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REVIEW AND VIDEO REPORT

# Anaesthesia for the child with a univentricular heart: a practical approach

**L Veronese,[1] J Swanevelder,[2] A Brooks[2]**

*[1] Logan Hospital, Australia*
*[2] Red Cross War Memorial Children's Hospital, University of Cape Town, South Africa*
***Corresponding author, email:*** *Lara.veronese@health.qld.gov.au*

Congenital heart disease (CHD) represents a major challenge for anaesthetists. This is due to the complexities in pathophysiology and the impact of surgery and anaesthesia, which predisposes to significant perioperative morbidity and mortality.[1] Univentricular heart (UH) consists of a large variety of rare and complex congenital cardiac malformations. There are rare cases of children having only a single functional ventricle from birth. More commonly, children may receive surgical palliation for complex cardiac defects by creating a univentricular heart when biventricular repair is not feasible. About 30% of patients with univentricular physiology have complications when undergoing non-cardiac surgery.[2]

During this review, we aim to provide a practical understanding of the pathophysiology of UH and outline a systematic approach to the perioperative management of these patients presenting for non-cardiac surgery. We conclude with an evaluation of the use of near-infrared spectroscopy, transoesophageal echocardiography and ventricular assist devices, as technologies with increasing application in the management of this challenging patient population.

Supplementary video available online: http://www.sajaa.co.za/index.php/sajaa/article/view/2572

**Keywords:** univentricular heart, Fontan, congenital heart disease, paediatric anaesthesia

## Learning objectives:

- Learn the epidemiology and classification system.
- Understand the pathophysiology and its evolution into completion of palliation.
- Understand and learn to evaluate the complications related to the pathophysiology, surgery and medical management of the UH.
- Form a perioperative plan for a child with a UH presenting for non-cardiac surgery.
- Consider the role of novel technology in the management of anaesthesia of the patient with a UH.

## Epidemiology

Current statistics show that worldwide there are 8 per 1 000 live births per year with CHD,[3] and the prevalence of this condition is increasing.[4] Of all cases of CHD, 1.8% to 9.4% consist of UH.[4,5] This prevalence varies markedly between countries.[3-5]

## Classifying the univentricular heart

A vast body of literature describes various classification systems and categories of UH. The International Society for Nomenclature of Paediatric and Congenital Heart Disease has taken several years to develop their current classification system which was released in December 2017. They state: *"The term 'functionally univentricular heart' describes a spectrum of congenital cardiac malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation. A heart may be functionally univentricular because of its anatomy or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass."*[6]

They include the following conditions as univentricular in nature:

- Double inlet right ventricle (DIRV)
- Double inlet left ventricle (DILV)
- Tricuspid and mitral atresia
- Hypoplastic left heart syndrome
- Hearts affected by a complex form of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other cardiovascular malformations may also sometimes be considered as a functionally univentricular.[6]

## Evolution of pathophysiology from birth to completion of surgical palliation

Due to the term "functionally univentricular heart" encompassing many distinct pathologies, a child may initially present with a cardiac anomaly that differs considerably from another child also classified as having a functionally UH. Despite this heterogeneity in presentation, children will fall into one of two categories. The child either has only one functional ventricle from birth (e.g. hypoplastic left heart syndrome) or has an anomaly that will not allow biventricular repair (e.g. pulmonary atresia with intact ventricular septum) and will be palliated with the construction of a functionally UH. Most often, if the child is presenting for non-cardiac surgery, they will usually have progressed through the first stage of surgical palliation.

The palliation process commonly involves three stages of therapeutic intervention.[7] The first stage is either increasing pulmonary blood flow through the creation of a shunt between a systemic artery and pulmonary artery, or diminishing excessive pulmonary blood flow by application of a pulmonary artery band. Today these palliative procedures are commonly done via

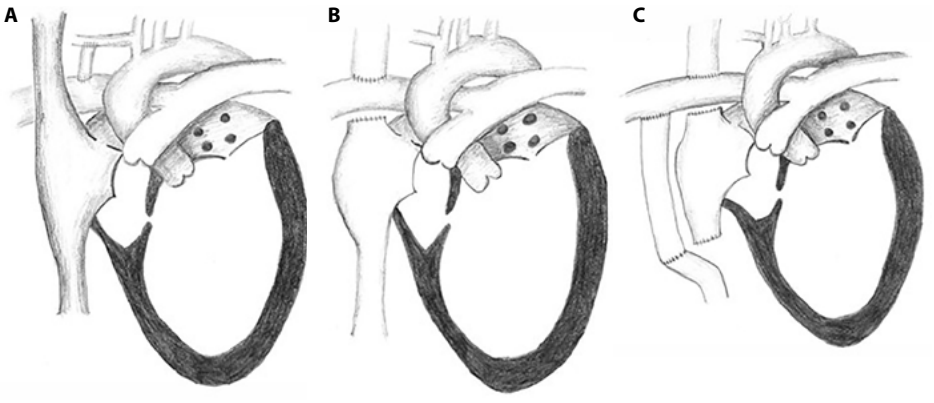



**Figure 1:** The stages of palliation in tricuspid atresia. (A) Stage 1: Creation of a shunt between a systemic artery and pulmonary artery (illustrated here as right subclavian artery to right pulmonary artery). (B) Stage 2: Connection of the superior vena cava to pulmonary artery. (C) Stage 3: Connection of the inferior vena cava to the pulmonary artery via a conduit (total cavopulmonary connection i.e. completion of the univentricular heart).

sternotomy, although in the past, a thoracotomy was frequently used. The second stage is the connection of the superior vena cava (SVC) to pulmonary artery, also known as bi-directional cavo-pulmonary connection, or bi-directional Glenn shunt. The third and final stage is connection of the inferior vena cava (IVC) to the pulmonary artery, with either an extracardiac or intracardiac conduit, which completes the formation of the UH (Figure1). This is performed with or without a fenestration. Different institutions may employ variations of these stages. This final stage is commonly referred to as the Fontan procedure or total cavopulmonary connection (TCPC).

