Paediatric Intensive Care Unit

Hypoplastic Left Heart Syndrome

Staff relevant to:	PICU
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Contents

1. Introduction and Who Guideline applies to

This guideline primarily concentrates on univentricular physiology of Hypoplastic Left Heart Syndrome (HLHS) on PICU.

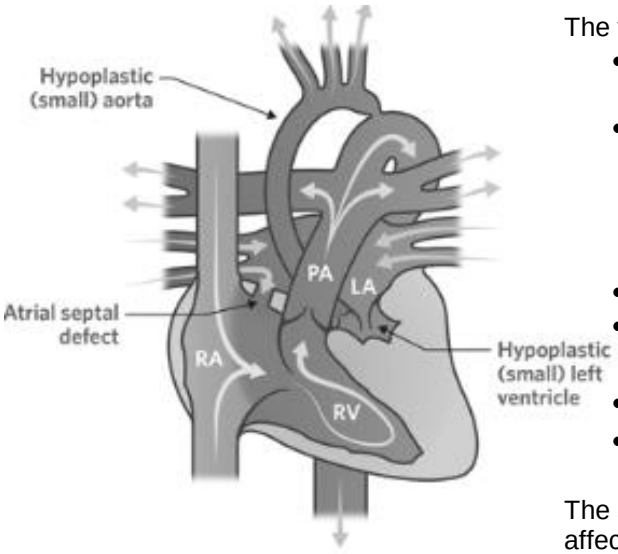
For the perinatal management of newborns with prenatally diagnosed duct dependent single ventricle circulation, please refer to Single Ventricle UHL Childrens Medical Guideline

Related documents:
Blalock-Taussig (BT) Shunt or Central Shunt UHL Paediatric Intensive Care Guideline

The term hypoplastic left heart syndrome, initially proposed by Noonan and Nadas in 1958, describes a spectrum of congenital cardiac abnormalities characterized by a functional single right ventricle with marked hypoplasia of the left ventricle and some degree of hypoplasia of the ascending aorta or arch. The aortic and mitral valves are atretic, hypoplastic, or stenotic. The right ventricle provides systemic blood flow by a right-to-left shunt via a patent ductus arteriosus. A wide-open ductus arteriosus and complete mixing of the venous returns at the atrial level are essential for postnatal survival.

HLHS represents 2–9% of congenital heart disease cases and accounts for 23% of neonatal deaths from congenital heart malformations and remain, despite the vast progress in HLHS treatment over the recent years, the highest risk and costliest group of lesions among congenital heart defects. HLHS is more common in males than in females, with a 55-70% male predominance.

2. Guideline Standards and Procedures



The term HLHS includes:

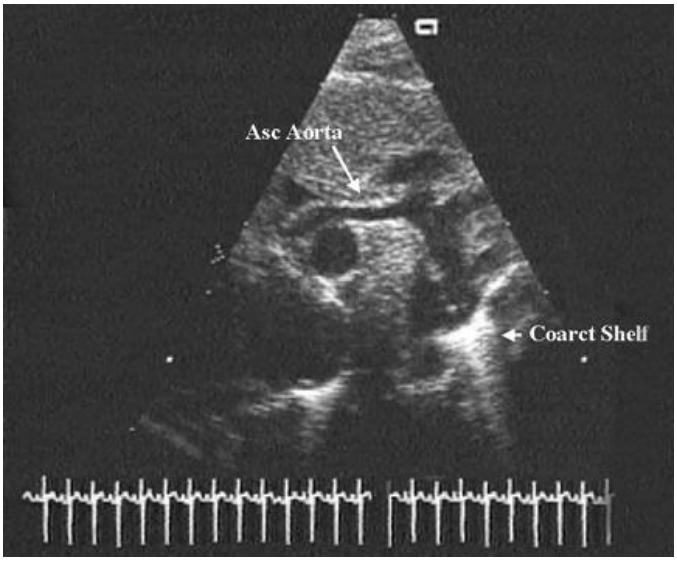
- Hypoplastic or small left ventricle, usually not forming the apex of the heart.
- Aorta and/or mitral hypoplasia or atresia.
    - 1. Mitral atresia (MA)/aortic atresia (AA)
    - 2. Mitral stenosis (MS)/aortic atresia (AA)
    - 3. Mitral stenosis (MS)/aortic stenosis (AS)
    - 4. MA/AS + ventricular septal defect (VSD)
- AA/MA or AA/MS represent the most severe form of HLHS
- AA/MS can be associated with ventriculo-coronary connections (sinusoids), which adversely affect outcomes and survival
- The ventricular septum is usually intact
- Coarctation of the aorta is also commonly present

The specific subtype of HLHS and individual patient factors markedly affect surgical planning (type of Norwood procedure vs hybrid procedure with stenting of the patent ductus arteriosus and placement of pulmonary artery bands).

