

Left Ventricular Function in Tricuspid Atresia

Angiographic Analysis in 28 Patients

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SUMMARY

Thirty-one left ventricular (LV) biplane angiograms were performed in 28 patients with tricuspid atresia. Measurements of left ventricular end-diastolic volume (LVEDV) and left ventricular end-systolic volume were obtained by the modified Simpson's rule and systolic ejection fraction (EF) calculated. Left ventricular volumes and ejection fractions were also obtained in 19 control patients with no significant heart disease. The patients with tricuspid atresia were classified according to the appearance of the pulmonary vascularity on initial radiologic examination: Group A, decreased pulmonary vascularity; Group B, increased pulmonary vascularity. In the 13 group A infants who were unoperated, LVEDV was increased and EF mildly diminished. In the group B patients LVEDV was increased and EF normal. In the 12 group A patients with surgical shunts LVEDV was elevated. The five group A patients with long-standing systemic artery to pulmonary artery anastomoses (> 10 years) showed the largest LVEDV and the poorest EF. The angiographic data indicate that patients with tricuspid atresia experience significant LV dysfunction as a consequence of longstanding LV volume overload. The early detection of LV dysfunction may be an indication for a right ventricular bypass procedure in these patients.

TRICUSPID ATRESIA is a rare but severely life-limiting malformation of the heart. We recently reported a series of 101 patients from the Children's Hospital Medical Center, Boston, Mass., in which survival to 17 years of age with palliative surgery was 50%.¹ Early survival was related to the success of surgical intervention and adequacy of pulmonary blood flow. There was an increase in mortality beginning in the middle of the second decade; two-thirds of patients alive at age 15 died by age 23. Causes for the late mortality in this group could be related to the state of the myocardium since only two patients beyond two years of age, free of congestive heart failure, died at operation. Long-term survivors have been noted to have normal to increased pulmonary blood flow and frequently exhibit congestive failure.^{2, 3} To investigate the role of left ventricular function in the survival of patients with

tricuspid atresia, analysis of left ventricular biplane angiograms performed in 28 patients was undertaken.

Materials and Methods

Thirty-one left ventricular (LV) biplane angiograms, obtained in 28 of 101 patients with tricuspid atresia, were available for analysis; the remaining 73 patients either did not undergo cardiac catheterization (24) or did not receive left ventricular angiography adequate for analysis (49). The selected 28 patients ranged in age from one day to 20 years (median six months). Systemic saturation in each patient was measured and pulmonary flow to systemic flow ratio (Q_p/Q_s) calculated.

The 28 patients were classified into their radiologic group according to appearance of the pulmonary vascularity, as previously reported.^{1, 4} Twenty-one group A patients had decreased pulmonary vascularity prior to surgical palliation. Volume studies were available prior to surgery in ten patients, after surgery in nine patients, and both before and after surgery in three patients. Six patients had normal or increased pulmonary vascularity (group B) and did not require shunt surgery. The radiologic appearance of the pulmonary vascularity was a reasonable estimate of pulmonary blood flow since both the mean Q_p/Q_s and systemic saturation were significantly different between the two unoperated groups (13 group A and six group B patients, $P < 0.02$, and $P < 0.001$ respectively). Table 1 relates the 31 volume studies to the patients' radiologic group and their surgical status.

Measurement of the left ventricular end-diastolic volume (LVEDV) and end-systolic volume was carried out by the method of Chapman based on modification of Simpson's rule.^{5, 6} Biplane cineangiography with a film speed of 60 frames/second was performed in 19 patients; the remaining 12 angiograms were obtained with cut film biplane angiography at six frames/second. Frames obtained during both extrasystolic and postextrasystolic beats were discarded. Left

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Table 1

31 Volume Determinations Related to the 28 Patients' Radiologic Group and Surgical Status*

Radiologic group	Surgical status	
	Not present	Present
Group A: 22 patients (Decreased pulmonary vascularity)	13	12
Group B: 6 patients (Normal or increased pulmonary vascularity)	6	0

*Includes volume determinations available on three patients before and after creation of a surgical shunt.

