PAEDIATRIC CARDIAC ANAESTHESIA

Introduction

The incidence of congenital heart disease is 6-8:1000. It can exist in isolation or be associated with chromosomal abnormalities or syndromes.

Congenital heart disease can be classified as follows:

1. 'Simple' left to right shunt: increased pulmonary blood flow
    - Patent Ductus Arteriosus (PDA)
    - Atrial Septal Defect (ASD)
    - Ventricular Septal Defect (VSD)
    - Atrioventricular Septal Defect (AVSD)
2. 'Simple' right to left shunt: results in cyanosis
    - Tetralogy of Fallot (TOF)
    - Pulmonary atresia
    - Tricuspid atresia
    - Ebstein's anomaly
3. Complex shunts: mixing of pulmonary and systemic blood flow with cyanosis
    - Transposition of Great Arteries
    - Total anomalous pulmonary venous drainage (TAPVD)
    - Hypoplastic left heart syndrome (HLHS)
    - Double outlet right ventricle (DORV)
    - Truncus arteriosus
4. Obstructive lesions
    - Coarctation of Aorta
    - Interrupted or hypoplastic aortic arch
    - Aortic Stenosis
    - Mitral Stenosis
    - Tricuspid Stenosis
5. Regurgitant lesions

Surgery for congenital heart disease can be

a. Corrective (eg. PDA ligation/ ASD closure/ VSD closure) or
b. Palliative (eg. Pulmonary artery (PA) banding/ Blalock-Taussig (BT) shunts/ cavo-pulmonary shunts)

Surgery can be either

a. Open (on cardiopulmonary bypass (CPB)) or
b. Closed (non-CPB)
    - ligation of PDA (thoracotomy)
    - Repair of coarctation of aorta (thoracotomy)
    - PA banding (thoracotomy or sternotomy)
    - BT shunt (thoracotomy or sternotomy)

Considerations for anaesthetic plan include:

1. The cardiac lesion:
    - the predominant one on the basis of pathophysiology
    - myocardial reserve /functional capacity (feeding, sweating, grunting, recurrent chest infections, failure to thrive, exercise tolerance, Hb, SpO_2) /rhythm /complications (neurological, heart failure, tet spells)
    - nature of shunt /obstruction
    - impact on pulmonary and systemic pulmonary vascular resistance (PVR/SVR)
2. Planned surgery
3. Management of CPB
4. Other patient factors
    - age
    - associated lesions /syndromes
    - respiratory tract infection
    - vascular access
    - airway and dental status
    - medications
    - previous surgery

Investigations and results

- FBC
- U/E/Cr
- PT/PTT
- CXR
- ECG
- 2D echo report /cardiac catheterisation report

Pathophysiology

The pathophysiology depends on the nature and size of the cardiac lesion. Where more than one cardiac lesion exist, usually one lesion will predominate.

Other factors that can impact on the overall behaviour of the cardiac lesion include:

1. Transition circulation:
    - This in the first few days to weeks of life.
    - The pulmonary circulation is very reactive and potential reopening of the ductus arteriosus can occur during this period. The circulation is extremely sensitive to hypoxia, hypercarbia, acidosis and prostagladins at this stage. Shunting or reversion to fetal circulation can occur.

2. Presence of a duct-dependent circulation. This could be either:
    - duct-dependent systemic circulation (eg critical aortic stenosis, hypoplastic left heart syndrome, coarctation of the aorta). It can present with collapse or cardiac failure.
    - duct-dependent pulmonary circulation will present with increasing cyanosis unresponsive to increasing oxygen concentrations. The duct must be kept open until further surgical management is possible by avoiding high FiO_2, allowing moderate hypercarbia and administering PGE_1 via an infusion. Cardiac output may have to be supported with fluids and inotropes.

3. Presence of non-restrictive/ balanced shunts
Balancing the systemic and pulmonary circulation is important to avoid cyanosis, cardiac failure or systemic hypotension.

