Spinal Anesthesia for Cesarean Section in a Parturient with Noonan Syndrome and Von Willebrand Disease

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Introduction: Noonan's syndrome is an uncommon congenital disorder which has multiple anesthetic considerations. This case describes the anesthetic management of a parturient with Noonan's syndrome and Von Willebrand disease undergoing cesarean section under spinal anesthesia.

Case: A 27 year-old primigravida parturient at 35+3 weeks gestation underwent cesarean section due to marginal placenta previa and vaginal bleeding. Her past medical history was significant for Noonan's syndrome, asthma, mild pulmonic stenosis, non-obstructive hypertrophic cardiomyopathy with an ejection fraction of 59%, and Von Willebrand disease. Her labs included a platelet count of 96,000, normal hemoglobin, and normal clotting factor levels and activity. A collagen/epinephrine closure time was normal which is consistent with normal platelet function. Physical exam revealed a 58 inch, 116 lb, female that displayed features consistent with Noonan's syndrome including a webbed neck, flattening of the midface, low-set ears, shield shaped chest, chronic holosystolic murmur, and mild kyphoscoliosis. The patient was premedicated with sodium citrate and prehydration with 500 ml of Lactated Ringer's. Immediately prior to spinal placement a phenylephrine infusion was started. A subarachnoid block was performed without difficulty and a T4 level of anesthesia was achieved. The patient remained comfortable and hemodynamically stable throughout the procedure. She had an estimated blood loss of 0.5 liters, and received 1.1 liters of crystalloid. On post-op day one the patient developed one episode of shortness of breath which responded positively to albuterol treatment. No other complications were noted in the perioperative period.

Discussion: Noonan's syndrome is an uncommon congenital disorder characterized by facial, cardiovascular, and skeletal abnormalities(1). Anesthesia providers should be aware of the potential for a difficult airway, difficult placement of regional anesthesia due to spinal deformities, the risk for malignant hyperthermia(2), limited cardiorespiratory reserve, risk of cephalopelvic disproportion and need for c-section, and an increased risk of coagulation disorders(1). In this parturient with Noonan's syndrome and Von Willebrand's disease we felt that a regional anesthetic was the optimal approach given her preoperative coagulation, echocardiographic, and physical findings. Few cases in the literature describe regional anesthesia for Cesarean section in parturients with Noonan's syndrome. However, adequate pre-operative assessment and laboratory testing can guide the anesthesiologist in choosing a safe anesthetic plan for parturients with Noonan's syndrome which may include regional anesthesia.

References: