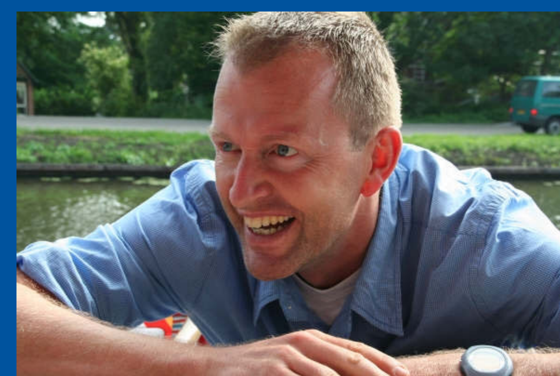
Wilhelmina Children's Hospital

Idiopathic scoliosis:
The growing spine and growing lung: chicken and egg……?

Bert Arets, MD, PhD
Associate Professor
Pediatric Pulmonologist
WKZ Children's Hospital
UMCU University Medical Center Utrecht



Conflict of interest disclosure

I have **no**, real or perceived, direct or indirect conflicts of interest that relate to this presentation.

………………but sometimes I, my wife and/or children have other interests!

To Take Home

- Lungs and thoracic cage/vertebrae grow dependently

- Pulmonary scoliosis exists……………… but in that case scoliosis is not idiopathic!

- Scoliotic lungs give problems, starting from 40 degree curves

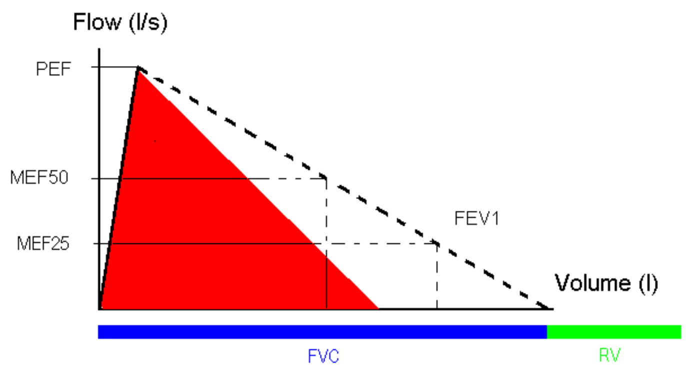
- Lung function is only part of "pulmonary well being"

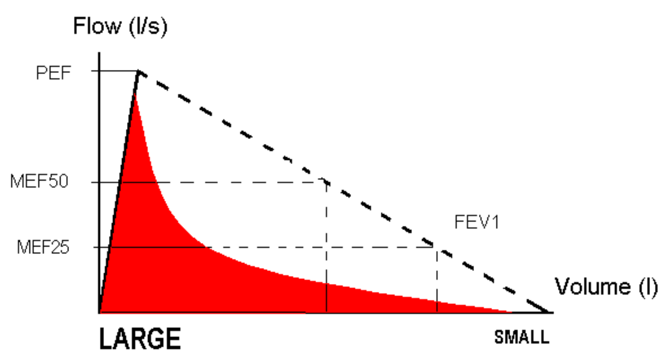
- The effect of scoliosis correction on lung function is
  - Limited
  - Better for "thoraco-abdominal surgery" than "anterior surgery"
  - small compared to "functional improvement" (QoL, pain, "nursing")

- No surgery to improve lung function…….

To Take Home: we have a lot in common!

- **Always look at curves!!!!!**





Scoliosis and lung (growth), what are the questions?

1. What is the relation between lung and bone development

   How does the lung grow??

2. What should we measure?

   Lung function testing

3. Chicken and/or egg?

   - does impaired/asymmetric lung growth lead to scoliosis?

     "pulmonary scoliosis"?

   - (how) does scoliosis lead to lung function abnormalities?

     "scoliotic lungs"?

4. Does scoliosis repair improve lung function?

How do the lungs grow?? (1)

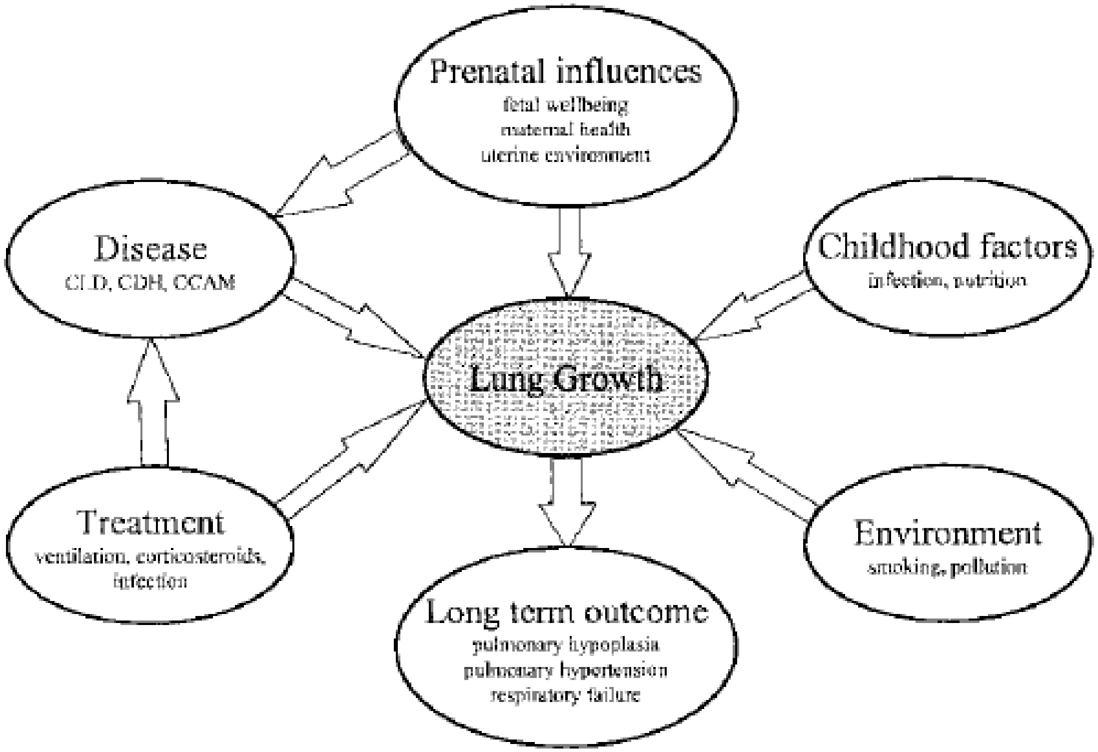


Figure 2 A model depicting the factors which may contribute to normal and abnormal lung growth. Please note the importance of both pre- and post-natal factors to lung growth.

