

Anaesthetic considerations in children with congenital heart disease undergoing non-cardiac surgery

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ABSTRACT

The objective of this article is to provide an updated and comprehensive review on current perioperative anaesthetic management of paediatric patients with congenital heart disease (CHD) coming for non-cardiac surgery. Search of terms such as "anaesthetic management," "congenital heart disease" and "non-cardiac surgery" was carried out in KKH eLibrary, PubMed, Medline and Google, focussing on significant current randomised control trials, case reports, review articles and editorials. Issues on how to tailor perioperative anaesthetic management on cases with left to right shunt, right to left shunt and complex heart disease are discussed in this article. Furthermore, the author also highlights special considerations such as pulmonary hypertension, neonates with CHD coming for extracardiac surgery and the role of regional anaesthesia in children with CHD undergoing non-cardiac operation.

Key words: Anaesthetic management, congenital heart disease, fontan physiology, left to right shunt, non-cardiac surgery, pulmonary hypertension, regional anaesthesia, right to left shunt

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INTRODUCTION

Congenital heart disease (CHD), still placed first among common birth defects, occurs approximately 1 in 125 live births. Thirty percent among these might require surgery during the first year of life due to extracardiac anomalies such as tracheoesophageal fistula, anorectal anomalies, cleft lip and palate, and renal and skeletal pathologies,^[1] and 85% of these CHD patients are expected to survive to adulthood in USA.^[2]

The challenge for anaesthesiologists in handling patients with CHD coming for extracardiac surgery relies on the patients' age, complexity of the heart lesion, coupled with patients' capacity to compensate, urgency of surgery and multiple coexisting diseases. In a clinical review of 191,261 patients less than 18 years old having one or more non-cardiovascular procedures, a diagnosis of CHD increases the mortality risk for both minor and major surgery, regardless of whether mortality is measured in a few days or after a month. Furthermore, with the same clinical group,

neonates and infants with CHD are associated with two fold increase in mortality from non-cardiac surgery.^[3] Anaesthesia-related paediatric cardiac arrest, according to a review, occurred in 75% of patients under 2 years of age with CHD during non-cardiac surgery.^[4] Nevertheless, under the experienced hands, anaesthesia for neonates with complex heart diseases like hypoplastic left heart syndrome (HLHS), unbalanced atrioventricular septal defects (AVSD), unstable Tetralogy of Fallot (TOF) and truncus arteriosus (TA) presenting to us for treatment of general surgical emergencies can still be conducted with manageable complications.^[5]

PREOPERATIVE CONSIDERATIONS

Children with CHD presenting for non-cardiac surgery can be grouped into three categories: Non-operated patient, with previous palliative surgery and with previous corrective surgery.^[6] Apart from categorising, to fully optimise the patient, it is a must for anaesthetist to obtain information about the cardiac

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lesion, its altered physiology and its implications under anaesthesia. This includes knowledge about whether the patient is on parallel or single ventricle physiology and relies deeply on relative resistance between systemic and pulmonary circulation.^[7]

Thorough preoperative preparation will include the following.

Anaesthetic history and physical examination

Effects of respiratory infection on pulmonary vascular resistance are more deleterious in patients with pulmonary hypertension (HTN) or cavopulmonary anastomosis.^[8] The decision on whether to delay the surgery must be discussed with the surgeon to weigh the risk benefit issues.^[9] Poor exercise tolerance is indicated by fatigue and dyspnoea on feeding, irritability and inability to gain weight. Previous cardiac and non-cardiac surgeries and prolonged intubation should be enquired about as they may suggest difficult IV insertion and subglottic stenosis, respectively.^[1]

Right ventricular function is equally important as the left ventricular function in the paediatric CHD patient, and thus should also be assessed. Patients with high pulmonary flow may present with tachycardia, tachypnoea, irritability, cardiomegaly and hepatomegaly.^[10]

Associated non-cardiac congenital anomalies include musculoskeletal abnormalities 8.8%, neurological defects 6.9% and genitourinary irregularities 5.3%. Down's syndrome patients may have atlanto-occipital subluxation that will warrant airway management precautions.^[6,10]

