TRICUSPID ATRESIA

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It is well known that a child with cyanotic congenital heart disease who shows left-ventricular hypertrophy (LVH) and left-axis deviation (LAD) on the electrocardiogram (ECG) probably has tricuspid atresia. Indeed this disease is often first diagnosed when the ECG is seen.

The clinical picture commonly resembles Fallot's tetralogy. Cyanosis is noted soon after birth, the apex is not displaced and a systolic murmur with single second sound is heard. On X-ray examination the heart is not enlarged and the lung fields are oligaemic.

Although this is the common presentation, it is by no means the only one. Tricuspid atresia, in fact, has a wide spectrum and this has not been sufficiently recognized. It seemed worth while therefore, to review our records of this condition and discuss the varying haemodynamic syndromes and their relation to clinical and laboratory findings. Several features are common to all cases of tricuspid atresia; differences being dependent upon anatomical variations.

A. Features Common to All Cases

1. *All blood from Systemic and Pulmonary Veins must pass through the Left Atrium and Left Ventricle*

(a) *Effect on chamber hypertrophy.* The left ventricle and mitral valve are invariably enlarged at necropsy.[1] The LVH is nearly always reflected in the ECG which also commonly shows LAD.

Left-atrial hypertrophy (LAH) would be expected to occur, especially in cases with high pulmonary blood flow, but is difficult to assess because the ECG criteria of LAH, particularly in children, are not clear and furthermore, right-atrial hypertrophy (RAH) commonly overshadows any changes produced by the left-atrial enlargement.

The frequent finding of a prominent left-atrial appendage in the postero-anterior chest film might be considered evidence favouring LAH. Astley *et al.*,[2] however, have shown that this sign is not accompanied by enlargement of the body of the atrium and postulated that it is caused by altered position, rather than enlargement, of the appendage.

(b) *Effect on oxygen saturation.* Systemic arterial desaturation with equal saturations in the aorta and pulmonary artery is inevitable, as in other conditions where complete mixing of the blood from the pulmonary and systemic veins occurs.

The degree of desaturation will clearly depend chiefly on the pulmonary blood flow (see below).

2. *Underdevelopment of the Right Ventricle*

This invariably accompanies tricuspid atresia[1] and contributes to the left-ventricular pattern in the ECG. It is also responsible for the important angiocardiographic sign called the 'right ventricular window',[3] which is a clear, triangular-shaped gap in the inferior-heart border, in the antero-posterior view, between the opacified right atrium and left ventricle.

B. Variable Features

1. *The Condition of the Atrial Septum*

The systemic venous blood crosses the atrial septum through an atrial septal defect (ASD) in one-third and a patent foramen ovale (PFO) in two-thirds of cases.[4] Except in the case of a very large septal defect, it is probable that the atrial communication always presents some degree of obstruction to blood flow and offers more resistance than the normal tricuspid valve.

Since it is the only route by which blood can leave the right atrium, a small inter-atrial communication should have the following effects:

(*a*) A large 'a' wave and slow 'y' descent in the right atrial and jugular venous pressure tracings;

(*b*) Electrocardiographic evidence of RAH;[5]

(*c*) Radiological evidence of right-atrial enlargement; and

(*d*) No evidence of a left-to-right shunt at atrial level.

There is evidence that tall P waves in the ECG are associated with small-orifice size and may decrease following its enlargement by surgical means.[6]

The barrier to egress from the right atrium is also presumably responsible for the commonly found angiographic sign of reflux of contrast medium into the hepatic veins.

2. *The Anatomy 'Downstream' of the Left Ventricle*

More than 50 years ago Kühne[7] first differentiated those cases with and those without transposition of the great vessels. This classification was enlarged by Edwards and Burchell[8] and subsequently by Keith *et al.*[4]

The latter's classification is depicted diagrammatically in Fig. 1 and is as follows:

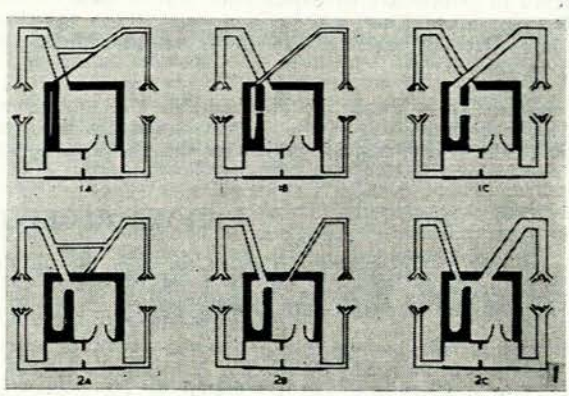

Fig. 1. Tricuspid atresia: anatomical types (after Edwards and Burchell) — see text.

