Operative Techniques in
Thoracic and
Cardiovascular Surgery

Absent Pulmonary Valve Repair

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The absent pulmonary valve syndrome (APVS) occurs in 3 to 6% of patients with tetralogy of Fallot (TOF). This syndrome is physiologically distinctive from other forms of TOF because of tracheobronchial compression resulting from massive dilation of the main pulmonary artery (PA) and its first- and second-order branches, and from the abnormal branching of segmental pulmonary arteries. Consequential tracheomalacia and bronchomalacia determine the timing and severity of respiratory compromise as well as the morbidity and mortality of these patients. Clinical presentation reflects the degree of respiratory distress secondary to airway obstruction and infections and heart failure as a result of the left to right shunt.

Controversy persists regarding the management of patients with TOF/APVS. The mortality of symptomatic newborns and infants remains considerable, secondary to airway obstruction by the dilated PA. 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.

A number of surgical techniques for reduction of bronchial obstruction have been proposed, with variable results. The method of choice, especially in symptomatic newborns and infants, is still 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.[2-4] An alternative approach is to bring the PA anterior to the aorta. Translocation of the PA anterior to the aorta displaces the dilated PA away from the trachea and bronchial tree.[5]

The illustrated approach incorporates the standard intracardiac portion of TOF repair (resection of right ventricular tract (RVOT) obstruction, patch closure of the ventricular septal defect) and translocation of the pulmonary artery anterior to the aorta. Standard cardiopulmonary bypass, moderate hypothermia, and cold crystalloid cardioplegia are used. 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 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.

Repair of the TOF is performed first. A short vertical incision is made in the infundibular portion of the right ventricle with a transannular extension of this incision. A limited amount of the infundibular septum (parietal band) is transected. The ventricular septal defect is closed with a knitted velour patch using a continuous suture technique and transatrial approach. A small patent foramen ovale is left open to allow right-to-left decompression if some degree of right-sided failure develops in the early postoperative period.

A transverse aortotomy is performed above the commissures, and a short tubular segment of the aorta is resected. This maneuver brings the ascending aorta down and to the left. The PA is transected above the annulus and brought anterior to the aorta. At this point, 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. Complementary anterior/posterior PA plication or homograft insertion can be done, if necessary.

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 ultimately 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 the pericardial reflections avoid 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 to 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 RVOT.

