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HOW I DO IT

Echocardiographic Assessment of Neonates with Congenital Heart Disease

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ABSTRACT

Echocardiographic imaging of neonates with congenital heart disease should be performed in a systematic manner, using the sequential segmental approach. The initial step involves determining the atrial arrangement, followed by assessment of the atrio-ventricular and ventriculo-arterial connections, and detection of any intracardiac or great arterial abnormalities. Finally, evaluation of systemic and pulmonary venous return to the cardiac chambers is undertaken. The identification of an obvious defect should not limit the systematic search in each segment of the heart for further abnormalities. Only by so doing can one avoid overlooking any potential lesions contributing to disturbed haemodynamic flow.

Key Words: *Doppler echocardiography, Echocardiography, Heart defects, congenital, Neonate*

INTRODUCTION

Congenital heart diseases presenting in the neonatal period are usually complex, often involving the chambers, great vessels, or veins. Conventionally, definitive diagnosis requires invasive cardiac catheterisation with cine-angiography. Recent advances in ultrasonic imaging allow a clear depiction of the detailed anatomy of the heart,[1] are non-invasive, and thus avoid many of the inherent risks associated with cardiac catheterisation.[2] Although magnetic resonance imaging is also non-invasive and can also reconstruct the cardiac anatomy 3-dimensionally,[3] the investigation requires the often-sick neonate to be placed in an enclosed chamber for a relatively long period. There is always the potential risk of deterioration during the investigation, which could be detrimental to the baby. Echocardiography thus remains the mainstay of diagnostic imaging for the sick neonate with a heart problem.

COMPLEMENTARY ROLE OF CLINICAL AND OTHER FINDINGS

In order to arrive at an accurate and definitive diagnosis by echocardiography, a thorough clinical examination followed by chest radiography and electrocardiography is recommended. Ultrasonic imaging should be guided by these initial clinical findings. For example, in the case of a cyanotic baby with an obvious heart murmur and an oligaemic lung field on chest radiography, the diagnosis of severe pulmonary outflow tract obstruction is suggested. Moreover, a simple chest radiograph shows the position of the heart within the thoracic cage; abnormal position is not uncommon in complex heart lesions. Echocardiography can then concentrate on demonstrating the relevant right heart anatomy.

SEQUENTIAL SEGMENTAL APPROACH TO ECHOCARDIOGRAPHIC ASSESSMENT

Symptomatic neonates with a congenital heart disease usually have abnormalities involving multiple sites. Thus, to avoid missing major lesions, it is prudent to perform echocardiography in a systematic manner, using the sequential segmental approach.[2,4] Much has been published on the logical sequence for performing the investigation.[2,5] The following description is my own approach to the task. The initial step involves determining the atrial arrangement, followed by assessment of the atrio-ventricular and ventriculo-arterial connections. The next major step is to look for any intracardiac or great arterial abnormalities. The final step involves ensuring that the systemic and pulmonary venous return are directed to the appropriate chambers.

