European Respiratory Society Annual Congress 2013

Abstract Number: 2709
Publication Number: 206

Abstract Group: 4.1. Clinical respiratory physiology, exercise and functional imaging

Keyword 1: Respiratory muscle Keyword 2: Imaging Keyword 3: No keyword

Title: Assessment of diaphragmatic function by ultrasonography (US) and opto-electronic plethysmography (OEP) in patients with Duchenne muscular dystrophy (DMD)

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Body: Variations of diaphragmatic thickness (DT) and excursion (DE) during different respiratory maneuvers can be measured by B- and M-mode US. To verify if and how these parameters provide useful insights for functional assessment in DMD, DT was measured in 44 DMD patients (age 16.3±4.6yrs, FVC 53.4±24.6 %pred) during quiet breathing (QB), inspiratory capacity (IC) and maximal inspiratory pressure (MIP) and DE in 21 DMD (age 14.9±4.6yrs, FVC 55.6±19.9 %pred) during QB and IC. On the same patients, corresponding changes in abdominal volume (Vab) were measured by OEP during QB and IC. In older DMD patients (>18 yrs) DT variation was higher and MIP lower compared to younger during MIP. Conversely, DE was not influenced by age. Vab variations during QB and IC were also similar between younger and older patients. In all patients a significant linear correlation (r²=0.618, p<0.001) was found between DE measured by US and the Vab by OEP. On the contrary, variations of DT were not correlated to variations of Vab during QB and IC (r²=0.015, p=ns) (see fig.). In conclusion, DT and DE are estimates of the level of diaphragm contraction and displacement, respectively. In addition, Vab can be considered an optimal index of diaphragm length and changing length. In older DMD patients a dissociation occurs between the drive to the muscle and the development of muscle strength.