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Operative Technique

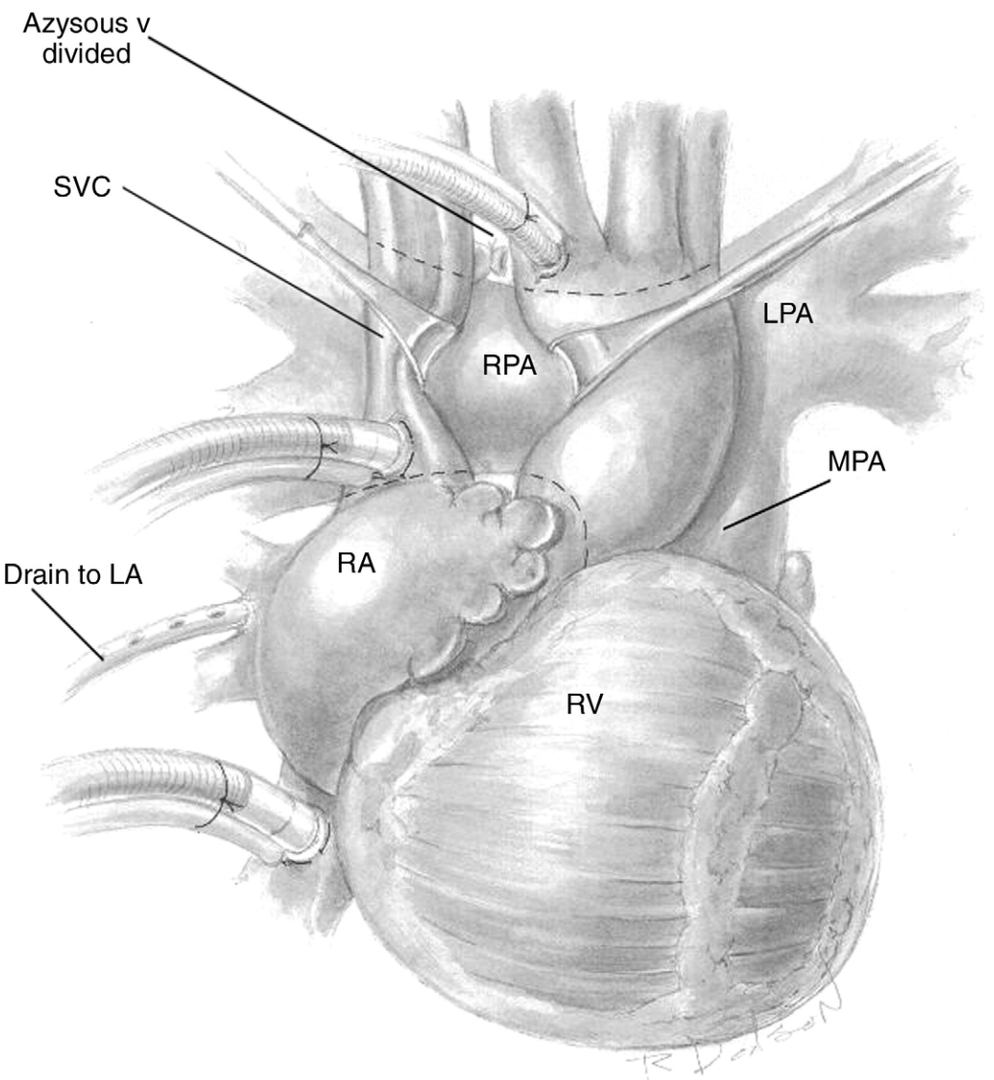

Figure 1 Before cardiopulmonary bypass is commenced, the aorta, proximal arch, and head vessels are widely mobilized. Standard cardiopulmonary bypass is instituted with direct bicaval cannulation. The aorta is cannulated as high as possible. A left atrial vent is inserted through the right pulmonary veins. Moderate hypothermia with cold crystalloid cardioplegia is applied. The SVC is dissected free and the azygos vein is transected to improve mobility of the SVC. The left and right pulmonary arteries, including the first pulmonary artery branches in the hilum of each lung, are dissected free and mobilized. Care is taken to stay away from the phrenic nerve on both sides. Traction of the aorta by a small retractor or tape facilitates exposure of both pulmonary arteries. Ao = aorta; LA = left atrium; LPA = left pulmonary artery; MPA = main pulmonary artery; RA = right atrium; RPA = right pulmonary artery; RV = right ventricle; SVC = superior vena cava; v = vein.

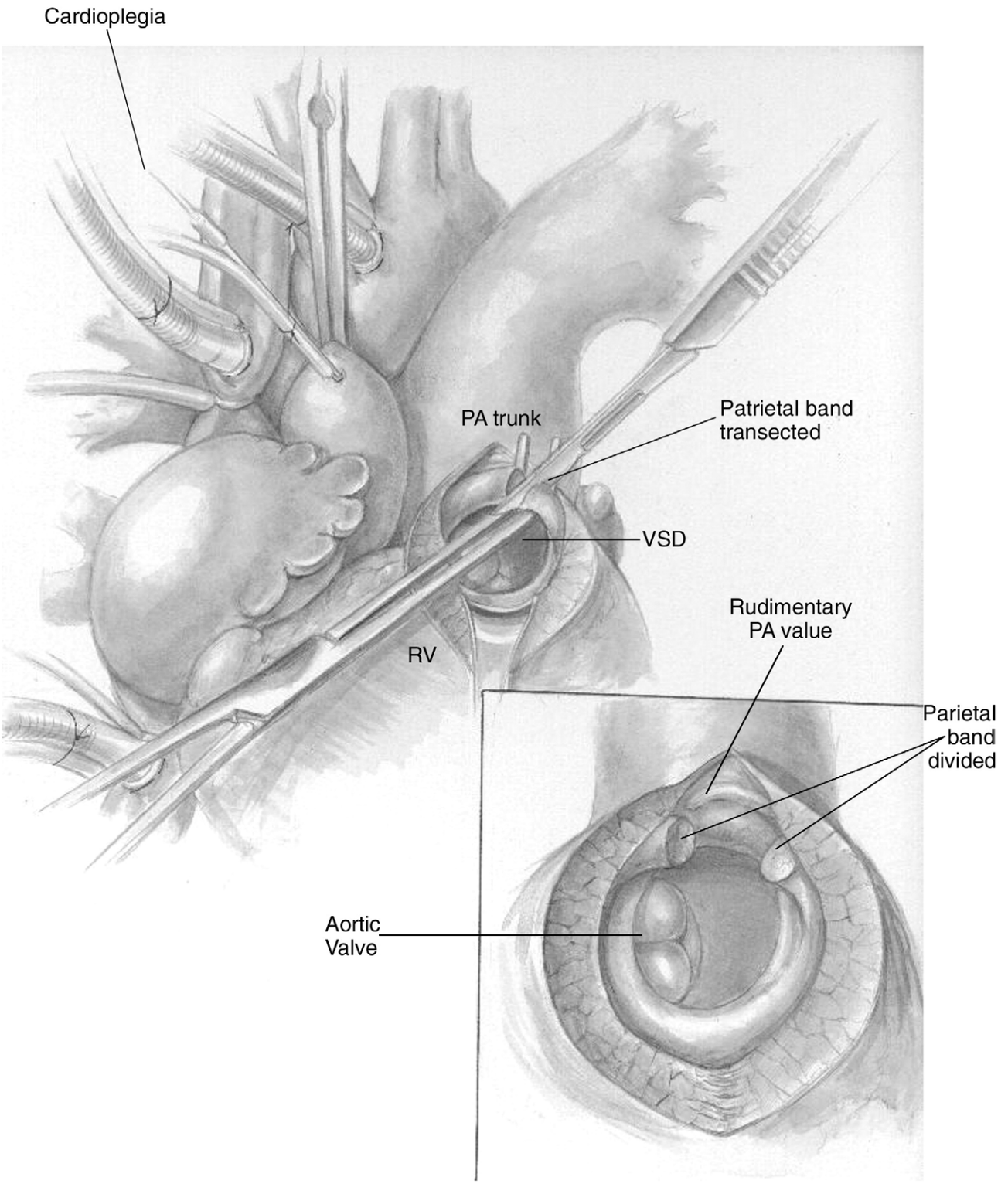

Figure 2 After clamping the aorta and delivering of cardioplegia, a short (10-15 mm) transannular longitudinal incision is made in the right ventricular outflow tract. The parietal band is transected to enlarge the right ventricular outflow tract. PA = pulmonary artery; RV = right ventricle; VSD = ventricular septal defect.

