EARLY MANAGEMENT OF INFANTS WITH HYPOPLASTIC LEFT HEART SYNDROME

Infants with hypoplastic left heart syndrome are one of the most challenging sub-groups of cardiac patients to care for. This challenge begins from birth, or earlier in those with a prenatal diagnosis, and the period of greatest acute risk often continues until the time of stage II palliation. Infants with hypoplastic left heart syndrome often respond poorly to seemingly minor external stresses, or to subtle changes in their medical management. These events can lead to significant morbidity and mortality.

The goal of these guidelines is to establish a streamlined approach to the early care of infants with hypoplastic left heart syndrome, with the aim of optimising their condition from birth until the postnatal period after stage 1 palliation.

Johnny Millar, PICU
Michael Cheung, Cardiology
Christian Brizard, Cardiac Surgery
Michael Stewart, PIPER
Carl Kuschel, NICU RWH
Elske Posma, FMU, RWH
Ricardo Palma-Dias, Director of Ultrasound Services, RWH
Ian McKenzie, Director of Anaesthesia
PRENATAL MANAGEMENT FOR THOSE WITH AN IN UTERO DIAGNOSIS

1. Where possible, delivery will be scheduled to occur in Melbourne at the Royal Women’s Hospital or Monash Medical Centre.

2. Soon after initial diagnosis, fetal echo scans will be reviewed by RCH/MMC cardiologists at mid-trimester, 30 and 36 weeks’ gestation, to monitor progression and to examine for adequacy of interatrial communication. The exact timing of these scans will be determined by the perinatal obstetrician responsible for overall management of the pregnancy.

3. For interstate patients, relocation to Melbourne will generally be scheduled for around 36 weeks’ gestation. It is expected that a perinatal obstetrician in Melbourne will be provided with comprehensive details of the pregnancy as soon as a decision for delivery in Victoria has been made.

For each patient, a joint meeting will be arranged with RCH care manager(s), a social worker, a consultant cardiologist and a consultant cardiac surgeon. At this time the early and long term management as well as potential outcomes will be discussed (Booklet for parents of RCH patients).

4. This meeting will be arranged by the Fetal Medicine Unit (for Royal Women's Hospital patients), or by the referring external cardiologist. The RCH ICU nursing unit manager will be informed of all pending deliveries at this time.

5. The attending perinatal obstetrician and RCH cardiologist are responsible for ensuring that significant new findings that arise during the pregnancy (eg diagnosis of another serious abnormality) are communicated to all management team members at the perinatal hospital and the RCH.

6. The attending perinatal obstetrician has primary responsibility for determining the timing of delivery. Normal vaginal delivery following spontaneous labour at term is the goal for otherwise uncomplicated pregnancies. In the case of induction of labour, the timing will be determined by consideration of clinical and social factors as well as availability of obstetric, cardiac surgical and intensive care resources. It must be remembered that following an induction of labour delivery may occur anywhere from 0-72 hours later. The agreed induction date will be communicated to the PICU AUM and the Koala AUM, by RCH cardiology / cardiac surgery staff.

IMMEDIATE POST-NATAL CARE AT REFERRING HOSPITAL

- Birth suite management will be directly supervised by a consultant neonatologist or fellow. The baby will be transferred to the local NICU for ongoing stabilization once initial resuscitation has been completed.

- Intravenous access: A double or triple-lumen UV line should be placed before transport (or two separate peripheral lines if a UV line cannot be inserted).

- Umbilical arterial access should be obtained at the discretion of the attending neonatologist, and in general only if the neonate is ventilated and/or requiring vasoactive drugs.

- Alprostadil (prostin) 10ng/kg/min should be commenced immediately after obtaining intravenous access.
The referring neonatologist or Fellow will inform ICU and the on-call cardiologist of the birth and the patient condition and will contact PIPER to arrange transfer to RCH. PIPER will liaise with PICU on the estimated time of transfer.

- **The target oxygen saturation (SpO\(_2\)) is 75-85%. Avoid additional oxygen unless SpO\(_2\) < 70%**.
- Observe prior to transfer for prostin-related apnoeas / other instability.
- Intubation and ventilation should not be undertaken in the stable, non-acidotic patient.
- Measure blood gas (umbilical venous or arterial, not capillary), glucose and lactate prior to transfer.
- All infants will be transferred to RCH PICU.

