Measuring inspiratory muscle strength in neuromuscular disease: one test or two?

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ABSTRACT: **Inspiratory muscle strength monitoring is crucial in patients with neuromuscular disorders. The sniff nasal inspiratory pressure (SNIP) and maximal inspiratory pressure ($P_{I,max}$) are usually measured. The present study investigated whether the test yielding the best value at baseline continued to yield the best value during follow-up.**

The present study included 25 patients with Duchenne muscular dystrophy (DMD) and 61 with myotonic muscular dystrophy (MMD). SNIP and $P_{I,max}$ were measured at baseline and then annually.

At baseline, SNIP was lower than $P_{I,max}$ in 20 (80%) DMD patients and 32 (52%) MMD patients. During follow-up in DMD patients, changes in the best method always occurred from SNIP to $P_{I,max}$. In MMD patients, when SNIP was better than $P_{I,max}$ at baseline, SNIP was usually (88%) better during follow-up, whereas a better $P_{I,max}$ than SNIP at baseline was frequently (50%) followed by a shift to SNIP.

Maximal inspiratory pressure may be sufficient for monitoring inspiratory muscle function in Duchenne muscular dystrophy adults. In myotonic muscular dystrophy, the marked variability in the test yielding the best value at baseline indicates a need for performance of both tests at baseline. However, when sniff nasal inspiratory pressure measurement yields the best value at baseline, using sniff nasal inspiratory pressure alone during follow-up may be appropriate.

KEYWORDS: Maximal inspiratory pressure, neuromuscular disease, sniff pressure

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Respiratory pump failure is one of the main causes of death in patients with neuromuscular disorders. Therefore, respiratory muscle strength must be monitored closely in order to evaluate the risk of respiratory failure. The static maximal mouth pressure measured during a maximal inspiratory effort that is sustained for ≥1 s against an occluded airway is the most widely used parameter for assessing inspiratory muscle strength [1]. However, this static maximal inspiratory pressure ($P_{I,max}$) manoeuvre is difficult to perform, and the extent to which low values reflect inspiratory muscle weakness and/or poor motivation or coordination of the patient cannot be determined. Moreover, several independent variables, such as technical factors (most notably the type of mouthpiece used) and the time allowed for learning the manoeuvre, may lead to variations across laboratories [2, 3]. Sniffing is a natural effort that many patients find easier to perform than static efforts, and sniff nasal inspiratory pressure (SNIP) is a recently suggested alternative to $P_{I,max}$ [4, 5]. In patients with neuromuscular disorders, each of these two volitional tests has its own advantages and disadvantages, such that the two tests are complementary rather than interchangeable [6–8].

Since patients require distinctive skills and features to perform well on both of these volitional techniques [4], it was hypothesised that the test yielding the highest value at baseline for each patient continues to yield the highest value during follow-up and, therefore, could be used alone during follow-up. This hypothesis was evaluated in two of the most common chronic neuromuscular diseases: Duchenne muscular dystrophy (DMD), a disease confined to the skeletal muscle; and Steinert myotonic muscular dystrophy (MMD), which is characterised by a slower progression of muscle weakness than in DMD patients and frequent central nervous system involvement, responsible for cognitive impairment.

METHODS

Patients

The present study was approved by the appropriate ethics committee (Ambroise Paré Teaching Hospital, Paris, France) and informed consent was obtained from all study participants. Patients

were recruited between January 2002 and December 2005 during routine follow-up visits at the Raymond Poincaré Teaching Hospital (Garches, France). Consecutive patients who had a diagnosis of DMD or MMD with at least a baseline visit and one follow-up visit 1 yr later were included in the study. In all study participants, the neuromuscular disease had been diagnosed $\geqslant 10$ yrs before study inclusion. None of the patients had performed the SNIP or $P_{I,max}$ manoeuvre prior to the study. Patients were included if they had been clinically stable for $\geqslant 1$ month. Noninvasive positive pressure ventilation was started when necessary according to international recommendations [9].

Experimental protocol

All pulmonary function tests were performed by the same experienced technicians in a single session for each patient at each visit. Patients were allowed time to recover fully between tests. Spirometry was performed in the sitting and supine positions. $P_{I,max}$, SNIP and maximal expiratory pressure were recorded with the patient seated with $P_{I,max}$ and SNIP measured in random order.

Spirometry

At each visit, spirometric variables and lung volumes were measured using the Vmax 229 SensorMedics system (SensorMedics, Anaheim, CA, USA) according to standard guidelines [10]. The best of three reproducible values was recorded. Results were expressed as a percentage of published values [10].