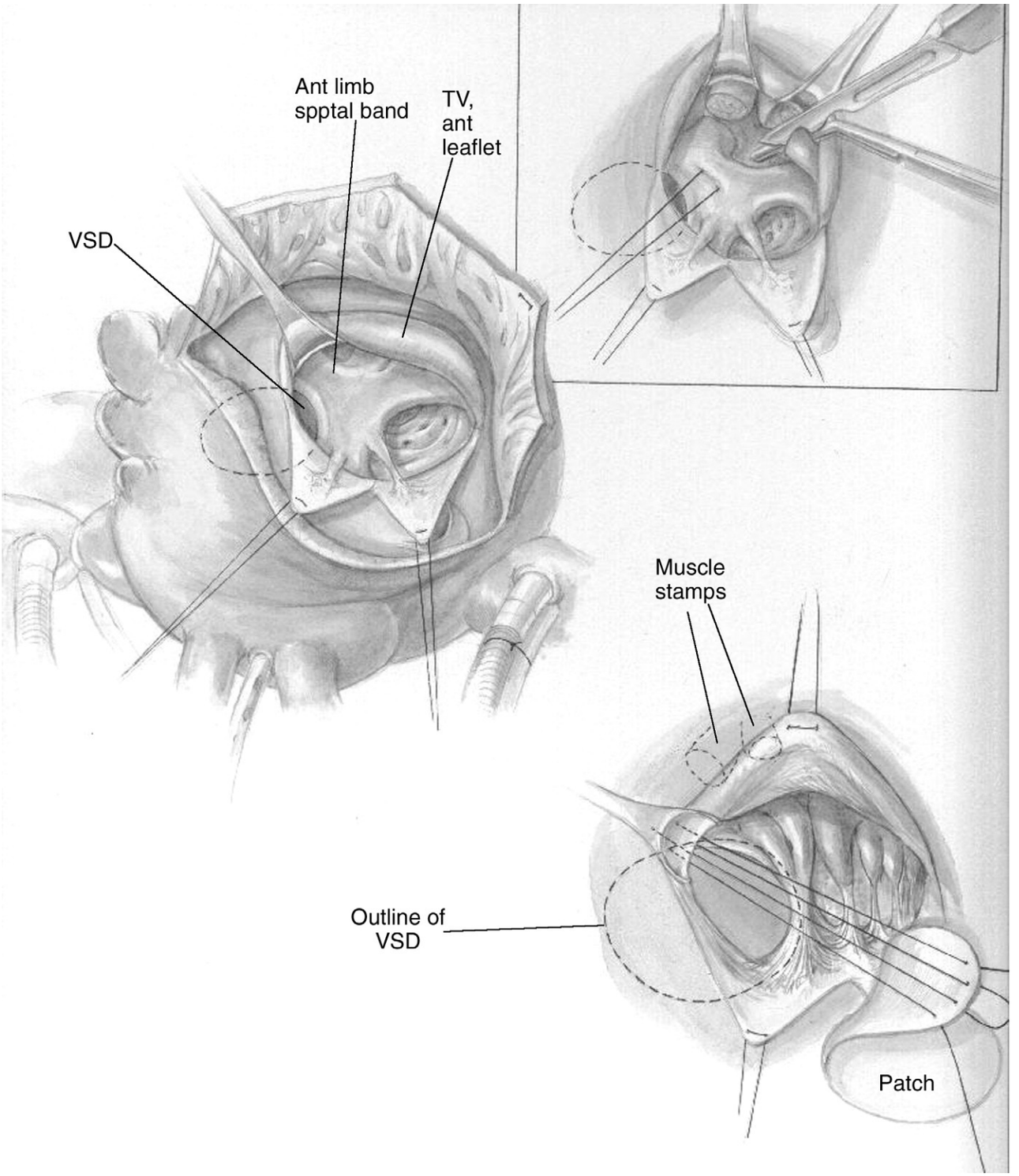

Figure 3 (A) If the parietal band is properly divided, a transatrial approach offers very good exposure for the closure of the ventricular septal defect, even in neonates. If necessary, extended resection of right ventricular outflow tract can be accomplished working through the tricuspid valve as well. (B) A continuous suture technique and Dacron patch are preferred. Alternatively, in neonates with a very fragile myocardium, a pledgeted suture technique should be used. The foramen ovale in newborns and small infants is left open. TV = tricuspid valve; VSD = ventricular septal defect. (Redrawn from Bove.)

