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RESEARCH ARTICLE

Expiratory Flow Maneuvers in Patients with Neuromuscular Diseases

ABSTRACT

Bach JR, Gonçalves MR, Páez S, Winck JC, Leitão S, Abreu P: Expiratory flow maneuvers in patients with neuromuscular diseases. Am J Phys Med Rehabil 2006;85:105–111.

Objectives: To compare cough peak flows (CPF), peak expiratory flows (PEF), and potentially confounding flows obtained by lip and tongue propulsion (dart flows, DF) for normal subjects and for patients with neuromuscular disease/restrictive pulmonary syndrome and to correlate them with vital capacity and maximum insufflation capacity.

Design: A cross-sectional analytic study of 125 stable patients and 52 normal subjects in which CPF, PEF, and DF were measured by peak flow meter and vital capacity and maximum insufflation capacity by spirometer.

Results: In normal subjects and in patients, the DF significantly exceeded PEF and CPF ($P \leq 0.001$). For normal subjects, PEF and CPF were not significantly different. For patients with neuromuscular disease/restrictive pulmonary syndrome, the CPF significantly exceeded PEF ($P < 0.05$). No normal subjects but 14 patients had DF lower than CPF. Thirteen of these 14 had the ability to air stack (maximum insufflation capacity greater than vital capacity), indicating greater compromise of mouth and lip than of glottic muscles. For 14 of 88 patients, maximum insufflation capacity values did not exceed vital capacity, mostly because of inability to close the glottis (inability to air stack). Nonetheless, for 11 of these 14 patients, the DF were within a standard deviation of the whole patient group; thus, bulbar-innervated muscle dysfunction was not uniform. CPF and PEF correlated with vital capacity ($r = 0.85$ and 0.86, respectively), and with maximum insufflation capacity ($r = 0.76$ and 0.72, respectively).

Conclusions: Measurements of CPF, PEF, and DF are useful for assessing bulbar-innervated, inspiratory, and expiratory muscle function. Care must be taken to not confuse them.

Key Words: Amyotrophic Lateral Sclerosis, Duchenne Muscular Dystrophy, Neuromuscular Disease, Cough Peak Flows, Peak Expiratory Flows, Dart Flows, Vital Capacity, Maximum Insufflation Capacity, Respiratory Muscles, Glottic Muscles

Both peak expiratory flows (PEF) and cough peak flows (CPF) have been described as useful clinical variables of respiratory muscle function.[1] "Dart flows" (DF) are generated by creating pressure behind the lips and tongue with the mouth closed. As the lips open and tongue releases the air, in a maneuver like spitting or projecting a dart through a narrow tube, these flows can also be measured by peak flow meter. These flows can be confused with PEF and CPF and cause the latter to be overestimated. They are largely a function of the ability to seal the lips and control the tongue and buccal muscles.

The main cause of morbidity and mortality in patients with neuromuscular disease/restrictive pulmonary syndrome (NMD) is respiratory muscle dysfunction and, in particular, cough dysfunction.[2–4] Inspiratory, expiratory, and bulbar-innervated musculature are required for effective coughing.[5,6]

Normal precough inspiration is to 85–90% of total lung capacity.[7] Thus, cough flows are diminished for patients who have decreased ability to inflate the lungs, especially when vital capacity (VC) is <1500 ml.[8] After a deep breath, the glottis is closed by intrinsic laryngeal (bulbar-innervated) muscles. The expiratory muscles (abdominal and intercostals) then contract, resulting in intrapleural pressures of 200 cm H_2O.[9] On full glottic opening with hypopharyngeal patency maintained by other bulbar-innervated musculature, there is an explosive decompression that normally generates flows of 300–1200 liters/min to expulse airway secretions.

In patients with NMD, weak inspiratory muscles can be assisted by providing deep lung insufflations or by the stacking of consecutively delivered volumes of air held with a closed glottis to approach a maximum insufflation capacity (MIC).[10–12] Expiratory muscles can be manually assisted by providing thoracoabdominal thrusts. The combination of applying an abdominal thrust to a maximally inflated lung is an assisted cough.[1,10] Unassisted cough flows depend on inspiratory, expiratory, and bulbar-innervated musculature. However, air stacking ability and, therefore, assisted cough flows depend only on glottic control or on bulbar-innervated muscle function alone. Thus, the greater the difference between the MIC and the VC and between assisted and unassisted CPF, the greater is bulbar-innervated muscle function by comparison with inspiratory muscle function. Patients who cannot close the glottis cannot air stack. They may "huff" but cannot cough. CPF better reflect the capacity to expulse debris from the airways (cough efficacy) than do PEFs. CPF not exceeding 160 liters/min are associated with extubation failure.[13]

There are no standard normal values for CPF or DF, but PEF range from 500 to 700 liters/min for men and from 380 to 500 liters/min for women, and from 150 to 840 liters/min for children and adolescents, with variations due to age, race, sex, and height.[14,15] For patients with asthma, their diminution generally indicates bronchospasm.[16] The purpose of this study was to compare the CPF, PEF, and DF, to see if they correlate with VC or MIC, and to consider their use in the evaluation of the respiratory muscles.

