Clinical and experimental aspects of tracheal stenosis
Dodge-Khatami, A.

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Complete Repair of Tetralogy of Fallot with Absent Pulmonary Valve Including the Role of Airway Stenting

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Abstract

Tetralogy of Fallot (TOF) with absent pulmonary valve (APV) represents an extreme form of tetralogy where pulmonary insufficiency and mild annular stenosis often results in massive pulmonary artery (PA) dilatation. The aneurysmal left and right PAs often compress the adjacent trachea and bronchi, leading to airway obstruction and respiratory failure in infancy. Between 1991 and 1997, 11 patients underwent a single stage repair of TOF and APV using a valved (10) or non-valved (1) homograft conduit and PA reduction arterioplasty. There was one perioperative (1/11=9.1%) and one late death (9.1%). Both deaths were related to airway complications. Morbidity associated with postoperative respiratory complications and ventilator-dependency due to underlying tracheo-bronchomalacia is a significant problem. Intermediate follow-up shows a high incidence of reintervention for conduit stenosis and/or insufficiency and tracheo-bronchial compression. These infants also required multiple hospitalizations for recurrent respiratory infections secondary to their tracheo-bronchomalacia. Stenting of the right and left main bronchi with balloon expandable metallic stents is a new experimental therapy that has been useful in 2 recent patients with respiratory failure despite satisfactory intracardiac repair. It may provide an attractive alternative therapy to prolonged mechanical ventilation with positive end expiratory pressure in patients with severe tracheobronchial malacia. Complete repair with a valved homograft conduit and reduction pulmonary arterioplasty in infancy at the time of diagnosis is the procedure of choice for infants with TOF with APV. With this approach the patient outcome is essentially determined by their airway status and airway management.
Introduction

Tetralogy of Fallot (TOF) with absent pulmonary valve (APV) was first described by Chevers in 1847. This accounts for 3-6.5% of all tetralogy cases, and represents an extreme form of tetralogy of Fallot. It is unique in that the congenital absence of the pulmonary valve produces an aneurysmal dilatation of the main, right, and left pulmonary arteries (PA) with resulting pulsatile compression of the tracheo-bronchial tree (Figure 1). This causes varying degrees of tracheo-bronchomalacia which yields the potential for severe respiratory insufficiency early in life. General consensus divides these patients into two groups according to the severity of symptoms and age at presentation which correlates well with operative survival and long-term prognosis. At one extreme are infants (less than 1-2 months of age) who present with severe respiratory insufficiency and the need for urgent intervention. At the other extreme are older children with only mild cyanosis and minimal airway symptoms who may have delayed repair of their cardiac lesion and who tend to have less postoperative and long-term respiratory morbidity. The purpose of this study is to review the outcome of 11 patients in whom one-stage complete repair of tetralogy of Fallot in infancy was performed, paying particular attention to our routine use of a valved homograft conduit and to the role of airway stenting as a new treatment of respiratory insufficiency from tracheo-bronchomalacia after satisfactory cardiac repair.

Figure 1. Preoperative pulmonary angiography revealing aneurysmal pulmonary arteries.
### Chapter 4

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Ao=aortic, Aox=aortic cross-clamp, ASD=atrial septal defect, A-V block=atrio-ventricular block, CA=circulatory arrest, CPB=cardiopulmonary bypass, C-E=Carpentier-Edward porcine bioprosthesis, FU=Follow-up, L=left, LOS=length of stay.
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PA=pulmonary artery, PDA=patent ductus arteriosus, PFO=patent foramen ovale, R=right, SVT=supraventricular tachycardia, PVR=pulmonary valve replacement, RIMA=right internal mammary artery, RV=right ventricle, VSD=ventricular septal defect
Materials and Methods

Patients

Between October 1991 and August 1997, 10 infants and 1 child were operated on for TOF with absent pulmonary valve syndrome at the Children's Memorial Hospital in Chicago (Table 1). Mean age was 122 days (range 4 days to 30 months) and mean weight was 4.53 kg (range 2.7-8.3 kg). Nine patients presented with signs of respiratory distress along with cyanosis. Two of these infants were transferred from other institutions intubated and mechanically ventilated for severe respiratory failure.

