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RESEARCH ARTICLE

ANESTHETIC MANAGEMENT OF A CESAREAN SECTION IN A PATIENT WITH NOONAN SYNDROME: A MULTIDISCIPLINARY APPROACH

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Abstract

Background: Noonan syndrome (NS) is a genetic disorder associated with congenital heart defects, coagulation disorders, and skeletal abnormalities, posing significant challenges in anesthetic management, especially during pregnancy.

Case Presentation: We report the case of a 29-year-old primigravida with NS who was admitted for severe intrauterine growth restriction at 30 weeks of gestation. The patient had hypertrophic cardiomyopathy with previous surgical correction, requiring chronic anticoagulation. A scheduled cesarean section was planned due to fetal and maternal risks, with careful perioperative management including anticoagulation bridging, invasive monitoring, and advanced anesthetic techniques. General anesthesia was chosen due to severe scoliosis, anticoagulation status, and the risk of hemodynamic instability. The procedure was uneventful, and both the mother and neonate were stabilized postoperatively in intensive care units.

Discussion: This case highlights the anesthetic challenges in pregnant patients with NS, emphasizing the need for a multidisciplinary approach. The anesthetic strategy was tailored to the patient's cardiac condition, respiratory function, and coagulation status, aligning with existing literature on NS-related obstetric and anesthetic management.

Conclusion: Multidisciplinary coordination, thorough preoperative assessment, and advanced perioperative monitoring are crucial in ensuring optimal maternal and fetal outcomes in NS patients undergoing cesarean delivery.

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Introduction:-

Noonan syndrome (NS) is an autosomal dominant genetic disorder associated with various congenital anomalies, including cardiac malformations and coagulation disorders [1]. The anesthetic management of patients with NS is challenging due to the frequently associated cardiovascular abnormalities, such as pulmonary stenosis and hypertrophic cardiomyopathy.

In pregnant patients with NS, anesthetic management becomes even more complex due to physiological adaptations of pregnancy that can exacerbate underlying hemodynamic disturbances. We present a case of a parturient with Noonan syndrome, emphasizing the necessity of multidisciplinary planning in managing such cases.

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Case Report:
The patient was initially admitted for intrauterine growth restriction at 30 weeks of amenorrhea.

Family History :
1. Sister with Noonan syndrome.
2. Brother deceased from sudden cardiac death.
3. Parental consanguinity (first-degree relatives).

MedicalHistory :
Cardiac:
- Hypertrophic cardiomyopathy due to Noonan syndrome diagnosed at age 7, initially treated with beta-blockers.
- Development of obstructive hypertrophic cardiomyopathy with subaortic septal obstruction at age 19, discovered during echocardiographic follow-up.
- Surgical treatment with valve replacement using a mechanical prosthesis and subaortic resection.
- Chronic anticoagulation therapy with warfarin.

Pulmonary:
Severe restrictive and obstructive pulmonary disease, bronchiectasis, scoliosis with a history of Harrington rod placement.

Other:
Splenomegaly, gestationaldiabetes.

ClinicalExamination :
1. **General condition:** Good, NYHA class I dyspnea.
2. **Vital signs:** BP 113/61 mmHg, HR 80 bpm.
3. **Measurements:** Height 1.59 m, weight 50 kg.
4. **Facial dysmorphism:** Hypertelorism, trapezius hypertrophy, low-set ears, retrognathism.

Gynecological and ObstetricExamination :
1. Uterineheight: 25 cm.
2. No uterine contractions.
3. Positive fetalheartsounds.
4. Satisfactoryamnioticfluidquantity.
5. Severe intrauterine growth restriction on obstetric ultrasound.
6. Estimatedfetalweight: 925 grams.

Preoperative Assessment
LaboratoryFindings:
1. Prothrombin time (PT): 52%.
2. Urea: 0.45 g/L.
3. Creatinine: 5 mg/L.

Cardiovascular Examination:
1. Systolic ejection murmur related to hyperdynamic circulation.
2. Audible prosthetic valve click at the mitral area.
3. **ECG:** Regular sinus rhythm, HR 94 bpm, PR interval 140 ms, left axis deviation.
4. Complete intraventricular block with secondary repolarization abnormalities.

Transthoracic Echocardiography:
1. Type III hypertrophiccardiomyopathy (Maron classification), non-obstructive at rest.
2. Good left ventricular function (LVEF 62%).
3. 5 mm septal defect without visible shunt.
4. Double-leafletmechanical mitral prosthesis, non-regurgitant, non-stenotic.
5. Intermediate probability of pulmonary hypertension.

Preanesthetic Evaluation

- **ASA classification:** II.
- **Respiratory function:** NYHA class I dyspnea, MET > 4.
- **Difficult intubation criteria:**
  - Retrognathism with limited mouth opening.
  - Flexible neck.
  - Scoliosis confirmed by thoracic X-ray.
- **Pulmonary function testing:** Vital capacity at 38%.
- **Hematologicassessment:** PT 52%, INR 1.80.
- **Current medications:** Warfarin and atenolol.