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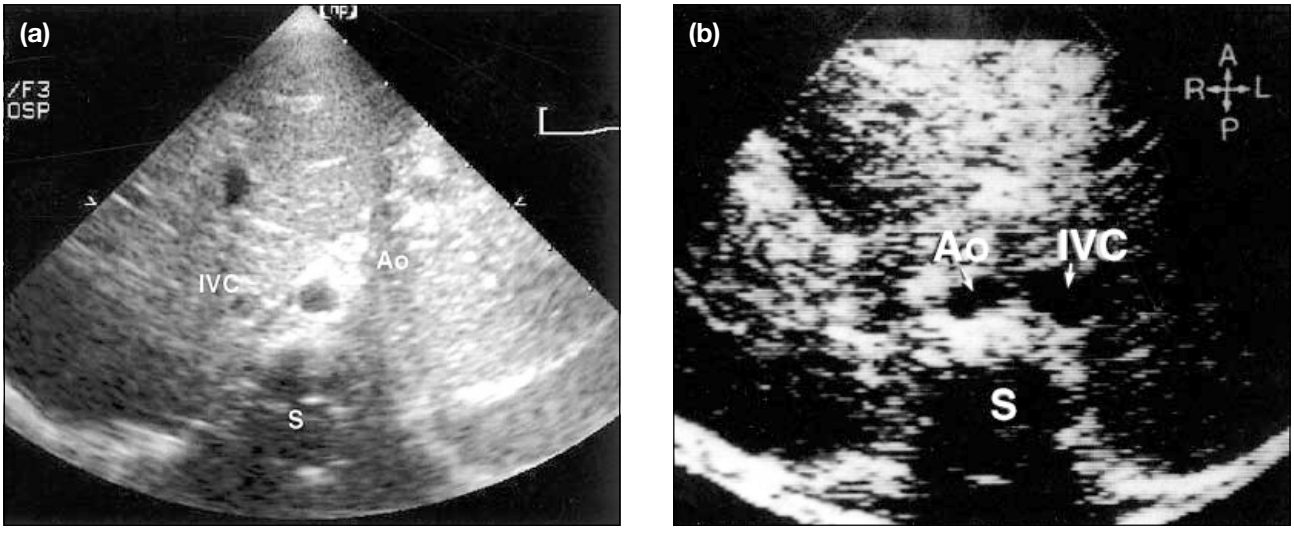

Figure 1. Atrial situs. (a) Cross-sectional view at the level of the 12th thoracic vertebra showing the inferior vena cava and the descending aorta on the right and left side of the vertebral body, respectively, inferring the usual atrial situs; (b) similar cross-sectional view showing the descending aorta to the right and the inferior vena cava to the left of the spine, inferring atrial situs inversus. The descending aorta can be recognized by its pulsatile nature. *Abbreviations:* Ao = aorta; IVC = inferior vena cava; S = spine.

Atrial Situs

To determine atrial arrangement, the subxiphoid cross-sectional and longitudinal cuts are utilised to view the position of the inferior vena cava and the descending aorta at the level of the 12th thoracic vertebra.[6] Normal situs arrangement is inferred from observing the inferior vena cava on the right and the descending aorta on the left of the vertebral body (Figure 1a). The inferior vena cava should be traced to empty into the right-sided atrium. Situs inversus is depicted by the opposite arrangement, with the inferior vena cava on the left and the descending aorta on the right of the vertebral body (Figure 1b). When the two vascular channels are parallel to one another on either side of the spine, right isomerism (a bilateral right atrial arrangement) is the most likely diagnosis.[6] In the situation of left isomerism (a bilateral left atrial arrangement), the inferior vena cava is interrupted and is replaced by the azygous vein posterior to the descending aorta, close to the vertebral body.[6] The presence of an atrial isomeric arrangement should alert the echocardiographer to look for complex heart abnormalities, such as atrioventricular septal defect and abnormal pulmonary or hepatic venous return.

Atrio-ventricular Connection and Intracardiac Abnormalities

Once the atrial arrangement is clarified, the next step is to define the connection between the atrium and ventricle. Using the subcostal and apical four-chamber views, one should be aware of the normal offsetting of the mitral and tricuspid valves with the tricuspid valve somewhat closer to the apex of the ventricles. Reverse of the offsetting in a heart with usual arrangement is indicative of atrio-ventricular discordance.[7] The degree of trabeculation within the two ventricles can provide further support for this diagnosis. The right ventricle would be heavily trabeculated and guarded by the tricuspid valve, which normally has septal insertion. Conversely, the relatively smooth left ventricle would be guarded by the mitral valve with parietal insertion. One should be aware that morphological and positioning anomalies do not necessarily concur. Hence in the setting of a normal atrial arrangement and discordant atrio-ventricular connection, the morphological right ventricle is on the left side and the morphological left ventricle on the right side. Alternatively, the atrio-ventricular connection can be univentricular (Figure 2). The mode of univentricular atrio-ventricular connection can be that of a double inlet via two separate valves, via a common atrio-ventricular valve(s), or with absent left or right atrio-ventricular connection (mitral or tricuspid atresia).

