Variations in clinical presentation of Fallot's tetralogy in infancy

Angiographic and pathogenetic implications

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Striking variability has been observed in the presenting features in infancy of patients subsequently shown to have tetralogy of Fallot. Some patients presented with severe cyanosis in the neonatal period while others had a systolic murmur and cyanosis only on crying. In these latter patients cyanosis became present at rest over the subsequent months. Yet others presented with episodic attacks of unconsciousness, and a final group presented with dyspnoea and heart failure, accompanying a left-to-right shunt.

Angiography demonstrates corresponding variations in the anatomy of the ventricular outflow tracts. In the severely cyanosed patients, the conus septum was deviated so as to obstruct the pulmonary outflow tract, and was best visualized in the lateral projection. In the patients with increasing cyanosis or episodic attacks of unconsciousness, the conus septum again obstructed the pulmonary infundibulum, but was obliquely orientated, not being seen clearly on either lateral or frontal projections. The episodic attacks were considered to be related to infundibular spasm, as previously shown to occur in Fallot's tetralogy. In contrast, increasing cyanosis was believed to be related to hypertrophy of infundibular musculature. In the patients with an initial left-to-right shunt, the arteries were orientated side-by-side so that the conus septum was observed in the frontal projection. Again, subsequent cyanosis was related to ventricular hypertrophy producing outflow tract obstruction.

These findings are interpreted in the setting of a recent study, indicating that Fallot's tetralogy is produced by rotation of the embryonic conus together with sinistro-anterior deviation of the conus septum.

Definition of the condition termed tetralogy of Fallot presents difficulty. When right ventricular outflow tract obstruction is minimal, distinction from ventricular septal defect with aortic overriding is hard to make, even using anatomical criteria (Van Mierop and Wiglesworth, 1963; Goor, Lillehei, and Edwards, 1971). Similarly, when obstruction is severe, the condition closely resembles pulmonary atresia with ventricular septal defect and presence of main pulmonary artery. Furthermore, it has been shown clinically that sub-valvar pulmonary stenosis frequently develops or increases within the first year of life (Gotsman, 1966). We have observed a wide spectrum of presentation in those infants subsequently diagnosed angiographically as Fallot's tetralogy who were catheterized in the first year of life. In this report

we demonstrate that clinical presentation can be explained on the basis of angiographic findings which in turn can be interpreted in the light of recent pathological studies (Becker, Connor, and Anderson, 1975). These demonstrated that tetralogy of Fallot represents varying degrees of conal rotation and sinistro-anterior deviation of the conus septum.

Clinical findings

Twenty-four infants, 16 male and 8 female, were referred, in whom the diagnosis of tetralogy of Fallot was confirmed by angiography and catheterization within the first year of life (Table). Angiographic criteria for diagnosis were the presence of infundibular stenosis, together with filling of the ascending aorta through a ventricular septal defect, in patients with atrioventricular concordance. When pulmonary stenosis was present, patients were considered to exhibit Fallot's tetralogy even though more than 50 per cent of the aorta

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TABLE Mode of presentation of Fallot's tetralogy in infancy

Mode of presentation	Case	Sex	Age at presentation	Age at catheterization	Somatic abnormalities
Severe cyanosis	1	M	1 d	2 d	
	2	M	1 d	2 d	
	3	M	2 d	10 d	
	4	F	10 d	2 w	Small head, mongoloid facies
	5	M	2 w	3 w	Small head, hypospadias
	6	M	2 w	2 w	Hare-lip, cleft palate, hypotonia
Cyanosis on crying cyanosis at rest	7	F	3 d	2 d	Multiple congenital anomalies. Died
	8	M	3 m	6 m	
	9	F	3 m	4 m	
	10	M	7 m	7 m	
	11	F	2 d	3 w	
	12	F	5 w	5 w	
	13	M	5 w	2 m	
	14	F	5 m	10 m	
	15	F	10 m	10 m	Rubella embryopathy
Episodic loss of consciousness	16	M	2 m	2 m	
	17	M	4 m	4 m	
	18	M	7 m	7 m	
	19	M	10 m	10 m	
Dyspnoea	20	F	7 m	11 m and 4½ y	
	21	M	2 w	11 m	
	22	M	2 w	8 m	
	23	M	12 d	5 m	
	24	M	3 m	12 m	

d, days; m, months; y, years; w, weeks.

