Surgical Treatment of Truncus Arteriosus in the First 6 Months of Life

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One hundred six infants were seen at the University of California Medical Center between 1974 and 1981 with the diagnosis of truncus arteriosus. One hundred of these underwent physiologic correction prior to 6 months of age. Six infants died prior to operation while undergoing intense medical therapy to improve their basic condition. There were 11 operative deaths with a mortality rate of 11%. Of the 86 long-term survivors, 55 have returned for conduit change because of either body growth or pseudointima proliferation of the conduit. There had been no mortalities at the time of conduit change, and 29 of these were repaired using a straight tube between the ventricle and pulmonary trunk, while 26 had valve conduits placed. Physiologic correction in the first 6 months of life has been accomplished with a low mortality rate and apparent good long-term results with none of the survivors having evidence of elevated pulmonary vascular resistance.

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THE PHYSIOLOGIC CORRECTION of truncus arteriosus was described by McGoon and associates in 1968.[1] This followed a frustrating and dismal experience of attempts to palliate patients with Collett and Edward's Type I or II truncus arteriosus by pulmonary artery banding as an attempt to reduce pulmonary bloodflow.[2] The results of banding were universally poor, with the most optimistic series showing operative mortalities in the range of 50%.[3] The follow-up of patients undergoing physiologic correction showed that a high percentage suffered from pulmonary vascular disease, which occurs at a fairly young age in a majority of patients with truncus arteriosus.[4,5] Thus, to determine if a significant reduction in the incidence of pulmonary vascular disease could be noted, physiologic correction would most likely have to be performed at a very young age.

Methods and Materials

The current series represents 106 infants under 6 months of age who presented with a clinical diagnosis of truncus arteriosus at the University of California, San Francisco between 1975 and 1981. All of these infants had either Collett and Edward's Type I or II lesions. The ages ranged from 6 days to 6 months, and the distribution is shown in Table 1. The clinical symptoms at the time of presentation are shown in Figure 1. The majority of the infants presented with congestive heart failure. One hundred four of the 106 children were receiving either diuretics and/or digitalis at the time of presentation. The weights at the time of presentation ranged from 1.9 to 5.2 kg, with the majority of the patients weighing from between 2.8 and 3.8 kg. In 87 patients, the diagnosis was confirmed by cardiac catheterization. In the remaining 19 patients, the diagnosis was obtained from clinical findings and echocardiography.

A supravalvular injection into the common trunk is the most important of the diagnostic studies. It determines the degree of truncal valve insufficiency, the size, and to a degree, the rate of flow through the pulmonary arteries (Fig. 2). In general, the intracardiac anatomy can be well defined by echocardiography (Fig. 3). Twenty-nine of the patients were considered to have abnormalities of the truncal valve with murmurs of truncal valve incompetence. Early in the experience, 18 patients were considered to be in profound heart failure of which it was thought that additional medical management might be able to improve their status and thus decrease the risk for operation. Fourteen of these 18 were patients with truncal valve insufficiency. Unfortunately, six of these infants died while undergoing medical therapy. It should be noted that in five of the six, improvement seemed to be dramatic, and one would have thought that, on clinical grounds, the objective was being obtained. The remaining 88 infants presented for a physiologic correction at the time of the admission.

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TABLE 1. *Clinical Presentation at Time of Admission (N = 106)*

Condition	No. of Patients
Heart failure	99
Decreased growth	103
Cyanosis	5
Normal	3

The operation was performed using cardiopulmonary bypass and employing an arterial line placed high in the ascending aorta or in the transverse arch, and a single atrial catheter. The flow rate was 125 ml/min. At the time of initiation of bypass, the pulmonary arteries are occluded to prevent overcirculation of the lung. The temperature is rapidly reduced to approximately 25°C. At this time, the aorta is cross-clamped, and the pulmonary trunk freed from the common trunk. The defect in the remaining aorta is closed. The ventriculotomy is performed and the aortic clamp released to inspect the degree of truncal valve incompetence. The clamp is reapplied and the ventricular septal defect closed with a continuous suture and a Teflon® felt patch. The aortic clamp is then released, and rewarming may be accomplished during the time that the conduit is placed between the pulmonary trunk and the right ventricular outflow tract.

