Two Lives on the Line: A Case Study in Obstetric Critical Care

Laura Lee Naylor Velez, RN, MS, CNS, CCRN
Kyle Toal, MD
Susan A. Goodwin, RN, MS, CNS, CPAN

The physiological changes of pregnancy can greatly exaggerate preexisting health conditions in the mother. One such condition is Marfan syndrome, a genetically inherited disorder characterized by weakened connective tissue. Persons with Marfan syndrome are very tall with disproportionately long extremities and are characterized by arachnodactyly and lax joints predisposed to dislocation. Marfan syndrome can lead to dislocation of the ocular lens, as well as weakness of the aortic root and aortic wall. The danger of aortic dissection in women with Marfan syndrome can be compounded by the hemodynamic changes of pregnancy. In pregnant women with Marfan syndrome, the highest risk for aortic dissection is from the beginning of the third trimester until shortly after delivery. Aortic dissection during pregnancy is an emergency that endangers the lives of both mother and fetus. We describe the successful medical and surgical management of a pregnant woman with Marfan syndrome. The complicated psychosocial issues associated with high-risk pregnancy and delivery and the need for accurate interdisciplinary communication are discussed.

NORMAL PREGNANCY

Pregnancy induces significant changes in the mother to promote uterine blood flow and to support the developing fetus. The changes include increases in blood volume, heart rate, stroke volume, and cardiac output. Pregnancy also produces decreases in systemic vascular resistance, pulmonary vascular resistance, and the colloid osmotic pressure of the vasculature. These changes begin as early as the seventh week of pregnancy and peak late in the second trimester (Table 1).

In pregnancy, the vasculature changes in ways that are poorly understood. These changes include hormone-induced changes, decreased amounts of mucopolysaccharides on the arterial wall, increased mass of the smooth muscle fibers, and changes in the protein content of the vessel walls. The normal hemodynamic...
ACUT changes of pregnancy and maternal hypertension are also thought to contribute to the weakening of the vasculature. Pressure changes and increased flow may produce structural changes in the arteries, which may directly lead to aneurysm and dissection. Hemostatic changes occur in the last trimester of pregnancy to protect the mother from hemorrhage as the placenta separates from the uterus at delivery. Levels of coagulation factors VIII, X, and IX increase at the same time that fibrinolytic activity decreases, leading to an overall hypercoagulopathy.

### ACUTE AORTIC DISSECTION

Acute aortic dissection is a sudden catastrophic event in which a tear in the intimal wall of the aorta allows blood to escape from the true lumen of the vessel, rapidly separating the inner layer from the outer layer of the tunica media. Patients with Marfan syndrome have an abnormal number of microfibers in the tissue of the aorta, which leads to progressive weakness of the tunica media. The parietal pericardium is attached to the ascending aorta just proximal to the origin of the innominate artery. Rupture of any part of the ascending aorta leads to extravasation into the pericardial sac. Rapid death results from the subsequent hemopericardium.

Dissections of the transverse arch of the aorta are more complex because the brachiocephalic, left common carotid, and left subclavian arteries are compromised.

The prevalence of dissecting aortic aneurysm is 1 in 10,000 general hospital admissions and 1 in 363 autopsies, suggesting that dissecting aortic aneurysm may be a cause of sudden death in the general population. Among patients with untreated acute dissection, the mortality rate is 20% at 15 minutes after dissection, 50% to 70% at 2 days, and 90% at 1 week. Aortic dissection occurs in 33% to 44% of patients with Marfan syndrome. The estimated mortality associated with pregnancy complicated by Marfan syndrome with aortic involvement is 50%.

The need for prenatal monitoring both by an obstetrician and a cardiologist is evident. The Medical Genetics Clinic of Johns Hopkins Hospital recommends an echocardiogram every 6 to 8 weeks to assess for increased dilatation of the aortic root during pregnancy for mothers with Marfan syndrome. If the diameter of the aortic root is greater than 4.0 cm, the risk for aortic dissection becomes significant.

### CLINICAL MANIFESTATIONS

A dissecting aortic aneurysm may mimic some of the signs and symptoms of early labor (Table 2). Pregnant women often experience epigastric discomfort that they interpret as burning in the chest. Although it is not a symptom of early labor, burning in the chest can be an early symptom of aortic dissection. Aortic dissection is rare in pregnancy and may be initially overlooked because its clinical findings are similar to those for early labor (Table 2). Blood pressures that differ from one arm to the other or radial pulses that differ in intensity from one arm to the other, the new onset of a diastolic aortic murmur, and increased severity of chest pain are important characteristics that can be used to distinguish early aortic dissection from early labor.

