SEVERE PULMONARY HYPERTENSION IN PREGNANCY FOLLOWING SUCCESSFUL REPAIR OF VENTRICULAR SEPTAL DEFECT IN CHILDHOOD

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***Background*: Because of advances in surgical repair, an increasing number of women born with structural cardiac disease now live to reproductive age. Patients treated successfully in childhood are followed for varying periods of time, then may be lost to follow-up or told that no follow-up is necessary because their condition is stable. However, the hemodynamic changes that accompany pregnancy may result in cardiovascular decompensation, even after years of apparently good health.**

***Cases*: We have recently cared for two women who had undergone repair of congenital heart disease in childhood. Although they thought that their repair was complete and they had been asymptomatic until the pregnancy, both presented with symptoms and signs of severe pulmonary hypertension, subsequently confirmed on cardiac catheterization. One patient elected to terminate her pregnancy, and the other died in the immediate puerperium.**

Conclusion*: Despite normal physical function and an absence of abnormal physical findings, a thorough cardiac evaluation including echocardiography should be considered for pregnant patients with a history of repaired congenital heart disease, especially if the original defect is known to lead to pulmonary hypertension. *(Obstet Gynecol 1993; 82:680–2)

Patients with repaired congenital cardiac lesions may have significant residual cardiac disease even when asymptomatic in adulthood. During pregnancy, there are increases in intravascular volume and cardiac output, a decrease in systemic vascular resistance, major fluctuations in cardiac output during labor and delivery, and significant shifts in intravascular volume immediately after delivery. Although functionally asymptomatic before pregnancy, the stresses of pregnancy can exceed the heart's ability to compensate, leading to a rapid and profound deterioration in maternal condition.

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We have recently cared for two such patients with critical cardiac disease in pregnancy despite an apparently complete repair of congenital lesions in childhood and an absence of physical restrictions in adulthood. These cases highlight the need for a careful cardiac evaluation of all pregnant patients with prior cardiac disease, regardless of their pre-conception physical status.

Case Reports

Case 1

A 32-year-old woman, gravida 2, para 1–0–1–1, was transferred to the University of Utah Hospital at 20 weeks' gestation. Previously in good health, she had been admitted to her local hospital with apparent pyelonephritis and treated with intravenous (IV) ampicillin and gentamicin. While hospitalized, she developed progressive shortness of breath with increasing oxygen requirements. An echocardiogram revealed severe tricuspid insufficiency, an enlarged pulmonary artery, and probable elevated pulmonary artery pressure.

Her past medical history was notable for a congenital ventricular septal defect, which was repaired at age 4 in her native Sweden. She was told at age 18 that her cardiac catheterization was normal, although the records of this examination are not available. At age 21, she was delivered by cesarean at 30 weeks' gestation for premature labor with a breech presentation. Her pregnancy and puerperium were otherwise uncomplicated. Her functional status was class I, without physical complaints until the current illness. She had a 15 pack-year history of cigarette smoking, but quit when her pregnancy was recognized.

At the time of admission, this thin woman had no respiratory distress receiving supplemental oxygen by face mask. Her pulse was 74 beats per minute and her blood pressure was 107/73 mmHg. On physical examination, minimal bibasilar crackles were present. Cardiovascular examination revealed a regular heart rate and rhythm. A grade III/VI holosystolic murmur was present, heard best at the left lower sternal border with radiation to the apex, and with moderately increased intensity during inspiration. At the left upper sternal border, a grade II/VI murmur with an ejection click could be heard. Jugular venous distention was 11 cm. On oxygen, 15 L/minute by face mask, her arterial blood gas had a pH of 7.52, an oxygen pressure of 70 mmHg, a carbon dioxide pressure of 22 mmHg, an HCO_3 of 16.6 mEq/L, and an oxygen saturation of 92%.

An echocardiogram revealed an enlarged right ventricle, severe tricuspid insufficiency, mild pulmonic insufficiency, a membranous ventricular septal defect, and a calculated pulmonary artery pressure of 100 mmHg. Cardiac catheterization documented a pulmonary artery pressure of 120/62 mmHg, a pulmonary artery wedge pressure of 10 mmHg, and bidirectional shunting across a high membranous ventricular septal defect, consistent with Eisenmenger syndrome.