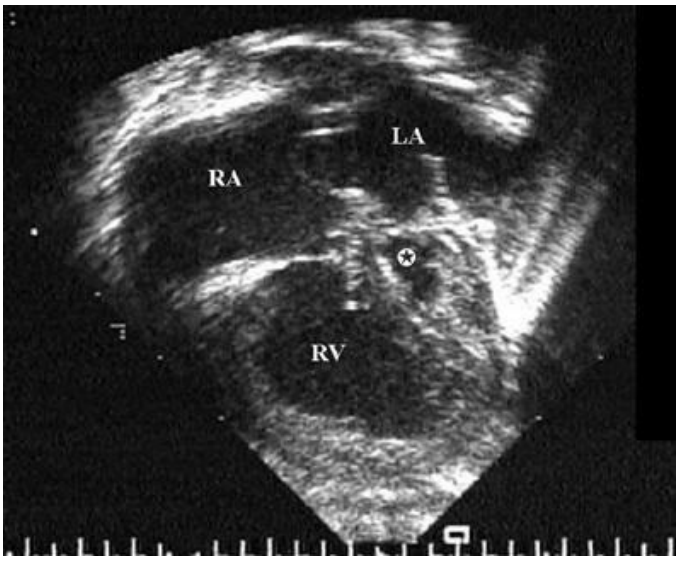
The postnatal circulation in hypoplastic left heart syndrome depends on a delicate balance between the pulmonary and systemic blood flow in order to ensure adequate oxygenation and tissue perfusion. The systemic blood flow is dependent on the overall cardiac output from the functional single ventricle and the relative pulmonary and systemic vascular resistances. Blood flow is inversely proportional to resistance (Ohm's law); thus whenever the resistance in blood vessels decreases, blood flow through those vessels increases. Following birth, as pulmonary vascular resistance decreases, a higher percentage of the fixed right ventricular output flows to the lungs instead of to the systemic circulation. This results in higher oxygen saturation on the cost of poor perfusion and metabolic acidosis and oliguria.

Coronary artery and cerebral perfusion also depends on blood flow through the ductus arteriosus and then retrograde aortic arch and ascending aortic flow. Therefore, increased pulmonary blood flow results in decreased flow to the coronary arteries and brain, with a risk of myocardial or cerebral ischemia.

It is vital that the pulmonary venous return is unobstructed by an unrestricted septum. The presence of significant obstruction to pulmonary venous return from a restrictive atrial septum may cause pulmonary venous & arterial hypertension and pulmonary oedema & hypoxia with systemic arterial saturations below 75-80%. In severe cases there is in utero lung damage and lymphangiectasia. These infants do not respond to oxygen and require prompt evaluation via echocardiography and discussion as to whether urgent relief of obstruction by either percutaneous balloon septostomy or operative atrial septectomy is appropriate. Venoarterial extracorporeal membrane oxygenation may be acutely lifesaving but given the very poor prognosis for recovery, is not always appropriate.



Long-axis view of the aortic arch in a patient with HLHS. The ascending aorta is markedly hypoplastic, serving only to deliver blood in a retrograde fashion to the coronary arteries. An echo-bright coarctation shelf is seen at the insertion of the ductus arteriosus



4-chamber view of the heart in a patient with HLHS. A large right ventricle (RV) and hypoplastic left ventricle (star) are seen.

Preoperative management:

Preoperative management of patients with HLHS aims to balance parallel circulations and maintain an optimal equilibrium between oxygen supply and demand. One should aim for the lowest effective HR and SVR to optimize diastolic filling time, with SaO2 maintained between 75% and 85% at the lowest tolerated FiO2.

Management strategies are aimed at addressing the 3 major causes of desaturation:

- Diminished pulmonary blood flow due to intact or highly restrictive atrial septum
- Low mixed venous oxygen saturation due to high Qp:Qs ratio attributing to low cardiac output
- Pulmonary venous desaturation (assuming a non-restrictive atrial septum) due to increased PVR related to pulmonary infection or pulmonary venous obstruction