Type 1. *Without transposition of the great vessels*
(*a*) With pulmonary atresia
(*b*) With pulmonary hypoplasia and a small VSD
(*c*) No pulmonary hypoplasia and a large VSD
Type 2. *With transposition of the great vessels*
(*a*) With pulmonary atresia
(*b*) With pulmonary or subpulmonary stenosis
(*c*) With a large pulmonary artery

These anatomical differences produce considerable variations in haemodynamics, the most important effect being the amount of blood that flows through the lungs. Other factors that augment pulmonary blood flow are a patent ductus arteriosus or extensive bronchial collateral vessels. Clearly patients in groups 1(a) and 2(a) rely entirely on such vessels for perfusion of their lungs.

Patients in groups 1(c) and 2 (c) have increased pulmonary flow, unless there is a high pulmonary vascular resistance. Since more oxygenated blood enters the left atrium and ventricle, these patients are less cyanosed than those with normal or reduced pulmonary blood flow.

Increased pulmonary blood flow results in: (*a*) a history of respiratory infections; (*b*) relatively mild cyanosis; (*c*) cardiomegaly; (*d*) splitting of the second sound; (*e*) a mitral-diastolic flow murmur; and (*f*) radiological evidence of pulmonary plethora.

The intensity of the systolic murmur is related to the pulmonary flow, but will also depend on the morphology of the VSD and the right ventricular outflow tract.

CLINICAL MATERIAL

19 patients with a diagnosis of tricuspid atresia have been seen in the Cardiac Clinic between 1950 and 1963. The diagnosis was established by cardiac catheterization and angiocardiography in 12 and in one child the diagnosis was made at necropsy. The remaining 6 patients were diagnosed on clinical grounds and are not included in the analysis except for determining the incidence of the condition.

Incidence, Sex, Race and Age

In the period under review 1,465 patients with congenital heart disease were seen, giving an incidence (if unproven cases are included) of 1·3%. There were 9 females and 4 males, 7 Whites, 5 Coloureds and 1 Bantu. Three patients were under 1 year at the time of first examination; the remainder showing a fairly even distribution up to 12 years.

History

Cyanosis had been observed in all but one child, an example of type 2(c). Of the 10 patients whose age of onset of cyanosis was available, 5 were cyanosed from birth, 2 more from 2 months or less, and 3 from between 6 months and 1 year. The example of type 1(c) fell into this last group.

The age of onset appeared to show some correlation with survival. Of the 5 patients cyanosed from birth, 2 were dead by the age of 13 months, and another aged 4 months is critically ill, whereas all whose cyanosis started later in life have survived 3 years or more. However, the oldest patient in this series, who had a Blalock-Taussig operation[9] at 12 years, was cyanosed from birth. Three patients had definite cyanotic attacks and squatted.

Dyspnoea and tiredness were common symptoms especially in walking children. Delayed milestones and failure of development were frequently mentioned. The only patient with frequent respiratory infections was the one in type-1(c) category. The sibling of one patient died of congenital heart disease of unknown type.

Findings

Cyanosis and clubbing. All patients were markedly cyanosed except for one patient with type-2(c) deformity. Clubbing was present in all cyanosed children over the age of 6 months.

Venous pulsation. A giant 'a' wave was noted in 2 patients, of whom 1 had a pulsating liver. A dominant 'a' wave was seen in a further 3 patients.

Auscultation. A systolic murmur was grade 4-5/6 in 3, grade 2-3/6 in 6 and soft or absent in 2 patients. Very loud systolic murmurs were present in 2 patients with type 1(b), and in the one patient with type 1(c). Where pulmonary atresia was present, the murmur was soft or absent.

Of the 10 patients in whom the second sound was analysed, this sound was single in 8 and split in 2. Both the latter had grade 4-5/6 systolic murmurs.

Polycythaemia. In 5 of the 10 patients the Hb level exceeded 16 G/100 ml., and in 5 it was between 11 and 16 G/100 ml.

ECG Investigations

ECG tracings were available in 11 patients.

The P wave. The tallest P wave in lead 2 was 2·5 mm. or more in 9 patients, and 4 mm. or more in 3. A rough correlation was found between this measurement and the height of the right atrial 'a' wave, but not with the 'y' descent nor with the appearance of the right atrium in the chest X-ray.

The width of the P wave was unremarkable, varying from 0·05 to 0·13 secs. and correlating well with age; being shorter in infants and longer in older children.

