

## Acute Aortic Dissection during Pregnancy

Walkiria Samuel Avila, Ricardo Dias, Renato Takeshi Yamada, Adriano Armelin

Heart Institute and University of São Paulo Medical School - FMUSP - São Paulo, SP - Brazil

Acute aortic dissection is one of the most dreaded clinical conditions during pregnancy. Difficulties in establishing a diagnosis and limitations regarding invasive studies increase mortality rates associated with the disease. The limited experience reported in the literature does not allow the determination of guidelines for clinical and/or surgical management of aortic dissection in pregnancy. The authors present a case of acute aortic dissection in a woman in her 33<sup>rd</sup> week of gestation and discuss the diagnostic approach considering the peculiarities of the disease's manifestation.

Acute aortic dissection is a rare but severe event during pregnancy and one of the most dreaded clinical conditions during this period. Difficulties in establishing a diagnosis and assessing of symptoms, that can be mistaken for those of a normal pregnancy, as well as limitations regarding invasive studies, due to potential risk to the conceptus, result in treatment delay and further increase mortality rates associated with the disease.

The heterogeneous presentation of cases and limited experience reported in the literature do not yet allow the determination of guidelines for clinical and surgical management of aortic dissection in pregnancy. However, treatment must always offer chances of survival for both the mother and fetus, taking into account the mother's clinical status, gestational age, and the opportunity for surgical intervention, if indicated.

The authors present a case of acute aortic dissection in a woman in her 33<sup>rd</sup> week of gestation and discuss the diagnostic approach considering the peculiarities of the disease's manifestation.

### Case Report

A 35-year-old physician, nullipara, primigravida, asymptomatic, in her 33<sup>rd</sup> week of gestation and with no personal history of heart disease or hypertension experienced severe retrosternal pain unrelated to exertion and accompanied by a drop in blood pressure (80/40mmHg), followed by gradual and spontaneous improvement. The patient was seen in the emergency room and treated with antispasmodic medication. Over the following 48 hours she presented respiratory discomfort, malaise, and lower extremity edema. At that time, the patient was advised to

have an echocardiogram, which showed a DeBakey type-I aortic dissection, mild aortic regurgitation and significant left ventricular diastolic dysfunction (Fig. 1), and was immediately referred to the Heart Institute.

The patient reported that routine prenatal tests performed during the first weeks of pregnancy were normal. In the 29<sup>th</sup> week, she presented brief palpitation, which prompted conventional electrocardiogram and transthoracic echocardiogram, both of which showed no abnormality. In the 32<sup>nd</sup> week, a urinalysis revealed leukocyturia (71,000/mL).

At admission, the patient was conscious, dyspneic +++++, acyanotic, afebrile, and pale +++++. Her blood pressure (BP) was 130/60 mmHg and her heart rate (HR) was 90 beats per minute (bpm); peripheral pulses were palpable and symmetrical, and pulse pressure was high. Pulmonary auscultation revealed normal vesicular breath sounds bilaterally, with no adventitious sounds. Cardiac examination showed regular rate and rhythm, without murmurs. She had lower extremity edema +++++, but there was no hepatomegaly. Her abdomen showed a pregnant uterus 30 weeks in size.

Computed tomography at admission showed increased diameters of the left heart cavities and confirmed the diagnosis of type-I aortic dissection, involving the brachiocephalic trunk and the ascending and descending aorta (Fig. 2). Obstetric and transabdominal ultrasonographic examinations demonstrated a single viable fetus showing vitality, gestational age of 33 weeks, and estimated weight of 2,300g.

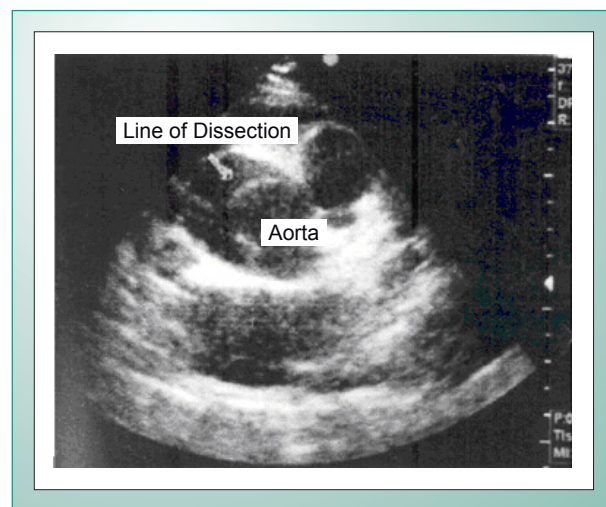


Fig. 1 - Two-dimensional echocardiography with color flow mapping of the aorta showing a type-1 aortic dissection, carried out by Dr. Luciana Braz Peixoto on 10/June/2004. Courtesy of the Echocardiography Department - Delboni Auriemo Diagnosis Medicine - America Diagnosis.