Stable neonate

- **Avoid excessive interventions (**minimal handling in a tranquil environment)
- Monitor RR and pattern, pre- and post-ductal pulse oximetry, continuous ECG and intermittent non-invasive blood pressure (BP) in the right arm (term neonates > systolic 65 and diastolic >35 mmHg measured in the upper body)
- Aim for spontaneous ventilation preferably in room air, accept 30-60/min, use non-invasive ventilatory support to reduce work of breathing and oxygen consumption, apply oxygen to maintain saturations in 75-85% pre-ductal
- Aim for normal pH & avoid alkalosis
- Continue Prostin @ 5-10ng/kg/min to maintain ductal patency, based on echo findings and systemic perfusion. Use the lowest effective prostaglandin dose to minimize dose-dependent side effects (hypotension and respiratory depression)
- The ECG may be normal (120-160bpm), with the typical neonatal findings of right-axis deviation and right ventricular hypertrophy, but in some cases T-wave inversion appears across all precordial leads.
- A complete echocardiogram to define cardiac anatomy
    - HLHS subtype
    - Ventricular function
    - Size of the interatrial communication and indications for a septostomy
    - Status of the patent ductus arteriosus
    - The presence and severity of tricuspid regurgitation
- Monitor clinical signs of perfusion, urine output and lactate & cerebral Near-infrared spectroscopy (NIRS)
- Accept Saturations in the 90's in room air if adequate urine output and low lactate. Watch closely for signs of systemic hypoperfusion (SvO2 >lactate>organ perfusion) as these neonates are in high risk for pulmonary overcirculation and need surgical repair sooner than later.
- Monitor fluid balance & administer fluids without restriction, according to standard neonatal recommendations
- Keep Hb 14-16 g/dl, increased concentration of haemoglobin decreases the ratio of pulmonary flow to systemic flow by increasing PVR
- Provision of nutrition to the neonate with HLHS is a controversial; EBM may be started if there is adequate flow in descending aorta (Doppler), monitor abdominal distension and stools. TPN would be an alternative if surgery needs to be delayed.
- Avoid central lines (other than umbilical) unless the patient is becoming unstable

Unstable neonate

- Patients with severe metabolic acidosis and cardiogenic shock require intubation and ventilation to reduce O2 consumption and to decrease afterload, which subsequently increases cardiac output.
- The PGE1 infusion should be started at a higher dose 20-50ng/kg/min to ensure duct patency
- Use catecholamines as a rescue therapy (with high-dose PGE1), aim for normal BP (systolic >65 and diastolic >35 mmHg) to support adequate perfusion of the coronary and cerebral circulation; ensure adequate preload.
- Coronary artery perfusion depends on blood flow through the duct. This is true for AA-MA, AS-MA, AA-MS. But if there is forward flow then depends more on antegrade flow.

- Place appropriate central venous and arterial line
- Adequate sedation and analgesia with morphine supplemented by regular paracetamol, consider alpha-2 agonists such as dexmedetomidine or clonidine.
- Monitor SvO2 & lactate & NIRS
- Management of a low Qp:Qs ratio
    - Systemic vasoconstriction with vasopressin/ or Noradrenaline
    - Check for blood flow through ductus arteriosus and take measures to decrease PVR
    - Improve functional residual capacity with adequate positive end-expiratory pressure (PEEP) and manipulating the PVR by using oxygen, inhaled nitric oxide, alkalosis and aggressive sedation
    - If there is evidence of low Qs with cyanosis and refractory hypoxaemia, despite adequate flow through ductus arteriosus, the patient should be reassessed for consideration of emergency atrial septostomy. Because due to restrictive ASD -> increased LA pressure-> increased PV pressure-> high PVR-> increased PA pressure low Qp.
- Management of a high Qp:Qs ratio
    - Milrinone to reduce afterload & increase cardiac output (be cautious as Milrinone may also reduce PVR with a potential undesired risk of increasing the Qp:Qs ratio in excess of total CO. Milrinone also uncovers any lack of volume , insufficient preload by hypotension which can drop to dangerous level, below perfusion pressure, which can worsen cardiac failure due to coronary hypoperfusion)
    - Add Adrenaline to increase contractility to augment CO
    - Keep ical>1.2
    - Aim Hb >14g/dl (increases PVR),

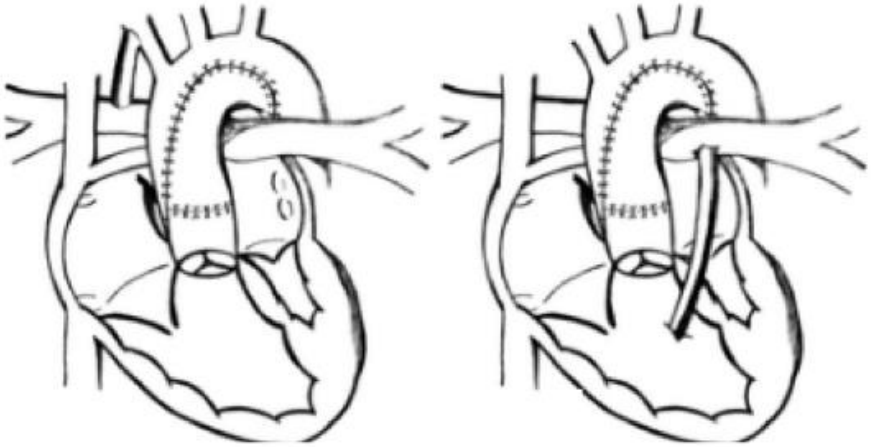
Surgical Palliation

HLHS is a uniformly lethal cardiac abnormality if not surgically addressed. The surgical palliation was first described by Norwood in the early 1980s. Multi-stage palliation is the current management strategy for the treatment of children with HLHS. In HLHS the single ventricle must be connected to both the systemic and pulmonary circulations. The Norwood stage 1 procedure is usually carried out around day 2-7 of age when pulmonary vascular resistance has dropped sufficiently. In patients who present with comorbidities or in LCOS, this may be delayed.

