

Superior vena cava to right pulmonary artery anastomosis

Results in 46 infants and children

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Superior vena cava to right pulmonary artery anastomoses have been constructed in 46 infants and children with cyanotic congenital heart disease associated with low pulmonary blood flow and low pulmonary vascular resistance. All children had lesions not currently amenable to corrective operative procedures. Good or excellent results, albeit with persistent mild to moderate cyanosis, followed operation in 31 children and have been maintained for up to eight years. There were 15 hospital and two late deaths. Although there has been a significant absence of late complications, it is probable that the caval anastomosis results in a fixed level of pulmonary blood flow and thus fixed effort tolerance. Thus, when puberty with its demands for increased physical activity occurs, these patients may require further surgery, such as a systemic to pulmonary artery anastomosis to increase pulmonary blood flow.

Inadequate pulmonary blood flow in the presence of normal or low pulmonary vascular resistance occurs with a variety of congenital cardiac lesions.

Cava-pulmonary circulatory bypass of the right heart, first suggested by Carlon, Mondini, and DeMarchi (1951) and subsequently developed by Glenn and Patiño (1954), Glenn (1958), and Bakulev and Kolesnikov (1959), has become an accepted palliative procedure, mainly of use in patients with tricuspid atresia. Robicsek, Sanger, Taylor, and Najib (1963) pointed out that a cava-pulmonary shunt increases systemic arterial oxygen saturation without increasing the volume load on the systemic ventricle, but though this is likely in conditions such as tricuspid atresia, there are no data regarding the circulatory adjustments following this operation.

This paper reports the results of cava-pulmonary shunts in 46 infants and children.

MATERIAL AND METHODS

The cava-pulmonary shunt was used in a series of 46 infants and children between 1960 and 1969. This operation was used as the procedure of choice in

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patients with basically incorrectable cardiac lesions with inadequate pulmonary blood flow. During this period some infants in whom investigation showed the pulmonary arteries to be very small were treated by systemic artery to pulmonary artery anastomosis.

The operative technique employed was that of Glenn (1958) and consisted of an end-to-side anastomosis of the right pulmonary artery to the superior vena cava followed by ligation of the superior vena cava at its junction with the right atrium. Interrupted 6/0 sutures were used for the anastomosis. Up to 1963, azygos vein ligation at the time of the caval anastomosis was used, but since then in selected patients the technique of delayed azygos ligation (Edwards and Bargeron, 1963) has been employed. In this technique, a ligature is passed twice around the azygos vein and the ends are brought out to a subcutaneous position using a separate stab incision. Six to eight days later, the ends are isolated and gentle retraction is applied to close off the azygos vein. The ligature is then tied.

Post-operatively, patients were nursed in a sitting position. Particular emphasis, since 1965, has been placed on the maintenance of an adequate right atrial pressure. Infusions of isotonic fluids were used to maintain right atrial pressure at approximately 10 cm. of water. This often required large volumes of fluid, e.g., 300 ml. of Ringer's lactate in the first two post-operative hours in a 5.4 kg. infant.

TABLE

DIAGNOSIS, AGE AT OPERATION, AND MORTALITY IN 46 INFANTS AND CHILDREN WITH CAVA-PULMONARY ANASTOMOSIS

Diagnosis	No. of Cases	No. Dead	Age							
			Under 1 Month		1-6 Months		6-24 Months		Over 2 Years	
			No.	Dead	No.	Dead	No.	Dead	No.	Dead
Tricuspid atresia	25	9	2	2	8	3	10	2	5	2
Common ventricle with pulmonary stenosis	6	1	-	-	2	-	1	-	3	1
Transposition with pulmonary stenosis	5	1	1	1	1	-	2	-	1	-
Hypoplastic right ventricle with pulmonary stenosis	3	1	-	-	1	1	1	-	1	-
Pulmonary atresia with ventricular septal defect	2	2	1	1	-	-	1	1	-	-
Common ventricle with pulmonary atresia	2	1	-	-	1	1	1	-	-	-
Tetralogy of Fallot	1	1	-	-	-	-	-	-	1	-
Pulmonary atresia with hypoplastic right ventricle	1	1	1	1	-	-	-	-	-	-
Situs inversus, laevocardia, pulmonary stenosis, and ventricular septal defect	1	-	-	-	-	-	-	-	1	-
Total	46	17	5	5	13	5	16	3	12	4

RESULTS

Forty-six infants and children, aged 1 day to 12 years, had surgical treatment. Twenty-six were girls and 20 boys.

