19

Total Anomalous Pulmonary Venous Connection

19.1 Morphology & Pathophysiology

Total anomalous pulmonary drainage or connection (TAPVC) is a congenital disorder in which all pulmonary venous blood drains into the right side of the heart through anomalous venous connections.

Supracardiac (Type I):

Pulmonary veins drain into common pulmonary vein which is connected to the innominate vein or rarely to SVC by ascending vertical vein (incidence of 50%) (see Figure 19.1).

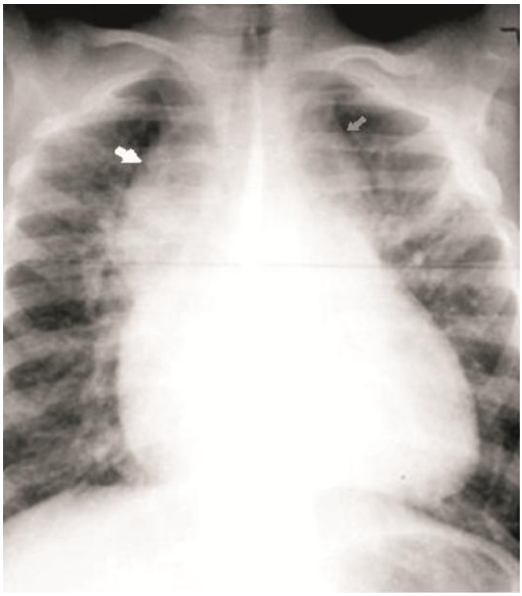

Figure 19.1 *Chest roentgenogram showing supracardiac type of anomalous pulmonary venous connection in a 7 month old infant. The heart has a "snowman shape." The pulmonary veins drain into an enlarged vertical vein (grey arrow), which in turn drains into the brachiocephalic vein and then to the superior vena cava (white arrow). The vasculature is of a shunt vasculature.*

Paracardiac (Type II):

Pulmonary veins drain directly into right atrium or into the coronary sinus either directly or through the common pulmonary vein (incidence of 25%).

Infracardiac (Type III):

Common pulmonary vein is connected to the hepatic veins or portal vein by descending vertical vein (incidence of 15-20%).

Mixed type:

Combination of above types of connection occurs in 3-5%.

Oxygenated pulmonary venous blood mixes with systemic venous blood in the right atrium, and an atrial septal defect (or the patent foramen ovale) is essential to maintain systemic arterial output. The patent ductus arteriosus is present in most patients. The mixing of systemic venous and pulmonary venous blood results in cyanosis of a varying degree, determined by the quantity of pulmonary blood flow and size of the atrial septal defect. Obligatory mixing of pulmonary venous blood and systemic venous blood in the right atrium also results in a similar oxygen saturation in all cardiac chambers, pulmonary artery, and systemic (femoral) artery.

Constriction of the anomalous pulmonary venous connections to systemic veins results in pulmonary venous hypertension, elevated pulmonary vascular resistance with predictable decrease in pulmonary blood flow, and increase in cyanosis. The balloon atrial septostomy (done for a restrictive atrial septal defect) will not alter oxygenation of the blood in the presence of pulmonary venous hypertension, but it would provide additional systemic cardiac output.

In the absence of obstruction of anomalous pulmonary venous connections to systemic veins and in the presence of a large atrial septal defect, pulmonary blood flow is increased with development of congestive failure, pulmonary

artery hypertension like in a large atrial septal defect, but with early rise in pulmonary vascular resistance.

19.1.1 Obstructed TAPVC

It presents immediately after birth with severe cyanosis and poor systemic perfusion, with varying degrees of increase in pulmonary arterial and venous resistances. The newborn or infant frequently has pulmonary edema. The patient's condition should be stabilized immediately by cardiovascular and metabolic standpoint, and provide the best possible preoperative status prior to a surgical repair.

Management:

1. Sedation, mechanical ventilation, or hyperventilation with 100% oxygen.
2. Keep the arterial PCO_2 < 30 mm Hg to improve the effective pulmonary blood flow.
3. Prostaglandin E_1 (alprostadil) infusion:

   It may allow some right-to-left shunting at the level of the ductus arteriosus. This may increase the systemic cardiac output although it may do so, at the expense of pulmonary blood flow. It may also allow patency of the ductus venous in TAPVC type III and may improve arterial oxygenation.
4. Aggressive correction of metabolic acidosis with sodium bicarbonate.
5. Transfusion: Packed cells or a whole blood is administered to improve the oxygen-delivery capacity.
6. Inotropes: To improve the cardiac contractility.
7. Other resuscitative measures:

   a) Balloon atrial septostomy.

Used to decompress the venous circuit and improve cardiac output in cases of a restrictive inter-atrial communication.

b) Stenting of obstructed vein.

Recently, used as a valid aid to provide an immediate relief of pulmonary hypertension and cardiogenic shock, allowing more effective hemodynamic stabilization prior to a surgical intervention.

c) Extracorporeal membrane oxygenation (ECMO).