When counseled regarding the grave prognosis of Eisen-

menger syndrome and pregnancy, the woman chose to terminate her pregnancy. After overnight dilation of the cervix with osmotic dilators, a dilation and evacuation was performed, initially under IV sedation. Because of an intraoperative episode of desaturation to 36% associated with hyperventilation, endotracheal intubation was performed, with prompt recovery. The surgical procedure was otherwise uncomplicated, with a total estimated blood loss of 50 mL. Her postoperative course was uncomplicated and she was discharged 4 days after surgery.

Eight weeks later, the patient returned for a follow-up examination. Systemic arterial pressure was 115/75 mmHg and severe pulmonary hypertension (122/75 mmHg) was still present, with a right-to-left shunt of 0.95 L/minute across the ventricular septal defect. The patient is currently being evaluated for a combined heart-lung transplant.

Case 2

A 23-year-old primigravid woman developed regular uterine contractions at 26 weeks' gestation. Her pregnancy had been uncomplicated to that point. Her medical history was significant for cardiac surgery at age 3 to repair "holes in the heart"; her parents had not sought a postoperative evaluation because they had believed that the surgical repair was complete. The woman had been asymptomatic after surgery and had not suffered any cardiopulmonary problems or limitation of physical activity.

At the time of presentation, she was found to be 6–7 cm dilated, with intact membranes and a viable fetus in a breech-funic presentation. A primary lower-segment cesarean delivery was performed under spinal anesthesia. The infant had Apgar scores of 5 and 7 at 1 and 5 minutes, respectively, and weighed 830 g.

Within 2 hours after surgery, the patient became pale and oliguric, with a heart rate of 120 beats per minute and a systemic arterial blood pressure of 95/60 mmHg. Hypovolemia was believed to be the likely cause of the symptoms, and IV fluids were administered. Dyspnea and hypoxemia then developed; systemic arterial oxygen saturation by pulse oximetry was 67% on room air, and rose to only 84% on 15 L/minute of supplemental oxygen.

Pulmonary embolism was suspected, and she was transferred to the intensive care unit. A ventilation-perfusion scan was interpreted as low probability for pulmonary embolism. Pulmonary artery catheterization revealed severe pulmonary hypertension with suprasystemic pulmonary artery pressures and no evidence of shunting; the pulmonary artery pressure was 135/75 mmHg and the pulmonary capillary wedge pressure was 22 mmHg. Despite aggressive cardiopulmonary support, the patient died on postoperative day 5 from progressive and intractable ventricular failure.

Records from childhood indicated that a holosystolic murmur was detected at 2–3 months of age. A membranous ventricular septal defect was closed with a Dacron patch at 3 years of age; severe pulmonary hypertension was suspected preoperatively. Following closure, the right ventricular pressure was 70/10 mmHg.

Discussion

With the exception of bicuspid aortic valve, an isolated defect in the ventricular septum is the most common congenital lesion of the cardiovascular system, accounting for approximately 20–30% of cardiac malformations, and with an overall incidence of 1.5–2.5 per 1000 live births. In about 80% of these cases, the defect is membranous or perimembranous, with extension into the muscular portion of the septum.[1] Spontaneous closure of ventricular septal defects is common, as one-quarter of all ventricular septal defects and about two-thirds of small defects eventually close. Of those ventricular septal defects that spontaneously close, 50% do so by age 3 and 85% by age 8.[2]

Small defects in the ventricular septum generally result in little or no abnormal hemodynamic function, and in almost all cases pulmonary vascular resistance is normal. Such defects are said to be restrictive, as the resistance to blood flow across the ventricular septal defect from left to right is at the level of the septum. With a small ventricular septal defect, right ventricular pressures are normal or only slightly increased. Moderate-sized lesions are also restrictive, but left-to-right shunting can lead to a mild increase in pulmonary vascular resistance. Large defects in the ventricular septum are nonrestrictive, as the resistance to flow from left to right is at the level of the pulmonary vasculature and not at the septal defect. This leads to elevated pulmonary vascular resistance and functional right ventricular outflow obstruction. Ultimately, right ventricular pressures can exceed those of the left side, and the shunt becomes right-to-left, a condition known as Eisenmenger syndrome.[1] Patients with Eisenmenger syndrome have a reported 50–70% mortality rate associated with pregnancy.[3] Pregnancy is also dangerous for patients with pulmonary hypertension without shunting, with maternal mortality rates of 30–40%.[4]

