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Great Ormond Street Hospital for Children



NHS Trust

CVS: ICU Management of HLHS and Single Ventricle Physiology

Allan Goldman, April 2006

Updated Adrian Plunkett, February 2007, Timothy Thiruchelvam, May 2009

Associated clinical guidelines/protocols:

- Cardiac ICU Manual

Fundamental Knowledge:

List of topics relevant to PIC that will have been covered in membership examinations. They will not be repeated here.

Information for Year 1 ITU Training (basic):

Year 1 ITU curriculum

- HLHS and Single Ventricle Physiology
- Post op management of HLHS, Glenn and Fontan

Curriculum Notes for Year 1:

Understanding surgical principles and management of children with single ventricle physiology including:

- The Norwood operation (stage 1)
- Bidirectional Glenn (stage 2 Norwood operation)
- Fontan operation (stage 3 Norwood operation)

Introduction

Physiology of the functionally univentricular heart

This term applies to a variety of congenital cardiac anomalies where there is only one ventricle pumping in parallel to both the systemic and pulmonary circulations. These patients are often sensitive and respond to common interventions such as supplemental oxygen, ventilation, and vasoactive drugs in a more radical fashion than patients with conventional two circulations. These patients also undergo multiple operations and are more vulnerable to intercurrent infections and illnesses.

Univentricular Physiology due to Systemic Outflow Obstruction

The focus of this module is on univentricular physiology secondary to congenital lesions causing left ventricular outflow tract obstruction (LVOTO). This encompasses a heterogeneous group of conditions including HLHS, critical coarctation, interrupted arch, critical AS, and variants of DILV.

The key features are:

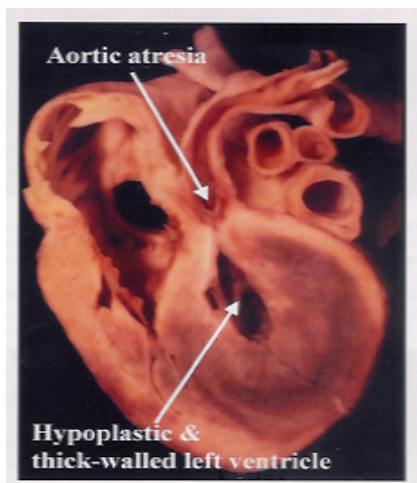
- There is complete mixing of the venous returns
- Systemic blood flow is provided by a right-to-left shunt via the PDA or a systemic to pulmonary shunt.

Thus with an unrestricted ASD and PDA, the systemic blood flow is dependent on the overall cardiac output from the functional single ventricle and the relative pulmonary and systemic vascular resistances (see summary figures at end). It is vital that the pulmonary venous return is unobstructed by a large ASD or there is incomplete mixing and elevated pulmonary venous pressure, pulmonary hypertension and pulmonary oedema.

HYPOPLASTIC LEFT HEART SYNDROME (HLHS)

This is a heterogeneous group of conditions characterised by under-development of the left-sided cardiac structures, resulting in various degrees of obstructed systemic cardiac output. The anatomic manifestations are variable but usually encompass a degree of mitral and aortic stenosis or atresia, poorly developed left ventricle (which may be almost absent) and hypoplastic ascending aorta and aortic arch (see figure 1). The commonest associated cardiac anomaly is aortic coarctation.

Figure 1:



Once considered a uniformly fatal condition; however, the outlook for newborns with HLHS has now been dramatically altered with staged reconstructive procedures.

1.) SUMMARY OF STAGED PALLIATION FOR HLHS:

This is based on the knowledge that an effective circulation is possible with univentricular physiology in the absence of a pulmonary ventricle.

Stage 1: The Norwood procedure provides:

- 1) Unobstructed systemic blood flow
- 2) Adequate coronary blood flow
- 3) Unobstructed pulmonary venous return across the atrial septum
- 4) Sufficient pulmonary blood flow from a systemic to pulmonary shunt without significant volume overload. (See fig. 1)

This leaves the pulmonary and systemic circulations in parallel with the right ventricle needing to perform increased work from a high volume load for both circulations simultaneously. It is in this context that it is imperative to understand the parallel circulation and balancing of the two circulations as described below.

Stage 2: The bi-directional Glenn operation or hemi-Fontan:

The systemic shunt is taken down and SVC anastomosed directly to the pulmonary artery resulting in a reduction of the ventricular volume load imposed by the systemic shunt. (Fig. 2) Important features of the Hemi-Fontan operation are augmentation of the central pulmonary arteries, and construction of a potential connection for the inferior vena cava to the pulmonary arteries making for an easier Stage 3 Norwood or Fontan operation.

Stage 3: The Fontan or Total Cavopulmonary Connection - TCPC procedure:

The inferior vena caval return is also channelled to the pulmonary artery to completely separate the pulmonary and systemic circulations and place them in series instead of parallel. (Fig. 3) This further reduces the volume load on the single ventricle. In this operation, a fenestration is often carried out between the IVC and the common atrium (particularly in high risk patients with borderline high pulmonary vascular resistance) in order to maintain systemic output in the face of a raised PVR. The price one pays for this may be desaturation.

