Anesthetic Considerations in Patients with Congenital Heart Disease undergoing Non-cardiac Surgery

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Outline

• Anesthetic considerations
• Type of CHD circulation
• Risk classification of CHD patients
• Cyanotic heart disease
• Infective endocarditis prophylaxis
• Case examples
Anesthetic Considerations

1. Knowledge of underlying lesion and type of circulation: change in SVR/PVR? SpO2?
Underlying Lesions

• Obtain information about cardiac lesion, altered physiology, its implication under anesthesia

• Group of patients with CHD
  - Non-operated patients
  - Previous palliative surgery
  - Previous corrective surgery
Types of Circulation

1. Normal circulation
2. Parallel or Balanced Circulation
3. Single Ventricular Circulation
1. Normal or Series Circulation
2. Parallel or Balanced Circulation

*Ex. Large ASD, VSD, truncus arteriosus.*
<table>
<thead>
<tr>
<th>Treatment</th>
<th>PVR</th>
<th>SVR</th>
<th>$Q_p/Q_s$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Increase $Fi_O_2$</td>
<td>Decrease</td>
<td>Increase</td>
<td>Increase</td>
</tr>
<tr>
<td>Increase $CO_2$</td>
<td>Increase</td>
<td>Decrease</td>
<td>Decrease</td>
</tr>
<tr>
<td>Increase pH</td>
<td>Decrease</td>
<td>Increase</td>
<td>Increase</td>
</tr>
<tr>
<td>PEEP</td>
<td>Increase</td>
<td>No effect</td>
<td>Decrease</td>
</tr>
</tbody>
</table>

*Abbreviations: $Fi_O_2$, fraction of inspired oxygen; PEEP, positive end-expiratory pressure; PVR, pulmonary vascular resistance; SVR, systemic vascular resistance.*
Atrial Septal Defect

Types:

Ostium seconum 75%

Ostium primum 15%

Sinus venosus 10%

Coronary sinus rare
Atrial Septal Defect

**Physiology:** left to right shunt

**Preop:** rarely cause symptoms, CHF

**Surgery:** sutures, pericardial patch, cardiac catheterization

**Postop:** SA node injury and dysrhythmias (immediate postop), post pericardiotomy syndrome (first few days)
Ventricular Septal Defect

Types:

Perimembranous 80 %

Subpulmonary 5 – 7 %

Muscular 5 – 20%

AV canal  5 – 8 %
Ventricular Septal Defect

**Physiology**: left to right shunt

**Preop**: identify type and size of VSD. The potential PHT needs to be followed closely to determine the timing of surgery

**Surgery**: sutures, pericardial patch

**Postop**: heart block, junctional ectopic tachycardia (in infants), residual VSD
Patent Ductus Arteriosus

• **Physiology:** Left to right shunt.

• **Preop:** CHF (child>1 year old), pulmonary vascular disease

• **Surgery:** ligated or divided and sewn, or cardiac catheterization

• **Postop:** Complications are rare.
Tetralogy of Fallot

- Four cardiac abnormalities
  - VSD
  - Subpulmonic stenosis
  - Overriding of aorta
  - Right ventricular hypertrophy
- Degree of cyanosis
- ↓Pulmonary blood flow
Tet spell

• Profound hypoxemia

• $O_2$ demand: crying, feeding, defecation

• Transient cerebral ischemia: paleness, unconsciousness

• Treatment: $SVR$, decreasing Rt to Lt shunt
TOF: Considerations

- Variable Rt to Lt shunt and pulmonary blood flow
- Associated conditions:
  - Paradoxical embolus – avoid air bubbles in lines
  - Polycythemia
  - 25% have another congenital abnormality
  - Tracheoesophageal fistula & trisomy 21
- IE prophylaxis
- **No air** in IV line
TOF: Considerations

• Maintain intravascular volume and SVR.