Determine if the following are present:

1. Cyanosis
   This results in chronic hypoxia, compensatory polycythemia with increased risk of thromboembolic phenomena, coagulopathy and metabolic acidosis. The severity is indicated by the baseline saturations in room air and polycythemia. These children benefit from hydration in the perioperative period and it is useful to order an intravenous drip for the duration in which they are kept nil by mouth. In selected cases, preoperative oxygen therapy, correction of acidosis and inotropic support may be required.

2. Reduced pulmonary blood flow
   Oxygen therapy, hyperventilation and avoiding a decrease in the systemic vascular resistance aid in reducing pulmonary vascular resistance and promoting blood flow to the lungs. Selective agents to reduce PVR eg nitric oxide may be beneficial especially in those infants with right ventricular hypertrophy.

3. Increased pulmonary blood flow and cardiac failure /duct-dependent systemic circulation /high pressure or volume shunts.
   These children benefit from moderate hypercarbia, room air or avoiding high FiO_2 and inotropes in some instances. Marked falls in SVR should be avoided.

Pre-operative visit

The aims of the pre-operative visit are to establish and optimize the child's clinical condition to facilitate the formulation and execution of an anaesthetic plan, and to establish rapport with the child and parents. Discussion with the parents should include the following: premedication if required, fasting, the induction process, invasive lines, the transfusion of blood/ blood products, inotropic support, analgesia/sedation, and the type of post-operative care (ICU in most instances). The insertion of trans-esophageal echocardiography (TEE) probe by the anesthetists in certain corrective surgeries (septal defect repairs, valvular surgery) and its associated risks should be conveyed and consent obtained.

Blood and blood products for cardiac surgery

1. Open heart surgery

   3 units of PCT (packed cells), 2 units of FFP and 10mls/kg platelets to be grouped and matched for each patient regardless of weight. More blood products including cryoprecipitate may have to be matched and confirmed as required (eg in redo cardiac surgeries).

   a. For children less than 15kg:
      - 3 units of packed cells (to be stored in MOT blood fridge prior to induction); **Fresh blood should be requested for neonates.**
      - 2 units of FFP; +/- Cryoprecipitate
      - 20 mls/kg platelets should be confirmed and available in the KKH blood bank. This is sent for only when instructed by the anaesthetist.

   b. For children more than 15 kg:
      - 3 units of packed cells (to be in MOT blood fridge at prior to induction), 2 more units in KK blood bank
      - 2 units of FFP; +/- Cryoprecipitate

- platelets 20 mls/kg (or 2 CSP for bigger children) to be confirmed and available in KKH blood bank. These are sent for only when confirmed by the anaesthetist.

2. Closed heart surgery

- 20 mls/kg of packed cells should be grouped and cross matched and available in KK MOT blood fridge
- 10 mls/kg of FFP should also be available for cyanotic patients in the KKH blood bank but this is sent for only when requested by the anaesthetist .

The blood and blood products are arranged by cardiac surgery resident. Cross check by anaesthesia resident at the time of anesthesia premed is mandatory.

Preparation of the operating theatre

1. Check anaesthetic machine and other appropriate equipment

2. Drugs (please print cardiac calculator)

A. Resuscitation:
   a) atropine
   b) adrenaline 1: 10,000 (or 1:100,000 for neonates)
   c) calcium chloride 10%
   d) 8.4% $NaHCO_3$ (4.2% for neonates)
   e) phenylephrine

B. Induction agents/ paralyzing agents/

C. Sedation (midazolam /dexmedetomidine infusion) and Analgesia (morphine/ fentanyl infusion)

D. Antibiotics: cefazolin 50 mg/kg (repeat after 8 hours)
   If patient is allergic to penicillin, please order IV Vancomycin

E. Inotropes (to confirm with anaesthesia consultant). Commonly used inotropes include adrenaline, milrinone, dobutamine, dopamine)

F. Vasodilators (to confirm with anaesthesia consultant) Commonly used vasodilators include sodium nitroprusside, GTN infusion, phentolamine

G. Heparin 300U/kg for open heart surgery

The ACT should be checked 3 minutes after injection of heparin and should be > 450 seconds before CPB is initiated. Check the dosage with anesthetist consultant for closed heart procedures like BT shunt or coarctation of aorta.