Fig 2: Norwood Stage 1 (conventional method)

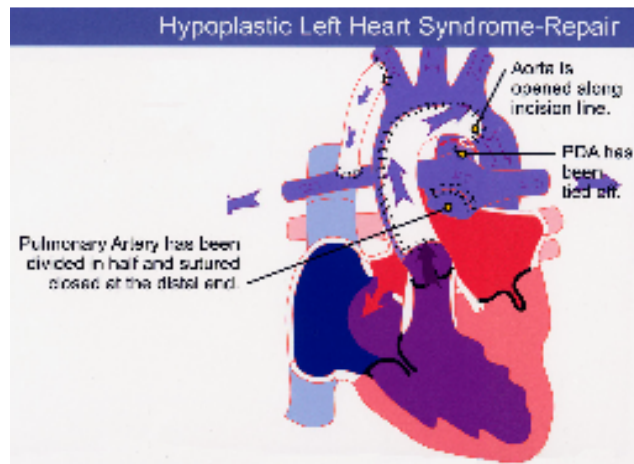


Fig3:Norwood stage2– Bidirectional Glenn

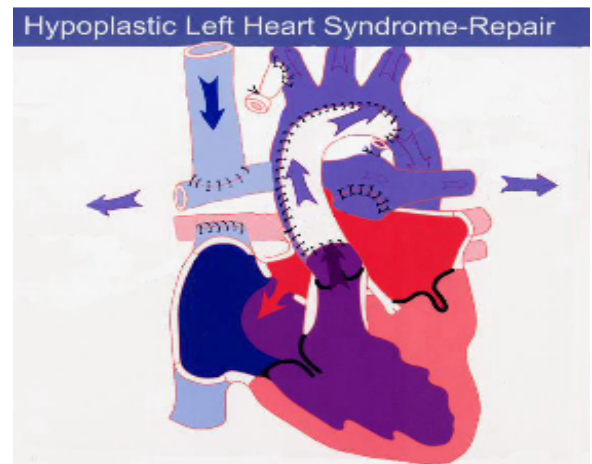
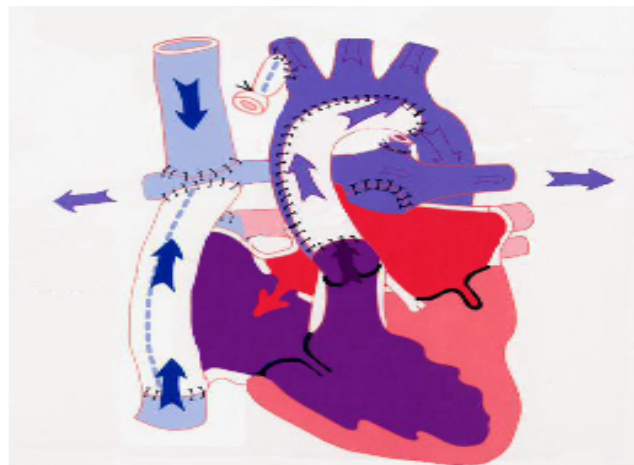


Figure 4: Norwood stage 3 - Fontan or TCPC



Refinements in operative technique and perioperative management have resulted in improved outcome in this high-risk group of patients. In fact, survival has now been reported in excess of 90% for the stage I Norwood operation for low risk patients with HLHS. Adverse survival in this group has most strongly been associated with significant non-cardiac congenital anomalies, shocked condition preoperatively, severe preoperative obstruction to pulmonary venous return, prematurity and low birth weight.

Survival of patients after the stage II Norwood with a bidirectional Glenn or hemi Fontan has been reported as high as 97% and 90% after the Fontan operation. The major advance in the treatment of these patients has been in the stage 1 Norwood in the first month of life.

2. KEY FEATURES OF PRE AND POST-OP MANAGEMENT OF THE NEONATE UNDERGOING NORWOOD STAGE 1 FOR HLHS:

General principles:

The focus should be on the use of prostaglandin infusion and reduction of the inspired oxygen concentration to avoid a high pulmonary to systemic blood flow and high systemic arterial saturations. This situation cannot, however, be sustained indefinitely as there will be a progressive deterioration in pulmonary function with increasing pulmonary oedema with time. These patients will therefore undergo a Norwood 1 stage operation from about 3 to 5 days of age.

The presence of significant obstruction to pulmonary venous return (usually from a restrictive ASD) leads to pulmonary venous and arterial hypertension and pulmonary oedema. It is therefore vital to ensure that the ASD is not restrictive. In this situation, the systemic arterial saturations are usually below 75-80%, and urgent intervention is often required to relieve this obstruction (usually with a percutaneous balloon septostomy). This can be difficult as the atrial septum is often thickened. Some centres will opt for operative atrial septectomy in this situation or an earlier Norwood operation.

Pre-operative management:

a) In the delivery suite:

Soon after birth with the first breath, there is a sudden drop in PVR and from this point onwards, the balance of the circulations in neonates with HLHS is governed by the ratio of pulmonary to systemic vascular resistance. Thus, stresses such as cold, which increase SVR, will reduce systemic oxygen delivery. The attending physician in the delivery suite should be aware of these issues and the primary determinant of PVR. $p\text{CO}_2$ should be kept within the **normal** range and oxygen should be given with caution (usually FiO_2 of 0.21 to 0.3) unless the baby has associated PPHN. The use of pulse oximetry is mandatory in this situation and the clinician should aim for saturations in the range of 80-85% in room air, which would correlate with a pulmonary to systemic blood flow ratio of 1.5-2:1.

b) On-going pre-operative management:

- Ideally these babies are managed with spontaneous ventilation in room air. The **focus should be on signs of adequacy of systemic oxygen delivery** such as lactic acidosis, urine output, clinical signs of perfusion. There is less focus now on SaO_2 in isolation and it's now acceptable to have higher SaO_2 in the 90's in room air provided all other clinical and lab signs point to adequate DO_2 .
- In stable patients we do allow for enteral feeds, but with low threshold for TPN if there are any signs of poor perfusion or NEC.

- Neonates with apnoea due to prostaglandin infusion or those with signs of inadequate systemic perfusion are usually ventilated. Echo should guide the need for a septostomy (restrictive ASD) and need for inotropes or inodilator agents. These babies are usually not fed and managed on TPN.
- The [Norwood stage 1 procedure](#) is usually carried out at 3 to 5 days at GOSH if patient stable. In patients who have presented in shock or LCOS this may be delayed until after full neurological assessment including serial EEG's if appropriate.

