Paediatric Intensive Care Unit

SINGLE VENTRICLE PATHWAY

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

Staff relevant to:	Neonatal, Cardiac, PICU, Obstetric & maternity staff caring for term babies with prenatally diagnosed duct dependant single ventricle circulation
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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 patients that might not be duct dependent, hence unless specified by the cardiologist the above guideline applies.

Contents

2. Guideline Standards and Procedures:

ANATOMY

Variations of Single left ventricle (e.g. Pulmonary atresia with intact septum, some with Tricuspid atresia, Double Inlet Left Ventricle (DILV), or unbalanced AtrioVentricular Septal Defect (AVSD)).
Variations of Single right ventricle, e.g. Hypoplastic Left Heart Syndrome (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 Patent Ductus Arteriosus (PDA) and additional source(s) of pulmonary blood flow.
- May (rarely) be complicated by Right Ventricle (RV) dependent coronary circulation.

c) **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)

3. Inadequate pulmonary blood flow – hypoxemia

A number of ventilatory and pharmacologic manoeuvres 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 found 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. Consider using advanced care planning documentation such as the RESPECT form where applicable. 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. It must be kept in mind that morbidity and mortality of neonates with complex congenital heart disease born at 39-40 weeks is lower compared to preterm. 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. Inform NNU nurse in charge about delivery plans and to check cot availability.
3. Refer to the antenatal Cardiologist’s letters, about the immediate management, and if necessary, seek clarification from them directly.
4. 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).
5. All further management and changes to be discussed with Cardiologist +/-Paediatric Intensivist.

POST-NATAL CARE AT REFERRING HOSPITAL

Babies with single ventricle will need to be delivered in a tertiary neonatal unit (QMC, LRI)

1. Delivery suite management will be directly supervised by a senior member of neonatal team. The baby will be transferred to the neonatal unit for on-going stabilization once initial resuscitation has been completed.

2. Intravenous access (by a senior clinician)– double lumen Umbilical Vein (UV) line should be placed before (two separate peripheral lines only if a UV line cannot be inserted).

3. Umbilical arterial (UA) 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 by the PICU team for transfer to EMCHC. If born outside of LRI neonatal services, then the referring neonatologist will contact Centre Transport to arrange transfer (in the event of poor mixing, urgent transfer to EMCHC may be required – see Management of restrictive Patent Foramen Ovale (PFO)).

6. Target oxygen saturations 75-85%; avoid additional oxygen unless Sats < 65%,please see below.

7. Avoid excessive interventions; invasive ventilation should not be routine in a stable patient - spontaneous ventilation is preferred.

8. In unventilated babies, they should have at least 1 hour's observation period for prostin-related apnoeas / other instability prior to transfer.

9. Blood gas (ideally arterial or umbilical venous,capillary not ideal but can be used), glucose, lactate prior to transfer

Indications for intubation

1. Apnoeas

2. Shock, severe circulatory disturbance

3. Evidence of systemic hypoperfusion (significant or progressive lactic acidosis);

NOTE this usually not present at birth; it evolves over time with the natural drop of PVR and evolving over-circulation with high saturations

Intubation can cause considerable instability to infants with a duct-dependent systemic circulation. Intubation should ideally be performed by a senior clinician; 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 in discussion with cardiac PICU and oncall cardiologist:

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.

    I. 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);
    II. Further adjustments done by manipulation Qs - afterload reduction - reduce stress & pain, hence oxygen consumption (deep sedation, minimal handling, keep warm, Milrinone under guidance if no previous experience with its use);

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)
4. Restrictive ASD (usually low sats and difficulties with ventilation) → Needs intervention (namely Atrial septostomy)
5. Restrictive duct (saturations not a good guide as vary with the anatomy) → Increase prostin (Serial monitoring with Echocardiogram with dose change)

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-focussed management plan without survival focussed 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.

A Senior paediatric/neonatal clinician with experience of newborn stabilisation 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 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 to maintain saturations around 75-85%, 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 EMCHC 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 (IF UNOBSTRUCTED 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 the paediatric cardiology ward 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 the paediatric cardiology ward - in the treatment room.

- Central lines

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 unable to access the 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 NasoGastric Tube to aspirate gastric contents prior to transfer.
Commence feeds in neonates with Duct dependent pulmonary blood flow with caution as high risk of NEC. Enteral feeds to be considered only with good evidence of gut (systemic) perfusion. (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

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Treatment algorithm followed and documented	audit	PICU consultant	As required	Clinical Practice Meeting

5. Supporting References

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6. Key Words

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

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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.

Contact and review details	
Guideline Lead (Name and Title) EMCHC CHD Clinical Practice Group Lead Dr Suhair Shebani	**Executive Lead:** Chief medical officer
REVIEW RECORD	
Description Of Changes (If Any) Ante-natal care section revised and updated: - Added 'Consider advanced care planning documentation such as the RESPECT form where applicable Peri-Natal Care section revised and updated: - Added 'It must be kept in mind that morbidity and mortality of neonates with complex congenital heart disease born at 39-40 weeks is lower compared to preterm. - Added '2. Inform NNU nurse in charge about delivery plans and to check cot availability' Post-Natal care revised and updated: 1. 'Local NICU' changed to 'the neonatal unit' 2. Specifies two separate peripheral lines only if a UV line cannot be inserted 8. Specifices unventilated babies to have at least 1hrs observation period for prostin related apnoeas/other instability prior to transfer 9. Changed to include capillary blood gas whilst acknowledging it is not ideal Indications for intubation revised & updated: 3. Changed to 'Evidence of systemic hypoperfusion' Management of low systemic cardiac output: Milrinone – added 'under guidance if no previous experience with its use 3. Intervention for Restrictive ASD specified as Atrial Septostomy 4. Addition of (serial monitoring with echocardiogram with dose change) Management of the infant with suspected or confirmed restrictrion of PFO : Changed from Consultant Neonatologist to senior paediatric/neonatal clinician with experience of newborn stabilisation should directly supervise delivery room management Pre-operative care: Addition of Enteral feeds to be considered only with good evidence of gut (systemic) perfusion.	