orphananesthesia

Anaesthesia recommendations for patients with

Hypoplastic left heart syndrome

Disease name: Hypoplastic left heart syndrome

ICD 10: Q23.4

Synonyms: HLHS

Disease summary:
Hypoplastic Left Heart Syndrome (HLHS) represents 1-2% of all congenital heart disease and is the most common lethal cardiac defect in neonates [3]. It is characterised by a variable degree of underdevelopment of the left heart structures, with hypoplasia or absence of the left ventricle, mitral stenosis or atresia, aortic stenosis or atresia and hypoplasia of the aortic arch. The result is an inability of the left side of the heart to maintain the systemic circulation. A duct-dependent circulation exists, with the right ventricle (RV) supporting the systemic circulation via right to left flow through the ductus arteriosus. Initial survival is dependent on continued ductal patency, unrestrictive atrial mixing to allow the pulmonary venous return to reach the systemic circulation and a balance between systemic (SVR) and pulmonary vascular resistance (PVR) to achieve adequate systemic and pulmonary blood flow.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

Disease summary

The incidence of HLHS is approximately 2 in every 10,000 births [1]. The precise aetiology is unknown. Research has indicated a genetic component with heritability seen in some families. There is an association with other genetic syndromes including Turners', Jacobsen's, Smith-Lemli-Opitz's, Trisomy 13 & 18, and chromosomes 17 & 18 deletion, amongst others. Extracardiac anomalies are rare in HLHS however, their presence, or the presence of genetic syndromes in association with a diagnosis of HLHS, usually carry a worse prognosis.

In regions with foetal ultrasound screening programmes the diagnosis of HLHS is often prenatal allowing for parental counselling, including options of elective termination or no intervention (comfort care), and allowing postnatal management planning. Neonates are often born in reasonably good condition and diagnosis by echocardiography may follow detection of a murmur, weak/absent femoral pulses (if there is significant arch hypoplasia) or cyanosis if clinically present. As the ductus arteriosus closes in the first week of life, rapid decompensation is likely and neonates may present in extremis with cardiorespiratory failure and shock. Without intervention, 90% will die in the neonatal period.

Once considered universally lethal, outcome has improved with primary cardiac transplantation or staged surgical reconstruction to create a viable circulation. With surgical intervention, 50-70% of neonates born with HLHS are now expected to survive to adulthood (Feinstein, 2012). Limited availability of suitably small donor organs and the high risk of mortality whilst awaiting a transplant dictates that most HLHS patients will undergo staged palliation. For all but the most borderline of cases, palliation is to a functionally univentricular pathway. The final stage is the creation of a total cavopulmonary univentricular circulation, the Fontan circulation, where pulmonary blood flow is supplied by passive systemic venous return, and the right ventricle is the single systemic ventricle. Right ventricular dysfunction is common, with myocardial ischaemia, chronic overload and the decreased ability of the morphological right ventricle to sustain systemic pressures all contributing factors. In addition, the Fontan circulation and associated elevated systemic venous pressures may impact on the function of other organ systems, such as the development of hepatic dysfunction. As cardiac function deteriorates, heart transplantation may re-emerge at a later stage as the only option for prolonged survival.

HLHS patients represent a high-risk population for morbidity and mortality under anaesthesia. Each reconstructive stage presents unique anatomical and physiological considerations for the anaesthetist. A clear understanding of the associated physiology and possible complications is of paramount importance in ensuring optimal patient care. Surgery should ideally be performed in centres with the necessary expertise and the availability of appropriate intensive care support.

