Uneventful Pregnancy and Delivery in a Patient with Idiopathic Pulmonary Hypertension: a Case Report

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Idiopathic pulmonary hypertension is a rare disease characterized by sustained elevation of the pulmonary artery pressure and pulmonary vascular resistance, normal pulmonary artery wedge pressure, in the absence of a known cause. Prior reports suggest a very high maternal mortality in patients with idiopathic pulmonary hypertension undergoing pregnancy, and for that the recommendation has been avoidance of pregnancy (or termination if the patient is already pregnant). On the other hand, there have been multiple reports of patients with idiopathic pulmonary hypertension sustaining pregnancy and labor without major complications. This case report illustrates the course of pregnancy and labor in a patient diagnosed with idiopathic pulmonary hypertension. At age 24, the patient started with symptoms of shortness of breath and chest pain, and upon evaluation she was found with moderately severe idiopathic pulmonary hypertension. One year and 8 months later the patient becomes pregnant, and begins follow up with gynecology and cardiology. During this time the patient was asymptomatic, and did not have any clinical evidence of pulmonary hypertension. The risks of pregnancy were discussed with the patient, and she decided to continue pregnancy. She had an uneventful pregnancy, complicated only by preterm labor at 34 weeks and 5 days of gestation. She had spontaneous labor and delivered vaginally a healthy baby boy, weighting 4 pounds and 12 ounces. No invasive monitoring was used. The mother and the baby were discharged home 48 hours postpartum. Seven months later the patient returned for evaluation, presenting evidence of severe pulmonary hypertension. She has been followed up ever since by a cardiologist and currently is stable but symptomatic. This report adds to the amount of evidence that suggests that pregnancy and labor in a patient with idiopathic pulmonary hypertension may have a better outcome than previously reported. The decision of undertaking and/or continuing pregnancy in a patient with idiopathic pulmonary hypertension relies ultimately on the patient's choice, but should be done on an individual basis after careful evaluation of the risks. Finally, the need of close follow up with a multidisciplinary team is mandatory in the patient with idiopathic pulmonary hypertension that wishes to undergo pregnancy.

Key words: Idiopathic pulmonary hypertension, Pregnancy, Maternal mortality, Multidisciplinary team.

Idiopathic pulmonary hypertension, previously termed primary pulmonary hypertension, is a rare disease characterized by sustained elevation of the pulmonary artery pressure and pulmonary vascular resistance, normal pulmonary artery wedge pressure, in the absence of a known cause. It is more common in women, usually from 10 to 40 years of age (childbearing age) (1). Prior reports suggest a very high maternal mortality in patients with idiopathic pulmonary hypertension undergoing pregnancy (2), and for that reason the recommendation has been avoidance of pregnancy (or termination if the patient is already pregnant). On the other hand, there have been multiple reports of patients with idiopathic pulmonary hypertension sustaining pregnancy and labor without major complications (3-11). This case report illustrates the course of pregnancy and labor in a patient with idiopathic pulmonary hypertension.

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Case report

On November 2001, a 24 year old Puerto Rican female without previous history of systemic illness, one previous pregnancy without complications, while living at New Orleans, Louisiana, presented to emergency room with episodes of syncope, chest pain and dyspnea on exertion of two weeks duration. No prior history of similar

symptoms. Upon evaluation the patient was found with stable vital signs (pulse 100 beats per minute, blood pressure 130/80, respiratory rate 20 per minute, afebrile), jugular venous distention (10 cm above the sternal angle), heart with regular rate and rhythm, systolic murmur III/VI intensity best heard at the left sternal border in the second intercostal space. The lungs were clear to auscultation bilaterally. The liver and spleen were not palpable. There were no edema, cyanosis or clubbing. The complete blood count, electrolytes and cardiac markers were normal. The chest x ray revealed an enlarged heart and prominent pulmonary arteries. Two dimensional echocardiography revealed a dilated right atrium and right ventricle, abnormal septal motion suggestive of right ventricular overload, normal left ventricular systolic function, moderately severe tricuspid regurgitation and moderately severe pulmonary hypertension (estimated peak systolic pulmonary artery pressure of 70 mm Hg). Contrast bubble test did not reveal right to left shunting. No evidence of septal defects. Extensive workup for secondary causes of pulmonary hypertension was done, and the results were negative, including rheumatologic workup, HIV test, pulmonary function tests, spiral computer tomography of the chest (CT angiogram), and lower extremity Doppler. In view of the above findings, the diagnosis of idiopathic pulmonary hypertension was made. Right heart catheterization was done revealing moderately severe pulmonary hypertension (mean pulmonary artery pressure of 50 mm Hg), responsive to calcium channel blockers (diltiazem). The patient improved clinically, and was discharged home on oral nifedipine. The patient at this time returned to Puerto Rico.

At age 26 (July 2003) patient became pregnant and was followed up at the high risk Gynecology Clinic and Cardiology Clinic. At this time the patient was feeling well, essentially asymptomatic, with a normal physical examination. The electrocardiogram of August 2003 (see Figure 1) did not show right chamber enlargement, and the chest x ray was within normal limits. The two dimensional echocardiogram revealed a mildly enlarged right ventricle, mild tricuspid regurgitation, no evidence of pulmonary hypertension, and normal left ventricular systolic function (August 2003, see Figure 2). The risks of pregnancy were discussed with the patient, and she decided to continue pregnancy. Since the patient was doing so well and there was no evidence of significant pulmonary hypertension at that time, there were doubts about the prior diagnosis of idiopathic pulmonary hypertension. She was followed closely by the cardiology and gynecology services, and had an uneventful pregnancy until 33 weeks and 2 days of gestation, when she presented with pelvic pain and preterm labor. The electrocardiogram was repeated at this time (March 2004, see Figure 3) and did

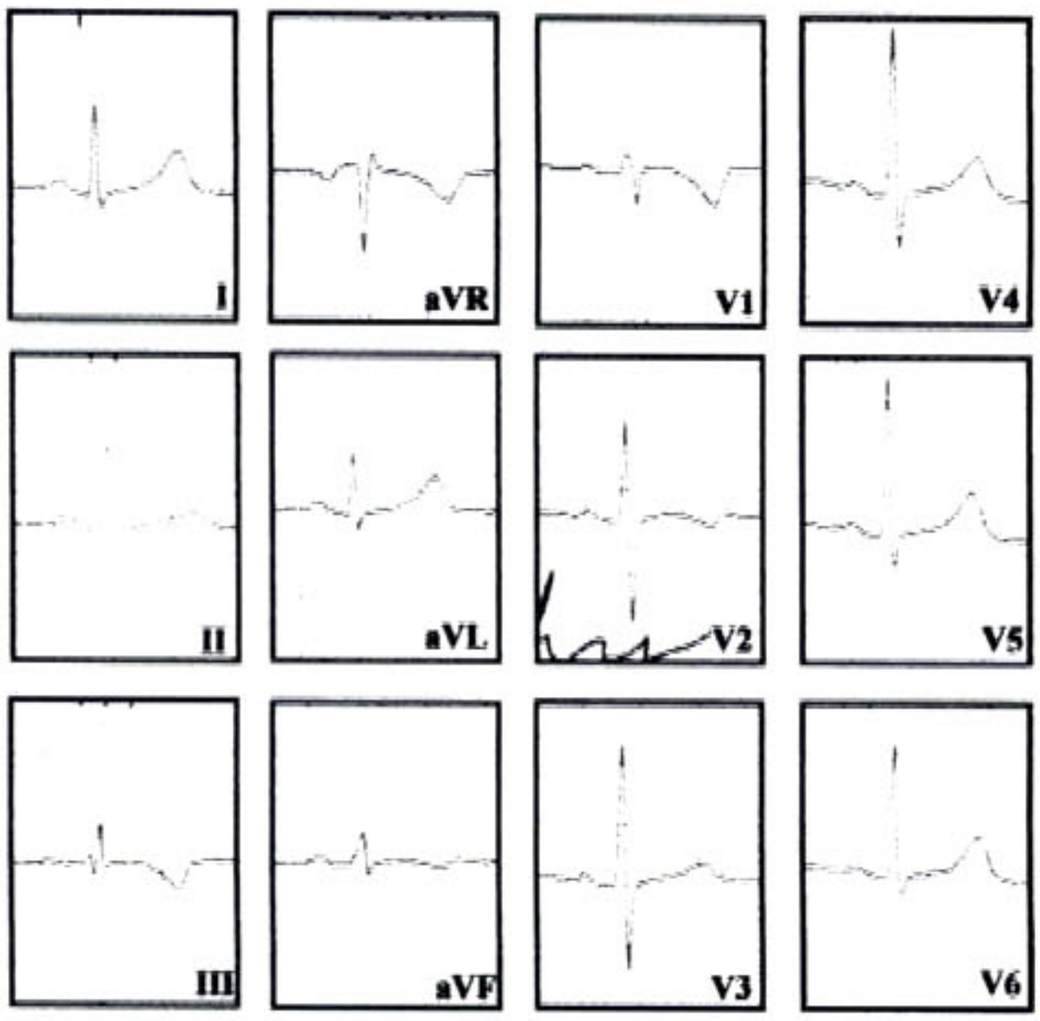

Figure 1. Electrocardiogram done on August 2003, early in pregnancy.

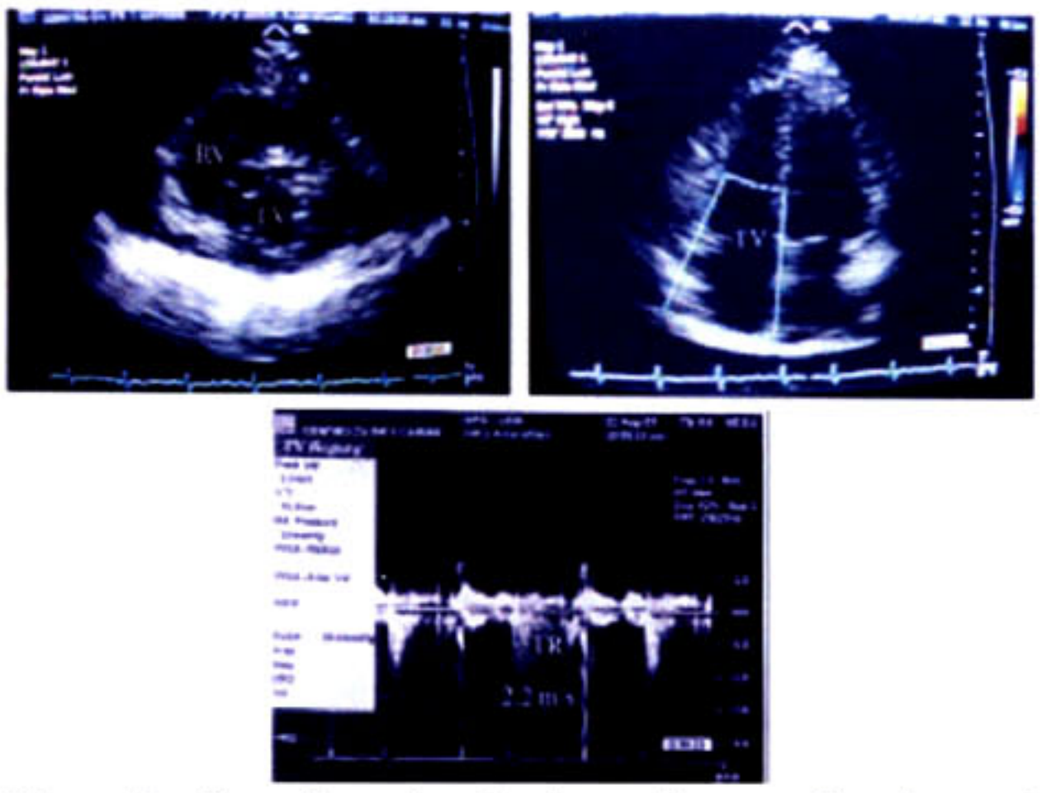

Figure 2. Two dimensional echocardiogram, Doppler, and color flow done on August 2003, early in pregnancy. LV = left ventricle, RV = right ventricle, TR = tricuspid regurgitation, TV = tricuspid valve.