- Congenital
  - Malformations
    - Agenesis
    - Hypoplasia
    - Diaphragmatic hernia
- Prenatal:
  - Fluid filling
  - Movements
  - Wellbeing
- Postnatal
  - Infections
  - Body and thoracic growth
  - Smoking
  - Injury (radiation, pollution,

Kotecha et al., Paed Resp Rev 2000

How do the lungs grow?? (2)

PRACTICE POINTS

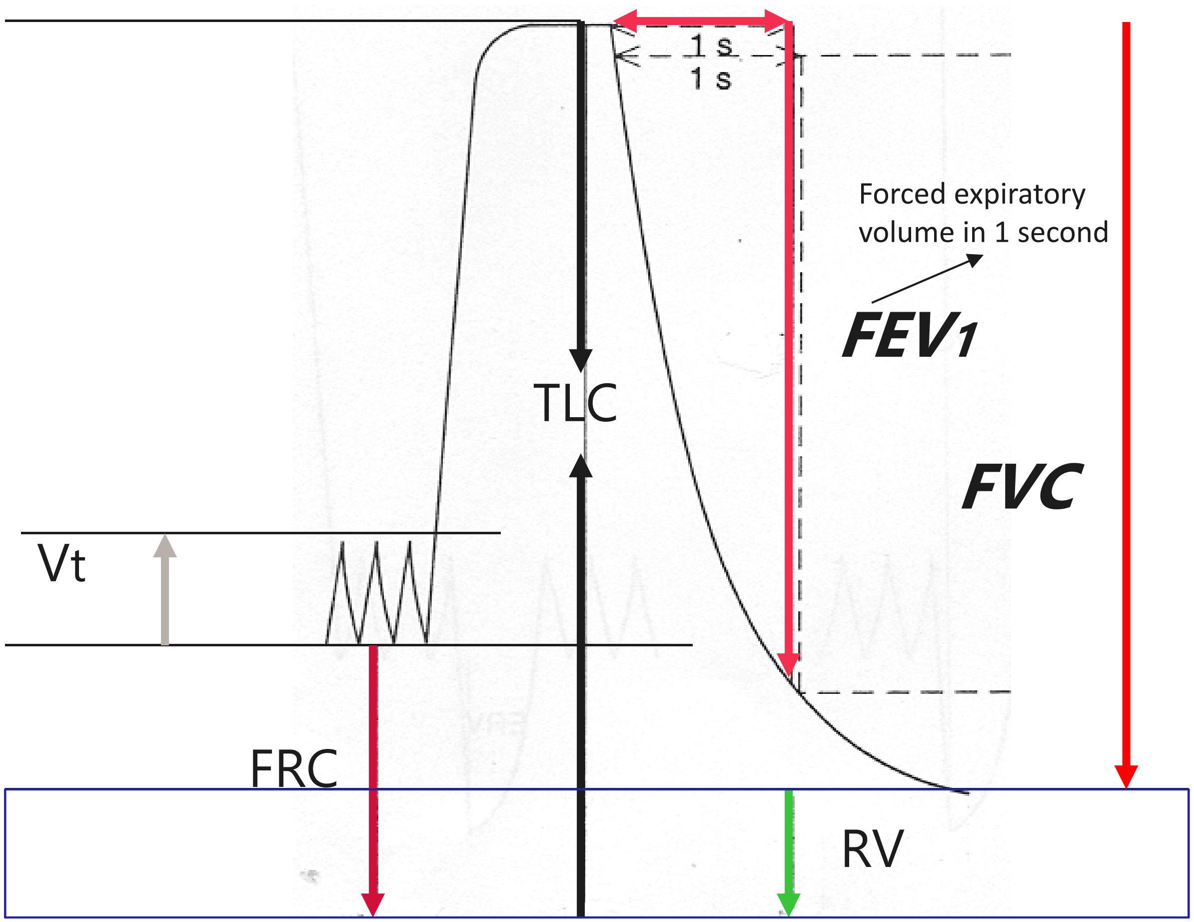
Normal lung growth depends on a number of inter-related factors, including:

- Normal embryonic and fetal development
- Genetic constitution
- Maternal and fetal nutrition
- Endocrine factors
- Fetal breathing movements
- Normal fetal lung fluid production
- Adequate intra-thoracic space
- Adequate extra-throacic space
- Normal post-natal adaptation

The above list is not exhaustive. Lung growth depends on many factors which may directly or indirectly affect lung growth.

Kotecha et al., Paed Resp Rev 2000

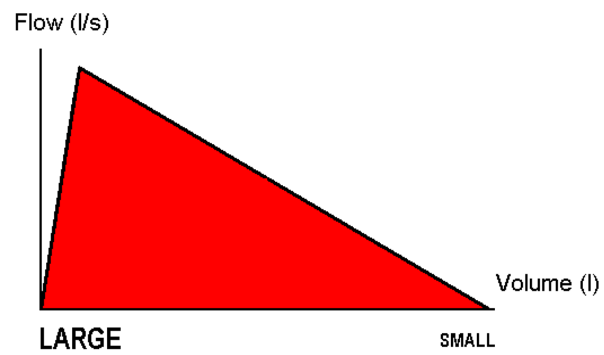
Lung function: more than spirometry!

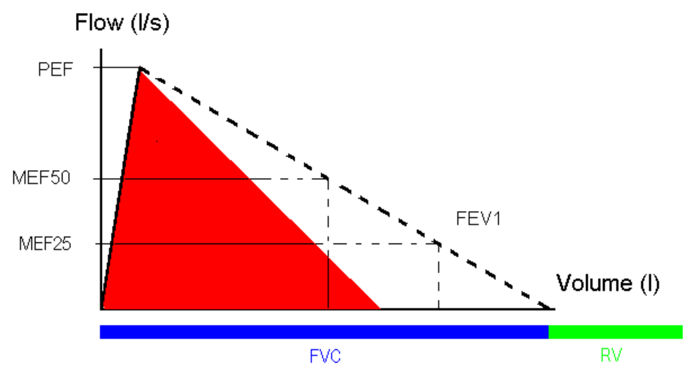


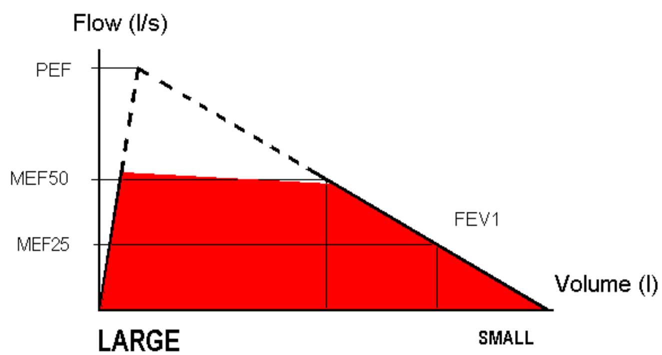
- Obstruction:
  - FEV1
  - FVC
  - FEV1/FVC
  - RV/TLC
- Restriction
  - TLC
  - (FVC)

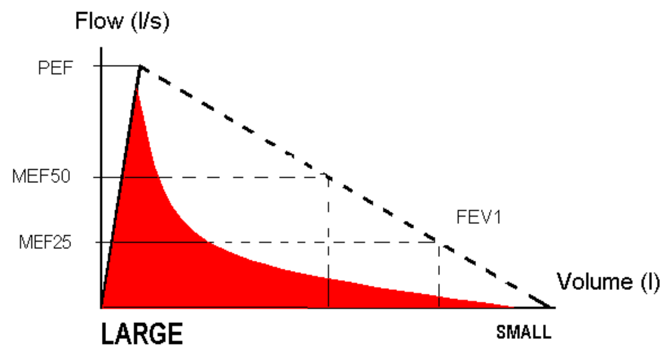
Lung function measurements like scoliosis assessment: always look at curves!!

Lung function measurements like scoliosis assessment: always look at curves!!





