150. Physiological monitoring during sleep and in neuromuscular disease in children

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Assessment of diaphragm thickness variations by ultrasonography in patients with Duchenne muscular dystrophy (DMD)

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Variations of diaphragm thickness (DT) during different respiratory maneuvers can be measured by B-mode ultrasonography (US). In order to verify if this parameter provides useful insights for functional assessment in DMD, we measured DT by a 7.5 MHz US linear probe in the 9th or 10th right intercostal space in 39 DMD patients (age 16.3±4.4 yrs, FVC 53.2±24.8%pred) in supine position at rest (end-expiration during quiet breathing, QB) and maximal inspiratory pressure (MIP) at residual volume. The contribution of the abdominal compartment to tidal volume (Vab%) was assessed by optoelectronic plethysmography during QB and an inspiratory capacity (IC) maneuver.

Patients were subdivided into 3 groups according to age (G1: n=11, age<14; G2: n=13, 14<age>18; G3: n= 15, age>18 yrs). MIP was significantly higher in G2 compared to both G1 and G3 (figure, left). The variation of DT during the MIP maneuver, expressed as % change of DT at rest, was significantly greater in G3

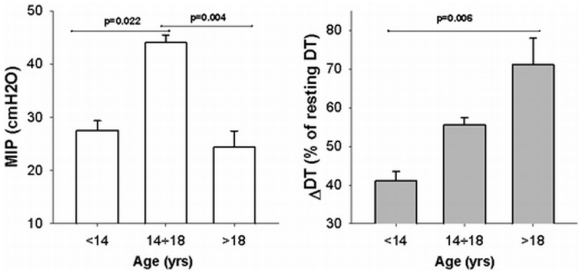

Figure 1

compared to G1. Patients belonging to G3 group were also characterized by lower Vab% during IC.
In conclusion, in DMD patients>18 yrs MIP decreases and maximal variation of DT increases. These results suggest that diaphragm impairment is expressed as a dissociation between muscle drive and muscle developed force. Measurements of DT by US allow a detailed noninvasive assessment on how alterations in diaphragm function progress in DMD.

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Thoraco-abdominal asynchronies during spontaneous breathing in Duchenne muscular dystrophy (DMD)

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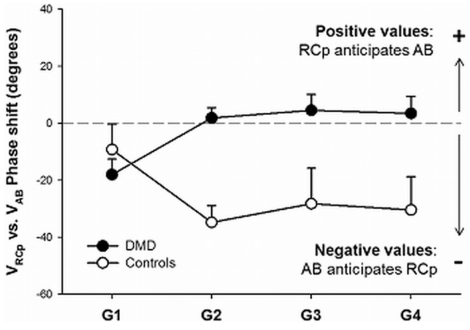
Measurement of thoraco-abdominal asynchronies (TAA) is a useful tool to evaluate the action and control of the respiratory muscles. Whether studying asynchronous chest wall movements can give better insight into progressive impairment of respiratory muscle function in DMD is debated.
We studied 100 DMD patients at different stages (mean age 14.1±5.8, range 4-32 yrs), subdivided into 4 age groups (G1<8(n=13), G2:8-12(n=27), G3:13-16(n=30), G4≥17(n=30) yrs) and 21 healthy age-matched male controls (CTR). Volume variations of chest wall compartments were measured during quiet breathing in supine position by opto-electronic plethysmography. Asynchronies between pulmonary rib cage(RCp) and abdomen(AB) were quantified in terms of phase shift between the relative compartmental volumes(V_{RCp}vs.V_{AB}).
In all CTR subjects volume variations of AB led RCp (negative phase shifts, figure1) independently on age. A similar behavior was found in G1 DMD patients, but not in older subjects(G2, G3 and G4) where RCp was the leading compartments($p<0.05$). With increasing age, the progressive changes in TAA were opposite between controls and DMD($p<0.01$).



In conclusion, in DMD patients lying supine the action of the inspiratory rib cage muscles progressively increases with age relative to the diaphragm. Accurate monitoring of TAA during spontaneous breathing is a valid indicator of diaphragm impairment and respiratory muscle dysfunction in DMD.

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Which are the most informative parameters to follow the respiratory decline in Duchenne muscular dystrophy?

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Rationale: Duchenne muscular dystrophy causes progressive respiratory muscle weakness. Few information is available on the natural evolution of respiratory parameters in boys with Duchenne muscular dystrophy.

