Target range of Qp/Qs in neonate with SV has not been elucidated. Barnea and colleagues [5] suggested that maximal oxygen delivery is attained with Qp/Qs of 0.5 to 1.0. Tweddell and associates [4] stated that their postoperative management after the Norwood procedure was aimed at achieving Qp/Qs of 0.8 to 1.2. Pearl and associates [6] indicated that optimal Qp/Qs is in the range of 0.7 to 1.0. Our policy is the following: when anaerobic metabolism is indicated by depleted SvO_2 as low as 0.3, accumulated blood lactate, and metabolic acidosis, then Qp/Qs is targeted at 1.0 to obtain maximal tissue oxygen saturation because mathematical model predicts that Qp/Qs of 1 provides maximal SvO_2 [2]; when SvO_2, lactate, and acid-base balance indicate aerobic metabolism, then Qp/Qs is targeted between 0.5 and 1.0 to prepare for increasing Qp/Qs.

Possible drawbacks of Qp/Qs-oriented management in neonates with TAPVC and SV are twofold. First, placement of caval blood sampling line or spectrophotometric catheter is invasive. It may cause infection or thrombosis. Frequent blood sampling increases transfusion dose, and thereby chance of its adverse effects. Second, calculated Qp/Qs may deviate from the true value. Pulmonary congestion or atelectasis may make pulmonary venous oxygen saturation below 0.96. Placement of caval catheter close to the atrium or in the cava connected to the pulmonary veins allows pulmonary venous blood contamination in venous samples, making measured caval oxygen saturation deviate higher. If pulmonary venous oxygen saturation is below 0.96 or caval oxygen saturation is incorrectly high, calculated Qp/Qs can be lower than the true value, leading to incorrect patient management. Nevertheless, we believe that Qp/Qs monitoring facilitates appropriate balancing between tissue perfusion and oxygenation, detects redundant ventricular volume-load, and will improve outcome of obstructive TAPVC repair in neonates with SV.

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CASE REPORTS

Anatomic Correction for Corrected Transposition After Pulmonary Unifocalization

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A Senning plus Rastelli operation was performed in a patient who had a rare combination of congenitally corrected transposition of the great arteries (S,L,L) with dextrocardia, major aortopulmonary collaterals, and diminutive central pulmonary arteries with arborization defects. The patient required four preparatory operations including bilateral unifocalizations of the aortopulmonary artery collaterals. Pulmonary artery to systemic pressure ratio after the double switch operation was 0.6. The patient demonstrates good biventricular function on echocardiogram at 3 months after the operation.

(Ann Thorac Surg 2003;75:1012–4)

The use of the double switch operation (DSO) for the management of congenitally corrected transposition of the great arteries (ccTGA) is becoming increasingly widespread [1]. Surgical management is especially difficult in the presence of diminutive central pulmonary arteries with arborization defects and major aortopulmonary collateral arteries (MAPCAs). We encountered this very rare disease entity, which required staged unifocalization of the MAPCAs before the DSO.

An 8-month-old male patient was referred with ccTGA (S,L,L) with a large ventricular septal defect, atrial septal defect, dextrocardia, pulmonary atresia with very small confluent pulmonary arteries and arborization defects. A total of five MAPCAs supplied nine pulmonary segments in the following pattern: two segments of the right upper lobe, three segments of the right lower lobe, two segments of the left lower lobe, and two segments of the left upper lobe. The central pulmonary arteries measured 1.5 to 2 mm on angiogram. Echocardiography demonstrated normal biventricular function without tricuspid or mitral regurgitation.

Four preparatory operations were performed: operation 1, end-to-side anastomosis of the main PA to the ascending aorta at 8 months old (Melbourne shunt); operation 2, unifocalization of the right lower lobe MAPCA, modified Blalock-Taussig (B-T) shunt using a 5-mm polytetrafluoroethylene (PTFE) graft at 15 months

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old; operation 3, unifocalization of two MAPCAs to the left lung, 5-mm left B-T shunt at 16 months old; and operation 4, extensive patch angioplasty of the branch pulmonary arteries, division of the three previous shunts, and insertion of a 6-mm PTFE shunt from right innominate artery to the reconstructed pulmonary artery at 23 months old. The systemic oxygen saturation (SaO_2) was maintained at 75% to 80% throughout these operations.

