

Background

Jane Johnston is a 28-year-old female who works from home as a contract web developer. She was diagnosed with cystic fibrosis (CF) at birth following failure to thrive. She has two copies of the F508del mutation, is chronically colonised with *Pseudomonas aeruginosa*, and attends the CF clinic for review every 3 months.

She commenced triple combination therapy, Kaftrio® (ivacaftor/tezacaftor/elexacaftor), 3 years ago. Prior to this, Jane experienced at least six exacerbations per year requiring intravenous antibiotics. These have since decreased in frequency to once per year on average.

Past Medical History

- Cystic fibrosis (F508del homozygosity)
- CF related bronchiectasis
- Secondary diabetes mellitus
- Pancreatic insufficiency
- Osteopenia

Jane has presented to the CF clinic with a 2-week history of progressive fatigue, an increase in breathlessness, and worsening cough productive of increased purulent blood-streaked sputum which is difficult to expectorate. She has tested negative for Covid-19 and influenza. Jane also reports constipation which has been getting worse over the past three to four months.

Observations

- Height 166cm and weight 50.2kg. Baseline weight 51kg -52.5kg
- Heart rate 76bpm, temperature 36.8°C, blood pressure 108/71mmHg, respiratory rate 18bpm and SpO2 98% on room air
- Pulmonary Function Tests (PFTs): FEV₁ baseline is 60%, current is 53%
- WBC: 8.4 (10⁹/l), CRP: 33mg/L
- HbA_{1c}: 6.5%

- Recent DEXA scan previous month showed osteopenia in femur and pelvis. This was unchanged from a DEXA 2 years earlier.
- Chest X-ray demonstrates coarse lung marking in keeping with diffuse bronchiectasis. There is no evidence of pneumothorax.
- CT thorax demonstrates diffuse bronchiectasis, with mucus plugging and resultant atelectasis in the lower lobes.
- Influenza, Pneumonia and Covid vaccinations are up to date.
- Sputum culture positive for *Pseudomonas aeruginosa*, sensitivity results are pending.
- Vitamin levels
 - 25-hydroxy Vitamin D: 16 nanograms/ml (20-40 nanograms/ml)
 - Vitamin A: 15 micrograms/dL (20-60 micrograms/dL)
 - Vitamin E: 4.5 microgram/mL (5.5-17 microgram/mL)
- Ultrasound liver – a mildly coarsened echotexture in keeping with fatty infiltration.

Medication

Drugs	Directions
Kaftrio® (ivacaftor/tezacaftor/elexacaftor) 75mg/50mg/100mg tablets PO	TT mane PO
Kalydeco® (ivacaftor) 150mg tablet PO	T nocte PO
Creon® (pancreatin) 25000 capsules PO	Two-five capsules during or immediately after each meal and snack PO
Salbutamol 100 microgram/dose inhaler	One to two puffs prn INH
Pulmozyme (dornase alfa) 2.5mg nebuliser liquid 2.5ml ampoules	T mane NEB
Levemir® (insulin detemir) FlexPen 100units/ml	12 units mane SC
NovoRapid® (insulin aspart) FlexPen 100units/ml	4 units before each meal SC
Alendronic acid 70mg tablets	T once weekly PO
Azithromycin 250mg capsules	T daily PO
Vitamins A, D, E and K tablets	mdu PO



Social History

Jane was recently married and lives at home with her husband Peter. She is a non-smoker and has a limited alcohol intake. Jane is a member of a local athletics club and usually runs twice per week. She has adjusted her diet since commencement of CFTR therapy as she is concerned with weight gain. Jane has expressed a desire to start a family and has asked about this at a recent outpatient appointment. She has expressed concern about the safety of her medicines in pregnancy and wonders if she should stop any of them. Jane and Peter worry if their baby would have CF. Jane works from contract-to-contract and does not have any remuneration when on sick leave or attending appointments. Jane is an identical twin, and her sister Emma has DF508 homozygous cystic fibrosis. Emma's lung disease is significantly worse, and she is on the lung transplant list. Emma has been advised that it would be high-risk to become pregnant and her physicians have advised against it.

Inpatient Journey

Jane is admitted to the respiratory ward for management of her acute presentation. She is reviewed by the multi-disciplinary team on a daily basis. By day 5 of her admission, Jane is feeling somewhat improved and is keen to get home and return to work.

Goals for the Team

1. Formulate an immediate management plan for her current hospital admission.
2. Outline how the team can execute an integrative discharge plan to optimise Jane's ongoing care.