A New Approach to Correction of Tetralogy of Fallot With Absent Pulmonary Valve

Viktor Hraška, MD

Department of Cardiovascular Surgery, Children's University Hospital, Bratislava, Slovakia

A new technique for primary repair of tetralogy of Fallot with absent pulmonary valve syndrome is suggested. In addition to the standard intracardiac portion of tetralogy of Fallot repair, this approach includes translocation of the pulmonary artery anterior to the aorta, shortening of the ascending aorta, and shortening and plication of the pulmonary artery. This new technique should eliminate compression of the tracheobronchial tree caused by impingement by the pulmonary artery.

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Although several techniques for correction of tetralogy of Fallot with absent pulmonary valve have been described, the method of choice is still controversial [1, 2]. In particular, symptomatic infants have a poor prognosis because of severe central pulmonary artery (PA) dilatation and bronchial compression. Primary repair with pulmonary arterioplasty has been proposed to reduce bronchial obstruction. A new technique that should eliminate compression of the tracheobronchial tree is described.

Technique

After median sternotomy, subtotal thymectomy is performed and the pericardium is opened. 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 using bicaval cannulation and distal cannulation of the ascending aorta. During cooling, the left and right PAs, including the first PA branches in the hilum of each lung, are dissected free and mobilized. At a rectal temperature of 25°C, the distal ascending aorta is clamped, and cold blood cardioplegia is delivered into the proximal ascending aorta. A vertical incision is made in the infundibular portion of the right ventricle with a short transannular extension of this incision. A limited amount of infundibular septum (parietal band) is resected. The large malalignment type of ventricular septal defect is closed with a polyethylene terephthalate fiber (Dacron) patch using a continuous suture technique either through a transatrial approach or through the ventriculotomy. 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.

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Address reprint requests to Dr Hraška, Department of Cardiovascular Surgery, Children's University Hospital, Limbova 1, 833 40 Bratislava, Slovakia; e-mail: hraska@dkch.dfnsp.sk.

A transverse aortotomy is performed above the commissures, and a tubular or triangular segment of the aorta is resected. This maneuver brings the ascending aorta down and to the left (Fig 1). The PA is transected above the annulus and brought anterior to the aorta [3]. At this point, and 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 (Fig 2). The aortic cross-clamp is released and rewarming is begun. A PA plication may be performed at this point if necessary. Before weaning from cardiopulmonary bypass, left atrial, right atrial, and PA catheters are placed for postoperative monitoring as are atrial and ventricular pacemaker wires. Modified ultrafiltration is performed as soon as the patient has been weaned from bypass.

Results

Between November 1998 and February 1999 this technique was used in 2 infants with a diagnosis of tetralogy of Fallot with absent pulmonary valve.

Patient 1

A 4,000-g infant was referred to our department at 7 months of age. She was hospitalized with a 3-month history of respiratory tract infections despite antibiotic treatment and a failure to thrive. Marked tachypnea, irritability, and failure to thrive were noted at the time of admission. The diagnosis was established by echocardiography. The chest roentgenogram revealed left lung hyperinflation. Angiography showed aneurysmal dilatation of both PAs. She tolerated the above operation without difficulties. She came off bypass on low inotropic support and remained hemodynamically stable. Postoperatively she required ventilatory support for 7 days. Echocardiographic examination before discharge revealed no signs of SVC or PA obstruction. Five months postoperatively she is clinically well and has no signs of respiratory infection, and her weight has doubled.

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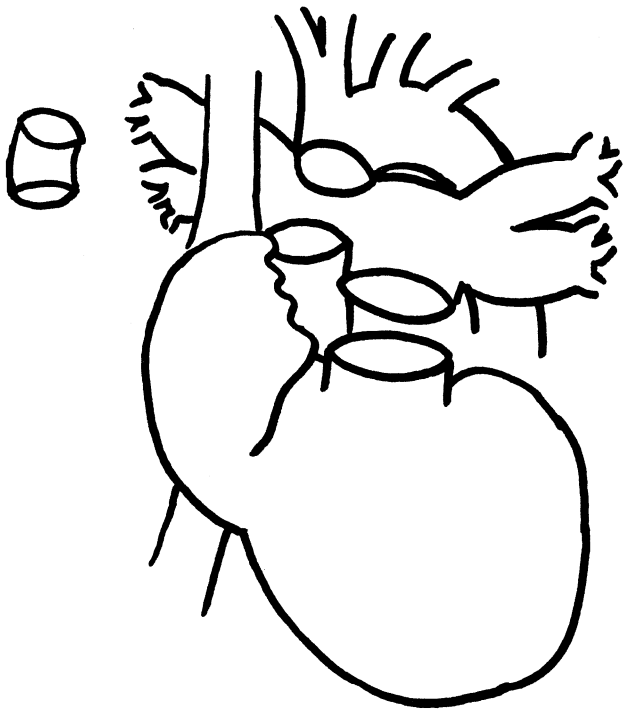

Fig 1. Tubular or triangular segment of the aorta is resected. This maneuver brings the ascending aorta down and to the left.

