

Introduction

Langerhans Cell Histiocytosis (LCH) is a rare neoplastic disorder of unknown aetiology which manifests as cystic interstitial disease, diagnosed by lung biopsy [1]. Treatment is with chemotherapy or lung transplant in severe cases [2].

Case Report

We present a 43-year-old, para 4 female with known LCH. She was reviewed in a high-risk anaesthetic clinic as she had previous bronchiolitis, deep vein thrombosis, pneumothorax, asthma, COPD, pregnancy induced hypertension, substance misuse (on long term Methadone) and was awaiting a lung transplant. Her pulmonary function tests showed severely reduced lung capacity (see table) with oxygen saturations of 94% on room air. Her echo showed a normal size left and right ventricle with an ejection fraction of more than 55%. Her medications include Carbocistein 750mg (TDS), Nifedipine 20mg (OD), Lufebec MDI inhaler, Methadone 70mg (OD) and Enoxaparin 20mg (OD). Her exercise tolerance was poor, she was getting short of breath at rest and unable to walk up a flight of stairs.

	Actual	Predicted
FEV1	0.47l	17%
FVC	1.66l	48%
FEV1/FVC	28.6%	38%
DLCO2	2.6	38%

Prior obstetric history was unremarkable; three vaginal deliveries and one emergency LSCS. Her last delivery was 18 years ago, prior to diagnosis. She presented at 29 weeks with worsening shortness of breath, treated with antibiotics and her usual inhalers. She was discussed in a multidisciplinary team meeting and was for planned LSCS aiming to avoid general anaesthesia (GA).

At 32 weeks gestation an elective LSCS was carried out under sequential combined spinal epidural anaesthetic with invasive arterial monitoring. A low dose spinal of 1.5mls of heavy Bupivacaine 0.5% was given along with Morphine 100mcg and Fentanyl 15mcg, which achieved a bilateral block to T7/8. This was topped up with 2% Lidocaine with Adrenaline, 13mls in aliquots, in a 40-degree ramped position. Remifentanyl TCI was used to treat additional pain. Neonatal Apgar scores were 9 & 10 at 1 & 10mins of life. Post delivery analgesic doses of ketamine (15mg) were also given. Offer of GA was declined. Respiratory parameters were maintained throughout with high flow oxygen therapy. Total estimated blood loss was 400ml. Post-operatively she was happy with her care and glad to have avoided a GA. The epidural was kept in for post-op pain relief and patient was monitored in HDU on delivery suite. Due to pain a PCA she was switched to a morphine PCA with 2mg boluses.

After delivery the patient remained on oxygen and had a persistent tachycardia of around 100-140bpm. A course of antibiotics was given for her a chest and a CTPA was carried out to rule out a PE, which was negative (see Fig 1). The patient was discharged home 13 days post LSCS.

Discussion

- The prevalence of LCH is 1-2 per 1,000,000 in adults [1], however the course has not been well characterised in pregnancy [2]. Her estimated morbidity and mortality were 40% and 5-15%, mainly associated with ventilatory failure after GA, hence our use of additional agents to avoid intubation and ventilation.
- Her weight, 48kg, limited the total volume of lidocaine available and her methadone made prediction of remifentanyl target levels difficult. Post operative pain required specialist team input, as epidural opioids and patient controlled analgesia (morphine) provided poor relief. Nefopam was suggested and worked well. We add to the limited literature available on delivery of pregnancies complicated by severe LCH.



Fig 1: CT Chest showing bilateral lung cysts with slight wall thickening (Nov 2025)

References:

- [1] Elzbieta Radzikowska. Update on Pulmonary Langerhans Cell Histiocytosis. Front. Med., 08 March 2021. Sec. Pulmonary Medicine. Volume 7 - 2020 | <https://doi.org/10.3389/fmed.2020.582581>
- [2] Bitar G, Shlossman P. Stable lung function with pulmonary Langerhans cell histiocytosis in pregnancy. J Case Rep Images Obstet Gynecol 2020;6: 100063Z08GB2020.