Severe Congenital Tricuspid Incompetence in the Neonate

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SUMMARY

Five neonates with congenital tricuspid incompetence due to severe tethering of the tricuspid valve to the right ventricle by abnormal chordal and papillary muscle attachments are described. The abnormality was called tricuspid valvular dysplasia (TVD) if the basal insertion of the valve was normal and the Ebstein malformation if it was displaced into the sinus portion of the ventricle.

In the two infants with isolated TVD and severe tricuspid regurgitation the functional obstruction to right ventricular outflow (ORVO) produced by the high perinatal pulmonary vascular resistance (PVR) made the exclusion of pulmonary atresia difficult, despite selective right ventricular angiocardiography.

In three infants the tricuspid valvular abnormality was associated with organic ORVO: pulmonary atresia in two and critical pulmonary valve stenosis in one. In the two infants with pulmonary atresia and intact ventricular septum (IVS) the severe tricuspid incompetence produced a clinical, radiological and hemodynamic profile which was clearly different from that usually seen in infants with pulmonary atresia and IVS and a normal right ventricular cavity (type 2 of Greenwold).

Additional Indexing Words:
Congenital heart disease
Tricuspid valvular dysplasia
Ebstein malformation
Pulmonary atresia with intact ventricular septum

Tricuspid Valvular Dysplasia (TVD) is characterized pathologically by the abnormal tethering of the tricuspid valve to the right ventricular endocardium by direct leaflet attachment and/or by short chordae tendineae. Similar pathological changes can occur in the Ebstein malformation, which therefore may differ from TVD only in that the basal attachment of one or more leaflets is displaced into the sinus portion of the right ventricle.1 TVD and the Ebstein malformation may occur as isolated anomalies or may be associated with pulmonary atresia and intact ventricular septum (IVS).1-7

The infant with either of these tricuspid valve malformations, whether isolated or associated with pulmonary atresia and IVS, may present in the neonatal period with cyanosis, massive cardiomegaly and heart failure, due to severe tricuspid regurgitation.2, 5, 6-13

Five neonates with severe congenital tricuspid incompetence form the basis of this report. In two infants with isolated TVD the angiocardiographic appearance simulated pulmonary atresia with IVS, probably because of functional obstruction to right ventricular outflow (ORVO). In three infants with organic ORVO, severe tricuspid incompetence was the predominant functional abnormality. Although two of the latter group had pulmonary atresia with IVS and a large right ventricular cavity (type 2 of Greenwold14), their close resemblance to neonates with isolated TVD is stressed.

Case Material

The five patients, four males and one female, were seen at the Royal Alexandra Hospital for Children between January 1970 and February 1973. There were two patients with isolated TVD; two with pulmonary atresia and IVS, one of whom had the Ebstein malformation and the other TVD; and one with critical pulmonary valve stenosis and TVD. They will not be described individually because they closely resemble one another. The prenatal history and delivery of each infant was uncomplicated. Their birth weights ranged from 2920 gm to 3530 gm. Central cyanosis was noted at birth in two infants, by four hours in two infants and on the fifth day of life in one infant. Four infants were admitted to our hospital within sixteen hours of delivery and one on the eighth day of life.

The cardiovascular physical findings were remarkably similar with the following being common to all the patients: central cyanosis, grade 3-4/4 in intensity; mild to moderate tachypnea; normal peripheral arterial pulses; hepatomegaly...
without systolic pulsation; marked cardiac enlargement with a prominent precordial right ventricular pulsation; an easily palpated precordial systolic thrill; a pansystolic murmur, grade 3-4/4 in intensity at the lower left sternal edge; a soft second heart sound. One of the infants with pulmonary atresia and IVS had impalpable peripheral pulses and profound low-output cardiac failure on admission to hospital, and the clinical features described above were only elicited following resuscitation by intermittent positive pressure ventilation and partial correction of a profound metabolic and respiratory acidosis. Additional cardiac murmurs were present in three infants: a mid-diastolic murmur at the lower left sternal border in two infants, one of whom had the high pitched early systolic murmur of a small ventricular septal defect at the mid left sternal border; and the continuous murmur of a persistent ductus arteriosus at the second left intercostal space in one infant.

The chest X-rays (fig. 1), like the clinical features, were strikingly similar. There was extreme cardiomegaly and the pulmonary vascular markings, where visible, were diminished. The electrocardiogram (fig. 2) in each case showed deviation of the mean frontal QRS axis to the right, evidence of right atrial enlargement and right bundle-branch block. Left atrial enlargement was evident in three infants.

