Truncus Arteriosus with Unilateral Absence of a Pulmonary Artery

Criteria for Operability and Surgical Results

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SUMMARY  In 15 of 126 (12%) patients with truncus arteriosus who were catheterized at the Mayo Clinic from 1967 through 1975, natural agenesis of one pulmonary artery was present. Ten truncus patients with either natural or acquired absence of one pulmonary artery have undergone definitive operation. The criteria for operability, based on a calculation of pulmonary resistance, are different in patients with single pulmonary artery than in patients with two pulmonary arteries. Study revealed that, if the calculated pulmonary resistance in the patient with single pulmonary artery is halved, this new value provides a more reliable assessment of the reactivity of the pulmonary arteriolar bed than does the total calculated pulmonary resistance value. Follow-up information suggests that the patient with single pulmonary artery may be at potentially high risk of continued progression of pulmonary vascular disease after surgical correction, possibly because of the postoperative obligatory increased flow through the single pulmonary arteriolar bed as a result of the entire cardiac output passing through it. Surgical correction of truncus arteriosus during infancy, before significant pulmonary vascular disease has developed, appears to be particularly desirable in this subgroup of patients with single pulmonary artery.

TETRALOGY OF FALLOT and persistent truncus arteriosus are the two congenital cardiovascular malformations most often associated with complete agenesis of one pulmonary artery. In tetralogy of Fallot, the pulmonary circuit is under low pressure and the risk of the development of pulmonary vascular obstructive disease is minimal. Therefore, assessment of suitability for operation, insofar as the status of the pulmonary resistance vessels is concerned, is not complicated in tetralogy by the additional feature of absence of one pulmonary artery. In truncus arteriosus, however, the pulmonary circuit is generally exposed to a pressure equal to that in the systemic circuit (the uncommon exception being the patient with stenosis at the pulmonary artery ostia), and as a result there is a tendency to develop early severe pulmonary arteriolar changes. Thus, the hemodynamic assessment becomes critical in the selection of patients for corrective operation.

The criteria for operability are different for truncus associated with single pulmonary artery than for truncus associated with two pulmonary arteries. We wish to report our experience regarding selection for surgery of patients with truncus arteriosus and associated unilateral absence of a pulmonary artery and the early and late results that have been achieved in such patients.

Material and Methods

This report is concerned exclusively with types I and II truncus arteriosus as defined by Collett and Edwards,1 conditions in which a single pulmonary artery arises or two pulmonary arteries arise from the truncus a short distance above the truncal valve. From August 1967 through
December 1975, 126 patients, ages one through 21 years, were evaluated by cardiac catheterization at the Mayo Clinic to assess their suitability for surgical correction. Fifteen patients (12%) had naturally occurring agenesis of one pulmonary artery. Two additional patients with truncus and natural agenesis of one pulmonary artery had been catheterized elsewhere just before coming to the Mayo Clinic for surgery and are included in this report, giving a total of 17 patients: seven with agenesis of the right pulmonary artery and 10 with agenesis of the left pulmonary artery. Eleven of the 17 (65%) had absent pulmonary artery on the same side as the aortic arch. Five patients had undergone previous banding elsewhere of their single pulmonary artery: four during the first year of life (ages 1½, 2, 4, and 10 months) and one at 11 years of age.

Three additional patients had complete acquired occlusion of one pulmonary artery, secondary to central pulmonary artery banding during infancy, and they are included in the series. These three patients with acquired absent pulmonary artery had had a single band placed on the pulmonary trunk near its origin from the truncus, proximal to its bifurcation, at ages 5, 6, and 9 months. The bandings were performed at other institutions. In one patient, this central band resulted in complete occlusion of the left pulmonary artery, with no narrowing of the right pulmonary artery, and in another, the right pulmonary artery was occluded, with no narrowing of the left pulmonary artery. In the third patient, the left pulmonary artery was completely occluded and the right pulmonary artery was narrowed and distorted. In the two patients who underwent definitive operation at the Mayo Clinic, the surgeon described the occluded pulmonary artery in each as being only a small fibrotic remnant with no evidence of a lumen.

Of the 20 patients (12 boys and eight girls) in the series, 17 were evaluated in our laboratory and three had studies elsewhere. In the 10 patients operated on, the longest interval between catheterization and surgery was one year, and in seven patients, the interval was less than one week.

In every study, the distal pulmonary artery was entered with a catheter where the pressure was determined and a blood sample for oxygen content obtained. Calculations of pulmonary flow and resistance were made in the usual manner. Oxygen consumption was measured in 13 patients, and an assumed value was used in seven.

Ten of the patients were candidates for surgical correction and were operated on between May 1969 and April 1975. In the four patients operated on before November 1972, an aortic homograft was used as the right ventricle-to-pulmonary artery conduit, whereas in the six patients undergoing surgery since that time, the Hancock conduit was used in the repair. Intraoperative postrepair pressure measurements were obtained on all of these patients (table 1). Pressures were recorded from the distal pulmonary artery, beyond the anastomosis to the conduit, and from the systemic circuit (aorta or left ventricle) in seven of the 10 patients. In three patients, the surgeon could not reach the pulmonary artery beyond the distal anastomosis to record pressures after completion of the repair.

There were two operative deaths, and postmortem examination on each included examination of lung sections with histologic grading of the pulmonary arteriolar disease according to the criteria of Heath and Edwards. The grade assigned corresponded to the most severe lesion seen in review of multiple sections irrespective of the actual number of such lesions observed. There have been two late deaths; the one autopsy was performed elsewhere.

All six surviving surgical patients and the 10 nonoperated patients had their present status determined either by re-examination at our clinic or by follow-up letter. Follow-up ranged from nine months to seven years. Of the 10 patients not offered surgery because of severe pulmonary vascular disease, two died subsequently; autopsy was not performed in either case.

### Hemodynamic Assessment

The accurate assessment of pulmonary vascular resistance is vital in truncus arteriosus if surgery is to be performed only on patients who are likely to survive and benefit from the corrective procedure. A previous report established the relationship between pulmonary resistance and surgical mortality in a series of 40 patients with truncus arteriosus (37 with two pulmonary arteries and three with single pulmonary artery) operated on at the Mayo Clinic between 1967 and 1972. The data revealed a direct, significant relationship between pulmonary resistance and operative mortality, and postoperative histologic examination of the lungs in surgical deaths showed excellent correlation between the calculated preoperative pulmonary resistance and the degree of pulmonary arteriolar disease. The study emphasized that different criteria are necessary to assess operability of patients with unilateral absence of a pulmonary artery, in contrast to those with two pulmonary arteries, if calculations of pulmonary flow and resistance are made in the usual manner and if an attempt is made to correlate these parameters with the degree of histologic pulmonary vascular obstructive disease present.

In a patient with truncus arteriosus and two pulmonary arteries, the right and left pulmonary circuits are in parallel and the total pulmonary resistance is therefore expressed by the following equation involving parallel resistances:

$$\frac{1}{R_T} = \frac{1}{R_L} + \frac{1}{R_R}$$

in which $R_T$ = total pulmonary resistance, $R_L$ = left lung resistance, and $R_R$ = right lung resistance.

If, for example, the resistance in each lung is 20 units $m^2$, the total pulmonary resistance would be:

$$\frac{1}{R_T} = \frac{1}{20} + \frac{1}{20} = \frac{2}{20} = \frac{1}{10}$$

$$R_T = 10 \text{ units } m^2$$

This is the calculation of total pulmonary resistance through two parallel pulmonary beds that correlated so well with the degree of histologic arteriolar disease. However, if the patient were to have his right pulmonary artery occluded or his right lung removed, and if the resistance in the left
Thus, special considerations apply to the question of single pulmonary artery. This, in fact, seems to be the only way of correlating the histologic findings in patients with two pulmonary arteries with the same results obtained in the patients with single pulmonary artery that have been used in angiocardiography. The method of having the collateral flow going through the lung, with a significant bronchial hypoplasia, is that of using a calculated collateral flow through the lung. This approximation is obviously an overapproximation if there is significant bronchial flow and the normal lung resistance is doubled.