The subcostal and apical four-chamber views are also utilised to look for atrial, ventricular,[8] and atrio-ventricular septal defects[9,10] (Figure 3). Abnormalities of the atrio-ventricular valves are also best revealed by the apical four-chamber views. Mitral stenosis[11] and Ebstein's anomaly of the tricuspid valve[12] are examples of such valvular lesions.

Ventriculo-arterial Connection

Tilting the transducer anteriorly from the subxiphoid four-chamber view reveals the outflow tracts of the

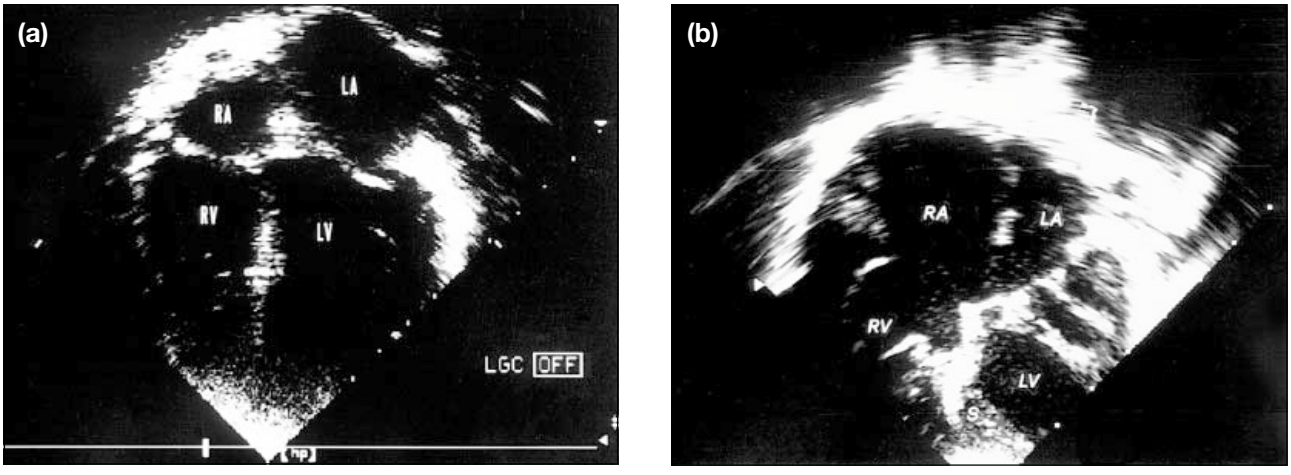

Figure 2. Apical four-chamber view of the heart. (a) A moderate-sized perimembranous ventricular septal defect can be seen. Note the slightly dilated left atrium and ventricle in the volume overloaded left heart; (b) a large atrio-ventricular septal defect with a common atrio-ventricular valve. Note the dilated right atrium and right ventricle in the setting of significant pulmonary hypertension. *Abbreviations:* LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

