Respiratory Medicine 113 (2016) 28–36

Contents lists available at ScienceDirect

Respiratory Medicine

journal homepage: www.elsevier.com/locate/rmed

Whistle and cough pressures in children with neuromuscular disorders

Sabrina Aloui [a], Sonia Khirani [a, b], Adriana Ramirez [a, c], Marina Colella [d], Bruno Louis [e], Alessandro Amaddeo [a, f], Brigitte Fauroux [a, e, g, *]

[a] AP-HP, Hôpital Necker Enfants-Malades, Pediatric Noninvasive Ventilation and Sleep Unit, 149 rue de Sèvres, 75015 Paris, France
[b] ASV Santé, 125 Avenue Louis Roche, 92230 Gennevilliers, France
[c] ADEP ASSISTANCE, 2 rue Benoit Malon, 92150 Suresnes, France
[d] Clinica Pediatrica, Istituto G. Gaslini, Genoa, Italy
[e] INSERM U 955, Equipe 13, Université Paris Est, 94000 Créteil, France
[f] Institute for Maternal and Child Health, IRCCS Burlo Garofolo, Pediatric Department, Trieste, Italy
[g] Paris Descartes University, 75000 Paris, France

ARTICLE INFO

Article history:
Received 14 April 2015
Received in revised form
15 February 2016
Accepted 18 February 2016
Available online 22 February 2016

Keywords:
Expiratory muscle strength
Maximal static expiratory pressures
Whistle
Cough
Forced vital capacity
Neuromuscular diseases

ABSTRACT

Rationale: Expiratory muscle strength is a determinant of cough function. Maximal static expiratory pressure (PEmax) manoeuvres are widely used but are limited by patient motivation and technique. The study hypothesized that whistle mouth (PmW) and cough gastric (PgasCough) pressures might provide additional tests of expiratory muscle strength in children and young adults with neuromuscular disease (NMD).

Methods: We retrospectively reviewed the data of lung function and respiratory muscle tests of all the patients with NMD followed in our centre between November 2001 and December 2013. PmW and PgasCough were compared to other common tests.

Results: Three hundred and four respiratory evaluations were performed in 143 patients, aged 3–29 years old. Seventy-two patients had 2 to 8 evaluations. Median [interquartiles] PEmax (38 [28–54] cmH_2O) did not differ significantly from PgasCough (45 [30–60] cmH_2O) and both were significantly greater than PmW (30 [19–44] cmH_2O). Significant good correlations were observed between all the expiratory muscle parameters. The best correlation was observed between PEmax and PmW ($r = 0.812$, $p < 0.001$). Moreover, good correlations were found between the percentage of predicted forced vital capacity and PmW ($r = 0.619$, $p < 0.001$) and PgasCough ($r = 0.568$, $p < 0.001$). Concerning the whistle test, the non-invasive measurement highly correlated with invasive measurements.

Conclusions: PmW and PgasCough are simple and valuable tests to assess expiratory muscle strength in children and young adults with NMD. These tests are particularly useful in children having difficulties to perform PEmax manoeuvre. They have the great advantage of their simplicity, but PgasCough is limited by its invasiveness.

1. Introduction

Measurement of expiratory muscle strength is of clinical importance in patients with neuromuscular disease (NMD) [1]. Indeed, as weakness progresses, cough may be impaired [2], leading to chest infection, a serious cause of morbidity and mortality [3–5]. Maximal static expiratory pressure (PEmax) is a volitional non-invasive manoeuvre widely recommended to assess expiratory muscle strength in patients with NMD [6,7]. However, PEmax manoeuvre may be technically difficult, particularly in young children. For this reason, some authors have used peak expiratory pressure in children [8]. But low values are also difficult to interpret, as they may result from poor effort, lack of cooperation, difficulties with the manoeuvre, or true expiratory muscle weakness. Peak expiratory flow (PEF) and peak cough flow (PCF) are also routinely used in adult patients for whom thresholds associated with an impaired coughing ability have been validated [9].

