Pediatric Cardiac Surgery Annual

# Surgical Repair of Total Anomalous Pulmonary Venous Connection

Kirk R. Kanter

**The diagnosis of total anomalous pulmonary venous connection (TAPVC) is made when all four pulmonary veins drain anomalously to the right atrium or to a tributary of the systemic veins. It constitutes between 1% and 1.5% of all children with congenital heart disease and can be categorized by the site of drainage into the systemic circulation (supracardiac, 45%; infracardiac, 25%; cardiac, 25%; mixed, 5%). The clinical presentation is different if the pulmonary venous drainage is unobstructed (heart failure, mild cyanosis) or obstructed (respiratory failure, severe heart failure). Surgical management depends on the anatomic type. Obstructed TAPVC requires urgent surgical intervention, whereas unobstructed TAPVC can be dealt with electively; although this is usually operated on once the diagnosis is made. Postoperative pulmonary artery hypertension can be problematic. Recent surgical results with isolated TAPVC have improved, with operative mortality consistently at less than 10%. A particularly challenging group of patients are those with single ventricle physiology and TAPVC with high operative mortality and poor long-term survival.**
**Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 9:40-44** 

**KEYWORDS: Total anomalous pulmonary venous connection, congenital heart disease, pulmonary hypertension, pulmonary veins**

The diagnosis of total anomalous pulmonary venous connection is made when all four pulmonary veins drain anomalously to the right atrium or to a tributary of the systemic veins. It constitutes between 1% and 1.5% of all children with congenital heart disease. Although total anomalous pulmonary venous connection (TAPVC) can be associated with other heart defects, especially in children with polysplenia or asplenia or with heterotaxy syndrome, for the purpose of this discussion we will mainly address those with isolated total anomalous pulmonary venous connection or those with other minor anomalies, such as patent ductus arteriosus or atrial septal defect.

Typically, TAPVC presents in infancy. The presentation depends on the site of drainage of the pulmonary veins and whether or not the veins are obstructed. Infants with unobstructed TAPVC will present with evidence of a large left-to-right shunt with heart failure, cardiomegaly, and mild desaturation. Children with obstructed TAPVC will present more with respiratory issues, hypoxia and even low cardiac output, as well as pulmonary artery hypertension. Occasionally, infants with obstructed TAPVC are referred for extracorporeal membrane oxygenation (ECMO) caused by respiratory failure or pulmonary hypertension. The cardiac diagnosis is made during a routine pre-ECMO echocardiogram. Children with obstructed TAPVC require urgent correction, whereas those with unobstructed TAPVC can have a more elective repair, although typically these children will be corrected once diagnosed.

TAPVC is categorized into four broad categories defined by the site of entry of the anomalous connection to the systemic venous circulation.[1] The most common is supracardiac TAPVC. The pulmonary veins drain to a confluence posterior to the heart and then to a vertical vein, most commonly on the left, which enters the innominate vein and then drains to the right atrium. The left heart receives oxygenated blood through a patent foramen ovale. These children typically present in infancy and are referred for operation once diagnosed. Supracardiac TAPVC accounts for roughly 45% of children with TAPVC.

Division of Cardio-Thoracic Surgery, Department of Surgery, Emory University School of Medicine and Children's Healthcare of Atlanta at Egleston, Atlanta, GA.

Presented at the AATS Congenital Heart Disease Symposium 85th Annual Meeting of the American Association for Thoracic Surgery, San Francisco, CA, April 10–13, 2005.

Address reprint requests to Kirk R. Kanter, MD, Division of Cardio-Thoracic Surgery, Emory University School of Medicine, 1365 Clifton Road, Atlanta, GA 30322; e-mail: kkanter@emory.edu


1092-9126/06/$-see front matter 
doi:10.1053/j.pcsu.2006.02.015

The second most common type of TAPVC is of the infracardiac variety, which accounts for approximately 25% of children with TAPVC. Most often, the pulmonary veins are obstructed and the child presents with respiratory distress and pulmonary artery hypertension. The pulmonary venous confluence is oriented more vertically than is typical with supracardiac TAPVC. The confluence drains to a vein that enters the systemic venous bed below the diaphragm, typically into the ductus venosus or, less commonly, the hepatic veins. Occasionally the pulmonary veins are small or have intimal proliferation, which makes the repair more challenging.

