Sniff nasal inspiratory pressure in children with muscular, chest wall or lung disease

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Abstract

Sniff nasal inspiratory pressure is proposed as a noninvasive test of inspiratory muscle strength. During this manoeuvre, the nasal pressure is supposed to reflect oesophageal pressure. The aim of the study was to compare the nasal pressure with the oesophageal pressure during a maximal sniff in children with neuromuscular disease (NM, n=78), thoracic scoliosis (n=12), and cystic fibrosis (CF, n=23).

A significant correlation was observed between the sniff nasal and oesophageal pressure. The ratio of the sniff nasal/oesophageal pressure was lower in patients with CF (0.72 ± 0.13) than in NM patients (0.83 ± 0.17), or patients with thoracic scoliosis (0.86 ± 0.10). In patients with CF and NM disease, this ratio was not correlated to age or spirometric data. The difference between the sniff oesophageal and nasal pressure exceeded 15 cm H2O in 17% of the NM patients, 33% of the patients with thoracic scoliosis, and 87% of the CF patients.

Sniff nasal pressure often underestimates the strength of inspiratory muscles in CF. Such an underestimation occurs more rarely in NM disorders and in thoracic scoliosis. A normal value excludes inspiratory muscle weakness but a low value requires the measurement of the oesophageal pressure.

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**Key words**: cystic fibrosis, nasal pressure, neuromuscular disease, oesophageal pressure, respiratory muscles.
Introduction

Classically, the strength of the inspiratory muscles is assessed noninvasively by the pressure measured at the mouth and sustained for at least 1 sec during a maximal inspiratory effort performed against an occlusion (PImax) [1, 2]. It is generally admitted that if three equal maximal efforts are obtained, then the subject is supposed to have realised a maximal effort. But it has been shown that reproducibility does not ensure maximality [3]. Since PImax is not easy to perform, the results are prone to important variations and low results may reflect not only inspiratory muscle weakness, but also a lack of motivation and/or coordination of the patient. Moreover, many other factors such as a training effect, chest wall configuration and stabilisation during the manoeuvres may contribute to the range of pressures observed in normal children [4-6].

Because sniff is a natural manoeuvre which many children find easier to perform than static efforts, sniff nasal inspiratory pressure (SNIP) has been proposed as an alternative, or as a complementary test to the Pimax [7-10]. The SNIP manoeuvre consists of measuring nasal pressure in an occluded nostril during a maximal sniff performed through the contralateral nostril from functional residual capacity (FRC) [11]. Transmission of the oesophageal pressure (Poes) to the nose is obtained considering that a transnasal pressure of 10 to 15 cm H₂O is necessary to obtain a collapse of the unplugged nostril valve in adults [12]. Normal values for the SNIP have been established for children as for adults [7-10]. Values in healthy children aged 6 to 17 years are similar to those measured in adults with a SNIP of 99 - 117 cm H₂O in boys and 92 - 97 cm H₂O in girls [8]. SNIP correlates with age and weight [8]. The main advantage of the SNIP manoeuvre is that it is a more pleasant
technique than Pmax for most of the subjects, and requires little practice. It solves the leak problems sometimes observed with a mouthpiece in neuromuscular patients [11, 13]. It reduces the risk of fatigue because the manoeuvre is natural, easy and shorter than the Pmax, which requires a sustained peak pressure of at least 1 sec.

A limitation of the SNIP manoeuvre is that it may underestimate Poes in subjects with nasal obstruction, significant lung or airway disease [14], and probably in severe neuromuscular patients considering that a transnasal pressure of 10-15 cm H$_2$O is necessary to obtain a collapse of the unplugged nostril valve to enable an accurate approximation of the Poes swing [12]. A comparison of SNIP with Poes during the same sniff manoeuvre (Sniff Poes) has been made in healthy adults, and in adult patients with neuromuscular disease [7] or chronic obstructive pulmonary disease (COPD) [14] but never in children with either neuromuscular or lung disease. Because most neuromuscular and lung diseases exhibit a progressive course, they are generally less severe in children. In this age group, recurrent rhinitis and upper airway infections are common. The relationship between Sniff Poes and SNIP may thus be different in children than in adults. The aim of the present study was thus to compare SNIP with Sniff Poes in children with neuromuscular disease, severe scoliosis, and cystic fibrosis (CF).

