Spontaneous Resolution of Massive Congenital Tricuspid Insufficiency

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SUMMARY Four cases of congenital tricuspid insufficiency (CTI) are presented in whom spontaneous improvement occurred. There was no evidence of Ebstein’s disease in three of the four. All presented with similar clinical and laboratory findings including massive cardiomegaly, mild to moderate congestive heart failure, and moderate to severe desaturation. Catheterization data demonstrated severe TI and right-to-left atrial shunting. During the period of follow-up, up to 3.5 years, all symptoms have spontaneously disappeared. Clinical and angiocardiographic evidence for minimal residual TI have remained. Right atrial and right ventricular volume measurements were markedly abnormal initially and returned to normal or near normal. These measurements may be a useful method for quantitating the severity of tricuspid insufficiency. The course of the three infants with massive CTI (without Ebstein’s disease) differs from previous reports and may be important both in formulating future management plans and in defining the etiology of the disorder.

CONGENITAL TRICUSPID INSUFFICIENCY (CTI) is an uncommon but recognized cause of neonatal congestive heart failure, massive cardiomegaly, and cyanosis. At least three distinct etiologies of severe CTI have been reported: dysplastic tricuspid leaflets or chordal attachments without their displacement, dysplastic and displaced leaflets (Ebstein’s anomaly), and TI secondary to pulmonary atresia with an intact ventricular septum. Four symptomatic infants with massive congenital tricuspid insufficiency were studied at the Children’s Hospital of Vanderbilt Medical Center during the immediate newborn period. Three of the four infants had no evidence for tricuspid valve displacement, and none had pulmonary stenosis or atresia. The fourth infant had suggestive evidence of tricuspid valve displacement. The three infants with severe CTI but without valve displacement had a clinical presentation and natural history similar to the infant with suspected valve displacement and to the reported cases of Ebstein’s anomaly presenting during the neonatal period. Evidence is presented that the prognosis of infants previously reported to have isolated massive CTI is not uniformly poor. Not only did all symptoms resolve in the four infants but overall heart size and the severity of the TI decreased as corroborated by serial right heart volume measurement. Since the course of these infants with severe CTI differs significantly from the experience of others, further consideration should be given both to the management and to the etiology of a seemingly heterogeneous patient group who have been considered to have massive CTI secondary to dysplastic tricuspid leaflets.

Case Material

Four neonates, two females and two males, were referred in the first week of life to the Vanderbilt Medical Center Children’s Hospital between October 1972 and March 1974 and have been followed from 1.75 to 3.5 years. Maternal pregnancy, labor and deliveries were uncomplicated. One of the neonates required resuscitation (G.H.). Weights of the four neonates at birth ranged from 3.3 to 3.7 kg (mean 3.5 kg). Within 24 hours after birth central cyanosis was evident, and in three of the four neonates there was associated evidence of congestive heart failure. Left ventricular oxygen saturations at the initial catheterization ranged from 30% to 91% (mean 74%).

On admission the cardiovascular examination revealed a harsh systolic murmur along the left sternal border that was
graded from I/VI to IV/VI in intensity. There were no third or fourth heart sounds except in one patient (E.S.). Nonpul- satile hepatomegaly, tachycardia, and respiratory distress were present in three of the four infants, and these infants were subsequently digitalized over 24 hours with parenteral digoxin (0.040 mg/kg/24 hr in three doses) and placed on oral digoxin. The digoxin was discontinued at six weeks in two infants (K.H. and G.H.). One infant (E.S.) is currently on maintenance digoxin. No additional therapy was required.

AP and lateral chest X-rays in the four patients revealed marked cardiomegaly as illustrated in figure 1. Pulmonary vascular markings were normal or diminished. Twelve lead electrocardiograms revealed right atrial enlargement (P wave > 2.5 mm) and right ventricular hypertrophy (qR and/or R > 28 mm in V_1, R and V_2) in the four patients. Two of the infants (E.S. and G.H.) also had suggestive evidence of biventricular hypertrophy as evidenced by a top normal R wave in V_6, and > 45 mm R and S in the mid-precordial leads. The P-R intervals ranged from 0.08 to 0.12 sec and the maximal QRS duration ranged from 0.04 to 0.08 sec (mean 0.06 sec). One infant (E.S.) had possible right ventricular pre-excitation.

All four infants have been followed from 1.75 to 3.5 years (mean 2.7 yrs). During this period all four infants became acyanotic and asymptomatic. Each has had normal physical and mental development. At the end of the follow-up period, one of the four infants had a normal physical examination and three (K.H., M.S., E.S.) had residual systolic murmurs Grade I-II/VI. The electrocardiograms of two infants (K.H., M.S.) became normal during the follow-up period, and two infants (E.S., G.H.) had persistent evidence of right ventricular hypertrophy (rSR' pattern in V_1, R and V_2). The cardiothoracic ratio on AP chest films reverted to normal in two infants (K.H., G.H.) and decreased in all four (fig. 2). The initial C/T ratio ranged from 0.77 to 0.99 with a mean of 0.87 and the follow-up C/T ratios ranged from 0.56 to 0.71 with a mean of 0.61.

