

# Obstructive Lung Disease in Patients with Congenital and Syndromic Scoliosis

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Obstructive Lung DZ



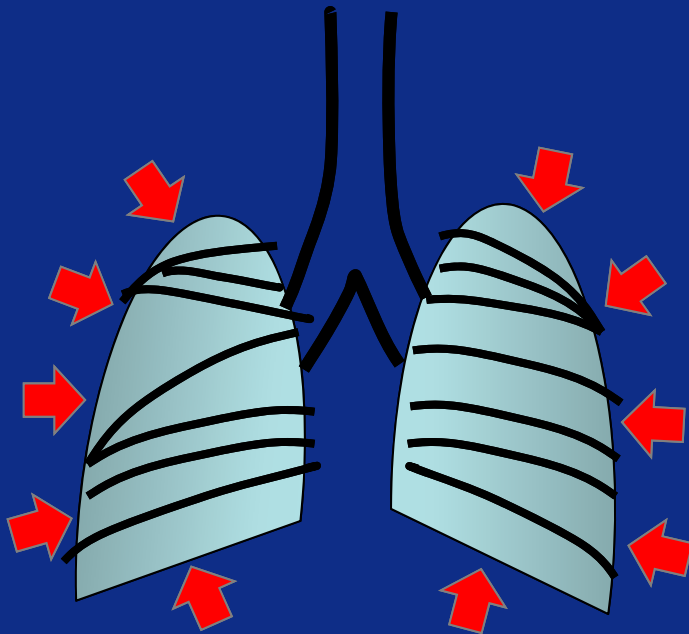
# Is Scoliosis Associated with Airway Disease?

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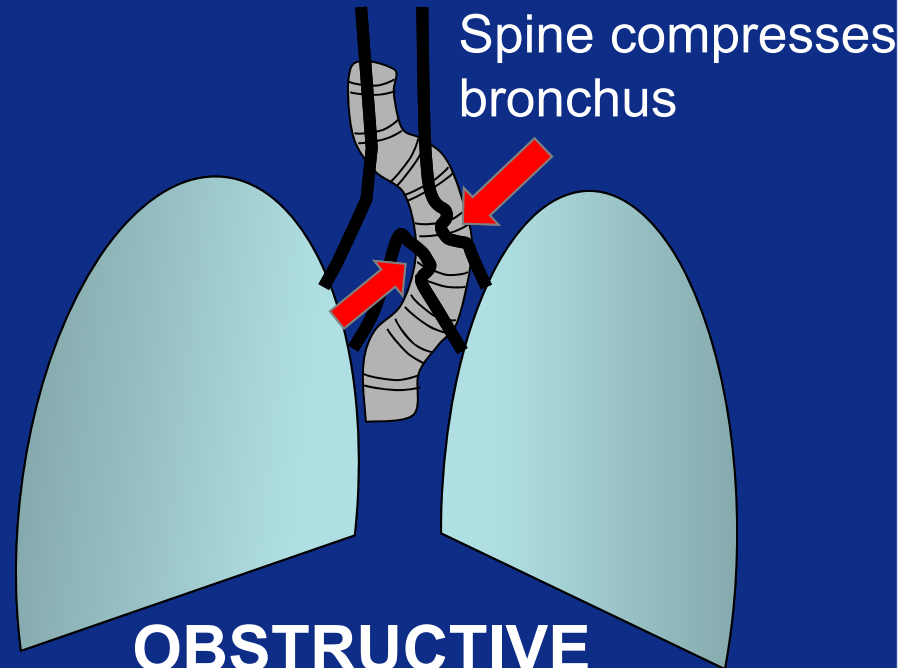


↓ FVC



RESTRICTIVE  
(Small lung cage  
restricting lung  
capacity)