With the third type of TAPVC, the pulmonary veins drain to the coronary sinus (cardiac TAPVC). This accounts for $\leq$ 25% of cases of TAPVC. It is uncommon for these children to have an obstructed picture; more often they will present with heart failure or cyanosis. Occasionally these children can survive into adulthood before the diagnosis is made.

The final type of TAPVC is the mixed type, which is a combination of any of the above three types of TAPVC. Presentation depends on the dominant form of the drainage to the anomalous veins. Mixed TAPVC occurs in approximately 5% of cases.

The clinical picture of obstructed pulmonary veins with TAPVC varies with the anatomic subtype. For supracardiac TAPVC, it can occur in up to 40% of patients when the vertical vein drains to the innominate vein. This is a particularly common if the vertical vein courses between the left pulmonary artery and the left main stem bronchus, where a vascular vise-type effect will occur in contrast to the typical course of the vertical vein anterior to the left pulmonary artery. We have seen one such infant presenting with obstructed TAPVC and concurrent respiratory syncitial virus pneumonia. We managed this child by stenting the obstructed vertical vein percutaneously in the catheterization lab and allowing the child to get over his pulmonary problems. After about a week, he was taken to the operating room and underwent uneventful repair of his supracardiac TAPVC with removal of the stent and ligation of the vertical vein.

Other anatomic subtypes of supracardiac drainage are more prone to obstruction. When the anomalous veins drain directly to the right superior vena cava (SVC), roughly 65% will be at least partially obstructed. When they drain to the azygos vein, almost all of these patients will present with an obstructed picture.

Infracardiac TAPVC almost always presents with an obstructed picture because the pulmonary venous confluence drains commonly to the sinus venosus, which constricts shortly after birth. These patients can present as a true emergency requiring immediate operation.

It is uncommon for children with cardiac-type TAPVC, where the anomalous veins drain to the coronary sinus, to have an obstructed picture; this accounts for only about 20% of these patients. Similarly, patients with mixed TAPVC will present with an obstructed picture only about 40% of the time.

Diagnosis of TAPVC is routinely made by echocardiography.[2] Only rarely is cardiac catheterization now necessary; it is typically reserved for patients with coexisting complex malformations requiring further definition or occasionally patients with mixed TAPVC in whom all of the pulmonary veins cannot be clearly delineated by echocardiography alone.[3] In fact, Serraf et al[4] advised against preoperative angiography in children with an obstructed presentation, arguing that this delays operation and causes clinical deterioration.

## Surgical Repair

The first surgical repair is attributed to Muller and colleagues at UCLA. This was done in 1951 without the aid of cardiopulmonary bypass by anastomosing the left atrial appendage to the pulmonary venous confluence. The first total repair was performed by Lewis and Varco at the University of Minnesota in 1956, using the technique of inflow occlusion. The first repair on cardiopulmonary bypass was by Kirklin at the Mayo Clinic in 1956.

In the past, almost all infants with TAPVC were repaired using profound hypothermia and circulatory arrest, although now it can be performed with bicaval cannulation and low flow hypothermic perfusion. Circulatory arrest has the advantage of allowing a bloodless field with excellent exposure of the pulmonary venous confluence without the need for unnecessary manipulation or clamping of the pulmonary veins. We tend to use bicaval cannulation in all but the smallest of neonates ($<$ 2.0 kg). Standard metal-tipped right angled cannulae are introduced directly in the inferior vena cava just above the diaphragm and the SVC near the junction with the innominate vein. In children with supracardiac TAPVC, the SVC is considerably larger than normal (because of the additional blood flow from the anomalously draining pulmonary veins), which facilitates direct cannulation. In those rare infants with supracardiac TAPVC draining to the azygos vein, rather than placing a tourniquet around the SVC cannula, we will place a delicate Yassergill neurosurgery clip inferior to the azygos vein but way from the sinus node to allow decompression of the pulmonary veins through the SVC cannula during cooling.