Operative management:

Stage 1 Norwood operation involves (see figure 2):

1) [Atrial septectomy](#) – allowing unobstructed pulmonary and systemic venous return and complete mixing in a low pressure, physiologically common atrium.

2) [Aortic arch reconstruction](#) – allowing unobstructed, low resistance, systemic outflow

3) [Systemic to pulmonary shunt](#) - to allow adequate but not excessive pulmonary blood flow

These principles are also true of the Sano procedure

Post-op Management:

- *Intra and postoperative echocardiography:* After the Norwood operation, an early echo should be performed (usually in theatre) to assess for the adequacy of the repair, in particular the aortic arch, and any obstruction to pulmonary venous return as well as myocardial function and A-V valve regurgitation.
- The chest is routinely left splinted open for first 3 –5 d post Norwood.
- Patients managed on Morphine infusion and neuromuscular blockers if unstable.
- Initial inodilator therapy is milrinone (0.5 to 0.7 mcg/kg/min) and low dose adrenaline (0.02 to 0.05 mcg/kg/min). Often converted to captopril (particularly if any tricuspid regurgitation) after patient extubated and stable. Milrinone continued until a few days after extubation, based on clinical condition and echo findings.
- Routine peri-op antibiotics
- Maintain Hb at around 14 g/ dl or HCT 0.4
- Not fed enterally for first 3 -5 days until chest closed and stable haemodynamics on low doses of inotropes. Early use of TPN.
- Usually managed with femoral central lines – if neck CVL present, try and remove as soon as patient stable.
- Start low dose heparin (10 u/kg/hr) after approximately 8-12 hours when sure no post-op bleeding. Convert to aspirin when tolerating enteral feeds.
- In unstable patients, urgent echo, consider paralysis and cooling to reduce metabolic demands if other conventional treatments fail including higher dose inodilator agents and balancing circulations.
- At time of chest closure, caution of desaturation from compression of shunt. May need to reopen chest. Remember to gently increase PIP (usually by 10-20%) to maintain inspired tidal volume (usually now measured on modern ventilators) as chest compliance usually reduced following sternal closure.
- If refractory low cardiac output consider T3 and physiological steroids.
- In patients who continue to deteriorate despite above manoeuvres consider mechanical support with ECMO. Must exclude anatomical lesions with echo in this clinical scenario.

SaO₂ falsely estimates Qp:Qs

- Irrespective of the anatomy, univentricular physiology involves mixing of the systemic and pulmonary venous returns, and then partitioning the cardiac output from this single ventricle to the systemic and pulmonary circulations in a parallel fashion. **The ratio of pulmonary blood flow (Qp) to systemic blood flow (Qs) is determined by the resistance in either circulation – blood flow favouring the path of least resistance.**
- **It has previously been accepted** that the SaO₂ reflects the ratios of these flows in the newborn with a duct dependent circulation or as well as in patients who have had a systemic to pulmonary shunt.
- Based on the Fick equation (with some very important assumptions),

$$Qp:Qs = (SaO_2 - SmvO_2) / (SpvO_2 - SaO_2)$$

Assuming the cardiac output is as predicted, assuming systemic and pulmonary arterial oxygen saturations are the same and assuming the lungs are normal and oxygen saturation in the PV (pulmonary vein) is 95% and assuming systemic arterial oxygen saturations (SaO₂) and mixed venous SmvO₂ are normal and that SaO₂-SmvO₂ = 25% then the equation can be further simplified to:

$$Qp / Qs = 25 / (95 - SaO_2)$$

Whilst theoretically this allows one to assess the effects of interventions, it is now well accepted that these assumptions are far too great an oversimplification of the issues particularly with respect to:

1. Presuming difference in arterio-venous oxygen saturation averages 25%: in patients with low cardiac output following the Norwood operation oxygen extraction may be much higher and it is now believed that the monitoring of mixed venous SmvO₂ gives the most accurate reflection of the adequacy of systemic perfusion. Tweddell et al's group, who arguably have the best short-term postoperative outcomes for the Norwood stage 1 operation, have shown that early postoperative goal directed therapy towards maintaining a normal SmvO₂ has had an important role in improving outcome of these patients. This has focused the management of these patients on improving cardiac output through lowering systemic vascular resistance with aggressive vasodilator strategy (see below) and the use of inotropes.
2. Tweed and colleagues have similarly shown that a significant number of patients post Norwood 1 have pulmonary venous desaturation which can result in a gross underestimation of the Qp to Qs ratio.

This has led to many centres to seeking information that confirms metabolic demands are met by systemic oxygen delivery through:

- continuous or intermittent monitoring of mixed venous saturation and/or
- non-invasive regional oximetry through cerebral or somatic near infrared spectroscopy

It remains crucial to appreciate that with univentricular physiology desaturation may be the result of:

1. systemic venous desaturation
2. pulmonary venous desaturation or

3. decreased pulmonary blood flow

3. BETWEEN STAGE 1 AND 2 NORWOOD

Preoperative cardiac catheter is routine required between stage 1 and 2 and 2 and 3 Norwood operations to assess factors that may harm a volume loaded, single, morphologically right ventricle functioning at systemic pressure and hinder safe progression to stage 2 and 3 including: 1) right ventricular function 2) tricuspid regurgitation, 3) residual arch obstruction 4) pulmonary artery anatomy 5) PVR. A tricuspid valvuloplasty is now performed more readily at the stage 2 operation whenever regurgitation of that valve is more than mild.

