**Title**

Cystic fibrosis in the adult patient: diagnosis and a review of new treatments for CF lung disease.

**Author and affiliations**

Author: **Melinda Choy1**

Co-authors: Peter Bye2

1Australian National University, Academic Unit of General Practice. Dr Melinda Choy is a second year GP and 2018 RACGP Academic Registrar. Before she started GP training, she was a BPT at Royal Prince Alfred Hospital.

2University of Sydney, Central Clinical School of Medicine. Professor Peter Bye is the head of the Cystic Fibrosis Unit at Royal Prince Alfred Hospital.

**Background**

Cystic fibrosis (CF) is a multi-system disorder, caused by mutations of the cystic fibrosis transmembrane conductance regulator (CFTR) gene. The defective CFTR results in decreased epithelial cell chloride secretion and increased sodium and water resorption. In CF lung disease, the result is drier, stickier mucus, which leads to chronic inflammation and infection.

Cystic fibrosis is traditionally considered a paediatric disorder, diagnosed in hospital. However, since 2015, over half of Australian CF prevalence is accounted for by adults. This is partly because the life expectancy for CF patients is now 38 years. Further, 10% of new CF diagnoses occur in the adult population. Adult CF can be an outpatient issue, initially seen in general practice rather than in the hospital.

Therefore it is of interest to general practice that treatment of CF lung disease is a rapidly evolving field which now involves CFTR modulators, a new class of medication targeting the specific gene mutation. When matched to patients’ particular CFTR mutation, CFTR modulators can improve lung function and reduce pulmonary exacerbations.

**Aims**

To provide an overview of cystic fibrosis diagnosis in the adult patient and an update on available treatments for CF lung disease, specifically the new CFTR modulators.

**Method**

A clinical overview of GP-relevant aspects of adult CF diagnosis and management will be undertaken through research of the most recent evidence.

**Results**

This presentation will provide an overview of initial CF diagnosis in an adult, and current and emerging treatments.

**Conclusion**

Adult CF is a growing area of outpatient medicine. This presentation will give an appropriate approach to clinical general practitioners for the recognition, diagnosis and referral pathways of CF lung disease, as well as an interesting overview of the available and emerging treatments for CF lung disease.