- Prognostic factors for a successful repair are:
    - Ventricular function: poor RV function is a predictor of mortality throughout surgical palliation
    - Tricuspid valve function: significant TR is a poor prognostic factor
    - Size of branch pulmonary arteries: Well-developed PAs, free of obstruction, are essential for an effective SV circulation
    - Pulmonary valve (PV) function: The PV (neo-Aortic Valve) should be competent and non-stenotic. Significant stenosis and/or regurgitation (but not a bicuspid PV alone) may preclude a Norwood operation
    - Size of ascending aorta: size is of theoretical importance due to the retrograde nature of coronary perfusion before and after Norwood procedure.

- Stage 1 Norwood procedure involves:
    - Atrial septectomy – allowing unobstructed pulmonary and systemic venous return and complete mixing in common atrium.
    - Aortic arch reconstruction – allowing unobstructed, low resistance, systemic outflow. The main pulmonary artery is separated from the pulmonary artery branches and connected to the ascending aorta. The remainder of the aorta is reconstructed using homograft material. Blood is now pumped from the single right ventricle out the “neo-aorta” to the systemic circulation.
    - Pulmonary circulation -since the pulmonary artery is now committed to the systemic circulation, a systemic to pulmonary shunt or Sano shunt provides a source of pulmonary blood flow. The Sano shunt is a Gore-Tex conduit that connects the lungs to the single ventricle via an incision made in the anterior wall of the right ventricle. The most common conduit size used is 5 mm for patients between 2.5 and 4 kg. The Sano shunt is increasingly used due to its better early haemodynamic stability; it eliminates the risk of diastolic run-off from the systemic (and coronary) circulations. The need for ventriculotomy associated with the Sano shunt is a theoretical concern due to the potential for late ventricular dysfunction and dysrhythmias, and there remains controversy over which type of shunt produces better long term pulmonary arterial ;growth’.
    - The alternative to a Sano shunt; the BT (Blalock-Tassig) shunt, is usually a 3.5 - 4mm synthetic tube from a systemic artery (typically the right subclavian) to the pulmonary artery. These infants have parallel circulations connected at the arterial level, likely contributing to a better RPA growth versus the Sano shunt.
    - Retrospective data comparing the two types of shunt suggest that early (<30 days) unfavourable events (death, acute shunt failure, cardiac arrest, ECMO, unplanned cardiovascular reoperation or NEC) may be less common with the Sano Shunt than the BT shunt; in 1 series [Ohye et al] 10% of infants in the Sano conduit group versus 14% in the modified BT shunt group. From postoperative day 30 to Glenn completion, events occurred in 12% in Sano shunt group versus 22% in BT shunt group respectively. Fewer patients in the Sano shunt group required early Glenn procedure for hypoxaemia after the Norwood procedure, reflecting the less vulnerable physiology in Sano shunt group.

- The chest is routinely left splinted open for first 3 –5 days post Norwood, reflecting the complexity of the surgery and the potential for haemodynamic instability in the early postoperative period. Data from the multicentre study from the Society of Thoracic Surgeons’ database shows that the chest was left open in 60% of cases [Hornik et al.].
- The general aim of the 1st stage is to achieve the following four objectives:
    - Unobstructed systemic cardiac output
    - Controlled source of pulmonary blood flow
    - Reliable source of coronary blood flow
    - Unobstructed egress of pulmonary venous return across the atrial septum
- Good development of the PAs in HLHS (central and peripheral) is crucial for the future effectiveness of the Fontan circulation.



BT shunt, Subclavian artery to PA

Sano shunt, RV to PA Conduit

Post-Operative Management Strategies

Cardiac output Monitoring

Cardiorespiratory monitor
CVP
SaO2
End-tidal CO2
Cerebral/Renal NIRS
Core/toe temperatures
SvO2

Lines and Tubes ·
Central venous catheter
Arterial line
PICC line
Foley catheter
Chest drains
NG tube
Atrial and ventricular pacing wires

Assessment

Vitals per PICU routine, Interval physical exam
Hourly urine output
Hourly chest tube output
Chest X-ray-admission
Daily ECG, Atrial wire study as necessary
Daily assessment of possibility for chest closure admission
Vascular US as needed if:
. Platelet count decreasing
. High volume chest tube output
. Signs of CVL malfunction

Labs on admission:

FBC, U&E, Mg, Phos, Clotting (consider TEG if bleeding), SVO2 if IJ present: ABG, lactate, iCal, glucose

Every 2 hours for 8 hours: SVO2, ABG, lactate, iCal, glucose. Then consider increasing interval, but not less frequently than every 6 hours

Daily at 0500: FBC, U&E, Mg, Clotting albumin, ABG, lactate, iCal, glucose

A baseline TEG (thromboelastograph test of clotting) will be ideal if the drain loss is sanguineous and excessive (>4ml/kg/h).