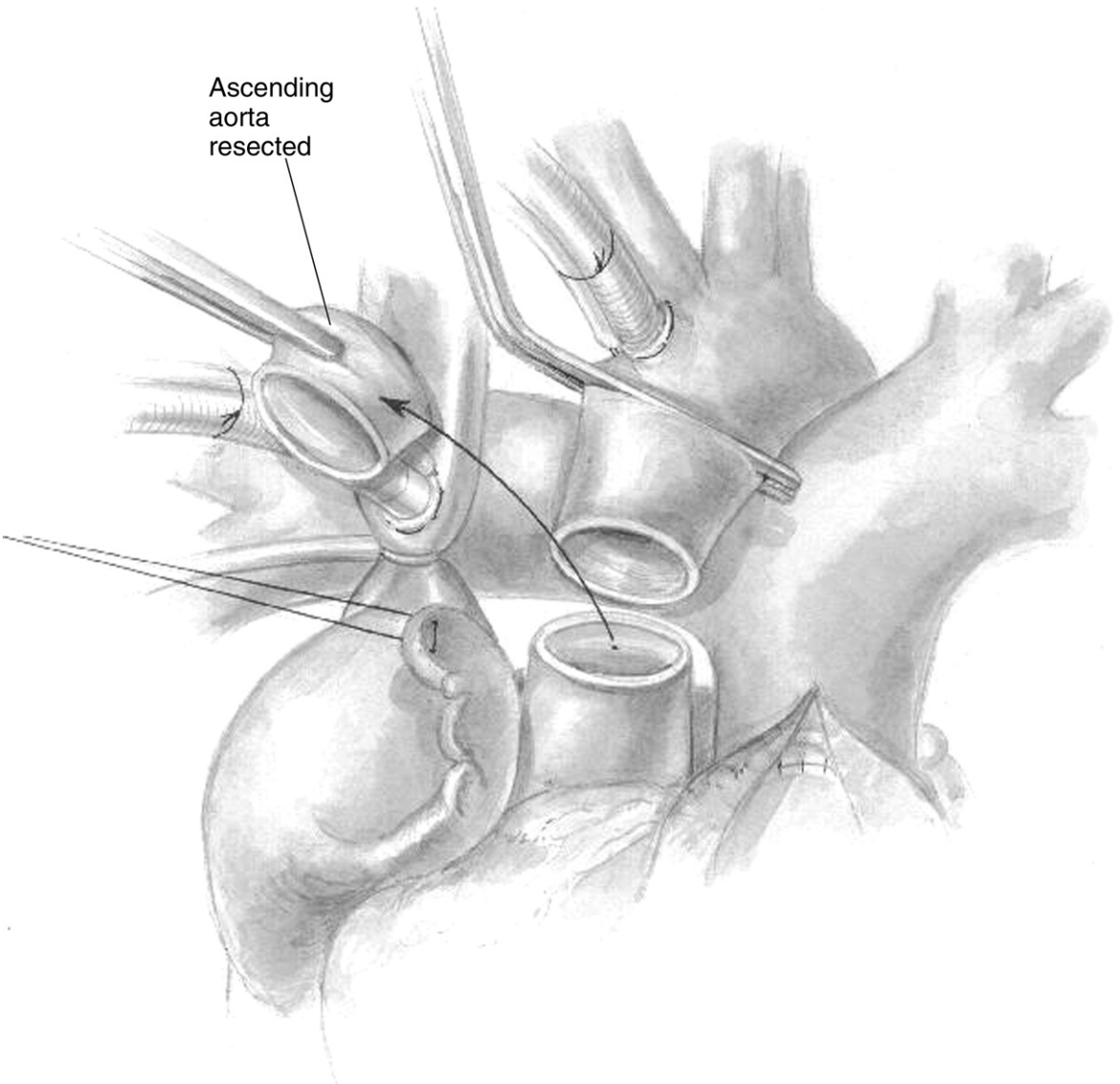

Figure 4 The transverse aortotomy is performed above the aortic valve commissures. A short tubular segment of the aorta is resected. This maneuver brings the future ascending aorta down and to the left. Resection of aorta, especially in newborns, might be omitted, to avoid too close a relationship between the aorta and the trachea and left bronchus.

