심폐재활
발표일시 및 장소 : **10 월 26 일(금) 13:39-13:51 Room E(5F)**

## OP- Scientific 1-3

# Significance of regular follow-up before noninvasive ventilation in Duchenne muscular dystrophy

Jang Woo Lee[1*], Won Ah Choi[2,3], Eun Young Kim[2,3], Yewon Lee[2,3], Jun Min Cha[1], Seong-Woong Kang[2,3†]

National Health Insurance Service Ilsan Hospital, Department of Physical Medicine and Rehabilitation[1], Gangnam Severance Hospital, Yonsei University College of Medicine, Department of Rehabilitation Medicine and Rehabilitation Institute of Neuromuscular Disease[2], Gangnam Severance Hospital, Pulmonary Rehabilitation Center[3]

**Objective**
Duchenne muscular dystrophy (DMD), an X-liked recessive and progressive muscular disorder, is the most common type of genetic muscular dystrophy. The development of mechanical ventilation and medical management of dilated cardiomyopathy have prolonged their lives. For determining the necessity of initiating noninvasive ventilation (NIV), various factors such as hypoventilatory symptoms, forced vital capacity (FVC), maximal inspiratory pressure (MIP), and end-tidal carbon dioxide (EtCO2), are considered. We aim to reveal the clinical importance of regular follow-up before initiation of NIV by comparing patients with and without regular medical surveillance in DMD.

**Materials and Methods**
This retrospective research analyzed successful applications of NIV in the pulmonary rehabilitation center in a single tertiary university hospital from March 2000 to April 2018. The patients were categorized into three groups according to follow-up status before NIV initiation. Group 1 patients had been followed regularly in the outpatient clinic before the initiation of NIV. The patients in group 2 were lost during follow-up or were never followed before NIV. And in group 3 patients were transferred from another hospital or another department with inadequate ventilatory support. We analyzed various parameters such as partial pressure of CO2 (PCO2) in arterial blood gas analysis (ABGA) and nocturnal non-invasive continuous monitoring, ventilatory support time for each day after completion of NIV setting, and various respiratory function including FVC, maximum insufflation capacity (MIC), peak cough flow (PCF), assisted peak cough flow (APCF), MIP and maximal expiratory pressure (MEP).

**Results**
Total 239 cases of DMD patients with NIV were included in this study. The mean age of the patients was 19.7 ± 4.6 years (Table 1). The number of the patients in group 1, 2, and 3 are119, 82, and 38 respectively. All parameters except for the duration of ventilator use, differed significantly. Age was lowest at group 1. Initial pCO2 of ABGA was highest in

group 2. Nocturnal maximum CO2 and mean CO2 were highest in group 2 and lowest in group 3. This trend was due to the inclusion of some patients undergoing NIV in group 3. All respiratory functions were highest in group 1 and lowest in group 3. Although age did not differ between group 1 and 3, respiratory function decreased remarkably in group 3 compared with group 1 and group 2, indicating that appropriate NIV setting exert a positive effect on preservation of respiratory function.

**Conclusion**

Regular follow-up before the onset of ventilatory failure is crucial for timely application of NIV. Because NIV itself does not guarantee correction of abnormal ventilation, accurate correction of hypoventilation should be confirmed using monitoring system. Appropriate NIV exert preventive effect on the deterioration of respiratory function.

Table 1. Demographic characteristics of the patients

| | **Total (N = 239)** |
|---|---|
| Age | 19.7 ± 4.6 |
| ABGA pH | 7.395 ± 0.046 |
| ABGA $PCO_2$ (mmHg) | 44.6 ± 12.6 |
| Initial maximum $CO_2$* | 52.5 ± 8.5 |
| Initial mean $CO_2$* | 45.4 ± 7.5 |
| Final maximum $CO_2$*† | 39.9 ± 5.7 |
| Final mean $CO_2$*† | 33.0 ± 5.3 |
| FVCsit (%)‡ | 22.6 ± 16.2 |
| FVCsup (%)‡ | 21.4 ± 15.2 |
| MIC (mL) | 1391 ± 579 |
| PCF (L/min) | 132 ± 80 |
| APCF (L/min) | 260 ± 91 |
| MIPsit (%)‡ | 22.3 ± 15.9 |
| MIPsup (%)‡ | 22.0 ± 14.9 |
| MEPsit (%)‡ | 14.9 ± 9.3 |
| MEPsup (%)‡ | 14.8 ± 9.1 |