Results

One hundred of these infants underwent physiologic correction with an operative mortality of 11% (Table 2). Ninety-eight of the infants had repair using a valve conduit of which ten were allografts and 88 were Dacron® tubes with a heterograft valve. In two infants, it was possible to anastomose the pulmonary trunk directly to the right ventricular outflow tract. Of the 11 infants who expired, eight had evidence of truncal valve insufficiency with one of the deaths in a child who had an associated truncal valve replacement. Two of the infants had cyanosis and evidence before surgery of pulmonary vascular disease. At the conclusion of operation, the pulmonary artery pressure exceeded that of the aortic pressure. Twenty-six of the 98 surviving infants leaving the operating room were returned for postoperative bleeding. Of the 89 patients discharged from the hospital, there have been three late deaths: one with bacterial endocarditis unrecognized, and the other two unrelated to their cardiac conditions. Of the 86 infants surviving over 2 years, there have been 55 returns for conduit changes. At the time of conduit change, 26 have received a valve conduit, and 29 a non-valve conduit. Five of these patients had associated aortic truncal valve replacement at the time of conduit change (Fig. 4). There were no mortalities among the children at the time of their conduit change. No infant at the time of recatheterization has shown evidence of elevated pulmonary vascular resistance.

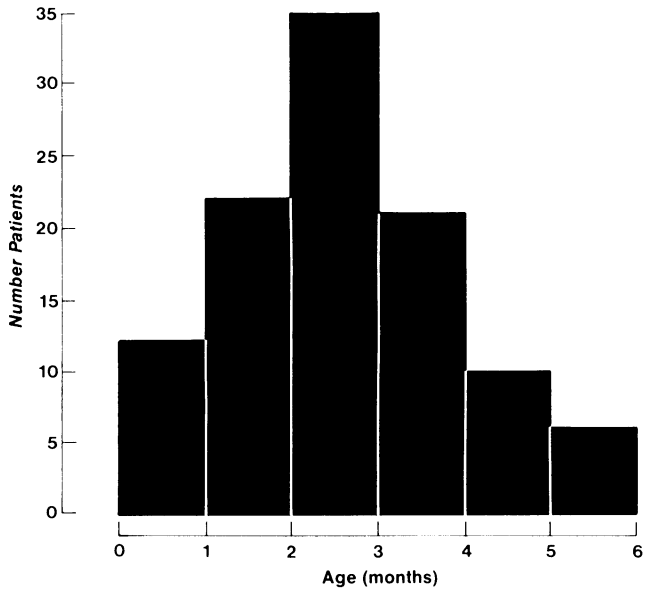

FIG. 1. Age distribution of the 106 patients at the time of initial presentation.

Discussion

It is clear that the majority of infants with truncus arteriosus presenting in heart failure in the first year of life will require some type of operative intervention for survival. The approach using a physiologic correction appears to have a lower mortality than that associated with pulmonary artery banding. As well, it does not incur deformity or destruction of the branch pulmonary arteries (Fig. 5). In all of the infants undergoing corrective surgery, it has always been possible to detach the pulmonary artery trunk from the posterior wall of the aorta and close the aorta directly. One can excise an adequate portion of the aortic wall in most instances to be certain that a confluence of pulmonary arteries is present. When making the initial incision in the posterior wall of the aorta, one must be cautious to avoid the suspension system of the truncal valve and also to identify the orifice of the left coronary artery. The commissures of the truncal valve are often shortened, and thus the depth of the sinus of valsalva is less than normal. In this situation, it is possible that the left coronary orifice may be fairly high in the sinus and can either be cut at the time of the excision of the pulmonary cuff or be incorporated into the suture line at the time of closure of the aortotomy.