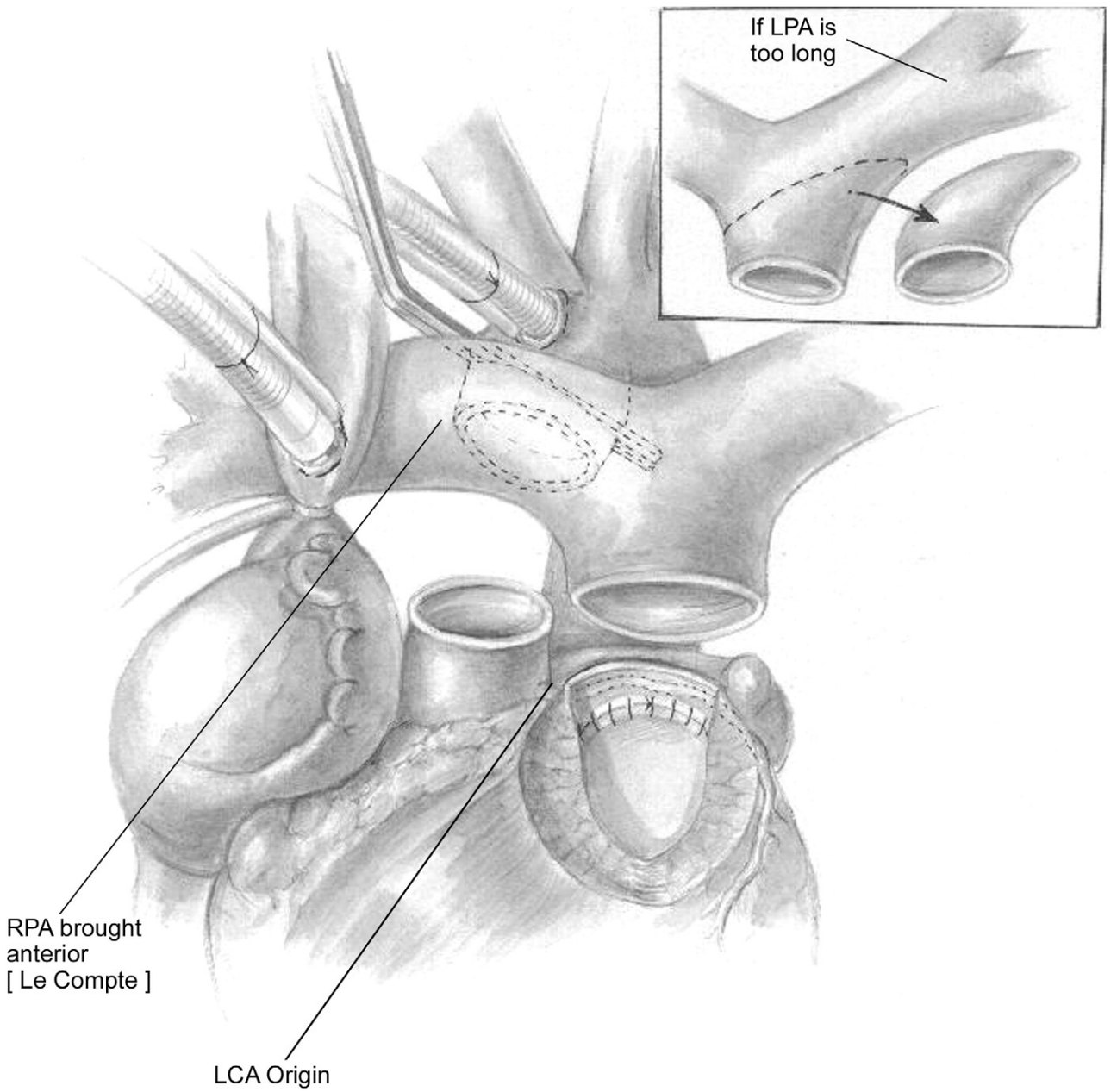

Figure 5 The pulmonary artery is transected above the annulus. Care is taken to stay away from left coronary artery. If the left pulmonary artery is too long, the pulmonary trunk is obliquely cut toward the left pulmonary artery. LCA = left coronary artery; LPA = left pulmonary artery; RPA = right pulmonary artery.

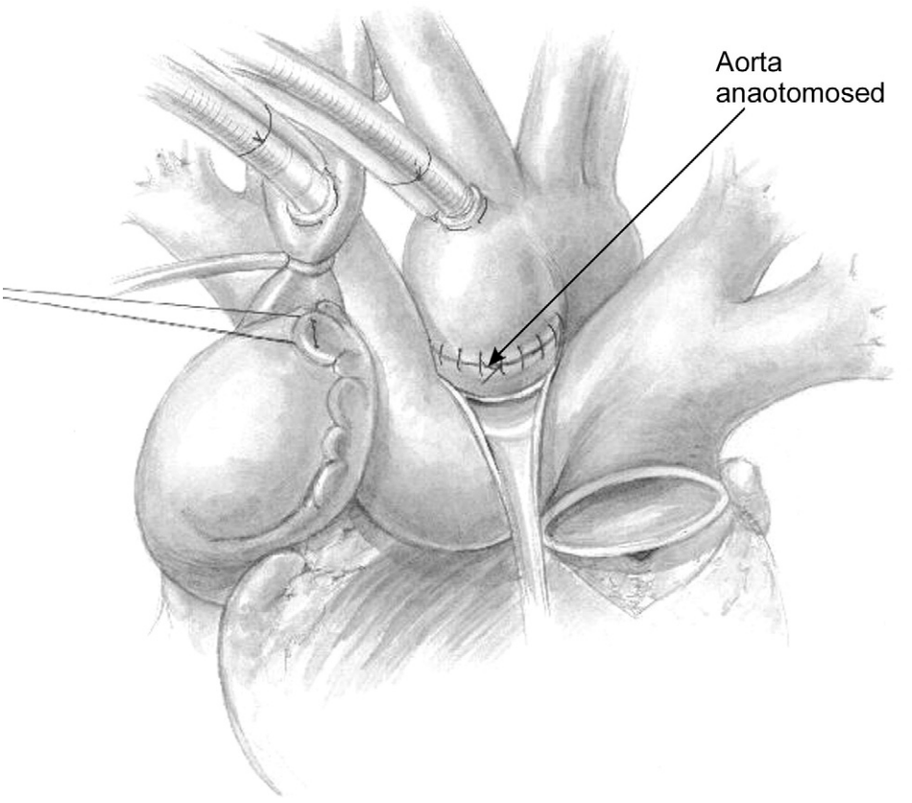

Figure 6 The transected pulmonary artery is mobilized, if necessary, and brought anterior to the aorta. At this point, end-to-end anastomosis of the ascending aorta is performed. Care is taken not to compress the right coronary artery by the translocated pulmonary artery. In particular, the right pulmonary artery must be mobilized adequately to avoid undue tension on the right coronary artery.