### DIAGNOSIS AND TREATMENT

Acute aortic dissection may be apparent on a chest radiograph as a widened mediastinum, particularly in the upper part of the mediastinum and toward the left side of the thorax. Cardiomegaly and pericardial effusions are also common radiographic findings in patients with ascending aortic dissection. An echocardiogram is obtained primarily to evaluate left ventricular function, aortic valve competence, and size of the aortic root. However, neither a chest radiograph nor an echocardiogram is sufficient for a definitive diagnosis of aortic dissection to be made. Computed tomography is the emergency diagnostic procedure of choice for aortic dissection. Having the patient lie flat, as required for thoracic computed tomography, could lead to compression of the descending aorta by the fetus in a pregnant woman. From descriptions in the literature, it is not clear how this position and the compression of the descending aorta might affect the dissection process in the ascending aorta. After a definitive diagnosis is made, the treatment of choice is surgical repair of the
Table 2 Comparisons of signs and symptoms of aortic dissection and early labor

<table>
<thead>
<tr>
<th>Signs and symptoms common to aortic dissection and early labor</th>
<th>Signs and symptoms distinct to aortic dissection</th>
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</thead>
<tbody>
<tr>
<td>Nausea, Mild hypertension, Restlessness, Anxiety, Epigastric discomfort, Diaphoresis</td>
<td>Blood pressure differs from one arm to the other, Intensity of radial pulse differs from one arm to the other, New onset of diastolic aortic murmur, Burning sensation in the chest.</td>
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Aorta and other structural damage. Marfan syndrome presents an additional surgical challenge because of the fragile nature of the surrounding tissue.

CASE STUDY
Preoperative Presentation
Mrs A. was 34 years old, gravida 2, para 1, at a gestation of 39 weeks, with previously diagnosed Marfan syndrome. She was brought to the obstetric unit by ambulance at 9:50 pm because of chest pain, “heartburn” all the way through to her back, and achy arms. By auscultation, the fetal heart rate was 120/min to 130/min; normal fetal heart rate is 120/min to 160/min. Mrs A. also had lower back pain and was having mild uterine contractions of 60 to 70 seconds’ duration. She had shortness of breath when lying supine and appeared increasingly anxious. She was transferred to the emergency department for further evaluation.

Mrs A. was admitted to the emergency department at 10:41 pm, where a chest radiograph and blood work were completed. The fetal heart rate was unchanged; the mother’s blood pressure was 110/70 mm Hg. Permission were obtained for blood transfusion and cesarean delivery. Mrs A.’s shortness of breath was worsening, as was the severity of her chest pain. Mrs A. had a grade III/VI diastolic murmur heard best at the second intracostal space on the right sternal border, a new clinical finding. Findings on the chest radiograph were suggestive of a dissecting aortic aneurysm, so a cardiologist was consulted. The cardiologist on call, unfamiliar with this patient, performed a rapid assessment, then ordered a computed tomography scan to confirm the diagnosis of dissecting aortic aneurysm.

An esmolol infusion was started to diminish shear forces within the aorta. Esmolol, a short-acting β-blocker, has a negative inotropic effect on the heart, thus reducing the left ventricular ejection fraction and decreasing the mean arterial pressure. Its rapid onset and short half-life allow nimble clinical maneuvering for control of blood pressure. An initial bolus of 500 μg/kg was administered, followed by a maintenance infusion of 50 μg/kg per minute. The decision was to do a cesarean delivery and then emergently repair the aortic dissection.

After her clinical condition had stabilized, Mrs A. was transferred from the emergency department to the operating room with the cardiovascular surgeon in attendance. With epidural anesthesia, a cesarean delivery was performed via a transverse uterine incision; at 5:01 AM, a baby boy weighing 3.1 kg (6 lb 13 oz) was delivered. As the husband and new father attended, the infant’s Apgar scores were 8 and 9. The infant was given to the parents to hold and nurture while the surgical team prepared for thoracic surgery. Hemostasis was achieved after Mrs A. had an estimated blood loss of 500 mL.

Surgical Care
According to the preoperative transesophageal echocardiogram, Mrs A. had a dilated aortic root with a Stanford type A dissection flap that involved the ascending aorta, the aortic arch, and the descending aorta (Figure 1). Left ventricular function appeared normal, with mild aortic valve insufficiency. Fourteen minutes after her son was delivered, Mrs A. was given a general anesthetic, and cardiopulmonary bypass was started. An extreme aortic dissection that severed the right coronary artery with a circumferential tear was found. By means of a modified Bentall procedure, the ascending aorta and the aortic valve conduit were replaced with a Saint Jude Medical aortic valve graft. The native coronary arteries were then reimplanted (Figures 2 and 3). Cerebral circulation was cross-clamped for 12 minutes for reimplantation of the brachiocephalic and the left common subclavian arteries. The aortic repair was complicated by the extremely fragile state of the surrounding tissue, typical of Marfan syndrome (Figure 4).

