Immediate Post-operative Management of Blalock- Taussig Shunt (BTS)

Royal Hospital for Sick Children
PICU

1. Rationale/Purpose/Objective

- To allow standardised management of anticoagulation in patients with a Blalock-Taussig Shunt in PICU
- Provide evidence base for management of a suspected blocked BT shunt.
- Provide educational material to assist staff managing a child with a BT shunt in the PICU.

2. Scope

- This guideline applies to the immediate post-operative management of any patient with a Blalock-Taussig Shunt in PICU.

3. Roles and responsibilities

- All healthcare professionals involved in caring for a post-operative cardiac patient in the PICU should be aware of this guideline.

4. Evidence

- The guidelines are constructed after consultation with standard textbooks, a medline search (BT shunt, systemic to pulmonary shunt, blocked shunt, occluded shunt, shunt failure, Streptokinase and shunt, TPA and shunt, thrombolysis, aspirin, heparin, warfarin, anticoagulation and shunt, anticoagulation in children) and local expert opinion. The best available levels of evidence were used to construct these guidelines. Level 1 evidence is lacking in this area.

1. Background

The Blalock- Taussig shunt (BTS) was first described in the 1940's. The Classical BTS was a direct anastomosis of the subclavian to the pulmonary artery(PA). This developed into the Modified BT shunt in the 1970's - Right or left subclavian artery to branch PA with a Gore-Tex shunt, this is the type of shunt that will be found on the unit today. [1-3]

The annual number of BT shunts being performed has fallen over the last 20 years. This is largely due to advancing surgical technique. BT shunts were originally used predominately in the management of Tetralogy of Fallot (TOF), now most TOF go straight to a full repair. BTS however remains an option for infants; particularly those that are unstable at presentation or who have anatomical considerations that prevent early total correction.[1-3]

Although the overall numbers of BT shunts being inserted has fallen, increasing proportions are being used in the management of the single ventricle patient.

This has coincided with an increased length of stay of shunt babies in the ICU. The mean length of stay for an uncomplicated shunt is 3 days.[4] A BT shunt may be placed in isolation or increasingly may be part of a more complex operation such as the Stage 1 Norwood for the hypoplastic left heart.[2,3]

The BT shunt is usually inserted to increase blood to flow to the lungs. The size and length of the shunt in part determine the amount of blood flow to the lungs. If the shunt is too big this may lead to relatively excessive pulmonary blood flow and high saturations described as pulmonary overcirculation. This may reveal itself with oedematous lungs, heart failure and poor systemic perfusion. The current trend would be to place as big a shunt as possible and allow the baby to grow into it, meanwhile managing the circulation with diuretics. If a shunt is too big this may lead to difficulties when ventilation is weaned. The shunt may occasionally be clipped or taken-down. This may need to be done quickly if low diastolic pressure is compromising the coronary circulation. Pulmonary overcirculation may also present with systemic hypoperfusion. This may be revealed with low blood pressure, low mixed venous saturations and a rising base excess and lactate. [1 2,3,5,5-7]
If the shunt is too small the baby will be desaturated and inadequate pulmonary perfusion will lead to hypoxia and poor oxygen delivery to tissues.[5-7]
Obviously there is a careful balancing act between pulmonary and systemic perfusion. When a BT shunt is inserted there is a dramatic change in physiology from the pre-operative state. "It's just a shunt" should be a phrase banned from the intensive care. In the postoperative period attention to detail is required as there can be frequent haemodynamic shifts as the cardiovascular system readjusts.[5]

The immediate post operative period is a time where the incidence of shunt failure is high. This can present acutely with precipitously dropping saturations. Acute shunt failure is usually secondary to the shunt clotting off or kinking. This is an emergency and the management is discussed below.[1,5,8]

The incidence of shunt thrombosis is reported at 12%.[9] Prevention of clot formation within shunts is controversial. Some studies have suggested that aspirin may reduce rate of shunt thrombosis while others have failed to prove this.[9-11,11,12] The use of heparin post-operatively is also controversial, some believe it is unnecessary and may be linked to causation of seromas.[10,13] Shunts all have an attrition rate; a study looking at the histopathology of shunts electively taken down found 21% had a 50% stenosis at a median age of 8 months. Smaller shunts were more likely to stenose.[3,7] Many centres anti-coagulate. Our current anticoagulation guideline is displayed on the following pages.

