

Repair of Truncus Arteriosus

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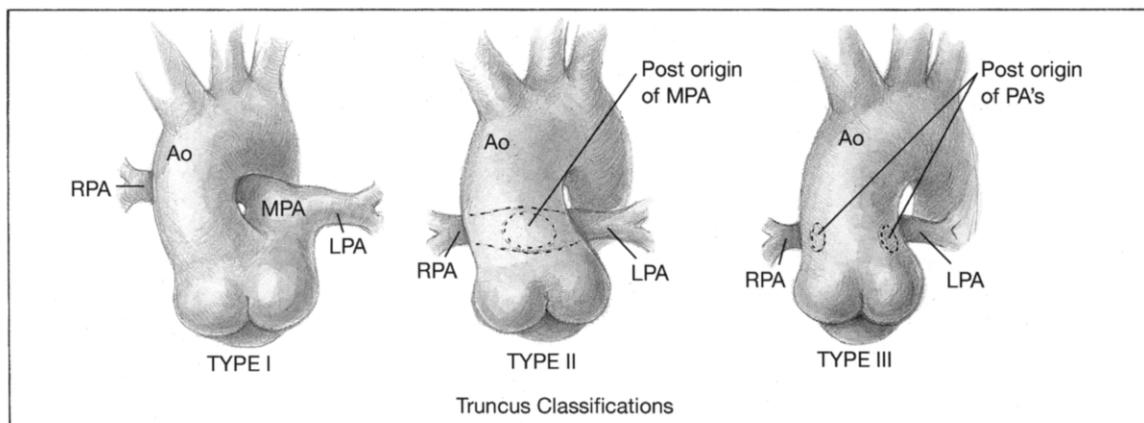
The repair of truncus arteriosus requires separation of the pulmonary artery bifurcation from the single arterial trunk, closure of the ventricular septal defect, and establishment of an unobstructed pathway from the right ventricle to the pulmonary arteries. This operation is now performed routinely in newborns at clinical presentation, which is usually within the first days or weeks of life. Waiting for the infant to grow is no longer advised. It is now recognized that leaving the pulmonary vascular bed unprotected for even a few weeks results in an augmented vasoconstrictor response that significantly increases the risk of acute pulmonary hypertensive crises during the postoperative period.¹ Significant improvements in the perioperative treatment of neonates requiring complex cardiac surgery have also contributed to a marked decrease in morbidity and mortality in truncus arteriosus repair.

Improved survival rates for these infants have also resulted from recognition and improved treatment of significant risk factors: (1) truncal valve insufficiency, (2) coronary artery anomalies, and (3) the presence of an interrupted aortic arch.² Moderate or severe truncal valve insufficiency is now better treated with techniques of valve repair rather than replacement.³⁻⁵ Recognition of the potential presence of coronary artery anomalies in infants with truncus helps avoid inadvertent injury to these vessels during separation of the pulmonary arteries from the truncal root or during the right ventriculotomy.⁶ The presence of a concomitant interrupted aortic arch has also become less of a risk factor as newer regional perfusion techniques permit

arch reconstruction without a period of circulatory arrest.^{2,3}

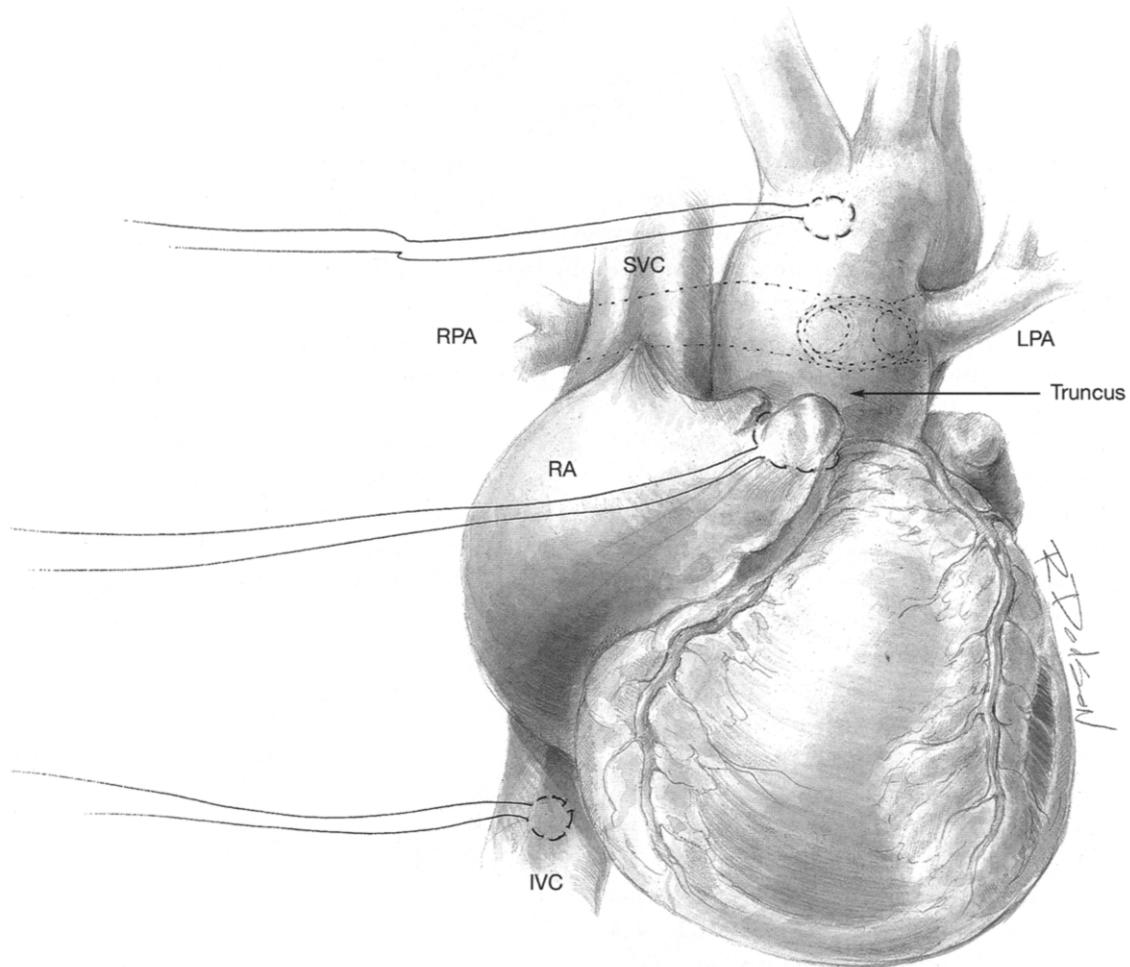
A variety of techniques for establishing continuity between the right ventricle and the pulmonary artery bifurcation has been described. Ideally, the connection should be widely patent, have potential for growth, and provide valve function, especially during the early postoperative period when pulmonary hypertensive crises are most likely. Although techniques of direct anastomosis between the right ventricle and pulmonary artery potentially answer these ideals, experience in comparison with conduits has not always been favorable.⁷⁻⁹ Historically, Dacron () valved conduits have served as satisfactory connections, especially for older and larger infants. However, the stiffness of the Dacron graft material limits its suitability for anastomosis to the fragile neonatal right ventricle.

