

Anaesthetic management for caesarean section in a patient with Sjogren's syndrome

Sir,

Sjogren's syndrome is an autoimmune disease of the exocrine glands with an incidence of 0.1-4.8%. It is more common in females. Patients present with dry eyes and mouth. Autoantibodies anti-SS-A (anti-Ro) and anti-SS-B (anti-La) circulating in maternal serum can cross the placental barrier and cause a congenital heart block (CHB).^[1] We present the anaesthetic management of a parturient with Sjogren's syndrome posted for elective caesarean section.

A 30-year-old second gravida at 27 weeks gestation was referred to our obstetric department with a diagnosis of CHB detected during a routine antenatal check-up. She gave history of proximal muscle weakness and low serum potassium levels in her first pregnancy. She was not further evaluated and underwent an uneventful caesarean delivery at term under spinal anaesthesia three years back. She also gave a history of dryness of the mouth for the last two years.

Blood tests revealed positivity for anti-SS-A/anti-Ro and borderline positivity for anti-La antibodies. Rheumatology consultation was done and she was diagnosed with Sjogren's syndrome. Foetal echocardiogram showed a complete CHB with an atrial rate of 134-143/min and a ventricular rate of 54-60/min. There were no structural heart defects and ventricular function was normal. She was started on oral dexamethasone 4mg once daily and salbutamol 4mg thrice daily. Due to persistently low levels of potassium,

she was started on syrup potassium chloride 10ml thrice daily. She was posted for an elective caesarean section at 37 weeks of gestation. Blood tests on the day of surgery revealed serum potassium of 2.8mEq/L. An 18-gauge external jugular vein was secured and potassium was corrected with 20 mEq potassium chloride.

Repeat serum potassium level was 3.5 mEq/L. Caesarean section was performed under subarachnoid block with 1.8 ml of 0.5% heavy bupivacaine and 10µg of fentanyl. A neonate with a heart rate of 55 beats/min was born. No active resuscitation was required, and the baby was shifted to neonatal intensive care unit (ICU) for observation.

Most of these patients deliver by caesarean section as it is difficult to monitor the foetal status during labour. The choice of anaesthesia depends on the urgency of delivery. Regional anaesthetic techniques are preferable over general anaesthesia.^[2] Preoperative assessment should rule out associated rheumatoid arthritis, interstitial pneumonitis, nephritis, vasculitis, peripheral neuropathies and lymphomas. Renal tubular acidosis with positive urine anion gap and hypokalemia require potassium and sodium bicarbonate replacement.^[3] Drugs with anticholinergic side effects like atropine, phenothiazines and tricyclic antidepressants are avoided. Hypothyroidism and sleep disorders are common. Hoarseness of voice is suggestive of crico-arytenoid joint involvement. Difficult airway has to be anticipated because of the enlarged parotid, submandibular glands and temporomandibular joint arthritis. Warm and humidified inspired gases have to be used to prevent mucus plugs from causing bronchospasm and dyspnoea. Cold intravenous fluids are avoided. Autonomic nervous system dysfunction may be present and careful titration of induction agents is required to avoid haemodynamic instability.

Care of fragile skin and eye protection with ocular lubricants is necessary.

Pregnancy with Sjogren's syndrome is associated with an increased rate of spontaneous abortion and congenital abnormalities.^[4] CHB, neonatal lupus rash, thrombocytopenia and elevated liver enzymes are seen in affected neonates.^[5] Foetal CHB occurs at 16-24 weeks of gestation with an incidence of 1-2%.^[1] The neonate usually has a structurally normal heart. Most of the mothers are asymptomatic and the disease is identified after the birth of the affected child. There is a 10 times risk of recurrence of CHB in subsequent pregnancies.^[5] Prenatally diagnosed CHB is treated with corticosteroids.^[6] Continuous monitoring of the electrocardiogram of the neonate in the ICU is necessary.^[5] CHB is associated with 15-20% mortality in the first three months. More than 50% of neonates will require pacemaker implantation.^[7] A beta-adrenergic agonist is used to increase the heart rate temporarily and a pacemaker is implanted if the heart rate is less than 55 beats/min.^[5] Multidisciplinary management involving obstetricians, rheumatologists, paediatric cardiologists and neonatologists is required in the management of these parturients for successful obstetric and neonatal outcomes.

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Conflicts of interest

There are no conflicts of interest.

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