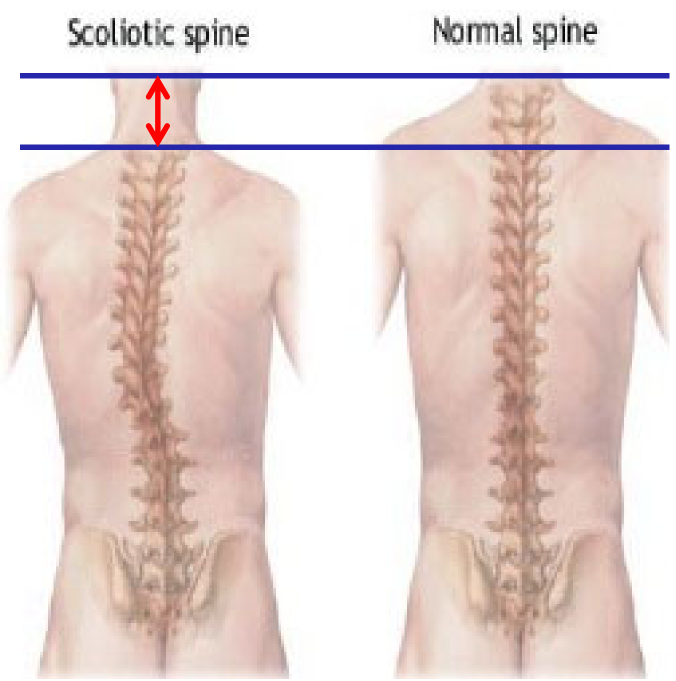




Lung function

Warning!!

- Most predictive values for lung function depend on height!!!!!
- Scoliotic people are “smaller”, so predicted values are too low!!!!!!
- → use span width to predict lung function



Pulmonary scoliosis……

How idiopathic is idiopathic scoliosis??

- No cause for idiopathic scoliosis (by definition)
  - But associations might show etiology??

- Moderate evidence for
    - impaired gait control (e.g. after polio)
    - decreased bone minerals
    - Breast asymmetry
    - Cortical thinning of right hemisphere

No data on lung growth or -function……

Schlosser et al. PLoS One, 2014

Non-idiopathic scoliosis

- Several congenital or acquired lung abnormalities are related to abnormal thoracic/vertebral development
  - Asymmetric lung development
    - (congenital)
      - Lung agenesis
      - Lung anomalies
    - acquired
      - After surgery or damage

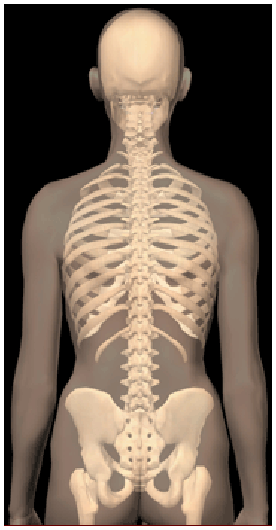
Table I congenital malformations or diseases associated with abnormal lung development and the stage of their development.

Embryonic
Pulmonary agenesis
Tracheal or laryngeal agenesis or stenosis
Tracheo- or broncho–malacia
Bronchial malformations
Ectopic lobes
A-V malformations
Congenital lobar cysts
Pseudoglandular
Cystic adenomatoid malformation
Pulmonary sequestration
Lung hypoplasia
Lung cysts
Congenital pulmonary lymphangiectasia
Congenital diaphragmatic hernia
Canalicular
Lung hypoplasia
Respiratory distress syndrome
Acinar dysplasia
Saccular/Alveolar
Pulmonary hypoplasia
Respiratory distress syndrome/Chronic Lung disease of prematurity
Acinar dysplasia
Alveolar capillary dysplasia

Pulmonary scoliosis?

Kotecha et al., Paed Resp Rev 2000

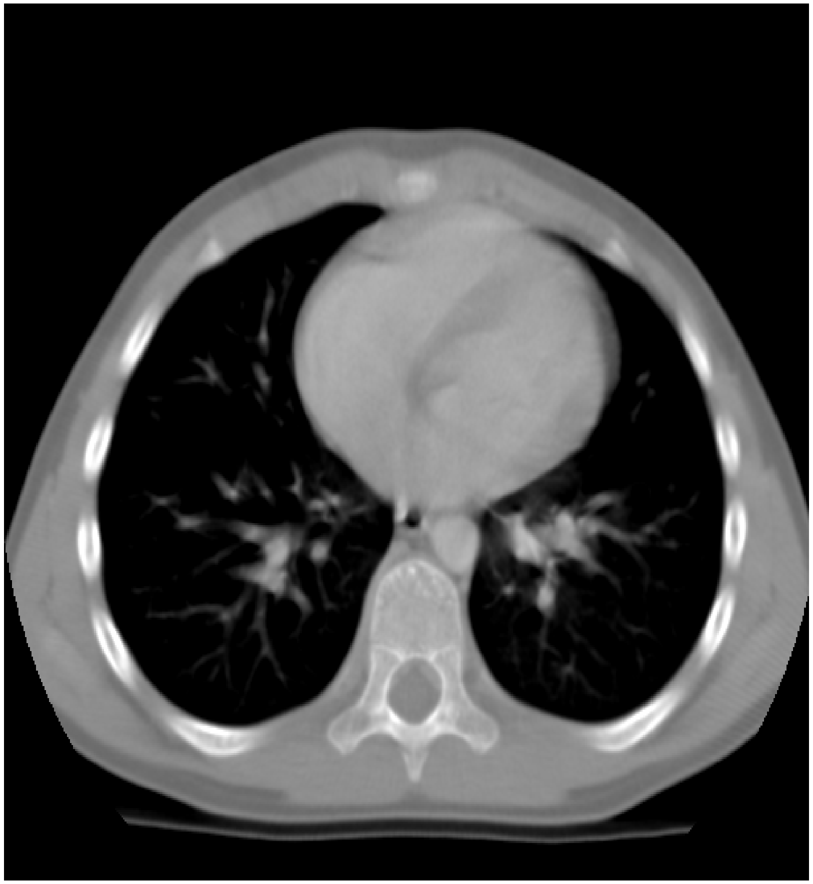
Deformed vertebral columna and the thoracic cage