P-R interval and P-R segment. The P-R interval varied from 0·08 to 0·16 secs. showing a similar relation to age as the width of the P wave. The P-R segment was 0·03 secs. or less in 7 patients, and never over 0·05 secs.

QRS. The directions of the mean QRS vectors in the frontal plane are shown in Fig. 2. LAD with an anti-clockwise loop was present in 8, in 7 of whom the axis lay between −10° and −60°. The one patient in whom both great vessels arose from the right ventricle, had an axis of −100°. Two patients had normal axes of +35° and +80° and both had clockwise loops. A very complex loop was found in one patient who had mesocardia (this case is excluded in Fig. 2).

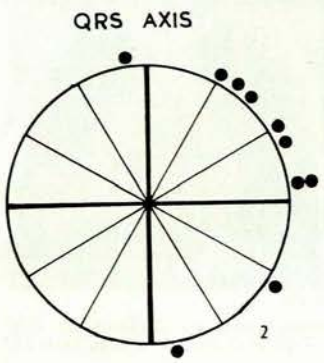

Fig. 2. Mean QRS vectors in the frontal plane. Note the majority lie between −10° and −60°.

Amplitude of R and S waves in chest leads. The tracings were evaluated, using Keith's criteria for LVH based on the magnitude of the R wave in leads V5 and V6, the S wave in V1, and the R/S ratio in V1.[4] Nine of the 11 patients had at least one positive criterion; lead V1 being the commonest site for this. One of the 2 patients without evidence of LVH had a mesocardia, and in the other, both great vessels arose from the right ventricle.

T wave. In the left praecordial leads the T wave was inverted in 4 patients, including the 2 who failed to show evidence of LVH by criteria mentioned above. A typical ECG

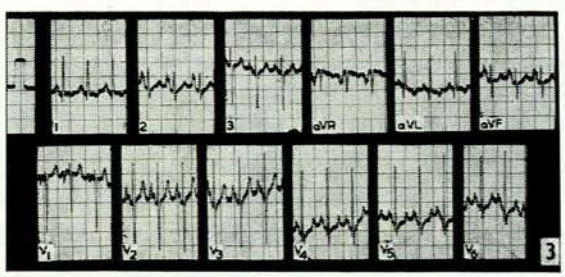

Fig. 3. Typical ECG, in a 6-months-old child, showing LVH, LAD and RAH.

tracing showing, in a 6-month-old child, LVH and LAD and tall P waves is seen in Fig. 3.

X-ray of the Chest

10 patients were studied in the postero-anterior view. The plate of the patient with mesocardia is not included except as regards the lung fields.

Cardiothoracic ratio. Of 9 patients, 3 had a ratio of 55% or less, 2 of 56 - 60% and 4 over 60%. The latter group includes the examples of type 1(c) and 2(c).

The right atrium was considered abnormally flat in 3, unduly prominent in 3, and normal in 3 patients each. Those with a flattened right border gave the appearance of disproportionate enlargement to the left.

Left cardiac border. The pulmonary-artery segment was prominent in 3 subjects, all of whom had pulmonary plethora. The fourth patient with pulmonary plethora had a very large globular heart obscuring a huge pulmonary artery which was shown at necropsy. The left-atrial appendage was considered prominent in 5 subjects. When this was associated with a 'bay' in the pulmonary artery region and a rather prominent left ventricle, the characteristic 'square' shape was seen. However, this appearance was rare, being shown in only 1 patient and being doubtfully present in another 2. One patient had a typical 'sabot' shape.

Pulmonary vessels. Five patients had pulmonary oligaemia, 1 had normal lung fields and 4 had pulmonary plethora. Of the latter, 2 were examples of type 1(c) and 2(c) and 2 had

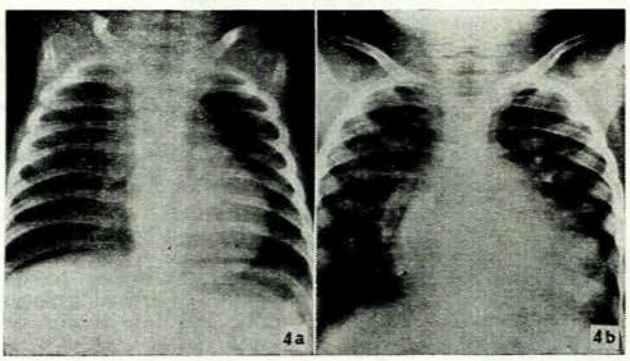

Fig. 4(*a*). Example of type 1(b) showing pulmonary oligaemia and small heart with flattened right border. (*b*) Type 2(c) showing a large heart and plethoric lung fields.

had previous Blalock-Taussig anastomoses. Figs. 4(a) and (b) show examples of types 1(b) and 1(c) respectively.

Findings at Cardiac Catheterization

Twelve patients were catheterized with the following results:

Inability to enter the right ventricle. In no case was the right ventricle entered.

Atrial-pressure tracings. Right atrial 'a' waves of 5 mm. or more were present in 4 patients; all but one of the others being over 3 mm. The 'a' wave in the right atrium was nearly always much higher than the left-atrial 'a' wave, when the latter could be recorded.

The rate of the 'y' descent in the first 0·08 secs. was measured in the 9 subjects in whom the duration of the 'y' descent was 0·08 secs. or more. It tended to be rather slow; being less than 10 mm./sec. in 5 cases and between 11—20 mm./sec. in 2 others. It was always considerably less than the same measurement from the left atrium which averaged 36 mm./sec.

Saturation. Arterial oxygen saturation varied between 37 and 88%. The only 2 patients with saturations of over 80% had pulmonary plethora.

In 5 patients, right atrial samples indicated a left-to-right shunt across the atrial septum during some phase of the cardiac and respiratory cycles. There was no difference between these and the 4 who failed to show a left-to-right shunt as regards P wave in ECG or right atrial 'a' wave.

Dye Studies

These were performed in 4 cases only. With systemic arterial sampling, the identity of curves obtained by injecting dye into vena cava, left atrium and left ventricle was characteristic. The curves showed evidence of large left-to-right shunts in 2 patients who had pulmonary plethora.

Angiograms were recorded in 13 subjects. Venous angiography only was used in the earlier studies, but more recently, selective angiograms from the left ventricle and usually also a vena cava were obtained, using either a rapid change biplane angiogram or cine-angiograms.

Of 9 patients who had venous angiograms, 7 showed a triangular 'window' between right atrium and left ventricle (Fig. 5). The patient with mesocardia had a similar radiolucent area on the right side, possibly of the same significance. In the remaining subject, the angiogram was of inadequate quality. Reflux of the dye into the hepatic veins was seen in all 6 patients where the angiograms were adequate for the assessment of this sign.

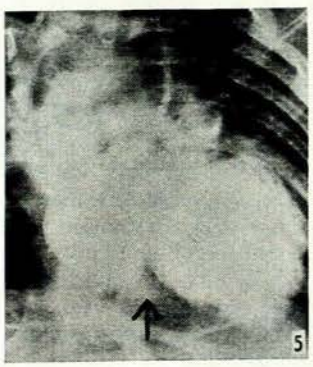

Fig. 5. Venous angiogram showing 'right ventricular window' (arrow) and hepatic reflux.

Left ventricular angiograms (including both patients studied by selective angiography and those in whom late pictures following venous injection were of adequate quality). The findings can best be summarized by using them as a basis of classification into the types discussed above. Eight patients were classified in this way.

Of type 1 (without transposition) there were 6 patients of type 1(b), one each of 1(a) and 1(c) and a further case of either 1(a) or 1(b).

Two patients had transposition. One, a variant of type 2(b), had a fairly large right ventricle with both great vessels arising from this chamber and aortic and pulmonary valves at the same level, indicating a 'double-outlet' syndrome. The aorta

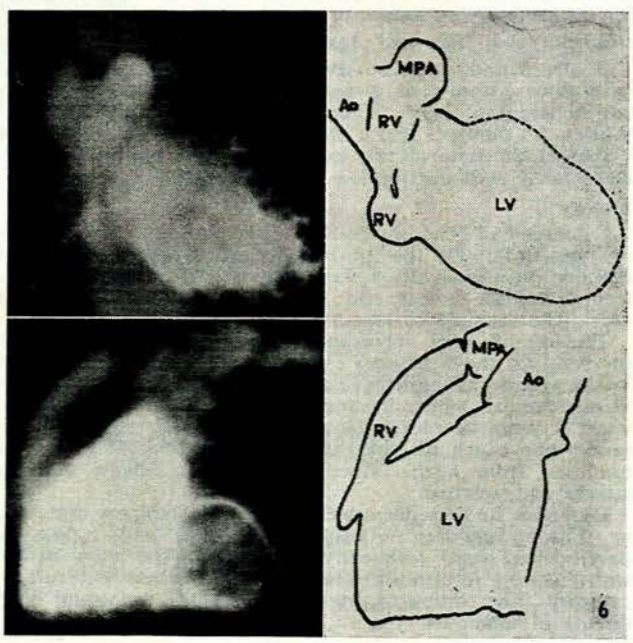

Fig. 6. Selective angiogram from left ventricle in type 1(b) (A) PA and (B) lateral view. Note large left ventricle and dye passing across ventricular septal defect into diminutive right ventricle.

arose anteriorly to the pulmonary artery. There was pulmonary valvar stenosis.

