Format: Abstract

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Peak Cough Flow in Children with Neuromuscular Disorders.

Kotwal N[1,2], Shukla PJ[3], Perez GF[4,5].

Author information

Abstract

PURPOSE: Patients with neuromuscular disease (NMD) experience weakened cough due to progressive respiratory muscle weakness. Peak cough flow (PCF) measurements derived from adult populations are used to recommend initiation of assisted cough therapies. The objective of this study was to characterize PCF values among pediatric patients with NMD.

METHODS: Retrospective chart review was performed for patients seen in the multidisciplinary pediatric muscular dystrophy clinic from 2010 to 2016. Clinical and demographic variables included age, gender, ambulation status, and PCF measurements.

RESULTS: 366 patients with an established diagnosis of NMD (median age 11.8 years) were included in this study. 102 (27.8%) out of the 366 patients were affected by Duchenne muscular dystrophy (DMD), 42 (11.5%) by congenital muscular dystrophy (CMD), 42 (11.5%) by Charcot Marie Tooth disease (CMT) and 24 (6.5%) by Becker's muscular dystrophy (BMD). The mean PCF values in DMD (255.8 L/min) and CMD (249.1 L/min) were lower than CMT (321.5 L/min) with p-values of 0.007 and 0.02, respectively. The mean PCF of BMD (333.3 L/min) was higher than that of DMD and CMD but the difference was not statistically significant. PCFs were not statistically different between ambulatory and non-ambulatory status (263.0 L/min versus 290.8 L/min, $p = 0.12$). Children under 10 years of age had lower PCF relative to older subjects (179.5 L/min versus 300.9 L/min, $p < 0.0001$).

CONCLUSION: Baseline PCF values in young children are below the adult-specific values suggested for starting assisted cough techniques. Further longitudinal trials are required to derive pediatric-specific reference values for PCF in patients with NMD.

KEYWORDS: Becker muscular dystrophy; Charcot Marie Tooth disease; Congenital muscular dystrophy; Duchenne muscular dystrophy; Neuromuscular disease; Peak cough flow

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