Other drugs that may be required include

i) Tranexamic Acid: Loading dose 25 mg/kg IV after induction in 1 hour followed by 2.5 mg/kg/hr. Continue the infusion for 4-6 hrs in ICU after shifting.

ii) esmolol

iii) amiodarone

iv) magnesium

v) methylprednisolone

3. Thermoregulatory equipment
4. Fluids
5. Blood/ blood products
6. Defibrillator
7. Rotem
8. Monitoring equipment

These include:

i) Standard anaesthetic monitors including 2 SpO_2 probes

ii) Temperature: nasal or esophageal, rectal

iii) Invasive lines

a. arterial line: site (consider the size of the patient, previous surgery, current surgery)

b. central venous line (CVL)
Possible sites (IJV/ femoral vein / direct atria).
Considerations include:

- left IJV should be avoided in children with persistent left SVC

- avoid IJV for infants with univentricular physiology
- cava-pulmonary shunts: single lumen IJV CVL to monitor PA pressures after shunt and triple lumen femoral CVL
- transthoracic lines may be placed to measure LA/ PA pressure post bypass

c. PA line (double switch operation)

iv) Urinary catheter
v) NIRS
vi) TEE probe

Conduct of anaesthesia

Either inhalation with sevoflurane or intravenous induction with ketamine, fentanyl, midazolam, thiopentone or propofol is used. The choice of induction is dependent on the functional status of child. At the very least, a pulse oximeter should be placed before induction. Other monitors should be placed as soon as the child tolerates it For duct dependent lesions, avoid high FiO_2 and avoid hyperventilation. The child is then paralyzed, IPPV commenced, vascular access lines are placed and anaesthesia is maintained with a sevoflurane: air:oxygen mixture.

In open heart surgery, cardiopulmonary bypass is used. This may be conducted under normothermic or hypothermic conditions. Under certain circumstances where reconstruction of the aortic arch or better surgical exposure of intracardiac defects is needed, total circulatory arrest (TCA) is employed. In this case, the patient is cooled to less than 20°C. Ice packs on the head and cooling of the operating theatre to 18°C are indicated.

When rewarming, think of the following:

i) Use of vasodilators to aid in rewarming. This is more commonly practiced for deep hypothermia in < 25 degree Celsius. The drugs which can be used for this purpose include IV SNP and GTN infusion.
ii) Turn on warming devices eg warming blanket, bair hugger. Remove ice pack from patient's head.
iii) Start inotropic infusions when patient's temperature is at least 32 degree Celsius
iv) Re-zero invasive lines
v) Call for blood/ blood products

Before coming off bypass, check for the following:
i) Temperature: core temperature should be at least 36.5ºC
ii) Stable cardiac rate and rhythm
iii) Stable hemodynamics
iv) Normal electrolytes (Ca^+ / K^+), base excess<-5mmol/L, gas exchange, Hb / Hct levels (check the last blood gas done on pump with perfusionist)
v) Blood products in OT

Use of blood from CPB: The unused blood from bypass circuit maybe used for transfusion after it is hemoconcentrated by perfusionist. Ideally, this should be used if the bypass time is less than 3 hours and excessive use of suction had been avoided during CPB by the surgeons.

***Please refer to our bypass checklist attached at the end of this chapter.**

Re-sternotomy

Re-sternotomy may be required in certain instances. Potential problems include bleeding and arrhythmias.

