freedom from failure of 92% at 10 years.1 Obstructed pulmonary venous pathway was described as an early complication of the lateral tunnel cavopulmonary connection as a result of improper baffled creation.2 Technical improvements have reduced this complication, and in our experience it is rare. Previous reports3-4 underline the relative greater frequency of this complication in heterotaxy when associated with anomalous pulmonary and systemic venous connections, which can make surgical repair complicated with the need to create a tortuous and potentially obstructive pathway. Despite this concern, a recent report from our institution5 has shown excellent outcome with cavopulmonary connection in patients with heterotaxy syndrome and anomalous pulmonary venous return. This is likely due to improved surgical technique and use of extracardiac conduit in cases with complex atrial and venous anatomy (18%).

None of the 3 patients had evidence of obstruction at the pulmonary venous pathway early after the Fontan procedure, as determined by 2-dimensional echocardiography. Pulmonary venous obstruction developed slowly through a period of several months to years. In patient 2, pulmonary venous obstruction developed 4 months after device closure of the fenestration, suggesting that the additional scarring from device implantation contributed to the obstruction. The obstruction was due to fibrotic tissue at either the interatrial septum or in the interatrial course of the pulmonary veins (patient 2).

To prevent this complication, and to ensure a wide open pulmonary venous outflow, particularly in patients with mitral atresia or stenosis, we currently enlarge the communication between the pulmonary veins and the right atrium by unroofing the coronary sinus posteriorly into the left atrium, in addition to resection of the interatrial septum.

In conclusion, late onset obstruction of pulmonary venous pathway after lateral tunnel cavopulmonary connection is a rare but serious complication that may be prevented by unroofing the coronary sinus at the time of Fontan procedure.

References

Complex double-outlet right ventricle repair in a neonate with complete tracheal agenesis

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T racheal agenesis is a rare malformation with fatal consequences in very early life. We report the case of a baby girl with tracheal agenesis who during early infancy underwent complete repair of her associated (S,D,D) double-outlet right ventricle with supracardiac total anomalous pulmonary venous connection and is still alive at the age of 10 months.

Clinical Summary
The patient was born spontaneously at a gestational age of 35 weeks. Birth weight was 2 kg, and Apgar score at 1 minute was 2, increasing to 6 after 2 minutes of mask ventilation. Critical clinical conditions required intubation, but the tube failed to pass the larynx under direct laryngoscopy. A blind end of the larynx was visualized with the aid of a flexible endoscope, whereas the tracheal bifurcation could be visualized from the distal esophagus through a tracheoesophageal fistula. A Portex 3 tube (Portex, Inc, Keene, NH) was positioned through the esophagus into the fistula, achieving a satisfactory bilateral ventilation.

Helical TC scan showed complete agenesis of the trachea (Floyd type II) with a distal tracheoesophageal fistula (Figure 1). Echocardiography showed (S,D,D) double-outlet right ventricle (with subaortic ventricular septal defect), total anomalous pulmo-
nary venous connection to the right superior vena cava, and unrestricted atrial septal defect.

After 30 days, the patient underwent a complex palliative operation to separate the airway from the alimentary tract, consisting of (1) division of the esophagus below the fistula and gastrostomy (for alimentation), (2) gastroenteroanastomosis (to resolve pylorus and proximal duodenal atresia), (3) Merkel diverticulum resection, and (4) double cervical esophagostomy. The proximal esophagostomy was created to divert salivary secretion protecting the airway; the distal esophagostomy was used as the entrance of the newly created airway, after positioning a 10 by 50-mm Rusch Polyflex esophageal stent (Rusch Inc, Duluth, Ga) to keep the esophagus air patent. A tracheostomy 5.5-mm (external diameter) Shiley cannula (Shiley, Inc, Irvine, Calif) was positioned through the lower esophagostomy into the stented esophagus, connecting the infant to the ventilator (Figure 2).

Because of failure to thrive and severe cyanosis, the infant underwent catheterization, which confirmed the cardiac diagnosis. Complete surgical repair was carried out, consisting of (1) intracardiac left ventricle–aorta pericardial baffling, (2) right superior vena cava division (above the drainage of the four pulmonary veins); (3) intra-atrial pericardial patch baffling of the right superior vena cava orifice (draining all four pulmonary veins) to left atrium through the atrial septal defect, and (4) anastomosis of the proximal right superior vena cava to the right atrial appendage. Cardiopulmonary bypass and cardioplegic cardiac arrest times were 166 and 94 minutes, respectively, and body temperature was lowered to 26°C. The child was taken to the postoperative cardiac intensive care unit and could be weaned from mechanical ventilation at 7 days.

Currently the patient is 10 months old and is waiting for airway and alimentary tract surgical reconstruction. Her hemodynamic status is unremarkable, and she has gained weight, being now in the 50th percentile.

