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Original Article

The essential echocardiographic features of tetralogy of Fallot*

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Abstract This presentation will demonstrate the essential features of tetralogy of Fallot in the infant and child before surgery, as well as some noteworthy features in the foetus. The four features, namely, subpulmonary stenosis, ventricular septal defect, aortic override, and right ventricular hypertrophy, can all be easily demonstrated by echocardiography. In addition, morphology of the pulmonary valve and the main and branch pulmonary arteries can be seen. The position of the coronary arteries and the major variants of proximal coronary anatomy can be defined. The arch anatomy and the presence of associated major aortopulmonary collateral arteries can be defined. All these features can be demonstrated in the foetus as well, after the first trimester, and the presence of major aortopulmonary collateral arteries can be seen more clearly because the lungs, being fluid filled, aid in ultrasound and do not provide the barrier that the air-filled lung presents after birth.

Keywords: Ultrasound; history of echocardiography; cardiac surgery

ECHOCARDIOGRAPHY HAS BECOME THE STANDARD modality in the diagnosis of tetralogy of Fallot,[1–3] the fundamentals of which are described in this paper. Diagnosis by echocardiography is often supported by other ultrasound and imaging modalities, and by cardiovascular haemodynamics, when necessary, to deliver the most effective care for patients who present with this condition. Although echocardiographic techniques and evaluations are very detailed, this overview is limited to a discussion of the salient features of tetralogy of Fallot. The echocardiographic features mirror the pathological features of the disease found at surgery,[4,5] making echocardiography the surgeon's reference point.

The most prominent and, historically, the earliest echocardiographic feature of defects with aortic override of the ventricular septum are seen from the classical long-axis and subcostal coronal views (Figs 1 and 2). In these views, a single great artery overrides the ventricular septal defect to a variable degree – usually about 50% – after further differentiation has excluded common arterial trunk. If the vessel appears to be a pulmonary artery, a diagnosis of transposition with ventricular septal defect or double-outlet right ventricle would have to be considered. Once the vessel is recognised as being the aorta, a variable degree of aortic override can be demonstrated to define whether there is left ventricular outflow narrowing. Other features, such as septal, left, and right ventricular hypertrophy, as well as the presence of a left superior vena cava to coronary sinus connection can also be defined. Thus, three of the four features of tetralogy can be defined immediately with these echocardiographic cuts, namely, the ventricular septal defect, the aortic override, and right ventricular hypertrophy. The ascending aorta and the arch laterality can usually be seen in smaller infants and children from the subcostal coronal view (Fig 2). Further scanning from these planes in different directions will demonstrate other features of tetralogy of Fallot and the remaining feature of the tetrad, namely, the pulmonary outflow tract obstruction.

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A video of this presentation can be viewed at the following hyperlink: [http://www.allkids.org/wcpccs].

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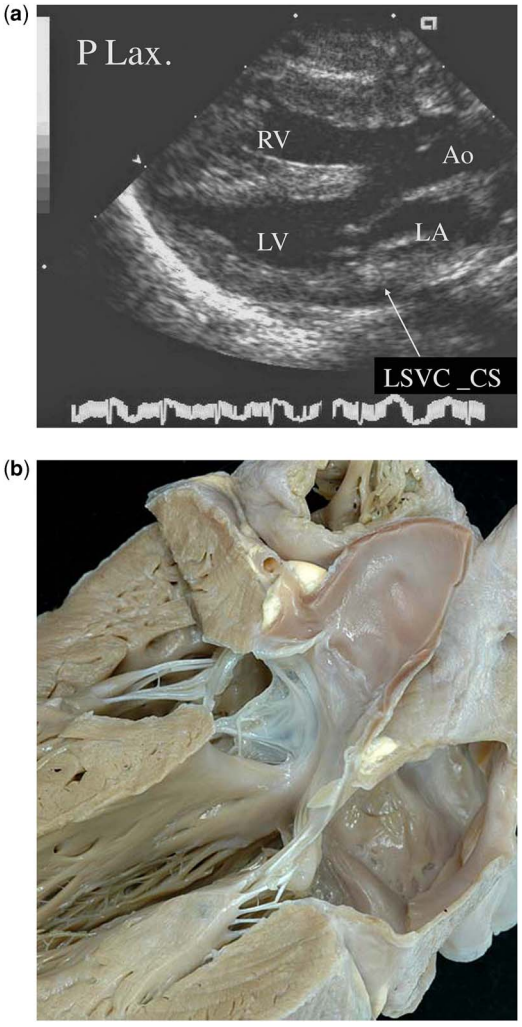

Figure 1.
*(**a**) A parasternal long-axis view of a neonate with tetralogy Fallot is represented. The Ao, overrides the ventricular septal defect and the muscular ventricular septum, lying between the LV and RV. A LSVC to CS connection is suggested from this view by the enlarged coronary sinus. The thickness of the anterior wall exceeds the thickness of the LV, indicating RV hypertrophy. (**b**) This figure from a pathological specimen is matched with the echocardiogram to show the Ao overriding the ventricular septal defect and muscular septum. Note that there is an area of mitral to aortic fibrous continuity, which is one of the features of this lesion (see text for details). Ao = aorta; CS = coronary sinus; LA = left atrium; LSVC = left superior vena cava; LV = left ventricle; RV = right ventricle; LSVC-CS = a left superior vena cave to coronary sinus connection.*

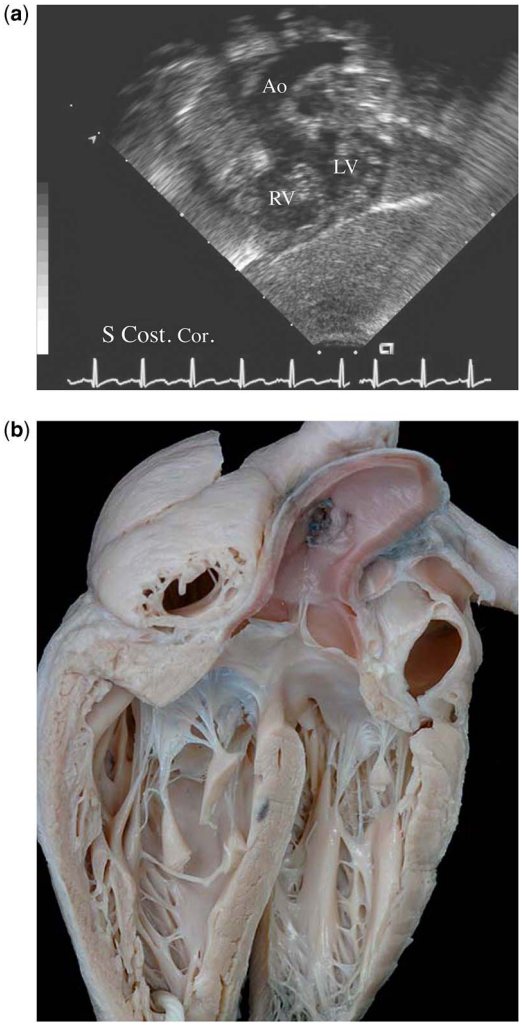

Figure 2.
*(**a**) Subcostal coronal view of a neonate with tetralogy of Fallot is represented. The RV and LV are shown and the Ao overrides the ventricular septal defect. The aortic arch traced arching to the leftward. (**b**) This figure from a pathological specimen is matched with the echocardiogram to show the aorta overriding the ventricular septal defect and muscular septum. Note the fibrous continuity between the aortic, mitral, and tricuspid valve and the membranous flap, the remnant of the membranous septum. This perimembranous extension occurs in a large proportion of cases of tetralogy of Fallot. Ao = aorta; LV = left ventricle; RV = right ventricle; S Cost Cor = subcostal coronal.*