arose from the right ventricle. According to Kirklin and Karp (1970) cases in which more than 50 per cent of the aorta arises from the right ventricle would be classified as double outlet right ventricle in the absence of pulmonary stenosis, even in the presence of aortic-mitral continuity. In this investigation the conus septum will be defined as the muscular structure dividing the conal origins of the aorta and pulmonary artery. Other investigators have termed this structure the 'crista supraventricularis' (Kjellberg *et al.*, 1959). However, as explained elsewhere (Anderson *et al.*, 1974; Becker *et al.*, 1975), confusion surrounds its usage in this context, and for this reason we have avoided the term. All the patients studied exhibited situs solitus.

Eleven patients (average 8 days) were seen in the first month. The remaining 13 patients were seen between 1 month and 1 year (average age 4.4 months). All infants were referred from other hospitals, 13 being emergency transfers. On reviewing the mode of presentation, it became evident that four patterns could be distinguished. These were: a) severe cyanosis in the neonatal period; b) initial presentation with cyanosis on crying and subsequent development of cyanosis at rest; c) episodic loss of consciousness; d) dyspnoea and signs of heart failure.

We will now amplify the description of these patterns.

a) Severe cyanosis in the neonatal period

Six patients presented with severe persistent cyanosis in the first month of life. The average age of presenta-

tion was 7 days; arterial PO_2 on admission was less than 35 mmHg (4.7 kPa). Three had no murmur and 3 had short grade 2/6 ejection systolic murmurs maximal at the left sternal edge. Chest x-ray showed slight to moderate cardiac enlargement in 4 cases and pulmonary vascular markings were reduced in all. The electrocardiogram was normal for age (Liebman, 1966).

Diagnosis was confirmed by right ventricular angiography which demonstrated an extremely small outflow tract with severe infundibular stenosis and a small pulmonary valve ring (Fig. 1). A ductus arteriosus, demonstrated in 5 of the infants, appeared to provide the major pathway for pulmonary blood flow. A ductus was not visualized in the sixth case.

The specific angiographic features of this group were as follows: an anteroposition of a small pulmonary artery relative to the aorta; the conus septum was usually visible dividing the arterial outflow tracts, and was generally best seen as a filling defect in cross-section on the lateral projection (Fig. 1A). A comparable specimen from our cardiopathological collection confirmed the presence of these morphological features (Fig. 1B).

Anastomoses of right pulmonary artery to ascending aorta were performed in all 6 infants in the first month of life.

b) Infants presenting with increasing cyanosis

In 9 infants cyanosis on crying was noted in the first 3 months of life. The cyanosis gradually increased,

FIG. 1 A) Right ventriculogram, lateral view, in a patient presenting with severe cyanosis in the neonatal period. The conus septum (CS) is seen in section between the narrowed right ventricular outflow tract (Inf) and the aorta (Ao) which fills through a ventricular septal defect (VSD). Though pulmonary atresia is apparently present in this frame, direct pulmonary artery filling was observed in later frames. B) A comparable specimen which has been dissected to correspond with the angiogram shown in (A). The aorta is seen to be overriding a ventricular septal defect. The conus septum is deviated sinistro-anteriorly and encroaches upon the pulmonary outflow tract producing conspicuous pulmonary stenosis. Note that the trabecula septomarginalis (TSM) is posterior to the conus septum and is situated inferiorly to the ventricular septal defect (VSD). RA = right atrium; TV = tricuspid valve.

becoming evident at rest by the second half of the first year. A systolic murmur, though present in 7 patients, was not detected in 2 patients. Cardiac failure was a feature of 2 patients. Chest x-ray revealed cardiomegaly in 4 patients; pulmonary vascular markings were within normal limits. Electrocardiogram showed right ventricular hypertrophy. Hypercyanotic attacks subsequently developed in 3 infants in this group. One of the patients exhibiting the features of the group died in the first week of life owing to the presence of multiple congenital anomalies.