Respiratory muscle testing

$P_{I,max}$ and SNIP were measured from functional residual capacity in a standard manner, as previously described elsewhere [4]. $P_{I,max}$ is an isometric manoeuvre and SNIP a quasi-isometric one [2, 5]. $P_{I,max}$ was measured using a flanged mouthpiece with the manoeuvres repeated at least three times or until two identical readings were obtained [3]. Once the operator was satisfied, the maximum value of three manoeuvres that varied by $<20\%$ was recorded [3]. SNIP was measured during $\geqslant 10$ and $\leqslant 20$ maximal sniffs in a standard manner, according to previously described methods [5]. Briefly, the plug used to obstruct the nostril was an eartip designed for recording auditory evoked potentials (Eartips, 13 mm; Nicolet Biomedical Inc., Madison, WI, USA). The plug was connected to a pressure transducer *via* a catheter, which was as short as possible. Air leakage was detected by obstructing the other nostril during an inspiratory manoeuvre and, when present, was eliminated by adding earplug wax. Detailed instructions as to how to perform the sniff manoeuvre were not given, as they were found to be unnecessary and possibly counterproductive in an earlier study [4]. Patients were encouraged vigorously during all test manoeuvres.

All pressure signals were measured using a differential pressure transducer (Validyne, Northridge, CA, USA), amplified by a carrier amplifier (Validyne) and passed through an analogue–digital board to a computer running AcqKnowledge software (Biopac Systems, Inc., Santa Barbara, CA, USA), which provided visual feedback to improve sniff efficiency. The signal was digitised at 100 Hz. Patients received strong verbal encouragement in addition to the visual feedback, as

suggested in a previous study [11]. The best values of $P_{I,max}$ and SNIP, expressed in cmH_2O, were recorded.

Analysis

Data from the DMD and MMD groups were compared using unpaired nonparametric Mann–Whitney U-tests. Changes in $P_{I,max}$, SNIP and vital capacity (VC) in both populations were assessed using paired Wilcoxon tests.

The method, between SNIP and $P_{I,max}$, that gave the highest value was considered the best method for the patient. Agreement between SNIP and $P_{I,max}$ was evaluated by constructing Bland–Altman plots [12]. Bias was represented by the mean differences between SNIP and $P_{I,max}$. Upper and lower limits of agreement were defined as the 2.5 and 97.5% limits of the distribution of the differences. Precision (the ability to reproduce the same measurement) was represented by the interval (bias $\pm$ SD), where the SD was that of the distribution of differences.

Univariate and multivariate logistic regression analyses were performed in order to evaluate whether SNIP being better than $P_{I,max}$ was associated with the diagnosis (DMD or MMD) and/or the severity of respiratory muscle weakness.

A p-value of <0.05 was considered significant.

RESULTS

The present study included 86 patients, 25 with DMD and 61 with MMD. All patients made two visits (representing 1 yr of follow-up), 63 three visits (2 yrs of follow-up), 29 four visits (3 yrs of follow-up) and four five visits (4 yrs of follow-up).

TABLE 1	Patient characteristics at baseline (BL) and at the second visit 1 yr later	
	MMD	**DMD**
Subjects n	61	25
Age yrs[#]	42.2 ± 11.6	22.1 ± 3.7
Males/females n	42/19	25/0
BMI kg·m^{-2} [#]	25.8 ± 6.2	17.3 ± 5.4
Noninvasive mechanical ventilation n		
BL	5	11
Visit 2	21	17
VC % pred		
BL	63.4 ± 16.9	21.9 ± 9.1
Visit 2	62.0 ± 18.0	$17.7 \pm 7.6^*$
SNIP cmH$_2$O		
BL	45.0 ± 21.1	20.2 ± 9.5
Visit 2	48.9 ± 22.2	$16.3 \pm 9.0^*$
***P*$_{I,max}$ cmH$_2$O**		
BL	43.8 ± 20.0	24.2 ± 9.7
Visit 2	41.5 ± 19.0	$21.3 \pm 7.6^*$

Data are presented as mean $\pm$ SD, unless otherwise stated. MMD: myotonic muscular dystrophy; DMD: Duchenne muscular dystrophy; BMI: body mass index; VC: vital capacity; % pred: % predicted; SNIP: sniff nasal inspiratory pressure; $P_{I,max}$: maximal inspiratory pressure. [#]: determined at BL. *: $p<0.05$ *versus* BL (Wilcoxon test).

