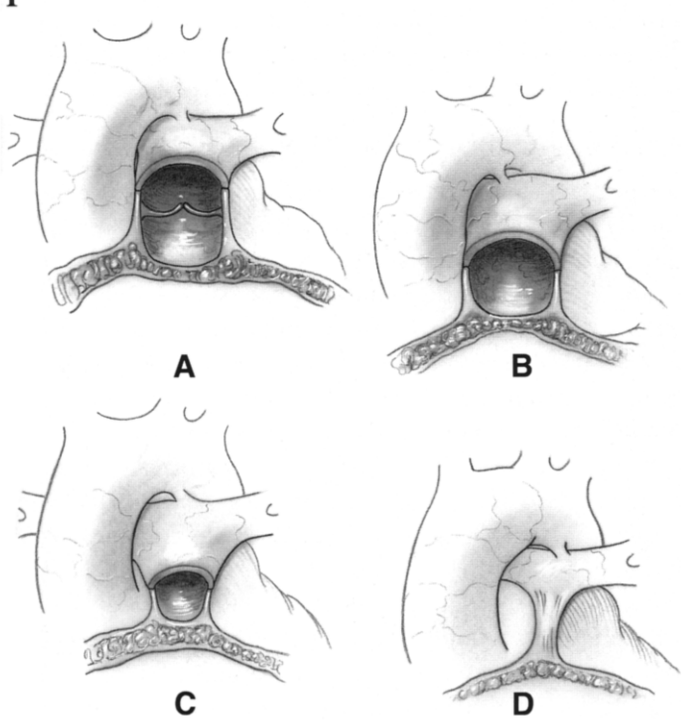
Conduit Repair of Tetralogy of Fallot With Pulmonary Atresia

Carin van Doorn and Marc R. de Leval

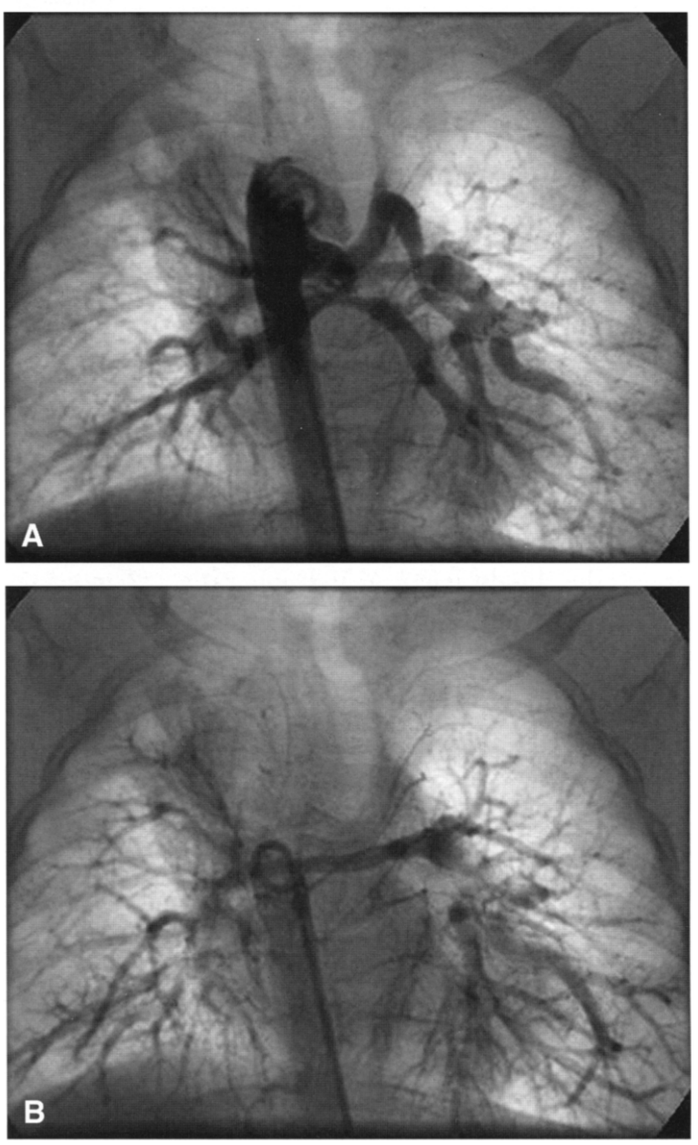
For the purpose of this article, tetralogy of Fallot with pulmonary atresia (ToF-PA) is defined as a congenital cardiac abnormality in which there is a lack of luminal continuity between the right ventricle and the pulmonary artery. The intracardiac anatomy is characterized by marked anterior and leftward displacement of the infundibular septum, which is often fused with the anterior wall of the right ventricle, and a large outlet, malaligned ventricular septal defect (VSD). Characteristically, the pulmonary arterial anatomy is extremely variable. The diameter and length of the atretic segment can range from a vessel that is patent down to the heart, to a fibrous strand between the right ventricle and pulmonary arterial confluence (Figure I). The central pulmonary arteries may be normal in size or hypoplastic, confluent or nonconfluent, and may be unilaterally or bilaterally absent. The pulmonary arteries are supplied from a patent ductus arteriosus, major aorticopulmonary collateral arteries (MAPCAs) or surgically created arteriopulmonary shunts, in isolation or combination. The distal pulmonary artery branches may have stenoses and connect to a variable number of parenchymal segments (Figure II).

The number and distribution of MAPCAs, when present, are also highly variable. They usually arise from the anterior wall of the descending thoracic aorta but may also originate from the aortic arch, subclavian artery, carotid artery or, occasionally, coronary arteries and connect to either central or parenchymal pulmonary arteries. Communications with central pulmonary arteries may be restrictive but can be high flow and high pressure, in which case, there is a risk of the

I



II



development of pulmonary hypertension. MAPCAs connecting to parenchymal segments enter the posterior aspect of the pulmonary hilum, running posteriorly to the bronchus of the segment they supply. They may constitute the only blood supply to the pulmonary segment, or an additional supply from central pulmonary arteries may be present (i.e., dual blood supply).

INVESTIGATIONS AND SURGICAL STRATEGY

A detailed anatomic and hemodynamic study is necessary early in life to plan treatment. The following information should be obtained (1) size and location of all VSDs; (2) site(s) and severity of obstruction in the right ventricular outflow tract (RVOT); (3) origin and branching pattern of the coronary arteries; (4) origin, course, and distribution of the central pulmonary arteries, MAPCAs, and any surgical shunts and the proportion of lung parenchyma supplied. Direct pressure measurements should be performed to identify important arterial stenoses. A combination of echocardiography and angiography, including selective injection of MAPCAs, is used to obtain this information. Magnetic resonance imaging is also increasingly able to provide these details.

The aim of complete surgical repair is to establish unrestrictive right ventricle to pulmonary artery continuity and separate the systemic from the pulmonary circulation. The most powerful factor for survival after surgery is the peak right ventricular/left ventricular (RV/LV) pressure ratio after repair. A ratio ≥ 1 has been associated with a considerable risk of dying early or late after repair,[1] and if there is no residual RVOT obstruction, then the small size of the pulmonary arteries and larger number of MAPCAs are the most important determinants.[2] The RV/LV ratio after repair can be predicted from the McGoon ratio, which is calculated from the sum of the diameter of the immediate prebranching left and right pulmonary artery divided by the diameter of the descending aorta, just above the diaphragm.[3] A McGoon ratio >2 indicates unrestrictive pulmonary blood flow. For a McGoon ratio of 1, the predicted RV/LV ratio after repair is 0.7.

Most patients with ToF-PA have restricted pulmonary blood flow and show cyanosis. Occasionally, they are well balanced or may have heart failure due to pulmonary overflow. In patients with good-sized central pulmonary arteries that require surgery during the first 3 months of life, we perform a systemic to pulmonary artery shunt, most commonly a lateral or central interposition shunt. Such a preliminary palliative operation is also considered for patients younger than 3 to 4 years, in whom an extracardiac conduit is necessary for repair. In patients with very hypoplastic central pulmonary arteries with a predicted ratio after repair of > 1, complete repair is precluded, and palliative procedures such as a central interposition shunt or outflow tract patch enlargement on cardiopulmonary bypass (CPB) are performed. Repeat catheterization is required in due course to see if the pulmonary arteries have increased in size.

Patients with anatomic continuity between the right ventricle and pulmonary artery, valvar atresia and good-sized pulmonary arteries (predicted RV/LV ratio after repair < 0.7) are treated very much like patients with tetralogy of Fallot (ToF), with a transannular patch repair. If there is a gap between the right ventricle and pulmonary artery or if the coronary anatomy precludes the placement of a transannular patch, an extracardiac valved conduit is used. If at the completion of the repair the RV/LV ratio is > 1 and the RVOT is wide open, CPB is reinstituted, and a central fenestration is made in the VSD patch. The fenestration could be closed with a device in the catheter laboratory if right ventricular (RV) pressures decrease at follow-up.

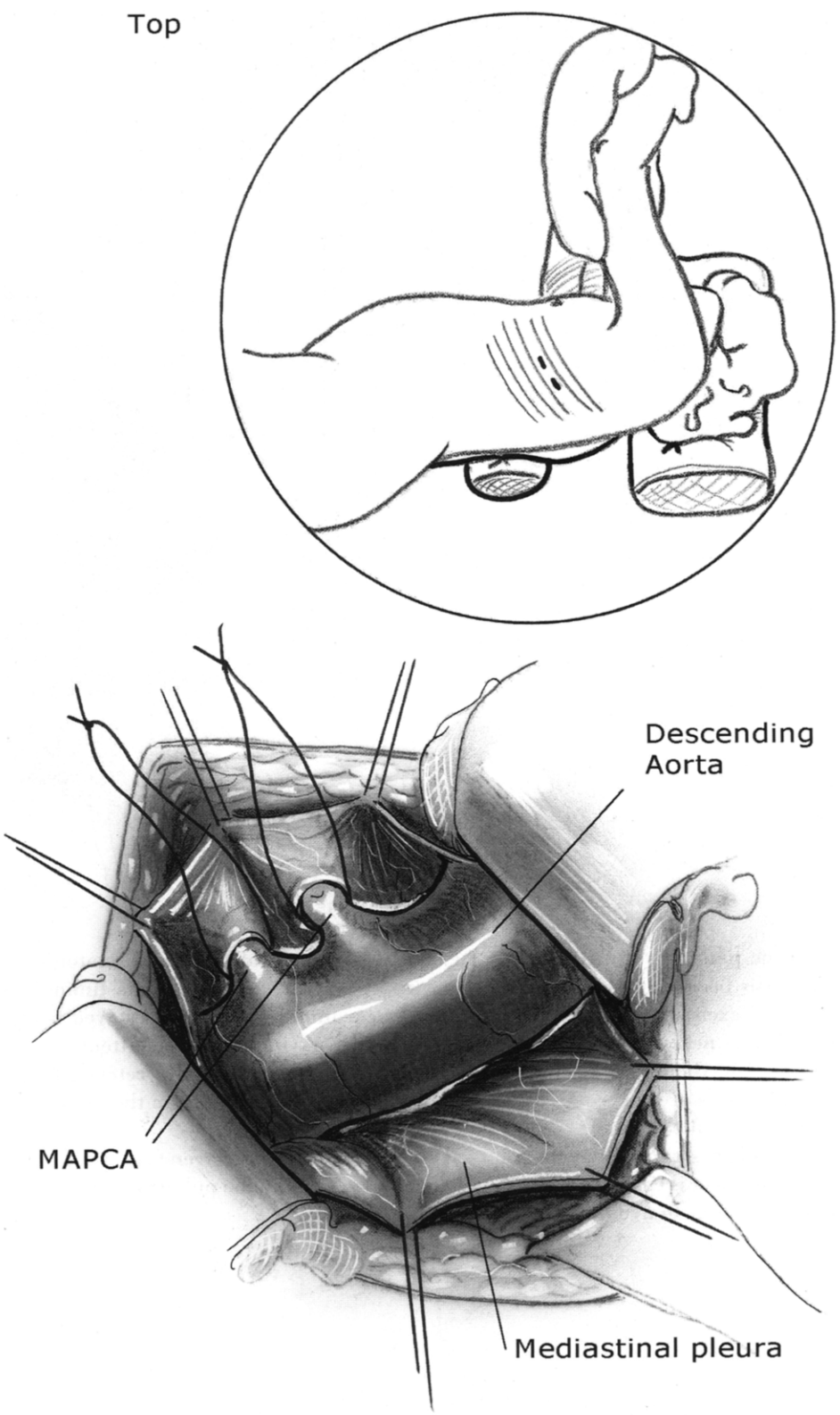
In the presence of MAPCAs that provide the blood supply to pulmonary segments, which are also served by central pulmonary arteries, the aforementioned surgical strategy also applies, and the MAPCAs are closed at the time of repair. However, MAPCAs that are the sole supply of significant bronchopulmonary segments must be connected to the central pulmonary arteries because of the risk of pulmonary infarction after closure. If MAPCAs are the sole blood supply to more than one-third the lung parenchyma, then unifocalization procedures are necessary to achieve complete repair. Unifocalized MAPCAs can be connected to an arterial shunt or to the right ventricle using a restrictive conduit and leaving the VSD open. If the capacity of the pulmonary arterial bed is judged adequate, either at unifocalization or at a later date, complete repair with a nonrestrictive extracardiac valved conduit and closure of the VSD are performed. Repeat cardiac catheterization is necessary to monitor the development of the pulmonary arterial bed, and interventional procedures may be required to deal with vascular stenoses. Because of the wide variation in anatomy, the principles of surgical repair have to be individualized for each patient.

