Case Report: Anesthetic Management of a Patient with Hepatopulmonary Syndrome, Prolonged QTc Interval and Panhypopituitarism for Liver Transplantation

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Introduction: Hepatopulmonary syndrome (HPS) is the triad of liver disease, arterial deoxygenation while breathing room air, and evidence of intrapulmonary vascular dilation. Liver transplantation patients with HPS have an increased risk of perioperative morbidity and mortality. There are no clear criteria established for which of these patients should proceed to transplantation. We present a unique case of successful orthotopic liver transplantation (OLT) in a patient with end stage liver disease, HPS, and QTc prolongation but also complicated by panhypopituitarism secondary to a benign suprasellar mass.

Case Report: A prepubertal 16-year-old, 81 kg (body mass index of 26.6 kg/m$^2$) male presented with orthopnea, dyspnea on exertion, and cyanosis. He had clear lung fields, an unremarkable precordium, significant hepatomegaly, mild splenomegaly and mild digital clubbing. Oxygen saturations ranged from as low as 60% on room air to 94% on 3LNC O$_2$. His past medical history was notable for a history of diabetes insipidus (DI) treated intermittently with intranasal desmopressin, asthma and depression. Work-up revealed cryptogenic cirrhosis, HPS, benign suprasellar mass resulting in panhypopituitarism and long QT interval (QTc 490 ms). The QT prolongation was attributed to advanced hepatic disease. He was placed on 2 to 5 liters of oxygen to maintain his saturations above 90%. His endocrinopathies were treated with thyroxine, growth hormone, corticosteroid replacement, testosterone and desmopressin.

Seven months after initial presentation, the patient presented for OLT. The perioperative management included taking his standard desmopressin and L-thyroxine at home preadmission, adding stress dose steroids and avoiding drugs that have a risk of causing Torsades de Pointes. The anesthetic was induced with midazolam, fentanyl, thiopental, and succinylcholine. The anesthetic was maintained with fentanyl, midazolam, pancuronium, and isoflurane in 60% to 100% O$_2$. After induction, saturations were 91% on 100% O$_2$ with end tidal CO$_2$ (ETCO$_2$) as high as 66 mmHg. Albuterol (metered dose inhaler) was given and ventilation parameters were given such that the saturations for the remainder of the case were between 98 – 100% and ETCO$_2$ ranged from 31 to 44 mmHg. The A-a gradient ranged from 467 down to 160 during the case. Infusions of low dose dopamine, epinephrine, glucose and insulin were started pre-reperfusion of the cadaveric liver to optimize cardiopulmonary, electrolyte and metabolic status.

Additional intraoperative medications included calcium, magnesium and sodium bicarbonate. Blood loss was estimated to be approximately half of the patient’s total blood volume. Fluid replacement included 6 units of packed red blood cells, 720 ml of cell saver blood, 7 units of fresh frozen plasma, 500cc of 5% albumin, and 8 liters of crystalloid. Because of the potential competing issues of massive fluid shifts and blood loss from the OLT with the potential for hyponatremia from vasopressin for the treatment of DI, intraoperative vasopressin was avoided until sodium levels (and plasma osmolality) during the case rose from an induction level of 146 mmol/l (292 mosm/kg) to a high of 155 mmol/l (322 mosm/kg) approximately 2 hours post-reperfusion of the cadaveric liver. Vasopressin was then started at 40 units/hour such that at the termination of the anesthetic 1.5 hours later the sodium level decreased to 150 mmol/l (318 mosm/kg). Urine output for the 8.5-hour anesthetic case was 2100 ml (160 ml of urine output during the 1.5 hours infusion of vasopressin). Immediate postoperative course was uncomplicated with the QTc interval returning to normal and resolution of the hypoxemia over several weeks. The patient was discharged on the twelfth postoperative day.

Discussion: This case combines the rare findings of HPS, panhypopituitarism and prolonged QTc interval in the setting of advanced hepatic disease, all of which compound the potential for perioperative instability. Severity of hypoxemia in HPS increases perioperative transplant mortality. However, there
are no effective medical treatments and mortality is 50% at 41 months post diagnosis of HPS. Additionally, in the setting of liver disease, patients with prolongation of the QTc had a decreased survival rate over a mean follow-up of 19 months. DI further complicates the perioperative management because of the potential for free water shifts complicated by either hypernatremia or hyponatremia in the setting of a liver transplant with large fluid and blood product requirements. Although in our experience, the sodium level in liver transplant patients tend to trend to mild hypernatremia, we decided that the best approach was to follow the sodium level, urine output, and osmolality and intervene when necessary. Despite this patient’s complex, multiple medical issues including his moderate to severe HPS, the decision was to proceed with OLT given the patient’s poor prognosis otherwise. As supported in the literature, both the patient’s HPS and prolonged QTc resolved post-transplant.

**Conclusion:** HPS remains a significant clinical challenge posing many medical and ethical issues. There are no clear criteria for which patients with HPS should proceed to OLT nor is there a good understanding on what factors increase perioperative risk. Until more defined criteria or viable medical alternatives become available, OLT with HPS is an acceptable option, even with moderate to severe HPS and confounding medical issues of prolonged QTc interval and panhypopituitarism.

**References:**