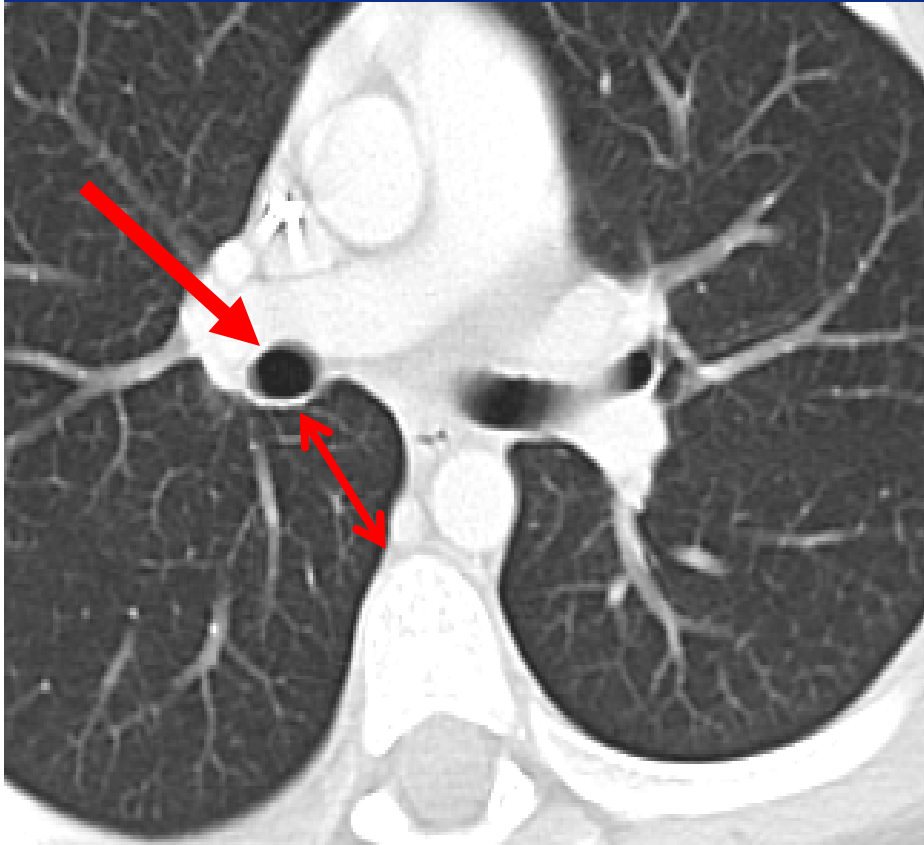
↓ FEV1/FVC



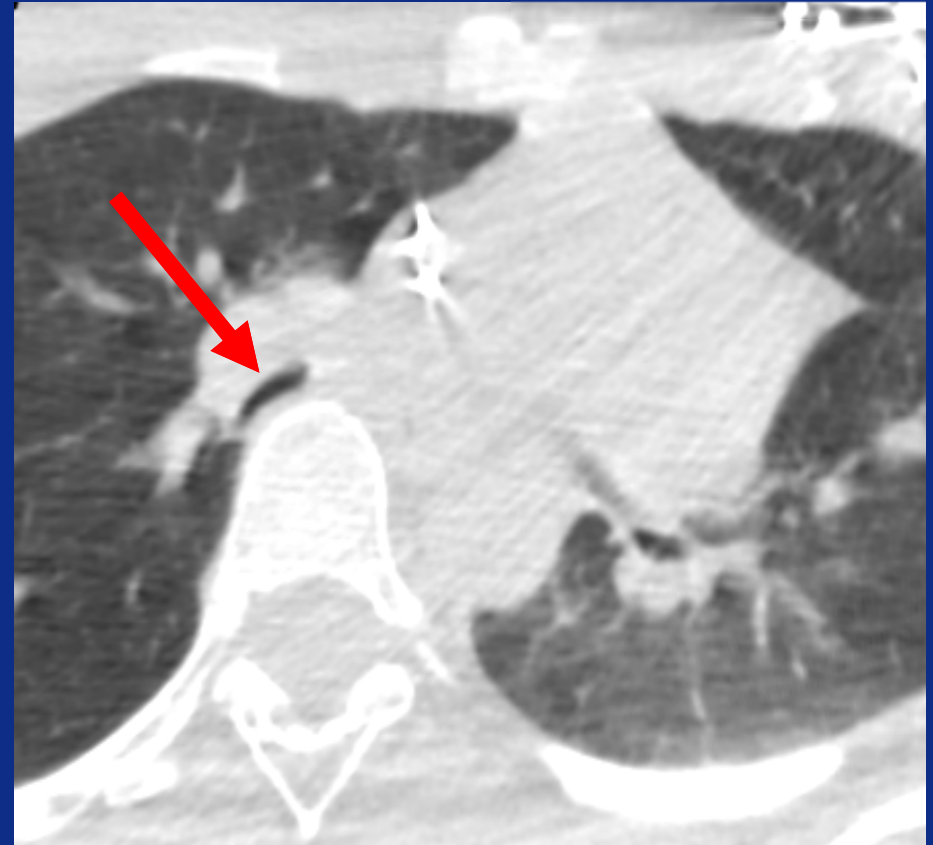
OBSTRUCTIVE  
(Normal size  
lungs, but partial  
bronchus  
obstruction)

