The Association for Clinical Biochemistry & Laboratory Medicine

Better Science, Better Testing, Better Care

Summary of NICE Guidelines

Title	Cystic Fibrosis: diagnosis and management
NICE Reference	NG78
Date of Review:	July 2018
Date of Publication	October 2017
Summary of Guidance (Max 250 words)	**Diagnosis** Cystic fibrosis can be diagnosed based on the following: • Positive test results in asymptomatic people, for example infant screening followed by sweat and gene tests for confirmation **or** • Clinical manifestations, supported by sweat or gene tests for confirmation **or** • Only clinical manifestations, in the rare case of symptomatic people with normal sweat or gene test results **Management** Care for cystic fibrosis patients should be provided at specialist cystic fibrosis centres by a specialist multidisciplinary team. The aim of care is to prevent or limit symptoms and complications of the condition. Annual patient review should include: • Pulmonary assessment • Nutritional assessment: -Total nutritional intake review -Testing for exocrine pancreatic insufficiency, using a non-invasive technique such as faecal elastase estimation -Offer pancreatic enzyme replacement therapy to patients with exocrine pancreatic insufficiency • Liver disease assessment: -Clinical assessment and liver function blood tests -Liver ultrasound scan to be performed if liver function tests are abnormal and consider ursodeoxycholic acid treatment -Consider referral to a liver specialist if liver function tests are persistently abnormal despite treatment with ursodeoxycholic acid • Testing for cystic-fibrosis-related diabetes, from 10 years of age -Cystic-fibrosis-related diabetes can be diagnosed using continuous glucose monitoring (CGM) **or** serial glucose testing over several days **or** oral glucose tolerance testing (OGTT) • Assessment for other cystic fibrosis complications • Psychological assessment • Exercise review • Assessments by a specialist nurse, physiotherapist and social worker More frequent reviews should be provided for cystic fibrosis patients immediately after diagnosis and in early life.
Impact on Lab (See below)	☐ Moderate

Lab professionals to be made aware	✓ Laboratory Manager ✓ Chemical Pathologist ✓ Clinical Scientist ✓ Biomedical Scientist
Please detail the impact of this guideline (Max 150 words)	Laboratories should be aware of the following: • The use of sweat tests to support diagnosis of cystic fibrosis • The use of faecal elastase estimation to test for exocrine pancreatic insufficiency in known cystic fibrosis patients • Cystic-fibrosis-related diabetes and its diagnosis

Impact on Lab

- **None**: This NICE guideline has no impact on the provision of laboratory services
- **Moderate**: This NICE guideline has information that is of relevance to our pathology service and may require review of our current service provision.
- **Important:** This NICE guideline is of direct relevance to our pathology service and will have a direct impact on one or more of the services that we currently offer.

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