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# Repair of Tetralogy of Fallot With Absent Pulmonary Valve Using a New Approach

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**Although several techniques for the correction of tetralogy of Fallot (TOF) with absent pulmonary valve have been described, the method of choice is controversial. Symptomatic infants, in particular, have a poor prognosis because of severe central pulmonary artery dilatation and bronchial compression. An alternative technique for primary repair of TOF with absent pulmonary valve syndrome is suggested. Apart from correction of TOF, this approach includes translocation of the pulmonary artery anterior to the aorta and away from the airways. This technique has a potential to reduce or eliminate bronchial compression by the pulmonary artery. Insertion of valve homograft with anterior and/or posterior plication of the pulmonary artery is considered especially in symptomatic newborns and infants. The intermediate-term functional outcomes have been encouraging, with zero mortality events even in the youngest age group of patients and with the disappearance of respiratory symptoms in the majority of them.**
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Tetralogy of Fallot (TOF) with absent pulmonary valve syndrome (APVS) is physiologically distinctive from other forms of TOF because of tracheobronchial compression resulting from massive dilatation of the main pulmonary artery (PA) and its first- and second-order branches and from the abnormal branching of segmental arteries.[1] Consequential tracheomalacia and bronchomalacia determine the timing and severity of respiratory compromise and the morbidity and mortality of these patients. Mostly in symptomatic infants, mortality remains considerable. In the 2004 Pediatric Cardiac Surgery Annular, Kirshbom[2] elaborated in detail all aspects of TOF-APVS. Surgical modification of the traditional approach that could eliminate or reduce compression of the tracheobronchial tree is described.

## Surgical Background

A number of surgical techniques for reduction of bronchial obstruction have been proposed with questionable results. The method of choice, especially in symptomatic newborns and infants, is controversial. All strategies have focused on plication and reduction of the anterior or posterior wall of the normally positioned PA with or without pulmonary valve replacement.[3-7] An alternative approach is to bring the PA anterior to the aorta and away from the tracheobronchial tree.[8,9]

## Surgical Technique

After median sternotomy, a subtotal thymectomy is performed, and the pericardium is opened. The ascending aorta, aortic arch, and brachiocephalic vessels are widely mobilized. The superior vena cava (SVC) is dissected free, and the azygos vein is transected to improve the mobility of the SVC. Cardiopulmonary bypass is established. During cooling, the left and right PAs, including the first PA branches in the hilum of each lung, are dissected free and mobilized. At a rectal temperature of 28°C, the distal ascending aorta is clamped, and cold cardioplegia is delivered.

Repair of TOF is done first. A vertical incision is made in the infundibular portion of the right ventricle with a short transannular extension of this incision. A limited amount of the infundibular septum (parietal band) is transected. The ventricular septal defect is closed with a polyethylene terephthalate fiber (Dacron) patch using a continuous suture technique and transatrial approach. A small patent foramen ovale

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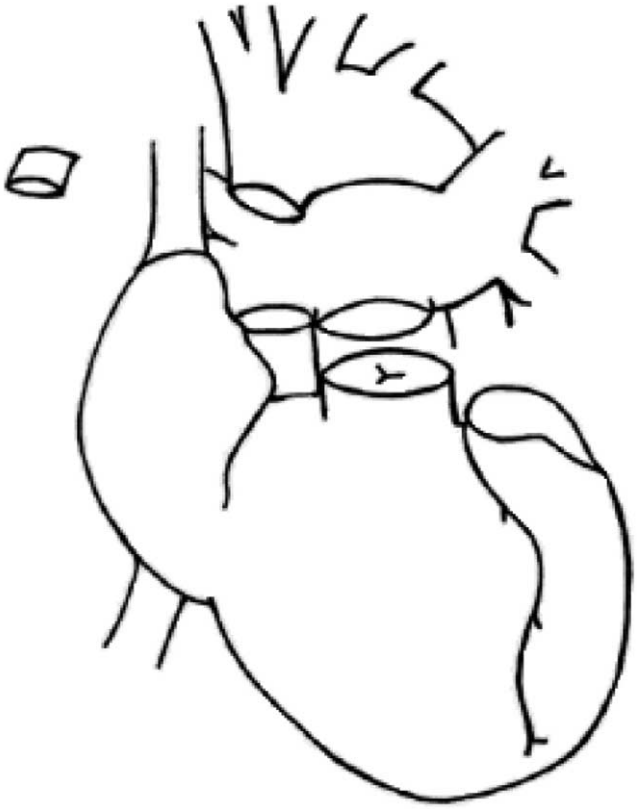

**Figure 1** A short segment of aorta is resected. The pulmonary artery is transected above the annulus.

is left open to allow right-to-left decompression if some degree of right-sided failure develops in the early postoperative period.

After TOF repair, a transverse aortotomy is performed above the commissures, and a short tubular or triangular segment of the aorta is resected (Fig 1). This maneuver brings the ascending aorta down and to the left. The PA is transected above the annulus and brought anterior to the aorta.[3] An end-to-end anastomosis of the ascending aorta is performed. Finally, a direct connection between the PA and the right ventricular outflow tract is accomplished (Fig 2). Complementary anterior/posterior PA plication or homograft insertion can be done, if necessary. The aortic cross-clamp is released, and rewarming is begun.

There are several technical pitfalls to keep in mind during the performance of this procedure. It is essential to gain adequate room between the SVC and ascending aorta for the translocated right PA. In addition to SVC mobilization, appropriate shortening of the ascending aorta, if necessary, allows the aorta to reside posteriorly and to the left of its usual location. This maneuver calls for a thorough mobilization of the aortic arch and brachiocephalic vessels. Shortening of the ascending aorta and mobilization of the PA beyond pericardial reflection avoids the potential compression of the right coronary artery and the SVC. On the other hand, it is not always necessary to make the aorta shorter and to risk too close a relationship of the ascending aorta with the trachea. Another relevant detail is shortening of the left PA (which is always too long) by oblique transection of the PA trunk with connection to the right ventricular outflow tract (RVOT).

