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Perioperative Management of Total Anomalous Pulmonary Venous Drainage

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Abstract

Total anomalous pulmonary venous drainage (TAPVD) is a congenital heart disease involving abnormal drainage of all the four pulmonary veins in to systemic venous drainage or right atrium. TAPVD can occur in isolation or as an additional component of complex congenital heart disease with univentricular or biventricular physiology. If corrected at the right time it allows the baby to have essentially normal growth and development with good quality of life but delay in diagnosis and referral can result in grave consequences like severe pulmonary hypertension, cardiogenic shock and mortality. It can present in the neonatal period or infancy depending upon obstruction to the abnormal pulmonary venous drainage. Obstructed TAPVD when presenting in the neonatal period constitutes a significant pre operative challenge and the outcome is often determined by the positive synergy in the transport and referral hospital team. The current review is targeted to stream line perioperative management strategy based on the currently available evidence in literature.

Key words: Total anomalous pulmonary venous drainage (TAPVD): Supracardiac; Infracardiac; Cardiac; Pulmonary hypertension.

Introduction

Total anomalous pulmonary venous drainage (TAPVD) consists of an abnormality of blood flow in which all 4 pulmonary veins drain into systemic veins or the right atrium with or without pulmonary venous obstruction[1].

The presence of an interatrial communication is necessary to sustain life, and therefore an atrial septal defect (ASD) or patent foramen ovale (PFO) is considered part of the complex. Also, the young age of the patients makes the presence of a Patent Ductus Arteriosus (PDA) usual, and this is not considered a complicating defect. Rarely Ventricular septal defects (VSD) are associated with the anomaly[2].

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Types of total anomalous pulmonary venous drainage

Darling proposed the most commonly used classification system for total anomalous pulmonary venous connection based on the site of pulmonary venous drainage[3]. In type I (i.e. supracardiac connection), the 4 pulmonary veins drain via a common vein into the right superior vena cava, left superior vena cava, or their tributaries.

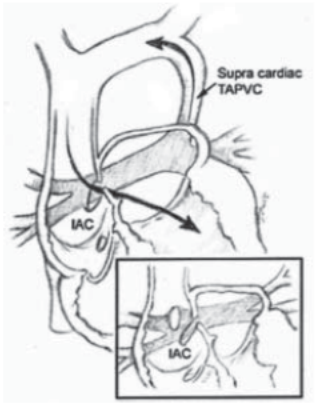

Figure 1: Supra Cardiac TAPVC

In type II (i.e. cardiac connection), the pulmonary veins connect directly to the right heart (e.g. coronary sinus or directly to the right atrium). In type III (i.e. infra diaphragmatic connection), the common pulmonary vein travels down anterior to the esophagus through the diaphragm to connect to the portal venous system. In type IV (i.e. mixed connections), the right and left pulmonary veins drain to different sites (e.g. left pulmonary veins into the left vertical vein to the left innominate, right pulmonary veins directly into the right atrium or coronary sinus.[4]

