# IAPA Advisory on Anaesthetic Management of the Neonate with Congenital Heart Disease for Non-Cardiac Surgery

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## Introduction

Neonates present a unique challenge for the anaesthesiologist due to various physiological and pathological factors. The presence of congenital heart disease (CHD) further complicates the administration of safe anaesthesia in this age group. The focus of this review is on how to identify neonates with congenital heart disease scheduled for non- cardiac surgery and plan appropriate and safe anaesthesia techniques for them. The prevalence of significant congenital heart disease varies from 3.1 to 8/1000 live births.[1] The incidence of neonates requiring surgical intervention in the first month of life is 4.6/1000 as per data from a single center.[2,3] The common indications for surgery in 80% of neonates are congenital anomalies especially those causing gastrointestinal obstruction and neural tube defects.

## Identification of congenital heart disease in neonates

Prenatal identification of foetal cardiac abnormalities has improved with advances in imaging such as foetal echocardiography, 3D-4D ultrasonography and MRI. A mid-gestational 4-chamber echocardiography can detect >50% of critical cardiac malformations. Newer techniques for foetal cardiac catheterization and surgery may theoretically offer mortality benefit in certain conditions (*e.g.,* hypoplastic left heart), but these have not yet been optimized and validated.[4]

A delayed or missed diagnosis of critical CHD occurs in 7 per 100 000 live births. Regular screening of neonates for congenital heart disease has been advised. In neonates with anomalies with ductus-dependent circulation, closure of the ductus arteriosus and changes in the pulmonary vascular resistance, can lead to rapid clinical deterioration.[5] Clinical examination along and using a pulse oximeter help in early identification of congenital heart disease in a neonate.[6]

The presence of congenital heart defects should be suspected in neonate whose mother is diabetic. Neonates with gross physical congenital anomalies, decreased/absent femoral pulses, heart murmurs, cardiac arrhythmias and evidence of cardiomegaly in a chest X-ray need to be assessed by a paediatrician/neonatologist or paediatric cardiologist. A comprehensive echocardiographic examination may be required.[7]

## Neonatal Circulatory Physiology

### Normal or 'series' circulation

Two separate circuits of body circulation have been described, the systemic and pulmonary systems, which together in series. Some congenital heart diseases also have this type of circulation with shunting of blood between the systemic and pulmonary circulations via defects, atrial septal defect (ASD) and

ventricular septal defect (VSD)]. Flow across this shunt takes place down the pressure gradient. Left-to-right shunts increase the pulmonary blood flow (PBF) and potentially decrease systemic blood flow; right-to-left shunts divert deoxygenated blood into the systemic circulation, causing cyanosis and reduced PBF.

Changes in pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR) secondary to anaesthesia have their greatest effect in patients with large, unrestrictive defects. Infants with a large unrestricted defect, such as VSD may exhibit 'balanced' circulation physiology.

**Parallel or 'balanced' circulation**

In a balanced circulation, the pulmonary and systemic circulations are connected by a large ASD or VSD defect and the blood flow to the lungs and body is a 'balance' between SVR and PVR. This is due to the pulmonary and systemic circulations being in parallel, with flow in the two parallel circuits (systemic and pulmonary) varying, depending on the relative resistance in each circuit. Hence, the term 'balanced' circulation. Excessive pulmonary blood flow can cause pulmonary oedema, and poor systemic and insufficient flow can lead to profound cyanosis. Examples of children with 'balanced' circulation are infants with a large unrepaired ASD or VSD, a modified Blalock–Taussig (BT) shunt, truncus arteriosus, and hypoplastic left heart syndrome. It is very difficult to optimally manage these neonates, and surgical care in a tertiary care cardiac center is advised.

