

ANAESTHESIA AND EISENMENGER'S SYNDROME

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Introduction:

Pulmonary vascular pathology relating to a ventricular septal defect was first described by Eisenmenger in 1897. Eisenmenger's syndrome (ES) is now used to describe the sequelae consequent upon irreversible changes in the pulmonary vasculature and a high pulmonary vascular resistance, secondary to a large congenital communication between pulmonary and systemic circulations that has resulted in a reversed (pulmonary-to-systemic) or bidirectional shunt and chronic hypoxaemia. Advances in medical care now allow these patients to survive well into adulthood.

Epidemiology:

The prevalence of adult patients with pulmonary arterial hypertension associated with congenital heart disease in Western Europe ranges between 1.6 and 12.5 cases per million adults, with 25–50% of this population affected by ES. The incidence is higher in developing countries.

Pathology:

In patients with a large non-restrictive ventricular septal defect or ductus arteriosus, shunt volume and direction are determined mainly by the pressure gradient between systemic and pulmonary circulations. In contrast, patients with a large atrial communication may have right-to-left shunting due more to reduced right ventricular compliance (a consequence of right ventricular hypertrophy) than pulmonary hypertension. This latter group of patients develops ES rarely and only later in life. All forms of pulmonary arterial hypertension have a common characteristic histopathological pattern of vascular remodelling. As pulmonary blood flow increases, endothelial cells in the pulmonary vasculature are subjected to increased shear stress that leads to:

- stimulation of collagen deposition
- smooth muscle proliferation within vessel walls
- endothelial dysfunction

Disease progression is associated with overproduction of vasoconstrictors, promoters of smooth muscle and fibroblast proliferation, and pro-coagulant factors.

Vital organ function in patients with ES deteriorates progressively over time. Cyanosis and secondary erythrocytosis are frequently observed. Impaired exercise tolerance and dyspnoea on exertion tend to increase in severity. Haemoptysis may occur as a result of rupture of dilated bronchial arteries. Partial thrombosis of enlarged proximal pulmonary arteries is found in up to 20% of adults with ES and may cause peripheral embolization and pulmonary infarctions. Cerebrovascular accidents may occur as a result of paradoxical embolization, venous thrombosis of cerebral vessels or intracranial haemorrhage. In addition, affected patients are at risk of brain abscesses, bacterial endocarditis and pneumonia. ES patients may have syncope as a result of inadequate cardiac output, or as a result of an arrhythmia. Symptoms of heart failure, which are uncommon until the disease is far advanced, also portend a poor prognosis. Renal dysfunction, secondary to heart failure and/or glomerular abnormalities (as a consequence of hypoxaemia), is common and progressive.

ANAESTHETIC MANAGEMENT FOR ES PATIENTS HAVING EMERGENCY OR MAJOR ELECTIVE SURGERY:

Preoperative assessment and optimization:

History:

- Nature of lesion
- Past medical & surgical history; recent anaesthetic history
- Present medication, e.g. epoprostenol, warfarin, sildenafil, bosentan
- Current condition; exercise tolerance, haemoptysis, syncopal episodes
- Other problems; e.g. Down's syndrome

Examination:

- Cyanosis, clubbing, plethora
- Hepatomegaly, oedema, ascites, jugular venous pressure
- Volume status; heart rate, blood pressure, capillary refill time, mucous membranes
- Chest auscultation; heart murmurs; air entry, added sounds, etc.

