day. The postoperative computed tomography scan showed antegrade perfusion of the left leg as a result of decompression of the false lumen and expansion of the true lumen as well as patency of the crossover bypass (Fig 2). The patient was discharged as ambulatory on the eighth postoperative day.

Comment

Malperfusion is a common problem in patients with acute type A dissection, with a reported incidence of malperfusion of a lower limb in 15% of patients [1]. Usually the malperfusion resolves after repair of the type A dissection as a result of decompression of the false lumen and expansion of the false lumen. On the other hand, immediate treatment is favorable because complete arterial occlusion will lead to irreversible tissue damage within 6 hours. This is the reason why several authors suggest a staged approach in patients with type A dissection complicated by malperfusion that addresses first the malperfusion and, in a second step, the repair of the type A dissection. As the risk of mortality from an acute type A dissection is approximately 1% per hour in the first 24 hours [4], this approach carries a substantial risk of mortality as it delays treatment of the dissection [3]. A concomitant procedure avoids this risk. This approach allows the beginning of reperfusion of the ischemic leg already during cooling of the patient for deep hypothermic circulatory arrest. This should reduce ischemia–reperfusion injury because experimental studies have shown that local hypothermia during early reperfusion protects skeletal muscle from ischemia–reperfusion injury [5]. Additionally, reperfusion of the leg starts significantly earlier and is not dependent on the decompression of the false lumen.

In conclusion, our approach is a safe and effective method to address the aortic dissection and the lower limb malperfusion simultaneously without the risk of limb loss caused by prolonged limb ischemia or the risk of dissection-related mortality attributable to delayed dissection repair with the benefit of protective cold reperfusion of the ischemic leg.

References


Valve-Sparing Neoaortic Root Replacement Late After the Norwood and Fontan Procedures

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Neoaortic root dilatation is a significant and inevitable late complication in patients after the Norwood operation. In this report, we describe a patient who had left pulmonary artery compression and severe neoaortic regurgitation from a severely dilated neoaortic root 11 months after the Norwood and Fontan operations and after palliation for pulmonary atresia.

Fig 2. Postoperative computed tomography scan with antegrade perfusion (blue arrow) of the left iliac artery and patent crossover bypass (red arrow).

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It is generally known that neoaortic root dilation is a common late complication after the definitive operation for congenital heart defects such as tetralogy of Fallot or transposition of the great arteries. With improvement in late results for hypoplastic left heart syndrome and its variants, neoaortic aneurysmal dilation and neoaortic valvular dysfunction were also reported in these patients late after the Norwood operation [1–3]. Surgical intervention for neoaortic root dilation has been performed in these patients [4, 5]. We performed valve-sparing, neoaortic root replacement in a patient who had aortic aneurysmal dilatation combined with pulmonary artery compression and severe aortic regurgitation 11 years after the Norwood procedure.

A male neonate diagnosed with double-inlet left ventricle, transposition of the great arteries, coarctation of the aorta, and subvalvular aortic stenosis underwent the Norwood procedure at 12 days of age. The neoaorta was reconstructed without a prosthesis. Two years after the Norwood operation, he underwent a subsequent Fontan operation. Cardiac catheterization at 6 years (4 years after the Fontan operation) demonstrated neoaortic root dilation with a diameter of 33 mm at the level of the sinus of Valsalva (Z value, +8.6; calculated from normal aortic sinus diameter). Neoaortic regurgitation was not detected. During follow-up, the neoaortic root gradually dilated. The diameter at the level of the sinus of Valsalva at 11 years was 39 mm (Z value, +8.7) with mild neoaortic regurgitation. The left pulmonary artery was severely compressed from the dilated neoaortic sinus of Valsalva (Fig 1). Left pulmonary arterial blood flow was partially interrupted, and the Fontan circulation was barely able to be maintained by single, right-lung perfusion. Central venous pressure was 13 mm Hg, but left pulmonary artery pressure was 20 mm Hg. Arterial oxygen saturation was 90% in room air.