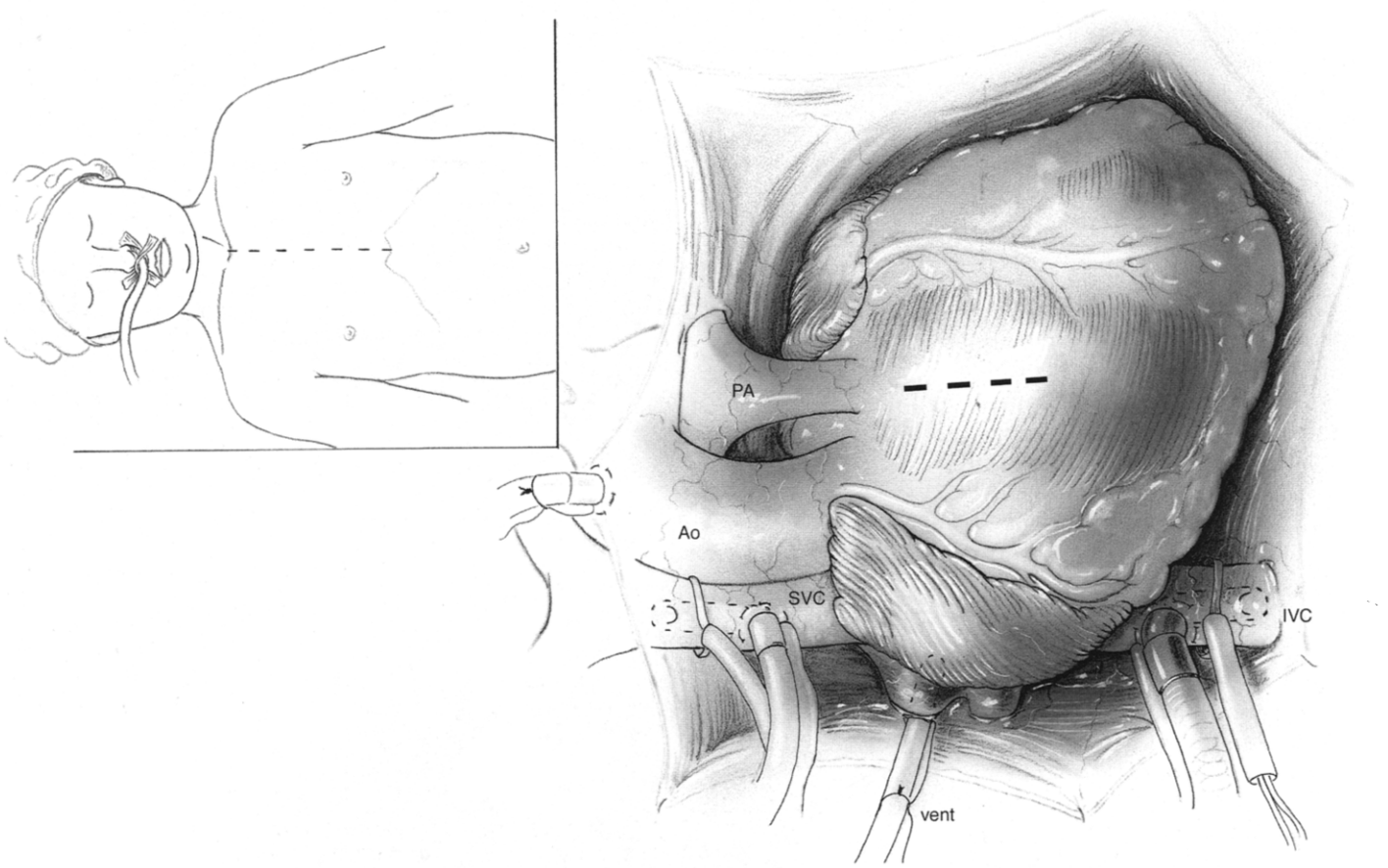
Choice of Extracardiac Conduit

Various types of valved conduits can be used to connect the RV to the central pulmonary arteries, including aortic and pulmonary homografts, porcine valved Dacron () conduit, or valved bovine jugular vein conduit. The choice of conduit depends on the circumstances at the time of the operation, local conduit availability, and surgeon preference. For example, a bifurcated pulmonary homograft may be advantageous if reconstruction of the central pulmonary arteries is also required. An aortic homograft and valved Dacron conduit may provide extra length if a large gap between the RV and pulmonary artery has to be bridged. Durability of the currently available conduits is limited, with approximately half the homografts and valved Dacron conduits failing at 10 years.[4,5] Bovine jugular vein conduits have only recently become available,[6] and their long-term durability is not yet known.

SURGICAL TECHNIQUE

1 The presence of MAPCAs can cause significant run-off in the pulmonary vascular bed during CPB, with resultant systemic hypoperfusion as well as excessive pulmonary venous return to the heart with the risk of cardiac distension. Ideally, MAPCAs should be dissected before the establishment of CPB but if this is not possible, it should be performed on normothermic or mildly hypothermic bypass, allowing the heart to continue to beat. The best exposure depends on the course of the MAPCAs. Those running in the upper part of the posterior mediastinum can be reached via median sternotomy, followed by an incision in the posterior pericardium above the left atrium. At the level of or above the pulmonary hilum, MAPCAs can be exposed by opening the anterior pleura and dissecting the hilum with the lung retracted towards the midline. Alternatively, a preliminary lateral thoracotomy is performed, the level of which is directed by the course of the MAPCAs. This procedure is particularly useful for access to vessels that arise below the hilum, which can often be identified where they arise from the anterior wall of the descending aorta. A silk tourniquet is placed loosely around the vessel with their ends positioned toward the anterior mediastinum. They can later be retrieved via median sternotomy and tightened once CPB has been established. The chest is closed in layers, and a drain is left behind. Once median sternotomy has been performed, the chest drain should be clamped to avoid excessive blood loss down the drain during CPB.

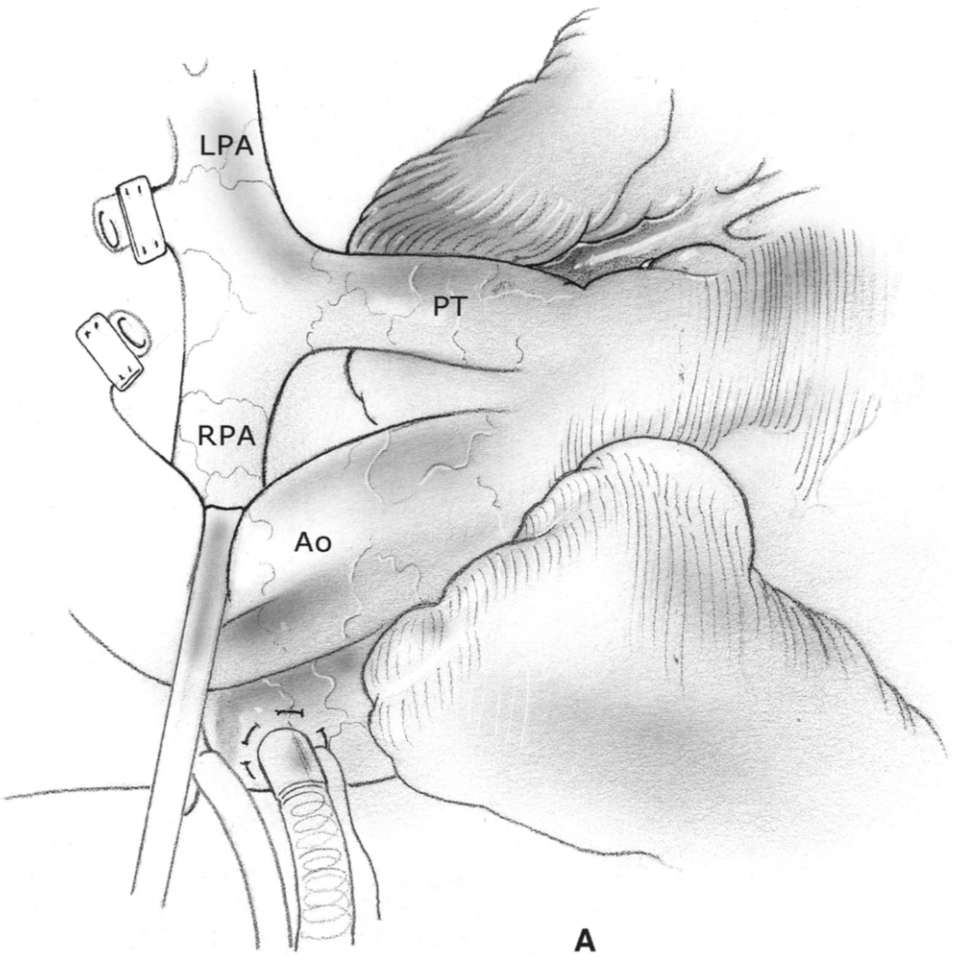




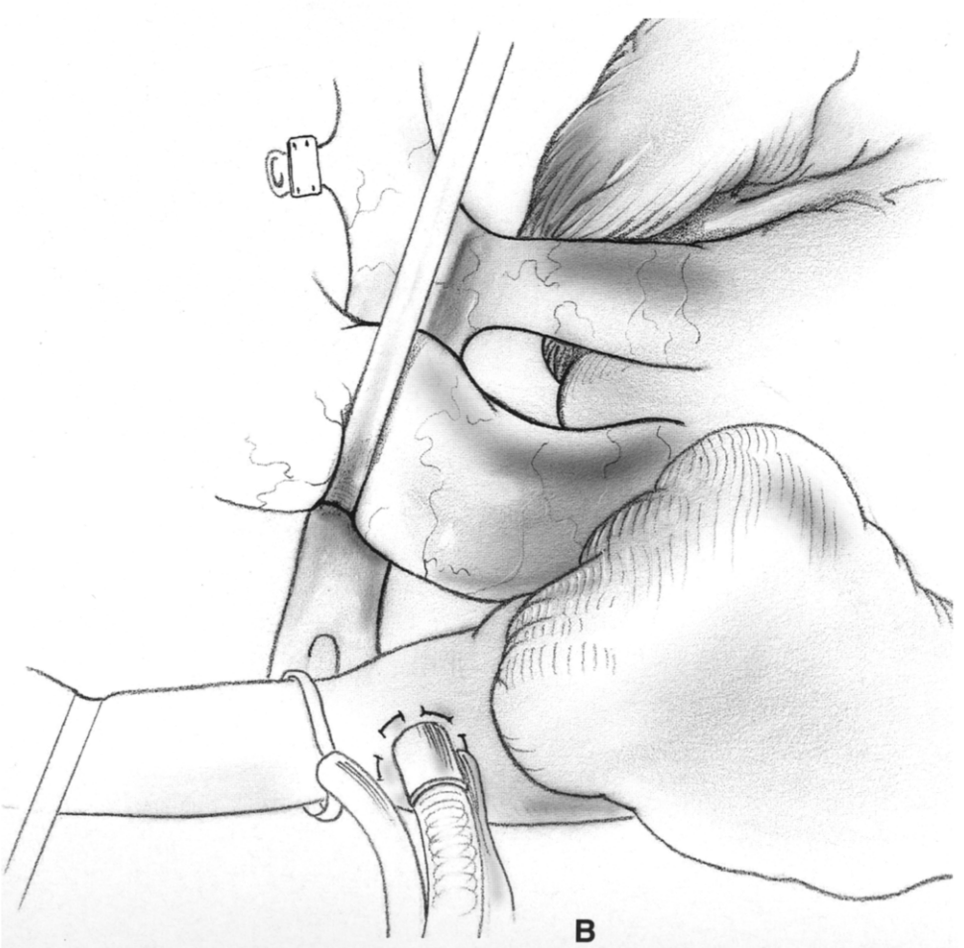
2 The patient is now turned to the supine position and median sternotomy performed. Particular attention is given to hemostasis because multiple small vessels in the pericardium and mediastinum may cause excessive bleeding. The thymus is subtotally excised to improve access. The pericardium is opened and stayed back, and any MAPCAs or surgical shunts are identified as necessary. The anatomy is inspected, paying particular attention to the coronary anatomy, size of pulmonary arteries, and the length of the atretic segment. The tourniquets are retrieved from the chest, and any further MAPCAs or surgical shunts are dissected. The pulmonary arteries are freed from the back of the ascending aorta and mobilized as far as possible toward the hilum.

