Congenital Tricuspid Insufficiency

Definition and Review*

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Simple congenital tricuspid insufficiency (CTI) has, up to now, been loosely included with the larger Ebstein's anomaly (EA) group. Certain pathologic and clinical features of CTI deserve definition and a place apart from EA. A review of the literature has produced 20 clinical cases for analysis to which we add two cases successfully treated surgically. The high mortality in the neonatal age group (14 of 14) continues to be a formidable challenge, but the older survivors (2 of 8) would appear to have better prospects, based on our limited experience.

In 1866, Ebstein described a congenital malformation of the tricuspid valve in which a portion of the valve (septal and posterior leaflets) arose from the right ventricular wall distal to the anatomic atrioventricular ring. Well over 300 cases have been reported to date, and in recent years, emphasis has shifted to the surgical repair.1-3 Although the spectrum of Ebstein's anomaly (EA) has diverse features, the hallmark, from a pathophysiologic standpoint, has generally been severe tricuspid insufficiency, so that for years, many clinicians made no distinction between Ebstein's anomaly and isolated congenital tricuspid insufficiency (CTI).

The advent of open heart surgery and the increasing diagnostic accuracy of cardiologists have revealed a subgroup of patients with CTI which is distinctly not of the EA variety and indeed has features which warrant further elucidation and definition. We have tabulated clinical cases in the literature available for analysis and added two cases of successful correction of CTI in adults.

Table 1 depicts the clinical cases reported in the literature as CTI documented by post-mortem examination. Additionally, Green et al4 reported 15 cases, only 4 of which were autopsied, and due to lack of detail other than catheterization data, these were not included in our summary. The cases reported by Becker et al4 were part of an autopsy series with no clinical correlation supplied, and so were not included in the tabulation.

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CASE REPORTS

CASE 1

This 18-year-old black man was evaluated at the Medical College of Virginia for increasing shortness of breath and fatigue of one year's duration. He showed no cyanosis, had distended neck veins with a trace of V-wave accentuation, a nonpulsatile liver enlarged to 5 cm below the costal margin, and moderate ankle edema. The heart was massively enlarged, a grade 4/6 systolic ejection murmur, and grade 2/6 diastolic murmur at the right sternal border were heard. The ECG showed first degree atrioventricular block, right bundle branch block, left superior hemiblock and right atrial enlargement. Chest x-ray film showed massive cardiac enlargement. Cardiac catheterization revealed elevation of the right atrial pressure with a pronounced V-wave of tricuspid insufficiency, and normal right ventricular pressures.

On June 3, 1974, surgery revealed a massive enlargement of the right atrium and a muscular, vasty hypertrophied right ventricle with uniform contraction. The tricuspid annulus was dilated to 8-10 cm in diameter and the valve was based on the anatomic annulus. All leaflets of the tricuspid valve were present, but the septal and posterior leaflets were shortened and bound down by abnormal short chordae which attached to the right ventricular wall. The anterior leaflet contained two 3-mm fenestrations. The atrial septum was intact. The tricuspid valve was replaced with a 35 mm stented porcine xenograft, and a reduction atrioplasty was performed.

Six months following surgery, the patient was largely asymptomatic and the chest x-ray film showed substantial reduction in size of the cardiac silhouette.

