Peripartum Cardiomyopathy

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DR. MICHAEL O'DELL *(Assistant Professor, Department of Family Practice):* Acute medical illnesses arising in the peripartal period are an important concern to the physician with an obstetrical practice. Peripartum cardiomyopathy is a rare primary myocardial disease that develops during pregnancy or within the first five months following delivery. The case presented outlines the clinical course of a patient who develops pulmonary edema shortly after delivery, and it should serve to heighten awareness of this uncommon but potentially devastating illness.

A 31-year-old, gravida 4, para 4, housewife was admitted to the hospital with complaints of increasing cough, sputum production, and progressive dyspnea five days after delivery. Her illness had started 15 days before delivery, when she developed symptoms of hoarseness, sore throat, productive sputum, and pleuritic chest discomfort suggestive of an upper respiratory tract infection. The patient was started on erythromycin but returned to the Family Practice Clinic six days later with persistent symptoms. A chest roentgenogram was obtained that revealed patchy pneumonitis involving both lower lobes. The patient was admitted two days prior to delivery because of persistent pleuritic chest discomfort. A pericardial friction rub was heard, but no clinical evidence of congestive heart failure was noted at that time. Her chest discomfort, dyspnea, and sputum production markedly improved following delivery. The delivery was complicated by meconium aspiration by the infant. A follow-up x-ray examination, one day after delivery and four days before the onset of congestive heart failure, showed resolution of the pneumonitis.

At the time of admission, five days after delivery, her blood pressure was 150/100 mmHg, pulse 120 beats per minute, and her respiratory rate was 24 per minute. The patient was febrile. Moderate respiratory distress was noted, and bibasilar crackles were heard over the lower lung fields. First and second heart sounds were soft, and no murmurs, rubs, or gallops were appreciated. Her chest x-ray films revealed fluffy perihilar infiltrates. A ventilation-perfusion scan revealed no evidence of pulmonary embolus.

Respiratory distress continued to worsen after admission, and the patient exhibited marked air hunger requiring intubation. The chest roentgenogram revealed progression to a diffuse five-lobe infiltrate. A Swan-Ganz catheter was promptly placed, which revealed a pulmonary artery pressure of 50/20 mmHg with a pulmonary capillary wedge pressure (PCW) of 24 mmHg. A multigated (MUGA) cardiac scan revealed an ejection fraction of 13 percent, implicating severe acute left ventricular dysfunction as a cause for respiratory distress.

The patient slowly improved with digitalis, diuresis, and ventilatory support. She was extubated 12 days after admission, and further recovery continued as the patient was able to participate in a cardiac rehabilitation program.

The patient was discharged 31 days after admission. Dr. Ruth from the Division of Pulmonary Disease will discuss the treatment of acute respiratory distress in this postpartum patient.

DR. WILLIAM RUTH *(Professor, Department of Medicine):* Dr. O'Dell has outlined a picture of a patient who had rapid deterioration of gas transfer. Soon after she was intubated, her alveolar-arterial (A-A) gradient was noted to be 565 mmHg, which indicates that her pulmonary blood was either being shunted through nonventilated segments or that there was a marked impairment of gaseous diffusion caused by alveolar and interstitial pulmonary edema. Starling's law uses the balance of hydrostatic and oncotic pressures to explain why pulmonary edema occurs. The increase in pulmonary capillary wedge pressure leads to the conclusion that an increase in pulmonary capillary hydrostatic pressure secondary to left ventricular failure is one of the causative factors that led to this patient's deterioration.

In addition to left ventricular failure, it is possible that sepsis could have contributed in part to this patient's respiratory distress. The systemic vascular resistance calculated from the hemodynamic data obtained from her Swan-Ganz catheter was consistently

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low, in the range of 600 to 700 dynes/sec/cm-5. This reading is more characteristic of sepsis than of heart failure. Moreover, the patient was febrile at the time of decompensation, and a small amount of necrotic decidual tissue was obtained several days later at the time of curettage.

No organisms were cultured from this tissue, but the patient was on broad-spectrum antibiotics at the time of curettage.

Finally, in consideration of the prodrome that the patient experienced prior to delivery, it is possible that a viral illness was in part responsible for triggering respiratory decompensation. The adult respiratory distress syndrome may develop as a sequela of certain viral infections or viral pneumonitis.

In conclusion, we are certainly dealing with left ventricular heart failure as a cause for respiratory failure. Concurrent pelvic infection and viral pneumonitis may also have been important factors.

DR. O'DELL: Other considerations could have included preeclampsia; however, no evidence of chorionic villi was present on the curettage specimen, making this unlikely. Urinalysis revealed only trace protein as well. Dr. Steven Gollub from the Division of Cardiovascular Diseases will discuss the diagnostic and therapeutic approach to the postpartum patient with cardiomyopathy as well as a natural history of the disease.

DR. STEVEN GOLLUB *(Assistant Professor of Medicine, Cardiovascular Diseases):* The present case is instructive because it directs attention to the differentiation of the cardiac and noncardiac causes of respiratory distress within the peripartal period. Such patients are critically ill, and appropriate therapeutic decisions depend upon a diagnostic rationale to define the underlying problem. Additional information, besides that which is provided by the history, physical examination, chest x-ray examination, electrocardiogram, and routine laboratory tests, is frequently required.

Dr. Ruth has already stressed the importance of the hemodynamic data obtained from the Swan-Ganz catheter. While elevated pulmonary artery pressures commonly result from hypoxia, in the absence of mitral value disease, an increase in pulmonary capillary wedge pressure reflects an increase in left ventricular end diastolic pressure. This indicates that left ventricular dysfunction is responsible for pulmonary edema and respiratory distress.

Further assistance in differentiating respiratory distress secondary to cardiogenic pulmonary edema from noncardiac causes can be gained from MUGA scans and the echocardiogram. Portable MUGA scans are available and can be performed on critically ill patients in an intensive care unit setting. While acute right ventricular heart failure could result from hypoxia and pulmonary hypertension of any cause, left ventricular dysfunction confirmed by MUGA scan would implicate cardiac failure as a cause of respiratory distress. In the patient under discussion, a portable MUGA scan was obtained within two days of admission and revealed a left ventricular ejection fraction of 13 percent, confirming respiratory failure secondary to cardiogenic pulmonary edema.

Corroborative data can also be obtained from M-mode and two-dimensional echocardiography. M-mode echocardiography might indicate dilated chamber dimensions with indices of impaired left ventricular function. Two-dimensional echocardiography might reveal decreased wall motion with or without dilatation. In this particular patient, echocardiograms were obtained on several occasions and did reveal chamber enlargement and decreased left ventricular function.

While confirming the presence of impaired left ventricular function, the echocardiograms were of marginal quality, probably a reflection of the patient's body habitus and ventilator dependence during the first days of her hospitalization.

A strong case for cardiogenic pulmonary edema has been established, yet the patient had no history of cardiac disease prior to her most recent conception. Peripartum cardiomyopathy is recognized as a distinct clinical entity, although its cause remains unknown. Speculation exists in the literature that the disease may be secondary to viral infection, immunologic injury, physiologic stress, toxic insult, hormonal imbalance, or a combination of such factors.[1-4] Demakis and Rahimtoola[1] restrict peripartum cardiomyopathy to include only those women who develop congestive heart failure of undetermined etiology in the last month of pregnancy or within the first five postpartum months and who have no previous history of heart disease. The patient under discussion meets the criteria for this diagnosis.

A review of the natural history of peripartum cardiomyopathy provides valuable information for formulating a rational approach toward the treatment of this disease entity. In 1971 Demakis and colleagues[5] reported the natural history of 27 women with the diagnosis of peripartum cardiomyopathy that were followed for as long as 21 years. The authors found that cardiomyopathy was associated with multiparity, toxemia, twins, and a maternal age above 30 years. Patients were divided into two groups depending on whether cardiomegaly had resolved within a six-month period. Prognosis was good for patients with resolved cardiomegaly. No cardiac deaths were reported, and the majority were functional class I.

In addition, no permanent detriment occurred with future pregnancy. In contrast, 11 of 13 patients with persistent cardiomegaly died within an average of 4.7 years. Three died of exacerbation of congestive heart failure during a future pregnancy.

Recently several groups have reported cases of inflammatory myocarditis that appeared responsive to

immunosuppressive treatment with corticosteroids and azathioprine.[6-8] These have included cases of peripartum cardiomyopathy.[7,8] Diagnosis is confirmed by percutaneous transvenous endomyocardial biopsy of the right ventricle. This procedure can be performed in the catheterization laboratory with an extremely low incidence of complications. Excellent specimens are obtained for light and electron microscopy. Patients who demonstrate inflammatory myocarditis are candidates for immunosuppression. The histologic picture of inflammatory myocarditis reveals lymphocytic infiltration of the interstitium and perivascular regions with or without myocytolysis. Treatment is generally started with prednisone and azathioprine, continued until there is histologic improvement, and then gradually tapered over a period of months as the clinical course remains stable.

It is important to maintain proper perspective when discussing endomyocardial biopsy diagnosis of inflammatory myocarditis and subsequent treatment with immunosuppressive therapy. This represents an innovative but as yet unproven approach toward a specific type of myocarditis of which certain cases of peripartum cardiomyopathy may be included. There are no large randomized series studying the effect of immunosuppression on myocarditis.