The last major feature of the tetrad is associated with deviated outlet septum and infundibular hypoplasia.[6] This finding was initially reported by Niels Stensen in 1671, renamed by Van Praagh the "Monology of Stensen".[7] In 1888, Etienne-Louis Arthur Fallot published five serialised contributions in the *Marseille Médical* concerning the "blue malady" in which he described the now classical tetralogy of pulmonary outflow tract obstruction, ventricular septal defect, aortic overriding, and right ventricular hypertrophy. It was only in 1924 that Maude Abbott coined the term "tetralogy of Fallot".[7]

Deviation of the outflow septum can be demonstrated from a variety of subcostal, coronal, sagittal, and parasternal views (Fig 3a–c). These views show not only the deviated outlet septum, but also the narrowed right ventricular outflow tract and the pulmonary valvar pathology, additionally providing assessment of both the main pulmonary artery and

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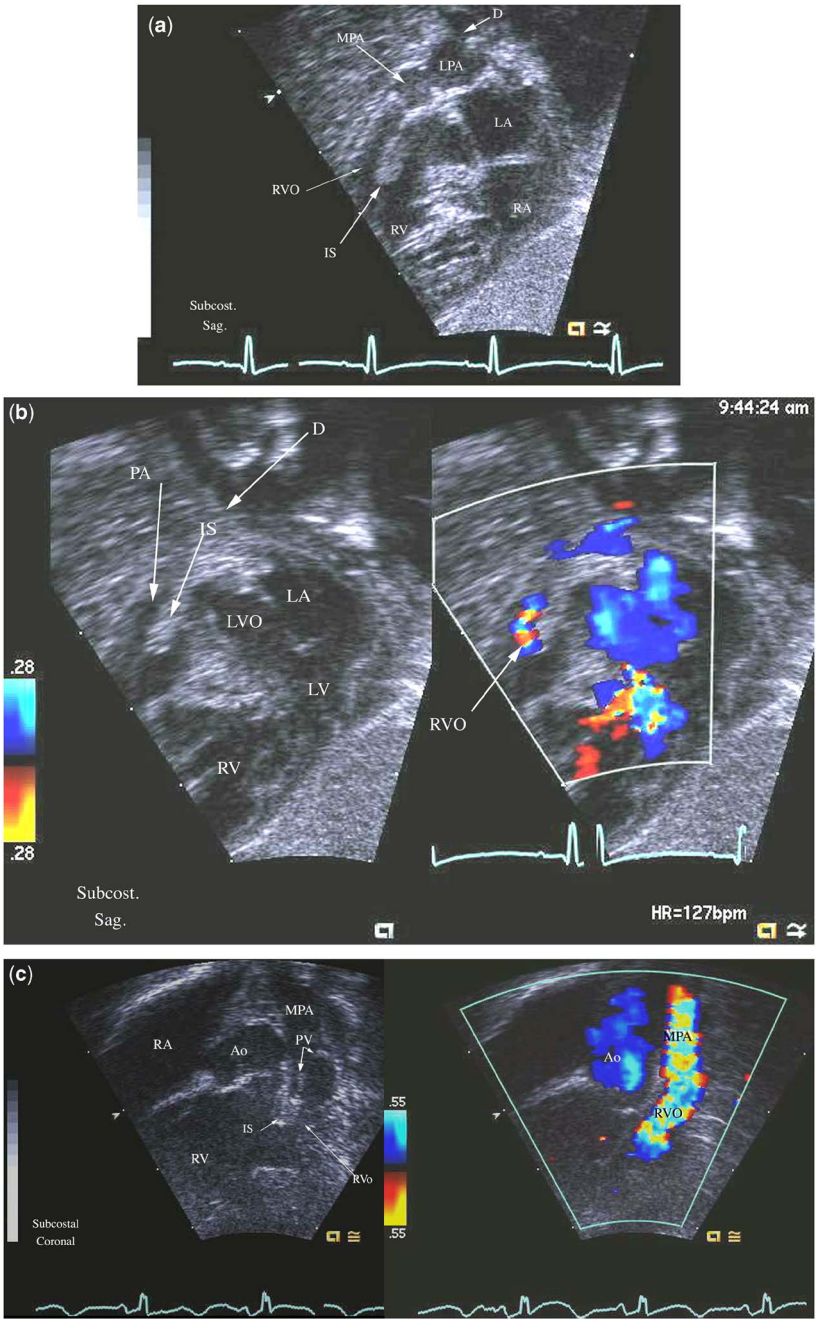

Figure 3.

*(**a**) This subcostal sagittal cut shows the narrow RVO tract anteriorly. It is narrowed by the anteriorly deviated IS, separating the infundibulum from the body of the RV. The MPA, above, is separated from the RVO by a doming pulmonary valve. The MPA is separated from the origin of the LPA by a ridge of tissue. The D enters the roof of the LPA from the descending Ao. (**b**) This subcostal sagittal cut with accompanying Doppler colour flow image is from a different patient with slightly different angulation from Figure 3a. Note that although these are simultaneously recorded images, the two images highlight and display different but complementary information, which, when synthesised, yield a more complete representation of the anatomy and physiology. The narrowed RVO tract anteriorly is only filled with colour, suggesting forward flow across the valve, whereas the deviated IS in the non-colour image shows only a small infundibulum, pulmonary valve, and PA. The MPA is separated from the RVO by a doming pulmonary valve, and the MPA is separated from the origin of the left PA by a ridge of tissue. The calibre of the duct is also noted in the colour image to be a small turquoise streak. The D enters the roof of the LPA from the descending Ao. (**c**) This figure is taken from a subcostal coronal cut at right angles to Figure 3b. The cut passes anteriorly through the right ventricle and both great vessels. The aorta (Ao) and the main pulmonary artery (MPA) are arising from the right ventricle (RV). The infundibular septum (IS) is deviated rightward narrowing the right ventricular outflow tract. The frame was systolic and the pulmonary valvar leaflets (PV) are opened and domed (Arrows). The right hand figure shows the superimposition of the Doppler colour flow image with the narrowed flow extending through the right ventricular outflow. Ao = aorta; D = arterial duct; IS = infundibular septum; LA = left atrium; LPA = left pulmonary artery; LV = left ventricular; LVO = left ventricular outflow; MPA = main pulmonary artery; PA = pulmonary artery; PV = pulmonary valve; RA = right atrium; RV = right ventricle; RVO = right ventricular outflow; Subcost sag = subcostal sagittal.*

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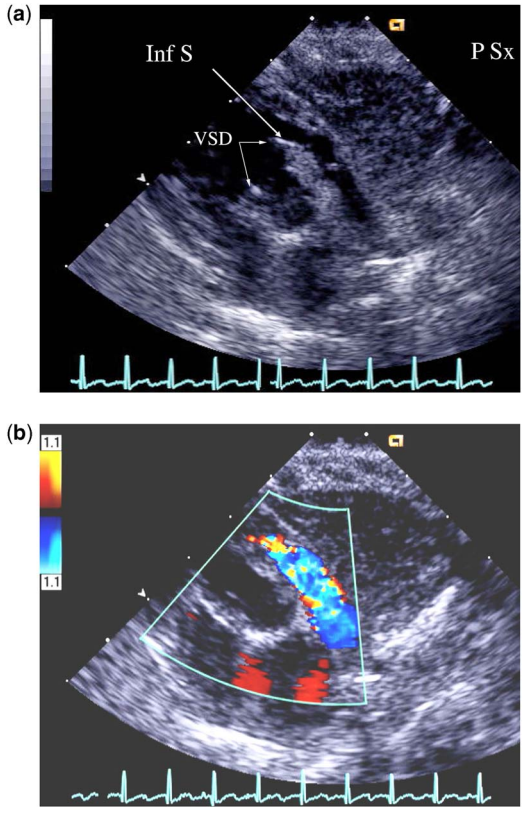

