Improving early and intermediate results of truncus arteriosus repair: a new technique of truncal valve repair

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Improving Early and Intermediate Results of Truncus Arteriosus Repair: A New Technique of Truncal Valve Repair

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Background. **Despite improved surgical results for truncus arteriosus, overall mortality rates, remain higher than those reported for other complex congenital heart diseases, especially with truncal valve regurgitation or an interrupted aortic arch.**

Methods. **Seventeen patients had complete repair of the truncus arteriosus at the Cleveland Clinic Foundation between August 1993 and June 1997. The age at operation ranged from 2 days to 4.5 years. Associated abnormalities included interrupted aortic arch in 3 patients and abnormal coronary artery anatomy in 3. Four patients had more than moderate truncal valve insufficiency requiring concomitant truncal valve repair.**

Results. **There were no early deaths and only one late death at a mean follow-up of 24 months. The death occurred 3 months postoperatively and resulted from refractory pulmonary vascular obstructive disease in a patient who was referred at 1 year of age. Reoperation was required in 4 patients.**

Conclusions. **Even in the presence of associated anomalies complete repair was performed with a low mortality rate. Truncal valve repair can be performed safely in the neonate with good results.**

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In patients with truncus arteriosus, primary repair in early infancy is recommended to prevent the development of irreversible pulmonary vascular obstructive disease [1, 2]. Whereas high mortality rates were reported after staged palliative procedures, such as main pulmonary or branch pulmonary artery banding [3, 4], recent studies found improved outcome for primary complete repair in early infancy [5–8]. The overall mortality rate for truncus arteriosus, however, remains higher than that reported for other congenital heart diseases [5–10]. Treatment of truncal valve regurgitation and associated abnormalities, such as interrupted aortic arch and abnormal coronary artery anatomy, still negatively affect survival [6, 7].

Patients and Methods

Seventeen patients with truncus arteriosus had complete repair between August 1993 and June 1997. The age at operation (Table 1) ranged from 2 days to 4.5 years (median, 7 weeks). Seven patients were less than 1 month old. Three patients were more than 12 months old and were referred at this late age. Of the 3 older patients, 1 had a hemitruncus and major aortopulmonary collaterals, and 1 was 4.5 years old with an aneurysm of the truncus and compression of the right pulmonary artery, which protected the right pulmonary vasculature from systemic pressure. The third patient was 12 months old and had a high pulmonary vasculature resistance still responsive to oxygen.

Weight at operation ranged from 1.3 to 6.8 kg (median, 3.5 kg). Five premature infants were less than 3 kg. There was a slight male preponderance of 9 patients. Associated noncardiac abnormalities included partial DiGeorge syndrome in 4 patients, of which only 1 had significantly low calcium levels. Two patients had type I truncus arteriosus, 5 patients with anatomy intermediate between type I and type II, and 8 patients had type II, and 2 patients had hemitruncus with associated cardiac diagnoses (Table 2) included three patients with a type B interrupted aortic arch, and 3 patients with an abnormal coronary artery pattern.

Preoperative echocardiography was done in all patients. Five patients had cardiac catheterization before complete repair. Only two of these studies were performed at our institution to assess preoperative pulmonary vascular resistance. Four patients had more than moderate truncal valve regurgitation. The morphological characteristics of the truncal valve and associated regurgitation are shown in Table 3.

Surgical Technique

After stabilization of hemodynamics and control of congestive cardiac failure, repair was done through a midline sternotomy. The pericardium was opened to the right side to provide cover for conduit with pericardium

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Table 1. Age at Operation and Associated Features

Age	No. of Patients	Associated Feature[a]
< 1 week	2	IAA(1), TVR(1)
1–4 weeks	5	IAA(2), TVR(2)
1–12 months	7	TVR(1), hemitruncus(1)
> 1 year	3	hemitruncus(1), trunk aneurysm(1)

[a] Number in parentheses indicates number of patients with associated anomalies.