Our choice of conduit for repair of truncus arteriosus has been the cryopreserved pulmonary allograft. Despite its limited availability in small sizes, the pulmonary allograft provides excellent handling properties, a low resistance to ejection, and a competent valve. Although tissue allografts are essentially inert and do not have true growth potential, the relatively thin-walled pulmonary allograft often dilates enough to permit the infant to reach an age of 2 years or older before a larger conduit is required. When a suitably sized, pulmonary allograft is unavailable, our next choice is an aortic allograft, which typically has a thicker wall, and is more likely to calcify and require earlier replacement.¹⁰

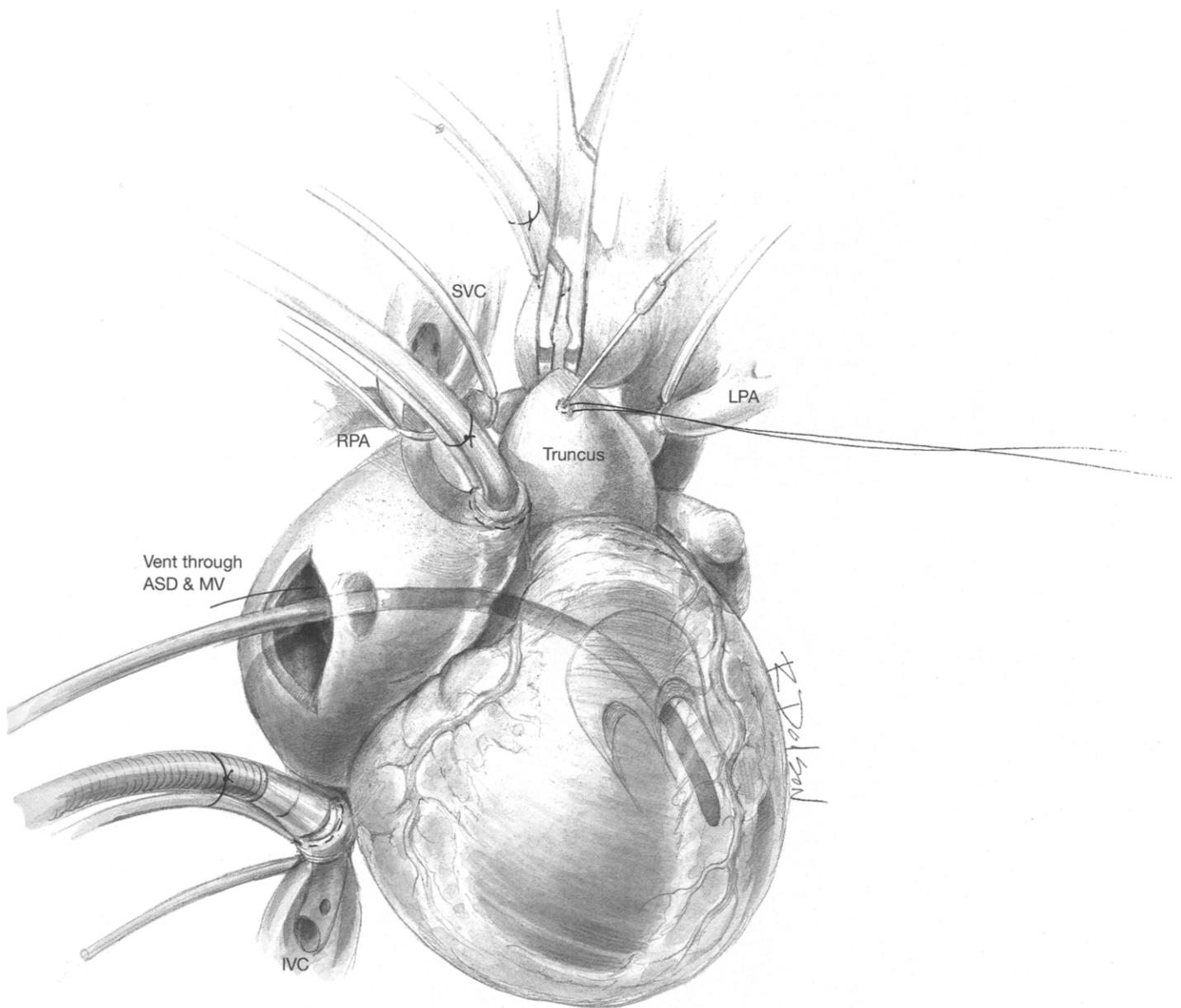


1a The origin of the pulmonary arteries is noted and conceptualized according to a classification such as that of Collet and Edwards (Figure 1a).¹¹