On right ventricular angiography, the aorta was deviated in a rightward direction relative to the ventricular septum and was observed to override a ventricular septal defect (Fig. 2A). The conus septum was generally obliquely orientated, and encroached upon the pulmonary outflow tract (Fig. 2A). In the patient who died, subsequent necropsy examination revealed rotation of the conus relative to the ventricles together with sinistro-anterior deviation of the conus septum (Fig. 2B). The position of the septum was such that in most cases it would not be visualized in direct cross-section on either frontal or lateral projections, though it would

be visible as a filling defect in those projections in some frames of the angiographic sequence.

c) Infants presenting with episodes of loss of consciousness

Four infants were seen in whom the first reported symptom was an episode of loss of consciousness. In each case a murmur had been heard in the first month of life, but cyanosis was not detected. The episodes began between 2 and 6 months of age and all the patients eventually became cyanosed at rest. One infant, however, remained pink between the attacks for several months and an arterial Po_2 (in air) of 94 mmHg (12.5 kPa) was measured. In another infant the episodes, beginning at 5 months, resembled convulsions to such an extent that he was treated with increasing doses of anticonvulsants until he was 10 months of age when cyanosis became evident at rest.

Chest x-ray at the time of catheterization showed normal heart size and diminished pulmonary vascular markings. Electrocardiogram showed right ventricular hypertrophy.

FIG. 2 *Angiocardiogram and specimen from a neonate who presented with cyanosis on crying and other congenital anomalies. A) A right ventriculogram viewed in the lateral projection, with slight rotation of the heart within the thorax. The aorta (Ao) and pulmonary artery (PA) are separated by the conus septum (CS), and infundibular obstruction is produced between the conus septum and a filling defect on the anterior infundibular wall (AW). A similar anterior defect can also be seen in Fig. 1A. Note the additional filling defect towards the ventricular apex (TSM). (RA – right atrium, RV – right ventricle.) B) A photograph of the necropsy specimen from this case after dissection to simulate the angiogram. The aorta can be seen overriding a ventricular septal defect (VSD). The conus septum is clearly visible, and the infundibular obstruction can be related to the septal extension of the conus septum together with hypertrophied trabeculae of the infundibular free wall. The apical filling defect is the trabecula septomarginalis. Note that this structure extends superiorly to the inferior rim of the defect, giving rise to the conal papillary muscle (CPM) of the tricuspid valve (TV). This structure, often termed the 'septal band', is not instrumental in producing infundibular obstruction.*

Right ventricular angiography confirmed the presence of Fallot's tetralogy and showed considerable contraction of the infundibulum during systole (Fig. 3A and B). In the majority of patients the conus septum was observed as a filling defect encroaching upon the pulmonary outflow tract. However, the orientation of the great arteries relative to each other was such that the conus septum was not usually visualized in *direct* cross-section on either frontal or lateral projections except in selected frames.

d) Infants with dyspnoea and signs of heart failure

Five acyanotic infants presented with dyspnoea, hepatomegaly, and a systolic murmur. The chest radiographs revealed cardiomegaly and pulmonary plethora (Fig. 4A), and the electrocardiogram showed evidence of biventricular hypertrophy.

Only one infant was subjected to cardiac catheterization during the phase of left-to-right shunting. In the others, originally thought to have isolated ventricular septal defects, the gradual development of cyanosis on crying, together with electrocardiographic evidence of increasing right ventricular hypertrophy and regression of left ventricular hypertrophy, suggested reversal of the shunt, as did regression of plethora on chest x-ray (Fig. 4B). They were catheterized at this stage.

The duration of the phase of left-to-right shunting varied from 3 weeks to 4 years. Three of these infants also developed hypercyanotic attacks before the age of 1 year.

In the patient studied during the phase of left-to-right shunting, angiography showed side-by-side orientation of the great arteries in the frontal plane, indicating dextroposition of the aorta. Since the arteries were in the same lateral plane the conus septum was visible in cross-section on the frontal projection (Fig. 5A).

