

ANESTHETIC MANAGEMENT OF A PREGNANT PATIENT WITH JERVELL AND LANGE-NIELSEN LONG QT SYNDROME



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INTRODUCTION

The Jervell and Lange-Nielsen (JLN) syndrome is a rare and severe autosomal recessive variant of the congenital long QT syndrome (cLQTS) associated with sensorineural deafness. Individuals with the JLN syndrome are at high risk of developing torsades de pointes, ventricular fibrillation and sudden cardiac death. These cardiac events can be precipitated by sympathetic stimulation due to exercise, pain or emotional distress.¹ Patients with syncope are particularly prone to sudden death and AICD implant has been suggested as early as at age two.²

CASE REPORT

The patient was an 18 year old female with poor prenatal care and known JLN syndrome that presented to our institution at 37 weeks gestation. She had syncope two years earlier but refused AICD placement at that time. Electrocardiogram demonstrated a normal sinus rhythm of 67 bpm and a QTc of 492ms. The obstetric team chose Cesarean section as the mode of delivery that would put the patient at the lowest risk for having a cardiac event. The electrophysiology team deferred AICD placement until delivery. The management plan was communicated to the patient by means of a sign language interpreter. She was brought to the operating room for elective cesarean section with electrophysiologists present and defibrillator pads in place. Following premedication with Labetalol 10 mg IV and adequate preload, a combined spinal-epidural (CSE) anesthetic technique was performed. Intrathecal dose consisted of 12 mg hyperbaric 0.75% bupivacaine, 250µg of preservative-free morphine and 25µg of fentanyl. The administration of neuraxial anesthesia and subsequent cesarean section proceeded without adverse maternal cardiac events. A healthy baby boy was delivered with Apgar scores of 9/9. Epidural catheter was used for postoperative analgesia. An AICD was placed 3 days after the Cesarean section.

DISCUSSION

The use of epidural³ and general⁴ anesthesia for cesarean section in patients with JLN syndrome has been documented in case reports. Although spinal and CSE anesthetics have been used successfully in patients with Romano-Ward syndrome⁵ (a more common, autoso-