**Material and methods**

**Patients**

The study protocol was approved by our institutional review board, and informed consent was obtained from all the patients and their parents.
The patients were recruited on a consecutive basis from our outpatient clinic. The paediatric patients belonged to three categories, neuromuscular disease (n=78), thoracic scoliosis (n=12), and CF (n=23) (Table 1). None of the patients had obvious nasal obstruction or congestion by checking the ability of the patient to breathe through one nostril while the other was occluded (Table 1).

**Measurements**

Poes was measured using a catheter mounted pressure transducer system (Gaeltec, Dunvegan, Isle of Skye, UK) [15] inserted pernasally after careful local unilateral anaesthesia (lidocaine 2%, Astra Zeneca, Rueil-Malmaison, France). Appropriate placement of the Poes transducer was assessed with the usual method [16]. The plug used to obstruct the other nostril was an eartip used for auditory evoked potentials (Eartips, 13 mm, Nicolet, Madison, WI). It incorporated the distal 1-2 cm of a 90 cm polyethylene catheter with a 2 mm internal diameter (Intersurgical Scientific Instruments, Oxford, UK). The other extremity of the catheter was connected to a differential pressure transducer (Validyne DP15, Northridge, CA), which was wired to a carrier demodulator (Validyne CD15) and passed through an analogue-digital board to a computer running an adequate software (Biopac System, Goleta, CA). The absence of air leak around the eartip was ascertained by occluding the controlateral nostril during an inspiratory effort.

The sniffs manoeuvres were performed in a single session with the patient seated in front of the computer screen. The patient was instructed to perform short sharp sniffs with closed mouth, starting from the end expiratory volume after a quiet breath. Each sniff was associated by a strong verbal encouragement with visual feedback.
Twenty sniffs were at least performed, separated by 30 sec, until a consistent value was reached [17]. Sniff Poes and SNIP represented the amplitudes of pressure changes, and were expressed in absolute values. For each patient, the highest sniff value was taken.

All the patients were asked to perform at least 3 physician-accepted forced vital capacity (FVC) curves, and the curves with the highest FVC were used for the final analysis [18]. Results were expressed as a percent of published values (% pred) with height calculated as the arm span for the patients with neuromuscular disease and scoliosis [19, 20].

In order to validate our bench, SNIP was compared to Sniff Poes in 8 healthy adults, mean age 28.5 ± 5.6 yrs, who were free of any known respiratory, ENT, or neurological disease. Mean Sniff Poes was 93 ± 27 cm H2O and mean SNIP was 86 ± 27 cm H2O. The SNIP/Sniff Poes ratio was 0.93, which is comparable to the value reported in the literature (Figure 1 on line) [7].

Statistical analysis

The agreement between SNIP and Sniff Poes was assessed by the method of differences against the means, according to Bland and Altman [21]. The relationships between the SNIP/Sniff Poes ratio and age and spirometric data were assessed by linear regression analysis. For quantitative variables, comparisons between the patient groups were conducted using ANOVA. A p value < 0.05 was considered as significant.

Results
Patients

The characteristics of the patients are represented in Table 1. Forty two neuromuscular patients had Duchenne muscular dystrophy, 14 patients had spinal muscular amyotrophy and the others had another congenital myopathy. Four patients (3 patients with Duchenne muscular dystrophy and one patient with spinal muscular amyotrophy) required long term nocturnal noninvasive positive pressure ventilation. FVC and forced expiratory volume in one second (FEV1) were markedly reduced in all the patient groups.

