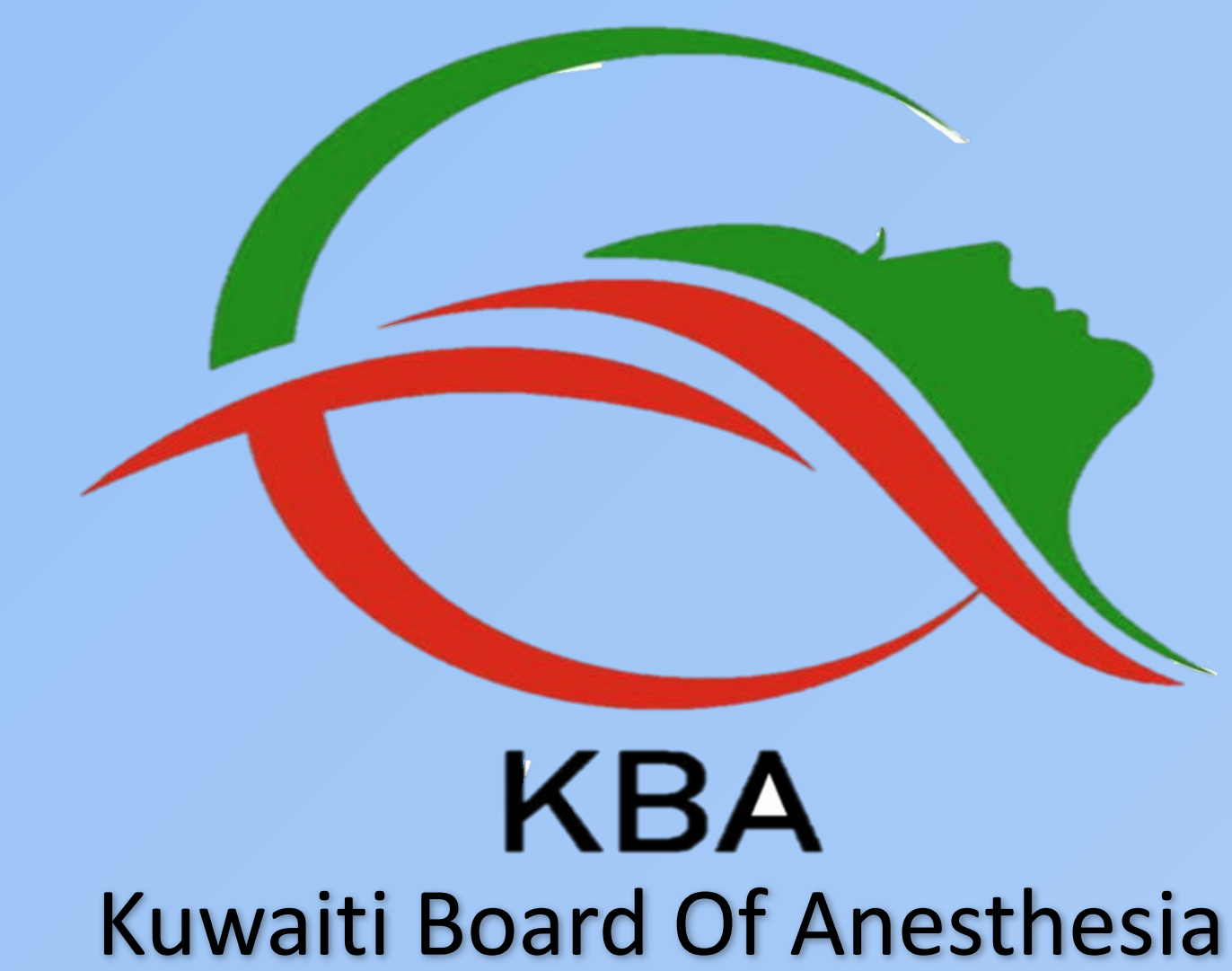




Anesthetic management of an obstetric patient with congenital long QT syndrome type 2 undergoing ovarian dermoid cyst excision: A Case Report

Ali Alwadani¹, Abdelrady Ibrahim², Fatemah Qasem¹²

Kuwait Board of Anesthesia, Kuwait Institute for Medical Specialization, Kuwait
Department of Anesthesia and Intensive Care, Maternity Hospital, Ministry of Health, Kuwait



