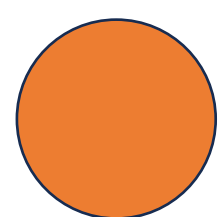


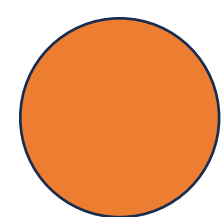
Congenital heart disease, associated pulmonary arterial hypertension and pregnancy: a triple challenge for the obstetric anaesthetist

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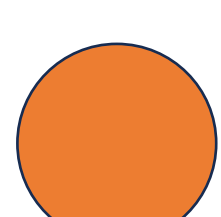
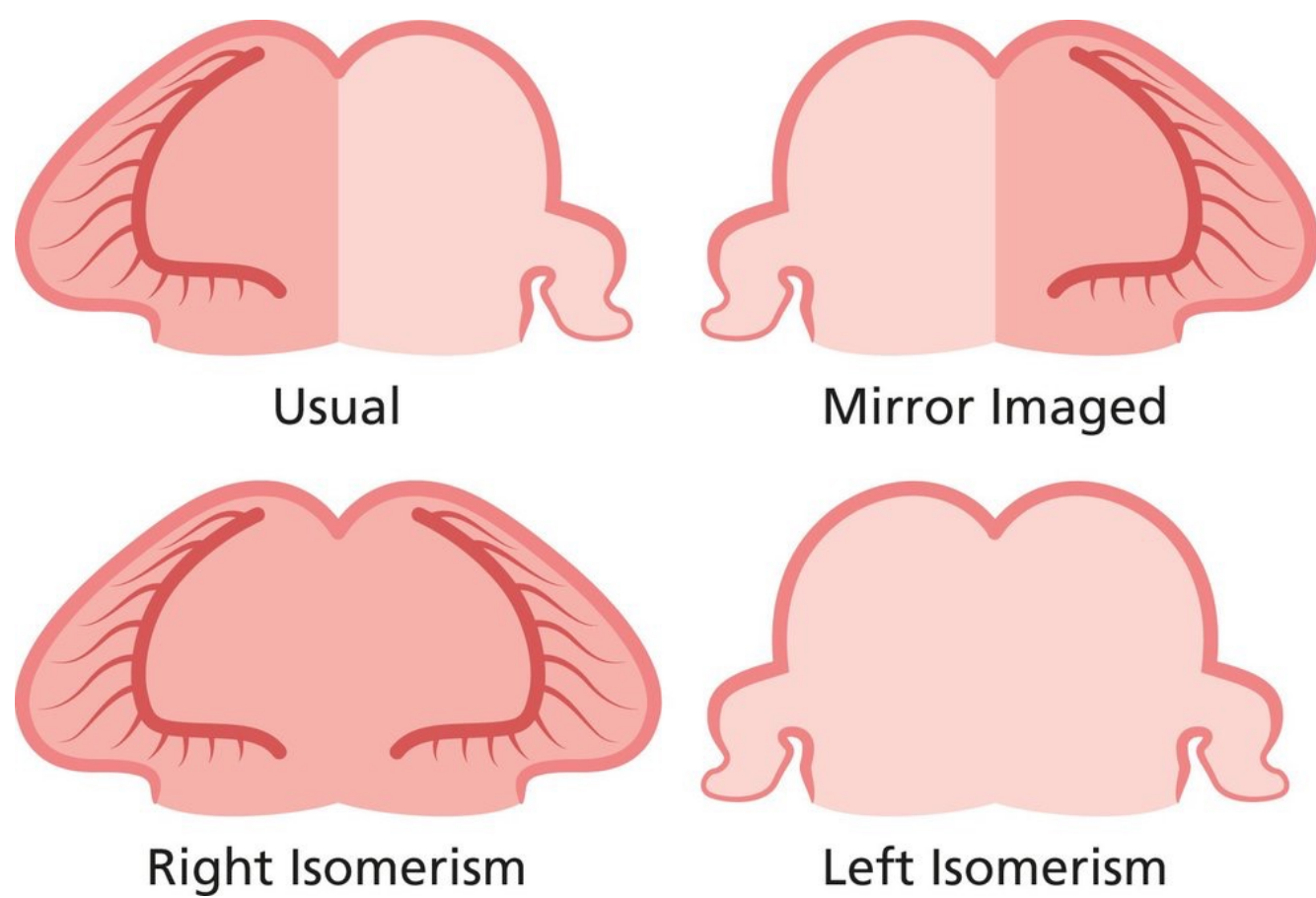
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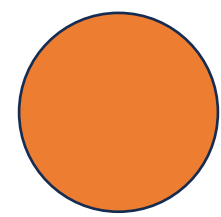
Pulmonary hypertension in pregnancy has a mortality of 9-25%¹