Cardiac Catheterization

Cardiac catheterization was performed within six hours of admission to hospital in each case and the results are shown in table 1. The pulmonary artery was not entered in either infant with isolated TVD. In the infant with critical pulmonary valve stenosis, it was thought that the main pulmonary artery had been entered and that there was no systolic pressure difference across the pulmonary valve. This resulted in an erroneous diagnosis of isolated tricuspid incompetence in this infant. In view of the necropsy findings it is likely that this pressure recording was artefactual. Oxy-metry demonstrated a right-to-left shunt at atrial level in each case. The right ventricular peak systolic pressure was normal in three patients and only slightly elevated in two, and was less than a serially recorded left ventricular peak systolic pressure in each case. In the right atrial phasic pressure recording of each infant, an “s” wave interrupted the “x” descent (fig 3). The mean right atrial pressure was elevated in only one case. Simultaneously recorded intracavitary electrograms and pressures across the tricuspid valve did not demonstrate atrialization of the right ventricle in the infants with TVD. An intracavitary electrogram was not done in the infant with the Ebstein malformation.

Angiography

Biplane angiography as AOT serial cut films were obtained by the selective injection of Urografin 60%,* 2 ml/kg body weight, into the right ventricle. The contrast medium was delivered within one second by a Vibrant/Hobbs pressure injector.† In the three infants with organic ORVO the right ventricular cavity was large. The heavy trabecular pattern, produced by the encroachment of muscle bands on the cavity of the right ventricle, which is usual in pulmonary atresia with IVS type 2, was not present in these infants (figs. 4 and 5). The contrast medium did not pass forward beyond the level of the pulmonary valve but regurgitated from the right ventricle through the widely dilated tricuspid valve annulus and into the aneurysmally dilated right atrium (fig. 4). The normal size of the main pulmonary artery and its major branches was established by their retrograde opacification from the aorta via a persistent ductus arteriosus.

In the two infants with isolated TVD there was no evidence that contrast medium had passed beyond the level of the pulmonary valve in the early frames of the angiograms (figs. 5A and B). There was, however, very faint opacification of a normal sized main pulmonary artery in a later frame of the angiogram (fig. 5C). The opacification of the pulmonary artery occurred before that of the descending aorta, indicating that the former had filled from the right ventricle and not retrograde from the aorta via a persistent ductus arteriosus. There was massive regurgitation of the contrast medium from the right ventricle through the dilated tricuspid valve annulus into the hugely dilated right atrium (fig. 5).

Figure 2
Electrocardiogram in the infant with critical pulmonary valve stenosis showing rightward deviation of the mean frontal QRS axis, right atrial enlargement and right bundle-branch block.

Figure 3
Right atrial phasic pressure recording in an infant (P.D.) with isolated TVD showing the “s” wave of tricuspid regurgitation.

Urografin 60% (Schering) — a mixture of the sodium and methylglucamine salts of 3, 5-bis-acetamido-2,4,6-triodobenzoic acid.

†Barber-Colman Company, Electrical-Mechanical Products Division, Rockford, Illinois.
In one of the infants with isolated TVD the failure of the pulmonary artery to opacify in the early frames of the angiocardiogram led to the erroneous diagnosis of pulmonary atresia with IVS. In the other infant, the faint opacification of the main pulmonary artery was appreciated, but because the definition was so poor, organic ORVO could not be absolutely excluded.

Management

The two infants with isolated TVD underwent exploratory thoracotomy, but once it was found that their pulmonary valves were normal, they were managed conservatively with digoxin and furosemide. Postoperatively they deteriorated rapidly with increasingly severe hypoxia, hepatomegaly and peripheral circulatory failure. One infant died on the second day of life and the other on the fourth.

The infant with critical pulmonary valve stenosis, who was thought at catheterization to have isolated tricuspid incompetence, was not operated upon and died in a similar manner to the infants with isolated TVD.

The infant with pulmonary atresia and the Ebstein malformation died during an attempted transventricular pulmonary valvotomy. In the remaining infant with pulmonary atresia and IVS, open pulmonary valvotomy and tricuspid valve replacement with a Bjork-Shiley tilting disc prosthesis,* were performed. This patient, who was in severe low-output cardiac failure on admission to hospital, did not survive the operation.

*A. Shiley Laboratories, Inc., Santa Ana, California

### Table 1

<table>
<thead>
<tr>
<th>Case</th>
<th>Pathology</th>
<th>RA Pressures (mm Hg)</th>
<th>LV</th>
<th>SVC</th>
<th>IVC</th>
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<th>RV</th>
<th>PV</th>
<th>LA</th>
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Abbreviations: TVD = tricuspid valvular dysplasia; PA = pulmonary atresia; IVS = intact ventricular septum; PVS = pulmonary valve stenosis; RA = right atrium; RV = right ventricle; LA = left atrium; LV = left ventricle; SVC = superior vena cava; IVC = inferior vena cava; PV = pulmonary vein.
Pathology

The status of the pulmonary valve was confirmed at operation in the infant who underwent pulmonary valvotomy and tricuspid valve replacement, and at necropsy in the four remaining infants.