4. BIDIRECTIONAL CAVOPULMONARY SHUNT

This intermediate step is introduced to decrease the volume load on the single right ventricle until the Fontan procedure can be safely performed. This involves removing the systemic to PA shunt/Sano conduit and connecting the SVC directly to the pulmonary arteries. A prerequisite for successful Glenn is low pulmonary vascular resistance. The IVC drainage continues into the RA where mixing takes place with oxygenated blood returning from the SVC through PA, through LA via the septectomy.

Postoperative Care

Head high, patient at 45-degree angle

Maintain adequate preload

Early extubation to improve cavopulmonary blood flow

Avoid respiratory alkalosis and use mild respiratory acidosis – PaCO₂ around 45 mmHg to improve hypoxaemia.

Inhaled NO has been shown to be unhelpful in the management of hypoxemia post Glenn shunt.

5. TOTAL CAVOPULMONARY CONNECTION

Criteria for Identifying 'Ideal' Candidates for an Extracardiac Fontan

▪ Age	2 years old
▪ Common or systemic AV valve	continent
▪ Lung/Heart Compliance	normal
▪ Pulmonary arteries	absence of congenital or iatrogenic abnormalities of proximal or distal branches
▪ Pulmonary artery resistance	2 Woods Units
▪ Mean pulmonary artery pressure	15–18 mm Hg
▪ Ejection fraction	55%
▪ MAPCAs	absent

Key Points in the Post-op Management after the Fontan Operation

Special issues / risks

- High PVR
- Obstructed venous pathways

- Impaired ventricular function
- Systolic / diastolic
- Inodilators (milrinone)
- AV asynchrony
- AV valve incompetence

Postoperative Monitoring

- SVC, 'pulmonary' arterial pressure
- SVC - LAP = trans pulmonary gradient (**TPG**)

Echocardiogram

- Assess ventricular function
- SVC + IVC to PA connections

Post-operative Care of TCPC

Adequate Central Venous Driving Pressure

- Intravascular volume repletion
- "The Fontan Position" (head up 45 degrees, knees bent to facilitate venous return)

Be aware of effects of positive pressure ventilation:

- Decreases systemic venous return
- Decreases ventricular filling
- Increases PVR
- Early extubation, consider negative pressure ventilation in some patients with LCOS and possibly inhaled nitric oxide if pathophysiology of LCOS is high PVR.

Other postoperative complications after Fontan operation

- **Bleeding**
Redo's / chest drains
- **Effusions**
Chylothorax
Drains should stay for at least 2-3 days

Phrenic nerve palsy

Thrombosis

- Strictly observe the anticoagulation policy
- Heparin to warfarin
- CVL to be removed as soon as possible as at all stages

Conclusion:

Staged reconstruction for HLHS is now a viable treatment option with good intermediate term outcome. This does however remain a palliative treatment strategy and viable alternative techniques at stage one reconstruction gaining popularity. The staged approach may yet be tailored to suit high-risk morphological sub-groups and low birth weight newborns. It remains vital that we get more information on the long-term neurological outcome of these patients.

Information for Year 2 ITU Training (advanced):

Year 2 ITU curriculum

To understand the Post operative management of Norwood Stage 1,
Bidirectional cavopulmonary shunt, Fontan circulation

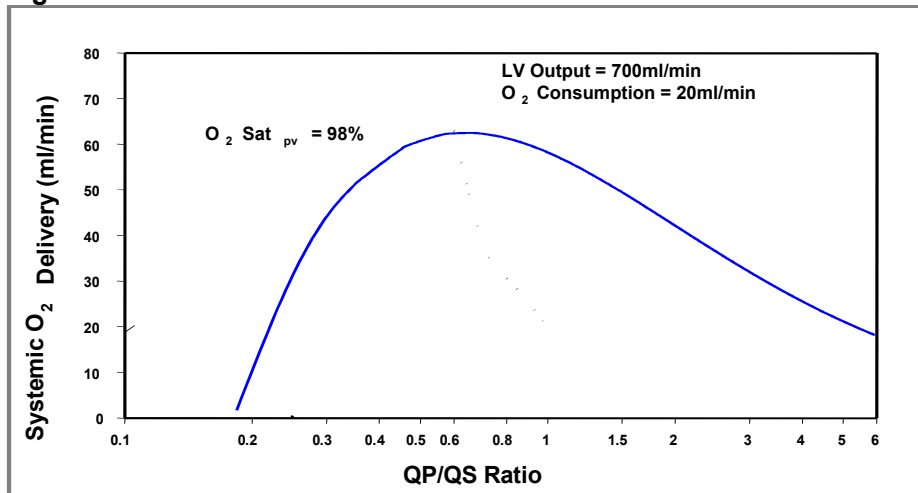
Curriculum Notes for Year 2:

Notes on balancing systemic and pulmonary circulation, *and optimising DO_2* in the immediate postoperative period

In the post-operative Norwood stage 1 patient, the cardiac output from both ventricles is combined and the systemic and pulmonary circulations are in parallel. Therefore, any increase in pulmonary blood flow will result in a reduction in systemic blood flow and systemic oxygen delivery (DO_2).

Figure 5 shows that the maximum DO_2 occurs at a $Q_p:Q_s$ of around 0.5 to 1:

Figure 5:



More importantly, however, is that DO_2 is increased more effectively by augmenting cardiac output as opposed to altering $Q_p:Q_s$ ratios – see figure 6.

Figure 6:

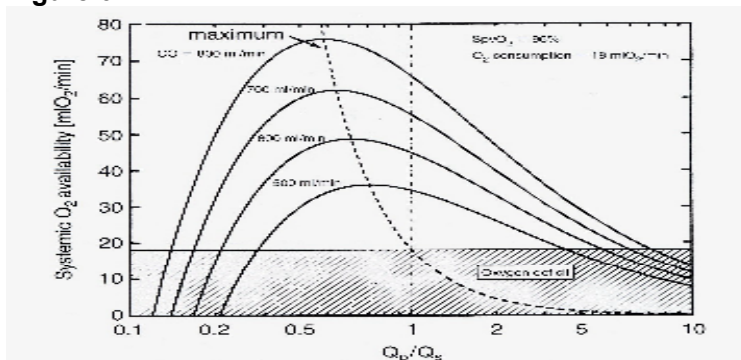


Figure 1.