Following systemic heparinization, the inferior and superior vena cava and ascending aorta are cannulated for CPB. The ascending aorta is cannulated high up to facilitate access to the pulmonary arteries. The left heart is vented via the right upper pulmonary vein to avoid distention.

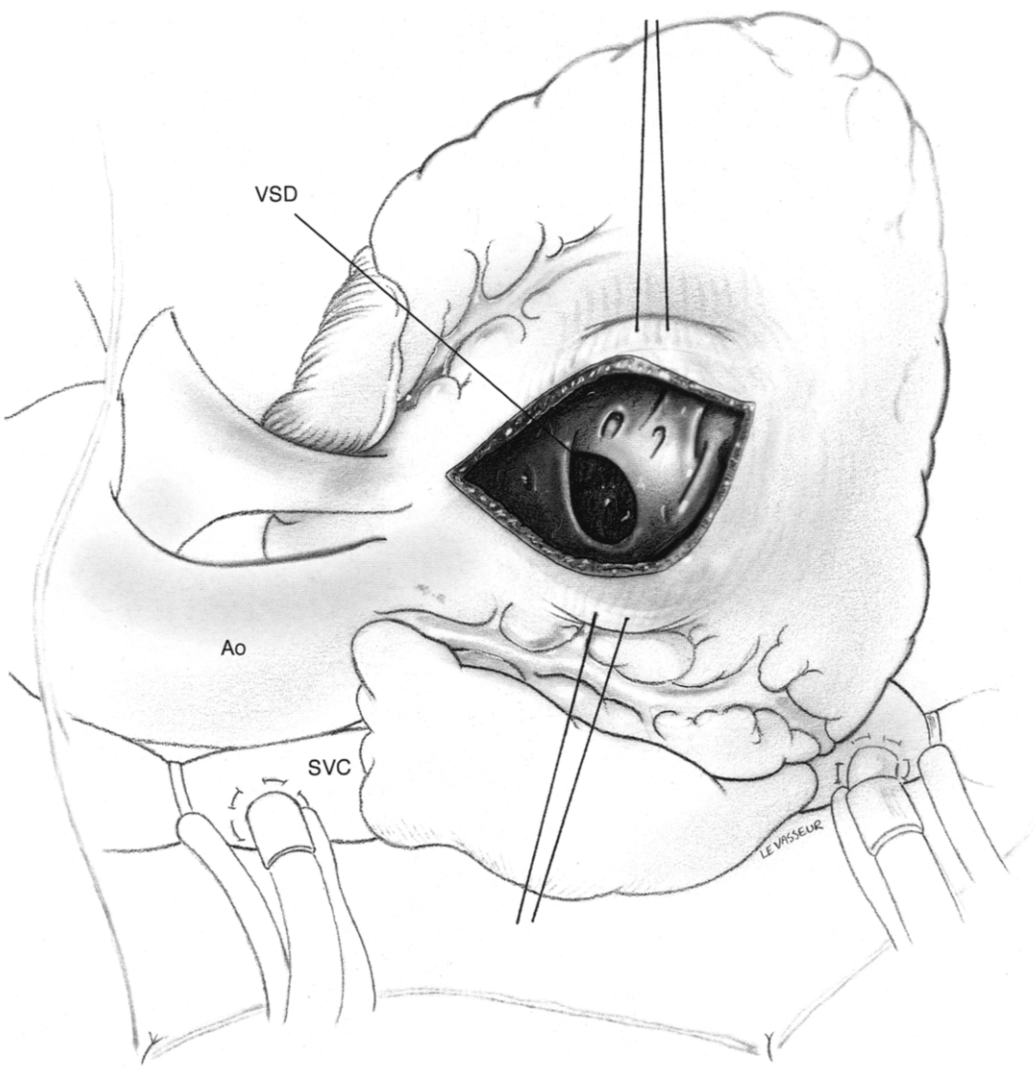
3 On commencement of CPB, the MAPCAs and any shunts are occluded, and the patient is cooled to 28°C. There may still be a large, left heart return but, as long as the heart continues to beat, this is ejected into the systemic circulation. The central pulmonary arteries are dissected further until they are fully freed from the mediastinal tissues down to the hilum on either side. In addition, good mobilization of the ascending aorta provides access to the central pulmonary arteries, and it is only in exceptional circumstances that a transection of the ascending aorta is necessary. To help reduce the gap between the pulmonary arteries and right ventricle, the ductal ligament (or patent ductus arteriosus) is divided between ligatures.



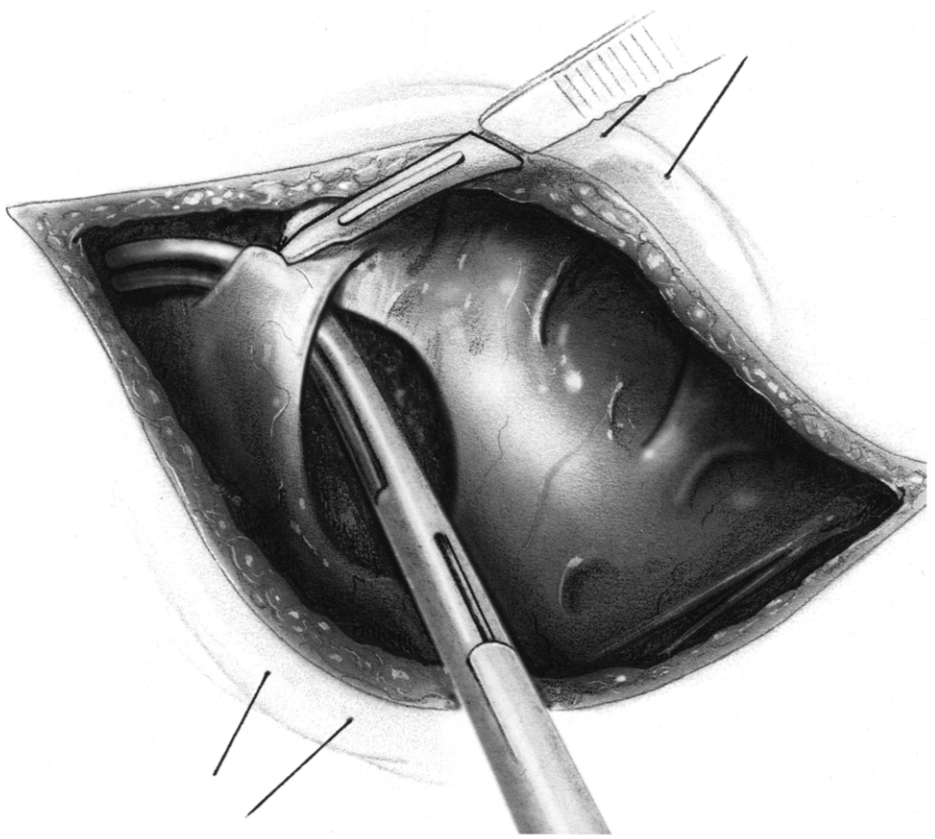
A



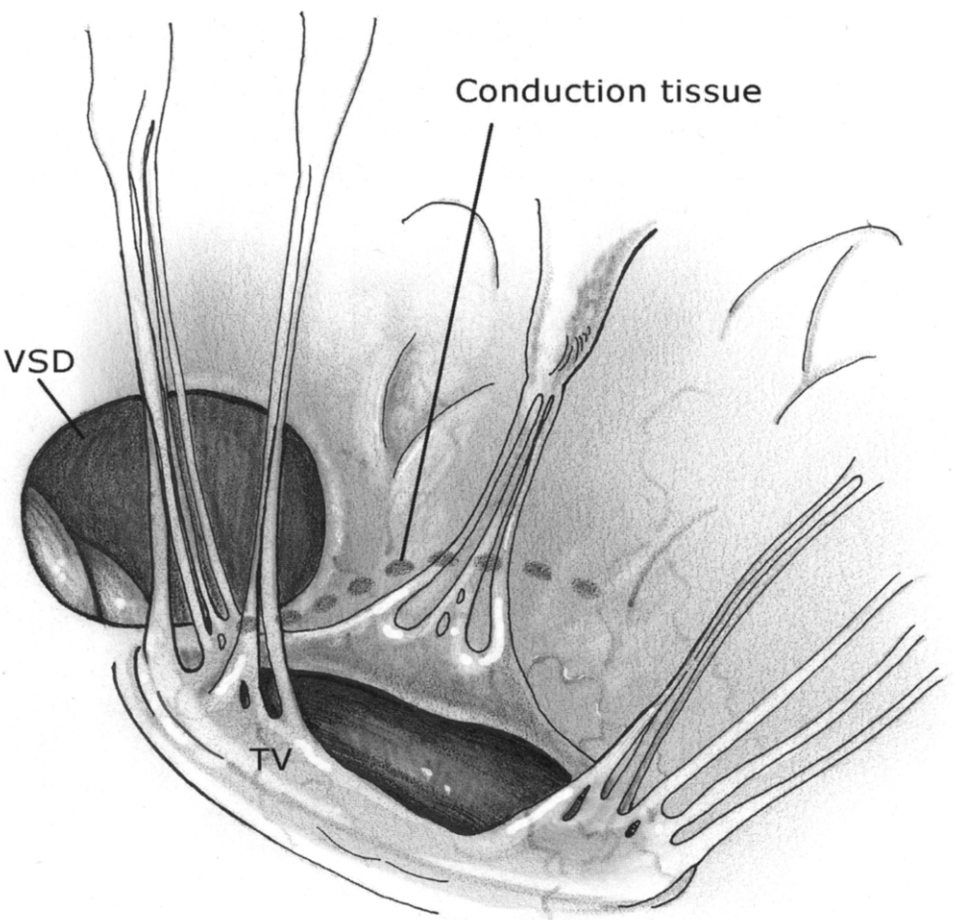
B



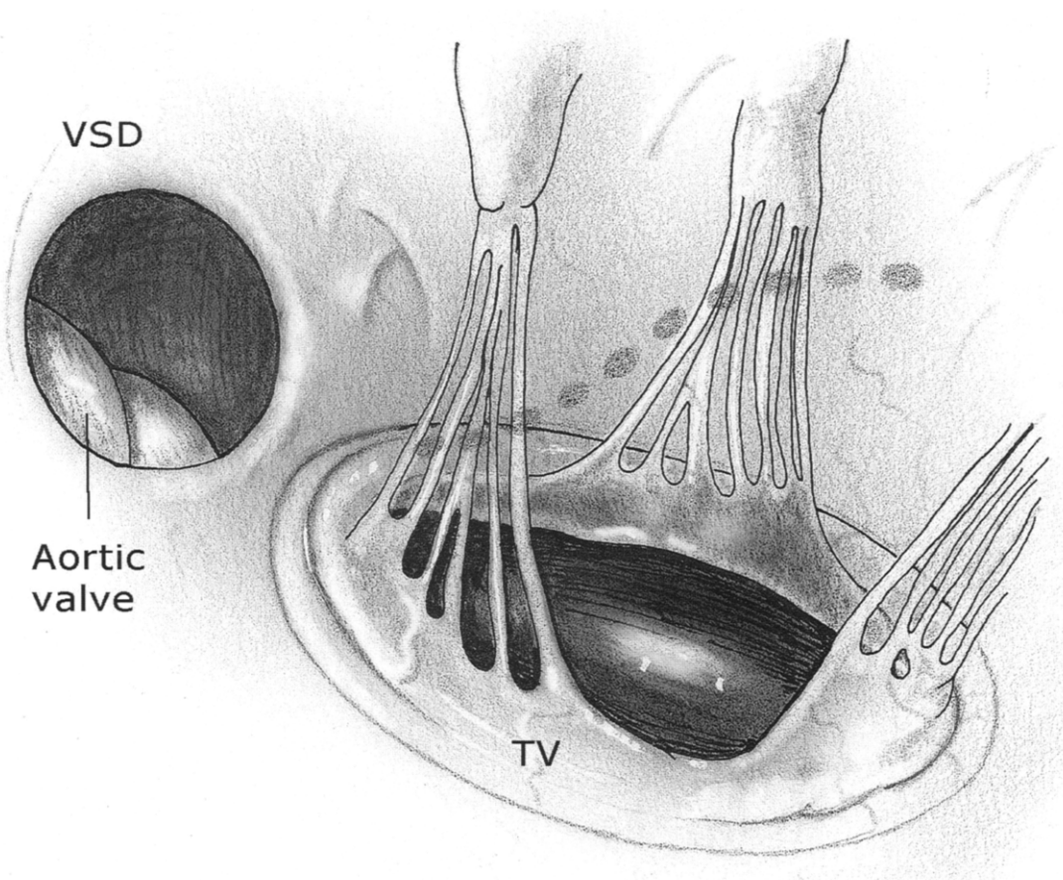
4 The aorta is cross-clamped, and the heart is arrested with 30 mL/kg hyperkalemic blood cardioplegia, with a further dose of 15 mL/kg every 20 to 30 minutes. Once the heart has stopped, it is important to ensure adequate venting to avoid ventricular distension. If left heart return is excessive, further cooling helps to protect end organs from ischemia, and also allows the CPB flow to be lowered to reduce return and improve visibility inside the heart. The caval snares are tightened. A vertical incision is made over the RVOT, and the intracardiac anatomy is inspected.



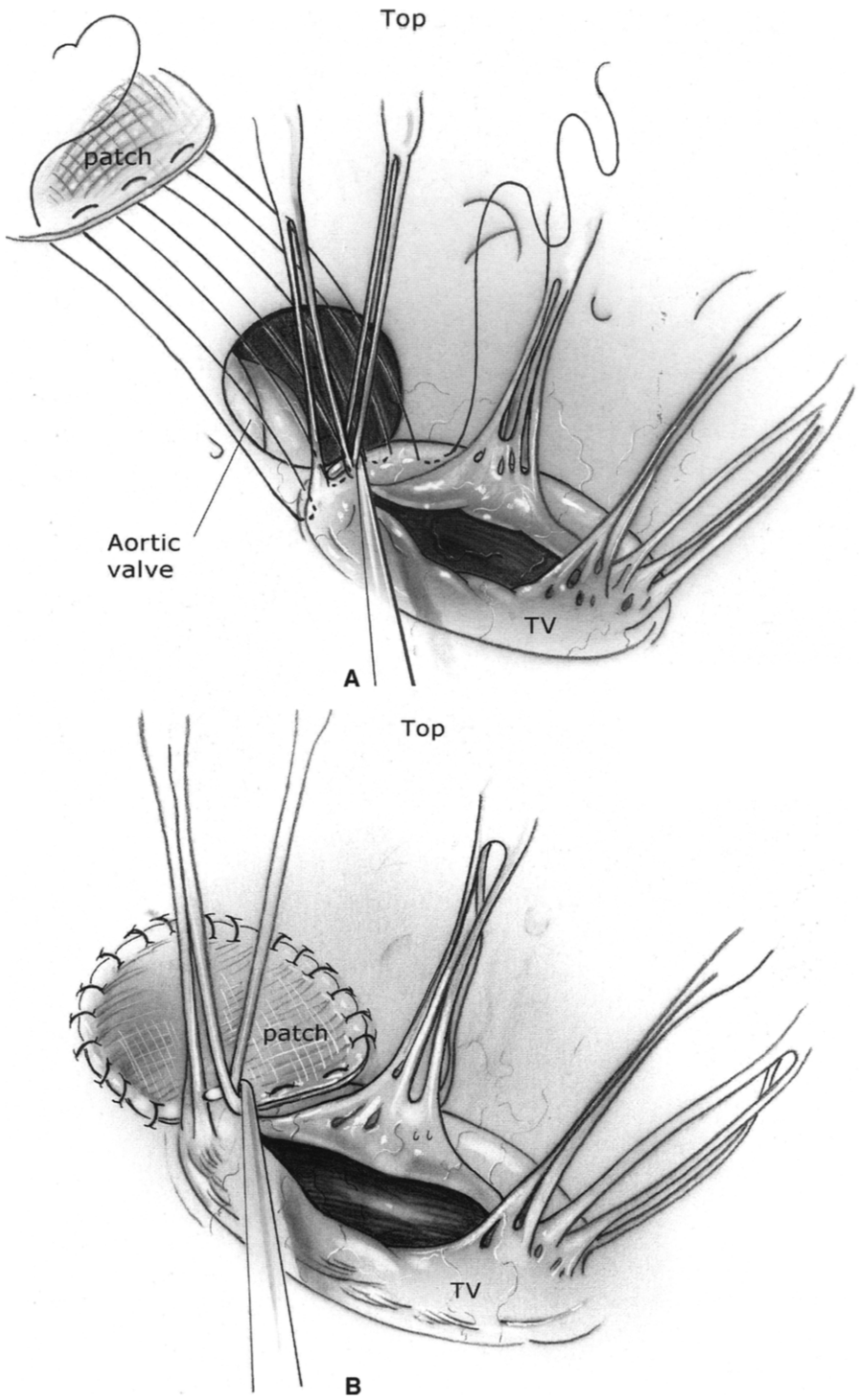
5 The infundibular stenosis is relieved by division and partial excision of the heavy trabeculations that bind the infundibular septum to the anterior right ventricular wall at both the parietal and septal ends.



