ACHD: ANESTHETICS CONSIDERATIONS AND PITFALLS

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FIXED BUT NOT CURED

- With appropriate timing, few ACHD may be considered cured
  - ASD corrected before 12yo
  - VSD and PDA corrected before 2yo
- All other lesions carry residual problems
  - Structural, contractile, haemodynamic and electrophysiologic changes
  - End-organ dysfunction

# Anaesthesia Risk Assessment

<table>
<thead>
<tr>
<th></th>
<th>0</th>
<th>1</th>
<th>2</th>
</tr>
</thead>
<tbody>
<tr>
<td>CHD diagnosis</td>
<td>Simple (ASD)</td>
<td>Moderate (ASD + PS)</td>
<td>Severe (TOF)</td>
</tr>
<tr>
<td>CHD intervention</td>
<td>Corrected, no residual</td>
<td>Corrected with residual</td>
<td>Palliated</td>
</tr>
<tr>
<td>Ventricular obstruction</td>
<td>None</td>
<td>Yes, gradient &lt;49 mm Hg</td>
<td>Yes, gradient &gt;50 mm Hg</td>
</tr>
<tr>
<td>Ventricle, number, and position</td>
<td>2 LV is systemic Ventricle</td>
<td>1 LV is systemic ventricle</td>
<td>1 RV is systemic ventricle</td>
</tr>
<tr>
<td>Dysfunction systemic ventricle</td>
<td>Mild</td>
<td>Moderate</td>
<td>Severe</td>
</tr>
<tr>
<td>PVR</td>
<td>Normal, &lt;2 wood units</td>
<td>2–4 wood units</td>
<td>&gt;4 wood units</td>
</tr>
<tr>
<td>SAO₂</td>
<td>&gt;90%</td>
<td>75%–90%</td>
<td>&lt;75%</td>
</tr>
<tr>
<td>Hct</td>
<td>30%–45%</td>
<td>25%–30%, or 45%–65%</td>
<td>&gt;65%</td>
</tr>
<tr>
<td>Arrhythmias</td>
<td>Seldom</td>
<td>Atrial level</td>
<td>Ventricular level</td>
</tr>
<tr>
<td>No. of cardiac medications</td>
<td>1</td>
<td>2</td>
<td>≥3</td>
</tr>
</tbody>
</table>

Add points for each line and then assess the risk according to the total sum: 0–6 points, minimal perianesthesia risk, (outpatient), 6–13 points, moderate perianesthesia risk (possible same day admit), 14–20 points, high perianesthesia risk (possible ICU admit).

Modified from Mosad E, presented during Society of Cardiothoracic Annual meeting 2002.

ASD, atrial septal defect; Hct, hematocrit; LV, left ventricle; PS, pulmonary stenosis; PVR, pulmonary vascular resistance; RV, right ventricle; SAO₂, systemic oxygen saturation; TOF, tetralogy of fallot.

WHAT DO WE NEED TO KNOW?

- Significant intracardiac shunt
- More than mild PHT
- More than moderate LV or mild RV dysfunction or failure
- Functional univentricular heart
- Systemic right ventricle
- More than mild obstructive valvular or vascular disease
- Significant congenital coronary artery abnormalities
- New-onset of arrhythmias

PREOPERATIVE EVALUATION

- Accurate history: 25 to 50% of patients can not name or describe their diagnosis
- Spontaneous exercise limitation
- Long-term cardiac or non-cardiac complications
- Recent TTE, laboratory data, functional lung testing
- Multidisciplinary evaluation

(Foster E, et al. JACC 2001;37:1176-83)
ANAESTHETIC PLAN

• Clear understanding of pathophysiology
• Premedication according to pathology: cave opioids, deshydration
• Avoid systemic embolization and air bubbles
• Restricted venous and arterial accesses
• Defibrillator pads in case of arrhythmias
• AB prophylaxis following guidelines: previous endocarditis, unrepaired cyanotic CHD, up to 6 mo following implantation of extraneous material, residual defect at the site of a patch or device

