



Anesthetic Considerations in Patients with de Bary Syndrome

Karen Izquierdo, MD; Kalpana Tyagaraj, MD, FASA; Evan Salant, MD

What is it?

- ❖ de Bary syndrome: autosomal recessive genetic disorder that affects multiple organ systems
- ❖ Manifestations include: cataracts, cutis laxa, small mouths, wide foreheads, and mental delays
- ❖ Since 2011, only 27 cases worldwide have been reported



Patient and Pre-Op

- ❖ Six month old male, in NICU
- ❖ Medical history: de Bary syndrome, prematurity
- ❖ Surgical history: ileostomy creation, no anesthetic or surgical complications
- ❖ Family history: mother and father first cousins

Intraop

- ❖ Mask induction followed by intubation with a C-MAC 0 blade and size 3 cuffed ETT
- ❖ IV access was lost after the catheter was noted to be kinked and after multiple attempts, a 24 G was secured on the right foot
- ❖ More blood loss than expected with the patient tachycardic to 240 led to us transfusing a total of 100 ccs of pRBC; patient responded well.
- ❖ Given the hemodynamic instability and occasional desaturations, we decided to keep patient intubated and transport back to the NICU.

Discussion

- ❖ Mask induction was feasible, however, more complicated given the patient's small mouth and short thyromental distance. Two handed techniques for masking, mask with straps, LMA or RSI are all the considerations for airway management.
- ❖ Intubation was successful, however, may not have been as simple with direct laryngoscopy given how anterior the vocal cords were. Video laryngoscopy should be highly considered for these patients.
- ❖ IV access was extremely difficult in this patient given the laxity of the skin and fragility of the blood vessels. This later became our only line to be able to transfuse through. Preparations should be made for difficult IV placement as well as properly securing whatever access is obtained.
- ❖ Given the rarity of this condition, any insight is important to report for proper preparation for good outcomes for future anesthesia teams.