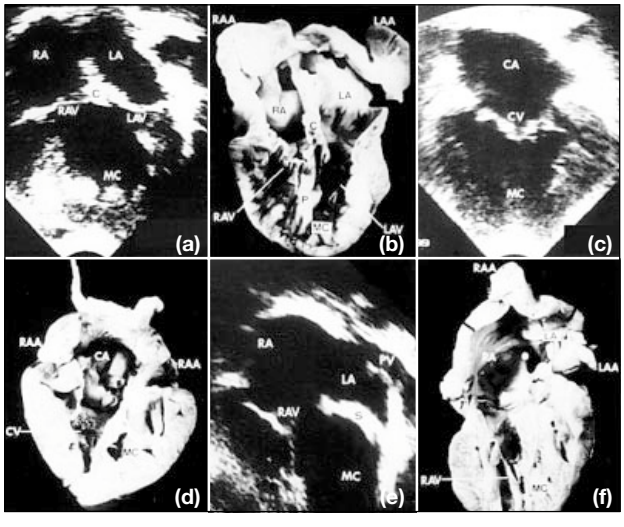

Figure 3. Apical four chamber views showing the various modes of univentricular atrio-ventricular connection correlating with pathological heart specimens cut to illustrate similar echocardiographic planes. (a and b) These show a double inlet univentricular atrio-ventricular connection via two separate atrio-ventricular valves; (c and d) these show double inlet univentricular atrio-ventricular connection via a common atrio-ventricular valve; (e and f) these show absence of left atrio-ventricular connection (mitral atresia). *Abbreviations:* C = central fibrous body; CA = common atrium; CV = common atrio-ventricular valve; LA = left atrium; LAA = left atrial appendage; LAV = left atrio-ventricular valve; MC = main ventricular chamber; PV = pulmonary vein; RA = right atrium; RAA = right atrial appendage; RAV = right atrio-ventricular valve.

ventricles.[13,14] This view should be interpreted alongside the parasternal long and short axial views. Typically, the left ventricle gives rise to the aorta, which has a relatively straight course towards the right shoulder, while the right ventricle gives rise to the anteriorly directed pulmonary trunk which bifurcates into the right and left pulmonary arteries. The two great vessels normally cross one another (Figure 4), so if

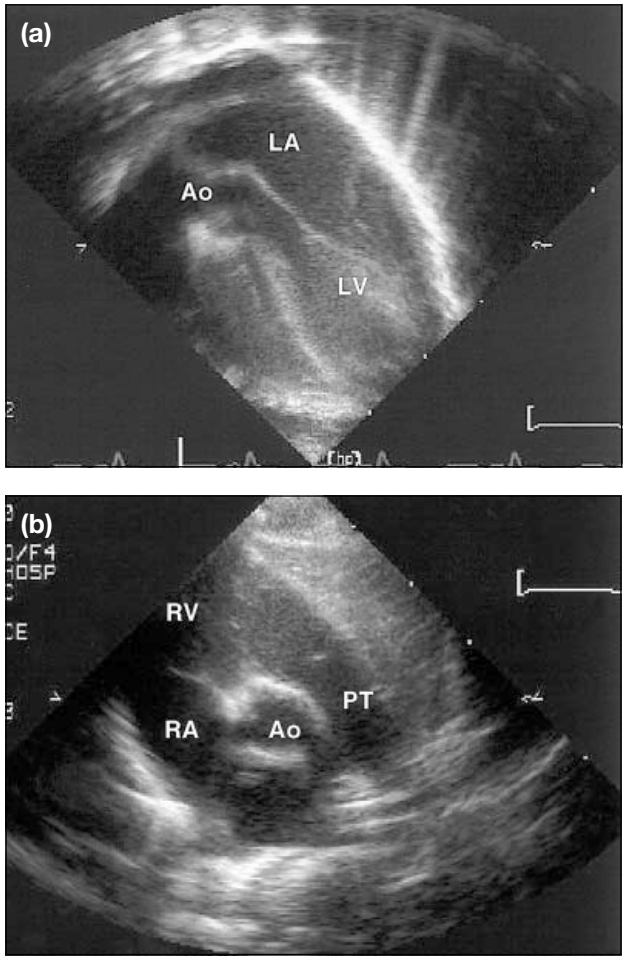

Figure 4. Echocardiograms showing normal crossing of the path of the two great vessels. (a) Apical long axial view which shows the left ventricle giving rise to the aorta. The vessel runs a relatively straight course towards the right shoulder; (b) parasternal short axis view showing the aorta in cross-section, being encircled by the anteriorly directed pulmonary artery which bifurcates into the left and right pulmonary arteries. *Abbreviations:* Ao = aorta; LA = left atrium; LV = left ventricle; PT = pulmonary trunk.

