

## ANAESTHETIC IMPLICATION OF THE PARTURIENT WITH JERVELL-LANGE SYNDROME COMING FOR CAESAREAN SECTION

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### PRESENTATION OF CASE

24 years old female, deaf from childhood, primigravida, presented for elective caesarean section. She had prolonged QT interval in ECG. Further history and examination revealed that she had been hospitalised for repeated syncope. She was diagnosed to have congenital long QT syndrome after Holter monitoring. Familial history of long QT syndrome was present. She was started on Tab. Propranolol 20 mg twice daily. She was also advised by cardiologist to undergo procedure for implantable cardiac defibrillator, but the patient did not undergo the procedure due to personal reasons.

She conceived spontaneously and was on regular follow up with the obstetric and cardiology teams. She was scheduled for elective caesarean section in view of underlying condition. All routine investigations were done and found to be normal. Echocardiogram was also done and it revealed normal study with ejection fraction of 69%. Electrocardiogram - sinus rhythm with QT prolongation. She was advised nil per oral as per fasting guidelines. She was premedicated with Inj. Ranitidine 50 mg, which was given slowly in intravenous line on morning of procedure and intravenous fluids with 0.9% normal saline of 100 mL per hour was started from morning. She was also advised to take morning dose of Tab. Propranolol with sips of water.

After obtaining consent from the patient, the patient was shifted to operating room. Subarachnoid blockade was planned. Monitors such as 5 lead ECG, noninvasive blood pressure and pulse oximetry were connected. Biphasic defibrillator ECG cables were also connected and kept ready. Patency of existing large bore intravenous line was checked and another large bore intravenous line secured. Baseline vitals were recorded. Goals were to maintain sinus rhythm with heart rate ranging from 60-80 beats per minute and maintaining mean arterial pressure of 70 mmHg. Under aseptic precautions, subarachnoid blockade was performed

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and 10 mg of 0.5% heavy bupivacaine was given and level achieved was T<sub>4</sub>. Caesarean section was done and boy weighing 2.1 kg with 8/10 and 9/10 Apgar at 0 and 5 minutes were extracted. Post extraction of baby, 10 units of Inj. Oxytocin was given as slow infusion. Blood pressure fell to mean arterial pressure of 62 mmHg and maintained with help of Inj. Ephedrine 3 mg and intravenous fluids. Post procedure, the patient was shifted to high dependency unit for observation and the period was uneventful. Paracetamol 1 gm intravenous was given in our case for management of pain. Cardiology review was obtained and suggested to continue same line of management as she was on and added implantable cardiac defibrillator device at the earliest. Both mother and baby was fine and discharged.

### DIFFERENTIAL DIAGNOSIS

Electrolyte imbalance.

Myocardial ischaemia.

Raised intracranial pressure.

Hypothermia.

Congenital syndromes.

### CLINICAL DIAGNOSIS

She had long QT syndrome and associated with deafness. She has features suggestive of Jervell-Lange Syndrome. Jervell-Lange Syndrome is a type of long QT syndrome associated with bilateral sensorineural hearing loss.<sup>1</sup>

### PATHOLOGICAL DISCUSSION

Various organ system undergoes dramatic physiological changes during pregnancy and each change in each system plays important factor in management of all cases of pregnancy from anaesthetic point of view.<sup>2</sup>

There is decompensation of heart due to physiological changes in cardiovascular changes in pregnancy in structural heart diseases whereas there are only few known effects in electrical activity during pregnancy.<sup>3,4</sup>

Jervell Lange-Nielsen syndrome is a type of long QT syndrome associated with bilateral sensorineural hearing loss.<sup>1</sup> This syndrome has potency to cause polymorphic ventricular tachycardia, which is known as Torsades de pointes.<sup>5,6</sup> The principal danger of this polymorphic ventricular tachycardia is deterioration into pulseless ventricular tachycardia or ventricular fibrillation and death.<sup>5,6,7,8</sup> The factors such as surgical stress, anxiety,

bradycardia, tachycardia, hypotension, hypercarbia, hypothermia, inadequate analgesia and anaesthesia may cause sympathetic stimulation increasing the risk of polymorphic ventricular tachycardia.<sup>5,6,7,8</sup>

There are only few reports of a parturient with Jervell Lange-Nielsen syndrome scheduled for caesarean section. Hence, we aimed at bringing forth the anaesthetic implication of the parturient with Jervell Lange-Nielsen syndrome coming for caesarean section.