Ensure:

- Arrange for 5 units of PCT, 2 units FFP, 2 CSP and cryoprecipitate available in KKH blood bank

- Bring 3 units of PCT to MOT blood fridge (check 1 unit of PCT before skin incision but do not warm – keep in blood box with cool packs. If it is not required after chest is opened, remember to put blood back in the MOT blood fridge)
- Defibrillation pads – apply to back, making sure no direct contact between each pad or with ECG pads. Rememeber to apply extra set of ECG pads for Defib machine. Check that the ECG waveform obtained on the Defib machine is good.
- Groin exposed and prepared for fem-femoral bypass

An *oscillating saw* is used for sternal opening (no need to let lungs down in this instance).

Transfer of patient to ICU

- Period when hemodynamic instability can occur
- Make sure that patient is stable before transfer
- Transfer one monitoring device at a time and make sure someone constantly looks at the numbers
- Make sure all lines are free and of sufficient length
- Ensure resuscitation drugs and fluid boluses are available at all times
- Make sure you are able to ventilate and oxygenate appropriately
- Transfer patient from table to bed when all the above fulfilled
- Ensure patient has adequate sedation and analgesia (midazolam/ dexmedetomidine/morphine/fentanyl infusion)
- Call ICU to inform of patient's impending arrival
- The cardiothoracic registrar should accompany the child to ICU
- Hand over to ICU registrar /consultant concisely and precisely about perioperative events. The SBAR

handover form has to be duly filled and signed by both PAN and ICU after handover

Chest left open

In certain instances e.g. when there is myocardial oedema, the chest may be left opened. Hemodynamic instability would result if attempts to close the chest were made at this point. The following should be ensured:

1. the handover to ICU staff must include the fact that the child's chest has been left opened.
2. when the chest wound has been covered, there must be obvious stickers to indicate that the chest has been left opened
3. the child must be kept sedated and paralyzed for the duration that the chest is left opened. This is usually achieved by making a "3-in-1" cocktail of the following drugs in a 50 ml syringe: midazolam 3mg/kg, fentanyl 250mcg/kg and rocuronium 25mg/kg.
   Running this cocktail at 1ml/hr would then give
   Midazolam 1 mcg/kg/min
   Fentanyl 5 mcg/kg/hr
   Rocuronium 0.5mg /kg/hr
4. antibiotics prophylaxis should be continued for the duration that the chest is left opened.

The chest is usually closed within 24-48 hours when the patient is hemodynamically stable and the oedema has settled. This is usually done in the ICU.

Re-opening Chest in the ICU

This may be required for rapid access to heart when there is:

- Cardiac tamponade from bleeding
- Open resuscitation

2 possible scenarios:

- chest already wired and skin layers closed
- chest has been left “open”

In latter is more common and access to the heart is rapid as only the sutures stitching the clear PVC to skin needs to be taken out. Maintain “3-in-1” cocktail whilst ensuring the following:

- Adequate access for anaesthetist to ventilator, IV lines and head end of patient
- Adequate resuscitation drugs available
- Continue ongoing fluid / blood / blood product replacement
- Additional sedative / hypnotic / muscle relaxant bolus may be required

In cases where the sternotomy has to be re-opened, prepare as for chest opening in OT.
Ensure:

- Adequate access for anaesthetist to ventilator, IV lines and head end of patient
- Adequate resuscitation drugs available
- Continue ongoing fluid / blood / blood product replacement

Additional sedative /hypnotic /muscle relaxant bolus may be required.

Chest closure in ICU

In cases where the sternotomy has been left open due to hemodynamic instability, myocardial oedema or bleeding, chest closure may be carried out once patient in stable. This is usually within 24-48h post operatively.
Paediatric Intensivists often take over care of the patient during this time as part of continuing care. In the event that they are not able to, the anaesthetist may also look after the patient during the procedure.

Ensure:

- Adequate access for anaesthetist to ventilator, IV lines and head end of patient
- Resuscitation drugs available (but not opened / drawn up as patient should be stable)
- Fluid boluses /blood products available
- "3-in-1" cocktail / sedation, analgesia and muscle relaxant infusions are in progress
- Additional sedative /hypnotic /muscle relaxant bolus may be required
- Constant monitoring of cardiovascular status when sternotomy is closed – fluid bolus may be required, and ventilation parameters may need to be adjusted for change in filling pressure requirements and chest compliance respectively.