Investigations:

- FBC (high Hb expected); INR (high?)
- U & E; renal dysfunction?
- Lactate; may be elevated due to low cardiac output
- CXR: large heart & prominent main pulmonary artery; oligoemic lung fields
- ECG: right axis deviation; right ventricular hypertrophy; right bundle branch block; ± large RA
- Echocardiography & cardiological opinion if recent deterioration

Optimization:

- Oxygen (to keep O₂ sats >80%) for at least 4 h prior to surgery
- Order FFP if INR >2.5; stop long-acting anti-thrombotics
- Start short-acting anti-thrombotic regimen
- Book ICU bed (if not there already)
- Give anxiolytic (e.g. midazolam orally) as indicated
- Apply local anaesthetic cream (e.g. Ametop) to cannulation sites (both arterial and venous)
- Explanation of risks given to patient and/or parents; obtain informed consent

Anaesthetic room management

- Insert peripheral venous cannula (s); give antibiotics
- Insert arterial cannula; take baseline ABGs
- Insert CVP but not PAC (increased risk of haemorrhage, arrhythmias, embolization, infection) Preoxygenation (5 min 100 %)
- Give fluids as required; aim for CVP >10 mm Hg; special care to avoid air bubbles in venous line
- Induction with ketamine 2 mg/kg & opioid (e.g. fentanyl 5 µg/kg)
- Avoid thiopental, propofol, etomidate
- Vecuronium (probably) relaxant of choice; intubate and ventilate
- Insert urinary catheter
- Avoid spinal; epidural may be appropriate in some cases

Intra-operative management

- Maintain preload; expect high CVP
- Maintain anaesthesia using opioid & isoflurane (≤ 1 MAC) or TIVA (?)
- Maintain high normal HR; use anticholinergics to prevent opioid-induced bradycardia
- Intensive monitoring (routine +):
 - IABP; CVP; urine output
 - ABGs, electrolytes, pH; lactate
 - BIS (to optimise anaesthetic depth)
 - TOE/Doppler (if relevant expertise)
- Keep Hct > 30%; transfuse as needed
- Use IPPV to control PaCO₂ (aim for 5 kPa); remember that EtCO₂ is unreliable
- Avoid hypoxia, acidosis, hypercarbia

Treatment of deterioration in PaO₂:

- Increase FiO₂ to 1.0
- Ensure anaesthesia & analgesia are adequate

- Check both lungs being inflated
- Check adequate filling (CVP >10 mm Hg)
- Hyperventilate
- Fully correct any acidosis using i.v. sodium bicarbonate
- Start inhaled nitric oxide (5-20 ppm)
- Infuse norepinephrine (start @ 0.1 µg/kg/min)

Postoperative management:

- ICU care; continue CVS monitoring for ≥48 h
- Maintain high FiO₂ for at least 48 h; respiratory support as indicated
- Maintain adequate analgesia with PCA or PCEA as appropriate
- Resume antithrombotic agents as soon as possible after surgery
- Preoperative vasodilator medication continued, e.g. nebulised epoprostenol, sildenafil
- N.B. Most deaths in ES patients are postoperative!

Prognosis:

In a series of 100 patients with Eisenmenger's syndrome listed for transplantation, the survival of patients who did not receive transplants was 97% at 1 year, 89% at 2 years and 77% at 3 years. Pregnancy in Eisenmenger's patients is associated with an increased rate of deterioration, death (in up to 50%) and spontaneous abortion (in up to 40%). Perioperative mortality rates for elective non-cardiac surgery & anaesthesia in ES patients is about 10%; for emergency surgery up to 30%.

References

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Eisenmenger's syndrome is defined as the process in which a long-standing left-to-right cardiac shunt caused by a congenital heart defect (typically by a ventricular septal defect, atrial septal defect, or less commonly, patent ductus arteriosus) causes pulmonary hypertension and eventual reversal of the shunt into a cyanotic right-to-left shunt. Because of the advent of fetal screening with echocardiography early in life, the incidence of heart defects progressing to Eisenmenger's has decreased. The Eisenmenger syndrome is a form of cyanotic congenital heart disease that does not usually respond to corrective surgery, and its occurrence is very rare in pregnant women. There are ongoing...^Â Anesthesia Eisenmenger syndrome Pregnancy Surgery. This is a preview of subscription content, log in to check access. References.