H22 Differences in Skeletal Pathology as Seen in an Individual With Quadriplegia Secondary to Duchenne Muscular Dystrophy

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After attending this presentation, attendees will see the distinction between a human adult skeleton displaying the effects of Duchenne Muscular Dystrophy (DMD) with normal adult skeletons. This will aid anthropologists in differentiating between normal bone variation and bone pathology. Although the skeleton is similar in size to a subadult or young female skeleton, the systemic loss of bone mass and density of the postcranial skeleton will reveal that this is not normal human variation but a pathological condition.

This presentation will impact the forensic science community by bringing awareness of what effect Duchenne Muscular Dystrophy has on the skeleton and how this may be distinguished from other bone disorders such as osteoporosis or normal bone variation.

The Mechanostat Model describes the direct correlation between muscle use and bone strength and density. If the muscle is active, applying force to bone with a strain greater than 1500μ, the bone will respond by increasing in strength and mass. Bones of individuals who have sustained some level of paralysis, and thus strain less than 800μ, will have a reduction in strength and mass.

In this study, the body of a deceased 63-year-old White male was donated to the Southeast Texas Applied Forensic Science (STAFS) facility with quadriplegia secondary to DMD, was compared to the bones of four White males of similar age with no paralysis history. Comparisons included the examination of the gross morphology, three-dimensional morphometry, histomorphometry, and bone density analysis. Three-dimensional data was collected using the Microscribe® G2X and analysis was completed using Morpheus *et al.* software. Bone histomorphometry examined osteon size, osteon number, osteon area, and osteon density. Dual Energy X-Ray Absorptiometry (DEXA) was used to determine Bone Mineral Density (BMD). Postcranial measurements were also taken using conventional methods (i.e., sliding caliper, osteometric board).

Except for the cranium, the gross morphology of the DMD skeleton showed systemic reduction in bone mass and density with the overall size similar to that of a young adult or a female. The skeleton was gracile with small articulating surfaces (e.g., femoral and humeral head sizes).

Results for cranial morphometrics show that the majority of the mean measurements were statistically significant ($p<0.05$) between the DMD subject and the four normal subjects. With the exception of MDH, OBH, FOL, OCF, CDL, and MAN, all other cranial measurements were substantially larger for DMD than the other subjects.

All postcranial mean measurements between DMD and normal subjects showed statistical significance ($p< 0.05$) with substantially larger postcranial measurements on the four normal subjects.

Studies using osteon size, area, and density to estimate age show osteon area for individuals older than 50 years of age typically range between 12,868μm^2-37,762μm^2 (Crowder 2012). Osteon diameter of bones with disuse, osteoporosis, or age-related osteoporosis is larger and the density lower (Black 1974). The study subject (DMD) had fewer osteons (n=3), larger osteon diameter (317μm) and area (78,923μm^2), and a lower concentration of osteon population density (1.46/mm^2).

Previous research on White males from the United States show the average BMD is approximately 0.790g/cm^2 (Looker 1995). Average BMD T-scores for females in the United States ranges between -1 and 1. The Dual-Energy X-Ray Absorptiometry (DEXA) scan for the DMD male showed a BMD of 0.466 g/cm^2 and a T-score -4.2. The T-score of the DMD male is significantly lower than found in his ancestry and age range and significantly lower than the average female. Conversely, one of the normal male subject's BMD score was 1.088 g/cm^2 and T-score 2.1 showing a slightly higher density than the average.

Results of the Pearson's correlation (to determine if there was a relationship between cranial and postcranial measurement of DMD) showed no dependent relationship. This lack of correlation underscores the limitations of the Mechanostat Model in that it does not adequately explain the difference in bone biomechanics and remodeling between the skull (non-weight-bearing) and postcranial bones (weight-bearing).

Bone Pathology, Duchenne Muscular Dystrophy, Mechanostat Model

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