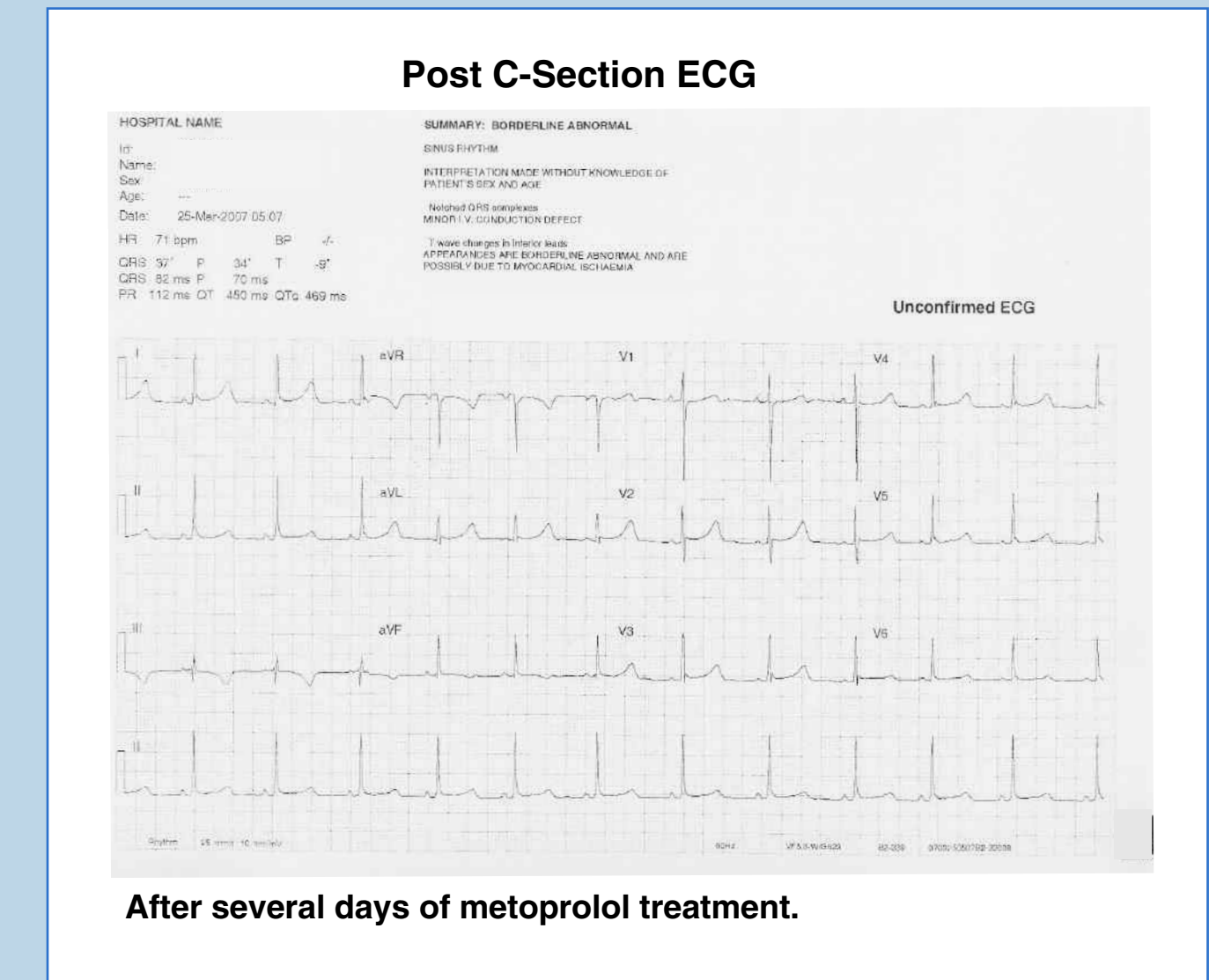
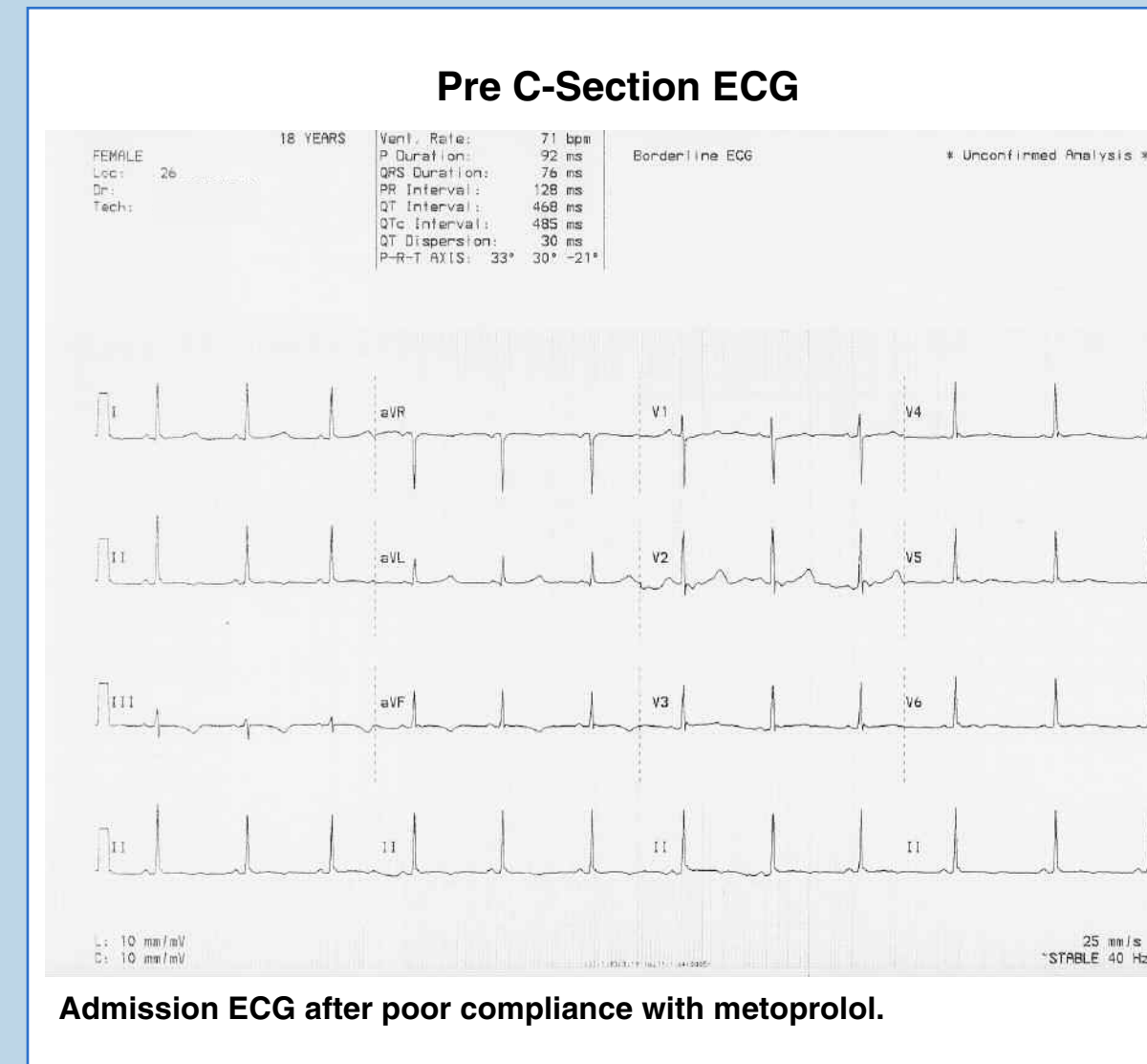
Table 1. Drugs That Prolong the QT Interval and/or Induce Torsades de Pointes

