

## Regional Anesthesia for Cesarean Section in a Parturient with Noonan's Syndrome

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Noonan's syndrome is a rare clinical entity. It is also known as female pseudo-Turner syndrome or Turner phenotype with normal chromosomes. These patients have facial, cardiovascular, and skeletal abnormalities which may pose a problem to the anesthesiologist during surgery. There are no reported cases of Noonan's syndrome requiring anesthesia for cesarean section.

## CASE REPORT

A 20-yr-old, full-term, primigravida woman was admitted in active labor. She was known to have Noonan's syndrome without evidence of cardiac disease, and had been followed throughout her pregnancy in prenatal clinic. Because of a grossly contracted pelvis, it was considered unlikely that she could deliver vaginally. She had no history of previous surgery, and her family history was non-contributory. Physical examination revealed an alert, cooperative female, whose height was 120 cm, weight 57 kg, arterial blood pressure 108/60 mmHg, heart rate 120 bpm, and respiratory rate 24/min. The typical findings of Noonan's syndrome that were present included flattening of the midface, downward slanting of the palpebral fissures, low-set ears, high palatal arch (without a visible uvula), webbing of the neck, and a "shield-shaped" chest. In addition, she exhibited two of the typical skeletal abnormalities, namely kyphoscoliosis and "spade-shaped" hands with deformity of the fifth metacarpal bone. Physical examination of the chest revealed accentuated first and second heart sounds and a grade II/IV systolic murmur at the lower left sternal border, findings which the cardiologist believed were consistent with cardiac displacement secondary to pregnancy. The pelvis was flat and contracted with a diagonal conjugate of less than 8 cm, resulting in cephalopelvic disproportion. Laboratory data were unremarkable, and the ECG and echocardiography were within normal limits. Because of the cephalo-pelvic disproportion, the patient had been scheduled for cesarean section.

The only premedication was sodium citrate 30 cc (0.3 M), which was given orally 30 min before the anticipated induction of anesthesia. Regional anesthesia was planned, but a fiberoptic laryngoscope was readily available in case emergency endotracheal intubation became necessary. Due to technical difficulty in locating the epidural space, subarachnoid block was performed atraumatically with a 22-gauge spinal needle inserted at the L<sub>3-4</sub> interspace with the patient in the right lateral position. Following the injection of 10 mg of hyperbaric

tetracaine, the patient was placed in the supine position with left uterine displacement, though positioning was difficult due to the patient's kyphoscoliosis and lumbar lordosis. A T<sub>4</sub> level of anesthesia was achieved initially, it slowly rose to T<sub>2</sub> intraoperatively; and, by the end of the surgical procedure, it had risen to T<sub>1</sub>. The systolic blood pressure decreased initially from 100 to 90 mmHg, but, through the aggressive intravenous administration of crystalloids (1350 ml) and two small, 10-mg doses of intravenous ephedrine, arterial blood pressure stabilized at just above 100 mmHg systolic and remained there throughout the procedure. The ECG showed a normal rate and rhythm during the entire procedure. Although ventilation was spontaneous and adequate throughout, oxygen was still administered *via* a mask until after the delivery of the baby. A normal-appearing female infant was delivered with an APGAR score of 7 and 8 at 1 and 5 min, respectively. At the completion of surgery, the blood loss was estimated to be 780 cc, and a total of 1850 ml of fluids had been administered. Vital signs at the end of the procedure were stable, and the subsequent hospital course of mother and infant was uneventful.

## DISCUSSION

Noonan's syndrome was first described by Noonan and Ehmke in 1963.<sup>1</sup> The principle clinical features<sup>2,3</sup> consist of flattening of the mid-face, hypertelorism, ptosis, downward slanting of the eyes, a high-arched palate, dental malocclusion, low set ears, webbing of the neck, a low posterior hairline, and a "shield-shaped" sternal deformity of the chest which gives the illusion of widely spaced nipples. In addition, most patients are of short stature, exhibit kyphoscoliosis, and have hyper-extensible joints with an increased carrying angle of the elbow (cubitus valgus). The most common congenital cardiac lesion is pulmonary stenosis, either alone or in combination with a septal defect, usually atrial. Both males and females may exhibit Noonan's syndrome, but males have cryptorchidism and are rarely fertile, while females are fertile, though they have a delayed menarche. Renal anomalies can occur, and the skin may be prone to keloid formation. Intellectual development is extremely variable, with intelligence ranging from superior to retarded; but the majority of patients with Noonan's syndrome show mild or borderline mental retardation. The inheritance may be familial,<sup>4</sup> autosomal dominant,<sup>5,6</sup> or sporadic,<sup>3,7</sup> but no chromosomal abnormalities have been found. The familial cases show a wider range of clinical expression than the sporadic cases. The variation of clinical features in the familial cases suggests that the spectrum of Noonan's syndrome may be much wider than is currently believed.

While it is important for anesthesiologists to be aware

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that both Turner's and Noonan's syndromes have associated cardiac disease, it is equally important to differentiate between the two, because each usually presents with different types of cardiac lesions. The clinical features common to both syndromes are the short stature, the skeletal anomalies, the webbed neck, and the facial characteristics that include epicanthal folds, ptosis, and prominent low-set ears. What differentiates most patients with Noonan's syndrome from those with Turner's syndrome is the fact that patients with Turner's syndrome have normal intelligence, left-sided cardiac lesions (e.g., aortic stenosis, coarctation of the aorta), and chromosomal abnormalities, such as partial or complete absence of X chromosomes. Since they are infertile, obviously these patients will not require anesthesia for obstetric delivery. Patients with Noonan's syndrome, on the other hand, usually have borderline or overt retardation, have right-sided cardiac lesions (e.g., pulmonary stenosis), have normal chromosomes (XX or XY), and are fertile. Since they are fertile, they may present for cesarean section, and, hence, they are of concern to the anesthesiologist.

