Cystic Fibrosis

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Cystic Fibrosis (CF): An autosomal-recessive, multisystem disorder that affects the lungs, digestive system, reproductive system and sweat glands.

Cause: mutation in the gene that encodes for the chloride channel cystic fibrosis transmembrane conductance regulator (CFTR)

Pathophysiology:

- Genetic defect → impaired sodium and chloride transport across epithelium cell surface → less water leaves cell via osmosis → viscous secretions
- In the bronchi: goblet cell hyperplasia, submucosal gland hypertrophy → production of viscous mucus combined with mucus plugging → airway inflammation → activation of neutrophil elastases → tissue distruction → increased thickness of airway walls and V/Q mismatch → hypoxaemia

Presentation

- Variable presentation depending on type of genetic mutation and environmental factors
- Chronic, productive cough
- Dyspnoea
- Nail clubbing
- Auscultation: basal crackles and expiratory wheeze
- Increased A-P chest diameter
- Susceptible to: sinusitis, Pseudomonas aeruginosa, allergic bronchopulmonary aspergillosis, bronchiectasis

Diagnosis

- Chest X-ray: hyperinflation, air trapping, atelectasis, flattened diaphragm, tram-track opacities (thickened bronchial walls)
- Chest CT: thickened bronchial secretions, mild - moderate bronchiectasis
- Sweat chloride test: increased sweat chloride concentration
- Pulmonary function test: reduced FEV1/FVC ratio, increased RV/TLC ratio

Management

Chest Physiotherapy:

- Postural drainage: placing the patient in a position which allows gravity to assist in draining mucus from the periphery of the lungs to the central upper airway
- Active cycle of breathing: consists of Breathing Control, Thoracic Expansion Exercises, and Forced Expiration Technique
- Prevent cross-infection: use microbiological surveillance and a local infection control strategy that includes cohorting
- Percussion: use cupped hands to rhythmically and firmly percuss the posterior chest wall over a towel
- Oscillating PEP: (flutter, cornet, acapella) encourages airflow behind secretions and oscillations induce vibrations that decrease the viscosity of secretions
- Aerobic exercise: cycling/ swimming/ brisk walking

Medical: Inhaled hypertonic saline, anti-inflammatory medications, antibiotics, bronchodilators, annual influenza and pneumococcal vaccine

Want to learn more?

With AcePhysio the learning journey doesn't stop here! Take a look at our further reading recommendations below to become an expert in CF:

1. Rodriguez Hortal, M. C., Nygren-Bonnier, M., and Hjelte, L. (2017) Non-invasive Ventilation as Airway Clearance Technique in Cystic Fibrosis. Physiother. Res. Int., 22: e1667
2. Rafeeq MM, Murad HAS. Cystic fibrosis: current therapeutic targets and future approaches. J Transl Med. 2017 Apr 27;15(1):84
3. Mckoy NA, Wilson LM, Saldanha IJ, Odelola OA, Robinson KA. Active cycle of breathing technique for cystic fibrosis. Cochrane Database Syst Rev. 2016 Jul 5;7:CD007862