METHODS

A cross-sectional study was conducted on all NMD patients entering an outpatient clinic between August 2003 and May 2004. The charts of the patients were reviewed for anthropometric data (age, sex) and for diagnosis. All cooperative NMD patients whose VCs were <80% of predicted normal were studied. No one meeting these criteria was excluded. Normal subjects were recruited, informed about the purpose of the study, and signed consent forms that were approved by the hospitals' ethics committee. The patients and controls received a written description of the maneuvers and had a 3-min training period before the measurements were taken.

The following variables were measured: PEF according to the recommendations of the American Thoracic Society,[17] CPF, and DF, all via an Access Peak Flow Meter (model 710, Health Scan Products, Cedar Grove, NJ), and VC (sitting and supine) and MIC via a spirometer (Mark 14, Ferraris Development and Engineering, London, UK). All of these measurements were done by a specifically trained respiratory therapist who was unaware of the study and recorded the highest value of four or more correctly performed efforts. The peak flow meter measured flows from 60 to 880 liters/min. Flows of <60 liters/min were recorded as 0 and flows of >880 liters/min were recorded as 881 liters/min. No patients had been hospitalized during the previous 30 days.

Statistical Analysis

Data for the categorical variables are expressed as number and percentage of patients. Data for the continuous variables are reported as median with dispersion of minimum, maximum, and interquartile range and range. The use of median values rather than mean values eliminated the effect of imprecisely measured ceiling and floor data.

Normally distributed continuous variables were compared using the unpaired and paired Student's *t* test, as appropriate, and nonparametric continuous variables using Wilcoxon's signed-

ranks test and Mann–Whitney *U* test, as appropriate. The statistical analyses were repeated assuming that all flows of <60 liters/sec were 59 liters/min, except for CPF, which occur without glottic closure and which, by definition, could only be 0 liters/min. PEF of >880 liters/min were estimated as 881 and then re-estimated as 1008 liters/min, which is 20% greater than 840 liters/min or 2 SD greater than maximum normal PEF. In addition, a univariable linear regression analysis was conducted to compare expiratory flows with pulmonary capacities. All statistical tests were two tailed. A P value of <0.05 was considered to indicate statistical significance. Statistical analysis was conducted with the use of Stata, version 7.0, and SPSS, version 12.0.

RESULTS

There were 125 patients with a mean age of 41 ± 21 (range, 7–82) yrs; 64% were men (n = 80) and 100 were >18 yrs old (80%). The patients' diagnoses are listed in Table 1. The 52 normal subjects were 28.6 ± 9.8 (range, 19–58) yrs of age, 65.4 ± 11.4 (range, 49–100) kg, and 165.8 ± 8.4 (range, 152–183) cm tall. CPF, PEF, and DF data are presented in Table 2. Two patients were unable to attain any measurable flows, two had measurable PEF and CPF but not DF, nine had measurable CPF and DF but no measurable PEF, six had measurable DF but not CPF or PEF, and one patient had measurable PEF and DF but not CPF.

The DF were significantly greater than CPF and PEF ($P < 0.001$) for both the normal subjects and the patient group. The CPF and PEF were not significantly different for the normal subjects. For the patient group, assuming unmeasurable flows to be 0 liters/min, the CPF were significantly greater than PEF ($P < 0.01$) (Table 2). The differences remained significant ($P < 0.01$) when considering adults only but not children only. The patients' CPF remained significantly greater than PEF ($P < 0.05$) when the eight patients with unmeasurable CPF and PEF were eliminated and when PEF were estimated to be 59 liters/min for the nine patients with measurable CPF but not measurable PEF.

