SINGLE VENTRICLE PATHWAY

For the Perinatal Management of Newborns with Prenatally Diagnosed Duct Dependent Single Ventricle Circulation.

1. Introduction and who this guideline applies to:

This document provides the overview for the pathway of patients born with duct dependent single ventricle physiology diagnosed antenatally, with emphasis at perinatal and immediate postnatal care recommendations for teams involved – neonatal and cardiac (including cardiologist, PICU, cardiac surgeon). This pathway covers roles, responsibilities and routine tasks of the pathway within the normal working practice of the referring NICU, accepting PICU, EMCHC and CenTre transport team.

This guideline is only applicable for term babies NOT for preterm babies and babies <2 kg although the principles apply.

These infants have heart conditions dependent not only on complete mixing of the systemic and pulmonary venous return, but with a functionally univentricular heart and effectively a single outlet from the heart; ventricular output is divided between the pulmonary and systemic arterial circuits via the arterial duct, depending on the size of the duct and the relative resistance to flow into the two circuits.

There will be some single ventricle patient that might not be duct dependent, hence unless specified by the cardiologist the above guideline applies.

2. Guideline Standards and Procedures:

ANATOMY

- Variations of Single left ventricle (e.g. Pulmonary atresia with intact septum, some with Tricuspid atresia, DILV, or unbalanced AVSD)

- Variations of Single right ventricle, e.g. HLHS, some unbalanced AVSDs

- Indeterminate single ventricle with either aortic or pulmonary atresia

PHYSIOLOGY varies based on duct dependency and the degree of restriction of pulmonary venous return at the atrial septum

A. Duct dependent Systemic blood flow

- Prostin necessary to maintain ductal patency and systemic blood flow
- Potential for a number of complicating factors of which the commonest is restriction of flow across the interatrial septum, this is uncommon and usually (but not always) anticipatable from the late prenatal imaging. Severely affected babies may profoundly desaturate after delivery, or be 'impossible to oxygenate'.

B. **Duct dependent pulmonary blood flow**

- Cyanosis is dependent on size of PDA and additional source(s) of pulmonary blood flow.
- May (rarely) be complicated by RV dependent coronary circulation.

**Not definitely duct dependent.**

It is not always possible prenatally to be certain that the systemic or indeed pulmonary circulation will be secure as the fetal circulation changes to the postnatal state. The change in afterload and increase in pulmonary venous return may alter the configuration of the flap valve of the foramen ovale to change interatrial flow. As the duct closes and the PVR drops there is also the potential for more severe right (or indeed left) ventricular outflow obstruction to be unmasked. Therefore some fetuses may be labeled as possibly duct dependent and either started prophylactically on prostin until can be assessed in more detail, or (often if a local baby) to be scheduled for an early scan and ‘observed’ ductal closure with a low threshold for starting prostin if clinical concerns.

The goal of initial stabilisation and preoperative care is to maintain the infant with balanced blood flow between the systemic and pulmonary vascular bed with optimal oxygen delivery and blood flow.

If this is not achieved, patients may become destabilized, with either:

1. Progressively increasing pulmonary blood flow, resulting in congestive heart failure and inadequate systemic blood flow.
2. Late diagnosis - cardiogenic shock (ductal constriction, pulmonary over circulation) or
3. Inadequate pulmonary blood flow - hypoxemia

A number of ventilatory and pharmacologic maneuvers may be used to “balance” the circulation attempting to optimise systemic blood flow and oxygen delivery; however, it only temporises the need for surgical intervention.

**ANTE-NATAL CARE:**

There will be a detailed written plan for location, timing and mode of delivery for these patients, agreed between the fetal cardiologist, the responsible obstetric and neonatal consultants and the family, which will be kept in the MOTHER’S own notes, (their hand held notes, and can be find in a computer system Heartsuite) and will be copied to all involved clinicians.

Occasionally parents will have opted in advance for ‘comfort care’, without resuscitation after delivery. The details of individual management of these infants will have been agreed in detail.
with the neonatal teams but a flexible approach must be maintained as circumstances can change post-delivery.

PERI-NATAL CARE:

The attending Obstetrician has the primary responsibility for determining the timing of delivery. Normal vaginal delivery following spontaneous labour at term is the goal for otherwise uncomplicated pregnancies. When labour is induced, the timing will be influenced by consideration of clinical and social factors as well as the availability of a neonatal stabilisation cot, EMCHC beds and (occasionally) cardiac surgical resources. Please bear in mind that delivery may occur up to 72 hours after induction and therefore ‘bed planning’ is frequently not either feasible or accurate. It is therefore critical that all parties are informed of any necessary alterations to the agreed plans.