(Seal R. Ped Anesth 2011;21:615-22
CYANOTIC LESIONS

- Inadequate pulmonary blood flow with R-L shunt or excessive pulmonary blood flow with L-R shunt and low CO
- Multisystemic effects
  - Erythrocytosis, hyperviscosity, iron deficiency (rigidity $\uparrow$)
  - $\uparrow$ hemolysis -> biliary lithiasis; thrombosis
  - Platelets $\downarrow$, von Willebrand $\downarrow$, platelets survival $\downarrow$
  - False values of Quick, PTT (citrate)
  - CNS dysfunction (abscess, stroke, embolizations)

CYANOTIC LESIONS

- Increased risk of ventricular dysfunction and myocardial ischemia
  - Impaired oxygen transport
  - Decreased diastolic perfusion pressure (shunts, aorto-pulmonary collaterals)
  - Impaired myocardial microvascular perfusion (erythrocytosis and hyperviscosity)
- Blunted hypoxic ventilatory response
- Preservation of response to hypercarbia

(Seal R. Ped Anesth 2011;21:615-22)
ANAESTHESIA AND CYANOTIC LESIONS

- Maintain favorable PVR
- Decrease R-L shunt: normovolemia, ↑ SVR
- Haemodynamic equilibrium and goal: Qp ≅ Qs
- Optimize hematocrit
- Modulate RVOTOT if present
- High FiO2
- $P_{ECO_2}$ underestimates PaCO$_2$

EISENMENGER

- Most frequent cause of cyanosis in adults
- Reversal of long-standing untreated L-R intra- or extracardiac shunt resulting in a R-L shunt
- Fixed PHT, RV dysfunction + TR
- Precarious balance of systemic, pulmonary and myocardial perfusions
- Perioperative mortality rate 4 to 30%

(Bennett JM, et al. J Clin Anaesth 2014;26:286-93
EISENMENGER

- Increase in BP  ➔  R-L shunt
- Sudden increase in PVR  ➔  acute RV failure and  ➔  R-L shunt
- R-L shunt worsens cyanosis  ➔  CV collapse and death
- Symptomatic RV failure, arrhythmias and every acute changes in pre- or afterload associated with high morbidity and mortality
- High risk of perioperative bleeding

ANAESTHESIA AND EISENMENGER

- PVR less reactive as in PHT patients
- Most anesthetics have only small effect on the PVR except NO$_2$, desflurane and ketamine (adults)
- Maintain the awake haemodynamic of the patient: PVR, SVR, HR, contractility
- Allow max haemodynamic fluctuation of 15% of initial value
- Prevent arrhythmias

OPTIMIZE VENTILATION

31yo man with Eisenmenger

- Eisenmenger secondary to large aorto-pulmonary window
- Acute onset of severe chest pain
- PaO2 6.9kPa, SaO2 90%, Ht 42%
- TTE and CT: PA dissection

31yo man with Eisenmenger

- Emergent repair: graft replacement and reconstitution of aorto-pulmonary window
- Hemodynamic collapse by induction, emergent CPB
- Severe RV dysfunction and hypoxemia (SaO2 50%) under high dose of inotropes and NO -> central ECLS
31yo man with Eisenmenger

- Emergent repair: graft replacement and reconstitution of aorto-pulmonary window
- Hemodynamic collapse by induction, emergent CPB
- Severe RV dysfunction and hypoxemia -> ECLS
- Unsuccessful weaning attempts at 3 and 5 days
- Vv-ECMO from day 5 to day 17 (Avalon)
- Extubation after 3 days on vv-ECMO
31yo man with Eisenmenger

- Precarious hemodynamic balance by Eisenmenger
- Emergent surgery on CPB bound with very high risk
- Few previous case reports
  - Immediate listing for heart-lung transplantation
  - Medical management
- Diagnosis is rare in living subjects

(Westaby S: NEJM 2007;356:2110-2
CONCLUSION

- Patients with bicuspid AV, small atrioventricular defects, repaired ASDs and mild pulmonic stenosis may be cared in peripheral hospital.
- Patients with moderate or complex ACHD should be managed in a dedicated ACHD center.
- High risk patients: poor functional class, PHT, cyanosis, functional single ventricle or heart failure.
THANK YOU