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Can Dynamic MRI Depict Change in Thoracic Function after Operation for Thoracic Insufficiency Syndrome?

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Conflicts of interest are listed at the end of this article.

See also the article by Tong et al in this issue.

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Thoracic insufficiency syndrome (TIS) is a rare childhood disorder that is defined as the inability of the thorax to support normal respiration or lung growth. It affects approximately 4000 children in the United States, with an incidence of approximately one in 100 000 to one in 300 000 live births. TIS develops as a consequence of a heterogeneous group of neuromuscular, syndromic, structural, and idiopathic diagnoses that affect the chest wall and spine (1). Abnormal growth of the lungs and thoracic cage can impair respiratory function. TIS often leads to respiratory failure, with an increased risk of early death (2). A reconstructive spine and chest wall operation can be performed in growing children with TIS when there is progressive thoracic deformity that may limit future lung development or thoracic function. However, there is currently no widely accepted and validated outcome measure to assess postoperative changes in respiratory function in patients with TIS. In this issue of *Radiology*, Tong et al (3) describe a quantitative MRI method for comparing the regional dynamics of the lungs, chest wall, and hemidiaphragms in children with TIS before and after surgical treatment with the vertical expandable prosthetic titanium rib (VEPTR) device.

Standard assessment of patients with spinal and chest wall deformity involves measurement of scoliosis and determination of lung volumes with pulmonary function testing. Whereas there is variable correlation between these measures in patients with more common disorders (eg, adolescent idiopathic scoliosis), the correlation between these measures is poor in patients with TIS. Cobb angle measurements of spinal curvature are considered the standard of reference for quantitative assessment of surgical corrective procedures. However, no meaningful correlation has been shown between Cobb angle and pulmonary function before or after operation (4). Forced vital capacity (the maximum amount of air expelled from the lungs after a maximum inhalation) and total lung capacity (the total amount of air in the lungs after a maximum inhalation) reflect overall thoracic function, but cannot provide information regarding regional lung, chest wall, or diaphragmatic function.

Nuclear medicine ventilation perfusion studies (5) in children with early onset scoliosis have shown that the lung in the concave hemithorax usually has a smaller volume than the lung in the convex hemithorax, with asymmetric ventilation in approximately 50% of patients. There are important clinical implications of asymmetric lung function in TIS, such as inadequate clearance of mucus from a poorly ventilated lung that causes atelectasis from mucous plugging (1). Children with TIS become primarily dependent on diaphragmatic contraction for inspiration, which leads to increased abdominal motion and may result in thoracoabdominal asynchrony.

Tong et al (3) evaluated the use of quantitative dynamic MRI to depict changes in regional dynamic thoracic function before and after surgical correction of TIS in 25 pediatric patients. They showed that average left and right lung volumes at end inspiration and end expiration increased substantially after operation, and that average lung tidal volume (ie, the normal volume of air displaced between normal inhalation and exhalation when extra effort is not applied) also improved, whereas conventional lung function testing demonstrated no change. Preoperative studies were performed close to the date of operation, whereas postoperative studies were performed approximately 1.5 years after operation. Four-dimensional imaging construction was performed by obtaining a subset of the two-dimensional free-breathing dynamic MRI sections representing one four-dimensional volume over one respiratory cycle. The left and right lungs were separately segmented in the four-dimensional volume at end inspiration and end expiration. By subtracting end-expiration images from end-inspiration images for each lung, lung excursion difference images were created. From these difference images, the chest wall and diaphragmatic components of lung excursion were separately derived.

Eleven volumetric parameters were extracted from image segmentation of the lungs, chest wall, and diaphragms obtained during free breathing, such as bilateral lung volumes at end inspiration and end expiration. The authors compared pre- and postoperative changes in these MRI-derived measures of lung volume and component tidal volumes to changes in clinical parameters (eg, total lung capacity and Cobb angles measured on frontal radiographs of the spine, and the assisted ventilation rating). The assisted ventilation rating is an ordinal scale ranging from +0 to +4, indicating increasing pulmonary compromise and reflecting the degree of external respiratory support needed to maintain oxygenation in children with TIS (6).