Some centers have advocated its use during resuscitation, and in some cases it has been adopted after the repair, to support the neonates with a residual pulmonary hypertension.

19.1.2 Unobstructed TAPVC

Medical therapy is directed at managing right ventricular failure, hypoxia, and congestive heart failure.

1. Hypoxia: Assisted ventilation or a normal ventilation with low levels of FIO_2.
2. Inotropic support: Improves ventricular function and contractility.
3. Diuretics.
4. Phenoxybenzamine: α-blockade rarely is used to reduce the pulmonary hypertension.

19.2 Operative Procedures

The principle of surgery is to redirect the pulmonary vein flow entirely to the left atrium, by creation of a large and unobstructed anastomosis between the pulmonary venous confluence and the left atrium (LA) using fine sutures

(running versus interrupted stitches, use of absorbable versus non-absorbable suture material is controversial).

Usually, an incision is created in the pulmonary venous confluence to match a corresponding incision in the posterior wall of LA and extending into the LA appendage.

The operation is performed on CPB with moderate hypothermia and cardioplegic arrest. Deep hypothermia and a short period of low-flow cardiopulmonary bypass or circulatory arrest may be desirable, for a meticulous anastomosis.

The pulmonary venous confluence is exposed in the posterior pericardium by retracting the heart rightward and anteriorly or by a superior access between the ascending aorta and the SVC. Alternatively, it can be accessed in the right pericardial fossa by a vertical incision through the right atrium, the inter-atrial septum, and the left atrium (LA).

19.2.1 Supracardiac TAPVC

The common pulmonary vein is opened wide and connected side to side to the left atrium. The foramen ovale is closed. Though routine ligation of the vertical vein is controversial at the time of TAPVC repair, it is ligated next to its connection to the systemic vein.

19.2.2 Infracardiac TAPVC

Technique is similar to that of the supracardiac. As the pulmonary venous confluence tends to be oriented vertically, typically in a Y - shape, the incision into the LA is relatively vertical or Y - shaped. The vertical vein may be left open to provide an access to a low pressure system, in cases of a particularly small and restrictive LA. The high resistance of the liver parenchyma eventually

leads to complete cessation of flow through this low-pressure venous channel. Some ligate the vein immediately above the diaphragm, and use the intra-pericardial portion of this vein to produce a larger anastomosis.

19.2.3 Cardiac TAPVC to Coronary Sinus

Unroof the coronary sinus through an incision between the coronary sinus and the fossa ovalis, thus creating a large ASD (atrial septal defect). Then, a patch is used to reconstruct the atrial septum, leaving the pulmonary venous drainage to flow through the unroofed coronary sinus into the LA.

19.2.4 Cardiac TAPVC to Right Atrium

If the anomalous veins drain directly into the right atrium, the atrial septum is detached posteriorly, and is reattached to the right of the right sided pulmonary vein ostia.

19.3 Postoperative Management

The postoperative course is variable and depends on the preexisting pulmonary venous obstruction. The hospital stay averages 8 to 12 days.

19.3.1 Hemodynamic Management

Pulmonary artery hypertensive crisis is a major cause of mortality in the postoperative period after repair of the TAPVC in early infancy.

19.3.2 Pulmonary Edema

During the postoperative period, patients with obstructed TAPVC, even after surgical relief of the pulmonary venous obstruction, frequently have pulmonary edema with varying degrees of an increase in pulmonary arterial and venous resistance.

Manage pulmonary edema by diuretics, assisted ventilation with a high fraction of FIO_2, and PEEP (positive end-expiratory pressure).

19.3.3 Episodic Pulmonary Hypertension

Episodic pulmonary hypertension occurs due to reactive pulmonary vascular bed in the postoperative period, resulting in a low cardiac output.

Manage with pulmonary vasodilators such as nitric oxide, magnesium sulfate, or prostaglandin.

19.3.4 Hypercyanotic Episodes

Severe cyanosis or severe hypercyanotic episodes may also occur postoperatively in the obstructed TAPVC.

Manage these episodes with one or more pulmonary vasodilators as above to decrease pulmonary vascular resistance and pulmonary vascular reactivity.

19.3.5 Poor Lung Compliance

Obstructed TAPVC patients often have a difficult postoperative course secondary to high pulmonary vascular resistance with a poor lung compliance. ECMO is used routinely in some centers after the surgical correction for these patients.

19.3.6 Hemodynamic Instability

Hemodynamic instability can occur with elevations in pulmonary vascular resistance, especially in patients with uncorrected pre-existing pulmonary venous obstruction. ECMO may be lifesaving in these patients.

Poor hemodynamic performance following a repair of TAPVC should raise concern of a potential obstruction at the site of the repair.

To rule out an obstruction at the site of the repair perform:

a) Chest radiography: It often reveals pulmonary edema.

   (Note this finding is nonspecific because it is seen in many patients with preoperative pulmonary venous obstruction despite normal hemodynamics and adequate correction of the obstruction).

b) Echocardiogram: Rules out any residual postoperative pulmonary venous obstruction, and examines the pulmonary venous anastomosis. If it is possible, cardiac MRI is indicated to refine the diagnosis.