Baseline characteristics

Table 1 reports the main patient characteristics at baseline; 16 patients used noninvasive mechanical ventilation. VC, SNIP and $P_{I,max}$ each differed significantly between DMD and MMD patients (Mann–Whitney test; p<0.0001). A single DMD patient had a VC >40% predicted, whereas only two MMD patients had VC <40% pred.

Figures 1a and b show Bland–Altman plots of the difference between $P_{I,max}$ and SNIP against the mean for each population. The bias (mean difference between SNIP and $P_{I,max}$) in MMD patients at baseline was 1.2. The upper and lower limits of agreement were -29.5 and 32.0, and the precision limits were -14.1 and 16.6. The bias for DMD patients at baseline was -4.0. The upper and lower limits of agreement were -15.4 and 7.4, and the precision limits were -9.7 and 1.7. In summary, the limits of agreement were larger in MMD patients than in DMD patients. $P_{I,max}$ was better than SNIP in 20 (80%) of the 25 DMD patients and 30 (49%) of the 61 MMD patients.

The univariate logistic regression analyses performed to identify factors associated with better SNIP than $P_{I,max}$ at baseline showed correlations with VC and diagnosis (MMD or DMD) (p=0.02 and 0.004, respectively). However, on multivariate logistic regression, only a diagnosis of MMD was significantly associated with SNIP being better than $P_{I,max}$ (p=0.03).

Changes over the first year

Table 1 reports the main patient characteristics at the second visit. Mechanical ventilation was started during follow-up in 16 MMD and six DMD patients (table 1). VC, SNIP and $P_{I,max}$ each remained significantly different between MMD and DMD patients (p<0.0001; Mann–Whitney test).

VC, SNIP and $P_{I,max}$ were significantly lower at the second visit in DMD patients (p<0.0015, 0.03 and 0.01, respectively; Wilcoxon test) but not in MMD patients (p=0.09, 0.06 and 0.18, respectively; table 2).

Figures 1c and d show Bland–Altman plots of the difference between $P_{I,max}$ and SNIP against the mean for each population. The bias for MMD patients at visit 2 was 7.4. The upper and lower limits of agreement were -23.9 and 38.8, and the precision limits were -8.2 and 23.1. The bias for DMD patients at visit 2 was -5.0. The upper and lower limits of agreement were -20.0 and 10.0, and the precision limits were -12.5 and 2.5. SNIP was less than $P_{I,max}$ in 23 (92%) of the 25 DMD patients and 19 (31%) of the 61 MMD patients. In summary, the limits
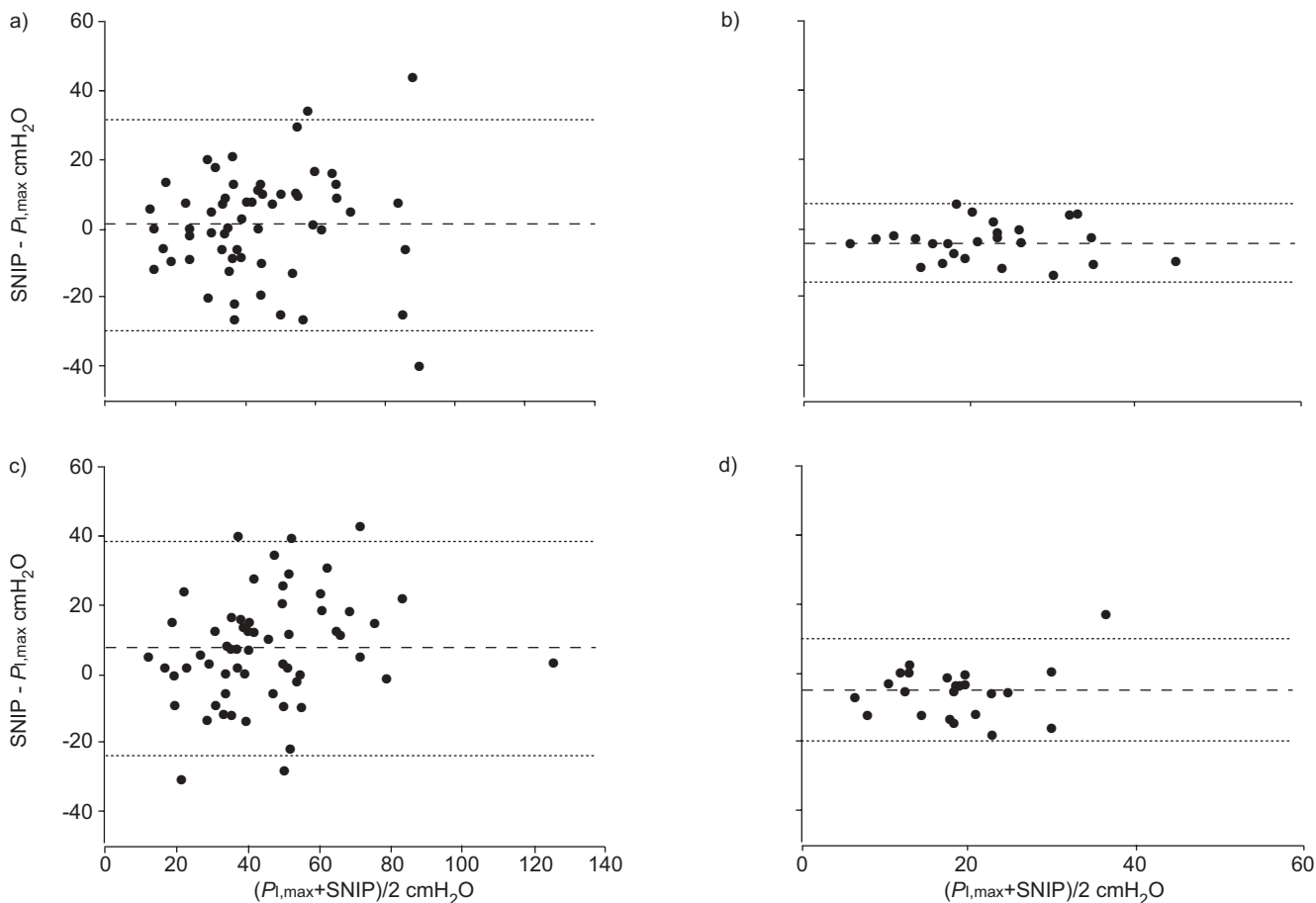
FIGURE 1. Bland–Altman plots showing the difference between sniff inspiratory nasal pressure (SNIP) and maximal static inspiratory pressure ($P_{I,max}$) plotted against the mean of these two variables in a, c) Steinert myotonic muscular dystrophy patients (n=61) and b, d) Duchenne muscular dystrophy patients (n=25) at baseline (a, b) and visit 2 (c, d). -----: mean; ·······: ±2SD.

