Anesthesia for a Child with Sengers Syndrome

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INTRODUCTION

- Sengers syndrome is a metabolic disorder
- Patients present with hypertrophic cardiomyopathy, mitochondrial myopathy, lactic acidosis with exercise, and congenital cataracts.
- Gene mutation of mitochondrial aglycerol kinase
- Only about 40 patients ever diagnosed with the disease
- Disease onset and severity varies greatly

CASE PRESENTATION

A ten year old male with a history of Sengers syndrome presented for electrophysiology study and ablation of Wolff-Parkinson-White accessory pathway. He was active despite his mitochondrial myopathy with no history of syncope. He did not have an implantable defibrillator as the parents declined to have one placed at this time.

Electrocardiogram showed sinus rhythm, biventricular hypertrophy and short PR interval.

Transthoracic echocardiography showed severe septal hypertrophy, systolic anterior motion of mitral valve, and ejection fraction 79%.

Parents reported no anesthetic complications with his prior dental exam and cataract surgery.

DISCUSSION

- All medications including beta-blocker, coenzyme Q and levocarnitine continued, clear liquids until 2 hours prior to procedure
- Awake IV placed with oral midazolam and topical anesthetic
- Standard ASA monitors, defibrillation pads, arterial line, BIS monitor
- Induction with ketamine, remifentanil, rocuronium
- Endotracheal intubation
- Maintenance with dexmedetomidine and remifentanil
- D10 maintenance fluid infusion
- At conclusion of procedure, no NMDB reversal needed, extubated, same day discharge

ANESTHETIC MANAGEMENT

- Volatile agents and propofol avoided due to mitochondrial disease
- Arterial line allowed for hemodynamic monitoring and close monitoring of acid/base status and glucose levels
- Maintain adequate preload and afterload given HOCM and SAM
- In the event of SVT, avoid AV nodal blocking agents due to WPW

REFERENCES