The final resultant physiology functions on two main principles: passive venous return carries deoxygenated blood straight to the lungs from the rest of the body, without being pumped by the heart; and a single ventricle pumps oxygenated blood to the systemic circulation. In this state, successful circulation is dependent on good ventricular function and low pulmonary vascular resistance (PVR). Venous return and blood flow to the lungs is augmented by negative intrathoracic pressure from the patient's own active inspiration. It is vital to understand the physiology of these three stages of palliation, since children may present for non-cardiac surgery at any point in their palliation process and need carefully planned anaesthesia which is tailored to their current situation.

### *Stage one: Systemic artery to pulmonary artery shunt or pulmonary artery band*

The aim of this initial stage of palliation is to alleviate hypoxia by providing adequate oxygen delivery to the tissues, whilst also preventing excessive pulmonary blood flow, which would raise the afterload on the struggling ventricle. Immediately after birth and diagnosis of the pathology, a higher PVR is present, which persists for the first few weeks of life. Non-pulsatile venous blood flow will not provide adequate pulmonary perfusion and a systemic artery-to-pulmonary artery Blalock–Taussig (BT) shunt is required. Although the "Classic" BT-shunt consisted of an end-to-side anastomosis, a synthetic conduit is now used to generate a restrictive shunt between a systemic artery (e.g. subclavian artery) and a proximal pulmonary artery.[8] Blood at systemic pressure then passes from the aorta into the subclavian artery, through this restrictive 3–4 mm conduit into the lungs,[8] where it is oxygenated and enters the ventricle to be returned to the systemic circulation. The increase in pulmonary blood flow causes the pulmonary vasculature to grow, which is essential for a successful palliation. Patients who suffer from a hypoplastic left heart may have an alternative procedure, entailing a conduit rather placed between the right ventricle and left pulmonary artery.[8] These children then have blood flow from the right ventricle entering the pulmonary and systemic circulations in parallel.[8] This may result in a higher systemic diastolic pressure, as there is no aorto-pulmonary run-off, so there might be better perfusion of the coronary arteries and systemic capillary beds. In the case of excessive pulmonary artery blood flow, a pulmonary artery band is often applied to protect the pulmonary vasculature by limiting blood flow.

This first stage of palliation is the most fragile, as the single ventricle is responsible for both systemic and pulmonary blood flow, and the ratio of cardiac output delivered to each is determined by the relative vascular resistances.[1] The arterial oxygen saturation for these individuals depends on the specific underlying condition, but if there is a state of complete mixing (i.e. a 50:50 split of blood flow between pulmonary and systemic circulations), a saturation of around 80% is expected. It is best to postpone elective non-cardiac surgery until after stage two palliation, as the anaesthetic risk is highest before the bidirectional cavo-pulmonary connection (Glenn operation) has been performed.[1]

### *Stage two: Superior vena cava to pulmonary artery connection*

Because the PVR continues to decrease over the first few months after birth, pulmonary blood flow increases. If this high circulation through the lungs continues, the pulmonary vascular tone will increase, leading to pulmonary vascular hypertrophy and pulmonary hypertension. Therefore, stage two of patient palliation should be considered by three to six months of age. This operation has traditionally been named the "bi-directional Glenn procedure" or "hemi-Fontan procedure". It involves the anastomosis of the SVC to the right pulmonary artery. This procedure can be done with or without cardio-pulmonary bypass (CPB). Blood flows from the SVC into the right pulmonary artery and then into the left and right lungs (i.e. bi-directional Glenn procedure). This blood is now oxygenated and enters a common atrium that is formed by an atrial septal defect (congenital or

surgically created). Deoxygenated blood from the IVC mixes with the oxygenated blood from the lungs in the common atrium and then enters the systemic circulation. The mixing results in systemic saturations of approximately 80–85%.[1,8]

This procedure can only be done once the pulmonary arteries have grown sufficiently,[8,9] and the PVR has decreased adequately after the neonatal period, to allow non-pulsatile flow from the SVC directly into the lungs via the right pulmonary artery. Intra-pulmonary arterio-venous shunts can develop in up to 25% of patients after this procedure. This is due to some known and other unknown factors, including lack of inhibition of angiogenesis by hepatic factors which, in normal physiology, enter the lungs via the IVC.[10] This can worsen cyanosis, with increasing severity over time. Some of these shunts will regress once stage three of palliation is completed. Others persist and contribute to a failure of palliation.[10]

### *Stage three: Completion of the univentricular heart/ Fontan procedure (IVC to pulmonary artery connection)*

This final stage of the palliative surgery is usually performed between one to five years of age, to allow the pulmonary arteries to grow and the PVR to decrease further.[8] However, the philosophy of some units is to postpone the Fontan procedure to a later stage, if the particular physiology of the patient allows for this. The IVC blood flow now also enters the pulmonary circulation, usually via a conduit. A small fenestration (hole between the conduit and the systemic atrium) may be created, which allows a "pop-off valve", to release pressure in the caval system, should it increase excessively.[1] In other words, it allows cardiac output to be maintained at the cost of slight desaturation due to the right to left shunt. It is critical that the anaesthesiologist is aware of the presence of such a fenestration, since an intraoperative increase of PVR may result in a shunt of deoxygenated blood into the systemic circulation, causing acute desaturation.