TABLE 2	Baseline and visit 2 cross-tabulation			
	DMD patients		MMD patients	
	$P_{I,max}$-1 > SNIP-1	SNIP-1 > $P_{I,max}$-1	$P_{I,max}$-1 > SNIP-1	SNIP-1 > $P_{I,max}$-1
$P_{I,max}$-2 > SNIP-2 n	20	3	15	4
SNIP-2 > $P_{I,max}$-2 n	0	2	15	27

DMD: Duchenne muscular dystrophy; MMD: myotonic muscular dystrophy; $P_{I,max}$-1: maximal inspiratory pressure ($P_{I,max}$) at baseline; SNIP-1: sniff nasal inspiratory pressure (SNIP) at baseline; $P_{I,max}$-2: $P_{I,max}$ at visit 2; SNIP-2: SNIP at visit 2.

of agreement remained larger at visit 2 in MMD patients than in DMD patients. In addition, although $P_{I,max}$ remained better than SNIP in DMD patients (compared with baseline, the percentage of patients for whom $P_{I,max}$ was best increased from 80 to 88% of the DMD population), the number of MMD patient for whom $P_{I,max}$ was better than SNIP decreased from 30 (49%) to 19 (31%).

In order to further evaluate changes in the best method ($P_{I,max}$ or SNIP) between the baseline visit and visit 2, a contingency table was built with two variables (baseline and visit 2; table 2). In the 20 DMD patients with $P_{I,max}$ as the best method at baseline, $P_{I,max}$ was still the best method after 1 yr. Of the five DMD patients with SNIP as the best method at baseline, three changed to $P_{I,max}$ being the best method after 1 yr. The best method changed between baseline and visit 2 in 15 of the 30 MMD patients with $P_{I,max}$ as the best method at baseline and in four of the 31 MMD patients with SNIP as the best method at baseline.