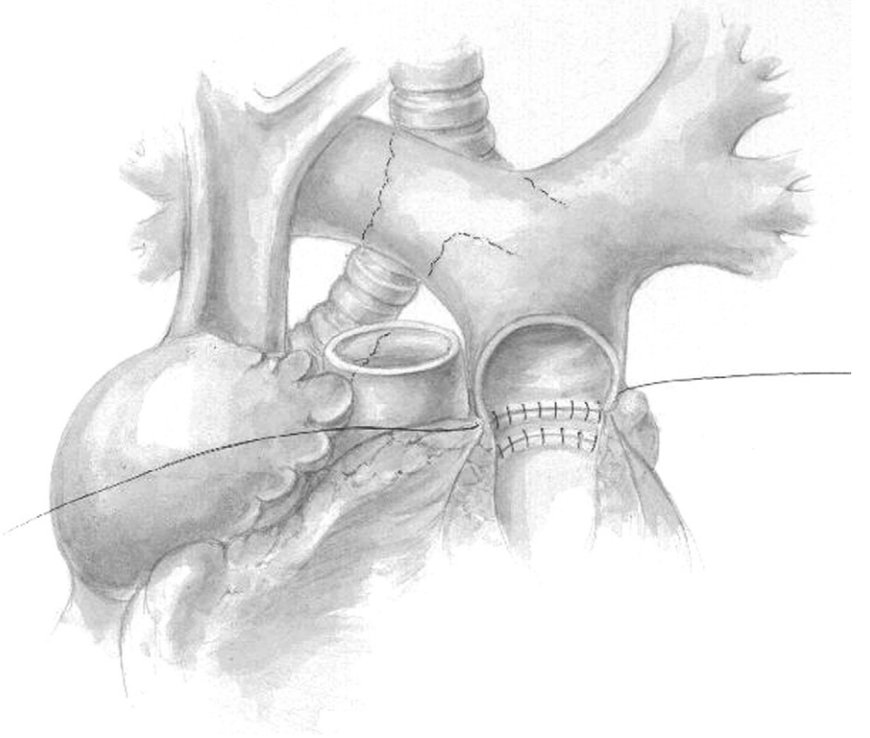

Figure 7 A direct connection between the pulmonary artery and right ventricular outflow tract is accomplished using a continuous suture technique (the aorta is cut off for clarity).

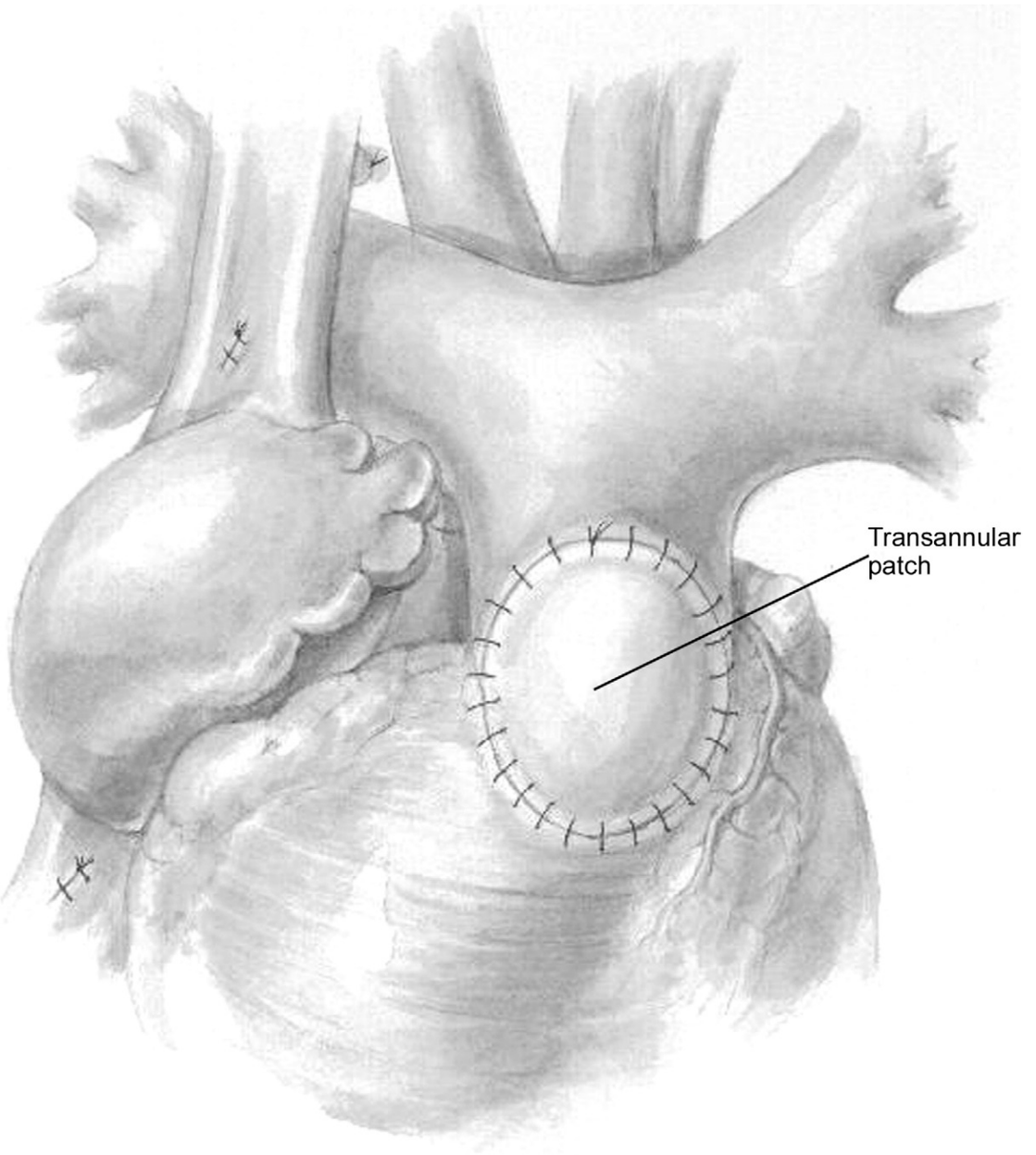

Figure 8 The right ventricular outflow tract is enlarged and reconstructed by use of a transannular pericardial patch treated with glutaraldehyde, with the aim of achieving a normal Z-value for the annulus. The newly created right ventricular outflow tract should have growth potential.

