

## **SKELETAL MUSCLE DISORDERS**

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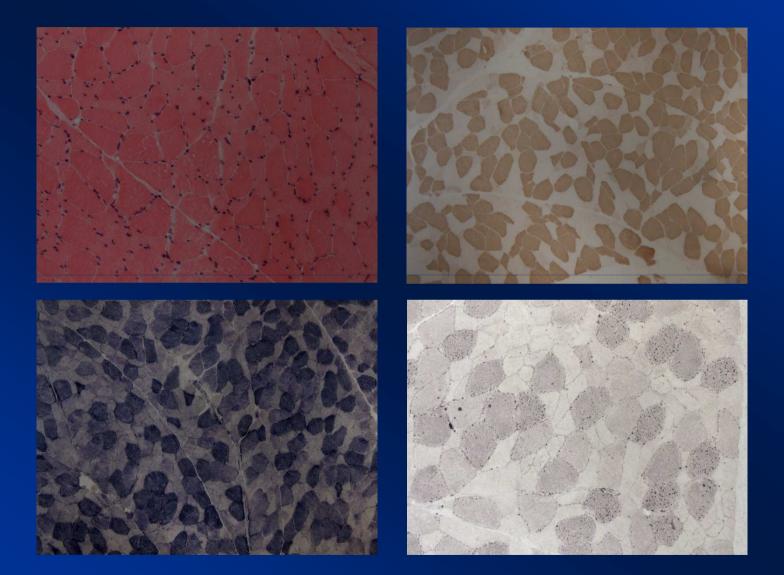
### **Topics of scientific research**

- Idiopathic inflammatory myopathies
- Sporadic inclusion body myositis (s-IBM)
- Myofibrillar myopathies
- Brody disease and Brody syndrome
- Mitochondrial diseases

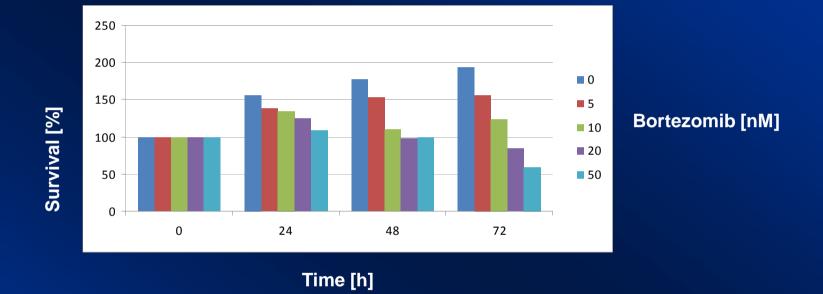
#### **Current research interests**

- Identification of muscle specific antigens in patients with dermatomyositis, polymyositis and paraneoplastic myositis
- Involvement of RNAPII-Associated Proteins (RPAPs) in muscle diseases
- Characterization of protein aggregates in myofibrillar myopathies using LCM (Laser Capture Microdissection)
- Zebrafish as animal models of myofibrillar myopathies
- The pathogenetic mechanisms of Brody Disease and Brody Syndrome
- •The fate of ragged-red fibers and the mechanisms of mitochondrial proliferation in mitochondrial diseases

# **Bortezomib and muscle toxicity**



### Primary cultures of normal human muscle



Three culture sets which were established from satellite cells derived from a portion of diagnostic muscle biopsy of three different patients who were considered free of muscle diseases

## **Future directions**

Generation and characterization of a reliable animal model (mice, rat) of bortezomib myotoxicity

Understanding the mechanism of action in skeletal muscle