

CF Care Beyond Modulators: A Case Study

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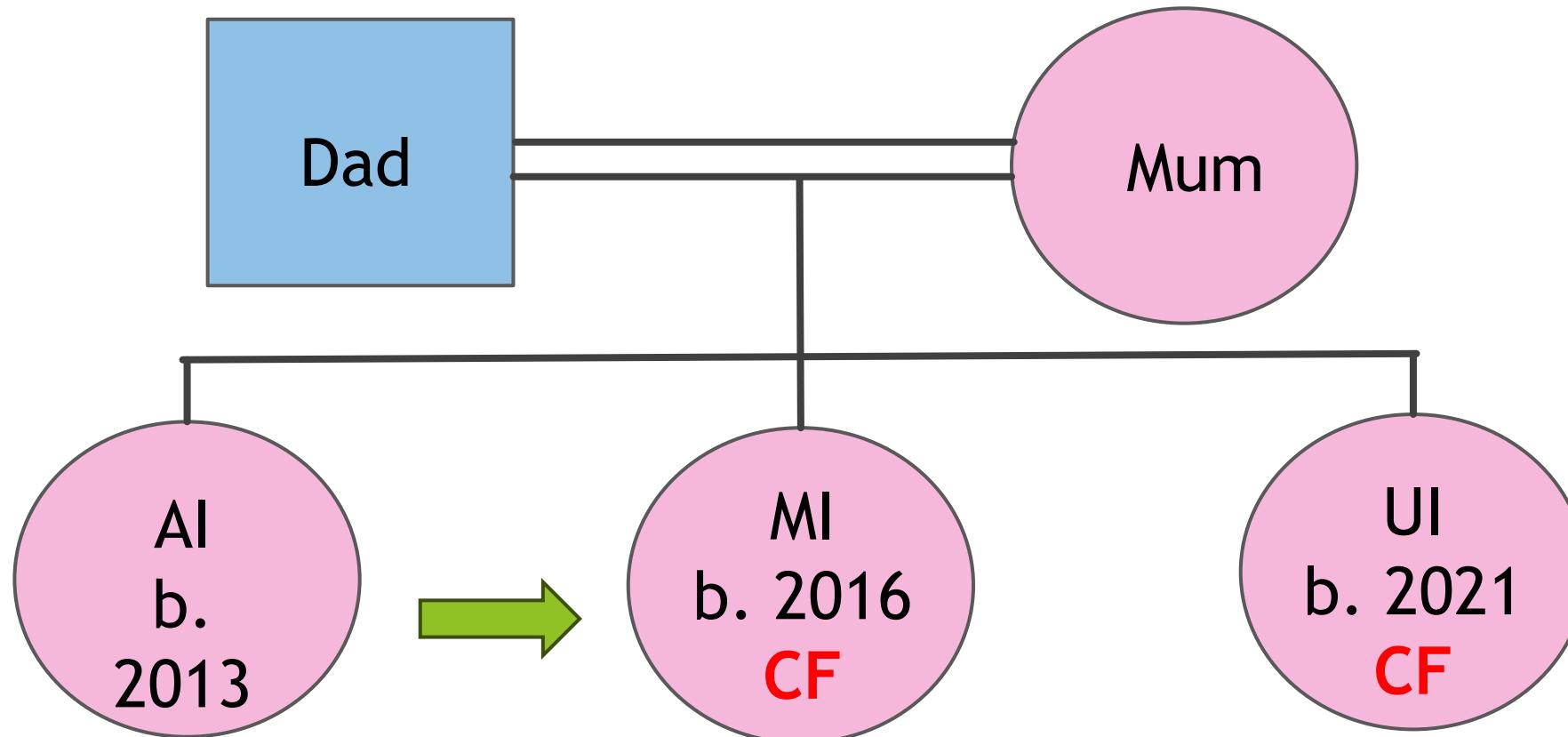
West Midlands (South & Central)
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Introduction

- ▶ Case study of a non-eligible patient and her journey
- ▶ Brief overview of family history and diagnosis
- ▶ Parallel planning
- ▶ Thoughts for staff



Family Tree





2023 - Stepping Up Support

- ▶ 3 monthly IVs
- ▶ Port, gastrostomy, physio vest
- ▶ Palliative care discussions started
- ▶ ? Transplant
- ▶ Advanced care planning discussions commenced



2024 - Escalation and Funding Challenges

- ▶ Niv for airway clearance
- ▶ Line infections - Port removed
- ▶ Individual funding request commenced
- ▶ ACP moved forward
- ▶ NIV overnight commenced
- ▶ Large Pnemothorax
- ▶ Decision made to stop active treatment



2025 - Transitioning to Comfort Care

- ▶ Palliative care continues at home
- ▶ No active treatment, no physio, no nebs and no antibiotics
- ▶ Rallied so MDT and decision made to use nebulisers and physio
- ▶ Pain increased so subcut pain relief started



Thoughts for staff

- ▶ Unsuccessful IFR what does this mean for sibling's future?
- ▶ How do parents maintain trust and communication with the team after a tough decision?
- ▶ What's the emotional impact when there are other family members with CF? (2 cousins with same genes)
- ▶ CF-related deaths in childhood are rare now - which means teams might feel less prepared for end-of-life care
- ▶ Last centre death was 2019 so emotional support and processes in place for discharge are unfamiliar.
- ▶ This case showed just how important timely ACPs are - they really help align care with family wishes
- ▶ Sibling



Conclusion

- ▶ We've learnt lots, ACPs, IFRs
- ▶ Processes in place for end of life care
- ▶ Care of children not eligible a focus for our centre
- ▶ CF is not fixed for everyone