Surgical management of tricuspid atresia

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Tricuspid atresia is one of the less common forms of congenital heart disease. The results of palliative surgery in 72 children are presented. Cardiac catheterization and angiocardiology are essential for precise definition of the anomaly. There is a 20% incidence of obstruction at atrial septal level. Closed atrial septostomy is of value in such cases in infancy. In most there is a reduced pulmonary blood flow. This may be increased by anastomosis of either the superior vena cava or a systemic artery to the pulmonary artery. The caval anastomosis, while having theoretical advantages, is not always possible, especially in small infants. In this series systemic arterial shunts have given at least comparable results, suggesting that both techniques are of value with this anomaly.

Atresia of the tricuspid valve cannot be corrected by operation; however, palliative surgery has given encouraging results. This severe form of cyanotic heart disease is usually fatal in early childhood (Nadas, 1963; Keith, Rowe, and Vlad, 1967).

The classification of Edwards and Burchell (1949) is generally accepted. They described four variations depending on the presence of normal or transposed great arteries and normal or obstructed pulmonary outflow. Figure 1 represents this in diagram form. Keith *et al.* (1967) found that in a large series 69% were type I and 31% type II.

MATERIAL

Seventy-two children, in whom a diagnosis of tricuspid atresia was made, were admitted to this hospital in the years 1955 to 1967 inclusive. They ranged in age from 2 days to 7 years. Seventy-six operations were performed in 66 patients; six had no operation.

Preoperative clinical assessment, cardiac catheterization data, operative techniques, mortality, morbidity, and follow-up observations have been recorded and studied.

The following operative techniques were employed: Subclavian artery to pulmonary artery (Blalock and Taussig, 1945); subclavian autograft to pulmonary artery (classed as Blalock-Taussig); descending aorta to left pulmonary artery (Potts, Smith, and Gibson, 1946); ascending aorta to right pulmonary artery (Waterston, 1962); superior vena cava to right pulmonary

artery (Glenn, 1958); and exploratory thoracotomy.

OVERALL RESULTS

The age distribution and frequency of surgical procedures are presented in Table I. Early and late mortality in the whole series, in relation to age, is shown in Figure 2. Early mortality is defined as that occurring during the child's first hospital admission.

Thirty-two of the 72 patients died. Six of these had no surgical treatment and none survived without operation. Of the 26 deaths following operation, 19 were early and seven were late deaths.

Thirty-six operations were carried out before the age of 6 months with a combined mortality rate of 57%. The remaining 40 operations in older children had a mortality rate of 12.5%. The average mortality overall was 34%.

Figure 3 shows the relation between the preoperative haemoglobin concentration, age, and the outcome of surgical treatment. The mean haemoglobin concentration in each age group for survivors is shown, as is the mean for those who died.

The observation period of survivors after operation varied between six months and 13 years (mean 4.3 years).

Transposition of the great arteries was diagnosed in eight of 72 patients (11.5%). Not all had complete investigation. Angiocardiology evidence is the only proof of this anomaly and was not performed in every one.

TABLE I
AGE DISTRIBUTION AND OPERATIVE TECHNIQUE

Operation	Age (yrs)												Total
	6/52	3/12	6/12	1	2	3	4	5	6	7	8	9	
Left Blalock ..	5	5	4	3	6	1	2	3	2	—	—	1	32
Right Blalock ..	3	2	—	2	2	1	1	1	—	1	—	—	14
Left subclavian autograft ..	—	—	—	—	—	—	—	1	1	—	—	—	2
Potts ..	1	—	2	2	2	—	—	—	—	—	—	—	7
Waterston ..	1	—	1	—	—	—	—	—	—	—	—	—	2
Glenn ..	1	4	1	3	1	—	1	1	—	—	1	—	16
Thoracotomy ..	—	2	1	—	—	—	—	—	—	—	—	—	3
Total ..	11	13	12	10	11	2	4	6	3	1	2	1	76

FIG. 1. Four principal varieties of tricuspid atresia (Edwards and Burchell, 1949): type I, great arteries normally related; type II, great arteries transposed. (Reproduced, with slight modifications, from Cardiovascular Pathology by R. E. B. Hudson (1965, vol. 2, p. 1945, London: Edward Arnold) with the kind permission of the author.)

FIG. 2. Early and late mortality in relation to age at operation.

FIG. 3. Comparison of average haemoglobin concentration in survivors and non-survivors.

The only invariable radiological finding was the presence of oligæmic lung fields with the exception of one patient who had transposed great arteries and increased pulmonary flow. The electrocardiographic findings were of normal or left axis deviation with increased left ventricular activity in all but five patients; these had dextrocardia and in them a right axis was associated with predominance of the lateral ventricle. P pulmonale was present in 28 cases, left bundle-branch block in seven, and the Wolff-Parkinson-White syndrome in one.

