Anesthetic Management of Catecholaminergic Polymorphic Ventricular Tachycardia in Pregnancy

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Learning Objectives

1. Describe the pathophysiology and presentation of catecholaminergic polymorphic ventricular tachycardia (CPVT).
2. Outline the goals of management of CPVT, particularly in pregnant females during labor.
3. List treatment options for parturients experiencing CPVT during labor.
4. Explain the fetal implications of maternal CPVT.

Introduction

CPVT Pathophysiology

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare familial or de novo mutation in either the cardiac RyR2 Ryanodine gene (CPVT1) or CASQ2 calsequestrin gene (CPVT2).[1] Both of these mutations alter normal cardiac myocyte calcium regulation, enhancing the propensity of sarcoplasmic reticulum calcium to be released. These spontaneous calcium discharges cause unpredictable malignant arrhythmias presenting as syncope or sudden cardiac death.

Presentation

Arrhythmic events are triggered by exercise, stress, or emotions, and more than 60% of affected individuals experience symptoms by age 20 years.[2] Patients will have a normal ECG and a structurally normal heart, and unfortunately cardiac arrest is the first manifestation of the disease in 13% of patients.[2] The risk of cardiac events at a young age is significantly higher in males with RyR2 mutations than in females (relative risk = 4.2).[2]

Treatment

Beta-blockers are first line treatment.[3] Refractory cases may require the addition of calcium channel blockers, flecainide, left cardiac sympathetic denervation, and ICDs.

Pregnancy and CPVT

Hormonal changes of pregnancy may increase the sensitivity of adrenergic receptors, increase overall arrhythmia burden, and expose the parturient to high levels of stress. Despite this, the limited literature on parturients with this condition does imply CPVT is well tolerated during pregnancy. Cheung et. al. retrospectively concluded no difference in cardiac event rate in women during pregnancy versus non-pregnant, and the overall event rate of a major cardiac event in 96 CPVT patients was 2%. [3]

Case Presentation

We describe a 30yo G5P3013 at 37wk1d admitted for induction of labor for IUGR. She carries a diagnosis of CPVT causing one prior cardiac arrest resulting in ICD placement. Her first three vaginal deliveries were prior to her diagnosis and ICD placement. Since receiving the ICD, it has discharged four times, induced by arguments, public speaking, exercising, and anxiety. Her ICD was functioning properly at the time of presentation and was set to deliver tachytherapies at a VT or VF rate of 200bpm or higher. She was also medically managed on propranolol 80mg twice daily. Cardiology was consulted who recommended continuing beta blocker therapy throughout the peri-partum period, and minimizing stress as much as possible during labor. Prior to starting the oxytocin IOL the patient was placed on 24 hour telemetry monitoring with a dedicated telemetry nurse and a magnet was placed at bedside. A lumbar epidural was placed by anesthesia and ran initially at 5cc an hour with 4cc bolus every 12 min to keep the patient as comfortable as possible. The infusion mixture consisted of 0.2% ropivacaine mixed with 2mcg/mL of fentanyl. During epidural placement a smaller test dose of 2mL 1.5% lidocaine with 1:200k epinephrine was given to prevent tachyarrhythmias in the event of unintentional intravascular injection. The patient delivered a healthy baby boy after just 1 hour of labor. She was monitored on telemetry for two additional days prior to discharge home.

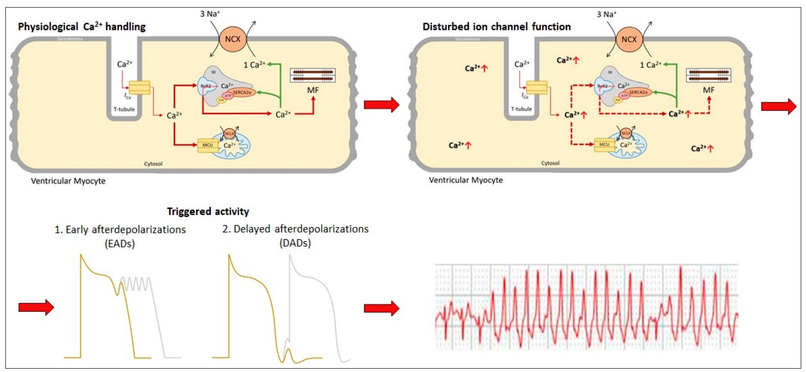

Figure 1. Calcium homeostasis of cardiac myocytes.[4]

Discussion

Goals of Management

1. Minimize parturient stress and pain
- Consider early neuraxial techniques and evaluate epidural/pain scores regularly
- Discuss delivery plan with OB, may need to avoid extensive pushing/laboring

2. Monitor closely
- Continuous telemetry monitoring throughout intra-partum and two days post-partum

3. Avoid arrhythmogenic medications
- Use direct acting alpha constrictors for hypotension over ephedrine, epinephrine, norepinephrine
- Caution with terbutaline
- Consider a lower test dose during epidural placement or omitting epinephrine all together

4. Continue beta blocker therapy throughout pregnancy and delivery
- Metoprolol and propranolol are the most appropriate during pregnancy and lactation
- Atenolol is a class D teratogen

Fetal Implications

1. All newborns born to a CPVT mother should undergo genetic screening
- RyR2-associated CPVT1 is inherited in an autosomal dominant manner, with a mean penetrance of 83%.[2]
- CASQ2-related CPVT2 is inherited in an autosomal recessive manner.[2]

Conclusion

CPVT is a rare and life threatening condition but well tolerated during pregnancy if precautions are taken. Goals for parturients should center around minimizing catecholamine release and continuing beta blocker therapy through the peri-partum period. Multidisciplinary management with anesthesia, obstetrics, cardiology, and neonatology will ensure the adverse event rate in these patients remains low.

References

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