In infants, increasing cyanosis or repeated cyanotic spells were indications for operation. These children were severely ill and some were moribund. Older children, usually of pre-school age, were operated on for similar reasons or because increased effective pulmonary blood flow was necessary to permit more normal school activities.

The diagnosis, age at operation, and mortality are shown in the Table. Diagnosis was made by cardiac catheterization and angiocardiography in all patients. Thirty-one children were discharged from hospital and have been observed for periods ranging from one month to eight years (Fig. 1).

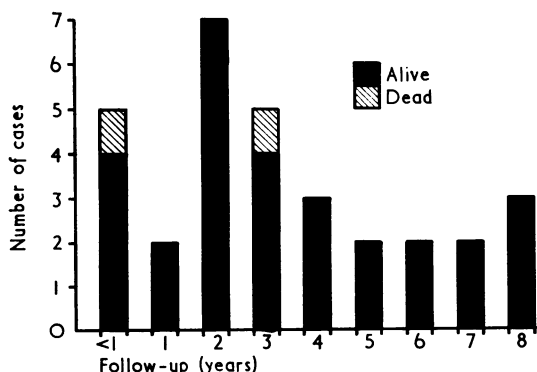


FIG. 1. Follow-up after hospital discharge in 31 infants and children with cava-pulmonary anastomosis.

MORTALITY Of the 15 hospital deaths, 14 occurred within three days of operation and one one month after operation. There were two deaths following discharge from hospital, one one month after operation and one three and a half years after operation. A necropsy was performed in 15 of the 17 non-survivors.

All 5 patients under 1 month of age died. Between 1 and 6 months of age, 8 out of 13 patients survived. Over 6 months, 23 out of 28 patients survived to leave the hospital, but 2 of these died later.

Of the 10 deaths in infants less than 6 months of age, 5 were due to respiratory complications and 3 resulted from a small right pulmonary artery being unable to accept superior vena caval blood flow without serious elevation of caval venous pressure or were related to technical failure to obtain an adequate anastomosis. One patient (aged 5 months), who had transposition of the great arteries, ventricular septal defect, and subvalvular pulmonary stenosis, had an atrial septal shift procedure at the same time as the caval anastomosis. One patient, despite an apparently satisfactory anastomosis, failed to improve and necropsy revealed that a left superior vena cava was present, thus preventing adequate flow to the right lung by permitting systemic venous drainage via the coronary sinus to the right atrium.

Over 6 months of age only one of seven deaths was due to respiratory complications. One patient, having made initial satisfactory progress, died suddenly one month after operation. No necropsy was performed. One patient died during reoperation for chylothorax. One patient with transposition of the great arteries and a single ventricle died three and a half years after operation. After

an initial improvement she developed tricuspid valvar insufficiency and did not survive a second operation directed at alleviating this insufficiency. One patient died from haemorrhage during operation. There was one anaesthetic death due to a misplaced endotracheal tube. The final death occurred in an infant in whom a Potts' anastomosis was initially attempted. This failed to function, and a cava-pulmonary anastomosis was performed with the child in a moribund condition 20 hours later, but the child died.

COMPLICATIONS Chylothorax occurred on six occasions. The effusion was right-sided in four patients and bilateral in two. Each patient was treated initially by repeated chest aspirations, but operative intervention was finally required in all six patients. Three patients required one operation, two patients required two operations, and in one child three attempts to control the chyle leak were necessary. In this patient, cardiac arrest and death occurred during the third procedure.