## Results

Since 1998, seven patients with TOF/APV have been operated on using this approach. All patients had severe respiratory problems and congestive heart failure with cyanosis. Surgery in neonates was performed on a semi-urgent basis due to serious respiratory compromise. Persistent bronchopulmonary infection requiring prolonged and repeated hospital admission and failure to thrive were the main indications for surgery in infants. No patient required extracorporeal membrane oxygenation before surgery. Median age at surgery was 35 days (range 14 to 256 days). Median weight at the time of operation was 3.5 kg (range 2.9 to 4 kg). Three patients underwent neonatal repair. The RVOT was reconstructed with a valved homograft in two newborns; a monocusp valve was used in one infant. Combined anterior and posterior plication of the pulmonary artery was performed in four patients.

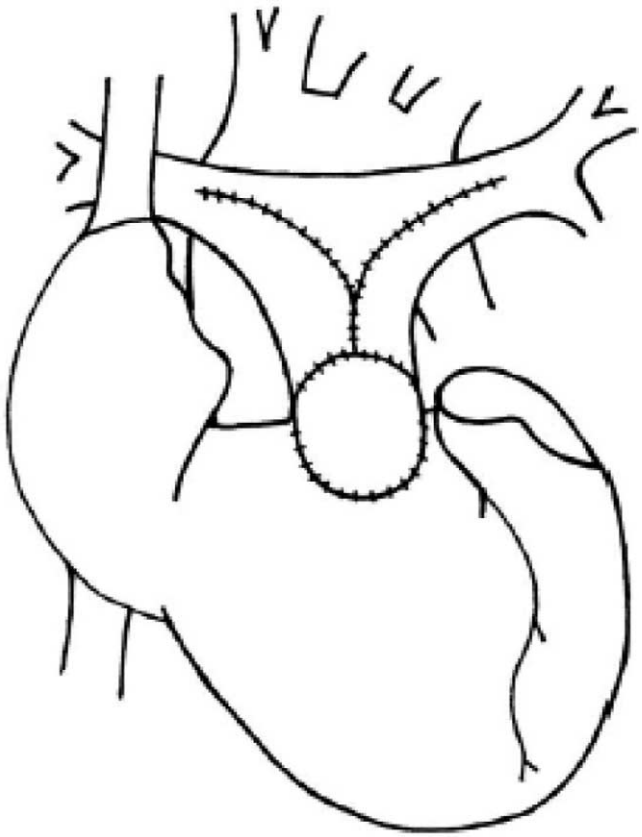

**Figure 2** The pulmonary artery is brought anterior to the aorta. The direct connection between the pulmonary artery and right ventricular outflow tract, or homograft insertion, is accomplished. Anterior and/or posterior plication of the pulmonary artery can decrease wall tension and prevent development of aneurysmal dilatation of the pulmonary artery.

### Early Results

There was no early death. Hemodynamically, the postoperative course was generally uneventful. The duration of mechanical ventilation varied from 2 to 13 days (median 7 days), and the intensive care unit stay varied from 9 to 25 days (median 12 days).

### Late Results

There was no late death. Three months after neonatal repair, one patient required reoperation for failure to thrive. A valved conduit was inserted into the pulmonary position. His postoperative course was uneventful. At median follow-up of 3.8 years, respiratory symptoms had completely disappeared (five patients) or were significantly reduced. Postoperative CT scans showed no compression of the trachea and main bronchi, and the pulmonary artery was at a distance to the tracheobronchial tree in all patients.

## Discussion

Controversy persists regarding the management of patients with TOF/APV. The mortality for symptomatic newborns and infants remains considerably secondary to airway obstruction by dilated PA.[2,5-7] Massively dilated right and left PAs up to the hilum are amenable to surgical intervention. Unfortunately, abnormalities of arborization, with tufts of arteries encircling and compressing the intrapulmonary bronchi, cannot be addressed during surgery.[1] This could partially explain the high rate of failure with the treatment of the youngest, symptomatic group of patients.

The suggested technique applies a well-known Lecompte maneuver.[8] Translocation of the PA anterior to the aorta displaces the dilated the PA anterior away from the trachea and bronchial tree.[9,10] Insertion of a valve homograft with anterior and posterior plication of the PA can decrease wall tension and prevent the development of aneurysmal dilatation of the PA. This management strategy is consistent with the report of McDonnell et al[6] who recommended not only anterior and posterior plication of PA but also insertion of a homograft, especially in newborns and infants.

Another alternative approach is removal of the entire main pulmonary artery and placement of a bifurcated pulmonary homograft.[7] This technique decreases the size of the pulmonary vessels, minimizing potential compression of the bronchi.

There is no consensus on the timing of surgery for babies presenting with TOF+APVS. Early primary repair should be considered. Symptomatic patients need to proceed directly to surgery. Early repair in asymptomatic patients can eliminate the potentially harmful effect of a dilated PA on the tracheobronchial tree.[6,7,10]

## Summary

In 1998 we started to use a new technique that has the potential to eliminate or reduce bronchial compression by the PA. Apart from correction of TOF, this approach includes translocation of the PA anterior to the aorta and away from the airways. Insertion of valve homograft with anterior and/or posterior plication of the PA is always considered, especially in symptomatic newborns and infants.

The intermediate-term functional outcomes have been encouraging, with zero mortality events even in the youngest age group of patients and with the disappearance of respiratory symptoms in the majority of them.

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