Surgical intervention was performed using moderate hypothermic cardiopulmonary bypass. The left femoral artery and vein were exposed before the sternotomy in case of unexpected bleeding. Arterial cannulas were inserted in both the distal aortic arch and the left femoral artery. Venous cannulas were inserted directly into both venae cavae. Because the original aorta with coronary arteries could be separated from the neoaorta and cross-clamped, retrograde cardioplegia was not required. After cross-clamping the original aorta and inducing cardioplegic arrest, the wall of the expanded neoaortic root was completely resected. Degenerative changes in the neoaortic cusps were not detected. Horizontal mattress sutures were passed from inside of the neoaorta and placed through the base of a 24-mm Valsalva graft (Terumo; Cardiovascular Systems Corporation, Ann Arbor, MI). The residual neoaortic wall was fixed inside the graft. We obtained good cusp coaptation by adjusting the height of the commissures and attaching each nodule of Aranti by a 7-0 monofilament suture. The original aorta, from which the coronary arteries arose, was anastomosed to the side hole of the graft in an end-to-side fashion. The distal end of the graft was anastomosed to the ascending aorta proximal to the brachiocephalic trunk. Concomitantly, the compressed area of the left pulmonary artery was augmented using an expanded polytetrafluoroethylene patch (Gore-Tex; W.L. Gore & Associates, Inc, Flagstaff, AZ).

Postoperative computed tomography demonstrated a straight aortic root and good coronary perfusion. The stenosis of the left pulmonary artery was relieved (Fig 2). Histologic analysis demonstrated edematous changes of the intima and fragmentation of elastic fibers in the vascular wall. Inflammation of the vessel or a connective tissue disorder was not detected. Postoperatively, the Fontan circulation was maintained.
adequately. Arterial oxygen saturation was 96% in room air at the time of discharge.

Comment

The origin of the pulmonary artery is used as a channel for systemic blood flow in patients with congenital heart disease, such as transposition of the great arteries or hypoplastic left heart syndrome, and patients after the Ross operation. Also, the ascending aorta in tetralogy of Fallot is sometimes dilated preoperatively. In such patients, neoaortic root dilatation or neoaortic valve regurgitation develops gradually late after the operation and often requires surgical intervention [1–3].

The mechanism of neoaortic dilatation is incompletely understood. It is generally thought that the dilatation is caused mainly by a structural difference between the aorta and pulmonary artery, and exposure of the pulmonary artery to systemic pressure [1]. However, it has also been reported that dilatation of the neoaorta is related to semilunar valve regurgitation, the type of material used during arch reconstruction, and arterial wall disease associated with a bicuspid valve [5, 6]. Cohen and colleagues [3] reported that the neoaortic root progressively dilated out of proportion to body size with time in 98% of hypoplastic left heart syndrome patients who underwent staged reconstruction. It seems that the dilatation of the neoaorta is inevitable in this population. Neoaortic valvular dysfunction is an absolute indication for reoperation, but reoperation for only neoaortic root dilatation is still controversial. It is unknown whether neoaortic root dilatation may cause neoaortic root rupture.

In general, surgical intervention is recommended for an aortic root aneurysm larger than 55 mm in diameter to avoid rupture in adult patients [7]. A neoaortic diameter of 55 mm might also be used as a tentative criterion in pediatric patients; however, because of different underlying mechanisms, it is difficult to make a decision for reoperation based only on the diameter of the neoaorta. Severe compression of surrounding tissues such as the pulmonary artery, superior vena cava, and bronchus caused by a dilated neoaorta should be a determining factor for reoperation.

In our case, neoaortic regurgitation was progressive and the left pulmonary artery was compressed by the dilated neoaorta. Pulmonary artery stenosis is a serious complication of the Fontan circulation. Because the preoperative circulation may have been maintained by only right pulmonary perfusion, early salvage of the left pulmonary artery was required before serious complications developed. There have been only a few reports of aortic valve-sparing surgery after the Fontan operation [4, 5], but it is expected that neoaortic dilatation may be detected in more patients and require surgical repair in the future. It is necessary to discuss the surgical strategy for these patients including technical maneuvers to prevent late aortic root dilatation. Especially in Fontan candidates, early reoperation is recommended before neoaortic root dilatation and regurgitation cause severe pulmonary stenosis and Fontan failure.

References

Biventricular Repair of Double-Outlet Right Ventricle and Closing Ventricular Septal Defect

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We report an infant with double-outlet right ventricle and progressive left ventricular failure caused by a closing apical muscular ventricular septal defect detected during follow-up, who achieved successful biventricular repair after surgical opening and enlargement of a closed perimembranous ventricular septal defect.