Over the last decade, some centres have moved from the above stage one strategy, to using a hybrid approach for the hypoplastic heart. The Giessen approach performs surgical bilateral pulmonary artery banding in addition to percutaneous stenting of the ductus arteriosus. This hybrid technique may be better tolerated in small neonates with brittle physiology.

## Complications of the univentricular heart

### *Complications of the pathophysiology*

It is useful to employ an anatomical approach as a cognitive aid for assessing these patients (Figure 2). This will direct history-taking and clinical examination.

Children with UH may have neurological deficits from multiple factors, including thrombotic events, chronic hypoxaemia, hyper-viscosity syndrome, and cerebral abscess.[11,12] They may also exhibit developmental delay due to multiple episodes of CPB and chronic illness. Plastic bronchitis is a rare condition which is described in patients who have undergone a Fontan procedure. The estimated prevalence is 4% of patients after TCPC.[13] Post-Fontan procedure, venous blood from the SVC as well as now from the IVC is re-directed towards the pulmonary circulation, which increases pulmonary pressures. This causes protein and lipid molecules from the lymphatics to leak into the bronchopulmonary tree. Tubular bronchial casts form, which

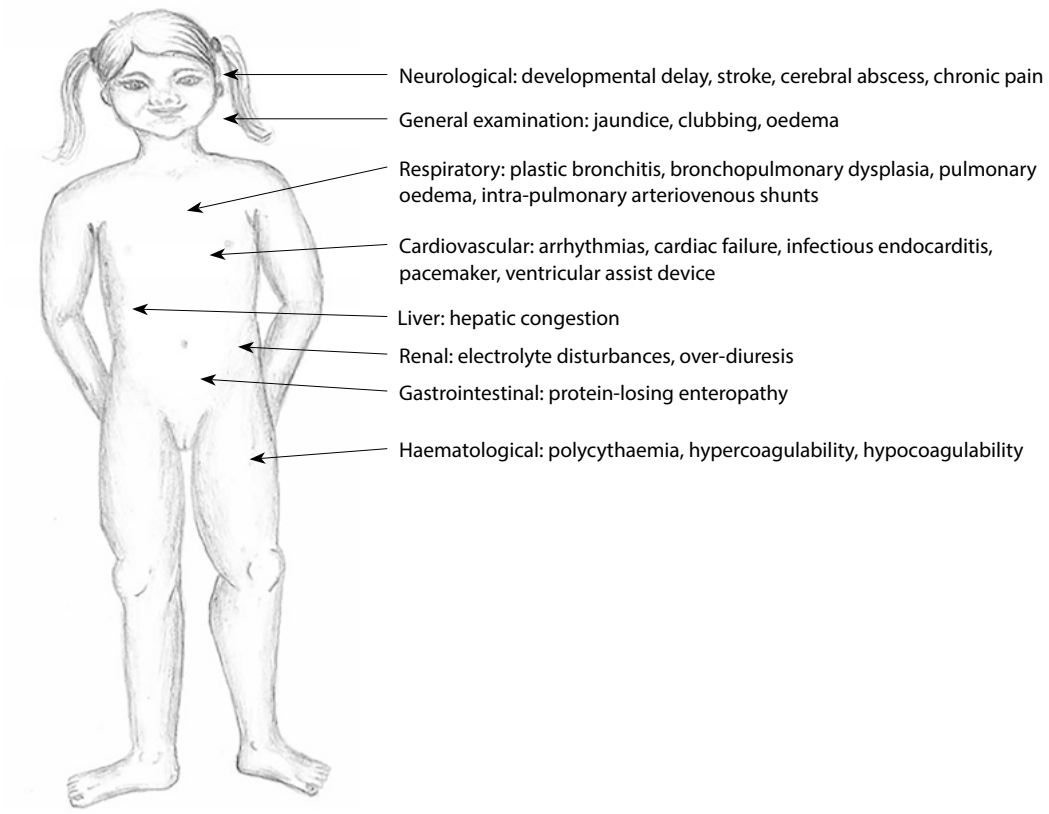



**Figure 2:** Anatomical approach as a cognitive aid for assessing children with UH

may cause airway obstruction soon after surgery. This condition has a long-term mortality of up to 30% after CHD repair.[11]

Current literature suggests that preterm babies are more likely to have CHD, and that fetuses with CHD are more likely to be born preterm.[14] Compounded by the dysfunctional cardiovascular system, this results in a higher requirement for prolonged positive pressure ventilation. It is therefore important to identify symptoms and clinical signs of bronchopulmonary dysplasia.

Cyanosis may be present due to worsening hypoxaemia, for reasons including poor ventricular function, shunt through the fenestration, worsening pulmonary hypertension, abnormal pulmonary arterial venous connections, and co-existing pulmonary disease.[11]

It is important to be aware that a child with UH may also have varying degrees of diastolic as well as systolic dysfunction, since this occurs in up to 50% of cases.[11] Diastolic dysfunction worsens with age, as it is related to hypertrophy and occasionally fibrosis of the myocardium, with uncoordinated relaxation.[11] Pacemaker considerations should be borne in mind for children with sinus node dysfunction (present in almost 60% of patients), and atrioventricular heart blocks precipitated by prior cardiac surgery.[11]

Protein-losing enteropathy (PLE) can occur, possibly due to high systemic venous pressure resulting in distension of the lymphatics, allowing loss of protein into the gut. Although this only occurs in about 4% of children with UH, it has a high mortality of 60%.[11] This condition should be suspected in children with hypoalbuminaemia, pericardial or pleural effusions, ascites, or pedal oedema.[12]

