fontan surgery.

DISCUSSION

In an embryo, the heart is first organ to develop, which develops from an embryonic heart tube-formed by fusion of the endocardial tubes. After the formation of the heart tube, position of ventricle in relation to atria is determined by looping of the heart tube. It may loop to the left (L-loop), or loop to the right (D-loop). With L-loop, the morphological right ventricle is positioned to the left of the left ventricle [12]. If a D-bulboventricular loop fails to migrate into the left hemithorax, with the heart in the right hemithorax, it can result in dextrocardia. In most cases,

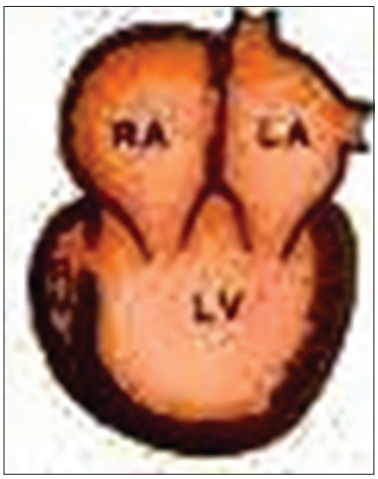

Figure 9: Diagrammatic presentation of double-inlet left ventricle. Both atrioventricular valves are opening into single ventricle of LV morphology

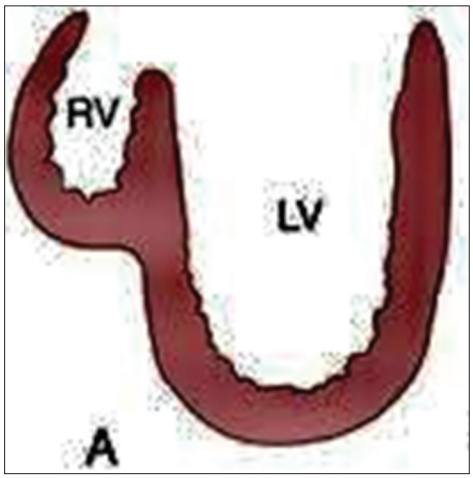

Figure 10: Diagrammatic presentation of double-inlet left ventricle with a characteristic small pouch of rudimentary RV

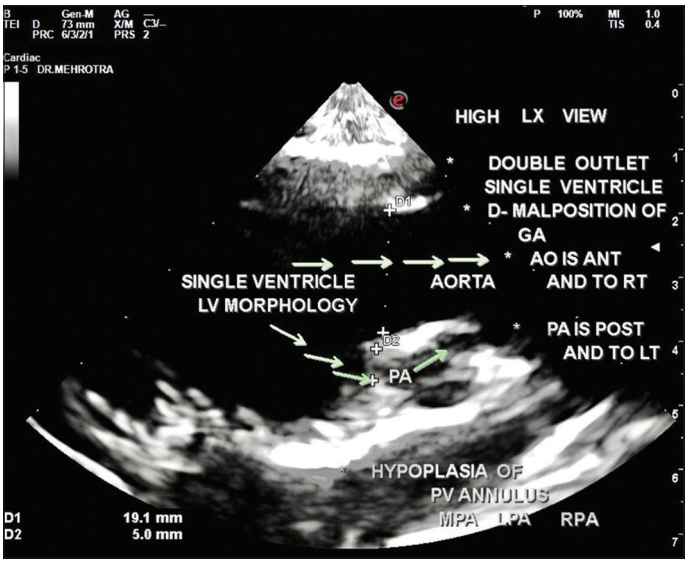

Figure 11: High LAX View: (1) Double-outlet left ventricle, (2) D-Malposition of Great Arteries – Ao is anterior and Pa is posterior, and (3) Hypoplasia of PV annulus and branch pulmonary arteries