Typical surgery

Cardiac surgery:

- Staged reconstruction surgery
  - Stage I: Norwood
  - Stage II: Glenn or hemi-Fontan
  - Stage III: Fontan or Total Cavopulmonary Connection (TCPC)
  - Hybrid procedures

 - Heart transplantation: Primary or as a rescue therapy at any stage of palliation

Non-cardiac surgery/procedures:

- Imaging (e.g. cardiac catheterisation, computed tomography (CT), angiography and magnetic resonance imaging (MRI) – to assess function and suitability for progression through staged reconstruction.
- Interventional catheter procedures (e.g. balloon arterioplasty for residual coarctation)
- Long-term central venous access (e.g. prostaglandin infusion, inotropic support)
- Gastrostomy (patients often have high nutritional requirements)
- Nissen fundoplication (high incidence of gastro-oesophageal reflux disease)
- Laparotomy and bowel resection (increased incidence of bowel ischaemia/necrotizing enterocolitis due to poor mesenteric flow)

Staged reconstruction surgery

A clear understanding of the functional cardiac anatomy and physiology before and after each palliative stage is essential to safely manage patients with HLHS. Surgical management varies, tailored to specific needs of morphological sub-groups and patient size, and continues to evolve with introduction of new techniques. A summary of the conventional stages of palliation is outlined below.

- Stage I: Norwood procedure

Performed in the neonatal period; establishes the right ventricle as the systemic ventricle. Consists of atrial septectomy (to ensure unimpeded pulmonary venous return to the right side), reconstruction of the aortic arch using the main pulmonary artery to create a neo-aorta, (relieves systemic outflow obstruction and allows the right ventricle to eject directly into the systemic circulation) and a systemic to pulmonary artery shunt (typically a modified 'Blalock-Taussig' (BT) shunt from a branch of the aorta to the pulmonary artery or a 'Sano' shunt from the right ventricle to the pulmonary artery) to provide pulmonary blood flow.

- Hybrid Stage I procedures

Performed in the neonatal period as an alternative to the Norwood procedure; involves joint interventional cardiology and surgical procedures to band the pulmonary arteries, stent the ductus arteriosus and create an unrestrictive atrial communication. Avoids the need for cardiopulmonary bypass at this stage, although long-term outcome benefits are yet to be demonstrated. Subsequent surgery would additionally include aortic arch reconstruction and ligation of the patent ductus arteriosus (PDA).

- Stage II: Glenn/Hemi-Fontan procedure

Usually performed at 4 to 6 months of age when the PVR has reduced and venous pressures are adequate to provide pulmonary blood flow. The systemic to pulmonary artery shunt placed during the Norwood procedure is removed. The superior vena cava (SVC) is disconnected from the right atrium and anastomosed directly to the right pulmonary artery in the 'Glenn' procedure or alternatively a 'hemi-Fontan' procedure is performed, where the SVC is again anastomosed to the pulmonary artery, however the SVC is left in continuity with the right atrium with a patch placed in the upper right atrium to prevent entry of SVC blood into the heart. Both procedures divert SVC blood directly to the pulmonary artery, providing a passive pulmonary blood flow. This reduces the volume load on the single functioning ventricle and improves circulatory system efficiency. The inferior vena cava continues to drain blood to the right atrium where it mixes with oxygenated blood from the left atrium, before being ejected into the systemic circulation.

- Stage III: Fontan/Total Cavopulmonary Connection surgery

Usually performed between 18 months and 5 years. There is completion of the total cavopulmonary connection (TCPC) with the inferior vena cava (IVC) also connected to the pulmonary artery via an intracardiac or extracardiac conduit. The blood circulation now exists in series. All systemic venous return is delivered passively to the pulmonary arteries, and pulmonary perfusion and adequate atrial filling is reliant on a low PVR and relatively high central venous pressure (CVP). A small fenestration is sometimes created between the IVC conduit and the right atrium, which allows a 'pop off' for systemic venous blood to enter the common atrium should the resistance rise in the cavopulmonary connection or the pulmonary vascular bed, thus maintaining cardiac output but at the expense of some arterial desaturation.

Type of anaesthesia

HLHS patients can be extremely fragile with a high incidence of peri-operative morbidity and mortality. Choice and conduct of anaesthesia must be put into the context of each patient's specific physiology with regard to their stage of palliation, as well as any comorbidities, and should be delivered by a team with the necessary expertise.