Introduction

- Congenital long QT syndrome (LQTS) is a genetic disorder characterized by an extended QT interval in the electrocardiogram (ECG).
- LQTS predisposes affected individuals to arrhythmic syncope, ventricular torsades-de-pointes (TdP), and sudden cardiac death (SCD) at a young age.
- About 90% of LQTS cases are caused by pathogenic variants in three genes (KCNQ1, and hERG found in potassium channel and SCN5A found in sodium channel) , leading to different types of the syndrome (LQT1, LQT2, and LQT3).
- **Triggers** such as emotional stress and audible stimulation E.g ringing phone , alarm clock or crying baby can provoke ventricular fibrillation (VF) in LQTS patients, and hormone concentrations, particularly during pregnancy and the postpartum period, can impact QT duration and the risk of arrhythmias.
- During pregnancy, the risk of arrhythmias is **reduced** in women with congenital LQTS, but it becomes **particularly high** during the 9-month postpartum period.
- up to 10% of women with LQTS experience their first cardiac event during the postpartum period

Case Summary

- A 26-year-old woman who is 16 weeks pregnant and has a history of previous **uncomplicated normal vaginal delivery** was diagnosed with congenital Long QT Syndrome (LQTS) Type 2.
- This diagnosis was reached after the patient experienced **multiple syncopal attacks**, triggered by various stimuli. The diagnosis was further supported by the **electrocardiogram (ECG) findings**, which showed significantly prolonged QTc intervals, abnormal repolarization (bifid T waves), and frequent Torsades de Pointes. Additionally, an echocardiogram revealed depressed cardiac function attributed to tachycardia-induced cardiomyopathy. The patient's **family history**, including the sudden cardiac deaths of her mother and aunt at young ages, also contributed to the diagnosis.
- At 16 weeks of gestation, the patient required surgery to remove an ovarian dermoid cyst due to severe abdominal pain and to avoid the complications. The diagnosis was confirmed through ultrasound examination.
- The patient was admitted to the ICU to ensure **thorough preparation** for the upcoming surgery. This preparation encompassed preoperative patient education and readiness, operating theater (OT) preparation, selection of anesthesia, and pain management.
- **Combined spinal epidural anesthesia** was chosen as the preferred anesthesia method to mitigate potential complications, while intra-operative sedation was administered to minimize patient stress.

Discussion

This case report emphasizes the importance of considering Long QT Syndrome (LQTS) in patients with recurrent syncopal attacks and a family history of sudden cardiac death (SCD). Diagnosis.

- Patients with LQTS are at higher risk of **malignant Torsades de Pointes** during the perioperative period.
- **Proper management includes** : recognizing the syndrome, minimizing sympathetic response , providing premedication, avoiding factors impacting cardiac repolarization, ensuring pain relief, and closely monitoring the QT interval.
- **Patient preparation** is a crucial step that requires the involvement of a multidisciplinary team consisting of surgeons, anesthesiologists, and cardiologists.
- **Operation theater preparation** should encompass the creation of a calm and quiet environment to minimize stress, maintaining a normal temperature, ensuring the availability of ASA monitors with 5-lead ECG, having a readily accessible supply of anti-arrhythmic medications, and removing any medications that may potentially prolong the QT interval.
- **Combined spinal epidural anesthesia** along with intra-operative sedation was chosen and performed under strict sterile conditions. Aims to maintain normal hemodynamics.
- **Postoperative care** Postoperative care should involve vigilant monitoring, the avoidance of stress or triggers for arrhythmia, and ensuring adequate pain management.

Conclusion

- Congenital Long QT Syndrome (LQTS) should be considered in patients with recurrent syncopal attacks and a family history of sudden cardiac death.
- Proper management requires a multidisciplinary approach.
- Minimizing sympathetic activity, avoiding medications that prolong the QT interval, providing adequate pain relief, and closely monitoring the QT interval are important in mitigating the risk of life-threatening arrhythmias.
- Combined spinal-epidural anesthesia represent a suitable choice to reduce sympathetic response in these cases.
- There is a lack of clinical trials on optimal anesthetic management in these cases, but this case report suggests the efficacy of combined spinal-epidural anesthesia in pregnant patients with LQTS.

References

- Anesthesiology 2005; 102:204 –10 , 2004 American Society of Anesthesiologists, Inc. Lippincott Williams & Wilkins, Inc. Anesthesia for Patients with Congenital Long QT Syndrome Susan J. Kies, M.D.,* Christina M. Pabelick, M.D.,† Heather A. Hurley, Pharm.D.,‡ Roger D. White, M.D.,§ Michael J. Ackerman, M.D., Ph.D.
- Seth R, Moss AJ, McNitt S et al (2007) Long QT syndrome and pregnancy. J Am Coll Cardiol 49:1092–1098
- RashbaEJ,ZarebaW,MossAJetal(1998) Influence of pregnancy on the risk for cardiac events in patients with hereditary long QT syndrome. LQTS Investigators. Circulation 97:451–456.