• Avoid increases in PVR

• Ketamine is recommended for induction

• Right to left shunt tends to slow the rate of inhalational agents

• Problems after surgery: residual RVOT obstruction, RV failure, heart block, residual VSD, late arrhythmia, sudden death
# Hemodynamic Goals

**TABLE 14.6** Cardiac grid for common congenital heart diseases (desired hemodynamic changes)

<table>
<thead>
<tr>
<th></th>
<th>Preload</th>
<th>PVR</th>
<th>SVR</th>
<th>HR</th>
<th>Contractility</th>
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</thead>
<tbody>
<tr>
<td>ASD</td>
<td>↑</td>
<td>↑</td>
<td>↓</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>VSD (right-to-left)</td>
<td>N</td>
<td>↓</td>
<td>↑</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>VSD (left-to-right)</td>
<td>↑</td>
<td>↑</td>
<td>↓</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>PDA</td>
<td>↑</td>
<td>↑</td>
<td>↓</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>Infundibular pulmonary stenosis</td>
<td>↑</td>
<td>↓</td>
<td>N</td>
<td>↓</td>
<td>↓^{a}</td>
</tr>
</tbody>
</table>

^{a} - over-riding consideration \(N\) - normal or no change
3. Single Ventricular Circulation

Ex. Blalock–Taussig, (BT)

Glenn shunt, HLHS, Tricuspid Atresia
Fontan Circulation

- Divert blood from IVC to PA
- Passive pulmonary blood flow
- Single ventricle to pump blood to the whole body
- Complications: atrial thrombus, atrial arrhythmia, ventricular dysfunction, chylothorax, protein-losing enteropathy
Anesthetic Considerations

1. Knowledge of underlying lesion and type of circulation: change in SVR/PVR? SpO₂?

2. Evidence of long-term complications or high-risk category
### Risk Classification of Children with Heart Disease undergoing Non-Cardiac Surgery

<table>
<thead>
<tr>
<th></th>
<th>Intermediate risk</th>
<th>Low risk</th>
</tr>
</thead>
<tbody>
<tr>
<td>High risk</td>
<td>Physiologically normal or well compensated</td>
<td>Physiologically normal or well compensated</td>
</tr>
<tr>
<td></td>
<td>Simple lesions</td>
<td>Simple lesions</td>
</tr>
<tr>
<td></td>
<td>Major surgery (intraperitoneal, intrathoracic, anticipated major blood loss requiring transfusion)</td>
<td>Minor (or body surface) surgery</td>
</tr>
<tr>
<td></td>
<td>Under 2 yr old Emergency surgery</td>
<td>Over 2 yr old Surgical</td>
</tr>
<tr>
<td></td>
<td>Preoperative hospital stay more than 10 days</td>
<td>Elective surgery</td>
</tr>
<tr>
<td></td>
<td>ASA physical status IV or V</td>
<td>Preoperative hospital stay less than 10 days</td>
</tr>
</tbody>
</table>

#### Table 1: Risk classification of children with heart disease undergoing non-cardiac surgery

- **Physiologically poorly compensated and/or presence of major complications**
  - (a) Cardiac failure
  - (b) Pulmonary hypertension
  - (c) Arrhythmias
  - (d) Cyanosis
- **Complex lesions (single-ventricle or balanced circulation physiology, cardiomyopathy, aortic stenosis)**
- **Major surgery (intraperitoneal, intrathoracic, anticipated major blood loss requiring transfusion)**
- **Under 2 yr old Emergency surgery**
- **Preoperative hospital stay more than 10 days ASA physical status IV or V**
Risk Classification

High risk: Long term complications of CHD

• Cardiac failure
• Pulmonary hypertension
• Arrhythmias
• Cyanosis
High Risk: Cardiac failure

- Volume overload: shunt, incompetent valve
- Pressure overload: outflow tract obstruction (AS, PS)
- S/S: tachypnea, tachycardia, sweating, cool peripheries, feeding difficulties, failure to thrive

- Prolong IV induction time
- Avoid prolonged use of 8% sevoflurane, propofol
- Ketamine = agent of choice, (but controversial in PHT)
• PAP ≥ 25 mmHg at rest or ≥30 mmHg during exercise

