Childhood Idiopathic Pulmonary Hypertension: a Case Report

Nathaniel J. Balmert1 and Joseph A. Prahlow, M.D.1

1 Western Michigan University Homer Stryker M.D. School of Medicine, Department of Pathology

Introduction
Pulmonary arterial hypertension (PAH) is characterized as average pulmonary artery pressure of greater than 25mm Hg at rest (normal =14 mm Hg). Childhood idiopathic pulmonary arterial hypertension (CIPH) is distinguishable from PAH by the absence of family history or underlying disease. Symptoms of CIPH include dyspnea, exercise limitation, and syncope. Common organ-specific changes include right ventricular hypertrophy and right ventricular strain. Diagnosis of the disease is poor with one, three, and five year survival rates of 68%, 48%, and 35%, respectively.1

In this report, we present the case of a six-year-old girl with idiopathic pulmonary arterial hypertension who died following an exacerbation of her disease, aggravated by inadvertent esophageal intubation.

Case Report
Clinical Presentation
A six-year-old girl was first conclusively diagnosed with pulmonary arterial hypertension approximately 3 years after she exhibited the first signs of any abnormality, which encompassed syncopal episodes on exertion. During the interim, she progressively developed right heart failure. Shortly following the diagnosis, she was readmitted due to seizures secondary to pulmonary hypertensive crises and hypoxia. She was eventually released to home on oxygen therapy.

Two months later, the child’s mother found her daughter seizing and called Emergency Medical Services (EMS). On initial exam, the child was found with a heart rate of 40 bpm and labored respirations, at which time chest compressions were initiated, and respirations were assisted using a bag-valve-mask (BVM) airway. During transport to the emergency department (ED), a second EMS unit met the ambulance en route, and, despite adequate ventilation via the BVM, intubation was attempted. The first attempt was unsuccessful, but a second attempt was reportedly successful. Transport continued to the ED, where resuscitation attempts continued. Following the second intubation attempt in the field, lung sounds were reported as present and equal with one, three, and five year survival rates of 68%, 48%, and 35%, respectively.

An additional gross autopsy finding was the identification of an esophageal intubation (Figure 4). The endotracheal tube was found in the patient’s esophagus with the cuff inflated (Fig. 5 & 6) and blanched underlying mucosa (Figure 7).

A six-year-old girl with idiopathic pulmonary hypertension is shown. The heart displays marked right ventricular dilation and as present and equal

Figure 11: A micrograph after anterior chest plate removal, showing the enormous heart. Also seen is the right lung (photograph). Figure 9: Microscopic stained pulmonary artery branch within the lung, highlighting the concentric medial fibrosis (blue) (trichrome stain). Figure 10: Autopsy photograph after anterior chest plate removal, showing the enormous heart (photograph). Figure 8: Microscopic appearance of pulmonary artery branch within the lung, showing medial cellular hypertrophy and wall thinning (hematoxylin & eosin, original magnification 20X).

Toxicology tests showed therapeutic levels of diazepam (Figure 1).

Final Diagnosis
Given the autopsy findings and the history as presented, the cause of death was ruled as primary pulmonary arterial hypertension with a contributing factor of esophageal intubation.

References