CASE REPORT

Postnatal diagnosis of maternal congenital heart disease: missed opportunities

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SUMMARY

A 30-year-old primigravida with no known comorbidities presented to the emergency department at 29+6/40 gestation, with breathlessness. The initial diagnosis was pulmonary embolism, which was later revised following initial investigations and considered to be pre-eclampsia/HELLP (haemolysis, elevated liver enzymes, low platelets) syndrome. Following caesarean section and delivery of a live baby, the patient had episodes of cyanotic hypoxia and was admitted to intensive care. A provisional diagnosis of idiopathic pulmonary hypertension was performed. Decompensation led to transfer to a specialist intensive care unit for extracorporeal membrane oxygenation, where a diagnosis of patent ductus arteriosus and Eisenmenger's syndrome was made. Heart disease is the leading indirect cause of maternal death, and Eisenmenger's syndrome in pregnancy carries a 50–65% mortality. A literature review demonstrated that this is the only reported case of a postnatal diagnosis of Eisenmenger's syndrome. We considered missed opportunities to make an earlier diagnosis, so that patients and doctors will benefit from the lessons we learnt.

BACKGROUND

Cardiovascular disease affects 0.5–4% of pregnant women^{1–3} and is a major cause of morbidity and mortality. The last CMAE (Centre for Maternal and Child Enquiries) report showed cardiac disease was the leading cause of indirect maternal death, and the leading cause of death overall.^{1–4} Specifically, the number of women with congenital heart disease in pregnancy has increased due to earlier diagnosis and treatment in childhood.⁵

Patent ductus arteriosus (PDA) occurs in 1 in 2000 births and accounts for 5–10% of congenital heart disease.⁶ The ductus arteriosus normally closes within the first few days of birth⁷ and failure of the ductus to close results in a spectrum of signs and symptoms. Patients may be asymptomatic, or present with symptoms of chronic hypoxaemia and polycythaemia, including headache and visual disturbance.² Clinical signs include a machinery murmur at the left sternal edge that radiates to the back, clubbing and cyanosis.⁶ Pulmonary oedema is a late sign and occurs due to right ventricular dysfunction.⁸

Eight per cent of all patients with congenital heart disease develop Eisenmenger's syndrome.² Eisenmenger's syndrome describes the reversal of blood flow through a cardiac shunt as a result of pulmonary hypertension. Patients with Eisenmenger's syndrome can present with cyanosis, clubbing,

polycythaemia and heart failure. The mortality is quoted as being 30–70%,⁹ and women with Eisenmenger's syndrome are advised against pregnancy.^{2 10–12} Despite this, there are several reported cases of successful pregnancies and deliveries. Women with Eisenmenger's syndrome should have thorough pre-pregnancy counselling and care should be provided by a multidisciplinary team including a cardiologist, in a specialist centre.¹³

CASE PRESENTATION

A 30-year-old primigravida who had moved to the UK from Pakistan 24 months prior to her booking visit at 12 weeks, was deemed a low risk pregnancy. She had no known comorbidities and all her booking bloods were normal except her raised haemoglobin (17.1 g/L) and low platelets ($133 \times 10^9/L$). Further blood tests were normal and she continued her antenatal care in the community.

Operative day minus 1: Initial presentation

The patient presented to our district general hospital emergency department at 29+6 weeks gestation. She described a 7-day history of shortness of breath with no chest pain or palpitations. Examination demonstrated tachycardia at 104 bpm, a respiratory rate of 20, oxygen saturations of 93% on air and a blood pressure of 145/86 mm Hg. Auscultation of the patient's chest and her chest X-ray were normal. With a provisional diagnosis of pulmonary embolus (PE), she was given a therapeutic dose of low molecular weight heparin in the emergency department.

Review of her admission blood tests showed haemoglobin of 156 g/L, thrombocytopenia (platelets $67 \times 10^9/L$) and deranged liver function tests (alkaline phosphatase 186 U/L, aspartate aminotransferase 66 U/L). She was also found to have brisk reflexes and proteinuria. The clinical picture was of evolving pre-eclampsia/HELLP (haemolysis, elevated liver enzymes, low platelets) syndrome spectrum and she was immediately transferred to the delivery suite. Her Early Warning Score (EWS)¹⁴ in the emergency department had been four, but when recalculated, her Modified Obstetric EWS (MOEWS)¹⁵ was 10.

On delivery suite, a magnesium sulfate infusion and oral labetalol were started to treat severe pre-eclampsia and prevent seizures. Further examination demonstrated a loud second heart sound and an audible third heart sound. Evidence of right ventricular strain was present on the patient's ECG, so an urgent echocardiogram was requested. This showed right ventricular hypertrophy with normal

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left ventricular function and normal pulmonary artery pressures. The findings were inconsistent with a PE, and objective testing was recommended to exclude PE.

