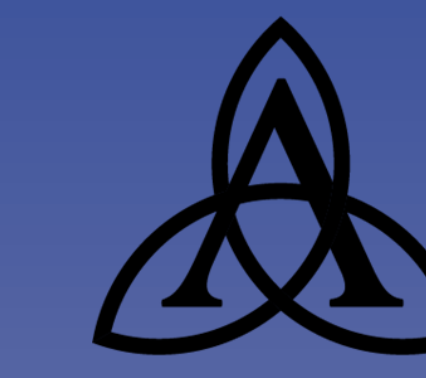


# A Rare and Difficult Case in Anesthesia: Moebius Syndrome



College of Osteopathic Medicine  
MICHIGAN STATE UNIVERSITY

Jason Greib MS-3, Michael Danic DO  
Ascension Genesys Hospital  
Grand Blanc, MI, USA



Ascension  
Genesys Hospital

## Learning Objectives

1. Understand what Moebius Syndrome is and the various abnormalities associated with the condition.
2. Recognize how features of Moebius Syndrome can provide difficulties in anesthetic care.
3. Identify steps and strategies that can be taken in order to minimize anesthetic risk in patients with Moebius Syndrome.

## Introduction

- Moebius Syndrome is a rare congenital neurologic condition that is estimated to affect between 2 to 20 newborns per 1 million births.<sup>1</sup>
- The classic feature of the condition is paralysis of multiple cranial nerves, primarily the 6<sup>th</sup> and 7<sup>th</sup> cranial nerves. The 5<sup>th</sup>, 8<sup>th</sup>, 9<sup>th</sup>, 10<sup>th</sup>, 11<sup>th</sup>, and 12<sup>th</sup> cranial nerves can also be involved though less commonly.
- In addition, many born with Moebius Syndrome suffer from craniofacial malformations including but not limited to micrognathia, microstomia, and dental abnormalities. Congenital cardiac anomalies and orthopedic malformations may also be seen.
- The combination of cranial nerve deficits and craniofacial malformations creates many potential difficulties in anesthetic care that must be given special consideration as demonstrated by this case.

## Case

- **56-year-old male with a history of Moebius Syndrome presented with complaints of sharp epigastric abdominal pain and obstipation for 3 days.**
- PMHx: HTN, HLD, Anxiety, IBS, Diverticulosis, Sleep Apnea, Moebius Syndrome
- PSHx: Adenoidectomy, Uvelectomy, Skin Graft - Right Jaw Recipient
- Social Hx: Denies EtOH, illicit drugs, tobacco use
- Meds: Atenolol, Rosuvastatin, Gemfibrozil, Omeprazole, Alprazolam, Hydrocodone/Acetaminophen, Gabapentin, Dicycloverine
- Allergies: NKDA
- **Imaging: CT Abdomen/Pelvis showed high-grade small bowel obstruction to the level of the distal Jejunum.**

ANESTHESIA PHYSICAL EXAM	
Vital Signs: HR 21, BP 120/80, RR 18, Temp 98.2	
Heart: <i>ran</i>	Lung: <i>CTU</i>
Airway Class: I II III IV (IV)	
<input type="checkbox"/> Check box if airway WNL	<input checked="" type="checkbox"/> Check box if patient obese (BMI > 30)
<input type="checkbox"/> Check box if patient underweight (BMI < 19)	
ROM Neck: <input checked="" type="checkbox"/> Full <input type="checkbox"/> Limited	Teeth: <input type="checkbox"/> Good <input checked="" type="checkbox"/> Chipped
<input type="checkbox"/> Edentulous	<input checked="" type="checkbox"/> Poor
<input type="checkbox"/> Caps	<input type="checkbox"/> Dentures
Neurologic: <input checked="" type="checkbox"/> Alert and Oriented X3	<input checked="" type="checkbox"/> No Gross Deficits
For Anesthesia Provider: ASA Physical Status 1 2 3 4 (3)	Plan: <input checked="" type="checkbox"/> General <input type="checkbox"/> MAC <input type="checkbox"/> Local <input type="checkbox"/> Epidural <input type="checkbox"/> Block (Type)
Check all tests reviewed: <input type="checkbox"/> N/A <input checked="" type="checkbox"/> Labs	<input type="checkbox"/> EKG <input type="checkbox"/> Chest X-ray <input type="checkbox"/> Medical Clearance <input type="checkbox"/> Cardiac Clearance