• PHT: ↓pulmonary compliance, ↑increased airway resistance

• Respiratory tract infections: poorly tolerated and have a greater impact on PVR

• 8 times major complications

• Treatment: 100% O₂, inhaled nitric oxide, i.v. prostacyclin, inotropic support of RV
High Risk: Arrhythmias

- **RBBB** is common but unlikely to generate heart block

- **Ventricular ectopics (VEs)** = ominous sign (30% die suddenly)
  Increased risk of VEs in patient who have undergone ventriculotomy or RV-PA conduit

- Mortality rate: 30% in single-ventricle circulation
  ∴ refer the children to the specialized centre
High Risk: Cyanosis

- Recognized when deoxyhemoglobin > 2.38 g/dL
- Worse: agitation, crying, exercise
- Blunted ventilatory response to hypoxia
Cyanotic Heart

Normal

Hypoxemia
Low $\text{PaO}_2 / \text{SaO}_2$

More hemoglobin (carriages)
(this needs iron, a healthy bone marrow, and takes time)

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Cyanotic Heart

Hyperviscosity

Chronic hypoxia

Erythrocytosis

Dehydration

Hydration
Continue ASA

Mucosal bleeding

Visual change

Neurologic symptoms

Cyanotic Heart

Impaired liver function
VitK deficiency

↓ Coagulation factors

Platelet dysfunction

↑ Platelet microparticle
↓ Platelet aggregation

Platelet dysfunction

Local microcirculation sluggish

Hyperviscosity

Hyperviscosity

Bleeding caution

Coagulopathy

Cyanotic Heart

↓ Hepatic perfusion
↓ Glucose production
1969 Aug;40(2):209-16, Simultaneous Hypoglycemia and Acute Congestive Heart Failure

Produce more insulin
University of Copenhagen 2013

↓ Hypoglycemia

May aggravate myocardial failure
Practical Approach to Cardiac Anesthesia 5 ed 2013

Dextrose containing solution
Congenital Heart Disease & Hypoglycemia

- Prevalence of low FPG (≤80 mg/dl) was higher in the unrepaired (58%), Fontan (47%), and biventricular group (33%) than in the healthy control (11%)

- Lower FPG independently predicted the hospitalization (FPG ≤ 84 mg/dl) and mortality (FPG ≤ 80 mg/dl)
High risk: Complex lesions

- Single ventricle
- Balanced circulation physiology
- Cardiomyopathy
- Aortic stenosis
Risk Classification

Intermediate risk: Type of Surgery

• Intraperitoneal surgery
• Intrathoracic surgery
• Vascular reconstructive surgery
• Hypovolemia with massive blood transfusion
Risk Classification

Intermediate risk:

• Lesion: simple
• Age: under 2 years old
• Type of Surgery: Emergency
• Preoperative hospital stay: >10 days
• ASA physical status: IV or V
Risk Classification

Low risk:

• Lesion: physiologically normal or well-compensated
• Age: more than 2 years old
• Type of Surgery: Elective, Minor or body surface area
• Preoperative hospital stay < 10 days
• ASA physical status I-III
Anesthetic Considerations

1. Knowledge of underlying lesion and type of circulation: change in SVR/PVR? SpO₂?

2. Evidence of long-term complications or high-risk category

3. Venous access & IV hydration
Infective Endocarditis Prophylaxis

- Procedures on infected skin or musculoskeletal tissues

- Urinary tract procedural manipulation (e.g., cystoscopy) in the presence of enterococcal urinary infection or colonization

**BOX 38-7 Cardiac Conditions Associated With the Highest Risk of Adverse Outcome From Endocarditis, for Which Prophylaxis Is Recommended**

- Previous infective endocarditis
- Congenital heart disease*
  - Unrepaired cyanotic congenital heart disease, including palliative shunts and conduits
  - Completely repaired congenital heart defects with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure†
  - Repaired congenital heart disease with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibits endothelialization)
  - Cardiac valvulopathy that develops in cardiac transplantation recipients
# Infective Endocarditis Prophylaxis