Other major, but non-fatal, post-operative complications were atelectasis of the left lung (one patient), respiratory insufficiency requiring prolonged assisted ventilation (one patient), tracheal stenosis (one patient), and a right hemiparesis (one patient). Marked improvement occurred in this latter patient during the subsequent two months.

Acute superior vena caval obstruction syndrome occurred during operation in one child and was associated with a marked fall in cardiac output. Loosening of the superior vena caval ligature was necessary and this was followed by a satisfactory post-operative course.

One patient, who initially did well, developed increasing cyanosis and effort intolerance five years after operation. Venous angiocardigraphy (which must be performed from the left arm) showed prominent collaterals between the superior vena cava and the coronary sinus. These collaterals were tied at a second operation and the child has again done well. Figure 2 shows the angiograms of a similar patient who had been operated on initially at another hospital and was therefore not included in our series.

FOLLOW-UP Thirty-one patients were discharged from hospital. There were two late deaths.

The results were assessed as excellent if the child could participate in all but the most stren-

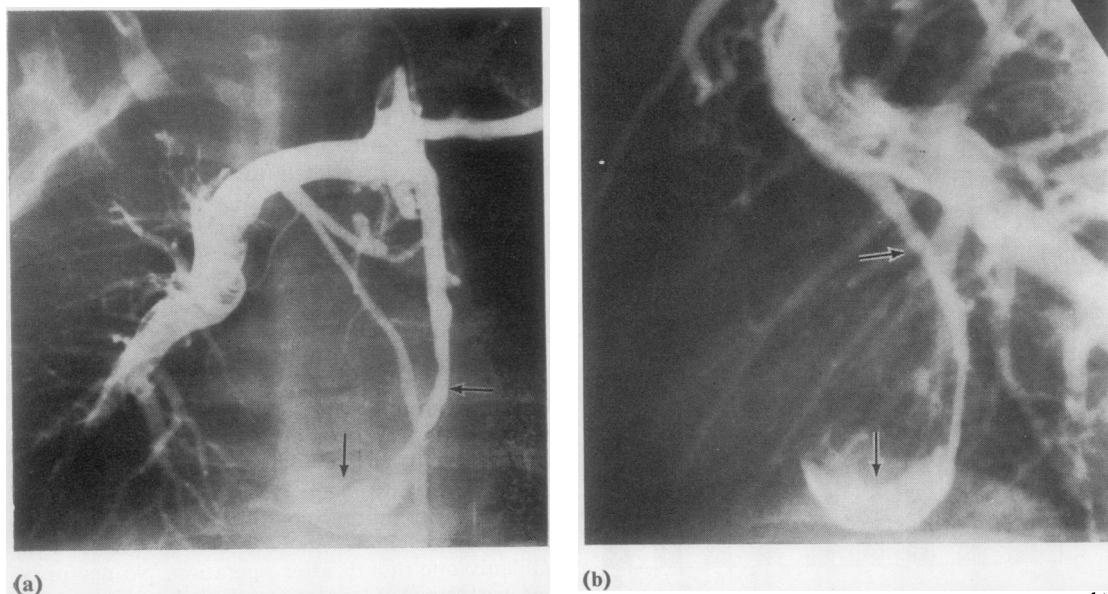


FIG. 2. Antero-posterior (a) and lateral (b) venous angiograms following left subclavian vein injection in a child with tricuspid atresia who became dyspnoeic six years after a cava-pulmonary anastomosis. The right pulmonary artery is well filled through the anastomosis. A venous collateral (upper arrow) is seen passing from the innominate vein to the coronary sinus (lower arrow). An internal mammary vein passes up anteriorly.

uous childhood activities appropriate to his age, and good if moderate effort restriction was present.

Mild to moderate cyanosis has persisted in all survivors.

Seven (22%) survivors had an initial excellent result and five have maintained this outcome. The two exceptions were the child who developed venous collaterals and the child with tricuspid valvar insufficiency.

Twenty-four (78%) have a good result, maintained in all except the child who died unexpectedly after hospital discharge.

Although these patients have maintained their initial improvement, it is our impression that they are more limited at around the time of puberty, when demands for increased effort tolerance occur, as compared to a similar group of children with well-functioning Blalock-Taussig anastomoses. Children with the caval anastomosis seem to have a relatively fixed level of pulmonary blood flow and therefore a fixed level of effort tolerance and have little or no reserve to permit a sustained increase in the level of activity.

To our knowledge there has been no late obstruction of the anastomosis. Further palliative procedures have not, as yet, been performed in these children.

DISCUSSION

Corrective surgery is not currently available for tricuspid atresia, pulmonary atresia with intact ventricular septum or common ventricle with pulmonary stenosis. These lesions are usually characterized by reduced pulmonary blood flow with a normal or low pulmonary vascular resistance. Palliative operations to increase pulmonary blood flow and systemic arterial oxygen saturation and thus alleviate symptoms are beneficial. Pulmonary blood flow may be augmented by anastomosis of either a systemic artery or systemic vein to the pulmonary artery and we have, in 46 cases, elected to use the caval anastomosis (Glenn, 1958), usually with modification of delayed azygos vein ligation (Edwards and Barger, 1963).

It is our practice to measure the pressure in the superior vena cava immediately after the anastomosis is completed and before the chest is closed. If the pressure is below 200 mm. of water, as occurs when the pulmonary artery and superior vena cava are about the same size, then the azygos vein can be ligated. If caval pressure is between 200 and 300 mm. of water, then delayed azygos vein ligation is advisable. If caval pressure is above 300 mm. of water, the azygos vein should be left

open and the caval ligature loosened enough to allow the caval pressure to drop below 250 mm. of water. One of our patients, who had become oedematous and hypotensive shortly after the chest was closed, was reoperated on immediately and found to have a caval pressure of 400 mm. of water. The caval ligature was loosened until the pressure fell and the child has since done well. Prior to our use of delayed azygos vein ligation there was a 35% incidence of superior vena cava obstruction syndrome, but since then this complication has been seen in only 2 of 28 patients (7%). Fluid sequestration in the obstructed venous drainage area may cause a fall in effective circulating blood volume with inadequate filling pressure in the heart. This was emphasized by Barger and Edwards (1965), who advocated measuring right atrial pressure post-operatively and the infusion of sufficient isotonic fluid to maintain an adequate central venous filling pressure. Glenn, Browne, and Whittemore (1966), in a collected series of 537 cava-pulmonary operations, reported a 5.6% incidence of mild and an 11.7% incidence of severe superior vena caval obstruction. The timing of azygos vein ligation was not mentioned but was presumably at the time of the anastomosis. Two patients in our series died after an attempted shunt into a pulmonary artery less than half the size of the superior vena cava despite adequate fluid administration and an open azygos vein. Both these patients had the superior vena caval obstruction syndrome. Late obstruction of the superior vena cava, reported by Boruchow, Bartley, Elliott, and Schiebler (1969), has not been seen in our patients. Glenn, Browne, and Whittemore (1966) considered that a small pulmonary artery and/or raised pulmonary vascular resistance precluded the venous anastomosis. Most authors believe the pulmonary artery should be at least half the diameter of the superior vena cava (Glenn *et al.*, 1966; Deverall, Lincoln, Aberdeen, Bonham-Carter, and Waterston, 1969).

The overall mortality in our series is 37%, being similar to the figures quoted by Glenn *et al.* (1966) and Young and Flemma (1966)—37% and 30% respectively. The relation between age and mortality is clear regardless of the underlying cardiac lesion. With tricuspid atresia comparative figures in the series of Glenn *et al.* (1966) and in this series are very similar with a very high mortality in young infants.