ventricular mass was calculated using the method of Rackley.⁷ Systolic ejection fractions were calculated. Left ventricular volumes and mass were determined in 19 patients with no significant heart disease and used as controls. Arbitrary division of patients below two years of age from those above two years of age was carried out in keeping with correlations previously observed between cast and angiographic volumes.⁸ When possible the tricuspid atresia patients and controls were matched for age, sex and body surface area. Nine control patients were under one year of age and were catheterized with the following findings: 1) persistent hypoxemia as a result of lung disease and persistent right to left shunt through a patent ductus arteriosus (4), 2) small patent ductus arteriosus (no significant left to right shunt) (2), 3) primary hypertension (120/80) at six months of age (1), 4) trivial pulmonary stenosis (gradient 13 mmHg) (1), 5) spontaneously closed ventricular septal defect at 12 months of age (1). The remaining ten control patients were greater than one year of age at the time of study; six had trivial pulmonary stenosis (gradient < 20 mm Hg at rest); four had no significant heart disease but were catheterized because of cardiac murmurs.

Statistical analyses were performed using the group and paired Student's *t*-test.

Results

Thirteen Unoperated Group A Infants

LVEDV and EF were determined in 13 unoperated group A infants, and were compared to the findings in nine infants without heart disease. As may be seen in figure 1, the LVEDV was significantly higher ($P < 0.02$) and the EF significantly lower ($P < 0.02$) in the tricuspid atresia group than in the controls. By contrast the left ventricular mass was not significantly different between the two groups. Within this subgroup of patients there was no correlation between either LVEDV or EF and Q_p/Q_s or systemic saturation.

Six Unoperated Group B Patients

Six unoperated group B patients, one week to 14 years of age, were studied. Three had normally related great arteries (Type I) and three transposed great arteries (Type II).⁹ The LVEDV was also increased compared to the matched controls ($P < 0.05$, fig. 2).

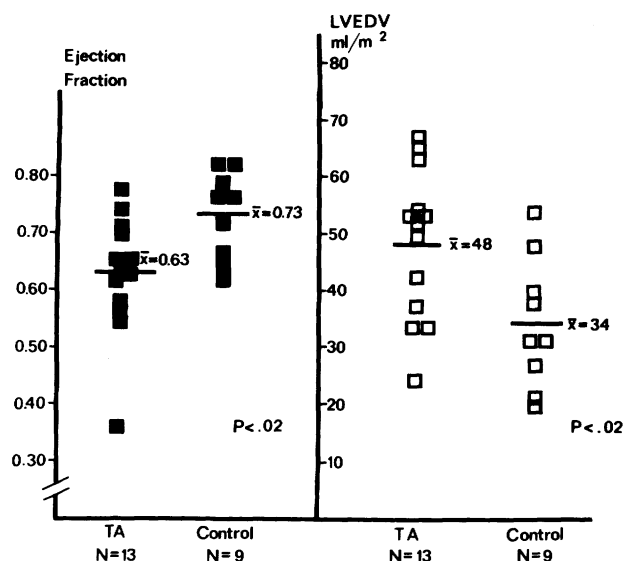


Figure 1

Left ventricular end-diastolic volume and ejection fraction in 13 group A patients compared to nine controls grouped for age and body surface area. Both the mean left ventricular end-diastolic volume (48 ml/m²) and ejection fraction (0.63) of the group A patients are significantly different than those of the control group ($P < 0.02$). EF = ejection fraction; LVEDV = left ventricular end-diastolic volume; TA = tricuspid atresia; X = mean.

This may be attributed to the volume overload on the left ventricle, enhanced in this instance by the (chest X-ray) increased pulmonary blood flow. Two of these six patients had greatly increased LVEDVs at age six and 14 years (one with d-transposition of the great arteries and one with normally related great arteries). Unfortunately serial data are not available in these two children so the influence of the duration of

