Br Heart J 1982; 47: 563–72

Two dimensional echocardiographic assessment of communications between ascending aorta and pulmonary trunk or individual pulmonary arteries

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SUMMARY The value of two dimensional echocardiography in identifying communications between the ascending aorta and pulmonary trunk or individual pulmonary arteries was assessed in 24 children, all of whom had either angiocardiographic and surgical or angiocardiographic confirmation alone. Fourteen cases had truncus arteriosus, four aortopulmonary window, four anomalous origin of the left pulmonary artery from the ascending aorta, and two anomalous origin of the right pulmonary artery from the ascending aorta. It was possible to identify reliably each individual abnormality with a combination of suprasternal, precordial, and subcostal cuts. Problems only arose in differentiating truncus arteriosus from pulmonary atresia and ventricular septal defect when the main pulmonary artery and infundibular region of the right ventricle were extremely hypoplastic.

There are a number of abnormal communications in congenital heart diseases in which the pulmonary trunk or one or both pulmonary arteries communicate directly with the ascending aorta. These are truncus arteriosus, aortopulmonary window, and anomalous origin of one pulmonary artery from the ascending aorta. All of these conditions are unified morphologically by the presence of a defect between the ascending aorta and the pulmonary trunk or one or other pulmonary arteries. The aims of this study were to determine how these different conditions might be recognised and distinguished by two dimensional echocardiography.

Subjects and methods

The patients were drawn from those seen routinely at The Hospital for Sick Children, Great Ormond Street. There were 14 patients with truncus arteriosus. In all the diagnosis was prospective and had been confirmed by both angiocardiography and surgery. There were four patients with anomalous origin of the left pulmonary artery from the aorta. All had had angiocardiographic and surgical confirmation of the diagnosis. There were two patients with anomalous origin of the right pulmonary artery from the ascending aorta. In one the diagnosis was prospective and was confirmed at surgery; in the other it was retrospective after angiocardiography. There were four patients with aortopulmonary window; the diagnosis was prospective in two and retrospective in two others investigated elsewhere. All had angiographic confirmation of the diagnosis and in three surgical correction was undertaken. The fourth child died of severe pulmonary vascular disease before surgery. They were studied with an Advanced Technology Laboratory Mechanical Sector Scanner using either a 3·0 or 5 MHz scan head.

The transducer was initially placed so as to obtain a subcostal four chamber cut to assess the inlets of the heart and the size of the ventricular chambers. With clockwise rotation of the scan head a long axis of the left ventricle was obtained to assess its outlet and the presence of a ventricular septal defect. With continued clockwise rotation of the transducer the outlet of the right ventricle could be assessed.

Next the transducer was rotated anticlockwise back into the four chamber cut. With further anticlockwise rotation of the transducer, the outlet(s) from the left and right ventricles could also be assessed.

The scan head was next placed in the precordial position, so that a long axis cut of the left ventricle and its outflow tract was obtained. With slight clockwise rotation from the long axis the outlet of the right

*R H A and F J M are supported by the British Heart Foundation and, respectively, the Joseph Levy and Vandervell Foundations.

Accepted for publication 2 March 1982

ventricle could also be seen. The transducer was then placed so as to obtain a short axis cut at the level of the papillary muscles of the left ventricle. Then, with anterosuperior angulation the mitral valve and outlet of the left ventricle could be visualised in their short axis. The scan was continued superiorly above the region of the semilunar valve to visualise any potential communication at this point.

Next, an apical four chamber cut was obtained and with anterosuperior angulation the outlet(s) of the heart and a ventricular septal defect, if present, could be seen.

The transducer was then placed in the suprasternal region so that the beam was parallel to the frontal and perpendicular to the sagittal plane of the body. In this cut the right pulmonary artery could be seen in its long axis, and its communication with the arterial trunk(s) assessed. The presence of an interrupted right pulmonary artery was also sought in this view. The transducer was next rotated clockwise or anticlockwise depending on the side of the arch. When the arch was visualised in its long axis, the head and neck arteries were identified as were any other arteries arising from it. In the long axis of the aortic arch the transducer was angled slightly downwards to visualise the aortic or truncal valves. With this manipulation any potential communication close to the semilunar valves could be identified. From the long axis position the transducer was angled towards the patient's left side in an attempt to visualise the left pulmonary artery.

Results

TRUNCUS ARTERIOSUS (14 cases)

In the subcostal four chamber cut a dilated left ventricle and left atrium were seen in all cases, reflecting the large left to right shunt. In the standard subcostal long axis cut of the left ventricle the truncus appeared to originate from the left ventricle alone (Fig. 1). Where one would expect to see a perimembraneous ventricular septal defect, a thick band of muscle (the ventriculo-infundibular fold) separated the truncal and tricuspid valves (Fig. 1). Only when the transducer was rotated slightly clockwise did the ventricular septal defect appear, reaching up to the truncal valve (Fig. 1). The truncal root was large and overrode the crest of the ventricular septum. With slightly further clockwise rotation it was evident that the same great artery formed the outlet of the right ventricle. In no case was a subpulmonary outflow tract found originating from the right ventricle, nor could the pulmonary trunk be traced as a separate structure running parallel to the main arterial trunk towards the base of the heart. In this cut the left pulmonary artery was seen in all patients originating from the posterior part of the trunk (Fig. 1). The left atrial appendage was seen in close proximity to the left pulmonary artery and trunk (Fig. 1). It was only with anticlockwise rotation of the transducer following the trunk towards the patient's right, that the take-off of both pulmonary arteries could be seen. The right pulmonary artery took an acute turn towards the hilum, while the left pulmonary artery followed a more gentle course (Fig. 2). Here again the left atrial appendage could be seen. The descending aorta could be seen in its short axis sitting between the two pulmonary arteries (Fig. 2). In this view it was again evident that there was no separate subpulmonary infundibulum in the right ventricle.

