Do we have a practical method for monitorization of pulmonary functions in non-ambulatory patients with neuromuscular disease?

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- No disclosure!

GFT outcome

- EOS patients should be assessed not only clinically and radiologically but also pulmonologically capacity @
  - The time of diagnosis
  - During the disease process
  - The end of treatment
- The 6-min walk test for ambulatory patients
  - Practical
  - Easily applicable
  - Reproducible standard test
- A similar test for non-ambulatory patients
  - Not described yet!

Aim

- To investigate the applicability of respiratory muscle and cough strength tests (RMCST) in non-ambulatory neuromuscular EOS patients
- To correlate with standard PFT

Methods

- Ten low-tone collapsing spine patients scheduled for deformity surgery
  - 5 SMA2
  - 3 DMD
  - 1 neuropathy
  - 1 CMD
- 9 M, 1 F
- Mean age 12.7(10-17) years

Methods

- Standard PFT tests (Vyntus Spiro, Carefusion, Germany)
  - Large and small airway functions

Methods

- Respiratory muscle and cough strength tests(RMCST)
  - Maximal Inspiratory Pressure-MIP
  - Maximal Expiratory Pressure-MEP
  - Peak Cough Flow-PCF
  - SNIFF
- Portable mouth pressure device (Micro MPM; Micro Medical Ltd, England)
- Mini Wright peak flowmeter (Clement Clark International, Edinburgh, UK)

Correlation between RMCST and PFT tests was statistically investigated

Results

FVC (L)	FVC PRED (%)	FEV 1 (L)	FEV1 PRED (%)	FEV1/FVC	MEF 25-75 (L)	MEF 25-75 PRED (%)
1,355 (0,44–1,99)	52,6 (23–95)	1.194 (0,36–1,74)	55,5 (22–99)	88,547 (81–100)	1,525 (0,29–2,62)	59,9 (14-116)

MIP (cmH2O)	MEP (cmH2O)	PCF (ml/mn)	SNIFF (cmH2O)
48,77 (23-85)	29,8 (13-43)	186 (110-270)	51 (24-82)

Results

	MIP	MEP	PCF
FVC Sig. (2 tailed) Correlation coefficient	 0.015 0.770	 0.032 0.675	 0.001 0.881
FVC PRED Sig. (2 tailed) Correlation coefficient	 0.050 0.832	 0.013 0.746	 0.034 0.671
FEV1 Sig. (2 tailed) Correlation coefficient	 0.019 0.753	 0.066 0.602	 0.002 0.850
FEV1 PRED Sig. (2 tailed) Correlation coefficient	 0.017 0.762	 0.026 0.693	 0.022 0.710

PFT

- Sophisticated and relatively expensive
- Needs a well-trained technician
- Relatively longer time is required

- Usability/ Practicality/Reliability ???
  - Young
  - Mentally retarded
  - Mouth disfunction

Conclusion

- Respiratory muscle and cough strength tests(RMCST) are significantly correlated with PFT in non-ambulatory patients with neuromuscular scoliosis
- RMCST is a fast, practical and easy-to-run test
- Measuring the RMCST at EOS outpatient clinics gives an idea about respiratory functions of non-ambulatory neuromuscular patients without need for sophisticated PFT tests
- They can be used for monitorization