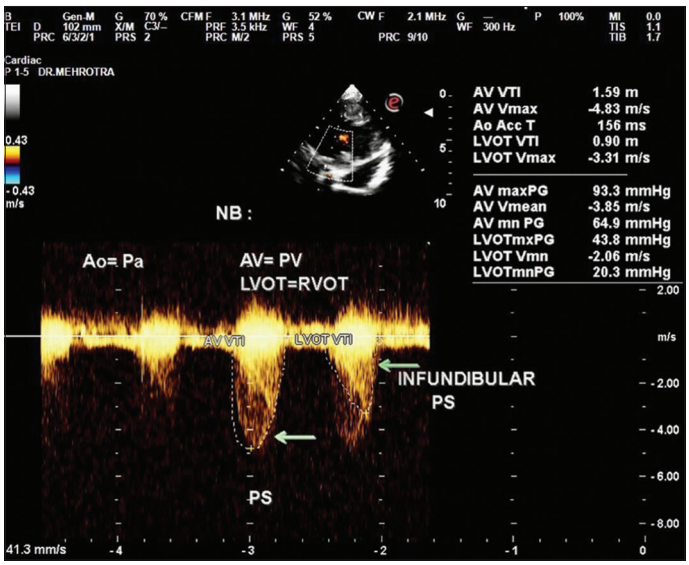

Figure 12: CW Doppler across RVOT – depicting severe pulmonary valvular and moderate infundibular stenosis

it is diagnosed incidentally. Some development anomalies are often associated with this. Dextrocardia can be accompanied by a reversal in the position of other organs which are termed as situs inversus totalis [13]. When there is association of dextrocardia with a normal position of other thoracoabdominal structures, it is termed as situs solitus.

DILV is a condition, where the atrioventricular valve is connected to a main single ventricular chamber. Eighty percent of double-inlet ventricles are DILV, where both left and right atrium are connected to the left ventricle (main SV) through mitral and tricuspid valve, respectively [14]. The right ventricle is underdeveloped/hypoplastic. Usually, the condition is combined with ventricular septal defect [15].

Many affected patients have associated defects in the heart or main arteries [16]. Eighty-five of patients of DILV are associated with transposition of great arteries, where aorta arises from rudimentary right ventricle and pulmonary artery from the left ventricle [14]. However, in our case, we found malposition of great arteries, whereby both aorta and pulmonary arteries arose from the left ventricle. This configuration is also known as DOLV. Double-outlet ventricle is an uncommon cardiac anomaly and comprises 1% of congenital heart disease. Meanwhile, DOLV accounts for less than 5% of double-outlet ventricle cases [17].

Since the well-documented case of DOLV by Sakakibara *et al.* [18], several cases have been reported, most of them summarized in the comprehensive review of pathological specimens of DOLV by Van Praagh *et al.*, [19] and the combination of DOLV and DILV, in situs solitus, was observed in only three of 109 cases. Dextrocardia, DILV, and DOLV in situs inversus have been diagnosed and reported by echocardiography, in the past [10,11]. However, none have been reported in situs solitus by color Doppler echocardiography.

The strengths of our approach are organizing, urgently, a color Doppler echocardiography by an advanced four-dimensional XStrain Color Echocardiography system of Esaote, Italy,

echocardiography being carried out by in house, extremely experienced and skilled pediatric echocardiographer – the author himself and transferring of the patient to a tertiary care hospital, immediately, on accomplishing the diagnosis, for a probable suitable palliative surgery. Limitations of our approach are unorthodox positioning of the patient in the right lateral decubitus posture, while performing the color Doppler echocardiography and placement of the transducer on the right chest by the left hand of the operator, to obtain optimal quality images, was a challenging task and encountering of limited windows, angles, and depth of access, and moreover, many parts of right ventricle, pulmonary arteries, pulmonary veins, and aorta could not be interrogated, to the fullest satisfaction. This was highly technically demanding case due to the presence of dextrocardia and other complex cardiac malformations. The ideal technique in such a situation would have been a cardiac magnetic resonance imaging (MRI), in the support of diagnosis, because, currently, cardiac MRI is considered to be the gold standard for imaging in the assessment of congenital heart disease [20,21]. Our institution does not possess a MRI.

CONCLUSION

To the best of our knowledge, this patient represents the first reported case of dextrocardia, DILV, and DOLV in situs solitus with D-malposition of great arteries and pulmonary stenosis, detected by color Doppler echocardiography. The unusual features of this case are the rare segmental anatomy. Moreover, this was an exceptionally challenging case for the echocardiographer, and vigorous and painstaking efforts had to be employed to obtain optimal echocardiographic images. An experienced and skilled pediatric echocardiographer is a fundamental requirement to bring out a comprehensive diagnosis of complex cyanotic congenital heart disease, particularly with dextrocardia. The imaging should be performed utilizing a high end 3D/4D color echocardiography system for excellent resolution of images and furthermore the cardiac echo to be conducted in an unorthodox right lateral decubitus position, with the transducer placed on the right chest, in patients with dextrocardia.

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