PAPER

Limitations of sniff nasal pressure in patients with severe neuromuscular weakness

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Background: Inspiratory muscle strength in patients with neuromuscular disorders can be assessed using sniff inspiratory nasal pressure (P_{nsn}) and maximum inspiratory mouth pressure (PI_{max}). However, the relative merits of P_{nsn} against PI_{max} are not known in patients with severe neuromuscular disease.

Objective: To investigate whether severity of disease modifies the relation between P_{nsn} and PI_{max} .

Methods: Vital capacity (VC), P_{nsn} , and PI_{max} were measured in 258 patients with neuromuscular disorders.

Results: Data were analysed from 241 patients, 17 being unable to perform PI_{max} or P_{nsn} manoeuvres. The correlation between P_{nsn} and PI_{max} was +0.94 ($p < 0.0001$), with a mean (SD) difference between P_{nsn} and PI_{max} of -4.8 (21.2) cm H_2O (the limits of agreement were 37.6 and -47.2 cm H_2O). VC (% predicted) was positively correlated with P_{nsn}/PI_{max} ($r = +0.86$; $p < 0.0001$), with a lower P_{nsn}/PI_{max} value in patients with a VC $< 40\%$ of predicted than in those with a VC $> 40\%$ (0.80 (0.35) v 1.04 (0.41); $p < 0.0001$).

Conclusions: PI_{max} is greater than P_{nsn} in patients with a severe restrictive ventilatory defect caused by neuromuscular disease. P_{nsn} may not accurately reflect inspiratory muscle strength in such patients and it is thus advisable to use both tests.

Although sniff inspiratory nasal pressure (P_{nsn}) and maximum inspiratory mouth pressure (PI_{max}) are used to assess inspiratory muscle strength in patients with neuromuscular disorders,^{1,2} it is unclear whether the values for P_{nsn} are higher than PI_{max} . Stefanutti *et al* reported that the airway pressure during a sniff manoeuvre was higher than the pressure generated during a maximal static inspiratory manoeuvre,¹ whereas a recent study by Iandelli *et al* suggested the opposite, especially in patients with severe neuromuscular disorders.² However, in those studies less than 20% of the patients had a vital capacity (VC) below 40% of predicted, which is a level that can be considered to indicate a severe restrictive ventilatory defect (all patients with symptoms of nocturnal hypoventilation and a VC less than 50% of predicted should be considered for long term non-invasive ventilation³). Thus our aim in the current study was to examine the affect of moderate to severe lung restriction on the relation between P_{nsn} and PI_{max} in patients with neuromuscular disease.

METHODS

We studied 258 patients with neuromuscular disorders as part of routine clinical evaluation over a three year period at the Raymond Poincaré hospital. Seventeen patients were excluded: nine were unable to perform PI_{max} manoeuvres and eight were unable to perform P_{nsn} manoeuvres.

All the tests were conducted in a single session, with patients in a seated position. P_{nsn} and PI_{max} were both measured from functional residual capacity (FRC) in a standard manner, according to previously described methods (PI_{max} is an isometric manoeuvre, while P_{nsn} is a quasi-isometric manoeuvre).^{4,5} P_{nsn} was measured during 10 maximal sniffs,⁶ while PI_{max} was measured with a flanged mouthpiece with the manoeuvres repeated at least three times or until two identical readings were obtained.⁷ All 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 System, Goleta, CA, USA). Patients received strong verbal encouragement with visual feedback, as previous studies have suggested.⁸ Spirometry and lung volumes were measured according to standard guidelines and reported as per cent predicted.⁹ Severity of disease was defined according to VC (% predicted), with a VC $< 40\%$ being arbitrarily defined as severe restrictive lung disease. PI_{max} and P_{nsn} are expressed as positive values although they are negative pressures with respect to atmosphere.

STATISTICAL ANALYSIS

All results are expressed as mean (SD). The correlation between P_{nsn} and PI_{max} was assessed using linear regression analysis. Agreement between P_{nsn} and PI_{max} was determined using a Bland and Altman plot.¹⁰ P_{nsn}/PI_{max} was used to evaluate the relations between P_{nsn} , PI_{max} , and VC using linear regression analysis. The difference between P_{nsn}/PI_{max} in the patients with a VC $< 40\%$ of predicted and patients with a VC $> 40\%$ of predicted was assessed using an unpaired *t* test.