not reveal any definite abnormality. Two dimensional echocardiography study revealed normal sized chambers, normal left ventricular systolic function, mild tricuspid regurgitation, and no evidence of pulmonary hypertension. On March 2004, at 34 weeks and 6 days of gestation, she had spontaneous labor and delivered vaginally a healthy baby boy, weighting 4 pounds and 12 ounces. The patient received epidural anesthesia. She was monitored during labor with the electrocardiogram, pulse oximetry, and blood pressure by cuff. No invasive monitoring was used. Vital signs during labor and post partum were stable. The patient sustained laparoscopic sterilization the next day with

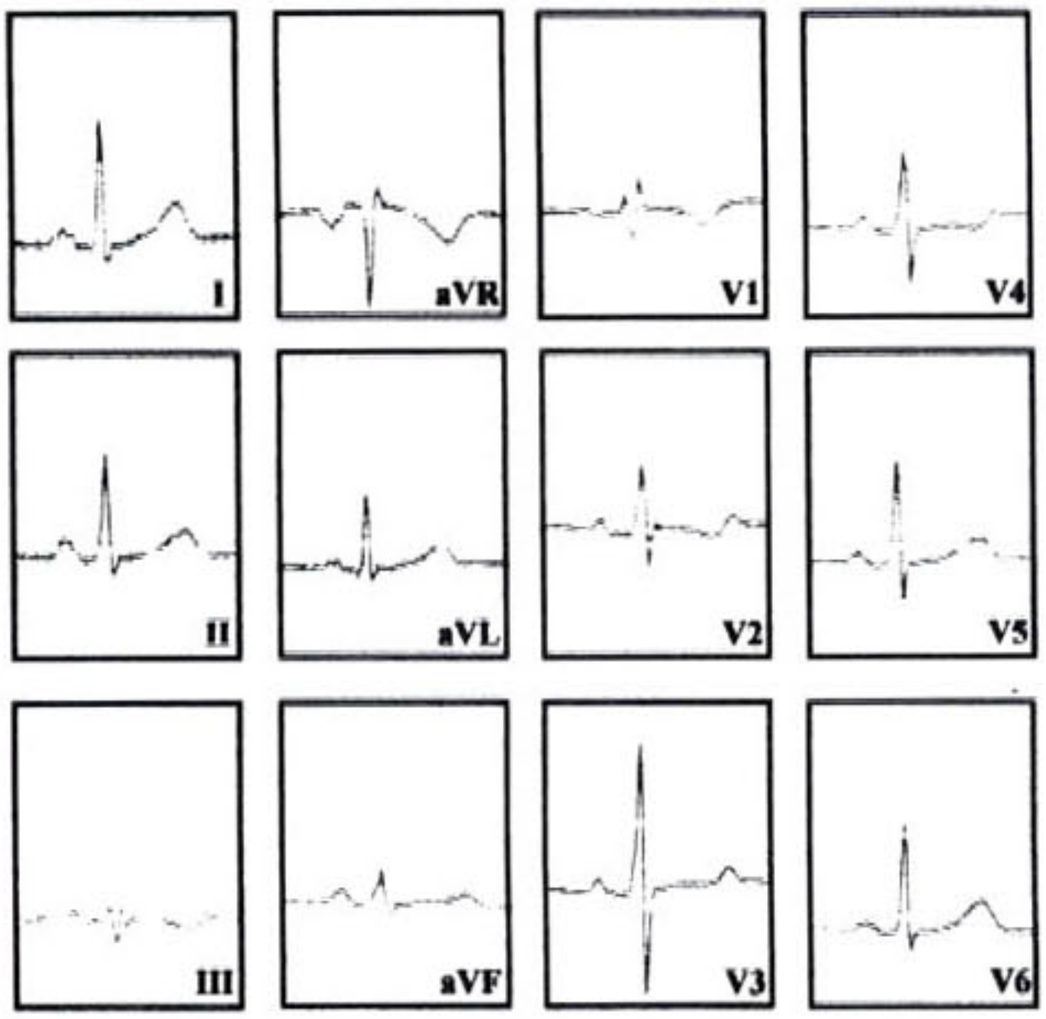

Figure 3. Electrocardiogram done on March 2004, pregnancy at term.

spinal anesthesia without any complications. The mother and the baby were discharged home 48 hours postpartum. The patient was not taking any medications at this time.