Day 0: Operative day

There were concerns about the increasing oxygen requirement and developing pulmonary oedema, so a multidisciplinary decision was made for a caesarean section. The patient received platelets prior to a spinal anaesthetic and remained cardiovascularly stable throughout an uneventful caesarean section. She delivered a live male child weighing 1207 g, who was admitted to neonatal intensive care due to prematurity.

Postoperatively, the patient remained stable in a high care environment on delivery suite. She had frequent arterial blood gas monitoring, and continued to receive magnesium sulfate, labetalol and low molecular weight heparin.

Day 1

The patient's oxygen requirements continued to increase, so consultant respiratory and cardiologist opinions were sought. A repeat chest X-ray was normal and arrangements were made for a CT-pulmonary angiogram (CTPA).

Later that day, the patient had two acute self-limiting episodes of severe cyanotic hypoxia and hypotension with decreased levels of consciousness, so was transferred to the intensive care unit (ICU). The CTPA did not show a PE, but there was evidence of ascites, hepatomegaly, pulmonary hypertension and right heart failure. The patient was managed with oxygen and continuous positive airway pressure for the first 24 h in the ICU.

Day 2

Periods of acute decompensation would resolve spontaneously, alerting the ICU consultant to suspect a right to left cardiac shunt. The cardiology team re-reviewed the initial echocardiogram images taken on admission. It was noted that the pulmonary artery was enlarged at 2.6 cm, the right ventricle was pressure overloaded and pulmonary artery pressures were extremely high at 100 mm Hg. No shunt was seen. The cause of hypoxia was considered to be idiopathic pulmonary hypertension (IPH).

The patient was intubated due to cardiorespiratory instability and had trials of iloprost and milrinone. She also required nor-pinephrine and dobutamine infusions, along with epinephrine boluses to maintain cardiovascular stability.

A referral was made to a regional unit and an extracorporeal membrane oxygenation (ECMO) transfer team arrived. The bedside transoesophageal echocardiogram (TOE) with a bubble test performed by the transfer team was negative, indicating that there was no apparent cardiac shunt. ECMO was inappropriate with no reversible cause and a working diagnosis of IPH was performed.

The patient was transferred for specialist care and inhaled nitric oxide was successful in decreasing the pulmonary artery pressures. A further TOE at this stage showed a PDA with right to left shunt. The patient was diagnosed as having Eisenmenger's syndrome.

DIFFERENTIAL DIAGNOSIS

The most likely initial diagnosis in this patient was thought to be PE. A primigravida with no known medical history presented with acute respiratory symptoms. The hypercoagulable state in pregnancy, in combination with acute shortness of breath, a sinus tachycardia and hypoxaemia point towards a possible PE.

The initial diagnosis was reviewed when blood results showed thrombocytopaenia and abnormal liver function tests. Pre-eclampsia was then suspected due to the accompanying hypertension, proteinuria and brisk reflexes.

Initially, the worsening hypoxaemia was attributed to be pulmonary oedema, part of the pre-eclampsia/HELLP spectrum. It was not possible to rule out a PE, and therapeutic doses of low molecular weight heparin continued postoperatively until the CTPA was performed on day 1.

These are the most likely diagnoses and were considered early, but as the clinical picture continued to change, the differentials were continually questioned.

OUTCOME AND FOLLOW-UP

Day 7

Following the diagnosis of Eisenmenger's syndrome, the patient was referred to a specialist unit with an interest in adult congenital heart disease. She first had venoarterial inserted, followed by insertion of venovenous ECMO lines.

Day 22

The outcome of a multidisciplinary team meeting was that it was unlikely that the patient could be weaned off ECMO and would require a heart-lung transplant.

Day 44

The patient was taken to theatre for a heart-lung transplant. Unfortunately, she did not survive to have the procedure.

DISCUSSION

This is the first case, to our knowledge, of a patient who had survived into adulthood, through pregnancy and delivery and had only been diagnosed with a PDA and Eisenmenger's syndrome postnatally. There are rare cases where Eisenmenger's syndrome has been diagnosed antenatally.^{2 13–16} The paucity of data and case reports is likely to be due to advances in modern medicine with earlier diagnoses and treatment, high fetomaternal mortality and the contraindication of pregnancy in patients with Eisenmenger's syndrome.^{2 10–12} Mortality with Eisenmenger's syndrome is associated with late diagnosis and late hospital admission.³ Below, we explore the timely investigations and management of our case, and what could be carried out differently in the future.

Signs and symptoms explained

Patients with a PDA, as described in our case, may be asymptomatic with no clinical signs during childhood.⁶ Symptoms develop later with the most noticeable being a change in exercise tolerance. Our patient was previously well, and we did not elicit any problems with exercise tolerance until she was on ICU.

