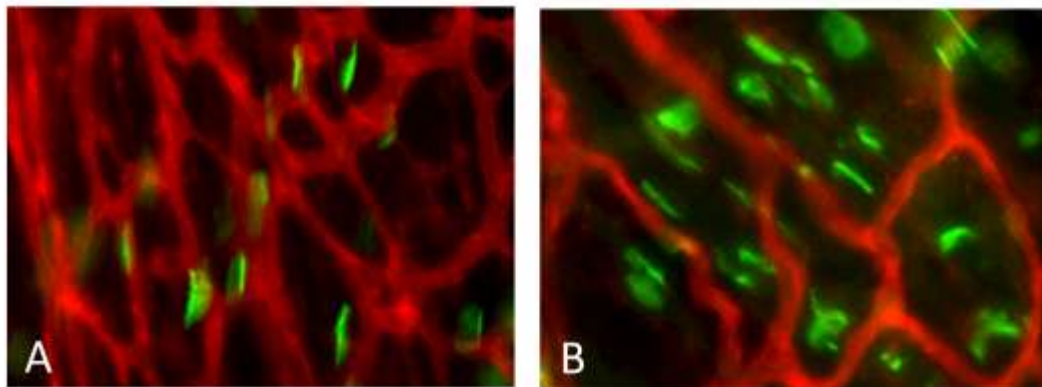


Is defective spinal cord and peripheral nerve vasculature a contributory factor to pathology in Spinal Muscular Atrophy?

Spinal Muscular Atrophy is a predominately childhood form of motor neurone disease which affects 1:6,000 live births Worldwide, and as such is one of the commonest genetic causes of infant mortality. The disease appears to target motor neurones which supply skeletal muscles, and when these become damaged the patient loses control of functions such as walking and eventually the muscles involved in breathing are affected with fatal consequences.

Much work has focussed on rescuing motor neurones to affect a cure for the disease. However, it has recently become increasingly apparent that many other cells and tissues within the body are also affected, and that these become significant when life is prolonged by medical intervention. We are particularly interested in defects in the circulatory system and have shown significant defects in the blood supply to skeletal muscles present at early stages of the disease.



The image shows blood vessels stained red in skeletal muscle, with muscle endplates indicated in green. A, shows healthy muscle, while B shows severe SMA muscle. The dramatic capillary bed defect is immediately apparent.

This project will build on our work in skeletal muscle to examine the blood supply to peripheral nerve and spinal cord, to determine if these two regions where motor neurone cell bodies and axons are present are also affected. This work will lead us toward future combinatorial therapies for this terrible disease.

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