TABLE 1 Diagnosis of neuromuscular patients

Diagnosis	n	%
Amyotrophic lateral sclerosis	41	32.8
Duchenne–Becker muscular dystrophy	27	21.6
Other muscular dystrophies	18	14.4
Postpoliomyelitis	12	9.6
Myopathies (nonmuscular dystrophy)	8	6.4
Myasthenia gravis	6	4.8
Spinal muscular atrophy	5	4.0
Myotonic dystrophy	4	3.2
Neuropathies	2	1.6
Spinal cord injury	1	0.8
Obesity hypoventilation syndrome	1	0.8
Total	125	100

The flow data for the six patients (5.5%) with unmeasurable PEF and CPF but measurable DF are in Table 3. Thus, these patients had relatively well-preserved bulbar-innervated musculature, despite severe inspiratory and expiratory muscle weakness. This was consistent with their diagnoses of postpoliomyelitis[1] and congenital muscular dystrophy,[5] conditions that, relatively speaking, spare bulbar musculature.

There were two patients with unmeasurable DF who had severe bulbar amyotrophic lateral sclerosis yet had mean CPF of 205 liters/min. In fact, the CPF exceeded the DF for 12 patients: two children with non-Duchenne muscular dystrophy and ten adults, of whom seven had amyotrophic lateral sclerosis, two had fascioscapulohumeral dystrophy, and one had myotonic dystrophy. Only one was unable to air stack. These 12 patients had a greater capacity to air stack, as seen by a greater MIC–VC difference, than in the general group (Table 4), suggesting less compromise of glottic muscles than of the cheeks, lips, and tongue. The 16 patients whose PEF exceeded CPF and, thus, whose expiratory muscles were relatively preserved by comparison with bulbar-innervated muscles are considered in Table 5. The PEF of 11 normal subjects (21.1%) also exceeded their CPF.

The MIC was measured for 88 patients. For 14 of the 88, the MIC did not exceed the VC because of inability to firmly close the glottis or prevent air leakage out of the nose or mouth during the air-stacking process; their mean VC was 1518.6 ± 764.8 ml, significantly lower than the mean VC of the whole population (2000 ml, $P < 0.03$), suggesting more advanced disease. Eight of these 14 had CPF equal to or lower than PEF, confirming more severe compromise of facial and glottic musculature. In only three of these 14 cases were the DF lower than or equal to CPF, indicating that bulbar musculature was variably involved with relative sparing of the tongue and lips.

Good correlation was found between CPF and MIC (r = 0.76) (Fig. 1) and between PEF and MIC (r = 0.72) (Fig. 2). Correlation was also found between MIC and DF (r = 0.73). For the remaining 37 patients, MIC was not measured because the VC was too close to the normal range (3155 ± 1091.7 ml in 27 adults and 2861 ± 997.9 ml for the ten children) and bulbar musculature was clinically intact. There was also a direct correlation between CPF and PEF with VC (r = 0.85 and 0.86, respectively) and with MIC (r = 0.76 and 0.72, respec-

TABLE 2 Expiratory maneuvers for normal subjects and patients with neuromuscular diseases

Group	Statistics	Measurement Variable: CPF	PEF	DF
Normal patients (n = 52)				
	Median, liters/min	455	445	881
	Minimum, liters/min	290	320	370
	Maximum, liters/min	880	720	881
	Interquartile range, liters/min	225	160	148
	Range, liters/min	590	400	510
	Measurement above reference range of >880 liters/min, n (%)[a]	0	0	34 (65.4)
	Measurement below reference range of <60 liters/min, n (%)[b]	0	0	0
Patients (n = 125)				
	Median, liters/min	250	220	335
	Minimum, liters/min	0	0	0
	Maximum, liters/min	710	635	881
	Interquartile range, liters/min	210	188	270
	Range, liters/min	650	575	820
	Measurement above reference range of >880 liters/min, n (%)[a]	0	0	5 (4.0)
	Measurement below reference range of <60 liters/min, n (%)[b]	9 (7.3)	17 (13.5)	5 (4.0)
	≥18 yrs of age (n = 100), mean ± SD	280.1 ± 167.6	225.6 ± 159.9	395 ± 260.3
	<18 yrs of age (n = 25), mean ± SD	248.4 ± 108	234.6 ± 98.5	332.8 ± 158.7

CPF, cough peak flows; PEF, peak expiratory flows; DF, dart flows.
[a] Flows of >880 liters/min were recorded as 881 liters/min.
[b] Flows of <60 liters/min were recorded as 0 liters/min.

TABLE 3 Patients with unmeasurable cough and expiratory flows *vs.* group as a whole

	Patients with Unmeasurable CPF and PEF (n = 6)	All 125 Patients	P
Age, yrs	29.5 ± 14.6	41 ± 21	0.06
DF, liters/min	128.3 ± 50.9	382.6 ± 244.1	<0.01
VC, ml	358.3 ± 115.5	2000.0 ± 1245.6	<0.01

CPF, cough peak flow; PEF, peak expiratory flow; DF, dart flows; VC, vital capacity. Data (except for significance) provided as mean ± standard deviation.

tively) for the entire patient group (Fig. 3). This is not surprising because CPF and PEF are dependent on the ability to take a deep breath. DF also correlated with VC (r = 0.79), indicating relative preservation of tongue and lips in early disease.