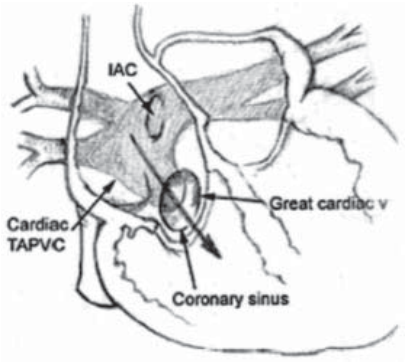

Figure 2 A: Coronary Sinus TAPVC

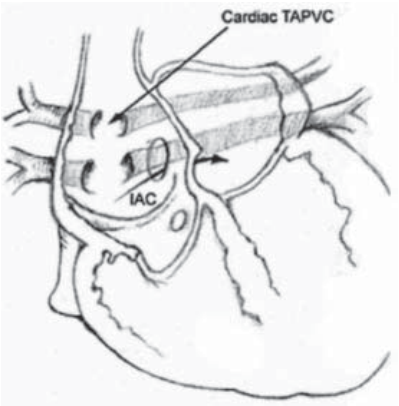

Figure 2 B: Cardiac TAPVC

Smith et al.[5] provided an alternative classification for TAPVD: Supra cardiac (without pulmonary venous obstruction) and infra diaphragmatic (with pulmonary venous obstruction). All the pulmonary veins drain into the systemic venous circulation and hence right atrial and right ventricular enlargement occur along with dilation of the pulmonary artery. In case of significant pulmonary venous obstruction, right ventricular hypertrophy occurs.

The left ventricle is of normal size, and left ventricular volume measured in life is usually within the normal limits. Left atrial size usually is diminished because it lacks the contribution of the common pulmonary vein. Total anomalous pulmonary venous connection occurs in isolation in two thirds of patients or manifests as part of a group of heart defects (e.g. heterotaxy syndromes) in approximately one third of patients.[1]

In infra diaphragmatic connection, severe obstruction almost inhibits pulmonary venous flow with obstruction of the common pulmonary vein. This obstruction manifests either as it courses through the diaphragm, at its junction with the portal vein system, or as an obstruction of pulmonary venous flow as the ductus venosus closes and pulmonary vein flow is forced to cross the liver portal sinusoid system. Finally, in all types, obstruction may occur because of restrictive atrial septal defect size and because of small left atrial size.

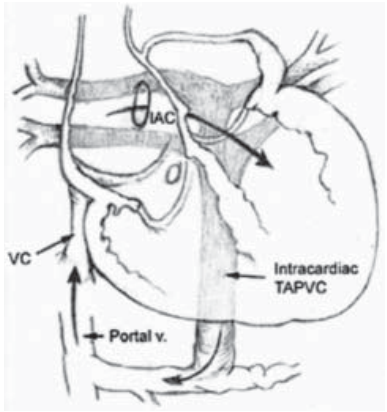

Figure 3: Infra Cardiac TAPVC

Presentation

Patients with pulmonary vein obstruction

Pulmonary venous obstruction occurs in virtually all patients with subdiaphragmatic drainage and in approximately 50% of patients with supracardiac drainage[6]. Patients with obstruction develop symptoms early, usually in the first 24-36 hours of life, including tachypnea, tachycardia, and cyanosis with or without cardiogenic shock. Signs of pulmonary hypertension progress with decreasing pulmonary blood flow and worsening cyanosis. Natural history is that of progressive clinical deterioration and early death in the first week or month of life, depending on the degree of pulmonary venous obstruction.[6]

Physical examination findings include severe cyanosis with significant respiratory distress. Cardiac impulse is prominent anteriorly, but, usually, the heart is not clinically enlarged. The pulmonary component of the second heart sound is accentuated, and occasionally a gallop rhythm may be present. A murmur usually is not detectable, yet a systolic murmur over the pulmonary area or a tricuspid insufficiency murmur at the mid and lower left sternal border may be present. Peripheral pulses are usually normal after birth but may deteriorate rapidly as heart failure progresses. Liver enlargement commonly occurs, especially in total anomalous pulmonary venous connection (TAPVD) type III with sub diaphragmatic drainage.[7]

Patients without pulmonary venous obstruction

Patients with unobstructed pulmonary venous flow present with symptoms more similar to a very large atrial septal defect. Mild failure to thrive with increased respiratory effort than normal with activity or recurrent respiratory infections may be present.

Physical examination findings include right ventricular volume loading with increase in right ventricular impulse, a wide split-second sound (usually with normal-intensity pulmonary closure), and pulmonary outflow murmur with or without a tricuspid diastolic murmur. Cyanosis infrequently occurs in the first year of life.

