ANIMAL MODELS OF DUCHENNE MUSCULAR DYSTROPHY, WITH SPECIAL REFERENCE TO THE *MDX* MOUSE

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Abstract:

Duchenne muscular dystrophy (DMD) is a progressive muscle wasting disease that affects approximately 1 in 3500 male births. We describe animal models of DMD with special reference to the *mdx* mouse. We also describe some of the standard operating procedures (SOPs) developed by the TREAT-NMD neuromuscular network (http://www.treat-nmd. eu/) for assessment of the *mdx* mouse, with a focus on techniques for assessing cardiac function that are used in our lab, including the cardiac conductance catheter. We have also recently developed cardiac MRI as a novel cardiac assessment technique for mouse models of muscular dystrophy. We describe how this technique can be used both in the assessment of ventricular function and in the investigation of the role of abnormal calcium influx in muscular dystrophy-associated cardiomyopathy.

Keywords: duchenne muscular dystrophy, *mdx* mouse, cardiomyopathy, animal models, MRI, cardiac catheter, standard operating procedures