In the parasternal long axis cut the trunk overrode the crest of the ventricular septum in all cases. In one case the origin of the left pulmonary artery from the trunk was seen in this cut.

With slight clockwise rotation of the transducer from the long axis, the origin of the left pulmonary artery could be seen in all cases. With further rotation into the short axis both pulmonary arteries could then be assessed as they arose from the trunk (Fig. 3).

From the suprasternal cut the right pulmonary artery was seen in its long axis, with the aorta in its short axis in the initial cut in all but one case. In the latter there was absence of the right main pulmonary artery confirmed at cardiac catheterisation (Fig. 4a). The origin of the right pulmonary artery could be seen as the transducer was rotated anticlockwise towards the long axis of the aortic arch (Fig. 4b). In the long axis cut of the aorta, the origin of the left pulmonary artery could be seen in all cases as it arose above the valve (Fig. 5). The vessel was situated on the left posterior aspect of the trunk. The right pulmonary artery was not seen in this cut as it angled acutely back behind the aorta towards the right hilum. In no case was a pulmonary valve, or a pulmonary trunk separate from the aorta, identified. Surgical or necropsy confirmation of the diagnosis was available in all cases.

ORIGIN OF THE LEFT OR RIGHT PULMONARY ARTERY FROM THE AORTA: LEFT PULMONARY ARTERY FROM THE ASCENDING AORTA (4 cases)

Two patients had pulmonary atresia and ventricular septal defect with anomalous origin of the left pulmonary artery from the ascending aorta. In one the right pulmonary artery was only present in the hilar region, being supplied by major aortopulmonary collateral arteries. In the other case the right and main pulmonary arteries were hypoplastic.

Two cases had anomalous origin of the left pulmonary artery from the aorta, the right pulmonary artery being connected to the right ventricle via a normal outflow tract and the ventricular septum being intact.

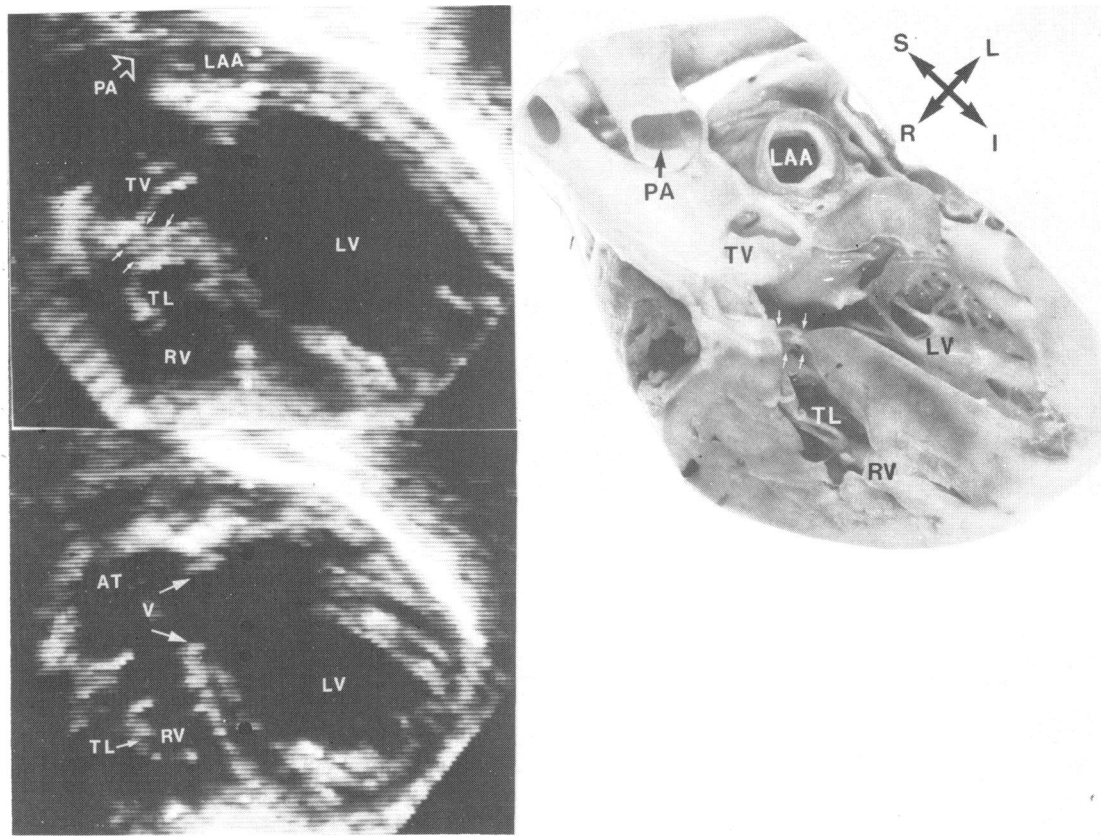

Fig. 1 *The upper left picture is a subcostal long axis cut in a case with truncus arteriosus. The small arrows show the ventriculo-infundibular fold separating the tricuspid valve and truncal cusps. Note the pulmonary artery arising from the lower aspect of the trunk. Also note the left atrial appendage. The specimen on the right is from the same case cut in the same plane. The lower picture is from the same patient with slight clockwise rotation to show the overriding trunk and the ventricular septal defect. AT, ascending trunk; LAA, left atrial appendage; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; TL, tricuspid leaflet; TV, truncal valve; V, ventricular septal defect.*

In one patient there was an associated persistent ductus arteriosus supplying the right pulmonary artery.