In double-outlet right ventricle (DORV) with remote ventricular septal defect (VSD), restriction of the VSD results in left ventricular (LV) hypertension leading to a fatal condition [1, 2]. Left ventricular hypertrophy caused by the restrictive VSD is a risk factor for early death in patients undergoing definitive biventricular repair [3]. In rare cases, progressive VSD restriction may require emergency palliative interventions for LV decompression. Herein we report a patient with DORV and a rapidly closing apical VSD who underwent successful biventricular repair after LV decompression by surgical opening and enlargement of a closed perimembranous VSD.

A male patient with prenatal diagnosis of DORV with a body weight of 2,738 g was referred to us immediately after birth. On admission, symptoms of congestive heart failure were absent, and the oxygen saturation was 90% under ambient air. Transthoracic echocardiography revealed [S,D,D] (Situs Solitus, D-loop ventricles, D-malposition of the great arteries) DORV, remote VSD, mild valvular pulmonary stenosis, and a patent foramen ovale. The VSD, 6 mm wide, was located in the apical muscular portion of the septum (Fig 1). The peak blood flow velocity through the VSD was 0.7 m/s from left to right. No other VSD was recognized. The LV end-diastolic volume (LVEDV) was 76% of the expected normal value, and the LV ejection fraction (LVEF) was 0.78. The peak blood flow velocity through the pulmonary valve was 3.1 m/s with adequate pulmonary blood flow. He was discharged at the age of 23 days.

During follow-up, the muscular VSD had become progressively restrictive, and he was readmitted at the age of 2 months because of heart failure. Transthoracic echocardiography revealed marked LV enlargement with a suprasystemic LV pressure (peak VSD flow, 3.3 m/s). Besides the closing 2-mm-wide apical VSD, shunt flow across two pinholes in a bulging fibrous tissue at the subaortic perimembranous portion was detected (Fig 2). The restrictive interatrial communication caused pulmonary congestion and pulmonary hypertension. Emergency balloon atrial septostomy was unsuccessful. The LVEDV was 256% of normal, and the LVEF was 0.21. Plasma brain natriuretic peptide level was 8,850 pg/mL. Electrocardiogram showed inverted T waves in the left precordial leads. A palliative surgery for decompression of the left heart was planned.

Through a median sternotomy, cardiopulmonary bypass was initiated. Atrial septectomy was performed. The perimembranous VSD was obscured by accessory valvular tissue. The tissue was totally resected to make a 3-mm interventricular communication in the perimembranous portion. The communication was further enlarged by incising the ventricular septum anteriorly by 3 mm. The pulmonary trunk was banded to achieve a mean pulmonary artery pressure of 20 mm Hg. The patient was weaned from cardiopulmonary bypass without difficulty. Postoperative transthoracic echocardiography at the age of 3 months showed a 5-mm-wide perimembranous VSD with the peak flow velocity of 1.5 m/s. The LVEDV was 60% of normal, and the LVEF was 0.69. There was no atrioventricular valve regurgitation, and the interatrial communication was widely open. High echogenicity along the LV endocardium was noted, suggesting endocardial fibroelastosis. Electrocardiogram still showed inverted T waves in the left precordial leads. Plasma brain natriuretic peptide level decreased to 110 pg/mL. The patient was discharged on maintenance therapy of carvedilol and angiotensin-converting enzyme inhibitor.

Cardiac catheterization at the age of 8 months showed equal right ventricular and LV pressures, and good LV function. The T-wave abnormality in the left precordial leads was normalized. The LVEDV was 88% of normal, and the arterial oxygen saturation was 77%. A modified Blalock-Taussig shunt was performed.

Repeated cardiac catheterization at the age of 2 years showed equal right ventricular and LV pressures with the LVEDV of 111% of normal and the LVEF of 0.64. The perimembranous VSD was 8 mm in width, and the muscular VSD was ignorable. Elective biventricular repair was planned.

At the age of 2.5 years and a body weight of 10 kg, the patient underwent surgery under cardiopulmonary bypass. The perimembranous VSD, 7 mm in diameter, was further enlarged anteriorly by 5 mm. Intraventricular rerouting was performed using a piece of an expanded...