To the contrary, only a few, small, select series describing a histologic response with clinical improvement in response to immunosuppressive therapy have been reported. Yet the approach appears attractive and rational, especially when the response to conventional therapy is slow.

The patient under discussion was treated conventionally, and her clinical progress was measured by serial MUGA scans, which revealed slow and incomplete improvement. Endomyocardial biopsy was performed but did not reveal evidence of an active inflammatory myocarditis. Light microscopy revealed only minimal changes with no lymphocytic infiltration of the interstitial and perivascular areas. Electron microscopy, however, did reveal focal abnormalities with dilation of the sarcotubulary system, elongation of mitochondria, and disruption of the orderly sequence of light and dark bands. Since there was no evidence of current inflammatory myocarditis, treatment with digoxin and diuretics was continued, and immunosuppressive therapy was withheld.

In the absence of evidence indicating a distinct cause for heart failure and considering the time course, this patient's illness does conform to the diagnosis of peripartum cardiomyopathy. It is certainly conceivable that sepsis or viral pneumonitis as suggested by Dr. Ruth also contributed to her fulminant respiratory distress, although the initial severity of the cardiomyopathy as indicated by her MUGA scan would lead me to believe that this was the principal factor responsible for her decompensation.

CANDACE GORTNEY *(Nurse Practitioner and Teaching Assistant, Department of Family Practice):* As the family practice nurse educator, I had become acquainted with the family several months prior to the onset of the cardiomyopathy. I had been teaching parenting skills to the patient and her spouse. The family had been experiencing relatively severe behavioral difficulties with two of their children. It had become apparent that ineffective parenting was causing the problem. We had quickly discovered that much of the difficulty resulted from the father's inconsistent discipline and lack of other parenting skills. Unfortunately it was felt that the chances of improving the father's skills were minimal. He seemed to function intellectually at a level similar to his children. I feel that this mother's illness was complicated by her attempts to keep a family of three children, with an ineffective spouse, functioning.

During her illness several caretakers were developed from within the family. This occurred spontaneously and without much intervention on our part. Again, it was clear that the patient was the caretaker for not only her family but possibly for the extended family as well. It was only in the event of her illness that other members of the family assumed a more responsible role.

A number of potential difficulties in establishing a good maternal-infant bond were present in this case. The pregnancy had been unplanned, although it was well accepted by the mother. The infant's meconium aspiration at delivery necessitated treatment of the infant in an intensive care unit setting. This was followed by the mother's own hospitalization. Every effort was made, when the patient stabilized, to allow her to hold her infant while in the intensive care unit. Subsequent follow-up has shown that the maternal-infant interaction is appropriate.

DR. O'DELL: The patient's subsequent outpatient course has been one of steady improvement. She has improved symptomatically and has regained her prepregnancy exercise tolerance. Although a third heart sound was noted intermittently during the early posthospitalization months, this is now absent. She has developed a first-degree atrioventricular block, which seems to be of only minor significance. Her cardiac ejection fraction as measured by MUGA scanning is now normal (Figure 1). In all, her clinical response has been gratifying. With the rapid return of her heart size to normal, her long-term prognosis is excellent.[9]

She has been counseled regarding the 50 to 88 percent risk of developing congestive heart failure with future pregnancies.[10] She and her spouse elected vasectomy as a means of permanent sterilization.

Deterioration of her children's behavioral problems was noted during her hospitalization. With improvement in her clinical status, she has again regained con-

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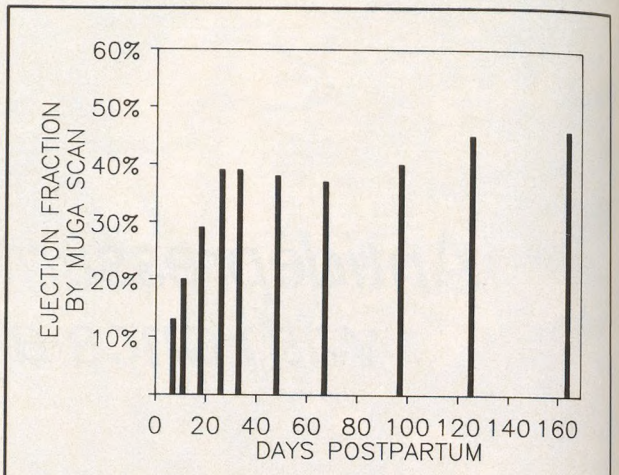

Figure 1. Improvement in the cardiac function with time as measured by several multigated (MUGA) scans

trol of the family. Her family situation remains difficult but manageable by her.

At this point I feel the patient has recovered completely from her illness and its effects. This seems remarkable in view of the 30 to 60 percent mortality reported by various authors.[10] She has also been able to resume her full activities, including the care of her youngest child. Each of the various disciplines involved contributed substantially to this outcome.

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