In all cases the left pulmonary artery arose from the posterior aspect of the ascending aorta, just above the valve (Fig. 6). It was visualised in the subcostal long axis cut and in the precordial long axis cut with slight clockwise rotation. The aortic origin was also seen in both cases in the suprasternal view (Fig. 7).

In the patients with pulmonary atresia a large perimembranous outlet ventricular septal defect was present, with the aorta overriding the septum and providing the outlet of both ventricles (Fig. 6). When the transducer was rotated further clockwise an atretic pulmonary valve could be seen in one case supported by a subpulmonary infundibulum, with a small pulmonary trunk above it. In the other patient it was not possible to find a pulmonary trunk or subpulmonary infundibulum. In neither patient could the right pulmonary artery be seen.

In the two cases without pulmonary atresia, the pulmonary valve, pulmonary trunk, and right pulmonary artery were all seen subcostally. In the precordial short axis cut the right ventricular outflow tract was visualised as it wrapped around the aorta in both cases. The persistent ductus arteriosus in one was visualised from the suprasternal approach. Angiocardiographic and surgical confirmation of the diagnosis was present in all cases.

ANOMALOUS ORIGIN OF RIGHT PULMONARY ARTERY FROM THE ASCENDING AORTA (2 cases)

In both cases there was associated pulmonary atresia with ventricular septal defect. The pulmonary trunk and subpulmonary infundibulum were extremely

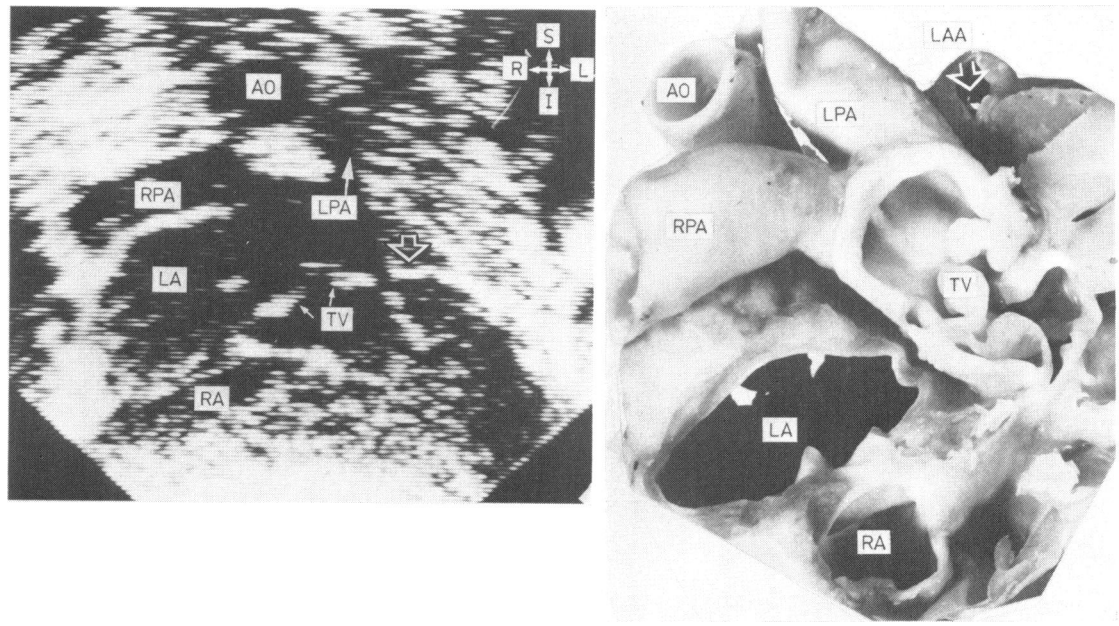

Fig. 2 *The picture on the left is a subcostal cut with anticlockwise rotation to visualise the origin of the pulmonary arteries. Note the right pulmonary artery takes an acute bend, while the left has a more gentle course. The specimen on the right is shown in the same plane. AO, aorta; LA, left atrium; LPA, left pulmonary artery; RPA, right pulmonary artery; TV, truncal valve.*

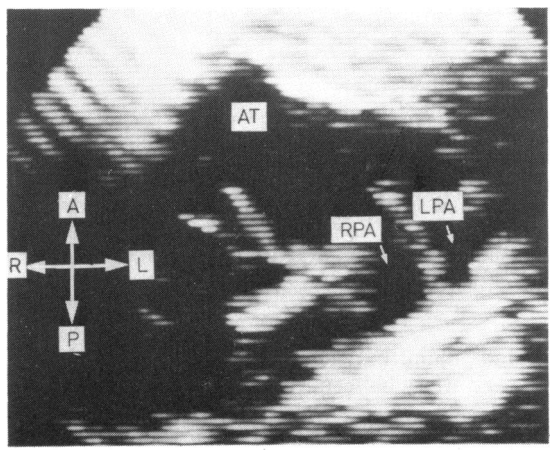

Fig. 3 *A precordial short axis cut to visualise the origin of the right and left pulmonary arteries. For abbreviations see Fig. 1 and 2.*

hypoplastic and could not be visualised echocardiographically. Angiocardiography disclosed a tiny pulmonary trunk with an atretic valve. In one case the left pulmonary artery was not connected to the pulmonary trunk, and was situated too distally to be visualised echocardiographically. The right pulmonary artery was seen arising from the ascending aorta in the suprasternal cut, originating more superiorly than in those cases with anomalous origin of the left pulmonary artery (Fig. 8). The origin of the right pulmonary artery could also be seen in the subcostal cut with anticlockwise rotation of the transducer.

