

European Respiratory Society Annual Congress 2013

Abstract Number: 3050

Publication Number: P1923

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

Keyword 1: Respiratory muscle **Keyword 2:** Physiology **Keyword 3:** No keyword

Title: Chest wall asynchronies during spontaneous breathing in spinal muscular atrophy (SMA)

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Body: In more severe types of SMA the intercostal muscles are weakened, while the diaphragm is relatively preserved. We studied chest wall asynchronies in SMA type II patients and in a recently proposed sub-classification of SMAIIIA and SMAIIIB to study chest wall distortion and respiratory muscle coordination. 4 SMAII (age 24.0 ± 14.2), 4 SMAIIIA (age 25.0 ± 14.9) and 9 SMAIIIB (age 21.5 ± 10.2) patients and 16 healthy age-matched controls (CTR, age 22.3 ± 11.4) were considered. In each subject pulmonary rib cage, abdominal rib cage and abdominal volumes (V_{RCp} , V_{RCa} and V_{AB} respectively) were measured in seated (ST) and supine (SP) positions by opto-electronic plethysmography. Rib cage distortion was quantified as the phase shift between V_{RCp} and V_{RCa} (θ_{RC}) and respiratory muscle coordination between V_{RCp} and V_{AB} (θ_{TA}). In ST chest wall asynchronies in all SMA were similar to CTR. The major differences were found in SP with SMAII showing highly negative values both in θ_{RC} and in θ_{TA} when compared to SMAIIIB and CTR (see fig). The change in posture had an effect on θ_{RC} and θ_{TA} in SMAII and SMAIIIA, with more negative values in SP. In SMAII the diaphragm strongly anticipates intercostal muscles action. This marked direct action of the diaphragm on RCa induces rib cage distortion especially in supine. Changing posture highlights a more similar behavior of SMAIIIA to SMAII confirming the suitability of sub-classification of SMAIII.