Cardiac Catheterization

A total of eight catheterizations provided the hemodynamic data for the four infants. Venous catheterizations of the four infants were performed without complications shortly after admission, and the results are shown in table 1. Repeat percutaneous catheterizations in these same patients were performed 4½ to 18 months after the first study. All pressure recordings were made with precalibrated Statham fluid-filled transducers and 5 or 6F NIH catheters. Oxygen saturations were obtained in rapid succession with a reflectance oximeter. Intracardiac electrograms were obtained in all infants during their second catheterization with a 6F end-hole catheter equipped with a unipolar electrode for simultaneous pressure recordings according to previously described techniques.

As indicated in table 1, there was no tricuspid or pulmonary stenosis in any of the infants. All infants had right-to-left shunting at the atrial level and were peripherally desaturated. All four infants had sharp and simultaneous transitions in the simultaneous pressure and electrograms during slow catheter withdrawal from right ventricle (RV) to right atrium (RA).

Angiocardiology

Biplane cineangiograms of the RA and RV were obtained following contrast injection of sodium and meglumine diatrizoates at 1-1.25 cc/kg. RV injections were made with special care taken to ensure that the catheter holes were well positioned in the body of the RV and that there was no catheter recoil into the RA during the injection. TI was defined as regurgitant contrast appearing in RA during RV injection that occurred with each systole excluding premature beats, according to previously described criteria.

![Figure 2. The cardiothoracic ratio as a function of age. The approximate normal value is depicted by the solid horizontal line at 0.55.](http://circ.ahajournals.org/content/54/5/796.full)
All four infants had severe TI on the initial catheterization (fig. 3) with right-to-left shunting across an atrial septal defect (or patent foramen ovale). On the follow-up catheterization, there were no atrial septal defects and only minimal TI in three infants (K.H., G.H. and M.S.). One infant (E.S.) had mild to moderate residual TI and in addition had suggestive evidence for ventricular displacement of a leaflet of the tricuspid valve (fig. 4) which is characteristically seen in patients with Ebstein's anomaly.9 Since this infant also had an ECG pattern compatible with right ventricular preexcitation and a prominent S₁ and S₂, she was considered possibly to have Ebstein's anomaly despite the normal intracardiac pressure-electrogram tracing. While there was questionable mild thickening of the pulmonary valves in all four infants on the initial catheterization, there was no pulmonary stenosis (MPA to RV pressure gradients < 20 mm Hg). At the initial catheterization, there was a small left-to-right shunting through a patent ductus arteriosus visualized in all four infants. The ductus was closed in all patients at the subsequent study.

Cineangiographic Volume Determinations

From the biplane cineangiograms, RA, RV, LA and LV volumes were determined according to previously described methods.13-15 Right atrial and ventricular volumes were determined using a modified Simpson's rule method with the AP and lateral images divided into 20 equal parts using 21 horizontal lines. The cross-section of the chamber when viewed from above was assumed to be elliptical, the volume of each segment was calculated, and the segment volumes summed using Simpson's rule. This method has been validated previously with in vitro studies of right atrial and right ventricular casts ranging in size from 16 to 134 ml for the right atrium and from 1 to 112 ml for the right ventricle. An equally good correlation between calculated and true volumes was obtained for both the very small and the large chambers.13, 14

In figures 5 and 6 the respective chamber volumes are expressed as a percent of predicted normal.13, 14 The RA maximal volume, expressed as percent of normal, was significantly increased as calculated from angiograms taken during the initial catheterization in all four infants and ranged from 225 to 759% of normal (fig. 5). The RA max volume ranged from 50 to 271 ml/m² (mean 161 ml/m²) (table 2). The RA max volume dramatically decreased during the follow-up period (fig. 5) to values that ranged from 113 to 330% of normal. Three of the four values were still above normal and ranged from 63 to 136 ml/m² (mean 93 ml/m²); one patient (G.H.) had a normal RA max volume of 51 ml/m². The RVEDV expressed as percent of normal was also significantly increased above the normal with one exception (G.H.) and ranged from 121 to 357% of normal (fig. 6). The RVEDV ranged from 45 to 137 ml/m² (mean 78 ml/m²). The RVEDV returned to normal in three of the four infants during the follow-up period and ranged from 40 to 56 ml/m² (mean 52 ml/m²). The RVEDV of the fourth patient (E.S.) showed a decrease from 357% of normal (137 ml/m²) to 147% of normal (67 ml/m²) (table 2).