Figure 1: Pre-op evaluation

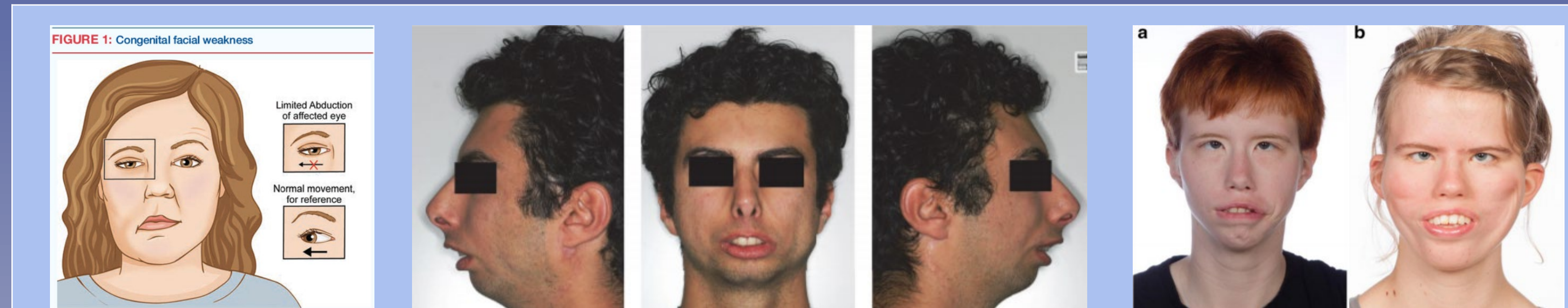


Figure 2: Examples of Moebius Syndrome craniofacial features<sup>1,2,3</sup>

## Pre-operative Anesthesia Plan

- **Mallampati class IV with potential for a difficult airway, perioperative aspiration, and high risk for exposure keratopathy.**
- Patient presented with a very small oropharyngeal opening due to micrognathia, poor dentition, and paralysis of the 5<sup>th</sup> and 7<sup>th</sup> cranial nerves.
- Awake fiberoptic intubation was planned in expectance of a difficult intubation via direct laryngoscopy.
- Avoid copious airway secretions and aspiration with use of a pre-operative antisialagogue.
- Ensure adequate control of post-operative nausea and vomiting with dexamethasone and ondansetron.
- Protect against exposure keratopathy by securing closure of the eyelids intra-operatively.

## Anesthesia Course

- **Initial vital signs:** BP 157/79, HR 81, RR 18, Temp 98.2, SaO2 97
- Prior to intubation, 0.2 mg of glycopyrrolate was administered.
- Awake fiberoptic intubation was performed using 20cc of lidocaine 1% sprayed into the mouth, nares, and trachea.
- A size 7.0 endotracheal tube was placed smoothly in 1 attempt with visualization of vocal cords and tracheal rings.
- After airway was secured and ETCO2 was verified, the patient was induced using 180 mg of propofol, 30 mg of rocuronium, and 200 mcg of fentanyl.
- Eyelids were secured shut with tape and isoflurane was started to maintain anesthesia.
- During the procedure 8mg of dexamethasone and 4 mg of ondansetron were administered at 30 minutes and 60 minutes respectively.
- The case proceeded uneventful with stable vital signs and ventilation.
- Reversal of neuromuscular blockade was achieved using 5 mg of neostigmine and 0.8 mg of glycopyrrolate, the patient was extubated and transferred to the PACU without complication.
- **PACU vital signs:** BP 151/86, HR 70, RR 16, Temp 97.7, SaO2 99 on 4 L NC

## Discussion

- This case demonstrates some of the anesthetic considerations that must be taken into account when caring for a patient with Moebius Syndrome.
- The most common complication encountered in this patient population is difficult intubation due to the altered anatomy and function of the face or airway. Cases of micrognathia and microstomia as in this case or paralysis of a cranial nerve 8-12 has been associated with difficult intubation using direct laryngoscopy.<sup>4</sup>
- Awake fiberoptic intubation provides a safer route to endotracheal intubation in patients with an anticipated difficult airway. It allows the patient to breath spontaneously and if difficulties are experienced it can be abandoned with minimal risk to the patient.<sup>5</sup>
- Patients with Moebius Syndrome are also at increased risk of perioperative pneumonia and respiratory failure from aspiration of airway secretions or gastric contents. This is due to palatopharyngeal dysfunction, tendency to retain oral secretions, and inability to protect the airway as a result of cranial nerve paralysis and facial malformations.<sup>6</sup>
- Pre-operative administration of an anticholinergic such as glycopyrrolate can help control airway secretions. In addition, the use of dexamethasone and ondansetron intra-operatively has been shown to reduce post-operative nausea and vomiting.<sup>7</sup>
- Lastly, inability to close the eye from paralysis of the 7<sup>th</sup> cranial nerve puts these patients at greater risk of exposure keratopathy and closure of the eyelid is imperative.

## Conclusion

- While Moebius Syndrome can provide difficulty to the anesthesiologist, this case shows that careful pre-operative evaluation and planning can overcome those difficulties.
- The use of awake fiber optic intubation in patients with Moebius Syndrome can decrease the risk of difficult intubation.
- While strategies discussed in this case were successful, an individualized anesthesia plan should be used for patients with Moebius Syndrome due to varying presentations.

## References

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