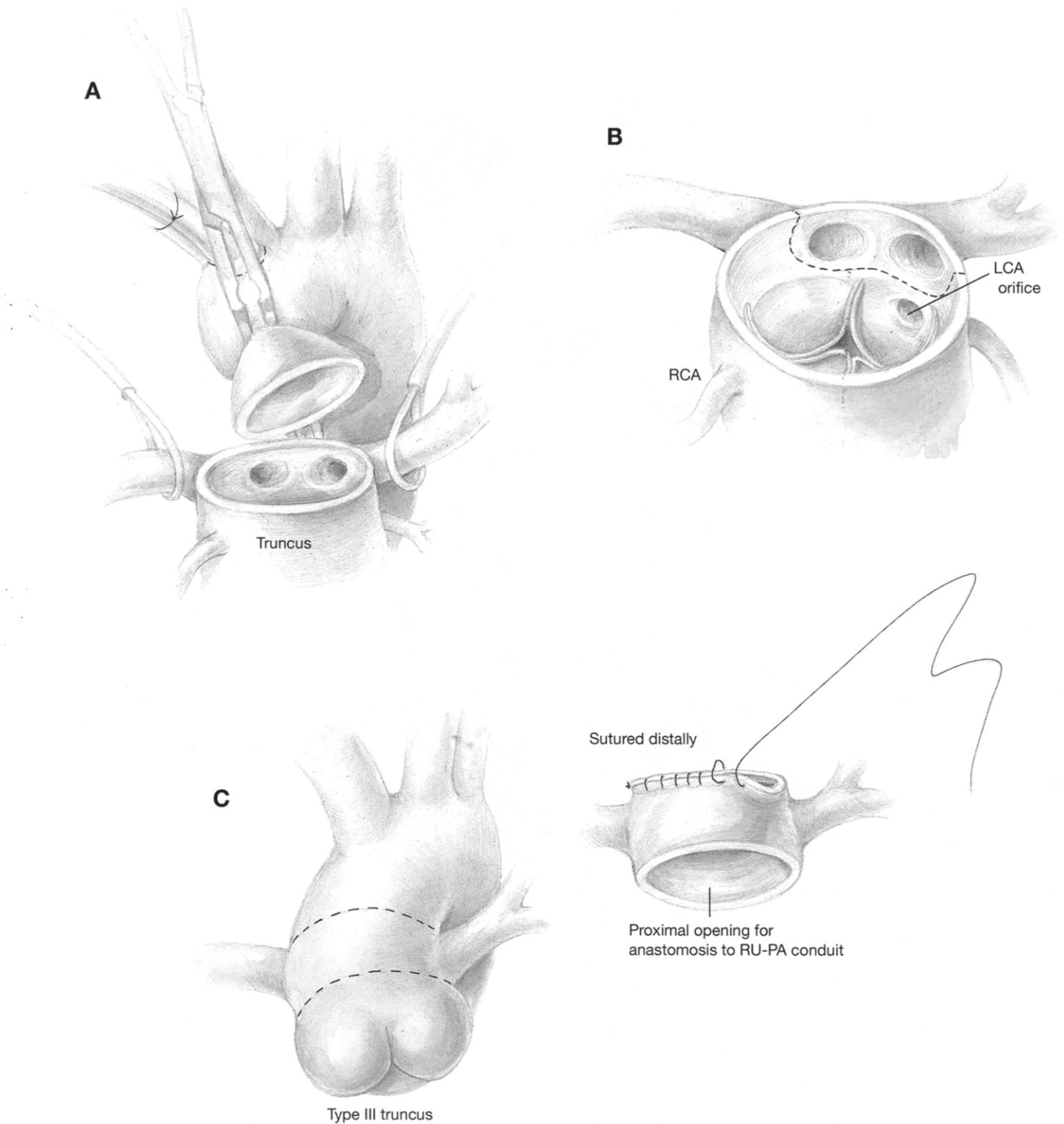
SURGICAL TECHNIQUE



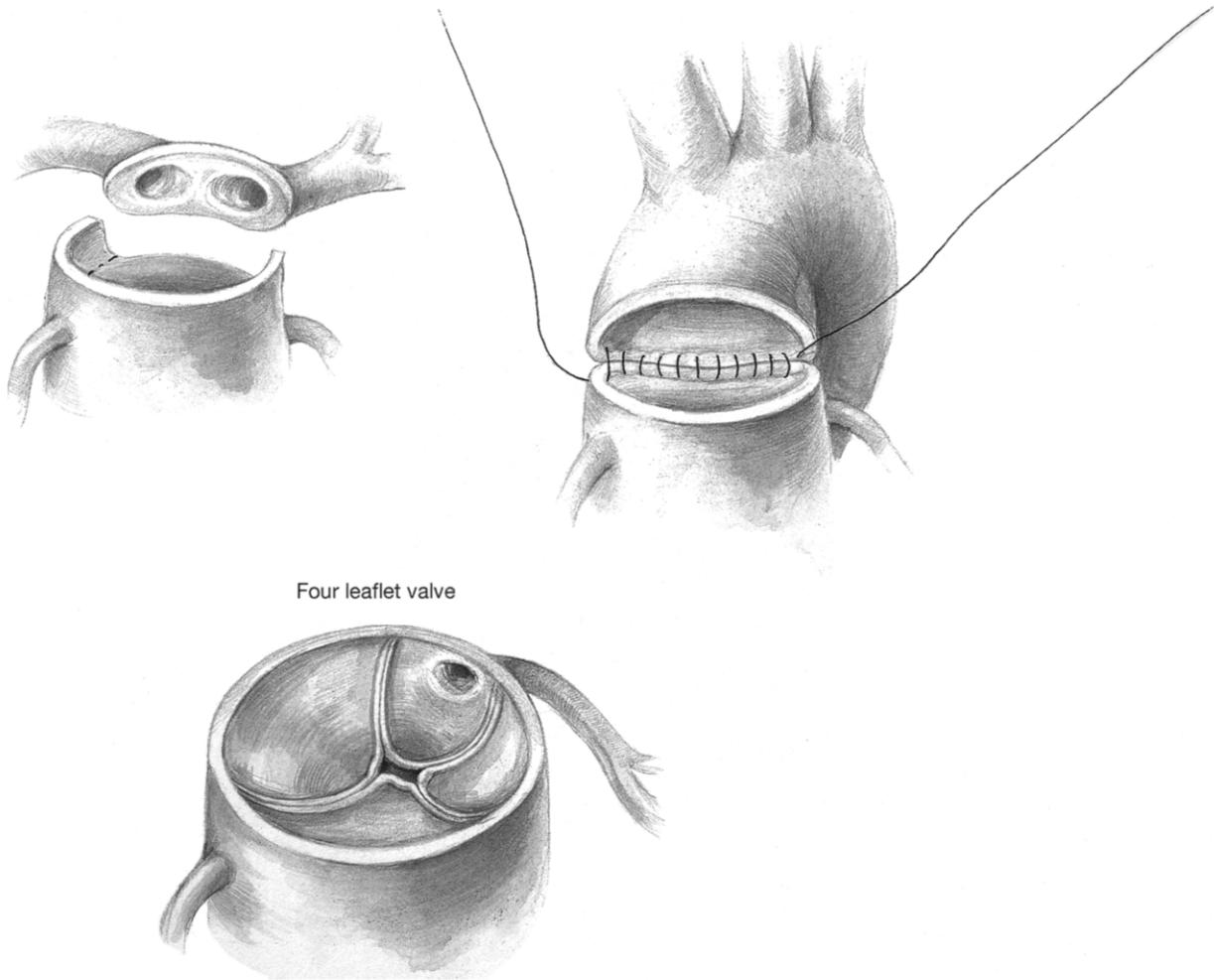
I Through a standard median sternotomy, the presence of thymic tissue is noted and, if present, partially or totally resected to optimize exposure. The pericardium is opened to the right of the midline, and a piece of pericardium is harvested and set aside for a 10-minute treatment with 0.9% glutaraldehyde. Sufficient autologous pericardium is taken for patch closure of the ventricular septal defect and for hood extension of the pulmonary homograft. In many cases, the pulmonary arteries show a type I½ configuration, as depicted here, with the confluence of the right and left branch pulmonary arteries coming off posteriorly and leftward, with a very short common trunk. An arterial purse-string is positioned at the base of the innominate artery to allow adequate space between the aortic cross-clamp and the pulmonary bifurcation. Purse-string sutures are placed in the right atrial appendage and inferior vena cava for double venous cannulation and continuous cardiopulmonary bypass. Although some surgeons prefer using circulatory arrest for this repair, excellent exposure with a near bloodless field can be provided without suspending circulatory support. Before initiating cardiopulmonary bypass, the left and right branch pulmonary arteries are dissected, and tourniquets are placed loosely. One of the branch pulmonary arteries can be snared before initiating bypass if the patient appears to be unstable from pulmonary overcirculation.