Figure 4.
*(**a**) This frame, taken in the PSx, shows the Inf S deviated superiorly and anteriorly towards the right ventricular outflow tract with the narrowing continuing into the right ventricular outflow tract, the pulmonary valve, and the main pulmonary artery. The VSD + arrows has a perimembranous extension. (**b**) Doppler colour flow equivalent demonstrates accelerated flow into the right ventricular outflow and main pulmonary artery. Inf S = infundibular septum; PSx = parasternal short-axis view; VSD = ventricular septal defect.*

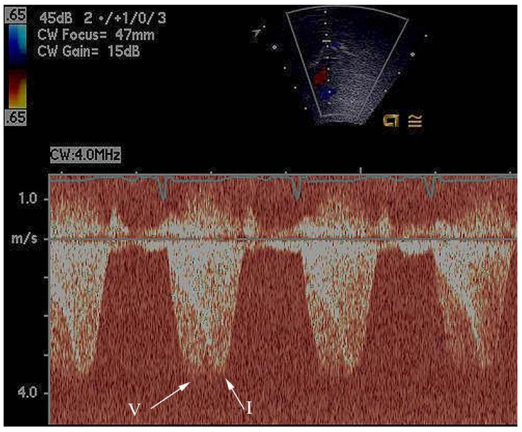

Figure 5.
This Doppler colour flow signal is obtained from a subcostal view from the reference image in the upper part of the image. The Doppler signal shows a dual-phased image in the continuous wave Doppler signal showing pulmonary flow moving away from the transducer. The peak velocity approaches 4 m/second – equivalent to a pressure drop of ~64 mmHg – as indicated by the scale markings on the left of the recording. There are two distinct peaks in this recording. The early peak is valvar in origin (V) and the later peak of equal velocity is infundibular in origin (I). The velocity from this latter peak increases as the right ventricle continues to contract and narrow the outflow towards the end of systole. The valvar velocity declines after the earlier occurrence of right ventricular peak ejection. Abd = abdomen; DAo = descending aorta.

the branching pulmonary arteries (Fig 4a and b). These subcostal views are complemented by parasternal views taken in the short axis that define ventricular size, hypertrophy, infundibular narrowing, annular size, pulmonary valvar and branch pulmonary anatomy, and physiology. In addition to imaging, continuous wave Doppler differentiates valvar from infundibular narrowing by the different timing of the velocity curves originating at these two sites (Fig 5). Thus, all the features of tetralogy of Fallot can be demonstrated with this complement of views.

Further echocardiographic interrogation demonstrates many other features of the disorder, augmenting our appreciation of its scope and providing essential information to the surgeon.

Knowledge about pulmonary valvar pathology is critical for surgical correction, particularly when the possibility of a transannular approach and patching of the outflow is required. Pulmonary annular size, valvar morphology, and branch pulmonary narrowing are demonstrated by echocardiography. Current surgical thinking about pulmonary valve repair in tetralogy of Fallot tries to spare the valve so that the degree of pulmonary valvar incompetence, which has long-term deleterious consequences, may be minimised. In this situation, where the anterior cusp lies across the anterior aspect of the right ventricular outflow area, a transannular incision may destroy a cusp as it overlies the anterior aspect of the pulmonary valve and infundibulum, whereas a bicuspid valve with a more anteroposterior orientation may present a commissure to the knife and thus spare the valve cusp from destruction.[8] Although direct inspection at the time of repair provides the following information, it is better done by echocardiography before commencement of the operative repair. The pulmonary valve is best detected by short-axis views (Fig 6a–c). Orientation of the probe in the appropriate plane so that it passes through the plane of the valvar leaflets is different from its orientation when defining the normal pulmonary valve, because right ventricular enlargement will push the valve annulus higher and deeper into the chest, making the pulmonary valve

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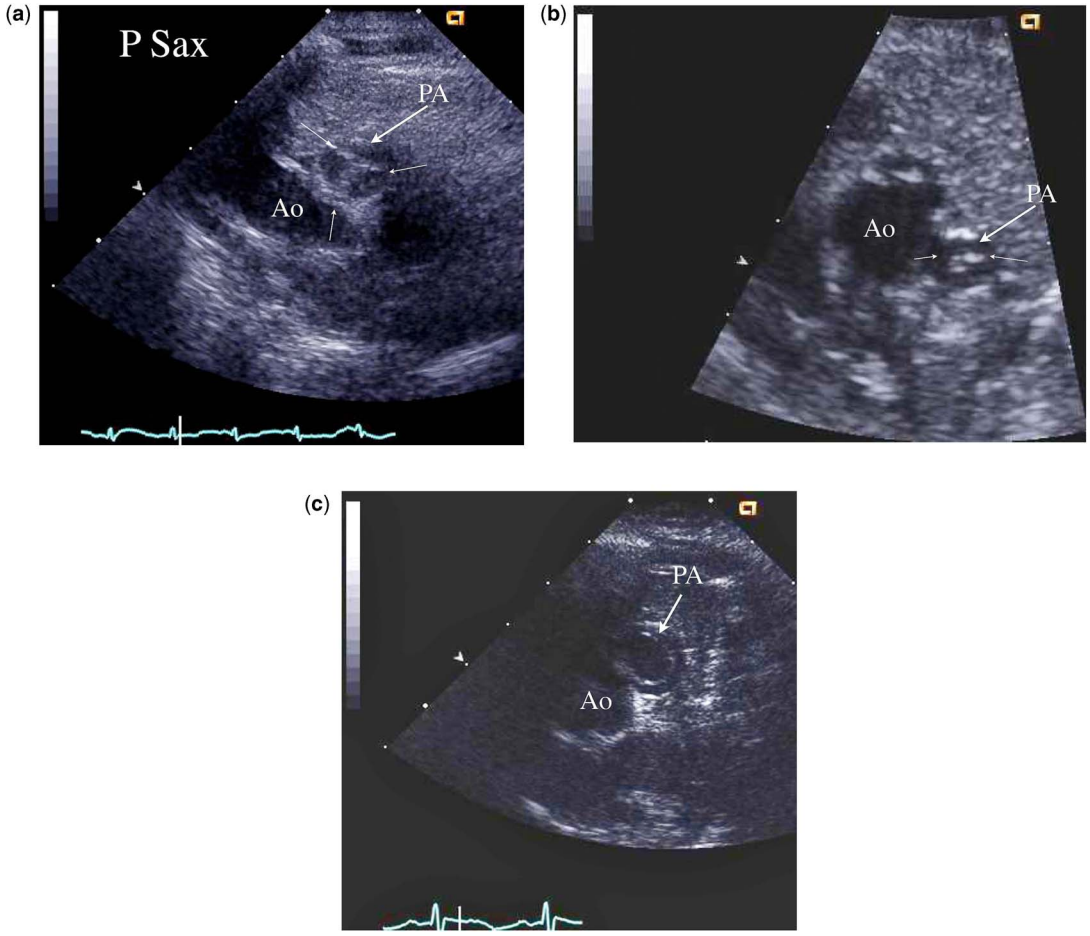

Figure 6.
*These three images represent three different forms of pulmonary valvar anatomy in tetralogy of Fallot taken in the PSx. (**a**) This echocardiogram shows an unusual form of tricuspid pulmonary valve; three small arrows at the commissures within the pulmonary artery. The large aorta is adjacent to the pulmonary annulus. (**b**) Shows a severe form of tetralogy of Fallot with a small bicuspid pulmonary valve and the cusp direction parallel to the chest wall, leading to possible obliteration by a transannular patch (arrows). The Ao is much larger in calibre than the pulmonary annulus. (**c**) Shows the so-called "volcano" or unicuspid form of pulmonary valvar stenosis. If transannular patching were required in this instance, it would create pulmonary valvar incompetence. Ao = aorta; PA = pulmonary artery; P Sax = parasternal short-axis view.*

