20. Annual assessment

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1. INTRODUCTION

- Annual review is an established feature of CF care:
  - It allows a global and systematic assessment of the different aspects of CF.
  - It facilitates collaboration and communication with the multidisciplinary team members.
  - It helps patient education on CF management.
- It can be conducted as
  - a '*specific annual review visit*', i.e. half-day to perform the investigations and a dedicated visit to discuss/summarize the results.
  - a '*continuous assessment',* i.e. evaluation of the various items/points during the regular patient visits throughout the year.
- An annual medical report is created for the patient's file and for the doctors implicated in the care of the patient.
- A list of points and items to be considered during the annual review or at least on an annual basis are presented in **Tables 1 and 2**.

Table 1: Items and points to consider during the annual review or at least annually

General Assessment	
Clinical Interview	– Medical and life events since previous annual review – Hospitalizations and use of health-care resources (including specialized consultations conducted since previous annual review) – History by systems • Respiratory • ENT • Digestive • Allergies • Endocrinology/metabolism: diabetes, bone disease • Renal and urinary (e.g. nephrolithiasis, incontinence) • Gynecology, contraception, maternity, paternity • Psychological problems • Social and insurance issues, work, education – Other according to clinical indications

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Clinical examination	– Vital signs including $SatO_2$, blood pressure, weight, height (BMI) – Cardiopulmonary examination – Basic ENT examination (throat inspection, good nasal breathing on both sides). – Abdominal examination – Other according to clinical indications
Treatment review	– Treatment modifications since previous annual review – Adherence to treatment – Number and type of antibiotic courses (oral, iv) since previous annual review – Review of vaccination status (flu vaccination confirmation, whooping cough, anti-pneumococcal vaccination) – Renewal of annual prescription – Prescription for travelling (on demand)
Venous access	– PAC function, intervals of care discussed with the CF nurse
Respiratory system therapy and Physiotherapy	– Annual review by an experienced respiratory physiotherapist (either the CF center physiotherapist or the patient's personal physiotherapist) – information on CF physiotherapists can be found in www.cf-physio.ch – Inhalation therapy review and adherence to treatment – Review of the sequence of inhalation therapies – Nasal/sinus therapy review – Review of physiotherapy techniques: competence – Review of material maintenance: function, cleanliness
Dietary assessment	– Annual assessment by the CF center dietician – Weight curve and BMI – Current diet - energy, protein and calcium intake – Adequacy of pancreatic enzyme replacement therapy – Bioelectric impedance analysis (BIA) for selected patients – Blood prealbumin levels for selected patients
Clinical pharmacology	For selected cases according to clinical indications
Psychologist	For selected cases according to clinical indications
Social worker	Suggested to all patients - information on social workers dedicated to adult CF patients can be found in the site of CFCH www.cfch.ch/fr/prestations/soutien-et-conseil/assistantes-sociaux
Respiratory functional tests	
Pulmonary function tests (PFTs)	– For all patients: Spirometry, plethysmography – Optional: Diffusing capacity – Reversibility testing if newly observed obstructive syndrome and/or in selected cases (at any time point – not necessarily during the annual assessment) – Lung clearance index, if available, for selected cases (e.g. adults with normal spirometry to detect early changes) – Arterial blood gases for selected cases

Exercise testing (6-min walk test, 3-min step test, ergospirometry)	For selected cases according to clinical indications
Oxymetry/ capnooxymetry	For selected cases according to clinical indications
Laboratory analysis	
Complete blood count	For all patients
Prothrombin time, INR	For all patients
Biochemistry	– Inflammatory markers: CRP, total IgG – Renal function and electrolytes: Creat, Urea, Na, K, Cl^-, $HCO3^-$, Ca, Mg – Liver function tests : AST, ALT, γGT, ALP, bilirubin – Pancreatic tests in selected cases: lipase, amylase – Fat soluble vitamins: A, D, E – Prealbumin in selected cases – Total cholesterol, LDL, HDL, triglycerides for patients >40 years-old or CF-related diabetes or family history of hyperlipidemia
ABPA screening	– Total IgE for all patients – In case of IgE elevation and ABPA suspicion assess *A. fumigatus* specific IgE, skin testing and precipitins
Glycemic control	– For diabetic patients: – assessment by a diabetologist – fasting glucose, random capillary glycemia (including 2h post prandial glycemia values), HbA1c, assessment for diabetes-related complications (retinopathy, nephropathy, neuropathy) – For non diabetic patients: Oral glucose tolerance test (OGTT) – For selected cases: The indication for continuous glucose monitoring system (CGMS) should be discussed with a diabetologist
Urine Analysis	– Urinary calcium excretion (24h urine collection or ratio of calcium/ creatinine in a fasting spot, morning urine sample) – For diabetic patients: glycosuria, albuminuria
Faecal analysis	– Faecal pancreatic elastase in selected cases
Imaging studies	
Chest X-radiography Chest CT-scan	– Annual chest X-radiography is NOT recommended for all patients routinely – A paired inspiratory/expiratory chest CT-scan, with minimal radiation protocol and without contrast medium is proposed at initial patient evaluation (e.g. at transition from paediatric care, or at diagnosis for patients diagnosed in adulthood)