Reverse difference cyanosis has been reported in the newborn period in total anomalous pulmonary venous connection to the superior vena cava (SVC). In this setting highly saturated blood in the SVC streams preferentially from right ventricle across ductus arteriosus to descending aorta; lower saturated blood in inferior vena cava streams across the foramen ovale to the left heart and aorta, resulting in higher saturation in the foot than in the right hand.[8]

If a restriction develops in the foramen ovale, some degree of pulmonary hypertension is more likely, with earlier onset of tachypnea, louder pulmonary closure sound, more prominent right ventricular impulse, and a greater likelihood of systemic and pulmonary venous congestion.

ECG

A tall peaked P wave in lead II or the right precordial leads characteristic of right atrial enlargement is a constant finding. Right-axis deviation is usual. Right ventricular hypertrophy is invariably present, usually manifested by high voltage in the right precordial leads, occasionally as an incomplete right-bundle-branch block pattern.[1,9]

Chest radiography

The lung fields reflect increased pulmonary blood flow. The right atrium and right ventricle are enlarged, and the pulmonary artery segment is prominent. The left-sided chambers are not enlarged.

A figure-of-8 or a snowman appearance of the cardiac show is seen in patients with TAPVD to the left innominate vein. The upper portion of the figure-of-8 is composed of the anomalous vertical vein on the left, the left innominate vein superiorly, and the SVC on the right. This diagnostic sign is not usually present in the first few months of life but is often present in the older child and adult. When the anomalous connection is to the right SVC, dilation of this structure results in a prominence at the upper right cardiac border.[1]

Echocardiography

- Apical and subcostal 4-chamber views usually best identify individual pulmonary veins and their confluence in patients with total anomalous pulmonary venous connection.
- The common pulmonary vein can be visualized to its point of entry to the systemic venous system or to the coronary sinus using multiple views.
- Right ventricular and pulmonary artery volume loading with flattened or paradoxic septal motion may be observed on M-mode imaging.
- Subcostal long - and short-axis views help in evaluating the size and flow patterns across the foramen ovale or atrial septal defect.[1]

Computed Tomography with contrast (CT scan) and Magnetic Resonance Imaging (MRI):

CT scan and MRI provide a wide-field imaging of the pulmonary veins and blood flow within them as well as adjacent cardiovascular structures. Non cardiac structures, such as airways, lungs, spine, and abdominal organs, also are visualized. MRI performed across various planes demonstrates

dynamic nature of blood flow, cardiac chambers, and AV and semilunar valves. Imaging techniques such as the CT MRI are particularly useful in complex conditions such as the heterotaxy syndromes, hypoplastic aortic arch and aortic arch interruption, when full delineation of anatomy has not been possible by echo[10].Usually, these techniques are not necessary in the initial evaluation of newborns and in the infant with TAPVD as echocardiography is accurate, portable, and readily available.[11]

Treatment

Corrective surgery is necessary for all patients with this condition. No palliative procedure exists. All infants with pulmonary venous obstruction should be operated on soon after diagnosis, in the newborn period. Infants who do not have pulmonary venous obstruction but do have heart failure with recurrent lower respiratory tract infection and failure to thrive are usually operated on between 4 and 6 months of age. Optimal stabilization the patient prior to surgery from a cardiovascular and metabolic standpoint is important.[9]

Pre operative Management

1. Intensive anti congestive measures with digitalis and diuretics should be provided for infants without pulmonary venous obstruction
2. Metabolic acidosis should be corrected if present after establishing mechanical/positive pressure ventilation
3. Infants with severe pulmonary edema (resulting from the infra cardiac type and from other types with obstruction) should be intubated and supported with positive pressure ventilation.
4. If the size of the interatrial communication appears small and immediate surgery is not indicated, balloon atrial septostomy or blade atrial septostomy may be performed to enlarge the communication[9]