All quantitative volume parameters at dynamic MRI increased after the surgical procedure. Lung volumes increased at end inspiration and end expiration, especially in the right lung, with a 22.9% and 26.3% volume increase, respectively, at end expiration (P = .001) and end inspiration (P = .002). Average lung tidal volumes increased 43.8% in the left lung ($P < .001$) and 55.3% in the right lung ($P < .001$). However, clinical parameters showed no statistically significant change between pre- and postoperative measurements. Thoracic and lumbar Cobb angle measurements were poor predictors of tidal volumes of the left and right chest wall and hemidiaphragms at MRI. However, assisted ventilation rating and forced vital capacity were moderately correlated with the tidal volumes of the left and right chest wall and hemidiaphragms at MRI. One interesting finding was that the increase in tidal volumes of the right chest wall and diaphragm was significantly greater than that of the other components. In one patient, an asynchronous motion was noted between the left and right lungs before the operation, which was corrected after the operation. This observation highlights the ability of quantitative dynamic MRI to depict abnormalities that would not be depicted at conventional imaging.

The introduction of growth-sparing surgical interventions as a treatment for TIS has altered the expected clinical trajectory of patients with severe complex congenital scoliosis, from one with significant thoracospinal deformity and respiratory morbidity to one with considerably better subjective outcomes. Objective methods are needed to assess the therapeutic effects of VEPTR and to optimize other treatment techniques in children with TIS. Radiography, US, CT, MRI, cine CT, and cine MRI have been used to depict spinal and chest wall deformities but not to depict local-regional changes. Cine four-dimensional CT (7) can depict thoracic motion but it has the disadvantage of radiation exposure. The dynamic thoracic MRI technique is a nonionizing radiation technique that has the advantage of a free-breathing acquisition and permits analysis of compartmental changes in lung volume that can be attributed to bilateral chest wall and diaphragmatic displacement (8). By quantifying these displacements, the overall mechanical function of the thorax as a ventilatory pump can be assessed with respect to the pre- and postsurgical structure of the spine, ribs, and implanted instrumentation.

Because this is a difficult condition to evaluate, the study by Tong et al (3) includes a small number of patients, and underlying disorders were heterogeneous. Nevertheless, the dynamic imaging technique that they describe could be used to help compare the results of treatment with the VEPTR device with other treatments for early onset scoliosis such as bracing, growing rods, or magnetically controlled rods that permit lengthening by means of an external remote controller (Magec System; Nuvasive, San Diego, Calif). It may be applicable not only to patients with rare syndromes, but also to patients with more common disorders associated with spine and/or chest wall deformities, in which pulmonary growth and function may be compromised such as in patients with idiopathic adolescent scoliosis.

In conclusion, quantitative dynamic MRI was able to depict a substantial improvement in thoracic function in patients with thoracic insufficiency syndrome (TIS) after vertical expandable prosthetic titanium rib (VEPTR) operation by helping to document postoperative increases in all components of tidal volume. Clinical parameters were unable to reflect these improvements because of their inability to assess local-regional changes in lung, chest wall, and diaphragmatic motion. This technique holds promise for improving our ability to accurately document the functional results of VEPTR in patients with TIS, and it may be applicable to a wider population of patients with impaired respiratory function associated with spinal and chest wall deformities.

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References

1. Mayer O, Campbell R, Cahill P, Redding G. Thoracic insufficiency syndrome. Curr Probl Pediatr Adolesc Health Care 2016;46(3):72–97.
2. Corona J, Matsumoto H, Roye DP, Vitale MG. Measuring quality of life in children with early onset scoliosis: development and initial validation of the early onset scoliosis questionnaire. J Pediatr Orthop 2011;31(2):180–185.
3. Tong Y, Udupa JK, McDonough JM, et al. Quantitative dynamic thoracic MRI: application to thoracic insufficiency syndrome in pediatric patients. Radiology 2019. https://doi.org/10.1148/radiol.2019181731. Published online May 21, 2019.
4. Mayer OH, Redding G. Early changes in pulmonary function after vertical expandable prosthetic titanium rib insertion in children with thoracic insufficiency syndrome. J Pediatr Orthop 2009;29(1):35–38.
5. Redding G, Song K, Inscore S, Effmann E, Campbell R. Lung function asymmetry in children with congenital and infantile scoliosis. Spine J 2008;8(4):639–644.
6. Nossov SB, Curatolo E, Campbell RM, et al. VEPTR: are we reducing respiratory assistance requirements? J Pediatr Orthop 2019;39(1):28–32.
7. Low DA, Nystrom M, Kalinin E, et al. A method for the reconstruction of four-dimensional synchronized CT scans acquired during free breathing. Med Phys 2003;30(6):1254–1263.
8. Cai J, Chang Z, Wang Z, Paul Segars W, Yin FF. Four-dimensional magnetic resonance imaging (4D-MRI) using image-based respiratory surrogate: a feasibility study. Med Phys 2011;38(12):6384–6394.