A cardiac catheterization immediately after the last operation revealed a pulmonary vascular resistance of 10 Wood units and systemic vascular resistance of 12.8 Wood units. The patient then underwent repeat cardiac catheterization at 4 years and 4 months. The final study revealed a Qp/Qs of 0.8, mean pulmonary artery pressure of 26 mm Hg, and a pulmonary vascular resistance that had fallen to 4 Wood units.

At 4 years and 10 months of age the patient underwent complete repair at a weight of 15.4 kg. Before the operation a "classic" repair was still an option with ventricular septal defect (VSD) closure and creation of a left ventricle to pulmonary artery connection. However, at operation, there was very limited space on the left ventricular free wall between multiple coronary arteries for conduit placement. Therefore, a Senning-Rastelli operation was performed using an 18-mm Hancock conduit for the right ventricle to pulmonary connection as described [2]. A Shumacker modification [3] of the Senning procedure was used for reconstruction of the pulmonary venous chamber. Patch angioplasties of the left and right upper pulmonary artery branches were also performed (Fig 1). The patient developed complete atrioventricular block after the procedure and a dual-chamber epicardial pacemaker system was implanted on postoperative day 5. The pulmonary artery to systemic pressure ratio was 0.6 after the repair as determined in the operating room.

The patient had right diaphragmatic paralysis and respiratory failure requiring tracheostomy as postoperative complications. The patient underwent right diaphragmatic plication but required mechanical ventilator support for 88 days postoperatively. A postoperative echocardiogram demonstrated mild right ventricular dysfunction with an estimated systolic pressure of 60 mm Hg at the time of discharge. Right ventricular function had improved to normal on an echocardiogram obtained 3 months after surgery.

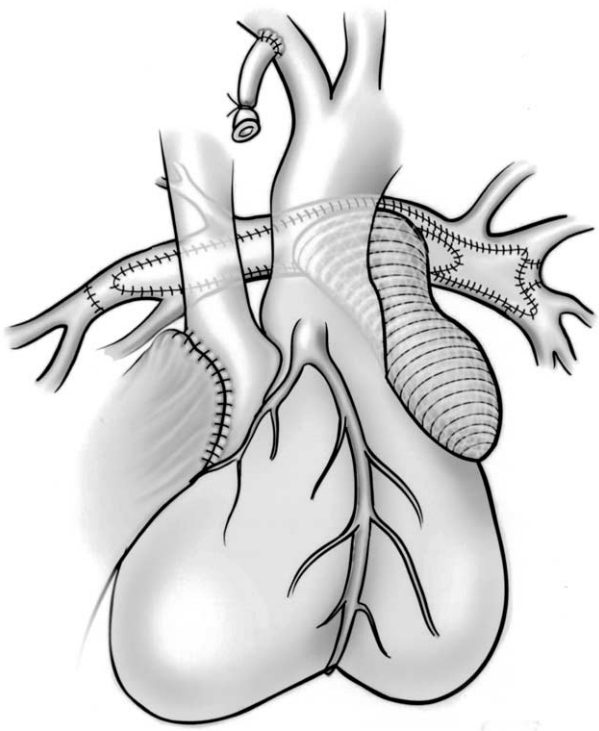

Fig 1. The scheme of the anatomy at the time of final repair.

Comment

The combination of pulmonary atresia with MAPCAs and ccTGA is extremely rare; complete repair has not been reported previously. The management strategy for MAPCA was derived from our experience with tetralogy of Fallot with pulmonary atresia. Our operative approach includes creation of a central end-to-side anastomosis of the diminutive main pulmonary artery to the aorta to optimize the chances for growth of the small central pulmonary arteries. Staged operations then are performed to unifocalize the MAPCAs to the central pulmonary arteries. This patient had severe central and first order branch pulmonary artery stenosis after unifocalization and required an additional operation for extensive PA reconstruction, take down of the previous shunts, and to insert a single central shunt before final repair.

The theoretical advantages of committing the morphologic left ventricle to the aorta for ccTGA has been extensively discussed [4–6]. In this case, the final decision to proceed with DSO was made intraoperatively because of the coronary distribution on the free wall of the left ventricle and not because of preexisting right ventricular dysfunction or tricuspid regurgitation. Maintaining the lowest possible pulmonary vascular resistance is an important point to ultimately allow right ventricle to pulmonary artery connection at the time of the DSO. Otherwise, the deleterious effects of a right ventriculotomy and future conduit failure can significantly diminish the theoretical benefits of DSO on right ventricular function. Low threshold for conduit replacement will be important for the long-term preservation of right ventricular function in this patient. Despite this patient's protracted postoperative course with right ventricular dysfunction and the requirement of mechanical ventilatory support, the hemodynamics seem satisfactory at 3 months after surgery with marked improvement of right ventricular function.