PDA Ligation in NICU OT

This elective operation is done in the NICU OT for premature ill babies. The baby is placed in the left thoracotomy position and operation is carried out in the open care Resuscitaire.

Ensure:

- The baby is reviewed preoperatively. Note the airway intervention and ventilation mode, location of invasive lines and peripheral IV cannula, medications, significant clinical findings and laboratory results.
- If the baby is not intubated or lined, arrange for NICU colleagues to intubate, set arterial line and insert peripheral IV cannula 2 hours prior to scheduled time for surgery.
- Have the NICU team shift the baby into NICU OT 30 minutes before operation (you may have to call NICU nurse in charge to confirm the exact time)
- Have one unit of PCT in KK MOT blood fridge
- Prepare the anesthetic and resuscitation drugs after discussing with the anesthesia consultant. Anesthetic tray prep may include: Ketamine, muscle relaxant, Fentanyl (ask from NICU), Cefazolin, 100cm extensions, anti-reflux valves

- Instruct AU nurse to bring: a bottle of 5% albumin, portable ETCO2 monitor, fluid warmer, 3M plastic, gamgees /gauzes, micropore eye tapes, masking tape.

Complications of PDA Ligations

- Bleeding - usually minimal unless the duct tears in which case it can be catastrophic.
- Inadvertent ligation of the aorta or pulmonary artery. The correlating signs of duct occlusion are the disappearance of murmur and rise in blood pressure (mainly diastolic). On the other hand if the aorta is ligated the lower limb SpO2 trace will disappear, right arm BP will be unrecordable. With PA occlusion the ETCO2 will disappear.
- Thoracic duct and recurrent laryngeal nerve injury
- Pneumothorax - usually a chest tube will be placed postoperatively to drain residual air.

ROTEM

The Rotational Thromboelastometry provides global information on the dynamics of clot development, stabilization and dissolution that reflect in vivo hemostatsis. Its use during cardiac surgery has been shown to significantly reduce the use of blood component therapy and overall blood loss.

The ROTEM test is performed during rewarming of the child. The graph generated by the machine indicates the requirement of various blood components that is required after heparin reveral with protamine after coming off bypass.

Depending on the bleeding status, a second ROTEM test can be done after transfusing the required blood products.

ECMO (Extracorporeal Membrane Oxygenation)

ECMO is a well-established therapy as a mode of cardiac and respiratory support in reversible cardiac and pulmonary failure in neonatal and paediatric patients.

ECMO is instituted in pediatric patients when conventional modes of cardiorespiratory supports have failed. Mechanically , blood is drained from the venous system, pumped through an artificial lung where oxygen is added and carbon dioxide removed and then,

depending on the configuration of the circuit, returned to either the venous or arterial circulation.

The role of anesthetist during institution or separation of ECMO would be:

a) To provide anaesthesia for the procedure.
b) Hemodynamic and respiratory monitoring of the child during the procedure.
c) Be ready for transfusion of blood products.
d) To maintain anticoagulation with heparin during institution of ECMO.

CARDIAC CATHETHERISATION STUDIES AND TRANS-THORACIC / TRANS-ESOPHAGEAL ECHOCARDIOGRAPHY

This is done in the Angiography suite located next to the Paediatric Major OT.

In our institution, cardiac catheterisation studies, TTE and TEE in children are done under GA. They can be diagnostic or interventional. Interventional procedures include PDA coiling, Amplatzer closure of ASDs or balloon valvuloplasties.

