We report the case of an acute type A aortic dissection occurring in a 35-year-old parturient. The initial diagnosis was missed; a subsequent emergency Caesarean section 3 weeks after presentation was followed by the development of left ventricular failure and pulmonary oedema in the early postoperative period. Echocardiography confirmed the diagnosis of aortic dissection and the patient underwent a successful surgical repair.

Keywords: arteries, aortic dissection; monitoring, echocardiography; pregnancy

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Acute aortic dissection occurring during pregnancy is a rare but recognized phenomenon. The results can be devastating for both mother and fetus with a reported mortality of 1% per hour if untreated. Most reported cases are associated with connective tissue disease (e.g. Marfan’s syndrome), systemic hypertension and congenital heart disease, including coarctation and bicuspid aortic valve. We report the case of an acute type A aortic dissection occurring in a fit and well 35-year-old parturient with no discernible risk factors. We discuss the clinicopathological features of acute aortic dissection and also review the literature concerning the management of peripartum aortic dissection.

Case report

A previously fit and well 35-year-old primigravid women at 37 weeks gestation presented to the maternity unit of her local hospital with sudden onset of interscapular back pain. Cardiovascular and neurological examination were normal with arterial pressure 130/80 mm Hg. A diagnosis of musculoskeletal back pain was made and the patient was discharged with simple analgesia. At follow-up 1 week later, the back pain had migrated to the right scapular area and the patient was complaining of mild exertional dyspnoea. At follow-up 1 week later, the patient had complained to the right scapular area and the patient was complaining of mild exertional dyspnoea. Cardiorespiratory examination was unremarkable: heart rate 85 beats min⁻¹, normal heart sounds, arterial pressure 130/70 mm Hg and an oxygen saturation of 97% in air. Peripheral pulses were not examined. There was no calf pain or swelling. A 12-lead ECG showed sinus rhythm. No chest radiography was performed. The possibility of a pulmonary embolism was raised but was felt to be unlikely in view of the unremarkable examination findings. The patient was discharged. Follow-up 3 days later by the midwife reported an improvement in her symptoms.

The patient re-presented urgently a week later with sudden onset of numbness and weakness of her left arm and leg. This had been immediately preceded by the development of right-sided neck and jaw pain during sexual intercourse. On examination she was pale and aphyreal with heart rate 80 min⁻¹, arterial pressure 100/60 mm Hg and tenderness over her right carotid artery. Auscultation demonstrated an ejection systolic murmur, an early diastolic murmur and a right carotid bruit. Neurological examination revealed decreased power (3/5) and sensation of the left arm and leg. Cranial nerve examination was normal and there was no meningism. A consultant neurologist diagnosed a cerebrovascular accident, probably of thromboembolic aetiology. CT scan of the brain and carotid Doppler studies were normal. A transthoracic echocardiogram was scheduled for the following morning. In view of the diagnosis and the possible requirement for anticoagulant therapy a decision was made to expedite delivery by Caesarean section. This was performed under combined spinal–epidural anaesthesia. The operation was uneventful, with the delivery of a healthy infant. Ten hours after Caesarean section the patient developed acute pulmonary oedema, which responded to diuretic therapy. At the same time it was noted that she had diminished pulses in her right arm with differential right and left non-invasive arterial pressures of 70/40 mm Hg and 100/50 mm Hg respectively. A transthoracic echo showed an enlarged aortic root, aortic regurgitation, a proximal dissection flap and a volume-overloaded left
A left radial arterial line was inserted and an intravenous infusion of labetalol was commenced to maintain systolic blood pressure $< 100$ mm Hg. The patient was transferred to the regional cardiothoracic centre. Following premedication with ranitidine 50 mg i.v., anaesthesia was induced using a modified rapid sequence with midazolam 0.1 mg kg$^{-1}$, fentanyl 15 $\mu$g kg$^{-1}$, thiopental 0.7 mg kg$^{-1}$ and suxamethonium 1 mg kg$^{-1}$ in order to minimize the combined risks of regurgitation and pressor response to intubation. A transoesophageal echocardiogram confirmed the previously documented findings (Figs 2 and 3). The patient was placed on atriofemoral cardiopulmonary bypass and thiopental 2 g was administered for cerebral protection. At operation an intimal tear was found in the ascending aorta with a dissection extending distally into the brachiocephalic artery and proximally involving the aortic valve. The right coronary ostium was unidentifiable, with complete occlusion of the right coronary artery. The ascending aorta was replaced with a 26 mm Dacron graft and the aortic valve was replaced with a 25 mm mechanical St Jude valve. The left coronary ostia was reimplanted, and the right coronary artery was grafted using the saphenous vein. Total cardiopulmonary bypass and aortic cross-clamp times were 220 min and 78 min, respectively, with a circulatory arrest time of 29 min at 14°C. Postoperatively the patient required dobutamine 5 $\mu$g kg$^{-1}$ min$^{-1}$ to maintain a mean arterial pressure of 70–80 mm Hg and intravenous oxytocin for 24 h to minimize the risk of postpartum haemorrhage. She was extubated 12 h later with full resolution of her preoperative neurology. Examination of the patient failed to reveal any Marfinoid stigmata. Histology confirmed the dissection with no evidence of cystic medial necrosis. The aortic valve showed myxomatous degeneration with changes characteristic of ‘floppy valve’ syndrome.

