Management of a Patient with Eisenmenger Syndrome Requiring Emergency Laparotomy in a District General Hospital - A Case Report

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Background

Eisenmenger Syndrome is a cyanotic heart condition in which long-standing left-to-right shunt causes pulmonary hypertension, pulmonary vascular remodelling, right ventricular hypertrophy and reversal of shunt¹. Surgery is usually managed in specialist centers.

Case Report

A 35 year old presented with an incarcerated umbilical hernia requiring urgent surgery.

Past medical history:

- Down syndrome, obesity and obstructive sleep apnoea requiring CPAP and home oxygen.
- Complete atrioventricular septal defect and established Eisenmenger Syndrome. A previous echocardiogram showed right ventricular systolic pressure of 90mmHg.

Multidisciplinary planning:

- Discussed early with patient's cardiologist and intensive care consultant at tertiary centre, as well as local intensive care consultant and surgeon. On balance of risks, a decision was taken to operate locally as was believed to be at immediate risk of bowel infarction.
- Pharmacy sourced specialist drugs, including IV sildenafil and nebulised iloprost.

Anaesthesia:

- Pre-induction noradrenaline and milrinone infusions.
- Fentanyl 200mcg, ketamine 100mg, rocuronium 70mg. Sevoflurane in O₂/air.
- 90 minute surgical time - incarcerated bowel re-perfused successfully avoiding resection.
- Having been stable intra-op, at end of surgery, patient deteriorated during transition to spontaneous ventilation with SpO₂ dropping to 55% and BP to 50/30.
- Noradrenaline, adrenaline, milrinone and selective pulmonary vasodilators sildenafil and nebulised iloprost were used to rebalance pulmonary and systemic circulations.
- Transferred post-operatively to ICU and switched to vasopressin and milrinone infusions. Iloprost and sildenafil were continued. Extubated on day two and discharged after a week.

Learning points

- Patients with specialist problems usually have a long-standing tertiary team who should be involved in any decisions made about their care.
- Eisenmenger physiology necessitates maintaining a delicate balance between systemic and pulmonary circulations. The specialist team was able to provide invaluable advice.
- Specialist drugs can be obtained by pharmacy from other hospitals at relatively short notice. In this case, they were probably life-saving.

References:

- 1) Intensive Care Medicine 2011;37:502-9.