FIG. 3 Right ventriculograms, lateral view, from a patient who presented with episodic loss of consciousness. In diastole (A) the right ventricular outflow tract (Inf) is widely patent, but during systole (B) it is narrowed by encroachment of a thickened anteriorly deviated conus septum (CS) and its extensions into the free wall of the infundibulum (arrowed). If comparison is made with the other specimens illustrated, it will be noted that this anterior obstruction cannot be the trabecula septomarginalis or 'septal band'. This structure would be inferior to the septal defect visible in (B) (VSD). RV = right ventricle; LV = left ventricle.

In the later angiograms of the same patient, obstruction to the pulmonary outflow tract resulting from hypertrophy of the pulmonary conus was shown to be present at infundibular level (Fig. 5B). A comparable specimen again confirmed the morphological features (Fig. 5C).

Discussion

Clinical presentation of patients with Fallot's tetralogy is dependent upon the degree of right ventricular outflow tract obstruction. While this fact has been well recognized (Gasul, Arcilla, and Lev, 1966; Rudolph, 1968; Gunteroth, 1968; Nadas and Fyler, 1972), the relation between clinical presentation and the underlying anatomical malformation has not to the best of our knowledge been clearly defined. In this investigation, therefore, we have reviewed the initial mode of presentation in infancy of patients with Fallot's tetralogy, and have attempted to determine their right ventricular anatomy from study of angiocardiograms. We have then interpreted this information in the setting of recent morphological studies of the anomaly (Goor

et al., 1971; Becker *et al.*, 1975) and on the basis of specimens from our own collection.

The basic morphological disturbance of Fallot's tetralogy has been shown to be rotation of the ventricular outflow tracts (conus) relative to the underlying ventricles, together with sinistro-anterior deviation of the conus septum, the latter deviation producing eccentric conal septation at the expense of the pulmonary artery (Fig. 6). As a consequence of these events, the septal insertion of the conus septum is situated sinistro-anteriorly to the trabecula septomarginalis (Fig. 7), this deviation also accounting for the presence of the ventricular septal defect. By virtue of its position inferior and posterior to the conus septum, it follows that the trabecula septomarginalis itself cannot be instrumental in producing pulmonary infundibular obstruction. Some authorities refer to the structure which we describe as the trabecula septomarginalis as the 'septal band' (Van Praagh, 1968). This definition can lead to confusion, since other investigators state that the 'septal band' produces pul-

FIG. 4 A) A chest x-ray of a patient at initial presentation with signs of a left-to-right shunt. Cardiomegaly and pulmonary plethora are demonstrated. B) A subsequent x-ray of the same patient taken 4 years later when right ventricular outflow tract obstruction (Fig. 5B) had developed. It demonstrates minimal cardiac enlargement and normal pulmonary vascular markings.

monary obstruction in Fallot's tetralogy (Kirklin and Karp, 1970). This is in no small way due to the fact that other workers describe a different structure as the 'septal band'. Thus, when Kjellberg *et al.* (1959) described their findings in Fallot's tetralogy, they equated the conus septum with the crista supraventricularis, and described its insertions and extensions as the 'septal band' and 'parietal band', respectively. Within this definition, the 'septal band' certainly produces pulmonary obstruction, but at the entrance to the pulmonary infundibulum and not in the position of the morphological 'septal band' as labelled by Kirklin and Karp (1970) (compare Fig. 2-3 and 2-17). Likewise, the 'septal band' described by Van Praagh (1968) and the structure labelled by Kirklin and Karp in their specimens (Fig. 2-17) is unlikely to produce infundibular obstruction, as it is related to the inferior margin of the septal defect and underlies the aortic outflow. Since usage of the term 'septal band' can clearly lead to confusion, it has been suggested that it would be preferable to dispense with its use in malformed hearts, together with the terms 'crista supraventricularis' and 'parietal band' (Anderson *et al.*, 1974; Becker *et al.*, 1975). We have, therefore, presently referred to the 'septal band' of Van Praagh (1968) as the *trabecula*

septomarginalis, following the precedent of Van Mierop *et al.* (1963). The crista supraventricularis and its extensions described by Kjellberg *et al.* (1959) have been described as the *conus septum* and its *septal and parietal insertions*. Thus, the pulmonary obstruction seen in Fallot's tetralogy is produced by the sinistro-anteriorly deviated conus septum together with its two insertions, and by hypertrophy of the muscular free and septal walls of the pulmonary infundibulum. The degree of obstruction in the cases studied anatomically was related to the degree of sinistro-anterior deviation of the conus septum.