Sniff Poes and SNIP values

The Sniff Poes and SNIP values were significantly lower in the patients with neuromuscular disease as compared to the patients with scoliosis and CF (Table 1).

The SNIP/Sniff Poes ratio was lower in patients with CF (0.72 ± 0.13) than in patients with neuromuscular disease (0.83 ± 0.17), or thoracic scoliosis (0.86 ± 0.10) (Table 1). Figure 1 represents the plots of the difference between Sniff Poes and SNIP against their mean for the 3 patient groups.

The mean difference between Sniff Poes and SNIP was significantly greater in the patients with CF than for the other groups. When choosing an arbitrary difference of 15 cm H₂O between Sniff Poes and SNIP, 17% of the patients with neuromuscular disease, 33% of the patients with thoracic scoliosis, and 87% of the patients with CF had a [Sniff Poes – SNIP] > 15 cm H₂O. The SNIP/Sniff Poes ratio was not correlated with age or spirometric data such as FVC, FEV1, and the FEV1/FVC ratio in any of the 3 patient groups.
Discussion

Our results show that SNIP may underestimate Sniff Poes in paediatric patients with obstructive lung disease such as CF, but also in patients with restrictive lung disease, such as neuromuscular disorders and scoliosis.

The SNIP manoeuvre has been shown to be both easy to perform and a reliable test of inspiratory muscle strength. Indeed, inspiratory muscle strength may be better reflected by SNIP than by Pimax [22], which is more difficult to perform, in particular in children [10]. During the sniff manoeuvre, the nasal valve located in the first 2.5 cm from the external orifice collapses when a critical transnasal pressure of 10-15 cm H₂O is reached [12, 23]. However, this value was measured in adults and may be different in children. In subjects without obstruction of the upper airways and normal lung and airway mechanics, there is only a small pressure gradient between the alveoli and extra-thoracic airways located proximally to the point of collapse. As such, SNIP has proven to be a reliable estimate of Sniff Poes in healthy adults and in adult patients with neuromuscular disease [7]. However, in adult patients with COPD, SNIP may underestimate Sniff Poes [14]. This difference is explained by the short and dynamic character of the sniff manoeuvre. Indeed, the transmission of pressure changes from the alveoli to the mouth depends on a time constant, which is the product of airway resistance and upper airway compliance. This time constant is increased in patients with COPD, explaining the dampening of the pressure changes during a short manoeuvre such as a sniff. Similar observations have also been observed with the occlusion pressure, i.e. the pressure change measured at the mouth 0.1 second after the onset of the inspiration [24-26]. CF lung disease is characterised by progressive airway obstruction, due to a vicious circle of bronchial
infection and inflammation. In agreement with the observation in patients with COPD, underestimation of Sniff Poes by SNIP was commonly observed in the patients with CF included in the present study [14]. But as in adult patients with COPD, the underestimation of Sniff Poes by SNIP in children with CF did not correlate with the degree of lower airway obstruction. One explanation may be that FEV1 and the FEV1/FVC ratio reflect expiratory flow whereas the sniff is an inspiratory manoeuvre [14].

Obstruction of the upper airways may also contribute to the difference between SNIP and Sniff Poes. Nasal obstruction, due to nasal inflammation or polyposis, affects 32% to 65% of CF patients [27-29]. Indeed, a systematic clinical and radiological ENT evaluation has been performed in 75 patients from our and other CF Parisian clinics and showed that 32% of the patients presented nasal obstruction and 43% nasal polyps [27]. In another French study, 50% of 78 CF patients aged 3 to 28 years, presented nasal polyps [28]. Even if nasal polyps are more common in adult CF patients [28], we did not observe a correlation between the SNIP/Sniff Poes ratio and age in the CF population of the present study. For the patients with NM disease and thoracic scoliosis, the most plausible reason for the underestimation of Sniff Poes by SNIP is nasal congestion and hypertrophy of the adenoids (and tonsils) which is very common in young children. The exclusion of the patients with obvious nasal obstruction or congestion seems to be insufficient to avoid this underestimation.

