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Letters to the Editor

Fractures and bone health in Duchenne muscular dystrophy in Scotland

I have read with interest the paper by Joseph et al. on Duchenne muscular dystrophy, and bone health monitoring [1]. Their patient cohort has peculiar properties. The median loss of ambulation is 10.2 years of age. About half of them were nonambulant at the time of the study. Sixty five percent of them were on steroids with entirely different regimens (daily deflazacort, pulsed deflazacort, daily prednisolone, pulsed prednisolone) for a median of 5.2 years at a median age of 5.5 years. In 10%, the treatment mode is unknown. Twenty-five percent are steroid naive or received steroids in the past. From their total of 91 cases, 44 (48%) had sustained at least one symptomatic radiographically confirmed fracture. This constituted nonvertebral 40% and vertebral 8%. There were also multiple fractures in time.

As a clinician, I have some difficulty in interpreting this data. We all agree that close monitoring of bone health is a prerequisite in boys with DMD, either ambulant or not (reduced mobility, osteoporosis, delayed puberty, fragile bones, extra bleeding from bone during scoliosis surgery, etc.). I have a few questions for the authors:

1. How many DMD boys are registered to the Scottish muscle network? Are these 91 cases all they have in their database?
2. Could these DMD boys be from a special sub-set with a referral (e.g. to the endocrinologist), because they had other early onset findings in addition? We know that about 10–15% cases with DMD can be quite resistant to the sort of treatment or therapy provided, whether steroids or physical therapy or even newer therapies approaching. In part this could be explained by a few genetic polymorphisms, but not all. I have personally seen DMD boys who lost the ability to walk by 6 years of age (fortunately not that often). Regardless of age, if a DMD case is noted to be hypotonic during any physical examination, the chance of that child becoming non-ambulant within 9–12 months is pretty elevated.
3. From this rather heterogeneously treated (or not treated) group of boys supplemented individually with varying supports such as vitamin D, bisphosphonate, testosterone, how did the authors calculate the probability and statistics aiming to talk about future to come? In their words “the probability of developing non VF was 50% by 13.6 years old”.

Reference

[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.

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