Perioperative Management of Refractory Chaotic Atrial Tachycardia as a Manifestation of Costello Syndrome

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CASE DESCRIPTION

A 13 month old, 6.3 kg female with Costello syndrome presented with bilateral hip subluxation.

Cardiac History:
- In Utero Diagnosis of Atrial Tachycardia
- After full term delivery, required propranolol and amiodarone for rate and rhythm control, diagnosed with chaotic atrial tachycardia (Fig. 1)
- Hospitalized at 1 month and 3 months old for cardiogenic shock secondary to breakthrough tachyarrhythmias
- Diagnosed with Costello Syndrome (Fig. 2)

Additional Past Medical History:
- Global developmental delay, Failure to thrive
- Diffuse hypotonia, ligament laxity
- Nystagmus, estropria

Past Surgical History: G-tube placement at age 7 months

Medications:
- Amiodarone 8.4mg/kg/day (dosed BID)
- Propranolol 1.9mg/kg/day (dosed TID)
- Flecainide 3.8 mg/kg/day (dosed TID)
- Prevacid, D-V-Sol

Pre-operative Testing:
- Echocardiogram: Mild LV dilation, mild decrease in LV systolic function, significant atrial arrhythmia
- Holter Monitor: Wandering atrial pacemaker, atrial ectopy is underestimated, frequent isolated PACs-multiple P wave morhologies, sometimes conducted aberrantly, atrial couplets, rare non-sustained episodes of atrial tachycardia.

Upon OR arrival, the atrial rhythm was stable at 130 bpm. ASA monitors were placed, and mask induction was with sevoflurane/nitrous oxide. IV access was obtained, Fentanyl 1mcg/kg was administered, and the child was intubated. Apparent high grade AV dissociation soon occurred. Due to the history of severe tachyarrhythmias leading to cardiogenic shock, vagolytics and sympathetic agonists were not immediately administered. Volatile anesthetic was changed to isoflurane, volume resuscitation was initiated with crystalloid and albumin, and PR intervals stabilized to baseline. Procedure time was 1 hour, and extubation was without incident.

DISCUSSION

Costello Syndrome, while still extremely rare, is being reported and diagnosed with increased frequency. To safely care for these children we must be prepared to address all manifestations of the disease. While most children with cardiac lesions will have stable structural defects, progressive disease, HOCM, and dysrhythmias may occur. A preoperative EKG, 24-hour Holter monitor, and echocardiogram are all indicated (3). A link between supraventricular tachycardias and sudden death in patients with Costello Syndrome has been suggested, but sufficient case numbers do not yet exist to show a statistical correlation (4).

Sevoflurane induced high grade AV block has been reported in children who have underlying conduction abnormalities (5), and even in healthy children (6). While sevofluranes effects on SVR and preload are well appreciated, its direct effects on the AV node and refractoriness are often overlooked (7). In the setting of significant conduction system abnormalities, and with a child on multiple antiaarrhythmics, rhythm disturbances should be anticipated. The decision to conservatively treat AV block in a well perfused patient will be controversial, and must be made in a clinical context.

Figure 1 (right): Chaotic atrial tachycardia, characterized by irregular atrial rate, multiple P waves morphologies, irregular P-P intervals, and isoelectric baseline between P wave

Figure 2 (left): Costello Syndrome, also known as faciocutaneouskeletal (FCS). Facial features include large mouth with thick lips, low nasal bridge, full nasal tip, downward slanting palpebral fissures, and forehead is sometimes hairy (1, 2).

Figure 3: Constellation of manifestations associated with Costello Syndrome

LITERATURE CITED