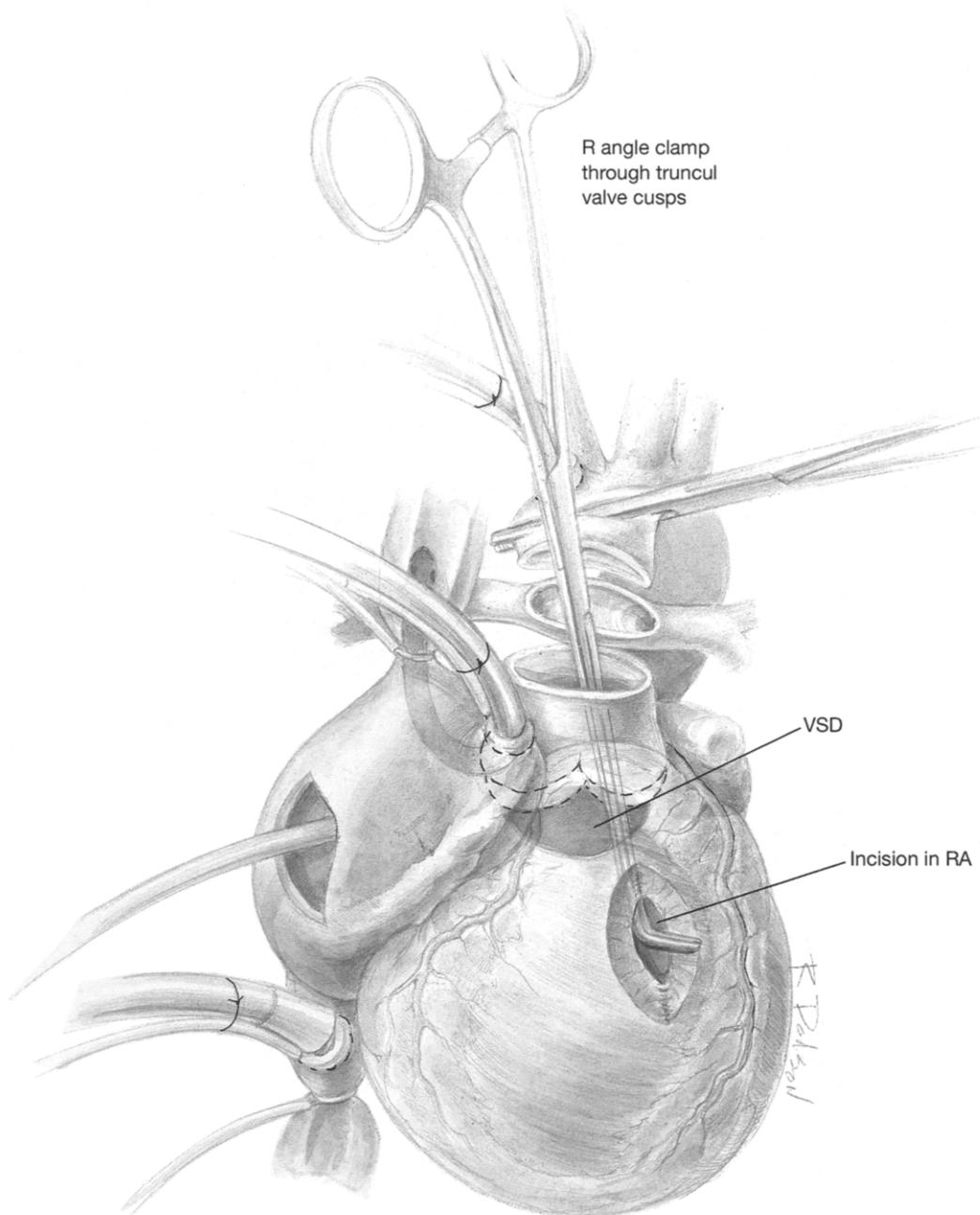
2 The patient has been placed on full-flow cardiopulmonary bypass, and the snares on the branch pulmonary arteries have been tightened. The infant is cooled to 28°C. One venous cannula has been advanced into the superior vena cava, and an angled venous cannula has been directed into the inferior vena cava. The aortic cross-clamp is applied, and 40 mL/kg of cold blood cardioplegia is administered into the truncal root. As the cardioplegia is being administered, caval snares are tightened, and a small right atriotomy is performed to expose the patent foramen ovale or atrial septal defect. A small vent is advanced through the interatrial septum and mitral valve into the left ventricle. When truncal valve insufficiency prevents satisfactory antegrade cardioplegic arrest, the coronary sinus is visualized through the atriotomy, and retrograde cardioplegia is administered.



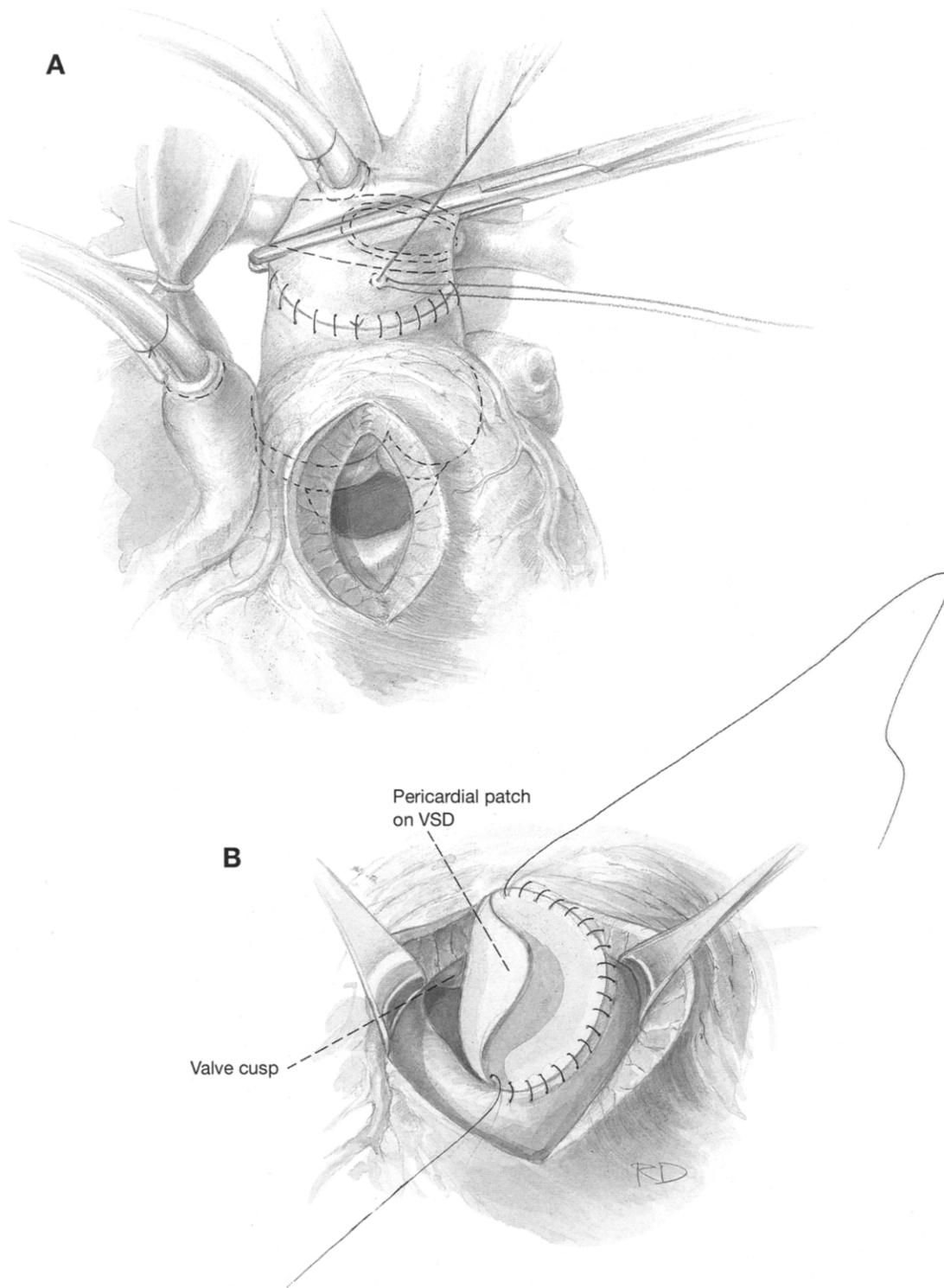
3 Although, classically, the pulmonary artery trunk or bifurcation is simply excised from the side of the truncal root, we prefer complete transection of the ascending aorta for optimal visualization. Before making any incision in the truncus, the origin of the coronary arteries must be assessed to prevent injury of an anomalously positioned vessel. The anterior portion of the transection is then performed, and the origins of the branch pulmonary arteries are identified from inside the truncal root. Transection is then completed posteriorly, leaving an adequate rim of tissue above the orifice of the pulmonary bifurcation. At this time, the surgeon carefully assesses the position of the coronary ostia and evaluates the truncal valve. With these vital structures in sight, the surgeon can carefully excise the pulmonary artery confluence (Figure 3B). When the branch pulmonary arteries emanate from opposite sides of the truncal root (Collet and Edwards type III) and sufficient tissue is present, a sleeve of the truncal root can be excised and the distal opening closed (Figure 3C).