2. Anticoagulation management

Introduction of anti-coagulant therapy with heparin should be instituted as soon as safely possible after theatre. Confirm with the consultant surgeon at handover that there are not specific reasons to avoid the following standard approach. Patients are usually given heparin in theatre.
Commence heparin when bleeding is not an issue (chest drains are bleeding less than 3 ml/kg/hr) **and** the post-op APTT is less than 60.
Generally heparin is **not** titrated to APTT unless there is a specific surgical request.

Heparin therapy should be managed as follows:

a) Standard heparin regime for prophylaxis[10,14]

Prepare infusion using 1000units/kg of heparin sodium, diluted to 50ml with 0.9% sodium chloride.

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

Monitoring:

- **Clotting profile** (APTT, PT and fibrinogen) should be checked As follows:
  - Prior to commencement of heparin
  - **4-6 hours** after INITIAL commencement
  - **One hour** after a syringe is changed
  - **Daily** whilst on heparin infusion

This is to ensure the patient's APTT does not rise too high (>80)

- **Platelets**
  - **Daily**

If platelets drop by >50% from baseline consider Heparin Induced Thrombocytopenia (HIT), this is most likely 5-10 days of treatment. However there are many other reasons for thrombocytopenia (NEC /infection etc) Notify consultant. Consider HIT antibody screen.

b) Therapeutic heparin regime[10,14]

Not routine - specific surgical request

Titrate heparin to achieve an APTT of 60-90
Prepare infusion using 1000units/kg of heparin sodium, diluted to 50ml with 0.9% sodium chloride.

Loading dose is not required if recently returned from cardiac theatre

- *ONLY IF APTT <50s*: Initial loading dose of 50units/kg (2.5ml) over ten minutes.
- *IF APTT 50-90s*: Run infusion at 20units/kg/hr (1ml/hr) and check APTT in **4 hours**. Manage as follows:

aPTT (s)	Bolus (units/kg)	Stop infusion	Rate Change	Recheck APTT (hr)
<50	50	[redacted]	+10%	4
50-59	[redacted]	[redacted]	[redacted]	4
60-90	[redacted]	[redacted]	[redacted]	Next day
91-99	[redacted]	[redacted]	-10%	4
100-120	[redacted]	30min	[redacted]	4
>120	[redacted]	60min	-15%	4

Most importantly any **change** in heparin dosage should be followed by a **repeat** clotting profile at 4 hours.
The need for more than 30 units/kg/hr of heparin to achieve APTT should be made known to the consultant.
Consider measuring anti-Xa level or anti-thrombin III level. This will be age dependent. It is not unusual for a child<1 to require 28units/kg/hr to achieve therapeutic APTT levels.
Once the APTT is achieved and the heparin infusion is stable – once daily clotting profile is acceptable.

- **Platelets**
  - Check **once a day** whilst on heparin
  - If platelets drop by >50% from baseline consider Heparin Induced Thrombocytopenia (HIT), this is more likely after 5 -10 days of treatment. However there are many other reasons for thrombocytopenia (NEC /infection etc) Notify consultant. Consider HIT antibody screen.

c) Aspirin
Must fulfill the following criteria to start aspirin:

1. Chest closed
2. Major intracardiac lines removed (pulmonary arterial/ left atrial lines)
3. Pacing wires out
4. Absorbing feed

Aspirin is commenced at 3-5mg/kg (max 75mg) once daily. Continue heparin until the second dose of aspirin is given.

