

Anesthetic Management of Bethlem Myopathy: A Multidisciplinary Approach

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CASE:

- 20-year-old G2P0101 at 37w6d presenting for a scheduled repeat cesarean section secondary to Bethlem myopathy
- Patient history complicated by severe restrictive lung disease requiring continuous noninvasive home ventilation, wheelchair bound, and family history of airway losses resulting in death
- Previous cesarean section performed under neuraxial, uncomplicated delivery
- Multidisciplinary team included: Obstetric Anesthesia, MFM, Cardiology, Pulmonology, and ENT

Pulmonary Function Tests/Interpretation:

Parameter Date	FEV1 (L)	FEV1 %Pred	FVC (L)	FVC %Pred	FEF25-75% (L/s)	FEF25-75% %Pred	DLCO_SB %Pred
6/9/2021	0.69	25.0	0.74	23.6	1.02	29.9	
6/9/2021	0.63	22.9	0.69	22.1	1.03	30.3	

Baseline
VBG

Specimen Source	Venous	
FIO2 Venous	Room Air	
pH, Venous	7.32 - 7.43	7.42
pCO2, Ven	40 - 60 mm Hg	35 ▼
pO2, Ven	30 - 55 mm Hg	91 ▲
HCO3, Ven	22 - 27 mmol/L	22
Base Excess, Ven	-2.0 - 2.0	-1.6
O2 Saturation, Venous	40.0 - 85.0 %	97.7 ▲

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Anesthetic Concern	Anesthetic Plan
Rhabdomyolysis, hyperkalemia	Plan A: Slow load epidural Plan B: Non-triggering anesthetic, full MH precautions
Poor respiratory reserve	Continue NIPPV throughout case and post-partum Slow load epidural > spinal Caution with narcotics – epidural morphine deferred Run epidural overnight for pain control – 0.125% bupivacaine + 10mcg/mL hydromorphone Aggressive respiratory therapy
Difficult positioning, contractures, pressure injuries	Epidural placed laterally, then patient assisted with positioning prior to loading epidural
Difficult airway	ENT on standby
Myoclonic contractures	Maintain normothermia, warm fluids
Exacerbated gastrointestinal paresis, and increased risk of reflux/aspiration	Pre-op famotidine, metoclopramide, and non-particular antacid

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Discussion:

- Bethlem Myopathy is a neuromuscular disorder caused by a mutation in type IV collagen
- Autosomal dominant inheritance, in rare circumstances can be autosomal recessive
- Symptoms can manifest as hypotonicity (infancy), proximal muscle weakness, progressive scoliosis, joint contractures, and decline in lung function
- Diagnosis includes clinical exam, lab abnormalities such as elevated CK, and muscle biopsy (definitive)
- Management in pregnancy is geared toward symptom optimization and requires a multidisciplinary care team to avoid progression of disease during pregnancy

