

General Anesthesia for Cesarean Section in a Patient with Noonan Syndrome

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Abstract

Introduction: Noonan syndrome is a rare congenital disorder characterized by abnormalities of the craniofacial, cardiovascular, and skeletal systems. Parturients with Noonan syndrome present potential significant challenges to anesthesia providers - namely difficult tracheal intubation, limited cardiorespiratory reserves, and technical problems in performing regional anesthesia due to short stature and skeletal anomalies¹. This case report illustrates the perioperative management of a parturient with Noonan syndrome undergoing general anesthesia for cesarean section.

Case: A 19-year-old term G₂P₁ patient with Noonan syndrome presented for cesarean section secondary to cephalopelvic disproportion. The patient exhibited characteristic features of Noonan syndrome i.e., short stature, webbed neck, pectus excavatum, and significant kyphoscoliosis. Despite kyphoscoliosis, she was without symptoms of restrictive pulmonary disease or other pulmonary problems. Cardiac evaluation revealed sinus tachycardia and right atrial enlargement. The patient, however, was NYHA Class I. Hematology studies and thromboelastography were normal. Airway examination was remarkable for limited mouth opening, Mallampati IV classification, protruding incisors, a high, arched palate, and thyromental distance <6.5cm.

Preparations were made for safe performance of general anesthesia that included awake intubation since the patient's examination indicated a potentially difficult airway. Regional anesthesia was excluded because of anticipated technical problems related to the patient's kyphoscoliosis. Adequate intravenous access was established. Standard ASA and left radial arterial invasive monitoring were applied. The airway was anesthetized via local anesthetic nebulizers and blockade of the glossopharyngeal, superior laryngeal, and recurrent laryngeal nerves. Oxygen was administered via nasal cannula during topicalization. Tracheal intubation was successfully achieved via awake fiberoptic technique and cesarean section proceeded uneventfully.

Discussion: Noonan syndrome is a rare disorder occurring in approximately 1 in 1000-2500 individuals. Inheritance is usually autosomal dominant, although many cases occur because of mutations involving the *PTPN11* and *KRAS* genes. Diagnosis is made mostly on clinical grounds¹. Characteristic features include short stature; ocular hypertelorism, low nasal bridge, micrognathia; webbed neck; abnormal chest shape; congenital heart defect; abnormal spine curvature; and developmental delay. Varied coagulation defects and lymphatic dysplasias are frequently observed. Congenital heart defects -namely pulmonary valve stenosis and/or hypertrophic cardiomyopathy- are found in 20 to 50% of individuals². The provision of anesthesia for parturients with Noonan syndrome requires extensive preparation, effective communication amongst caregivers, and early assessment with special attention to the airway, cardiorespiratory, and musculoskeletal systems. Depending upon the patient's condition, it may be possible to administer either regional or general anesthesia. In this case, an anesthetic plan of care was carefully formulated and executed as described previously after thorough overall evaluation and risks-to-benefits discussion with the patient, her family, and the obstetrics team.

References:
1. Gambling & Douglas. *Obstetric Anesthesia & Uncommon Disorders*. Pp427-9
2. Tartaglia M, Gelb. "Noonan syndrome and related disorders: genetics/pathogenesis." *Annu Rev Genomics Hum Genet*. 2005; 6:45-68.

Case Presentation

A 19-year-old term G₂P₁ patient with Noonan syndrome presented for cesarean section secondary to cephalopelvic disproportion. The patient exhibited characteristic features of Noonan syndrome i.e., short stature, webbed neck, pectus excavatum, and significant kyphoscoliosis. Despite kyphoscoliosis, she was without symptoms of restrictive pulmonary disease or other pulmonary problems.



The photos above illustrate the short extremities of the patient of interest. Short stature and short extremities are characteristic of individuals affected by Noonan Syndrome.

Cardiac evaluation revealed sinus tachycardia and right atrial enlargement. The patient, however, was NYHA Class I. Hematology studies and thromboelastography were normal.

Airway examination was remarkable for limited mouth opening, Mallampati IV classification, protruding incisors, a high, arched palate, and thyromental distance <6.5cm.



The photo above illustrates the airway examination of the patient of interest. It is remarkable for a Mallampati IV classification and limited mouth opening.

Preparations were made for safe performance of general anesthesia that included awake intubation since the patient's examination indicated a potentially difficult airway. Regional anesthesia was excluded because of anticipated technical problems related to the patient's kyphoscoliosis. Adequate intravenous access was established. Standard ASA and left radial arterial invasive monitoring were applied. The airway was anesthetized via local anesthetic nebulizers and blockade of the glossopharyngeal, superior laryngeal, and recurrent laryngeal nerves. Oxygen was administered via nasal cannula during topicalization. Tracheal intubation was successfully achieved via awake fiberoptic technique and cesarean section proceeded uneventfully.

Discussion

Noonan syndrome is a rare disorder occurring in approximately 1 in 1000-2500 individuals. Inheritance is usually autosomal dominant, although many cases occur because of mutations involving the *PTPN11* and *KRAS* genes. Diagnosis is made mostly on clinical grounds¹.

Characteristic features include short stature; ocular hypertelorism, low nasal bridge, micrognathia; webbed neck; abnormal chest shape; congenital heart defect; abnormal spine curvature; and developmental delay. Varied coagulation defects and lymphatic dysplasias are frequently observed. Congenital heart defects -namely pulmonary valve stenosis and/or hypertrophic cardiomyopathy- are found in 20 to 50% of individuals².

Technical difficulty may be encountered by those providing anesthesia to patients with Noonan syndrome. Specifically, the presence of a high-arched palate and small mandible may present difficulty in achieving tracheal intubation. Regional anesthesia may be difficult to perform as well due to short stature and musculoskeletal abnormalities such as lumbar lordosis and kyphoscoliosis¹. Even if the intrathecal space is engaged, the presence of a narrowed or otherwise abnormal spinal canal may affect anesthetic spread and lead to unreliable and unpredictable dosing and level of analgesia resulting in either high or total spinal anesthesia. Significant kyphoscoliosis and pectus deformities may further worsen the already decreased functional residual capacity of pregnant patients with Noonan syndrome. This further predisposes such patients to rapid hypoxemia. Additionally, pregnant patients with Noonan syndrome are at increased risk for operative delivery because of a contracted "male-" type pelvis and at increased risk for cephalopelvic disproportion.

Preoperative evaluation should be performed early and in a thorough manner in planning anesthetic management. Special efforts should be made to identify signs and symptoms of Noonan syndrome - especially as they may affect function of the cardiac, respiratory, renal, hematologic, and musculoskeletal systems.

Conclusion

The provision of anesthesia for parturients with Noonan syndrome requires extensive preparation, effective communication amongst caregivers, and early assessment with special attention to the airway, cardiorespiratory, and musculoskeletal systems. Depending upon the patient's condition, it may be possible to administer either regional or general anesthesia. In this case, an anesthetic plan of care was carefully formulated and executed as described previously after thorough overall evaluation and risks-to-benefits discussion with the patient, her family, and the obstetrics team.

References

1. Gambling & Douglas. *Obstetric Anesthesia & Uncommon Disorders*. Pp427-9
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