The remainder of the patient’s postoperative recovery was unremarkable. She was discharged 10 days after her initial admission. Currently both mother and baby are doing well.

**Discussion**

Aortic dissection during pregnancy is potentially lethal to both mother and fetus. The management is complex and depends on the type of dissection and gestational age. The Stanford classification system divides aortic dissections into two types: type A, involving the ascending aorta regardless of the entry site location, and type B, involving the aorta distal to the origin of the left subclavian artery. Type B dissections occurring during pregnancy are very rare. The treatment is medical with strict control of blood pressure as previously reported. Type A dissections require emergency surgery. There are cases in the literature detailing surgical repair at all stages of pregnancy and the post-partum period. Following a 12-year review of acute aortic dissection complicating pregnancy Zeebregts and colleagues have suggested the following management guidelines. In a pregnant woman with an acute type A dissection, treatment should be aimed at saving two lives. Before 28 weeks gestation, aortic repair with the fetus kept in utero is recommended. If the fetus is truly viable (i.e. after
primary Caesarean section followed by aortic repair at the same operation is the treatment of choice.
Between 28 and 32 weeks gestation, there are a dilemma, with the delivery strategy determined by the fetal condition. The fetal and maternal mortalities for cardiovascular surgery during pregnancy are 20–30% and 2–6%, respectively.\(^1\)\(^{15}\)

Our patient’s symptoms and examination findings were suggestive of aortic dissection but the diagnosis was not made until 3 weeks after presentation. Despite this, her patient survived and made a full recovery. A number of studies have shown this to be unlikely. Mészáros and colleagues\(^16\) showed a pre-hospital mortality of 21%. The mortality rate for untreated proximal aortic dissections increases by 1 to 3% per hour after presentation and is approximately 25% during the first 24 h, 70% at 1 week and 80% at 2 weeks.\(^2\) In our patient the dissection propagated slowly, with extension into the brachiocephalic trunk and hemiparesis occurring 3 weeks after her initial presentation. Crucially, she was not given excessive vasopressor or fluids during her Caesarean section. This could have resulted in rapid progression of the dissection, acute valvular disruption and intra-operative cardiac decompensation, with potentially disastrous consequences for mother and fetus.

Although the clinical manifestations of acute aortic dissection are well described, the diagnosis is often overlooked. A study over a 27 year period showed that misdiagnosis occurred in 85% of patients presenting with acute dissection.\(^16\) This is supported by a number of case reports in which the diagnosis was initially missed in the peripartum period.\(^5\)–\(^8\) The most common presenting feature is sudden onset of severe back or chest pain (up to 96% of cases) that is characteristically stabbing, tearing or ripping. The pain is frequently migratory, generally following the path of propagation of the dissection.\(^17\) In our case the initial back pain moved to the scapula.

Signs on physical examination may reflect the location of the dissection and its extent. Organ ischaemia results from obstruction of branch arteries originating from the aorta. Vessel occlusion can be caused by extension of the dissection into the artery wall or direct compression by the expanding false lumen. Our patient’s left hemiparesis was caused by propagation of the dissection into the brachiocephalic artery, probably brought about by the cardiovascular response that accompanies sexual intercourse.\(^18\)

Importantly, our patient was also noted to have a diastolic murmur and aortic regurgitation. Acute aortic valve incompetence accompanies 18–50% of proximal aortic dissections and is the second most common cause of death (after aortic rupture) in dissections. Although echocardiography demonstrated good ventricular function in our case, regional wall motion abnormalities occur in 10–15% of patients with proximal dissection and are primarily caused by low coronary perfusion. The right coronary artery is involved more commonly than the left, and occasionally dissection and myocardial infarction can occur together.\(^19\) Thrombolysis in this setting can be fatal.