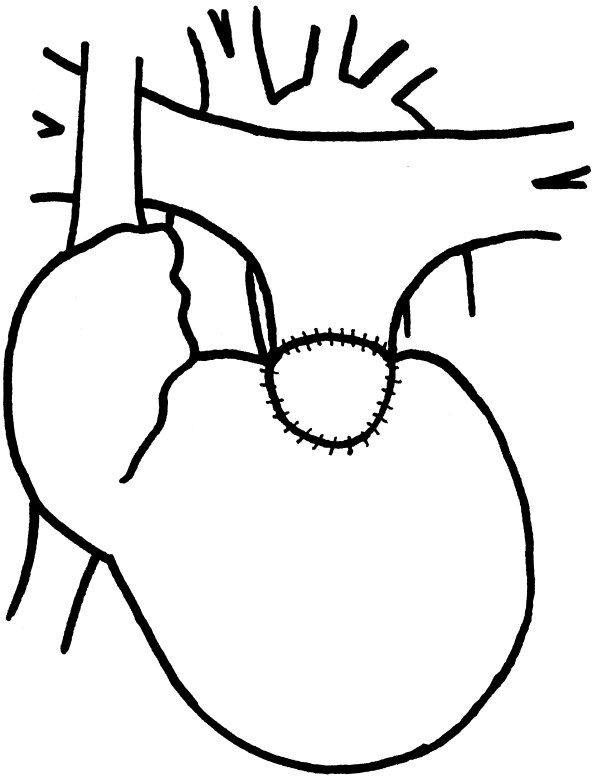

Fig 2. The pulmonary artery is transected above the annulus and brought anterior to the aorta.

Patient 2

A 3,300-g infant was admitted to our department at the age of 4 months with progressive respiratory distress. She required parenteral antibiotic therapy. Echocardiography, angiography, and chest roentgenogram showed typical massive dilatation of PAs with bilateral air trapping and hyperinflation of both lungs. She underwent the above correction. In the postoperative course, she was hemodynamically stable and required ventilatory support for 2 days. After weaning from mechanical ventilation, she required continuous chest physiotherapy for 2 weeks. No signs of SVC or PA obstruction were noted on postoperative echocardiographic examination. One month after operation she is doing very well.

Comment

Controversy persists regarding the management of patients with tetralogy of Fallot with absent pulmonary valve and progressive respiratory distress. A number of surgical techniques for reduction of bronchial obstruction have been proposed with variable results [1, 2].

A new technique described herein reduces or should eliminate bronchial compression by the PA. Translocation of the PA anterior to the aorta displaces the dilated PA anteriorly, away from the trachea and bronchial tree. 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, shortening the ascending aorta allows the aorta to ultimately reside posteriorly and to the left of its usual location. This maneuver calls for thorough mobilization of the aortic arch and brachiocephalic vessels. Shortening of the ascending aorta and mobilization of the PA beyond the pericardial reflection avoids potential compression of the right coronary artery and SVC. Another relevant detail is shortening the left PA (which is always too long) by oblique transection of the PA trunk with connection to the right ventricular outflow tract. Finally, plication of the PA can decrease wall tension and prevent later development of aneurysmal dilatation of the PA.

Addendum

From March 1999 to September 1999, two additional symptomatic infants with tetralogy of Fallot with absent pulmonary valve have undergone this repair, with good early results.

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References

1. Snir E, de Leval MR, Elliott MJ, Stark J. Current surgical technique to repair Fallot's tetralogy with absent pulmonary valve syndrome. Ann Thorac Surg 1991;51:979–82.
2. Godard F, Houyel L, Lacour-Gayet F, et al. Absent pulmonary valve syndrome: surgical treatment and consideration. Ann Thorac Surg 1996;62:136–42.
3. Lecompte Y, Zannini L, Hazan E, et al. Anatomic correction of transposition of the great arteries. J Thorac Cardiovasc Surg 1981;82:629–31.