Medication history must be elicited. In patients with CHD who may be on aspirin, warfarin, antidepressants, diuretics, angiotensin converting enzyme (ACE) inhibitors, and antiarrhythmics, the anaesthetic providers must be mindful of their associated side effects. Laboratory investigations should be tailored accordingly. In current practice, all cardiac medications should be given on the morning of surgery,^[11] with exemption of ACE inhibitors due to their hypotensive effects during anaesthetic induction as seen in adult cases.^[12] There is no issue with low-dose aspirin for simple superficial surgery. However, for major surgeries, aspirin is commonly discontinued 7-10 days prior to surgery.^[13] Children on warfarin must be admitted for anticoagulant monitoring and change over to intravenous heparin prior to surgery.^[1,12]

Fasting time orders should be clearly written with timing if possible. Dehydration should be avoided in cyanotic patients. If timing of surgery is uncertain, then an IV line should be placed and fluids started.

Sympathetic stimulation due to crying of an anxious and distressed patient can increase oxygen consumption and myocardial work; this might be poorly tolerated in a child with limited cardiac reserve. Midazolam is the preferred premedication to reduce oxygen consumption in the dose of 0.5 mg/kg orally half an hour before surgery. If IV line is present, then incremental doses of 0.1-0.25 mg/kg midazolam can be given, provided airway and breathing issues were addressed accordingly.^[14]

In the recent guidelines from American College of Cardiology/American Heart Association (ACC/AHA), underlying cardiac rhythm modes and settings of patients with implanted pacemakers and/or automated defibrillator must be investigated prior to and after operation. Intraoperative troubleshooting of the device must be done with trained personnel.^[15]

Endocarditis prophylaxis has recently been revised for dental procedures. AHA recommends antibiotic prophylaxis for following patients:

- When gingival tissue is manipulated, or periapical region of teeth or perforation of oral mucosa
- Prior history of infective endocarditis
- Non-repaired cyanotic CHD, including shunts and conduit
- Complete CHD repair within the previous 6 months
- Repaired CHD with residual defects.

Antibiotics for infective endocarditis prophylaxis is no longer indicated in patients with

- Aortic stenosis, mitral stenosis, or symptomatic or asymptomatic mitral valve prolapse.
- Genitourinary and gastrointestinal tract procedures (transoesophageal echocardiography, oesophagogastroduodenoscopy, colonoscopy, etc.) also do not warrant infective endocarditis prophylaxis unless active infection is present.^[16,17]

Investigations

Full blood count and coagulation profile should always be requested. Polycythemia increases blood viscosity which leads to thrombosis and infarction in cerebral,

renal and pulmonary regions.^[18] Prothrombin time (PT) and partial thromboplastin time (PTT) are usually deranged in a polycythemic patient. Coagulation abnormalities also occur due to platelet dysfunction, hypofibrinogenemia and factor deficiencies. Preoperative phlebotomy is performed in symptomatic hyperviscosity and hematocrit (HCT) more than 65%. However, dehydration must be corrected first before deciding about phlebotomy. On the other hand, WBC count and C-reactive protein (CRP) measurement provide potential diagnosis of infection.^[13]

Serum electrolytes as mentioned earlier should be checked in patients receiving diuretics. ECG may show ventricular strain or hypertrophy. ECHO is used for Doppler and colour flow mapping, while catheterisation is used for information about pressures in different chambers, magnitude of shunt and coronary anatomy.^[13]

The chest X-ray shows the heart position and size, atelectasis, acute respiratory infection, vascular markings and elevated hemidiaphragm.^[18]

Cardiologist evaluation

The need for cardiologist evaluation depends on the complexity of the lesion. In a patient with simple or moderately complex lesion that has been completely corrected and is well compensated, a standard pre-anaesthetic visit without cardiology consultation is acceptable. Moderately complex lesions accompanied with inability to compensate will warrant cardiologist evaluation and optimisation, but clearance must always be given by anaesthesiologist.^[13]

INTRAOPERATIVE CONSIDERATIONS

Whether or not the patients with complex heart disease are coming for minor day surgery or major operation, the anaesthetist must assure that the hospital is equipped to answer all potential problems the case might bring. If not working in a specialty institution, communication with other specialists and back-up support from other centres should be assured if complications arise.