NECROPSY FINDINGS

Necropsies were performed in 26 of the 32 children who died, and with one exception were performed at this hospital. The principal findings are recorded in Table II. Tricuspid stenosis of pin-hole type was present in four. These patients

TABLE II
POST-MORTEM FINDINGS IN 26 CASES

Post-mortem Findings	No.
Atrial septal defect	
Large	12
P.F.O.	9
Very small	5
Ventricular septal defect	
Present	19
Absent	7
Transposition of great arteries	3
Dextrocardia	3
Right ventricular outflow	
Obstructed	21
Normal	5
Ductus arteriosus	
Patent	12
Closed	13
Associated with preductile coarctation	1
Abnormal mitral valve	2

have not been excluded from the study, as clinically and functionally they are the same as those with atresia. Transposition of the great arteries was present in three (11.5%). Two of these had obstruction to pulmonary flow and one had a normal outflow.

Atrial septal defect was recorded as being large, small or very small. The small or very small defects were such as to obstruct the flow between the atria. This was found in five patients (19%).

SYSTEMIC ARTERY TO PULMONARY ARTERY ANASTOMOSIS

Fifty-five children had a total of 57 shunt operations of this type. One descending aorta to left pulmonary artery anastomosis was not performed at this hospital. Forty-three survived hospital treatment and 38 survived in the long term. The age distribution of the different techniques is presented in Table I and mortality rates in each age group are shown in Figure 4. Table III compares the mortality rates for operation before and after 6 months of age.

TABLE III

COMPARATIVE MORTALITY BEFORE AND AFTER 6 MONTHS OF AGE FOLLOWING SYSTEMIC ARTERY OR SUPERIOR VENA CAVA TO PULMONARY ARTERY SHUNTS

	Systemic Artery to Pulmonary Artery Shunt	Superior Vena Cava to Pulmonary Artery Shunt
Total % mortality (early and late)	31	43
Total % mortality (operation under 6 months)	62	44
Total % mortality (operation over 6 months)	3	42
% Mortality first admission	21	31

Ten of the 55 children had a second operation—a further systemic to pulmonary artery shunt in three, a Glenn procedure in five, and an exploratory thoracotomy in two. The outcome in these patients is shown in Table IV. The average

TABLE IV

RE-OPERATION AFTER SYSTEMIC ARTERY/PULMONARY ARTERY SHUNT

Re-operation	Survivors	Deaths	Average Length of Follow-up (yrs)
Systemic artery / pulmonary artery shunt	3	—	3
Glenn	3	2	4.5
Exploratory thoracotomy	—	2	—

FIG. 4. Early and late mortality following systemic artery to pulmonary artery shunts in relation to age at operation.

age at operation was 34 months and the average follow-up of survivors 4.7 years.

WATERSTON ANASTOMOSIS Two children had an ascending aorta to right pulmonary artery shunt (as described by Waterston, 1962). One child died on the first postoperative day having been moribund before surgical treatment.

POTTS ANASTOMOSIS There have been seven descending aorta to left pulmonary artery shunts (Potts) with four survivors. Of the three children who died, one developed intractable congestive cardiac failure and died at 4 months while two died in the first week, both with clinical evidence of shunt thrombosis. This was confirmed at necropsy in the child in whom this was performed.

BLALOCK-TAUSSIG ANASTOMOSIS Subclavian artery to pulmonary artery shunts have been performed on 48 occasions in 46 children, 14 on the right, 32 on the left, and two left subclavian autografts. Thirteen children died.

RESULTS

Congestive cardiac failure Six of the 38 survivors (16%) have developed congestive cardiac failure. One had a Waterston anastomosis and has been observed for five years. He has a mild degree of failure requiring continuous medical treatment. One child developed failure after a Potts anastomosis. The heart failure was relieved after a Glenn (superior vena cava to right pulmonary artery) shunt. Four patients have developed congestive cardiac failure following Blalock-Taussig shunts. The degree of failure is mild in three patients. One of these has had shunts on each side and both are functioning. One child has

moderate failure. In two further children observed for five years, there has been an abnormal increase in the transverse cardiac diameter with increased left ventricular activity assessed clinically and by electrocardiography, but without clinical evidence of heart failure.

Cerebral complications Cerebral complications in the follow-up period occurred in three patients. One developed a cerebral abscess at the age of 10, six years after a left Blalock anastomosis. One child, aged 14 months, developed a hemiplegia in the immediate postoperative period. Investigation revealed thrombosis of the left internal carotid artery. One child aged 14 months had slight mental changes, having suffered a cardiac arrest during operation.