In one of these cases there was an associated double aortic arch.

Regardless of which pulmonary artery originated from the aorta, the ventricular septal defect in patients with associated pulmonary atresia appeared in the subcostal long axis cut, and no ventriculoinfundibular fold was identified (Fig. 6).

AORTOPULMONARY WINDOW (4 cases)

Four cases were seen with aortopulmonary window, all with surgical or necropsy confirmation. In the suprasternal long axis cut of the aorta, the communication between the ascending aorta and main pulmonary artery could be seen in all cases (Fig. 9). The appearance was different from those cases with either truncus arteriosus or anomalous origin of the pulmonary artery from the aorta (compare Fig. 5, 6, and 9). As the lower border of the ascending aorta was followed superiorly the communication could be seen, appearing like a semicircle, instead of the usual circular pattern of the main pulmonary artery. In one patient a coexisting persistent ductus arteriosus (confirmed at surgery) was visualised by this approach.

In the precordial short axis cut with slight anterosuperior angulation, the window could also be visualised (Fig. 10). From the same cut the pulmo-

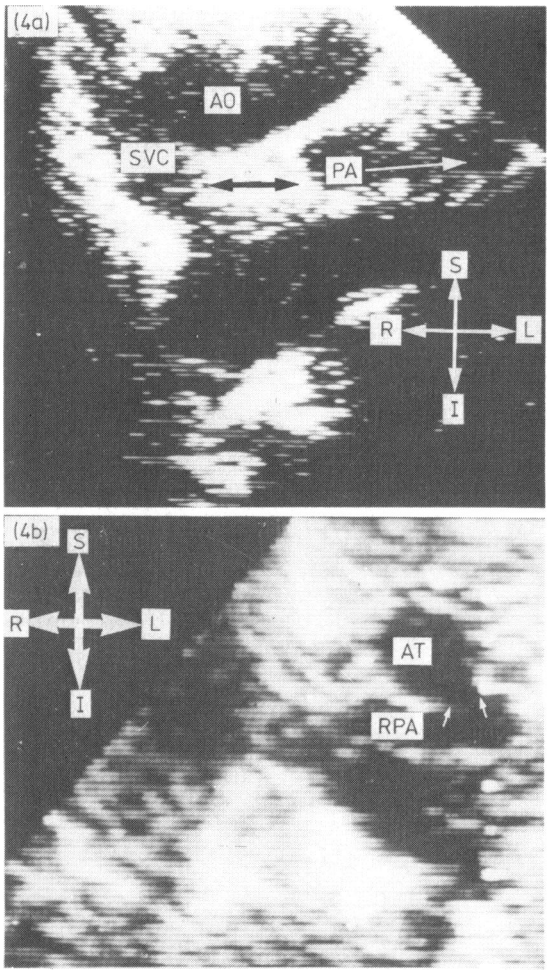

Fig. 4 (a) *Suprastructural cut in a case with truncus arteriosus and absence of a right main pulmonary artery. The black arrows indicate the region where the normal right pulmonary artery usually runs.* (b) *A suprasternal cut with slight anticlockwise rotation to visualise the origin of the right pulmonary artery from the ascending trunk. The small arrows indicate the site of communication. SVC, superior vena cava. For the other abbreviations see Fig. 1 and 2.*

nary trunk and right pulmonary artery could be imaged, along with the infundibular region. The widow was not seen in the parasternal long axis cut.

In the standard subcostal long axis cut the aortopulmonary window was seen in two cases. The window was reliably visualised in all four, only when the transducer was rotated anticlockwise into a position where the ascending aorta was seen in its long axis with the pulmonary trunk crossing it (Fig. 11). Care was required to avoid misinterpretation of the region where the right pulmonary artery and aorta crossed, as drop-out in this region could be mistaken for an aortopulmonary window. This was avoided by ensuring that only the region where the main pulmonary artery and aorta crossed was used to visualise the aortopulmonary window.

Discussion

The group of malformations described above has one anatomical feature in common, namely a communication between the ascending aorta and the pulmonary trunk or one or both pulmonary arteries. Angiographically, each would be characterised by direct opacification of either the pulmonary trunk, or the right or left pulmonary arteries from an ascending aortogram.[1 2] As it turns out, the logical process whereby these different anatomical substrates can be distinguished is precisely the same for cross-sectional echocardiography as for angiography.

The first step is to identify aortopulmonary window by demonstrating that two semilunar valves and two ventricular outflow tracts are present. To accomplish this, a precordial and subcostal cut should be used to demonstrate the ventricular outflow tracts and the separate arterial valves. The precordial cut in isolation[3] will diagnose aortopulmonary window in most cases, but has been misleading in our experience in a case of left coronary artery to coronary sinus fistula. This may seem at first sight surprising, but the appearances in the precordial short axis cut are remarkably similar. This experience emphasises the importance of always using multiple cuts, and identifying the origin of the coronary arteries from the aorta.[4]

The suprasternal cut is of great value as the appearance differs from cases with truncus arteriosus or anomalous origin of one pulmonary artery from the aorta. Normally the main pulmonary artery is seen as a circle, lying beneath the aortic arch. In patients with an aortopulmonary window a semicircle is present, the absent upper part representing the window.