### Key words

Aortic dissection, pregnancy, cardiac surgery.

## Case Report

Based on the progressive risk of aortic rupture and taking fetal viability into account, the medical team indicated immediate corrective surgery for the aortic dissection and termination of pregnancy with cesarean section prior to thoracotomy.

The patient participated in and agreed with the decision. She was conscious when she entered the operating room, with stable vital signs, as shown by invasive monitoring. During anesthetic induction for cesarean section, a sudden hypotension occurred, followed by asystolic cardiac arrest. Under external cardiac massage, the baby was immediately delivered by a cesarean section through median laparotomy. As the mother's heart rate was not restored, an exploratory sternotomy was performed, revealing significant hemopericardium. After pericardial drainage, the heart rate was restored and the patient stabilized hemodynamically. Cardiopulmonary bypass was established by antegrade perfusion through the brachiocephalic trunk and venous drainage by a single cannula advanced to the right atrium; myocardial protection was carried out with cold blood cardioplegic solution and hypothermia at 20°C. Aortic dissection repair was carried out by inserting a Dacron tube into the ascending aorta (fig. 3) and right subclavian artery, after it was detached from the brachiocephalic trunk during arterial line placement.

Pathological studies showed partial delamination of the aorta at the media layer and hemorrhage within the adventitia, with moderate accumulation of mucoid substance in de media and intima.

After surgery, the patient was maintained on assisted ventilation for 48 hours and received routine postoperative drugs and the usual doses of oxytocic agent. She remained hemodynamically stable, presenting no bleeding, with normal uterine contractility and lochia flow. During the first seven postoperative days she developed lower-extremity edema, significant arterial hypertension (mean BP 180/120 mmHg), and sinus tachycardia (mean HR 110 bpm), which were gradually controlled with atenolol 200 mg/day, captopril 150 mg/day, amlodipine 20mg/day, and furosemide 80mg/day. Low-molecular-weight heparin was also administered in

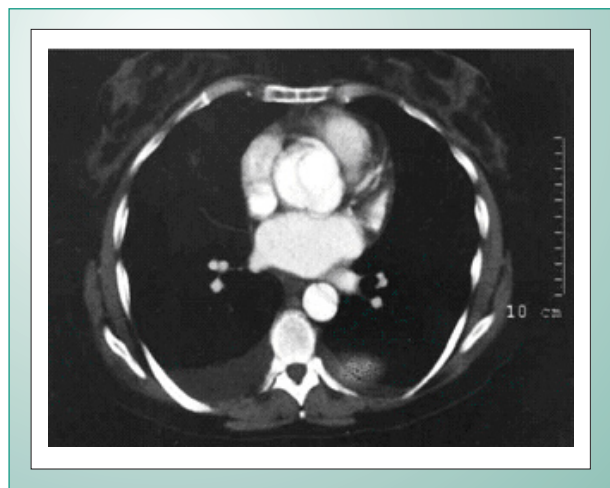
prophylactic doses for six weeks. In the days that followed, drug dosages were adjusted to maintain BP around 100/60 mmHg and HR at approximately 70 bpm. The patient was discharged on the 15<sup>th</sup> postoperative day. Atenolol and amlodipine dosages were maintained; captopril dosage was reduced (75mg/day). Due to the lack of aseptic and antiseptic techniques for emergency laparoscopy and thoracotomy, antibiotic prophylaxis was instituted with vancomycin 500 mg/day and ceftriaxone 1g/day for 21 consecutive days.

The baby boy was born the 33<sup>rd</sup> gestational week under general anesthesia and during maternal cardiac resuscitation maneuvers. He presented bradycardia, cyanosis, and pallor, with Apgar score 1 at one minute, 5 at five minutes, and 7 at ten minutes.

The infant was placed on mechanical ventilation with 100% oxygen in an anesthetic bag during the first minute, and his heart rate, skin color, and spontaneous breathing rapidly improved; however, he was hypotonic and had no reflex irritability. He progressed to respiratory failure, requiring intermittent mechanical ventilation and endotracheal surfactant administration for two hours, which improved ventilation parameters. The baby was extubated while still in his first day of life and was kept on oxygen nasal catheter with continuous positive airway pressure (CPAP).

On his second day of life, the baby was diagnosed with right pneumothorax and reduced oxygen saturation. He was then intubated again and maintained on mechanical ventilation and vasoactive agents. After two chest drainages and high-frequency ventilation, both his clinical condition and ventilation parameters improved, which allowed him to be extubated on his 9<sup>th</sup> day of life, although he was still kept in the incubator with oxygen catheter. As his respiratory condition enhanced, the administration of oxygen and vasoactive drugs was progressively reduced. Chest tubes were removed on the 12<sup>th</sup> day, when enteral feeding was initiated through orogastric tube.

Due to his difficulties to coordinate sucking and swallowing, non-nutritive stimulation was required, with gradual transition to oral feeding. Despite normal total blood count and



**Fig. 2** - Preoperative CT depicting the type-1 dissection. B) Postoperative CT depicting the ascending aorta repair with Dacron.



**Fig. 3** - Postoperative CT depicting the ascending aorta repair with Dacron.

negative blood culture, the neonate was kept on antibiotics for 10 days, because the radiographic examination showed an area of opacity at the right lung base, C-reactive protein levels were high for his age (6.8), and there was a history of maternal urinary infection at the time of delivery. Neurological examination was unremarkable, as were the Doppler echocardiogram and brain ultrasound; brain tomography showed mild attenuation of white matter in both cerebral hemispheres. Responsive to sucking stimuli and being fed all milk volume orally, the baby was discharged at 34 days of life in good condition.

## Discussion

Aortic dissection in young women is an uncommon disease, sometimes of unknown etiology, potentially catastrophic and variable in extension, beginning with a rupture of the aortic intima.

In young patients, the most common clinical conditions predisposing to aortic dissection are collagen diseases (Marfan, Ehlers-Danlos) or those that primarily affect the vessel, such as aortic coarctation and bicuspid aortic valve<sup>1</sup>. However, in healthy women, a strong correlation between aortic dissection and pregnancy has been reported, based on the high incidence of this event during pregnancy<sup>2,3</sup>. Indeed, in the Schnitker and Bayer review 24 out of 49 aortic dissections in women younger than 40 were associated with pregnancy, and 20 of them occurred before labor<sup>4</sup>. The cause of this association (pregnancy/aortic dissection) in women considered healthy at admission is still unclear.

At first, it was suggested that physiological increments in blood volume and cardiac output might be potential causes for the event. However, this hypothesis does not explain the lower incidence of dissection during labor<sup>5</sup>, when most circulatory impact occurs – as compared to other phases of the pregnancy-puerperal cycle – due to major hemodynamic oscillation.

It has also been argued that vascular structural changes observed during pregnancy – and which are similar to cystic medial necrosis and responsible for increased aortic diameter and compliance (more marked at pregnancy term) – may play a significant role in dissection<sup>5</sup>. Mucoïd degeneration of the media and intima layers described in the pathological examination of the aorta of the case under discussion, coincides with literature reports on dissections that occur during the third trimester of pregnancy. In this framework, a recent genetic study is worth noting. The study demonstrated variable expression of matrix metalloproteinase genes of the aorta that greatly influence physiological vascular remodeling during pregnancy<sup>6</sup>.

The exact influence of hormones on vascular connective tissue remains unknown. Animal studies, however, have demonstrated that estrogens inhibit collagen and elastin deposition in the aorta, whereas progesterone accelerates – although minimally – the deposition of noncollagenous proteins in the vessel walls<sup>7</sup>.

Clinical manifestations of aortic dissection – classically expressed through severe retrosternal and/or abdominal pain – were, in this case, underestimated and undifferentiated from the chest pain resulting from mechanical effects of the

pregnant uterus and constipation, which are common in normal pregnancies. Regarding differential diagnosis, amniotic fluid embolism, premature placental detachment, uterine rupture, and spontaneous pneumothorax are to be considered as frequent causes of chest pain during pregnancy.