Seven months later (October 2004) the patient returned for evaluation, presenting symptoms of shortness of breath and chest discomfort. The electrocardiogram at that time revealed right axis deviation, right atrial and right ventricular enlargement with secondary ST and T wave abnormalities (see Figure 4). Chest x ray revealed mild

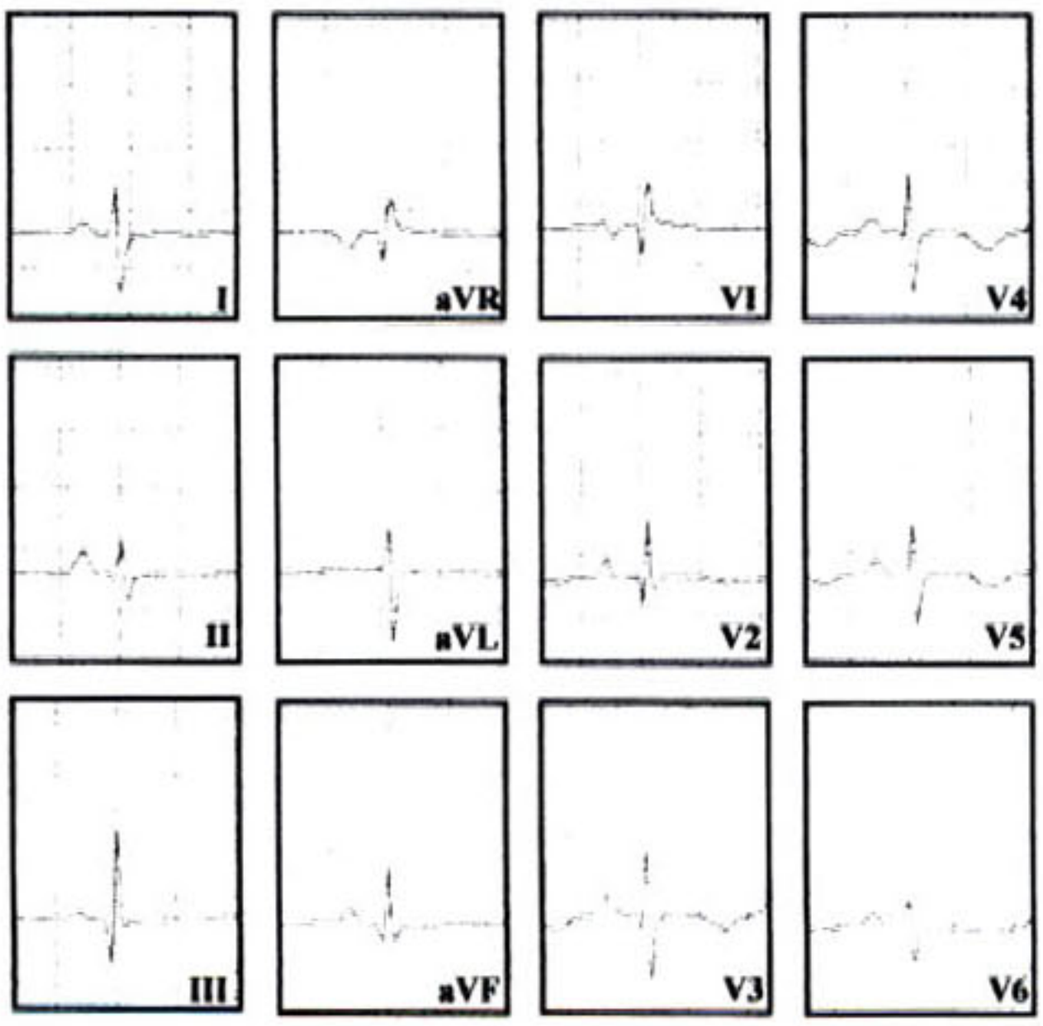

Figure 4. Electrocardiogram done on October 2004, seven months post partum.

