

Cystic Fibrosis Annual Review

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According to National Institute of Clinical Effectiveness (NICE) Quality standard [QS168] Published date: 18 May 2018:

“People with cystic fibrosis have the results of all assessments they have had during the past year reviewed annually by a specialist multidisciplinary team.”

An Annual Review is carried out for all patients with CF who are under the care of the Royal Alexandra Children’s Hospital, Brighton as the CF Centre. The Annual Review takes place as near as possible to the child’s birthday.

5 components:

- Home visit
- Hospital Visit
- Data correlation and referral
- Pre-annual review clinic multi-disciplinary team meeting
- Annual Review multi-disciplinary clinic.

1. Home visit

Undertaken by CF Nurse Specialist (Network/Centre) 6-8 weeks before clinic appointment.

- Annual review questionnaire (holistic review including lifestyle, treatment adherence, school, education and understanding of CF)
- Psychology screening questionnaires-PHQ8 and GAD 7 for parents and children - HADS for children.
- CF Quality of life questionnaire
- Transition – ‘Ready, Steady, Go!’ assessment (starting at age 11-12 yrs.)
- Specific mention when appropriate of puberty, fertility, transition, pregnancy, transplantation
- Life milestone planning (starting nursery, school, university)
- Treatment equipment review (nebuliser/inhaler techniques)
- 3 Day Food Diary
- Stool and Urine sample pots (Stool sample to coincide with timings of food diary whenever possible)
- Preparation for blood tests.
- Preparation for Modified Glucose Tolerance Test (aged 10 and over – CFRD screening guidelines)

2. Hospital Visit

With CF Nurse Specialist to coordinate approx. 4 weeks before clinic appointment.

- Blood tests with hospital Phlebotomist or for patients with a portacath by the CF Nurse Specialist.
 - FBC
 - U and E
 - Creatinine
 - LFT's (+GGT)
 - IgE
 - Ig G, A, M
 - Vitamin A,E and D
 - Zinc
 - HBA1C
 - Random glucose or modified GTT (over 10)
 - INR (with CF Liver disease)
 - Ferritin
- Spirometry & Body Plethysmography (pre and 15 minutes post bronchodilator)
- Chest X-Ray
- Food diary collection for CF dietitian
- Stool Sample for microscopy for fat globules
- Urine Sample for glucose urinalysis (in OPD)
- Urine Sample for urinary sodium & electrolytes (lab)
- CF Physiotherapist review:
 - Modified VO₂ Max exercise test – age appropriate
 - Physiotherapy review of airway clearance techniques and equipment
 - Discuss goal orientated plans for the year ahead
 - Sputum sample/Cough Swab (Sputum sample sent for Non-tuberculous mycobacterium)
 - Assessment of posture and review of continence issues

3. Prior to Annual review clinic

- Audiology (if received one or more aminoglycoside courses in previous year)
- Bone densitometry (DXA scans) 2 yearly over the age of 10yrs (annually if abnormal).
- Liver Ultrasound scan 2 yearly over the age of 5yrs (annually if abnormal).
- Data correlation and entry to local CF database
- Referral as necessary to Social care agencies
- Continuous Glucose monitoring if indicated from screening or as requested by Endocrine team (See CFRD guidelines)
- Update and collate information from other specialties: Liver (Kings), Gastro, CAMHS

4. Pre-Annual Review Multidisciplinary team meeting (Network & Centre teams)

- Review of issues raised at home visit
- Review of respiratory status for the year
- Review of investigations
- Chest x-ray review
- Assessment of nutritional status and evidence of malabsorption, following food diary analysis and enzyme use, nutritional support and vitamin supplementation
- Review of Psychology screening tools & discussion of psychosocial factors.
- Specialist pharmacist review and discussion of all medication, both prescribed and non-prescribed and review adequacy of supply arrangements.

5. Annual Review Multidisciplinary Clinic (Network & Centre teams)

(To be seen by CF Consultant, CF Nurse Specialist, CF Dietitian, CF Physiotherapist, CF Psychologist, CF Pharmacist)

- Pulse Oximetry
- Height & Weight measurement
- Growth Velocity recording and assessment
- Detailed assessment of the patient's condition, therapies, changes over the last year, treatment improvements, lifestyle and education issues
- Download of digital nebuliser data, monitoring treatment adherence, technique and efficacy (as appropriate)
- A management plan should be discussed with the patient / carers and agreed
- A report should be written by a consultant once all results are available, and sent to the GP and patient / carers.
- CFRD Patients – Annual review with Endocrine team and incorporated into CF Annual review management plan.
- UK CF Registry entry.