Systemic oxygen availability (ml O_2 /min) as a function of the ratio of pulmonary to systemic blood flows (Q_P/Q_S) at different levels of cardiac output. Pulmonary venous oxygen saturation (Sp O_2) is assumed to be 96%, and oxygen consumption (VO_2) is assumed to be 18 ml O_2 /min. (Used with permission from Barua et al.⁵)

Total Cardiac Output

- It is for this reason that the focus of postoperative care after the Norwood 1 operation has now shifted to improving cardiac output and away from the past obsession of balancing $Q_p:Q_s = 1$. (see below)
- The best monitor of adequacy of cardiac output or systemic oxygen delivery DO_2 is $SmvO_2$, which as mentioned earlier, is monitored continuously in some centres of excellence (Tweddell et al.).
- In the absence of $SmvO_2$, low cardiac output is manifest by poor perfusion, acidosis, lactic acidosis, poor urine output, high filling pressures, and often poor function on echo and often with low SaO_2 .
- Also need to be vigilant at maintaining adequate diastolic BP for adequate coronary perfusion and adequate filling pressures, particularly with the use of aggressive vasodilator therapies.

Therefore the goal of therapy in these patients is to optimise DO_2 to meet vital organ tissue demands rather than aiming for a specific SaO_2 per se.

This should encompass:

- Assessing that there is unobstructed inlet and outlet flows to and from the single ventricle by echo (particularly post surgery). All patients pre and post Norwood 1 should have an early echo (ideally in theatre) and this should be repeated if any concerns in the early post-op period or failure to wean from ventilation.
- The **goal is to optimise DO_2** by inotropes, afterload reduction, while balancing the two circulations and maintaining adequate blood pressure (especially Diastolic BP) and SO_2 s.
- As discussed above newer data suggests improved DO_2 is best achieved by focusing on the management of improving total cardiac output through reducing systemic vascular resistance rather than balancing $Q_p:Q_s$.
- In all instances it is also imperative to maintain adequate oxygen carrying capacity by ensuring adequate Hb of 13 to 16 mg/dl.

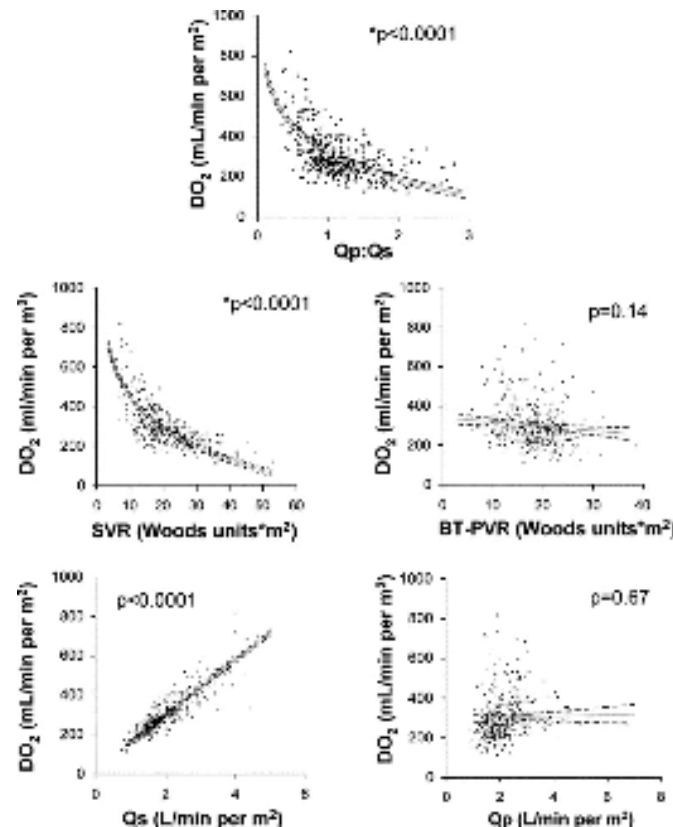
“Balancing the circulations?”

1. Altering SVR to favour Q_s

The favoured approach at the moment to improving DO_2 in patients with LCOS before and after the Norwood 1 operation is the use of pharmacological agents to reduce SVR. In this situation, where there is usually a high SVR and normal PVR, the use of systemic intravenous vasodilators appears to favourably influence DO_2 by having a greater effect on reducing SVR compared with PVR.

- Agents most commonly used include high dose milrinone (0.5 to 1.0 mcg/kg/min) (drug of choice at GOSH because of inodilator properties and proven value in large RCT); nitroprusside and phenoxybenzamine (long acting, difficult to reverse without the use of a vasoconstricting agent and therefore rarely used at GOSH).
- It is also important to recognise that when systemic vasodilators are used in patients with functional univentricular physiology that CO and BP are sensitive to adequate preload, which should therefore be maintained with adequate filling [and judicious titration of PEEP](#).
- [As there is invariably a degree of impaired myocardial contractility post Norwood stage 1, we routinely add low dose epinephrine 0.02 to 0.04 mcg](#)

/kg/min to the milrinone infusion. We try not to use high doses of epinephrine if possible because of the increased myocardial oxygen consumption and risk of myocardial fibrosis.



Li J, Zhang G, McCrindle BW, Holtby H, Humpl T, Cai S, Caldarone CA, Redington AN, Van Arsdell GS. Profiles of hemodynamics and oxygen transport derived by using continuous measured oxygen consumption after the Norwood procedure. J Thorac Cardiovasc Surg. 2007 Feb;133(2):441-8.