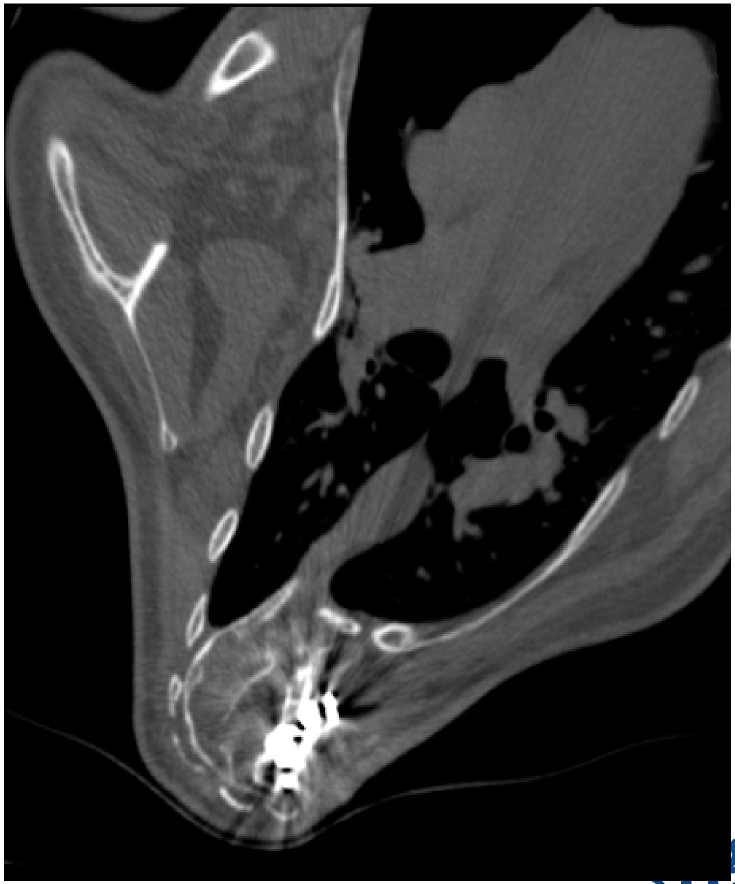
3% of the population

Scoliosis and the lung

- Normal

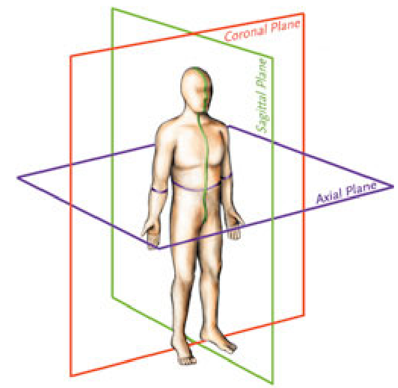


Severe (kypho)scoliosis



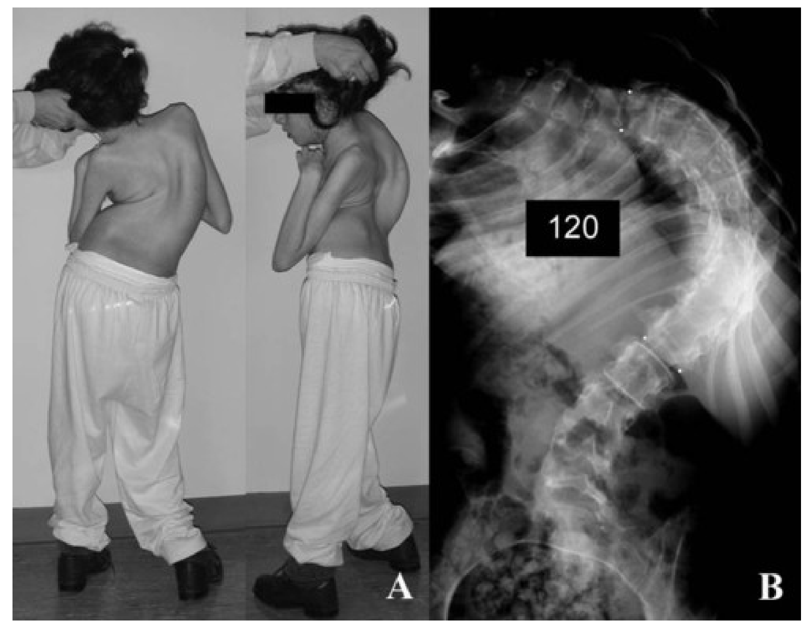
Scoliotic lungs?

Lung problems with scoliosis: Scoliosis is only part of the story!



- Idiopathic (kypho)scoliose
  - Decreased lung function (obstruction and restriction)

- +............?
  - (functional) muscle weakness
  - former lung damage (infections, growth, aspirations)
  - other congenital abnormalities
  - psychomotor retardation
    - Epilepsy
    - swallowing



Scoliotic lungs?

Lungproblems with increasing scoliosis

- Mild: no-hardly
- <40 degrees: sometimes swallowing difficulties
- >60 degrees:
  - Difficult breathing,decreased lungfunction/ventilation
    - Exercise intolerance
    - mucus clearance/coughing (low flows/stiff thorax)
  - swallowing difficulties/aspiration
  - Infections

- → respiratory deficiency during e.g. infections, sleeping/night (position)
- → ventilator deficiency
- → further “intrinsic” lung damage

Scoliotic lungs?

What does the patient experience with decreased lung function

- Restriction (decreased volumes)
  - difficult breathing
  - exhaustion

- Obstruction (decreased airway patency)
  - difficult breathing
  - wheezing/stridor
  - difficult sputum expectoration

Scoliotic lungs?

Scoliosis and lung function

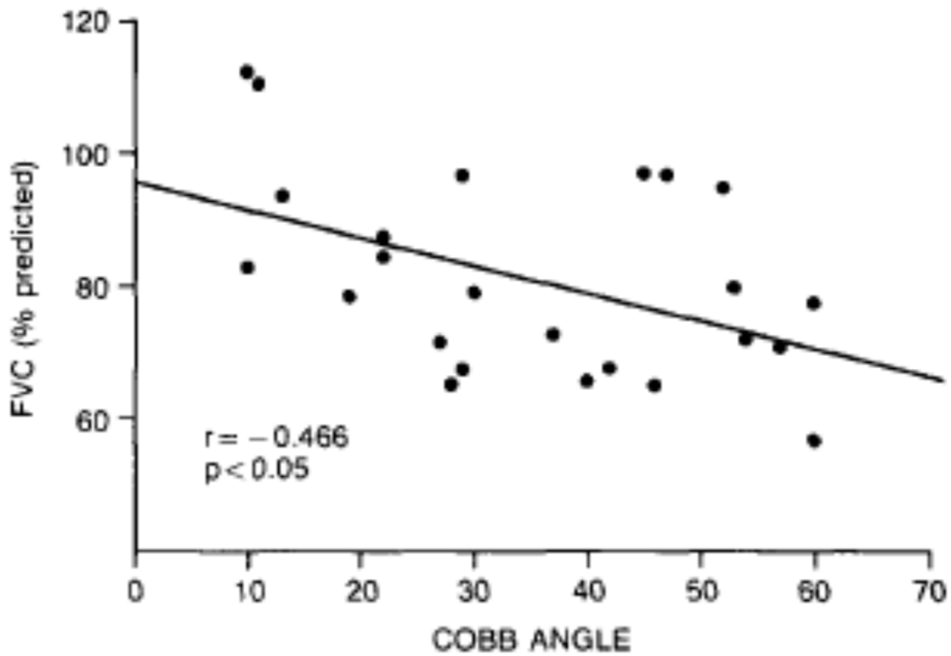


Fig. 1. Forced vital capacity (FVC, % predicted) versus the thoracic Cobb angle in adolescent female patients with idiopathic scoliosis.

Scoliotic lungs?

Szeinberg et al., Ped Pulm 1988

Lung function and scoliosis: about curves and angles……

- N=631 pts (85% females), 9 centers (1995-2003)
- Preoperatively lung function measurement
- Pulmonary impairment = lung function based…….
  - MILD: 65-80%pred
  - MODERATE: 50-65%pred
  - SEVERE: <50%pred

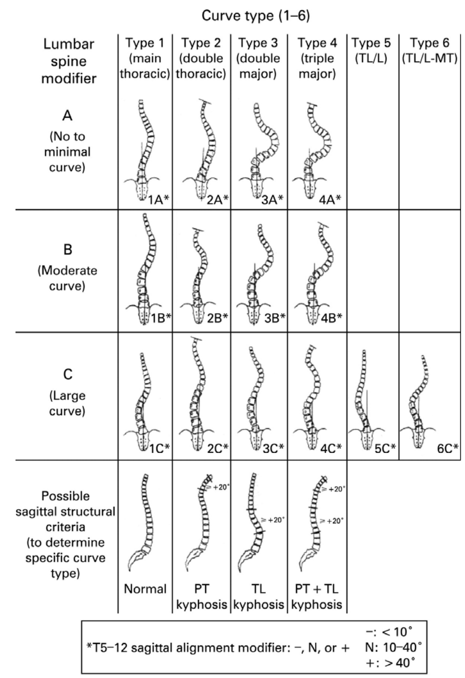


TABLE I Composition of the Study Cohort by Lenke Type and Associated Radiographic Measurements

Lenke Type	No. of Patients	Cobb Angle *(deg)* Cephalad Thoracic	Cobb Angle *(deg)* Thoracic	Cobb Angle *(deg)* Lumbar	Kyphosis *(deg)* T5 to T12	Kyphosis *(deg)* T2 to T12	Lumbar Lordosis *(deg)*
I	394	26 ± 8	52 ± 9	33 ± 9	23 ± 13	31 ± 13	–60 ± 12
2	92	40 ± 9	61 ± 13	31 ± 13	26 ± 15	34 ± 15	–60 ± 12
3	30	24 ± 7	62 ± 12	51 ± 10	28 ± 16	36 ± 16	–63 ± 12
4	16	41 ± 13	76 ± 20	58 ± 14	33 ± 14	36 ± 13	–55 ± 12
5	79	15 ± 7	47 ± 13	50 ± 10	23 ± 11	30 ± 12	–56 ± 14
6	20	19 ± 7	53 ± 6	62 ± 11	27 ± 13	34 ± 12	–54 ± 10

Scoliotic lungs?