With bicaval cannulation, the cannulae are well out of the way so one is able to get excellent exposure of the pulmonary venous confluence, especially after cardioplegic arrest. On occasion, it is helpful to introduce brief periods of circulatory arrest during the most critical portions of the operation to optimize surgical exposure with a nearly bloodless field. However, with bicaval cannulation and low-flow hypothermic perfusion, we find that this is rarely necessary. In these infants we have not recognized complications of direct caval cannulation such as superior vena caval thrombosis or postoperative chylothorax, as long as one takes the necessary precautions with meticulous technique in cannulating and decannulating these vessels. We do try to remove indwelling central venous lines such as internal jugular vein lines as early as possible postoperatively to avoid such line complications and tend to rely on intraoperatively placed intracardiac lines.

Many techniques have been described for repair of supracardiac TAPVC. Commonly, once on bypass and after car-

dioplegic arrest, the pulmonary veins are exposed in the posterior pericardium by dividing the pericardial reflection between the right atrium and the pulmonary venous confluence.[5] An incision is made in the pulmonary venous confluence which is typically oriented in a transverse fashion. A corresponding incision is made in the posterior wall of the left atrium, which sometimes can be quite small. It is helpful to properly orient this incision by first opening the right atrium and placing a right angle clamp through the patent foramen ovale tenting up the back wall of the left atrium near the atrial septum on the right. The incision is started at this point and then directed toward the base of the left atrial appendage making a transverse incision in the left atrial back wall that corresponds to the incision in the pulmonary venous confluence. A direct anastomosis is now made between these two incisions. Obviously, the anastomosis needs to be as large as possible. The anastomosis can be augmented by dividing the left-sided vertical vein as far superiorly as possible at its entry into the innominate vein. An incision is made on the right medial aspect of the vertical vein down to where it enters the pulmonary venous confluence. This vein is then flapped down medially onto the superior aspect of the anastomosis of the pulmonary venous confluence to the back of the left atrium. This flap of vein will serve to augment the anastomosis with a pedicled flap of autologous vein, as well as to increase the effective volume of the left atrium.

The anastomosis can be performed from the right side of the operating table starting at the base of the left atrial appendage connecting it to the pulmonary venous confluence near the left upper pulmonary vein. Then one works rightward, completing the anastomosis at the junction of the atrial septum and the pulmonary venous confluence near the confluence of the right-upper and right-lower pulmonary veins. The last portion of the anastomosis can use interrupted suture technique.

An alternative approach to this technique of repair of supracardiac TAPVC is to approach the anastomosis from the left side of the operating table with the apex of the heart tipped up and to the right.[6] The anastomosis starts at the right side and the surgeon works toward himself to the base of the left atrial appendage connecting the pulmonary venous confluence near the left-upper pulmonary vein. The patent foramen ovale is then closed either primarily or with a patch if there is concern about impingement on the pulmonary venous anastomosis. Then the vertical vein is typically ligated. The heart is closed and the child is rewarmed and weaned from cardiopulmonary bypass.

If there is concern about postoperative pulmonary artery hypertension and subsequent right ventricular failure, a lifesaving technique can be to leave the patent foramen ovale partially open to allow for decompression of the right side through the atrial septum with right-to-left shunting at the expense of some systemic arterial desaturation.

Hawkins et al[7] described a direct approach to the left atrial to pulmonary venous confluence anastomosis by first excising the fossa ovalis through the right atrium. A generous transverse incision is made in the back wall of the left atrium and a corresponding incision is made in the pulmonary venous confluence. The pulmonary venous confluence is now anastomosed to the back wall of the left atrium directly through the fossa ovalis which is then closed with a pericardial patch. The authors maintain that this technique eliminates the possibility of distortion of the anastomosis because it is performed with the heart in an anatomically correct position.

