Pulmonary Hypertension in a Premature Infant With Bronchopulmonary Dysplasia

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Pulmonary hypertension, premature infant, bronchopulmonary dysplasia

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Conflicts of interest: None to report.

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An almost 9-month-old former 23-week gestation preterm male infant, who was living at home with administration of 0.75 L of oxygen for bronchopulmonary dysplasia (BPD), presented to the high-risk neonatal clinic for routine follow-up. He was tachypneic and displayed moderate subcostal retractions, an increase from his baseline mild retractions. Crackles were audible throughout bilateral lung fields, with decreased air entry over the left lower lobe. His lips were dusky, and a pulse oximeter check revealed an oxygen saturation of 55%. The patient had a history of pulmonary hypertension (PH), with a recent echocardiogram 3 weeks ago demonstrating elevated but stable pulmonary pressures at 40 to 44 mm Hg. Current medications included spironolactone-hydrochlorothiazide, sildenafil, lansoprazole, and budesonide nebulizer treatments twice daily, albuterol sulfate nebulizer treatments three times daily, and furosemide as needed. The most recent dose of furosemide was given 2 weeks prior to this visit as a result of tachypnea and heightened work of breathing.

Because of the patient's low saturation levels and labored breathing, his oxygen was quickly increased to 2 L per minute (LPM) to achieve an oxygen saturation of 94%. An albuterol nebulizer treatment was administered, without noted improvement, and a stat echocardiogram was performed. This limited study revealed severe PH, with a pulmonary pressure of 70 mm Hg, in addition to septal wall flattening and pulmonary artery dilation. At this point, the patient was transferred to the local tertiary hospital for pediatric intensive care unit (PICU) admission.

CASE PRESENTATION

The infant was born at 23 weeks gestational age with a birth weight of 470 g to a gravida one para one

mother via emergency cesarean section because of a preterm premature rupture of membranes (PPROM) and prolapsed umbilical cord. At birth, his Apgar scores were 1, 4, and 4 at 1, 5, and 10 minutes, respectively. He was immediately intubated and given surfactant, and he required mechanical ventilation from that point until he was 152 days old. During that time, he frequently switched between conventional and high-frequency ventilators because of a pulmonary hemorrhage, pulmonary interstitial emphysema, BPD, and two episodes of ventilator-associated pneumonia. His initial echocardiogram on day 3 of life only revealed a small patent ductus arteriosus; however, by his third week of life, the presence of PH was confirmed.

The infant was started on inhaled nitric oxide (iNO) for 3 days to treat the PH. Upon improvement, the iNO was discontinued. It was restarted twice before he was successfully extubated. To wean the patient from iNO, sildenafil therapy was initiated at 0.5 mg/kg/dose twice per day to prevent rebound hypertension. He required three courses of dexamethasone, following the Dexamethasone: A Randomized Trial protocol, to be successfully extubated. According to research conducted by Doyle, Davis, Morley, McPhee, and Carlin (2006), low-dose dexamethasone therapy has been shown to improve oxygenation and ventilation and assist with earlier extubation among premature infants who either weigh less than 1,000 g at birth or are born at a gestational age of less than 28 weeks and who require ventilatory support beyond the first week of life. This patient also experienced significant retinopathy of prematurity and required bilateral bevacizumab therapy to prevent retinal detachment. During his neonatal intensive care unit (NICU) course, multiple head ultrasounds revealed only a left grade I intraventricular hemorrhage; no periventricular leukomalacia was visualized on magnetic resonance imaging.

When this patient was discharged home at 51 weeks corrected age, he required 0.5 LPM oxygen and was eating the majority of his enteral feedings by mouth, with supplementation of his 26 Kcal/oz fortified breast milk given via nasogastric tube as needed. During the next 2 months, this infant had increasing difficulty taking his feedings orally. He began arching upon feeding, became more irritable, and refused to take more than one third of the volume orally, sounding "wet in his nose" after his feedings. Lansoprazole therapy was initiated for presumed gastroesophageal reflux disease, which led to some improvement in symptomatology, but his oral intake did not increase. His weight for adjusted age improved, from the 4th to the 14th percentiles, with his adjusted head circumference hovering between the 1st and 2nd percentiles. His baseline oxygen requirement began to increase from 0.5 LPM, at NICU discharge, up to 0.75 LPM in the daytime and 0.85 LPM at nighttime, by the time of his presentation at the clinic.

Given his acutely increased oxygen requirement and the echocardiogram documenting severe PH, the patient was admitted to the PICU. Upon admission, a chest radiograph revealed lucency in the left hemithorax, consistent with a small to moderate-sized pneumothorax. Extensive coarse lung markings were present, compatible with chronic lung disease changes, along with left lower lobe lung consolidation. To improve both the hypoxia related to the pneumothorax and worsening PH, high-flow oxygen, with a fraction of inspired oxygen (Fio_2) content of 100%, was administered at 8 LPM for 1 day before the patient was titrated down to 1 LPM prior to hospital discharge 5 days later. Chest tube placement was ultimately unnecessary for pneumothorax resolution.

During his hospitalization, this infant received a one-time furosemide dose in addition to his usual medication regimen that included sildenafil, still administered at 0.5 mg/kg/dose twice daily. All feedings were given via a nasogastric tube and oral feeding attempts were held, related to microaspiration concerns, until his condition stabilized and an outpatient modified barium swallow study with upper gastrointestinal series could be completed.

Once he was home, his mother noted that the infant had increased alertness, activity, and endurance for in-home therapies when not attempting oral feedings. The patient underwent a battery of testing and specialist consultations to determine the best plan of care, given his chronic lung disease and concomitant PH. Repeat echocardiography demonstrated improved PH, compared with the study done at the time of PICU admission, but showed that pulmonary pressures remained moderately elevated, in the 50-59 mm Hg range. His oxygen requirement continued to rise, ranging between 1 to 1.5 LPM. A PH specialist was consulted, who tripled his sildenafil dose to 1 mg/kg/dose three times per day, with strict instructions to maintain oxygen saturations at 95% or higher. The modified barium swallow study with upper gastrointestinal series revealed silent intermittent microaspiration of all thickness liquids, but not purees, and normal gastrointestinal anatomy. A multichannel pH impedance study showed minimal reflux overall, with just one episode of increased severity in conjunction with emesis.

After acquiring second opinions from pulmonary and gastrointestinal specialists, the parents elected to have a gastrostomy tube surgically placed, without a Nissen fundoplication. The surgery was performed with assistance from the cardiac anesthesia team, and the patient tolerated the procedure well. He was extubated after surgery but then experienced an acute oxygen desaturation down to the 30% range, requiring 12 LPM of oxygen via high-flow nasal cannula to