This method, however, such results were not necessarily in our method. Consequently, the term 'single pulmonary artery' is obviously more precise than the term 'single pulmonary artery' at the time of this contribution. This is because a single pulmonary artery is not a single pulmonary artery, but two pulmonary arteries (that is, twice as many pulmonary arteries). Therefore, the patient with a single pulmonary artery without a significant bronchial hypoplasia, will have twice the total calculated pulmonary flow and hence twice the total hemodynamic situation would apply.
hypertrophy of the lung supplied by the pulmonary artery is a troublesome one because the additional lung volume present is difficult to quantitate. Clearly, in such a situation, the pulmonary resistance that best corresponded to the degree of arteriolar disease present would lie somewhere between the calculated value and the halved value. We have not made such an adjustment in any of our patients, because none of them had clearly significant compensatory hypertrophy of lung volume in the lung supplied by the pulmonary artery. If such existed, however, and its degree could be estimated, it would seem reasonable to assign as the appropriate value of pulmonary resistance, corresponding to the degree of arteriolar disease present, a value proportionate to the additional lung volume felt to present; that is, for example, to assign a value midway between the calculated resistance value and the halved value if the hypertrophied lung supplied by the pulmonary artery were considered to have 50% more volume than a normal lung.

Results

In the 10 patients who were surgical candidates, the calculated pulmonary resistances ranged from 5.3 to 18.4 units m² (table 1). When these calculated resistances were halved in an attempt to correlate them with the histologic arteriolar disease as observed in patients with two pulmonary arteries, the range became 2.7 to 9.2 units m².

Figure 2 shows the comparison between intraoperative postrepair peak systolic pressure ratio (distal pulmonary artery to systemic) and preoperative pulmonary resistance value in two groups of patients with trunci on whom both distal pulmonary artery and systemic (left ventricle or aorta) postrepair pressure measurements were obtained. Analysis of covariance reveals that, when groups I and II (see legend, fig. 2) were compared for a common regression line, no significant differences were detected. However, when groups I and III were compared, the hypothesis of a common regression line is rejected (P < 0.001). These data clearly support the premise that, if pulmonary resistance values in patients with a single pulmonary artery are to be correlated with findings in patients with two pulmonary arteries (that is, two pulmonary arteriolar beds), the halved pulmonary resistance in the single pulmonary artery patient will provide a better indication of the degree of arteriolar disease present — and hence serve as a predictor of the level of postrepair pulmonary hypertension that might be anticipated — than will the total calculated pulmonary resistance.

In three of the 10 patients who underwent operation (cases 4, 8 and 10), the surgeon could not reach beyond the distal conduit anastomosis to measure pressures after repair. Two of these patients (cases 4 and 8) were the only operative deaths. In case 4, the single left pulmonary artery was noted at operation to be hypoplastic. Although the distal anastomosis was made as large as possible, after repair the measured right ventricular pressure was greater than systemic, and the patient died in the operating room. Autopsy revealed that the diameter of the distal anastomosis in this 13-year-old child was only 7 mm, and histologic study of the lungs revealed only grade 1 Heath-Edwards changes in the pulmonary arterioles. Thus, this child’s postoperative right ventricular hypertension was caused by a small distal anastomosis rather than by severe pulmonary vascular disease.

In case 8, death occurred on the operating table because of uncontrollable bleeding from the area of the distal conduit anastomosis. This 20-year-old patient had undergone banding of her single left pulmonary artery at the age of 11 years and at operation at our institution it was noted that the band had migrated far distally to a position near the hilum of the left lung. Because of much scar tissue and distortion of the pulmonary artery at the band site, the distal anastomosis technically was extremely difficult to accomplish, and persistent bleeding from this area could not be controlled. Autopsy revealed that the distal anastomosis was 9 mm in diameter. In addition, examination of the lungs revealed grade 3 Heath-Edwards changes, indicative of moderately severe pulmonary vascular obstructive disease. This patient’s preoperative halved pulmonary arteriolar resistance...
of 9.2 units m$^2$ was the highest of any patient offered surgery, and the lung findings of grade 3 Heath-Edwards changes are entirely compatible with our previous histologic observations in patients with two pulmonary arteries and this level of pulmonary resistance.

