Conducted during menses showed a small area of bronchial mucosa thickening, particularly evident with the virtual bronchoscopy and not visible during the first examination. Comparisons between the two CT scans led to the correct diagnosis. On the basis of such findings, flexible fiberoptic bronchoscopy, performed on the first day of menses, disclosed a tiny submucosal red spot in the left upper bronchus with signs of recent bleeding.

Medical therapy has been recommended as the first choice in pulmonary endometriosis. It consists of the suppression of endometrial tissue with progesterone (ie, pseudopregnancy) or danazol (ie, pseudomenopause). Danazol is a synthetic steroid with antiestrogenic and light androgenic effects that affects ovarian hormone synthesis.11 It has proved to be effective in curing or controlling symptoms, even in patients who are nonresponsive to ovulation suppression,12 but a variable recurrence rate after the cessation of therapy has been reported.1,13 Furthermore, heavy side effects of the hormonal therapy often are observed, including climacteric symptoms, virilization, weight gain, and sterility.4 Surgery should be the preferred method if the patient wishes to become pregnant, if the side effects of hormonal therapy are intolerable, or in case of recurrence when the drug therapy is discontinued. Pulmonary resection is indicated when a single point of bleeding has been located definitively. For peripheral lesions, thoracoscopic wedge resections have been successfully performed.4,14 In patients with centrally located bronchial endometriosis, subsegmentectomy, segmentectomy, or lobectomy are required.3,15–17 Treatment with oophorectomy has been reported in the literature,15 but it seems to be an extreme solution and should be avoided.

The precise endoscopic identification of the tracheobronchial lesions brings new therapeutic options. With the combination of CT scanning and flexible fiberoptic bronchoscopy, the source of bleeding should be located in every tracheobronchial endometriosis.

In our patient, we decided to proceed to laser treatment because the lesion had been precisely located in a bronchus, which is easily reachable by the bronchoscope. We preferred the ventilating rigid bronchoscopy mainly for our extensive experience in the use of this procedure. Nevertheless, a successful laser ablation of the lesion also could have been performed through the flexible fiberoptic bronchoscope.

No previous endoscopic treatments of this condition have been reported in the literature. Endoscopic Nd-YAG laser can eliminate mucosal and submucosal lesions with a minimally invasive procedure, without significant operative risk. Such treatment could be the first line of therapy for central airway endometriosis, provided that the source of bleeding has been conclusively located and the lesions can be reached with the bronchoscope. Endoscopic ablation potentially can achieve good and probably long-term outcomes without the adverse effects of pharmacologic therapy and surgical therapy.

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Successful Management of Pregnancy in a Patient With Eisenmenger Syndrome With Epoprostenol*

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Pregnancy in the setting of pulmonary hypertension and Eisenmenger physiology is associated with a substantial maternal and fetal risk. Such patients are advised against pregnancy. We report a case of a woman with an Eisenmenger atrial septal defect diagnosed during the last trimester of pregnancy. On presentation, she was critically ill and there was evidence of fetal distress. She was emergently...
treated with IV epoprostenol, and her status improved. She underwent cesarean section and delivered a male infant with Apgar scores of 8 and 9. Her dyspnea improved, and she was characterized as World Health Organization functional class II on a subsequent clinical visit. Although pregnancy should be discouraged in women with Eisenmenger syndrome, we have demonstrated that IV epoprostenol successfully treated a woman with Eisenmenger syndrome diagnosed in the third trimester.

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Key words: Eisenmenger syndrome; epoprostenol; pregnancy; pulmonary hypertension

Eisenmenger syndrome consists of a congenital communication between the systemic and pulmonary circulation, with resultant pulmonary arterial hypertension and reversal of flow through the defect. In a review1 of outcome of pregnancy in patients with pulmonary vascular disease from 1978 through 1996, the maternal mortality rate in the Eisenmenger syndrome was 36%, which did not represent an improvement when compared to the previous review published in 1979. Over the past decade, the treatment of primary pulmonary hypertension has been revolutionized with the use of IV epoprostenol.2 Noncontrolled data suggest that epoprostenol is also useful in patients with congenital heart disease.3 We report a case of Eisenmenger syndrome diagnosed during the third trimester of pregnancy and successfully managed with IV epoprostenol. To our knowledge, this is the first report of the use of epoprostenol to manage pregnancy in a patient with Eisenmenger physiology.

CASE REPORT

A 21 year-old gravida 3 para 2 woman was transferred to our institution in August of 2001 at 34 1/7 weeks' gestation for the management of dyspnea and preterm contractions. The patient had two uncomplicated vaginal deliveries in 1997 and 1999 and had done well during this pregnancy until several weeks prior to hospital admission when she complained of increasing shortness of breath and bilateral lower-extremity edema. She denied symptoms of chest pain, light-headedness, syncope, and palpitations. Her pregnancy had been uncomplicated until her recent hospital admission. She had gained 70 lb during her pregnancy. Her medical history was unremarkable outside of the two pregnancies. Her only medications included prenatal vitamins.

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On initial physical examination, she was in respiratory distress with an oxygen saturation of 80% on a 100% nonrebreather mask. Her heart rate was 107 beats/min, respiratory rate was 24 breaths/min, and BP was 134/73 mm Hg. Her jugular venous pressure was elevated. Her carotid upstrokes were reduced. Her lungs were clear to auscultation. She had a palpable right ventricular heave. On cardiac auscultation, she was tachycardic with a normal S1, a loud pulmonic component to the second heart sound, 2/6 murmur of tricuspid regurgitation, and a right-sided S3. Her abdomen was gravid. She had trace bilateral lower-extremity edema.