DISCUSSION

Unlike in a previous report by Suarez et al.[18], although CPF were greater than PEF for normal subjects, we did not find the difference to be statistically significant. However, our patient population may have been too small to observe a significant difference.[18] As Suarez et al.[18] reported for patients with Duchenne muscular dystrophy, we did observe significantly greater CPF than PEF for patients with NMD.

The three flow maneuvers we studied are similar in that they are expiratory flows measured at the mouth using a peak flow meter. However, each method requires different respiratory muscle group combinations. With glottic closure, the greater transpulmonary pressures created by coughing rather than by PEF maneuvers resulted in greater flows measured at the mouth for 88.2% of patients and 78.9% of normal subjects. However, cough efficacy is dependent on the peak flow velocity, which is greater as airways narrow during

TABLE 4 Patients with cough peak flows (CPF) greater than dart flows (DF)

	CPF > DF (n =12)	All Group (n = 125)	P
VC	1220.8 ± 611.9	2000.0 ± 1245.6	<0.01
MIC	1974.0 ± 643.3	2179.8 ± 1097.9	NS
MIC – VC	822.0 ± 580.3	632.1 ± 474.7	NS
CPF	179.2 ± 49	273.8 ± 157.6	<0.01
PEF	113.8 ± 55.8	227.4 ± 149.4	<0.01
DF	107.9 ± 75.1	382.6 ± 244.1	<0.01

VC, vital capacity; MIC, maximum insufflation capacity; NS, not significant; PEF, peak expiratory flows. Data (except for significance) provided as mean ± standard deviation.

TABLE 5 Patients with peak expiratory flows (PEF) greater than cough peak flows (CPF)

	PEF > CPF (n =16)	Entire Group (n = 125)	P
VC	2042.3 ± 899.8	2000 ± 1245.6	NS
MIC	2324.2 ± 943.1	2179.8 ± 1097.9	NS
MIC – VC	588.5 ± 580.5	632.1 ± 474.7	NS
CPF	213.4 ± 103.2	273.8 ± 157.6	0.04
PEF	257.8 ± 92.0	227.4 ± 149.4	NS
DF	387.8 ± 168.4	382.6 ± 244.1	NS

VC, vital capacity; NS, not significant; MIC, maximum insufflation capacity; DF, dart flow. Values (except for significance) provided as mean ± standard deviation.

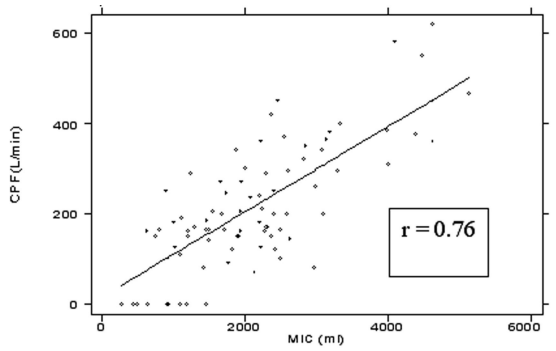

FIGURE 1 *Correlation between cough peak flows (*CPF*) and maximum insufflation capacity (*MIC*).*

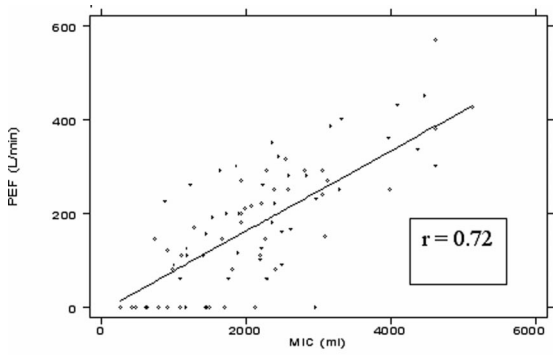

FIGURE 2 *Correlation between peak expiratory flows (*PEF*) and maximum insufflation capacity (*MIC*).*

coughing, making coughing more effective at expulsing airway secretions than huffing, even though PEF and CPF may be comparable when measured at the mouth.[8] The reduction of the cross-sectional area of the airways during coughing is due to smooth muscle constriction mediated by a vagal reflex (presumably preserved in these diseases) and due to dynamic compression of the airways generated by the expiratory (transpulmonary) pressure.[19,20] The reduction in the cross-sectional area of the airways increases five-fold the velocity of gas and 25-fold the kinetic energy of the airstream. This explains why the subgroup of 16 patients (12.8%) with CPF lower than PEF nevertheless coughed rather than huffed to expel secretions. Effective CPF and PEF share the need for deep lung volumes, explaining their good correlation with VC and MIC.