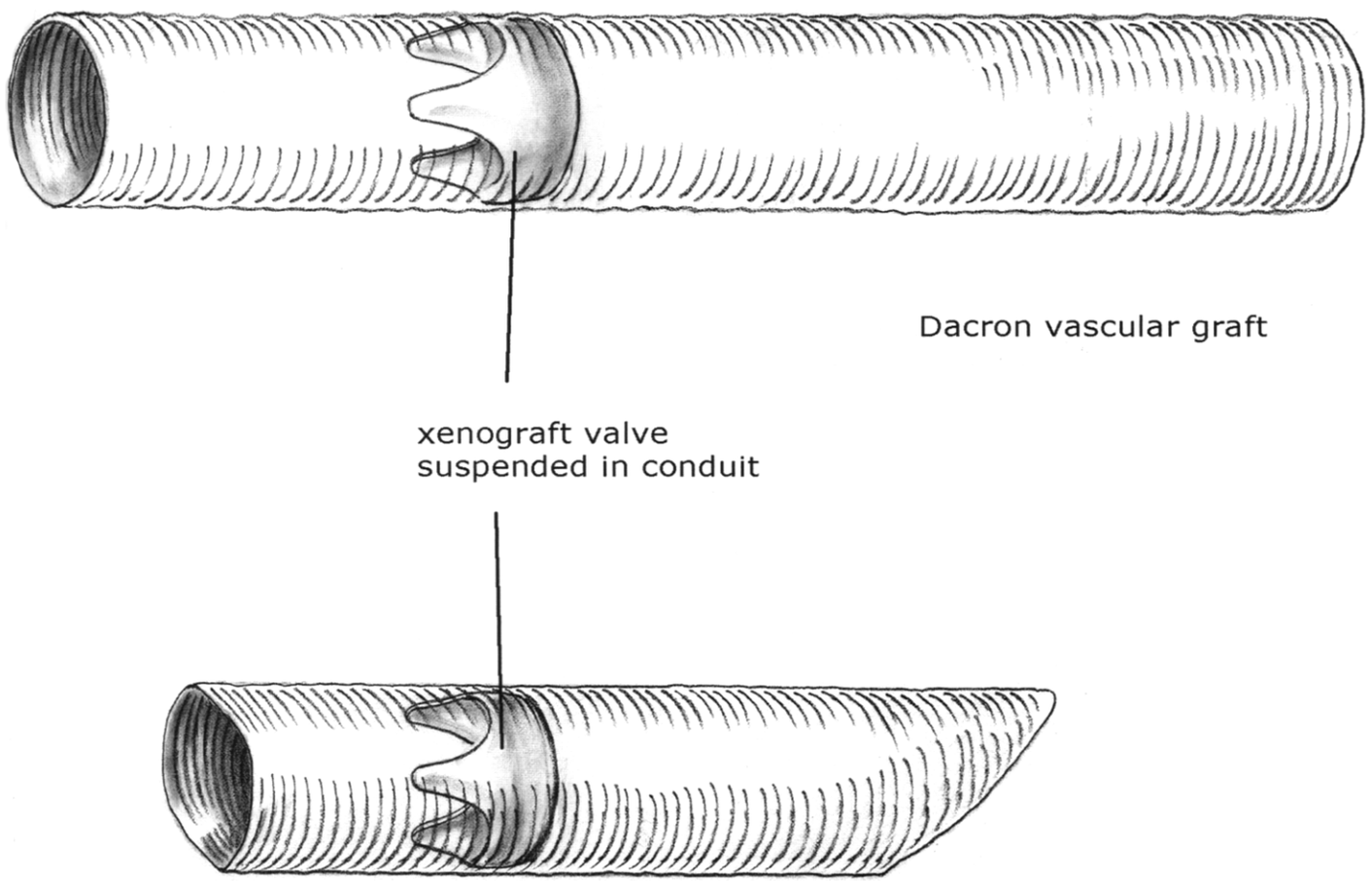
6 The assessment of adequate size of the right ventriculotomy and relief of RVOT obstruction is aided by the use of tables for normal pulmonary annulus size as a function of body surface area.[7] With the RVOT gradually opening up, the VSD becomes visible. This is most frequently a perimembranous defect and thus the penetrating and branching bundles of the conducting tissue are closely related to the posterior and inferior margin of the defect.



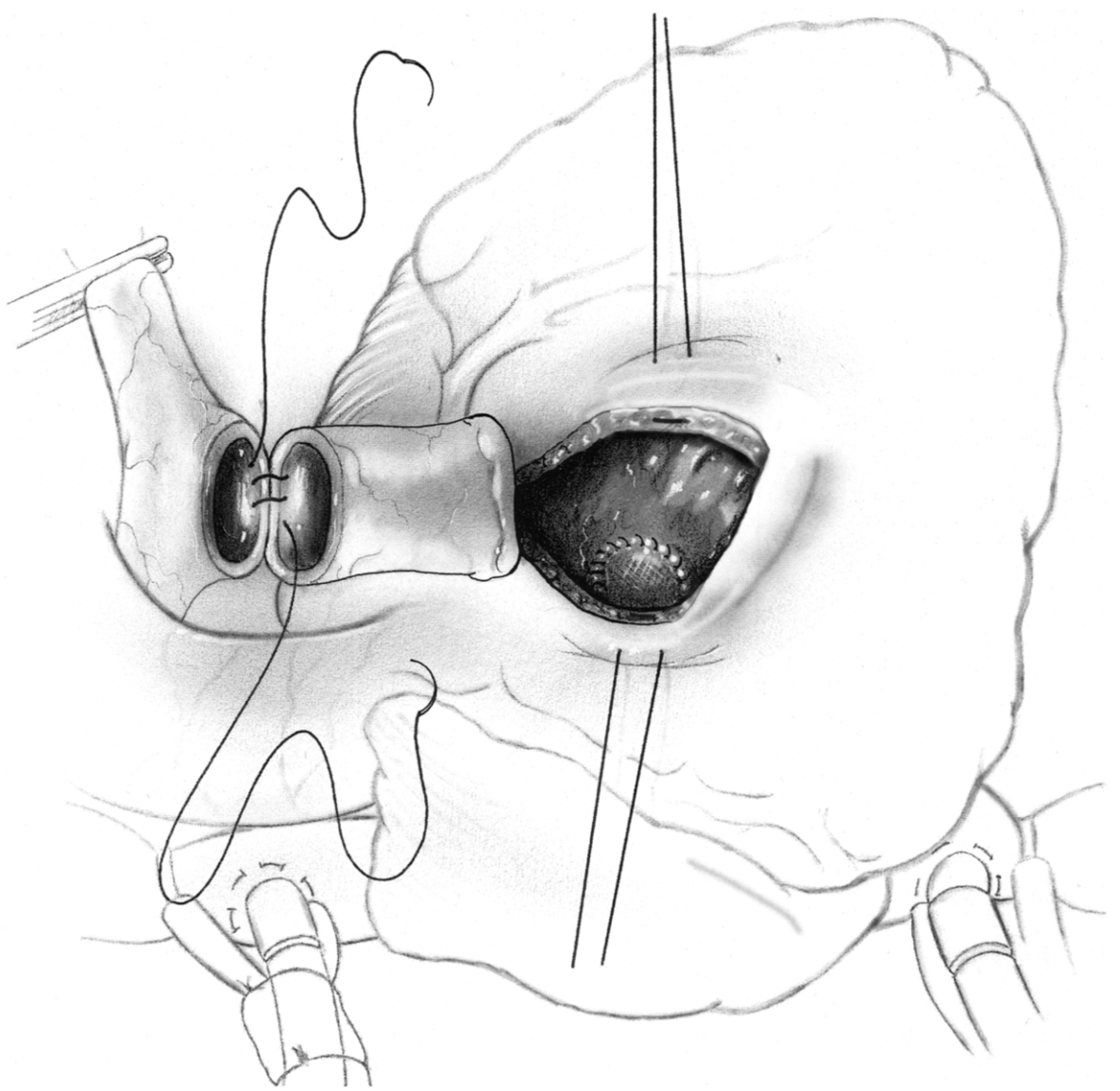
7 If a muscular VSD is present, the conduction tissue is remote from the margins of the defect.