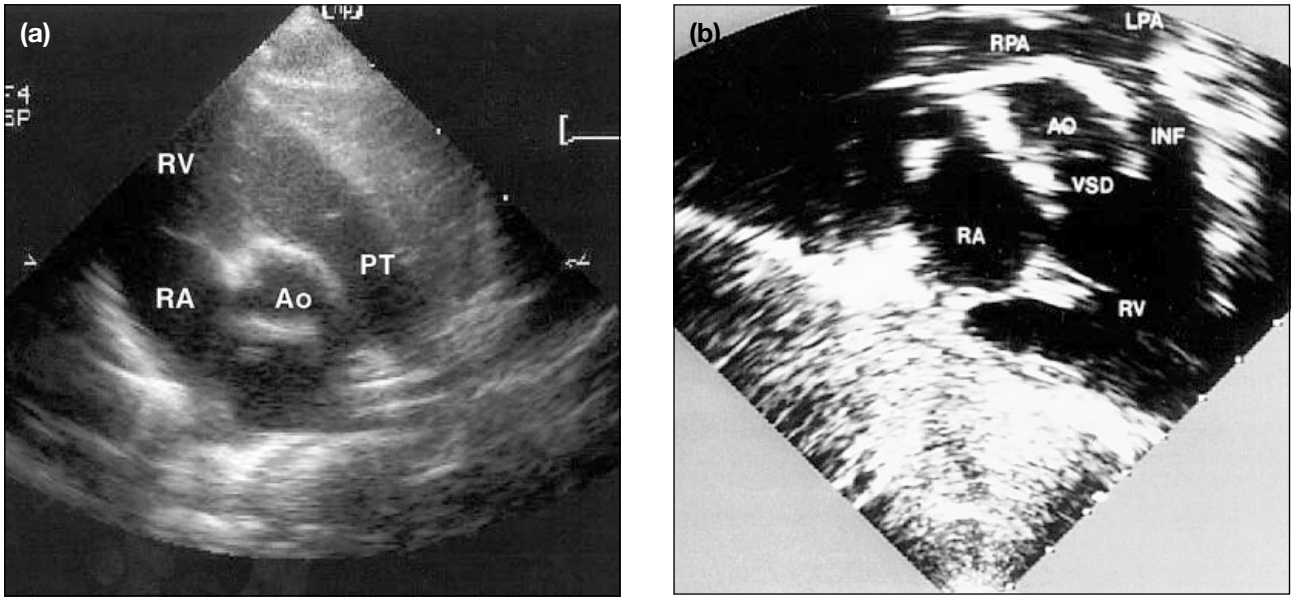

Figure 5. Anterior angulation from the subxiphoid view can demonstrate abnormalities of the ventricular outflow tracts. (a) This illustrates a single trunk which gives rise to both the aorta and pulmonary arteries; (b) subvalvar narrowing of the pulmonary outflow tract in the setting of Tetralogy of Fallot. *Abbreviations:* Ao = aorta; INF = infundibulum; LPA = left pulmonary artery; PA = main pulmonary artery; RA = right atrium; RPA = right pulmonary artery; RV = right ventricle; Trunk = truncus arteriosus; VSD = ventricular septal defect.

the great vessels appear parallel this indicates an abnormal ventriculo-arterial connection. For example, identification of the posterior course of the pulmonary trunk as bifurcating and parallel to the anterior aorta indicates transposition of the great vessels.[15,16] One should also be aware of other abnormal ventriculo-arterial connections, such as a double outlet ventricle and truncus arteriosus (Figure 5a). In addition, obstructive lesions involving the valvar and subvalvar levels of the semilunar valves (Figure 5b) of the great vessels, as in subpulmonary obstruction of Tetralogy of Fallot, should be considered.

Abnormalities of the Great Vessels and Venous Vascular Channels

The suprasternal short and long axial views along the aortic arch can reveal major arch abnormalities, including those of coarctation[17] and interruption[18] (Figure 6). In addition, by varying the angulation of the transducer anterio-posteriorly and to the left and right in the short axial view, the right superior vena cava or persistence of the left superior vena cava, as well as abnormal intracardiac or supracardiac pulmonary venous return, can be identified. In the setting of pulmonary outflow tract obstruction (e.g. Tetralogy of Fallot or pulmonary atresia with ventricular septal defect), additional abnormalities including hypoplasia, localized stenosis, or interruption of either left or right pulmonary artery near their origins of bifurcation may also be identified. If obstructive pulmonary venous return is suspected — usually in the clinical setting of severe cyanosis with chest radiography showing pulmonary venous congestion — the subxiphoid view should be used to locate an abnormal descending venous channel draining into the liver[19] (Figure 7). This latter situation of infradiaphragmatic total pulmonary venous connection emphasises the need for obtaining relevant clinical information prior to an intelligent search for cardiac abnormalities.

