

## **EARLY MANAGEMENT OF INFANTS WITH HYPOPLASTIC LEFT HEART SYNDROME**

Infants with hypoplastic left heart syndrome are amongst the most challenging subgroup 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.

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### 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, Mercy Hospital for Women or Monash Medical Centre.
2. Fetal echo scans will be reviewed by RCH cardiologists at 18, 30 and 36 weeks' gestation to confirm diagnosis, 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, in general be scheduled for around 35 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.
4. For each patient, a joint meeting will be arranged together with RCH care manager(s), a social worker, the consultant cardiologist and the consultant cardiac surgeon. This meeting will be arranged by the Fetal Management Unit (for Royal Women's Hospital patients), or by the referring cardiologist (for Mercy Hospital / Monash Medical Centre patients). 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 e.g 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 and Cardiac surgical 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 7 West AUM, by RCH Cardiology / Cardiac Surgery staff.

### IMMEDIATE POST-NATAL CARE AT REFERRING HOSPITAL

1. Birth suite management will be directly supervised by a Neonatal Consultant or Fellow. The baby will be transferred to the local NICU for ongoing stabilization once initial resuscitation has been completed.
2. Intravenous access – a triple lumen UV line should be placed before transport (or two separate peripheral lines if a UV line cannot be inserted).
3. 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.
4. Prostin delivery (10-20ng/kg/min) should be commenced immediately after obtaining first route of intravenous access.
5. Referring neonatologist will contact NETS to arrange transfer to RCH.
6. **Target oxygen saturations 75-85%; avoid additional oxygen unless sat < 70%.**

7. At least 1 hour's observation period for prostin-related apnoeas / other instability prior to transfer.
8. Ventilation should not be routine in the stable, non-acidotic patient.
9. Blood gas (arterial or umb venous, not capillary), glucose, lactate prior to transfer.
10. NETS team to inform on-call cardiologist + ICU of birth and patient condition, and likely time of transfer to RCH.
11. All infants will be transferred to RCH PICU in the first instance.

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

This may be suspected from in utero imaging; alternatively should be suspected in the infant with a known diagnosis of HLHS in whom there is severe metabolic acidosis, with poor oxygenation, and Xray 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.

Where suspected prenatally, a Consultant Neonatologist should directly supervise delivery room management of these infants. 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 *with supplemental oxygen therapy*, 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 available to transport them to RCH immediately after birth.

The on-call consultant cardiologist should be present to do an echo immediately on arrival at RCH. Cath lab and cardiac surgery should be on standby for immediate intervention.

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

1. All infants will initially be admitted to PICU.
2. Cardiology fellow and consultant, and 7West AUM or care manager, should all be informed when infant arrives on PICU. An echo should be performed within 2 hours of arrival (with the exception of infants with restriction of PFO). CXR & ECG in all.
3. *Bloods*  
On arrival, routine bloods – FBC, Gas + lactate, electrolytes, glucose, FBC, Coags, chromosomes, Group & Save.
4. Stable patients who are not acidotic, with a normal lactate, with secure central or peripheral intravenous access (two), may be transferred to 7West after review by the ICU consultant.
5. *Surgery*  
This will, where possible, be planned for day 3 or 4 of life, but earlier if there is circulatory instability –particularly when this related to excessive pulmonary blood flow.
6. *Other Health Professionals:*  
Families will meet with Care Managers and social workers after admission, either on 7West or in ICU.
7. *Associated abnormalities*  
Routine pre-operative ultrasound scans of head + kidneys.

## **OTHER ASPECTS OF PRE-OPERATIVE MANAGEMENT:**

### **1. Feeds**

Infants with HLHS are at high risk of gut ischaemia, and our current approach is not to feed them. Infants arriving at RCH within the first 24 hours of life, who have CV access should be given TPN until surgery.

Those without TPN should receive 10% dextrose / 0.45% saline as maintenance fluid.

### **2. Vascular access**

Insertion of even peripheral drips can 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 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 this being done.

Access should be attempted by a senior ICU doctor (senior registrar or consultant), in the treatment room on 7West or in intensive care.

#### *Central lines*

Central venous access should not be attempted in a non-anaesthetised patient.

If umbilical venous access is not present, and CV access required (for inotropes etc) then Femoral venous lines should be inserted, under anaesthesia, by a senior ICU doctor or cardiac anaesthetist. Neck 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 access in the right upper limb. Avoid multiple unsuccessful attempts before referring to the consultant cardiac surgeon re: cut-down Right arm will also provide optimal monitoring during intra-operative period of isolated cerebral perfusion..

### **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. At RCH – ensure that infant is adequately filled, and 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 ASD (usually low sats) → Needs intervention.
- iv. Restrictive duct (any sats) → Increase prostin.

### **4. Intubation (at referral hospital / RCH).**

#### *Indications:*

- i. Apnoeas,
- ii. Shock, severe circulatory disturbance

- iii. Pulmonary overcirculation (Saturations >90%) with systemic hypoperfusion (lactic acidosis)

Intubation can cause considerable instability to infants with a duct-dependent systemic circulation. Intubation should ideally be performed by a senior neonatologist / intensivist; using the guidelines outlined below.

❑ *Drugs*

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)

- ❑ Colloid (4% albumin) or saline should be available (20ml/kg)  
 ❑ Consider low-dose dobutamine infusion (up to 5mcg/kg/min) prior to intubation.

Target blood gases:

- ❑ pH – normal  
 ❑ PaO<sub>2</sub> – 35-45  
 ❑ PaCO<sub>2</sub> – 35-45  
 ❑ AT RCH: *Consider* additional nitrogen to reduce FiO<sub>2</sub> to minimum 0.18, only in the infant *with pulmonary overcirculation and evidence of systemic hypoperfusion* after appropriate circulatory resuscitation, with or without an intravenous systemic dilator. However, this is only a temporising measure, and infants with these clinical signs warrant urgent 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, 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 at surgery

**3. Arterial access** – as above

**4. Drug therapy:**

Vasodilators:

*α blockade:*

Phentolamine may be used intra-operatively, in favour of phenoxybenzamine which is much longer-acting.

*Nitrates:*

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, with the introduction of phenoxybenzamine when adequate haemodynamic stability. Captopril should be introduced once enteral feeds have been established.

Inotropes:

Dobutamine; escalation with low dose adrenaline IF poor contractility on echo. Noradrenaline should be weaned early.

### 2. Central 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 7West*

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
- ❑ Function of systemic RV
- ❑ Degree of Tricuspid Regurgitation
- ❑ Adequacy of ASD
- ❑ Neo-aortic obstruction & arch obstruction
- ❑ 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  
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