**Management of the infant with suspected or confirmed restriction of the PFO**

This may be suspected from in utero imaging. It should also be suspected in the infant with a known diagnosis of HLHS in whom there is severe metabolic acidosis, poor oxygenation and X-ray appearance suggesting obstructed pulmonary venous drainage. Pre-operative survival and neurological outcome of these infants depends on early surgical or transcatheter intervention to enlarge the restrictive communication.

If a restrictive PFO is suspected prenatally, a consultant neonatologist should directly supervise delivery room management. If the clinical findings immediately after birth are consistent with HLHS with restrictive PFO, the baby should be intubated, commenced on prostin, muscle relaxed and sedated (see below), and transferred as early as possible to RCH. Other standard resuscitative measures are generally ineffective in this situation. For these infants, the neonatal transport team should be informed in advance of birth and available to facilitate timely transport to RCH after delivery.

Early surgical intervention (Norwood procedure) may be required for these patients ie within the first 24-48 hours if the patient remains relatively stable. Emergency surgery for resuscitation in the setting of a severely restrictive atrial communication is not recommended.

**PRE-OPERATIVE CARE AT RCH (IF UNOBSSTRUCTED ATRIAL COMMUNICATION)**

1. All infants will be admitted to PICU.
2. **Communication:** The cardiology fellow, should be informed when infant arrives on PICU.
3. **Echo:** An echo should be performed within 2 hours of arrival.  
   *Other investigations:* Chest X-Ray, ECG, FBC, blood gas + lactate, electrolytes, glucose, coags, chromosomes, group & save. Cranial and renal ultrasounds
4. **Surgery**
   - This will, where possible, be planned for the first week of life.
5. **Other Health Professionals:**
   - Families will meet with care managers and social workers after admission.
OTHER ASPECTS OF PRE-OPERATIVE MANAGEMENT:

1. Feeds
Infants with HLHS are at high risk of gut ischaemia and should not be fed enterally. Infants with central venous access should be given TPN from day 2 of life until surgery. Those without central access should receive 10%dextrose / 0.45% saline as maintenance fluid.

2. Vascular access
Simple procedures may cause fluctuations in systemic vascular resistance secondary to agitation, pain etc. Multiple attempts at vascular access should be avoided, as this can be traumatic for the infant, and can increase the incidence of intravascular thrombus formation. Experienced personnel should obtain vascular access in these patients.

   *Peripheral lines*
   In a conscious patient, sucrose and / or paracetamol, or a small dose of sedation should be routinely given prior to line insertion.
   Access should be attempted by a senior ICU doctor (senior registrar or above).

   *Central lines*
   Central venous access is only required in the unstable patient who requires inotropic support. This should not be attempted in a non-anaesthetised patient.
   If umbilical venous access is not obtainable, and central access is required a femoral venous line should be inserted by a senior ICU doctor or cardiac anaesthetist. Internal jugular lines should be avoided wherever possible. Heparin (10u/kg/hr) should be routinely infused to central venous lines.

   *Arterial lines (RCH)*
   Where possible, aim for arterial access in the right radial artery. Avoid multiple unsuccessful attempts before referral to the cardiac surgery for cut-down. A right radial arterial line will also provide optimal monitoring during the intra-operative period of isolated cerebral perfusion. Bear in mind variations in arch anatomy in particular aberrant origin of the right subclavian artery.

3. Management of a low systemic cardiac output
*Four distinct causes should be sought & systematically excluded; echo is essential to guide management:*
   i. Excessive pulmonary blood flow (high saturations)
      • Control ventilation and modify pulmonary vascular resistance. Ensure that infant is adequately filled. Consider careful introduction of a low dose vasodilator (sodium nitroprusside 0.5-1mcg/kg/min).
   ii. Impaired systemic ventricular function +/- significant tricuspid regurgitation (normal or low sats)
       • Consider dobutamine +/- vasodilator
   iii. Restrictive PFO (usually low sats)
       • Needs intervention.
   iv. Restrictive duct (any sats)
4. Intubation (at referral hospital / RCH).

**Indications:**
- Apnoeas,
- Shock, severe circulatory disturbance
- Pulmonary overcirculation (Saturations >90%) with systemic hypoperfusion (lactic acidosis)

Intubation can cause considerable instability in infants with a duct-dependent systemic circulation. Elective intubation should be performed by a senior neonatologist / intensivist, using the guideline below.