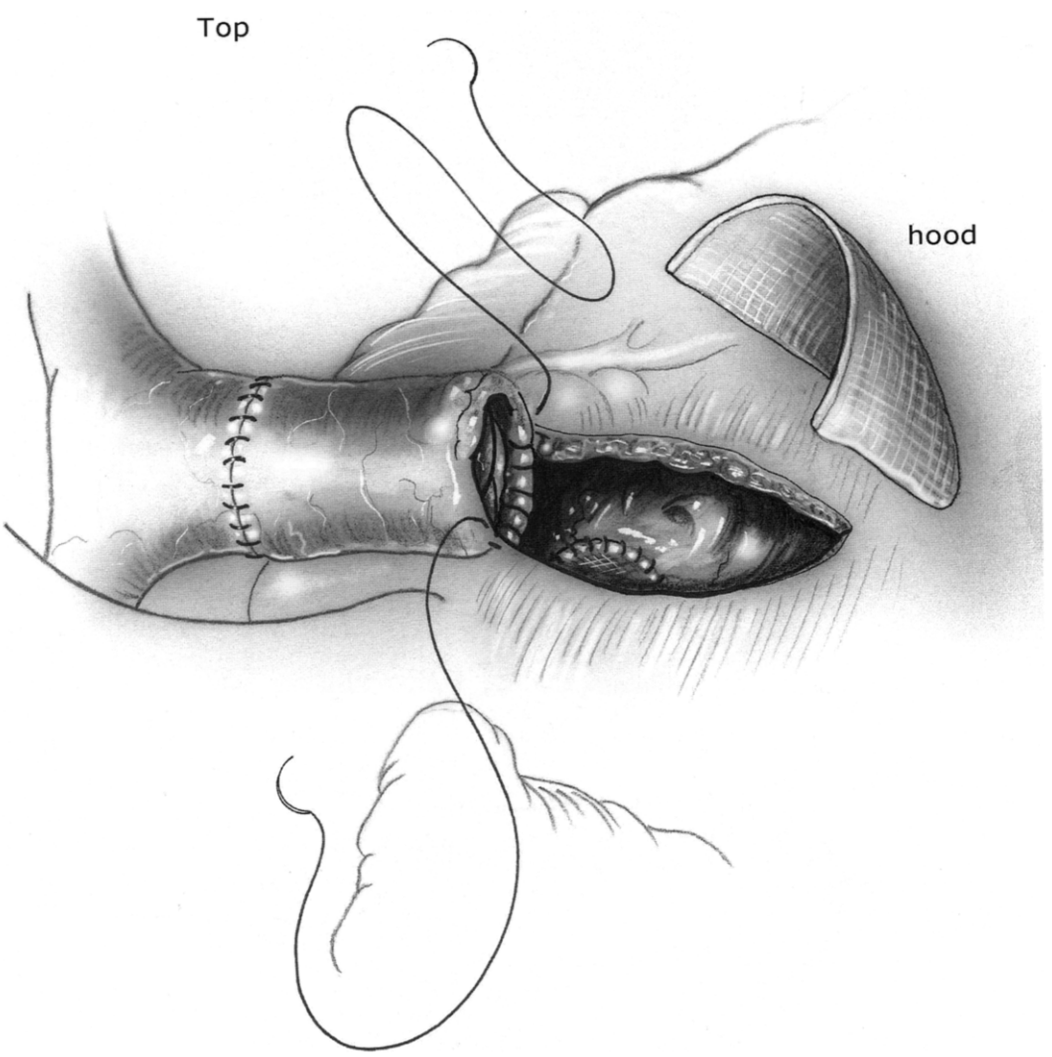
8 The perimembranous VSD is closed with a Gore-Tex (W.L. Gore & Associates, Phoenix, Arizona) patch, which is cut larger than the actual defect to cover the triangular area containing the conduction tissue. The patch can be fixed in place using a continuous suture (described below), or, alternatively, multiple interrupted pledgetted sutures can be used. For the continuous suture technique, the patch is clipped on the left upper corner of the sternal incision. We use a monofilament polypropylene double-armed suture with a semicircle needle. Suturing starts at the junction of the posterior limb of the trabecula septomarginalis and the base of the septal leaflet of the tricuspid valve. Initially, 3 or 4 mattress stitches are placed clockwise along the base of the tricuspid valve leaflet beyond the central fibrous body. The suturing in the area of the conduction tissue is now completed, and the patch is then lowered down. The remainder of the closure is performed using an over-and-over suture around the margin of the defect. There may be multiple trabeculations in the region of the aortic annulus, and the sutures should exit very near the aortic valve annulus to avoid leaving a residual VSD between the trabeculations. Finally, the suture line is reinforced with pledgetted mattress sutures in areas where the muscle is friable.