Our present angiographic observations are compatible with this morphological concept. Those patients presenting with severe cyanosis are shown angiographically to have considerable sinistro-anterior deviation of the conus septum. Rotation of the conus is not the prominent feature in this group, though it is present. The result of the preponderance of septal deviation is severe obstruction and better visualization of the conus septum in the lateral rather than frontal projections. The conus septum is oblique, but is closer to a coronal position than in the other groups (Fig. 8a). These patients also have small pulmonary arteries and valve rings. It is likely that the latter features are

FIG. 5 Angiograms from a patient presenting initially with dyspnoea and signs of heart failure. A) An angiogram taken at the time of initial presentation when the chest x-ray suggested left-to-right shunting (Fig. 4A). The injection is into the left ventricle (LV) and is viewed in the frontal projection. The right ventricle (RV) has filled through a ventricular septal defect, and both aorta (Ao) and pulmonary artery (PA) opacify simultaneously. The arteries are in the same frontal plane and are of similar diameters. They are separated by the conus septum (CS) which is clearly visible in this projection. B) An angiogram taken four years later after reversal of the shunt (Fig. 4B). The injection is into the right ventricle (RV) and is viewed in the lateral projection. Pulmonary outflow tract obstruction is now present and is produced posteriorly by the conoventricular flange and anteriorly by the extension of the conus septum free wall (arrows). C) A comparable specimen dissected to simulate the appearance seen in the frontal angiogram (A). The aorta and pulmonary artery are side by side and the aorta is predominantly above the right ventricle, though mitral-aortic fibrous continuity was present through the ventricular septal defect (VSD). In this particular specimen the conus septum is deviated both obliquely and leftwards. It encroaches upon the pulmonary outflow tract, and outflow tract obstruction (POTO) is produced by the conus septum and its extension into the infundibular wall. Note that the obstruction is not produced by the trabecula septo-marginalis (TSM) which is inferior to the level of obstruction and extends posteriorly to the inferior margin of the ventricular septal defect (VSD).

secondary to decreased main pulmonary artery flow in the fetus, as vessel size at birth appears to be proportional to flow through them during intra-uterine development (Rudolph, Heymann, and Spitznas, 1972; Shinebourne and Elseed, 1974). The patients presenting with either episodic loss of consciousness or progressive cyanosis had common angiographic features. Conal rotation was present, as evidenced by the aortic dextroposition relative to the pulmonary artery. As a consequence of this rotation, the conus septum was orientated in an oblique plane and was not always clearly visualized

in either frontal or lateral projections (Fig. 8b). Clearly the statement that the septum was oblique in these cases is made by a process of exclusion. Direct evidence for such an orientation can only be provided by performing oblique angiographic projections, which were not done in our investigation. However, examination of necropsied specimens as presently demonstrated and in the investigation of Becker *et al.* (1975) supports our contention. Initial infundibular stenosis was again a consequence of sinistro-anterior deviation of the conus septum. However, the degree of obstruction eventually

FIG. 6 Diagram illustrating the morphogenetic mechanisms producing Fallot's tetralogy (based on the investigation of Becker *et al.*, 1975). In the normal heart, inversion of the conus is approximately that indicated by the line X—X in (a). Absorption of the subaortic conoventricular flange (open arrow — C.A.) then carries the aorta (Ao) to a position above the left ventricle. The conus septum normally septates the conus in approximately equal parts between aorta and pulmonary artery (PA). Fallot's tetralogy (b) is produced after incomplete inversion of the conus, which can be described as conal rotation (note new position of conal axis X—X). Conal absorption then produces aortic-mitral continuity but fails to take the aorta above the left ventricle, giving dextroposition and overriding. Associated sinistro-anterior deviation of the conal septum (solid arrows) septates the conus in unequal parts at the expense of the pulmonary artery, producing pulmonary outflow tract obstruction.

present was dependent upon hypertrophy of the infundibular free wall, together with the conus septum. This increasing hypertrophy finally produced a permanent level of obstruction in the patients with increasing cyanosis.

