Palliative Surgery in Tricuspid Atresia

Forty-two Cases


ONLY one in three children with tricuspid atresia survives the first year of life. The structural abnormalities in this condition have so far precluded all but palliative surgery.

This communication discusses 42 children diagnosed as having tricuspid atresia, in each of whom at least one palliative operation was performed.

We define tricuspid atresia as atresia of the atrioventricular valve corresponding to the systemic venous atrium.

The palliative operations are (1) systemic artery–pulmonary artery anastomosis. (Blalock-Taussig anastomosis, Potts anastomosis and their variants.) (2) Systemic vein–pulmonary artery anastomosis.2-4

The purpose of these operations is to increase oxygenation of blood in the systemic arteries by shunting systemic blood through the lungs.

Clinical Material

Diagnosis

Each of these cases satisfies all or most of the following criteria for tricuspid atresia with diminished pulmonary blood flow:
1. Central cyanosis.
2. Electrocardiogram: (a) normal axis or left axis deviation, (b) preponderance of left ventricular activity over right ventricular activity.
3. X-ray: (a) diminished pulmonary vascularity, (b) heart size normal, except in six cases with heart failure, when the heart was enlarged.
4. Angiocardiogram: (a) opacification sequence: RA→LA→LV→Aorta
   ↓     ↓
   RV→PA, (b) RA–LV notch,
   (c) Pulmonary artery small or not seen.


Clinical Features

The characteristic signs were central cyanosis from birth, poor weight gain, and effort intolerance, usually shown in infants by slowness in feeding. (By “infant” we mean “child less than 1 year of age.”) Twenty-four had spontaneous cyanotic attacks and six had heart failure.

Indications for Surgery

The following indications were used, regardless of weight or age: heart failure, very poor weight gain, persistent cyanotic attacks, effort intolerance worse than grade 1. At least one indication was present in each case.

Surgery

Between 1955 and 1961 only Blalock-Taussig anastomosis, Potts anastomosis or one of their variants was done in this series. Since 1961 Glenn’s operation has been attempted in cases in which the preoperative angiocardiogram suggested that it was practicable. Forty-two children had 46 operations: 30 Blalock-Taussig anastomoses, five Potts anastomoses, one aorta-right pulmonary artery anastomosis, and 10 Glenn operations.

When the subclavian artery was too short for Blalock-Taussig anastomosis, the main subclavian trunk was resected and used as an autograft between the descending aorta and the left pulmonary artery. In this series, such cases are classified as Blalock-Taussig anastomoses.

The technic of systemic vein (superior vena cava)-pulmonary artery anastomosis was that described by Glenn.2 When the diameter of the right pulmonary artery was less than half that of the superior vena cava measured at the site where they crossed, the Glenn operation was not attempted and instead an arterial shunt was done.

During Glenn’s operation, when the end of right pulmonary artery to side of superior vena cava anastomosis had been done, pressures were measured in the superior vena cava and the right pulmonary artery. The effect on these pressures of complete occlusion of the superior vena cava below the anastomosis was then observed. If the superior vena caval pressure did not rise above 30 mm. Hg the shunt was considered satisfactory. There was never a gradient greater than 5
mm. Hg between superior vena cava and the right pulmonary artery.

In two cases the superior vena caval pressure rose to 35 mm. Hg; for this reason the superior vena cava was left only partially occluded. In this series, these cases are classified as Glenn operations. Although both children survived the operation one of them died later with a severe respiratory infection and the other has not apparently benefited from the operation. He is awaiting second operation. An angiocardiogram on this child shows that although some contrast medium does go into the right lung, most of it bypasses the lung and enters the right atrium.

In two other cases, also classified as Glenn operations, the azygos vein was left open to vent the superior vena cava. One of the two died soon after operation. In the other, an angiocardiogram shows that much of the contrast medium bypasses the right lung and enters the inferior vena cava via the azygos vein. The importance of this azygos "steal" has been emphasized by Robicsek et al.5

Second operations. Four children underwent second operations. In three, the initial Blalock anastomosis had not been followed by clinical improvement. Of these three, for the second procedure, one had a contralateral Blalock anastomosis and the other two had Glenn operations. The fourth patient developed heart failure following Potts anastomosis. Glenn's operation was then done in addition to the Potts anastomosis, this was followed by regression of heart failure.

Results

By operative death we mean death during operation, or death while still in hospital following operation. By late death we mean death occurring after discharge from the hospital.

Whatever operation was done, no infant who weighed less than 10 lb. at the time of operation survived; there are eight in this group.

Above the weight of 15 lb. whatever operation was done, there were no operative deaths, but there was one late death; there are 21 in this group.

Above the age of 1 year, there have been no deaths (follow-up period 1 to 9 years); there are 17 in this group.

On children who weighed between 10 and 15 lb. there were 17 operations. Among the 12 arterial shunt operations, there were four operative and three late deaths. Two of these operative deaths were in children just exceeding 10 lb. in weight, but who had heart failure. Among the five Glenn operations there were no operative deaths and one late death.

In the arterial shunt group as a whole, death was in association with heart failure, infection and, in four cases, including one with Potts anastomosis, occlusion of anastomosis by thrombus.

Following Glenn's operation all cases had some degree of superior vena caval obstruction, usually temporary, but two operative deaths were associated with a marked degree of obstruction and brain damage.