Newton et al. *Bone Joint Surg Am*, 2005

More pulmonary impairment in
1. primary thoracic than (thoraco-)lumbar scoliosis
2. more involved vertebrae

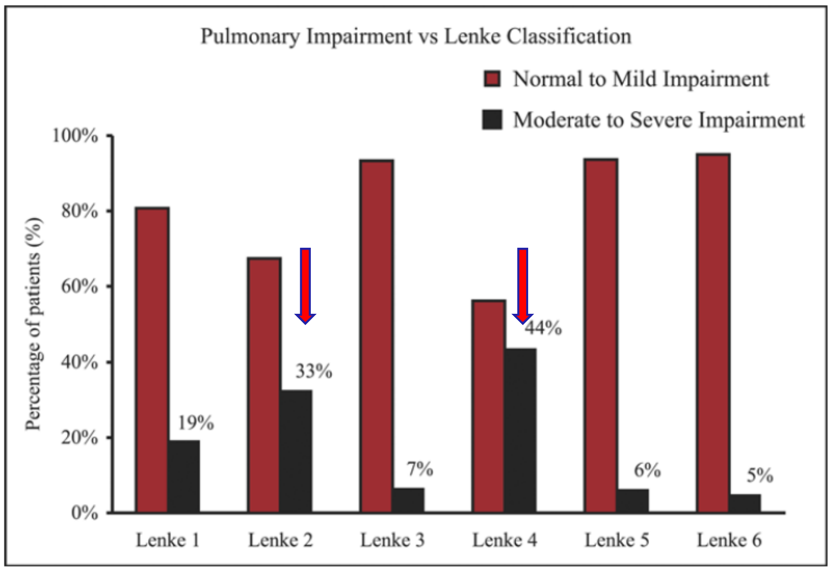


Fig. 4
Bar graph demonstrating the prevalence of moderate or severe pulmonary impairment for each Lenke curve type in the study cohort. Primary thoracic curves were associated with impaired pulmonary function, whereas patients in whom the primary curve was lumbar or thoracolumbar were less likely to have moderate or severe impairment.

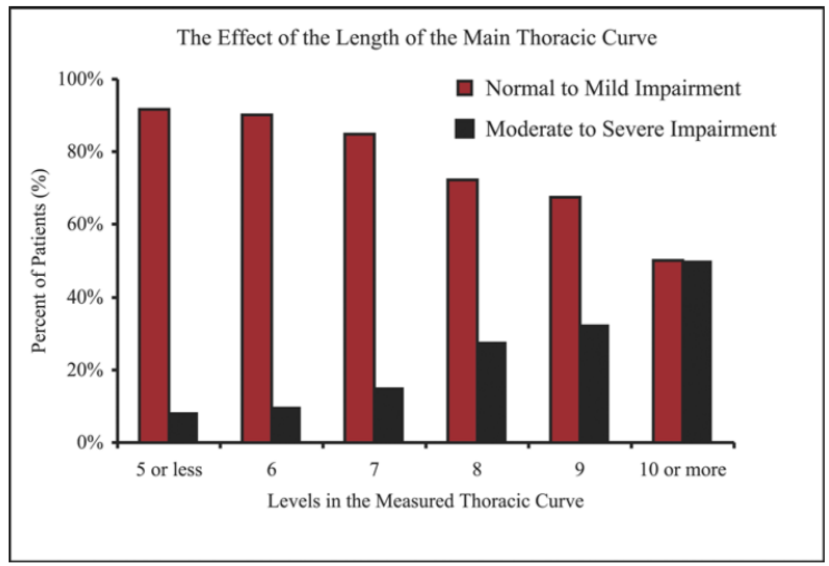


Fig. 2
Bar graph depicting the correlation between increased length of the thoracic curve (number of vertebrae over which the thoracic Cobb angle is measured) and pulmonary impairment.

Lung function

Newton et al. *Bone Joint Surg Am*, 2005

Angles and curves (cor)relate significantly but poorly to pulmonary impairment

Explaining <10% of variability.........90% comes form other factors!!

No relation with flexibility!?

TABLE III Correlations Between Pulmonary Function and Radiographic Measurements of Deformity

Radiographic Variable	Correlation: Percent of Predicted Value for Forced Vital Capacity	Correlation: Percent of Predicted Value for Forced Expiratory Volume	Correlation: Percent of Predicted Value for Total Lung Capacity
Cephalad thoracic curve magnitude	–0.202*	–0.221*	–0.152*
Cephalad thoracic curve flexibility	0.040	0.041	0.027
Main thoracic curve magnitude	–0.300*	–0.293*	–0.193*
Main thoracic curve flexibility	0.025	0.027	0.031
Lumbar curve magnitude	0.058	0.045	0.093
Lumbar curve flexibility	0.009	0.012	0.026
Thoracic apex displacement	–0.117†	–0.148*	–0.186*
Thoracic apex level	–0.010	0.000	–0.070
No. of vertebrae in thoracic curve	–0.342*	–0.305*	–0.192*
C7 displacement from central sacral vertical line	–0.182*	–0.149*	–0.139†
T5 to T12 kyphosis	0.157*	0.174*	0.153*
T2 to T12 kyphosis	0.230*	0.242*	0.211*
Lumbar lordosis	–0.100*	–0.127*	–0.076

*P < 0.002. †P < 0.01.

Scoliotic lungs?

Newton et al. *Bone Joint Surg Am*, 2005

Thoracic curve and lung function:

TABLE II Results of Pulmonary Function Tests Stratified by Magnitude of Thoracic Curve

Thoracic Curve *(deg)*	Forced Vital Capacity L	Forced Vital Capacity % of Predicted Value	Forced Expiratory Volume in 1 Sec L	Forced Expiratory Volume in 1 Sec % of Predicted Value	Total Lung Capacity L	Total Lung Capacity % of Predicted Value
≤20	3.31 ± 0.90	92 ± 15	2.84 ± 0.86	87 ± 21	4.89 ± 1.18	104 ± 14
21-30	3.45 ± 0.77	97 ± 16	2.94 ± 0.61	92 ± 14	4.50 ± 1.08	98 ± 17
31-40	3.10 ± 0.81	92 ± 16	2.66 ± 0.62	88 ± 16	4.31 ± 1.34	98 ± 20
41-50	3.01 ± 0.70	89 ± 17	2.55 ± 0.57	83 ± 17	4.07 ± 0.92	90 ± 16
51-60	3.03 ± 0.75	85 ± 16	2.57 ± 0.62	81 ± 16	4.00 ± 1.23	88 ± 20
61-70	2.79 ± 0.72	84 ± 14	2.33 ± 0.56	78 ± 13	3.71 ± 1.28	89 ± 22
71-80	2.40 ± 0.69	73 ± 18	2.01 ± 0.58	69 ± 18	3.92 ± 1.59	86 ± 22
>80	2.27 ± 0.72	69 ± 14	1.95 ± 0.56	65 ± 14	3.82 ± 0.98	83 ± 13
All	2.97 ± 0.77	86 ± 17	2.51 ± 0.64	81 ± 17	3.98 ± 1.31	90 ± 20

