Dental Management of a Patient With Catecholaminergic Polymorphic Ventricular Tachycardia: A Case Report

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ABSTRACT

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a type of cardiac arrhythmia that occurs in people with a structurally normal heart. Stress or anxiety-induced release of endogenous catecholamines causes a dysfunction in the myocytic calcium-ion channel, leading to ventricular arrhythmias that can cause dizziness, syncope, or sudden cardiac death. Since dental procedures can be anxiety-provoking, the main purpose of this paper is to report the dental management of a young patient with dental fear and CPVT. Several other issues are also discussed, such as the importance of continual collaboration with medical colleagues, the risk-benefit of using epinephrine-containing local anesthesia for dental treatment for patients with arrythmias, the potential risk of repeated general anesthesia in a patient with a cardiac arrhythmia, and the challenges of providing comprehensive dental treatment in a high caries-risk patient with extreme dental anxiety. (J Dent Child 2013;80(2):101-4)

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Catecholaminergic polymorphic ventricular tachycardia (**CPVT**) is one of the most severe of the inherited arrhythmogenic disorders that may present in a structurally normal heart.[1] This condition, which was first described in 1975, occurs as a result of mutations in genes encoding the Ryanodine receptor 2 (**RyR2**) and Calsequestrin 2 (**CASQ2**).[2] The modes of inheritance are autosomal dominant (RyR2) and autosomal recessive (CASQ2), and the responsible mutations are located on chromosomes 1q42-43 and 1p13-21, respectively.[1-4] New mutations are responsible for roughly half of the cases of CPVT, while RyR2 mutations account for approximately 50% and CASQ2 mutations account for approximately 1% to 2%.[2]

Although different genes are involved, RyR2 and CASQ2 mutations appear clinically identical. Both RyR2 and CASQ2 genes encode proteins that are involved in handling calcium, which is necessary for the regular contraction of myocytes. The RyR2 channel releases calcium ions from the sarcoplasmic reticulum to the cytosol in response to certain signals. The resulting increase in calcium ion concentration triggers the myocyte to contract. Calcium ions are then transported back to the sarcoplasmic reticulum and bound to CASQ2, allowing the cardiac muscle to relax. The mutations result in changes in the structure and/or function of the RyR2 channel or CASQ2. The disruption to the control of myocytic calcium ion flow leads to abnormal cardiac rhythm, resulting in arrhythmias.[5]

The prevalence of CPVT is estimated to be 1 in 10,000, affecting both men and women equally. Symptoms often appear in the first decade of life, usually after 3 years of age, with the average age of onset from 7 to 9 years.[3] The first manifestation of CPVT is syncope following emotional or physical stress.[5] Other symptoms include heart palpitations, light-headedness, and dizziness. Junttila et al.[2] reported that 30% of CPVT sufferers have a family history of sudden cardiac death before the age of 40 years. Electrocardiograms appear normal, and the diagnosis of CPVT may be missed unless an exercise stress test is

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performed.[6] Patients diagnosed with CPVT are usually treated with a beta-blocker, such as metoprolol or atenolol, to reduce the heart rate and antagonize the effects of catecholamines. By depressing adrenergic tone, beta-blockers reduce the occurrence of exercise and emotion-related arrhythmias. It is often recommended that competitive sports and strenuous exercise be avoided in CPVT patients.[2]

There is currently no dental literature regarding CPVT. This condition has dental relevance because some invasive dental procedures can be anxiety-provoking. Therefore, the purpose of this case report is to describe the dental management of a young patient with dental anxiety and CPVT.

CASE REPORT

A 13-year-old female presented with her grandmother to the pediatric dental clinic at the University of Florida, Gainesville, Fla., for a new patient exam. The grandmother, who had become the patient's adoptive mother since she was 2 years old, reported a medical history significant for "adrenalin-induced" ventricular tachycardia, which was controlled with 75 mg of atenolol twice daily. Additional medical history included hypothyroidism managed with Synthroid (AbbVie Inc., North Chicago, Ill.), stress-induced vasovagal syncope, diaphoresis, and mild developmental delay. The patient's developmental delay was associated with a neurologic injury from ventricular tachycardia that occurred before the diagnosis of CPVT was made.