General anaesthesia with endotracheal intubation is often employed due to the age of patients presenting for surgery, the complexity of surgical procedures and an increased incidence of gastro-oesophageal reflux. Anaesthesia for induction and maintenance may be volatile or intravenous. All anaesthetic agents have the propensity to alter haemodynamics and injudicious use may destabilise these patients. Anaesthetic agents must be used cautiously and titrated to effect. Positive pressure ventilation by decreasing venous return can significantly reduce pulmonary blood flow in patients with a passive pulmonary blood supply, with deleterious effects. Specific considerations for the conduct of general anaesthesia at different stages of palliation are discussed later (see 'Anaesthesiologic procedure').

IV access prior to induction may be helpful, allowing administration of fluid prior to induction, and an immediate route to treat any instability associated with induction. Previous surgeries and ICU admissions may make establishing IV access challenging. Venous hypertension associated with Fontan circulation can however often result in enlarged veins.

Isolated regional or local anaesthetic techniques are seldom used due to the young age of most surgical HLHS patients however may be considered in older patients. Regional anaesthesia (in the form of single shot caudals, spinal and epidurals), in conjunction with general anaesthesia, has been reported for non-cardiac surgery with improved post-operative pain and decreased post-operative pulmonary hypertensive crises (Sacrista, 2003). However these techniques carry a risk of significant and prolonged reductions in SVR, which have been associated with increased peri-operative instability and the need for intraoperative inotropic support, and this must be considered. Central neuroaxial blockade may be contraindicated by patients' anti-platelet and anticoagulant therapy.

Necessary additional diagnostic procedures (preoperative)

A thorough preoperative assessment is required to understand current anatomy and physiology and to identify any signs of diminished pulmonary or systemic circulation or cardiac dysfunction.

Investigations:
- **Bloods**
  - **Full Blood Count** – polycythaemia secondary to chronic cyanosis; thrombocytopenia.
  - **Urea & Electrolytes, Magnesium** – electrolyte disturbances particularly if taking diuretics.
  - **Liver Function Tests** – (unstable patients) hepatic congestion or hypoperfusion.
  - **Arterial Blood Gas** – (unstable patients) baseline & assess acid-base and oxygenation.
  - **Clotting screen** – anti-coagulated or presence of acquired coagulation factor abnormalities.
- **Electrocardiography (ECG)** – Often non-specific with right axis deviation and right ventricular hypertrophy. May detect undiagnosed arrhythmias.
- **Echocardiography (ECHO)** – If no recent results and/or change to patient's functional status. Useful to assess right ventricular function, atrial communication, atrio-ventricular valve adequacy, right ventricular outflow tract and neo-aorta. Atrio-ventricular valve insufficiency is common but if severe may impede pulmonary venous return and ultimately contribute to a rise in PVR.
- **Chest radiography (CXR)** – if signs of worsening cardiac function, hypoxia or chest infection.

Optional additional investigations: Cardiac Magnetic Resonance (CMR/MRI) scan / Cardiac catheterisation.

Particular preparation for airway management

There is an increased incidence of gastro-oesophageal reflux in patients with HLHS. A rapid sequence induction may be considered, although there is little evidence in the literature to support this and the abrupt physiologic shifts may be poorly tolerated.

There is no reported association with isolated HLHS and a difficult airway. Multiple previous intubations and prolonged ventilation may put some patients at increased risk of subglottic stenosis. Intubation in general, is a time of potential physiological imbalance with risk of hypoxia and hypercarbia. Such imbalance is likely to be poorly tolerated in these patients and the anaesthetic team should be well prepared to gain quick control of the airway, intubate and ventilate appropriately. Careful consideration needs to be given to the effects of hyperoxia and hypocarbia on pulmonary vascular resistance (PVR). A reduction in PVR in patients with a balanced circulation has the potential to compromise their systemic circulation, myocardial blood supply and precipitate cardiac arrest. (See 'Anaesthesiologic procedure').

Particular preparation for transfusion or administration of blood products

No specific transfusion requirements have been reported for patients with isolated HLHS, but exposure to blood products for prior reconstructive surgery may result in an increased risk of antibodies. Patients taking anti-coagulation or anti-platelet therapy will have an increased propensity to bleed and should be worked-up accordingly.

Particular preparation for anticoagulation

HLHS patients may require anticoagulation therapy to reduce the risk of stent or vessel occlusion, or to counteract coagulation disturbances resulting from protein-losing enteropathy (a potential long-term sequalae of the Fontan circulation), which may cause a pro-thrombotic state. Stopping anticoagulation therapy prior to surgery, if indicated, needs to be discussed with the relevant teams to evaluate the risks of stopping and plan an appropriate duration.

Particular precautions for positioning, transport or mobilisation

Following Glenn and Fontan palliation, a cavopulmonary connection is present and pulmonary blood flow is passive. Care must be taken with positioning to prevent impairment of systemic venous return. Lying Fontan patients completely flat for induction, particularly if dehydrated from fasting or illness, can significantly compromise their haemodynamics, particularly if the systemic vascular resistance then also falls during induction. 'Fontan positioning' has been described to optimise venous return, with 45-degree head up (improving superior vena cava drainage) and knees flexed (improving inferior vena cava drainage) but complete adoption of this position is usually more applicable to the post-operative period.

Probable interaction between anaesthetic agents and patient's long-term medication

No specific interactions with anaesthetic agents have been reported. One must consider other peri-operative implications of the patient's long-term medications such as the increased incidence of apnoeas and hypotension associated with prostaglandins, electrolyte disturbances associated with diuretics and the risk of prolonged hypotension under anaesthesia associated with ACE-inhibitors.

Anaesthesiologic procedure

A through understanding of the patient's underlying cardiopulmonary physiology is essential to ensure optimal anaesthetic management of patients with HLHS. Outlined below are some specific considerations for each of the conventional stages of palliation.

Uncorrected

The neonate remains duct-dependent for an adequate systemic circulation and a prostaglandin infusion should be continued to maintain ductal patency. The pulmonary and systemic circulations exist in parallel, both provided by the right ventricle, and flow to each must be balanced equally. This 'balance' in flow is predominantly determined by the pulmonary vascular resistance (PVR) and the systemic vascular resistance (SVR) and the anaesthetic goal should be to predict and minimise changes to avoid haemodynamic compromise of either circulation. Anaesthetic agents may cause the SVR to fall, which may increase systemic circulation at the expense of pulmonary blood flow, resulting in profound hypoxaemia. Conversely, a fall in PVR will increase pulmonary blood flow at the expense of systemic perfusion and can rapidly lead to a low cardiac output state. This may happen naturally in the pre-operative period as PVR falls following birth. Supplemental oxygen will reduce the PVR and may be deleterious. As low as feasible inspired oxygen concentration should be used peri-operatively, accepting SpO_2 75-80%. If higher concentrations are

required to cover induction/intubation, this should be decreased as soon as tolerated. Hypocarbia and alkalosis will also reduce PVR and should be avoided, and hypoventilation to achieve modest hypercarbia is generally employed. Hypoxic inspired gas mixtures and inhaled carbon dioxide, previously used in some centres to increase PVR, have largely gone out of vogue. Increases in afterload (SVR) should be avoided with consideration of the effects of pain, cold, distress and vasopressor medication. Hypotension after induction is not uncommon and inotropes should be available if required. General considerations of anaesthesia for neonates apply.

Post-Norwood stage I palliation (Interstage)