Her ECG demonstrated right-axis deviation and right ventricular hypertrophy. Her chest radiograph demonstrated an enlarged cardioiodiaphragmatic silhouette. Her chest CT did not demonstrate any evidence of intestinal lung disease or pulmonary embolus. Her echocardiogram demonstrated marked right atrial and right ventricular enlargement, and severe pulmonary hypertension with an estimated pulmonary artery pressure of 120 mm Hg (Fig 1). With color-flow Doppler echocardiography and an injection of agitated saline solution, she had evidence of right-to-left shunting through an atrial septal defect at rest (Fig 2).

The patient was managed in conjunction with the high-risk obstetric service who performed a biophysical profile (assessment of reactive nonstress test, fetal breathing movement, fetal body movement, fetal tone, and amniotic fluid volume), which suggested fetal distress. Because of her severe hypoxemia, respiratory distress, and newly diagnosed pulmonary hypertension, the patient was started on IV epoprostenol. No other therapies were instituted or altered. Over the ensuing 24 hr, the epoprostenol was titrated up to 9 ng/kg/min with an improvement in her oxygenation to a saturation of 90% and tachycardia. She was then taken to the operating room where she underwent a cesarean section and delivered a male infant with Apgar scores of 8 and 9. Postoperatively, she received anticoagulation with enoxaparin, 80 mg bid subcutaneously. One week postpartum, a right-heart catheterization was performed while the patient was receiving IV epoprostenol (Table 1). The atrial septal defect was easily crossed with a multipurpose catheter. Based on a measured oxygen uptake of 134 mL/min, her pulmonic blood flow calculated to 3.3 L/min and systemic blood flow to 3.5 L/min. Her pulmonary vascular resistance was 16.6 Wood units, and systemic vascular resistance was 28.2 Wood units.

Her hospital course was complicated by an infected hematoma at the site of her surgical incision, which required IV antibiotics. The patient was discharged after a 2-week hospitalization receiving warfarin anticoagulation and epoprostenol. At a subsequent clinic visit, she reported a marked improvement in her dyspnea and was World Health Organization functional class II receiving 12 ng/kg/min of epoprostenol.

DISCUSSION

Eisenmenger syndrome is one of the few cardiac conditions for which pregnancy is considered absolutely contraindicated. The poor outcome during pregnancy is a result of impaired hemodynamics and exercise capacity superimposed on the gestational cardiovascular demands to which there is insufficient adaptation of the right heart and poorly compliant pulmonary vasculature. Normal physiologic changes of pregnancy can worsen right-to-left flow across a congenital defect and result in worsening hypoxemia.

In general, therapy of Eisenmenger syndrome is supportive.4 Patients should avoid intravascular volume depletion, heavy exercise, and high altitude. Phlebotomy with isovolemic replacement should be performed in patients with moderate or severe symptoms of hypervisci-
cosity, but may result in iron deficiency anemia. More recently, IV epoprostenol has been successful in patients with congenital heart disease.3

To our knowledge, this is the first case report of treatment of a pregnant woman with Eisenmenger syndrome with IV epoprostenol. This patient received her diagnosis late in pregnancy, beyond the time at which a therapeutic termination could have been performed. She was managed with a multidisciplinary approach, and her care included cardiologists, high-risk obstetricians, and

![Figure 1. Two-dimensional, apical, four-chamber view demonstrating right atrial and ventricular enlargement. RA = right atrium; RV = right ventricle; LA = left atrium; LV = left ventricle.](image1)

![Figure 2. Color Doppler echocardiography demonstrating flow across the atrial septal defect. See Figure 1 legend for expansion of abbreviations.](image2)
anesthesiologists. The relatively short-term effects of epoprostenol likely included vasodilatation (both pulmonary and systemic) and increased cardiac output resulting in improved oxygenation and placental blood flow. General anesthesia and cesarean section were successfully performed in this patient, in part related to the multidisciplinary approach. She received aggressive anticoagulation, which is important as deep venous thrombosis and pulmonary embolism are important causes of postpartum mortality in patients with pulmonary arterial hypertension. Although pregnancy should be discouraged in women with Eisenmenger syndrome, we have demonstrated that IV epoprostenol successfully treated a woman with Eisenmenger syndrome diagnosed in the third trimester.

References

Table 1—Right-Heart Catheterization

<table>
<thead>
<tr>
<th>Variables</th>
<th>Pressure, mm Hg</th>
<th>O₂ Saturation, %</th>
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</thead>
<tbody>
<tr>
<td>Right atrium</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>98/40, 61</td>
<td>75.0</td>
</tr>
<tr>
<td>Left atrium</td>
<td>6</td>
<td></td>
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<tr>
<td>Systemic arterial</td>
<td>122/51, 74</td>
<td>90.4</td>
</tr>
<tr>
<td>Pulmonary vein</td>
<td></td>
<td>98.0</td>
</tr>
<tr>
<td>Inferior vena cava</td>
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<td>73.4</td>
</tr>
<tr>
<td>Superior vena cava</td>
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<td>63.0</td>
</tr>
</tbody>
</table>

Overline indicates mean pressure.