Long-term follow-up

Since follow-up duration varied between patients, the best method ($P_{I,max}$ or SNIP) at baseline was crossed with the method that was most often best at subsequent visits (table 3). In the 17 DMD patients with $P_{I,max}$ as the best method at baseline, $P_{I,max}$ remained the best method throughout follow-up. In contrast, both DMD patients with SNIP as the best method at baseline switched to $P_{I,max}$ being the best method during follow-up. Of the 20 MMD patients with $P_{I,max}$ as the best method at baseline, nine switched to SNIP during follow-up; of the 24 MMD patients with SNIP as the best method at baseline, only one switched to $P_{I,max}$.

DISCUSSION

The present study comparing $P_{I,max}$ and SNIP in patients with two of the most common chronic neuromuscular diseases showed that the test yielding the best value at baseline was usually $P_{I,max}$ in DMD patients, whereas a more balanced distribution was seen in MMD patients. The best test changed significantly more often (30%) in patients with MMD than in those with DMD (12%). The frequency of switching from one test to the other differed according to which test yielded the best value at baseline. Thus, when $P_{I,max}$ was better than SNIP at baseline in DMD patients (80%), $P_{I,max}$ remained better (100%) during follow-up. In contrast, when SNIP was better than $P_{I,max}$ at baseline in DMD patients, a switch to $P_{I,max}$ was often seen during follow-up. In MMD patients, when SNIP was better than $P_{I,max}$ at baseline (51%), SNIP usually (88%) remained better at the second visit; whereas when $P_{I,max}$ was better than SNIP at baseline, a switch was often (50%) noted at the second visit.

The severity of respiratory function impairment differed between the two groups. However, it is well known that hypercapnia and a need for mechanical ventilation do not occur at the same level of restrictive disease in MMD patients as in DMD patients [13–16]. The need and reasons for monitoring respiratory function in the two populations in the present study were roughly similar, despite the difference in respiratory muscle function.

$P_{I,max}$ is the most widely used volitional test for evaluating inspiratory muscle strength [4]. However, many patients with neuromuscular disease find the $P_{I,max}$ manoeuvre difficult to perform. In addition, air leaks around the mouthpiece may be difficult to eliminate. The sniff test relies on a natural effort that

TABLE 3	Baseline and subsequent visit cross-tabulation			
	DMD patients		MMD patients	
	$P_{I,max}$-1 > SNIP-1	SNIP-1 > $P_{I,max}$-1	$P_{I,max}$-1 > SNIP-1	SNIP-1 > $P_{I,max}$-1
$P_{I,max}$-FU > SNIP-FU	17	2	11	1
SNIP-FU > $P_{I,max}$-FU	0	0	9	23

During follow-up, the method indicated to be the best method is that that was more frequently the best method. DMD: Duchenne muscular dystrophy; MMD: myotonic muscular dystrophy; $P_{I,max}$-1: maximal inspiratory pressure ($P_{I,max}$) at baseline; SNIP-1: sniff nasal inspiratory pressure (SNIP) at baseline; $P_{I,max}$-FU: $P_{I,max}$ during follow-up; SNIP-FU: SNIP during follow-up.

most patients find easier to perform than the $P_{I,max}$ manoeuvre [17]. SNIP is a more pleasant technique than $P_{I,max}$ for most patients [6, 18, 19] and it does not raise the leak problems with a mouthpiece sometimes observed in neuromuscular patients. SNIP may carry less risk of fatigue, as the manoeuvre is shorter than $P_{I,max}$, which requires a sustained peak pressure for $\geqslant 1$ s. Predicted normal values adjusted for age and sex are available in the literature for both adults and children [5, 19–21]. Based on these normative values, it is generally considered that a SNIP of <-70 cmH_2O in males or <-60 cmH_2O in females is unlikely to be associated with inspiratory muscle weakness [4, 20, 22].

SNIP reflects the oesophageal pressure decrease during the sniff manoeuvre, which accurately reflects inspiratory muscle strength in normal individuals [5]. However, SNIP may underestimate inspiratory muscle strength in specific populations, such as individuals with nasal obstruction [21] and patients with chronic obstructive pulmonary disease (since sniff is a short manoeuvre and transmission of oesophageal pressure to the nose is reduced in these patients [23]). Underestimation probably occurred in patients with severe neuromuscular disease who were unable to produce an initial negative transnasal pressure of 10–15 cmH_2O, a necessary condition for collapsing the unplugged nostril valve [24]. In order to obtain this negative transnasal pressure, the patient must produce an oesophageal pressure swing of >10–15 cmH_2O, which depends on inspiratory peak flow and on nostril resistance when the valve is not collapsed. In addition, SNIP is generated in a ballistic manoeuvre during which the inspiratory muscles shorten to a greater extent and at a higher speed than during the $P_{I,max}$ manoeuvre, which is a more sustained isometric effort. Given both the force–velocity and force–length relationships of striated muscle, SNIP should be <$P_{I,max}$, as pressure generation falls when the operating length of the muscle decreases and when the velocity of muscle shortening increases. Accordingly, $P_{I,max}$ was >SNIP in patients with severe neuromuscular disease [7, 25]. The present results are in agreement with these previous studies; muscle weakness was more severe in the DMD group than in the MMD group, and $P_{I,max}$ yielded the best value at baseline in most DMD patients, whereas a more balanced distribution was noted in the MMD patients.