After cardiopulmonary bypass for 170 minutes, including a cross-clamp time of 123 minutes, the bypass was successfully discontinued. The estimated blood loss during surgery was 600 mL. Twelve units of pooled platelets, 10 units of cryoprecipitate, and a total of 20 g ofaminocaproic acid
were given intraoperatively to control bleeding. The hospital chaplain provided support to Mr A during Mrs A’s heart surgery. The cardiovascular surgeon called the husband in the waiting room with updates on Mrs A’s condition twice during the surgery.

Postanesthesia Care Unit
Mrs A was admitted to the postanesthesia care unit at 11:25 AM. The recovery team consisted of a critical care nurse and a postanesthesia care unit nurse. Mrs A was intubated and on mechanical ventilation. A mediastinal chest tube drained 45 mL of sanguineous fluid. A pulmonary artery catheter in the right internal jugular vein and a right radial arterial catheter were in place, with waveforms indicative of adequate hemodynamic pressures. Continuous monitoring of arterial blood pressure was required to maintain a mean arterial pressure less than 100 mm Hg because of the fragile state of the aortic tissue receiving the graft. Intravenous nitroglycerine and dopamine were titrated to maintain the mean arterial pressure in the prescribed parameters. The pulmonary artery catheter was used to measure the systemic vascular resistance and allow the

Figure 1 Aortic dissection.

Figure 2 Reimplantation of native coronary artery into aortic graft.
normal sinus rhythm with no changes in the ST segment.

Activity, respiratory effort, circulation, consciousness, and color were each evaluated every 15 minutes by using the Aldrete scale (scale, 0-10); her score at admission was 2. Initial pulmonary artery pressure was 32/19 mm Hg with a pulmonary capillary wedge pressure of 14 mm Hg, indicating normal function of the left ventricle. Initial results of arterial blood gas analysis were pH 7.316, Pco₂ 38.3 mm Hg, Po₂ 388 mm Hg, oxygen saturation 98.6%, hemoglobin level 79 g/L, hematocrit 0.23, and platelet count 198,000 x 10⁹/L. Other laboratory values were potassium level 4.4 mmol/L, serum urea nitrogen level 3.2 mmol/L (9 mg/dL), creatinine level 80 µmol/L (0.9 mg/dL), and magnesium level 0.49 mmol/L (1.2 mg/dL). When the emergent circumstances of the surgery were taken into account, the slight metabolic acidosis was minimal; the hyperoxygenation was a normal finding after cardiopulmonary bypass and its accompanying hypothermia. Hypomagnesemia is a common finding after cardiopulmonary bypass. Mrs. A received an additional 2 units of packed red blood cells, 12 units of pooled platelets, and 10 units of cryoprecipitate; 2 g of magnesium sulfate corrected the hypomagnesemia.

As Mrs. A began to awaken from anesthesia, the recovery team repeatedly reinforced that she had delivered a healthy baby boy. Oxygen diffusion was monitored by frequent assessment of breath sounds for pulmonary congestion. Pulmonary artery pressures were monitored during weaning from mechanical ventilation. Rapid weaning can cause an increase in the pulmonary artery pressure because of local vasoconstriction of the pulmonary vasculature. Three and one-half hours after admission to the post-anesthesia care unit, Mrs. A was weaned from mechanical ventilation, and she was extubated at 3 P.M. After extubation, she was transferred to the critical care unit.

Critical Care Unit

The critical care nurse consulted with the women’s center nursing staff about the physical assessment and plan of care after cesarean delivery. Fundal height and the quality and quantity of vaginal and incisional bleeding were assessed. The nursery staff transported the infant to the critical care unit on several occasions to promote bonding between mother and child. Although the critical care staff offered to notify the hospital’s lactation consultant to aid in breast feeding, Mrs. A decided to feed her child formula because of her sore sternal incision.

Mrs. A. dangled her legs on the side of the bed 4 hours postoperatively and was assisted to a chair 2 hours later. Because of the fragile nature of the aortic tissue, all activity was accomplished cautiously to prevent a sudden increase in intrathoracic pressure. Infusions of vasoactive agents were decreased in dosage and then discontinued the first evening. Invasive catheters were removed the morning of the first postoperative day, and a saline lock was established on the peripheral intravenous catheter.