Despite posing limitations for the diagnosis of aortic dissection, the echocardiography proved to be useful in early investigation, since it is a non-invasive and easy-to-perform examination. Computed tomography, combining the advantage of rapid evaluation and relatively low cost, confirmed the diagnosis and, especially, dissection classification – key to therapeutic approach definition.

The localization of aortic dissection in the ascending aorta and extending into the descending aorta has characterized the present case being as DeBakey type I, which prompted immediate surgical correction, since medical treatment for that kind of dissection is associated with increasing mortality rates, reaching 80% in the first week after the diagnosis<sup>8</sup>.

In recent years, three relevant data for treating this severe disease successfully have been identified: the early referral of these patients for surgery, surgeon's greater experience, and postoperative care. Nonetheless, corrective surgery for aortic dissection is still associated with severe cardiac complications and operative mortality rates between 9% and 36%<sup>9</sup>. As with the risk posed to the mother, heart surgery causes fetal morbidity and mortality risks of approximately 30% and 9%, respectively – which may vary according to the type of procedure, time of cardiopulmonary bypass, hypothermia, nonpulsatile flow, and previous maternal clinical conditions<sup>10</sup>.

In the present case, the priority indication for pregnancy termination prior to the aortic dissection repair aimed at sparing the lives of both the mother and conceptus, since the latter, already viable, would have better chance of survival than if it had been maintained inside the uterus during this highly complex surgery. While considering such approach, the high risk of acute fetal distress was taken into account, as it would require emergency cesarean section during intra- or postoperative period, thus seriously worsening the patient's prognosis.

Our approach was consistent with data described in the literature, in which the best survival rates (mother's and fetus's) are based on gestational age. Therefore, if the event occurs up to 28 weeks of pregnancy, surgical aortic dissection repair should be carried out during intrauterine life; after 32 weeks, when the fetus is viable, surgical repair should be performed after the cesarean section. A more difficult decision to be made is when aortic dissection occurs between 28 and 32 weeks of gestation. During this period, delivery must be decided based on the conceptus' conditions, and cesarean section should be indicated in case of fetal distress, or pregnancy may be continued if the fetus can tolerate maternal surgery<sup>11</sup>.

Cardiac tamponade and subsequent cardiac arrest are strong warnings for the risk and severity of delaying surgical procedure. In the present case, laparotomy prior to thoracotomy saved the conceptus and freed the surgeon to repair the aorta without the additional concern with the intrauterine fetus. The successful outcome was undoubtedly due to the indication of surgical intervention in the aorta immediately after diagnosis was confirmed.

## Case Report

The clinical conditions shown by the neonate presuppose acute intrauterine distress. The satisfactory response to early resuscitation maneuvers allows us to presuppose that this asphyxia did not last long; however, it was long enough to compromise the central nervous system, although temporarily, as shown by the length of time taken for the tonus and reflex irritability recovery. Although ultrasound examination was normal, in prematures with significant perinatal asphyxia intracranial hemorrhage<sup>12, 13</sup> is expected to occur.

At a gestational age of 33 weeks, surfactant production is not yet enough to keep appropriate fetal alveolar distension. The situation is aggravated by asphyxia, which favors higher inactivation and increased surfactant consumption. For that reason, its use has been indicated to improve respiratory conditions, thus favoring earlier weaning from mechanical ventilation<sup>13</sup>.

Pulmonary traumas resulting from pressure and volume variations are more closely associated with the use of surfactant during mechanical ventilation, while pneumothorax is less common in patients under positive alveolar pressure. Despite

that, complete lung re-expansion was obtained through bilateral drainage, and no pulmonary sequelae were reported.

The suck and swallow deficit after gestational week 34 may signal some degree of impairment of the central nervous system<sup>14</sup>. Nevertheless, the child responded well to sucking stimulus, and the neurological assessment was considered normal for his age, resulting in good prognosis.

After hospital discharge, counseling on family planning is mandatory due to the high risks of a new pregnancy in patients with aortic diseases. Vascular changes fail to normalize after pregnancy, resulting in predisposition to new cardiovascular events. Therefore, it is a consensus that aortic diseases are a contra-indication to pregnancy<sup>15</sup>, and tubal ligation for sterilization is the most effective contraceptive method for such cases.

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