

Management Of A Parturient With Congenital Fiber Type Disproportion Myopathy



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Introduction

Congenital Myopathies (CM) encompass a spectrum of inherited genetic disorders which affect muscle tone and contraction¹. This case report describes a particular CM known as congenital fiber-type disproportion (CFTD).

Case Report

A 28-year-old primigravida with h/o CFTD was admitted for labor induction at 37 weeks for preeclampsia with severe features. PMH was significant for OSA on CPAP, mild asthma, chronic HTN, and sleeve gastrectomy for a BMI of 58.8. She reported generalized weakness, difficulty climbing down, and even combing hair. HR was 88/min, BP 160/92mmHg, RR 27/min and SpO2 93%.

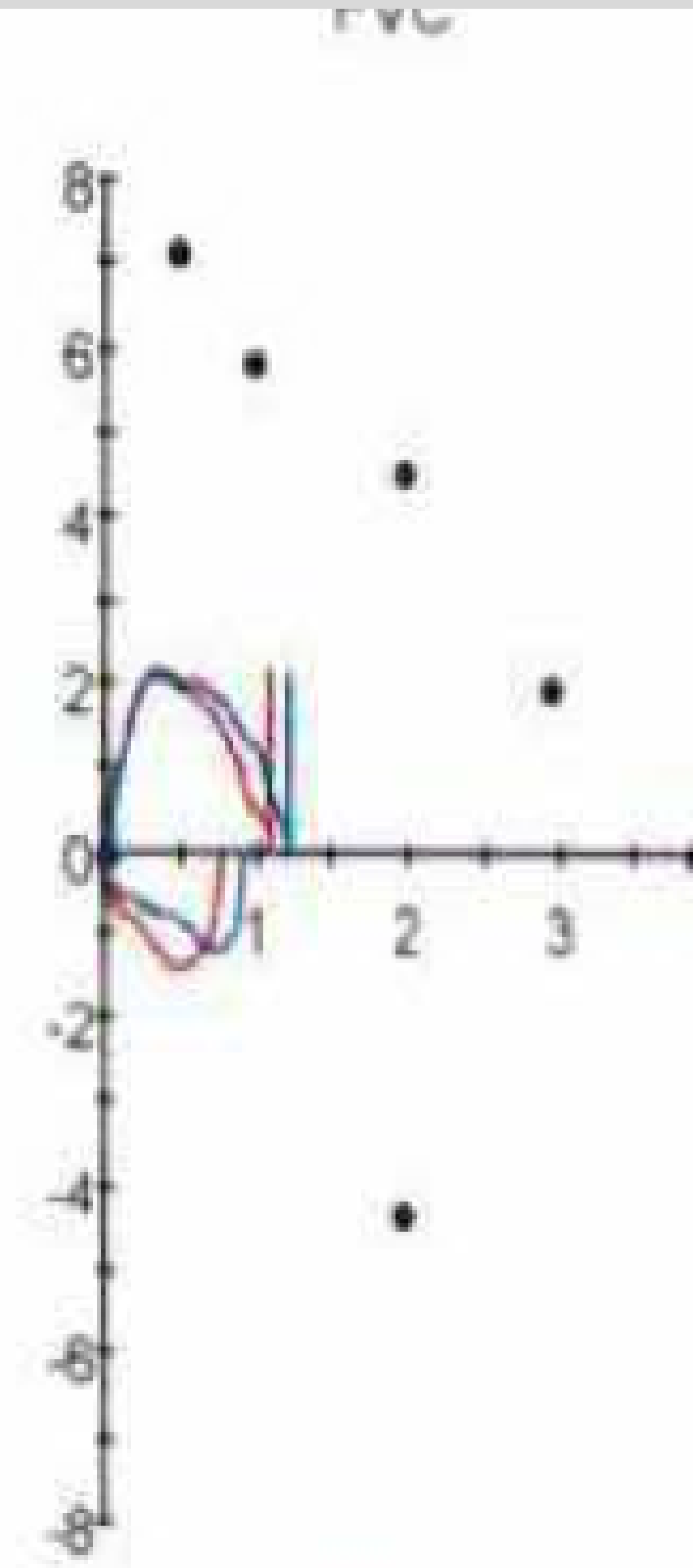
Preoperative Assessment

Airway exam revealed Mallampati class-III, adequate mouth opening, high-arched palate, TMD less than 3 fingerbreadths. Weakness of bilateral sternocleidomastoid, iliopsoas, and limitation in upward gaze was noted. VBG reported a baseline PCO2 of 72.8 mmHg. ECHO revealed an EF of 65%. Levetiracetam was used instead of MgSO4 for seizure prophylaxis because of concerns about respiratory depression. BiPAP was initiated with pressure support of 6 cm H2O and was titrated according to the PCO2 value and the patient's respiratory effort. ABGs were obtained every 2 hours. BP was managed with IV labetalol.

Anesthetic Management

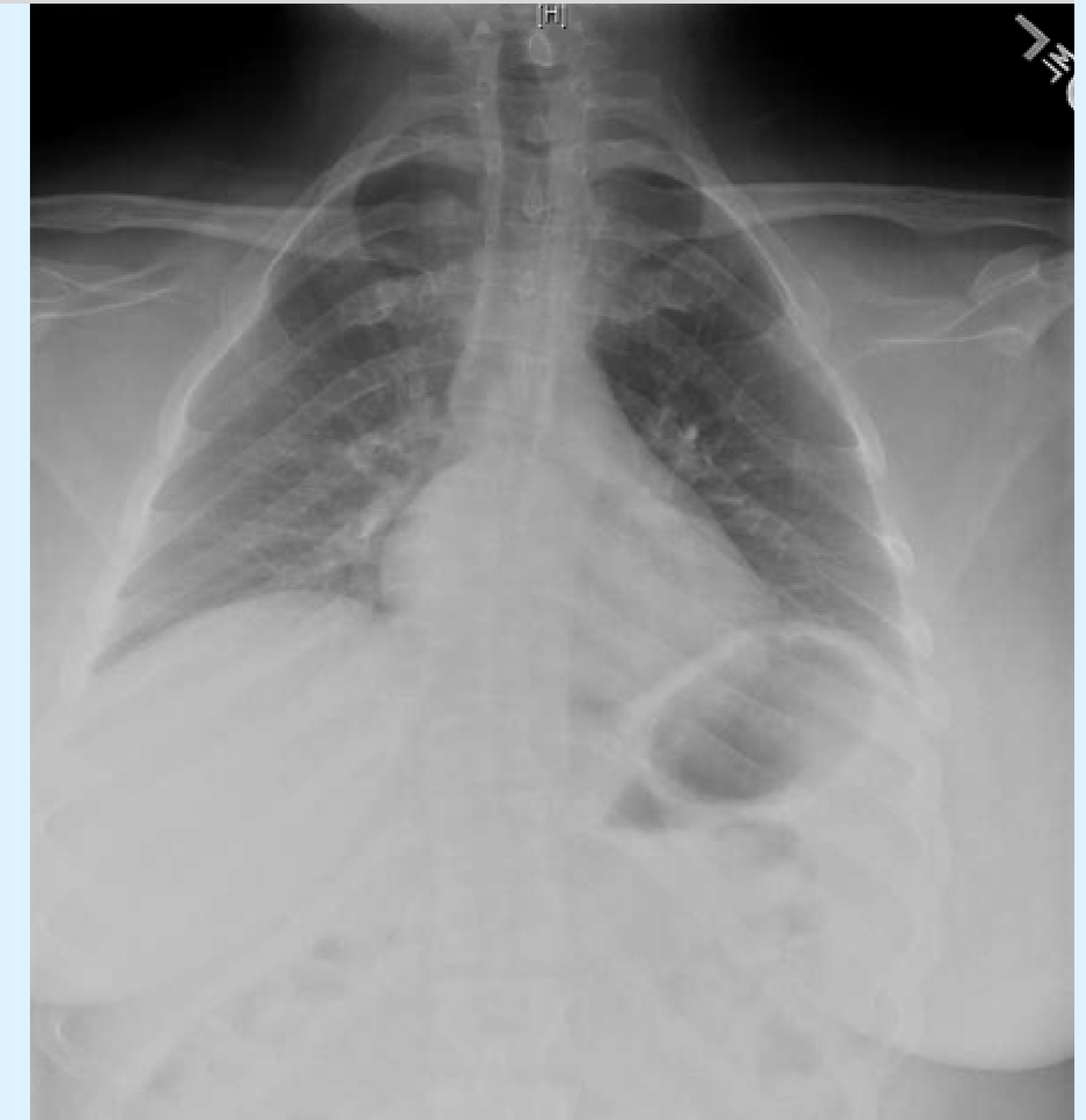
- Ultrasound-guided Dural puncture epidural was placed in a sitting position. The procedure was challenging due to her scoliosis and obesity.
- Because of bilateral iliopsoas weakness, Careful positioning was necessary as these patients are at higher risk of joint dislocation.
- Patient-controlled epidural analgesia was administered using 0.0625% Bupivacaine and Fentanyl 2mcg/mL at 12cc/hr.
- The patient received TXA and misoprostol for the risk of atony. She delivered vaginally and weaned off from BiPAP on postpartum day1.

PFT



PFT demonstrated severe restrictive lung disease (FVC 1.11L) with reduced diffusion capacity.

CxR



CXR reported atelectasis and scoliosis.

Discussion

- The incidence of CFTD is 1 in 50000. The condition is diagnosed with type 1 muscle fiber hypotrophy of at least 12% and clinical features of a myopathy2.
- Worsening of muscle weakness in the third trimester is attributed to rising progesterone on cellular potassium homeostasis.
- Severe symptoms include respiratory failure, ophthalmoplegia, dysphagia, dilated cardiomyopathy, and kyphoscoliosis3.
- CM patients are at risk of MH due to the possibility of RYR1 mutations, so succinylcholine and volatile anesthetics should be avoided, and an MH cart should be readily available.
- Complications include; premature labor, preterm delivery, spontaneous abortion, the prolonged first stage of labor, and uterine atony due to myopathy5.
- A multidisciplinary team approach is essential to discuss the route of delivery, method of analgesia, and anesthesia.
- FVC values above 1L can facilitate vaginal delivery even though there are no clear guidelines for a cut-off value.

Literature Cited

1. Curr Opin Neurol 2021; 34:727-737., 2.Neurology 1969;19:591-6053.Am J Obstet Gynecol 1995;173:1007-1014, 4.Eur J Anaesthesiol 2017;34:641-649 5.Obstet Gynecol 2007;109:501-4

Anesthetic considerations

- 1.Consensus regarding vaginal delivery
- 2.Tailored pharmacotherapy
- 3.Respiratory support- Intermittent NIV
- 4.Arterial line for serial ABG monitoring
- 5.Early epidural- Neuraxial Ultrasound
- 6.Preventive measures for Malignant Hyperthermia
- 7.A theoretical risk for Uterine atony-PPH
- 8.Close monitoring intra-post Partum for worsening of respiratory status