RESULTS

Two hundred and forty one patients with neuromuscular disorders were eligible for analysis (table 1). Their mean (SD) age was 45.1 (16.4) years and 58% were male. No problems were encountered with air leaks during the PI_{max} manoeuvre using the flange mouthpiece. Mean VC (% predicted) was 51.9 (26.0)%, mean PI_{max} was 45.6 (28.2) cm H_2O , and mean P_{nsn} was 41.5 (26.4) cm H_2O (35.2% of patients had a VC $< 40\%$ of predicted and 37.8% were receiving non-invasive ventilation).

Abbreviations: FRC, functional residual capacity; NIV, non-invasive ventilation; PI_{max} , inspiratory mouth pressure; P_{nsn} , sniff inspiratory nasal pressure; VC, vital capacity

Table 1 Patients with neuromuscular disorders eligible for analysis

	Diagnosis	N
Muscle	Myotonic dystrophy	64
	Duchenne muscular dystrophy	23
	Congenital myopathies	19
	Acid maltase deficiency	16
	Limb girdle muscular dystrophy	14
	Mitochondrial myopathy	3
	Becker's muscular dystrophy	3
	Facioscapulohumeral dystrophy	3
	Congenital muscular dystrophy	3
	Inclusion body myositis	1
	Polymyositis	1
	Poliomyelitis	41
Nerve	Guillain-Barré syndrome	8
	Spinal muscular atrophy	7
	Charcot-Marie-Tooth	2
	Bilateral diaphragm paralysis	1
	Myasthenia gravis	32

As expected, there was a positive correlation between Pn_{sn} and PI_{max} ($r = +0.94$, $p < 0.0001$). However, the agreement between Pn_{sn} and PI_{max} , assessed using a Bland and Altman plot¹⁰ was relatively poor. Figure 1 shows a Bland and Altman plot with a mean difference between Pn_{sn} and PI_{max} of -4.8 (21.2) cm H₂O and limits of agreement of 37.6 and -47.2 cm H₂O. Despite this, we observed a positive correlation between VC (% predicted) and Pn_{sn}/PI_{max} (fig 2; $r = +0.86$; $p < 0.0001$), which indicates that as VC falls the absolute value of Pn_{sn} declines more than PI_{max} —that is, PI_{max} is greater than Pn_{sn} . Furthermore, the mean Pn_{sn}/PI_{max} value was lower in patients with a VC $< 40\%$ of predicted than in patients with a VC greater $> 40\%$ of predicted (0.86 (0.35) v 1.04 (0.41); $p < 0.0001$). When expressed in absolute values (cm H₂O), Pn_{sn} was less than PI_{max} in 147 of the 241 patients.

DISCUSSION

As in previous studies of patients with neuromuscular disorders, we found a positive correlation between Pn_{sn} and PI_{max} , but with relatively poor agreement between these two tests.^{1,2} However, the present findings differ from previous studies^{1,6} in that the value of PI_{max} was at least the same as or even greater than Pn_{sn} , particularly in those patients with

Figure 1 Bland-Altman plot: difference between sniff inspiratory nasal pressure (Pn_{sn}) and maximum static inspiratory pressures (PI_{max}) plotted against the mean of these two variables ($n = 241$). Filled symbols, patients with VC $< 40\%$ of predicted; empty symbols, patients with VC $> 40\%$ of predicted. VC, vital capacity.

Figure 2 Relation between per cent predicted (% predicted) vital capacity (VC) and sniff inspiratory nasal pressure, normalised for maximum static inspiratory pressure (Pn_{sn}/PI_{max}) ($n = 241$; $r = 0.86$; $p < 0.0001$). Horizontal dashed line indicates VC (% predicted) of 40% and the vertical line indicates unity between Pn_{sn} and PI_{max} ($Pn_{sn}/PI_{max} = 1$).

severe ventilatory restriction caused by neuromuscular disease. In fact, the present data are more consistent with, and extend, the observations of Iandelli *et al*,² and highlight the limitation of Pn_{sn} in this particular patient population.

