Duchenne Muscular dystrophy (DMD) is a lethal X-linked genetic disease, characterized by absence of dystrophin (Dp427), a protein important for muscle integrity. The disease manifests with progressive muscle wasting in male children, although DMD patients are also afflicted by several neurological disorders, i.e. epilepsy, autism and mental retardation, among others. However, these disorders have never been considered relevant, mainly because of premature patients' death. To date, as new therapies prospect an increase in DMD patient life of about ten years, neurological disorders will paradoxically worsen the quality of life.

One major pharmacological treatment of DMD patients is administration of high doses of corticosteroids. However, besides the fact that prolonged exposures to GCs has several side effects, the signaling that these molecules elicit in the brain and in other tissues of dystrophic subjects, already pathologically compromised, has never been investigated. This research project is aimed at exploring the effects that increased levels of GCs exert in brain limbic regions, (i.e. hippocampus, amygdala and medial prefrontal cortex), in DMD mouse models. The results obtained demonstrate that in neurons of mdx mouse both genomic and intracellular signaling-mediated responses were affected compared to the wild type showing a characteristic response to a chronic exposure to glucocorticoids. These results supported us to continue to characterize the effects of glucocorticoids in brain under conditions of chronic administration. It will be interesting to investigate the response to chronic administration of glucocorticoids in other mouse tissues and in human and mouse cell lines.

References:
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