FOLLOW-UP: SYSTEMIC ARTERY TO PULMONARY ARTERY ANASTOMOSIS Of the 43 children who survived the first operative procedure, there was no clinical improvement in seven, all of whom had a Blalock-Taussig anastomosis and therefore a further operation was performed. Two had a similar shunt on the opposite side, three a Glenn, and two a thoracotomy with failed attempts at further anastomosis. Two had improvement lasting a few months. One died 10 months after a Blalock shunt and necropsy revealed thrombosis of the anastomosis. One child had a Potts anastomosis at the age of 6 months performed elsewhere and required a right Blalock-Taussig shunt at the age of 7 years.

In four more of the long-term survivors, at periods between 5 and 11 years postoperatively, there is clinical deterioration.

Twenty-six of the original group who had a systemic artery to pulmonary artery shunt (47%) are free of complications. Most of the children

are still cyanosed with mild to moderate clubbing but they are well developed for their age and able to lead satisfactory lives.

CAUSE OF DEATH There have been 13 deaths following subclavian artery to pulmonary artery anastomosis, 12 in children under the age of 6 months at the time of operation and one over this age. The older child died two and a half years after operation from acute tracheobronchitis. Nine of the former group died during the first hospital admission and three were late deaths.

The cause of death in this group was:

Cardiac arrest during operation (2). Their preoperative condition was critical.

'Intact' atrial septum found at necropsy (2).
Bronchopneumonia (2) (? excessive shunt in 1).

Preoperative congestive cardiac failure which was increased after operation (3).

The three late deaths in the young age group were due to shunt thrombosis (1), bronchopneumonia (1), and multiple cardiac and visceral anomalies (1).

SUPERIOR VENA CAVA TO RIGHT PULMONARY ARTERY ANASTOMOSIS (GLENN)

Systemic vein to pulmonary artery anastomosis was first attempted in this hospital in 1961. This operation was described by Carlon (Carlon, Mondini, and De Marchi, 1951), Glenn and Patiño (1954), and Bakulev (Bakulev and Kolesnikov, 1959). The technique used was that of Glenn (1958).

There have been 16 operations. The age distribution and mortality are shown in Tables I and III and in Figure 5. Four of the children had had a previous systemic artery to pulmonary artery anastomosis (3 Blalock; 1 Potts). The average age at operation was 15.5 months and the average follow-up of survivors 3.3 years.

From 1961 to 1965 the Glenn procedure was the operation of choice. This depended on the right pulmonary artery being at least half the

diameter of the superior vena cava. A small pulmonary artery precluded the Glenn procedure at thoracotomy in five of 21 cases. Complete occlusion of the superior vena cava below the anastomosis was attempted in all patients. In two, partial occlusion only was performed because when the superior vena cava was occluded the pressure in it rose above 30 mm. Hg. One of these children survived and angiography shows that most of the caval blood is passing to the atrium and not to the pulmonary artery. The azygos vein was divided in all but two cases. Delayed ligation was not attempted. In one of the two undivided cases an angiogram demonstrates the 'azygos steal' picture in which contrast bypasses the right lung shunting via the azygos system to the inferior vena cava.

POSTOPERATIVE COURSE The nine survivors have been observed from six months to seven years. Two children died after leaving hospital, one five weeks after discharge, and one eight months later. The first child had extensive venous thrombosis in the head and neck which was thought to have originated in the left arm, the Glenn anastomosis being patent. The second had bilateral bronchopneumonia.

Facial congestion occurred after operation in five of the nine survivors but in only one has this persisted. There have been no late complications. Three of the nine cases are assessed as very good results and five as good. One child is only slightly improved and this is the child with partial superior vena cava occlusion.

CAUSE OF DEATH DURING FIRST HOSPITAL ADMISSION Five children died during or soon after operation. One 7-year-old had congestive cardiac failure before operation and developed a cardiac arrest when the pulmonary artery was occluded. All the other children had marked congestion of the head and neck after operation and cerebral congestion at necropsy. One had encephalomalacia and one, who had a cardiac arrest during operation, had extensive cerebral venous sinus thrombosis.

FIG. 5. Early and late mortality following superior vena cava to pulmonary artery shunts in relation to age at operation.

ATRIAL SEPTOSTOMY Atrial septostomy using the Rashkind/Miller technique (Rashkind and Miller, 1966) has been carried out on one child in this series. This procedure was of benefit and the experience has prompted us to review our catheterization data and necropsy results.

In the years 1964 to 1967, of 15 children fully investigated, three (20%) have had manometrically distinct atria with the right atrial pressure at a higher level, the differences being 2 mm., 2 mm., and 3 mm. respectively. Of these three, one, aged 2 weeks, had a balloon septostomy; one has done well after a Blalock-Taussig anastomosis; and one died after a Glenn procedure. In this last patient a patent foramen ovale was present at necropsy.

Five examples of very small atrial septal defect were discovered in 26 post-mortem studies. None of these patients had a cardiac catheterization before death. The ages at death varied between 3 weeks and 18 months.