The potential anesthesia problems presented by a patient with Noonan's syndrome include problems related to impairment of cardiopulmonary function, the possibility of a difficult airway, and the problem of technical difficulty with regional anesthesia. The same type of cardiopulmonary embarrassment that occurs in dwarf parturients because of the normal physiological changes of pregnancy occurring with the limitations imposed by the skeletal anomalies may also be seen in patients with Noonan's syndrome. The functional residual capacity (FRC), which is normally decreased in pregnancy, is further compromised because of the "shield-shaped" chest, the pectus deformity, and the kyphoscoliosis. Similarly, the contracted pelvis usually encountered in these patients prevents the engagement of the fetal head, causing even greater encroachment of the uterus on the diaphragm and producing an even further reduction in FRC. Thus, the patient with Noonan's syndrome is extremely prone to develop hypoxemia during the induction of general anesthesia, especially when followed by a difficult tracheal intubation.

In Noonan's syndrome, 30–50% of the patients are prone to pulmonary stenosis. Although the incidence is lower in females, right ventricular failure could be precipitated by excessive intravenous administration of crystalloids prior to regional anesthesia. On the other hand, if the patient is not adequately hydrated prior to the spinal or epidural block, the hypotension that may occur from the sympathetic blockade, particularly if it is excessive, may produce an inordinate decrease in right ventricular output and cardiac arrest. Thus, patients with Noonan's syndrome should have cardiac evalua-

tion and, possibly, echocardiography prior to anesthesia. If pulmonary stenosis is diagnosed, the anesthesia of choice should be general, and central venous pressure and direct arterial pressure should be monitored.

The potential for airway difficulties in Noonan's syndrome due to the high palatal arch, the dental malocclusion, and the webbed neck make the rapid sequence induction of anesthesia and tracheal intubation usually utilized to prevent aspiration of gastric contents risky and inadvisable.

Non-achondroplastic dwarfs may have odontoid hypoplasia and atlanto-axial instability that can result in cervical cord compression.<sup>8</sup> Therefore, although dwarfs with Turner's syndrome usually have a normal cervical spine,<sup>9</sup> radiographic evaluation of the cervical spine in any dwarf with skeletal abnormalities (including Noonan's syndrome) is advisable.

Regional anesthesia may be technically difficult in Noonan's syndrome because of the kyphoscoliosis, lumbar lordosis, and relatively narrow spinal canal which contains a normal-size spinal cord. The epidural and subarachnoid spaces are thus more difficult to locate, and there is a greater possibility of a subarachnoid tap while attempting to locate the epidural space.<sup>8,10</sup> Even if the epidural space can be identified, insertion and advancement of a catheter may be difficult<sup>10</sup> and the dosage and spread of the local anesthetic unpredictable. If subarachnoid block is utilized, positioning of the patient after the block may be difficult because of the abnormal curvature of the spine, and the level of anesthesia may, therefore, be difficult to control. Thus, these patients may develop a high level of anesthesia, and require circulatory and/or ventilatory support.<sup>8</sup>

Our patient had the potential airway problem posed by a high arched palate and a short webbed neck, coupled with the positional problems provided by severe kyphoscoliosis and lumbar lordosis; but echocardiography indicated that there was no pulmonary stenosis.

In our patient, awake oral endotracheal intubation would have been difficult because the faucial pillars and uvula were not visible (Mallampati class III). Awake nasal intubation would have been equally problematic because of the flattening of the mid-face and the small nasal passages and high palatal arch. Awake oral fiberoptic intubation was a possibility which was discussed with the patient as an alternative to regional, as was the possibility of total spinal and ventilation *via* a mask with cricoid pressure. However, we advised, and the patient accepted, regional anesthesia.

A continuous epidural anesthetic was originally planned, so that the dose of local anesthetic could be titrated, the mother could be awake during the delivery of the baby, and the risk of difficult intubation and aspiration could be avoided. When technical difficulty

was encountered in locating the epidural space, subarachnoid block was elected to provide most, if not all, of these advantages.

The reason that we utilized a relatively large dose (*i.e.*, 10 mg) of tetracaine in such a short pregnant patient was that we were concerned that, because of her severe lumbar lordosis, little, if any, local anesthetic would make it over the lumbar curve to flow into the thoracic curve and provide an adequate level of anesthesia. Although we achieved the desired level of anesthesia initially, the sensory level rose higher than we desired intraoperatively; but hypotension was avoided by the intravenous administration of crystalloids and incremental doses of ephedrine. There was no respiratory distress. The positional problems provided by the musculoskeletal deformities were overcome by supporting the back, hips, and lower extremities with pillows and sheets.

In summary, patients with Noonan's syndrome may present with varying numbers and degrees of anomalies, some of which present challenging anesthetic problems. These patients should have a complete evaluation to determine the presence or absence of cardiac, renal, and musculoskeletal anomalies; and, if a patient with Noonan's syndrome becomes pregnant, an anesthesiologist should evaluate the patient and plan for the anesthetic management early in the prenatal period. Possible difficulties with regional and general anesthesia should be discussed fully with the patient, and this discussion should include the possible need for awake fiberoptic tracheal intubation. However, in spite of the

possible technical difficulties, regional anesthesia carried out very cautiously offers the advantage of avoidance of the risk of difficult intubation and aspiration. An ear, nose, and throat surgeon should be available until the level of regional anesthesia is determined to be safe. Spinal anesthesia probably should not be given if the patient has pulmonary stenosis or does not have a cardiac workup to rule out the presence of such a lesion.

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