## DISCUSSION OF MANAGEMENT

Jervell Lange-Nielsen syndrome is an inherited autosomal recessive condition.<sup>9</sup> This condition is a combination of congenital long QT with deafness. This condition as prevalence of 1 in 3000.<sup>10</sup> It is well-known fact that in pregnancy there is increase in cardiac output and heart rate. This in turn can cause increase incidence in arrhythmias in normal women, but this when associated with Jervell Lange-Nielsen syndrome, there is increase in morbidity and mortality significantly when compared to parturient without Jervell Lange-Nielsen syndrome.<sup>11</sup> This makes the job of anaesthesiology more challenging in management of such condition.

The drug which remains mainstay of treatment of this condition is beta blockers.<sup>10</sup> Our patient was also on beta blocker Tablet Propranolol 20 mg and continued perioperatively. There are no studies or evidence that reported incidence of teratogenicity with use of beta blockers, but they are prone to have foetal effects such as intrauterine growth retardation and foetal bradycardia.<sup>3,12</sup> In our case, the foetal heart rate was never below 140 bpm.

The other mainstay of management of this condition is implantable cardiac defibrillator along with beta blocker. In our case, the patient was also advised to undergo the same, but the patient denied for some personal reasons.

Regional anaesthesia is better compared to general anaesthesia as there is reduction of the stress response and provision of effective analgesia, moderate catecholamine release, reducing the risk of Torsades, but it also add disadvantages.<sup>10</sup> There are cases, which were reported was done under regional anaesthesia. Most of them avoided regional anaesthesia in view of potential hypotension, which might trigger arrhythmia.

The most important goals in management of this condition is prevention of pain, anxiety, hypoxia, hypercarbia, hypothermia, shivering and glycaemic control.<sup>5</sup> The other most important factor is to check and maintain the proper functioning of implantable cardiac defibrillator. The factors which interfere with proper functioning of implantable device should be avoided in the perioperative period.

All precautions should be made available and taken to prevent any arrhythmias during perioperative period. In our case, the patient was advised to continue propranolol on the day of surgery. Biphasic defibrillator was also kept ready and the ECG leads from the defibrillator was also connected along with routine 5 lead ECG.

Intraoperative period all the drugs to control rate, pressure, anxiety and arrhythmias are to be kept ready and used when needed appropriately. In our case, the rate was within normal limits throughout the period and variations in pressure was maintained by Inj. Ephedrine and intra venous fluids. Though there are less data on the effects of Ephedrine on the QT interval, this drug was used safely.

After the delivery of baby oxytocin was given to patient and close monitoring was done. There was no change in rhythm in our case. Oxytocin was considered as potentially dysrhythmogenic in patients with long QT, however, oxytocin has been used for induction of labour in long QT without effect.<sup>13,14</sup>

The patient was monitored in high dependency unit for any changes in rhythm and for other common complication in parturient post procedure. In our case, there was no significant events occurred in this period. Postoperative pain might trigger increase in rate. There are cases were epidural were used for both labour analgesia and postoperative pain management.<sup>10</sup> In our case, postoperative pain was alleviated by administration of intravenous paracetamol 1 gm every 8<sup>th</sup> hourly.

Cases like Jervell Lange-Nielsen syndrome can be effectively and safely administered with meticulous monitoring without any complications.

## CONCLUSION

All cases of pregnancy with Jervell Lange-Nielsen syndrome can be effectively carried out with multidisciplinary approach. Anaesthetic management of such cases aims in prevention of prolongation of QTc, also be prepared for management of any arrhythmias, which occur in perioperative period with main concern on Torsades. Both regional and general anaesthesia can be administered to such patients with very effective method in monitoring and careful administration of anaesthesia.

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