CASE 2

This 50-year-old white woman was first aware of having a heart problem at the age of 20 when she was rejected from the military service in Germany because of an enlarged heart shadow on chest x-ray examination and the presence of a
<table>
<thead>
<tr>
<th>Author</th>
<th>Age</th>
<th>Sex</th>
<th>Cyanosis</th>
<th>Associated Abnormalities</th>
<th>Treatment</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Hots (1923)</td>
<td>13 yrs</td>
<td>F</td>
<td>Yes</td>
<td>None</td>
<td>Medical</td>
<td>Died, CHF</td>
</tr>
<tr>
<td></td>
<td>12 yrs</td>
<td>M</td>
<td>Yes</td>
<td>PFO</td>
<td>Medical</td>
<td>Died, CHF</td>
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<tr>
<td>Ariel (1930)</td>
<td>2 da.</td>
<td>M</td>
<td>Yes</td>
<td>PFO</td>
<td>Medical</td>
<td>Died, pneumonia</td>
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<tr>
<td>Abbott (1936)</td>
<td>7 da.</td>
<td>M</td>
<td>Yes</td>
<td>PFO, PDA</td>
<td>Medical</td>
<td>Died</td>
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<tr>
<td>Dubin, Hollinshead</td>
<td>5 min.</td>
<td>F</td>
<td>Yes</td>
<td>PFO, PDA, anomalous septum right atrium</td>
<td>Medical</td>
<td>Died, CHF</td>
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<tr>
<td>(1944)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Palladino, Kinney</td>
<td>3 yrs</td>
<td>M</td>
<td>No</td>
<td>None</td>
<td>Medical</td>
<td>Died, CHF</td>
</tr>
<tr>
<td>(1948)</td>
<td>3 mos</td>
<td>M</td>
<td>No</td>
<td>None</td>
<td>Medical</td>
<td>Died, pneumonia</td>
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<tr>
<td>Barritt, Urich</td>
<td>28 yrs</td>
<td>M</td>
<td>Yes</td>
<td>None</td>
<td>Medical</td>
<td>Died, CHF</td>
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<tr>
<td>(1956)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kincaid et al (1962)</td>
<td>6 mos</td>
<td>F</td>
<td>Yes</td>
<td>PFO, PDA</td>
<td>Medical</td>
<td>Died, pneumonia</td>
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<tr>
<td>(1962)</td>
<td>5 da.</td>
<td>F</td>
<td>Yes</td>
<td>PFO, PDA</td>
<td>Medical</td>
<td>Died, CHF</td>
</tr>
<tr>
<td>Urban (1963)</td>
<td>16 yrs</td>
<td>P</td>
<td>No</td>
<td>None</td>
<td>None</td>
<td>Died, suddenly</td>
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<tr>
<td>Reisman et al (1965)</td>
<td>12 hrs</td>
<td>P</td>
<td>Yes</td>
<td>PFO</td>
<td>Surgical exploration</td>
<td>Died on table</td>
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<tr>
<td>Jordan, Taylor (1966)</td>
<td>6 hrs</td>
<td>F</td>
<td>Yes</td>
<td>PFO</td>
<td>Medical</td>
<td>Died, CHF</td>
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<tr>
<td>(1966)</td>
<td>2 da.</td>
<td>F</td>
<td>Yes</td>
<td>PFO, mitral valve dysplasia</td>
<td>Medical</td>
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<tr>
<td>Sanyal, et al (1968)</td>
<td>6 da.</td>
<td>M</td>
<td>Yes</td>
<td>PFO, PDA</td>
<td>Medical</td>
<td>Died, CHF</td>
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<tr>
<td>Antia, Osunkoya (1969)</td>
<td>3 yrs</td>
<td>M</td>
<td>No</td>
<td>None</td>
<td>Medical</td>
<td>Died, CHF</td>
</tr>
<tr>
<td>Barr et al (1974)</td>
<td>All</td>
<td></td>
<td>Yes</td>
<td>All PFO or ASD</td>
<td>Two explored;</td>
<td>All died</td>
</tr>
<tr>
<td>(1974)</td>
<td>neonate</td>
<td></td>
<td></td>
<td>One, small VSD</td>
<td>Pulmonary valvotomy, TV replaced with Bjork-Shiley prosthesis</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3 M</td>
<td></td>
<td></td>
<td>One, severe PS</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>1 F (?)</td>
<td></td>
<td></td>
<td>One pulmonary atresia</td>
<td></td>
<td></td>
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<tr>
<td>Aaron, Mills, Lower (present report)</td>
<td>18 yrs</td>
<td>M</td>
<td>No</td>
<td>None</td>
<td>TV replaced with porcine xenograft;</td>
<td>Survived</td>
</tr>
<tr>
<td></td>
<td>50 yrs</td>
<td>F</td>
<td>No</td>
<td>None</td>
<td>TV replaced with porcine xenograft;</td>
<td>Survived</td>
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ASD = Atrial Septal Defect  
CHF = Congestive Heart Failure  
PDA = Patent Ductus Arteriosus  
PFO = Patent Foramen Ovale  
PS = Pulmonary Stenosis  
TV = Tricuspid Valve  
VSD = Ventricular Septal Defect

Heart murmur. In the interim, she gradually developed severe dyspnea on exertion, orthopnea, pedal edema and easy fatigability. Cardiac catheterization at the National Naval Medical Center rendered the impression of Ebstein's anomaly. On examination, cyanosis was absent, the neck veins were minimally distended, the liver was not enlarged and there was minimal pretibial edema. The cardiac rhythm was irregular and there was a grade 3/6 blowing pansystolic murmur at the left sternal border. The ECG indicated atrial fibrillation and right intraventricular conduction delay. Chest x-ray film revealed a massive cardiac enlargement.

