

Advances in CF Testing

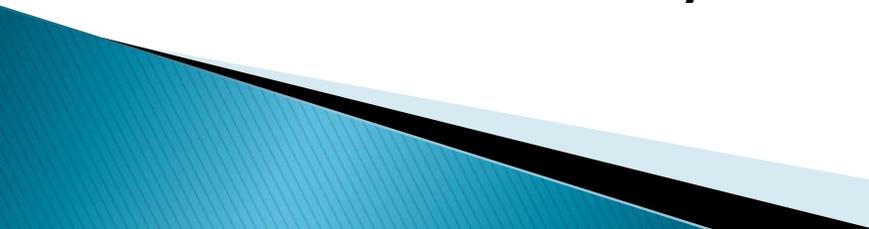
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Updates in Pediatric Pulmonary Care XII:

An Interdisciplinary Program

April 13, 2012

Objectives

- ▶ Recognize the importance of Pulmonary Function Testing in Cystic Fibrosis
 - ▶ Be aware of different types of Pulmonary Function Tests
 - Why and for what age groups
 - ▶ Be familiar with Bronchoscopy
 - ▶ Understand why and when we use Bronchoscopy in Cystic Fibrosis
 - ▶ Understand the reasons for using CT scans of the chest in Cystic Fibrosis
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Pulmonary Function Tests (PFTs)

- ▶ What are pulmonary function tests?
- ▶ A group of tests that measure:
 - How well the lungs take in and release air
 - How well they move gases such as oxygen from the atmosphere into the body's circulation



Pulmonary Function Tests (PFTs)

- ▶ Why do we perform pulmonary function tests?
 - To diagnose certain types of lung disease
 - Measure the progress in disease treatments
- ▶ Can diagnose obstructive airway disease or restrictive lung disease



Types of Pulmonary function tests

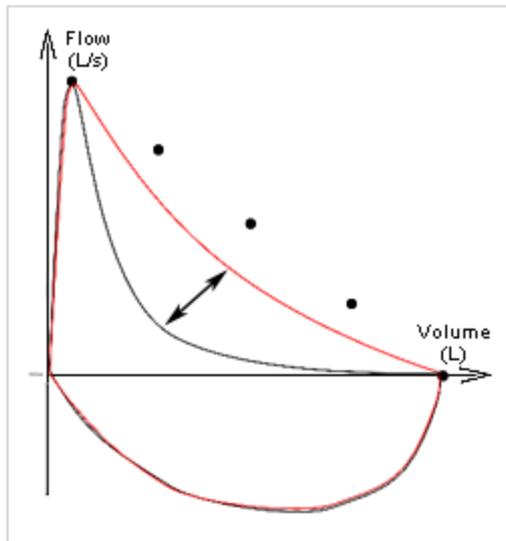
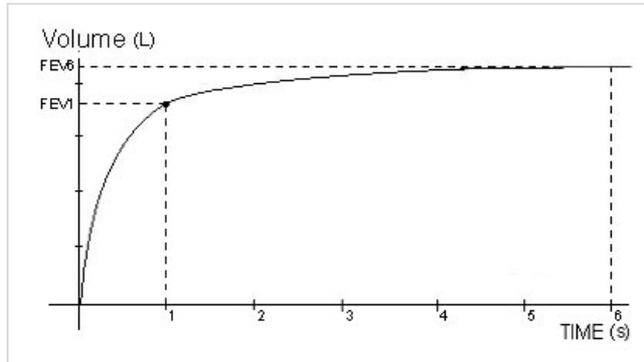
- ▶ Spirometry
 - ▶ Plethysmography
 - ▶ Impulse oscillometry
 - ▶ Infant pulmonary function testing
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Spirometry

- ▶ Measures airflow
- ▶ Measures how much air is exhaled and how quickly



Spirometry



- ▶ **Forced Vital Capacity (FVC):**
 - The amount of air which can be forcibly exhaled from the lungs after taking the deepest breath possible
- ▶ **Forced expiratory volume in 1 second (FEV1)**
 - The maximal amount of air you can forcefully exhale in one second

Spirometry normal and abnormal values

Measured Test	Normal	Mild	Moderate	Severe
FVC	More than 80%	60 - 80%	40 - 60%	Less than 40%
FEV1	More than 80%	60 - 80%	35 - 60%	Less than 35%
FEF25-75	More than 65%	50 - 65%	30 - 50%	Less than 30%