Interstage Norwood patients (between stage I and II) have the highest risk of mortality of any of the stages of palliation reported at 2-16% (Feinstein, 2012). These patients are dependent upon their systemic to pulmonary artery shunt; prolonged fasting and dehydration, which may lead to shunt thrombosis, should be avoided. Intravenous fluids may be necessary in the preoperative period. Patients are preload dependent and may require volume loading prior to induction and close attention should be given to peri-operative fluid losses or shifts. Coronary ischaemia, systemic ventricular failure, neo-aortic obstruction and inadequate or excessive shunt flow are common causes of death. Patents have limited cardiovascular reserve and a high incidence of instability at induction (69%) and emergence (48%) has been reported (Watkins, 2012). Additional monitoring and a low threshold for intensive care may be warranted. The circulation remains in parallel and requires careful balancing. Considerations outlined above still apply. Increases in SVR and/or reduction in PVR are poorly tolerated and risk cardiac instability and arrest. Conversely increases in PVR and/or reduction in SVR may lead to severe hypoxaemia. Excessive oxygenation should be avoided, accepting SpO_2 75-85% and aiming for normocarbia. Avoid increases in SVR. Low cardiac output states are common, particularly post-cardiopulmonary bypass, and pharmacological afterload reduction with phosphodiesterase inhibitors, sodium nitroprusside or alpha-blockers may be required.

Post-Glenn or hemi-Fontan palliation

The circulation remains in parallel, however patients with Glenn physiology are usually more resilient than those with Norwood physiology, with reduced incidence of peri-operative instability. Pulmonary blood flow is passive and dependent on a low PVR to permit a small pressure gradient between the SVC and common atrium. The CVP measurement (from a neck vessel) now provides a measure of the pulmonary artery pressure and should be maintained at higher than normal venous pressures, typically 10-15mmHg. The PVR should be kept low, avoiding hypoxia, hypercarbia and acidosis. Peri-operative management should ensure adequate venous return and maintenance of intravascular volume. Hypocarbia should also be avoided, as it may cause cerebral vasoconstriction reducing cerebral blood flow, venous return to the SVC and pulmonary blood supply, in turn. High intrathoracic pressures occurring during positive pressure ventilation may also impede venous return and subsequently pulmonary blood flow. Spontaneous ventilation creates a negative intrathoracic pressure with inspiration, increasing systemic venous return and cardiac output and is therefore preferable but not always appropriate. Inadequate spontaneous ventilation runs the risk of hypercarbia, hypoxia and reduced lung volumes increasing the PVR, which may also be deleterious. If mechanical ventilation is required, a ventilation strategy should be employed to give the lowest possible mean airway pressures for the desired minute ventilation and gas exchange parameters. Ventilating with higher volumes at a lower rate with a low inspiration to expiration ratio (e.g. I:E ratio 1:3) and avoidance of excessive positive end-expiratory pressure (PEEP) may help reduce overall pulmonary pressures. Early weaning from mechanical ventilation is recommended. Oxygen restriction is less important

than pre-Glenn and may be used to reduce PVR. Patients will remain cyanotic due to deoxygenated blood returning from the IVC returning to the atrium – accept SpO_2 80-85%.

Post-Fontan palliation

The cavopulmonary circulation is complete and the pulmonary and systemic circulations now exist in series. Cardiac output is critically dependent on right heart filling determined by pulmonary blood flow. Good systemic venous return is essential. Preload must be maintained, with attention to volume status, avoiding hypovolaemia, dehydration and fluid shifts. As discussed above, a higher 'CVP' of 10-15mmHg is often targeted. The PVR should be kept low and high intrathoracic pressures avoided. Patients should no longer be cyanotic with SpO_2 >95%. Oxygen saturation may however be significantly lower in the context of a fenestrated conduit. The presence of a fenestration is important to note, as peri-operative arterial desaturation may relate to increased 'right to left' shunt rather than pulmonary pathology.

Post-Fontan palliation HLHS patients often have the most robust physiology compared to other stages however they still represent a high-risk population. Long-term sequelae of the Fontan circulation may include problems relating to high venous pressures such as Fontan-associated liver disease, recurrent effusions, plastic bronchitis, ascites and protein-losing enteropathy. Right ventricular dysfunction is common, as with time the morphological right ventricle struggles to sustain systemic pressures. Signs and symptoms of cardiac and end organ dysfunction should be actively sought in the pre-operative assessment, as their presence is associated with a poorer prognosis.

Other considerations

Sedative-analgesic pre-medication may be useful in reducing patient anxiety and stress at induction, avoiding unwanted increases in sympathetic response, myocardial oxygen demand and afterload.

In most stages of palliation, patients are pre-load dependent. Prolonged fasting and dehydration may be poorly tolerated. Additionally, dehydration is association with polycythaemia and hyperviscosity, which may increase the risks of shunt thrombosis in Norwood stage 1 patients. Fasting should be kept to a minimum with clear fluids continued as long as possible, and IV maintenance fluid considered if delays are anticipated.

Atrial contraction is important in maintaining ventricular filling and adequate atrial emptying. Sinus rhythm should be maintained and arrhythmias diagnosed and treated appropriately. Avoidance of tachycardia and maintenance of adequate diastolic pressure will help ensure adequate coronary perfusion, reducing the risk of myocardial ischaemia.

As with all patients with intracardiac communications or shunts, care must be taken to avoid air bubbles and the risk of systemic embolization. For this reason, some also advise the avoidance of nitrous oxide (N_2O) in the maintenance of anaesthesia.

Standard antibiotic prophylaxis against infective endocarditis may be indicated for some procedures.

Particular or additional monitoring

- Central venous monitoring

- o Aim to preserve line sites avoiding cannulation of neck vessels where possible, as thrombosis may jeopardise future surgical options. Femoral vessel cannulation is often preferred. If neck cannulation is unavoidable, use of small calibre lines with fewer lumens, and early removal post-operatively is desirable. Neck cannulation is often warranted for the palliation surgery to create the cavopulmonary anastomoses, in order to provide an indicator of the pulmonary artery pressures.

  - o Note both venous and arterial occlusions are common due to previous lines and/or cardiac catheterisation.

  - o Continuous venous oximetry (SvO_2) if available - provides an indicator of systemic oxygenation.

- Blood pressure monitoring

  - o Invasive arterial monitoring and blood gas analysis should be considered for lengthy or cardiac procedures or if concerns about ventricular function or oxygenation.

  - o Non-invasive blood pressure (NIBP) measurement is often sufficient for brief procedures in stable patients.

  - o Note arterial pressure readings are likely to be lower in a limb involved with a BT shunt.

- Near-infrared spectroscopy (NIRS): cerebral or somatic

  - o Provide a measure of regional oxygen saturation as a non-invasive estimate of SvO_2.

- Peri-operative echocardiography: Epicardial or transoesophageal

  - o May provide valuable information on ventricular function, valve function and shunt flow.

- Left atrial pressure (LAP) line (may be inserted during Fontan surgery)

  - o Useful for assessing transpulmonary gradient (SVC – LAP).

Possible complications

- Arrhythmias – Most commonly supraventricular tachyarrhythmia. Junctional ectopic tachycardia (JET), ventricular tachycardia and complete heart block also reported.

- Myocardial ischaemia – particularly when coronary perfusion is provided by retrograde flow from a patent ductus arteriosus (PDA). Low PVR in the presence of a BT shunt can cause diastolic run-off reducing myocardial perfusion pressures.

- Shunt thrombosis – Increased risk with polycythaemia, dehydration and reduced or competitive shunt flow.

- Low cardiac output states – may be secondary to inadequate preload, elevated PVR, ventricular dysfunction, anatomical obstruction to venous return; May require extra-corporeal membrane oxygenator (ECMO) support.

- Infections – Increased risks of infection associated with low cardiac output, cyanosis, high pulmonary blood flow, mechanical ventilation and invasive lines.

- Prolonged post-op mechanical ventilation – secondary to pulmonary over- or under-circulation.

- Cardiac arrest.

Postoperative care

HLHS patients have a high risk of post-operative complications. Post-operative management needs to be a continuation of care tailored to their unique physiology. There should be a low threshold for post-operative High Dependency Unit (HDU) or Intensive Care Unit (ICU) admission.

Neonates receiving prostaglandins are at increased risk of apnoeas, particularly in conjunction with opioid medications. Post-operative apnoea monitoring is advisable in this group.

Patients with cavopulmonary connections will benefit from early transition to spontaneous ventilation and extubation. Adopting a 'Fontan position', 45 degrees head up with knees bent, may also facilitate venous return.