**Preoperative Assessment and Risk Stratification**

**Cardiac physiological assessment**: A focused examination to assess the presence of heart failure, pulmonary hypertension, arrhythmias and cyanosis can help identify neonates at high perioperative risk.[8,9]

**Heart failure:** Neonates with reduced cardiac function must be identified because they are at high risk of cardiac arrest (10%) and inotrope requirements (96%) during anaesthesia.[8,9] Clinical signs and symptoms of poor cardiac reserve and heart failure in neonates include; tachypnoea, tachycardia, sweating (*e.g.,* on feeding), failing to thrive, hepatomegaly and cool peripheries. Neonates with milder degrees of cardiac failure pose less risk. Anaesthetic induction time will be prolonged, a slow, titrated induction is needed to prevent excessive drug administration.

**Pulmonary hypertension:** Neonates with pulmonary artery pressures of more than 25 mmHg are at high risk and they should receive treatment in a tertiary center. Treatment to reduce pulmonary vascular pressures include providing 100% oxygen, transiently inhaled nitric oxide, intravenous (IV) prostacyclin and inotropic support of the right ventricle may be required. Respiratory infections are poorly tolerated in such patients.

**Arrhythmia:** It is desirable to have a preoperative ECG in all neonates with CHD. Neonates with single ventricle physiology are particularly prone to arrhythmias (as high as 30%) leading to death. [8,9]

**Cyanosis:** Cyanotic neonates often have concurrent cardiac failure, pulmonary hypertension and arrhythmias.

**Focused clinical examination:** Should be performed to identify respiratory illness and congenital anomalies affecting airway management.

**Cardiac medications** should be continued before surgery. Some anaesthesiologists prefer to omit angiotensin converting enzyme (ACE) inhibitors based on adult literature, but evidence in paediatric population is lacking. Neonates with ductus dependent circulation who are on an infusion of prostaglandin should have the infusion continued.

**Risk stratification*:*** The range of congenital heart diseases and their implications make it difficult to propose a standard risk stratification to identify neonates at greater risk of perioperative mortality. Important factors include the complexity of the cardiac lesion, physiologic compensation and type of surgery. Categorization into high, intermediate and low risk procedures can help in the level of planning and complexity of monitoring and post-operative care as shown in *Table 1*.[9]

***Table 1:*** **Risk stratification for surgery**

| **High risk** | **Intermediate risk** | **Low risk** |
|---|---|---|
| Poorly compensated disease | Physiologically normal | Physiologically normal |
| Complex lesions (single ventricle or balanced circulation physiology, severe aortic stenosis, cardiomyopathy) | Simple lesions | Simple lesions |
| Major surgery (major fluid shifts, blood transfusion anticipated) | Major surgery | Minor surgery |
| Preterm neonates | Preterm neonates | Term neonates |
| Hospital stay >10 days | Hospital stay >10 days | Hospital stay <10 days |
| Emergency surgery | Emergency surgery | Elective surgery |

Given the requirement for specialist support and increased perioperative morbidity, an evidence-based approach suggests all high-risk neonates be anaesthetized where specialist facilities are available, including a dedicated post-operative care unit capable of handling sick neonates.

**Anaesthetic Management**

**Premedication:** Sedative premedication is usually avoided. Titrated doses may be considered to avoid distress, minimize oxygen consumption, and reduce the amount of induction agent required (to minimize reductions in SVR). Cyanotic children and those with heart failure will require monitoring of vital signs after administration of sedation and may require oxygen therapy to maintain oxygen saturations.

**Endocarditis prophylaxis**: Appropriate guidelines must be followed.[10]

**Fasting guidelines:** As for other neonates (1 h for clear fluids and 4 h for breast milk). Maintenance fluid requirements during prolonged fasting in babies with obstructive lesions and cyanotic disorders should be met by appropriate IV fluid therapy.

**Induction of anaesthesia:** Sevoflurane induction can be used in neonates with congenital heart diseases, however, if cardiac function is poor, prolonged exposure to high concentrations (8%) should be avoided. It would be prudent to obtain IV access prior to induction or have an assistant experienced in paediatric IV cannulation available at hand. The disadvantages of inhalation induction include peripheral vasodilatation and the risk of aspiration in the case of full stomach.[11]

Ketamine and propofol have been extensively evaluated in pediatric patients with CHD undergoing non-cardiac surgery. Propofol reduces SVR and mean arterial pressure (MAP) which may cause a clinically significant reduction in oxygen saturation, by increasing the shunt flow. Ketamine has a minimal effect on SVR, MAP, PVR, and pulmonary arterial pressure (PAP); and hence is the agent of choice when a reduction in SVR is not desirable or in the presence of pulmonary hypertension.[12]

Difficulties in airway management are likely to result in physiological imbalances and a swift control of the airway, endotracheal intubation and ventilation are advised.