cardiomegaly and enlarged pulmonary arteries. Lung ventilation-perfusion scan demonstrated low probability for pulmonary embolism. Two dimensional echocardiography revealed a D- shaped left ventricle consistent with right ventricular overload, right sided chamber enlargement, severe tricuspid regurgitation, estimated peak systolic pulmonary artery pressure of 85 mm Hg (severe pulmonary hypertension), normal left ventricular systolic function, with an ejection fraction of

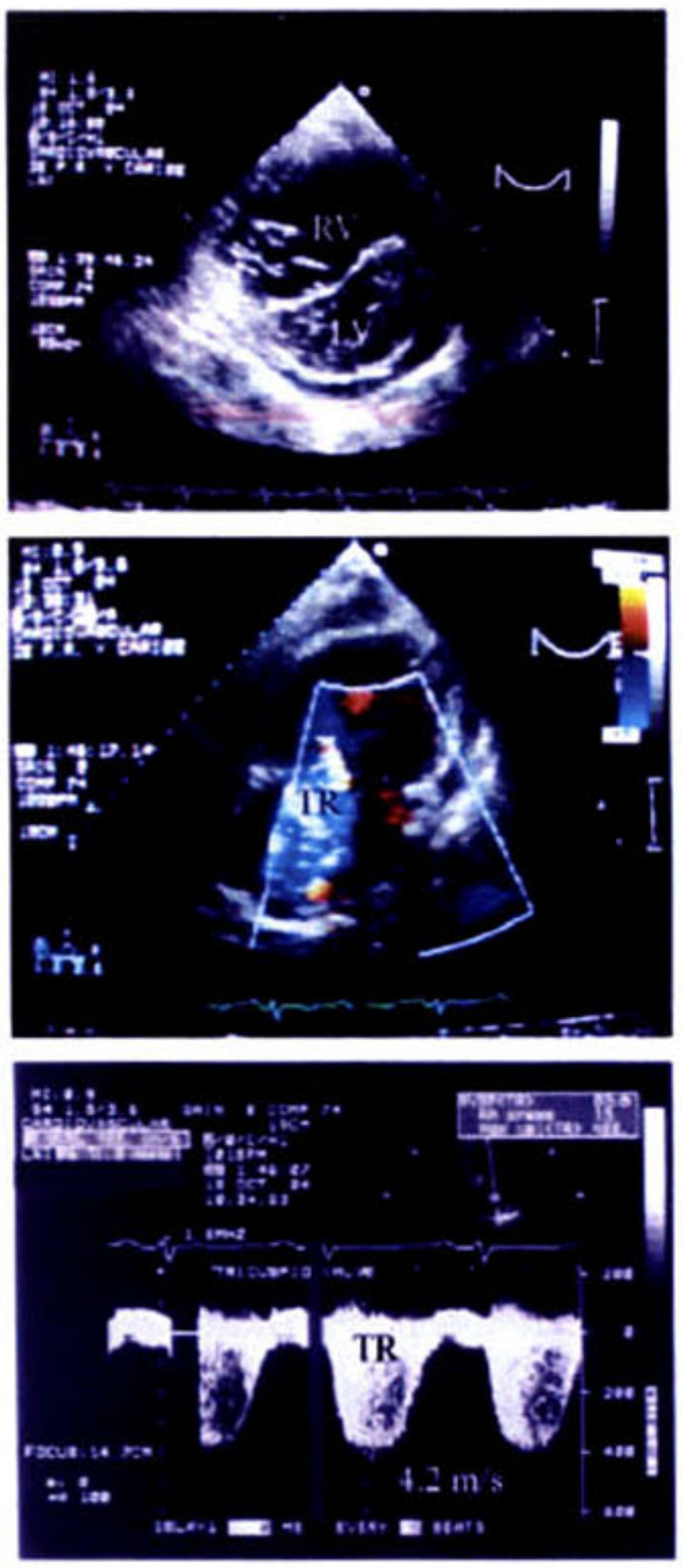

Figure 5. Two dimensional echocardiogram, Doppler, and color flow done on October 2004, seven months post partum. LV = left ventricle, RV = right ventricle, TR = tricuspid regurgitation.