Objectives: In order to identify respiratory parameters associated with the earliest decline, lung function, blood gases, respiratory mechanics, and respiratory muscle strength were measured in 48 boys with Duchenne muscular dystrophy (mean age 12.7±3.4, range 7.6 – 19.0 years), over a period of 10 years.
Main results: Only four parameters showed an important decline with age. Gastric pressure during cough was below normal in all patients with a mean 5.7±3.8 cmH_2O decline per year. Sniff nasal inspiratory pressure tended to increase before the age of 10 years followed by a rapid decline (mean decrease 4.8±4.9 cmH_2O or 5.2±4.4% predicted per year). Absolute forced vital capacity values peaked around 13-14 years of age and remained mainly over 1 liter but predicted values showed a mean 4.1±4.4% decline per year. Diaphragmatic tension time index increased above normal after the age of 14 years old with a mean increase of 0.04±0.04 point per year.
Conclusion: Repeated gastric pressure during cough, sniff nasal inspiratory pressure, and forced vital capacity measurements provide simple tools to assess the progression of respiratory muscle weakness in young boys with Duchenne muscular dystrophy. Endurance indexes decline at a later age. These indexes may help to monitor treatment effects.

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Inefficient cough in Duchenne muscular dystrophy (DMD)

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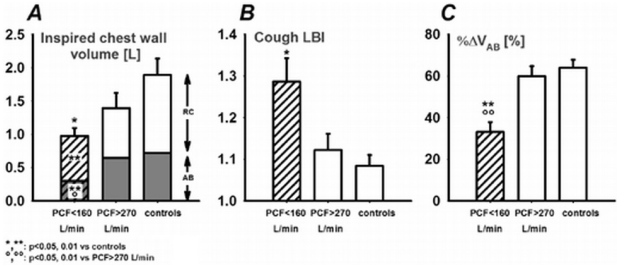
In DMD, impaired cough secondary to muscle weakness leads to serious respiratory complications, namely atelectasis, ineffective airway clearance, pneumonia and tracheal intubations. In order to study which factors influence and determine inefficient cough in DMD we studied 36 DMD patients and 15 healthy controls (C,age:16.3±5.4 yrs). Peak cough flow (PCF) was measured at the mouth while rib cage (RC), abdominal (AB) and total chest wall (CW) volume variations were measured by opto-electronic plethysmography during quite breathing and maximal cough (supine position). PCF was <160 L/min in 15 patients (inefficient cough, I:age:17.6±5 yrs,FVC:33.7±20%predicted) and>270 L/min in 9 (efficient cough, E:age:16.1±4 yrs,FVC:70.8±34%predicted). Tidal volume (V_T) was similar in I, E and C. In I, RC, AB and CW inspired volumes preceding cough were significantly lower than controls and inspired AB volume was lower than E (panel A). Thoraco-abdominal asynchrony during cough, quantified by labored breathing index (LBI), and percentage abdominal contribution to V_T ($\%\Delta V_{AB}$) were respectively higher and lower in I group (panel B and C).



In conclusion, in DMD inefficient cough is characterized by impaired inspiration, thoracoabdominal asynchony and lower abdominal contribution to volume variations due to diaphragmatic weakness. $\%\Delta V_{AB}$, that does not require patient's collaboration, seems to be a good predictor of inefficient cough.

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Adiposity but not severity of obstructive sleep-disordered breathing correlates with morning plasma TNF-α levels in Greek children

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Background: Sleep-disordered breathing (SDB) has been associated with increased frequency of excessive daytime sleepiness (EDS). In adults, increased TNF-α plasma levels probably mediate this association, but conflicting results have been reported in children. We hypothesized that: i) the higher the severity of SDB in childhood, the higher the frequency of EDS and morning TNF-α plasma levels; ii) subjects with high TNF-α levels are more likely to have EDS.
Methods: Children without and with snoring underwent polysomnography, EDS was determined by parental response to specific questions and TNF-α morning plasma levels were measured.
Results: Children with moderate-to-severe SDB [n=24; 5.7±2 yo; apnea-hypopnea index (AHI) 6-23.5 episodes/hour), but not participants with mild SDB (n=22; 6±2.5 yo; AHI 1.1-4.7) were at significantly higher risk for EDS compared to controls without snoring (n=26; 6.2±2.3 yo; AHI 0.2-1) [OR (95% CI): 7 (1.6-30.9) and 3 (0.6-13.8), respectively]. The 3 groups did not differ regarding TNF-α

levels (0.63±0.2 vs. 0.65±0.2 vs. 0.57±0.13 pg/mL; p>0.05). TNF-α levels were associated significantly with body mass index z-score (p<0.05), but not AHI or SpO_2 nadir (p>0.05). Subjects with high TNF-α levels (>0.57 pg/mL i.e. median in controls) were not at higher risk for EDS compared to those with low levels [OR (95% CI) adjusted for obesity: 1.9 (0.6-6.4)].
Conclusions: Increasing severity of SDB is related to increasing frequency of EDS but not with elevated TNF-α plasma concentrations which are positively correlated with the degree of adiposity. Children with high TNF-α levels are not at increased risk for EDS.