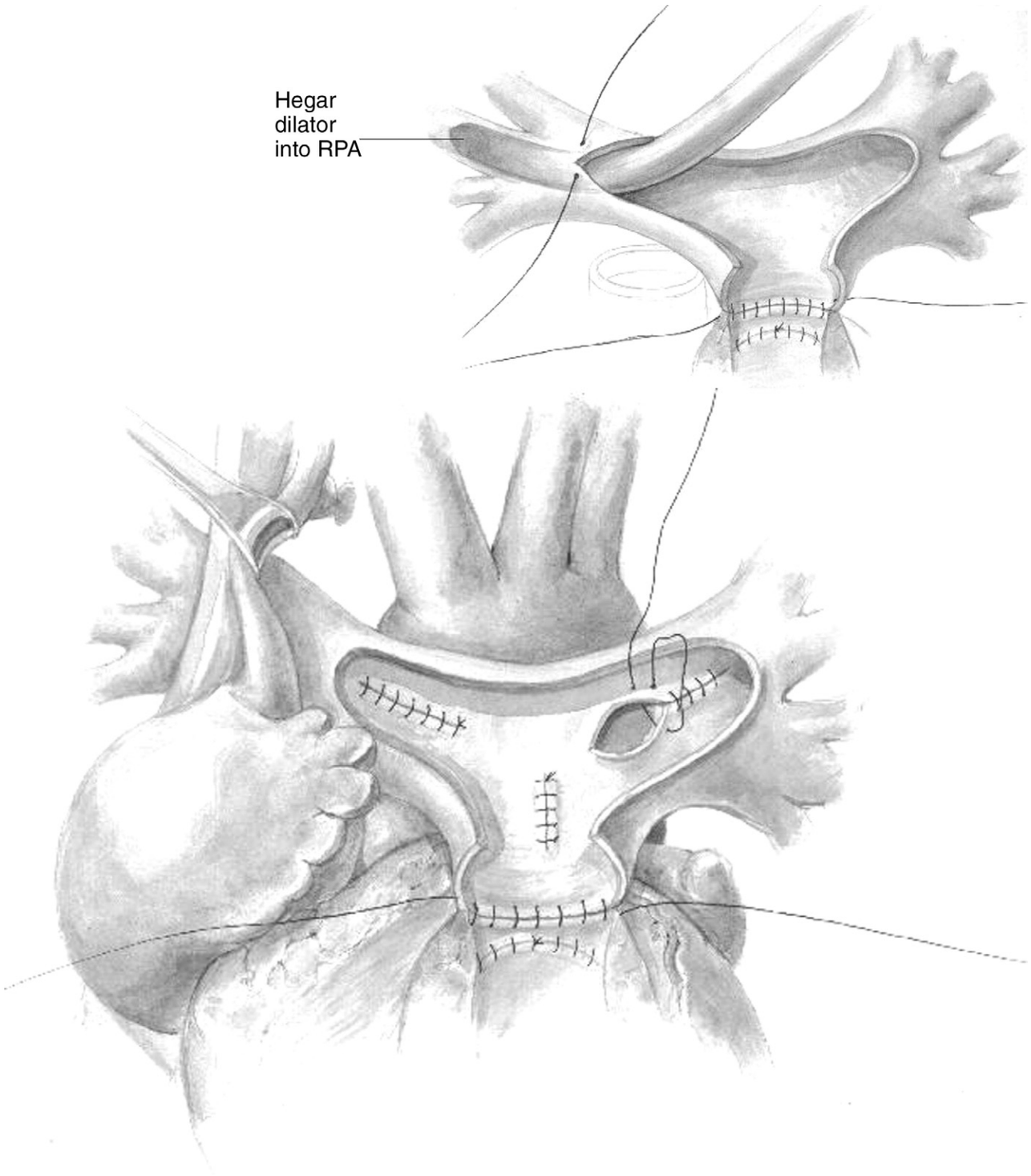

Figure 9 Especially in newborns and small symptomatic infants, anterior plication of the pulmonary artery is performed to decrease wall tension and prevent later development of aneurysmal dilation of the pulmonary artery. Triangular segments of the anterior wall of each branch pulmonary artery and part of the anterior wall of the pulmonary artery trunk are excised. An appropriately chosen Hegar dilator is used to guide the extent of the resection of the pulmonary artery and the magnitude of plication. An anterior wall resection may be combined with posterior wall plications of the main and both pulmonary arteries. RPA = right pulmonary artery.

