

# Assessment of pulmonary function in children with Duchenne Muscular Dystrophy

H Levine<sup>1,2</sup>, I Goldfarb<sup>2,3</sup>, H Musaffi<sup>1,2</sup>, D Prais<sup>1,2</sup>, S Aharoni<sup>2,3</sup> and Y Nevo<sup>1,4</sup>

<sup>1</sup>Pulmonary Institute, Schneider Children's Medical Center (SCMCI), Petach-Tikva, Israel,

<sup>2</sup>Sackler Faculty of Medicine, Tel Aviv University, Ramat Aviv, Tel Aviv, Israel

<sup>3</sup>Neurology Institute, Schneider Children's Medical Center, Petah-Tikva, Israel

<sup>4</sup>Hebrew University Hadassah Medical School, Jerusalem, Israel

## Introduction

Duchenne Muscular Dystrophy (DMD) is the most common neuromuscular disorder of childhood. Lung disease contributes significantly to morbidity and mortality. Forced Vital Capacity (FVC) is widely used to assess the progression of restrictive lung disease. Additional techniques, maximal inspiratory and expiratory pressures (MIP and MEP respectively) and peak cough flow (PCF), are used for follow up DMD patients. PCF is advocated for treatment adjustment in international guidelines, however, there is lack of standardization for children.

## Aims

To assess the changing and differences of all pulmonary function testing for respiratory disease severity and their correlation with age, ambulation and steroid treatment in DMD patients.

## Method and Subjects

A retrospective analysis of spirometry, lung volumes, respiratory pressures tests from DMD patients enrolled in a natural history cohort study. Data were analyzed for change with age, genetic mutations, ambulation status, and glucocorticoid treatment.

## Results

Out of 116 muscular dystrophy patients, 79 with DMD were enrolled (median age 13 years, range 4–27). Among them 44 (56%) completed successfully respiratory function tests. The %FVC, %TLC and %MEP predicted for age, but not PCF, showed a significant decrease with age, for 4% a year, and loss ambulation, with a better measurements in ambulatory patients treated with steroids. PCF values were randomly distributed without any difference or correlation with age and steroid treatments. Deflazacort treatments showed no significant added value on prednisone.

## Conclusion

FVC and TLC, are valuable measurements for respiratory follow up in patients with DMD, correlating with age, ambulation and steroid treatment, whereas PCF was unreliable to follow as it did not differ between groups and had no correlation to age or steroid treatment. The benefit of steroid treatment continues even in non-ambulatory patients.

Figure 1 (A-D): Correlation of pulmonary functional tests with age in ambulatory DMD (ADMD) and non-ambulatory DMD patients (NADMD); %FVC (A), %TLC (B), %RV (C), %RV/TLC (D).