ABGA: arterial blood gas analysis, $PCO_2$: partial pressure of carbon dioxide, FVC: forced vital capacity, MIC: maximum insufflation capacity, PCF: peak cough flow, APCF: assisted peak cough flow, MIP: maximal inspiratory pressure, MEP: maximal expiratory pressure, sit: at sitting position, sup: at supine position

* In the nocturnal noninvasive and continuous $CO_2$ monitoring

† After completion of noninvasive ventilator setting

‡ These values calculated to percentage of normal predictive values.

Table 2. Comparison of pulmonary function among the groups

| | Group 1 | Group 2 | Group 3 | Group 1 vs 2 | Group 2 vs 3 | Group 1 vs 3 | Among groups |
|---|---|---|---|---|---|---|---|
| Age (years) | 18.2 ± 3.8 | 21.7 ± 5.1 | 19.9 ± 5.1 | <0.001 | 0.206 | 0.066 | <0.001 |
| ABGA pH | 7.401 ± 0.341 | 7.370 ± 0.054 | 7.423 ± 0.054 | 0.078 | 0.048 | 0.719 | 0.004 |
| ABGA PCO2 (mmHg) | 41.6 ± 10.3 | 49.5 ± 13.9 | 43.4 ± 13.5 | <0.001 | 0.089 | >0.999 | <0.001 |
| Initial maximum CO2 (mmHg)* | 51.3 ± 6.2 | 55.7 ± 9.9 | 49.6 ± 9.7 | 0.002 | 0.007 | 0.978 | <0.001 |
| Initial mean CO2 (mmHg)* | 44.8 ± 4.7 | 48.9 ± 9.3 | 40.6 ± 10.7 | 0.040 | 0.016 | 0.406 | <0.001 |
| Ventilator time (hours/day) | 8.8 ± 2.4 | 9.8 ± 3.7 | 9.2 ± 4.4 | 0.137 | >0.999 | >0.999 | 0.138 |
| FVCsit (%)† | 27.8 ± 17.7 | 17.8 ± 11.7 | 16.0 ± 14.1 | <0.001 | >0.999 | 0.002 | <0.001 |
| FVCsup (%)† | 26.5 ± 16.8 | 16.9 ± 11.6 | 14.7 ± 10.5 | <0.001 | >0.999 | <0.001 | <0.001 |
| MIC (mL) | 1565 ± 598 | 1280 ± 511 | 1050 ± 448 | 0.004 | 0.088 | <0.001 | <0.001 |
| PCF (L/min) | 161 ± 73 | 110 ± 78 | 83 ± 73 | <0.001 | 0.291 | <0.001 | <0.001 |
| APCF (L/min) | 283 ± 86 | 237 ± 84 | 227 ± 105 | 0.003 | >0.999 | 0.012 | 0.001 |
| MIPsit (%)† | 28.6 ± 17.7 | 16.7 ± 10.9 | 13.1 ± 8.1 | <0.001 | 0.339 | <0.001 | <0.001 |
| MIPsup (%)† | 28.2 ± 16.0 | 16.6 ± 11.1 | 13.2 ± 7.8 | <0.001 | 0.394 | <0.001 | <0.001 |
| MEPsit (%)† | 18.0 ± 9.5 | 12.0 ± 8.3 | 10.8 ± 7.0 | <0.001 | >0.999 | <0.001 | <0.001 |
| MEPsup (%)† | 17.6 ± 8.9 | 12.6 ± 9.4 | 10.5 ± 6.4 | 0.002 | 0.765 | <0.001 | <0.001 |

ABGA: arterial blood gas analysis, $PCO_2$: partial pressure of carbon dioxide, FVC: forced vital capacity, MIC: maximum insufflation capacity, PCF: peak cough flow, APCF: assisted peak cough flow, MIP: maximal inspiratory pressure, MEP: maximal expiratory pressure, sit: at sitting position, sup: at supine position

* In the nocturnal noninvasive and continuous $CO_2$ monitoring

† These values calculated to percentage of normal predictive values.