

1. Introduction

The purpose of this document is to set out guidelines to ensure standardised care for children with cystic fibrosis looked after at the Royal Brompton & Harefield NHS Foundation Trust and District General Hospitals on a network care basis. They should be used as a guide only. The Royal Brompton Hospital is a Specialist CF Centre as defined by the Specialist Commissioners, NHS England.

With the development of the APP for our guidelines in 2017, we know that aside from the UK, the guidelines have been downloaded 53 countries - Afghanistan, Argentina, Australia, Austria, Bangladesh, Belgium, Brazil, Bulgaria, Ecuador, Egypt, France, Germany, Gibraltar, Greece, Hong Kong, Hungary, India, Iran, Ireland, Italy, Kazakhstan, Kenya, Latvia, Lebanon, Luxembourg, Macedonia, Malaysia, Malta, Mexico, Montenegro, Netherlands, New Zealand, Oman, Pakistan, Portugal, Philippines, Qatar, Romania, Russia, Saudi Arabia, Serbia, South Africa, Spain, Sweden, Switzerland, Thailand, Turkey, Seychelles, UAE, Ukraine, Uruguay, USA, and Yemen. This year we are going paperless, so the guideline will be available online and via APP only. We will only print out the 'What's new' section, contacts and drug formulary for use in our clinics.

Our philosophy of care for patients with cystic fibrosis is based on current guidelines laid down by the Royal College of Physicians, Royal College of Paediatrics & Child Health (formerly British Paediatric Association), CF Trust, British Thoracic Society, and NHSE Service Specifications. These have identified significant advantages in terms of survival and morbidity for patients receiving care from specialist centres. Specialist centres offer access to comprehensive care from a multidisciplinary team consisting of consultants with a special interest in CF, trainee doctors, nurse specialists, dietitians, physiotherapists, clinical psychologists, pharmacists and social workers. The team is also responsible for producing and distributing educational material and carrying out research to improve knowledge about this disease. Special procedures and investigations are provided that may not be available at District General Hospital level (such as formal lung function and bronchoscopy). We are happy to continue with a shared care policy, as long as the NHSE National Service Specification and our signed Service Level Agreement are adhered to. We also run several out-reach clinics whereby our MDT see CF patients in their local hospitals.

Details of the Service Specification can be found –
<https://www.england.nhs.uk/commissioning/spec-services/npc-crg/group-a/a01/>.

Death in childhood from CF is now rare, and UK children born today are likely to have a median life expectancy of 47 years, which will improve further if CF modulator therapy becomes available for use in all patients. There are approximately 10,500 people with CF in the UK and now 60% are adults. On average, large District General Hospitals will have a local CF population of between 10 and 20 patients (it may be less in the London region which has a higher density of hospitals); and General Practitioners between 0 and 2 patients. The Paediatric CF Service at the Royal Brompton Hospital has around 320 children under its care whilst there are about 650 patients in the Adult Service. The paediatric team normally sees children and adolescents until they finish their GCSEs and they will have made the transition to an Adult CF Service at the Royal Brompton Hospital or another Specialist Adult CF Centre of their choice by their 17th birthday.