<table>
<thead>
<tr>
<th>Situation</th>
<th>Drug</th>
<th>Single Dose 30-60 min Before Dental Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Adults</td>
</tr>
<tr>
<td>Oral</td>
<td>Amoxicillin</td>
<td>2 g</td>
</tr>
<tr>
<td>Unable to take oral medication</td>
<td>Ampicillin or</td>
<td>2 g IM/IV</td>
</tr>
<tr>
<td></td>
<td>Cefazolin/ceftriaxone</td>
<td>1 g IM/IV</td>
</tr>
<tr>
<td>Allergic to penicillins/oral</td>
<td>Cephalexin or</td>
<td>2 g</td>
</tr>
<tr>
<td></td>
<td>Clindamycin or</td>
<td>600 mg</td>
</tr>
<tr>
<td></td>
<td>Azithromycin/clarithromycin</td>
<td>500 mg</td>
</tr>
<tr>
<td>Allergic to penicillins/unable to take oral medication</td>
<td>Cefazolin/ceftriaxone or</td>
<td>1 g IM/IV</td>
</tr>
<tr>
<td></td>
<td>Clindamycin</td>
<td>600 mg</td>
</tr>
</tbody>
</table>

Vancomycin is an alternative for patients who are unable to tolerate a β-lactam or when the infective agent is considered to be methicillin-resistant *Staphylococcus aureus*.
Example 1: A 6-month-old boy with an unrepaired VSD presenting for emergency surgery for a scrotal swelling

**Physiology:** compensated

**Lesion:** simple

**Risk category:** intermediate (age<2 yr old, emergency surgery)

**Management:** depends on resource

**Anaesthesia considerations:**
- Possibility of poor cardiac reserve, avoid excessive anesthetic agents
- Avoid high FiO₂ (to minimize left-to-right shunt)
- Avoid air bubbles in venous lines (risk of paradoxical embolus)

**Endocarditis prophylaxis:** unnecessary
Example 2: A 6-yr-old boy with a repaired tetralogy of Fallot 8 months ago with no residual defect, presenting for dental extractions due to dental caries

**Physiology:** compensated

**Lesion:** simple

**Risk category:** low (elective, minor surgery, well-compensate simple lesion, age >2 yr old)

**Management:** perform procedure in local hospital.

  If VEs present on ECG—consider to be high risk

**Anaesthesia considerations:**

- Gas or i.v. induction

**Endocarditis prophylaxis:** unnecessary
Example 3: An 8-yr-old boy with Fontan circulation presenting for emergency surgery for reduction and fixation of a supracondylar fracture with neurovascular compromise.

**Physiology**: may be well-compensated

**Lesion**: complex single ventricle

**Risk category**: high (complex lesion, emergency)

**Management**: discuss with specialist.
- Transfer = risks limb loss from neuromuscular compromise
- Not transfer = risk life if local hospital anesthetist does not understand complex single-ventricle physiology
Example 3: An 8-yr-old boy with Fontan circulation presenting for emergency surgery for reduction and fixation of a supracondylar fracture with neurovascular compromise

Anaesthesia considerations:
- Full stomach may require rapid sequence intubation.
- Optimize pulmonary blood flow
  - Spontaneous ventilation
  - Avoid hypoxia, hypercarbia and atelectasis
  - Avoid high pressures, high PEEP, and long inspiratory times
  - Slight head-up position & raising legs

Endocarditis prophylaxis: unnecessary
Anesthetic Considerations

1. Knowledge of underlying lesion and type of circulation: change in SVR/PVR? SpO₂?

2. Evidence of long-term complications or high-risk category

3. Venous access & IV hydration

4. Recent URI/LRI: may cause changes in airway reactivity and PVR

5. Routine drug therapy: ACE-I, ASA, warfarin

6. Cautious sedative premedication: avoid distress, minimize oxygen consumption, not oversedation

7. Endocarditis prophylaxis
Intraoperative Considerations

1. Invasive monitorings: depend on the type of surgery & cardiac lesion
RA (Neuraxial block)

- Reduce stress response from pain better than opioids, benefit in thoracolumbar and lower extremities surgery.