In case 10, in addition to complete occlusion of the left pulmonary artery as a result of banding at the age of nine months, there was severe narrowing and distortion of the right pulmonary artery as a consequence of this procedure. Reconstruction of the damaged right pulmonary artery was accomplished to the greatest extent possible, but the surgeon believed that a significant gradient still existed across the distal anastomosis. The pressure beyond the anastomosis could not be measured after repair, but systolic pressure in the right ventricle and conduit was 85 mm Hg when left ventricular systolic pressure was 100 mm Hg. Despite the significant postoperative right-sided hypertension, however, the patient survived operation and, at last report, was doing well, although still receiving digoxin nine months after operation.

There have been two late deaths among the eight operative survivors. One patient (case 1) died unexpectedly nearly six years after operation, shortly after performing on the trampoline in physical education class. Immediately after getting off the trampoline, she complained of feeling ill and then slumped to the gymnasium floor. Vigorous resuscitation efforts by school officials were unsuccessful. No autopsy was obtained. On retrospective questioning, the parents stated that they felt she was still doing reasonably well up until the time of her sudden death but that her exercise tolerance perhaps had not been as good in the six to 12 months immediately preceding her death as it had been in the previous five years after corrective surgery.

One patient (case 2) did well for two years after operation but then began to develop increasing exercise intolerance. Three and one-half years after operation, recaherterization in her home community revealed severe pulmonary hypertension (pulmonary artery pressure 120/50 mm Hg when aorta equaled 98/50 mm Hg). During the next 2½ years, her condition gradually deteriorated, with increasing signs of right heart failure, and she died 6 years 2 months after operation. Autopsy revealed that the surgical repair was complete and intact and histologic examination of the lungs revealed grade 5 Heath-Edwards changes, indicative of very severe pulmonary vascular obstructive disease.

The six surgical survivors are doing well: one (case 3) more than five years after operation and four others (cases 5, 6, 7, and 9) between one and two years after operation. None of these patients is taking cardiac medications, and all are considered to have good exercise tolerance. One patient (case 10) was considered to be doing well nine months postoperatively at the time of follow-up, although still receiving digoxin. None of the six survivors has been recatheterized since surgery.

Ten patients were inoperable because of severe pulmonary vascular disease in the lungs supplied by their single pulmonary artery. Nine of these patients had naturally occurring agenesis of one pulmonary artery, and the tenth had acquired occlusion of the right pulmonary artery secondary to a pulmonary artery banding at the age of five months. The calculated pulmonary resistances in these patients ranged from 22.8 to 38.2 units m$^2$. Halving of these calculated values gives resistance figures ranging from 11.4 to 19.1 units m$^2$, values that suggest severe pulmonary vascular obstructive disease. All of these inoperable patients had systemic arterial saturations below 78% at the time of their catheterization at our clinic, with hemoglobin concentrations ranging from 16.4 to 22.0 g/dl — as compared with the systemic arterial saturation (while the patients were breathing room air) of the 10 operable patients of 80 to 93% achieved with hemoglobin concentrations ranging from 11.6 to 16.5 g/dl (table 1). Thus, a systemic arterial saturation less than 80% in the patient with truncus arteriosus and a single high-pressure pulmonary artery will usually indicate severe pulmonary vascular obstructive disease and inoperability.

Of the 10 nonoperated patients, two have died from progression of their pulmonary vascular disease. Eight are still alive from six months to seven years after catheterization. The present ages of the eight surviving, nonoperated patients range from eight to 19 years. All eight have markedly decreased exercise tolerance (NYHA Class III or IV), and three are nearly totally disabled. All eight patients are cyanotic, and six with recent follow-up are significantly polycythemic, with hemoglobin concentrations ranging from 17.6 to 22.9 g/dl.

