**Paper #19: The Etiology of Thoracic Insufficiency Syndrome in Neuromuscular Scoliosis Based on Quantitative Dynamic Lung MRI (QdMRI)**

*Robert M. Campbell, MD; Jay Udupa, PhD; Jack Flynn, MD; Hank Mayer, MD; Michael Nance, MD; Howard Panitch, MD; Wei-Hsun Wang, MD; Yubing Tong, PhD; Ivy Wu, PhD; Kieth Baldwin, MD; Joseph McDonough, Masters in Bioengineering; Andrew Mong, MD*


**Introduction:** Neuromuscular (NM) scoliosis is associated with poor pulmonary function felt due to muscle weakness, but thoracic insufficiency syndrome (TIS) due to complex spine/chest deformity also contributes, addressable by surgery, but difficult to assess with current technology. Standard dynamic Lung MRI (dMRI) can only qualitatively show TIS, but we have developed a new quantitative dMRI approach termed QdMRI, and in this pilot study have analyzed the dMRIs of 5 neuromuscular scoliosis patients. Based on these results, we propose new physiologic mechanisms for TIS in NM scoliosis that appear to improve with treatment.

**Methods:** Retrospectively the records, xrays, and pre-op dMRI scans of 5 NM scoliosis pts, 3 post op scans. All pts were treated with bilat rib to pelvis hybrid VEPTRs (Eiffel Tower). QdMRI analysis was by constructing a consistent 4D image from the free-breathing dMRI slice acquisitions, segmenting key anatomic structures in the 4D volume, and deriving quantitative dynamic parameters from the defined structures.

**Results:** Avg age 7.9 yrs, avg f/u 2.13 yrs. Scoliosis 75° preop, 45° at f/u. SAL .84 preop, .96 at f/u. Pelvic obliquity 19.6°, f/u 13.6°. One pt had substantial soft tissue remodeling of the kidney with increased excursion on dMRI. On QdMRI, preop concave Δrib cage volume (cc) was 46.5, Δdiaphragm volume 25.7, convex Arcv 40.5, Δdv 27.4. Kidney excursion (mm)concave was 2.58, convex 3.02. Two pts had concave Δdv less than convex, one concave greater than convex, two had equal volumes. For those three patients with post op scans, concave Δrib cage volume increased 57% (convex 72%), Δdiaphragm volume increased 128% (convex 109%). Concave kidney excursion increased 6.3mm(320%), convex 5.2mm(192%). Complications included two pts with migration of devices proximally.

**Conclusions:** QdMRI can measure separate contributions of diaphragm and rib cage to lung expansion during respiration for each hemi-thorax, enabling accurate assessment of thoracic function for the first time. VEPTR treatment appears to increase rib cage expansion more on the convex side of the thorax than the concave, and conversely increases diaphragm excursion on the concave side. With treatment, renal excursion increases bilaterally, concave> convex, probably reducing a blockade effect on the diaphragm. QdMRI has potential to increase the understanding of the anatomic mechanisms of spine/chest wall deformity that cause restrictive lung disease in NM scoliosis and may enable development of new treatments for it.