Although we acknowledge that the inclusion of healthy children could have strengthened our results, there is an international agreement among paediatricians on the impossibility to perform invasive studies, such as the introduction of an oesophageal catheter, in healthy children [30, 31]. For this reason, we evaluated
adult controls in our laboratory whose results were comparable to those observed by Uldry et al. [32].

In conclusion, our results show that SNIP often underestimates the strength of inspiratory muscles in CF. Such an underestimation occurs more rarely in neuromuscular disorders and in thoracic scoliosis. As such, the SNIP may be useful as a screening test, normal values excluding inspiratory muscle weakness in children. But in case of low values, the measurement of Poes is warranted to rule out an erroneous diagnosis of inspiratory muscle weakness.

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References


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30 National Health and Medical Research Council (NHMRC). National statement on ethical conduct in research involving humans.1999; Canberra.

Table 1: Characteristics of the patients.

<table>
<thead>
<tr>
<th></th>
<th>Neuromuscular disease n=78</th>
<th>Thoracic scoliosis n=12</th>
<th>Cystic fibrosis n=23</th>
</tr>
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<tbody>
<tr>
<td>Age (years) range</td>
<td>12.7 ± 3.7 4 - 18</td>
<td>14.5 ± 2.5 9 - 18</td>
<td>13.8 ± 2.9 7 - 18</td>
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<td>Female/Male</td>
<td>20/58</td>
<td>10/2</td>
<td>13/10</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>42.2 ± 3.7 146 ± 4</td>
<td>49.5 ± 11.6 163 ± 11</td>
<td>38.3 ± 9.7 150 ± 10</td>
</tr>
<tr>
<td>Height (cm) ‡</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>FVC (% pred)</td>
<td>54 ± 29</td>
<td>55 ± 25</td>
<td>38 ± 29</td>
</tr>
<tr>
<td>FEV1 (% pred)</td>
<td>64 ± 35</td>
<td>50 ± 24</td>
<td>30 ± 29</td>
</tr>
<tr>
<td>FEV1/FVC (% pred)</td>
<td>107 ± 29 (n=54)</td>
<td>103 ± 55 (n=10)</td>
<td>75 ± 17 (n=23)</td>
</tr>
<tr>
<td>Sniff Poes (cm H₂O)</td>
<td>49 ± 4 #</td>
<td>82 ± 25</td>
<td>93 ± 29</td>
</tr>
<tr>
<td>SNIP (cm H₂O)</td>
<td>41 ± 4 #</td>
<td>70 ± 25</td>
<td>66 ± 29</td>
</tr>
<tr>
<td>SNIP/Sniff Poes ratio</td>
<td>0.83 ± 0.17</td>
<td>0.86 ± 0.10</td>
<td>0.72 ± 0.13 §</td>
</tr>
</tbody>
</table>

Abbreviations: FVC: forced vital capacity, expressed as a percentage of predicted value, FEV1: forced expiratory volume in one second, expressed as a percentage of predicted value, Sniff Poes: oesophageal pressure during a maximal sniff manoeuvre, SNIP: sniff nasal inspiratory pressure during a maximal sniff manoeuvre. ‡ Height is given as the arm span for patients with neuromuscular disease and thoracic scoliosis. # p<0.0001 as compared to the patients with thoracic scoliosis or cystic fibrosis. § p<0.005 as compared to the patients with neuromuscular disease or thoracic scoliosis.
Legends of Figure 1

Difference between the sniff nasal inspiratory pressure (SNIP) and the sniff nasal oesophageal pressure (Sniff Poes) against the mean of these two variables in the 3 groups of patients.

The plain lines represent the mean values and the dotted lines the ± 2 standard deviations.

Figure 1