Hepatic congestion can also be a problem related to increased central venous pressure. It can present with cholestatic jaundice or coagulopathy, and if persistent can lead to liver fibrosis and failure.[11] Although hypercoagulability and hypocoagulability are both more prevalent in patients with the Fontan physiology, the balance may be shifted towards a hypercoagulable state. In combination with stagnation of blood flow, this makes thrombosis a common problem in UH.[11]

### *The long-term complications of surgery*

Arrhythmias are common due to the surgical manipulation of anatomy involved in cardiac conduction pathways.[15] Frequent episodes of atrial tachyarrhythmias may occur, the incidence increasing dramatically with time after the repair.[11] These are usually managed via trans-catheter ablation. Chronic pain post sternotomy/thoracotomy is a concern that is easily overlooked in these children.[16] They may be living with chronic pain and not expressing it adequately, depending on their age. Symptoms and signs of chronic pain should be identified, to enable an adequate perioperative analgesia plan, and ensure referral to a chronic pain clinic. Any active infection, including infectious endocarditis, should be diagnosed during the preoperative assessment.

### *Complications of medical management*

Complications of medical management are mostly related to prescribed medication. It is important to assess whether the medication and doses are appropriate, and identify electrolyte disturbances, dehydration due to over-diuresis, and hypotensive episodes.

## Practical approach to anaesthesia in a child with a univentricular heart presenting for non-cardiac surgery

### *Preoperative management: "the four Ps"*

Preoperative management can be divided into four categories, namely: Patient, Pathology, Procedure and Preoperative preparation. Routine assessment should be performed, along with these below-mentioned points pertinent to a case of a child with a UH.

#### *Patient*

- Assess for presence of recent respiratory tract infection, infective endocarditis and other infections.
- Look for evidence of cardiac failure and arrhythmias.
- Check coagulation status, including intrinsic coagulopathies, the presence of a thrombus, and anticoagulant medication.
- Assess hydration status – these children tolerate dehydration very poorly because they rely on adequate preload.
- Review investigations: baseline haemoglobin, white cell count (occult infection), serum albumin (may indicate PLE), liver and renal function. A recent echocardiogram is useful to obtain pulmonary artery pressure measurements, and parameters relating to systolic and diastolic function.[9]

#### *Pathology*

- Understand the pathology and associated syndromes/comorbidities.
- Check the stage of palliation that the child has received, and presence of a fenestration.
- Actively identify the previously-mentioned complications of the child's prior surgery, medical therapy or complications of the pathophysiology of a UH, in discussion with their cardiologist.

#### *Procedure*

- Carefully consider the scheduled operation, and the likely effect of the procedure upon the physiology, e.g. laparoscopic surgery, requirement for intraoperative positions such as anti-Trendelenburg.
- Classify the surgery as major, minor or intermediate surgery, and consider how this impacts the perioperative anaesthesia plan, the level of postoperative care, and the overall risk.
- Plan the timing of surgery to optimise human factors and intensive care unit (ICU) availability – often best first on the morning list.

*Preoperative preparation*

- Anxiolytics may be required in children who have had several surgical procedures, and supplemental oxygen should be considered after such premedication.[17]
- In order to ensure hydration, promote the drinking of clear fluids until one to two hours before surgery.[18]
- Obtain informed consent regarding the placement of invasive lines and use of transoesophageal echocardiography, and admission to ICU if necessary.
- Plan the perioperative medication regimen, depending upon the surgery envisaged, and the current status of the patient.
- Management of perioperative anticoagulation should be addressed in consultation with the surgeon, anaesthesiologist, cardiologist and intensive care team.

### *Intraoperative management*

We have chosen to use an "ABCDEF" approach to intraoperative management. These goals may be achieved in several ways, depending on the preference of the anaesthesiologist and the patient, and available resources.

*A and B: Airway and Breathing*

- **Careful airway control**
  An inhalational induction can be performed, with careful avoidance of any airway obstruction. Multiple intubation attempts with resultant hypercarbia or hypoxia are detrimental and lead to pulmonary hypertension.
- **Positive intra-thoracic pressure can impede venous return**
  Mean airway pressure has a near-linear inverse relationship with cardiac output in a child with UH.[19] The ideal scenario is to maintain spontaneous respiration. If not possible or practical, then the ventilator should be set in such a way as to minimise mean airway pressures.[20] This can be achieved by ventilating with tidal volumes 5–6 ml/kg, a short inspiratory time, low-normal respiratory rate and low positive end-expiratory pressure (PEEP), just adequate to prevent atelectasis and achieve oxygenation. The patient should ideally be extubated soon after completion of the surgery.
- **Maintain low pulmonary vascular resistance**
  Avoid triggers that may raise pulmonary arterial pressures, with the subsequent negative impact on venous return. Ensure a fine balance with adequate ventilation to maintain the arterial partial pressure of carbon dioxide ($PaCO_2$) within normal range (around 40 mmHg).[21] Ventilatory targets would also include targeting arterial oxygen tension of more than 82 mmHg.[21] Pain, anxiety, acidosis, hypovolaemia, use of vasoconstrictors and light level of anaesthesia may all cause pulmonary arterial hypertension and should be actively avoided.