Preoperative Assessment

The congenital heart disease ranges from simple lesions e.g. PDA and ASD to complex heart lesions. Cyanotic Heart Lesions include Tetraology of Fallot, Pulmonary Atresia/VSD or single ventricle pathology e.g. hypoplastic right ventricle lesions.
During the preoperative evaluation, the effects of the cardiac lesion on the general health of the child (e.g. failure to thrive, functional status, URTI) should be assessed.
The effects of concomitant drug therapy should also be noted and relevant drugs continued up to the day of cardiac studies. Drugs include anti-failure drugs e.g. digoxin and diuretics or ß blockers for cyanotic spells in FT.
Review previous GA and surgeries e.g. palliative shunts.
Establish the presence of co-existing congenital diseases.

Investigations and preoperative instructions

FBC, U/E/Cr, PT/PTT, CXR, GXM and ECG for those undergoing cardiac catheter studies.
In children having *only* TEE, investigations will only be done if indicated (by history, physical examination).
Previous Catheter and 2D Echo results should also be noted.

Blood should be cross matched and available in the OT blood fridge for interventional cases.

IV fluid hydration must be ordered and commenced for all cyanotic patients from the time of fasting.

Angiography suite preparation

Drugs:

- Appropriate anaesthetic drugs should be drawn
- Resuscitative drugs are drawn according to the patient's cardiac disease and general condition e.g. phenylephrine and esmolol in Tetralogy of Fallot with cyanotic spells or adrenaline in critically ill babies.
- Antibiotics are not routinely but when required, AHA guidelines are to be followed.
- Heparin may occasionally be requested by the cardiologist; be sure to confirm the doses with the anesthesia consultant and cardiologist.

Fluids:

- Lactated Ringers is the default solution with an extension tubing and 3 way tap attached.
- In neonates and patients at risk of hypoglycemia, a dextrose maintenance drip may be required.
- Albumin or boluses of normal saline are usually given for unexpected blood loss if the original haematocrit is acceptable. Otherwise, blood loss should be replaced with cross matched blood (packed cells).

Equipment:

- The anesthesia machine and drip stand should be positioned within the red floor markings to avoid obstructing the movement of the C-arm of the fluoroscopy machine.
- The physiologic monitors should be positioned such that a clear view of patient parameters is obtained at all times.
- Ensure proper taping of ETT to prevent dislodgement during TEE and kinking by antero-posterior arms of the X-ray machine. If TEE is to be carried out, a mouth guard / bite block should be put in before securing the ETT.

Positioning of patient:
The arms of the patient are positioned on either side of the head, so that unobstructed images of the heart may be obtained. Avoid over-stretching of the brachial plexus in the older patients by supporting the arms with a pillow, gamgees or sponge. All pressure points should be protected.

Temperature management:
When the anticipated procedure time is long or the patient is a small infant, a plastic sheet should be used as an occlusive drape to keep patient warm. If a Bair Hugger is used, ensure proper positioning of warm air hose to avoid thermal injury. Temperature should be monitored.

Conduct of anaesthesia
Following an intravenous or inhalational induction, anaesthesia is continued with volatile agent supplemented with boluses of fentanyl (0.5-1 mcg/kg) and an IPPV/muscle relaxant technique is used.
The FiO2 used depends on the pathophysiology of the patient and whether or not the cardiologist wishes to sample blood for calculation of intra-cardiac shunts. In these cases an air/O2 mixture as close to 21% is often required. IV paracetamol may be used as analgesic adjunct.

Reversal
Ensure that the cardiologist is satisfied with haemostasis of the femoral puncture sites and pressure bandages are applied before reversing the patient.

Recovery
Patients are transferred to Major OT Recovery Room for monitoring and thereafter to either the cardiac step down unit (CSDU) or the general ward.

If the patients are transferred directly to CICU intubated, you may require an air/oxygen blender to avoid high FiO2 during transfer. (High FiO2 may cause pulmonary vasodilatation hence flooding of the lungs in patients with single ventricle physiology).

Hybrid Procedures

Closure of certain heart defects like VSD may be carried out in Major OT by the cardiologist in an open chest setting. Preparation should be as for open heart surgery with the additional provision of Transoesophageal Echocardiography.