Although suggested by the clinical findings, the diagnosis of aortic dissection must be confirmed by investigation. Diagnostic modalities include chest radiography, echocardiography, contrast-enhanced computed tomography, aortography and magnetic resonance imaging. Although chest radiography lacks specificity it can be useful for the initial prediction of dissection when used in combination with the history and examination findings. The classic radiographic feature of mediastinal widening occurs in up to 50% of cases.\(^20\) Concern about fetal radiation exposure meant that our patient did not undergo chest radiography at first presentation. Transesophageal echocardiography diagnosed the dissection in our patient. This investigation is useful in the initial screening of patients with suspected aortic dissection. The sensitivity and specificity can be up to 75% and 90%, respectively. Transoesophageal echocardiography overcomes many of the limitations associated with transthoracic echocardiography and has sensitivity and specificity data as high as 99% and 98%, respectively.\(^1\) CT and aortography involves the risk of i.v. contrast administration and radiation.

The main predisposing factor for aortic dissection is degeneration of the collagen and elastin in the intima media. Systemic hypertension is the main risk factor. Other well-established risk factors include hereditary connective tissue disease (e.g. Marfan’s syndrome and Ehlers–Danlos syndrome), coarctation of the aorta, bicuspid aortic valve, aortitis and arch hypoplasia. On the basis that ~50% of aortic dissections in women under 40 years of age occur in pregnancy or the puerperium, it is frequently stated that pregnancy is an independent risk factor for aortic dissection.\(^21\)\(^22\) However, a recent review by Oskoui and Lindsay\(^23\) has questioned the causal relationship between pregnancy and aortic dissection. Hypertension and Marfan’s syndrome are the most commonly occurring risk factors in cases of aortic dissection during pregnancy. The most common site of pregnancy-associated dissection is the proximal aorta, and aortic rupture usually occurs during the third trimester or first stage of labour.\(^1\)

In our patient neither clinical nor histological examination showed any evidence of hereditary connective tissue disease. Syphilis serology was normal and there was no documented evidence of hypertension. The only abnormal finding was a floppy aortic valve with idiopathic myxoid degeneration. Floppy aortic valves occur less commonly than floppy mitral valves, but both can be associated with myxoid degeneration. This degeneration is defined as significant disruption of the valve fibrosa and its replacement by acid mucopolysaccharides together with cystic change. The underlying cause can be rheumatic, endocarditic or idiopathic, or connective tissue disorders. Idiopathic myxomatous degeneration is rare, with patients mainly in their fifties and sixties, although younger cases have been reported. There is a male preponderance. Most patients present with severe aortic regurgitation, with the diagnosis made after valve replacement.\(^24\)\(^25\) This is the first reported case of pregnancy and acute
aortic dissection occurring in association with this valvular pathology in a non-Marfanoid patient.

In conclusion, this case highlights some important learning points. Despite a clinical presentation suggestive of aortic dissection, our patient was initially diagnosed with a thromboembolic stroke. This may have been the result of a lack of awareness of peripartum aortic dissection or the unavailability of out-of-hours echocardiography. This culminated in a Caesarean section being performed in the presence of an acute type A dissection. As the consequences of missing an acute aortic dissection can be fatal, it is imperative that any patient presenting with a suggestive clinical history is thoroughly investigated. Echocardiography is an effective diagnostic tool with high specificity and sensitivity that avoids the added risk of fetal radiation exposure and is the investigation of choice in pregnancy. Furthermore, the American College of Cardiology and American Heart Association state that any patient under 45 years of age who is thought to have suffered a cerebrovascular event should be routinely investigated with echocardiography. This imperative that any patient presenting with a suggestive clinical history is thoroughly investigated. Echocardiography is an effective diagnostic tool with high specificity and sensitivity that avoids the added risk of fetal radiation exposure and is the investigation of choice in pregnancy. Furthermore, the American College of Cardiology and American Heart Association state that any patient under 45 years of age who is thought to have suffered a cerebrovascular event should be routinely investigated with echocardiography to exclude a cardiac cause.26 Once aortic dissection is diagnosed the definitive management is dependent on the gestation. This report is a reminder to anaesthetists that, although rare, acute aortic dissection can present during pregnancy.

References
17 Crawford ES. The diagnosis and management of aortic dissection. JAMA 1990; 264: 2537–41
21 Braunwald E. Heart Disease. Philadelphia: W.B. Saunders, 1992; 1536–57
23 Oskou R, Lindsay J Jr. Aortic dissection in women <40 years of age and the importance of pregnancy. Am J Cardiol 1994; 73: 821–3