Spirometry and Cystic Fibrosis

- ▶ Important to show us how lung function changes over time
 - ▶ Spirometry changes over time, between two clinic visits or over any period of time reveal how a person's cystic fibrosis is changing
 - ▶ However, we can only use the spirometry results if they can be done correctly and the results are 'reproducible'
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Spirometry and Cystic Fibrosis

- ▶ Clinically significant Spirometry changes:

Measured Test	% Change To Be Significant
FVC	10% or greater
FEV1	10% or greater
FEF25-75	30% or greater

- ▶ However, we want to stop any decline early so we may act if there are more subtle changes

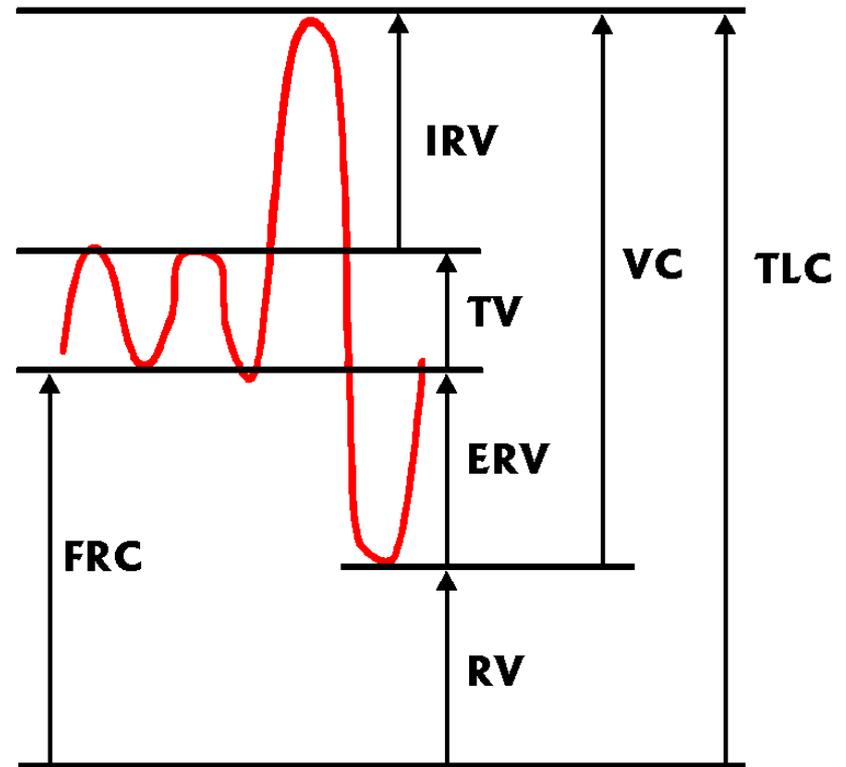
Plethysmography

Gives us information on more lung volumes, specifically total lung capacity and functional residual capacity



Plethysmography

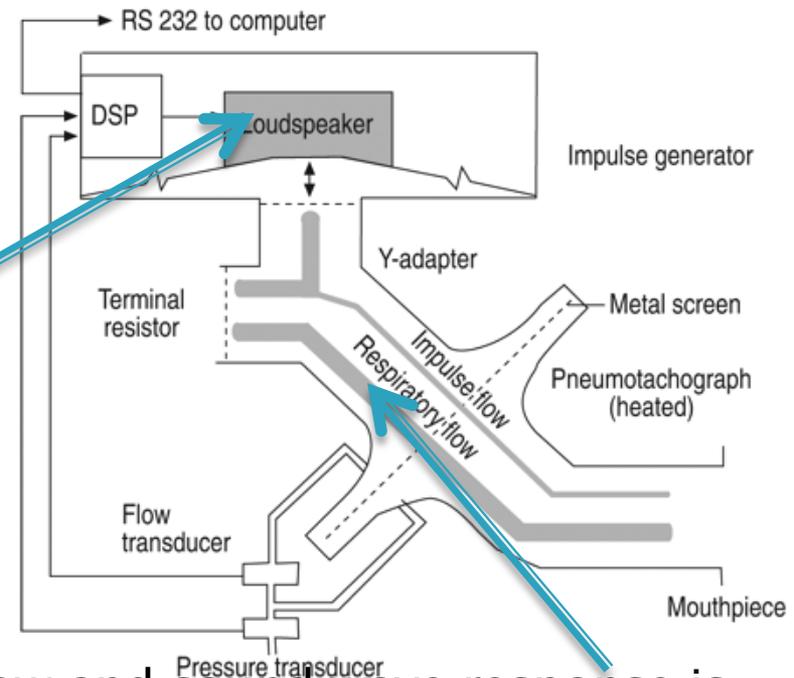
- ▶ **Functional residual capacity (FRC):**
 - The volume in the lungs when the muscles of respiration are relaxed – at the end of a normal exhalation.
 - If it's too high – air trapping: obstruction in the airways which can occur in cystic fibrosis
- ▶ **Total lung capacity (TLC):**
 - The total amount of air in the lungs after a maximal inhalation



