Cystic Fibrosis Foundation Evidence-Based Guidelines for Management of children with Cystic Fibrosis

Introduction

- Autosomal recessive
- Defect in long arm of ٧th chromosome
- Cystic fibrosis transmembrane conductance regulator(CFTR)
- More than ١٩٠٠ mutation in CFTR
- Active in epithelium of lungs,GI,liver, pancreas, urogenital system, seat glands

Cont

- Deletion of three nucleotids leads to loss of a single phenylalanin residue at codon ٥٠٨(ΔF٥٠٨)
- ٧٠% OF CF alleles in northern europea.
- Rare in native asians and africans

Cont

- Abberant function of CFTR leads to reduced chloride conductance and dysregulation of sodium hyperabsorption.
- Failure of chloride secretion and sodium hyperreabsorption leads to dehydration of airway surface.

Cont

- Decrease mucocilliary clearance
- Increase of bacterial colonization
- Exessive production of lung cytotoxins
- Chronic bronchitis
- Bronchiectasis
- Respiratory failure
- Death

Diagnosis

BOX 51-1 Diagnostic Criteria for Cystic Fibrosis (CF Foundation Consensus Panel)

One or more characteristic phenotypic features consistent with CF:

- Chronic sinopulmonary disease
- Gastrointestinal and nutritional abnormalities
- Salt loss syndromes
- Male urogenital abnormalities resulting in obstructive azoospermia

OR

A history of CF in a sibling

OR

A positive newborn screening test result

AND

An increased sweat chloride concentration

OR

Identification of two CF mutations

OR

Demonstration of abnormal nasal epithelial ion transport

Pulmonary Recommendations

The CF Foundation recommends:

- a smoke-free environment be provided and that all caregivers are informed that cigarette smoke exposure harms children with CF.

Infection Control, Surveillance and Treatment

The CF Foundation recommends:

- newly diagnosed patients should be separated from other patients cared for in CF clinics.

Infection Control, Surveillance and Treatment

The CF Foundation recommends

- use of nirsevimab be considered for prophylaxis of respiratory syncytial virus.
- Annual influenza vaccination is recommended for infants with CF > ٦ months of age, all household members, and all healthcare providers caring for these infants.

Airway Clearance

The CF Foundation recommends:

- airway clearance therapy(ACT) be initiated in the first few months of life.
- use of albuterol before percussion and postural drainage.

Infection Control, Surveillance and Treatment

The CF Foundation recommends:

- oropharyngeal cultures should be performed at least quarterly.
- bronchoscopy and bronchoalveolar lavage be considered in infants who fail to respond to appropriate intervention

Infection Control, Surveillance and Treatment

The CF Foundation recommends:

- against the prophylactic use of oral antistaphylococcal antibiotics in asymptomatic infants.
- there is insufficient evidence to recommend for or against active attempts to eradicate Staphylococcus aureus and MRSA in asymptomatic infants.

Infection Control, Surveillance and Treatment

The CF Foundation recommends:

- against the use of chronic antibiotics for prophylaxis to prevent Pseudomonas aeruginosa.
- new acquisition of Pseudomonas aeruginosa, should be treated with anti-pseudomonal antibiotics and increased airway clearance, regardless of the presence or absence of symptoms.

Infection Control, Surveillance and Treatment

The CF Foundation recommends:

- infants who remain persistently colonized with Pseudomonas aeruginosa after two attempts at eradication be treated chronically with alternate month tobramycin solution for inhalation.

Chronic Pulmonary Therapies

The CF Foundation recommends:

- there is sufficient evidence to recommend for the routine use of chronic azithromycin in patients colonized with Pseudomonas.

Chronic Pulmonary Therapies

The CF Foundation recommends:

- dornase alfa (recombinant human DNase) may be used in symptomatic infants.
- ٧% hypertonic saline may be used in symptomatic infants.

Chronic Pulmonary Therapies

The CF Foundation recommends:

- without airway reactivity or asthma,

  use of inhaled corticosteroids to improve lung function or reduce exacerbations is not effective.

CFTR molecular therapies

- Ivacaftor
- Lumcaftor/ivacaftor
- Tezacaftor/ivacaftor
- Eiexacaftor/tezacaftor/ivacaftor

Diagnostic Testing

The CF Foundation recommends:

- a baseline chest x-ray should be obtained within the first ٣ - ٦ months and once again within the first two years of life.

Diagnostic Testing

The CF Foundation recommends:

- against the use of chest CT scans for routine surveillance.
- chest CT scans be considered in infants with symptoms or signs of lung disease who fail to respond to appropriate interventions.

Diagnostic Testing

The CF Foundation recommends:

- PFTs be considered as an adjunctive tool to monitor respiratory status.

Diagnostic Testing

The CF Foundation recommends:

- there is insufficient evidence to recommend for or against use of pulse oximetry routinely as an adjunctive tool to detect lung disease.
- pulse oximetry measurements be obtained in the infant with CF with acute respiratory symptoms.