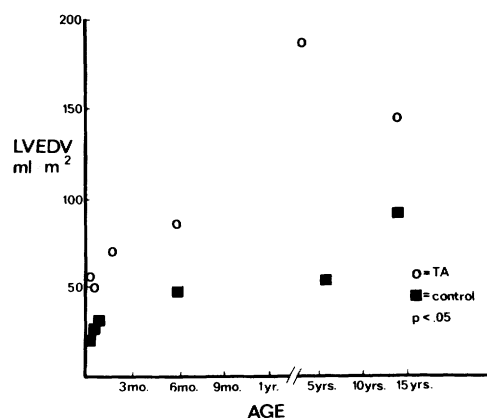


Figure 2

Left ventricular end-diastolic volume in six group B patients compared to six controls matched for age, sex, and body surface area. The left ventricular end-diastolic volume in the six group B patients is significantly greater than the controls ($P < 0.05$). LVEDV = left ventricular end-diastolic volume; TA = tricuspid atresia.

volume overload on the LVEDV cannot be evaluated. Ejection fractions in these six tricuspid atresia patients remained normal even at the older ages. There was no correlation between either LVEDV or EF and Q_p/Q_s or systemic arterial saturation.

Twelve Group A Patients with Surgical Shunts

LVEDV in the 12 patients (group A) with surgically created shunts was greater than normal (fig. 3). In addition, there was good correlation ($r = 0.74$) in this group of patients between the LVEDV and pulmonary to systemic blood flow ratio (fig. 4). However, the LVEDV of the five patients less than ten years of age was elevated to a lesser degree ($P < 0.01$) than the seven older patients ($P < 0.001$) when compared to control groups.

The ejection fraction was preserved in the five younger patients (< 10 years of age, $P > 0.05$, fig. 5) but was significantly decreased in the seven older children who had longstanding shunts ($P < 0.001$, > 10 years).

Seven Group A Patients with Longstanding Shunts

In the seven patients with longstanding surgical shunts (> 10 years), there were five systemic arterial shunts (4 Potts, 1 Blalock) and two systemic venous shunts (2 Glenn). Figure 6 shows a marked increase in LVEDV and a decrease in the EF in these seven

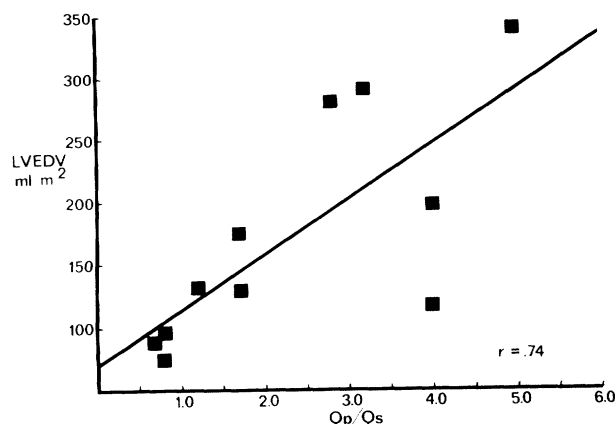


Figure 4

Left ventricular end-diastolic volume related to the pulmonary to systemic blood flow ratio in 11 of the 12 group A patients with surgical shunts. LVEDV = left ventricular end-diastolic volume; Q_p/Q_s = pulmonary to systemic blood flow ratio.

patients. In addition LV mass in these seven patients was significantly increased ($P < 0.005$). In these seven patients there was good correlation between LVEDV and Q_p/Q_s ($r = 0.80$). Fair correlation ($r = -0.60$) was obtained between EF and both Q_p/Q_s and systemic saturation. The two patients with Glenn anastomoses had the smallest LVEDV and least altered EF of the group. Table 2 shows the LVEDV

