Intraoperative Sudden Cardiac Death In Pediatric Patients - Calamity yet to Overcome?

We read the case report entitled - "Cardiac Arrest after induction of Anesthesia in a two-month old Infant with Undiagnosed Williams syndrome-Case report and comprehensive review'', in this issue.[1]

The authors have discussed Williams syndrome in detail. The risk of sudden cardiac death in patients with Williams syndrome increases manifold, and the causes are myocardial ischemia and cardiomyopathy.[2,3] In this case, the infant had preoperatively hypertension for the age which was an alarming finding but got neglected. Thorough investigation for this specific finding could have given diagnosis of the patient before elective bilateral congenital hernia reparative surgery and intraoperative death could be prevented. This child diagnosed later having supravalvular aortic stenosis which is a fixed cardiac output state. Anesthetic agents which reduce systemic vascular resistance and reflex tachycardia have detrimental effects on cardiac output, diastolic blood pressure, and coronary perfusion in such patients of fixed cardiac output state. Hence, the goals of anesthesia induction should be for maintaining systemic vascular resistance and avoidance of tachycardia. As authors could not diagnosed the disease, routine induction of anesthesia with their vasodialatory effects was not tolerated by this child.

Many such syndromic congenital heart diseases are associated with extracardiac anomalies and require early reparative procedures under anesthesia. Such patients may not show all specific signs or symptoms of the syndromes and can go undiagnosed. These patients have constant threat of sudden fatal cardiac events during perioperative period.

The prevalence of cardiac arrhythmias in pediatric population is 5%.[4] These diseases can present anytime from birth to 15 years of age and their recognition requires high index of suspicion.[5] Common syndromes with arrhythmic risks can go undiagnosed because of the absence of structural heart disease. These are as follows:

1. Wolff–Parkinson–White (W-P-W) syndrome: It is the syndrome with preexcitation and tachyarrhythmias due to atrioventricular accessory pathway.[6,7] Incidence of Sudden cardiac death events in this condition is 0.15 to 0.39 % in 10 year follow up study.[8] Atrioventricular reentrant tachycardias is the most common arrhythmia in this syndrome. Atrial fibrillation is potentially fatal and leads to ventricular fibrillation

The incidence of W-P-W syndrome associated with Ebstein's anomaly is 10%.[9]

Diagnostic features of W-P-W syndrome on electrocardiography (ECG) are[2] short PR interval (<0.12 s),[3] prolonged QRS complex (>0.12 s),[4] slow rising, and slurred QRS complex known as delta waves.

2. Brugada syndrome: This syndrome occurs because of genetic mutation in sodium channel.[10] Sudden cardiac death in this syndrome is 4% under 40 years of age. Diagnosis is made by only ECG. ECG findings are–[2] atypical right bundle branch block, polymorphic ventricular tachycardia (VT), and cove-shaped ST elevation in leads V1–V3 in the absence of structural heart disease. And so, it may go undiagnosed if we do not do preoperative ECG in infants[11]
3. Long QT syndrome: This is a myocardial repolarization disorder with prolonged QT interval (>0.46 s) and ventricular fibrillation triggered by Torsades de pointes. Torsades de pointes is sinusoidal twisting of QRS axis around isoelectric line of ECG.[12] Only 60% patients manifest with the symptoms such as syncope, seizure, and cardiac arrhythmia. It is a potassium channelopathy disorder. Other potassium channelopathy syndromes are Romano–Ward syndrome, Jervell and Lange-Nielsen syndrome, Andersen–Tawil syndrome, and Timothy syndrome.[13] Sudden Cardiac death occurs in 9% of patients of long QT syndrome
4. Lenegre's disease: In this syndrome, progressive conduction disorders occur due to genetic mutation in SCN 5A causing idiopathic ventricular fibrillation
5. Arrhythmogenic right ventricular (RV) dysplasia: Sudden cardiac death occurs before 5 years of age due to intractable VT in this syndrome. This is a familial and autosomal dominant inheritance disease with nonspecific clinical features which cannot be diagnosed easily. This condition constitutes structural, functional, and electrophysiological abnormalities of heart. Fibrofatty deposition in RV mass makes it paperthin (by replacing myocardium) and dysfunctional. The International Task Force has revised the criteria for diagnosis of this condition with 2D-echo, magnetic resonance imaging, and RV angiographic parameters[14]
6. Sengers syndrome: It is genetic disease combination of congenital cataract, hypertrophic cardiomyopathy (HOCM), and lactic acidosis after exercise. In these patients, bradycardias are resistant to atropine. Sudden death occurs in the 1st year of life because of cardiac failure.[15]

The list of syndromes with congenital cardiac and extracardiac anomaly is unending. DiGeorge syndrome with tetralogy of Fallot, HOCM, LEOPARD syndrome, sick sinus syndrome, and many congenital myopathic syndromes are associated with sudden cardiac death under anesthesia.

Cardiac arrhythmia can affect the quality of life and can lead to morbidity and mortality in infants and children. Vigilant preoperative assessment can give proper diagnosis and treatment. These diseases can be treated with pharmacotherapy, antiarrhythmic therapies such as drugs, pacing, radioablation, or surgical cardiac treatment before noncardiac procedures. Multidisciplinary approach by anesthesiologist, pediatrician, cardiac surgeon, and cardiologist plays a pivotal role for safe and successful management of such patients.

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