Atrial isomerism is a rare congenital disorder where a patient has two copies of either the left or right atrium



We present the case of a 33 year old woman with pulmonary arterial hypertension(PH) associated with congenital heart disease who underwent a planned caesarean section(CS) for her third delivery



The pregnancy was complicated by a persistent junctional bradycardia which precipitated a significant clinical deterioration

Case

- PMHx: established PH at presentation aged 19 due to congenital abnormalities: atrial septal defect(ASD), partial anomalous pulmonary venous drainage (PAPVD), left atrial isomerism
- Corrective surgery: baffling of PAPVD, fenestrated patch repair of ASD. Low risk PH at follow-up.
- Presented with a 3rd, unplanned pregnancy 3 years after her previous delivery.
- PH stable at booking. PASP 38mmHg with good biventricular function. Noted to have junctional bradycardia 40-45bpm.
- Clinical deterioration from 18 weeks with increasing breathlessness on exertion. Echocardiogram and BNP remained stable.
- Deterioration was attributed to the junctional bradycardia preventing augmentation of cardiac output in response to pregnancy and periods of increased activity.
- 24 hour tape showed some augmentation of heart rate on exertion. Electrophysiology(EP) team advised not for invasive pacing unless syncopal episodes occurring.
- Caesarean section(CS) planned for 34/40 gestation due to continuing deterioration.

Management

Pre-op

- Admitted two days prior to CS to administer steroids for fetal lung maturation, insertion of arterial and central lines and to commence isoprenaline infusion in intensive care.

Intra-op

- Anaesthesia established with incremental CSE: 2.5mg hyperbaric bupivacaine + 350mcg diamorphine. Subsequent epidural top-ups with 0.5% Levopbupivacaine.
- Heart rate maintained at 60-70bpm with isoprenaline infusion
- Fetus received CPAP on delivery
- Uterotonic = oxytocin 5u given as slow infusion.
- 500ml blood loss. 300ml crystalloid given in 50ml aliquots, titrated to CVP in addition to 1 unit of blood.

Post-op

- Transferred to ICU for weaning of isoprenaline. HR returned to 40-45bpm.
- Baby received further CPAP in NICU.
- Both discharged home after 7 days.

Discussion

- Pulmonary hypertension is defined as a mean pulmonary artery pressure greater than 20mmHg. ²
- Risk in pregnancy correlated to the degree of right heart failure.
- The normal physiological increase in blood volume (40%) and cardiac output (50%)³ is poorly tolerated by the right ventricle and may lead to catastrophic cardiovascular collapse.

- Atrial isomerism has an incidence of 1 in 10,000.⁴
- Conduction defects are common; absence of the sino-atrial node and/or atrioventricular node is typical in left atrial isomerism, resulting in brady-arrythmias.

- Achieving a safe CS delivery required a multi-disciplinary approach.
- EP team advised on the risk/benefit of invasive pacing and were on standby in case of need for emergency invasive pacing.
- PH team present in theatre to administer Epoprostenol or nitric oxide if acute decompensation occurred eg. when placental auto-transfusion and removal of aortocaval compression produce an increase in right ventricular filling.

- Isoprenaline enabled the augmentation of cardiac output.
- The incremental CSE technique was crucial to providing cardio-stable anaesthesia.⁵
- No vasopressor required but first choice would be Vasopressin due to limited effect on pulmonary vasculature.⁶

- The patient has returned to her pre-pregnancy exercise tolerance.
- Despite counselling after all 3 pregnancies, she declined contraception prior to discharge from hospital.
- She will be closely monitored for 6 months post-partum with regular echocardiography and BNP measurement.
- She is likely to need a permanent pacemaker in the future.

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