An alternative technique for repair of supracardiac TAPVC is to perform a bi-atrial transverse incision as first described by Shumacker and King[8] and others.[9] This consists of making a transverse right atrial incision across the crista terminalis then through the atrial septum and through the posterior wall of the left atrium. Under direct vision, the posterior wall of the left atrium can now be directly anastomosed to the pulmonary venous confluence. The atrial septal defect is typically closed with a piece of pericardium or other patch material. The incision in the right atrium may be closed primarily or with a separate patch if necessary. This technique has the advantage of enhanced exposure of the pulmonary veins, but has more suture lines on the atrium and raises the possibility of a higher incidence of atrial arrhythmias. Michielon et al[10] reported a significantly increased risk of late arrhythmias with this technique. There is a theoretical advantage of increasing the size of the left atrium with the double patch technique, but a study from Vanderbilt showed near normal left atrial volumes postoperatively with more standard surgical techniques.[11]

Some investigators have even advocated approaching the repair of supracardiac TAPVC superiorly between the SVC and the ascending aorta through the transverse sinus.[12–14] The pulmonary venous confluence is connected to the dome of the left atrium.

The surgical repair of infracardiac TAPVC is quite similar to that for supracardiac TAPVC. Often, however, the pulmonary venous confluence is oriented vertically so that the incision in the confluence is more of a hockey stick incision, running vertically and then on to the left upper pulmonary vein to correspond with a similar incision on the back wall of the left atrium from the left atrial appendage medially and inferiorly. It is not necessary to ligate the draining vein below the diaphragm and, in fact, some have advocated routinely leaving this vein open in patients with an obstructed presentation.[15] The advantage of leaving the vertical vein unligated is that it theoretically can allow decompression of the pulmonary venous channel in the early postoperative course through the sinus venosus in the face of a small, poorly compliant left atrium. Although Shah et al[16] reported on two patients with unobstructed supracardiac TAPVC managed without ligation of the vertical vein who subsequently developed significant left-to-right shunts, in our practice we have not recognized any problems with residual shunts by leaving this channel unligated as has been shown by Cope et al.[15]

On the other hand, if the left atrium is quite small, it can be helpful to divide the vertical vein and use this remnant as an onlay patch to augment to the size of the left atrium.[13,17] This technique involves wide mobilization of the pulmonary veins and pulmonary confluence. The repair is accomplished with the apex of the heart tipped up and to the right. After dividing

the vertical vein at the diaphragm, it is opened longitudinally. A parallel incision is made in the back of the left atrium extending into the left atrial appendage. The venoatrial anastomosis begins at the superior end of the venous confluence to the base of the left atriotomy. The anastomosis proceeds up both sides until the inferior portion of the confluence (the divided vertical vein) is anastomosed to the tip of the left atrial appendage.

With both supracardiac and infracardiac TAPVC, Hawkins et al[18] from Utah reported that the use of absorbable suture material results in a lower incidence of late venous obstruction. The incidence of pulmonary venous obstruction with absorbable monofilament suture material was 3.2% (1 of 32 patients) compared with a 17% incidence (4 of 23 patients) with absorbable monofilament suture material ($P < .05$). Unfortunately, the two groups were operated on during different times (the patients receiving absorbable suture material were operated on more recently), so the beneficial effect could have been confounded by an era effect.

With TAPVC draining to the coronary sinus (cardiac total anomalous pulmonary venous connection), the repair is performed by simply enlarging the existing atrial septal defect or patent foramen ovale and completely unroofing the coronary sinus until all four pulmonary veins are adequately visualized. The enlarged atrial septal defect is then closed with a patch of pericardium or other patch material with care taken to avoid the area of the conduction tissue.[6] Although some have advocated primary closure of the atrial septal defect, we have found that this sometimes can distort the anatomy and, therefore, believe the use of the patch is quite beneficial.

The surgical management of mixed TAPVC varies according to the particular anatomy of each patient.[3] A single anomalous pulmonary vein occasionally can be left uncorrected if the other three are satisfactorily repaired, but close observation is required during follow-up to identify a significant left-to-right shunt or the onset of pulmonary vascular obstructive disease.