Delivery Planning

Given the severe intrauterine growth restriction, underlying cardiopathy with risk of hemodynamic decompensation, and the need for close fetal monitoring, a **scheduled cesarean section at 30 weeks of amenorrhea** was decided.

- **PreoperativeOptimization:**
  - Warfarin discontinuation and bridging with low molecular weight heparin (LMWH) under strict biological surveillance.
  - LMWH cessation 12 hours before surgery.
  - Continuation of atenolol for heart rate control.
  - Close echocardiographic monitoring.
  - Blood request due to increased hemorrhagic risk.
  - Multidisciplinary consultation with obstetricians, cardiologists, and anesthesiologists.
- **ObstetricStrategy:**
  - Continuous fetal well-being evaluation via cardiotocography and ultrasound.
  - Administration of corticosteroids for fetal lung maturation in anticipation of preterm birth.
  - Neonatal team presence at delivery.
  - Possibility of neonatal intensive care admission due to estimated fetal weight of 925 g and severe prematurity risk.

A preoperative vascular access assessment was performed to anticipate the need for extracorporeal membrane oxygenation (ECMO) in case of maternal cardiac decompensation.

Anesthetic Management

- **Choice of Anesthesia:** General anesthesia was preferred over regional anesthesia due to:
  - Severeorthopnea.
  - Risk of hemodynamic decompensation due to systemic vascular resistance reduction.
  - Respiratory insufficiency risk due to scoliosis.
  - Chronicwarfarin anticoagulation.
- **AnestheticProcedure:**
  - ECG, HR, RR, SpO2, and NIBP monitoring.
  - Two 14G peripheral IV lines.
  - 4 Fr invasive arterial line placement.
  - Preoxygenation with 6 L/min O2 for 8 minutes until expiratory O2 fraction reached 93%.
  - Induction: Midazolam 0.15 mg/kg, lidocaine 1 mg/kg, propofol 2 mg/kg, rocuronium 0.6 mg/kg after ensuring mask ventilation feasibility.
  - Intubation via videolaryngoscopy without incident using a 6.5-mm endotracheal tube.
  - Ventilation: Volume Assist Control (VACi) mode, tidal volume 6 mL/kg, RR 12/min, FiO2 45%, PEEP 5 cmH2O.
  - Maintenance: Isoflurane (MAC 1 target).
  - Post-delivery: Fentanyl 2 µg/kg and oxytocin 15 IU administration.
  - Uneventfulsurgery.
  - Postoperative monitoring in PACU, extubation at H+1, admission to ICU.
  - Neonate admitted to neonatal intensive care due to low birth weight.

Postoperative Management

1. **ICU monitoring** due to risk of hemodynamic decompensation.
2. **Extubation at H+1** with close respiratory and hemodynamic monitoring.
3. **Strict anticoagulation control** before resuming warfarin.
4. **Echocardiographic surveillance** to assess cardiac function and detect complications.
5. **Neonatal intensive care follow-up** for the premature newborn.
6. **Favorable outcome**, with gradual discharge after stabilization of maternal and neonatal parameters.

Discussion:-

Noonan syndrome (NS) is an autosomal dominant genetic disorder with an estimated prevalence of 1 in 1000 to 1 in 2500 live births [1]. It is characterized by a distinctive facial appearance, short stature, skeletal abnormalities, and congenital heart defects, most commonly pulmonary stenosis and hypertrophic cardiomyopathy [2]. The anesthetic implications of NS are complex due to the associated cardiovascular, respiratory, and coagulation disorders, which can pose significant perioperative challenges.

Several case reports in the literature have described the challenges of performing a cesarean section in patients with NS, particularly in those with severe cardiac disease. The risk of perioperative cardiac decompensation is heightened in patients with hypertrophic cardiomyopathy, especially when associated with left ventricular outflow tract obstruction [3].

In our case, the patient presented with hypertrophic cardiomyopathy and subaortic obstruction, surgically treated with mechanical mitral valve replacement and anticoagulation therapy. This added complexity to the perioperative management due to the need for anticoagulation bridging and close coagulation monitoring. The decision to proceed with general anesthesia rather than regional anesthesia was based on the patient's severe cardiovascular involvement, the risks associated with hemodynamic fluctuations, and concerns about anticoagulation [4].

Advanced monitoring, including intraoperative echocardiography and invasive hemodynamic monitoring, was crucial in ensuring hemodynamic stability. Our multidisciplinary approach, involving obstetricians, cardiologists, anesthesiologists, and neonatologists, allowed for early anticipation and management of potential complications, ensuring both maternal and neonatal safety.

Compared to other cases in the literature, our management strategy was tailored to the patient's specific clinical profile, incorporating comprehensive risk assessment and optimization of both maternal and fetal outcomes. The use of advanced anesthetic and surgical techniques contributed to a favorable outcome without major complications [5].

Conclusion:-

This case highlights the necessity of a **multidisciplinary approach** for cesarean section in a patient with NS and severe cardiopulmonary involvement. **General anesthesia**, **advanced monitoring**, and **ECMO preparedness** allowed for a successful outcome.

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