Sniffing is more natural than the $P_{I,max}$ manoeuvre, a fact believed to explain the higher SNIP than $P_{I,max}$ in normal individuals [6] and patients with limited respiratory muscle weakness [7, 25], such as those in the present MMD group. In addition, the SNIP is measured from the peak pressure, whereas $P_{I,max}$ is the mean pressure sustained over 1 s, which includes an early pressure peak followed by a decline to a lower sustained pressure. Thus, the nature of the predominant effect influences the SNIP relative to the $P_{I,max}$. Finally, due to the differences in the type of effort and pattern of muscle activation between the two manoeuvres, SNIP and $P_{I,max}$ probably reflect different aspects of inspiratory muscle function. A more sustained manoeuvre may achieve greater inspiratory muscle activation in patients with more severe weakness.

In addition to the severity of respiratory muscle weakness, a number of other factors may contribute to the difference between the DMD and MMD groups regarding the method

producing the best value. Logistic regression analysis was performed to look for factors associated with a better SNIP than $P_{I,max}$ at baseline. When VC and diagnosis were entered in a univariate analysis, correlations were observed with both factors (p=0.02 and 0.004, respectively). However, when these factors were combined in an multivariate logistic regression analysis, only diagnosis was significantly associated with SNIP being better than $P_{I,max}$ (p=0.03). $P_{I,max}$ and SNIP are both volitional methods that require cooperation and motivation from the patient. The greater variability in the patients with MMD may be ascribable to lack of motivation and poor coordination. MMD is an autosomal dominant multisystemic disorder that often involves the brain and results in cognitive impairment [26], thereby possibly increasing the variability in volitional manoeuvres. Interestingly, the considerable variability in the MMD group regarding the best test was observed only when $P_{I,max}$ was better at baseline, suggesting that the ability to perform SNIP properly persisted over time. Conversely, some of the MMD patients who were unable to perform SNIP properly at baseline learnt the manoeuvre during follow-up. The ability to perform $P_{I,max}$, a more difficult and less natural manoeuvre than SNIP, was lost over time in some patients. Cognitive tests were not performed, and thus it was not possible to assess possible relationships between variability in the best test and cognitive impairment.

In order to assess the hypothesis that a better SNIP than $P_{I,max}$ is more likely to occur in patients with mild rather than severe muscle weakness, NICOT et al. [27], a group from a paediatric pulmonary department with extensive experience in respiratory muscle function testing in neuromuscular patients, were asked to compare SNIP and $P_{I,max}$ in 15 DMD children with VC >40% pred. SNIP was better than $P_{I,max}$ in eight patients, a proportion similar to that seen in the current MMD population. This finding supports the present hypothesis.

In conclusion, these two volitional tests are not interchangeable in patients with chronic neuromuscular disorders [6–8]. Nevertheless, the use of sniff nasal inspiratory pressure measurement has only been suggested for monitoring patients with amyotrophic lateral sclerosis [18, 28]. In the present adult patients with Duchenne muscular dystrophy, the maximal inspiratory pressure manoeuvre generally yielded the best value at baseline, and maximal inspiratory pressure assessment remained the best test in most patients during follow-up. Thus, maximal inspiratory pressure measurement alone may be sufficient in this adult population. In patients with myotonic muscular dystrophy, in contrast, the marked variability in the test yielding the best value at baseline indicates a need for performing both tests at baseline. During follow-up, when sniff nasal inspiratory pressure measurement yielded the best value at baseline, sniff nasal inspiratory pressure remained best during follow-up. Therefore, using sniff nasal inspiratory pressure measurement alone during follow-up may be appropriate in this subpopulation provided vital capacity remains >40% pred. Nevertheless, maximal inspiratory pressure and sniff nasal inspiratory pressure manoeuvres should both be performed when the patient shows poor cooperation or difficulty in understanding and/or performing the manoeuvre.

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