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Low morning serum cortisol levels in children with adenotonsillar hypertrophy and obstructive sleep apnea
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Adenoidal and tonsillar tissue in children with obstructive sleep apnea (OSA) has enhanced expression of glucocorticoid receptors (Goldbart 2005). Although this finding suggests a favorable profile for topical steroid therapy in children with OSA, its pathogenetic role in adenotonsillar hypertrophy is unknown. It is possible that overexpression of glucocortocoid receptors in pharyngeal lymphoid tissue reflects low endogenous cortisol levels. We hypothesized that children with OSA and tonsillar hypertrophy have lower morning serum cortisol levels compared to healthy control subjects.
Methods: Consecutive children with snoring and participants without snoring underwent polysomnography, grading of tonsillar size and measurement of morning serum cortisol.
Results: Children with moderate-to-severe OSA (n=17; 6.1±2.2 yo; AHI 14.7±10.6 episodes/h) had significantly lower morning serum cortisol levels than subjects with mild OSA (n=14; 6.8±2.3 yo; AHI 2.6±1.2 episodes/h) or control participants without snoring (n=14; 6.5±2.5 yo; AHI 0.7±0.2 episodes/h): 16.9±8.7 vs. 23.3±4.2 or 22.3±5.3 mcg/dL; p<0.05. In contrast, children with moderate-to-severe OSA (n=13; 5.1±1.1 yo; AHI 11.1±5.6 episodes/h) had similar cortisol levels relative to subjects with mild OSA (n=13; 6.8±2.4 yo; AHI 2.4±1.2 episodes/h) or control participants without snoring: 25.6±8.1 vs. 20.2±11 or 22.3±5.3 mcg/dL; p>0.05.
Conclusions: Low morning serum cortisol in children with OSA and tonsillar hypertrophy might be responsible for the enhanced expression of glucocorticoid levels in pharyngeal lymphoid tissue.

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Sleep disordered breathing in children with trisomy 21 and pulmonary hypertension
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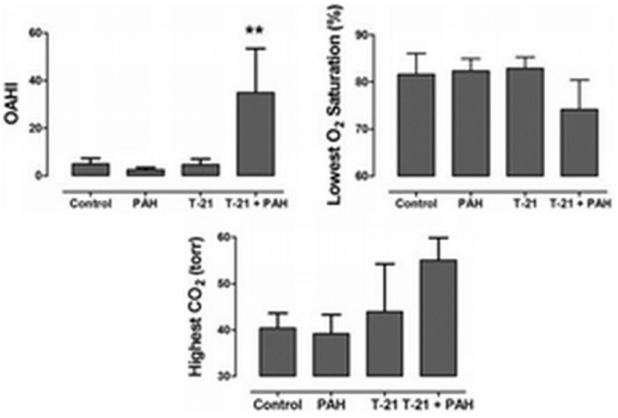
Introduction: Children with trisomy 21 (T21) have a significantly high prevalence of sleep disordered breathing (SDB), specifically obstructive sleep apnea (OSA). Children with T21 are also at risk for pulmonary arterial hypertension (PAH). It is unclear if PAH per se confers additional risk for OSA in this population.
Aim: To compare the prevalence of SDB in T21 children with PAH and without PAH and with controls.
Methods: This was a retrospective study where PSG data on all non-obese children at the Hospital for Sick Children, Toronto with T21 and/or PAH referred over a 5 year period, were reviewed and compared with PSG data in age-matched controls. The main outcome measure was the obstructive apnea-hypopnea index (OAHI).



Results: PSG of 33 children aged 0.2 – 8 years with a BMI < 25kg/m^2 were reviewed and compared in 4 groups; 1) controls (n=8), 2) children with PAH (n=9), 3) children with T21 (n=10), and 4) children with both T21 and PAH (n=6). In children with both T21 and PAH, there was a significantly higher OAHI (p=0.01) and a trend for lower oxygen saturations and a higher tcCO2 when compared with the other groups (see figures below).
Conclusions: Children with both T21 and PAH are at increased risk of severe OSA. All children with T21 who are known to have PAH should undergo sleep surveillance with PSG. Future research may be directed towards understanding the interaction of T21 and PAH predisposing to OSA.