Chronic hypoxia results in a compensatory polycythaemia,¹⁷ which was an early feature in our case, and was not considered abnormal. Clinical examination is very important. Clubbing and cyanosis can occur in the toes only and not the fingers, as the duct is distal to the subclavian arteries.⁶ Our patient's toes were clubbed, and had we noticed this earlier, we may have considered cardiac disease as a differential diagnosis prior to delivery.

Why did we not know earlier?

The normal drop in systemic vascular resistance (SVR) during pregnancy caused by progesterone increases the risk of right to left shunting and hypoxaemia.^{2 16} Together with the physiological anaemia of pregnancy, women with a shunt are therefore

more likely to decompensate.¹⁸ In our case, the patient was able to compensate for some of these changes in pregnancy, resulting in her presenting to the emergency department with hypertension, which was misinterpreted to be pre-eclampsia. Despite treatment with labetalol and magnesium, she did not decompensate preoperatively. It is likely that any drops in blood pressure were not significant enough to cause a reversal in blood flow through her shunt. Following delivery, there is a further fall in SVR, which can exaggerate the right to left shunt¹⁶ and is likely to be the reason that the patient decompensated after her caesarean section.

A patient's history and clinical examination are vital, and investigations are performed to help confirm or exclude diagnoses. Perhaps the initial echocardiogram report showing normal pulmonary artery pressures was misleading. This reiterates the importance of questioning the diagnosis and seeking senior multidisciplinary reviews. Therefore, respiratory and cardiology opinions were sought. The right ventricular strain pattern on ECG, together with right ventricular hypertrophy on echocardiogram, should have prompted earlier consideration of pulmonary hypertension as a diagnosis, even if the cause was unknown. Approximately 5–10% of patients with congenital heart disease will eventually develop pulmonary hypertension,¹⁷ and this carries a 30% risk of mortality in pregnancy.³

Congenital heart disease reduces the cardiac reserve of any patient, and heart failure may result due to the increased demand in pregnancy.¹⁰ This may explain this patient's presentation of acute breathlessness with oedema and worsening type 1 respiratory failure. Features of cardiac failure were evident on the CT report postoperatively, with evidence of right heart failure, ascites and hepatomegaly.

What would we have done differently?

CMACE's last report showed that, of the 50 women who died due to cardiac disease, 4 died due to congenital heart disease and their care was identified as being suboptimal.¹ The focus should be on identifying women with pre-existing heart disease, modifying risk factors and increasing the frequency of antenatal visits.⁴ Recent migrants are shown by the UK obstetric surveillance system to have a higher risk of morbidity.¹⁹ Our patient spoke reasonably fluent English, but the above reports indicate that translation services should be offered if English is not a patient's first language.^{1 19}

Prenatally, counselling for women with cardiac disease should be offered. There should be consideration for the high fetomaternal mortality in Eisenmenger's syndrome, including the option of termination of pregnancy. A multidisciplinary review is important, and correction of the PDA is possible. Severe pulmonary hypertension, as demonstrated in our case, may not fully reverse after surgical correction.⁸ Our patient's right and left ventricles were identical in size, meaning that correcting the PDA was not an option in our case. Survival rates are reported to be good in women with heart-lung transplants, but long-term effects are unknown.³

Antenatally, drug therapies have been attempted, but with no overall increase in survival.³ Inhaled nitric oxide has been reported to be successful in the peripartum period, but has significant side effects, including bleeding.²⁰

The management of patients with Eisenmenger's syndrome and pulmonary hypertension should focus on avoiding the cardiovascular stress of labour, and caesarean section is advised.^{9 12} There is no significant difference in outcome based on anaesthetic technique.^{9 21–26} It is suggested that women are admitted

from the second trimester³ or early in the third trimester¹⁷ for bed rest and optimisation.

These considerations may have led to an earlier diagnosis. The overall outcome may have been the same, but an earlier diagnosis would have altered our management. A patient with congenital heart disease would have access to early and frequent multidisciplinary visits, including being seen by a cardiologist and delivery in a tertiary centre with facilities for combined cardiac and obstetric care.^{3 12 13}

Learning points

- ▶ Early diagnosis is key to the multidisciplinary management of heart disease in pregnancy in an appropriate centre.
- ▶ Polycythaemia, particularly in the context of pregnancy and Asian ethnicity, should alert clinicians to the possibility of chronic hypoxia as a potential cause.
- ▶ Thorough clinical history and examination is vital—noting differential clubbing on toes, and a loud second heart sound may have led to an earlier clinical diagnosis and appropriate care.
- ▶ Modified Obstetric Early Warning Score (MOEWS) should be used in all pregnant women.
- ▶ Investigations are an adjunct to decision-making, based on the rest of the clinical picture. It is important to question the results of investigations, such as the echocardiogram, if they do not match the rest of the clinical picture.

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