The use of standard cardiopulmonary bypass with reduction of temperature to 25°C–27°C seems to be

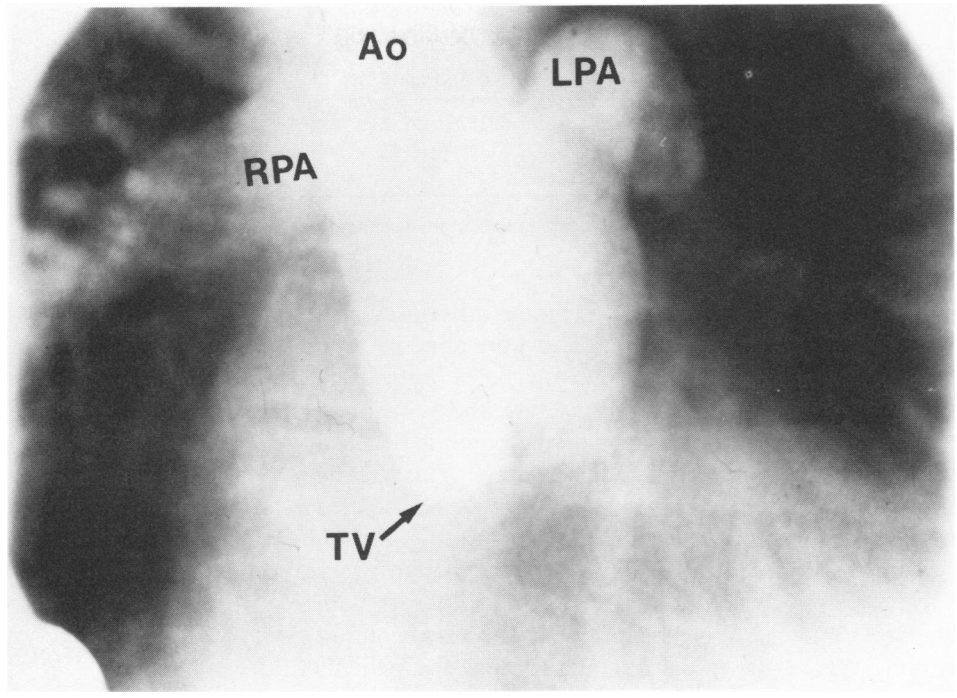

FIG. 2. A truncal root injection, showing a nice outline of the truncal valve (TV), with no evidence of truncus valve incompetence and good-sized pulmonary arteries (RPA and LPA).