*C: Circulation*

- **Understand the physiology**
  In a UH and after stage one surgery with a systemic-to-pulmonary artery shunt, the entire cardiac output is distributed from a single ventricular stroke volume to both the systemic and pulmonary circulations. The respective vascular resistances of the pulmonary and the peripheral circulation dictate this relative distribution of flow. In single ventricle physiology, there is an abnormal physiological response to manipulation of preload, afterload, and contractility. The pulmonary flow is dependent on the preload and relies on a low PVR.
- **Maintain preload**
  A patient with Fontan physiology is very sensitive to changes in preload. The patient should be adequately hydrated and the anti-Trendelenburg position should be avoided. Leg raising during induction of anaesthesia can limit the associated decrease in cardiac output.[9] In case of intraoperative hypotension, a fluid challenge is in principle preferable to inotropic support, because a high venous pressure is necessary to assure adequate cardiac output.
- **Maintain ventricular function**
  Avoid drugs with negative inotropic effects.
- **Maintain afterload**
  Maintain afterload to ensure good perfusion of the single ventricle. However, it is also important not to increase afterload by excessive peripheral vasoconstriction, as this may precipitate ventricular failure.
- **Maintain sinus rhythm**
  A defibrillator and anti-arrhythmic drugs should be readily available. In high-risk cases, external defibrillator pads should be in place.
- **Recognise the risk of haemorrhage**
  These patients may be at increased risk of bleeding due to their raised systemic venous pressures, coupled with coagulation deficits.[1] Since blood loss is poorly tolerated, blood conservation strategies should be employed, and a more liberal transfusion trigger of a haemoglobin of 10 g/dl may be preferred.[9]
- **Consider the use of invasive lines and monitoring**
  The decision to place an arterial line or central venous line will be dictated by the predicted insult caused by the surgical procedure, and patient factors such as ventricular function. Frequent arterial sampling can detect problems early on and allow for modifications of anaesthetic technique. A central line is useful if there is inadequate peripheral intravenous access and allows monitoring of trends in central venous pressures. Central access also allows easy administration of vasoactive drugs. If a Glenn shunt is in place (stage two), the central venous line pressure reading will represent the pulmonary artery pressure, and any drug administration will be directly into the pulmonary vasculature. Interfering with flow in the SVC may lead to obstruction or thrombosis of the Glenn shunt. Due to multiple previous cardiac catheterisations, access to femoral veins may be limited. The use of ultrasound to facilitate line placement can serve to protect veins from injury and avoid complications.

*D: Drugs*

- Good analgesia should be provided, and special attention paid to children with chronic pain.

- Muscle relaxation should be adequately reversed at the end of the procedure, so that respiratory function is not compromised after awakening. Sugammadex may be a preferable reversal agent if available, due to its superiority over neostigmine and glycopyrrolate with regards to haemodynamic effects.[22]
- Cardiac drugs such as inotropes, vasopressors, anti-arrhythmics and pulmonary vasodilators may be necessary in more severe cases and should be carefully titrated to effect, preferably monitored by transoesophageal echocardiography. All vasopressors cause an increase in pulmonary as well as systemic vascular resistance. The exception to this rule is vasopressin, which may be of use in these patients in certain circumstances, e.g. septic shock.[23] Of the commonly used vasopressors, vasopressin has the least impact on raising PVR and works directly on V1 receptors to increase systemic vascular resistance, whilst sparing the pulmonary circulation.
- Patients with complex CHD have a much higher incidence of infective endocarditis (IE) than the general adult population. Any clinical scenarios involving low- or turbulent flow, the use of prosthetic material, or invasive procedures, increase the risk.[24] It is therefore recommended that a prophylactic antimicrobial agent is administered in patients with a palliative shunt or conduit.[24]

*E: Exposure*

- Maintain normothermia: hyperthermia increases oxygen consumption and heart rate (which decreases diastolic filling time in children with diastolic dysfunction) and causes vasodilation. Hypothermia has several negative consequences, such as an increased incidence of infection, coagulation defects, and delayed awakening.[25] The sympathetic response triggered when deviating from normothermia, may also have the detrimental effect of increasing PVR.

*F: Focus on anaesthesia principles*

- Intravenous induction of anaesthesia should involve careful and slow titration of drugs.
- Regional anaesthesia techniques can cause widespread arteriolar and venodilation, causing a decrease in afterload and preload. Risk versus benefit must be carefully considered for each patient, and when employing a regional technique, steps should be taken to prevent these side effects. In many cases, these patients are on anticoagulants, which will preclude the use of neuraxial techniques. Regional nerve blocks and local anaesthesia can be an attractive alternative to general anaesthesia in certain situations, which can minimise physiological effects of general anaesthesia and mechanical ventilation.
- Air embolism is a significant risk in these children if a fenestration is present; therefore, meticulous attention should be paid to avoid the introduction of air bubbles in intravenous lines and syringes.

### *Postoperative management*

The largest study to date on patients with univentricular physiology undergoing non-cardiac surgery showed a complication rate of 31%.[2] Preoperative ejection fraction was the main predictor for the occurrence of complications, monitored care length of stay, and hospital length of stay.[2] Regarding day case surgery, if a small procedure is planned in an otherwise well child with a UH, one can consider same-day discharge. However, if there are any concerns, the child should be admitted for overnight observation.[8] Where the cardiac risk exceeds the surgical risk of the patient, it is necessary to monitor progress in an environment in which staff are familiar with such complex patients.

All major surgery cases require a postoperative ICU bed,[8] since the stress response and the changes in haemodynamics in such cases pose a risk to these patients with their fragile physiological reserves. The oxygenation and ventilation strategy should consider the stage of surgery and prioritise maintenance of normal lung function.[21] Some children may be able to be extubated at the end of the procedure and the use of lung ultrasound to assess for pleural effusion or significant atelectasis can assist in decision-making. Close monitoring postoperatively is essential.

