Eisenmengers Syndrome

The development of pulmonary vascular disease (pulmonary hypertension) in the presence of systemic to pulmonary communication with associated cyanosis. (must exclude other causes of pulmonary hypertension)

Anesthetic Considerations:

6. Anesthetic considerations for pulmonary hypertension: avoidance of hypoxemia, hypercapnia, acidosis, hypovolemia
7. Avoidance of hypovolemia
8. Maintain SVR to avoid R to L shunting
9. Strict avoidance of intravenous air injection (possible paradoxical embolus)
10. DVT prophylaxis
11. Increased risk of arrythmias (A Fib, A Flutter, sudden cardiac death)
12. Caution with laparoscopic surgery (increased CO2 absorption, pneumoperitoneum and associated increase in airway pressures secondary to abdominal insufflation)
13. Consider phlebotomy in patients with Hct >65% with isovolemic replacement to improve ssx hyperviscosity

Anesthetic Goals:

1. Maintenance of systemic vascular resistance and avoidance of R to L shunting
2. Avoid increases in PVR (hypoxemia, hypercapnia, acidemia, increased airway pressures and PEEP)
3. Maintenance of preload

PATHOPHYSIOLOGY:

- Initially there is increased pulmonary blood flow secondary to L → R shunt either via atrial communication/ ventricular communication/ aortic shunts
    1. Atrial Shunts
        i. ASD
        ii. Common atrium
        iii. TAPVD
        iv. Partial AVPD
    2. Ventricular Shunts
        i. VSD
        ii. Single ventricle
        iii. TGA with VSD
        iv. DORV
        v. AV canal
    3. Aortic Shunts
        i. PDA
        ii. Truncus arteriosus
        iii. Pulmonary atresia with VSD and collaterals
- Shunt fraction (Qp/Qs) indicates ratio of pulmonary blood flow: systemic blood flow
    1. Normal Qp:Qs = 1:1
    2. L to R shunt Qp:Qs = >1
    3. R to L shunt Qp:Qs= <1
- Most commonly develops as a result of increased shunt fraction (i.e. increased L to R flow); type of lesion correlates with frequency of occurance (i.e. VSD>ASD) and larger shunts also seem to carry increased risk
- Increased L to R flow leads to pulmonary hypertension (pulmonary arteriolar hypertension) and subsequent reversal of shunt such that shunt fraction <1 i.e. R to L shunt
- Prognosis is better than those with primary pulmonary hypertension, but ~20y decrease in life expectancy in those with simple unrepaired CHD, and ~40y decrease in those with complex congenital heart disease

PREDISPOSING AND/OR ASSOCIATED FACTORS (SEE ABOVE):

1. Congenital heart disease with systemic – pulmonary communication
    i. ASD
    ii. VSD
    iii. Aortic shunt

TREATMENT:

- Treatment of underlying cardiac lesion can be associated with reversal of pulmonary vascular hypertension.
- Symptomatic treatment of pulmonary hypertension:
    - Prostaglandins
    - Phosphodiesterase inhibitors
    - Inhaled NO
    - CCBs

PRIORITYRESUSCITATIONHISTORYEXAMLABSPFTS(ETC)REGIONAL

Preoperative:

(Emergent, Urgent, Elective)

5. History
    a. Type of systemic to pulmonary communication
    b. Functional capacity/ exercise tolerance
    c. Frequency of cyanotic spells
    d. Palpitations
    e. Ssx increased blood viscosity (including visual disturbances, headache, dizziness, paresthesias, pulmonary infarction, pulmonary arterial thrombosis)

6. Physical
    a. Airway exam
    b. Assessment of volume status

c. Focused cardiorespiratory exam (rule out ssx decompensated RV failure)
d. Auscultation of precordium for murmurs, abnormal heart sounds (loud P2)

7. Investigations
    a. Labs (CBCd and Hct, lytes, BUN, Cr INR, PTT, type and screen + crossmatch)
    b. ECG – RVH, right axis deviation, afib/flutter

Optimization

5. Echocardiographic estimation of RVSP and shunt fraction may be useful
6. No specific treatment has been proven to provide sustained decrease in PVR
7. IV epoprostenol (or inhaled?) may be useful
8. Consider phlebotomy if Hct>65% or ssx increased blood viscosity
9. Multidisciplinary preoperative optimization including involvement of pulmonologist/congenital heart disease specialist; assessment of oxygen responsiveness of hypoxemia
10. Eisenmengers is a contraindication to surgical correction of the congenital heart defect that was responsible for the original L to R intracardiac shunt
11. Lung transplantation with repair of the cardiac defect OR combined heart lung transplantation may be the only treatment option in end stage disease

Room Preparation/Setup

- standard emergency drugs
- standard machine check (including suction, gas line supply, emergency O2 supply, inhalational agents, CO2 absorbant, circuit leak test)
- standard CAS monitors, consider 5 lead ECG +/- art line
- consider central line for ability to administer norepineprhine or other alpha adrenergic/SVR maintaining agent

Induction:

- balanced induction with anticipation of vasodilation accompanied by administration of induction agents
- consider phenylephrine or afterload increasing agent to temporize
- caution with decreased SVR with epidurals/neuraxial techniques. Spinal would be relatively contraindicated but a slowly titrated epidural has been successfully reported in the literature
- strict avoidance of intravascular air injection

Maintenance:

- caution with high PEEP and increased airway pressures
- avoidance of systemic vasodilation and R to L shunt

Emergence:

- increased risk of paradoxical embolization
- treatment of postoperative pain, shivering, hypoxemia and hypercapnia as well as acidemia
- consider postoperative disposition – observation bed vs ICU

REFERENCES:

Stoeltings Anesthesia and Coexisting Disease, Chapter 3
UptoDate: Medical Management of EIsenmenger Syndrome
UptoDate: Evaluation and Prognosis in Eisenmenger syndrome