In the other patient the angiograms were adequate only to demonstrate the transposition. Near equality of pulmonary and systemic blood flows suggest that this was also an example of type 2(b). An example of type 1(b) is seen in Fig. 6.

TREATMENT

A subclavian-pulmonary artery anastomosis was performed in 3 patients. One was operated on at 4 months and died at 8 months. The other 2 were operated on at ages 5 and 13 and are alive at 11 and 17 years. All were considerably improved following operation.

A further patient had a Barrett's procedure[10] performed at 6 years and died aged 9 years, a few days after an attempted Blalock operation at which the left pulmonary artery was found to be absent.

DISCUSSION

Our incidence of 1·3% is comparable with those of Abbott[11] (1·6%), and Wood[12] (1·5%), and Campbell's[13] figure of 5% of cyanotic congenital heart cases. Thus, one patient with tricuspid atresia is likely to be seen for approximately 10 with Fallot's tetralogy.

There is little in the history to distinguish tricuspid atresia from other cyanotic congenital heart disease. We consider that cyanosis from birth usually implies a poor prognosis, but our oldest patient points to the fallacy of applying this to the individual case. A history of frequent respiratory infections suggests, as in other congenital heart diseases, a high pulmonary blood flow.

Subjects with a tumultuous pulmonary flow may be expected to be acyanotic, as in our example of type 2(c). A giant or dominant 'a' wave and occasionally a pulsatile liver may help to distinguish tricuspid atresia from other conditions, especially from Fallot's tetralogy. Absence of a murmur suggests associated pulmonary atresia while a split second sound indicates a high pulmonary flow.

The ECG is the most important bedside diagnostic procedure. Our finding of almost invariable LVH and frequent association with RAH, is in agreement with all reports of this condition.

LAD was present in all but 2, who both had normal axes. Somlyo and Halloran[6] found normal axis or RAD in 25% of their cases, and most of these were of types 1(a) or 2(c). Our 2 instances of normal axis did not appear to fall into either of these groups, though neither had angiograms of adequate quality for definite classification. The child with a double outlet from the right ventricle was the only subject to have an axis to the left of −90°.

Although tricuspid atresia is the commonest cause of LVH and LAD in cyanotic congenital heart disease, these findings have been described in several other conditions, notably in single ventricle and in transposition. Shaher[14] has recently reviewed the literature. In his own cases he found that tricuspid atresia accounted for only 20% of cyanotic patients with LVH or LAD. However his criteria for LAD were wide (+60° to −90° in children under 10) and in patients with both LVH and LAD tricuspid atresia was still the most frequent diagnosis, especially in the younger age groups.

The chest X-ray shows pulmonary oligaemia in most patients, as would be expected from the incidence of the various anatomical types. There have been conflicting reports concerning the cardiac outline, the right border being variably described as flattened[15] and prominent.[16] We found both types of silhouette as did Wittenberg[17] who suggested that the shape depends on the degree of dilatation of the right atrium and of its displacement to the left owing to the underdeveloped right ventricle. A prominent right border did not imply a small orifice at the atrial septum in our series.

PROGNOSIS AND TREATMENT

Campbell[13] has stressed the poor prognosis in this condition and urged surgical treatment.

The indication for operation is underfilling of the pulmonary vessels, and a systemic-pulmonary artery anastomosis, by either the Blalock or the Potts[18] procedure, has been recommended. A superior vena cava/pulmonary artery anastomosis[19] has the theoretical advantage of improving the blood supply to the lungs without adding to the burden on an already overworked left ventricle. The foramen ovale or atrial septal defect has been widened when the defect has been of inadequate size.

Three of our patients improved following Blalock operations, although all still have symptoms. Anastomotic operations should probably be performed more often on patients with tricuspid atresia, in order to lessen their disability and tide some of them over until a corrective operation has been developed. The situation is analagous to that of Fallot's tetralogy a decade ago.

SUMMARY

13 patients with tricuspid atresia are reviewed to illustrate the variable presentations of this condition. The constant and variable haemodynamic features and their relationship to clinical and laboratory findings are discussed.

The ECG is the most useful bedside diagnostic procedure, showing almost invariably LVH and commonly LAD and right atrial hypertrophy.

The prognosis is generally poor and an anastomotic operation is recommended when there is underfilling of the pulmonary vessels.

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