2. Management of a Suspected Blocked BT Shunt [5,6,9,15]

This is an emergency

Diagnosis
Consider in any patient who has a **significant sustained desaturation** with a systemic to pulmonary cardiac shunt, or whose saturations drop and a shunt murmur is no longer audible.
Most likely to occur in a new shunt or in a dehydrated patient known to have a shunt. It is also more likely if flow is competing with an open duct (PDA).

Check the obvious:
Desaturation can frequently be respiratory in nature.
DOPE (**D**isplaced ET position, **O**bstructed ET, **P**neumothorax, **E**quipment failure)
Auscultate – previous obvious shunt murmur will have disappeared
Check gas - ?low PaO2, rising lactate, correct acidosis
Review most recent APTT.
If the shunt is blocked the patient will profoundly desaturate.
This will be followed by hypotension.
In the worst case if not resolved this will lead to death

Principle of management …. Think shunt.
Think could shunt have blocked? Act quickly.

Management [5,6,15]
Resuscitate – A,B,C
Request Urgent Echo
Inform surgeon and cardiologist immediately.
Do not wait for Echo – if suspicious start management and escalate as necessary.
Call surgeon immediately there is concern shunt may have blocked – do not wait.

Meanwhile

1. Hand ventilate
2. Bolus sedation
3. Increase SVR – Stepwise
   a) 5ml/kg aliquots of 4% human albumin volume
   b) Phenylephrine
      (The dose is 3-10microgram/kg. Dilute 10mg in 50ml 5% glucose and give 0.02-0.05ml/kg as a slow IV injection)
   c) Start or increase dopamine infusion
   d) Start noradrenaline infusion
4. Reduce PVR - Sedate and consider paralysis
   Hand ventilate – decrease pCO_2 – aim alkalosis.
   Oxygenate

Consider bolus magnesium sulphate –
(0.4mmoll/kg magnesium sulphate 50%)

5. Bolus heparin 50 units/kg
6. Start heparin infusion at 20 units/kg/hr or increase heparin infusion if already running.
7. Restart prostaglandin in neonate.
8. Consider second bolus of heparin 50 units/kg
9. Consider Tissue Plasminogen Activator (alteplase) * Decision by Intensivist, Cardiologist and Consultant surgeon.
10. Decide for catheter or surgical intervention. **

*There are case reports that suggest recombinant tissue plasminogen activator (alteplase) may be successful in unblocking shunts. However its use must be considered carefully. A relative contraindication for use is recent surgery. Contra-indicated if surgery within 10 days or active bleeding.[10,14,16,17]

TPA (alteplase) is short acting, it binds selectively to fibrin-clot bound plasminogen and activates to form the fibrinolytic plasmin. It has a plasma half-life of 4-5 mins. Less than 10% remains 20 minutes after it is stopped, but bleeding tendency can persist for much longer. The case reports vary in method of administration between systemic treatment and directly administered into shunt. Local administration has been reported with 0.1mg/kg and 0.2 mg/kg boluses followed by a systemic infusion of 1mg/kg for 24 hours. However doses being used to clear arterial and venous thrombi in children have been much lower than this < 0.1mg/kg/hr for 12 hours. Dosage is therefore an issue.[3,10,14,17-19] The BNF-C recommends a dose of 100-500micrograms/kg/hour for 3 to 6 hours with follow up imaging. Max of 100mg/day.

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

On consultation with haematology the following dose schedule has been agreed:

Tissue plasminogen activator (alteplase). Stop heparin infusion. In neonate give 10ml/kg of FFP prior to commencing TPA(alteplase).