## Postoperative Considerations

Postoperatively, these children can have pulmonary artery hypertension, particularly those who presented with an obstructed picture. In fact, as noted above, some may have been misdiagnosed as persistent fetal circulation with pulmonary artery hypertension and may already be on ECMO support. After repair, we routinely measure pulmonary artery pressures. If they are elevated, then we will leave a transthoracic pulmonary artery pressure measuring line inserted through a purse string suture in the right ventricular outflow tract and then passed across the pulmonary valve into the pulmonary artery. This is very helpful in the postoperative period in terms of guiding appropriate management.[4] Occasionally, children with persistent pulmonary artery hypertension will require nitric oxide treatment or even postoperative ECMO in addition to the standard manipulations of hyperventilation, adequate oxygenation, and sedation.

Both in the operating room and in the early postoperative period, the left ventricle can be particularly non-compliant with a restricted stroke volume after repair. These children will be very sensitive to volume overload. They seem to have a fixed stroke volume and, therefore, increasing the heart rate can often compensate for this decreased stroke volume until the ventricle becomes more compliant. We also routinely use milrinone both to improve ventricular compliance as well as to decrease pulmonary vascular resistance. In these very critical patients, delayed sternal closure is a useful option.

## Results

The surgical results with repair of TAPVC have improved significantly over the years as a result of earlier diagnosis and treatment, improved preoperative and postoperative care, and refined surgical and anesthetic techniques. Early series reported surgical mortalities in the range of 13% to 25%.[4,6,7,11,19] More recently, there have been series with operative mortalities less than 10%, even in the face of obstructed TAPVC.[10,13,20–23] Although early reports indicated that patients with obstructed TAPVC were at greater risk,[11,19,20,22] more recent publications have not found preoperative pulmonary venous obstruction to be a risk factor for mortality.[4,6,20,21]

A particularly challenging group of patients with total anomalous pulmonary venous connection are those with single ventricle physiology. In these children, because of early pulmonary artery hypertension, regulation of pulmonary blood flow can be quite challenging, particularly if they need either a systemic to pulmonary artery shunt or pulmonary artery banding. What appears to be adequate pulmonary blood flow in the operating room can be excessive pulmonary blood flow later on as the pulmonary vascular resistance drops necessitating readjustment of the source of pulmonary blood flow. Even in the best of hands, these children are problematic. A report from The Children's Hospital of Philadelphia showed a 1-year survival in this group of children with total anomalous pulmonary venous connection and single ventricular physiology as low as 37%.[24] In the report by Gaynor et al,[24] early mortality was 58% if the TAPVC was corrected at the original operation. Furthermore, late pulmonary venous obstruction occurred in more than 50% of these survivors.[24] These sobering results have been confirmed by others.[21,25] In fact, Hashmi et al,[26] from The Hospital for Sick Children in Toronto, reported a 95% operative mortality for 20 children with right atrial isomerism in whom TAPVC repair was performed either alone or in combination with other procedures.

### References

1. Darling RC, Rothney WB, Craig JM: Total pulmonary venous drainage into the right side of the heart: report of 17 autopsied cases not associated with other major cardiovascular anomalies. Lab Invest 6:44-55, 1957
2. van der Velde ME, Parness IA, Colan SD, et al: Two-dimensional echocardiography in the pre- and postoperative management of totally anomalous pulmonary venous connection. J Am Coll Cardiol 18:1746-1751, 1991
3. Imoto Y, Kado H, Asou T, et al: Mixed type of total anomalous pulmonary venous connection. Ann Thorac Surg 66:1394-1397, 1998