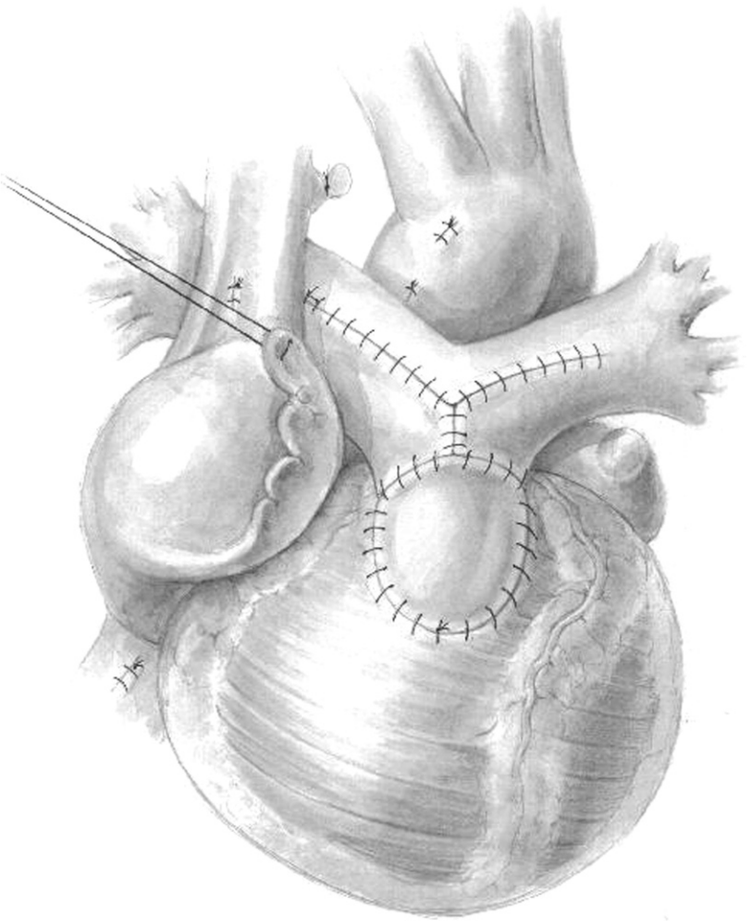

Figure 10 The final outcome of translocation of the pulmonary artery with direct connection to the right ventricle, patch reconstruction of the right ventricular outflow tract, and anterior plication of the pulmonary arteries. If a homograft is not used, the reconstruction has growth potential. The usual monitoring lines and temporary atrial and ventricular pacing wires are always placed.

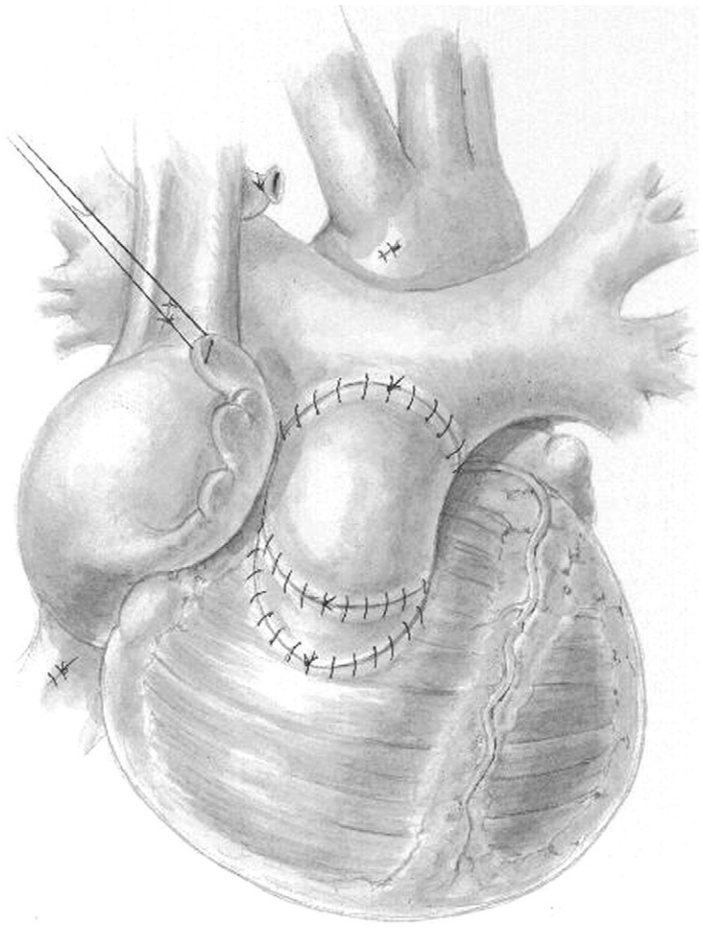

Figure 11 Homograft insertion (recommended in symptomatic newborns) or monocusp valve placement is performed, if necessary. A short pulmonary homograft is orthotopically placed; the proximal anastomosis is supplemented with a roof of autologous pericardium, which has been treated with glutaraldehyde.

Conclusions

Despite improvements in surgical techniques and critical care, controversy persists regarding the management of patients with TOF/APVS. The method of choice, especially in symptomatic newborns and infants, is still controversial. The above-described technique has the potential to eliminate or reduce bronchial compression by the PA. Apart from correction of TOF, this approach applies a well-known technique (Lecompte maneuver) of translocation of the PA anterior to the aorta and away from the airways. Insertion of a valved conduit and plication of the PAs are recommended, especially in symptomatic newborns and infants.

There is no consensus on the timing of surgery for babies presenting with TOF/APVS. 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. Failure of the treatment may occur in symptomatic patients, where the obstruction of the airways extends beyond the proximal PAs.

References

1. Rabinovitch M, Grady S, David I, et al: Compression of intrapulmonary bronchi by abnormally branching pulmonary arteries associated with absent pulmonary valves. Am J Cardiol 50:804-813, 1982
2. Godart F, Houyel L, Lacour-Gayet F, et al: Absent pulmonary valve syndrome: surgical treatment and consideration. Ann Thorac Surg 62: 136-142, 1996
3. Conte S, Serraf A, Godart F, et al: Technique to repair tetralogy of Fallot with absent pulmonary valve. Ann Thorac Surg 63:1489-1491, 1997
4. Donnell BE, Raff GW, Gaynor JW, et al: Outcome after repair of tetralogy of Fallot with absent pulmonary valve. Ann Thorac Surg 67:1391-1396, 1999
5. Hraska V: A new approach to correction of tetralogy of Fallot with absent pulmonary valve. Ann Thorac Surg 69:1601-1603, 2000