The correlation of CPF with MIC or MIC–VC difference is explained by their dependence on bulbar-innervated muscle (glottic) function. CPF are also dependent on hypopharyngeal patency being maintained by bulbar-innervated hypopharyngeal musculature. DF, on the other hand, are independent of laryngeal and hypopharyngeal dysfunction and usually exceed CPF and PEF. However, DF do not emanate from the airways and require little or no inspiratory or expiratory muscle effort. We have several patients who operate sip-and-puff motorized wheelchairs and generate high DF, despite

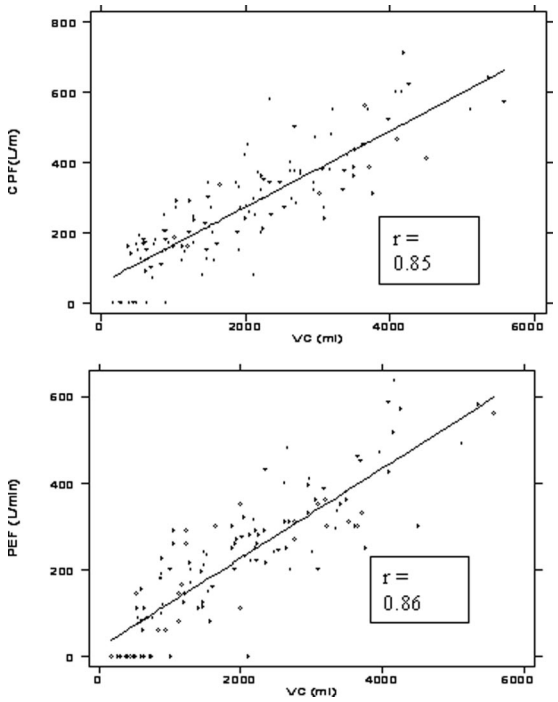

FIGURE 3 *Correlation between cough peak flows (*CPF*) and peak expiratory flows (*PEF*) with vital capacity (*VC*).*

having no measurable VC. DF, although also dependent on bulbar-innervated muscles but not glottic function, do not seem to reflect risk of respiratory complications. Inability to create measurable DF, however, is associated with ineffective saliva control and drooling. Thus, different patterns of bulbar-innervated muscle dysfunction occur. Figures 1 and 2 demonstrate that DF tend to correlate linearly with height and weight. In this way, DF are similar to PEF because these have also been reported to correlate with height and weight.[21,22] Wohlgemuth et al.[21] and Holcroft et al.[22] also pointed out the need to caution their subjects from spitting during PEF measurements.

Although all of the flow maneuvers are dependent on effort and motivation, we do not think this was a confounding factor in our study because the three measures were obtained in the same visit, in varying order, by the same examiner, and only the maximum value of many attempts was recorded.

Measurement "ceiling" and "floor" artifacts are common in empirical studies. There are elaborate statistical procedures that can be employed to estimate the range of plausible effects of the measurement limitation on actual *P* values. However, in this study, simpler and more direct logic suffices. This is because DF values exceeded 880 liters/min for 65% of normal subjects but for only 4% of patients and because DF values were significantly greater for normal subjects than for patients even when analyzing the data using ceiling DF of 881 liters/min when the actual values had to be greater than this figure. Likewise, for both patients and normal subjects, DF were significantly greater than PEF and CPF even when a ceiling value of 881 liters/min was used. The *P* values, already <0.001, were even more significant when the analyses were repeated using greater values for DF. Thus, the restriction of measurement range produced a conservative bias in the test of significance of DF group differences.

In summary, assisted and unassisted CPF, PEF, and DF are useful measures of bulbar-innervated and respiratory muscle function for patients with NMD,[1] permitting greater knowledge of the pattern of respiratory muscle compromise. DF, CPF, and MIC correlate with bulbar-innervated muscle function. It is important to pay special attention to the technique of each flow measurement because DF can be mistaken for CPF or PEF and respiratory risk can be underestimated. The techniques are simple, and the peak flow meter is inexpensive and widely available. Further study is warranted to determine standard values of PCF and DF by age, height, and weight. Peak flow meters with greater range need to be developed to more accurately measure high flows. Effective interventions to assist inspiratory and expiratory muscle function and the accurate characterization of risk of respiratory complications depend on accurate assessment of expiratory flow maneuvers.[23–25]

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