Cineangiocardiology in tetralogy of Fallot

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SUMMARY The advantages of angled angiographic projections are demonstrated in a review of 55 cases of tetralogy of Fallot. Particular attention was paid to the origins of the branch pulmonary arteries, whose diameter was measured and compared with that of the descending aorta, showing that in 12 per cent there was narrowing to below 40 per cent of descending aortic diameter. Right aortic arch was present in seven cases (13%) and a major coronary artery anomaly in two (4%).

The four features of tetralogy of Fallot are not a coincidental grouping. Deviation of the conotruncal septum to the pulmonary side results in its non-union with the ventricular septum, hence the ventricular septal defect. Malalignment between the conotruncal (infundibular) septum and the ven-

tricular septum results in aortic overriding. The compound pulmonary stenosis can be thought of as being the result of hypoplasia of the pulmonary side of the dividing conotruncus, with subsequent muscular hypertrophy. The pulmonary hypoplasia may extend to the point where the embryonic right and left branch pulmonary arteries join the sixth aortic

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Fig. 1 Normal right ventriculogram, right anterior oblique view. The normal crista supraventricularis is an unbroken line from its junction with the tricuspid valve ring (lower arrow) to its junction with the pulmonary valve ring (upper arrow). Right pulmonary artery is well shown passing across the spinal shadow. Left pulmonary artery is obscured.

Fig. 2 Normal right ventriculogram, left anterior oblique view. This view profiles the interventricular septum (straight arrows) and displays the left pulmonary artery (note the very mild narrowing of the origin of left pulmonary artery indicated by curved arrows—this is not unusual in the normal).

arch, and so may involve the origins of the right and left pulmonary arteries. Right ventricular hypertrophy is a secondary phenomenon.

Selective angiocardiology has been used for over 30 years in the investigation of patients with tetralogy of Fallot.¹ Traditionally biplane investigations have been performed with the patient supine, filming in the frontal (AP or PA) and lateral projections.^{2,3} In addition, techniques of oblique projection have been in use for a number of years (P W T Brandt, 1975, personal communication) culminating in the more recent concept of axial angiocardiology as described by Barger and Elliott and their colleagues.^{4,5} These projections have shown that in many patients with tetralogy of Fallot, frontal and lateral filming has furnished inadequate information particularly regarding the site and size of the ventricular septal defect or defects, and the size of the main and distal pulmonary arterial tree.

Since 1976 we have used angled projections for the angiocardiology investigation of all our

patients with congenital heart disease and the purpose of this report is to present our findings with specific respect to tetralogy of Fallot.

All studies were performed on a standard biplane cineangiographic installation with a ceiling mounted overhead intensifier and undercouch x-ray tube, giving posteroanterior and left lateral projections when the patient was supine. The simple oblique projections were produced by raising the patient's right side with radiolucent wedges, giving the ceiling intensifier a right anterior oblique (RAO) and the horizontal intensifier left anterior oblique (LAO). Added axial tilt in the LAO view was produced by angulating the patient to his left across the table, as described by Barger *et al.*⁴ in the "long axial technique"; this effectively angles the x-ray tube to emit x-rays in a caudocranial direction. Our patients were small enough to allow tilt of up to 40°. When tilt was required in the posteroanterior view, the supine patient was half sat up using wedges, to produce the view on the overhead intensifier caudocranial projection.

Fig. 3 Tetralogy of Fallot, right anterior oblique view. The tricuspid valve ring is reinforced by a dotted line. The line of the crista is broken; the infundibular septum (IS) is seen displaced away from the tricuspid ring and, by virtue of the ventricular septal defect, contrast outlines its posterior margin which is continuous with the aorta. Contrast in the left ventricular outflow tract defines the atrioventricular septum (straight arrows) and the belly of the aortic valve (hollow arrow) to confirm that the aorta is normally related to the left ventricle. The right ventricular outflow tract is stenosed by muscular hypertrophy of the infundibular septum and the free wall opposite it.