Impulse Oscillometry (IOS)

- ▶ Easy to use PFT
- ▶ Uses sound waves to detect airway changes

Breath through a pneumotachograph – a sound wave is generated by a loudspeaker superimposed over breathing



The airflow and sound wave response is transmitted to the apparatus and used to calculate various components of resistance to breathing

Impulse Oscillometry (IOS)

- ▶ Nothing more than quiet breathing is required from the patient
- ▶ Can be used for children 3 years old and above

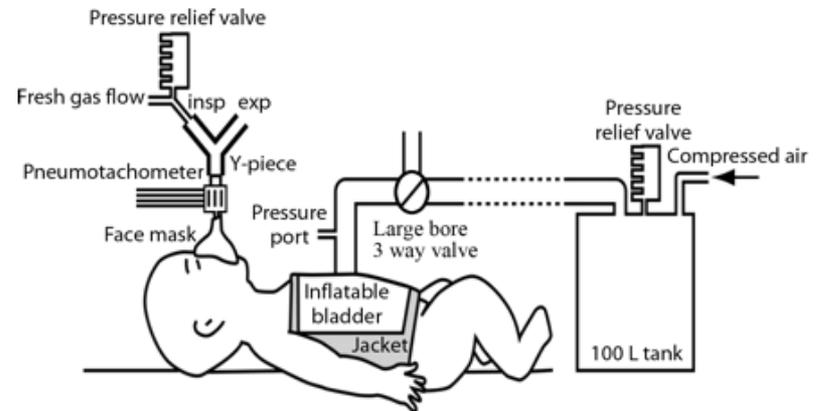


Impulse Oscillometry (IOS)



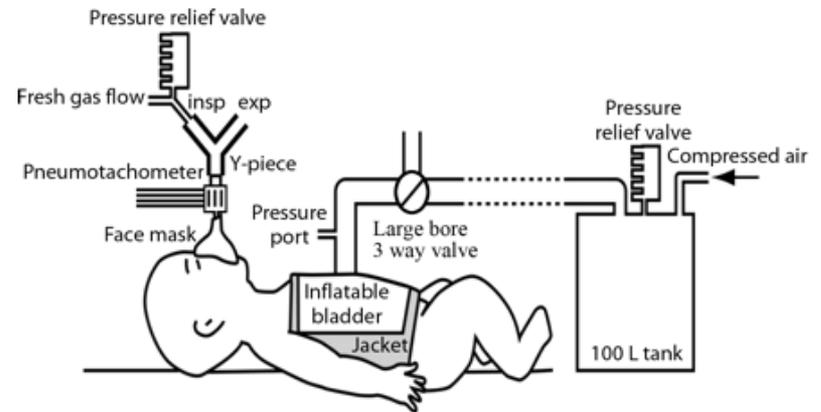
Infant Pulmonary Function Testing

- ▶ Measures breathing of babies and toddlers
- ▶ Raised volume rapid thoracoabdominal compression technique (RVRTC)



Infant Pulmonary Function Testing

- ▶ Infant is sedated
- ▶ Lungs are passively inflated towards total lung capacity
- ▶ Thoraco–abdominal compression pressure is applied to force expiration
- ▶ Performed twice
- ▶ $FEV_{0.5}$ and FVC