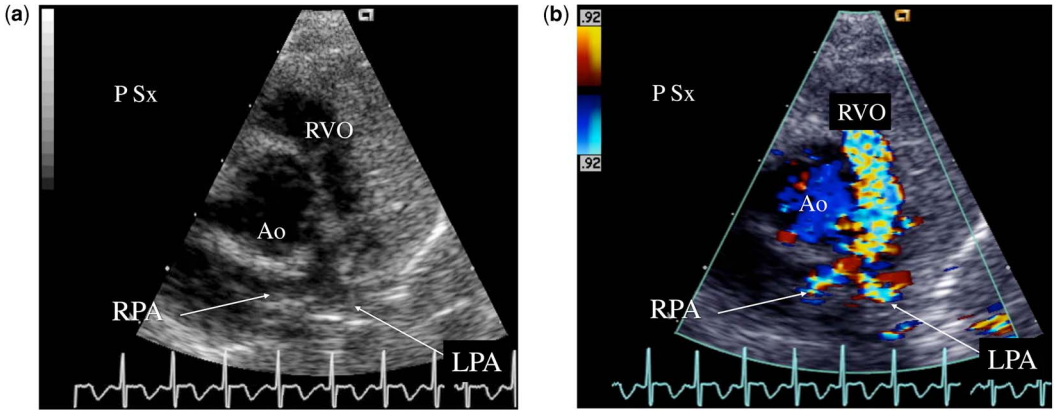

Figure 7.
*(**a**) This frame shows a parasternal short-axis view (PSx), lower than that seen in Figure 4, with more cranial angulation showing the RVO tract, pulmonary valve, main pulmonary artery, and narrowing of the origins of the left and right pulmonary arterial branches (LPA and RPA). The Ao lies medially. (**b**) The corresponding Doppler colour flow image here shows clear acceleration of the velocity from the right ventricular outflow tract into both branches, but with narrowing of the flow signal at the origins of the branch pulmonary arteries, indicating branch pulmonary arterial stenosis. Ao = aorta; LPA = left ventricular artery; PSx = parasternal short-axis view; RPA = right pulmonary artery; RVO = right ventricular outflow.*

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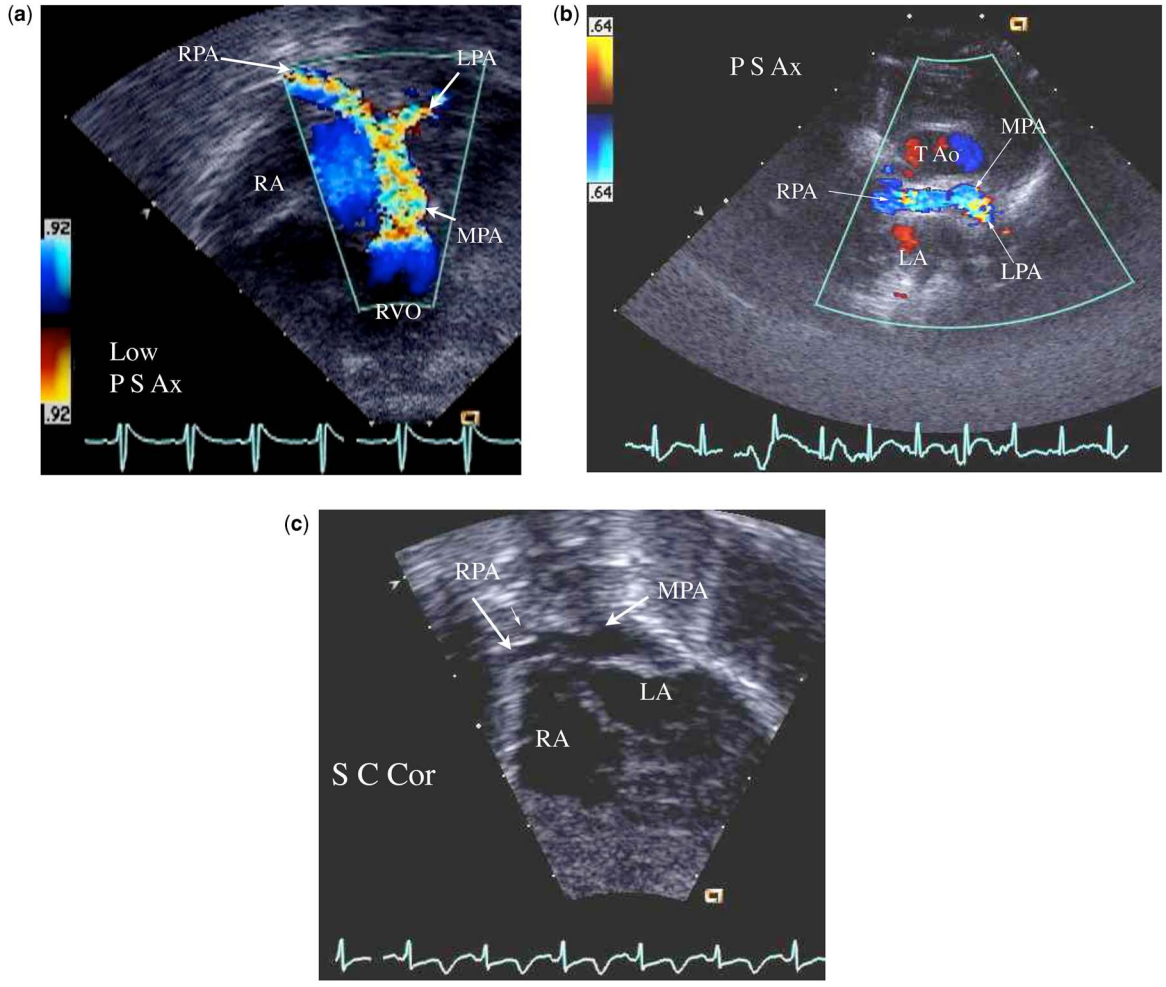

Figure 8.
*(**a**) This P S Ax taken with appropriate anatomical image orientation demonstrates the diminished size and more symmetrical orientation of the branch pulmonary arteries. It further explores the use of Doppler colour flow imaging in this condition. The acceleration begins in the RVO tract and continues through the MPA, LPA, and RPA branches. (**b**) This view is taken in a very high horizontal PSx plane. In this view, the frame shows the T Ao with a colour signal and the MPA, LPA and RPA branches into the hilums of both lungs. The difference between the colour signals in the pulmonary arteries over the low velocity of flow in the aorta represents an increase in the flow velocity within the proximal pulmonary circulation. (**c**) In this subcostal coronal cut, the transducer has been aligned with the RPA, originating from the MPA. The upper lobe artery is identified (small arrow). The left pulmonary artery runs more posteriorly and is not present in this tomographic cut. LA = left atrium; LPA = left pulmonary artery; MPA = main pulmonary artery; P S Ax = parasternal short-axis; RA = right atrium; RPA = right pulmonary artery; RVO = right ventricular outflow; S C Cor = subcoastal coronal; T Ao = transverse aorta.*

lie further from the precordium and positioned in a more vertical orientation.