Right ventriculography

The appearances of the normal right ventricle in right and left anterior oblique projections are shown in Fig. 1 and 2. Note that the origins of the right and left branch pulmonary arteries, the outflow tract, and the interventricular septum are shown to greater advantage than on frontal and lateral views. In general, the right anterior oblique view needs to be about 25°; with right anterior oblique on the overhead intensifier in installations where the intensifiers are fixed at right angles, the horizontal intensifier will show 65° left anterior oblique which is often too shallow to profile the ventricular septum which in tetralogy frequently needs 80° left anterior oblique. Some installations will allow the intensifiers to be set at angles greater than 90° to each other, particularly the modern U and C arm installations. Alternatively, the right anterior oblique may be kept at 20° and a mild degree of caudocranial tilt added to the left anterior oblique, lowering the overlapping apical septum, thus providing a better view of the basal structures, as will be seen from some of the illustrations.

Displacement of the infundibular septum is best demonstrated on the right anterior oblique view. The normal continuity of the crista between pulmonary valve ring and the upper limit of the tricuspid ring is broken; the line of the infundibular septum is continuous with the anterior wall of the aorta, and as such defines one aspect of the boundaries of the ventricular septal defect. The posterior limit of the discontinuity may be the tricuspid ring

Fig. 4 (a) Tetralogy, right ventricle, right anterior oblique view, early frame. As contrast fills the right ventricle, early flow into the base of the ventricle defines the tricuspid valve ring (black arrows) and a small shelf of residual bulboventricular flange (white arrow) between it and the overriding aorta. This flange is not a constant feature of tetralogy. (b) Later frame. Contrast has now spread via the ventricular septal defect to the left ventricular outflow tract (arrowed), obscuring the earlier features of Fig. 4a.

Fig. 5 Ordinary subcrystal ventricular septal defect, right ventricular injection, right anterior oblique view. The negative shadow of the ventricular septal defect, caused by non-opacified blood streaming through it, is arrowed. The line of the crista above the ventricular septal defect is convex upwards but is unbroken.

Fig. 6 Tetralogy variation. Occasionally the infundibular septum fails to hypertrophy (arrowed). The pulmonary stenosis is mostly valvar.

(Fig. 3) or remnants of the bulboventricular flange (Fig. 4a and b). One of the advantages of cine radiography is that this posterior limit can be defined before the area is obscured by the left ventricular outflow tract. Fig. 5 shows that in an ordinary subcrystal perimembraneous ventricular septal defect, the line of the crista, though deviated, is not broken. Discontinuity is present in the rare "malalignment ventricular septal defect" which can be thought of as tetralogy without pulmonary stenosis.

Fig. 7 Tetralogy variation. Low, fixed infundibular stenosis with virtually normal pulmonary valve and pulmonary arteries.

The right anterior oblique view also shows the right ventricular infundibulum clear of the aorta allowing an assessment of its size, muscular hypertrophy, and lability of stenosis. Since both sides of the infundibular septum are seen, its hypertrophy can be accurately determined. The normal crista gives a spurious impression of bulk (Fig. 1). Occasionally in tetralogy the infundibular septum fails to hypertrophy, disclosing its essential hypoplasia (Fig. 6). The infundibular stenosis is usually labile, but there is a wide spectrum of appearances and occasionally a fixed low infundibular stenosis may be seen (Fig. 7).

Deviation of the infundibular septum in the manner described requires not only the presence of a sizeable ventricular septal defect beneath it, but also some degree of overriding of the aorta across the ventricular septum. Furthermore, visualisation of continuity of the aorta with the atrioventricular

septum (Fig. 4) will confirm that, despite the override, the anatomical relation of the aorta with the left ventricle is not disturbed. As far as surgery is concerned, the degree of override is unimportant once it is established that the ventricular septal defect is in the subaortic position.

Role of the left anterior oblique projection is therefore as follows.

- (i) It confirms great vessel relations to the ventricles, the size of the ventricular septal defect, and the degree of aortic override, by profiling the interventricular septum (Fig. 8, 9).
- (ii) It demonstrates the origin of the left pulmonary artery (Fig. 10, 11).
- (iii) It detects ventricular septal defects in atypical positions (Fig. 12).
- (iv) It detects any relative hypoplasia of the left ventricle (Fig. 13).