# Obstructive Lung Disease (airways obstruction)

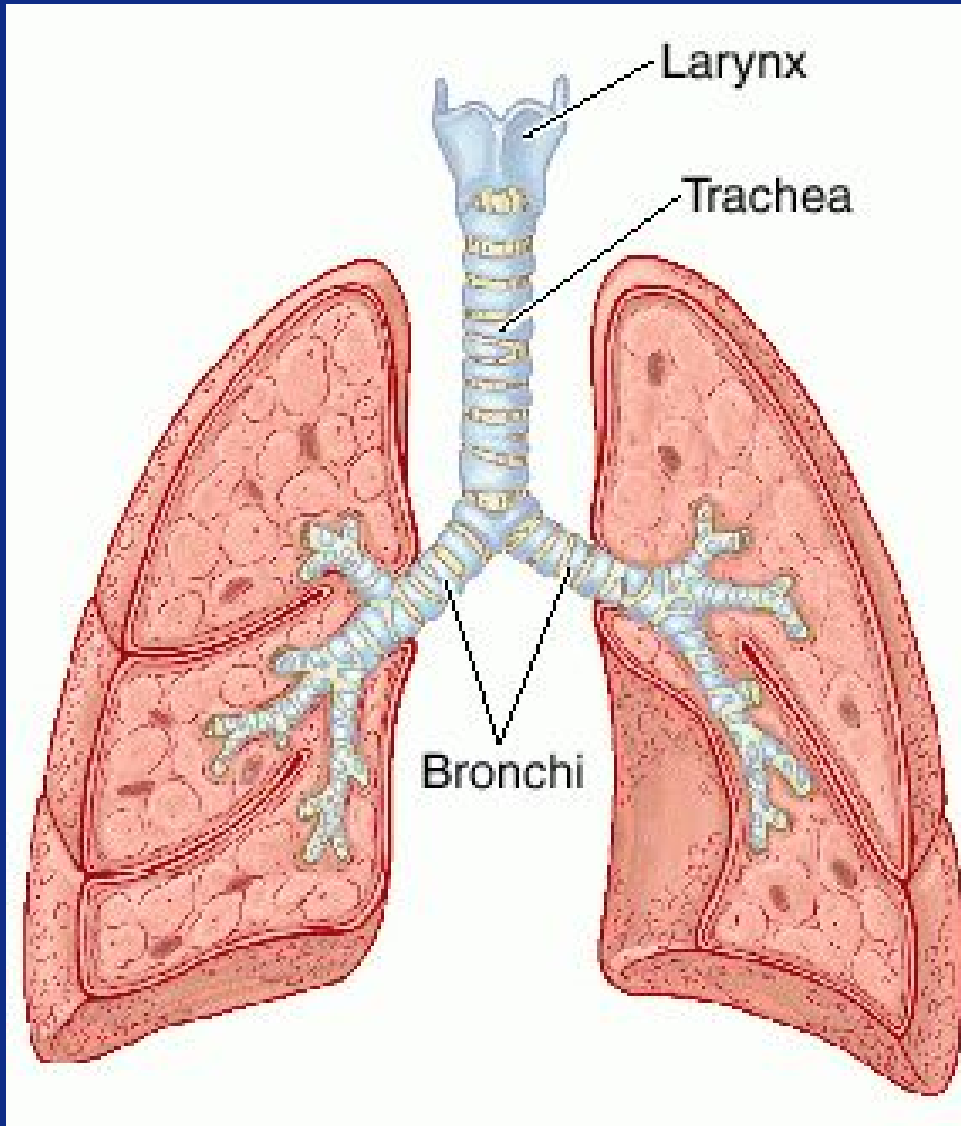
- Airways disease
  - Asthma
  - Cystic fibrosis
  - Airway compression
- Difficulty exhaling gas quickly from the lungs due to obstructed airways  
(air does not come out fast)