- Optimize volume status and coagulation status.

- Slowly titrated epidural is preferable to rapid spinal block.

- RA or RA combined with GA has been used successfully in patients with single ventricles, shunt physiology, left-sided obstructive lesions, and pulmonary hypertension.
Intraoperative Considerations

1. Invasive monitorings: depend on the type of surgery & cardiac lesion

2. Choice of anesthesia: GA vs RA

3. Induction agent: inhalation vs intravenous
Induction of Anesthesia

**Inhalation**

- Rt.-to-Lt. shunt prolong inhalation induction

- Avoid in patient with poor cardiac function (myocardial depression)

- High cardiac output promotes clearance of anesthetics from lungs (greater effect in soluble agent)

**Intravenous**

- Prolong inductive time in low cardiac output state ∴ slow titration & reduced dose
1. Invasive monitorings: depend on the type of surgery & cardiac lesion

2. Choice of anesthesia: GA vs RA

3. Induction agent: inhalation vs intravenous

4. Maintain hemodynamic goals: preload, PVR, SVR, HR, contractility

5. Hypercyanotic spell during anesthesia
Ventilation & Oxygenation

- Balance PVR & SVR, avoid PHT

- Avoid excessive PBF in Lt.-to-Rt. shunt lesions (pulmonary congestion)

- Pulmonary edema: require higher ventilator pressures

- In Rt.-to-Lt. shunt: pulse oximetry overestimate arterial oxygen saturation as saturation decreases; EtCO$_2$ underestimate PaCO$_2$, and discrepancy worsens with hypoxemia

Intraoperative Tet Spell

• May be unable to do “squat position”

Treatment

• Volume expansion

• Alpha agonists: phenylephrine, levophed (↑SVR)

• Beta blocker: cease infundibular spasm

Intraoperative Considerations

1. Invasive monitorings: depend on the type of surgery & cardiac lesion
2. Choice of anesthesia: GA vs RA
3. Induction agent: inhalation vs intravenous
4. Maintain hemodynamic goals: preload, PVR, SVR, HR, contractility
5. Ventilation & Oxygenation
6. Hypercyanotic spell during anesthesia
7. Fluid management
Fluid Management

• “Goal-directed fluid therapy” is recommended for major procedures with substantial blood loss or fluid shifts

• Avoid fluid & salt overload while avoid hypovolemia

• Based on parameters beyond HR and BP:
  
  stroke volume variation (SVV), pulse pressure variation (PPV), doppler corrected flow time (FTc), central venous oxygen saturation
Intraoperative Considerations

1. Invasive monitorings: depend on the type of surgery & cardiac lesion

2. Choice of anesthesia: GA vs RA

3. Induction agent: inhalation vs intravenous

4. Maintain hemodynamic goals: preload, PVR, SVR, HR, contractility

5. Ventilation & Oxygenation

6. Hypercyanotic spell during anesthesia

7. Fluid management

8. Plan for extubation
Extubation

• “Fast-tract extubation" to avoid respiratory complications

• Risk factors for reintubation in PACU

**Patient factors:** age <1 yr, chronic pulmonary disease, preoperative hypoalbuminaemia, and renal insufficiency

**Surgical factors:** emergency case, head&neck, CVT and airway surgery, and operative time >3 hr

**Anesthetic factors:** NMBA, ASA physical status III
Intraoperative Considerations

1. Invasive monitorings: depend on the type of surgery & cardiac lesion

2. Choice of anesthesia: GA vs RA

3. Induction agent: inhalation vs intravenous

4. Maintain hemodynamic goals: preload, PVR, SVR, HR, contractility

5. Ventilation & Oxygenation

6. Hypercyanotic spell during anesthesia

7. Fluid management

8. Plan for extubation