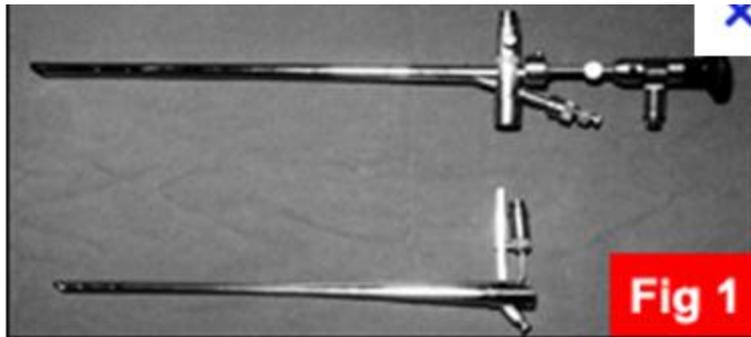
Pulmonary Function tests

- ▶ Infants
 - Infant PFTs
 - RVRTC
- ▶ Children 3–5
 - Impulse oscillometry
- ▶ Children older than 5
 - Spirometry
 - Plethysmography



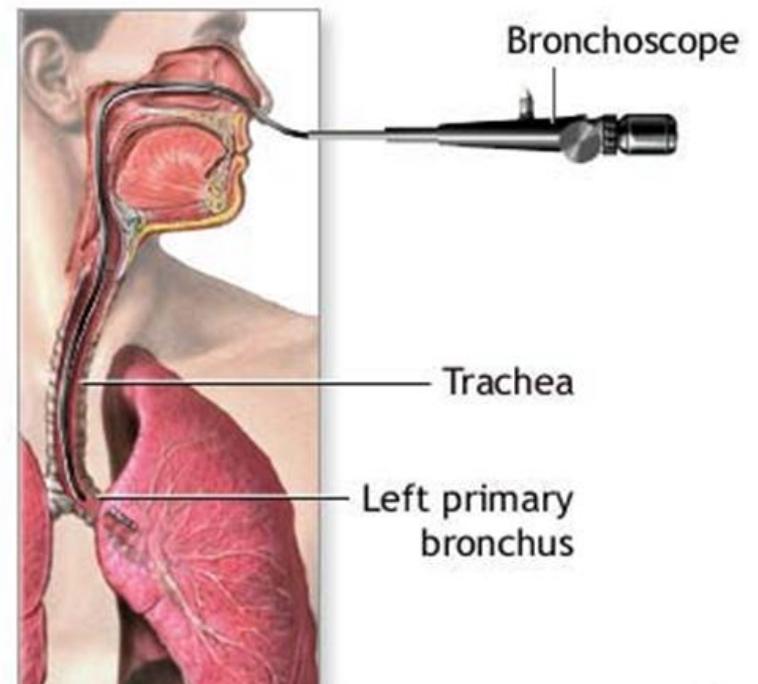
Bronchoscopy

- ▶ Flexible bronchoscopy
- ▶ Rigid bronchoscopy



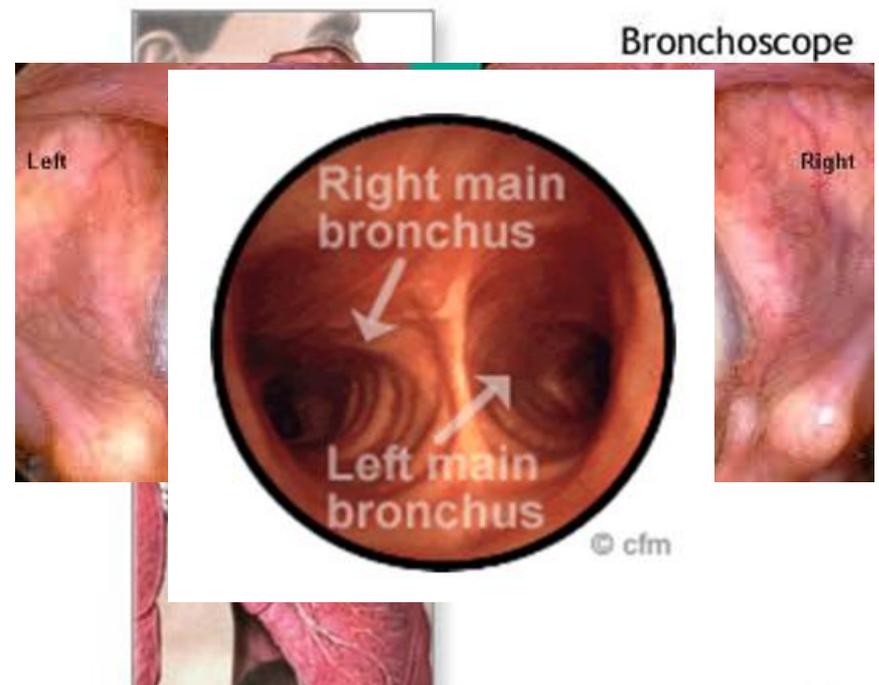
Flexible bronchoscopy

- ▶ Visualize the airways
- ▶ Relatively safe