Discussion

In the selection of patients for potentially corrective operation, attention must be paid to both operative and late results. Recently, Marcelletti and associates reviewed the early and late results through May 1975 in 92 patients with truncus arteriosus operated on at the Mayo Clinic between September 1967 and January 1975. Of the 92 patients, 85 had two pulmonary arteries (table 2). The data revealed that the operative mortality is directly related to pulmonary resistance. For patients with pulmonary resistance less than 10.0 units m$^2$, the operative mortality was 21% as compared with 50% for the 14 patients with pulmonary resistance greater than 10.0 units m$^2$. In addition, of the seven operative survivors in this latter group, one patient who had a preoperative pulmonary resistance of 11.4 units m$^2$ died three years after operation from severe progressive pulmonary vascular obstructive disease. Among the 56 operative survivors in the group with pulmonary resistance less than 10.0 units m$^2$, there has been only one late death secondary to progressive pulmonary vascular disease. This occurred four years postoperatively in a patient whose preoperative pulmonary resistance was 8.6 units m$^2$. These data, plus the observation that patients with truncus and two pulmonary arteries and a pulmonary resistance greater than 10.0 units m$^2$ often can be reasonably active for years, with systemic arterial oxygen saturations approximating 80% achieved with modest degrees of polycythemia, have led to our present position of not offering operation to patients with pulmonary resistance greater than 10.0 units m$^2$.

Patients studied in our laboratory who are found to have pulmonary resistances of 10.0 units m$^2$ or greater while breathing room air are always immediately assessed while breathing 100% oxygen to ascertain with certainty that this elevated resistance is on the basis of occlusive intimal
arteriolar disease rather than as a result of medial vasoconstriction secondary to hypoxia and acidosis. On occasion, pharmacologic vasodilators such as tolazoline and isoproterenol are also used in this evaluation. It has been our experience that, with the exception of an occasional child in the first year of life, patients with a pulmonary resistance greater than 10.0 units m⁻² rarely exhibit a significant reactivity of their pulmonary bed and resultant increase in pulmonary flow in response to either oxygen breathing or pharmacologic vasodilators.

Different criteria for operability are necessary in the group of patients with single pulmonary artery. In our series, one patient (case 3) with a calculated pulmonary resistance of 17.1 units m⁻² had a pulmonary circuit peak systolic pressure only slightly more than one-half systemic immediately after operation. In the patient with truncus arteriosus and two pulmonary arteries, experience has shown that a pulmonary resistance of this level is secondary to advanced pulmonary arteriolar occlusive disease, and as a consequence, the arteriolar bed is no longer a potentially reactive one. In contrast, the data on this patient indicate that at operation his single pulmonary arteriolar bed was still a reactive one, capable of dilating enough so that after operation it could accept the entire cardiac output with only a modest elevation in pulmonary artery pressure.

There were two late deaths (cases 1 and 2). Both patients probably died as a result of continued postoperative progression of their pulmonary vascular disease. Although patients with a single pulmonary artery and a pulmonary resistance calculated in the usual manner as great as 15 to 20 units m⁻² may still have reactive pulmonary arteriolar beds and pulmonary artery pressures immediately after operation that are compatible with survival, they seem to be at particularly high risk of continued postoperative progression of their pulmonary vascular disease and eventual death secondary to the resultant pulmonary hypertension. Experimental work in animals⁶,¹⁰ has suggested that ligation of one pulmonary artery after birth often leads to pulmonary arteriolar changes in the opposite lung. An extensive review by Pool¹¹ of both clinical experience and experimental work concluded that the presence of only a single pulmonary artery was important in the genesis of pulmonary arteriolar changes. After surgical repair the entire cardiac output of these patients with truncus arteriosus must pass through one lung, and the postoperative persistent obligatory increase in flow through this lung may make continued progression of pulmonary vascular obstructive disease in these patients more likely than in patients with two pulmonary arteries.

