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Title: Assessment of the work of breathing (sWOB) within different functional and genetic groups in patients with cystic fibrosis (CF)

Prof. Dr Richard 10185 Kraemer richard.kraemer@swissonline.ch¹ and Prof. Sabina 29733 Gallati sabina.gallati@insel.ch². ¹ Paediatrics, University of Berne, Inselspital, Berne, Switzerland, 3010 .

Body: Rational. Pulmonary ventilation necessitates the work of breathing (sWOB) to overcome the resistive forces of the airways. During inspiration sWOB is performed by respiratory muscles, and during expiration by the release of energy stored in the elastic tissue of the lung. sWOB can be obtained by the integral of the tidal volume – box shift volume in the plethysmograph. This parameter was not yet explored in CF so far. Objectives. Association between sWOB and lung function over a substantial age range in CF patients. Methods. The Bernese CF database was prospectively built-up during the past 30 years comprising spirometric, plethysmographic, multibreath washout and gas exchange data, patients genotyped by SSCP screening and sequencing. Data were analysed by standard deviation scores (SDS) using reference equations. Results. From 238 CF-patients (120 males, 117 females; mean age 13.8 ± 6.6 yrs; observation time: 7.9 ± 3.1 yrs) 3478 test sets were evaluated. Best correlation of sWOB was found with sR_{eff} ($t=13.3$; $p<0.001$), TLC ($t=9.2$; $p<0.001$), sR_{tot} ($t=6.5$; $p<0.001$), FRC_{pleth} ($t=5.3$; $p<0.001$), FRC_{MBNW} ($t=2.8$; $p<0.005$), and $MMEF_{75-25}$ ($t=2.0$; $p<0.05$). sWOB was highest in patients with pulmonary hyperinflation ($8.5 \pm 0.9.7$ SDS) compared to those with normal static lung volumes (3.4 ± 5.0 SDS; $p<0.001$) and those with lung restriction (3.1 ± 5.5 SDS; $p<0.001$). There was significantly different progression of sWOB within genetic groups. Conclusions. Energy expenditure of breathing, estimated by sWOB, features an interesting functional parameter, highly associated with the degree of pulmonary hyperinflation, and could well serve as a factor of progression in CF.