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Title: Alterations of thoraco-abdominal volumes in patients with spinal muscle atrophy (SMA)

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Body: SMA is an autosomal recessive disease characterized by degeneration of motor neurons of spinal cord resulting in muscle wasting and weakness. Although, there is a general consensus that intercostal muscles are weakened and the diaphragm is relatively spared, particularly in SMAI and SMAII, the most severe forms of the disease, very little attention has been paid to possible alterations in the respiratory pattern in the mild SMAIII form. Thoraco-abdominal volumes were measured in 4 SMAII and 14 SMAIII patients and 18 age-matched controls during quiet breathing (QB), maximal voluntary cough (CGH) and slow vital capacity (SVC) in supine position by Opto-Electronic Plethysmography. SMAIII patients were divided into the recently proposed sub-classification[1] into SMAIIIA (wheelchair bound) and SMAIIIB (ambulant). SMAIIIB patients and controls showed a similar ability to significantly increase the percentage contribution of pulmonary rib cage ($\Delta V_{RC,P}$(%)) with the voluntary maneuvers. Conversely, SMAII and SMAIIIA patients were both characterized by similar reduced thoracic expansion (figure). In conclusion, in SMAII and SMAIIIA patients intercostal muscles weakness results in a poor thoracic expansion during quiet breathing and maximal voluntary respiratory maneuvers. The sub-classification of type III is valid not only for motor ability, but also for respiratory function. [1] Mercuri et al, Lancet Neurol 2012; 11: 443–52.