The most striking abnormality in each case was the tethering of the tricuspid valve leaflets to the mural surface of the right ventricle by very short chordae tendineae (fig. 6). The chordae tendineae attached either to rudimentary papillary muscles or directly to the carinae trabeculae of the right ventricle. Direct adherence of the leaflets to the ventricle was not a feature. The tricuspid valve annulus was widely dilated in each infant and measured from 3.0 to 8.5 cm in circumference (fig. 6). The circumference of the mitral valve in these infants ranged between 3.0 and 3.5 cm. The basal attachment of the tricuspid valve was situated normally at the tricuspid annulus in the infants with TVD (fig. 6). In the infant with the Ebstein malformation, the basal insertion of the septal leaflet was displaced 5.0 to 7.0 mm into the sinus portion of the right ventricle (fig. 7). The anterior and posterior cusps were inserted normally at the annulus fibrosus in this infant. Three distinct cusps were present in four infants, while in the infant with critical pulmonary valve stenosis, the anterior and posterior cusps were represented by a single leaflet. The amount of leaflet tissue was normal in each case. The free margin of each cusp was thickened and deformed by nodular excrescences along its atrial surface. The remainder of each cusp was not thickened, and within the substance of the leaflet the abnormal continuation of the chordae tendineae to the tricuspid valve annulus was visible (fig. 8).

The right atrium was markedly dilated in each infant and in four it was the largest of the cardiac chambers. The muscleae pectinatae were prominent and the right atrial wall was from 2.0 to 3.0 mm in thickness. An interatrial communication was present in each infant: a probe patent but valvular competent foramen ovale in two, a fenestrated and valvular incompetent foramen ovale in two, and a large secundum atrial septal defect in one.

The right ventricle was moderately dilated in three infants and markedly dilated in two. Although right ventricular hypertrophy was present, with prominence of the carinae trabeculae, moderator band and crista supraventricularis, the cavity of the right ventricle was not severely encroached upon by muscle bundles. The right ventricular wall thickness ranged between 3.0 and 4.0 mm, while the left ventricular wall thickness was between 5.0 and 6.0 mm. A small and functionally insignificant ventricular septal defect was present in one of the infants with otherwise isolated TVD. This defect was not of the atrioventricular canal type. The systemic and pulmonary venous drainage, the mitral and aortic valves, and the left ventricle and aorta were normal in appearance in each infant. Histological examination of the hearts by light microscopy was unremarkable. Dysplasia of the right kidney in one of the infants with isolated TVD was the only significant extracardiac abnormality in the five cases.

Discussion

In the neonate with isolated TVD, the freely incompetent tricuspid valve may allow the right ventri-
cle to pump its systemic venous return retrograde into the low pressure right atrium rather than antegrade into the pulmonary circulation, if the pulmonary vascular resistance (PVR) is high. The tricuspid regurgitation, if severe, will result in massive cardiomegaly and heart failure, and the shunting of systemic venous blood across the interatrial septal defect will result in systemic arterial oxygen desaturation and cyanosis.

In these respects the two neonates with isolated TVD in this report do not differ from previously reported cases. However there is an important angiocardiographic difference, namely the difficulty in demonstrating an organically unobstructed right ventricular outflow in our cases. This diagnostically misleading finding was not encountered in previously reported cases studied by selective right ventricular angiocardiography. Nevertheless this angiocardiographic appearance could also be attributed to functional ORVO produced by a high perinatal PVR.

Although the exploratory thoracotomy in our infants with isolated TVD could have unfavorably influenced their natural history, their clinical course was not different from five of the six similar cases previously reported who died in the neonatal period without prior operative intervention. The unrelieved hypoxia and hepatomegaly and the terminal low output cardiac failure in these neonates with isolated TVD suggest to us that the expected decrease in their PVR which would have curtailed the tricuspid regurgitation and alleviated the severe volume-load on the right ventricle, did not occur. In addition, closure of the ductus arteriosus may have greatly intensified their hypoxia, since the persistence of the ductus arteriosus is essential for pulmonary blood supply when antegrade flow from the right ventricle is prevented.