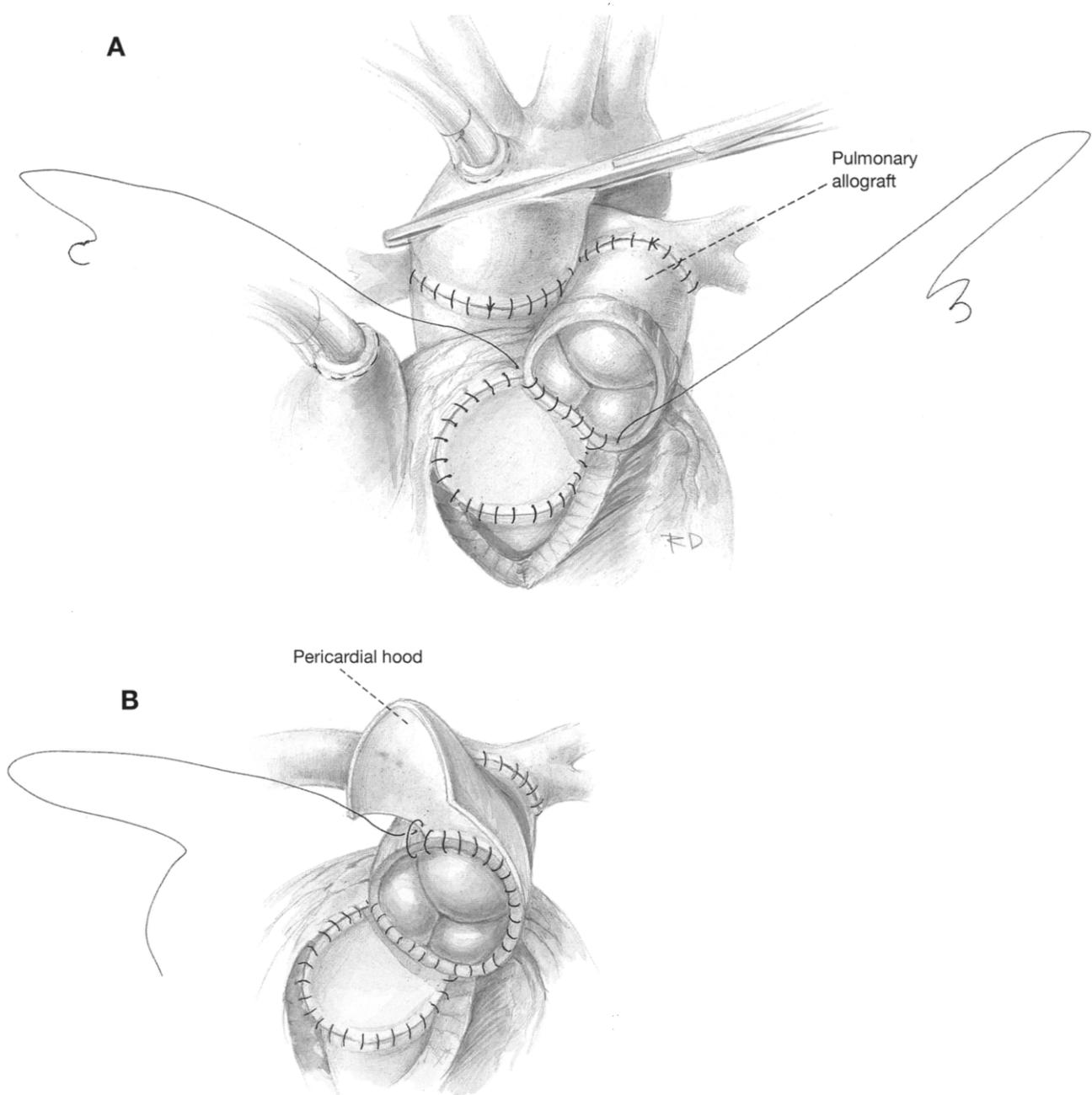
4 Once the pulmonary bifurcation is separated from the arterial trunk, the branch pulmonary arteries are mobilized to allow positioning to the left. The remaining proximal truncal root is trimmed to allow for a direct end-to-end anastomosis. If truncal valve repair or reconstruction is indicated, it is performed at this time.^{4,5}