The operative mortality for correction of truncus arteriosus using a right ventricle-to-pulmonary artery conduit has steadily decreased since the inception of the operation in 1967. Since November 1972, the Hancock conduit has been used exclusively at the Mayo Clinic for this operation, and the surgical mortality in 33 cases during the past three years in which this conduit was used has been 9%. The steady reduction in surgical mortality during the past eight years is related both to the evolution of improved operative techniques and to improved patient selection. At the present time in hemodynamically favorable patients, both with single and with two pulmonary arteries, who are more than three years old, the operative risk is about 10%. Unfortunately, the reported risk of definitive correction of truncus arteriosus in the very young patient has remained much higher than this at our institution¹ and at others.¹²,¹³ Obviously, correction during the first year or two of life is highly desirable in these patients because of the high risk of the early development of pulmonary vascular disease, and this would be particularly applicable to the patient with single pulmonary artery. Although the patients in this report who survived banding during the first year of life (cases 5, 6, 9, and 10) had apparently all had their pulmonary arteriolar bed effectively protected as a result of the palliative operation, the operative risk of pulmonary artery banding in the infant with truncus is very high, with most series¹⁴,¹⁵ reporting an operative mortality greater than 50%. Very recent results¹⁶ (J. Stark, P. Ebert, personal communications) involving definitive correction of infants with truncus arteriosus are more encouraging, however, and would now prompt us to attempt this rather than banding in the infant who is in refractory congestive heart failure. It would seem reasonable to anticipate that continued surgical advances will soon make elective surgical correction of all patients with truncus arteriosus during the first year or two of life the accepted mode of management in order to minimize the risk of significant pulmonary arteriolar changes developing either before or after definitive operation.

References

Myocardial Metabolism in Cyanotic Congenital Heart Disease Studied by Arteriovenous Differences of Lactate, Phosphate, and Potassium at Rest and during Atrial Pacing

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SUMMARY To study myocardial metabolism in chronic hypoxia due to cyanotic congenital heart disease, coronary arteriovenous differences of lactate (L), pyruvate (P), inorganic phosphate (P_i) and potassium (K) were measured in 14 cyanotic patients and seven controls, at rest and during atrial pacing. At rest, there was no difference in any parameter between cyanotic and noncyanotic patients. During atrial pacing (150–175/min) for 10 min, a moderate drop in L-extraction occurred in the control patients with some increase in L/P ratio in coronary venous blood.

Cyanotic patients fell into two groups: in nine (group I), the arterial oxygen saturation (SaO_2) dropped with pacing. Their L-extraction fell sharply, from $28.1 \pm 3.12$ to $-2.8 \pm 5.51$ and L production occurred in five. There was a significant increase in coronary venous L/P ratio. Five cyanotic patients (group II) showed no drop in SaO_2 with pacing, and L extraction as well as L/P ratio remained stable. Uptake of P_i was noted in all patients at rest; during pacing this disappeared in controls and group I cyanotics but not in group II. No K changes were seen in any patient.

Thus, myocardial metabolism is normal at rest in patients with cyanotic CHD; during atrial pacing, a shift toward anaerobic metabolism occurs if SaO_2 drops; cyanotic patients whose SaO_2 remains stable appear to withstand pacing better than controls.

In patients with cyanotic congenital heart disease (CCHD), the myocardium is constantly perfused with hypoxic blood. Little is known about the effect of this chronic hypoxia on myocardial metabolism and the possible effects on the heart muscle. Microinfarcts of the left ventricle in tetralogy of Fallot have been described, and might be the cause of the left ventricular dysfunction which has sometimes been found after surgical correction of the defect. The question arises whether hypoxia due to CCHD might induce changes in myocardial metabolism similar to those seen in ischemia of coronary heart disease, i.e., decreased lactate extraction, and release of phosphate and potassium into coronary venous blood. The present study was designed to assess whether such changes occur in CCHD either at rest, or during the stress of atrial pacing. *

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*Preliminary accounts have been presented to the American College of Cardiology, the British Cardiac Society and the European Society for Clinical Investigation.
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