2. Manipulating PVR to favour Qs:

Strategies to increase PVR in situations where the Qp:Qs is deemed too high and where residual anatomic lesions have been ruled out, oxygen consumption, haematocrit and afterload reduction are being optimised include:

- Reduce FiO₂ to room air (0.21). The trend has now moved away from using subatmospheric O₂ levels as an alternative to hypercarbia to increase PVR. There is increasing evidence that hypercarbia is a more effective way of improving DO₂ and in particular cerebral oxygen delivery which may be compromised by hypoxic gas mixtures. The latter (subatmospheric O₂) should thus be used with extreme caution in these patients (NOT used at GOSH).
- Induced respiratory acidosis (hypercarbia) by either hypoventilation or exogenous CO₂. The latter is rarely used now, particularly with the emphasis now being on reducing SVR.
- Increases in PVR can also be achieved with the use of high PEEP. This increases PVR by compressing inter-alveolar pulmonary arteries. *It is important to appreciate, however, that the nadir of LOW PVR occurs at FRC and thus if the patient has poor lung compliance and atelectasis then increasing PEEP to achieve FRC will in fact drop PVR by aiding lung recruitment. Another word of caution is that by using excessive PEEP one*

may reduce systemic venous return, which may be problematic particularly if patients are receiving aggressive vasodilator therapy.

- High haematocrit or Hb around 16, which increases blood viscosity through the pulmonary bed thus reducing pulmonary blood flow.

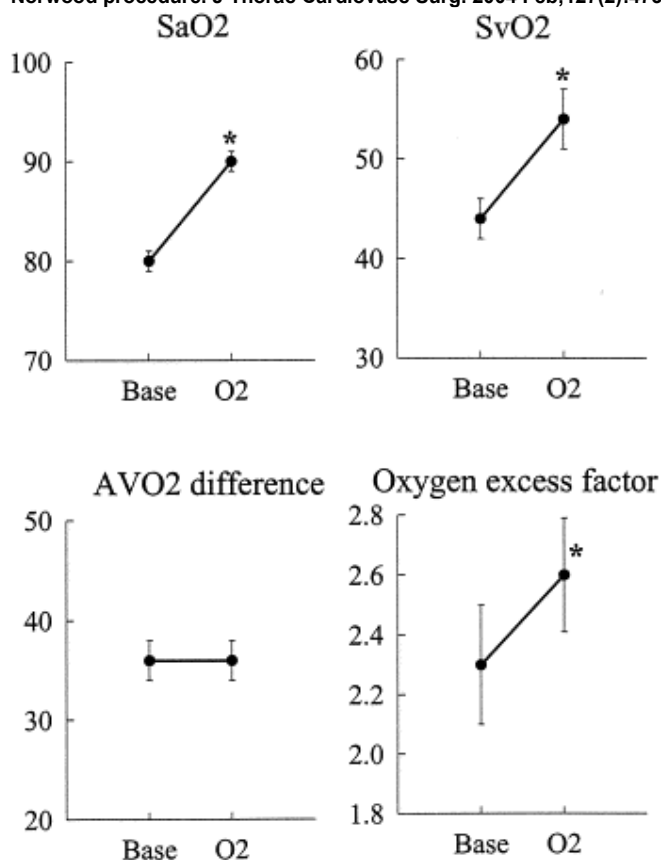
In contrast, some patients' pre and post-op course may be complicated by a low pulmonary blood flow causing severe hypoxaemia. A high PVR in the newborn may be caused by PPHN or secondary insult, which may in turn compromise DO_2 . It is vital in this situation to first exclude an anatomical problem such as a blocked or kinked shunt or a closing duct.

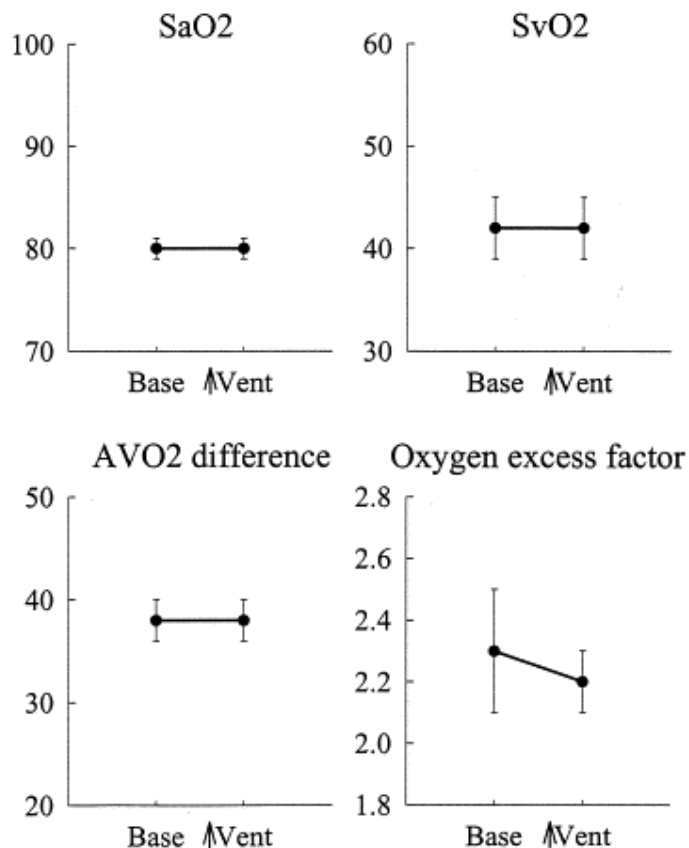
Where the problem is physiological increase in PVR, then the following strategies to reduce PVR are used:

- Gently increase FiO_2 (titrate up slowly as precipitous increase in FiO_2 can cause a sudden fall in PVR and loss of cardiac output from high pulmonary run off and inadequate coronary perfusion), normocarbica, mild alkalosis, appropriate PEEP and occasionally HFOV to achieve adequate lung recruitment in patients with poor compliance (from ARDS/capillary leak from SIRS, atelectasis etc). When all these conventional manoeuvres fail, consider inhaled nitric oxide (an unusual, but not unheard of clinical situation).