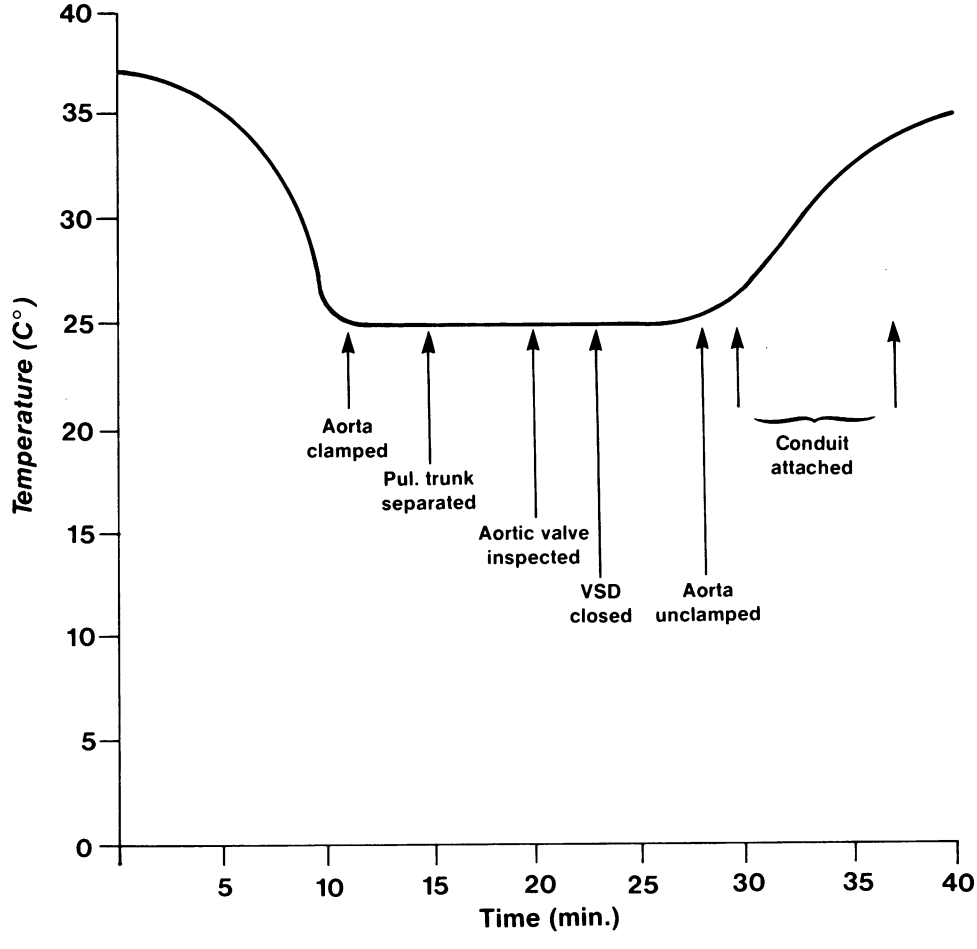

FIG. 3. Diagrammatic representation of the time sequence and conduct of the operation for truncus repair. Note that the initial temperature drop is rapid when cardiopulmonary bypass is initiated, and that the conduit can be attached during the rewarming period. Thus, the entire operation can be accomplished in approximately 40 minutes of cardiopulmonary bypass time.

TABLE 2. *Results of Operation for Physiologic Correction of Trunchus in the First 6 Months of Life (N = 106)*

	Total	Mortality
Before surgery		6*
Operation	100	11
Late	89	3

* Six infants died before surgery undergoing medical management.

adequate for the correction of truncus arteriosus. Although the first four infants reported were done by total circulatory rest,[6] the operation can be shortened by maintaining flow during the procedure and rewarming during the time of attachment of the conduit between the pulmonary artery and right ventricle. This greatly shortens the total bypass time as no specific isolated time is required for rewarming alone.

The question continually arises as whether to use a valve or nonvalve conduit in repair of infants with truncus arteriosus. It is our opinion that a valve conduit is preferable at the initial operation. Since the patients have great swings in pulmonary artery pressure due to unstable pulmonary vascular resistance common in all infants. An infant incurring a rapid increase of pulmonary artery pressure in a non-valve conduit will significantly increase the amount of pulmonary regurgitation and may prompt right-sided heart failure, whereas, at the time of conduit change, if the pulmonary artery size is good and right ventricular performance near normal, then the placement of a non-valve conduit seems reasonable. Whether these children will incur right-sided ventricular damage from prolong pulmonary regurgitation remains unclear. It has been our policy to place a smaller non-valve graft at the time of conduit change than if we were using a valve conduit. Thus, to a degree, the amount of pulmonary regurgitation can be restricted, and the rigid form of the conduit may control the amount of pulmonary regurgitation. The unfortunate circumstances of right ventricular deterioration seen in Tetralogies of Fallot with a dilated outflow tract may not occur in the presence of a rigid conduit, in which the outflow tract remains unchanged and the amount of pulmonary regurgitation is consistent.[7] Concurrently, the conduit itself is nonexpansile, and thus does not project as a right ventricular aneurysm, extending out from the right ventricle since it will not expand.

