EXTRACELLULAR MATRIX AND MITOCHONDRIA OF THE SKELETAL MUSCLE

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BACKGROUND In normal skeletal muscle a portion of the extracellular matrix (ECM), the perimysium, and the subsarcolemmal accumulations of mitochondria in the myofibres are co localized at numerous points of contact between the perimysium and the sarcolemma of myofibres (the points called Perimysial Junctional Plates) [1]. Using different myopathic muscles we decided to track the reasons of the co localisation.

METHODS Muscles observed were: (i) the EDL from tenascin null (Tnx) mice, (ii) muscle biopsies from Ulrich (collagen VI deficient) human disease, (iii) the EDL from Mdx 8 week old mice. The two first bearing defaults of their ECM the third of their sarcolemma and myofibres cytoskeleton. Scanning electron microscopy was used to observe density and spatial distribution of the perimysium into Tnx and Mdx muscles only. Transmission electron microscopy was used to observe the fine structure of the perimysium and the morphology of structures at the level of PJPs into Tnx, Mdx and Ulrich's muscles.

RESULTS Scanning electron microscopy has shown that the perimysium is over expressed and is modified in its spatial distribution into Tnx and Mdx muscles. Transmission electron microscopy has shown that the perimysium of the three muscles contains a mixture of modified and intact type I collagen fibres. When modified, they reach myofibres with any associated mitochondria or some apoptotic ones. In contrast, intact collagen type I fibres are mixed with fine reticular collagen material when they reach myofibres at points where the cytoplasm always contains subsarcolemmal accumulations of mitochondria. However, muscles differ in the area of their contacts between intact types I collagen fibres and the area of co localised mitochondria: (i) Tnx muscles have homogenous large regions of contacts between collagen and their myofibres and homogenous large corresponding accumulations of subsarcolemmal mitochondria. This leading to an extremely abundant mitochondrial content into some of their myofibres; (ii) Centronucleated myofibres in Mdx muscles have large but non homogenous regions of contacts and large but non homogenous subsarcolemmal mitochondria. In such myofibres, contacts and subsarcolemmal mitochondria are co localised at points of large sarcolemmal densifications spreading out along their sarcolemma; (iii) Ulrich's muscles have only some small contacts and a few corresponding mitochondria.

CONCLUSIONS Amount of perimysium and its associated reticular network controls the amount of subsarcolemmal mitochondria into the myofibres. Collagen VI is one of the components of the reticular network which is requested for control [2]. The defect in the sarcolemma of Tnx myofibres and the corresponding defect in subsarcolemmal mitochondria indicate also that correct sarcolemma is needed.

REFERENCES

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