Surgery

1. The goal of surgery is to redirect pulmonary vein flow entirely to the left atrium. In patients with a supracardiac or infracardiac connection, the common pulmonary vein is opened wide and connected side to side to the left atrium. The foramen ovale is closed, and the ascending or descending vein is usually ligated. In a cardiac connection (to right atrium or coronary sinus), the atrial septum is resected partially and a new septum is surgically created, directing pulmonary veins to the left atrium. A coronary sinus may be separately tunneled to the right atrium or left to drain with the pulmonary veins to the left atrium. In selected cases with severe pulmonary hypertension or right ventricular dysfunction, the vertical vein may be left open.[9]

Complications

1. In the early post operative period, pulmonary hypertension due to a small and poorly compliant left heart leading to cardiac failure and pulmonary edema is common and may require prolonged respiratory and medication support postoperatively.
2. In patients with obstructed total anomalous pulmonary venous connection, the pulmonary arteries may be hypoplastic and respond poorly to the vasodilators.
3. Postoperative arrhythmias are usually atrial. [9]
4. The complication of pulmonary venous obstruction occurs later in 5-10% of patients and is usually evident in the first 6 months following surgery. This obstruction is more easily treated surgically if it involves only the pulmonary venous confluence and anastomosis area. Sutureless marsupialization with pericardium has seemed to help improve surgical results.[12]
5. If the pulmonary venous obstruction involves intimal fibrotic hyperplasia of individual pulmonary veins extending deeper back into the veins then surgery may not be successful.
6. Surgical mortality remains higher in repair of mixed form of total anomalous pulmonary venous connection, especially in patients with more complex patterns of pulmonary venous connection, which have persistently small or thickened individual pulmonary veins.[13]

Post operative care

Following surgery, the child is transferred to the PICU team with detailed information on various parameters listed below (Table 1)

Table 1: Information to be given to the Critical Care team during transfer of child into PICU

Surgery	• Type of lesion • Procedure-correction/palliation • CPB time, Aortic cross clamp time
History	• Previous medical illness • Medications • Allergies • Previous surgeries / medical history
Anesthesia	• Intra operatrive problems - surgical, anesthetic, CPB • Respiratory parameters • Airway (difficulties, ET tube size, fixed, leak) • Ventilator settings and parameters
Haemodynamics	• Cardiac rate and rhythm • Filling pressures (central venous pressure-CVP) • Vaso active agents • Most recent vital signs • Most recent laboratory data - Hct, K^+, ABG, temperature • Mixed venous oxygen saturation (MVO_2)

Initial assessment in the PICU

A rapid initial assessment of the child is made soon after transfer into the PICU. This includes a detailed physical examination (Table 2) and laboratory studies (Table 3).

Table 2: Physical Examination

Respiratory	• Breath sounds • ET tube size, fixed, leak • Chest excursion
Cardiovascular	• Heart rate and rhythm • Blood pressure - waveform • Filling pressures - LA/RA • Pulse volume • Heart sounds and cardiac murmurs • Temporary Pacemaker/settings
Skin	• Cyanosis turgor
Central nervous system	• Awake status, pupils
Per Abdomen	• Addominal Distention, liver size, spleen • Ascites

Table 3: Laboratory studies

Complete blood count	• Hematocrit • platelets
Electrolytes	• Sodium • Potassium • Calcium • Magnesium
Arterial Blood gas	• Acidosis (Metabolic/Respiratory) • pH, PCO_2, PO_2, anion gap, MVO_2, Lactate
Coagulation	• PT, APTT, ACT

PT: Proth Rombin Time, APTT: Activated Partial Thromboplastin Time, ACT: Activated Clotting Time

A chest radiography (AP view) is performed to determine the following

1. Position of the tip of the endo tracheal tube
2. Location of the central vascular catheters and other monitoring lines
3. Position of the chest tubes
4. Heart size
5. Lung vascularity, presence of atelectasis, pneumothorax or pleural effusion
6. Pulmonary vascular markings

Principles of post operative management

Continuous monitoring and surveillance of the clinical data are mandatory to direct further treatment following cardiac surgery. Following the repair of TAPVD, medical therapy is directed towards augmenting cardiac output and minimizing pulmonary vascular resistance (PVR) using β adrenergic agonists and pulmonary vasodilators such as dobutamine, isoproterenol, and milrinone.

