



NiSTAR Clinical Guideline

Congenital heart disease

1. Introduction

All referrals for neonates with suspected congenital heart disease should be discussed with the cardiology registrar and/or consultant at RBHSC. The diagnosis will often be facilitated by telelink to the children's cardiac ward.

The baby may be transferred to RBHSC, or if ventilated to either RMH or PICU RBHSC depending on cot availability. The exact destination must be established before transfer.

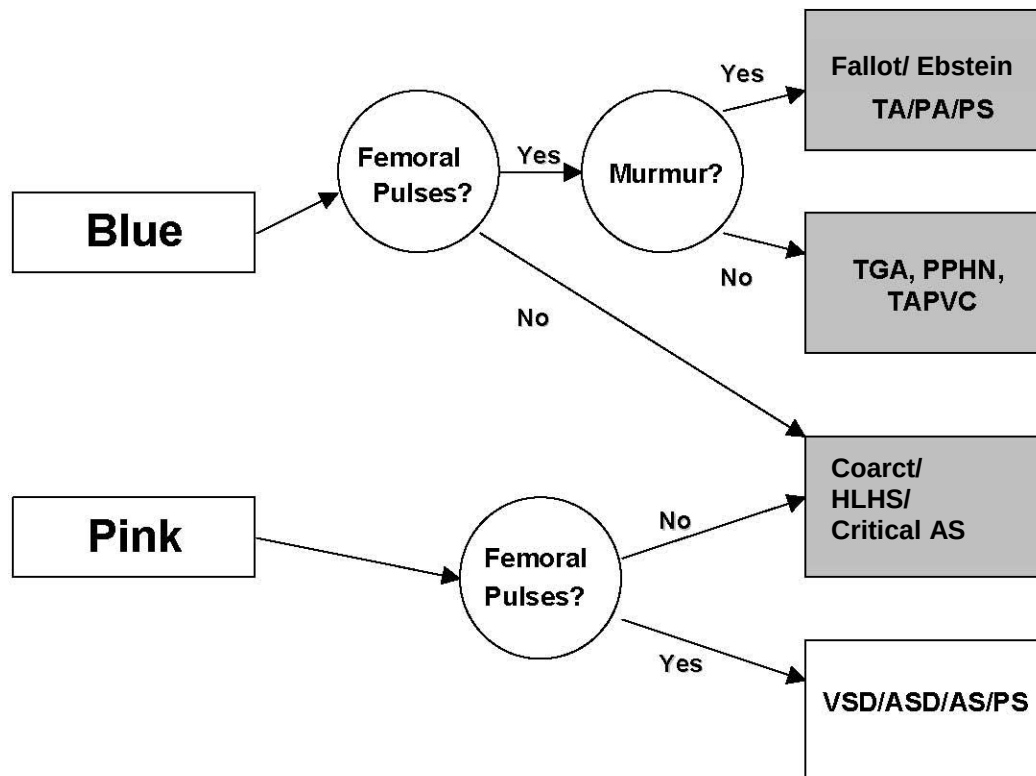
2. Assessment

Most congenital cardiac disease requiring intensive care will present either as cardiogenic shock or cyanosis. A diagnosis of congenital cardiac disease should always be considered in any shocked neonate.

Ask about:

- Antenatal scans and family history
- History of labour, delivery and resuscitation
- Cyanosis - present from birth or time of onset, and severity?
- SpO₂, pre- and post- ductal
- Respiratory distress
- Perfusion and pulses
- Four limb blood pressures
- Cardiac murmur
- Hepatomegaly
- Arterial blood gas and lactate levels if available. Venous or capillary if no ABG available.
- 12-lead ECG
- CXR: cardiac ratio, contour and vasculature
- Echocardiogram (and who performed it). Was a telelink performed?

Approach to diagnosis



Grey box = potential duct dependent lesion: **START PROSTIN**
Consider iNO if pulmonary hypertension likely

3. Immediate management

Airway and breathing

- The nitrogen washout (hyperoxia) test is helpful in trying to distinguish cardiac and respiratory causes of cyanosis. The test is carried out by placing the infant in 100% oxygen for ten minutes. If the infant remains cyanotic, s/he is likely to have cyanotic heart disease.
- Clearly there are exceptions to this rule - severe respiratory disease or PPHN may result in persistent cyanosis even in 100% inspired oxygen. Note that most cardiologists accept that the nitrogen washout test performed properly is unlikely to close the duct, and thus is not contraindicated in duct dependent lesions.
- Once the hyperoxia test is completed aim to keep saturations in mid-80s if supplemental oxygen is required. Do not exceed a FiO_2 of 0.30-0.40 in babies with cyanotic CHD, unless Prostin is being infused and a consultant cardiologist has advised this. If critical left heart outflow tract obstruction is suspected (eg HLHS, critical aortic stenosis) aim to ventilate in air in order to avoid pulmonary overcirculation. Aim for SpO_2 in the 75-80% range.