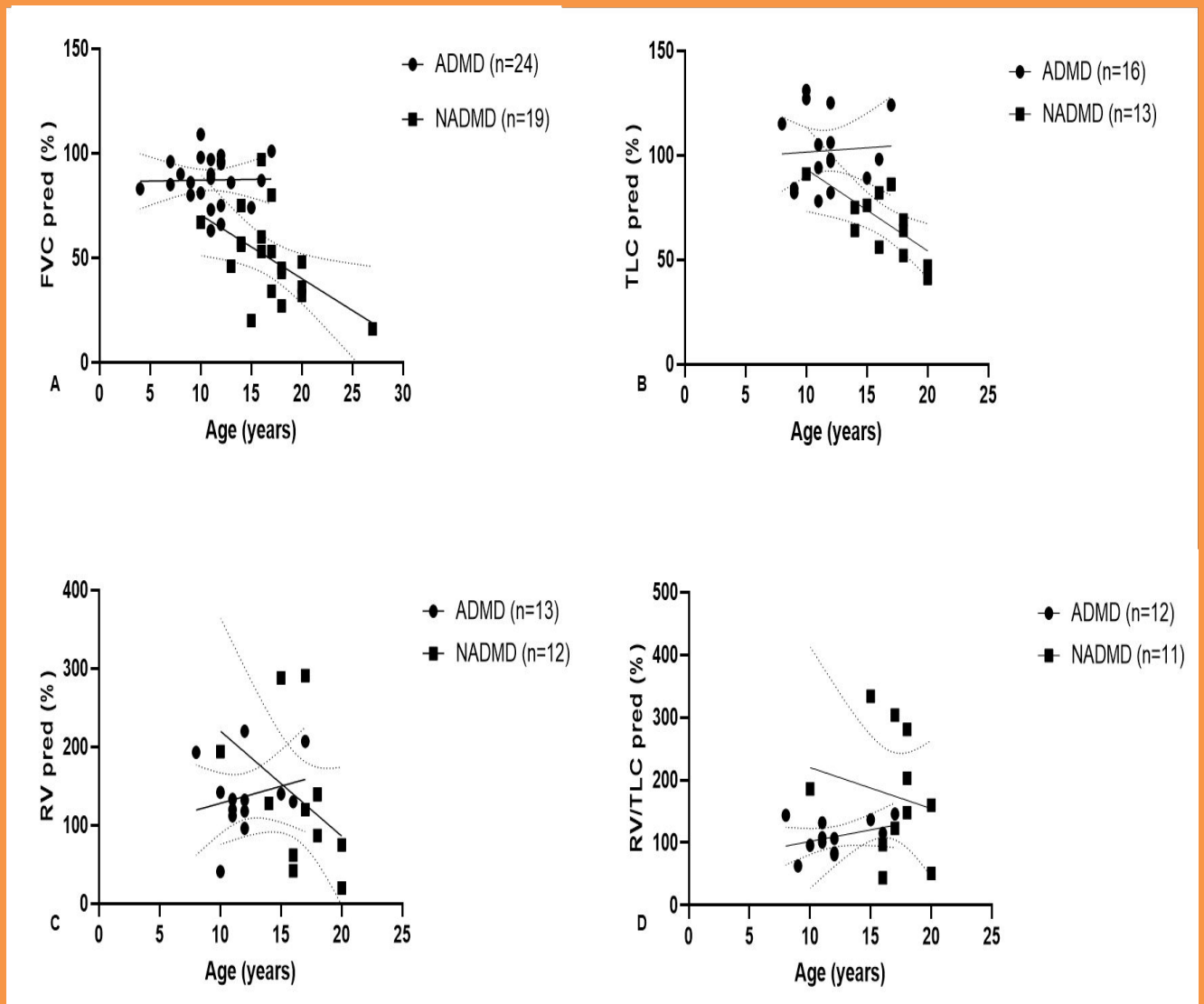


Figure 2 (A-C): Correlation of pulmonary function tests with age in ambulatory and non-ambulatory DMD patients; %MEP (A), %MIP (B), PCF (C) by age (Years) in DMD patients.

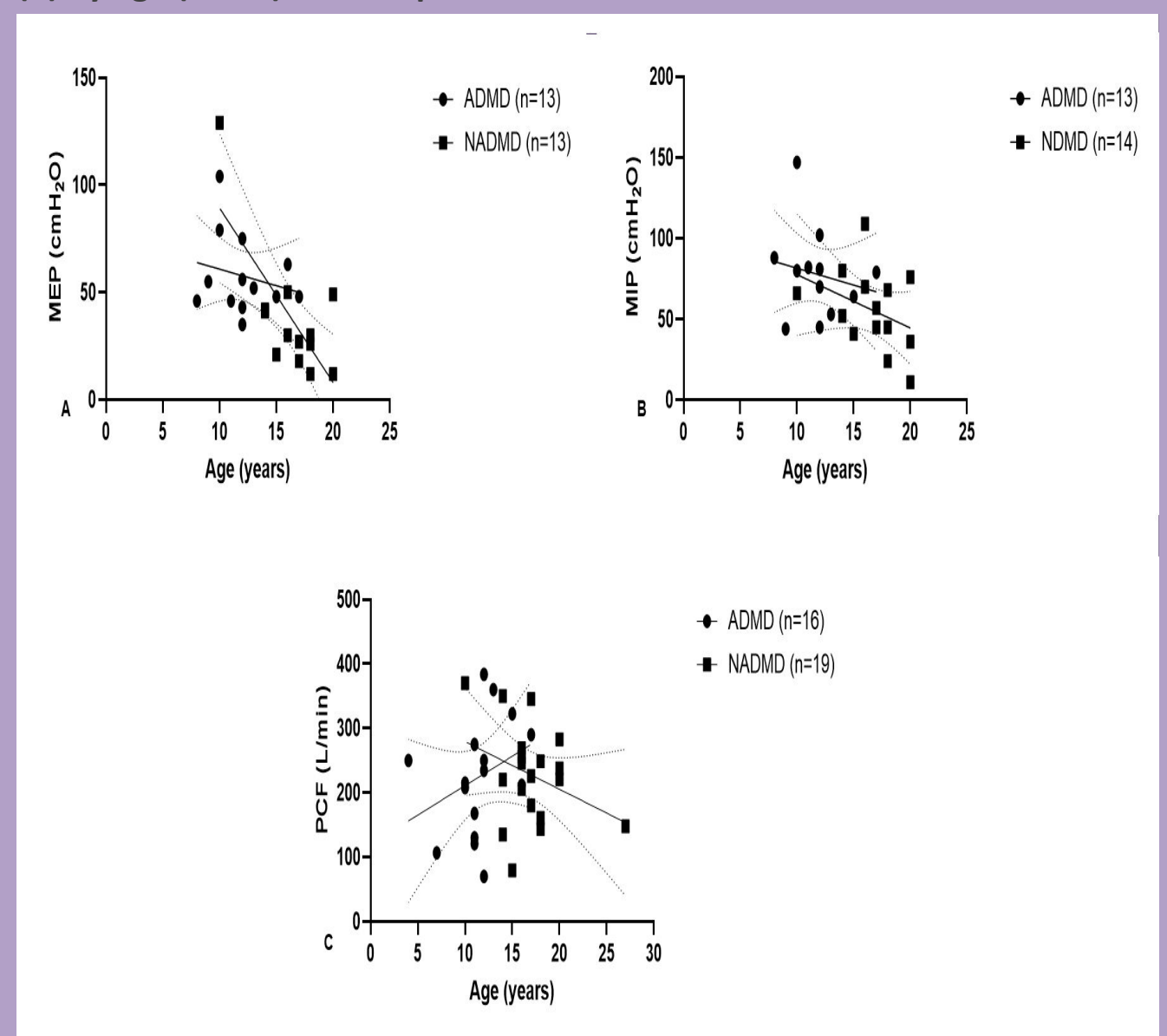


Figure 3 (A-G): Pulmonary Functional Tests in treated and untreated DMD patients; %FVC (A), %TLC (B), %RV (C), %RV/TLC (D), %MEP (E), %MIP (F) and PCF (G).