## Novel techniques and developments in the management of anaesthesia for children with a univentricular heart

### *Near-infrared spectroscopy*

Considerable controversy exists amongst anaesthetists concerning the clinical value of near-infrared spectroscopy (NIRS) in paediatrics. A recent study has shown a strong correlation between cerebral regional oxygen saturation and cardiac function during low cardiac output states in neonates.[26] In 2017, Hoffman et al. studied a population of children with hypoplastic heart syndrome, and found that NIRS measurements in the early postoperative period were strongly predictive of mortality.[27] They concluded that using NIRS postoperatively as a monitor to assist in goal-directed interventions may be warranted in this population.[27]

A systematic review of the evidence for the value of NIRS as a monitor in critically ill children showed Class II, level B evidence to support the conclusion that NIRS offers a favourable risk-benefit profile and can be effective and beneficial as a haemodynamic monitor for the care of critically ill patients.[28]

Since these children present for surgery with a low cardiac output state, the use of a non-invasive, rapidly responding monitor such as NIRS may be very useful in alerting the anaesthetist to changes in oxygenation and cardiac output.

### *Transoesophageal echocardiography*

Transoesophageal echocardiography (TOE) appeared in paediatric practice in the late 1980s and has since become an indispensable monitor to the paediatric cardiac anaesthetist and

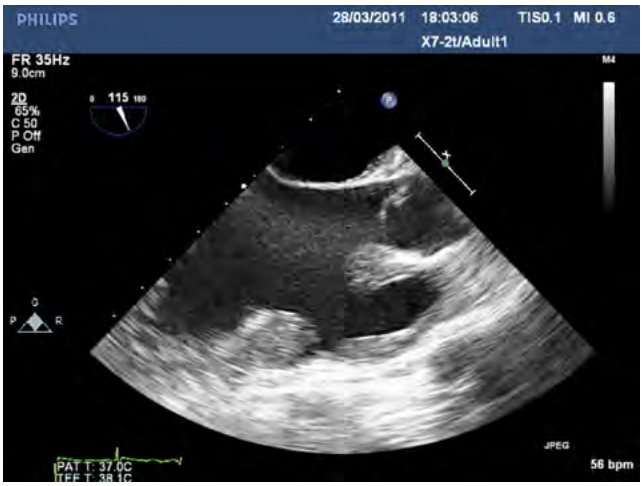
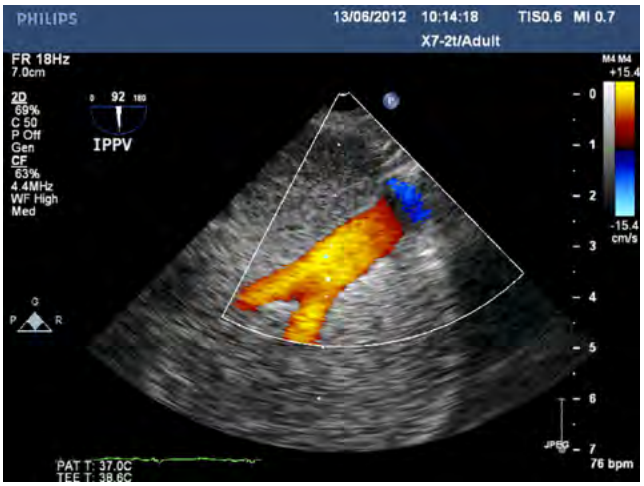



Figure 3: A 20-year-old female with tricuspid atresia, rudimentary RV, large VSD, atrial septectomy. The patient had a previous lateral tunnel Fontan performed, now planned for a non-cardiac procedure, hysterectomy. This 2D midoesophageal echocardiography image of the left ventricular outflow tract clearly demonstrates the VSD.



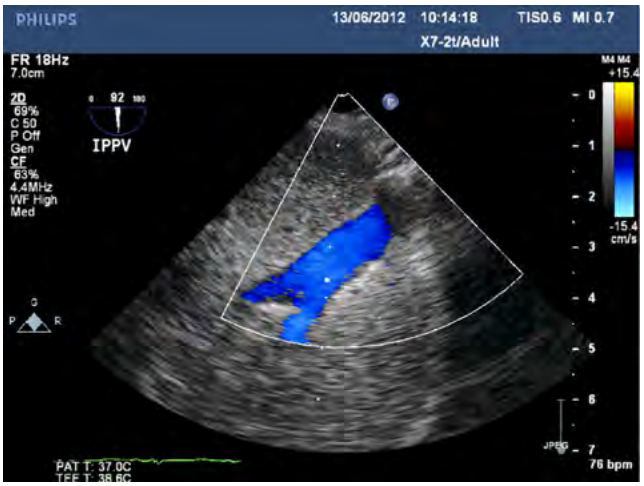




Figure 4.1 and 4.2: A 28-year-old male with double inlet-double outlet single ventricle, severe infundibular- and pulmonary valve stenosis. This patient had a previous lateral tunnel Fontan, and is experiencing cardiac failure and arrhythmias. He now presents for an electrophysiological study and radiofrequency ablation in the catheter laboratory. These 2D colour-flow Doppler images of the inferior vena cava demonstrate impaired venous return through the yellow-red (Figure 4.1 towards) and blue (Figure 4.2 away) colour of bidirectional flow within one cardiac cycle.

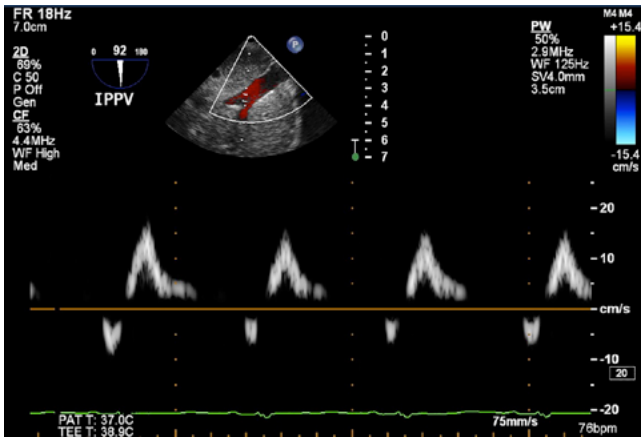



Figure 5: The same patient as in figure 9. This still continuous wave Doppler (CWD) image demonstrates impaired venous return velocities and cyclic episodes of flow reversal.