- ▶ Sedation is necessary



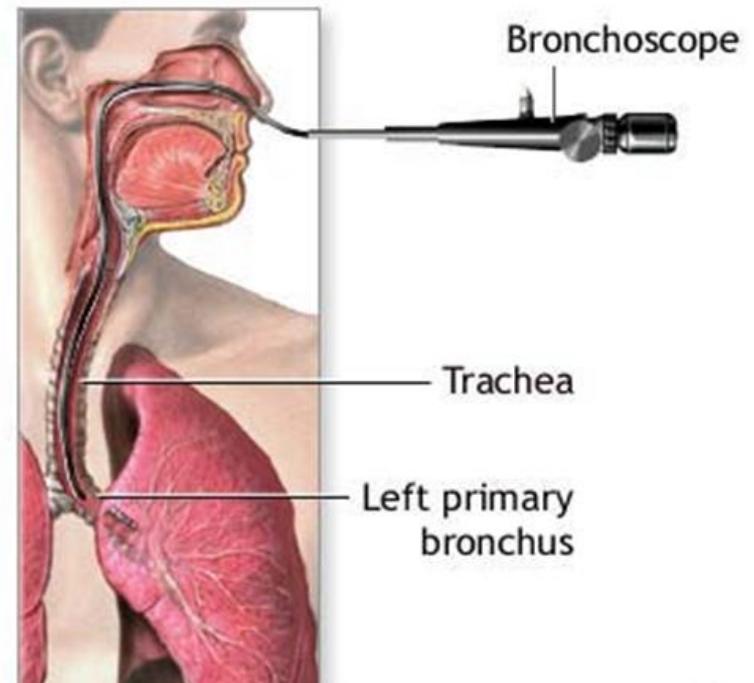
Flexible bronchoscopy

- ▶ Bronchoscope is passed through the nose
- ▶ The nose and upper airway is visualized
- ▶ The vocal cords are seen
- ▶ Passes through the vocal cord to the lungs
- ▶ Samples broncho-alveolar lavage (BAL) obtained
- ▶ The procedure usually takes 10–30 minutes



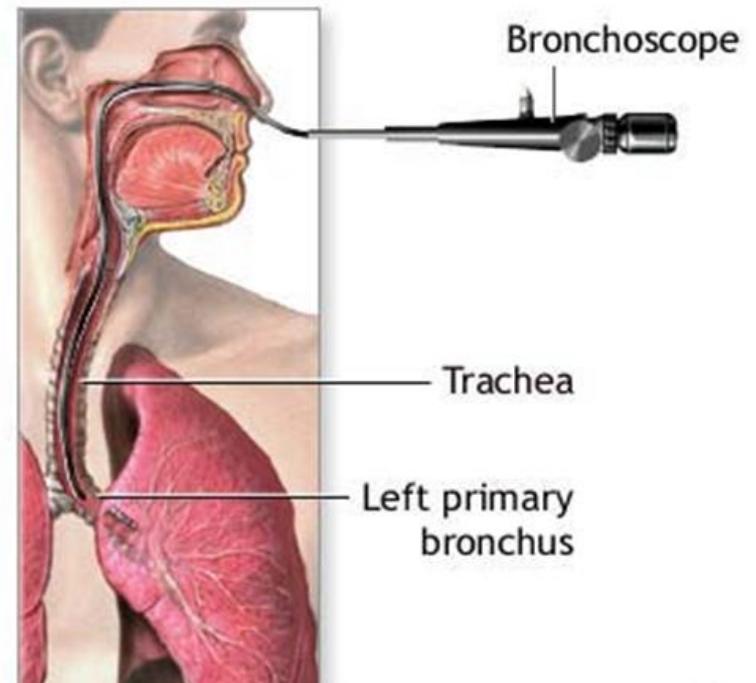
Flexible bronchoscopy and Cystic Fibrosis