Hemodynamic management

Cardiac output depends on the heart rate and stroke volume. Stroke volume in turn depends on adequate preload, minimizing afterload, contractility, adequate heart rate and optimal cardiac rhythm. Warm peripheral extremities, brisk capillary refill and a urine output of atleast 0.5 mL/kg/hr are good indicators of adequate cardiac output.

If the heart rate is slow enough to compromise the cardiac output, temporary atrial pacing at higher rate. A few centres still consider isoproterenol in such scenarios. Sinus tachycardia may be due to pain, awakening or hypovolemia and will respond to specific

treatment. Junctional tachycardia and junctional ectopic tachycardia are commonly observed in the immediate post operative period. Atrial overdrive pacing usually helps in inducing systematic conduction. Amiodarone, β blocade and IV Magnesium sulphate may help in controlling the heart rate by reducing the excessive conduction rate or by inducing arterio venous block, thereby slowing the ventricular rate. In case of hemodynamic instability cardioversion has to be attempted. Conservative measures such as aggressive sedation, avoiding higher doses of sympathomimetics, digitalization and avoiding pyrexia along with inducing hypothermia may help in controlling the heart rate. Atrio ventricular sequential pacing may be necessary when the conduction is disrupted in conditions like A-V dissociation.

Myocardial contractility is impaired after open heart surgery due to ischemia, hypoxia and Cardio pulmonary bypass (CPB) induced myocardial dysfunction. Usage of following drugs may improve the contractility and thus cardiac output.[14]

Inotropes

Dopamine – 5 – 10 µg/kg/min

Dobutamine – 5 – 10 µg/kg/min

Epinephrine – helps in improving the blood pressure with its α and β adrenergic effects, particularly when the diastolic BP remains low. (Dose 0.03 - 0.1 µg/kg/min)

Inodilators

Isoproterenol – 0.03 – 0.07 µg/kg/min

Milrinone – a phosphodiesterase III inhibitor provides inotropy along with peripheral vasodilatation (Dose - 0.3 – 0.7 µg/kg/min). As a result, ventricular filling pressures decrease while stroke volume and CO increase.[15]

Vasodilators

Sodium Nitroprusside (SNP) and Nitroglycerin (NTG) - may help in afterload reduction (0.1 - 2 µg/kg/min).[16]

Vasopressin - produces a non-catecholamine-mediated elevation in systemic vascular resistance and mean arterial pressure without depressing cardiac output and with little effect on pulmonary vascular tone. Vasopressin is beneficial in patients with catecholamine refractory vasodilatory shock as well.[17,18] This scenario is particularly witnessed if the child is operated in an emergency with latent or evolving sepsis.

Vasodilators

Inhaled Nitric Oxide (NO)

Nitric oxide stimulates Guanylate Cyclase to form cyclic GMP, which causes relaxation of vascular smooth muscle. It can be delivered by inhalation directly to alveolar units and is rapidly inactivated by hemoglobin making it the most selective of currently available selective pulmonary vascular dilators (except for oxygen). NO has a half life ranging from 0.1 - 5 seconds in physiologic systems. In case of severe pulmonary hypertension as in obstructed TAPVD, NO is very useful and leads to clinical improvement in majority of neonates and infants.[19,20]

Sildenafil

A phosphodiesterase V inhibitor increases the concentration of cGMP leading to smooth muscle relaxation and dilatation of pulmonary arteries. A dose of 0.5mg/kg/Q6H was found to be efficacious in lowering the pulmonary vascular resistance in children with pulmonary hypertension.The drug has minimal systemic effects. Sildenafil should be avoided in acute pulmonary hemorrhage or in acute respiratory distress syndrome (ARDS) with gross V- Q mismatch.