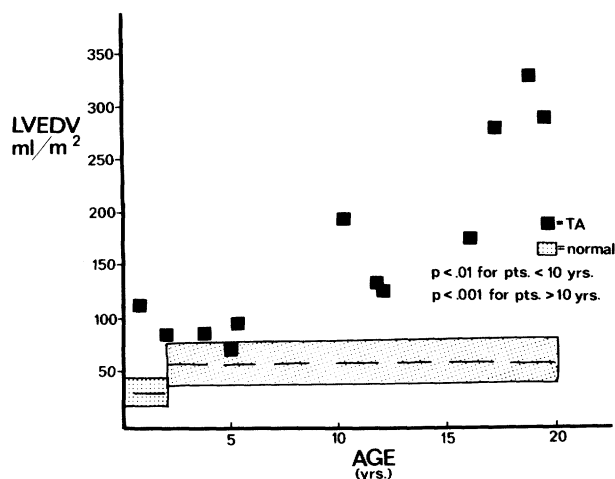


Figure 3

Left ventricular end-diastolic volume in 12 group A patients with surgical shunts compared to normal. The stippled rectangle shows the normal left ventricular end-diastolic volumes obtained from the 19 controls with mean and one standard deviation for children less than and greater than two years of age. The P values shown are based on comparison of tricuspid atresia patients with individual controls matched for age, sex, and body surface area. The left ventricular end-diastolic volume is elevated in the patients less than ten years of age ($P < 0.01$) but to a lesser degree than the patients greater than ten years of age ($P < 0.001$). LVEDV = left ventricular end-diastolic volume.

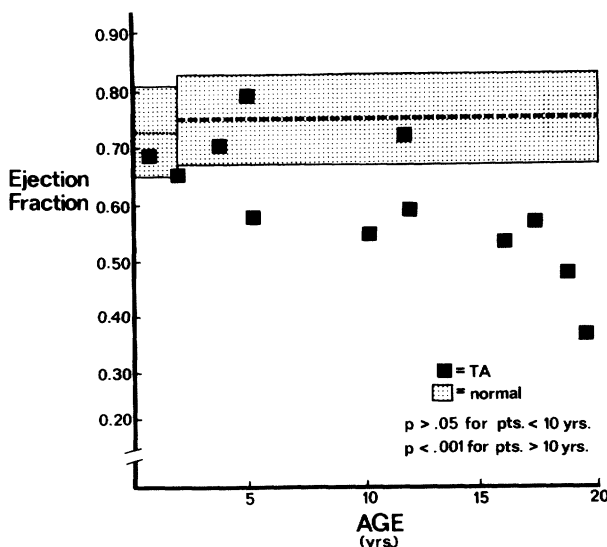


Figure 5

Ejection fraction in 12 group A patients with surgical shunts compared to normal. The stippled rectangle shows the normal ejection fraction obtained from the 19 controls with mean and one standard deviation for children less than and greater than two years of age. The P values were based on comparison of tricuspid atresia patients with individual matched controls. The ejection fraction in the patients less than ten years of age is preserved within the normal range ($P > 0.05$) but is significantly decreased in the patients over ten years of age ($P < 0.001$). TA = tricuspid atresia.

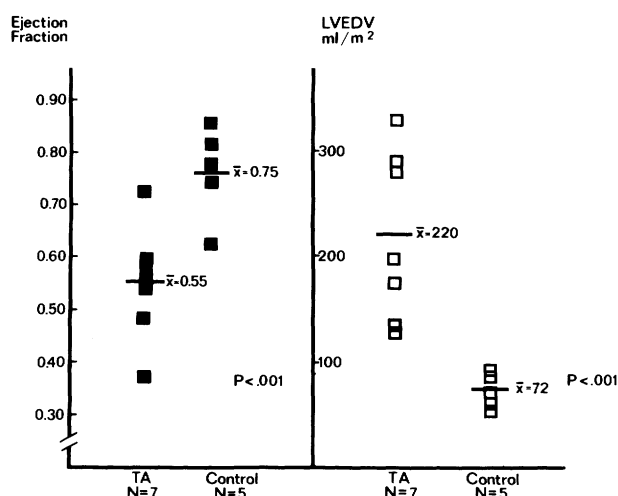


Figure 6

Left ventricular end-diastolic volume and ejection fraction in seven tricuspid atresia patients with longstanding surgical shunts compared to five controls. The mean left ventricular end-diastolic volume (220 ml/m²) and the mean ejection fraction (0.55) in the tricuspid atresia group is significantly different than the control group ($P < 0.001$). EF = ejection fraction; LVEDV = left ventricular end-diastolic volume; TA = tricuspid atresia; X = mean.

and EF in the five patients with systemic arterial to pulmonary artery shunts, and compares them with the two Glenn shunts, the five controls, and the total group of seven shunted patients. The mean LVEDV (254 ml/m²) for the five patients is greatly elevated and the mean EF is 0.50.