To display the valve more clearly, the probe must be placed in a higher precordial position and in a more clockwise rotation to achieve a view through the plane of the pulmonary valve, which is most frequently bicuspid. If the valve is tricuspid, placing the infundibular incision between the cusps of the valve will prevent valvar destruction. If it is unicuspid, there is no option but to incise the leaflet, thereby creating greater pulmonary valvar incompetence with all of its consequences (Fig 6a–c). In smaller children, it is sometimes possible to examine the pulmonary valve from the subcostal sagittal and the parasternal short-axis view.

The main and branch pulmonary artery sizes are of critical importance in tetralogy, as well as in situations associated with pulmonary atresia. From the subcostal sagittal view, the parasternal short-axis and equivalent planar view will display this morphology (Figs 7 and 8). Complete definition of the diameters of the main and branch pulmonary arteries to the hilum of each lung may make cardiac catheterisation unnecessary if the pulmonary artery and its branches can be traced well. In addition, using a combination of views, it is

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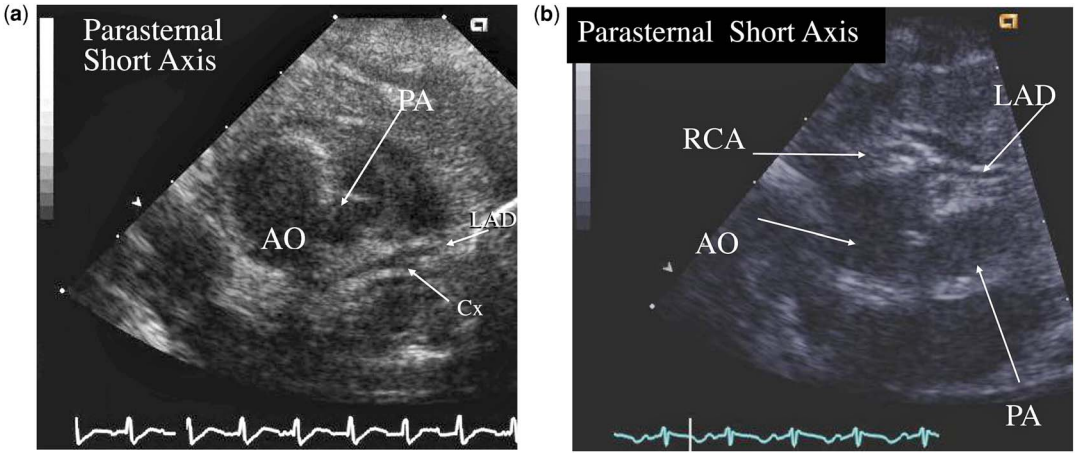

Figure 9.
(a) This parasternal short-axis cut concentrates on the usual position of the left main coronary artery, which is seen arising from the Ao, and bifurcating into the LAD and Cx coronary arteries. The arteries all run behind the main pulmonary artery (PA) and are therefore safe from incision at surgery. (b) In this cut, the RCA arises off the aorta (AO) and gives rise to a large LAD coronary artery, which runs in front of the PA. The LAD artery lies anterior to the right ventricular outflow tract and is therefore exposed to the surgical field at the time of surgery. Ao = aorta; Cx = circumflex coronary artery; LAD = left anterior descending coronary artery; PA = pulmonary artery; RCA = right coronary artery.

possible to define stenosis at the origin of the branches, the branch size, as well as acceleration of the Doppler colour flow signal, which can indicate stenosis (Fig 8).

Another valuable morphologic feature in tetralogy of Fallot that needs definition is coronary arterial anatomy. In most cases of tetralogy of Fallot, the left main coronary artery runs in a proximal course behind the main pulmonary artery (Fig 9a). Such a pattern indicates that the left anterior descending coronary artery is protected from the surgeon's knife. Coronary arteries that run across the right ventricular outflow tract are not protected, particularly if buried in pericardial scarring from prior surgery. When the left anterior descending arises from the right coronary artery and runs in front of the right ventricular outflow tract, it is at risk of being cut. Although the surgeon can usually see this artery, it may be buried or run in a pericardial adhesion, thus becoming covert. Using this high parasternal view, the artery is easily recognisable echocardiographically (Fig 9b). Large conus branches or dual left anterior descending coronary arteries are also at risk from surgical trauma. The artery can usually be differentiated both from the large conus branch artery or the dual left anterior descending artery that run across the right ventricular outflow tract because the left anterior descending coronary artery runs its usual course behind the pulmonary artery in these situations.

One of the more problematic issues for surgical repair in tetralogy of Fallot arises when pulmonary atresia is present. In this instance, pulmonary blood flow arises either from an arterial duct (Fig 10a and b) or from major aortopulmonary collateral arteries. When the source of flow arises solely from the duct arising from the base of an innominate artery, it is easily identified from standard views (Fig 10c). The arterial duct may also arise from the base of a right aortic arch, where it is usually small, tortuous, and oriented in a vertical direction (Fig 10d and e). When major aortopulmonary collateral arteries are present, the differences in pulmonary blood supply can also be defined by echocardiography, particularly with the addition of Doppler colour flow information. The major aortopulmonary collateral arteries generally arise off the descending aorta but may arise elsewhere from the arch vessels or even from the abdominal aorta. They usually have a slightly cranially oriented course, running from below to above, while the arterial duct is usually caudally directed. The major aortopulmonary collateral artery vessels run this course because of the embryonic ascent of the lung and the systemic arteries that take their blood supply as the lung ascends, whereas the vertical orientation of the duct relates to the reversed foetal flow direction into the pulmonary arteries via the arterial duct. When the intrapulmonary pulmonary arteries anastomose with the sixth arch vessels, they usually lose contact with the major aortopulmonary collateral arteries. Acherman et al noted that when major aortopulmonary collateral arteries and/or central pulmonary

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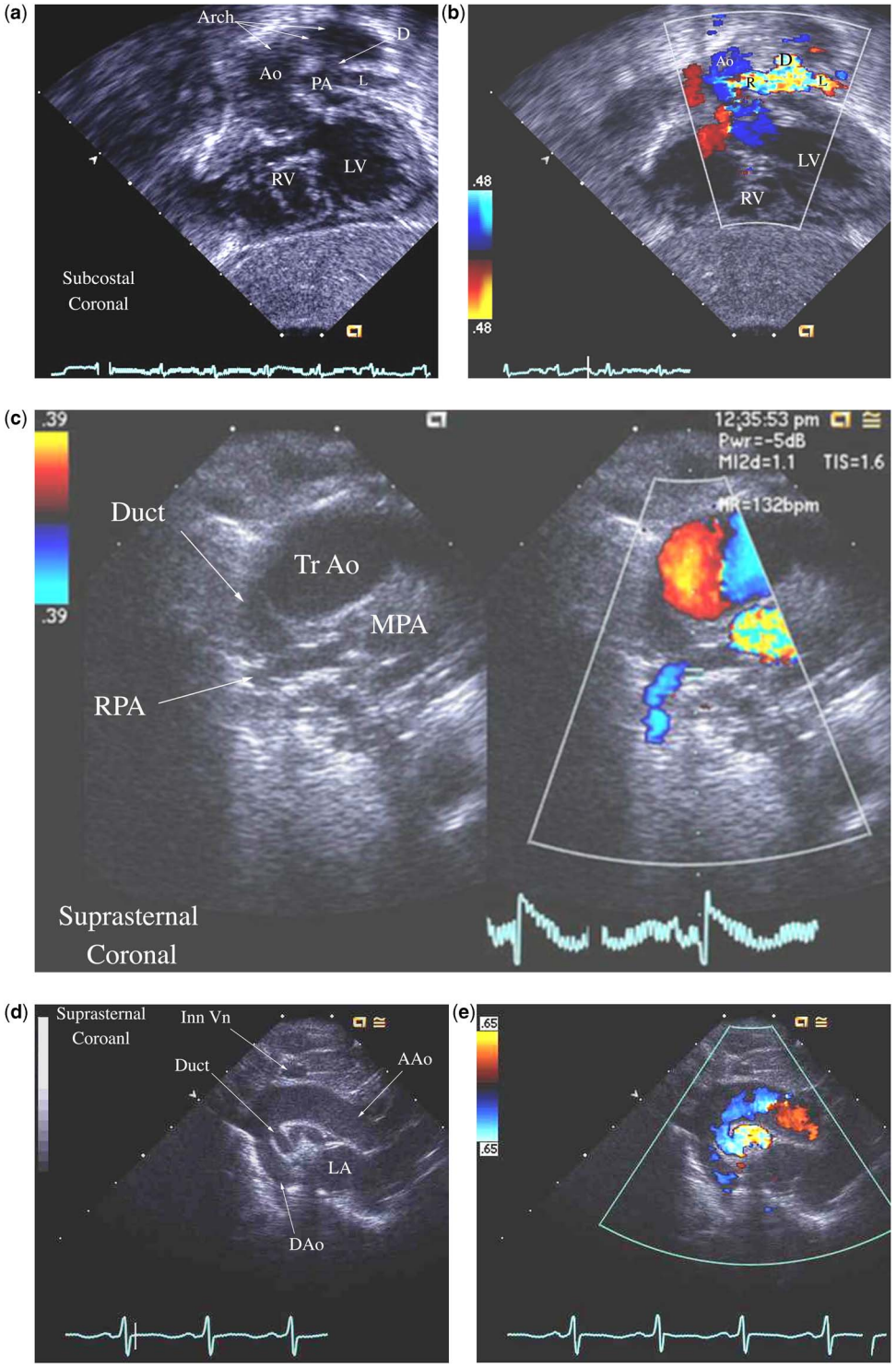

