Sheffield Thoracic Institute

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Sheffield Adult Cystic Fibrosis Centre

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Cystic Fibrosis Secretary: 0114 226 9069

Cystic Fibrosis Ward
Ward Manager: 0114 305 2138
Nurses Station: 0114 226 6580 / 0114 305 2137

FPE/MJW/jr
Date as postmark

Dear Doctor

Your patient _____ has cystic fibrosis (CF) with a specific genotype and has been prescribed Kalydeco (Ivacaftor, Vertex pharmaceuticals) 150mg bd for the foreseeable future. Ivacaftor is now licensed for the following genotypes; G551D, G178R, S549N, S549R, G551S, G1244E, S1251N, S1255P, G1349D.

CF is an autosomal recessive life shortening condition arising from failure of the cystic fibrosis transmembrane regulator (CFTR) protein and abnormal transfer of salt and water across the cell membranes of exocrine glands, notably the lung (recurrent infection and respiratory failure), pancreas and gut (maldigestion, poor growth and weight) and the male reproductive tract (infertility).

The listed genotypes result in a Class II defect which can be partially corrected by the "potentiator" drug ivacaftor. This has been shown to correct the sweat test, improve weight, lung function and quality of life in patients with CF for as long as the drug is taken.

Supplies will be from the hospital via BUPA homecare, so there will be **no** requirement for GPs to prescribe. Prescriptions will be initiated and continued only by Drs Edenborough and Wildman at the Sheffield Adult CF Centre (Authorised Prescribers) who will also coordinate monitoring.

Monitoring requires an initial sweat test, a repeat test 6-8 weeks later and annually to monitor efficacy and adherence. Liver function tests will also be monitored 3 monthly and all other parameters of well being will be recorded as usual and audited via the CF Trust Port CF database. Other treatment will continue as the clinical condition dictates. Patients will be counselled as to how to take this medicine in relation to food etc.

Caution is required with macrolides, antifungals, rifampicin, antiepileptics and benzodiazepines. New medication in general should only be initiated in consultation with the CF team**.** The summary of product characteristics should be referred to for more details. This is available here**:**
http://www.medicines.org.uk/emc/medicine/27586/SPC/Kalydeco+150+mg+film-coated+tablets/

GPs are encouraged to add ivacaftor as a hospital issued drug in order to pick up potential interactions. There is a useful reminder on how to do this here: intranet. We will write to you again if treatment with ivacaftor is stopped so that this can be removed from the patient's record.

All queries (from patient or GP) should be directed to the Sheffield Adult CF Centre via the address or telephone numbers shown above. Further details are available from the specialist commissioning website "Ivacaftor page" http://www.england.nhs.uk/commissioning/wp-content/uploads/sites/12/2015/07/ivacftr-cystic-fibrosis-a01pc.pdf

Yours sincerely

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