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Systemic-Pulmonary Artery Shunt

The systemic-pulmonary artery shunt is an anastomosis or a connection between a branch of systemic artery and pulmonary artery (PA), which is done in an attempt to increase pulmonary blood flow.

22.1 Types of Shunts

Waterston shunt (ascending aorta and right PA anastomosis), Potts shunt (descending aorta and left PA anastomosis), and Blalock Taussig shunt (subclavian artery to PA anastomosis).

Classical BTS. It is a direct anastomosis of the subclavian artery to the pulmonary artery (PA). The end of the divided subclavian artery is anastomosed to the side of the branch pulmonary artery. It is usually performed on the side of the chest opposite to the aortic arch (i.e., subclavian artery branch of the innominate artery).

Modified BT (Blalock-Taussig) shunt: It is an anastomosis between the side of the right or left subclavian artery to the side of the branch PA, with an interposition polytetrafluoroethylene (Gore-Tex) graft. It is usually performed on the side ipsilateral to the aortic arch, using a 4 mm graft.

BTS remains an option for infants who have anatomical considerations that prevent an early total correction, particularly, those that are unstable at presentation.

Currently, an increasing number of shunts is being used in the management of the single ventricle.

22.2 Pathophysiology

The size and length of the shunt determines the amount of blood flow to the lungs. It is important to balance pulmonary and systemic blood flows by creating an appropriate shunt. It is critical to balance the perfusion of the

systemic and pulmonary bed due to cardiovascular and hemodynamic changes occurring in the postoperative period.

22.2.1 Too Large a Shunt

If the shunt created is too large, it may lead to a relatively excessive pulmonary blood flow (pulmonary overcirculation) and high O_2 saturations. It manifests with heart failure, pulmonary edema, and poor systemic perfusion. This may also lead to difficulties when ventilation is weaned.

The systemic hypoperfusion manifests with a low blood pressure, low mixed venous saturations, and rising base excess and lactate. The low diastolic pressure may compromise the coronary circulation, and the shunt may be emergently clipped or taken-down.

22.2.2 Too Small a Shunt

If the shunt created is too small, the systemic O_2 saturations fall due to inadequate pulmonary perfusion, which will lead to systemic arterial hypoxia and poor oxygen delivery to the tissues and later development of metabolic acidosis.

22.3 Blocked BT Shunt / Shunt Failure

The shunt may be blocked, requiring expeditious management of the problem.

22.3.1 Clinical Diagnosis

Significant and sustained desaturation of blood occurs and a shunt murmur is no longer audible. It will be followed by hypotension. If not resolved, this will lead to death.

Its incidence is high in the immediate postoperative period and presents acutely with a precipitious drop in oxygen saturations. It is usually secondary to a shunt clotting or kinking. This is an emergency. Most likely to occur if the shunt is new or the patient is dehydrated, or if the shunt flow is competing with an open ductus arteriosus.

22.3.2 Management of Shunt Failure

Implement the following ABCD-s expeditiously.

1. Rule out the following causes of arterial desaturation:
   - Displaced ET (endotracheal tube) position.
   - Obstructed ET tube.
   - Pneumothorax or hemothorax.
   - Ventilator equipment failure.
2. Auscultate for the shunt murmur that has disappeared.
3. Re-check blood gas for a low PaO_2, rising lactate, and correct metabolic acidosis.
4. Review most recent APTT.

22.3.3 Treatment

If one is assertive that the shunt could have blocked, implement following ABCDs:

A. *Initiate A, B, C* of cardiopulmonary resuscitation.

B. *Perform* an urgent echocardiogram.

C. *Call a* surgeon and / or cardiologist immediately.

D. *If highly suspicious* that shunt may have been blocked, do not wait for the results of echocardiogram. *Immediately act and do the following:*

1. Hand ventilate and give a bolus of sedation.
2. Increase SVR (systemic vascular resistance) in a stepwise fashion by:
   a) 5 mL/kg aliquots of 4% to 5% of human albumin as a volume.
   b) Phenylephrine:

      Dose 3-10 micrograms/kg.

      (Dilute 10 mg in 50 mL 5% DW and give 0.02-0.05 mL/kg as a slow IV injection).
   c) Start dopamine infusion or increase the rate of infusion (if already is on).
   d) Start levophed infusion.
3. Reduce PVR (pulmonary vascular resistance) by :
   a) Sedation and paralysis.
   b) Hand ventilate: - Oxygenate with 1.0 FIO_2. Decrease $PaCO_2$ - Aim alkalosis.
4. May institute the following additional measures:
   a) Bolus of magnesium sulphate - (0.4 mmol/kg of 50% magnesium sulphate).
   b) Bolus of heparin 50 units/kg followed by heparin infusion at 20 units/kg/hr, or increase the rate of heparin infusion if already is on.
   c) Restart prostaglandin in the neonate.
   d) Consider a second bolus of heparin (50 units/kg).

e) Implement the following actions decided by a surgeon or a cardiologist:

- Administer *tissue plasminogen activator* (alteplase).
- Surgical or catheter intervention.