On September 4, 1974, cardiac exploration demonstrated a hugely dilated right atrium with an intact atrial septum. The tricuspid annulus was 10 cm in diameter and the valve was based on the anatomic tricuspid annulus. The anterior and posterior leaflets were fused and bound down by shortened chordae to the right ventricular wall, and the septal leaflet appeared normal. The right ventricle was large and muscular and did not show the saccular segment characteristic of Ebstein's anomaly. The defective tricuspid valve was replaced by a 35 mm stented porcine xenograft and the atrial size was reduced.

Ten months after surgery, the patient had mild fatigability, functions within class 2 (New York Heart Association) limitations and the cardiac size is appreciably decreased. Recent right heart catheterization demonstrated mean right atrial pressure of 7 mm Hg and pulmonary artery pressure at 28/8 mm Hg.

**DISCUSSION**

The report of Becker et al succinctly established the difference between Ebstein's anomaly and pure congenital tricuspid insufficiency. Eleven of 36 (31
percent) of the specimens reviewed had no displace-  
ment of the tricuspid valve, and thus no atrial- 
ized ventricle. To be sure, the valves that were  
displaced also regularly had various manifestations  
of dysplasia, but the division of the right ventricle  
was the critical distinction. Their findings were sim- 
ilar to those of others concerning the deficiencies of  
the tricuspid valve,11*18JB-18  
1) focal or diffuse  
thickening of the valve leaflets; 2) deficient devel- 
opment of the chordae tendineae and papillary mus- 
cles, most often binding down or "tethering" the  
valve margin; 3) improper separation of valve com- 
ponents from the ventricular wall; 4) focal agenesis  
of valvular tissue. These faults, present in varying  
degrees and combinations, result in a valve which is  
grossly insufficient, occasioning tremendous dilata- 
tion and hypertrophy of both the right ventricle and  
atrium (Fig 1).

The pathologic-anatomic distinctions between  
CTI and EA then, are these: 1) The functional right  
ventricular pumping chamber in the more severe  
forms of EA is typically small and limited to the  
outflow tract, whereas in CTI the right ventricle is a  
huge muscular structure with no volume limitations.  
2) That part of the right ventricle in EA that is  
proximal to the valve, the so-called atrialized ven- 
tricle, has been shown by contrast studies to impede  
right atrial emptying by competing with the ven- 
tricular inflow orifice,7,18 while in CTI it is simply a  
wide open insufficiency of the atrioventricular valve.  
3) Following surgical repair wherein the tricuspid  
valve is replaced in the anatomic location, the atrial- 
ized ventricle in EA may become a liability to the  
remaining small volume right ventricular pump, act- 
ing in effect as a paradoxing aneurysm. No such  
problem exists in CTI, as the entire ventricle is an  
active pump. 4) In EA, the large sail-like anterior  
leaflet may, by its abnormal attachments, obstruct  
the outflow tract.8 This problem does not occur in  
CTI.

Associated anomalies in CTI are mainly confined  
to an interatrial communication (patent foramen  
owale, atrial septal defect), present in 14 of the 22  
collected cases. A small ventricular septal defect, of  
no functional importance, was reported in one case.  
The mitral valve was deficient or dysplastic in three  
of the 22 cases. The ductus was patent in four. In the  
postmortem series of Becker et al,4 some form of  
pulmonary obstruction (stenosis or atresia) was  
found in 80 percent of their cases of simple CTI, 33  
percent in EA, yet this combination has been docu- 
mented in only three of the 22 cases reported, and  
these all in one series.17

The major and consistent clinical sign in CTI is  
congestive heart failure, usually manifest in the neo- 
natal period, with attendant marked cardiac en- 
largement, a prominent precordial right ventricular  
pulsation, systolic thrill, pansystolic murmur at the  
left sternal edge and a soft second heart sound. By  
contrast, in EA congestive heart failure occurs in  
only 30 percent of the reported cases. Cyanosis is  
present and profound in all of the early crisis cases

**FIGURE 1.** At left is depicted dysplasia of the tricuspid valve in congenital tricuspid insufficiency. The leaflets are poorly differentiated, fused, retracted with margins "tethered" by abortive chordae. The illustration at right shows the deformed tricuspid valve translocated into the ventricle to diminish the size of the effective right ventricle and create the third chamber that distinguishes Ebstein's anomaly.
due to right-to-left atrial shunting, but those who escape the early congestive heart failure generally remain free of cyanosis. This conclusion derives from the fact that all but one of the post-neonatal survivors in this series had intact atrial septa. 

Chest x-ray films in CTI consistently show marked enlargement of the cardiac silhouette, indicating a globular or box-shaped heart, and decreased perfusion of the lungs where right-to-left shunting occurs. In EA, the enlargement of the cardiac silhouette varies from marginal to huge. 

The electrocardiogram typically shows right axis deviation, right atrial dilatation and right bundle branch block, as is seen in EA. None of the collected cases of CTI showed the type B Wolff-Parkinson-White syndrome as is seen in about 10 percent of EA, but the occurrence of rhythm disturbances, i.e., paroxysmal supraventricular tachyarrhythmias, ventricular arrhythmias, atrial fibrillation and frequent premature beats of many types would indicate a similar hyperexcitability in both entities. Our two cases illustrate this well, showing a bizarre combination of defects in the first, and atrial fibrillation with a slow ventricular response in the second patient.

In both CTI and EA, cardiac catheterization usually shows normal or slightly elevated right atrial and right ventricular pressures with replacement of the "X" descent in the atrial pressure curve by a positive systolic curve. The right-to-left shunt at the atrial level can be bidirectional, or absent as seen in the older patients, and right ventriculogram shows wide open tricuspid insufficiency with little or hesitant forward flow through the pulmonary artery due to dye dilution in the large chambers. The absence of ventricular depolarization proximal to the tricuspid valve demonstrated by utilizing an intracardiac electrocardiogram probe, should confirm that EA is not present in simple CTI. This is likely to be the only distinguishing feature to separate the two entities preoperatively.

CTI is decidedly a lethal condition judging by the fact that every case in the literature, so far reported, has died. The bulk of the deaths are in the neonatal and infancy period (14 of 22) and result from unrelenting congestive heart failure or related pneumonia. The real mystery surrounds the group that somehow gets through this period only to develop gradual but progressive congestive heart failure at some later date. It is tempting to think that these delayed cases are a group with minimal insufficiency who develop acquired valvular lesions secondary to rheumatic fever or infection and thus succumb as the valve deteriorates. Indeed, at least one author has postulated that the acquired tricuspid insufficiency seen occasionally in rheumatic mitral valve disease occurs in congenitally abnormal valves. Review of the delayed cases in this collected series, with rare exception, fails to reveal either a history of rheumatic fever or endocarditis, or other valve lesions of consequence, and the tricuspid valves have been markedly abnormal with gross chamber enlargement, a feature not seen in the acquired form of tricuspid insufficiency.

The single case of sudden, unexpected death would excite little interest save for the fact that in the closely related EA, sudden death on a rhythm basis is not unusual, especially in those cases with intact atrial septum, and CTI has a similar propensity for rhythm disturbances.

Medical treatment of the congestive heart failure stemming from CTI has been ineffective in the infant group. In the older group, some response was seen, though the ultimate resolution was death after a protracted worsening course.

Besides our two cases, four patients have been subjected to surgery with only one attempt to replace the tricuspid valve. These ended fatally. Our two cases had hazardous and difficult recoveries, reflecting the advanced nature of their illness, but have survived for 10 to 12 months now and show signs of continued improvement. The availability of the stented porcine xenograft (Hancock Laboratories), which seems to be a very adequate tricuspid replacement, has been a substantial asset in managing these difficult patients. This valve, with considerable experience in all valve positions, has enjoyed singular success in five years of trials now.

Our success in treating the older age group is indicative that this entity can be approached surgically with confidence in those with hearts large enough to pump safely. The moribund neonates still present a formidable challenge to the cardiologist and cardiac surgeon alike.

REFERENCES

1 Brekke JC: Congenital tricuspid insufficiency. Am Heart J 29:647-649, 1945
8 Dubin IN, Hollinshead WH: Congenitally insufficient tricuspid valve accompanied by an anomalous septum in the right atrium. Arch Path 38:225-228, 1944
9 Palladino VS, Kinney TD: Cardiac hypertrophy and congenital tricuspid insufficiency; report of two cases. Bull Internat AM Mus 28:23-33, 1948