19.3.7 Ancillary Measures

(I) Ultrafiltration

Both the conventional and modified ultrafiltration may be used during surgery of infants and newborns with an obstructed TAPVC.

(II) Peritoneal Dialysis

It is occasionally used postoperatively to remove the excess of extracellular fluid.

(III) Left to Right Shunting

If the vertical vein is not ligated at the time a repair of TAPVC, persistent patency of the vertical vein (ascending or descending) may lead to a significant left to right shunting, requiring a reoperation or a catheter intervention for closure.

Systemic arterial oxygen saturation (SaO_2) should be normal after an uneventful operation.

19.3.8 Pulmonary Vasodilators in TAPVC

(I) Prostaglandin E_1 (PGE_1)

It dilates the vascular smooth muscle in the ductus arteriosus, ductus venosus, pulmonary arteries and viens, and systemic arteries.

In obstructed TAPVC, PGE_1 is usually used as a pulmonary vascular dilator, but one should exercise caution to observe its effects in the complex circulation of TAPVC.

In sub-diaphragmatic TAPVC, PGE_1 dilates the ductus venosus and improves pulmonary venous flow.

In other types of TAPVC with obstruction, PGE_1 dilates pulmonary arteries and increase pulmonary flow, dilates ductus arteriosus and systemic arteries, and increases right-to-left shunting and worsens cyanosis.

(II) Magnesium Sulfate

$MgSO_4$ has both systemic and pulmonary vascular dilating effects.

Mechanism of action: Direct action on the vascular muscle cells, but may also increase the formation or release of nitric oxide.

Lower-dose $MgSO_4$ with a slow infusion is advised to avoid systemic hypotension.

(III) Nitric Oxide

Mechanism of action:

Stimulates guanylate cyclase to form cyclic GMP that causes relaxation of the vascular smooth muscle.

Delivery:

It is delivered by inhalation directly to the alveolar units, and is rapidly inactivated by the hemoglobin. It is the most selective of currently available pulmonary vascular dilators.

(IV) Sildenafil

Nasogastric (oral) administration of sildenafil may be used as an alternative to nitric oxide, or sildenafil may be used during the weaning from nitric oxide.

19.3.9 Management of Pulmonary Artery Hypertensive Episodes

It is usually required to minimize pulmonary artery hypertensive episodes by institution of the following measures:

1. Sedation with infusions of a fentanyl.
2. Paralysis with a neuromuscular blocking agent.
3. Oxygenation and hyperventilation.
4. Inhaled nitric oxide, magnesium sulphate, PGE_1, and sildenafil.

The above drugs are used for reduction of both pulmonary vascular resistance and PA pressure.

19.3.10 Invasive Pressure Monitors

Arterial, central venous, and PA (pulmonary artery) line.

An oximetric catheter placed in the PA assesses the cardiac output and pulmonary artery pressure.

19.3.11 Vasoactive Drug Infusions

Dopamine or dobutamine, milrinone, epinephrine, isoproterenol, nitroprusside and/or phenoxybenzamine, magnesium sulphate, and inhaled nitric oxide (see Section I Chapters 4 & 16).

19.3.12 Postoperative Bleeding

It is occasionally encountered from the anastomotic suture lines.

19.3.13 AV Conduction Abnormalities

AV (atrioventricular) conduction defects are potential, especially in coronary sinus type TAPVC as the AV node in the AV septum may be injured during an atrial septal incision between the coronary sinus and fossa ovalis.

Temporary atrioventricular pacing should be readily available.

19.4 Surgical Reinterventions During the Postoperative Period

Though the pulmonary vasodilators reduce pulmonary artery hypertensive crises, the long term outcome depends on the preexisting pulmonary venous stenosis and/or a recurrent venous obstruction.

The preexisting pulmonary venous obstruction was more common in the coronary sinus type of TAPVC, and early postoperative restenosis may occur. It occurs in 5-10% of patients, and is usually evident in the first 6 months following surgery.

The recurrent venous obstruction should be managed, only by an early aggressive approach of reoperation and repeated balloon dilatation. The obstruction is more easily treated surgically if it involves only the pulmonary venous confluence and anastomosis area.

If the pulmonary venous obstruction is due to intimal fibrotic hyperplasia of individual pulmonary branch veins extending deeper back into the veins, then surgery may not be successful.

In mixed form of TAPVC, with more complex patterns of pulmonary venous connection, individual pulmonary veins are small or thickened, and the mortality remains higher after repair.

In a smaller number of patients, a diffuse stenotic process involves the whole length of the pulmonary veins including their intrapulmonary course. This process may progress to an almost complete vein occlusion. If this lesion is bilateral, either sutureless techniques and/or lung transplantation would only improve the outcome.

If the vertical vein is not ligated at the time of repair of TAPVC, persistent patency of the vertical vein (ascending or descending) may lead to a significant left to right shunting. It requires a reoperation, or a catheter intervention for closure.