BT Shunt Considerations:

Goal diastolic BP>35mmHg (coronary perfusion)
Avoid hyper-oxygenation
Avoid hyperventilation and alkalosis
Blender on resuscitation bag to 50%

Please refer to Blalock-Taussig (BT) Shunt or Central Shunt UHL Paediatric Intensive Care Guideline

Sano Shunt Considerations:

Goal diastolic BP>35mmHg (coronary perfusion)
Avoid hyper-oxygenation
Avoid hyperventilation and alkalosis
Blender on resuscitation bag to 50%

- **Intervention by system**

Respiratory:

- Mechanical ventilation-SIMV/PC, Target: PaCo2 4.5-5.5 kPa, PaO2 5.3-6 kPa/pH 7.3-7.4, ventilate in air with Saturation range 75-85%
- If SaO2 < 75%or paO2<5.3 kPa: CXR to rule out pulmonary venous desaturation due to a malpositioned endotracheal tube, pulmonary consolidation, pleural effusion/haemothorax or pneumothorax, based on findings +/- ↑PEEP, ↑FiO2, +/- initiate iNO. If more profound hypoxaemia (SaO2 < 70%), urgent Echo to rule out anatomic obstruction to pulmonary blood flow **such as** blocked, compressed or kinked BTS or Sano shunt, and adequacy of septectomy. This scenario requires emergency investigation and treatment to increase the shunt perfusion pressure.
- If SaO2 >85%: decrease FiO2 to 0.21, consider increasing Milrinone to reduce afterload

Cardiovascular:

- Common atrial pressure (cardiac preload) should be maintained at an adequate level (aim CVP around 10) with judicious volume administration (boluses of 5–10 ml/kg), tailored on a case by case basis considering specific factors such as degree of TR
- Milrinone infusion at 0.25 to 0.75 mg/kg/min to reduce systemic vascular resistance. A dose at the lower end of the range should be considered as the starting point early after Norwood procedure, because of reduced renal clearance of the drug in this setting
- Low dose Adrenaline infusion 0.02 to 0.05 mcg/kg/min to support the myocardial contractility of the single ventricle and to maintain a MAP (typically > 40 mmHg) to promote diuresis. Try not to use high doses of Adrenaline if possible because of the increased myocardial oxygen consumption and associated tachycardia
- Noradrenalin or Vasopressin if diastolic pressures consistently below 25mmHg. It is important to maintain adequate diastolic BP for adequate coronary perfusion and adequate filling pressures. Patients with modified BTS have greater systolic and diastolic flow into the Qp and therefore may have diastolic hypotension, requiring the addition of a noradrenaline infusion.
- Calcium infusion is beneficial to increase contractility without further provoking tachycardia Target iCal 1.2-1.3mmol/L
- Early echo to assess for unobstructed Tricuspid valve and outlet flows to and from the single ventricle. Rule out residual anatomic lesions.
- Maintain Hb around 14 g/dl or HCT 0.4 to maintain adequate oxygen carrying capacity
- For signs and symptoms of Low Cardiac Output, please follow LCOS algorithm below

Fluids, Electrolytes and Nutrition, please follow postop management guidelines:

- Enteral feeds should be started by nasogastric tube when there is reasonable CO, follow high risk feeding protocol
- GI prophylaxis- standard IV dosing. Patient should be given GI prophylaxis until they are at 50% of their goal enteral feeds
- Fluid overload and acute kidney injury are relatively common after Norwood procedure, due to LCOS. In patients with preserved renal function, start low dose furosemide infusion at 0.2

mg/kg/hr around 12 hours post op to avoid excessive fluid overload. Patients with fluid overload who do not respond to diuretics and those with more severe acute kidney injury should be managed with dialysis.

- Consider TPN if not tolerating feeds and concerns with distended abdomen

Haemostasis:

- Primary objective: Normalize coagulation profile to reduce bleeding
- Normalize coagulation profile to reduce bleeding, target normal parameters for: INR, PTT, Fibrinogen, Platelet count, and HCT
- Monitor chest drain output. High volume chest tube output or tension of the membrane covering the open chest. Bleeding is considered to be severe when drain/measured losses are sanguineous and exceed:
    - 4ml/kg/hr in the first hour or tension of the membrane covering the open chest
    - 2ml/kg in the second hour
    - 1ml/kg in subsequent hours or any bleeding below this threshold associated with haemodynamic instability
    - A baseline TEG (thromboelastograph) will be ideal if the drain loss more than expected and FFP, Cryo & Platelets administered accordingly. Ideally, if the bleeding is excessive (>4ml/kg/h), the drain losses should be evaluated every 15 minutes.
    - Notify surgeon for bleeding > 4mL/kg/hr for 2 consecutive hours
    - If haemostasis achieved, begin low dose heparin infusion (10 units/kg/h) after approximately 4 hours for shunt/line prophylaxis. Convert to aspirin 5mg/kg when tolerating enteral feeds.
- No need to hold prophylactic heparin prior to chest closure. Stopping therapeutic heparin prior to chest closure is discussed with surgeons and usually held 2h before chest closure.