Where afterload reduction is optimised the DO_2/VO_2 balance may benefit from a controlled increase in inspired fraction of oxygen.

Bradley SM, Atz AM, Simsic JM. Redefining the impact of oxygen and hyperventilation after the Norwood procedure. J Thorac Cardiovasc Surg. 2004 Feb;127(2):473-80.





Summary of Management of Patients with LCOS and Univentricular physiology

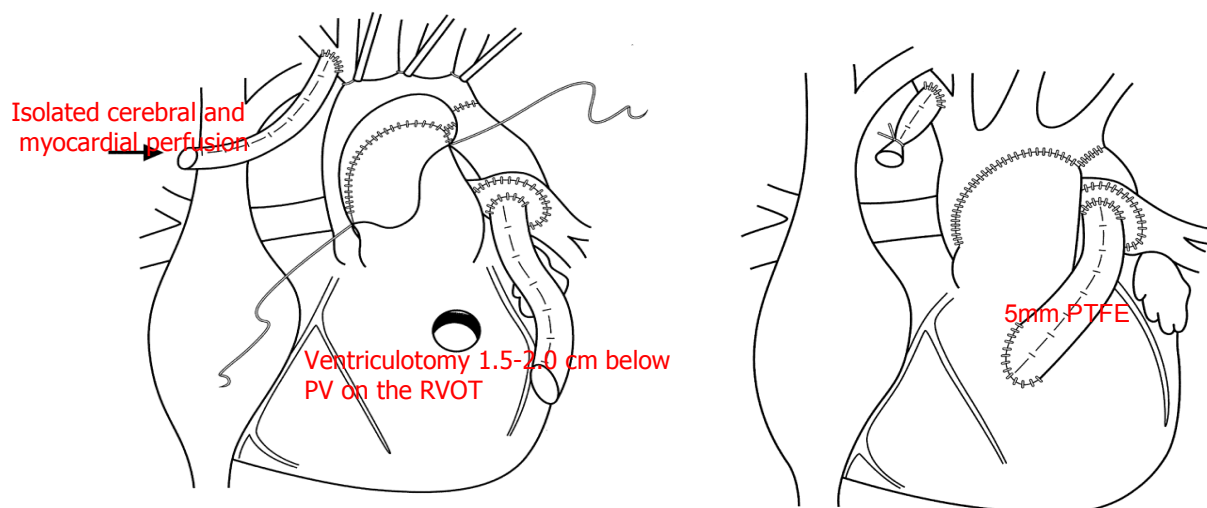
- Minimise oxygen consumption
- Exclude anatomic lesions, in particular, obstruction to pulmonary venous return (adequate atrial septectomy) and neo-aorta / aortic arch, with further assessment of RV function and right AVVR
- Increasing CO is the most effective way of improving DO_2 . This is best achieved by ensuring A-V synchrony reducing SVR with aggressive vasodilator therapies and modest inotropy. Adequacy of perfusion assessed by $SmvO_2$, Lactate, acidosis and clinical criteria.
- Maintain adequate O_2 carrying capacity (Hb)
- Ensure adequate lung function recruitment if hypoxaemic due to atelectasis.
- Optimise ratios of $Q_p:Q_s$ by manipulation of PVR.
- If this fails consider reducing oxygen consumption (VO_2) by cooling
- If all above fails consider mechanical support. It is worth noting that Ungerleider and colleagues have reported the successful use of semi-elective mechanical support with NOMOVAD (no membrane oxygenation ventricular assist device) to support all neonates after the Norwood 1 operation for 48 to 72 hours postop. This is based on the principle that many of these patients suffer significant long-term neurological morbidity and that this morbidity may be reduced by the use of high cardiac output using a VAD / ECMO during the period when these neonates brains are most susceptible to HIE, i.e. during the first 48 hours post bypass. It should be noted that they remain the only large centre using this strategy, and the long-term neurological outcome of these patients is eagerly awaited.
- It is important to also note that the patient's post-op clinical course might be further compromised by:

- The deleterious effects of cardiopulmonary bypass with systemic inflammatory response.
- A period of deep hypothermic arrest with the risk of ischaemic reperfusion injury

Sano modification:

Poor outcomes in early Norwood stage 1 operations were often attributable to large systemic to pulmonary shunts causing excessive pulmonary blood flow and right ventricular overload. To counter this, Sano et al developed a modified operation where the pulmonary blood flow is supplied directly by a 5 mm Gortex tube / conduit from the RV to the pulmonary trunk, thus obviating the need for a BT shunt (see figures a and b):

Figure a and b – Sano modification of the Norwood procedure



The theoretical advantages over the conventional Norwood 1 are:

- 1.) A more stable pulmonary blood supply; resulting in decreased diastolic “run-off” from the systemic circulation (“steal” phenomenon), thus allowing a higher diastolic pressure, optimising coronary perfusion.
- 2.) Decreased pulmonary blood flow; resulting in decreased ventricular overload and ventricular dilatation
- 3.) Pulsatile flow in the pulmonary arteries; facilitating better symmetrical growth of these vessels.

Early reports suggested a more stable post-operative course following the Sano operation. However, subsequent reports (ref a) have not shown a significant difference in terms of post-operative haemodynamics in Norwood operations using the Sano vs. conventional (BT shunt).

