Although results of repair or palliation of congenital heart disease have steadily improved in the last two decades, anaesthesia for paediatric cardiac surgery remains a sub-specialty with a number of complex high-risk procedures. The anaesthesiologist is confronted with a wide range of operations, with many of them done infrequently. Simple analysis of mortality rates and complications may be misleading when other factors are not considered, cases complexity in particular. Surgical modifications and management improvements have led to increased survival after first-stage Norwood palliation of the hypoplastic left heart syndrome. However, mortality still exceeds that of most reconstructive procedures for complex congenital heart defects. Peri-operative management and outcome of HLHS palliation varies dramatically from institution to institution [1]. Nevertheless, the anaesthesiologist has to know exactly the principles of the circulation in univentricular heart to ensure best care during the peri-operative period.

One of the primary goals of pre- and immediate postoperative management is maintenance of “balanced” systemic and pulmonary circulations. It is generally assumed, that pulmonary to systemic blood flow ratio (Qp/Qs) close to 1 is associated with the best clinical presentation for neonates with HLHS [2]. As pulmonary vascular resistance can change, sometimes widely, after induction of anaesthesia for stage I palliation, Qp/Qs will also vary, despite adequate management. Imbalances of Qp/Qs and limited myocardial reserve of the morphological right ventricle compounded with inadequate systemic oxygen delivery and end-organ perfusion accounts for most of early mortality. To optimize systemic oxygen delivery we, like some centres, have adopted continuous monitoring of systemic venous oxygen saturations (SvO₂) with or without oximetric catheters to allow continuous Qp/Qs measurements [3]. For induction of anaesthesia it is important to keep the circulation as it was before induction if the patient was in stable condition. That means avoiding all anaesthetics that will affect the peripheral vascular resistance. In our centre we use only high dose fentanyl and pancuronium for induction of anaesthesia. We never use inhalational agents in HLHS before initiation of bypass. After endotracheal intubation the FiO₂ was adjusted to 0.21. During the whole induction period it is important to maintain the body temperature of the infant to ensure that temperature induced vasoconstriction is avoided. Our current concept is to facilitate high-flow cardiopulmonary bypass to ensure maximum organ perfusion even when cooling down to 18°C core body temperature. Vasoconstriction is a natural reaction of the body to cold, causing the blood vessels to contract. Such a reaction would lead to a clear rise in blood pressure on cardiopulmonary bypass (CPB). Since the blood pressure is regulated in the arteriolar loop of the capillary system, there is a marked increase in blood pressure and a passage of free water into the surrounding tissue which, in turn, may lead to the intra-operative development of oedema. Such „mechanical“ development of oedema is possibly misconstrued as a capillary leak syndrome. For this reason, many heart centres reduce the flow on cardiopulmonary bypass. A reduction in the blood flow may lead to an inadequate supply of oxygen to the tissue, however, despite the decrease in body temperature. This oxygen debt is detected during rewarming as a measurable increase in acidic metabolic end products (e.g. lactate).
Advocates of such a method claim that a reduced supply under hypothermic conditions can be tolerated by the body in a similar way to hibernation. Another approach is to counteract the natural reaction of the body and use medication to compensate for the cold-induced vasoconstriction. In such a way, the surgeon has perfect conditions in which to operate while all the organs remain perfused even during hypothermia. In order to maintain the blood pressure, or rather organ perfusion, during vasodilation, the flow on CPB must be increased (up to 20% of baseline). In a recent study we have published results of the “high-flow, low resistance technique [4]. The most critical period is termination of CPB. We routinely use continuous infusion of dopamine 4-6 µg kg⁻¹ min⁻¹ as inotropic support only. All other therapy was guided by determination of the Qp/Qs ratio and the result of transoesophageal echocardiographic examination in infants down to 3 kg/BW.

**Balanced circulations with low total cardiac output**

SaO₂ of 75–80% and SvO₂ of 40–55% with corresponding Qp/Qs below 1: If lactate levels are rising, inadequate total cardiac output and low systemic oxygen delivery is assumed. Since Qp/Qs is already in optimal range, measures to improve systemic oxygen delivery concentrate on elevation of haematocrit and on augmentation of inotropic support (norepinephrine starting with 0.05 µg kg⁻¹ min⁻¹ up to maximum of 0.1 0.05 µg kg⁻¹ min⁻¹). Before further decision transoesophageal echocardiographic examination is done to exclude anatomical problems. If aerobic conditions are still not achieved, ECMO support should be instituted.

**Unbalanced circulations with high systemic oxygen delivery**

SaO₂ of 75–80% and SvO₂ above 60% with corresponding Qp/Qs ≤1: with rising lactate levels, measures that increase systemic vascular resistance or decrease pulmonary vascular resistance should be initiated to adjust Qp/Qs between 1 and 2.

**Unbalanced circulations with pulmonary overcirculation**

SaO₂ of 75–80% and SvO₂ below 40% with corresponding Qp/Qs ≥2: with rising lactate levels, measures that decrease systemic vascular resistance or increase pulmonary vascular resistance should be commenced to adjust Qp/Qs between 1 and 2

Fig. 1. Qp/Qs ratio in survivors and non-survivors: calculated Qp/Qs ratio was higher in survivors than in non-survivors [3].
In every case the time course of lactate is repeatedly measured to determine perfusion deficits early. We are aware that lactate is an unspecific parameter for estimation of circulation, but we know from our own as well as studies from other groups that an increase in lactate in the early postoperative period is a clear indicator to look carefully for the reason of that increase of lactate.

Conclusion

Anesthetic management in patients with HLHS is still challenging. Basic principle in planning anaesthesia is to maintain “balanced” systemic and pulmonary circulations. During the early post-bypass period the drug therapy as well as respirator settings should be guided by the determination of the $\text{Qp/Qs}$ ratio, as well as close measurements of haemodynamic and laboratory parameters such as lactate, $\text{SaO}_2$, $\text{SvO}_2$.

References