All commonly used induction agents are well tolerated depending on the rate and dose of the drug, whether it is inhalational or intravenous. Systemic vascular resistance (SVR) and peripheral vascular resistance (PVR) balance should be considered when using intravenous agents. Inhalation induction is acceptable

in CHD patients with uncomplicated cardiac lesion. Patients with poor cardiac function, who require inotropes preoperatively, may not tolerate inhalational induction, and favour the use of ketamine. In a review of 18 neonates with complex cardiac defects undergoing major general surgery, ketamine was the most common induction agent in those not intubated at the time of surgery.^[5] Inotropes should be continued and IV induction agents titrated. The need for invasive monitoring should depend on the type of surgery and cardiac lesion.^[7]

Left to right shunts are the most common lesions representing over 50% of children with CHD. Examples include Atrial septal defects, Ventricular septal defects, Patent ductus arteriosus, AV canal defects, PAVD and Blalock Taussig shunt (BT Shunt). They have minimal effect on inhalation or intravenous induction and decrease with the drop in SVR or an increase in PVR. Left to right shunts lead to excess pulmonary blood flow. Patients are acyanotic, but deterioration in gas exchange may result from pulmonary congestion. 1-1.5 minimum alveolar concentration (MAC) of isoflurane, halothane and sevoflurane has no effect on Qp/Qs ratio in patients with isolated ASD or VSD during mechanical ventilation.^[19] 100% oxygen and hyperventilation in patients with L-R shunt will result in pulmonary vasodilation, which in turn will further increase pulmonary congestion, and thus should be avoided.

Right to left intracardiac shunts prolong inhalation induction, while IV induction is faster. R-L shunt or shunt reversal occurs when SVR decreases or PVR increases.^[20] Hypercyanotic “tet” spell under anaesthesia responds to volume, increase in SVR with alpha agonists such as Phenylephrine or Ephedrine and ceasing infundibular spasm with beta blockade.^[19] Another significant caveat in managing these patients is to be aware that pulse oximetry overestimates arterial oxygen saturation as saturation decreases, end-tidal carbon dioxide readings underestimate PaCO₂, and discrepancy worsens with hypoxemia.^[21] When in doubt, obtain an arterial blood gas preoperatively for baseline.

Mean pulmonary artery pressure greater than 25 mmHg at rest and 35 mmHg during exercise is defined as *pulmonary hypertension*. High pulmonary flow as occurs in unrestricted L-R shunt will lead to congestive heart failure (CHF) and pulmonary HTN. Initially pulmonary HTN is reactive and responds

to hypothermia, stress, pain, acidosis, hypercarbia, hypoxia and elevated intrathoracic pressure, but later pulmonary HTN becomes fixed.^[22] The anaesthetic goals in managing such a patient are to prevent increase in PVR and depression of myocardial function. Pulmonary HTN crisis intervention measures such as 100% oxygen, inhaled nitric oxide, phosphodiesterase inhibitors, prostacyclin analogues, inotropes and other measures to maintain cardiac output and pulmonary blood flow must be prepared and administered accordingly as the need arises. On the other hand, despite the potential airway and ventilatory issues during sedation or anaesthesia, the choice of airway device dictates the incidence of complications in children with pulmonary HTN undergoing non-cardiac surgery. Reports regarding incidence of pulmonary hypertensive crisis among tracheally intubated adult patients with severe pulmonary HTN encourage anaesthetists to use less-invasive airway management if the surgical procedure permits.^[23]

Patients with single ventricle pathway may have gone through stages of palliation. Following completion of BT Shunt and Glenn shunts, oxygen saturation is expected to be 75-85%. Always note that after these two stages, residual shunts, increased sensitivity to SVR and PVR pressure changes, and arrhythmia are still not uncommon.^[24] Previous surgery scar, and atrial or ventricular overdistension can trigger arrhythmia. Anaesthetists must diagnose the rhythm first before administering antiarrhythmics.^[1,24]