IAA = interrupted aortic arch; TVR = truncal valve regurgitation.

after the repair. The aortic cannula was placed as cephalad as possible, and bicaval cannulation was used in all except four cases. High-flow cardiopulmonary bypass at a flow rate of 150 to 200 mL/kg/per minute with phenoxybenzamine (1 mg/kg) was used. Deep hypothermic circulatory arrest at 18°C nasopharyngeal temperature was used in seven cases. After initiation of bypass, the branch pulmonary arteries were occluded with vessel loops to prevent overcirculation of the lungs and to preserve systemic and coronary blood flow. This process usually stabilized an unstable patient immediately after initiation of bypass. The truncus arteriosus was cross-clamped above the main pulmonary artery, and crystalloid cardioplegia was administered in a prograde fashion through the truncal root. In cases with significant truncal valve regurgitation, cardioplegic solution was administered directly into the coronary ostia. The left atrium was vented through the interatrial septum in all cases. The pulmonary trunk was divided from the truncus arteriosus, and this defect in the aorta was usually closed primarily. A right ventriculotomy was done, and the ventricular septal defect was closed with an albuminized Dacron patch and a combination of a few interrupted pledgetted and continuous running polypropylene sutures. The surgically created atrial septal defect was closed directly and the cross clamp released. During rewarming, the conduit was placed between the right ventricle and the pulmonary arteries.

Cryopreserved homograft valved conduits (11 aortic, one pulmonary) and Dacron polyester Hancock conduits (5 patients) were used. When an aortic homograft was used, a portion of the aortic arch was used for the proximal gusset. In 3 patients type B interrupted aortic arch was repaired by end-to-end direct anastomosis. Mean cardiopulmonary bypass time was 141 minutes (range, 100 to 206 minutes). Mean aortic cross clamp time was 49 minutes (range, 33 to 81 minutes).

Table 2. Associated Cardiac Diagnoses

Diagnosis	No.
Interrupted aortic arch	3
Trunk aneurysm	1
Nonconfluent pulmonary artery	2
Hypoplastic right pulmonary artery	1
Major aortopulmonary collaterals	1
Major coronary anomaly	3
Single ostium	1
Double left anterior descending artery	1
High take off left coronary	1

Table 3. Relationship Between Truncal Valve Anatomy and Regurgitation

Morphologic Characteristic	No.	No. (%) of Patients With Regurgitation
Bicuspid	3	0 (0)
Tricuspid	9	1 (11)
Quadricuspid	5	3 (67)

Four patients required truncal valve repair. A quadricuspid valve was remodeled to a tricuspid valve in 3 patients, and subcommissural sutures were placed in 1 patient. The remodeling technique from quadricuspid to tricuspid truncal valve excised the smallest cusp of the truncal valve (Fig 1) and closed this sinus by a subcommissural suture approximating the adjacent cusps (Fig 2). In two cases with interrupted aortic arch and one case with type II truncus arteriosus, the Lecompte maneuver was done with the primary repair [10]. Left atrial and pulmonary artery pressures were monitored continuously postoperatively. The patients were mildly hyperventilated and the hematocrit was kept between 30% and 35%. Nitric oxide, although available, was not required for postoperative treatment.