Fig. 8 Tetralogy, left anterior oblique. The interventricular septum is profiled and the ventricular septal defect is shown to be between the crest of the septum (lower arrow) and the mildly overriding aortic valve cusps (upper arrows). rv, right ventricle; lv, left ventricle.

Fig. 9 Tetralogy, left anterior oblique view. Similar view to Fig. 8, showing severe aortic overriding.

Fig. 10 Tetralogy, left anterior oblique view, showing a moderate narrowing of the origin of left pulmonary artery (arrowed).

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Occasionally, right ventricular pressure is supra-systemic, indicating a restrictive ventricular septal defect. This appears to be nearly always the result of partial membranous closure of the defect. At operation we have noted that redundant tricuspid tissue is responsible. In this situation it is desirable to examine the ventricular septal defect closely at angiography; two right ventricular injections may have to be made to show the "wind-sock" of membranous tissue which often is best seen in the lateral projection (Fig. 14). The view can be combined with an anteroposterior view with tilt for the pulmonary arteries as described below.

Pulmonary arteries

The increasing awareness of hypoplasia of the main and proximal pulmonary arteries in tetralogy lends support to the unifying theory that all the features

Fig. 11 *Tetralogy, left anterior oblique view. This frame illustrates that added caudocranial tilt to the left anterior oblique view can enhance visualisation of the pulmonary arteries, and also that early ectopics during contrast injection can cause the pulmonary arteries to fill before the aorta. Note that the angle of tilt brings the right hemidiaphragm up to mid-frame (arrows).*

Fig. 12 *Muscular ventricular septal defect. A small muscular ventricular septal defect (small arrows) clearly fills the left ventricle separately from the main ventricular septal defect (large arrow) which is in the usual tetralogy position. Early frame, right ventricular injection, left anterior oblique view.*

Fig. 13 *Relative hypoplasia of the left ventricle in a case of severe tetralogy of Fallot.*

Fig. 14 Obstructed ventricular septal defect. Membranous tissue obstructs the ventricular septal defect, the contrast sharply outlining its margins (arrowed). There is no flow into the left ventricle. The original cine film clearly showed the flexible "wind-sock" appearance of the membranous tissue. Right ventricular injection, lateral view.

Fig. 15(a)

Fig. 15(b)

Fig. 15(c)

Fig. 15 Three views of the pulmonary arteries, posteroanterior view with caudocranial tilt. (a) Well formed pulmonary arteries. Right and left pulmonary arteries are only fractionally smaller than the main pulmonary artery. (b) A typical tetralogy; main pulmonary artery is short and narrow but the branch pulmonary arteries are well formed. Right ventricular outflow tract injection; note the doming of the pulmonary valve. (c) Severe pulmonary artery hypoplasia, extending well into the branch pulmonary arteries. Note that the branch arteries regain a reasonably normal size at the hilar regions.

Fig. 16 Histogram showing the sizes of the right and left branch pulmonary arteries at the hilum, relative to the descending aorta; note a relatively close distribution around the 1:1 ratio.

Fig. 17 Histogram showing the sizes of the origins of the right and left branch pulmonary arteries to the descending aorta. Note the wider distribution compared to Fig. 16 and the bias towards a smaller than 1:1 relation.

Fig. 18 Wedged pulmonary venous angiography shows a good sized isolated left pulmonary artery which did not opacify from either the right ventricle or aortic angiograms.

of tetralogy stem from hypoplasia of the pulmonary components of the bulbus, truncus arteriosus, and sixth aortic arch. Narrowing of these vessels has long been recognised on routine frontal and lateral angiocardiology.