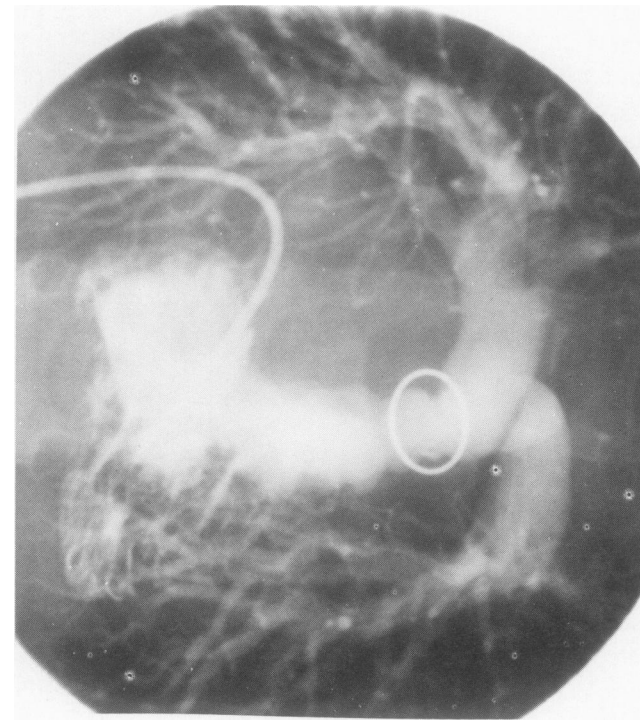

FIG. 5. Postoperative right ventricular injection in a child 5 years following the repair of truncus. Note the good growth of the pulmonary arteries with a narrow column of contrast material going through the valve conduit. It can also be noted that the ring of the conduit is considerably larger than the flow column, indicating pseudointima proliferation in the conduit.

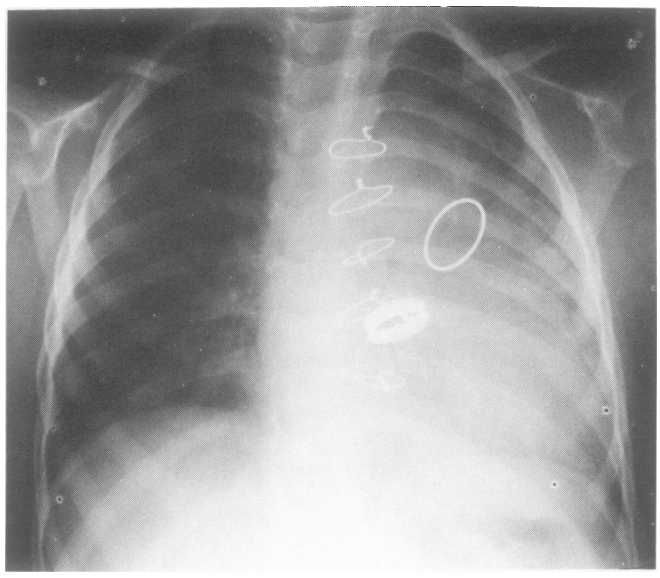

FIG. 4. Postoperative chest x-ray following a conduit change in a 4-year-old child with massive truncal valve incompetence. Note the increased heart size remains, and the ring of the Bjork–Shiley aortic valve as well as the ring of the valve conduit are visualized.

The basic operation as described by McGoon is essentially unchanged to this date.[1] The ventricular septal defect in the truncus anomaly is usually high, muscular in rim, and no conduction injuries have been noted in our group of patients.

Truncal valve insufficiency remains a major problem of the entity, and if significant at the time of initial operation, probably is best managed by associated truncal

valve replacement at the time of repair. Since the majority of infant mortality in this series (eight of 11) had truncal valve insufficiency, it is clear that a more aggressive approach might have increased the survival rate. Five children at the time of conduit change had residual truncal valve insufficiency–significant in degree that a truncal valve was replaced with a mechanical prosthesis at this time. All five of these children survived surgery and were greatly improved by the combination procedure. In theory, closing the ventricular septal defect in patients with significant truncal valve insufficiency actually may worsen the strain on the left ventricule since the same amount of regurgitation can be anticipated per beat, and now it must be directed entirely to the left ventricular chamber, rather than dispersed over the ventricular volume of both the right and left ventricle when the ventricular septae defect was open. Therefore, severe left ventricular failure has been seen in some of the infants, and almost all have required inotrophic support from several days to weeks if they were left with accompanying truncal valve insufficiency.