- Indications for intubation are recurrent apnoea, shock or respiratory failure. Do not over ventilate; aim for a low-normal arterial pH. Following intubation, paralyse and sedate well with fentanyl or morphine.

Circulation

- An umbilical venous catheter or PICC line, or at least 2 peripheral venous lines, must be inserted. This is to ensure constant delivery of critical infusions.
- Resuscitate with up to 30 mls/kg volume if shocked.
- Commence prostaglandin (PgE₂) infusion if a duct dependent lesion is likely:
 - poor perfusion
 - diminished pulses
 - cyanosis
 - metabolic acidosis

Commence Dinoprostone (PgE₂) infusion at 5-10 ng/kg/min. You may use higher doses (up to 20 ng/kg/min), but watch for hypotension and fever.

Always discuss the use of higher doses with a consultant cardiologist.

To make up PgE₂ infusion:

- Take 500 micrograms of Dinoprostone and add to 550 ml of 5% glucose. Mix well.
- Put 50 ml of this solution in a 50 ml syringe (= 1 microgram per ml)
- Syringe pump infusion rate is calculated as follows:

$$\text{Drug dose (ng/kg/min)} \times \text{baby's weight (kg)} \times 60/1000 \text{ ml/h}$$

Side effects of prostaglandin are hypotension, apnoea and fever.

Consider intubation if using doses >15 ng/kg/min or if the baby is apnoeic on lower doses of PGE₂.

- If inotropes are required, add dopamine or dobutamine then adrenaline.
- Consider diuretics (furosemide 1-2mg/kg IV) in those with cardiac failure
- Check a capillary glucose and keep blood sugar in the normal range.

Transport considerations

All babies on a Dinoprostone infusion must be accompanied by a doctor in view of the above mentioned side effects

- Insert a nasogastric tube and keep the baby nil by mouth.
- Ensure sufficient air cylinders are available for babies ventilated in low FiO₂. You are likely to require 3 E size air and 1 E size oxygen cylinders on the transport trolley.
- PgE₂ should preferably be given by PICC line or UVC but can be given peripherally. In the latter case, ensure adequate venous access to allow continuous delivery of PgE₂ infusion should a line tissue.
- Babies who have been resuscitated from shock should have invasive BP monitoring in place.
- Ensure that hypotension, acidosis, hypoglycaemia are corrected before transfer.

Supraventricular Tachycardia (SVT)

Most SVTs in the neonate are re-entry atrio-ventricular SVTs utilising an accessory pathway.

SVT may initially be asymptomatic, but when prolonged may result in congestive cardiac failure and shock after a variable period. Sustained/paroxysmal SVT may develop in-utero and may result in hydrops.

A paediatric consultant cardiologist should be contacted regarding diagnostic and therapeutic management options.

Differential Diagnosis

- sinus tachycardia at rates of up to 230 beats per minute can occur in the neonate, especially in association with fever, anaemia, sepsis and pain
- ventricular tachycardia in the neonate may have a relatively narrow QRS complex and must always be considered
- SVTs other than atrio-ventricular SVTs utilising an accessory pathway are always a possibility, e.g., atrial flutter.

Investigation

- physical examination including general appearance, pulse, BP, murmur, heart failure
- 12 lead ECG shows narrow QRS complexes, retrograde or no P waves, HR>220bpm. (Later a 12 lead ECG in sinus rhythm may show pre-excitation or evidence of structural heart disease).
- Echocardiography should be performed after stabilisation. A number of important structural associations of SVT may not be picked up on clinical examination. These include
 - Ebstein's malformation
 - congenitally corrected transposition of the great arteries
 - dilated and hypertrophic cardiomyopathy
 - myocardial tumours

Initial management of re-entry atrio-ventricular tachycardia

1. Confirm that the tachycardia is narrow complex on 12 lead ECG
2. If patient relatively **stable** (not shocked): Perform vagal manoeuvres
 - icepack for infants (either commercially available or place crushed ice inside 2 plastic bags) - apply icepack to face; be careful of eyes and do not hold on face too long as ice can "burn" an infant's skin.
 - do not put pressure on eyeballs as this can result in retinal detachment
 - do not use carotid sinus massage as this may compromise cerebral circulation