Comparing our study with that of Stefanutti *et al*,¹ there are two major differences. The first is that overall Pn_{sn} is less than PI_{max} ; the second is that advanced lung restriction from severe neuromuscular disease is associated with a lower value for Pn_{sn} than for PI_{max} . This disparity between the current study and Stefanutti's could be the result of technical differences, as both Pn_{sn} and PI_{max} manoeuvres in the earlier study were done without visual feedback. As sniffing is a more natural manoeuvre than PI_{max} ,¹¹ it is conceivable that without visual feedback Pn_{sn} would be greater than PI_{max} . In addition, in the Stefanutti study there were only three attempts at the unfamiliar PI_{max} manoeuvre, but 10 sniff manoeuvres were done. In our clinical practice we get the patient to do at least three PI_{max} manoeuvres or continue until two identical readings are obtained,⁷ and our greater PI_{max} value may therefore be attributable to the learning effect. Furthermore, the current study differs from previous studies of patients with amyotrophic lateral sclerosis where facial muscle weakness and leaks around the mouthpiece were a major problem causing a reduction in PI_{max} .^{12,13}

The results of our study are supported by the findings of earlier studies in patients with chronic stable inspiratory muscle weakness² and in patients with acute respiratory failure,¹⁴ showing that Pn_{sn} underestimates inspiratory muscle strength in patients with severe neuromuscular disease. Although we acknowledge that VC is an indicator of both respiratory muscle function and lung and chest wall compliance, it is more commonly used than Pn_{sn} or PI_{max} in clinical practice to monitor sequential changes in respiratory impairment as neuromuscular disease progresses. Furthermore, in patients with advanced restrictive ventilatory defects secondary to neuromuscular disease there are marked reductions in VC with relatively small changes in maximum pressures, owing to the curvilinear relation between VC and maximum inspiratory pressures¹⁵—that is, VC is probably a more sensitive marker of disease progression in advanced disease than in mild disease.¹⁶

To explain the greater decline of Pn_{sn} than of PI_{max} in patients with severe neuromuscular disease, we must

consider the mechanism of sniffing compared with the maximal static inspiratory manoeuvre. Sniff is generated during a ballistic manoeuvre where the inspiratory muscles shorten to a greater extent and at a higher speed than during a $PI_{\max}$ manoeuvre, which is a more sustained isometric task. Thus as a result of both the force–velocity and force–length relations in striated muscle, Pn_{sn} should be less than $PI_{\max}$ —that is, pressure generation falls with a reduction in the operating length of the muscle and also with an increase in the velocity of muscle shortening. However, as sniffing is more natural than the maximum inspiratory static manoeuvre, it is believed that this accounts for the higher Pn_{sn} than $PI_{\max}$ value in normal subjects.⁶ Also, the value for Pn_{sn} is measured from the peak pressure, whereas $PI_{\max}$ is the average pressure sustained over one second, which includes an early peak pressure before falling off to a lower sustained pressure. Thus whichever of these two effects is greatest will influence the value of Pn_{sn} more than $PI_{\max}$. Nevertheless, patients with severe neuromuscular disorders may not be able to perform a rapid sniff manoeuvre owing to significant muscle atrophy. Furthermore, because of the differences in the type of effort and the pattern of muscle activation in the two manoeuvres, the Pn_{sn} value and $PI_{\max}$ value probably reflect different aspects of inspiratory muscle function. Thus a more sustained manoeuvre may achieve a greater pattern of inspiratory muscle activation in severely affected patients.

CONCLUSIONS

In patients with moderate to severe neuromuscular disease, overall $PI_{\max}$ yielded similar values to Pn_{sn} . Although there was a close correlation between Pn_{sn} and $PI_{\max}$, there was a relatively poor agreement between the two variables, and the value for $PI_{\max}$ was higher than Pn_{sn} , particularly in patients with a severe restrictive ventilatory defect. Pn_{sn} may overestimate the level of inspiratory muscle weakness, which challenges the view that this is the most appropriate test in this particular group of patients. We therefore suggest that $PI_{\max}$ and Pn_{sn} are not interchangeable but are complementary tests and should be used in combination with VC for a complete sequential assessment of inspiratory muscle strength in patients with neuromuscular disease.

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