The number of times conduit changes will be required in these patients is unknown. Clearly, the infants who had the allograft valves performed the best, in that none have had to have their conduit changed. It would seem more reasonable to consider an allograft at the time of the first conduit change, as one could most likely place an adult-sized allograft at this time. If a non-valve conduit proves feasible, then the composition of the material may not be as important in the straight tube.

The most optimistic side of early correction in the truncus anomaly is related to the absence of pulmonary vascular disease and to the growth of the pulmonary arteries, which to date has been normal in children that we have followed.[8] No late mortality has been related directly to the truncus anomaly, and the one child with unrecognized bacterial endocarditis who was not treated for several weeks possibly could have been saved with earlier intervention.

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Discussion

Dr. John A. Waldhausen (Hershey, Pennsylvania): This is an excellent report on a major problem in congenital heart disease. It is not only one of the largest series in the management of truncus arteriosus but it has the best early and late results.

We too have been interested in the problem of intimal proliferation seen in these conduits, and my associates, Dr. William Pierce and Dr. Wayne Richenbacher, have come up with a possible solution derived from the artificial heart program.

(Slide) This is a Dacron fabric graft that has been coated with a thin layer of filler-free silicone rubber which gives the prosthesis a very good blood surface interface. There has been no clot formation or platelet aggregation on the inner wall.

(Slide) When you place an ordinary Dacron graft into a goat as a main pulmonary artery replacement, at 8 months you see this tremendous intimal proliferation similar to what is seen in children.

(Slide) On the other hand, if you place the silicone rubber coated graft into the goat's pulmonary artery, you see very little tissue ingrowth, and the tube remains open.

Based on these studies, we have started to use these filler-free silicone rubber coated Dacron tubes in patients, with very good results. We agree with Dr. Ebert that replacement of conduits is suprisingly easy, and the results are very good with a very low mortality.

I would like to ask Dr. Ebert: When does he think we need to place a pulmonary valve in the conduit in older children? I think it is evident that infants do need valves, but in older children there is a question for this need when the pulmonary vascular resistance is low.

Dr. Gordon K. Danielson (Rochester, Minnesota): I would like to compliment Dr. Ebert and his colleagues on their excellent results.

Dr. Ebert has, I believe, performed a great service in showing that the mortality for the repair of truncus arteriosus can actually be lower in the first 6 months of life than at any later time. Previously, our own philosophy, and that of many surgical centers, was to try to bring the infant with congestive heart failure to an older age with maximal medical therapy, operating only when this therapy failed and the infant was either in end-stage heart failure or had developed pulmonary vascular disease. Dr. Ebert has shown that this philosophy was in error.

I would like to contrast his series with our own, which dates from September 1967, when Dr. Dwight McGoon performed the first successful surgical repair of truncus arteriosus, using an aortic homograft. This experience extends over a 16-year period. The mean age of these patients was older—6 years. It is our impression also that several adverse effects develop with increasing age. The first is elevation of pulmonary vascular resistance, which was present in 80% of these patients. Indeed, 27% had a resistance greater than 10 units, a level that we would now consider inoperable.

Furthermore, truncal valve insufficiency is progressive in many patients, and occurred in a total of 37% of our patients. The final factor that appears to be related to age is progressive ventricular dysfunction, but this has been more difficult to analyze.

Now, the operative mortality (slide) for this series was 28%, which contrasts markedly with the 11% reported by Dr. Ebert for his patients under 6 months of age. It is my recollection that the operative mortality at his institution has run rather consistently about twice this in patients