3. If the patient is **unstable**: appropriate resuscitative measures should be instituted including

- consideration of D/C cardioversion
- intubation and assisted ventilation
- review of blood gases, glucose, electrolytes (sodium, potassium, ionised calcium, magnesium) and lactate

4. **Intravenous adenosine**

- need full resuscitation facilities available
- babies should have continuous ECG monitoring throughout administration of adenosine
- Adenosine can be given in a starting dose of 50 micrograms/kg, with doses increasing by 50 micrograms/kg up to 250 micrograms/kg (see BNFc). Need to wait 2 minutes between doses and check patient's vital signs.
- Adenosine should be given very quickly and as close to the baby in the intravenous set-up as possible. A three-way tap should be used so that the adenosine can be quickly flushed with normal saline. When the volume of adenosine is small it should be diluted with normal saline so that the drug has the chance to reach the body.
- If 250 micrograms/kg fails this can be repeated once if the patient is stable. Consult with cardiology.
- The recorded strip immediately after conversion to sinus rhythm has occurred should be saved and inspected for concealed pre-excitation or flutter waves which may be revealed during the first few beats after conversion to sinus rhythm.
- After a patient has been reverted a 12 lead ECG should be performed to look for pre-excitation and other abnormality.
- Rapid re-initiation of tachycardia is not uncommon, mostly due to premature atrial contractions stimulated by the adenosine. If this occurs it is reasonable to try adenosine again.
- On occasions adenosine may cause a major pause or initiate atrial fibrillation.
- If the diagnosis is an atrial tachycardia this may be revealed by adenosine which causes transient atrio-ventricular block and multiple P-waves will be seen in a row indicating atrial flutter. Some atrial ectopic tachycardias are sensitive to adenosine so that the atrial impulse will stop.

Direct Current Cardioversion

This should be considered for unstable patients after intubation and initiation of ventilation.

Safety is paramount

This is best undertaken after consultation with a cardiologist unless the urgency is extreme.

- Infants must have intravenous access and suitable sedation and analgesia.
- **For tachycardias with a regular well-defined QRS complex** the discharge should be synchronised with the QRS complex to avoid ventricular fibrillation.
- **For ventricular fibrillation or polymorphic VT** an unsynchronised shock is necessary; otherwise the device will continuously delay the shock whilst trying to track the QRS complex.
- Connect the defibrillator ECG leads to the patient to obtain an ECG recording. Confirm QRS synchronisation (not T wave) by the on-screen signal.
- Select appropriate sized paddles, ensuring there is complete coverage of the paddle by gel or gel pads. Avoid excess gel on the skin as this may form a conductive bridge between the paddles.
- Preferred Position; sternum paddle at the base of the heart, apex paddle at the apex/axilla. If there is insufficient area the sternum paddle may be applied to the front of the chest, while the apex paddle may be placed over the back, giving a front to back shock.
- SVTs usually require 1 joule per kg and ventricular arrhythmia 2 - 4 joules per kg (see current APLS protocol).

Further management

The prescription of maintenance medication should be made in consultation with the Consultant Cardiologist.

Complete heart block (complete AV block or 3rd degree heart block)

This is very uncommon but may be seen where there is maternal Lupus and other connective tissue disorders with anti-Ro or anti-La antibodies. Antibody mediated damage to conducting tissue is permanent.

Complex structural congenital heart disease, particularly with right atrial isomerism may also cause heart block.

Most babies will have been identified in utero and birth arranged in RMH.

Investigation

- physical examination including general appearance, pulse, BP, murmur, heart failure
- 12 lead ECG: P waves appear regularly at normal rate (100-170/min.) but are not followed by QRS. QRS occurs at a much slower rate.

Initial management

- Manage Airway and Breathing as appropriate
- The baby may be hydropic if the in-utero ventricular rate has been slow enough. Emergency management of hydrops with pleural and peritoneal drainage may be necessary immediately after birth.
- Do a 12-lead ECG
- Consider starting an isoprenaline infusion if there are signs of circulatory failure or congestive cardiac failure. This is more likely if the ventricular rate is <50/min. The starting dose is 0.02 microg/kg/min., increasing to a maximum dose of 0.5 microg/kg/min).
- Refer to Paediatric Cardiology for further management. This may include insertion of pacemaker, although it is extremely rare for this to be required as an emergency.