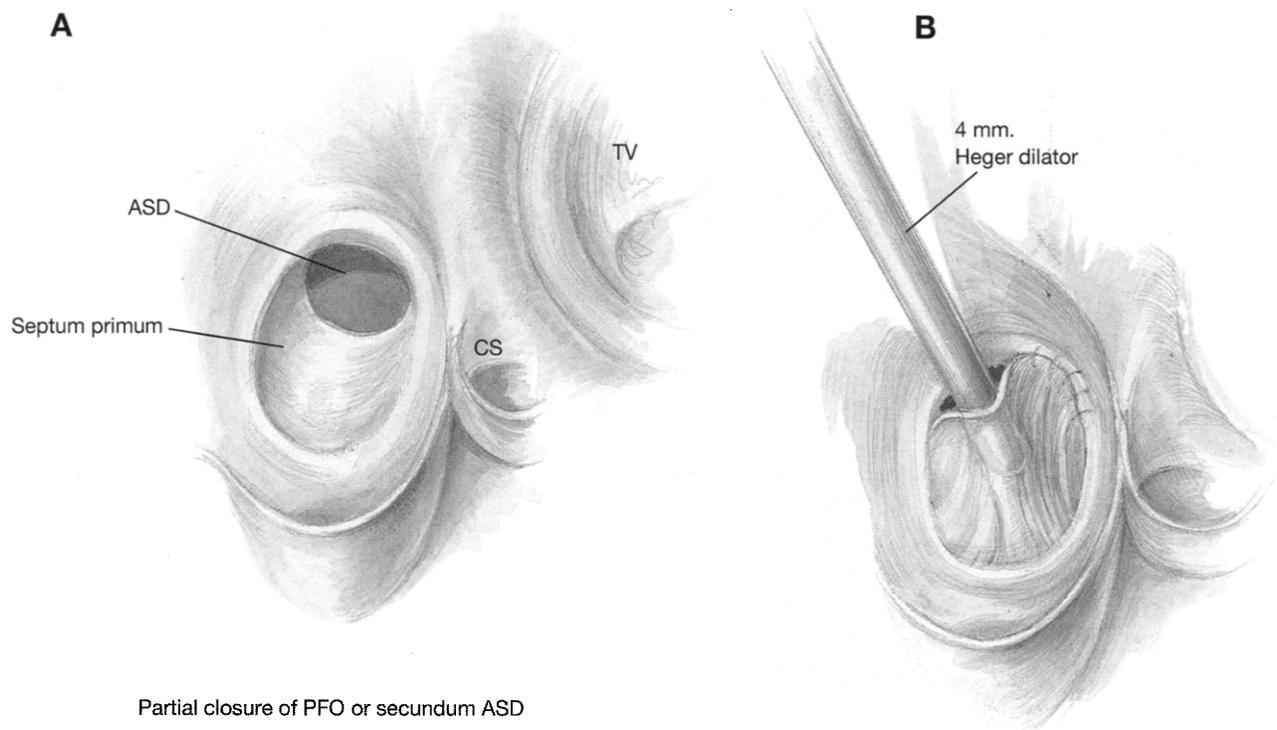
5 Before anastomosing the truncal root to the distal ascending aorta, a right ventriculotomy is made. By looking through the divided truncal root and through the truncal valve, the crest of the interventricular septum is visualized, and the position of the ventricular septal defect is identified. While noting the distribution of coronary arteries on the surface of the right ventricle, a right angle clamp is placed through the truncal valve, and the optimal site for incision is identified. Of course, the ventriculotomy must be rightward of the left anterior descending coronary and positioned between major conal branches. Although the incision is placed near the base of the truncus, care must be taken to prevent extension superiorly into the annulus of the truncal valve.



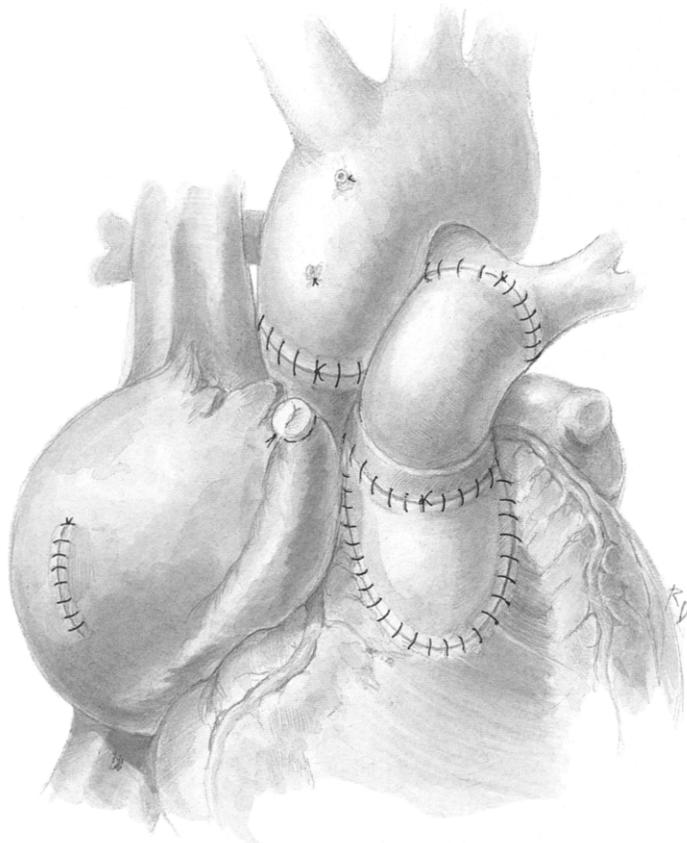
6 Once the ventriculotomy is completed, the truncal root is anastomosed to the ascending aorta with 6-0 Prolene sutures (Ethicon, Inc., Somerville, NJ). End-to-end anastomosis creates a concentric plication of the truncal root as well as upward traction on the truncal commissures. These effects often result in improved competence of a mildly insufficient truncal valve. An infusion of cardioplegia into the reconstructed aortic root provides additional myocardial protection and permits observation of the competence of the truncal valve from below (Figure 6a). An ovoid patch of glutaraldehyde-treated autologous pericardium is used to close the ventricular septal defect (Figure 6b). Superiorly, the patch is secured to the epicardium and carefully transitioned into the ventriculotomy onto the muscular margin of the defect. A bridge of muscle commonly protects the conduction tissue as the suture line is brought inferiorly. However, when the inferior portion of the defect consists of the base of the septal leaflet of the tricuspid valve, sutures must be carefully placed in the valve tissue near the annulus to avoid injury to conduction tissue.