Bosentan

An oral endothelin receptor antagonist that binds with both ETA and ETB receptors. Though limited studies are available, it was found to improve hemodynamics and relieve symptoms of pulmonary hypertension.[1]

Phenoxy benzamine

A non specific alpha receptor antagonist, causes vasodilatation. Intravenous phenoxy benzamine has proven to reduce the afterload on the right ventricle in the post operative period. A dose of 0.5 - 2 mg/kg/day is used. In the current era, it is sparingly used though available as a part of standard formulary in most countries.

Respiratory Management

The oxygen consumption of the children may be increased following surgery secondary to hyperthermia, increased respiratory and circulatory system activity, muscle system activity and endogenenous secretions of catecholamines. Hence it is important to improve the oxygen delivery while decreasing the oxygen consumption. This may be achieved by using sedatives, analgesics and muscle relaxants.

Various respiratory complications such as stridor, mucous plugging, diaphragmatic paralysis, and pulmonary edema can occur as a result of mechanical ventilation and endo tracheal tube. Adequate care can reduce the incidence and risks of these complications. When uncuffed ET tubes are used, a leak of 15 -20 cm H2O is recommended. When cuffed ET tube is used, the cuff is inflated until the leak just disappears. Warming and humidification of inspired gases will avoid heat loss and thickening of secretions. The temperature of these gases should be kept below 37° C. The children who have undergone TAPVD rerouting are usually hyperventilated in view of pulmonary hypertension where in the pH is maintained on the alkalotic side (>7.45) with a low pCO2 (25-30 mmHg). Reasonable physiological positive end expiratory pressure (PEEP) improves oxygenation and decreases atelectasis.

Fluids and electrolytes

The optimal filling pressure varies from time to time depending on the myocardial function. Post CPB, patients are total body fluid overloaded with capillary leak and depleted intravascularly. Although acute volume expansions are often required, patients should be administered only one half to two thirds of the normal maintenance. Factors such as infant bed warmer, rewarming, post operative fever will increase insensible losses by 10 -20% and need to be compensated. Thus, a fluid requirement of about 1 -1.5 mL/kg/hr with an additional compensation for insensible water loss needs to be administered to maintain adequate filling pressures.

Hyperkalemia usually occurs as a consequence of decreased cardiac output, poor perfusion, blood transfusion and altered renal function. Care is taken to maintain the serum potassium levels in the range of 3.5 -4.0 mmol/dL. Hyperkalemia is treated with Glucose Insulin (0.1IU/kg), 10% Calcium chloride solution (0.2 – 0.5 mL/kg), sodium bicarbonate (1-2 mEq/kg). Potassium is supplemented if the serum potassium is less than 3.0 mmol/dL.

Hypomagnesemia is commonly reported post cardiac surgery and may lead to ventricular arrhythmias.

Hematologic management

Children have high incidence of intra operative and post operative bleeding as a result of platelet dysfunction and abnormalities of platelets release. Coagulation cascade factors are inhibited by heparin administered during surgery. Though the heparin effects are reversed by Protamine, "Heparin rebound" occurs several minutes later characterized by bleeding and prolonged activated clotting time. Administration of platelets and fresh frozen plasma or cryoprecipitate may help in achieving hemostasis.

Neurologic complications

Non specific neurologic complications such as involuntary movements, choreoethytosis may result following cardiopulmonary bypass and total circulatory arrest (TCA). Hence close observation and time to time assessment for gross abnormalities, abnormal movements and seizures are necessary.

Gastro intestinal complications

Initial evaluation includes the presence or absence of bowel sounds and measurement of abdominal girth. The liver and spleen should be percussed for position and span to assess the degree of right ventricular failure. Necrotizing Enterocolitis (NEC) may be seen in infants within 24 hours of surgery characterized by abdominal distention, absence of bowel sounds, and radiographic evidence of bowel distention.

Nutrition

Feeding may be started after extubation. In case the child requires mechanical ventilation for more than 48 hours and hemodynamically stable, then minimal enteral feeds may be started on the first post operative day after gastro intestinal complications are ruled out. In children with low output state and unstable hemodynamics, the feeding may be delayed till stability is achieved.