Neurology:

- All opiate analgesics, the majority of sedatives and all muscle relaxing agents have a negative inotropic effect, at the same time adequate pain control decreases oxygen demand, and minimises need for additional sedatives. Fentanyl infusions are more appropriate for some patients (those with pulmonary Hypertension)
- Morphine infusion to titrate per ICU comfort protocol supplemented by regular paracetamol. Spikes of lactate during cares and handling may suggest inadequate pain control.
- Muscle relaxant: - Patients who are less stable or who have an open sternum may require muscle relaxants to minimise their oxygen demand. Normal default is Atracurium to run at 10-40 mcg/kg/min - dilute in sodium chloride 0.9%. As Atracurium causes histamine release, some prefer to use Rocuronium instead.
- Consider clonidine or dexmedetomidine infusion if no rhythm concerns.

Infectious Disease:

- Antibiotic prophylaxis (ensure timing related to most recent dose in operating room)
- 1. Surgical antibiotic prophylaxis: in case of open chest to be continued 24 hours after chest closure
- Nystatin drops 100 000 units (= 1ml) 6 hourly orally to stop when extubated & lines out – continue if on antibiotics.

Lines

- Usually managed with femoral central lines – if internal jugular central line present, try and remove as soon as patient stable in view of importance to prevent any upper body veins occlusion or narrowing which can affect future palliative pathway; ideally keep flushed with low dose heparin infusion till in situ.

Low cardiac out algorithm: cardiogenic shock

Commonly after Norwood palliation, the nadir for Cardiac Output occurs up to 12 hrs after cardio pulmonary bypass (CPB); therefore patients should be monitored very closely until this period has passed. Monitoring includes a combination of clinical and laboratory parameters. It is important to identify any existing or anticipated problems (bleeding, pulmonary hypertension crisis, arrhythmia). Remember the desaturation may be the result of systemic venous desaturation (SvO2) or pulmonary venous desaturation (SpvO2) or decreased pulmonary blood flow.

- LCOS Definition: inadequate systemic O2 delivery, low mixed venous oxygen saturation with SvO2 difference greater than 25%
- Clinical picture: Tachycardia, hypotension, desaturation, mottling, oliguria and lactic acidosis, elevated arterial oxygen saturation with poor perfusion, a wide pulse pressure, pulmonary oedema, and high atrial pressure. Additionally, diminished right ventricular function may be evident on echocardiography
- Concerns: High lactate not responding to management, increasing inotropic support, persistently low mixed venous oxygen saturations & low Systemic NIRS readings are warning signs.
- Common causes:
    - Anaemia/Haemorrhage
    - Ventricular dysfunction
    - Residual cardiac lesion
    - Arrhythmia
    - SIRS/Infection
    - Pericardial tamponade
    - Pneumothorax

Goal: to achieve normal systemic oxygen delivery. This requires that the Pulmonary to systemic blood flow ratio (Qp:Qs= (SaO2 – SvO2) / (SpvO2 – SpaO2) is close to 1. SvO2 to SaO2 difference of ~25% suggests adequate O2 delivery, the O2 amount that the peripheral tissues will extract for their aerobic metabolism. Based on the Fick principle, the amount of oxygen that the tissue consumes (VO2 = (C0 x Ca)-(CO x Cv)) is the difference between the amount of O2 in the arteries and the amount of O2 in the veins. Ca is O2 content in arterial blood, versus Cv is the O2 content in the mixed venous blood. So if the demand increases, the SvO2 to SaO2 difference will increase. In patients with low cardiac output post stage 1 procedure the oxygen extraction may be much higher and the monitoring of mixed venous SmvO2 gives an accurate reflection of the adequacy of systemic perfusion. Thus, a Low cardiac output state is the result of increased consumption & decreased O2 delivery, causing a decrease in SmvO2.

Management strategies:

- Urgent echo: exclude anatomic lesions including
    - Obstruction to pulmonary venous return
    - Flow through the neo-aorta / aortic arch,
    - Shunt flow

- RV function and right atrio ventricular valve regurgitation (AVVR)

