ELSEVIER

Brain & Development xxx (2018) xxx–xxx

BRAIN & DEVELOPMENT
Official Journal of the Japanese Society of Child Neurology

www.elsevier.com/locate/braindev

Original article

The assessment of sniff nasal inspiratory pressure in patients with Duchenne muscular dystrophy in China

Shu Zhang [a,b], Qian-qian Mei [b], Jing Xin [b], Hong-ying Zhang [b], Shi-wen Wu [b,*], Chun-feng Liu [a,*]

[a] *Department of Neurology and Suzhou Clinical Research Center of Neurological Disease, The Second Affiliated Hospital of Soochow University, China*
[b] *Department of Neurology, General Hospital of Chinese People's Armed Police force, China*

Received 6 December 2017; received in revised form 23 January 2018; accepted 27 January 2018

Abstract

Objective: Progressive weakness of respiratory muscles remains one of the leading causes of death among patients with Duchenne muscular dystrophy (DMD). Currently, there are few pulmonary function data among Chinese DMD patients. This study was carried out to evaluate the sniff nasal inspiratory pressure (SNIP) change among a group of Chinese DMD patients, and compare it with the SNIP value of patients with neuromuscular disorders in other countries.

Methods: SNIP data were collected in three research groups that consists of 581 subjects: 125 DMD boys who have taken steroid (Age 5.0–13.3, DMD-steroid group), 145 DMD steroid-naive boys (Age 5.0–13.9, DMD-nonsteroid group), and 311 healthy controls (Age 5.0–14.0, Control group).

Results: The SNIP for DMD-nonsteroid group, DMD-steroid group and Control group were: 56.5 (±14.3) cm H_2O,66.4 (±15.5) cm H_2O and 78.9 (±21.5) respectively. The SNIP in the DMD-nonsteroid group became significantly different from that of the healthy controls since age 7.0–8.9. The significant difference of SNIP between DMD-steroid group and DMD-nonsteroid group at age 7.0–10.9. The peak value of SNIP in the DMD-nonsteroid group appeared at age 8.7, and decreased dramatically thereafter, while in DMD-steroid group and the Control group peaked at 10.2 years and 12.2 years respectively. There was a bit difference between SNIP in this group and that in previous researches which may be due to geographical distribution and ethnic backgrounds.

Conclusion: This study strengthens the previous findings that SNIP can be used to evaluate respiratory dysfunction during the early stage of young patients with neuromuscular disorders, and demonstrates that steroid is effective in slowing the decrease of SNIP in this group of Chinese DMD boys.

Keywords: Duchenne muscular dystrophy (DMD); Sniff nasal inspiratory pressure (SNIP); Respiratory dysfunction

Abbreviations: DMD, Duchenne muscular dystrophy; FVC, forced vital capacity; MIP, maximal inspiratory pressure; SNIP, Sniff nasal inspiratory pressure

* Corresponding authors at: Department of Neurology, General Hospital of Chinese People's Armed Police Force, 69 Yongding Road, Haidian District, Beijing, China (S.-W. Wu). Department of Neurology and Suzhou Clinical Research Center of Neurological Disease, The Second Affiliated Hospital of Soochow University, 1055 Sanxiang Road, Suzhou, China (C.-F. Liu).

E-mail addresses: wu_shiwen@yahoo.com (S.-w. Wu), liuchunfeng@suda.edu.cn (C.-f. Liu).

https://doi.org/10.1016/j.braindev.2018.01.008

Please cite this article in press as: Zhang S et al. The assessment of sniff nasal inspiratory pressure in patients with Duchenne muscular dystrophy in China. Brain Dev (2018), https://doi.org/10.1016/j.braindev.2018.01.008

1. Introduction

Duchenne muscular dystrophy (DMD), caused by deficiency in dystrophin, is one of the most common neuromuscular diseases in children. It is an X-linked recessive disorder affecting one in 3500 live male births, which causes muscle weakness leading to wheelchair life in childhood and early death. Progressive respiratory muscle weakness is a major cause of death in DMD patients [1]. The two classical indicators, forced vital capacity (FVC) and maximal inspiratory pressure (MIP), are often used to evaluate the pulmonary function for adults. However, respiratory dysfunction is difficult to evaluate in young children [2,3]. Pulmonary function test using FVC is often used for evaluating respiratory function [4,5], but it is not sensitive in the early stage and can be affected by daytime hypercapnia in the advanced stage of disease [6]. MIP is the most widely used classical method to assess inspiratory muscle strength [7]. However, it requires mouthpiece tubing, which easily causes air leakage during measurement [8], and MIP is difficult to be carried out in young children, especially in those with cognitive dysfunction [1,9].

