A case of Costello Syndrome

Stanlies D’Souza MD
Baystate Health, dsouzastan@yahoo.com

Follow this and additional works at: https://scholarlycommons.libraryinfo.bhs.org/all_works
Part of the Medicine and Health Sciences Commons

Recommended Citation
INTRODUCTION
We present a case of the anesthetic management of a patient with Costello syndrome. Costello syndrome is a rare autosomal dominant faciocutaneoskeletal syndrome which has important anesthetic implications.

CASE DESCRIPTION
An eight year old boy with a rare genetic disorder of Costello syndrome presented for heel cord lengthening for bilateral club feet. The patient had moderate hyperdynamic, hypertrophic left ventricular outflow tract obstruction. The patient also presented with a DDD pacemaker for complete AV block following left ventricular outflow tract myectomy and ablation of intracardiac anomalous pathway. The patient was induced by mask inhalation, a peripheral intravenous line was obtained, direct laryngoscopy showed grade 4 Cormack-Lehane view. The airway was secured with an endotracheal tube using a fiberoptic bronchoscope. He had an uneventful general anesthesia course with no perioperative complications.

Our patient did present with all of the typical faciocutaneoskeletal features of Costello syndrome (see discussion) along with hypertrophic cardiomyopathy and a previous known difficult intubation.

REFERENCES

DISCUSSION
Costello syndrome is a rare (estimated 200-300 cases worldwide) autosomal dominant syndrome associated with:
• mental retardation
• coarse facies
• macrocephaly
• redundant skin especially of hands and feet
• unusually flexible joints
• gastrointestinal reflux.

This syndrome has been known to be associated with cardiac abnormalities especially hypertrophic cardiomyopathy and structural heart defects.

Some of these structural heart defects include:
• hypertrophic cardiomyopathy
• idiopathic subaortic stenosis
• asymmetric septal hypertrophy
• valvar pulmonic stenosis
• aortic dilation
• atrial tachycardia.

Securing the airway may pose a challenge due to the following characteristics associated with this syndrome:
• short neck
• macroglossia
• hypertrophied tonsillar and supraglottic tissue
• laryngeal papilomata
• choanal atresia.

In addition, a cardiac evaluation is helpful in the perioperative management of these patients. Hemodynamic goals of hypertrophic cardiomyopathy are to maintain preload and afterload, and avoid tachycardia to prevent left ventricle outflow tract obstruction.

CONCLUSION
One should be aware of clinical implications of rare genetic diseases and associated anesthetic risks and complications like Costello syndrome.