Pain can cause a sympathetic response with unwanted tachycardia and increases in SVR and PVR. A good multi-modal analgesic strategy should be employed for these patients.

Information about emergency-like situations / Differential diagnostics

Shunt stenosis or thrombosis
- Increased risk associated with polycythaemia, dehydration, and reduced or competitive shunt flow.
- May present gradually or suddenly with acute obstruction to pulmonary blood flow leading to severe hypoxia, cyanosis and acidaemia. Diagnosis is usually clinical. Lack of shunt murmur and echocardiography may aid diagnosis.
- Treatment involves immediate resuscitation and pharmacological treatment with heparin or thrombolytic agents or mechanical disruption by catheter intervention or surgery to restore shunt flow. In extreme cyanosis, extra-corporeal membrane oxygenator (ECMO) support may be initiated prior to further intervention.

Ambulatory anaesthesia

Due to the high incidence of peri-operative morbidity and mortality in patients with HLHS, ambulatory anaesthesia is often inadvisable. Post-Norwood infants will rarely be appropriate for ambulatory anaesthesia, while uncomplicated patients with Glenn, hemi-Fontan or Fontan circulations that are clinically stable may be considered.

With 70% of patients with HLHS now expected to survive into adulthood, obstetric management of these patients is likely to become increasingly more relevant. Cardiac disease remains a common cause of maternal death and congenital heart disease makes up a significant portion of this. The haemodynamic changes associated with pregnancy and delivery may be detrimental to Fontan physiology, especially in the presence of ventricular dysfunction and arrhythmias. There is significant risk of maternal morbidity and mortality. Chronic maternal hypoxaemia is also associated with a higher incidence of spontaneous abortion, intrauterine growth restriction and preterm labour.

Women should undergo preconception counselling to be informed of the risks, and also to be optimised and managed appropriately throughout their pregnancy. Parturients are ideally managed in centres where obstetricians have experience of congenital heart disease and there is appropriate cardiology and neonatal services.

Method of delivery is usually defined by obstetric considerations. Elective caesarean section may be indicated in parturients with severe ventricular dysfunction. Vaginal delivery is generally associated with less blood loss and is often the preferred mode in patients with reasonable cardiac function. An active second stage with shortened duration and/or assisted delivery is preferable where possible, reducing the time where Valsalva manoeuvres during 'pushing' may impede venous return and increase afterload (Jooste, 2013). Parturients should have continuous electrocardiography (ECG) monitoring and large bore IV access early in labour. Small fluid boluses may be required to maintain preload. Supplementary oxygen during labour may also help to reduce the PVR and improve foetal oxygen delivery.

Labour analgesia with epidural, if not contraindicated, may help to reduce the sympathetic response and tachycardia associated with the pain of labour. A graduated technique using lower doses of local anaesthetic may help to prevent sudden changes in SVR. An epidural or combined spinal-epidural (CSE) with a low dose spinal, is usually preferred for caesarean section over spinal anaesthesia alone which is associated with more precipitous falls in SVR. Again doses should be modified to prevent sudden changes in SVR. Caution should be applied with the use of vasopressors to increase SVR after central neuroaxial blockade, as hypotension may be due to low systemic cardiac output, which will be worsened by increasing the afterload. Principles of general anaesthesia for caesarean section are as outlined previously. Left uterine displacement is crucial in this population in order to maintain preload. Regardless of mode of delivery, meticulous attention must be paid to any peri-partum blood loss, as hypovolaemia is likely to be poorly tolerated. The prothrombotic state of pregnancy, particularly in combination with protein-losing enteropathy, may increase the risk of intra- or extra-cardiac thrombosis. Appropriate prophylaxis should be discussed with the relevant teams.

Literature and Internet links

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Patient/Family information

1. Understanding your child's heart – Hypoplastic Left Heart Syndrome.
   British Heart Foundation. www.bhf.org.uk
2. Hypoplastic Left Heart Syndrome. Children's Heart Federation. www.chfed.org.uk

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