**Monitoring:** Pulse oximetry, electrocardiography, noninvasive blood pressure monitoring an using appropriately sized cuff, core temperature monitoring, end tidal carbon dioxide estimation, fluid requirement need to be continuously monitored in all the children. Its useful to have two pulse oximeters, one for lower limb and one for upper limb to detect abnormalities in the peripheral circulation. Depending upon the complexity of the lesion, invasive arterial line monitoring should be established. This will not only facilitate beat to beat blood pressure monitoring but will also extrapolate pulse pressure variation. An arterial line can also be used for blood gas analysis. Alternatively, non-invasive Masimo® monitor can be used for multi wavelength pulse oximetry estimation and Pleth Variability Index (PVI).[13] If available, intraoperative echocardiography can be used in these patients.

**Maintenance of anaesthesia:** Isoflurane and sevoflurane are widely used for maintenance of anaesthesia. High dose opioids provide stable haemodynamics, but require mechanical ventilation and postoperative intensive care. Analgesia with opioids is commonly used. Milrinone and adrenaline infusions may be required if cardiac failure is present.

Adjustments in the fractional inspired oxygen concentration ($FiO_2$) need to be made depending on the pathophysiology of the cardiac disease. Hypoxia due to low $FiO_2$ can increase PVR and exposure to high $FiO_2$ can reduce PVR. Changes in PVR can result in a neonate with balanced circulation physiology becoming unstable. It is advisable to provide the lowest $FiO_2$ which will maintain an oxygen saturation of 95% in neonates with acyanotic heart disease and 75-85% in cyanotic neonates.[14] Mechanical ventilatory settings for neonates with balanced circulation should be adjusted to avoid pulmonary over inflation and pulmonary vasoconstriction or vasodilation. Positive end expiratory pressure (PEEP 4–6 $cmH_2O$), adjusting inspiratory pressures, respiratory rate, or tidal volumes to achieve an arterial carbon dioxide ($PaCO_2$) tension of 30-40 mmHg and avoiding respiratory alkalosis can help achieve these aims.[15] Nitrous oxide as an adjuvant during induction of anaesthesia is well tolerated by neonates with compensated heart disease. It is criticized for its effect of expansion of air embolism in the presence of shunt lesions. It is a mild myocardial depressant and can cause elevation of PVR, which is undesirable in pre-existing pulmonary hypertension.

Ensuring that the intravenous line is free of air bubbles, however minute, is especially important in neonates with ASDs because of the potential for the air bubble to cross from the right to the left heart and into systemic circulation, leading to paradoxical shunt.[16]

**Regional Anaesthesia**

Evidence for regional anaesthesia for neonates with congenital heart disease is limited. Safe use of spinal anaesthesia has been described for neonates with single ventricle physiology, premature neonates and neonates with decompensated and complex cardiac lesions. Minimal changes were noted

in haemodynamics and oxygen saturation.[17-20] Use of epidural analgesia has also been described to facilitate lower dose of anaesthetics and early extubation.[11] Despite the theoretical disadvantages in neonates with Tetralogy of Fallot (TOF) or atrio-ventricular septal defects, regional anaesthesia and analgesia maybe well tolerated, when sufficient expertise in performing the various techniques is available and correct drug dosages are followed.

## Conclusion

While evidence for the ideal management of neonates with congenital heart diseases undergoing non-cardiac surgery is minimal, data extrapolated from older children and adults and an understanding of cardiovascular physiology and pharmacology can guide our practice in the care of this vulnerable population. Planning and team work with a multi-disciplinary approach involving neonatologists, paediatric cardiologists and a competent surgical and anaesthetic team are the cornerstones to a successful outcome.

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