Clinical findings included moderate to severe cyanosis, wheezing, tachypnea, and a harsh pansystolic murmur over the precordium. A diastolic murmur at the left sternal border and an absent second sound of severe pulmonary insufficiency were equally noted. Diagnostic features on chest radiography included cardiomegaly and greatly dilated central pulmonary arteries, while peripheral pulmonary vascularity was reduced. Preoperative bronchoscopy was performed in all patients. All patients underwent two-dimensional echocardiography (n=11) and 5 had cardiac catheterization. Two patients had a discontinuous left pulmonary artery supplied by a patent ductus arteriosus (PDA). Two patients had DiGeorge Syndrome. Surgical intervention was undertaken on an urgent or emergent basis in almost all cases.

Operative Technique

Operative repair was performed through a median sternotomy with aortobicaval cannulation, continuous cardiopulmonary bypass, hypothermia at 26-28°C and cold blood cardioplegia. A right superior pulmonary vein vent was used in all cases. After the aortic cross-clamp was applied and cardioplegia delivered, a vertical right ventriculotomy was performed. Right ventricular outflow tract relief was achieved by resecting hypertrophied infundibular muscle bands. The VSD was then closed using an elastic Dacron patch anchored with interrupted plegetted 5-0 Ticron sutures. This was done through the right ventriculotomy in all but 3 cases in which transatrial closure of the VSD was performed. The right atrium was entered to locate a concommitant patent foramen ovale (PFO) or ostium secundum atrial septal defect (ASD). An ASD was closed by direct suture in 4 cases and a PFO or small ASD was knowingly left open in the remaining 7 cases. This was done in anticipation of possible high right-sided pressures at the end of repair.

An attempt was made to relieve extrinsic airway compression by performing an anterior reduction pulmonary artery arterioplasty when necessary (n= 9). Not surprisingly, only the
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two asymptomatic infants did not require a PA arterioplasty, as their pulmonary vascular anatomy did not appear to produce airway compression at the time of surgery. Bilateral quadrangular incisions were then made anteriorly in the right and left pulmonary arteries with excision of the largest possible surface area of aneurysmal PA (Figure 2). The posterior wall of the PA and its adhesions to the bronchi were left undisturbed. Caution was taken to correctly extrapolate the future anastomosis with the valved conduit as to not create even minimal stenosis at the PA bifurcation. The openings in the right and left PA's were closed primarily. After full deairing maneuvers, the aortic cross-clamp was released and the remaining conduit reconstruction was performed with a fibrillating or beating heart. A cryopreserved valved pulmonary (n=4, sizes 10-17) or aortic (n=6, sizes 9-16) homograft was anastomosed first distally to the PA bifurcation and then proximally to the RVOT using a running polypropylene suture. These smaller sized homografts were related both to the very low weight of neonates and limited supply of homografts at the time of surgery, most of which were done on an emergency basis. The proximal homograft reconstruction was performed with an aortic or pulmonary homograft hood (n=8) or a pericardial hood (n=2). In one case, transatrial infundibulectomy and VSD closure was done along with PA arterioplasty without a homograft or conduit insertion. Retrospectively, this proved to be a problem as this patient was a

Figure 2. Repair of ventricular septal defect through an infundibulectomy with a Dacron patch and excision of excess aneurysmal pulmonary artery tissue in view of reduction arterioplasty. Insert shows proposed incisions in the pulmonary arteries.
postoperative outlier in regards to hospital stay and respiratory morbidity and eventually required valve insertion. In 2 patients, the left PA was discontinuous with the main pulmonary artery and supplied by a patent ductus arteriosus. Reimplantation of the left PA to the pulmonary trunk and PDA ligation were performed in both of these patients. Intraoperative transesophageal echocardiography was done in 5 patients to assess adequacy of repair, and is currently a standard part of the operation.

Results

There were two deaths in this series (18.2%), one perioperative (9.1%) and one late (9.1%). One child who required an open sternum covered by a silastic patch for 9 days because of hyperinflated lungs, developed Candida and Pseudomonas sepsis, and died on postoperative day 50. This patient was transferred to our institution intubated and ventilated after presenting with severe respiratory distress from diffuse tracheo-bronchomalacia. The other died on postoperative day 202 from generalized sepsis during her second hospitalization. She had been discharged after an uneventful first postoperative stay on day 33, but required readmission after an accident at home involving chest trauma with fractured ribs. After reintubation, increasingly higher levels of PEEP were required to maintain airway patency. Persistent respiratory failure from airway collapse due to severe tracheo-bronchomalacia required placement of intraluminal Palmaz balloon expandible metallic stents in both mainstem bronchi. She eventually underwent reoperation for homograft insufficiency and received a larger sized homograft with repeat reduction pulmonary arterioplasty, but ultimately expired from generalized sepsis.

