One lung ventilation in a patient with Seckel Syndrome

Aimen Houni MD
Baystate Health, aimen.houni@baystatehealth.org

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

This Presentations, Research is brought to you for free and open access by Scholarly Commons @ Baystate Health. It has been accepted for inclusion in All Scholarly Works by an authorized administrator of Scholarly Commons @ Baystate Health.
One Lung Ventilation in a Patient with Seckel Syndrome
Aimen Houni MD, Matthew Harris DO, Toni Chahla MD, Stanlies D’Souza MD
Department of Anesthesiology, Baystate Medical Center/ Tufts University School of Medicine, Springfield, MA

INTRODUCTION
A patient with Seckel syndrome, with dysmorphic features likely presents with difficult mask ventilation and difficult intubation with direct laryngoscopy. We report a patient with such syndrome who needed one lung ventilation.

CASE DESCRIPTION
A 58 years old female with Seckel syndrome and severe mental retardation presented with a persistent cough and bilateral pulmonary nodules. She was scheduled for right video-assisted thoracoscopic lung biopsy. Pertinent physical exam findings included: a nonverbal patient with low-set ears and dwarfism (52 inches & 27kg), a narrow and receding mandible, prominent nose and eyes. Face mask induction was performed maintaining spontaneous ventilation and intravenous access was obtained. A 28 French double lumen tube was placed using a glide scope and position was confirmed with a flexible fiberoptic bronchoscope. Surgery was performed with an uneventful postoperative course.

DISCUSSION
Seckel syndrome, also known as bird headed dwarfism, is a rare autonomic recessive disorder with an approximate incidence of less than 1;10,000.1 These patients display severe growth retardation, microcephaly and mental retardation. Hematological and cardiovascular abnormalities have been reported in many patients with Seckel syndrome. Malignant hypertension, atrioventricular canal defect, tetralogy of Fallot, anemia, and pancyopenia are some of the other conditions described.1,2,3 As per published case reports, micrographic, receding jaw and crowded dentition can make direct laryngoscopy difficult, while beak like protruding nose can make mask ventilation difficult.1

When compared to bronchial blockers, double-lumen tubes (DLT) have been shown to provide faster lung isolation and need far fewer repositionings.5 Placing a DLT under direct laryngoscopy may prove very difficult in these patients. Video laryngoscopes or flexible fiberoptic bronchoscopes (FFB) can be used to place a DLT in the setting of a difficult airway, but smaller DLT’s (28 French) will not accommodate even a pediatric FFB.

Placing a single lumen tube, then replacing it with a DLT over an exchange catheter is also an option. However, in patients of extremely small stature even the smallest available DLT may be too large. Age and weight have shown to be poor predictors of appropriate endotracheal tube size6 and larger DLT may result in airway damage. When a DLT is not an option a bronchial blocker may be necessary to achieve lung isolation, but this technique also carries a risk of injury such as bronchial rupture due to balloon overinflation.

CONCLUSION
Understanding the constellation of features present in Seckel syndrome and their impact on airway management is crucial for practitioners approaching these cases. With proper preparation and anticipation of a difficult airway even a DLT can be safely placed in these patients.

REFERENCES

Figure 1 Facial profile of a 5 year old boy with Seckel syndrome.7 ©Arora S, Ghi B, Rattan V. Journal of Anaesthesiology Clinical Pharmacology.