1. Inform the On-Call Consultant Cardiologist or the Cardiac Registrar as soon as possible.
2. Refer to the antenatal Cardiologist’s letters, about the immediate management, and if necessary, seek clarification from them directly.
3. Cardiac team to communicate to Cardiac Surgeon, Paediatric Intensivist and Ward / PICU Manager to confirm the availability of ward or intensive care beds as anticipated to be required (PICU capacity is not always a requirement).
4. All further management and changes to be discussed with Cardiologist +/- Paediatric Intensivist.

POST-NATAL CARE AT REFERRING HOSPITAL

1. Delivery suite management will be directly supervised by a senior member of neonatal team. The baby will be transferred to their local NICU for on-going stabilization once initial resuscitation has been completed.
2. Intravenous access – a triple (preferred) or double 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 inotropes.
4. Dinoprostone (Prostaglandin E2) should be commenced immediately after obtaining first route of intravenous access. (Usually start at 3 - 5 nanogram/kg/min; range 5 - 50 nanogram/kg/min; above 15 nanogram/kg/min please discuss with the cardiologist). Alprostadil (Prostin VR) can be used in the same dosage as above.
5. Once the neonate is accepted to Glenfield Hospital, referring neonatologist will contact Centre Transport to arrange transfer (in the event of poor mixing, urgent transfer to Glenfield may be required – see Management of restrictive PFO).
6. Target oxygen saturations 75-85%; avoid additional oxygen unless sats < 65%.
7. At least 1 hour’s observation period for prostin-related apnoeas / other instability prior to transfer.
8. Avoid excessive interventions; invasive ventilation should not be routine in a stable patient - spontaneous ventilation is preferred.

9. Blood gas (arterial or umbilical venous, not capillary), glucose, lactate prior to transfer.

**Indications for intubation**

1. Apnoeas

2. Shock, severe circulatory disturbance

3. Pulmonary over circulation (Saturations >90%) with systemic hypoperfusion (lactic acidosis); NOTE this usually not present at birth; it evolves over time with the natural drop of PVR postnatally.

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.

1. Induction Drugs as per the local policy

2. Volume should be available (usually 0.9% Sodium chloride 10ml/kg)

3. Consider supporting circulation prior to intubation (low dose adrenaline or dobutamine infusion). If patient is compromised please do not delay intubation.

**Management of a low systemic cardiac output**

Four distinct causes should be sought & systematically excluded; echo is essential to guide management:

1. Impaired systemic ventricular function with/without significant tricuspid regurgitation (normal or low sats) → Consider dobutamine or low dose Adrenaline

2. Excessive pulmonary blood flow (high saturations) WITH systemic hypoperfusion - clinical signs: elevated arterial sats, poor perfusion, wide pulse pressure, oliguria, lactic acidosis, low SvO2

   2.1. Maximising cardiac output is the most important step - ensure adequate preload and support cardiac function, optimise Hb (give volume, low dose adrenaline or dobutamine, RBC transfusion);

   2.2. Further adjustments done by manipulation Qs - afterload reduction - reduce stress & pain, hence oxygen consumption (deep sedation, minimal handling, keep warm, Milrinone, SNP);

3. Maximising PVR is of limited use, supplemental CO2 or N2 are obsolete - ventilate with PEEP, titrate rate and other settings to target higher CO2 levels 5 - 7 kPa (mild hypercarbia increases cardiac output), titrate oxygen to achieve sats 75-85%, strictly avoid alkalosis;(7,8,9) Restrictive ASD (usually low sats and difficulties with ventilation) → Needs intervention
4. Restrictive duct (saturations not a good guide as vary with the anatomy) → Increase prostin

Management of the infant with suspected or confirmed restriction of the PFO (Subset with a particularly poor prognosis)

Confirmed Antenatal Diagnosis - Please confirm plan made in the antenatal letter and documented in Maternal notes regarding parents' wishes. They may have chosen comfort care only without intervention.

Should be suspected in the infant with a known antenatal diagnosis of HLHS in whom there is severe metabolic acidosis. Prognosis for this particular set of patients with hypoplastic left heart is very poor and these infants require early surgical or emergency transcatheter intervention to enlarge the restrictive communication.

Where suspected prenatally, a clear delivery plan will be in place and these babies will if at all possible, be delivered at the Leicester Royal Infirmary or possibly even at Glenfield.