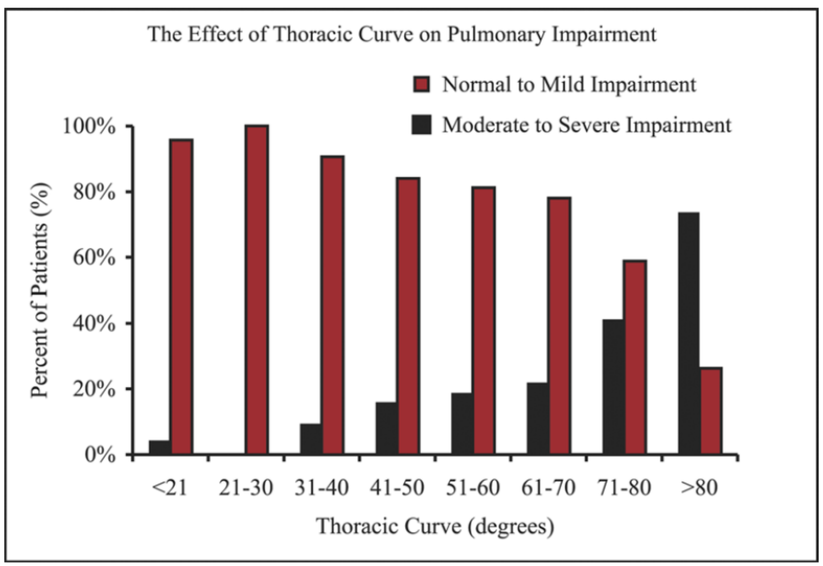


Fig. 1

Bar graph illustrating that increased coronal deformity is associated with increased pulmonary impairment.

Scoliotic lungs?

Newton et al. *Bone Joint Surg Am*, 2005

Number of involved vertebrae and lung function

TABLE IV Radiographic Markers Significantly and Independently Contributing to Variability in Pulmonary Function

	Coefficient of Multiple Determination (R^2)		
	Percent of Predicted Value for Forced Vital Capacity	Percent of Predicted Value for Forced Expiratory Volume (%)	Percent of Predicted Value for Total Lung Capacity
No. of vertebrae in thoracic curve	0.108	0.086	—
Thoracic curve magnitude	0.036	0.047	0.036
T2 to T12 kyphosis	0.043	0.041	0.038
C7 displacement from central sacral vertical line	0.010	0.006	—
Thoracic apex displacement	—	—	0.014
Total	0.197	0.180	0.088

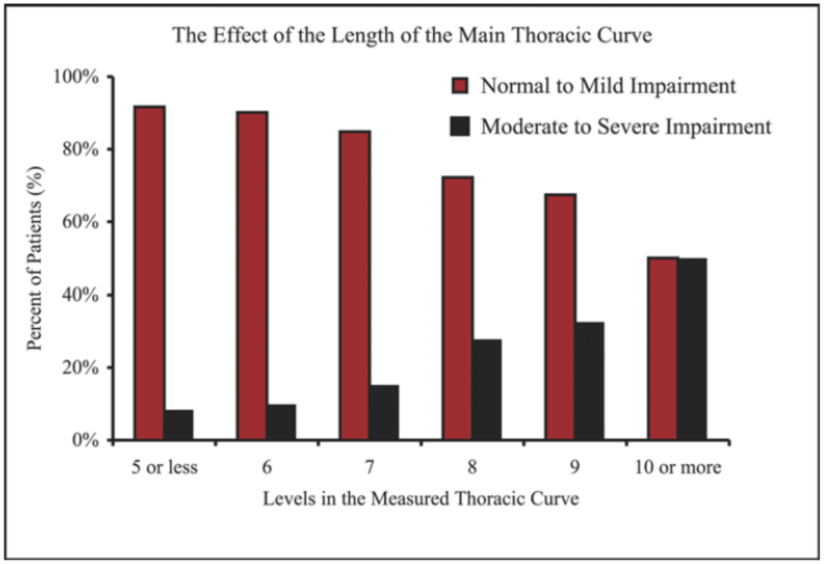


Fig. 2

Bar graph depicting the correlation between increased length of the thoracic curve (number of vertebrae over which the thoracic Cobb angle is measured) and pulmonary impairment.

Newton et al. *Bone Joint Surg Am*, 2005

Scoliotic lungs?

Thoracic kyphosis: the less the worse???