Postoperative complications are listed in table 1. Of the 25 children who survived, 23, who underwent a total of 26 operations, have been followed; the follow-up period was from 1 to 9 years. A good result is one with improved weight gain, grade-1 effort intolerance, diminution in cyanosis, and cessation of any cyanotic attacks. A moderate result is one with distinct postoperative improvement but still with effort tolerance not better than grade 2. A poor result is one with no improvement. Thirteen arterial shunts have a good result, four a moderate and four a poor result. Four Glenn operations have a good result and one a poor result.

Postmortem Examination

Postmortem examinations were made in 15 of the 17 dead children. In addition to tricuspid atresia with atrial septal defect, all of these had a ventricular septal defect and some form of obstruction to pulmonary flow, i.e., subvalvular stenosis, pulmonary valve stenosis, pulmonary valve atresia, hypoplastic or atretic main pulmonary artery. Only one atrial septal defect was small. There were four cases with a distinct patent ductus arteriosus and three in whom the duct was only probe patent.

Discussion

At present corrective surgery is not available for tricuspid atresia. Various palliative procedures have been suggested. They include (1) systemic artery-pulmonary artery anastomosis (Blalock-Taussig and Potts); (2) systemic vein (superior vena cava)-pulmonary artery anastomosis;2 (3) enlargement
of small atrial septal defect in addition to shunt operation;⁶ (4) enlargement of small ventricular septal defect.⁷

Procedure 3 is a technical possibility but would seem to have been appropriate in only one child in our postmortem series.

Enlarging a small ventricular septal defect as suggested by Brock would be appropriate if there was also an adequate channel from the right ventricle to the lungs. In our postmortem series no such cases were found. The size of the right ventricle and the channel from it to the lungs is related to flow. In cases of tricuspid atresia with a small ventricular septal defect extensive remodeling of the right ventricle and pulmonary artery would be required to increase significantly the flow through them after enlargement of the ventricular septal defect.

In this series only systemic artery-pulmonary-artery and systemic vein (superior vena cava)-pulmonary artery shunts were done. Robicsek et al.⁸ have put forward theoretical reasons for expecting Glenn’s operation to be superior to arterial shunts. Wijffels et al.⁹ have brought forward further theoretical and experimental considerations supporting the same view. We have found no published clinical results confirming that expectation.

In the 10- to 15-lb. group of 17 operations, four operative and three late deaths were associated with 12 arterial shunt operations, whereas no operative deaths and only one late death was associated with five Glenn operations. The numbers are too few to draw conclusions about the relative merits of these shunt operations. Furthermore, Glenn’s operation was done only on the more favorable cases, i.e., only those with a sizable right pulmonary artery.

Table 1 shows a high incidence of heart failure developing after arterial shunt operations, but none after Glenn operations. This is offset by the occurrence of superior vena caval obstruction in every case of Glenn’s operation, leading in two cases to death with brain damage.

The relative value of Glenn’s operation and arterial shunts could be accurately assessed only by a controlled trial.

The weight of 10 lb., below which there were no survivors, is put forward only as a prognostic guide. It is not suggested than an operation, otherwise indicated, should not be done because the infant weighs less than 10 lb. However, we recognize that our indications may need revision for this group. Two operative deaths in the 10- to 15-lb. group would probably come into this group if allowance were made for the weight of the excess fluid of heart failure.

The single case in which heart failure followed a Potts anastomosis and regressed after Glenn’s operation is of interest. The increase in cardiac output associated with this particular Potts anastomosis may have caused the heart failure. By doing Glenn’s operation, probably the same perfusion of body and lungs was obtained with a lower cardiac output. This cardiac output was maintained without heart failure.

Summary

Forty-two children diagnosed as having tricuspid atresia with diminished pulmonary

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Table 1

Palliative Surgery in Tricuspid Atresia: Postoperative Complications*

<table>
<thead>
<tr>
<th>Complications</th>
<th>Arterial shunt</th>
<th>Glenn operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart failure</td>
<td>9†</td>
<td>0</td>
</tr>
<tr>
<td>Superior vena caval obstruction</td>
<td>0</td>
<td>10†</td>
</tr>
<tr>
<td>Respiratory infection</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Shunt failure</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>Cerebral</td>
<td>3 (1 late)</td>
<td>2</td>
</tr>
</tbody>
</table>

*Arterial shunt: 32 children had 36 operations, 18 with complications. Glenn operation: 10 children had 10 operations.
†Usually temporary.
blood flow are discussed. Palliative shunt surgery was done in each of these children. None of the eight infants who weighed less than 10 lb. at the time of operation survived. In 17 operations on infants who weighed between 10 and 15 lb. there were four operative and four late deaths. Glenn’s operation is practicable and has been found relatively satisfactory in this group. In 21 operations on children who weighed above 15 lb. there were no operative deaths and one late death. The 17 children aged over 1 year at the time of operation are alive. In one child heart failure followed a Potts anastomosis, but regressed after Glenn’s operation.

References

Padua

The school of Padua was the most celebrated in Europe. There, in the sixteenth century, two anatomists, Vesalius and Realdus Columbus, contributed further to the elaboration of the concept of the passage of blood through the lungs.—ANDRÉ COURNAND, M.D. Circulation of the Blood. Edited by Alfred P. Fishman, M.D., and Dickinson W. Richards, M.D. New York, Oxford University Press, 1964, p. 23.
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