- Minimize oxygen consumption. Consider aggressive sedation, paralysis and cooling (35 C) to reduce metabolic demands if on higher dose inotropic support. Central temperature should be monitored.
- Increasing CO is the most effective way of improving oxygen delivery
    - Adequate O2 carrying capacity, Keep Hb around 14g/dl
    - Ensuring A-V synchrony
    - Reduce SVR with Milrinone up to 0.75 mcg/kg/min (remember Milrinone accumulates in oliguria and has a half-life of 3-4 hrs)
    - Modest inotropic support with Adrenaline & add calcium chloride infusion (keep ical>1.2) to optimize CO
    - If hypotension triggers the use of escalating catecholamine doses, ECMO should be considered
- Ensure adequate pulmonary gas exchange if hypoxaemic due to atelectasis.
    - Optimize lung recruitment with adequate positive end-expiratory pressure of 6-8 to prevent pulmonary venous desaturation
    - Optimize ratios of Qp:Qs by manipulation of PVR with Oxygen and consider NO
- Consider Hydrocortisone in systemic inflammatory response secondary to the deleterious effects of cardiopulmonary bypass with
- Cardiac arrest, which is usually a complication of LCOS, is more common after Norwood procedure than after most other operations, aim for early discussion with Surgeon and ECMO team about ECMO initiation to rest the heart and the lungs.

Factors contributing to hypoxaemia and ventilator dependence after stage 1 palliation

Factor	Possible cause	Diagnostic measures
Pulmonary venous desaturation	• Ventilation/perfusion mismatch: malpositioned endotracheal tube, pleural effusion, haemothorax, pneumothorax, chylothorax, pulmonary oedema, atelectasis, pneumonia, pneumononitis and AV malformation	• Chest radiogram • Lung ultrasound • Bubble echocardiography/angio-CT/cardiac catheter for arteriovenous malformation
Systems venous desaturation	• Low Do_2: anaemia, LCOS, impaired coronary perfusion, ventricular dysfunction, RT or TS, neo AR or AS, CoA, pericardial effusion/tamponade, arrhythmia • Increased VO_2: sepsis, hyperthermia, pain, agitation, seizures	• NIRS, SVO_2 • Haemoglobin • Temperature • ECG, atrial EC, Holter monitor • ECHO • Cardiac catheter
Inadequate Qp	• Low SVR (in patient with MBTS): sepsis, hyperthermia, systemic vasodilators • High OVR: lung disease, pulmonary, vascular disease, pulmonary vein stenosis, r-FO/IAS, TR, TS, RV, arch obstruction • Obstruction shunt-conduit stenosis • PA branches: stenosis, kinking, tenting, thrombosis, thromboembolism • Small MBTS or RV-PA conduit	• Echocardiogram • Cardiac catheter, angio-CT • Anatomical evaluation of MBTS, RV-PA conduit, PC branches, pulmonary veins • PVR study

Signs of LCOS in patient's status-post Norwood procedure

Note: all signs may not be present in all patients; numeric values are guidelines and must be considered in the clinical context.

- Tachycardia (HR > 160 bpm)
- SaO2-SvO2 > 30
- Urine output < 1mL/kg/hr
- Hypotension (SBP < 55 mmHg, MAP < 40 mmHg)
- Serum lactate > 2 mmol/L
- Cerebral or renal oximetry by NIRS < 40%
- Cool or mottling of extremities (Toe temp < 28 degrees)

Evaluate PB

SBP > 75 mmHg (MAP > 55 mmHg)

- Consider weaning vasoconstrictors
- Ensure sedation and analgesia are adequate
- Consider increasing milrinone

Evaluate Heart rate

HR < 130

- Evaluate for dysrhythmia
- Consider atrial or atrioventricular sequential pacing
- Wean excessive sedation

HR > 160

- Primary fever: Antipyretics, cool to low-normal temperature.
- If peripheral vasoconstriction, efforts should be focused on decreasing afterload and improving cardiac output.
- Evaluate and treat for dysrhythmia
- If SBP > 65 mmHg & DBP> 35 mmHg) consider weaning chronotropic infusions
- Consider chest X-ray and ECHO to rule out intrathoracic air/ fluid collection

Evaluate SaO2

SaO2 > 85%: try to optimize balance of Qp: Qs by:

- Decrease FiO2 as tolerated to 0.21
- Consider echocardiogram to assess:
  a. Pericardial effusion
  b. Ventricular function
  c. AV valve regurgitation
  d. RV-PA conduit/ PA stenosis
  e. Residual systemic outflow/arch obstruction
- If elevated BP, consider increasing milrinone, consider SNP??? & titrate to target goal

Evaluate CVP

CVP ≤ _8 mmHg

- Consider fluid administration (10 mL/kg). Repeat up to 2 times (e.g. total 30 mL/kg) as needed, and then consider increasing Adrenaline infusion
- Reassessment of CVP

CVP > 8 mmHg

- Consider an initial fluid bolus (10 mL/kg), and reassess hemodynamics. If no improvement and CVP remains > 8 mmHg, consider increasing Adrenaline infusion

No Improvement despite Adrenaline?