Discussion

Tracheal agenesis, or congenital absence of the trachea, is a very rare malformation accounting for less than 1:50000 live births. It was first described in 1900 by Payne. Since that time, fewer than 100 cases have been reported in the literature. According to Floyd and coauthors tracheal agenesis is classifiable into three anatomic subtypes: type I, agenesis of the proximal trachea, with a normal distal trachea connected to the esophagus through a tracheoesophageal fistula; type II, complete agenesis of the trachea, with the two main stem bronchi fused in the midline at the carina, which arises from a tracheoesophageal fistula; and type III, complete agenesis of the trachea, with the two main stem bronchi arising separately from the esophagus. Associated congenital malformations and congenital heart disease account for 90% and 70%, respectively, of the cases reported in the literature. Some authors refer to tracheal agenesis as part of multimalformation syndromes, such as VA(C)TER(L) (vertebral anomalies, anal atresia, cardiac abnormalities, tracheoesophageal fistula or esophageal atresia, renal agenesis and dysplasia, limb defect) and TACRD (tracheal agenesis or atresia, complex congenital cardiac malformations, radial ray defects, duodenal atresia) syndromes.

Clinical presentation is characterized by rapid onset of severe respiratory distress at birth, absence of audible crying, and impos-
sibility of endotracheal intubation. Survival is due to either ‘inci-
dental’ endoesophageal intubation or prompt surgical tracheo-
tomy (Floyd type I).

Surgical repair or palliation of associated congenital cardiac
lesions has been never described in the literature, because most
affected patients usually die before any attempt at surgical inter-
vention. This is the first patient to our knowledge to survive
palliation of the respiratory tract malformation as well as definitive
intracardiac repair. She is now thriving and is currently being
evaluated for definitive surgical reconstruction of the airway and
alimentary tract.

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Formation of a stenotic fibrotic membrane at the distal anastomosis of
bovine jugular vein grafts (Contegra) after right ventricular outflow tract
reconstruction

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T he surgical correction of many congenital right ventric-
ular outflow tract (RVOT) anomalies necessitates the
interposition of an extracardiac valved conduit. A vari-
ety of conduits have been used, and a few years ago the
xenogenic valved conduit Contegra (Medtronic, Inc, Minneapolis,
Minn) was introduced into clinical practice.1-3 The graft consists
of a jugular bovine vein segment containing a native trilea-
fl et valve. It presents several advantages relative to other available
conduits because it demonstrates a high pliability of the graft
tissue, provides abundant conduit material proximal and distal to
the valve for RVOT reconstruction, and is readily available in a
large range of sizes.

Although a few reports have been published, little information
exists regarding the fate of the conduit after implantation. The
resection of 2 Contegra grafts in our experience amounting to 67
implantations prompted us to report these cases and to analyze
the reason for conduit failure.

Clinical Summary
Between May 2001 and January 2003, the Contegra conduit was
implanted in 67 patients for RVOT reconstruction, including cor-

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rection of tetralogy of Fallot and pulmonary atresia (n = 34), Ross
procedure (n = 17), correction of truncus arteriosus (n = 7),
Rastelli procedure (n = 4), and other procedures (n = 5). Rou-
tinely performed intraoperative and postoperative echocardiogra-
demonstrated excellent conduit performance in all patients. At
12 and at 8 months after implantation, however, 2 patients (3%)
showed increasing pressure gradients as great as 50 mm Hg at the
distal anastomoses and eventually required reoperation with graft
replacement (Figure 1, A). Both patients had undergone repair of
tetralogy of Fallot with a 12-mm Contegra conduit, which was
selected because of the patients’ diminutive pulmonary arteries.

Pathologic Findings
Macroscopic inspection revealed the formation of an internal annular
membrane anchoring at the level of the distal anastomosis, with
proximal extensions into the valve sinuses in both explanted grafts.
The valve leaflets were unaffected and well preserved (Figure 1, B).
Histologic analysis demonstrated a fibrinous composition of the
membranes partially covered with granulation tissue and the pres-
ervation of the collagen structures of all three vessel wall layers of
the conduits with, however, a reduced density of the medial
collagen fibers. The infiltration with lymphocytes and macro-
phages was observed in all layers but was predominantly in the
adventitia and on the internal lumina of the grafts.

Discussion
Because of the recent introduction of the Contegra conduit, expe-
riences regarding its long-term performance are limited, and until
now only a single conduit failure, an aneurysmal dilatation, has
been reported.4 Here we report 2 cases of conduit failure from the
formation of an intimal membrane that led to severe RVOT ste-
nosis at 8 and 12 months after implantation. Both patients had been
treated for tetralogy of Fallot and severe hypoplasia of the pulmo-
nary arteries and had received the smallest available conduit size

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