60% (see Figure 5). Right heart catheterization done in November 2004 revealed severe pulmonary hypertension: pulmonary artery pressure 84/47 mm Hg, mean 60, versus an aortic pressure of 91/76 mm Hg. An oximetry run did not reveal oxygen step up. The pulmonary arterial tree was challenged with 6, 12 and 18mg of adenosine intravenously (every 2 minutes) and no change in

pulmonary artery pressure was obtained. The patient was discharged on warfarin, digitalis, ACE inhibitor, diuretics and oxygen, and has been followed by the cardiology service ever since. She is now 29 years old (July 2006), she remains stable, but symptomatic. Her baby boy is now 2 years and 5 months old and is healthy. An echocardiogram done in December 2005 still reveals severe pulmonary hypertension, with an estimated peak systolic pulmonary artery pressure of 84 mm Hg.

Discussion

Prior reports described maternal mortality associated with pregnancy in idiopathic pulmonary hypertension as high as 50% (12). More recent reports describe a maternal mortality as low as 23% (3). Also, there have been reports of regression of pulmonary hypertension (13-15) and improvement of patient's symptoms during pregnancy (16). Normal gestation represents a state of endogenous vasodilatation, and one study documented improvement of cardiac output and decrease in pulmonary hypertension during pregnancy in a patient with idiopathic pulmonary hypertension (4). In the case described in this report, the patient presented with severe pulmonary hypertension, which improved during pregnancy up to the point of questioning the initial diagnosis of pulmonary hypertension since she had no clinical evidence of this condition while pregnant. She had an uneventful pregnancy, complicated only by preterm labor. Also, there were no complications during delivery or postpartum period, being this time the period of highest risk for death in the pregnant patient with pulmonary hypertension (17). Less than one year later, the patient again developed severe pulmonary hypertension. These findings suggest regression of pulmonary hypertension during pregnancy. Unfortunately, there were no definitive studies to document status of pulmonary hypertension just prior to pregnancy, so no assessment can be made about improvement of pulmonary hypertension in relation to the gravid state.

This report adds to the amount of evidence that suggests that pregnancy and labor in a patient with idiopathic pulmonary hypertension may have a better outcome than previously reported. A relatively recent review reports that under favorable circumstances and with adequate medical management the chances of maternal survival could be expected to increase in these patients (18). The decision of undertaking and/or continuing pregnancy in a patient with idiopathic pulmonary hypertension relies ultimately on the patient's choice, but should be done on an individual basis after careful evaluation of the risks. Finally, the need of close follow up with a multidisciplinary team is mandatory in the patient with idiopathic pulmonary hypertension that wishes to undergo pregnancy.

Resumen

Estudios previos sugieren una alta mortalidad asociada al embarazo en pacientes con hipertensión pulmonar idiopática, y la recomendación ha sido evitar y/o terminar el embarazo. Sin embargo, ha habido estudios que informan pacientes con esta condición que han tenido el embarazo y parto sin grandes complicaciones. Este informe ilustra el curso de la preñez y parto en una joven con hipertensión pulmonar idiopática. A los 24 años la paciente fue diagnosticada con hipertensión pulmonar idiopática moderadamente severa. Un año y 8 meses después la paciente sostiene un embarazo. En este momento la paciente estaba asintomática, y no tenía ninguna evidencia clínica de hipertensión pulmonar. Esta tuvo un embarazo sin complicaciones, y dio a luz prematuramente por vía vaginal a un niño saludable. No se utilizaron medidas invasivas para seguimiento en el parto. Madre e hijo fueron dados de alta 48 horas posterior al parto. Siete meses más tarde la paciente regresa con hipertensión pulmonar severa. La paciente actualmente continúa su seguimiento con un cardiologo, y se encuentra estable pero sintomática. Este estudio añade a la evidencia actualmente disponible que sugiere mejores resultados y sobrevida de lo antes informado en pacientes con hipertensión pulmonar idiopática que deseen llevar a término un embarazo y parto. Finalmente, el seguimiento frecuente y multidisciplinario es mandatario para paciente con hipertensión pulmonar idiopática que desee sostener un embarazo.

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