DISCUSSION

Abbott (1936) found 21 cases of tricuspid atresia in 1,000 necropsies in congenital cardiac cases. There were 16 cases diagnosed clinically in 1,000 cases of congenital heart disease. Wood (1956) assessed the incidence of tricuspid atresia as 1.5% of congenital cardiac cases. Campbell (1961) reported an incidence of 5% of cyanotic congenital heart disease.

The results of palliative surgery, particularly in children over the age of 6 months, are most encouraging.

The basic anomaly is a deficient pulmonary circulation with a small volume of oxygenated blood returning to the left atrium with the exception of tricuspid atresia associated with transposed great arteries and normal or excessive pulmonary blood flow (Figure 1). An atrial septal defect is essential for survival.

In most cases the aim of surgical treatment is to increase pulmonary blood flow. This is achieved by systemic artery to pulmonary artery anastomosis or by superior vena cava to pulmonary artery anastomosis.

Robicsek, Sanger, Taylor, Najib, and Tavana (1963) have emphasized the theoretical advantage of the caval shunt. Tissue oxygenation is improved without an increase in left ventricular output, the improvement depending on a change in the ratio of oxygenated and non-oxygenated blood within the heart. The systemic artery shunt, on the other hand, increases tissue oxygenation by adding to left

ventricular output and work. Edwards and Barger (1968) report the Glenn procedure to be the operation of choice, quoting a mortality rate of 8.6% in their last 23 cases.

In the years 1961 to 1965, in this series, the selection factor was operating in favour of the Glenn shunt, as the systemic artery anastomosis was reserved for those cases in which the size of the pulmonary artery precluded the caval anastomosis.

Despite these arguments, however, our experience shows at least comparable results with the systemic artery shunt. No child over the age of 1 year has died following this type of procedure whereas there have been two deaths after caval shunts. We accept that the number of these latter shunts in older children is small, and that a figure for mortality rate is apt to be misleading, but we consider that the results with systemic artery shunts are as good as any alternative method can provide.

We have not found a significant incidence of late heart failure, increasing cardiac size or left ventricular activity in cases with systemic arterial shunts. Our findings may to some extent reflect the experience of this type of shunt in this unit, nearly 600 such procedures having been performed since 1953.

The Glenn procedure has not always been possible in view of a discrepancy in size between the pulmonary artery and the superior vena cava. We have, however, been disappointed by the degree of venous congestion even with adequate vessel size and without a gradient across the anastomosis. Azygos vein ligation is important (Robicsek, Sanger, Taylor, and Najib, 1963). It may be that delayed ligation (Edwards and Barger, 1968) is a valuable technique. These authors have also suggested that postoperatively a diminished blood volume can be fatal. Three of our cases had the clinical picture which could be similar to that which they described following an apparently adequate shunt.

In the older age group it may well be that, given adequate anatomy, a modification of technique and management will be attended by an improvement in results using the Glenn procedure. The survivors in this series have not developed late complications and have with one exception done well clinically. It is in terms of the early post-operative mortality that the technique may compare unfavourably with the systemic arterial shunt.

The problems in infancy are greater and the results similar with each technical approach. In addition to the small size of the pulmonary artery the pulmonary vascular resistance is higher and

infants present early because the haemodynamic abnormality is more serious.

It may be that development of the pulmonary arterial tree is more adequate if subjected to pulsatile flow. The higher resistance to flow makes the low pressure caval anastomosis less likely to function. We have, however, one very good result after a Glenn anastomosis performed at 2 months. A systemic arterial shunt to be followed later by a Glenn procedure may be the approach of choice (Glenn, Ordway, Talner, and Call, 1965; Hallman, 1968).

Adequate cardiac investigation, including catheterization and angiocardiology, is mandatory. Atrial septostomy (Rashkind and Miller, 1966) is indicated if the right atrial pressure exceeds that in the left atrium. Farnsworth, Ehlers, Levin, Ho, and Engle (1967) report obstructive atrial communication in half of 30 cases. Keith *et al.* (1967) reviewed the literature and quote a 33% incidence of significant obstruction. Our incidence is 20% in 15 consecutive cases.

Rashkind, Friedman, Waldhausen, and Miller (1967) described four ill infants in whom atrial septostomy was performed as a prelude to systemic arterial shunt. They emphasized that a 2 to 3 mm. Hg gradient is significant. This accords with our experience. Closed septostomy is confined to infants in the first six months of life.

Out of 92 cases of tricuspid atresia reviewed in the literature, 20 had transposition of the great arteries and a normal pulmonary outflow (Keith *et al.*, 1967). In this situation pulmonary blood flow is increased.

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ADDENDUM

We have recently treated a baby with tricuspid atresia, transposition of the great arteries, and increased pulmonary blood flow. A balloon septostomy was necessary and banding of the pulmonary artery was then performed. The child, aged 3 weeks, has done well.