PALLIATIVE CARE CASE STUDY

A Case of Pulmonary Arterial Hypertension: If Only!

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The patient presented originally in 2010 to a pulmonary hypertension (PH) clinic with a complicated medical history. She was a 36-year-old female with a history of pyruvate kinase deficiency resulting in chronic hemolytic anemia since birth. She has had chronic blood transfusions, a splenectomy, and history of iron overload, for which she was treated with iron chelation. She has a history of hepatitis C from a blood transfusion but her viral load is negative. Further, she is paraparetic and wheelchair-bound due to a spinal cord infection at birth. Despite this, she had lived an active life, living independently and maintaining her household including her husband, 3 children, and a small farm with little assistance. In 2010, she developed edema and dyspnea; an echocardiogram was concerning for PH. Her initial right heart catheterization revealed a mean pulmonary artery pressure of 42 and a pulmonary vascular resistance of 6 Wood units. She was initially thought to have idiopathic pulmonary arterial hypertension and was started on sildenafil with improvement in exercise capacity.

She was lost to follow-up for insurance reasons but returned nearly 3 years later feeling worse. At that time she had several admissions with ascites and volume overload and was transferred from an outside hospital. She was also requiring more frequent transfusions to maintain a hemoglobin of 7 g/dL. She had trouble propelling her wheelchair up inclines and had a recent syncopal event. A repeat right heart catheterization showed a right atrial pressure at 16, a mean pulmonary artery pressure of 58, a pulmonary vascular resistance of 10.4 Wood units, and a pulmonary artery saturation of 40% with a hemoglobin of 6.9 g/dL. Echocardiogram showed a large right atrium, right ventricular (RV) diastolic diameter of 5.1 cm, and a tricuspid annular plane systolic excursion of 1.6 cm. N-terminal proBNP was 1754. She was markedly jaundiced with a bilirubin of 11.6. A transjugular liver biopsy showed iron-overload cirrhosis. She required supplemental oxygen, had clubbing, and a bubble echocardiogram showed intrapulmonary shunting. She was now diagnosed with portopulmonary hypertension, hepatopulmonary syndrome, and iron-overload cirrhosis in the setting of chronic hemolytic anemia and paraparesis. She was treated with intravenous (IV) inotropes, diuresed aggressively, and begun on IV prostacyclin infusion and an endothelin receptor antagonist (ERA) in addition to the phosphodiesterase type 5 (PDE-5) inhibitor. She improved markedly on this regimen and was discharged from the hospital after a 14-day stay.

Approximately 2 months later she was readmitted with septic shock from spontaneous bacterial peritonitis, decompensated RV failure, and hypoxemic respiratory failure. She was treated with antibiotics, inotropes, and vasopressors; high-dose garlic was begun for hepatopulmonary syndrome. She improved and was discharged after a 1-month stay.

Four months later, she was readmitted for volume overload and decompensated RV failure for approximately 14 days. On each of these admissions she required inotropic support and aggressive diuresis. Her prostacyclin infusion was further increased and midodrine was added for blood pressure support once vasopressors/inotropes were weaned. It appeared that each time she decompensated when her hemoglobin drifted below 8.5 g/dL and a higher transfusion goal was adopted. Further uptitration of prostacyclin was limited by chronic hypotension. An extensive evaluation for transplant possibilities was pursued. It was determined that bone marrow transplant for pyruvate kinase deficiency could not be done because of her cirrhosis and PH; lung transplant could not be concurrently done because of her paraparesis, chronic transfusions, and donor-specific antibody load; and liver transplantation could not be done because of her degree of PH. Referrals around the country to assess whether any center would offer liver, lung, and bone marrow transplant were denied.

Palliative care was discussed, but the patient reported that between admissions she was still able to take care of her children and her farm and was happy with her quality of life. Subsequently, she has been admitted approximately every 1 to 3 months for various issues, usually decompensated RV failure. Her renal function then began to decline. A home dopamine infusion was started at 5 mcg/kg/min, which improved her renal function and blood pressure and she was maintained for 6 months without hospitalization.

She then developed hemoptysis (small amounts of blood) and was readmitted. Shortly thereafter, she had 500 mL of hemoptysis and was emergently intubated. Her family was notified that she would likely not survive this insult but wanted to continue aggressive care at

this point. The patient went to interventional radiology and was found to have extensive collateralization and a massively dilated right internal mammary artery. She underwent right bronchial artery embolization and the hemoptysis resolved. She was extubated and discharged to home after a 1-month stay in the hospital.

In the subsequent year palliative care has continued to follow the patient. She returns to the hospital every 1 to 2 months with decompensated RV failure, volume overload, and renal dysfunction. In the hospital with vasopressors and diuretics her BUN will fall to 40 and creatinine to 1 (decreased muscle mass) but will slowly rise to a BUN of 130 and creatinine of 4 as an outpatient. She is now DNR/DNI but is still happy with her quality of life between admissions. She requires an extensive diuretic regimen to remain out of the hospital for 1 to 2 months, and at her directive this is continued despite loss of renal function. Because she intermittently develops sufficient ascites that it interferes with her ability to transfer from the wheelchair independently and her quality of life, a palliative PleurX catheter to drain the ascites has been offered, but given the tenuous renal function, hypotension, and infection risk, she understands that such an intervention would likely precipitate further renal failure and may shorten her life. She has chosen to keep returning to the hospital as long as possible. She now has developed cardiac cachexia. She remains on continuous dopamine, prostacyclin infusion, ERA, PDE-5 inhibitor, chelation, midodrine, and monthly transfusions. It has been over 7 years since the initial diagnosis.