Sniff nasal inspiratory pressure (SNIP), an approach used to diagnose respiratory dysfunction in neuromuscular disorder subjects, can be easily performed by children older than 5 years old [10–13]. In 2012, it was reported that SNIP is often impaired early in patients with neuromuscular diseases [10]. Véronique Nève et al. also found that SNIP decreased earlier than vital capacity and peak expiratory flow in subjects with DMD [11,12]. For patients with amyotrophic lateral sclerosis, SNIP value less than 40 cm H_2O was significantly associated with higher occurrence of nocturnal hypoxia and lower survival rate [14,15]. Many countries have established their own reference SNIP value for children at different age [16–19]. Currently, there is no published study that focused on the assessment of SNIP in Chinese DMD children. In the present study, we investigated the decline of SNIP measurements in a group of Chinese DMD subjects by comparing them with normal controls. We also evaluated the effects of steroid treatment on SNIP values in DMD by comparing those received steroid treatment with those did not.

2. Materials and methods

2.1. Clinical data

Two groups of DMD subjects were recruited from our multidisciplinary DMD outpatient clinic in General Hospital of the Chinese People's Armed Police Force from October 2014 to October 2017. Of those, one hundred and twenty-five DMD subjects who took prednisolone 0.75 mg/kg/d for more than three months (ages 5.0–13.9 years) were designated as DMD-steroid group; and 145 DMD steroid-naive subjects (ages 5.0–13.6 years) were designated as DMD-nonsteroid group. All subjects were diagnosed with DMD and confirmed by genetic testing and/or muscle biopsy showing dystrophin deficiency. We included 311 healthy boys (ages 5.0–14.0) as a Control group. The exclusion criteria were: (a) upper respiratory tract infection or pneumonia within 2 weeks; (b) adenoid hypertrophy determined by X-ray and/or nasopharyngeal endoscopy; (c) asthma, chronic pulmonary disease or chronic rhinitis; (d) malformation of the nose or nasal septum; (e) taking medication affecting breathing and/or respiratory muscles during the study; and (f) follow-up not possible. Informed consents were obtained from parents of the subjects before study inclusion. Ethics Committee approval was obtained by the Medical Ethics Committee of General Hospital of the Chinese People's Armed Police Force. Body height (arm span was taken as the body height in subjects who could not stand or had severe scoliosis), weight, and other demographic and clinical data were recorded.

2.2. Methods

SNIPsit was assessed using a MICRO RPM respiratory pressure meter (Micro Medical Ltd., Kent, England) as described below. With the subject in sitting position, the probe was placed into one nostril while his mouth and contra-lateral nostril were closed. The subject was instructed to inhale forcefully after normal exhalation and maintained for three seconds. These tests were performed successfully at least three times in each subject, and the maximum value was recorded.

2.3. Statistical analysis

For the cross-sectional comparisons, data are presented as mean ± standard deviation and range values, and were compared by one-way analysis of variance (ANOVA) with SPSS 23.0 software. If the ANOVA was significantly affected, further comparisons were made using the Least Significant Difference test. We also used Student's *t*-test to test the difference in SNIP across groups by SPSS 23.0 software, and GraphPad Prism 5.0 was used to show the change trend of SNIP with age. A significance of $P < .05$ was considered statistically significant.

2.4. Results

In this study, SNIP was measured in all 581 children (Table 1) assigned to the 3 research groups. The average SNIP observed in DMD-nonsteroid group, DMD-steroid group and Control group were 56.5 (±14.3) cm H_2O, 66.4 (±15.5) cm H_2O and 78.9 (±21.5) respectively. There were significant differences among these

Please cite this article in press as: Zhang S et al. The assessment of sniff nasal inspiratory pressure in patients with Duchenne muscular dystrophy in China. Brain Dev (2018), https://doi.org/10.1016/j.braindev.2018.01.008