The next step is to identify typical truncus arteriosus, in which both pulmonary arteries originate from the solitary arterial trunk sometimes via a short main pulmonary artery. This is easily achieved with angiography projections, and is equally readily shown echocardiographically. In the subcostal cut this is usually obvious. The subcostal long axis cut allows visualisation of the origin of the left pulmonary artery from the aorta. Care must be taken not to mistake the left atrial appendage for the other pulmonary artery (Fig. 12). The left atrial appendage lies in close proximity to the left pulmonary artery and can be seen entering the left atrium lying behind the ascending trunk. The appendage is also seen in patients with ventriculoarterial concordance, but the presence of the left pulmonary artery arising from the trunk with

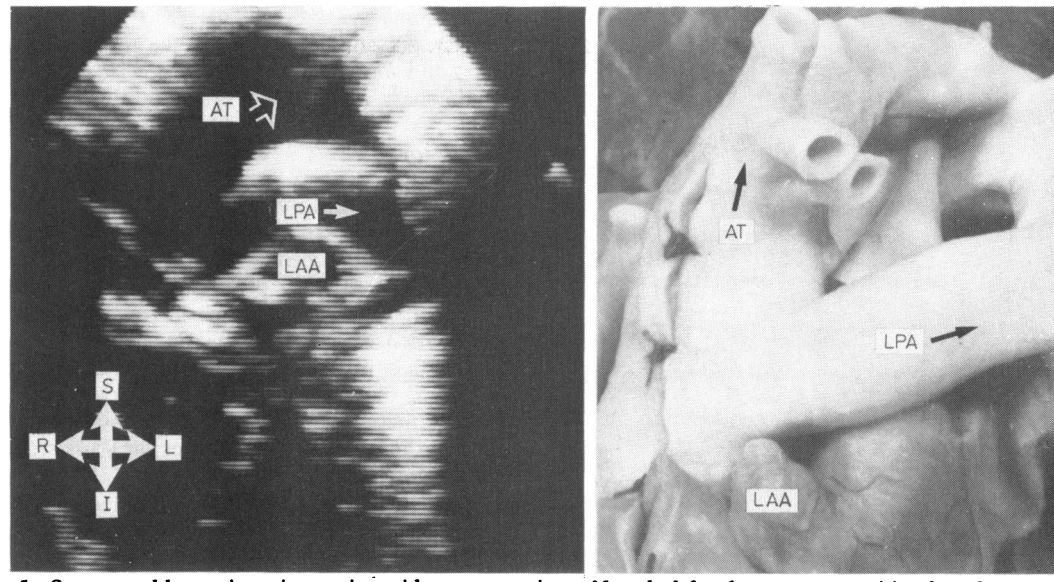

Fig. 5 *Suprasternal long axis cut in a patient with truncus arteriosus. Note the left pulmonary artery arising from the posterior aspect of the ascending trunk. In addition note the relation of the left atrial appendage to the left pulmonary artery. The specimen on the right is from the same case showing the echocardiographic features. For abbreviations see Fig. 1 and 2.*

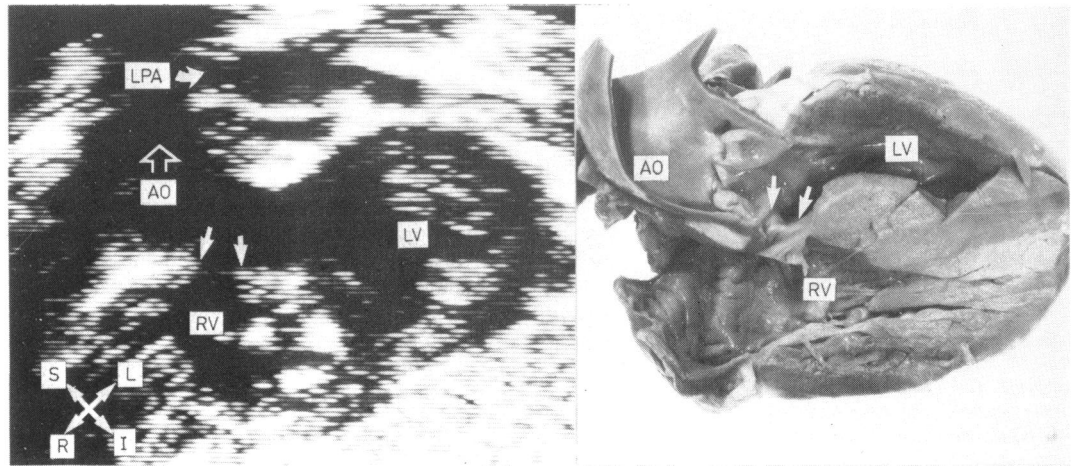

Fig. 6 *The picture on the left is from a case with pulmonary atresia and ventricular septal defect, with anomalous origin of the left pulmonary artery from the aorta. The arrows point to the ventricular septal defect. The specimen on the right is from a different case with pulmonary atresia and ventricular septal defect. Note the region of continuity between the tricuspid valve and aortic valve. For abbreviations see Fig. 1 and 2.*

the appendage overlapping it provides this misleading picture. Both pulmonary arteries may be visualised simultaneously when the transducer is rotated clockwise following the origin of the trunk. Likewise, further confirmation of bilateral truncal origin can be obtained in the precordial short axis cut.