- Consider additional vasopressor (vasopressin or Noradrenaline)
- Consider muscle relaxation
- Consider initiating hydrocortisone 1 mg/kg/dose IV every 6 hours
- Transfuse pRBC's to maintain Hb @ 14
- Consider additional fluid boluses especially if CVP remains < 8 mmHg Consider echocardiogram to assess:
  a. Pericardial effusion
  b. Ventricular function
  c. AV valve regurgitation
  d. RV-PA conduit/ PA stenosis
  e. Residual systemic outflow/arch obstruction
- Maintain ical >1.2

Important points for chest closure management

- Therapeutic heparin is witheld 2 hours prior to chest closure if surgeons agree. Prophylactic heparin do not to be witheld prior to chest closure
- At time of chest closure, be aware of desaturation and hypotension from compression of shunt
- May need to reopen chest
- Gently increase PIP (usually by 10- 20%) to maintain inspired tidal volume as chest compliance usually reduced following sternal closure
- Avoid overly negative fluid balance and assess fluid status frequently
- Restart heparin one hour post chest closure

Prognosis & major complications

Overall survival to the time of hospital discharge after the Norwood procedure is approximately 75%. Success rates are higher (85%) in patients with low preoperative risk and lower (45%) in patients with important risk factors. Survival after the bidirectional Glenn/hemi-Fontan and Fontan operations is nearly 90-95%. The actuarial survival rate after staged reconstruction is 70% at 5 years. Institutional success rates vary.
Important preoperative risk factors are:

- Prematurity, birth during the early term period of 37 to 38 weeks' gestation is associated with worse outcome
- Birth weight <2.5kg
- Significant preoperative tricuspid insufficiency
- Pulmonary venous hypertension
- Associated major chromosomal or non-cardiac abnormalities
- Preoperative mechanical ventilatory or circulatory support

Note the following:

- Poor RV function is a predictor of mortality throughout surgical palliation
- Low cerebral near-infrared spectroscopy oxygen saturations during the first 48 hours after Norwood procedure are strongly associated with adverse outcomes
- The need for mechanical preoperative ventilation has been shown to be a risk factor for poor outcome
- Severe preoperative lactic acidosis, and a need for inotropic support, which further increases the risk for right ventricular dysfunction and tricuspid regurgitation are important risk factors for survival of stage I Norwood palliation
- Neurodevelopmental prognosis is not known; however, differences are well described.

-Major complications following the Norwood procedure include aortic arch obstruction at the site of surgical anastomosis and progressive cyanosis caused by limited blood flow through the shunt. An inadequate atrial communication contributes to progressive cyanosis.

Transfer to ward criteria

- SaO2 > 75% with stable respiratory status for >24 hours
- Stable haemodynamic for >24 hours without need for vasoactive infusions
- Tolerating intermittent diuretics
- Tolerated 24 hours of feeding protocol

3. Education and Training

None

4. Monitoring Compliance

None identified at present

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements

5. Supporting References

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6. Key Words

Congenital cardiac abnormalities, Norwood procedure, Ventricular

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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 & review details	
Guideline Lead (Name and Title) Samira Neshat – PICU Consultant	**Executive Lead** **Chief Medical Officer**
Details of Changes made during review: **New guideline**	

Appendix 1 Post –Operative Norwood/Sano Management Algorithm

Note: all signs may not be present in all patients; numeric values are guidelines and must be considered in the clinical context.

Cardiac output monitoring

- Cardiorespiratory monitor: CVP
- SaO_2 End-tidal CO_2
- Cerebral/Renal NIRS
- Core/toe temperatures, SvO_2

Goal: adequate CO

- Sats 75%-85% avoid hyperoxygenation
- HR 130-160 bpm
- CVP 8-12
- BP 65/35 mmHg, map >40mmHg
- Ph 7.35-7.45, pCO_2 4.5-5.5
- Blender on resuscitation bag to 50%

Evaluate for Pulmonary Venous Desaturation

- ABG/MVO_2/Chest x-ray
- Adequate chest rise? Tidal volume? ET CO_2 trend?
- Suction patient (after bolus sedation)
- O_2 challenge, if 10% increase in saturations, then lung disease is present, increase MAP + judicious increase in airway pressure to treat V/Q mismatch

Evaluate for Low Pulmonary Blood Flow

- Minimal response to O_2 challenge
- +/- hypoperfused chest x-ray/ET CO_2
- Usually normal BP

Evaluate for Low SvO_2

- Low BP or evidence of compromised oxygen delivery, consumption or not optimised ventilation
- Haemoglobin <14g/dl?

Evaluate PVR

- At risk: if high sats pre-op, <38 wks, lung disease, restrictive ASD, pulmonary vein obstruction
- Start INO, alkalosis, sedation and paralysis, aim $PaCO_2$ 4.5, increase FiO_2, increase milrinone

No improvement

Small, Kinked or Thrombosed Shunt

- Shunt murmur?
- Echo, contractility? Valvar competency? Already on adrenalin low dose? iCal?
- Increase BP to drive shunt flow (Vasopressin or Noradrenaline)
- Start Heparin

Title: Hypoplastic Left Heart Syndrome
V: 1 Approved by: PICU Clinical Practice Group: April 2021 Next Review: April 2024
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