Analysis of 3D Lung Volumes of Children with Asphyxiating Thoracic Dystrophy undergoing Lateral Thoracic Expansion Surgery

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Purpose

• To further the understanding Asphyxiating Thoracic Dystrophy (ATD)
• To develop an understanding of the success and failures of the surgery.
• Develop tools for evaluation of pulmonary disease that may combine physiologic and anatomic data.
Asphyxiating Thoracic Dystrophy (Jeune Syndrome)

• AR

• Broad Spectrum of clinical manifestations
  – Ranges from neonatal death from respiratory failure to cosmetic deformity
  – Often a progressive nephropathy
  – Occasionally hepatic fibrosis
  – Occasionally polydactyly
Radiologic Observations

- Small “bell” shaped chest
- Small pelvis with flared iliac bones, “trident” acetabulum
- Cone shaped epiphyses
- Short long bones
Inclusion

• The LTE Surgery was an IRB approved Protocol
  – Diagnosed with ATD
  – Ventilator dependent
  – All were thought to have a very poor prognosis and most were assigned hospice care
Materials

• 5 ATD patients who survived the LTE surgery and who were evaluated with Volumetric CT imaging were included in the evaluation.

• 1 patient who had Jeune's Syndrome but never required ventilation or surgery. He underwent volumetric CTs during pulmonary follow-up and evaluation.

Figure 1

Figure 2

A. S. Baker Medical Illustrator
During a Single Sedation

- PFTs were done using Whole Body Plethysmography, and Nitrogen Washout
- All CTs were done with controlled ventilation as described by Long, Castile et al.
  - Full inspiratory volumes were obtained at 25 cm of water.
  - Controlled exhalation was achieved in less than 1/2 of the visits and obtained at 0 cm of water.
Positive pressure is generated by occluding the expiratory port. A continuous supply of fresh air is attached to the inspiratory port. Pressure release valve is set to 25 cm water pressure.
CT Parameters

- GE Lightspeed Plus and GE Hispeed
- 5-7mm, a pitch of 1 on the single slice scanner
- A speed of 13cm/sec on the 8 slice scanner
- MA was initially 140mas, but was cut to 40mas and .5 rotation.
- Kv 120
- Data was reconstructed to 2.5mm
CT Volume Calculation

• GE Advantage Windows 4.0
  – Histogram mode

• Volumes were calculated from density thresholds of -200 to -1024 HU, and the lung volume was isolated

• Trachea, esophagus and stomach were “cut” from volume
Mean: -623, Std.: 161.8, min = -1023, max = -200
Total Volume = 276.64 cc (for entire object w/o cut planes)
Clinical Considerations

• Pre-op CTs were obtained 1 day prior to LTE

• Post-op CTs between 10 days and 10 months. Determined by indications for follow-up
  – unresolved pneumonia
  – or preparatory for contralateral LTE.
Data Analysis

- CT volumes vs. PFT Total Lung Capacity
- Each lung was evaluated in relation to surgery
- Mean Density and Standard Deviation
  - To determine a standard for inflation
Data

• Because of sedation failure PFTs only 70%
• CT Volumes obtained 100%
• The CT lung volumes had mean densities of
  – -609 HU and SD 152
  – One “Jeune without symptoms”
    • -708 HU and SD 160
    • -800 HU in normal children
PFTs Vs. CT volume

CT Lung volume cc’s

\[ y = 0.9712x - 27.208 \]

\[ R^2 = 0.8239 \]
However

- The *average* difference between volume is 10%
- $R^2 = 0.82$
- Good correlation but few data points
Mean: -499. Std.: 113.3. min = -871, max = -200
Total Volume = 95.96 cc (for entire object w/o cut planes)

Mean: -444. Std.: 121.3. min = -1001, max = -200
Total Volume = 51.67 cc (for entire object w/o cut planes)
Healthy Jeune

Post-Op on vent
What happened after surgery?

- Volume of the side operated on grew by 14% by the post operated CT
- The contralateral side grew by 13%
- Though the children got healthier both gaining weight and length in the interval
Conclusions about the surgical procedure

• The operation did not affect solely side operated on, but allowed both lungs to enlarge (mediastinum shifts.)

• Did the procedure or the care make the difference?
Outcome of the Surgery

• The patients in this evaluation represent 5 surviving patients of 7 operated on.
• All had significant improvement in quality of life, 4/5 are off ventilation at least part of the day
Conclusions about the CT

- The CT lung volumes can be used to evaluate the outcomes of pulmonary surgery.
- More data is needed to determine if the mean volume densities, or density curves can be used to evaluate lung expansion, or provide other qualitative information about the lung.