What is harder to demonstrate by angiocardiography or echocardiography is the presence or absence of a short main pulmonary arterial segment. Echocardiographically, this is because the left and right pulmonary arteries form a sharp angle with one another, just as they do when they originate from a normal pulmonary trunk. In all the cases with right and left pulmonary arteries studied, the arteries arose directly from the solitary trunk, albeit in close approximation to one another. The echocardiographic techniques also all showed a single semilunar valve which in these cases overrode the ventricular septum. It must be

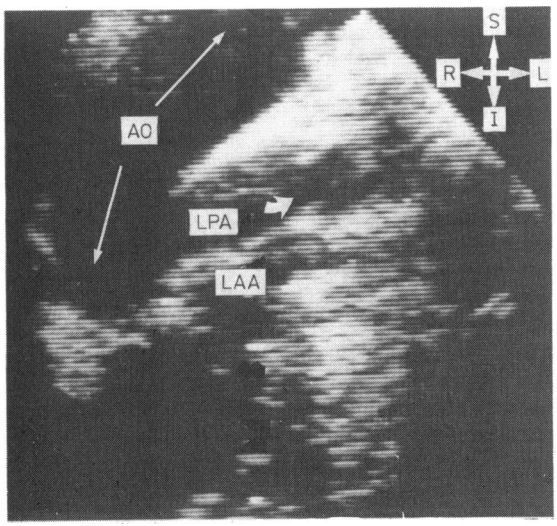

Fig. 7 *A suprasternal cut in a case with pulmonary atresia and ventricular septal defect with anomalous origin of the left pulmonary artery from the aorta. Note the appearance is similar to that seen in Fig. 5 in a case of truncus arteriosus. For abbreviations see Fig. 1.*

remembered, however, that monoventricular origin of a truncus has been described,[5 6] so overriding need not necessarily be present. In the hearts studied no evidence of a blind infundibulum from either ventricle was identified, nor was a second arterial trunk found extending from the base of the heart.[5]

In the suprasternal cut, it has been suggested[7] that the main pulmonary artery segment is imaged originating from the ascending trunk. Our anatomical studies, together with the well-recognised observation of a high arching left pulmonary artery in truncus arteriosus, suggest that it is usually the left pulmonary artery that gives rise to this appearance. This is important, since the very same feature is seen in anomalous origin of the left pulmonary artery alone in the absence of truncus arteriosus (compare Fig. 5 and 6). This of course would not apply in the case where there was a reasonable sized main pulmonary artery segment.

Aortopulmonary window and typical truncus arteriosus are therefore readily recognised. When only one pulmonary artery originates from the ascending aorta, however, two problems arise, one related to recognition of discontinuity of the pulmonary arteries, and the other to hypoplasia of one pulmonary artery.

The first problem is exemplified by those cases in which the left pulmonary artery arose from the ascending aorta, and the right from the right ventricle

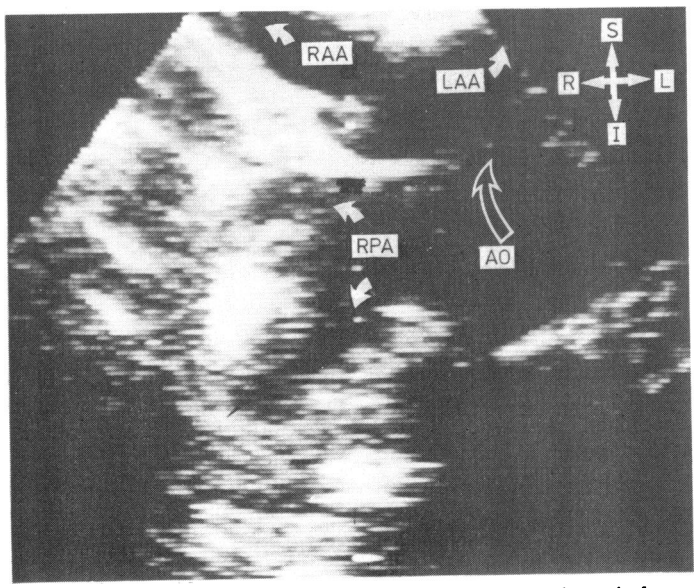

Fig. 8 *A suprasternal cut in a case with pulmonary atresia and ventricular septal defect with anomalous origin of the right pulmonary artery from the ascending aorta. This patient also had a double aortic arch. LAA, left aortic arch; RAA, right aortic arch. For other abbreviations see Fig. 2.*

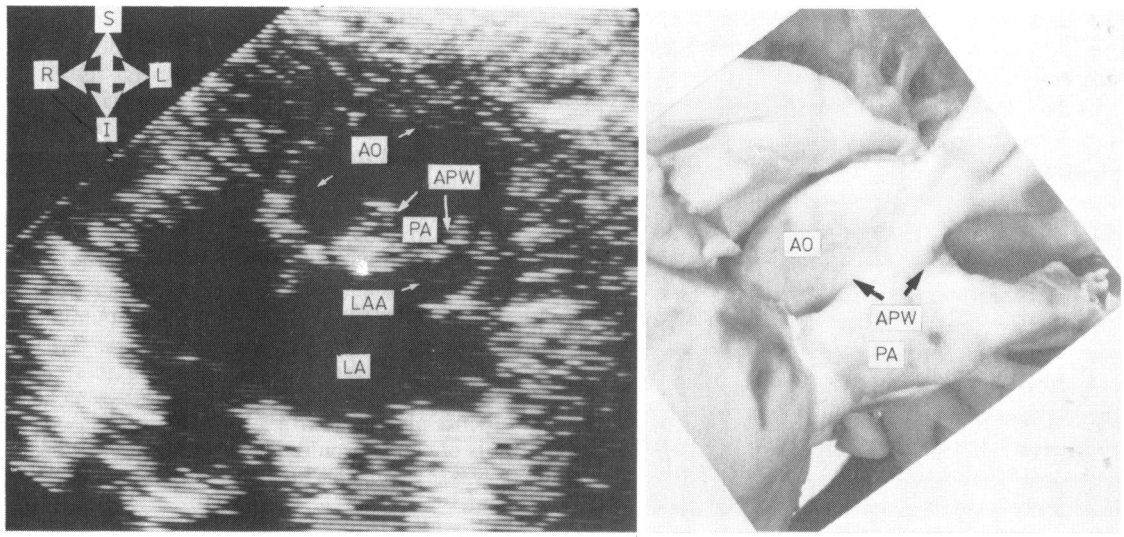

Fig. 9 *The picture on the left is a suprasternal cut in a case with aortopulmonary window. Note the pulmonary artery has a semicircular appearance, instead of its normal circular shape. The specimen on the right from a different case shows the aortopulmonary window. This patient also had an interrupted aortic arch. APW, aortopulmonary window. For other abbreviations see Fig. 1 and 2.*