- ▶ Useful technique to obtain lower airway specimens (broncho-alveolar lavage (BAL)) from children who are unable to expectorate
- ▶ Pathogens sometimes found in BAL which are not present on throat culture/sputum



Flexible bronchoscopy – Indications

- ▶ New or increased symptoms with no identifiable pathogen on throat swab or sputum
 - Or who have not responded to oral and IV antibiotics
- ▶ To culture atypical mycobacteria



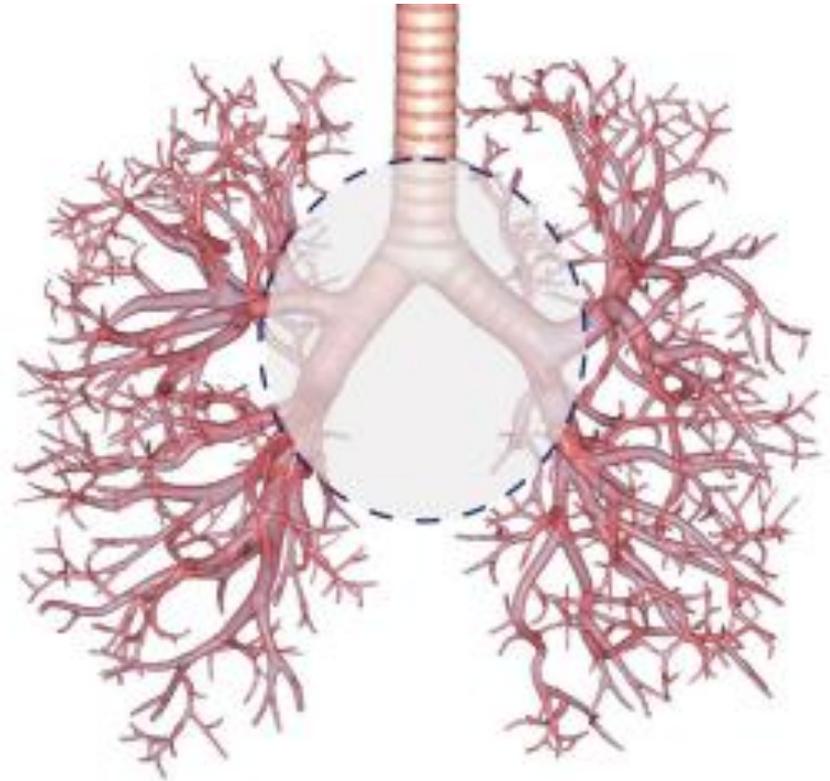
Flexible bronchoscopy – Indications

- ▶ Lobar or segmental collapse in children with CF
 - Remove mucus plugging
 - Exclude unsuspected foreign bodies
 - Obtain samples for microbiology



Flexible bronchoscopy – Indications

- ▶ Children or adults who have undergone lung transplantation
 - Biopsies help to monitor rejection



Chest Computed Tomography (CT)

- ▶ Creates precise pictures of the structures in the chest, including the lungs
- ▶ Painless and non-invasive
- ▶ Chest CT scanning machine takes many pictures/slices of the lungs and inside of the chest
- ▶ Chest CT's can be performed with or without contrast