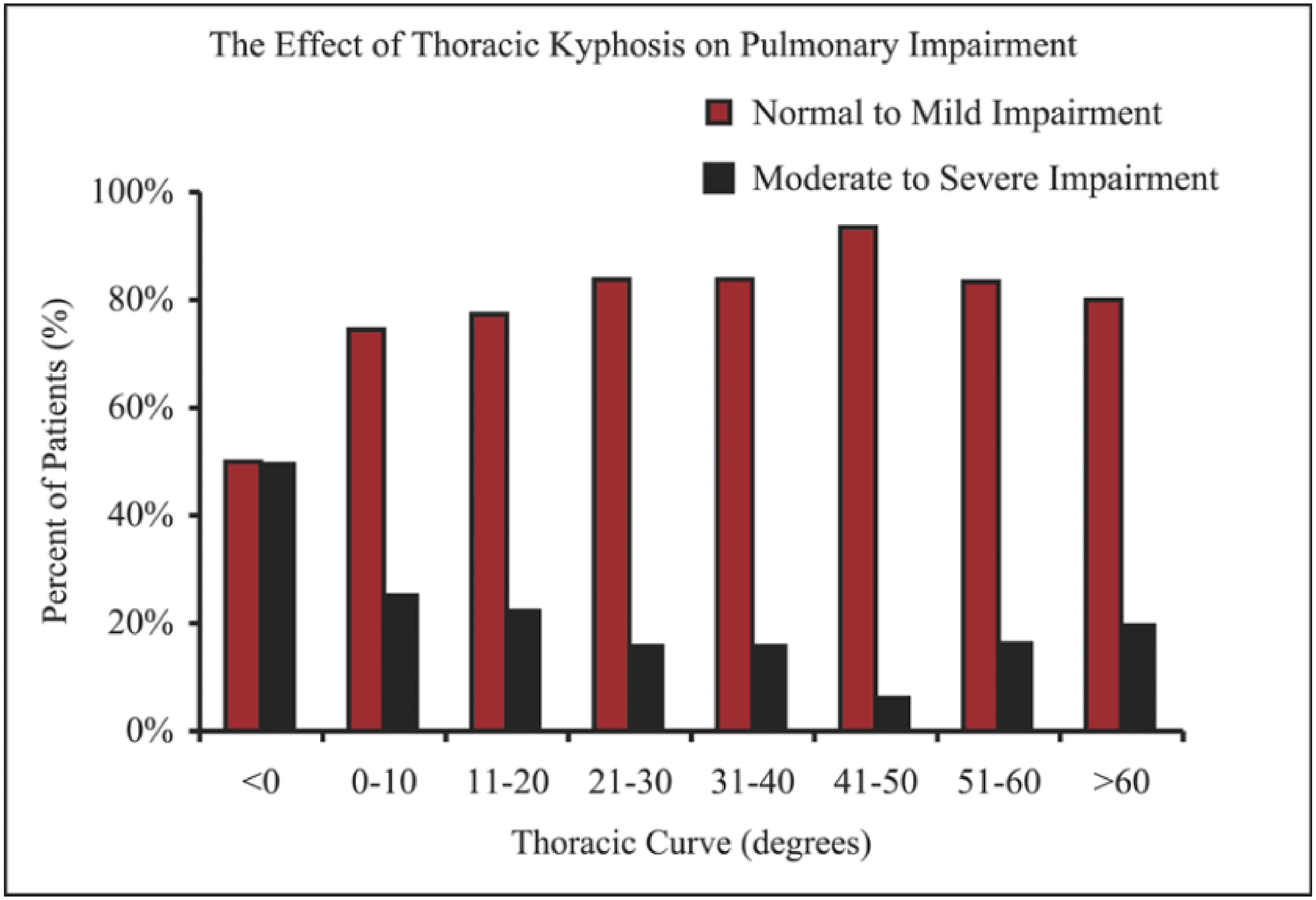


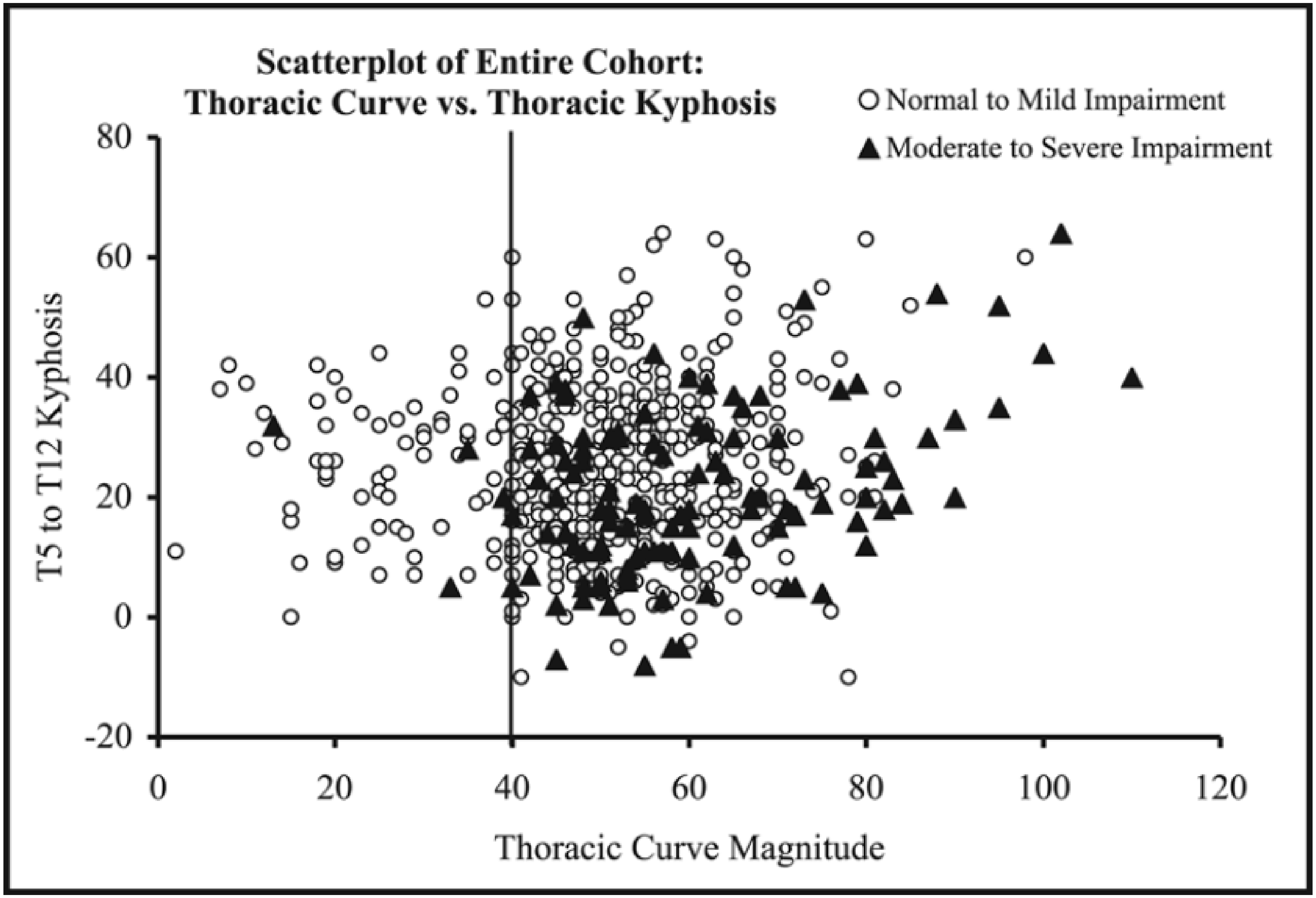
Fig. 3

Bar graph demonstrating the effect of the sagittal plane deformity (as measured by the Cobb angle from the fifth to the twelfth thoracic vertebra) on pulmonary function. The prevalence of moderate or severe pulmonary impairment is significantly higher ($p \leq 0.01$) in patients with thoracic hypokyphosis of ≤10°.

Scoliotic lungs?

Newton et al. *Bone Joint Surg Am*, 2005

Pulmonary problems start at 40 degrees?

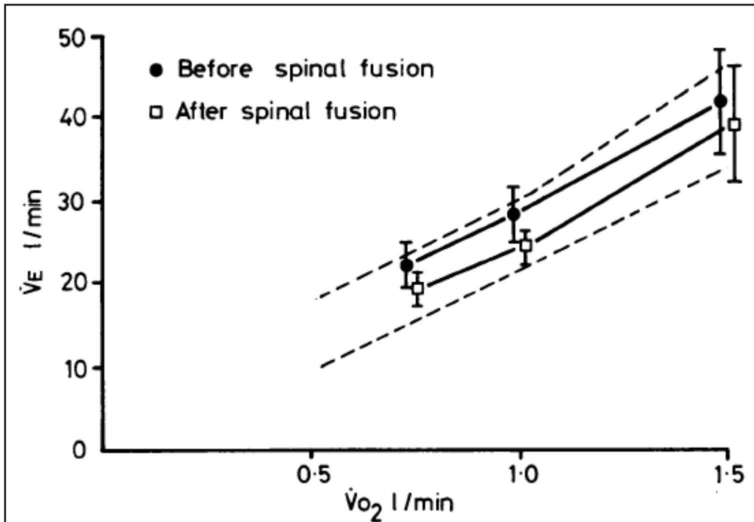


Scoliotic lungs?

Newton et al. *Bone Joint Surg Am*, 2005

Scoliosis repair and lung function: back to the seventies............

- N=10 girls: adolescent idiopathic scoliosis
- lung function and exercise testing before and 17-23 months after spinal fusion
- Cobbs angle: form 65->27
- Functional improvement
- Significant decrease in submaximal minute ventilation.
- No change in lung function



Effect of spinal fusion on minute ventilation ($\dot{V}_E$). Normal values ±1 SD (Jones et al, 1975) are shown by broken lines and observed values are shown ±1 SD. Postspinal fusion values significantly improved at V_{O_2} of 0·75 l (P<·05) and 1·0 l (P<·01).

Shneerson et al. Thorax 1979

Ventral derotation (VDS) + Harrington gives little improvement of FVC.....