She had been under the care of the University of Florida Pediatric Cardiology and Electrophysiology Clinic since she was diagnosed at the age of 3 years. Her cardiologist's recommendations included limiting vigorous physical activity, avoiding dehydration, reducing anxiety-provoking situations, and regularly using a Holter monitor. The Holter monitor, also known as ambulatory electrocardiography, is a portable device commonly used for 24-hour monitoring of electrical activity of the heart. Its extended recording period is sometimes useful for observing occasional cardiac arrhythmias that would be difficult to identify within a shorter period of time.[6]

Prior to attending the university's pediatric dental department, the patient had been under the care of a private pediatric dentist who had performed previous dental treatment under general anesthesia (**GA**). The growing complexity of her medical history was the main reason for referral for dental treatment in a hospital setting.

At the initial dental evaluation, despite being knowledgeable about the medical condition, the patient's mother had an extremely nervous disposition. It was apparent that the patient was influenced by her mother's anxiety. The oral examination revealed a permanent dentition with multiple carious lesions on the molars and maxillary incisors. Though asymptomatic, the maxillary left central incisor showed extensive carious involvement and had a negative response to both cold and electric pulp testing. Her oral hygiene was poor, with extensive plaque accumulation and marked gingival inflammation. She was also consuming a highly cariogenic diet. Dental radiographs confirmed the clinical carious lesions and revealed a periapical radiolucency around the maxillary left central incisor.

Considering her complex medical condition, extent of treatment required, and extreme dental anxiety, it was decided that the dental treatment be completed under GA. After a preoperative consultation with her cardiologist, the recommendations for dental treatment were as follows: preoperative cardiology and anesthesia support, postoperative admission for cardiac monitoring, adequate pain control, and management of patient anxiety. In the event that polymorphic ventricular tachycardia (**PVT**) occurred, the cardiologist recommended intravenous esmolol and lidocaine but not defibrillation. If performed, defibrillation would likely create the release of endogenous catecholamines and further promote the ventricular tachycardia.

On the day of dental treatment, the patient was given 10 mg of midazolam orally by the anesthesiology team to reduce preoperative anxiety. The following dental treatment was performed in the operating room under rubber dam isolation: dental prophylaxis; root canal therapy of the maxillary left central incisor followed by anterograde filling with gutta-percha (Obtura III Max Heated Gutta Percha System, Obtura Spartan Endodontics, Earth City, Mo); raising of a full-thickness buccal mucoperiosteal flap; a retrograde filling, following apicetomy, using mineral trioxide aggregate (ProRoot MTA, Dentsply and Tulsa Dental, Tulsa, Okla); and closure of the flap with 3-0 chromic gut sutures (Ethicon Inc, San Angelo, Texas).

Resin-based composite material (TPH, Dentsply International Inc, Milford, Del.) was used to restore the carious teeth and the access cavity of the maxillary left central incisor. Following an initial uneventful postoperative recovery, the patient was admitted by the pediatric cardiology team for supplemental intravenous hydration, cardiac monitoring, and further observation. Her usual oral medications were resumed later that day on the pediatric ward, and her vital signs remained stable throughout admission.

Discharge from the hospital occurred after 24 hours, and the patient was given instructions for meticulous oral hygiene and daily topical fluoride (PreviDent 5000 Plus Colgate Palmolive, New York, N.Y.). The patient returned to the dental clinic 1 week postoperatively, and no complications were reported. The patient was placed on a regular 3-month recall schedule in view of her high caries risk.

Nine months after the dental treatment was performed in the operating room, and in spite of intensive preventive regime, the patient had poor oral hygiene and presented with new and recurrent carious lesions in the

maxillary incisors. At the time, our GA waiting list was protracted and there was genuine concern about the risk of repeated dental treatment under GA. These concerns were shared with her cardiologist, who supported performing future dental procedures in the dental chair in a hospital setting. The cardiologist proposed that the amount of exogenous epinephrine from the local anesthesia (**LA**) solution was negligible when compared with the endogenous epinephrine release from anxiety, so the use of epinephrine-containing LA was not contraindicated.

On the day of dental treatment, however, the patient's mother expressed serious concerns about the use of LA with epinephrine and was afraid "that her child might have a heart attack and die." Under these circumstances, the dentist felt it prudent to administer 3% mepivacaine plain (Carbocaine Cook-Waite Carestream Dental, Rochester, N.Y.) to reduce both the parental and the patient anxiety. Since the main objective was to complete treatment safely with particular emphasis on anxiolysis, parental absence was an additional behavioral management technique that was crucial to achieving a successful outcome.