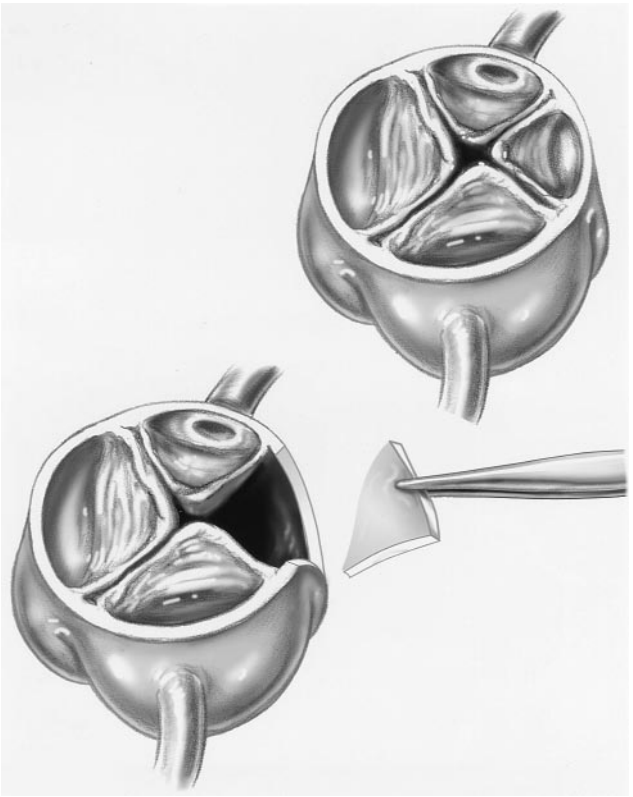

Fig 1. Truncal valve repair, in which the number of leaflets is reduced. The smallest cusp is excised as illustrated.

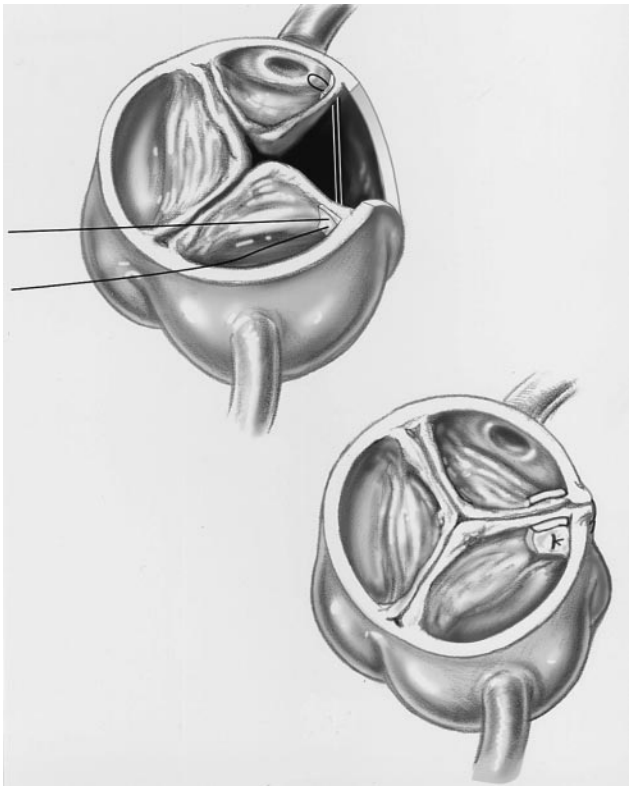

Fig 2. A defect of sinus was closed with a subcommissural suture approximating the adjacent cusps.

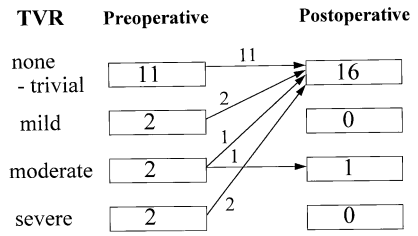

Fig 3. Degree of truncal valve regurgitation before and after complete repair of truncus arteriosus in 17 patients. (TVR = truncal valve regurgitation.)

Results

There were no early deaths. A patient from a foreign country operated on at age 1 year died 3 months postoperatively. Clinically, her postoperative course was uneventful except for a low systemic saturation, which was responsive to oxygen. Postoperative echocardiography showed moderately severe pulmonary hypertension. Cardiac catheterization after repair showed a pulmonary artery pressure 86% of systemic, partially responsive to oxygen and nitroglycerin. She was discharged on home oxygen and a nitroglycerin patch; however she died in her home country where follow-up treatment was not readily available. An autopsy was not available.