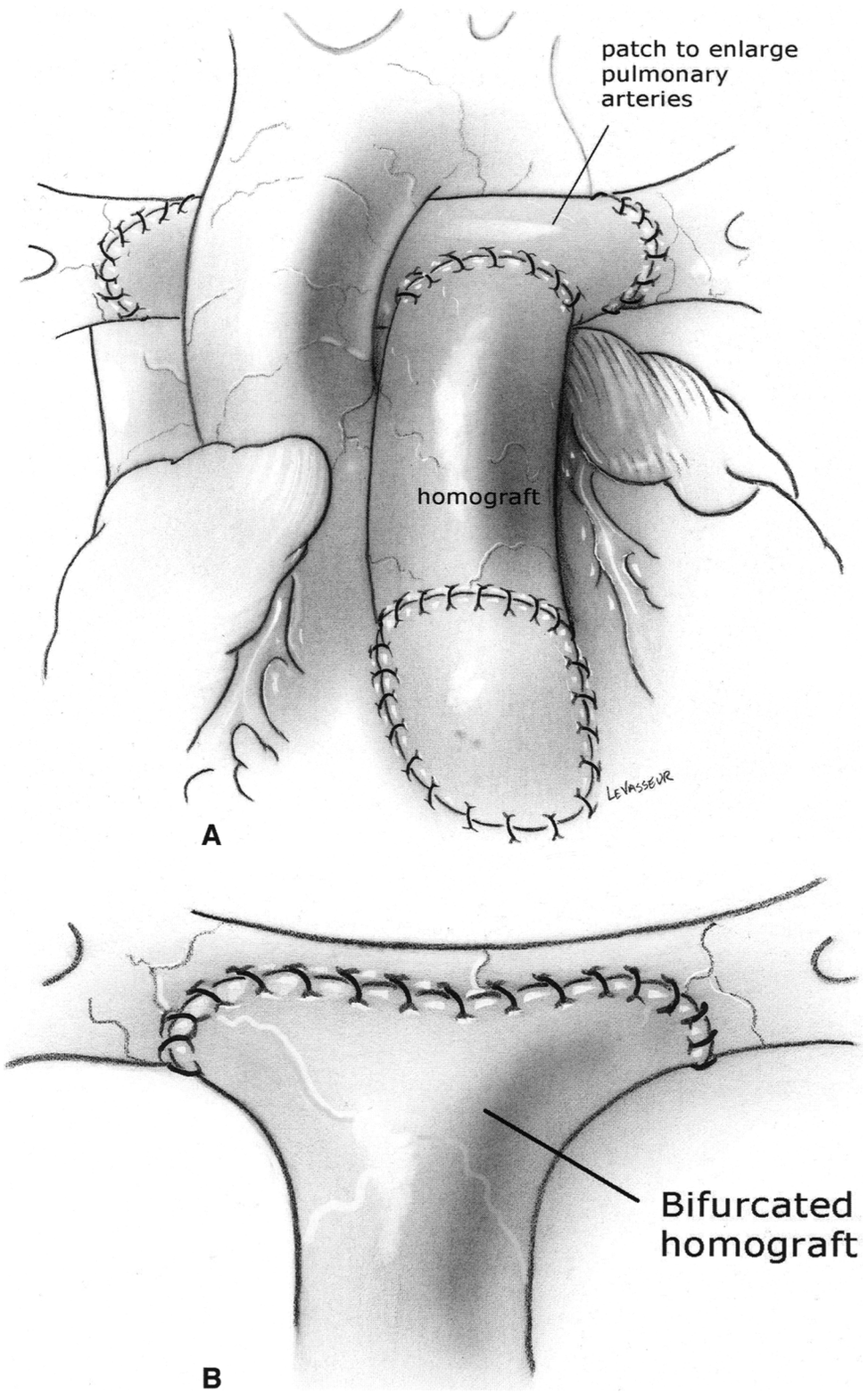
9 If the main pulmonary artery is of good size, the vessel is transected horizontally, just cephalad to the atretic part. The aortic cross clamp can now be removed, and rewarming of the patient can begin. Once the heart starts to beat, the size of the ventriculotomy and RVOT needs to be reinspected, and may require further widening now that the heart has regained tone. The extracardiac conduit is now prepared. If a homograft is used, the distal end is cut horizontally, and, at the proximal end, the subvalvar muscle is trimmed, leaving 3 to 4 mm behind. For a porcine-valved Dacron conduit, the distal end is cut across 5 to 10 mm above the valve. The conduit is stretched, and the distance between the pulmonary artery and upper end of the right ventriculotomy determined. A rectangular section is cut out at the back of the conduit, and a hood is fashioned on the front. It is important to try and place the conduit away from the back of the sternum by curving it gently to the left to avoid conduit compression or accidental entry during any future sternotomies.



10 A clamp is now placed across the pulmonary arteries to help exposure and control back bleeding. The distal anastomosis is performed first with a running 5-0 or 6-0 polypropylene suture.



11 The proximal anastomosis is placed in an extra-anatomic position, with the lower half of the conduit sewn directly on the upper half of the margin of the right ventriculotomy.



12 A hemostatic suture line is important because this part of the repair is difficult to access at a later time. In the case of a pulmonary homograft conduit, the upper half of the anastomosis is completed with a hood of bovine pericardium or Gore-Tex, or, in the case of an aortic homograft, the anterior leaflet of the mitral valve can be used. Starting at 3 o'clock, 1 arm of a double-armed 4-0 polypropylene suture is taken, and the surgeon starts working counterclockwise around the posterior margin of the ventriculotomy towards him/herself. The other arm of the polypropylene suture is used to fashion the hood with a running suture. Patients with moderately hypoplastic or nonconfluent pulmonary arteries may require enlargement or reconstruction of the central pulmonary artery confluence with a patch, bifurcated homograft, or the use of an additional conduit to reconstruct the branch pulmonary arteries.

CLOSING COMMENTS

Weaning From Bypass

When the surgery is completed and the patient is rewarmed, ventilation is commenced, and the patient is weaned off CPB. The RV/LV peak pressure ratio is determined. If RV peak pressure is > 70% systemic peak pressure and the RVOT is wide open, the VSD needs to be fenestrated. Transesophageal echocardiography is an invaluable tool to help decide on the level of any residual obstruction. To create the VSD, CPB is recommenced and the caval snares tightened. The right atrium is opened, the CPB flow briefly lowered, and a 4- to 5-mm hole is cut in the middle of the VSD patch.

Once satisfactory hemodynamics after repair have been achieved, the heparin is reversed with protamine. Any MAPCAs that were snared during CPB are ligated. Hemostasis is secured, which may take some time because of the multiple small collateral vessels in the mediastinum. Two atrial and 2 ventricular epicardial pacing wires are placed, and the chest is closed.

Outcome

For patients with good-sized pulmonary arteries, the operative mortality approaches that for ToF at < 5%. Patients who do not progress following surgery should have an early examination to look for any residual defects, such as residual VSD, RVOT obstruction, or the presence of MAPCAs.

Conduit Failure

The durability of homograft conduits is a case for concern. In our institution, Stark and coworkers reviewed the fate of 405 homografts placed in the subpulmonary position, and found that freedom from failure at 5, 10, and 15 years was 84%, 58%, and 31%, respectively.[4] No significant difference was shown between aortic and pulmonary homografts, and between cryopreserved and antibiotic preserved homografts, but first conduits lasted longer than second and subsequent conduits. Repeat conduit placement was associated with low mortality, and the long-term survival of patients who were alive 90 days after the initial homograft placement was 95% at 5 years and 85% at 15 years. In a recent series from the Mayo Clinic, the overall long-term survival of subpulmonary porcine-valved Dacron conduits, homograft conduits, and non-valved conduits was 55% at 10 years and 31% at 20 years.[5] The investigators reported inferior durability of a homograft compared with a porcine-valved Dacron conduit.

Reducing the Number of Reoperations

Most patients receiving a subpulmonary conduit are likely to have this replaced at least once during their lifetime. Therefore, alternative treatment strategies are being sought to limit the number of reoperations. Percutaneous stenting of stenosed conduits has emerged as a successful technique for delaying surgical replacement. Treatment for incompetence with the percutaneous implantation of a stented bovine jugular valve was successfully introduced recently in clinical practice by Bonhoeffer and coworkers from our institution.[8] If the conduit cannot be improved and reoperation is unavoidable, then alternative techniques for creating an unobstructed and competent subpulmonary pathway need to be considered. In this respect, the early results of construction of a pericardial roof over the bed of the explanted conduit with insertion of a bioprosthesis seem promising.[5]

REFERENCES

1. Kirklin JW, Blackstone EH, Shimazaki Y, et al: Survival, functional status, and reoperations after repair of tetralogy of Fallot with pulmonary atresia. J Thorac Cardiovasc Surg 96:102-116, 1988
2. Blackstone EH, Shimazaki Y, Maehara T, et al: Prediction of severe obstruction to right ventricular outflow after repair of tetralogy of Fallot and pulmonary atresia. J Thorac Cardiovasc Surg 96:288-293, 1988
3. McGoon DC, Baird DK, Davis GD: Surgical management of large bronchial collateral arteries with pulmonary stenosis or atresia. Circulation 52:109-118, 1975
4. Stark J, Bull C, Stajevic M, et al: Fate of subpulmonary homograft conduits: Determinants of late homograft failure. J Thorac Cardiovasc Surg 115:506-516, 1998
5. Dearani JA, Danielson GK, Puga FJ, et al: Late follow-up of 1095 patients undergoing operation for complex congenital heart disease utilizing pulmonary ventricle to pulmonary artery conduits. Ann Thorac Surg 75:399-411, 2003
6. Breymann T, Thies WR, Boethig D, et al: Bovine valved venous xenografts for RVOT reconstruction: Results after 71 implants. Eur J Cardiothorac Surg 21:703-710, 2002
7. Rowlatt UF, Rimoldi H, Lev M: The quantitative anatomy of the normal child's heart. Pediatr Clin North Am 10:499, 1963
8. Bonhoeffer P, Boudjemline Y, Qureshi SA, et al: Percutaneous insertion of the pulmonary valve. J Am Coll Cardiol 39:1664-1669, 2002

From the Cardiothoracic Unit, Great Ormond Street Hospital for Children National Health Service Trust, London, United Kingdom.

Address reprint requests to Carin van Doorn, MD, FRCS (C/Th), Cardiothoracic Unit, Level 2 Cardiac Wing, Great Ormond Street Hospital for Children NHS Trust, Great Ormond St., London WC1N 3JH, United Kingdom. E-mail VandoC@gosh.nhs.uk.

1522-2942/03/0803-0000$30.00/0
doi:10.1053/S1522-2942(03)00037-2