Dose	Investigations	Ensure
0.1 -0.6mg/kg/hr Start at low dose and titrate up if no complications. Can be given for>6hrs closely monitored	3 hourly FBC, Coag screen including d-dimers (d-dimers should climb as thrombolysis occurs)	 Platelets >100 Fibrinogen>2
	6 hourly Review of clot with imaging	

Standard thrombolytic therapy should be followed with a heparin infusion aiming for an APTT of 60-90.[10,14,17]

**Cardiac catheter may be useful from a diagnostic and interventional viewpoint. ECHO does not always demonstrate clot, angiography may help if there is doubt regarding diagnosis. Clot destruction via catheter has been demonstrated and may be an option in some specific circumstances.[16,19,22 -25]

However if there is any concern regarding surgical repair or a deteriorating child despite above interventions– return to theatre should not be delayed.

3. Management of Pulmonary Over-circulation

The management of pulmonary over-circulation can be complex and is dependent on the anatomy and physiology of the underlying lesion. The following is only a beginners guide to recognising and managing this difficult problem. Consultant input is essential.

If the BT shunt is too big this may lead to relatively excessive pulmonary blood flow and high saturations described as pulmonary over-circulation. This is more common if the ductus arteriosus is still open and may resolve as the duct closes.
Pulmonary over-circulation may reveal itself in the early post-operative period or become more problematic when ventilation is weaned.

Diagnostic clues
Relatively high saturations
CXR- oedematous lungs
Low mixed venous saturations
Rising lactate
Increase in base deficit
Often tachycardic
May have relatively low mean blood pressure
Widening toe core gap
Review NIRS – decreased splanchnic and then cerebral saturations
Signs of right heart failure e.g large liver, ascites(late sign)

Important to review diastolic pressure – if low this may compromise coronary flow.
Look for signs of ischaemia –ECG changes

Treatment
Mild form may be treated simply with fluid restriction and diuretics.
As this becomes more problematic, manipulation of pulmonary and systemic vascular resistance(PVR and SVR) is required.

If ECG changes are present this is an emergency. Inform intensive care consultant
Repeat Echo and 12-lead ECG.

Increase PVR
Reduce oxygen
Increase PEEP
Allow pCO_2 to rise gently

Reduce SVR
Consider reducing vasopressor therapy slowly
Consider vasodilation – eg. Milrinone or SNP (discuss with intensive care consultant)
Overcirculation may also be present in conjunction with a low cardiac output state therefore inotropy may be required.

The shunt may occasionally need to be clipped or taken-down. This may need to be done quickly if low diastolic pressure is compromising the coronary circulation. [1] [2,3,5,5-7]

This guideline will be reviewed every 2 years.

Reference List

1. Williams,J.A. *et al.* Two thousand Blalock-Taussig shunts: a six-decade experience. *Ann. Thorac. Surg.* **84**, 2070-2075 (2007).

2. Bove,E.L. *et al.* The modified Blalock-Taussig shunt: analysis of adequacy and duration of palliation. *Circulation* **76**, III19-III23 (1987).

3. Gladman,G., McCrindle,B.W., Williams,W.G., Freedom,R.M. & Benson,L.N. The modified Blalock-Taussig shunt: clinical impact and morbidity in Fallot's tetralogy in the current era. *J. Thorac. Cardiovasc. Surg.* **114**, 25-30 (1997).

4. Swain,S.K. *et al.* Neonatal Blalock-Taussig shunt: technical aspects and postoperative management. *Asian Cardiovasc. Thorac. Ann.* **16**, 7-10 (2008).

5. Schindler,M. Care of the Critically Ill Child. Macnab,A., Macrae,D. & Henning,R. (eds.), pp. 198-199 (Churchill Livingstone,2002).

6. Lok,J.M., Spevak,P.J. & Nichols,D.G. Critical Heart Disease in Infants and Children. Nichols,D.G. *et al.* (eds.), pp. 806-807 (Mosby Inc, Philadelphia,2010).

7. Wells,W.J. *et al.* Obstruction in modified Blalock shunts: a quantitative analysis with clinical correlation. *Ann. Thorac. Surg.* **79**, 2072-2076 (2005).

