CASE REPORT

Postoperative Cardiorespiratory Arrest in Down Syndrome

David Cha[1]*, Stella Pak[2], Yan Yatsynovich[2]

A 24-year-old male with Down Syndrome (DS) presented with 1-day duration of right upper quadrant pain, nausea, and vomiting. He was timely diagnosed with acalculous cholecystitis and treated with laparoscopic cholecystectomy. His immediate postoperative period was complicated by acute onset bilateral pulmonary edema and subsequent respiratory failure which triggered a type II myocardial infarction. Patient's course was further complicated with acute metabolic acidosis and acute kidney injury resulting in continuous veno-venous hemodialysis. Family members collectively decided to transition patient to comfort care. Over the next few hours the patient expired from cardiogenic shock as no further vasopressor support was administered.

Discussions: It is well known that patients with DS rarely suffer from coronary artery disease. In this case, we believe the patient's residual right ventricular dysfunction from a previous significant ventricular septal defect partially contributed to the patient's decline. Furthermore, findings in previous reports describe general anesthesia and sevoflurate's role in possibly inducing cardiac arrest due to autonomic dysfunction. Due to low cardiopulmonary reserves, DS patients are at higher risk of complications than the normal patient population. Considering many patients with DS either have existing or repaired structural anomaly, perioperative assessment and vigilant monitoring may help reduce cardiopulmonary complications in DS patients.

Keywords: down syndrome; myocardial infarction; MI; cardiac arrest; postoperative; sevoflurate; perioperative; general anesthesia; complications; young adult

Down syndrome (DS) is one of the most common birth defects in the United States as the national prevalence is approximately 14 per 10,000 live births. Congenital heart diseases are present in about 40%-60% of children with DS [1-3]. In the setting of an atrioventricular canal defect, there is a common association with a cleft mitral valve due to the lack of a common atrioventricular junction [4-6]. Although heart defects are common in DS patients, coronary artery disease and myocardial infarctions (MI) are thought to be rare [7-8].

In DS patients undergoing procedures, bradycardia and hypotension have been reported with use of sevoflurane and general anesthesia (GA) [9]. The sympathetic nervous system dysfunction induced by sevoflurane and GA may also be an accelerating factor for cardiac arrest in these patients [10]. Herein, we report a case of myocardial infarction in a young DS patient with repaired congenital heart defect.

Case Report

A 24-year-old male with history of Down syndrome complicated by atrioventricular canal defect and cleft mitral valve, presented to our facility with 1-day duration of right upper quadrant pain, nausea, and vomiting. His additional past medical history included a history of stroke, prior myocardial infarction, prolonged QT syndrome, and obstructive sleep apnea. He had surgical correction of ventricular septal defect with remnant right ventricular enlargement from prolonged pressure overload. Initial vital signs included a hypotension of 80/54 mmHg, tachycardia of 100/min, tachypnea of 20/min, temperature of 37.3 °C, and oxygen saturation of 100 % on room air. He was promptly diagnosed with acalculous cholecystitis and treated with laparoscopic cholecystectomy.

Laparoscopic cholecystectomy was successfully performed without any intraoperative complication, but his immediate postoperative period was complicated by acute onset bilateral pulmonary edema and subsequent respiratory failure. It appeared that the physiological stress from surgery and respiratory failure had triggered a type II myocardial infarction, which in turn, led to an acute cardiogenic shock with right ventricular failure and resultant cardiopulmonary arrest (pulseless electrical activity) of approximately 10 minutes of cardiopulmonary resuscitation (CPR) was performed with 4 doses of epinephrine and successful return of spontaneous circulation. Hypothermic protocol was initiated promptly. His Troponin I collected in 3-hour intervals, peaked at 64.4 ng/mL. He was subsequently intubated for ventilatory support. Because of his cardiogenic shock, epinephrine drip was also started. Transthoracic echocardiography demonstrated markedly dilated right ventricle with severe systolic dysfunction. Pulmonary artery and central venous pressure were estimated to be 62 mmHg

From the [1] Department of Medical Education, Wright State University Boonshoft School of Medicine, Dayton, OH, USA
[2]Department of Internal Medicine, Kettering Medical Center, Kettering, OH, USA
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*Corresponding author: David Cha, Department of Medical Education, Wright State University Boonshoft School of Medicine, Dayton, OH, USA. E-mail: cha.4@wright.edu

and 20 mmHg, respectively.

His post-arrest course was further complicated with acute onset of metabolic acidosis with pH of 7.1 and acute kidney injury with creatinine rapidly rising from 0.9 to 5.6 mg/ dL. In the light of critically ill condition, he was placed on continuous veno-venous hemodialysis. He also developed intermittent myoclonic movement in the distal portion of upper and lower extremities.

In the view of progressive deterioration and poor prognosis, the family members collectively decided to shift goal of care from survival to comfort measures. In accordance with the family's decision, no further vasopressor support was given. Subsequently, the patient deteriorated over next few hours and expired from cardiogenic shock.

Discussion

It is well known for patients with DS to be without atherosclerosis, lowering their chances of coronary artery disease [8]. This patient had a history of significant VSD leading to Eisenmenger syndrome. Although he had surgery to close the VSD, his right ventricle remained dilated with residual right ventricular dysfunction, which we believe partially contributed to his rapid deterioration. This case highlights that DS patients with repaired congenital heart defects are still at an increased risk for cardiac events.

Findings in previous reports show that GA and sevoflurane may induce cardiac arrest in DS patients due to autonomic dysfunction, leading us to believe this patient's recent anesthesia induction played a role in expediting further decline [9]. Cardiogenic shock likely has developed due to his low preload from underlying right ventricular anomaly.

Autonomic dysfunction and anatomic anomaly predispose DS patients to development of cardiac complications from even low to moderate risk procedures. Due to rarity of DS patients, the incidence rate of cardiorespiratory complication in perioperative period has not been reported; however, perioperative cardiopulmonary complication is known to occur far more frequently in DS in comparison to general population [11]. Incorporating DS as a risk factor in perioperative assessment process might help reduce the cardiac complication of DS patients. Due to low cardiopulmonary reserve, DS patients also experience rapid deterioration from complications that would have not been detrimental in patients without pre-existing comorbidities. Thus, vigilant perioperative monitoring and thorough assessment are warranted to minimize the risk of cardiopulmonary complication in patients, regardless of the general risk associated with the procedure.

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