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	– If a chest CT-scan with a minimal radiation protocol is available, it is recommended every 2-3 years. If not, plain chest radiography is recommended every 1-2 years depending on the clinical status.
Abdominal ultrasound	Annually. Elastography may be added in selected cases.
Bone densitometry	Dual energy X-ray absorptiometry scanning every 2-5 years
Microbiological analysis	
Review of microbiology results	– Colonizing and intermittently present microorganisms – Last positive culture for *P. aeruginosa*, MRSA, mycobacteria, rare microorganisms
Sputum microbiology	– Standard microbiology (including *B. cepacia* complex) and mycology – Culture for non-tuberculous mycobacteria at least annually

Table 2: Specialist consultations to consider

ENT specialist	– In selected cases – Audiometry for patients receiving IV aminoglycosides ≥1x/year and for selected patients treated with azithromycin
Diabetologist	– At least annually for diabetic patients
Ophtalmologist	– Annually for diabetic patients – Before initiation and during treatment with ivacaftor (evaluation for cataract)
Bone disease specialist	– Annually for patients with low trauma fracture, osteoporosis treatment or difficult to treat vitD deficiency despite good dosing and adherence
Gastro-enterologist	– Colonoscopy screening: after 40 years of age (see also Chapter ''Aging in Cystic Fibrosis''). Follow-up at the discretion of the gastro-enterologist.

2. REFERENCES

1. Kerem E, Conway S, Elborn S, Heijerman H. Standards of care for patients with cystic fibrosis: a European consensus. Journal of cystic fibrosis : official journal of the European Cystic Fibrosis Society 2005;4:7-26.
2. Debray D, Kelly D, Houwen R, Strandvik B, Colombo C. Best practice guidance for the diagnosis and management of cystic fibrosis-associated liver disease. Journal of cystic fibrosis : official journal of the European Cystic Fibrosis Society 2011;10 Suppl 2:S29-36.
3. Chuang S, Doumit M, McDonald R, Hennessy E, Katz T, Jaffe A. Annual Review Clinic improves care in children with cystic fibrosis. Journal of cystic fibrosis : official journal of the European Cystic Fibrosis Society 2014;13:186-9.

4. Sermet-Gaudelus I, Bianchi ML, Garabedian M, et al. European cystic fibrosis bone mineralisation guidelines. Journal of cystic fibrosis : official journal of the European Cystic Fibrosis Society 2011;10 Suppl 2:S16-23.
5. Sinaasappel M, Stern M, Littlewood J, et al. Nutrition in patients with cystic fibrosis: a European Consensus. Journal of cystic fibrosis : official journal of the European Cystic Fibrosis Society 2002;1:51-75.
6. Alexander CL, Urbanski SJ, Hilsden R, Rabin H, MacNaughton WK, Beck PL. The risk of gastrointestinal malignancies in cystic fibrosis: case report of a patient with a near obstructing villous adenoma found on colon cancer screening and Barrett's esophagus. Journal of cystic fibrosis : official journal of the European Cystic Fibrosis Society 2008;7:1-6.
7. Hernandez-Jimenez I, Fischman D, Cheriyath P. Colon cancer in cystic fibrosis patients: is this a growing problem? Journal of cystic fibrosis : official journal of the European Cystic Fibrosis Society 2008;7:343-6.
8. Quittner AL, Abbott J, Georgiopoulos AM, et al. International Committee on Mental Health in Cystic Fibrosis: Cystic Fibrosis Foundation and European Cystic Fibrosis Society consensus statements for screening and treating depression and anxiety. Thorax 2016;71:26-34.