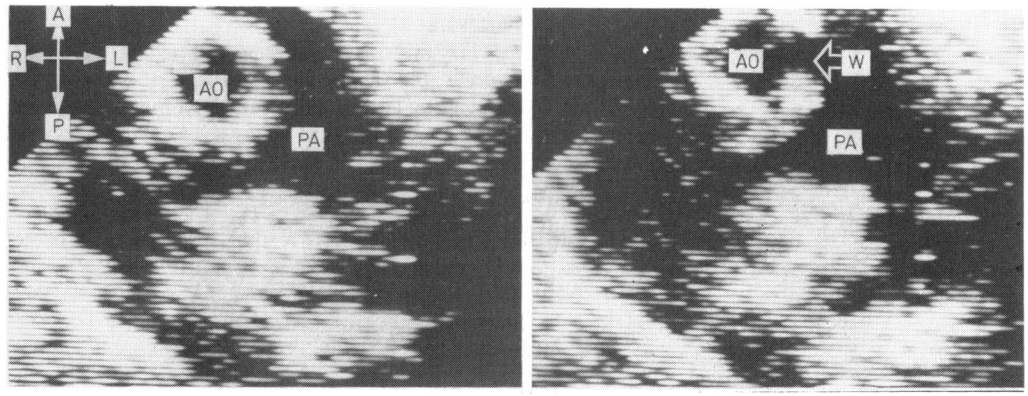

Fig. 10 *Precordial short axis cut in a case with aortopulmonary window. The picture on the left shows a normal right ventricular outflow tract. The image on the right is obtained with slight clockwise rotation to visualise the window. W, window. For other abbreviations see Fig. 2.*

in the presence of an intact ventricular septum. In the subcostal and precordial short axis cuts, the right pulmonary artery is well visualised in its normal position, and lack of demonstration of bifurcation of the pulmonary trunk may be incorrectly ascribed to the frequent and normal manner in which the left pulmonary artery turns out of the plane of the tomographic cut as it leaves the right pulmonary artery. This error may be avoided by using the suprasternal or high short axis view, provided that care is taken to trace the right pulmonary artery to the expected position of the left pulmonary artery. As the presumed site of the bifurcation is reached, the right pulmonary artery, instead of continuing into the left, appears to terminate abruptly. Though we have not observed cases with isolated anomalous origin of the right pulmonary artery from the aorta, we presume that this diagnosis would be readily made because of the abnormal appearance of the right pulmonary artery in the standard subcostal and precordial cuts.

The second problem, that of extreme hypoplasia of the pulmonary artery not arising from the aorta, occurs when there is an interventricular communication and pulmonary atresia. If the pulmonary outflow

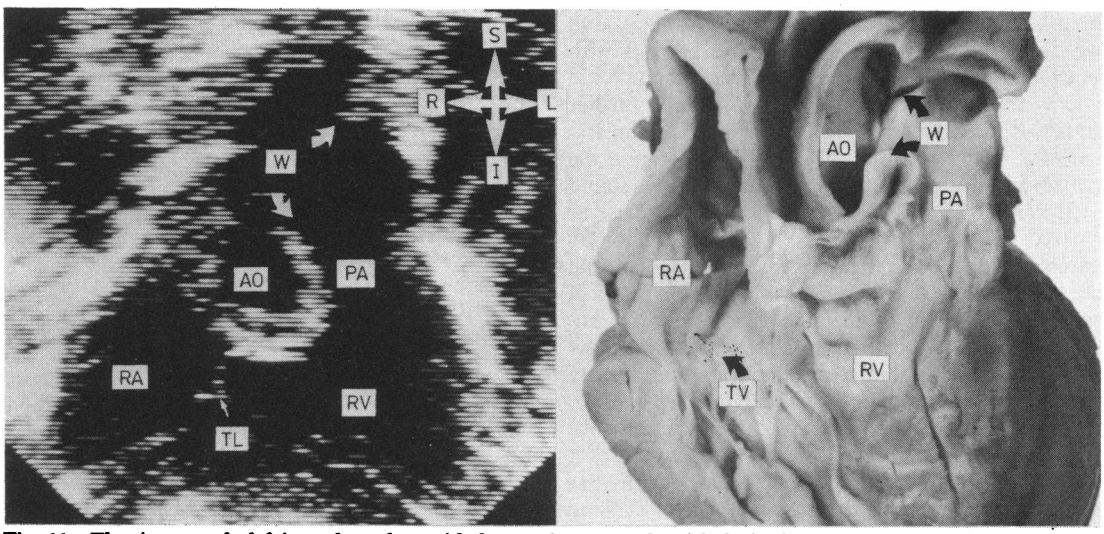

Fig. 11 *The picture on the left is a subcostal cut with the transducer rotated anticlockwise following the outlets of heart. Note the large window indicated by the arrows. The specimen on the right is from a different case, but cut in the same way to visualise the aortopulmonary window. RA, right atrium. For other abbreviations see Fig. 1, 2, and 10.*