8. Ahmad,U. *et al.* Modified Blalock-Taussig shunt: immediate and short-term follow-up results in neonates. *Heart Lung Circ.* **17**, 54-58 (2008).

9. Li,J.S. *et al.* Clinical outcomes of palliative surgery including a systemic-to-pulmonary artery shunt in infants with cyanotic congenital heart disease: does aspirin make a difference? *Circulation* **116**, 293-297 (2007).

10. Monagle,P. *et al.* Antithrombotic therapy in neonates and children: American College of Chest Physicians Evidence-Based Clinical Practice Guidelines (8th Edition). *Chest* **133**, 887S-968S (2008).

11. Motz,R., Wessel,A., Ruschewski,W. & Bursch,J. Reduced frequency of occlusion of aorto-pulmonary shunts in infants receiving aspirin. *Cardiol. Young.* **9**, 474-477 (1999).

12. Tweddell,J.S. Aspirin: a treatment for the headache of shunt-dependent pulmonary blood flow and parallel circulation? *Circulation* **116**, 236-237 (2007).

13. Mullen,J.C., Lemermeyer,G. & Bentley,M.J. Modified Blalock-Taussig shunts: to heparinize or not to heparinize? *Can. J. Cardiol.* **12**, 645-647 (1996).

14. Andrew,M. Anticoagulation and thrombolysis in children. *Tex. Heart Inst. J.* **19**, 168-177 (1992).

15. Booker,P.D. Pediatric Cardiac Anesthesia. Lake,C.L. & Booker,P.D. (eds.), pp. 654-681 (Lippincott Williams and Wilkins, Philadelphia,2005).

16. Singh,V. *et al.* Thrombolysis with percutaneous transluminal balloon angioplasty of a blocked modified Blalock-Taussig shunt. *Indian Heart J.* **56**, 673-676 (2004).

17. Williams,M.D. Thrombolysis in children. *Br. J. Haematol.* **148**, 26-36 (2010).

18. Manco-Johnson,M.J. *et al.* Recommendations for tPA thrombolysis in children. On behalf of the Scientific Subcommittee on Perinatal and Pediatric Thrombosis of the Scientific and Standardization Committee of the International Society of Thrombosis and Haemostasis. *Thromb. Haemost.* **88**, 157-158 (2002).

19. Suda,K., Matsumura,M. & Matsumoto,M. Recanalization of occluded modified Blalock-Taussig shunt using topical recombinant tissue plasminogen activator with balloon angioplasty. *Cardiol. Young.* **14**, 322-323 (2004).

20. LeBlanc,J.G. *et al.* Treatment of grafts and major vessel thrombosis with low-dose streptokinase in children. *Ann. Thorac. Surg.* **41**, 630-635 (1986).

21. Rajani,R.M., Dalvi,B.V., Kulkarni,H.L. & Kale,P.A. Acutely blocked Blalock-Taussig shunt following cardiac catheterization: successful

recanalization with intravenous streptokinase. *Am Heart J.* **120**, 1238-1239 (1990).

22. MacMillan,M., Jones,T.K., Lupinetti,F.M. & Johnston,T.A. Balloon angioplasty for Blalock-Taussig shunt failure in the early postoperative period. *Catheter. Cardiovasc. Interv.* **66**, 585-589 (2005).

23. Ormiston,J.A., Neutze,J.M., Calder,A.L. & Hak,N.S. Percutaneous balloon angioplasty for early postoperative modified Blalock-Taussig shunt failure. *Cathet. Cardiovasc. Diagn.* **29**, 31-34 (1993).

24. Sivakumar,K. *et al.* Emergency transcatheter balloon recanalization of acutely thrombosed modified Blalock-Taussig shunts. *Indian Heart J.* **53**, 743-748 (2001).

25. Sreeram,N., Walsh,K. & Peart,I. Recanalisation of an occluded modified Blalock-Taussig shunt by balloon dilatation. *Br. Heart J.* **70**, 474-475 (1993).