

Letter to the Editor

Fractures and bone health in Duchenne muscular dystrophy in Scotland



Response

We thank Professor Topaloglu for his comment on our retrospective study of radiologically confirmed fractures in all boys with Duchenne muscular dystrophy (DMD) managed in the Scottish Muscle Network [1]. Professor Topaloglu has three main queries which we will address.

1. There were 91 boys with DMD (18 years and younger) in the Scottish Muscle Network in December 2015 when the study was conducted, all of whom were included in our report [1]. All boys with DMD in Scotland are managed in seven paediatric centres within the Scottish Muscle Network. Responsible clinicians report new diagnosis of DMD to the Clinical Audit System of the Scottish Muscle Network. The network also sends 3–6 monthly reminders to all lead clinicians in the seven centres. In addition, we contacted all the clinicians in the seven centres at the inception of the study to ensure we did not miss any cases.
2. The study population is not a special sub-set of boys referred to paediatric endocrinology. We are confident that our study captured all boys managed in Scotland, and not just from one specialist neuromuscular centre.
3. The analysis of probability of fracture in relation to age and glucocorticoid duration was performed using Kaplan–Meier analysis. As pointed out by Professor Topaloglu, there are numerous factors associated with bone health and fractures in DMD, including vitamin D, bisphosphonate and testosterone. Only one boy received prophylactic treatment with bisphosphonate; whilst the majority of the others were following identification of fractures. Given the relatively small sample size of this retrospective study, we chose not to evaluate the relationship of clinical risk factors with fractures. Larger nationwide studies of fractures and factors associated with fractures in DMD are needed and underway.

Our study which reported radiologically confirmed fractures highlights the fact that fragility fractures are extremely common in DMD in this national cohort with complete case ascertainment. Our results also point to the inconsistency of bone health monitoring in this population, which mirrors a recent report from the Muscular Dystrophy Surveillance, Tracking, and Research Network (MD STARnet) database in the United States [2]

References

- [1] Joseph S, et al. Fractures and bone health monitoring in boys with Duchenne muscular dystrophy managed within the Scottish muscle network. *Neuromuscul Disord* 2019;29(1):59–66.
- [2] Weber DR, et al. Bone Health and Endocrine Care of Boys with Duchenne Muscular Dystrophy: Data from the MD STARnet. *J Neuromuscul Dis* 2018;5(4):497–507.

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