There have been cases of successful conservative management of some, but not all, neonates with the Ebstein malformation and severe tricuspid regurgitation, and presentation beyond the neonatal period of patients with isolated TVD. These would suggest that the circulatory status peculiar to the newborn, rather than the tricuspid valve malformation per se, determines whether severe tricuspid regurgitation with heart failure manifests itself in the neonatal period and whether it can be managed conservatively.

We believe that the poor prognosis plus the possible difficulty in excluding organic ORVO in neonates with isolated TVD and severe tricuspid regurgitation suggest that prosthetic replacement of the tricuspid valve, and, where indicated, direct inspection of the pulmonary valve, should at least be considered in the management of this rare anomaly. However, as mentioned above, the right ventricle of these infants is functionally adapted to pumping blood retrograde into the low pressure right atrium, and may not be able to sustain an adequate pulmonary circulation following tricuspid valve replacement.

Pulmonary atresia with IVS has been subdivided according to the size of the right ventricular cavity into two types: type 1 in which the cavity is small, and type 2 in which it is either normal or large. The size of the right ventricular cavity is probably determined by the competence of the tricuspid valve in utero and therefore it is not surprising that infants with pulmonary atresia and IVS and severe tricuspid regurgitation have been reported. Massive tricuspid regurgitation in the neonate with pulmonary atresia and IVS produces a recognizable, but hitherto poorly categorized constellation of clinical, radiological, hemodynamic and angiocardiographic findings. These infants resemble those with isolated TVD rather than infants with pulmonary atresia and IVS and a normal right ventricular cavity — type 2 of Greenwold.

The prominent precordial thrill and loud tricuspid regurgitant murmur, the extreme cardiomegaly and perhaps the electrocardiographic right bundle-branch block indicate the massive tricuspid regurgitation and dilatation of the right heart chambers which were present in the neonates with pulmonary atresia and IVS in this report. Keith, Rowe and Vlad described nine patients with similar clinical findings. A precordial thrill is, however, usually absent in patients with pulmonary atresia and IVS type and when present, the tricuspid regurgitant murmur is usually soft: grade 2-3/6 in intensity or less.

Although roentgenographic cardiomegaly is a characteristic of pulmonary atresia with IVS type it is uncommonly of the magnitude seen in the patients in this report. Two of the patients described by Schrire, Sutin and Barnard are very similar both clinically and radiologically to the neonates in this report.

The finding at cardiac catheterization of a right ventricular peak systolic pressure which is less than the left ventricular peak systolic pressure suggested a severely incompetent tricuspid valve and therefore unobstructed retrograde flow from the right ventricle in our patients. Although similar pressure findings have been recorded in patients with pulmonary atresia and IVS type it is an uncommon finding. Whether severe tricuspid regurgitation was present in these reported cases is not stated.

In a right atrial phasic pressure recording the presence of an "s" wave interrupting the "x" descent is said to denote tricuspid regurgitation. This in-

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dicator of tricuspid regurgitation was present in both our patients with pulmonary atresia and IVS. There is no reference in the literature to the right atrial phasic pressure recording in pulmonary atresia with IVS type 2; however an analysis of the last eight cases seen by us did not reveal this pattern of tricuspid regurgitation in any patient (unpublished data).

The size of the right ventricular cavity in patients with pulmonary atresia and IVS is best assessed by selective right ventricular angiocardiography. The infants in this report had large right ventricular cavities and they did not have the prominent trabecular pattern usually visible in patients with pulmonary atresia and IVS type 2 and produced by the encroachment of large muscle bands on the cavity of the right ventricle. The appearance of the right ventricle, together with the widely dilated tricuspid valve annulus and the massively enlarged right atrium, reflects the severe tricuspid regurgitation present in these patients.

Green, Zeigler and Micher recognized that severe tricuspid regurgitation in the infant with pulmonary atresia and IVS could preclude successful pulmonary valvotomy. It seems likely that in these infants functional ORVO prevented the amelioration of the tricuspid regurgitation which ordinarily would have followed pulmonary valvotomy. It was for this reason that tricuspid valve replacement was performed in one of the patients with pulmonary atresia and IVS in this report. Although this patient, who was in profound low output cardiac failure with severe acidemia prior to operation, did not survive, the procedure proved to be technically practicable.

We would only consider tricuspid valve replacement in patients with pulmonary atresia and IVS and tricuspid incompetence when the tricuspid regurgitation was judged to be massive by the clinical, radiological, hemodynamic and angiocardiographic criteria outlined above. Even so, there remains, as in the neonate with isolated TVD and severe tricuspid regurgitation, the uncertainty as to whether the right ventricle of these infants can withstand the acutely increased afterload which might follow tricuspid valve replacement.

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