Progressive Care Unit

Mrs. A. was transferred from the critical care unit to the progressive care unit the morning of the second postoperative day. The surgeon removed the chest tube 48 hours after the surgery. Mrs. A.
and her baby were discharged to home after a 5-day stay.

NURSING CONSIDERATIONS

Aortic dissection, surgical repair, and the effects of cardiopulmonary bypass stress every organ system. Complications include dysrhythmias, hemorrhage, and renal, cardiac, pulmonary, and neurological dysfunction. The primary considerations of the recovery team are to maintain adequate oxygen-carrying capacity, oxygen transport, and oxygen diffusion. Hemoglobin levels are affected by dilution of plasma during cardiopulmonary bypass and by blood loss during and after surgery. The increase in clotting factors that occurs late in pregnancy must also be considered. Close monitoring of the fluid balance and laboratory results is required. Factors affecting cardiac output during recovery from any cardiac surgery are vasoconstriction from hypothermia and then vasodilatation as the patient is warmed. Fluid volume, vasoactive medications, left ventricular function, heart rate, and pain also affect postoperative cardiac output.

Before Mrs A. was admitted to the postanesthesia care unit, the critical care nurse reviewed similar case studies obtained through a literature search. Thus, the nurse was aware of the special considerations of postpartum fluid shifts and the delicate tissue state associated with Marfan syndrome. The recovery team reviewed a textbook on physiological changes in pregnancy. Doing so enabled the team to have a clear understanding of the pathophysiological conditions unique to this patient.

DISCUSSION AND RECOMMENDATIONS

Life-threatening aortic dissection and the psychosocial factors associated with a high-risk pregnancy combined to create a complicated clinical picture. Mrs A. and her husband made the decision to risk this pregnancy because of the unexpected and tragic death of their first child. Their first child, a son, was delivered at full term without complications. He died of acute lymphocytic leukemia when he was approximately 2 years old. Mr and Mrs A. were aware of the 50% chance that their children would inherit the gene for Marfan syndrome.

The second pregnancy was inherently more dangerous to the mother and fetus because of the progressive deterioration of the connective tissue associated with Marfan syndrome. During her pregnancy, Mrs A. was followed up closely by a cardiologist and had an echocardiogram every 4 to 6 weeks. She missed her eighth-month echocardiogram and did not reschedule the procedure. A progression in the dilatation of her aortic root might have been noted at that time. In one case study, however, findings on serial echocardiograms were not predictive of aortic dissection in a pregnant woman with Marfan syndrome.

Communication between the critical care staff, the cardiologist, and the cardiac surgical team was excellent. Few obstetric patients are treated in the combined medical-surgical critical care unit in this community hospital. Thus, communication between the critical care nurses and the maternal/child health nurses was essential to ensure that adequate postpartum care was provided.

The critical care nurses addressed the psychosocial issue of mother-infant bonding. Detailed instructions for care after the cardiac surgery were completed.

Mr A. was given support during the early hospitalization of his wife and was encouraged to attend the delivery of his son. Because of the rapid deterioration of his wife’s condition, he was allowed to see his wife and child for only a few minutes after delivery. A significant stressor for many men is fear of injury or death of their wife or partner. Even when a man values the pregnancy and desires the child, his first concern is for the well-being of his partner. Mr A. chose not to see the infant after delivery until he was certain his wife had survived the heart surgery. Postoperatively, the critical care unit encouraged open visitation; Mr A. then spent time with both his wife and their infant.

FUTURE IMPLICATIONS

The loss of the first child and the replacement of that child with the new infant were not addressed. Discharge teaching on maternal/child health issues was not completed because the maternal child nurses were not notified of Mrs A.’s impending discharge. Mrs A. would have benefited from a comprehensive plan of care that combined both maternal/child health and critical care issues. As a result of this case, the plan of care has been coordinated with discharge instructions for critically ill mothers.

When a patient has complex diagnoses that cross specialty boundaries, communication between disciplines becomes essential. This type of communi-
ducation could be facilitated by referral to a clinical nurse specialist. The clinical nurse specialist can monitor the patient closely through the entire pregnancy and delivery and coordinate an interdisciplinary plan of care.

**CONCLUSION**

In this era of specialized medicine, patients with complex conditions may not fit into obstetrical, critical care, pediatric, or medical/surgical categories. Any pregnant woman with Marfan syndrome should be considered at high risk. A successful outcome hinges on rapid diagnosis, excellent surgical intervention, and communication among all members of the healthcare team. The active involvement of a clinical nurse specialist could improve the outcome of such a complicated pregnancy.

**References**