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CASE REPORTS

Retroaortic Innominate Vein With Coarctation of the Aorta: Surgical Repair and Embryology Review

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A newborn girl with a retroaortic innominate vein, coarctation of the aorta, ventricular septal defect, and subaortic stenosis underwent a complete repair at 8 days of age. The ascending aorta was transected and the innominate vein was brought in front of it. We review the anatomy and embryology of this rare entity and describe the surgical technique to avoid recoarctation and innomante vein compression.

(Ann Thorac Surg 2003;75:1014–6)

A left retroaortic brachiocephalic (innominate) vein is a rare condition. It has an incidence of 0.2% to 1% among congenital cardiac anomalies [1–3]. Although is was first described by Kerschner [4] more than 100 years ago, the clinical experience with this anomaly is limited with less then 100 cases described worldwide [1–5]. It is frequently associated with right aortic arch or high left aortic arch [1, 6]. We believe that this association is not accidental. Herein we review the embryology to support our theory and provide a basis for surgical correction.

A newborn girl weighing 3.12 kg was noted to have a murmur at birth with weak femoral pulses. Her arm-to-leg blood pressure gradient was 20 mm Hg. Echocardiogram demonstrated an unrestrictive ventricular septal defect (VSD) with posterior malalignment of the outlet septum, subaortic stenosis, restrictive patent foramen ovale (PFO), almost closed ductus, bicuspid aortic valve and good biventricular function. Aortic annulus was 0.5 cm, pulmonary annulus 1.1 cm, tricuspid annulus 1.2 cm, ascending aorta proximal to left carotid artery (LCA) was 0.45 cm. There was a juxtaductal coarctation at the level of the left subclavian artery (LSA). There was 1-cm and 0.31-cm in diameter narrow segment of the arch between the LCA and LSA The patient was started on prostaglandin (PGE1) at 0.05 $\mu g \cdot kg^{-1} \cdot min^{-1}$. She remained stable and underwent elective surgery on March 20, 2002 at the age of 8 days.

The surgery was performed through the midline sternotomy. There was an incidental finding of the retroaortic innominate vein (Fig 1A). The patient was place on cardiopulmonary bypass (CPB) in standard fashion using ascending aortic and bi-caval cannulation. Patient was cooled to 25°C. Descending aorta and arch vessels were mobilized. After cardioplegic arrest, the PDA was ligated and circulatory arrest was established for 25 minutes. During the period of circulatory arrest, ascending aorta was transected and the innominate vein was brought in front of the ascending aorta. The hypoplastic aorta was ligated just beyond the LSA and ductal tissue was resected. The continuity of the ascending aorta was reestablished by a "semilunar" anastomosis and the descending aorta was anastomosed into the ascending aorta end-to-side (Fig 1B). CPB was resumed. A subaortic myectomy was performed and the ventricular septal defect was closed with a Dacron Sauvage patch. PFO was closed with direct suture. Total aortic cross-clamp time was 93 minutes. Total CPB time was 79 minutes. Upon weaning off pump transesophageal echocardiogram (TEE) was performed and demonstrated unobstructed flow in the ascending aorta and through the end-to-side aortic anastomosis. There was flow acceleration in the subaortic area with a mean gradient of 8 mm Hg. Patient developed complete heart block and required sequential pacing for 5 days but regained normal sinus rhythm on postoperative day 5. She was discharged home 10 days after surgery.

Comment

In our patient, there was a significant distance between the origin of the left common carotid and the left subclavian artery, and a high aortic arch. These findings are typical of "pseudocoarctation" of the aorta, which distinguishes itself from a true coarctation by a high aortic arch and the absence of clinical signs or hemodynamic abnormalities, with the trans-stenotic pressure gradient remaining below 25 mm Hg [7, 8]. Embryologically, this anomaly results from a lack of shortening of the left dorsal and ventral aortic roots of the left fourth aortic arch [8]. At the eighth week both anterior cardinal veins are interconnected by the superior and inferior capillary plexuses (Fig 2A). Normally, the aortic arch shortens during the embryological development and occupies the space of the inferior transverse capillary plexus, thus causing its regression, while the rest of venous blood shunts into the superior transverse capillary plexus. This facilitates normal development of the innominate vein

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