- N=33
- mean thoracic curve 70 ($\pm$20)degrees
- Mean lumbar curve of 72.9 ($\pm$15)degrees
- VC before and 1 year after VDS and Harrington
- → correction curvature was 50 % (thoracic) and 68% (lumbar)
- → FVC: from 70%pred to 74% (ns)
  - Bigger interval related to better the improvement of the VC.
  - Younger age at VDS related to better F VC improvement
  - No relation to number of involved vertebrae

Scoliosis repair

Korovessis et al. Clin Orthop 1992

Harrington procedure

- meta-analysis effect Harrington instrumentation on lung function
- Adolescents with idiopathic scoliosis
- 38 studies, 5 proper studies
- N=173

- Increased FVC%pred +2-11%

Kinear et al. Spine 1993

Scoliosis repair

Lung function after growing rod surgery for progressive early-onset scoliosis: a preliminary study.

- N=8
- Group 1 finished “growing rod procedure” + definite fusion surgery
- Group 2 at the beginning of this procedure
- Lungfunction and X-photo’s before and after.
- No correlation between change in Cob angle and change in lung function

CMJ **Table 2.** Lung function change in group 1

Items	Pre-implantation of growing rod	Lengthening surgery stage	Pre-finial fusion	*P* values
FVC (L)	0.97±0.40	1.20±0.33	1.45±0.41	0.01
FVC/predicted FVC (%)	68.5±15.7	70.0±8.7	71.8±13.5	0.94
FVE1 (L)	0.89±0.34	1.07±0.26	1.28±0.30	0.05
FEV1/predicted FEV1 (%)	74.2±16.5	72.2±8.1	75.0±15.2	0.43

Table 3. Lung function change in group 2

Items	Pre-implantatin of growing rod	First lengthening surgery	Second lengthening surgery	*P* values
FVC (L)	0.97±0.14	1.19±0.20	1.31±0.26	0.04
FVC/predicted FVC (%)	67.8±15.7	64.8±11.8	70.2±14.0	0.08
FVE1 (L)	0.94±0.12	1.10±0.09	1.19±0.24	0.02
FEV1/predicted FEV1 (%)	77.8±16.2	69.8±9.7	73.4±12.8	0.09

Jiang et al. Chin Med J 2011

Effect scoliosis repair:
Summary of studies from last 40 years...

- Many studies, many techniques

- Esthetic and probably functional improvement
  - Improved mobilisation, pain,
  - Better ventilation

- Little, no or even negative effect on lung function and "pulmonary volumes"
  - Thorax remains stiff
  - Abnormalities in muscle function: "vertebral-muscular" connections?
  - No extra intrathoracic room?

Korovessis et al. *Clin Orthop* 1992

Kinear et al. *Spine* 1993

Shneerson et al. *Thorax* 1979

Jiang et al. *Chin Med J* 2011

Newton et al. *Bone Joint Surg Am*, 2005

Scoliosis repair

Thoraco-abdominal or thoracic surgery??

Thoraco-abdominal!

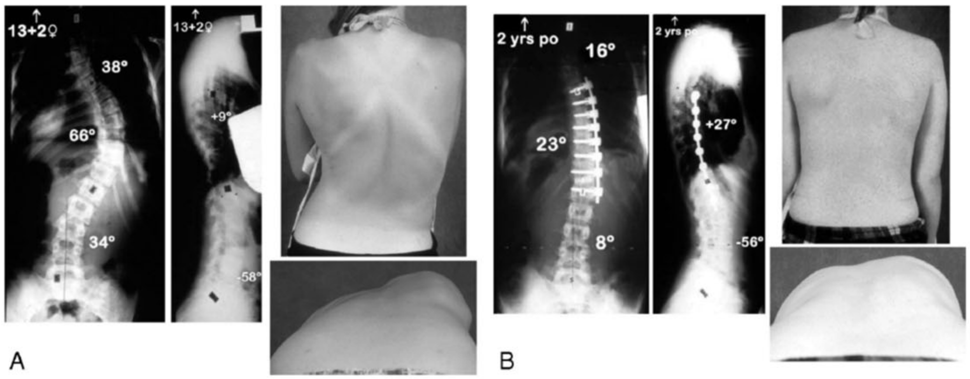


Table 1. Comparison Between the Thoracotomy and Thoracoabdominal Approaches

	TC Group (n = 64)	TA Group (n = 55)	P
No. of fused vertebrae	7.6 ± 0.8	5.1 ± 1.1	<0.0001
Age at surgery (yr)	14.9 ± 2.3	15.2 ± 1.8	0.75
Risser sign	3.1 ± 1.9	3.4 ± 1.8	0.48
Major Cobb			
Preoperative	55 ± 8.5°	52 ± 11.5°	0.07
2 yr Postoperative	29 ± 8.5°	20 ± 10.0°	<0.0001
Correction	48 ± 13%	61 ± 17%	<0.0001
Thoracic			
Preoperative	22 ± 14.5°	23 ± 9.1°	0.58
2 yr Postoperative	31 ± 11.1°	26 ± 9.6°	0.51
Amount of increase	9 ± 12.7°	3 ± 8.7°	0.008

*Statistically significant difference if $P < 0.05$.

Table 2. Comparison of PFTs Between the Thoracotomy and Thoracoabdominal Groups

	TC Group (n = 64)	TA Group (n = 55)	P
FVC absolute value			
Preoperative	3.05L ± 0.70	3.27L ± 0.67	0.08
2 yr Postoperative	2.74L ± 0.61	3.21L ± 0.64	<0.0001*
Difference	−0.31L ± 0.43	−0.06L ± 0.32	<0.0001*
% Difference	12% ± 17.2	2% ± 9.7	<0.0001*
FVC % predictive value			
Preoperative	87% ± 13.9	95% ± 18.2	0.01*
2 yr Postoperative	74% ± 12.2	87% ± 14.6	<0.0001*
Difference	−13% ± 10.9	−8.4% ± 15.0	0.045*
% Difference	−15% ± 11.8	−8% ± 15.0	0.002*
FEV_1 absolute value			
Preoperative	2.56L ± 0.55	2.82L ± 0.60	0.015*
2 yr Postoperative	2.35L ± 0.49	2.81L ± 0.52	<0.0001*
Difference	−0.21L ± 0.38	−0.02L ± 0.29	0.002*
% Difference	−7% ± 13.1	−0% ± 10.5	0.001*
FEV_1 % Predictive value			
Preoperative	81% ± 12.1	91% ± 17.7	<0.0001*
2 yr Postoperative	72% ± 11.6	85% ± 13.3	<0.0001*
Difference	−10% ± 9.9	−7% ± 14.6	0.022
% Difference	−11% ± 11.8	−6% ± 11.1	0.021*

% differences = difference/preoperative value.
*Statistically significant difference if $P < 0.05$.

Scoliosis repair

Yongjung et al. Spine 2008

Why not anterior thoracic?

- (more) damage to:
  - Muscles (diaphragm, latissimus dorsi, serratus, intercostals)
  - Ribs and thoracic cage

- more adhesions

- more pleural fluid

Scoliosis and lung function: Wrap up and conclude!

- Lung and thorax grow dependently and lung abnormalities can cause scoliosis (non-idiopathic)
- Expect ventilatory restriction in more severe scoliosis (>20% of patients when Cobb >50 degrees)
- Lung function impairment only partially explained by scoliosis
  - Increased susceptibility for infections and aspiration
  - Not volumes or obstruction but functionality is the issue!
  - Lung function is only part of pulmonary well being
- In general hardly any effect of surgery on lung function:
  - choose anterior and thoracoabdominal approach?

Conclusions and THM

Greetings from the pediatric pulmonologists UMC Utrecht

Questions??