Well Documented Association	Moderate Potential/Case Reports	Increased Potential Risk in Congenital QTS Patients*	Unclear Association† (at therapeutic doses)
CARDIAC Amiodarone Sotalol Dofetilide Dofetilide Procainamide Quinidine Sotalol	Flecainide Moxipril/HCTZ Nicardipine Isradipine Procainamide Telithromycin	Dobutamine Dopamine Epinephrine Epinephrine Isoproterenol Misdoline Norepinephrine Phenylephrine	Mexiletine
ANTI-MICROBIALS Azithromycin Erythromycin Foscarnet Grepafloxacin Levofloxacin Moxifloxacin Sparfloxacin Telithromycin	Azithromycin Foscarnet Grepafloxacin Levofloxacin Moxifloxacin Sparfloxacin Telithromycin		Ampicillin Ciprofloxacin Fluoroquinolone Itraconazole Ketoconazole TMP/SMX
PSYCHOTROPICS Chlorpromazine Haloperidol Mesoridazine Pimozide Thioridazine	Lithium Quelapine Risperidone Venlafaxine Ziprasidone		Amitriptyline** Amoxapine Clomipramine Desipramine Doxepin Fluoxetine Galantamine Imipramine Nortriptyline Paroxetine Protriptyline Sertraline
MISCELLANEOUS Aranic trioxide Chlorpromazine Claspridol Compendonol Droperidol Halofantrine Haloperidol Levomethadyl Mesoridazine Methadone Ongentophosphates Pimozide Thioridazine	Amantadine Asterizole Chloral Hydrate Doxastrom Felbamate Fosphenytoin Granisetron Indapamide Octreotide Ondansetron Salmeterol Tacrolimus Terfenadine Temozolomide Tizanidine Voriconazole	Albuterol Cocaine Fenfluramine Levulbuterol Metoprolol Pentameterol Phenylephrine/epinephrine Propofol Pseudoephedrine Ritodrine Terbutaline	

* These agents are in addition to those in the well documented and moderate risk associations lists, to be avoided in patients with congenital QTS. † Limited case reports available. ‡ Products which have been withdrawn from and/or not available on the United States market. § Onset may be delayed. ¶ FDA black box warning. # Proposed risk, marked with an asterisk of decreased cardiac ionic effects. ** Prolongation seen in overdose. A continuously updated list of drugs is available at www.torsades.org (accessed July 6, 2004).

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mal dominant form of cLQTS),¹ this is the first documented use of CSE in a patient with JLN syndrome. Avoiding sympathetic stimulation and tachycardia induced arrhythmias to the greatest extent possible is of paramount importance in the choice of anesthetic technique for patients with JLN syndrome. If the regional block is not adequate, the patient can experience pain with subsequent sympathetic stimulation; if the regional block is too high, hypotension and reflex tachycardia can put the patient at risk. Induction of general anesthesia and laryngoscopy can also cause sympathetic stimulation and increase the risk of torsades de pointes and ventricular fibrillation. Keeping this in mind, a combined spinal-epidural technique was chosen to provide both adequate anesthesia for the procedure and effective post-operative analgesia. Ensuring adequate communication of the treatment and anesthetic plans to these patients with a sign language interpreter is essential. Involvement of electrophysiologists in the operating room, to aid in management of potential cardiac arrhythmias, was also an important aspect in the care of this patient.



CONCLUSION

We conclude that combined spinal-epidural technique could be the preferred option for parturient patients with JLN syndrome presenting for Cesarean section.

REFERENCES

- P. D. Booker, S. D. Whyte and E. J. Ladusans : Long QT syndrome and anesthesia. *British Journal of Anesthesia*, 2003, Vol. 90, No. 3 349-366
- Denjoy I, Lupoglazoff JM, Villain E, Vaksman G, Godart F, Lucet V, Leenhardt A, Guicheney P, Schwartz P: The Jervell and Lange-Nielsen syndrome. Natural history, molecular basis and clinical outcome. *Arch Mal Coeur Vaiss*. 2007 May;100(5):359-64
- Ryan H: Anesthesia for cesarean section in a patient with Jervell, Lange-Nielsen syndrome. *Can J Anaesth*. 1988 Jul;35(4):422-4
- Yamashita K, Tomiyasu S, Fujie T, Sumikawa K, Akamine S, Ayabe H, Akiyama Y, Hayano M: Endoscopic resection of the thoracic sympathetic trunk for the treatment of frequent syncopal attack of idiopathic long QT syndrome. *Masui*. 1999 Apr;48(4):399-403.
- Ganta R, Roberts C, Elwood RJ, Maddineni VR: Epidural anesthesia for cesarean section in a patient with Romano-Ward syndrome. *Anesth Analg*. 1995 Aug;81(2):425-6