Figure 10.

*(**a**–**e**) Patients with tetralogy of Fallot where the sole source of pulmonary blood supply is from a patent arterial duct. (**a** and **b**) These images were taken within seconds of each other in the same infant with pulmonary atresia and sole source ductus-derived pulmonary blood supply. Figure 10a is a subcostal coronal view identifying the D arising from the base of the left-sided aortic arch (triple arrow). The MPA and the left branch are also noted. The LV and RV are labelled. (**b**) The Doppler colour flow image shows the arterial duct, and the LPA and RPA are seen to be of good size and arising from the duct. The disturbed flow at increased velocity in yellow and green is at a higher velocity than on the aorta. The labelled items are the same as Figure 10a. (**c**) This cut in simultaneous black-and-white and colour images was taken from the suprasternal notch in a coronal plane. The Tr Ao is identified behind the thymus, showing low-velocity blood flow, indicating that the flow is travelling towards the left side. Therefore, the aortic arch is left sided. The MPA and RPA are identified posterior and inferior to the aorta. The pulmonary arteries here are supplied exclusively from a*

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Figure 10. *Continued*

*right-sided duct from the base of the right innominate artery. (**d** and **e**) These images are simultaneously recorded in black-and-white and colour flow images in the same patient with pulmonary atresia, and sole source supply from a patent arterial duct. Both images are represented in the oblique coronal plane with the aortic arch descending to the right. In contradistinction to the image in Figure 10d, the arterial duct arises from the base of the aorta, rather than from an innominate artery. The duct is tortuous and demonstrates a vertical origin from the base of the underside of the aortic arch. The Inn Vn is noted anteriorly and the AAo and DAo are labelled, as is the LA. In Figure 10e, the complementary image and flow into the pulmonary artery is shown. Note the colour change indicates acceleration of flow in the duct. AAo = ascending aorta; Ao = aorta; D = patent arterial duct; DAo = descending aorta; Inn Vn = innominate vein; L = left; LA = left atrium; LV = left ventricle; MPA = main pulmonary artery; PA = pulmonary artery; R = right; RPA = right pulmonary artery; RV = right ventricle; TrAo = transverse aorta.*

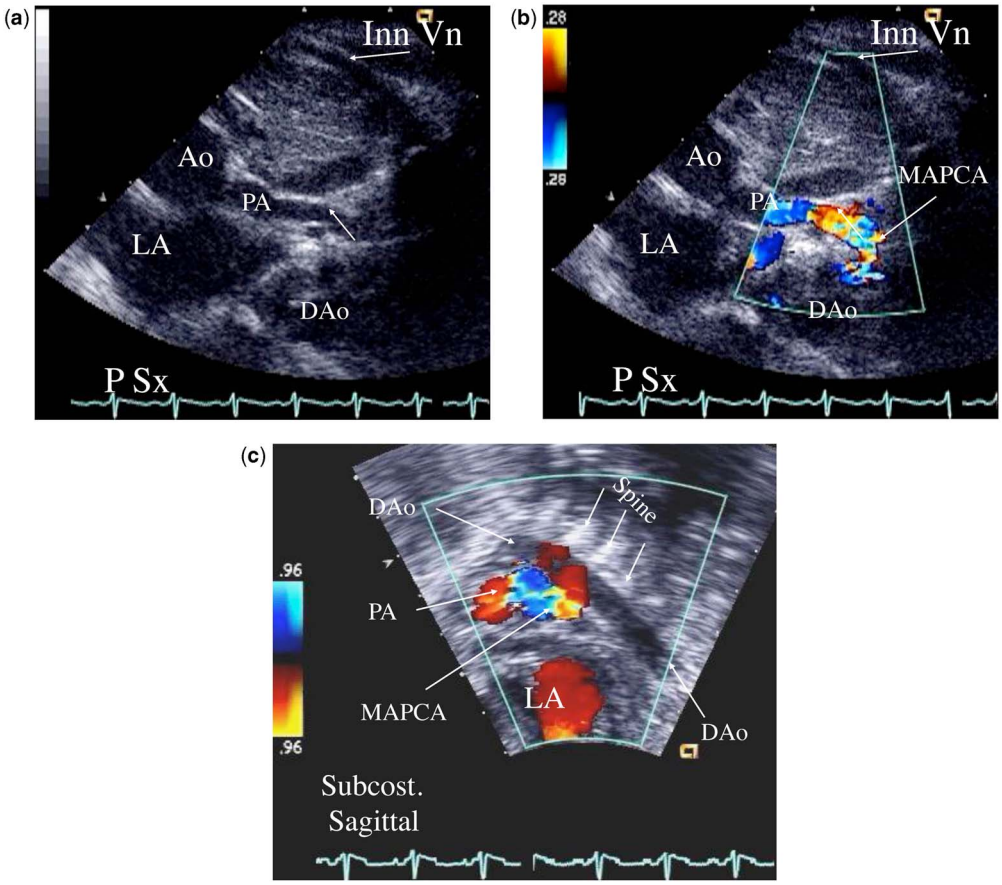

Figure 11.

*These figures demonstrate the MAPCAs in pulmonary atresia associated with tetralogy of Fallot. (**a** and **b**) These images were simultaneously black-and-white (11A) and colour images (11B) of the native pulmonary arteries in the PSx in one patient. The small MPA with its left branch is seen (arrow) behind the Ao. The LA and the DAo are posterior relations of the pulmonary artery. In the colour image, the disordered flow is seen entering the pulmonary artery peripherally from the DAo into a MAPCA, proceeding into the small left PA and then becoming more uniform as it enters the proximal part of the LPA. This distal entry of vascular flow is typical of flow from MAPCAs, as opposed to the more central flow typical of an arterial duct. (**c**) This subcostal sagittal cut demonstrates Doppler colour flow. The plane passes through the heart from anterior to posterior. The anterior part of the heart is excluded from this magnified view that passes from anterior to posterior through the LA, DAo and then the bodies of the spine (Spine) posteriorly. Flow from the descending aorta in red turns at the entrance of the MAPCA and follows a sinuous course represented by yellow and blue flow until it enters the PA in red. Ao = aorta; DAo = descending aorta; LA = left atrium; LPA = left pulmonary artery; MAPCAs = major aortopulmonary collateral arteries; PA = pulmonary artery; PSx = parasternal short-axis view; Subcost = subcostal.*

arteries exist, Doppler colour flow initiates at the hilum of the lung rather than centrally, as is the situation with an arterial duct.[9] These features can be identified from a variety of views (Fig 11). It is clear that when major aortopulmonary collateral arteries are defined by echocardiography, their complete evaluation requires that angiography be performed for complete delineation of the complex nature of the disorder and its surgical repair.[10]

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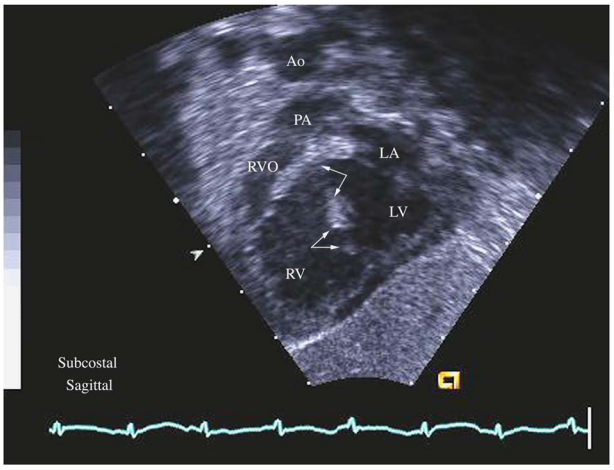

Figure 12.
This subcostal sagittal cut shows the ventricular septum in section, with the RV being anterior and the LA and LV being posterior. The transverse Ao is seen above the PA and the RVO is anterosuperior. The arrows indicate a usual ventricular septal defect in the superior position and an additional muscular ventricular septal defect is identified by the lower set of arrows. Ao = aorta; LA = left atrium; LV = left ventricle; PA = pulmonary artery; RV = right ventricle; RVO = right ventricular outflow.

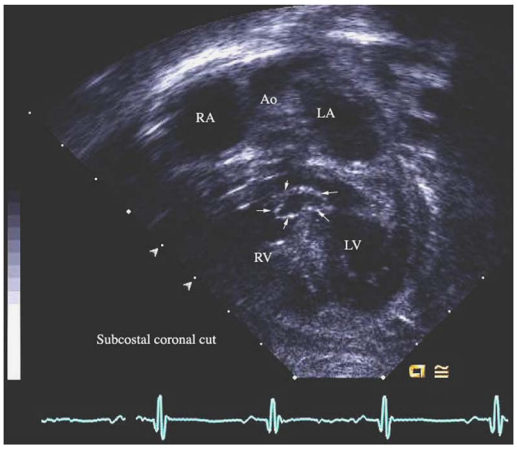

Figure 13.
This subcostal coronal cut, similar to Figure 2a, shows the ascending Ao arising above the ventricular septal defect. This space is also occupied by the common valve leaflet (arrows), straddling between the RV and LV. Note that the anterosuperior leaflet has no septal attachment, classified as a Rastelli Type C defect, the usual finding in conotruncal defects. Ao = aorta; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

If additional ventricular septal defects are present, the echocardiographer needs to relay this information to the surgeon. Imaging techniques, including angiography, may fail to demonstrate additional defects because the tendency is for blood to flow through the larger hole, and because the ventricular pressures are equal in the two ventricles. Consequently, the additional defects may be recognised by intraoperative trans-oesophageal echocardiography only after the patient has been separated from bypass, when the blood is forced through a smaller defect orifice or orifices, and the pressure between the ventricles assumes a more normal pressure differential. Defects that are larger in size are, however, easily detected (Fig 12).

Defects in the atrioventricular canal associated with tetralogy may be readily appreciated from a variety of echocardiographic views, as the ventricular septal defect is usually large and extends from the inlet septum to the area of the infundibulum of the right ventricle. With this association, the atrioventricular valvar morphology usually shows the anterior bridging leaflet extending across the ventricular septum without the attachment of the anterior bridging leaflet to the crest of the septum – also termed Rastelli Type C atrioventricular canal defect[11] (Fig 13).

Foetal echocardiography

In addition to corroborating the diagnosis postnatally, foetal echocardiography has allowed part of the natural developmental history of the disease to be shown, and many papers have been written on the essential features of the disease.[12–14] Views, similar to those described above, can be obtained to demonstrate the essential features of tetralogy of Fallot by foetal echocardiography (Fig 14a and b). The deviated outlet septum projecting towards the right ventricular wall, producing subpulmonary stenosis and narrowing of the main pulmonary artery and its branches, can also be assessed in utero (Fig 15).

It is notable that, although there is pulmonary obstruction, flow acceleration is not always present, depending on the foetal physiology. When the arterial duct is widely patent, pulmonary arterial pressure will be at systemic systolic levels, as will the right ventricular pressure because of the ventricular septal defect. The pressure drop therefore will be small or absent. Consequently, the flow velocity recorded across the pulmonary valve will be minimal (Fig 16). Of course, when the arterial duct is not present, as exists in a small percentage of foetuses – and neonates – with tetralogy of Fallot, velocity will be accelerated across the stenotic pulmonary valve.

A unique opportunity to evaluate the presence major aortopulmonary collateral arteries exists in utero. Owing to the fact that the lungs contain

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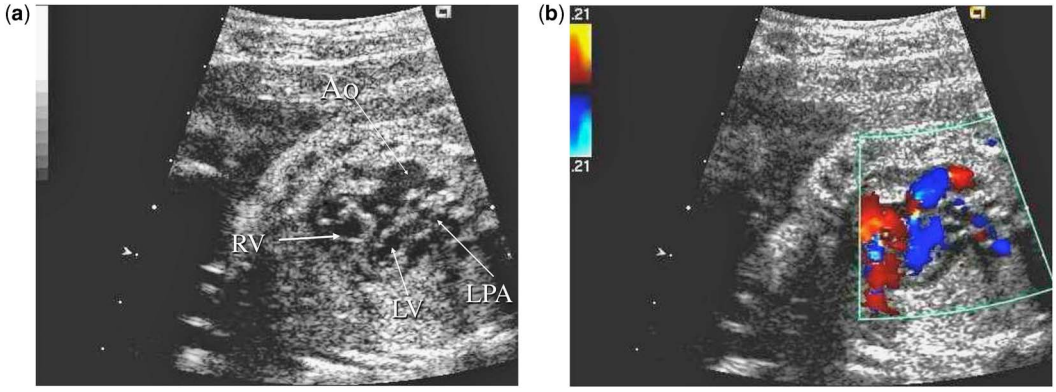

Figure 14.
*(**a**) This foetal echocardiogram was taken in a sagittal plane so that the right side of the image is more anterior and the left side of the image is more posterior. The Ao is seen to override both the LV and RV even in this 20-week gestational-aged foetus. The LPA can be seen to arise separately, not from the Ao, excluding the diagnosis of common arterial trunk. (**b**) The colour picture was taken nearly simultaneously and shows flow into both the Ao and separately into the LPA. Ao = aorta; LPA = left pulmonary artery; LV = left ventricle; RV = right ventricle.*

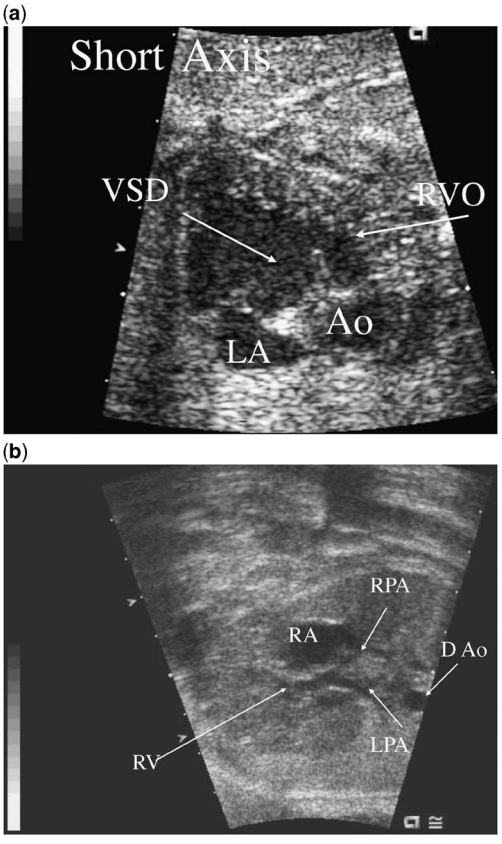

Figure 15.
*(**a**) This slightly oblique angle to the short-axis view of a foetus shows the monology of Stensen.[7] This outlet septum is deviated anteriorly towards the RVO tract, bringing the Ao to lie above the VSD and both ventricles. This feature is easily recognised by foetal echocardiography. (**b**) In this 21-week-old foetus, the outflow from the RV and narrowed ventricular outflow, small pulmonary valve, and left and right branch pulmonary arteries can already be identified. These branch pulmonary arteries bifurcate to the left and right, respectively, of the larger descending aorta (DAo). Ao = aorta; LA = left atrium; RVO = right ventricular outflow; VSD = ventricular septal defect.*

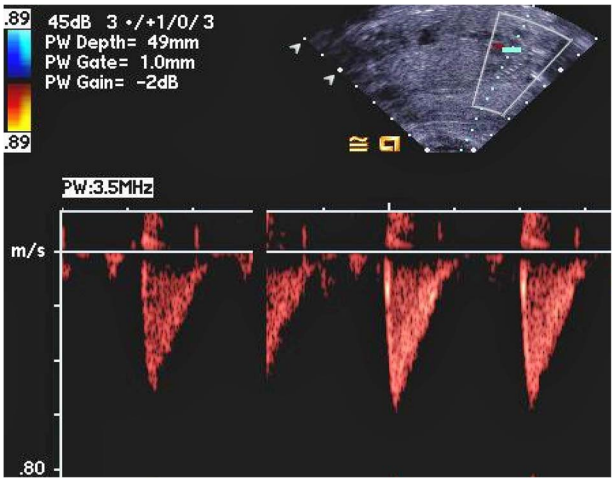

Figure 16.
This Doppler tracing was recorded in the main pulmonary artery from a foetus having a milder degree of the disorder with a widely patent arterial duct as well. The peak velocity of the signal in the main pulmonary artery is not different from the normal peak velocity because of the near equal pressures in the right ventricle and pulmonary artery.

fluid and are therefore not a barrier to sound penetration, as they will be after birth, the use of Doppler colour-flow imaging can be used to define the presence of these small vessels as they arise from the aorta (Fig 17). In the foetus, these may be identified using images parallel to the lumbar spine, but the short-axis views may also demonstrate major aorto-pulmonary collateral arteries to both lung fields.

Although the echocardiographic evaluation of tetralogy of Fallot requires complete imaging from all views, with additional information from the other modalities of ultrasound, a comprehensive description and discussion is beyond the scope of this presentation.

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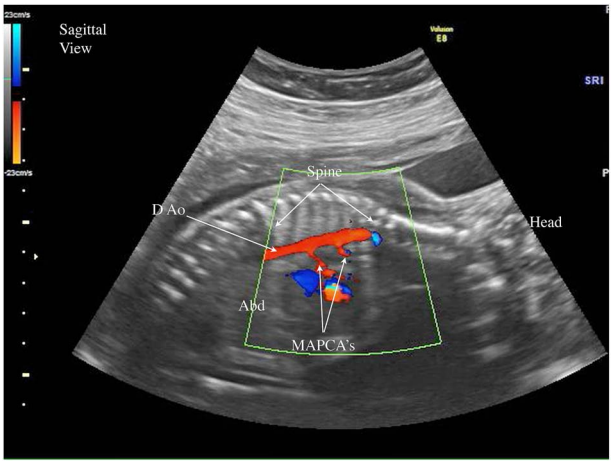

Figure 17.
This example of tetralogy of Fallot with pulmonary atresia and MAPCAs arise off the DAo. The foetus is lying with its spine facing the transducer placed on the maternal abdominal wall. Doppler colour flow imaging shows these pulmonary feeding MAPCAs to the lungs arising off the DAo and travelling their serpiginous course into the lungs. Abd = abdomen; MAPCAs = major aortopulmonary collateral arteries; DAo = descending aorta.

References

1. Silverman NH. Pediatric Echocardiography. Williams and Wilkins, Baltimore, MD, 1993: 195–204.
2. Eidem BW, Cetta F, O'Leary P. Lippincott Echocardiography in Pediatric and Adult Congenital Heart Disease, Vol. 201.Williams Wilkins, Philadelphia, PA, 2010: 237–248.
3. Lai WW, Mertens LL, Cohen MS, Geva T. Echocardiography in Pediatric and Congenital Heart Disease, Chapter 22. John Wiley and Sons, Oxford, UK, 2009: 362–384.
4. Lev M, Eckner FAO. The pathologic anatomy of tetralogy of Fallot and its variants. Dis Chest 1964; 45: 251–261.
5. Anderson RH, Paul M, Weinberg PM. The clinical anatomy of tetralogy of Fallot. Cardiol Young 2005; 15 (Suppl 1): 38–47.
6. Van Praagh R, Van Praagh S, Nebesar RA, Muster AJ, Sinha SN, Paul MH. Tetralogy of Fallot: underdevelopment of the pulmonary infundibulum and its sequelae. Am J Cardiol 1970; 26: 25–33.
7. Van Praagh R. The first Stella Van Praagh Memorial Lecture: the history and anatomy of tetralogy of Fallot. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2009; 12: 19–38.
8. Mainwaring RD, Pirolli T, Punn R, Hanley FL. Late repair of the native pulmonary valve in patients with pulmonary insufficiency after surgery for tetralogy of Fallot. Ann Thorac Surg 2012; 93: 677–679.
9. Acherman RJ, Smallhorn JF, Freedom RM. Echocardiographic assessment of pulmonary blood supply in pulmonary atresia and ventricular septal defects. J Am CollCardiol 1996; 28: 1308–1313.
10. Malhotraa SP, Hanley FJ. Surgical management of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals: a protocol-based approach Semin. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2009; 12: 145–151.
11. McElhinney DB, Reddy VM, Silverman NH, Brook MM, Hanley FL. Atrioventricular septal defect with common valvar orifice and tetralogy of Fallot revisited: making a case for primary repair in infancy. Cardiol Young 1998; 8: 455–461.
12. Poon LYC, Huggon IC, Zidere V, Allan LD. Tetralogy of Fallot in the fetus in the current era. Ultrasound Obstet Gynecol 2007; 29: 625–627.
13. Allan LD, Sharland GK, Milburn A, et al. Prospective diagnosis of 1006 consecutive cases of congenital heart disease in the fetus. J Am Coll Cardiol 1994; 23: 1452–1458.
14. Hornberger LK, Sanders SP, Sahn DJ, et al. In utero pulmonary artery and aortic growth and potential progression of pulmonary outflow tract obstruction in tetralogy of Fallot. J Am Coll Cardiol 1995; 25: 739–745.

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