In these patients where loss of consciousness accompanied a hypercyanotic attack, infundibular shutdown can be incriminated, as first suggested by Wood (1958). The mechanism of infundibular spasm is as yet uncertain, but is possibly related to increased sympathetic tone (Honey, Chamberlain, and Howard, 1964). In the patients presenting with dyspnoea and signs of heart failure, initial absence of outflow tract obstruction allows a large left-to-right shunt through the ventricular septal defect. In the single patient of our series studied at the stage of left-to-right shunting, the angiograms illustrated a side-by-side position of the great arteries (Fig. 8c). As would be expected with this arterial orientation, the conus septum was visible in direct cross-section on the frontal projection (Fig. 5A). A specimen in our collection showed similar features, but the septum was more obliquely orientated in this case (Fig. 5C). We interpret this as reflecting predominant conal rotation with minimal septal deviation, similar to that reported in double

outlet right ventricle (Goor and Edwards, 1973; Anderson *et al.*, 1974). Indeed, it could be argued that the case presently reported was an example of double outlet right ventricle. However, in this case, aortic-mitral fibrous continuity was present. Though more than half the aorta was above the right ventricle, Kirklin and Karp (1970) consider it preferable to regard these cases with aortic-mitral valvular continuity as Fallot's tetralogy, and we are following this precedent. Subsequent pulmonary obstruction in these cases can again be considered as the result of hypertrophy of the infundibular musculature (Fig. 5B). In this group, the ostium to the pulmonary outflow tract would be entirely muscular. In contrast, the aortic valve would still possess a partly fibrous conus. When right ventricular hypertrophy develops as a result of the left-to-right shunt it is not surprising that muscular outflow tract obstruction is produced.

We have interpreted the angiographic observations reported here as evidence of rotation of the conus relative to the ventricles. An alternative explanation would be that the entire heart was rotated within the thorax. We considered this possibility, but would expect the latter contingency to produce concomitant rotation of the ventricular

FIG. 7 A specimen of Fallot's tetralogy dissected so as to demonstrate the structures producing infundibular obstruction. A constriction ring is produced at the entrance to the infundibulum (Inf) by the conus septum (CS) and its insertion into the septum (SICS). These structures were termed the 'crista' and the 'septal band' by Kjellberg et al. (1959). However, the 'septal band' of Van Praagh (1968) is the trabecula septomarginalis (TSM). This structure reinforces the septum beneath the septal defect (VSD). As Van Praagh pointed out, it is not part of the crista. This dissection also shows that it does not produce infundibular obstruction, and is a different 'septal band' from that described by Kjellberg et al. (1959). Other abbreviations: Ao=aorta; PA=pulmonary artery; RA=right atrium; RVS=right ventricular sinus; TV=tricuspid valve.

sinuses. This was observed to a certain extent in one of the specimens studied but was of insufficient magnitude to account for all the rotational features observed. We, therefore, submit that we have demonstrated that variations in the degree of either conal rotation or deviation of the conus septum can account for variations in presentation of Fallot's tetralogy in infancy.

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FIG. 8 Diagram illustrating the manner in which alterations in orientation of the great arteries affect the plane of visualization of the conus septum. In the diagrams the line X-X indicates the degree of conal rotation. In diagram (a) lack of rotation produces an anteroposterior orientation of the great arteries. In diagram (c), however, clockwise conal rotation has produced side-by-side orientation. Diagram (b) illustrates the 'classical' orientation of Fallot's tetralogy which is intermediate between (a) and (c). It will be noted that in situation (a) the conus septum is seen more readily in cross-section on the lateral view. In contrast, in situation (c) the conus septum is seen in cross-section on the frontal view. In the classical situation (b) the conus septum is oblique in both lateral and frontal views and is, therefore, not readily visualized in either plane, though it would be visible in cross-section in selected frames on either projection. Ao = aorta; PA = pulmonary artery; X-section = cross-section.