Mean cardiopulmonary bypass time was 140.9 minutes (range 109 to 182) and mean aortic cross-clamp time was 60.2 minutes (range 46 to 110). Pathologic analysis showed degenerative myxoid changes of the vestigial pulmonary valve components in 5 out of 9 specimens. Patients remained intubated a mean of 16.9 days (range 5 to 50). Mean hospital stay was 13.4 days (range 14 to 50). There was one outlier (did not have a valved homograft at initial repair) who remained hospitalized for 273 days and who eventually required a tracheostomy before reoperation to insert a valved homograft. This patient was not used for the intubation or hospital stay calculations.

Mean follow-up was 25.5 months (range 1.7 to 67 months). Of the nine surviving patients, NHYA functional class is markedly improved with 4 patients in class I and 5 in class II. All
patients are in sinus rhythm and seven have right bundle branch block (RBBB). Reoperation for conduit insufficiency or stenosis was thus far required in 5 patients. After homograft valve removal, pulmonary valve replacement (PVR) was performed in three patients using a porcine Carpentier-Edwards (C-E) valve (two valved-conduits sizes 14 and 18 and one simple #19 valve). One of these three patients had previously undergone a balloon angioplasty dilatation of the distal conduit for stenosis at 306 days post-operatively. Carpentier-Edwards prostheses were chosen for their relative rigidity to better sustain the extrinsic compression produced by the surrounding tissues and sternum. One infant had a larger size aortic homograft (size 14) inserted to replace a freely regurgitant size 11 homograft 153 days after the initial procedure. At the same time, repeat PA reduction arterioplasty for recurrent bilateral aneurysmal PA dilatation and closure of a previously left open PFO were performed. The fifth reoperation involved the only patient without an initial homograft and who is not included in the statistical analysis. This infant had persistent and severe ventilator-dependent respiratory failure requiring a tracheostomy on postoperative day 27. Reoperation was performed on postoperative day 224 with RV patch takedown, insertion of a size 13 aortic homograft and a right PA reduction arterioplasty. She was discharged on day 273 but still requires night-time ventilation.

Of note is a technical adjunct that was performed after the fifth reoperation in regards to the proximal conduit/homograft anastomosis to the ventriculotomy. After the homograft takedown, we initially inserted a valved porcine C-E conduit. The child failed to wean from the cardiopulmonary bypass with significant ECG changes and ventricular tachycardia compatible with severe acute left coronary insufficiency. The C-E conduit was taken down and the most

Figure 3. Incision into the right ventricular infundibulum to provide a wider outflow tract and implantation base for the new homograft. Note the posterior sutures at the pulmonary annulus in proximity of the left main coronary artery.
superior aspect of the right ventriculotomy was brought together with interrupted pledgeted sutures to lessen any possible tension in the proximity of the left main coronary artery (Figure 3). We then inserted an aortic homograft whose proximal anastomosis to the RV was well away from the coronary artery and the child managed to come off cardiopulmonary bypass easily with no evidence of coronary ischemia. This technique was repeated on our last reoperation and yielded a good result. We therefore recommend that after pulmonary artery transection, the free superior edges of the right ventriculotomy (site of the previous homograft valve suture line) be sutured directly together, allowing the future proximal conduit/homograft anastomosis to be at a safe distance from the left main coronary artery. Median interval to reintervention was 283 days (Kaplan-Meier estimation, range 153 to 719). The probability of freedom from reintervention at 1 year was 75%.

Several patients had severe tracheo-bronchomalacia with episodes of recurrent respiratory distress requiring re-intubation and ventilation with high levels of PEEP. In two of these patients Palmaz balloon expandable metallic stents were placed in both mainstem bronchi with a rigid bronchoscope under fluoroscopic guidance (Figure 4). In both patients the stents employed were 10 mm in length. The balloon sizes used (which determines the intraluminal diameter) were 4 mm, 5 mm, 5.5 mm and 6 mm. One patient, a 7 month old girl with DiGeorge syndrome died on postoperative day 202 from generalized sepsis despite bronchoscopic evidence of improvement in her airway patency. The other child, now 2.2 years of age, is doing well at home.