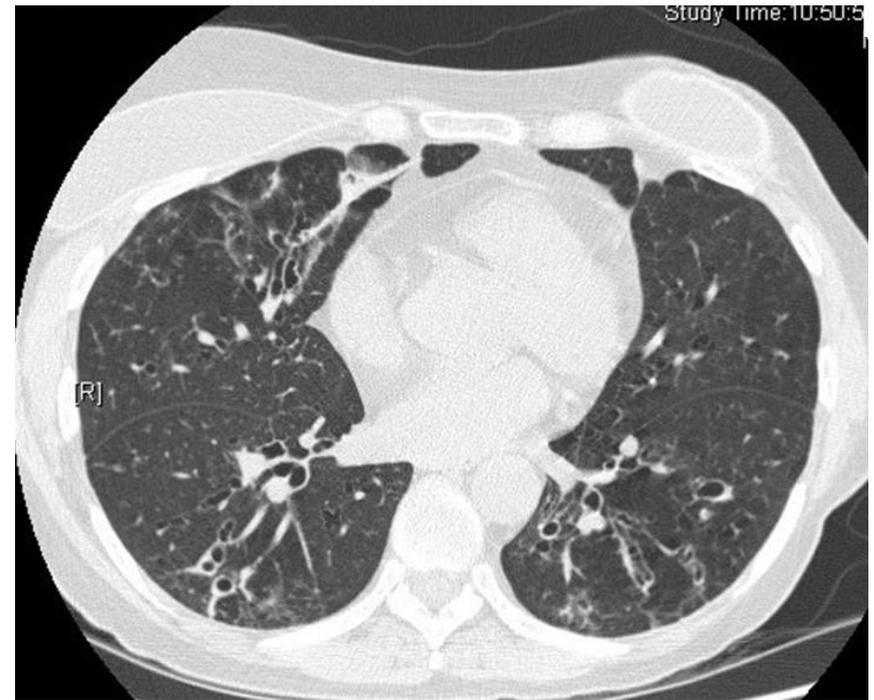
Chest Computed Tomography (CT) and CF

- ▶ CT provides information on the lung structure
- ▶ It is helpful to identify early signs of disease
- ▶ It is more sensitive in detecting changes in small airways



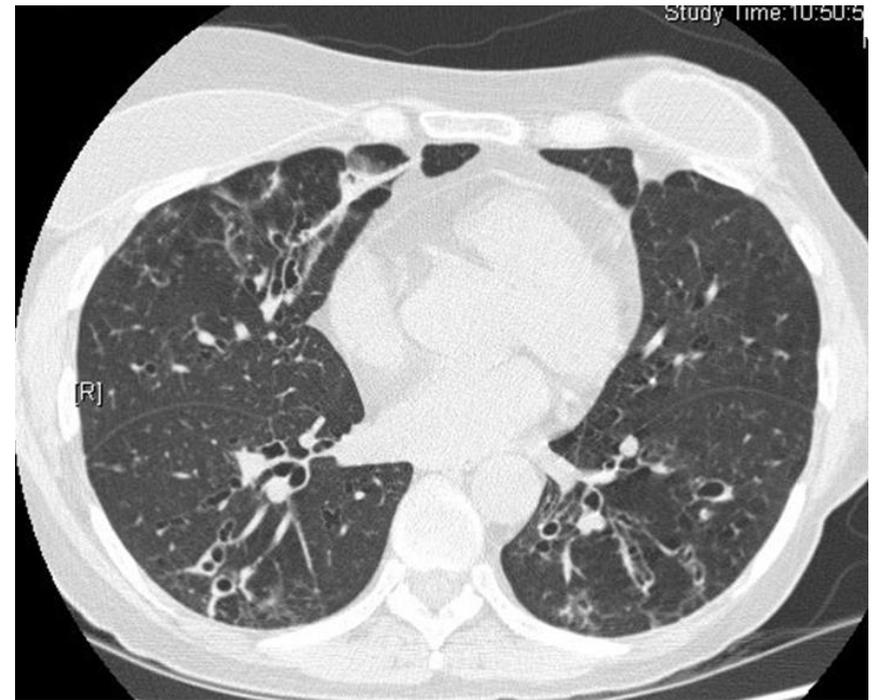
Chest Computed Tomography (CT) and CF

- ▶ The CT is scored using various scoring systems
- ▶ Presence, extent and severity of bronchiectasis, peribronchial thickening, mucous plugging, atelectasis, air trapping.



Chest Computed Tomography (CT) and CF

- ▶ Baseline CT to study the extent of the lung involvement
- ▶ Every few years to examine the lungs and decide if current treatment is optimal
- ▶ or if there is progression of disease, if more treatment is needed



Conclusion

- ▶ Pulmonary Function Testing (PFT) is important to monitor the extent of lung involvement in children with cystic fibrosis
 - ▶ Different types of PFTs are available depending on the age of the child
 - ▶ Bronchoscopy is an important way to look at the airways and obtain cultures when a child cannot expectorate sputum or is not responding to oral or IV antibiotics
 - ▶ Chest CTs are helpful to evaluate and follow the extent of lung involvement and are more sensitive for detecting small airway involvement
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Thank you