Infection

Low grade fever post CPB is common, but persistent high grade fever or other features of sepsis like leucocytosis or leucopenia, thrombocytopenia, persistent hypoglycemia, hypothermia and warm shock should be worked up with appropriate cultures (blood, urine, ET secretion/sputum).[14]

Guidelines to post op management

Unobstructed TAPVD

1. Routine examination on receival from OR
2. CXR, ACT and ABG
3. Crystalloids 1.5 - 2ml//kg/hr (maximum). Volume may be restricted to less than 2ml/kg/hr
4. Minimal transfusion of blood products; avoid unless needed
5. Elective ventilation (TV - 10 ml/kg, PEEP - 4-6 cm H_2O; To maintain a pH >7.4, pCO_2 <40; a lower FiO_2 < 0.4 in new born and neonates)
6. Inotropes and Inodilators support for myocardial dysfunction
7. ABP to be maintained at minimum optimal level for initial few hours
8. Sedation maintenance IV with Fentanyl / Dexmetedomedine
9. Deep sedation, muscle relaxants may be used in specific cases
10. IV Vasodilators (SNP/NTG/Phenoxy benzamine to control systemic / pulmonary pressures)
11. Keep Serum Potassium less than 4 mmol / L
12. Initiate Peritoneal dialysis if hyperkalemia / low urine output / ascites
13. Routine echocardiography for biventricular function, TR (tricuspid regurgitation), PR (pulmonary regurgitation) and TAPVD confluence.
14. Lasix infusion at the earliest (once peripheries warm, no acidosis, consistently stable hemodynamics)
15. Temporary pacing as required. (A-V sequential pacing in case of A-V dissociation, atrial pacing for sinus bradycardia and nodal rhythm.
16. Heart rate control (Digitalis/hypothermia for sinus tachycardia/nodal tachycardia)
17. IV Metaprolol or Amiodarone for junctional ectopic tachycardia and A-V dissociation.
18. Watch for drainage, urine output and acidosis along with vital parameters
19. Watch for Endo tracheal bleeding during ET suctioning
20. NG feeds from POD 1 (if ventilation planned for more than 24 hours).
21. Vasodilators – Bosentan /Sildenafil with a gradual increment in the dosage based on PA pressures in children older than 3 months of age
22. Fluid balance – Target output atleast 15 – 25% more than intake

Obstructed TAPVD

Usually neonates and infants with obstructed TAPVD have severe pulmonary hypertension and are likely to have high risk of post operative complications such as pulmonary hypertensive crisis and low cardiac output state. It is a common practice to have a delayed sternal closure in such children

1. Routine examination on receival from OR
2. CXR, ACT and ABG
3. Crystalloids 1 – 1.5ml//kg/hr (maximum). Volume may be restricted to less than 2ml/kg/hr
4. Minimal transfusion of blood products; avoid unless needed
5. Elective ventilation for 24 hours (TV – 10 ml/kg, PEEP – 4-6 cm H_2O; To maintain a pH >7.45, pCO_2 <40, a FiO_2 of 0.5 – 0.7 except in case of new borns)
6. Inotopes and Inodilators support for myocardial dysfunction
7. ABP to be maintained at minimum optimal level for initial few hours
8. Sedation maintenance IV with Fentanyl / Dexmetedomedine and intermittent boluses of sedation along with muscle relaxants may be used.
9. IV Vasodilators (SNP/NTG/Phenoxy benzamine to control systemic / pulmonary pressures)
10. Keep Serum Potassium less than 4 mmol / L
11. Initiate Peritoneal dialysis if hyperkalemia / low urine output / ascites
12. Routine echocardiography for biventricular function, TR, PR and TAPVD confluence, rule out pulmonary venous obstruction.
13. Lasix infusion at the earliest (once peripheries warm, no acidosis)
14. Temporary pacing as required. (A-V sequential pacing in case of A-V dissociation, atrial pacing

