

A rare case of lymphangioliomyomatosis (LAM) in pregnancy

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Background and Objective

- Lymphangioliomyomatosis (LAM) is a rare systemic condition (4 in 1,000,000).
- LAM is oestrogen driven affecting almost exclusively reproductive age females.
- Progressive cystic destruction of the lungs occurs.
- Risks in pregnancy include deteriorating lung function, spontaneous pneumothorax and preterm birth.
- We report a successful pregnancy outcome in a primigravida with LAM who had a Caesarean birth at 36 weeks’ gestation.
- Care was managed at a local hospital in East London with the support of the North-East London Maternal Medicine Network.

Case Report

- 31-year-old, no previous pregnancies
- LAM diagnosed 3 years prior to pregnancy after a spontaneous pneumothorax
- Under specialist care at the UK national LAM centre.
- Pre-pregnancy management included a left-sided bullectomy and pleurodesis, and maintenance sirolimus.
- Pre-pregnancy FEV1 and FVC 50-80% predicted.

Pregnancy care (see fig 1):

- Sirolimus discontinued.
- Respiratory infection in first trimester with wheeze managed with oral and inhaled corticosteroids.
- Echocardiogram: normal LV and RV function.
- Cervical length monitoring for previous large loop excision of the transformation zone (LLETZ).
- Borderline shortened cervix managed with vaginal progesterone.
- Regular obstetric medicine review.
- Fetal medicine scanning.
- Progressive dyspnoea at 31 weeks: normal chest radiograph, no pneumothorax (fig 2a).
- VTE prophylaxis in the third trimester.
- Fetal growth restriction (3rd centile) on fetal scanning.
- Planned Caesarean birth at 37 weeks with aim of reducing pneumothorax risk.

- Caesarean delivery at 36+2 weeks after spontaneously ruptured membranes.
- Female infant weighing 1775g (0.4th centile) born in good condition.
- Postnatal desaturation. Right apical pneumothorax confirmed on imaging (Fig 2b).
- Stable, but persistent pneumothorax despite CT-guided drain.
- Discharged with home oxygen.
- Definitive management of pneumothorax under regional cardiothoracic unit.

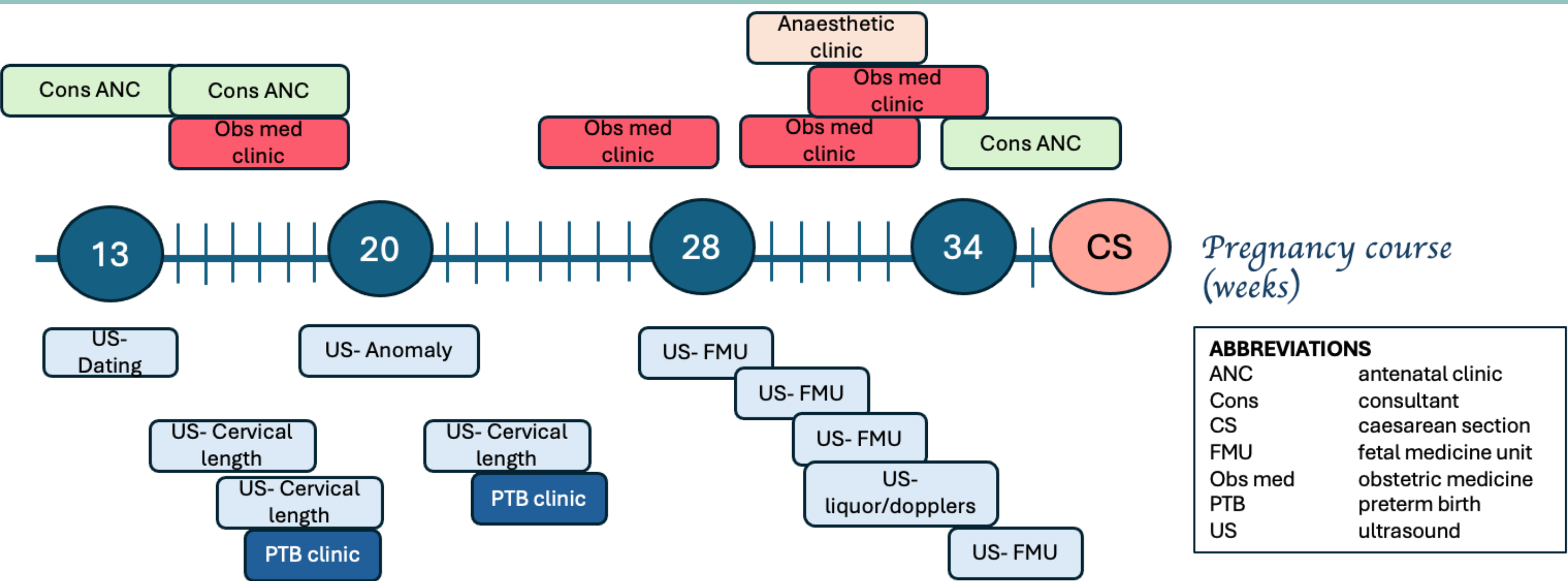


Fig 1. Summary of pregnancy care including consultant-led clinics and imaging.