Reoperation was required in 4 patients. Two patients with concomitant interrupted aortic arch had right pulmonary artery compression with decreased pulmonary blood flow to the right lung, suspected radiographically and confirmed by bedside lung perfusion scanning. This problem was relieved by the Lecompte maneuver at reoperation.

One patient had truncal valve replacement twice because of infective endocarditis and homograft failure at 1 and 4 months after initial repair. The other patient with a homograft conduit, which had aneurysmal degeneration 3 months postoperatively, required homograft replacement. Two patients had infective endocarditis, one of whom also had partial DiGeorge syndrome. One patient had truncal valve replacement and another was treated successfully with medication.

The relationship between the number of leaflets and truncal valve regurgitation is shown in Table 3. We found a high incidence of more than moderate truncal valve regurgitation in the quadricuspid truncal valve group. In this series, bicuspid or tricuspid truncal valves with more than moderate regurgitation were uncommon.

All patients with more than moderate preoperative truncal valve regurgitation had valve repairs, and only 1 patient had residual moderate truncal valve regurgitation postoperatively (Table 4). The patient who had infective endocarditis was a 2-day-old infant who initially had a good result. Immediate postoperative transesophageal echocardiogram showed minimal to trivial truncal valve incompetence. His postoperative course was complicated by *Staphylococcus* endocarditis leading to severe truncal valve regurgitation, which ultimately necessitated emergency truncal valve replacement for refractory congestive cardiac failure and compromise. Figure 3 shows the relationship of truncal valve regurgitation before and after complete repair; only 1 patient had moderate truncal valve regurgitation after complete repair.

Table 4. Summary of Truncal Valve Repair

Patient No.	Age	Degree of TVR Preoperatively	Valve Morphology	Valve Repair Technique	Degree of TVR Postoperatively	Outcome Follow-up
1	2 days	Severe	Quadricuspid	Quadricuspid to tricuspid	Trivial	IE and TVR (POD 20)
2	14 days	Moderate	Quadricuspid	Quadricuspid to tricuspid	Trivial	22 months
3	2 months	Severe	Tricuspid	Subcommisural suture	Trivial	13 months
4	20 days	Moderate	Quadricuspid	Quadricuspid to tricuspid	Moderate	1 month

IE = infective endocarditis; POD = postoperative day; TVR = truncal valve repair.

Comment

With recent improvements in neonatal cardiopulmonary bypass techniques and small body technology, primary complete repair should be performed before development of pulmonary vascular obstructive disease and compromise of ventricular function [11]. In symptomatic neonates poorly controlled with diuretics, operation should be scheduled as soon as possible. If symptoms are well controlled by diuretics and fluid restriction alone, the operation can be electively scheduled for a few weeks of age. In this period, meticulous follow-up is essential even if the child is in a stable condition, because as pulmonary vascular resistance decreases, severe congestive cardiac failure can develop rapidly. There is no reason to delay repair by more than 2 to 3 months, as the risk of refractory pulmonary hypertension increases after 3 months of age [2]. If the patient presents after 3 months of age, an operation should be performed electively as soon as possible. In our series, 3 patients more than 12 months old were referred for surgery. In one patient, pulmonary vascular resistance was high but was responsive to oxygen. This patient died 3 months postoperatively probably from pulmonary vascular obstructive disease. Late presentation mandates precise assessment of pulmonary vascular resistance. Our three late referral cases had preoperative cardiac catheterization to assess the pulmonary vascular resistance and exclude the presence of major aortopulmonary collaterals.

In the neonatal period, proper preoperative treatment is critical. At diagnosis, patients in severe congestive cardiac failure are treated with diuretics, fluid restriction, and tube feedings. If these measures are inadequate, the patient is intubated, ventilation is used to limit pulmonary blood flow, and surgical repair follows within days. In the present series, 7 patients (41%) required intubation and ventilation to treat congestive heart failure optimally.