DOPPLER FLOW STUDY

Doppler echocardiography should only be performed when most or all of the cardiac abnormalities have been identified. Although less experienced echocardiographers may anticipate that colour flow mapping would pinpoint a specific diagnosis, not only is the relatively rapid motion of the colour flow confusing, but frequently abnormal anatomy can be masked by the simultaneous flashing of colours.

At times the identification of vascular channels can be guided by the direction of flow, especially with vessels running unusual courses. In general, arterial flow is usually directed away from the heart, while venous drainage is towards the cardiac chambers. Pulsed Doppler further differentiates pulsatile arterial flow from non-pulsatile venous flow that varies with respiration. Colour flow Doppler can be employed to confirm haemodynamic disturbances suggested by identified abnormalities of cardiac anatomy. For example, the

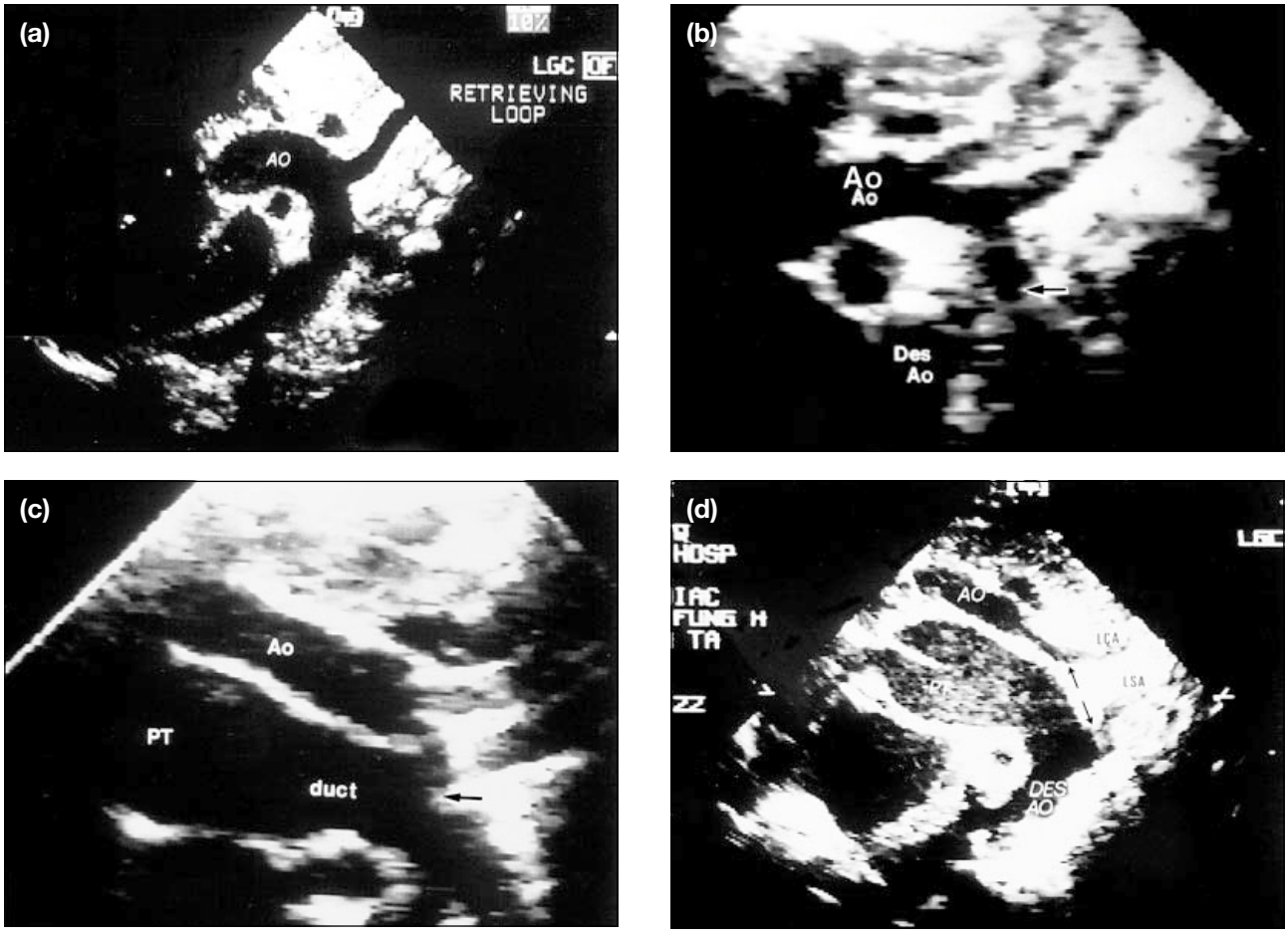

Figure 6. Suprasternal notch long axial views along the aortic arch. (a) A normal aortic arch; (b) coarctation of the aorta with discrete narrowing at the isthmus; (c) isthmal hypoplasia of the transverse arch; (d) interruption (between arrows) of the aortic arch. *Abbreviations:* Ao = aorta; Des AO = descending aorta; duct = arterial duct; LCA = left carotid artery; LSA = left subclavian artery; PT = pulmonary trunk.

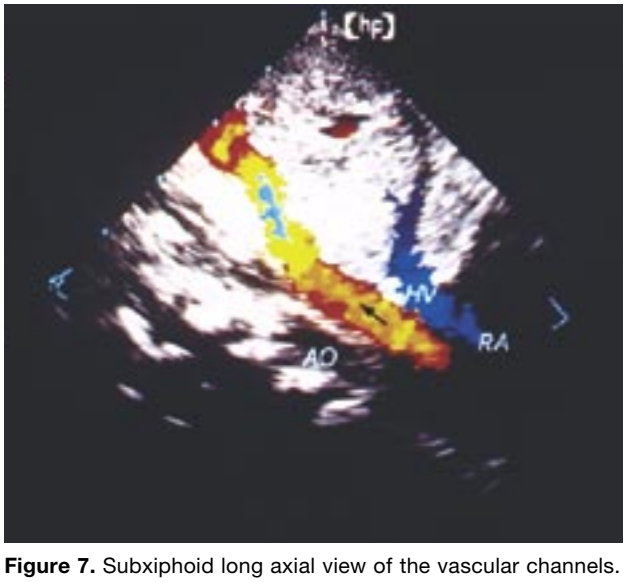

Figure 7. Subxiphoid long axial view of the vascular channels. This illustrates that in addition to the descending aorta, in this case there is a vein entering from above the diaphragm and descending into the liver (infradiaphragmatic total anomalous pulmonary venous connection), with an increase in venous influx to the hepatic veins which return to the right atrium. *Abbreviations:* AO = aorta; HV = hepatic vein; RA = right atrium.

detection of a ventricular septal defect by imaging alone would imply the presence of a left to right shunt. Colour flow mapping does not add to the diagnosis of the defect. However, valuable information on the severity of pulmonary hypertension can be determined — if the colour jet crosses the defect with little turbulence, or if right to left shunting is present. The application of continuous wave Doppler studies then allows estimation of the transventricular gradient and calculation of the pulmonary arterial systolic pressure using the Bernoulli's equation.

CONCLUSION

In summary, the diagnosis of a complex congenital heart disease in the neonate requires a systematic approach using a combination of echocardiographic views. The identification of an obvious defect should not limit the systematic search in each segment of the heart for further abnormalities. Only by so doing can one avoid overlooking any potential major lesion contributing to disturbed haemodynamic flow.

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