7 A cryopreserved pulmonary allograft is selected soon after the heart is exposed. Although some surgeons select the largest allograft that can fit in the chest, we prefer to match allograft size to the estimated size of the pulmonary bifurcation. The typical range for a neonate is 8 to 12 mm. Allografts of this size are technically easier to implant, are less likely to be compressed by the chest wall, and have not been a risk factor for early replacement.¹⁰ Excessive length and the potential for kinking are avoided by trimming the allograft 3 mm distal to the pulmonary valve commissures. After the distal pulmonary anastomosis is completed with a running suture of 6-0 Prolene, the proximal portion of the allograft is sewn to the superior aspect of the ventriculotomy. Approximately one-third of the circumference of the allograft is attached to the edge of the ventriculotomy (Figure 7a). Placing a gusset or hood of glutaraldehyde-treated autologous pericardium completes the proximal right ventricle to allograft anastomosis. To preserve the configuration of the pulmonary valve, the hood is first secured to the remaining circumference of the pulmonary allograft (Figure 7b). The remaining pericardial gusset is trimmed to fill the ventriculotomy and sutured to the epicardial edge with 5-0 Prolene sutures.



8 Before de-airing and removing the cross-clamp, the vent is removed, and the interatrial septum is visualized. If only a small patent foramen is present, it is left open. Closing the interatrial septum around a 4-mm Hegar dilator narrows any communication larger than 4 mm. Leaving a small atrial communication is important to minimize the risks of right ventricular failure and/or acute pulmonary hypertensive crises during the early postoperative period. Providing a calibrated opening for right to left shunting at the atrial level permits adequate left-sided cardiac output at the expense of a mild decrease in arterial oxygen saturation. Failure to narrow a significant atrial communication may result in an excessive amount of right-to-left shunting, with dangerously low levels of oxygen saturation.



9 After the atriotomy is closed, and the left ventricle and aortic root are de-aired with needle aspiration, the aortic cross-clamp is removed. During rewarming and reperfusion, a loading dose of milrinone (50 $\mu\text{g}/\text{kg}$) is administered, and continuous infusions of milrinone (0.5 $\mu\text{g}/\text{kg}/\text{min}$) and dopamine (3 to 5 $\mu\text{g}/\text{kg}/\text{min}$) are begun. The infant is then weaned from cardiopulmonary bypass. After approximately 10 minutes of modified ultrafiltration, the heart is decannulated. The left pleural space is opened widely to encourage rotation of the heart to the left so that the sternum does not compress the conduit. A piece of Gore-Tex (W.L. Gore & Associates, Phoenix, AZ) membrane is loosely tacked to the pericardial edges to allow safe repeat sternotomy when conduit replacement becomes necessary. Sternal closure is then performed in a standard manner. If sternal closure results in any compromise in blood pressure or oxygen saturation, the sternum is left open with delayed sternal closure performed in 2 to 3 days.

Postoperative Treatment

After the repair of truncus arteriosus, treatment involves minimizing pulmonary vascular resistance and supporting right ventricular function. For the first 24 hours, the infant is paralyzed, sedated, and ventilated to maintain a level of hypocapnia and respiratory alkalosis. The milrinone infusion is continued for its combined inotropic and vasodilatory effects. If the infant is hemodynamically stable after 24 hours, paralysis is discontinued, and sedation is decreased. If hemodynamic remains stable as the infant awakens, mechanical ventilation is weaned. After successful ex-

tubation, the inotropic agents (ie, milrinone and dopamine) are weaned off.

COMMENTS

The technique described incorporates several strategies designed to achieve a high rate of success for what was once a daunting congenital heart defect. The performance of the procedure during the neonatal period may be the most important factor. The use of continuous cardiopulmonary bypass rather than deep hypothermic circulatory arrest allows the surgeon to proceed through the repair in a deliberate yet unrushed fashion. Double venous cannulation and a carefully placed vent provide excellent exposure and an essentially bloodless operative field. Complete transection of the aorta compliments the exposure and helps the surgeon avoid the potential pitfalls of pulmonary artery, coronary artery, or truncal valve injury. The use of a pulmonary allograft eliminates the need to insert an oversized conduit that may not match the anatomy and space limitations of a neonate. Applying these principles, surgeons can now achieve early survival rates in excess of 95%.³

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