End-Stage Cystic Fibrosis in Pregnancy

Abstract Type: Case Report/Case Series
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Introduction: Cystic fibrosis (CF) is an autosomal recessive disease of exocrine glands that results in progressive obstructive pulmonary disease, pancreatic exocrine deficiency and abnormally high sweat electrolyte concentrations. The underlying mechanism in this disease is a defect in cAMP-mediated activation of chloride conductance in epithelial cells required to regulate the components of mucus, sweat, and digestion.

Case Report: A 19yr old G1P0 at 19 weeks gestation with end stage cystic fibrosis was admitted to labor and delivery in respiratory distress. Prior to pregnancy, she was under consideration for a double lung transplant. Her pre-pregnancy FEV-1 was 20% of predicted. Baseline CO2 values ranged between 60-80 mmHg. She was BiPAP dependent at home, with settings of PS 18, PEEP of 10, and FiO2 of 70%. On admission, her initial arterial blood gas on a nonrebreather mask was pH 7.30, PaCO2 89, PaO2 178, HCO3 43, Base Excess +16, and SaO2 99.

The patient was admitted to our medical intensive care unit, where she remained on BiPAP and multiple antibiotic regimens for chronic bacterial pneumonia. In preparation for delivery, multidisciplinary meetings were held. A decision was made for an assisted vaginal delivery at 28 weeks under neuraxial analgesia. An intrathecal catheter was selected for reliability and careful titration. However, at 27 and 5/7 weeks, she was transferred to the labor intensive care room for induction of labor secondary to further respiratory decompensation. An intrathecal catheter was placed at interspace L3-L4 under sterile conditions in the sitting position for early labor pain. Analgesia was achieved with 0.8 ml of 0.125% bupivacaine followed by a continuous infusion of 0.1% bupivacaine at 1ml/hr. Secondary to increasing vaginal edema and non-reassuring fetal heart tracings, she was taken urgently for cesarean section (CS). Surgical anesthesia was achieved with her indwelling intrathecal catheter. The patient tolerated the CS on BiPAP in a 30° head up position without incurring further respiratory compromise. Although she remained stable for over a week in the ICU, she suffered acute respiratory failure and septic shock on POD 11 and was unable to be resuscitated.

Discussion: Cystic fibrosis is a lethal genetic disease. Due to dramatic advancements in diagnosis and treatment, an increasing number of women are surviving into their reproductive years. This has led to a subsequent rise in pregnancy in these patients. Today, an understanding of the disease and its vast anesthetic implications in pregnancy is imperative.

References: