Sudden Cardiac Death after Induction of General Anesthesia in a Patient with Undiagnosed Williams Syndrome

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Introduction

Williams Syndrome is a complex syndrome characterized by craniofacial dysmorphic features, developmental delay, and cardiac abnormalities. Sudden death has been described for this syndrome during induction of anesthesia, surgical procedures, and cardiac catheterizations. We present a case of sudden cardiac death refractory to resuscitative efforts in a previously healthy infant with undiagnosed William’s Syndrome undergoing an elective surgical procedure.

Case Presentation

A 2 month-old, full-term 4.8 kg male infant presented for elective inguinal hernia repair. His medical and birth histories were remarkable only for an episode of brushtwo weeks prior. There was no history of anesthetic complication in the family. The patient was brought to the operating suite and a smooth mask induction was performed using sevoflurane after application of standard ASA monitors. A 22g angiocath was placed and 2 mg/kg of propofol was administered to facilitate a smooth intubation.

Approximately 5 minutes later, the patient developed a wide-complex tachycardia with a heart rate of approximately 150 beats per minute. The patient became hemodynamically unstable with loss of palpable pulses. PALS resuscitation was initiated including several rounds of epinephrine, cardioversion, and amiodarone with persistent tachycardia. An arterial line was placed, and several arterial blood gases were obtained showing a worsening metabolic acidosis attributable to hypoperfusion but without significant electrolyte abnormalities. CPR was continued until the initiation of ECMO approximately one hour later for a return of adequate perfusion. A brief echo done immediately prior to cannulation by cardiology showed poor cardiac function and supported the need for ECMO. The patient was then transported to the cardiac intensive care and more extensive workup could be undertaken.

Echocardiography showed moderate-to-severe systolic function and a hypokinetictachycardia. A diffusely hypoplastic and thick ascending aorta suggestive of supravalvular aortic stenosis was also appreciated. The patient was placed on neurointervention and continued on ECMO while awaiting recovery of heart function. The patient unfortunately suffered a series of worsening seizures and had little appreciable improvement in cardiac function. A head CT demonstrated devastating diffuse hypoxic ischemic injury and the family opted to withdraw care. Cardiac arrest from suspected coronary ischemia in the setting of supravalvular aortic stenosis was highly suggestive of William’s syndrome and a cytogenetics panel was sent with consent of the parents. In-situ coronary angiography and echocardiography showed severe, long-segment supravalvular aortic stenosis (black arrow) and marked dilation of the coronary arteries (black arrow). Reproduced from Salahuddin et al

Discussion

Case reports of sudden death in Williams syndrome patients date back to the first published cases in 1961; however, there does not appear to be a significant amount of literature on Williams syndrome and sudden cardiac death with anesthesia in this age group. Anesthesia-related deaths due to cardiac arrest in WS have also been reported. The sudden and rapid downhill course as well as the lack of response to resuscitation seem to be almost pathognomonic in these deaths.

Coronary artery abnormalities are often associated with congenital supravalvar aortic stenosis. Some investigators have recommended that every patient with WS should be considered at risk for myocardial ischemia. Coronary artery involvement in patients with WS may manifest as coronary ostial stenosis, diffuse coronary artery stenosis, coronary artery dilation, or obstruction to coronary artery inflow by the aortic valve, the sinotubular ridge, or a combination of both. It has been reported in a review of congenital supravalvar aortic stenosis that impaired coronary blood flow has been reported frequently in these patients.

Although some of these cases have been reported following hypotension and bradycardia, there are reports of sudden cardiac death with stable hemodynamics as well. Anesthetic regimens involving high-dose opioids similar to those employed for adult patients with severe coronary disease may be of benefit. It is suggested by many authors that pre-surgical testing should include a minimum 12-lead ECG and echocardiogram with some authors suggesting coronary angiography to evaluate for abnormalities. However, as this procedure itself would require general anesthesia for children, it is of questionable utility in this population. In addition, deaths have occurred when the coronary arteries appear normal and when echocardiography demonstrates seemingly minor pathology.

Ultimately, a strong clinical suspicion by the anesthesiologist of an underlying syndrome is necessary to help prevent anesthetic risk in a child with undiagnosed Williams Syndrome. Facial morphology and behavioral clues may help, although these are much less pronounced or even absent in infants and small children. The risk of death with any general anesthetic must be respected in this population, particularly for elective procedures.

References