**Figure 4.** Postoperative bronchogram demonstrating intrabronchial Palmaz metallic stents
Discussion

In our series of infants with tetralogy of Fallot with absent pulmonary valve, we had one operative mortality and one late mortality, both patients with airway complications expiring from generalized sepsis. The incidence of reoperation in our infants who had an initial homograft insertion was 40% (4/10). In some respects, this reoperation rate is to be expected, given the inherent problem of using a non-growing prosthesis. We feel the use of a pulmonary valve as part of the repair has resulted in an improvement of clinical symptoms, increased exercise tolerance, improved right ventricular function, and less airway symptoms.

Tetralogy of Fallot with absent pulmonary valve is a rare and distinct clinical entity with its own unique set of clinical findings and management pitfalls. The pathogenesis of this syndrome is still controversial, as the structural microscopic findings of the pulmonary artery wall are variable according to different studies. Embryologic studies implicate an absent ductus arteriosus which causes fetal pulmonary regurgitation, leading to massively dilated pulmonary arteries, and tracheobronchial compression 9,10,13.

Historically, the two clinical extremes in the spectrum of TOF with absent pulmonary valve have been described and correlate well with age 2-12. At one end, neonates present with severe respiratory insufficiency and may already have at birth severe tracheo-bronchomalacia due to poor in utero airway growth from extrinsic compression by the aneurysmal PA's 9,10,13. These patients present with cyanosis, failure to thrive, episodic choking, and difficulty eliminating secretions because of an inefficient cough mechanism. The recurrent pulmonary infections can lead to bronchiectasis, chronic infection, lobar emphysema and parenchymal scarring. Ultimately, fixed hypercapnic respiratory failure and associated obstructive sleep apnea may ensue 14. At the other end of the spectrum, older children may present with much milder symptoms of cyanosis and dyspnea on exertion with a clinical course similar to "standard" TOF. Currently, this bipolar description is no longer of as much practical significance, as nearly all cases of TOF with APV are now diagnosed early. The distinction between infant and adult types is therefore no longer so important, and the type and rapidity of management is dictated by the presence or not of respiratory symptoms from tracheobronchomalacia.

In severely ill neonates, respiratory stability is the first priority. Postural drainage, humidity, oxygen, nebulizer therapy, intubation and mechanical ventilation are initiated as required. Continuous high PEEP (10 cm H2O) helps keep airways open throughout the respiratory cycle and is routinely used at our institution for these patients 14. Some infants have been reported to have an excellent response to prone ventilation 3, as this position results in less posterior compression on the
airways by the enlarged PA's, which fall forward toward the sternum.
The type of surgical intervention for TOF and absent pulmonary valve has evolved considerably.
Initially, two-stage procedures were recommended for severely symptomatic neonates and infants. Pulmonary artery ligation with a systemic-to-pulmonary shunt was done to allow patient and airway growth until elective definitive repair. This approach, however, yielded a high mortality rate in infants with severe respiratory compromise. Other attempted surgical procedures reported in the 1960s and 1970s included suspension of the dilated PA to the posterior sternal fascia, reimplantation of the pulmonary arteries, lobectomy, and superior vena cava-to-right PA anastomosis. Complete correction before the 1980s was mostly reserved for older, less symptomatic children. More recently, advances in neonatal surgical and cardiopulmonary bypass techniques with better myocardial preservation allow a more aggressive approach with complete primary repair, even in severely ill neonates. Although most authors agree that reduction PA arterioplasty is needed, controversy still exists as to the necessity of a valved or non-valved right outflow conduit.

Godart, Planché et al. reported nine infants in whom one-stage repair included reduction PA arterioplasty, VSD closure, RVOT excision and transannular patch without pulmonary valve insertion. Overall mortality was 22.2%, and mortality correlated with longer CPB and aortic cross-clamp times, reflecting more complex anatomy. No relation was found to age at operation, residual RV / PA pressure gradients, or RV to LV systolic pressure ratios. At a mean follow-up of 30 months, there was one late death and one reoperation. Although the lack of a valve in the pulmonary position leaves patients with pulmonary insufficiency, that group does not believe that valved conduits are needed in these patients. They recommend pulmonary valve insertion only if there is distal pulmonary stenosis or pulmonary hypertension.

Waterson, Mee et al. reported a series of 19 infants without pulmonary valve insertion with a perioperative mortality of 16% and one late death. Their follow-up includes 5 reoperations. They also do not believe in using a pulmonary valve at initial repair, as only one of their reoperations has required a homograft for free PL. They argue that the systematic insertion of a valved conduit at initial repair constitutes an increased risk for reoperation for conduit change in a population already anticipated to have a high incidence of reoperations.