Normal bronchus remote  
from spine



Obstructed bronchus  
compressed over spine



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<http://www.volny.cz/martinam/im.v/trachea.jpg>



# Cincinnati Children's Data 2004-2009

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# Inclusion Criteria

- Congenital or Syndromic Scoliosis
- Cobb angle  $\geq 40^\circ$
- Pre-operative Pulmonary Function Testing



# Exclusion Criteria

- Asthma
- Cystic Fibrosis
- Airway anomalies

# Methods

- Queried local database for patients with scoliosis X-rays and Pulmonary Function Testing from 2004-2009
- Reviewed Pulmonary Function Testing for all patients with Congenital or Syndromic Scoliosis and Cobb angles  $\geq 40$  degrees

# Methods

- Defined Obstructive Lung Disease per consensus American Thoracic Society criteria ( $FEV_1/FVC$  ratio below the 95% confidence interval)
- Prevalence of Obstructive Lung Disease in a reference population using this definition is 2.5%

# Results

- N = 26 patients
- Syndromic diagnoses included:
  - NF-1
  - Marfan Syndrome
  - Klippel-Feil
  - Schwachmann-Diamond Syndrome
  - Diastrophic Dysplasia

# Results

- Median age at scoliosis diagnosis: 11 years
- Median primary Cobb angle: 55 degrees

# Results

- Prevalence of Obstructive Lung Disease (airway disease) was 27% (7/26)
- Prevalence of Restrictive Lung Disease (small lung volume) was 53% (14/26)

# Is a Mechanism of the Obstructive Lung Disease (OLD) Large Airway Compression?

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# Methods

- 5 of 7 patients with OLD had flexible bronchoscopy and CT scan evaluations
- Flexible bronchoscopy reports and videos were reviewed to define the anatomy of the trachea and mainstem bronchi
- CT scans were reviewed to compare large airway findings on CT with large airway findings on flexible bronchoscopy



# Findings-Bronchoscopy

- Each of the 5 bronchoscopies revealed mainstem bronchial compression from the posterior airway wall on the side of the major spinal curve



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# Findings-Chest CT

- Patients with right-sided major curves had compression of the right mainstem bronchus between the spine (posterior) and the right pulmonary artery (anterior).
- Patients with left-sided major curves had compression of the left mainstem bronchus between the spine (posterior) or descending aorta (posterior) and left

# Conclusions

- Obstructive lung disease (airway disease) is common in children with Congenital or Syndromic scoliosis who have a Cobb angle  $\geq 40^\circ$
- Airway compression from lordoscoliosis a common possible mechanism for this finding



Thank you

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Peter Strurm