In the current literature, the hospital mortality rate is still above 10% [6–10]. If repair of truncus arteriosus is combined with interrupted aortic arch or truncal valve regurgitation, there is a significantly higher mortality rate [6, 7]. In this study, there were no hospital deaths; the late mortality rate of 5% represents 1 patient probably with pulmonary vascular obstructive disease. Although pulmonary vascular obstructive disease was the only risk factor for mortality in this series, our series is small, and no real significance or inferences can be drawn realistically from hazards and risk factors for survival. The major difference between our perioperative treatment and that of other groups is the use of continuous high-flow cardiopulmonary bypass with phenoxybenzamine. We believe that phenoxybenzamine greatly improves peripheral organ perfusion during cardiopulmonary bypass and postoperatively [12].

Truncal valve insufficiency remains a challenging problem. Some investigators advocated that a significantly insufficient truncal valve should be replaced at the time of primary repair [6, 7, 13], whereas others, like us, prefer to attempt repair if the anatomic structure is not too unfavorable [14, 15]. In the present series, less than moderate truncal valve incompetence was managed conservatively. All truncal valves were preserved, even in patients with moderate to severe truncal valve regurgitation. Only 1 patient required truncal valve replacement after repair, resulting from infective endocarditis.

We currently use two techniques to treat truncal valve regurgitation. One technique is subcommissural suture placement [16] to correct prolapse and improve central cusp coaptation. In highly incompetent quadricuspid valves the smallest leaflet is excised and the corresponding segment of annulus is plicated with a subcommissural suture. The benefit of this technique is to avoid suturing the leaflet itself, which usually has myxomatous change in neonates with severe truncal valve regurgitation. This technique was used successfully in 3 neonates. Elami and coworkers [14] reported five cases of truncal valve repair, all in patients older than 1 month [14]. Our experience included 3 neonates. However our follow-up is short and a longer period of follow-up is necessary to assess the durability of the repair. There have been no recent studies of the relationship between the number of leaflets of the truncal valve and incidence of truncal valve regurgitation. We found that quadricuspid leaflets have a higher incidence of truncal valve regurgitation compared with either bicuspid or tricuspid valves.

Recently, several authors reported anterior translocation of the pulmonary artery bifurcation, in cases of truncus arteriosus repair associated with a interrupted aortic arch, when the right pulmonary artery had become compressed by the reconstructed neoaortic arch [16, 17]. After a similar experience we used the Lecompte maneuver as part of the repair in three subsequent cases, one with type II truncus arteriosus and two with a type B interrupted aortic arch. The Lecompte maneuver has been helpful in avoiding right pulmonary artery compression, in combined truncus and interrupted aortic arch repair.

We were unable to analyze the durability of conduits placed between the right ventricle and pulmonary artery because of the small number of patients. There was a single case with aneurysmal degeneration of a homograft conduit placed in the pulmonary artery position. Three months after initial repair, this patient had conduit replacement with another homograft, which has been satisfactory thus far.

In conclusion, complete early repair of truncus arteriosus can be accomplished with a minimal risk of mortality even in the presence of associated lesions, such as interrupted aortic arch and marked truncal valve incompetence. Truncal valve repair in the neonate has yielded good early results at short-term follow-up. The Lecompte maneuver is an important component of truncal repair with interrupted aortic arch because it eliminates right pulmonary artery compression. Pulmonary vascular obstructive disease remains an important obstacle to long-term success in patients referred later in life.

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In place of a cognitive examination, candidates for recertification will be required to complete both the general thoracic and cardiac portions of the SESATS VI syllabus (Self-Education/Self-Assessment in Thoracic Surgery). It is not necessary for candidates to purchase SESATS VI booklets prior to applying for recertification. SESATS VI booklets will be forwarded to candidates after their applications have been accepted.

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