[NM-182] Perioperative Management of Refractory Chaotic Atrial Tachycardia as a Manifestation of Costello Syndrome

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Introduction

Costello Syndrome is a rare chromosomal abnormality characterized by coarse facial features (Fig. 1), developmental delay, musculoskeletal abnormalities, and a variety of cardiac abnormalities, including interconnected structural and conduction system defects which complicate perioperative management.(1) We report a case of an child with Costello Syndrome complicated by refractory chaotic atrial tachycardia presenting for bilateral hip reduction.

Case Scenario

A 13 month old, 6.3 kg female with Costello syndrome presented with bilateral hip subluxation. She was born full term, with a diagnosis of atrial tachycardia in utero. Within days of birth she required propranolol and amiodarone for rate and rhythm control, and was diagnosed with chaotic atrial tachycardia (Fig. 2). She was discharged, but returned at age 1 month and 3 months in cardiogenic shock secondary to breakthrough tachyarrhythmias. A diagnosis of Costello syndrome was reached; she was stabilized on propranolol, amiodarone, and flecainide, and arrived at 13 months for bilateral hip reduction. Pre-operative evaluation included an echocardiogram, and holter monitor reading the day before surgery.

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. Complete AV block ensued, with ventricular rate of 60 bpm, and systolic BP 60 mmHg. Due to the history of severe tachyarrhythmias leading to cardiogenic shock, vagolytics and sympathomimetics were not immediately administered. Volatile anesthetic was changed to isoflurane, volume resuscitation was initiated with crystalloid and albumin, and PR intervals stabilized. Procedure time was 1 hr, and extubation was uneventful.

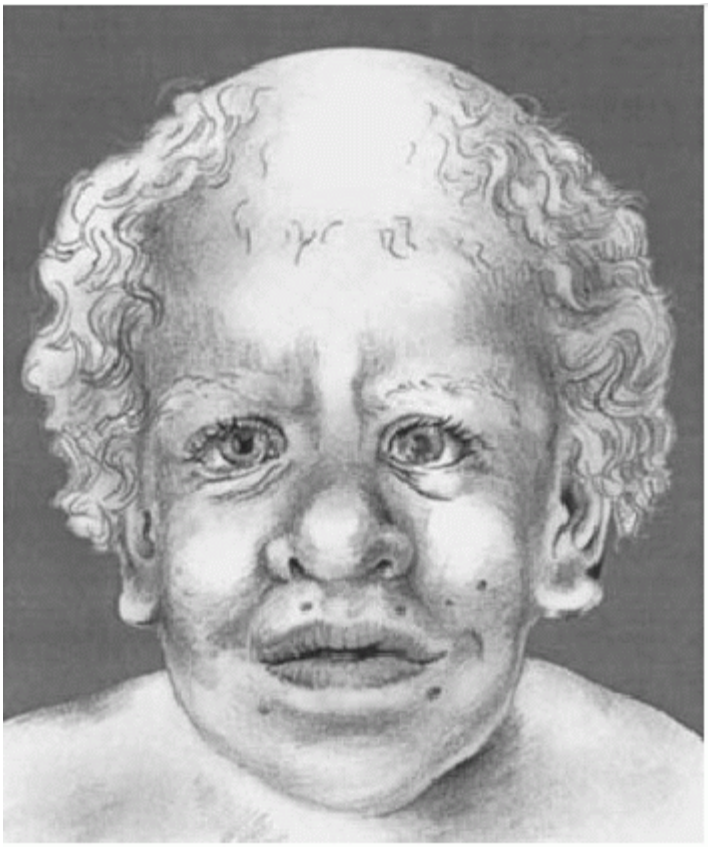
Discussion

Costello Syndrome, while extremely rare, is being reported and diagnosed with increased frequency, and to provide safe care we must address all manifestations of the disease. While most children with cardiac lesions will have stable structural defects, progressive disease, HOCM, and dysrhythmias occur. Preoperative EKG, 24-hr Holter monitor, and echocardiogram are indicated.(2) A link between SVTs and sudden death in patients with Costello syndrome has been suggested.(3)

References

1)Hennekam RC. Costello Syndrome: an overview. Am J Med Genet C Semin Med Genet. 2003 Feb 15;117C(1):42-8.

2)Katcher K, et al. Anesthetic implications of Costello syndrome. Paediatr Anaesth. 2003 Mar;13(3):257-62.
3)Siwik ES, et al. Cardiac disease in Costello syndrome. Pediatrics. 1998 Apr:101(4 Pt1):706-9.



Vent. rate	179	BPM	** ** ** ** * Pediatric ECG Analysis * ** ** ** **
PR interval	*	ms	Chaotic atrial tachycardia with aberrant IV conduction
QRS duration	72	ms	Right axis deviation
QT/QTc	182/314	ms	QTc difficult to measure
P-R-T axes	* 171	38