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Noninvasive Respiratory Care Received by Individuals With Duchenne Muscular Dystrophy Since 1979—Reply

In reply:

We commend Dr Bach and his colleagues for advancing and optimizing noninvasive respiratory care for individuals with neuromuscular disease since the late 1970s. However, we believe he may have misinterpreted the aim and purpose of our study results,¹ and we would like to clarify. In recent years, emphasis on the use of noninvasive respiratory aids has only increased, and this emphasis has been endorsed in numerous consensus statements on Duchenne muscular dystrophy (DMD) respiratory care. Despite the emphasis on noninvasive respiratory management in the published guidelines, we found significant variability in monitoring for respiratory muscle weakness and in the use of noninvasive respiratory care devices among the neuromuscular centers in our study.¹ Furthermore, there is a paucity of literature on adherence to these published guidelines and very little information on reasons why providers may deviate from the recommended care. Our data suggest that the guidelines for respiratory care have not been universally embraced, at least in the centers that we examined. This “care gap” might be due to potentially correctable barriers (eg, the need for further clinician education, a shortage of therapists trained in noninvasive aids, or the need for improved insurance coverage for the necessary equipment). Alternatively, the gap might represent a decision by providers to deviate from the recommendations (eg, due to local expertise and preference for ventilation via tracheostomy).

Our study reported descriptive data on information collected retrospectively for a surveillance network from medical records. Our sole intention was to compare data available with the care management guidelines published and determine whether any

trends or changes could be identified. The aim of our paper was to report on DMD respiratory care management from medical record data available through 2011 for individuals cared for in select areas that are affiliated with MD STARnet and to compare those data with the published care management guidelines. We very specifically did not set any standards of care, attempt to criticize local practice, nor imply any perspectives on the use of tracheostomies in DMD, since this was not the purpose of our work. One idea that our article does suggest is that standards of care need to be widely enacted—and if they cannot be achieved locally they may not achieve their intent, which is to benefit large groups of patients.

We appreciate the opportunity to respond to the letter of Dr Bach et al regarding our manuscript. We hope that their letter and our response will spark a discussion on how to address barriers to guideline implementation. The best way to ventilate patients with DMD is a complex issue that must respect the voice and choice of patients. For additional perspectives from individuals with DMD using noninvasive and invasive ventilation, we encourage readers to review *Ventilation and Duchenne*² from DMD Pathfinders (<http://www.dmdpathfinders.org>, Accessed May 17, 2017), a user-led organization whose mission is to “promote choice and control, and quality of life for teenagers and adults with DMD.”

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