We believe that, in the first two months of life, systemic artery to pulmonary artery anastomosis offers the best chances for survival (Glenn *et al.*, 1966; Hallman, 1968; Deverall *et al.*, 1969). The

Waterston shunt is the procedure of choice at this time (Waterston, 1962). Series of cases have been reported in which systemic artery to pulmonary artery anastomosis has been the procedure of choice (Hallman, 1968 ; Deverall *et al.*, 1969 ; Paul, Greenwood, Cole, and Muster, 1969) and mortality statistics comparable to those reported herein have been reported. However, with the exception of the infant group, comparable mortality, and particularly an absence of significant late complications, leads us to believe that the cava-pulmonary anastomosis is preferable in the pre-pubertal years if technically possible.

All the survivors of the cava-pulmonary shunt in our series had an initial good or excellent result. Although there were two late deaths, the majority of patients have no late complications. Shunt thrombosis has not occurred and there does not seem, with the technique used, to be any stenosis of the anastomosis, as sometimes occurs with systemic arterial anastomosis. All children are still mildly to moderately cyanosed and all have some limitation of exercise tolerance.

Chylothorax has been a serious post-operative complication and has not responded to repeated chest aspirations. The difficulty in achieving control of a chyle leak at reoperation is evidenced by the number of reoperations required and is probably due to the leak being of a generalized nature, perhaps accentuated by elevated superior vena cava pressure rather than a local thoracic duct laceration. In a number of patients, efforts were made to identify the thoracic duct as it passed through the diaphragm, but they were not successful. In several cases the tissues behind the oesophagus and to the right of the aorta were ligated at the time of the cava-pulmonary anastomosis in the hope of obstructing the thoracic duct and thereby preventing the diffuse leak of chyle. There was no chylothorax in the patients in whom this was done.

Respiratory complications in infancy have been an important cause of post-operative morbidity and mortality. A similar experience with other cardiac lesions treated surgically in infancy has been reported. Intensive respiratory management has resulted in better results in such series (Downes, Nicodemus, Pierce, and Waldhausen, 1970).

The development of marked effort intolerance or increasing cyanosis after an initially satisfactory result is uncommon. When present these features indicate a progressive reduction in pulmonary blood flow. Apart from occlusion of the anastomosis and/or pulmonary arterial thrombosis, the

development of venous collaterals or the development of the azygos steal picture (Robicsek *et al.*, 1963) should be considered. We are not aware that this has occurred in our patients. Boruchow *et al.* (1969) have suggested that a reduction in left lung blood flow might result from a relative increase in intracardiac obstruction to pulmonary blood flow as a result of disproportionate growth in the heart and this might initiate a cycle of events resulting in decreased pulmonary blood flow.

Observation of these patients over the years has resulted in certain conclusions, admittedly largely subjective, regarding the proper use of the cava-pulmonary anastomosis. It is our impression that growth of the anastomosis occurs, but, despite patency, the symptomatic palliation seems to be relatively fixed and does not permit as much exercise tolerance in the individual patient as does an optimally functioning Blalock-Taussig shunt. This may reflect the capacity of the latter shunt to permit an increase in pulmonary blood flow at times of increased demand. Such an increase does not seem to occur with the venous shunt.

If, as puberty approaches with its demands for increased activity, the pulmonary blood flow is not adequate to permit reasonably normal activity, a Blalock-Taussig shunt can be performed on the opposite side. By this age, this anastomosis can be constructed with every expectation of its remaining patent for many years.

At this time our practice is to advise the cava-pulmonary anastomosis as the treatment of choice in children between 1 month and 10 years of age. Younger infants, often in urgent need of palliation, will usually be more suitably treated by arterial anastomosis. Children older than 10 years, in whom the size of the subclavian artery makes the construction of a reliable Blalock-Taussig shunt more likely, will probably achieve better palliation with this type of shunt.

Full cardiac catheterization and angiocardiography are essential in the investigation of the infant with serious cardiac disease, and it is important to search for the left superior vena cava if the possibility of a palliative cava-pulmonary anastomosis exists. If tricuspid atresia is found at the initial study, the presence of an adequate atrial septal defect must be assured at that time. If the defect is inadequate, an immediate balloon septostomy should be performed in infants or the defect enlarged at the time of palliative surgery in older children.

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