The major factor influencing whether pulmonary hypertension is present after surgical repair of a ventricular septal defect is the preoperative status of the pulmonary vascular bed. The larger the ventricular septal defect and the later in life that it is repaired, the more likely are pulmonary artery muscle hypertrophy and proliferation with resultant pulmonary hypertension at the time of surgery. Conversely, early repair before the development of increased pulmonary resistance is likely to be followed by normal cardiac and pulmonary function, and correction before age 2 is associated with an excellent prognosis. Patients with significantly elevated pulmonary vascular resistance in the immediate postoperative period generally suffer a gradual but progressive increase in pulmonary pres-

sures to suprasystemic levels. Although death in severe cases usually occurs within 10 years of surgery, patients with only moderately increased pulmonary vascular resistance can have a stable course, with a much slower progression of pulmonary disease.[5]

In the first case above, the report of a normal catheterization at age 18 and the successful navigation of pregnancy 3 years later suggest that the repair was either complete and later failed, or that it was actually incomplete and the residual defect was small and not noticeable. At the time of surgery, pulmonary vascular resistance was probably normal or only slightly increased. However, it is likely that after failure of the septal repair or simply with time and a residual defect, pulmonary hypertension developed and right-sided pressures eventually exceeded those of the systemic circulation. The decreased systemic vascular resistance that accompanies pregnancy resulted in development or worsening of a right-to-left shunt across the septal defect, producing Eisenmenger syndrome and decompensation. In the second case, right ventricular pressure was elevated immediately after childhood repair, and pulmonary hypertension was likely progressive over time until the pregnancy.

Despite apparently successful repair of a ventricular septal defect in childhood, residual cardiac disease may be present and progress into adult life. The hemodynamic alterations associated with pregnancy can result in cardiovascular deterioration of a patient who was functionally normal but at the limit of her ability to compensate before pregnancy. Patients with a history of repaired congenital cardiac disease should have their records carefully reviewed with attention to the diagnosis, type of repair, and postoperative evaluation. The patient's functional status should be closely assessed and the history and physical findings suggestive of residual cardiac disease sought. All patients with an abnormal physical finding or functional impairment should have an echocardiogram. These reports further suggest that, in cases in which the original lesion may lead to pulmonary hypertension, even patients with a normal physical examination and functional capacity should be strongly considered for echocardiography.

References

1. Perloff JK. The clinical recognition of congenital heart disease. 3rd ed. Philadelphia: WB Saunders, 1987:365–403.
2. Li MD, Keith JD. Spontaneous closure of ventricular septal defect. Am Heart J 1970;80:432–3.
3. Gleicher N, Midwall J, Hochberger D, Jaffin H. Eisenmenger's syndrome and pregnancy. Obstet Gynecol Surv 1979;34:721–41.
4. Elkayam U, Gleicher N. Primary pulmonary hypertension and pregnancy. In: Elkayam U, Gleicher N, eds. Cardiac problems and pregnancy. 2nd ed. New York: Alan R. Liss, 1990:189–97.
5. Kaplan S, Perloff JK. Survival patterns after surgery or interventional catheterization. In: Perloff JK, Child JS, eds. Congenital heart disease in adults. Philadelphia: WB Saunders, 1991:60–90.

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AMYOTROPHIC LATERAL SCLEROSIS IN PREGNANCY

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***Background*: Amyotrophic lateral sclerosis is rare in the obstetric population; only one case has been described in the last 18 years. Two pregnant women with this disease were observed within a 1-year period in Minneapolis.**

***Cases*: A twin pregnancy was diagnosed in a woman during hospital admission for evaluation of a pure lower motor neuron degenerative process. The disease was rapidly progressive, with maternal death occurring 6 weeks after the delivery of healthy twins at 34 weeks' gestation. A second patient with amyotrophic lateral sclerosis conceived in the early stages of respirator dependency; her disease remained stable throughout the pregnancy, and she delivered at 33 weeks. The neonates had a good outcome in both cases.**

Conclusion*: Amyotrophic lateral sclerosis, though rare in pregnancy, does occur and can result in good neonatal outcome. Maternal disease does not regress during pregnancy and may worsen under the increased respiratory and weight-bearing demands; whether this represents actual disease progression cannot be determined definitely. Labor management should include pulse oximetry determination and is facilitated by lack of disease involvement of the uterine sensory and motor nerves and a lack of resistance of the pelvic floor musculature. *(Obstet Gynecol 1993;82: 682–5)

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