Fontan physiology relies on transpulmonary gradient to direct blood into the pulmonary circulation, and thus dictates cardiac output. Therefore, if a patient with Fontan physiology comes in for a laparoscopic procedure, one must take into consideration the capacity of the patient to handle the insult of hypercarbia, pneumoperitoneum and extremes of positions.^[25] Studies have shown that insufflation pressures less than 8-12 cm H₂O did not decrease cardiac output, while a decrease in cardiac performance was seen in pressures of 15-20 cm H₂O. Case reports show neonates with well-compensated Fontan physiology can have an uneventful laparoscopic abdominal surgery. In another case report, an adolescent with non-fenestrated Fontan went through two laparoscopic procedures with 3 years interval and successfully tolerated incision, CO₂ insufflation and Trendelenberg position. On both occasions, intraabdominal pressures were maintained less than 10 cm H₂O, and there were no significant haemodynamic changes and anaesthetic management

alteration. Patient was extubated immediately after the operation and was discharged the next day.^[26]

Pain management is a critical factor that an anaesthetist must cover during intra- or postoperative management. Opioid infusion or patient-controlled analgesia for major operations has been the primary postoperative intervention for pain for patients with CHD.^[1,5] The use of regional anaesthesia for well-compensated patients with CHD was reported with no complications.^[3] In the study by Shenkman *et al.*, 44 unsupplemented spinal anaesthetics for inguinal hernia correction were done with 1 mg/kg of either hyperbaric tetracaine or bupivacaine in premature or former premature infants with non-cyanotic CHD; even when fluid restricted, causes minimal cardio-respiratory complications.^[27] In a related investigation, infants undergoing minor non-cardiac surgery under awake spinal anaesthesia were not different from controls without CHD in terms of percent decrease in mean arterial pressure and heart rate. Fourteen patients in the study group were even classified as high risk, having pulmonary HTN, congestive heart failure, combined cardiac lesions, multiple cardiac surgeries, and on drug therapy.^[28]

POSTOPERATIVE CONSIDERATIONS

Even with favourable outcome, patients with CHD coming for non-cardiac surgery are still under high-risk category after operation.^[5] Observing them in high-dependency bed or intensive care unit will warrant assurance of catching arrhythmia, cardiac ischaemia, dehydration, pain, ventilator issues and other complications before they cause detrimental effects.

CONCLUSION

As there are many complex factors involved, anaesthetic management of patients with CHD coming in for non-cardiac surgery is based on individual experience, and confidence in handling the case. Precision in planning and good foundation in physiological and pharmacological principles will contribute to the successful completion of the procedure.

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Conference Calendar Details

Name of the conference: 60th Annual National Conference of the Indian Society of Anaesthesiologists, ISACON 2012

Date: 26th to 29th December 2012

Venue: Labh-Ganga Convention Centre, MR 10-Bypass Junction, Indore, M. P., India

Organizing Secretary: Dr. Kishore Arora, Associate Professor of Anaesthesiology, M. G. M. Medical College & M. Y. Hospital, Indore 452 001, Madhya Pradesh, India

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Website: www.isacon2012.com

Name of the conference: 5th Annual Conference of Indian Association of Pediatric Anesthesia IAPA CON 2013

Workshop: Pediatric Regional Anesthesia and Difficult Airway - 8th February 2013

Date: 9th and 10th February, 2013

Venue: Hotel Taj gateway , Vadodara, Gujarat

Organising Secretary: Dr. Falguni Naregal, Neel Surgical Hospital for Children,

Near Atithi Gruh, Akota, Vadodara - 390020, Gujarat, India

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Website: www.iapacon2013.org

Name of the conference: 10th Congress SAARC Association of Anaesthesiologists

Date: 22nd to 24th February, 2013

Venue: Bangabandhu International Conference Centre, Agargaon, Sher-E-Bangla Nagar, Dhaka-1207, Bangladesh

Organising Secretary: Prof. Debabrata Banik, SAARC-AA Congress 2013

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