Furthermore, the Sano operation has several potential disadvantages:

- 1.) A ventriculotomy is required. This may render the RV susceptible to arrhythmias and contractile dysfunction.

- 2.) *Free “pulmonary” regurgitation through the conduit may result in ventricular dilatation.*
- 3.) *Central pulmonary stenosis may occur at the site of conduit insertion.*

Currently the choice of BT shunt vs. Sano modification depends on the institutional / surgical preference. However, a multicentre, randomised trial comparing the techniques sponsored by the National Heart, Lung, and Blood Institute has completed recruitment with analysis underway. Outcome measures in this study will include mortality and neurodevelopmental outcome, as well as morbidity and cardiac function measures.

“Hybrid” palliation

Hybrid palliation is a recently proposed alternative to Norwood stage 1. The intention of the procedure is to reduce the amount of time the patients are exposed to intra-operative circulatory arrest and cardiopulmonary bypass (which contribute to mortality and morbidity).

The procedure involves stenting the arterial duct and banding the pulmonary artery. The procedure is carried out partly by the surgical team (PA banding), and partly by the interventional cardiologists (PDA stenting) – hence the term “Hybrid”. In order to be suitable for the procedure, the ASD must be non-restrictive (although atrial septostomy has been included in the procedure in some cases).

The second stage of palliation in these cases involves aortic arch reconstruction, Glenn shunt and PDA ligation / stent removal).

The theoretical advantages are a reduced need for cardiopulmonary bypass and circulatory arrest during the neonatal period which is hoped to lead to improved long-term neurological development and myocardial function.

However, there are potential complications of this approach; including stent migration, stent thrombosis, over- and under-banding of the pulmonary arteries.

At present there is insufficient data to draw firm conclusions on the long-term outcomes of this procedure (but data is being collected and reported in small series). *It should be remembered that in the absence of an arch repair the blood supplied to head and neck vessels may be variable or more susceptible to ‘run off’ to the pulmonary and somatic vascular bed.*

A potential development for the future is the use of flow restrictors in the pulmonary artery which may be inserted via catheter, in place of the PA band. This would allow the stage 1 palliation to be completed without the need for surgery.

Foetal Intervention

As early as March 2000, foetal intervention has been carried out on a select cohort of fetuses deemed to have evolving HLHS in a bid to better engage the left ventricle and where possible, improve the chances of a biventricular repair. The lesions targeted for intervention include the stenotic aortic valve and the restrictive atrial septum.

Transplantation

Orthotopic cardiac transplant has been performed in neonates with HLHS, with good outcome. However, donors are in severe shortage, and the mortality of HLHS patients awaiting transplantation has been reported as 21-37%. In view of this shortage of donors transplantation as a primary treatment for neonates with HLHS is not routinely offered.

COMPLICATIONS OF FONTAN SURGERY

Arrhythmia

Thrombosis

Consequences of systemic venous hypertension

- Chylothorax
- PROTEIN LOSING ENTEROPATHY

This generally occurs as an important complication after the Fontan operation. It is characterised by depressed serum albumin concentration with no obvious cause. Clinical suspicion may be aroused by late postoperative onset of generalised oedema and pleural or peritoneal fluid collections. Patients may or may not have diarrhoea. The diagnosis is characterised by Alpha 1 anti trypsin in the stool, as clearance of this protein is abnormally high. These patients also classically have low total serum calcium due to low serum proteins. Specific studies show excessive loss of serum proteins from the GIT and endoscopic examination shows prominent lymph vessels in the jejunal mucosa. Approximately 13% of Fontan patients develop evidence of protein losing enteropathy by 10 years. It is associated with a near 50% mortality by 5 years after diagnosis.

Protein losing enteropathy (PLE) has been reported in almost all situations in which there is chronically elevated central venous pressure including chronic heart failure, chronic constrictive pericarditis and following atrial switch operations. From a pathophysiological point of view, it is presumed the combination of increased lymph production from increased IVC pressure and impaired lymph drainage from increased SVC pressure contribute to this complication.

Treatment is geared towards re-operations that result in a lower right atrial pressure. There is, however, a considerable early mortality after re operation. The use of valves in the IVC orifice are almost never carried out today.

Other strategies aimed at correcting haemodynamic abnormalities when right atrial pressure is low has resulted in some resolution. Creating a late fenestration has been reported with some success. Non-surgical treatments have included the use of steroids and heparin. It is now also believed that GI protein loss occurs before clinical onset of PLE.

Treatment strategies tailored to the severity of the disease include:-

- Symptomatic relief with diuretics and supplemental protein
- Strategies to halt intestinal protein leak using steroids or heparin
- Alteration of cardiovascular physiology via fenestration, atrial pacing or heart transplant

Other sources of information:

Websites.

http://www.pediheart.org/practitioners/defects/ventriculoarterial/TOF_PA.htm
<http://www.emedicine.com/radio/topic685.htm>
<http://www.emedicine.com/ped/topic2539.htm>
<http://www.emedicine.com/EMERG/topic575.htm>
http://www.rch.org.au/cardiology/defects.cfm?doc_id=3011
<http://www.emedicine.com/ped/topic2550.htm>
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Operative Cardiac surgery Edited by TJ Gardner and Thomas L Spray (used as source for many of pictures of surgical procedures)

Ref a) **Norwood procedure for hypoplastic left heart syndrome: BT shunt or RV-PA conduit?** Edwards L, Morris K, Siddiqui A, Harrington D, Barron D, Brawn W. Arch. Dis. Child. Fetal Neonatal Ed. Published online 26 Sept 2006

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Applied physiology to the contemporary management of the neonate with hypoplastic left heart syndrome. Atik FA. Arq Bras Cardiol. 2006 Sep;87(3):e16-26