for sinus bradycardia and nodal rhythm.
15. Heart rate control (Digitalis/hypothermia for sinus tachycardia/nodal tachycardia)
16. IV Metaprolol or Amiodarone for junctional ectopic tachycardia and A-V dissociation.
17. Watch for drainage, urine output and acidosis along with vital parameters
18. Watch for Endo tracheal bleeding during ET suctioning
19. NG feeds from POD 1 (if ventilation planned for more than 24 hours).
20. Vasodilators-Bosentan /Sildenafil with a gradual increment in the dosage
21. Inhaled Nitric oxide if required due to recurrent PAH crisis.
22. Fluid balance-Target output atleast 25-50 % more than intake

Weaning criteria

The following indicators of hemodynamic stability should be considered when planning to wean from ventilator support.

1. PAP/SAP <0.75 (on invasive monitoring or echo)
2. Mean BP > related age normal
3. Adequate Peripheral perfusion
4. No increase in inotropic requirements
5. No significant metabolic acidosis
6. Urine output > 1 ml/kg/hour
7. Absence of pulmonary hypertensive episodes while weaning the ventilation.
8. No neuromuscular blockade, no or minimal IV sedation
9. Tolerate PS/CPAP ventilation without any adverse events for a considerable period
10. Normal neurologic status
11. Normal movement of diaphragm

PAP: Pulmonary artery pressure, SAP: Systemic arterial pressure

Management of pulmonary hypertensive crisis

Pulmonary arterial hypertensive crisis (PAH) is defined as pulmonary artery pressures rising to atleast 2/3 rd of the systemic arterial pressure. It is usually characterized by desaturation and systemic hypotension. The presence of a decompressing channel in the form of a patent foramen ovale or an atrial septal defect or an unligated vertical vein or a ventricular septal defect may prevent acute RV failure and low cardiac output. The presence of a pulmonary artery pressure monitoring catheter can help to establish the diagnosis of pulmonary hyptertensive crisis. In the absence of a decompressing channel, an acute raise in the right atrial pressure may be indicative of pulmonary arterial hypertension.

Clinical manifestations include tachycardia, a sudden desaturation with or without bronchospasm. Most commonly, systemic hypotension is seen. Cyanosis may be witnessed if the shunting through the decompressing channel is significant. The most effective strategy to manage PAH (Pulmonary artery hypertension) is prevention. The following measures are to be followed for prevention and management of pulmonary hypertension.

- Maintenance of adequate sedation and analgesia, particularly during ET suctioning
- Muscle relaxant and deep sedation if hemodynamics are stable
- Maintaining a pH of 7.45 -7.5, i.e. respiratory alkalosis in acute pulmonary hypertensive crisis situation, by hyperventilation.
- Bosentan/Sildenfil in the absence of inhaled nitric oxide
- Inotropes to maintain cardiac output
- Extra corporeal Membrane oxygenation if recurrent crisis and hemodynamically unstable.[14]

Extra corporeal membrane oxygenation (ECMO)

Extracorporeal membrane oxygenation (ECMO) has become the most widely used mode of mechanical cardiopulmonary support for children after cardiac surgery[21,22]. Children with cardiac dysfunction and pulmonary hypertensive crisis unresponsive to conventional pharmacologic therapy may be supported with ECMO in the immediate post op period. The hospital survival rates in children receiving ECMO after heart surgery have ranged between 35% and 60%.[23-29]

Conclusion

Management of a neonate or infant with TAPVD involves a major perioperative connect between the referring physician and the pediatric cardiologist, pediatric cardiac surgeon and intensivist. The outcome in major centre is entirely dependent upon early detection rapid perioperative stabilization and timely surgical interventions. An intervention at the

right time ensures a essentially normal cardiac status after recuperation, but it is desirable to have a long term growth and neuro developmental follow up.

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