A Consultant Neonatologist should directly supervise delivery room management of these infants. It may also be appropriate for a consultant cardiologist / cardiology team to be in attendance. In any case there should be a detailed pre-delivery plan agreed between the Cardiologist, Neonatologist and Paediatric Intensivists, as well as interventional cardiologists as required.

If the clinical findings immediately after birth are consistent with HLHS with importantly restrictive mixing at the foramen ovale, the baby should be intubated, commenced on prostin, muscle relaxed and sedated (as above), and transferred with supplemental oxygen therapy, immediately to PICU at EMCHC.

Other standard resuscitative measures are generally ineffective in this situation, so for these infants, the neonatal transport team should be available to transport to Glenfield immediately after birth.

The on-call consultant cardiologist should be present to do an echo immediately on arrival at EMCHC (if not at delivery) and liaise with the MDT to decide on further management.

PRE-OPERATIVE CARE AT GLENFIELD (IF UNOBSSTRUCTED ATRIAL COMMUNICATION)

- Echo by Paediatric cardiologist and CXR and ECG
- Monitor the balance of parallel circulations; continuous monitoring and repeated clinical examination of perfusion is critical (concern if HR >160/min and RR >60/min); a trend in cerebral NIRS might be helpful (9); biochemical parameters are late indicators of systemic hypoperfusion (SvO2 > lactate > organ dysfunction)
- If stable these babies may well be managed on ward 30 rather than PICU. However, especially babies at risk of congestive heart failure or pulmonary over-circulation with systemic hypoperfusion as PVR falls and Qp/Qs rises, should have close observation and regular blood gas estimation, with a low threshold to keep on PICU / admit there if concerns.
- 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 doctor (registrar or consultant), if on ward 30 - in the treatment room on ward 30.

- 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.

- Arterial lines

Where possible, aim for access in the right radial. If one can't access right radial then try different sites. Avoid multiple unsuccessful attempts. Do not attempt a right brachial artery line; leave it for anaesthetist for intra-operative monitoring (during any anticipated period of isolated cerebral perfusion).

- Bloods

On arrival, routine bloods – FBC, Gas + lactate, electrolytes, glucose, FBC, Coagulation, chromosomes, Group & Save.

- Associated abnormalities

Routine pre-operative ultrasound scans of head + kidneys are needed only in patients with postnatally diagnosed cardiac problems or if there are clinical concerns.

- Feeds

Insert NGT to aspirate gastric contents prior to transfer.

Commence feeds in neonates with Duct dependent pulmonary blood flow with caution as high risk of NEC (refer to Feeding UHL Childrens Intensive Care Guideline C90/2016).

If enteral feeding cannot be established, TPN via UVC/PICC or central line is indicated.

### 3. Education and Training

Training and raising awareness are on-going processes. On-going awareness is promoted through the induction and continuous bedside teaching. Training is provided for medical staff during lunchtime teaching (Wednesdays) and other sessions, and at junior doctors’ induction training. Nursing education is supported by the Practice Development teams, and nursing educators.

### 4. Monitoring Compliance

<table>
<thead>
<tr>
<th>What will be measured to monitor compliance</th>
<th>How will compliance be monitored</th>
<th>Monitoring Lead</th>
<th>Frequency</th>
<th>Reporting arrangements</th>
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<tbody>
<tr>
<td>Treatment algorithm followed and documented</td>
<td>audit</td>
<td>PICU consultant</td>
<td>As required</td>
<td>Clinical Practice Meeting</td>
</tr>
</tbody>
</table>
5. Supporting References


2) http://www.signavita.com/articles/review-articles/100-preoperative-management-of-hypoplastic-left-heart-syndrome

3) http://www.rch.org.au/uploadedFiles/Main/Content/picu/HLHS.pdf


5) Reducing the incidence of necrotizing enterocolitis in neonates with hypoplastic left heart syndrome with the introduction of an enteral feed protocol. Del Castillo SL1, McCulley ME, Pediatr Crit Care Med. 2010 May;11(3):373-7


6. Key Words

Single ventricle, single ventricle physiology, Hypoplastic left heart syndrome, HLHS, Prostin, restrictive PFO, Univentricular heart

The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs. As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.
### REVIEW RECORD

**Description Of Changes (If Any)**

*Added:*

- Anatomy and physiology explanation
- Management of low cardiac output syndrome extended
- Preoperative care at GH extended:

  Monitor the balance of parallel circulations; continuous monitoring and repeated clinical examination of perfusion is critical (concern if HR >160/min and RR >60/min); a trend in cerebral NIRS might be helpful (9); biochemical parameters are late indicators of systemic hypoperfusion (SvO₂ > lactate > organ dysfunction)

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