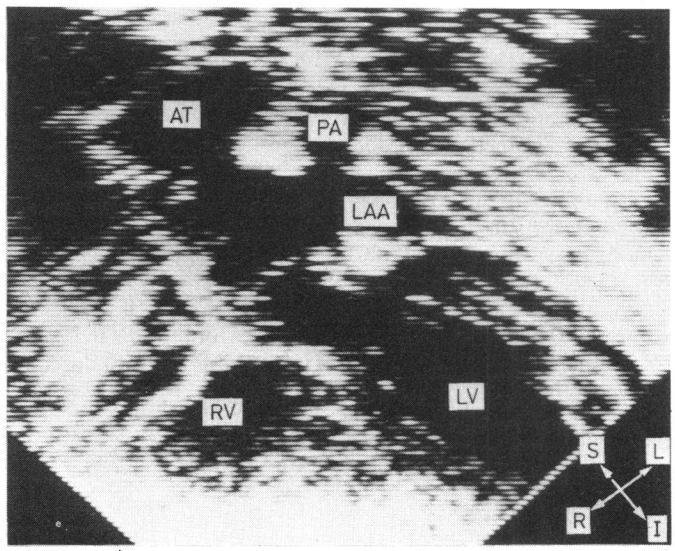

Fig. 12 *Subcostal long axis cut in a case with truncus arteriosus. Note the origin of the left pulmonary artery. Observe the left atrial appendage which overlaps the left pulmonary artery giving a false impression of truncal origin. For abbreviations see Fig. 1 and 2.*

tract is merely stenotic, then though the pulmonary trunk will be hypoplastic, it should be possible to identify it separately from the aortic trunk.

When there is pulmonary atresia, however, the pulmonary trunk may be so grossly hypoplastic (or atretic) as to be unidentifiable. Under these circumstances, just as with angiocardiography, it may be impossible to differentiate between pulmonary atresia with ventricular septal defect and truncus arteriosus.[5] Identification of a blind right ventricular outflow tract will then establish the diagnosis as pulmonary atresia with ventricular septal defect.[7] Failure to demonstrate such an outflow tract, however, does not necessarily establish the diagnosis of truncus, both because of the

limits of resolution of the equipment and because in two out of 22 specimens of pulmonary atresia with ventricular septal defect studied at necropsy, no blind right ventricular outflow tract was found.[5] A further guide to differentiation between the two is demonstration of a ventriculo-infundibular fold separating the tricuspid from the arterial valve, since this was found in 51 out of 66 (77·3%) of cases with truncus, as opposed to four out of 24 (16·7%) with pulmonary atresia.[5] Finally, the demonstration of more than three semilunar valve cusps[8] favours the diagnosis of truncus. In cases of anomalous origin of one pulmonary artery from the ascending arterial trunk, however, the distinction between truncus and pulmonary atresia with ventricular septal defect is academic. What *is* important is to distinguish between absence of the contralateral pulmonary artery and atresia of it somewhere proximal to the hilum. If echocardiography cannot demonstrate the pulmonary artery arising from the ascending trunk, or show a persistent ductus running to the hilum on that side, then angiocardiography, possibly including pulmonary vein wedge angiography, will be the only method by which a hilar pulmonary artery, if present, can be demonstrated, short of exploratory surgery.

Thus, by the application of cross-sectional echocardiography, it is possible reliably to identify a defect between the ascending aorta and pulmonary trunk or one or other pulmonary arteries. Truncus arteriosus and aortopulmonary window are readily separated, as are those cases where a pulmonary artery arises anomalously from the ascending aorta in the presence of a normal main pulmonary artery and valve. Problems, however, may arise in differentiating truncus arteriosus from pulmonary atresia and ventricular septal defect when the main pulmonary artery and infundibular region of the right ventricle are extremely hypoplastic. This difficulty may also be encountered when using more traditional techniques of investigation.

References

1 Edwards JE, Carey LS, Neufeld HN, Lester RG. *Congenital heart disease; correlation of pathologic anatomy and angiocardiography*. Philadelphia & London: W B Saunders, 1965: 289–305.

2 Calder L, Van Praagh R, Van Praagh S, *et al.* Truncus arteriosus communis: clinical, angiocardiographic, and pathologic findings in 100 patients. *Am Heart J* 1976; **92**: 23–38.

3 Satomi G, Nakamura K, Imai Y, Takao A. Two-dimensional echocardiographic diagnosis of aorticopulmonary window. *Br Heart J* 1980; **43**: 351–6.

4 Yoshikawa J, Yanagihara K, Owaki T, *et al.* Cross-sectional echocardiographic diagnosis of coronary artery aneurysms in patients with the mucocutaneous lymph node syndrome. *Circulation* 1979; **59**: 133–9.

5 Crupi G, Macartney FJ, Anderson RH. Persistent truncus arteriosus: a study of 66 autopsy cases with special reference to definition and morphogenesis. *Am J Cardiol* 1977; **40**: 569–78.

6 Thiene G, Bortolotti U, Galluci V, Terribile V, Pellegrino PA. Anatomical study of truncus arteriosus communis with embryological and surgical considerations. *Br Heart J* 1976; **38**: 1109–23.

7 Hagler DJ, Tajik AJ, Seward JB, Mair DD, Ritter DG. Wide-angle two-dimensional echocardiographic profiles of conotruncal abnormalities. *Mayo Clin Proc* 1980; **55**: 73–82.

8 Houston AB, Gregory NL, Murtagh E, Coleman EN. Two-dimensional echocardiography in infants with persistent truncus arteriosus. *Br Heart J* 1981; **46**: 492–7.

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