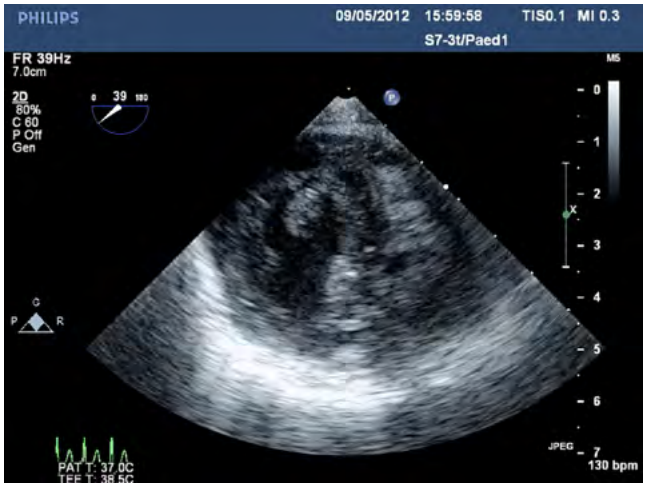



Figure 6: A 5-year-old male with hypoplastic left heart syndrome and atrial septal defect, no ventricular septal defect (VSD), presently with Glen shunt, now presenting for a completion of Fontan circulation procedure through an extracardiac conduit. The TOE examination assists intraoperative haemodynamic decision-making. This 2D transgastric short axis view demonstrates a rudimentary left ventricle, and preserved systolic function of a hypertrophic, right systemic ventricle.

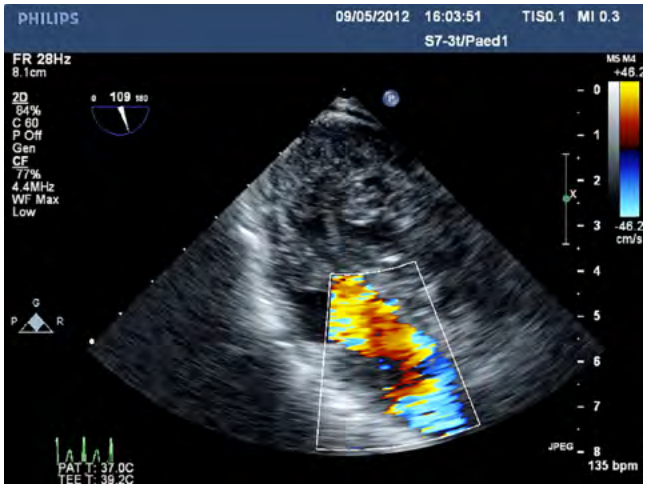



Figure 7: The same patient as in Figure 3. This 2D colour-flow Doppler image of the right ventricular outflow tract demonstrates good flow across this patient's pulmonary-systemic valve.

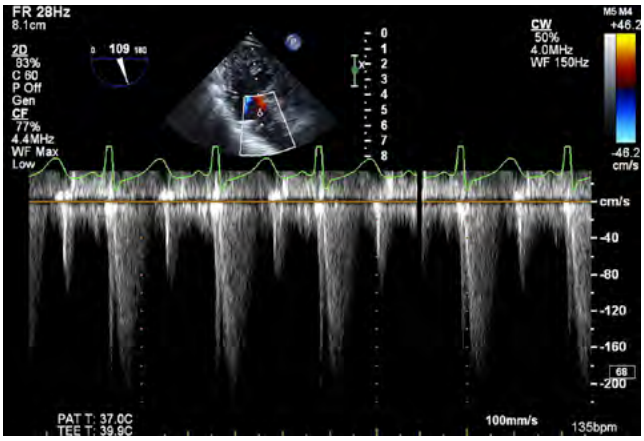



**Figure 8:** The same patient as in Figure 6. This still CWD image of the right ventricular outflow tract helps to quantify the good flow and cardiac output across this patient's pulmonary-systemic valve.

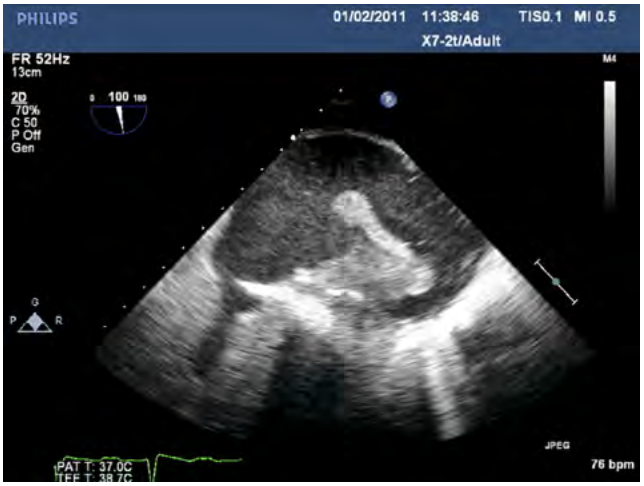



**Figure 9:** A 36-year-old male with tricuspid atresia, rudimentary right ventricle, large ventricular septal defect, transposition of the great arteries, severe pulmonary stenosis and a previous modified atrio-pulmonary Fontan. Now presenting for a laparoscopic mesenteric lymph node biopsy and hernia repair. This 2D TOE image of the right atrium demonstrates a large organised thrombus and the spontaneous echo contrast of sluggish flow.