4. Serraf A, Bruniaux J, Lacour-Gayet F, et al: Obstructed total anomalous pulmonary venous return. Toward neutralization of a major risk factor. J Thorac Cardiovasc Surg 101:601-606, 1991
5. Kirklin JW: Surgical treatment of total anomalous pulmonary venous connection in infancy. In: Barratt-Boyes BG, Neutze JM, Harris EA (eds): Heart Disease in Infancy: Diagnosis and Surgical Treatment. Edinburgh, Scotland, Churchill Livingstone, 1973, pp 89-96
6. Cobanoglu A, Menashe VD: Total anomalous pulmonary venous connection in neonates and young infants: Repair in the current era [see comment]. Ann Thorac Surg 55:43-48, 1993
7. Hawkins JA, Clark EB, Doty DB: Total anomalous pulmonary venous connection. Ann Thorac Surg 36:548-560, 1983
8. Shumacker HB, King H: A modified procedure for complete repair of total anomalous pulmonary venous drainage. Surg Gynecol Obstet 112:763-765, 1961
9. Corno A, Giamberti A, Carotti A, et al: Total anomalous pulmonary venous connection: surgical repair with a double-patch technique. Ann Thorac Surg 49:492-494, 1990
10. Michielon G, Di Donato RM, Pasquini L, et al: Total anomalous pulmonary venous connection: Long-term appraisal with evolving technical solutions. Eur J Cardiothorac Surg 22:184-191, 2002
11. Hammon JW Jr, Bender HW Jr, Graham TP Jr, et al: Total anomalous pulmonary venous connection in infancy. Ten years' experience including studies of postoperative ventricular function. J Thorac Cardiovasc Surg 80:544-551, 1980
12. Tucker BL, Lindesmith GG, Stiles QR, et al: The superior approach for correction of the supracardiac type of total anomalous pulmonary venous return. Ann Thorac Surg 22:374-377, 1976
13. Lupinetti FM, Kulik TJ, Beekman RH III, et al: Correction of total anomalous pulmonary venous connection in infancy. J Thorac Cardiovasc Surg 106:880-885, 1993
14. Balakrishnan KR, Parvathy U: Modified septosuperior approach for the repair of supracardiac total anomalous pulmonary venous return in infants. Ann Thorac Surg 80:1140-1142, 2005
15. Cope JT, Banks D, McDaniel NL, et al: Is vertical vein ligation necessary in repair of total anomalous pulmonary venous connection? Ann Thorac Surg 64:23-28, 1997
16. Shah MJ, Shah S, Shankargowda S, et al: L→R shunt: A serious consequence of TAPVC repair without ligation of vertical vein. Ann Thorac Surg 70:971-973, 2000
17. Phillips SJ, Kongtahworn C, Zeff RH, et al: Correction of total anomalous pulmonary venous connection below the diaphragm. Ann Thorac Surg 49:734-738, 1990
18. Hawkins JA, Minich LL, Tani LY, et al: Absorbable polydioxanone suture and results in total anomalous pulmonary venous connection. Ann Thorac Surg 60:55-59, 1995
19. Mazzucco A, Rizzoli G, Fracasso A, et al: Experience with operation for total anomalous pulmonary venous connection in infancy. J Thorac Cardiovasc Surg 85:686-690, 1983
20. Bando K, Turrentine MW, Ensing GJ, et al: Surgical management of total anomalous pulmonary venous connection. Thirty-year trends. Circulation 94:II12-II16, 1996 (suppl)
21. Hancock Friesen CL, Zurakowski D, Thiagarajan RR, et al: Total anomalous pulmonary venous connection: An analysis of current management strategies in a single institution. Ann Thorac Surg 79:596-606, 2005
22. Kirshbom PM, Myung RJ, Gaynor JW, et al: Preoperative pulmonary venous obstruction affects long-term outcome for survivors of total anomalous pulmonary venous connection repair. Ann Thorac Surg 74:1616-1620, 2002
23. Raisher BD, Grant JW, Martin TC, et al: Complete repair of total anomalous pulmonary venous connection in infancy. J Thorac Cardiovasc Surg 104:443-448, 1992
24. Gaynor JW, Collins MH, Rychik J, et al: Long-term outcome of infants with single ventricle and total anomalous pulmonary venous connection. J Thorac Cardiovasc Surg 117:506-513, 1999
25. Sadiq M, Stumper O, de Giovanni JV, et al: Management and outcome of infants and children with right atrial isomerism. Heart 75:314-319, 1996
26. Hashmi A, bu-Sulaiman R, McCrindle BW, et al: Management and outcomes of right atrial isomerism: A 26-year experience. J Am Coll Cardiol 31:1120-1126, 1998