We suggest that the routine use of oblique views for right ventriculography provides a more reliable demonstration of these narrowings. Furthermore we believe that in cases where the pulmonary arteries seem particularly small, or when they are not well seen on the initial angiogram, a specific view is advisable. To date, we have found that the postero-anterior view with up to 45° caudocranial tilt is most appropriate (the "sitting up" view). Direct injection into the main pulmonary artery is desirable, but this usually is only advisable when a surgical shunt is present to help avoid the possibility of an induced cyanotic spell. Failing this, the injection should be made close to the right ventricular outflow tract, and the production of ectopic beats should be viewed as an advantage as this usually causes the pulmonary arteries to fill first (Fig. 11). Fig 15 shows how the extent of hypoplasia of the pulmonary arteries can vary considerably. This hypoplastic effect very rarely extends as far as the hilum.

Fig. 16 shows the distribution of distal branch pulmonary artery size at the hilum relative to the size of the descending aorta in our group of patients.

Fig. 19 Anterograde aortic angiogram, lateral view. The coronary arteries are well demonstrated; left anterior descending is usually smaller than normal (arrowed).

Fig. 20 Selective right coronary angiogram, left anterior oblique view, showing a single coronary artery; the left coronary (arrowed) arises from the proximal right coronary and passes in front of the right ventricular outflow tract to divide into left anterior descending and left circumflex arteries.

The ratios were derived by measurement on the angiogram of the vessels and dividing the pulmonary diameter by aortic. The ratios are tightly distributed around the 1:1 mark. Fig. 17 shows the same format applied to the origins of the branch pulmonary arteries; note the biased distribution and the frequency of severe narrowing.

Occasionally the branch pulmonary arteries may be isolated from the main pulmonary artery and be fed from primary aortic-pulmonary collaterals or by a persistent ductus arteriosus. We have found the wedged pulmonary venous angiogram to be the best method of visualising these vessels when they are not well seen on aortography (Fig. 18). The principle of this procedure is as described by Singh and colleagues,⁶ but we use a firmly wedged end-hole catheter and a hand injection of contrast.

Aorta and coronary arteries

Aortography is of considerable value in tetralogy of Fallot and has become a routine in our unit. We find that either the patient is a likely candidate for a shunt procedure or for total correction. In the first

Fig. 21 Anterograde aortogram, lateral view, showing a single coronary artery arising from the left posterior sinus. The right coronary artery (arrowed) arises from left anterior descending and passes anterior to the right ventricular outflow tract to gain the right atrioventricular groove.

instance it is desirable to document the size and position of the aortic arch and its branches, and in the latter, prior knowledge of coronary artery distribution can facilitate the surgical repair. In our series of 55 cases, seven had a right aortic arch, and one had a left arch with an aberrant right subclavian artery. Though only two had a major coronary artery anomaly, our experience is that a normally arising but unusually large conal branch can also complicate a right ventriculotomy.

Our present aortographic technique is to position the transvenous catheter just above the aortic sinuses via the right ventricle and ventricular septal defect. Using the largest convenient catheter, power injection, and electrocardiographic triggering, a satisfactory angiogram can be produced with 0.75 ml contrast medium/kg (Fig. 19). Postero-anterior and lateral views are employed, the former with a wide field to display the branches of the aorta, and the lateral with greater magnification to demonstrate the coronary arteries.

From time to time we have performed retrograde aortography, both with standard catheters and with ring catheters designed to sit in the aortic sinuses. Arterial puncture, however, is seldom necessary in tetralogy because of easy transvenous access to the aorta, and we now consider that a retrograde study simply for coronary anatomy is seldom indicated.

Any coronary artery anatomy that places a large branch artery across the high right ventricular outflow tract will complicate the right ventriculotomy at total correction. Typically, the vessel is an anomalous left anterior descending branch arising from the right coronary artery that either replaces the normal branch from the left or shares its distribution. Most serious are the single coronary arteries, examples of which are shown in Fig. 20 and 21.

Comment

The many aspects of tetralogy of Fallot discussed here make it more of a challenge to the cardio-

angiographer than most congenital cardiac pathologies. The fine detail required for the coronary arteries and the pulmonary arteries will probably ensure that radiology will remain the only adequate diagnostic method, despite the considerable improvement in ultrasonic and other non-invasive techniques in the past few years. In turn, the angiographic techniques employed must continue to be critically appraised in the light of the current therapeutic approach.

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