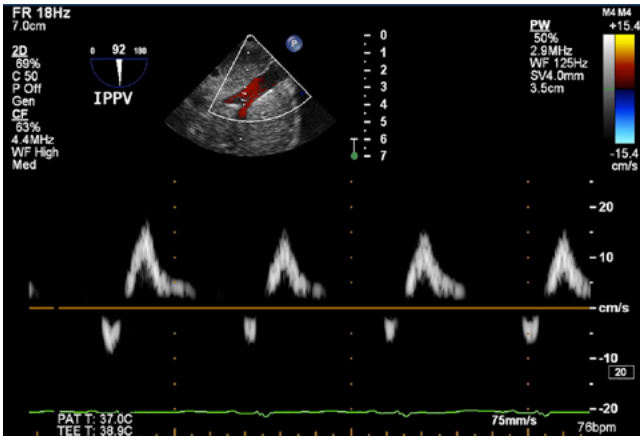



**Figure 10:** The same patient as in Figure 9. This 3D TOE image shows the same thrombus in 3 dimensions.

surgeon. In most paediatric cardiac surgery centres, the use of intraoperative TOE is now considered essential for congenital heart surgery. It has also developed into a helpful tool for the anaesthetist in non-cardiac surgery (Videos 1, 2 and Figures 3, 4.1, 4.2, 5). It is essential that the physician performing the intraoperative TOE examination is experienced and has a proper understanding of congenital cardiac pathology. Since smaller paediatric probes were introduced to the intraoperative congenital cardiac surgical arena in the early 1990s, TOE has become not only a diagnostic tool, but also an important monitor.[29]

Intraoperative echocardiography provides assessment of cardiac output (CO), biventricular function, pulmonary artery pressure (using the tricuspid regurgitant jet), intravascular volume status, and the degree of intracardiac shunting[23] (Videos 3, 4 and Figures 6, 7, 8). In non-cardiac surgery, TOE assists intraoperatively with the haemodynamic "balancing" of the systemic and pulmonary circulations in children who have unrepaired anatomy or stage one palliation of their univentricular physiology. Intravascular volume status can be optimised using TOE as a dynamic monitor of fluid responsiveness, which is integral to the anaesthesia management of children with UHs undergoing surgery with expected significant blood loss or fluid shifts. In the perioperative setting in patients with univentricular cardiac anatomy, TOE is superior to transthoracic echocardiography in determining the presence of an intramural thrombus before cardioversion,[30] which is frequently present in these patients due to atrial flutter or fibrillation[31] (Videos 5, 6 and Figures 9, 10).

Paediatric multiplane TOE probes can safely be used in children who weigh 4 kg or more. The probe should be inserted with great care in small babies, to avoid oesophageal damage. Serious complications during paediatric TOE are rare. Stevenson assessed TOE complications in this specific group of patients and found a complication rate of 3.2%.[32] Airway complications, compression of vascular structures, and haemodynamic instability were the most frequent; therefore, great care is required when performing TOE in paediatric patients.[32] The possibility of compression of the airway and major vessels by the probe, leading to desaturation and haemodynamic compromise, is therefore a clinically significant problem when using TOE in small children.[33]

### *Ventricular assist devices*

Mechanical circulatory support as a bridge to recovery is a major component of cardiogenic shock management in the adult population, and its use in CHD is emerging. Extracorporeal membrane oxygenation (ECMO) and percutaneous ventricular assist devices (VAD) may provide partial left ventricular and right ventricular support, but experience in complex CHD, especially in children, is lacking.[21] There has been considerable development in the mechanics of VAD in the past decade. A VAD system does not include an oxygenator, therefore, it assists only one or both ventricles. ECMO provides biventricular and pulmonary assistance. There have been several case studies of the use of VAD in the scenario of the failing UH,[34-36] and in future many children may present for non-cardiac surgery with

a device in situ. Employing these devices in paediatrics has lagged behind their use in the adult population but has been steadily gaining momentum. Such a child is a challenging case, with multiple factors such as the underlying univentricular pathology, functioning of the VAD and the associated obligatory anticoagulation, as well as the current surgical problem. A thorough understanding of the mechanics of the VAD is essential when anaesthetising one of these patients, who should ideally be managed in a centre that is familiar with such devices.[37] Intraoperative monitoring can be problematic due to the lack of significant pulsatile flow, and pulse oximeters may not function well.[37] NIRS can be a useful adjunct in this instance, to facilitate the maintenance of baseline values. Non-invasive blood pressure cuffs may malfunction, and an ultrasound-guided placement of an arterial line should be strongly considered before induction of anaesthesia, to enable continuous monitoring of blood pressure, as well as oxygenation and acid-base status. Since these children may not respond to pain or inadequate depth of anaesthesia with changes in heart rate and blood pressure, a monitor of depth of anaesthesia should be considered.[37]

## Conclusion

When faced with a child with a UH presenting for non-cardiac surgery, thorough planning and preparation of drugs and equipment are vital to the success of the case. Understanding the pathophysiology and related complications is crucial since these directly impact perioperative management. Children are now having corrective surgery at an earlier age and lower weight, and operative mortality rates have improved. The use of novel technologies can assist in optimising physiology and in preventing decompensation of the fragile haemodynamics of these patients. The current era of VAD will result in prolonging survival in many more patients with their univentricular physiology and will pose a unique set of challenges to the anaesthetist. With the ever-increasing number and complexity of technological advances, it will be our role to ensure that an understanding of these new techniques is combined with a thorough grasp of basic sciences.

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### *ORCID*

L Veronese https://orcid.org/0000-0002-4069-2991
J Swanevelder https://orcid.org/0000-0001-6986-4470
A Brooks https://orcid.org/0000-0002-6034-6105

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