Discussion

Tricuspid atresia consists of complete absence of the right-sided atrioventricular valve, and underdevelopment of the right ventricle. There is an obligatory interatrial communication, and a ventricular septal defect connecting the left ventricle to the diminutive right ventricle. Transposition of the great arteries is found in 30% and obstruction to pulmonary blood flow is frequently present (70%) at either the ventricular (obstructive VSD) or pulmonary valve level.¹⁰⁻¹²

Table 2

LVEDV and EF in 7 Group A Patients with Longstanding Shunts (4 Potts, 2 Glenn, 1 Blalock)

	Mean LVEDV (ml/m ²)	Mean EF
5 control patients	72	0.75
7 patients with longstanding shunts (>10 years)	220	0.55
2 patients with Glenn shunts	130	0.66
5 patients with systemic to PA shunts	254	0.50

Abbreviations: EF = ejection fraction; LVEDV = left ventricular end-diastolic volume; PA = pulmonary artery.

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Such a combination of morphologic abnormalities imposes unique hemodynamic demands on the heart. The most significant alteration is that the left ventricle in tricuspid atresia functions as the major pump for both the systemic and pulmonary circulations. The left atrium receives both the pulmonary venous return and the systemic venous return from the right atrium and delivers this increased volume to the left ventricle. This in turn dilates the left ventricle as our volume data confirm. Such an increase in left ventricular volume is present as early as one week of age (four patients in this study).

Congestive heart failure is a common complication in tricuspid atresia patients with surgically created systemic artery to pulmonary artery anastomosis.^{2, 13-15} Furthermore, there are patients with tricuspid atresia who have died of congestive heart failure in the third and fourth decade of life who have never undergone palliative surgery.^{2, 3}

A number of factors appear to play a role in the development of LV dysfunction in patients with tricuspid atresia. The mildly diminished ejection fraction in the 13 unoperated group A infants may be explained by the presence of marked hypoxemia (mean saturation = 67%).¹⁶ In the ten children less than ten years of age (five group B and five operated group A patients), preservation of LV function may perhaps be explained by both a higher systemic saturation (mean = 84%) and the lack of volume overload of sufficient duration to cause significant LV dysfunction.

There were eight patients over ten years of age who showed variable LV function related to the duration and magnitude of the volume overload. Five of these eight patients had had surgical systemic artery to pulmonary artery shunts for longer than ten years (table 2) and all five had abnormal LV function as evidenced by both greatly increased LVEDV and LV mass, and moderately decreased ejection fraction (all less than 0.58). Although serial data are lacking in 25 of the 28 patients, the findings in these older patients suggest that the duration of the LV volume overload contributes to LV dysfunction in tricuspid atresia.

Two of these eight patients over ten years of age had longstanding Glenn anastomoses and their ejection fractions were preserved, a finding which indicates that LV function is less impaired in systemic venous to pulmonary artery shunts and may be explained by the absence of a LV volume overload.¹⁷ In the one remaining patient over ten years of age, without a surgical shunt, the normal ejection fraction may be related to the absence of a greatly increased pulmonary blood flow ($Q_p/Q_s:1.7/1$), a conclusion supported by the correlation obtained between both LVEDV ($r = 0.80$) and EF ($r = -0.60$) and the Q_p/Q_s in the patients with longstanding shunts. Thus,

the magnitude, as well as duration of the volume overload, appears to play an important role in producing LV dysfunction.

Ross and Somerville¹¹ have stated that in patients with tricuspid atresia, severe hypoxia with one or more inadequate shunts in the absence of pulmonary hypertension is an indication for a Fontan operation; they recommend this procedure for patients over ten years of age. Our survival data also suggest that a right atrial to pulmonary artery anastomosis should be a consideration in patients with tricuspid atresia in the early part of the second decade of life.¹ In addition, the angiographic data indicate that patients with tricuspid atresia experience significant LV dysfunction as a consequence of longstanding LV volume overload. The creation of a right atrial to pulmonary artery anastomosis with closure of the atrial septal defect results in the elimination of the obligatory right to left shunt and thereby the volume overload of the LV.^{11, 18-20} In patients with tricuspid atresia early angiographic evidence of decreased LV function may be an additional indication for right ventricular bypass procedure.

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