Supporting our approach, Snir, Stark et al. report a series of eight infants in whom complete repair included insertion of a cryopreserved valved homograft. Postoperative mortality was 25%, and at a mean follow-up of 3.6 years there were no reoperations. More recently, McDonnell et al. reported 28 patients undergoing one-stage repair including anterior and posterior PA reduction arterioplasty and RV outflow tract reconstruction with either a patch
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(n=19), a valved conduit (n=5), or a monocusp valve (n=4). Their early mortality was 21.4% and they had one late mortality. Reoperation consisted of repeat PA plication and placement of a valved conduit and was necessary in 10.7% of cases. Only preoperative intubation constituted an independent risk factor for reoperation or death. They concluded that insertion of a valved homograft or monocusp in the RVOT associated with anterior and posterior reduction PA arterioplasty is advisable at the initial repair, although they acknowledge an increased incidence of reoperation with this approach. In an earlier series from our hospital, 10 patients with TOF and APV were repaired. Four had pulmonary valve insertion at the time of repair. Five of the 6 patients not receiving a pulmonary valve remained symptomatic and 3 needed a subsequent pulmonary valve insertion. All patients receiving a valve were symptom-free. Improvement in RV function, exercise performance, decrease in PA size, and cardiothoracic ratio were more marked in those patients who had primary insertion of a pulmonary valve. The conclusion of this review was that pulmonary valve insertion should be performed at the time of the intracardiac repair.

Despite advances in surgical techniques for repair of the cardiac lesions in TOF and APV, survival remains guarded in many instances due to the respiratory compromise from tracheobronchomalacia. The standard medical therapy for this has been positive pressure ventilation with or without a tracheostomy. The use of balloon expandable metallic stents (Palmaz, Johnson & Johnson) in a series of children was reported by Filler and associates in 1995. Subramanian and colleagues reported the specific use of Palmaz stents in a child with tetralogy of Fallot with absent pulmonary valve. That child had recurrent episodes of cyanosis and respiratory distress after complete repair of tetralogy of Fallot with absent pulmonary valve at 2 weeks of age. At 5 months of age she had placement of bilateral Palmaz bronchial stents with complete relief of respiratory symptoms. Hagl and associates described a technique of external stabilization using ring-reinforced polytetrafluoroethylene prostheses in 7 children. Multiple pledgedet suture were placed extramucosally to the dysplastic tracheal wall, through the prosthesis, and tied under video-assisted bronchoscopy. This achieved immediate reexpansion of the collapsed segments without tracheal resection and thus allowed for more rapid postoperative extubation. At a mean followup of 38 months, patients are reported in excellent clinical condition and free of stridor. We have reported our results with placement of 12 balloon expandable metallic stents in 6 children. Two of these children had tetralogy of Fallot with absent pulmonary valve and are also reported in this review. The stents in properly selected patients certainly allow earlier weaning from mechanical ventilation with ensuing earlier discharge from the hospital. The primary early complication as well as a mid-term
problem remains granulation tissue formation, and we are still concerned enough about long-term growth potential to be cautiously optimistic about their role in patients with tetralogy of Fallot with absent pulmonary valve. It is emphasized that expandable metallic angioplasty stents represent an experimental therapy in tracheobronchomalacia. Currently we feel their use should be limited to infants failing conventional therapy.

Tetralogy of Fallot with absent pulmonary valve is a well-recognized distinct clinical entity that may present early in life as a surgical emergency, in respect to both cardiac and respiratory systems. Currently, we recommend complete surgical repair in infancy with a valved homograft, VSD closure, and reduction pulmonary arterioplasty. We believe early reduction pulmonary arterioplasty is a crucial step which allows early relief of the respiratory insufficiency which often becomes the life-threatening priority in these neonates. Operative mortality can be reduced and postoperative hemodynamics are optimized with the use of a valved homograft in the pulmonary position. We have used intra-bronchial balloon expandable metallic angioplasty stents as an adjunct to reduction PA arterioplasty in maintaining large airway patency in selected severely symptomatic patients. High levels of PEEP may thus be avoided on a long-term basis, allowing for less ventilator dependency and earlier extubation. In these patients, serial bronchoscopy is mandatory to assess airway status and allows dilatation or intraluminal débridement of granulation tissue as required. Long term follow-up is necessary for both the cardiac and respiratory systems.

References