Following informed consent, oral administration of 75 mg hydroxyzine, 40% nitrous oxide, and LA infiltration of 153 mg (equivalent to 3 carpules) of 3% mepivacaine plain, dental restorations were successfully completed under rubber dam isolation. Although the patient is on a 3-month recall/preventive regime, compliance remains a challenge.

DISCUSSION

In managing a CPVT patient, it is important to take active measures to reduce stress and anxiety for patients and their family. Achieving profound anesthesia is an important part of alleviating stress and pain both during and after any dental procedure. In spite of this, the use of epinephrine-containing LA for dental procedures in certain cardiac-compromised patients remains controversial. The use of epinephrine in LA has several advantages. By causing local vasoconstriction, epinephrine increases the duration and depth of anesthesia, provides hemostasis, and decreases the systemic toxicity of LA solution.[7] The benefits gained from adding epinephrine to a local anesthetic, however, should be weighed against the risks.

According to Malamed,[7] plasma levels of epinephrine double from resting levels after 1 cartridge of 2% lidocaine with 1:100,000 epinephrine. This linear elevation in plasma epinephrine is dose-dependent and may persist from a few minutes to about half an hour. Contrary to a previously held position that the intraoral administration of usual volumes of epinephrine produces no cardiovascular response, and that patients were more at risk from endogenously released epinephrine than they were from exogenous epinephrine, published evidence has proven otherwise. Venous plasma concentrations associated with the injection of 100 to 150µg of epinephrine using 2% lidocaine with 1:100,000 epinephrine, as often occurs in oral and periodontal surgery, produces concentrations equivalent to that present during heavy exercise.[8] These levels of plasma epinephrine can lead to moderate increases in cardiac output and stroke volume.

Even with aspiration and slow injection during the administration of LA for dental procedures, a sufficient amount of epinephrine can be absorbed into the bloodstream, producing sympathomimetic reactions such as tachycardia, sweating, and palpitations.[7] Little et al.[9] postulated that the use of local anesthetics containing epinephrine could precipitate tachycardia and arrhythmias in cardiac patients. In this case, epinephrine-containing LA was avoided due to the concern expressed by the patient's mother, which was taken into consideration given its potential to increase patient anxiety and promote endogenous catecholamine release, thereby posing a significant risk of a tachycardia. In weighing the risks and benefits of using epinephrine-containing LA for patients with CPVT, it is recommended that dental practitioners always consult with the patient's cardiologist and consider the anxiety of the individual patient regarding its use. Furthermore, dentists must ensure that patients with CPVT are in strict compliance with their beta-blockers before providing dental treatment. A long-term follow-up study by Hayashi et al.[4] on 101 affected patients found that fatal and near-fatal cardiac events still occurred in a third of patients on beta-blockers. According to those authors, missing even a single dose can provoke lethal arrhythmias.

The American Heart Association published guidelines on the dental management of patients with structural heart defects; however, given that heart is structurally normal in CPVT, antibiotic prophylaxis is not required since the risk of infective endocarditis is negligible.[10]

Familial PVT is caused by a mutation of the myocardial isoform of RyR2. Mutations of the corresponding gene in skeletal muscle ryanodine receptor 1 (RyR1) predispose its carriers to malignant hyperthermia when exposed to halogenated anesthetics or succinylcholine.[11] Swan et al.[11] reviewed case histories of 30 patients with familial PVT and found that succinylcholine and volatile anesthetics did not have a clinically significant effect on RyR2 defects. In spite of the relatively low risk of malignant hyperthermia from GA in these patients, it was crucial to consider the cost of repeated GA in view of this patient's high caries risk. Regular consultation with her cardiologist was beneficial in considering future treatment in the dental chair, albeit in the hospital.

In conclusion, when providing dental treatment for patients with CPVT, it is imperative that dental practitioners gather a thorough medical history to identify the

patient's cardiac risk and pay careful attention to their anxiety levels. In spite of the issues presented in this paper, the greatest challenge with our patient remains the poor compliance with preventive recommendations, resulting in a continual risk of dental disease in an anxious patient with a demanding parent.

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