- **Drugs**
  - Start low-dose dobutamine infusion (up to 5mcg/kg/min) prior to intubation. Pancuronium 0.1mcg/kg then Fentanyl 5mcg/kg (give muscle relaxant first to avoid chest wall rigidity & bradycardias which may be associated with intravenous fentanyl)
  - Volume for bolus infusion should be ready to administer

**Target blood gases:**
- pH – normal
- PaO$_2$ – 35-45 mmHg
- PaCO$_2$ – 35-45 mmHg

Management of excessive pulmonary blood flow and inadequate systemic perfusion at RCH

Intubate as above
Manage pulmonary vascular resistance:
- PEEP 8-10 cmH$_2$O
- Ventilate to allow mild respiratory acidosis (PaCO$_2$ 50s, pH>7.3) 
- FiO$_2$ 0.21. may be incrementally reduced to minimum 0.18 using additional N$_2$.

Inotropes – use dobutamine 5-10 mcg/kg/min
Vasodilators – add sodium nitroprusside if blood pressure tolerates Early surgery

---

**INTRA-OPERATIVE MANAGEMENT**

1. **Intubation** – as above

2. **Central Venous access:**
   - Where possible, existing central lines (UV/femoral) will be used for induction of anaesthesia and pre-bypass management. Otherwise, a single lumen jugular line will be inserted for this, and removed as early as possible
   - A direct atrial line will be left in situ by the cardiac surgeon.

3. **Arterial access** – as above

4. **Drug therapy:**
   - **Vasodilators:**
   - **Nitrates:**
     - Sodium nitroprusside rather than GTN, as it is more effective arterial dilator. This may be used in preference to phentolamine as its effects can be rapidly reversed by discontinuing the infusion.
   - **Inotropes:**
Dobutamine should be routinely used on weaning from bypass. Noradrenaline – consider low-dose noradrenaline in the event of excessive systemic vasodilation on separation from bypass.
POST-OPERATIVE MANAGEMENT

1. Drug Therapy
   **Vasodilators**: as above initially. Captopril should be considered once enteral feeds have been established.
   **Inotropes**:
   Dobutamine: escalation with low dose adrenaline IF poor contractility on echo.
   Noradrenaline should be weaned early.
   Milrinone: if additional inotrope required and tolerating vasodilation. 0.5mcg/kg/min.
   **Anticoagulation and platelet drugs**:
   - Heparin 10 units/Kg/hour started immediately on return from theatre.
     - If the patient is bleeding ask the surgeon to confirm the timing of starting heparin.
     - A different dose may be specifically requested by the surgeon. Document the request, the rationale and any anticoagulation target
   - APTT checked daily. day one, day two
   - Do not chase APTT target (unless specifically requested to), but decrease heparin dose if APTT>80s.
   - >3.5mm shunt: Aspirin (3-5mg/kg/dose daily) when feeding.
   - 3.0mm shunt: aspirin (3-5mg/kg/dose daily) and clopidogrel (0.2mg/kg daily) when feeding.
   - Can consider stopping heparin once commenced on aspirin.

2. Central Lines
   Neck lines or femoral venous lines should be removed as early as possible. Systemic heparin (10u/kg/hr) should be given early after surgery, if no major bleeding.

3. Principles of management of ventilation and circulation:
   *These are detailed in the existing ICU protocol*

4. Echos **On ICU or Koala**
   All infants should have at least weekly echos (on a specified day of each week) after the Norwood operation looking for the following:
   - Patency of BT shunt / Vmax across RV-PA conduit and branch PA flow
   - Function of systemic RV
   - Degree of Tricuspid Regurgitation
   - Adequacy of ASD
   - Neo-aortic obstruction & arch obstruction
   - Patency of the DKS anastomosis
   - Thrombus (intracardiac, SVC)
   - Effusions

5. Line insertion for extubated, non-sedated patients
   The same guidelines should be followed as for pre-op patients, to minimise the risk of sudden decompensation due to changes in systemic vascular resistance.

6. Long term Central venous Access
   Broviac lines (single lumen, 3F) should be considered early in infants likely to require longer term inotropes or parenteral nutrition.
This document has been read, and approved by the following department heads:

Dr Johnny Millar, Director of Cardiac Intensive Care, RCH
Dr Michael Cheung, Director of Cardiology, RCH
Dr Christian Brizard, Director of Cardiac Surgery, RCH
Dr Ian McKenzie, Director of Anaesthesia, RCH
Dr Michael Stewart, Director of PIPER, RCH
Dr Elske Posma, Head of Fetal Medicine Unit, RWH
Dr Carl Kuschel, Head of Neonatology, RWH
Dr Ricardo Palma-Dias, Director of Ultrasound Services, RWH