22.3.4 Thrombolytic Therapy

Recombinant tissue plasminogen activator (alteplase) (TPA):

It may be successful in unblocking shunts. It is contra-indicated if operated < 10 days or in the presence of active bleeding.

Mechanism of Action:

Binds selectively to the fibrin clot-bound protein, plasminogen, and activates to form plasmin which has fibrinolytic action. It has a plasma half-life of 4-5 mins. Only < 10% remains for 20 minutes after it is stopped, but the bleeding tendency can persist for much longer.

Method of administration:

It is given both systemically and directly into the shunt.

The dosage is controversial.

Local administration has been reported with 0.1 mg/kg and 0.2 mg/kg boluses, followed by a systemic infusion of 1 mg/kg for 24 hours.

The dose in children is reduced to < 0.1 mg/kg/hr for 12 hours.

The other recommended dose is: 100-500 micrograms/kg/hour for 3 to 6 hours with a follow up imaging. Maximum of 100 mg/day.

In view of the dose controversies, TPA (alteplase) should only be given for a shunt blockage with agreement of a consultant cardiologist, surgeon and a hematologist.

Following TPA protocol is commonly adopted:

1. Stop heparin infusion.
2. *Give 10 mL*/kg of FFP (fresh frozen plasma) prior to commencing TPA in a neonate.
3. Dose: 0.1 - 0.6 mg/kg/hr. Start at a low dose and titrate up if no complications. Can be given for > 6 hrs and
4. Closely monitor: 3 to 4 hourly, fibrinogen, clotting tests (screen) including d-dimers (d-dimers should rise as thrombolysis occurs).
5. Make sure platelet the count is > 100,000 and serum fibrinogen is > 200 mg%.
6. Review of the clot with * an echocardiogram or angiography every 6 to 8 hours.

Standard thrombolytic therapy should be followed with a heparin infusion aiming for an APTT of 60-90.

* Echocardiogram does not always demonstrate the clot. Catheter angiocardiogram may be useful, both from a diagnostic and an interventional stand point. Clot destruction via a catheter may be an option in some specific circumstances.

If there is a concern regarding surgical technique, like kinking of the shunt or deterioration of an infant despite above interventions - return to an operation theatre.

22.4 Anticoagulation of Modified BT Shunt

Prevention of clot formation within BT shunt postoperatively with use of anticoagulants is controversial. Studies have showed that 1/5 of the shunts

electively taken down had 50% stenosis at a median age of 8 months. Many centers now anticoagulate BT shunts postoperatively. Aspirin may reduce the rate of shunt thrombosis while others have failed to prove this. The postoperative heparin usage may be linked to causation of seromas.

Patients are usually given heparin in an operation theatre or commence heparin when bleeding is not an issue:

(i.e., chest drains are bleeding < 3 ml/kg/hr and APTT < 60 postoperatively).

Heparin dose is usually not titrated to APTT unless there is a specific reason to do so. It is given in two methods.

22.4.1 Routine Heparin Prophylaxis

One should ensure APTT does not rise too high (> 80).

1000 units (10 mg) of heparin per kg body weight of an infant. It is diluted to 50 mL with 0.9% sodium chloride or 5% dextrose in water (D5W). The solution has 20 units/per kg BW/mL..

Infusion is commenced at 10 units/kg/hr (0.5 mL/hr).

Monitor: APTT, PT and serum fibrinogen. Monitoring is done as below to ensure APTT does not rise too high > 80.

1. Prior to commencement of heparin.
2. 4-6 hours after commencement of the infusion.
3. One hour after a syringe is changed.
4. Daily while on heparin infusion.

22.4.2 Therapeutic Heparin Regimen

It is not routine -only administered by a surgeon’s decision.

Heparin is titrated to achieve an APTT of 60-90.

Prepare infusion using 1000 units of heparin sodium/kg BW of an infant. It is diluted to 50 mL with 0.9% sodium chloride or D5W.

Loading dose is not required if recently returned from an operation theatre.

If APTT is < 50s: Give a loading dose of 50 units/kg (2.5 mL) over ten minutes.

If APTT is 50-90s: Run infusion at 20 units/kg/hr (1 mL/hr) and check APTT in *4 hours* (see Table 21.1).

Table 21.1 ** Therapeutic Heparin Protocol.*

APTT	Bolus dose	Stop infusion	Rate change	Recheck APTT
<50	50 (u/Kg)		+10%	4 hrs
50-59				4 hrs
60-90				Next Day
91-99			- 10%	4 hrs
100-120		for 30 minutes	-10%	4 hrs
>120		for 60 minutes	-15%	4 hrs

*Adapted from J. Richardson, Immediate Postoperative Management of Blalock-Taussig shunt (BTS). Royal hospital for sick children. July 2010.