* Corresponding author. Pediatric Noninvasive Ventilation and Sleep Unit, AP-HP, Hôpital Necker, 149 rue de Sèvres, Paris F-75015, France.
E-mail address: brigitte.fauroux@aphp.fr (B. Fauroux).

http://dx.doi.org/10.1016/j.rmed.2016.02.009
0954-6111/

Abbreviations

PEmax	maximal static expiratory pressure
PmW	whistle mouth pressure
PgasCough	cough gastric pressure
NMD	neuromuscular disease
PEF	peak expiratory flow
PCF	peak cough flow
Pgas	gastric pressure
PesW	oesophageal whistle pressure
PgasW	gastric whistle pressure
CD	congenital muscular dystrophy
CM	congenital myopathy
DMD	Duchenne muscular dystrophy
SMA	spinal muscular atrophy
FVC	forced vital capacity
FVC %pr	predicted forced vital capacity
Pes	oesophageal pressure
R	coefficient of correlation
ALS	amyotrophic lateral sclerosis
M	male
F	female
Pdi	transdiaphragmatic pressure
PdiW	transdiaphragmatic whistle pressure

Standard values for PCF have been published for children [10] but thresholds associated with respiratory complications are not available.

Two additional tests using a natural, simple manoeuvre, i.e. the measurement of mouth pressure during a short, sharp and maximal expiration through a whistle (PmW) and the gastric pressure during a maximal cough (PgasCough) have been evaluated in healthy adults and adults with NMD [11,12]. They have been shown to accurately reflect expiratory muscle strength, however PgasCough requires the measurement of gastric pressure (Pgas) which limits its routine use. We hypothesized that these tests might also be useful and simple additional tests of expiratory muscle strength in children and young adults with NMD. Table 1 summarizes the advantages and limits of the different tests.

The aim of our study was to compare PEmax, PmW and PgasCough measurements between each others and with the standard lung volume measurements in children and young adults with NMD to assess the potential advantage/disadvantage of these measurements. We also assessed the relationship between the non-invasive PmW and invasive measurements of oesophageal (PesW) and gastric (PgasW) whistle pressures.

2. Material and methods

2.1. Patients

We retrospectively reviewed the charts of all the patients with congenital muscular dystrophy (CD), congenital myopathy (CM), Duchenne muscular dystrophy (DMD), and spinal muscular atrophy (SMA) who were referred to our centre for an assessment of respiratory muscle function between November 2001 and December 2013. The study was approved by the Institutional Review Board of the French learned society for respiratory medicine (Société de Pneumologie de Langue Française), and patients and parents gave their informed consent.

2.2. Procedure

Lung function and respiratory muscle tests were recorded at each outpatient visit. The tests were performed only if the patient was in a clinically stable state (absence of infection, acute medical condition, or unscheduled medical visit in the previous month). The frequency of visits varied according to the clinician's assessment, but was generally every 1–2 years. Data of patients treated with non-invasive ventilation were excluded from the study. All the patients were evaluated in the seated position.

The patients were asked to perform at least 3 acceptable forced vital capacity (FVC) curves. The highest FVC was retained and predicted FVC (FVC %pr) was calculated [13,14].

Afterwards, an oesogastric catheter (Gaeltec, Dunvegan, Isle of Skye, UK) was inserted pernasally after careful local anaesthesia (lidocaine 2%, Astra Zeneca, Rueil-Malmaison, France) [15], and appropriate placement was checked [16].

The strength of the expiratory muscles was assessed by asking the patient to perform maximal expiratory efforts from total lung capacity. Although no specific instructions were given for cough manoeuvres, coughs were usually made near to total lung capacity. The pressure tracings were displaced on a computer screen to

Table 1
Advantages and limits of the different respiratory maneuvers.

	Advantages	Limits
Forced vital capacity (FVC)	Largely used in children >4–8 years old; sensitive for assessing progress in moderate to severe global respiratory muscle weakness	Requires collaboration; poor specificity for the diagnosis of respiratory muscle weakness
Peak expiratory flow (PEF)/Peak cough flow (PCF)	Largely used in children >4–8 years old	Requires collaboration and coordination with the peak flow meter
Maximal static expiratory pressure (PEmax)	Largely used in children >6–8 years old; reference equations	Requires full collaboration, coordination and comprehension
Mouth pressure during a maximal whistle (PmW)	Natural manoeuvre, easy to perform, can be performed in very young children (>2 years of age); playful, audible feedback	Requires collaboration; highly sensitive to mouth closure; lack of reference values in children
Gastric pressure during a maximal cough (PgasCough)	The most natural manoeuvre, easy to perform, can be performed at any age (from newborn)	Invasive and mildly uncomfortable; requires collaboration; lack of reference values in young children
Oesophageal pressure during a maximal whistle (PesW)/Gastric pressure during a maximal whistle (PgasW)	Natural manoeuvre, easy to perform, can be performed in very young children (>2 years of age); playful, audible feedback	Invasive and mildly uncomfortable; requires collaboration; lack of reference values in young children