Fig 2. Chest radiographs: a) Normal, 31/40 pregnant
b) Right apical pneumothorax on postnatal day 1

Discussion

- LAM in pregnancy is rare and presents unique challenges to the obstetric, anaesthetic, and wider medical team.
- Effective multidisciplinary teamwork is essential for management of risk and complications in pregnancy.
- High index of suspicion for pneumothorax is needed.
- Mode of birth is often by Caesarean, but this can be individualised. Fetal growth restriction in a first pregnancy informed decision-making in this case.
- Local delivery in a unit of the woman’s choice was facilitated with support of the North-East London Maternal Medicine Network (fig 3) and the UK National LAM Centre.

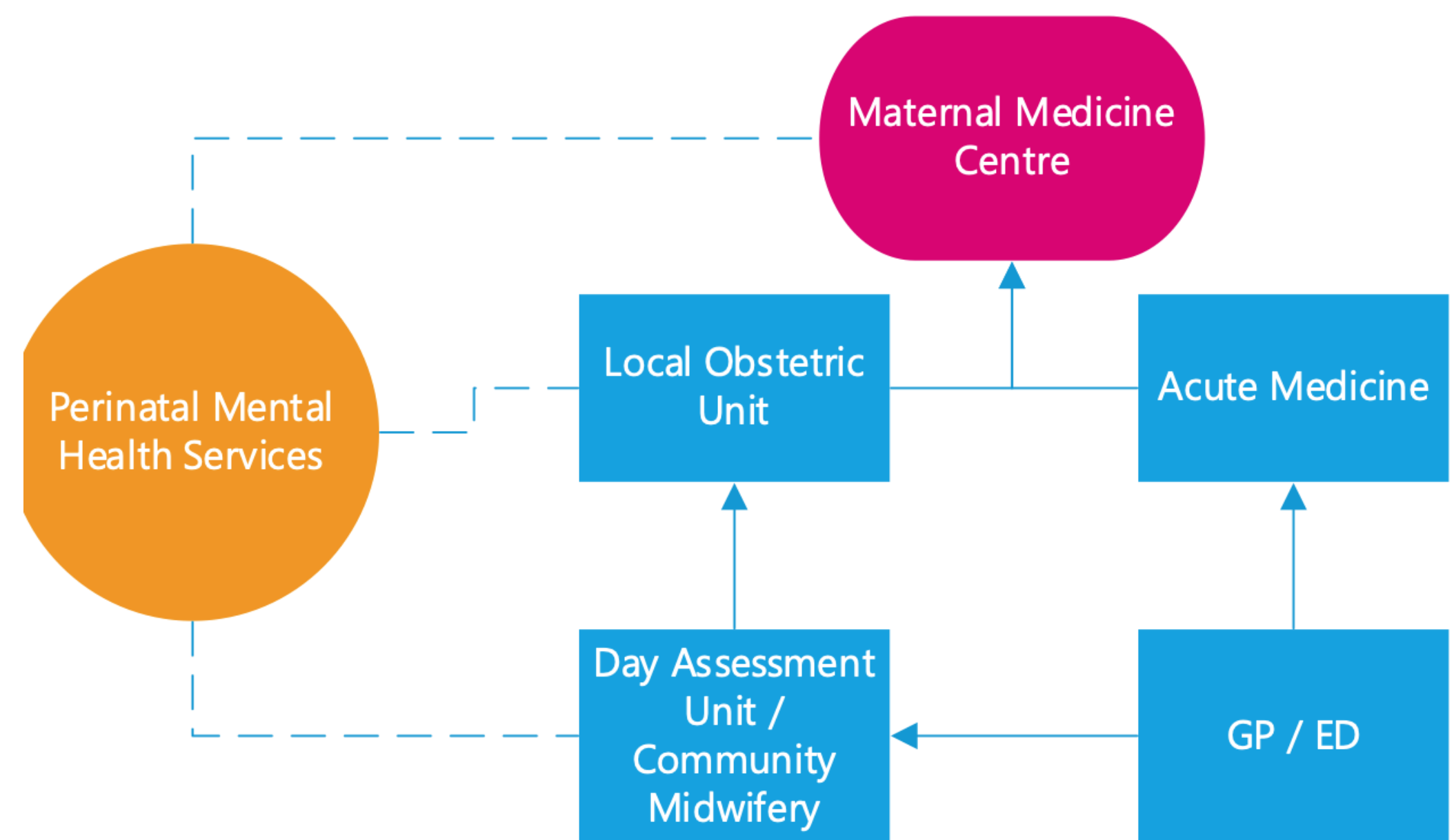


Fig 3. The maternal medicine network: defined pathways for equal access to specialist care for women with medical problems in pregnancy.

Conclusions

- Successful pregnancy outcome is possible in LAM
- There is an increased risk of pneumothorax, particularly peripartum.
- Complex and rare diseases require multidisciplinary teamwork aided by maternal medicine networks in UK.

References

<https://www.england.nhs.uk/publication/maternal-medicine-networks-service-specification/>