Abstract # 252

Anesthetic Management of a Parturient with Noonan Syndrome and Intra-operative Arrhythmia

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Case Presentation: A 21 year-old primigravida with Noonan's syndrome presented at 39 weeks in active labor. She had a class 2 airway with a short webbed neck, micrognathia, scoliosis and a mild developmental delay with anxiety disorder. As a child, she underwent pulmonary angioplasty and echocardiography revealed mild pulmonary valve stenosis with mild pulmonary hypertension.

Although technically challenging, epidural catheter placement was attempted. After 8 hours in the first stage of labor, fetal decelerations developed and a cesarean section was needed. Titrated dosing of the epidural was used to provide surgical anesthesia and the patient remained stable until a healthy baby was delivered. Immediately after delivery, the patient's developed supraventricular tachycardia which was treated with β-blockers and sedation.

Discussion: Pulmonary stenosis is common in patients with Noonan syndrome resulting in increasing right ventricular afterload. Excessive IV fluids, prior to epidural block, could lead to right ventricular failure. On the other hand, if the patient is not adequately hydrated, the hypotension resulting from the sympathetic block may lead to decrease in venous return and cardiac output. Our patient had a mild degree of pulmonary stenosis and a preserved cardiac function, so epidural technique was successfully utilized. But with severe lesions, general anesthesia with invasive monitoring is warranted.

Difficult airway equipment was available in case emergency intubation became necessary. The physiologic changes of pregnancy, in addition to the associated anatomical airway abnormalities, increase the risk of a difficult intubation.

Coagulation profile was checked prior to epidural placement and was normal. Bleeding diatheses including coagulation factor deficiencies and abnormal platelet count have been described which may contraindicate regional anesthesia. Varying degrees of mental impairment may present a challenge from consent to cooperation with procedures, although most individuals with Noonan syndrome are of normal intelligence. Finally, technical difficulties with neuraxial placement may be experienced due to exaggerated lumbar lordosis, short stature, and a narrow spinal canal.

To summarize, parturients with Noonan syndrome can present with an array of anomalies that may present difficulties to the anesthesiologist including impairment of cardiopulmonary functions, possibly difficult airway, bleeding disorders, and a technically difficult regional anesthesia. These anomalies may be further exacerbated by the normal physiologic changes of pregnancy. Careful preoperative assessment and determination of the associated anomalies can help preparation for potential problems.