As calculated from the volume data, the RVEF (fig. 7) and the RV outputs (fig. 8) were either normal or increased. The LVEDV was increased in two infants (G.H. = 61 ml/m² and K.H. = 48 ml/m²) and was normal in the remaining two patients at the initial catheterization (27 and 35 ml/m²). LVEDV was normal at the time of recatheteri-
Surgical Technique

Placement of the intra-atrial baffle in these patients was similar to that used for corrective operations. High-flow cardiopulmonary bypass (3.3 L/min/m²) was instituted after direct cannulation of the venae cavae, and perfusate temperature was rapidly dropped to the 25–28°C range. This permitted a 50% reduction in flow rate, if necessary, for visualization of the left atrium in the presence of a massive bronchial collateral return. In three patients, profound hypothermia was required (17–20°C) to permit intermittent (10–15 min) intervals of total circulatory interruption in order to obtain a completely clear operative field for the accurate placement of sutures.

Elastic dacron fabric was used to construct the baffle around the cavae, excluding the pulmonary veins. The suture line was brought below the coronary sinus in six patients, diverting this flow into the new pulmonary venous atrium. The two patients (2 and 3) with a large left superior vena cava draining into the coronary sinus, absence of an innominate vein, and left juxtaposition of the atrial appendages (JAA) required special attention. In patient 2 the anterior margin of the baffle was sutured into the base of the septal leaflet of the tricuspid valve and the coronary sinus was fenestrated adjacent to the mitral valve, diverting coronary sinus and left superior vena cava drainage into the systemic venous atrium. In patient 3, because of the JAA and the high position of the entrance of the left superior pulmonary vein, particular care was taken to avoid obstruction to atrial venous inflow; again the coronary sinus was fenestrated, directing coronary sinus and LSVC flow to the new systemic venous atrium. In seven of eight cases, the new pulmonary venous atrium was enlarged with a pericardial patch.

The large patent ductus arteriosus, present in three patients, could not be approached safely through the median sternotomy incision because of tremendous enlargement of the main and the left pulmonary arteries. Consequently, these short friable ducts, each 1.0–1.5 cm in diameter, were approached through a separate left thoracotomy incision prior to the median sternotomy. In the first patient encountered with this anomaly (6), the PDA was divided, chest closure completed, and the patient returned to the intensive care unit with recatheterization planned prior to the Mustard procedure, 5–7 days later. However, severe hypoxemia and complete ventilator dependence, presumably on account of abolition of a major site of mixing of the systemic and pulmonary circulations, necessitated undertaking the baffle procedure on the third postoperative day. On the basis of this experience, the two subsequent patients with large PDAs underwent division of the PDA and the Mustard baffle operation on the same day. Aortic coarctations, present in two patients, were not resected because the obstruction appeared mild by angiography (fig. 3). In patient 3 the hemodynamically insignificant ductus was closed through the pulmonary artery after instituting cardiac bypass.

Three of the eight patients required reoperation for bleeding within the first 24 hours postoperatively. No specific coagulation defects were identified prior to operation in these three patients, although mild prolongation of both the prothrombin time and activated partial thromboplastin time was present in two of the three. All patients received platelet transfusions and fresh frozen plasma at the

right-to-left shunting through a patent ductus (fig. 3); two presented angiographic evidence of coarctation of the aorta (table 1, fig. 3). Three of the eight patients had undergone surgical atrial septectomy, and one of these, an ineffective pulmonary artery band. All patients at surgery were noted to have evidence of large bronchial collaterals.
those patients with more severe TI both values would be significantly increased.

The three cases described with isolated tricuspid insufficiency had suggestive evidence of altered hemodynamics in utero secondary to tricuspid insufficiency since all manifested marked cardiomegaly and ECG evidence of right ventricular hypertrophy at the first exam within 24 hours of birth. All the cases demonstrated that in the neonate with tricuspid insufficiency, systemic venous return may be partially shunted across the patent foramen ovale resulting in peripheral arterial desaturation. In regard to the clinical presentation, the three neonates with isolated congenital tricuspid insufficiency without displacement presented here do not differ from: (a) previously reported cases of severe isolated CTI without Ebstein's, (b) the single patient in this group with suspected Ebstein's anomaly of the tricuspid valve, (c) previously reported cases of Ebstein's in the neonate, (d) previously reported cases of TI associated with pulmonary atresia with intact ventricular septum. Since clinical distinction may be impossible, catheterization is indicated realizing that angiocardiographic distinction between these lesions may also be difficult. The importance of visualizing the pulmonary artery during a right ventricular angiocardiogram to exclude pulmonary atresia with intact ventricular septum should be emphasized since prompt surgical management is indicated in this group.

Excluding those neonates with TI secondary to pulmonary atresia, the management of CTI is unclear. Medical management of neonates with Ebstein's anomaly and massive TI is currently recommended since the severity of the TI usually decreases, and surgical replacement of the tricuspid valve carries high morbidity and mortality. In the one infant (E.S.) considered to have Ebstein's anomaly and managed medically, spontaneous improvement in the symptoms and degree of TI was observed. The three other infants had no evidence of valve displacement angiographically on either the initial or follow-up catheterizations, and the etiology of their TI is unclear. The normal RV and LV ejection fractions and systolic outputs of these infants do not suggest a generalized cardiomyopathy which might result in a dilated tricuspid annulus and secondary TI. Congenital dysplastic valve leaflets or shortened chordae have been documented by several pathological studies of children with massive TI who died. It is likely that these infants had a mild degree of tricuspid valve dysplasia. If so, then the natural history of CTI in certain infants with dysplastic leaflets may be considerably better than had been previously reported and may resemble those patients with true Ebstein's anomaly of the tricuspid valve. This has important therapeutic consequences since surgical replacement of the tricuspid valve had been advocated in infants with severe CTI secondary to dysplastic valve leaflets.

It is possible that the three infants without evidence of valve displacement do not have a primary tricuspid abnormality but have tricuspid insufficiency secondary to in utero RV pressure overload with premature (in utero) closure or constriction of the ductus arteriosus. Such a case of premature ductal closure has been reported; however, tricuspid competence was not assessed. The infant had severe cyanosis, congestive heart failure and massive cardiomegaly which spontaneously improved shortly after birth. All infants in this study had a patent ductus arteriosus at the time of their initial catheterization and this possible etiology remains speculative.

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Right Ventricular Volume Characteristics in Ventricular Septal Defect

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SUMMARY Right and left ventricular volume characteristics were determined from biplane cineangiocardiology in 37 patients with isolated ventricular septal defects. Patients were divided into three categories as determined by the degree of left-to-right shunt: small shunt—less than 35% of pulmonary blood flow (N = 9); moderate shunt—35–49% (N = 8), and large shunt—greater than 50% (N = 20). Right ventricular (RV) end-diastolic volume was increased above normal in 15 of 20 studies performed in patients with large left-to-right shunts and averaged 159 ± 10% of normal (P < 0.001). In contrast, only one of the patients in the small shunt group and only half of the patients in the moderate shunt group showed increases in RV end-diastolic volume. The increase in RV volume was proportional to the corresponding increase in left ventricular end-diastolic volume, with the right ventricle ranging from 48 to 116% of LV end-diastolic volume (average 83%). Right ventricular ejection fraction was normal in all patient groups. Right ventricular output was increased commensurate with the increases in the RV end-diastolic volume. These data indicate that substantial augmentation in RV end-diastolic volume does occur in patients with isolated ventricular septal defects and large left-to-right shunts. These data can be explained by the significant diastolic and "isovolumic" shunting from left ventricle to right ventricle which occurs in these patients.

IN AN ISOLATED VENTRICULAR SEPTAL DEFECT (VSD), the major hemodynamic overload has been considered to be on the left side of the heart with consequent increases in left atrial and left ventricular size. These increases in left heart volume variables have been documented and agree well with the degree of left-to-right shunting in patients with VSD. In contrast, however, the right ventricular response to an isolated VSD in terms of ventricular dilatation as a function of degree of left-to-right shunt has not been documented. The purpose of this investigation, therefore, was to determine right ventricular end-diastolic volume, ejection fraction, and systolic output in patients with isolated VSD and to relate these variables to similar left ventricular volume characteristics as well as to other hemodynamic variables reflecting the size of the left-to-right shunt.

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Methods

Infants and children undergoing cardiac catheterization at Vanderbilt University and having the diagnosis of an isolated ventricular septal defect constitute the study population. Patients with a significant atrial shunt (mixed venous blood to right atrial O2 saturation stepup of >7%), semilunar valvular incompetence, or atroventricular valvular incompetence were excluded from this study. All data were obtained during diagnostic cardiac catheterization. Patients less than six weeks of age received no premedication, but occasionally were given small doses of morphine (0.05 mg/kg). Patients from six weeks to two years of age were sedated with meperidine, 1 mg/kg, and hydroxyzine, 1 mg/kg i.m., given 30 min before the beginning of the catheterization procedure. Occasionally, additional doses of meperidine of 0.1 to 0.5 mg/kg were required for sedation during the procedure. Patients above two years of age were sedated with Innovar, 0.025 ml/kg, up to a maximum of 1 cc i.m. given 30 min prior to the procedure. Rarely, additional doses of Innovar of ¼ to ½ of the original dose were required for sedation. Right and left heart pressures were obtained with NIH catheters with zero
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