Any change in heparin dosage should be followed by a repeat clotting profile at 4 hours. If there is a need for > 30 units/kg/hr of heparin to achieve APTT, it should be made known to the consultant. Consider measuring anti-Xa level or anti-thrombin III level. It is not unusual for a child < 1 year to require 28 units/kg/hr to achieve therapeutic APTT levels.

22.4.3 Thrombocytes

Check *once a day* while on heparin.

If platelets drop by > 50% from the baseline, think of heparin induced thrombocytopenia (HIT); HIT is more likely after 5-10 days of treatment.

Evaluate for other causes for HIT such as necrotizing enterocolitis, infection, etc.

Consider HIT antibody screen.

22.4.4 Aspirin

Must fulfill the following criteria to start aspirin:

1. Chest closed.
2. Major intracardiac lines are removed (pulmonary arterial/ left atrial lines).
3. Pacing wires are out.
4. Tolerating and absorbing oral feeds well.

Dose: 3-5 mg/kg (maximum 75 mg) once daily.

Discontinue heparin after a second dose of aspirin is given.

22.5 Pulmonary Overcirculation

If the BT shunt is too large, this may lead to relatively excessive pulmonary blood flow and high oxygen saturations. It is more common if the ductus arteriosus is still open and may resolve as the duct closes.

The definitive management is dependent on the anatomy and physiology of the underlying lesion and surgeon’s decision.

The following is only a guide in diagnosis and initial management.

22.5.1 Clues for Diagnosis

Pulmonary overcirculation may manifest in the early postoperative period or becomes more problematic and becomes a severe issue when ventilation is weaned.

Patient has tachycardia. Mean arterial blood pressure is relatively low. Widening toe to core temperature gap, relatively a high arterial oxygen saturation, low mixed venous oxygen saturations, a rising lactate, and an increase in base deficit are noted.

Diastolic blood pressure may be low; if it is low, it may compromise coronary flow. Therefore, identify for the signs of myocardial ischemia (by EKG changes). Chest x-ray shows pulmonary edema or congestion. Signs of right heart failure (large liver, ascites) appear late.

22.5.2 Management

(I) Fluid Restriction & Diuretics

Mild form may be treated simply with restriction of fluids and use of diuretics.

(II) Manipulation of Pulmonary and Systemic Vascular Resistances

It is required if pulmonary overcirculation becomes problematic and doesn't respond to diuresis.

Emergency situation may also arise if myocardial ischemia (by EKG changes) is also present. One should repeat echocardiogram and perform 12-lead EKG.

Steps to ↑ PVR:

Reduce inspired (FIO_2) oxygen, increase PEEP, and allow arterial PCO_2 to rise steadily and slowly.

Steps to ↓ SVR:

- Consider reducing vasopressor dose slowly.
- Consider vasodilatation: I.e., use of milrinone or sodium nitroprusside.
- Consider cardiac inotrpes: Low cardiac output state may be associated with pulmonary overcirculation; therefore, use of inotropes may be required.

Clipping of a shunt:

The shunt may occasionally need to be clipped or taken-down and should be done quickly if low diastolic pressure is compromising the coronary circulation.

22.6 Miscellaneous Issues with Systemic - Pulmonary Artery Shunt

22.6.1 Respiratory Insufficiency

Ventilator dependent respiratory failure in newborns or young infants after shunt operation may be attributed to a phrenic nerve injury during surgical dissection. If persists, it may require diaphragmatic plication.

22.6.2 Intrathoracic Bleeding

Intrathoracic bleeding: Injury to aortopulmoanry collateral vessels may occasionally result in significant hemorrhage.

22.6.3 Vocal Cord Paralysis

Vocal cord paralysis: Very rare and may occur in a newborn due to injury to the recurrent laryngeal nerve.

22.6.4 Horner's Syndrome

Horner's syndrome: Rarely, occurs due to injury to cervico-thoracic sympathetic fibers in the vicinity of great vessels.

22.6.5 Chylothorax

Chylothorax: It is a rare complication due to injury of the thoracic duct. Limited lymphatic leak is more common due to the disruption of thoracic lymphatics.

22.6.6 Gangrene

Gangrene: The arm may rarely become ischemic due to division of the subclavian artery (in classical BT shunt). Diminished growth of the arm may be noticed after many years. Ischemia may also rarely occur in the modified shunt due to diminished blood flow in the subclavian artery distal to the shunt.

22.6.7 Seroma

Seroma: Leakage of serous fluid from PTFE graft may result in prolonged chest drainage or a seroma formation.

22.6.8 Pulmonary Hemorrhage

Pulmonary hemorrhage: Hemoptysis or major pulmonary hemorrhage occurring late after BT shunt is most probably due to pseudoaneurysm formation at the shunt site, with rupture into the pulmonary parenchyma.