

# Pediatric Specialists *of Virginia*



## Understanding Tests in Juvenile Myositis

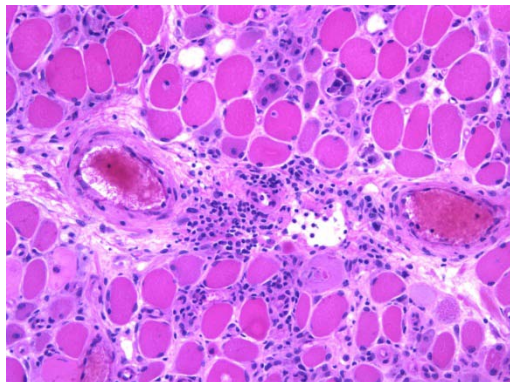
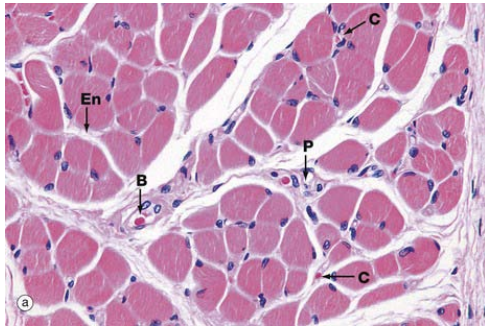
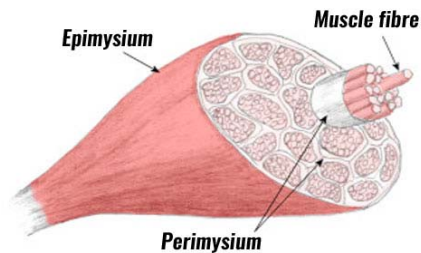


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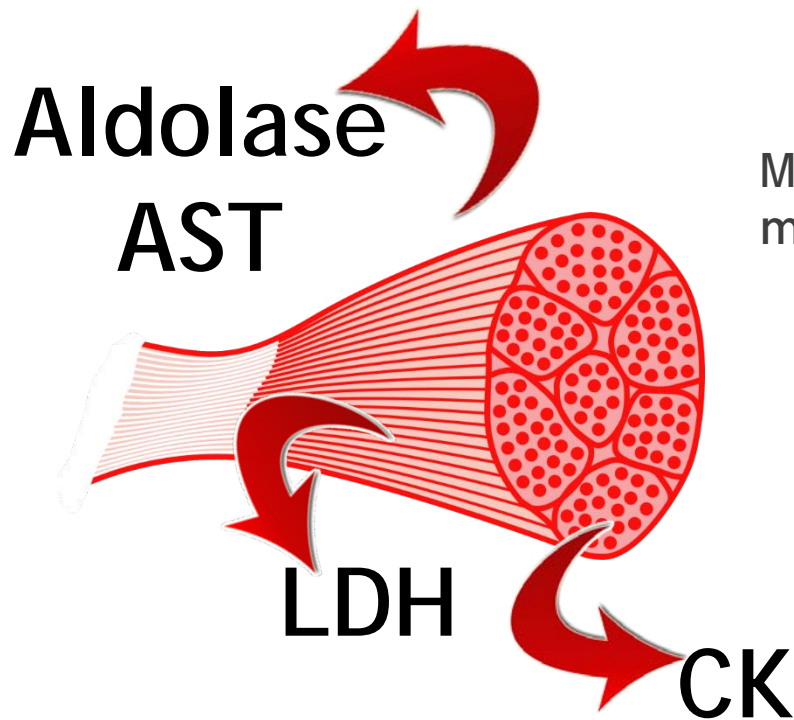
Inova Children's Hospital

# Anatomy of Normal vs Inflamed Muscles



- Normal skeletal muscles:
  - Polygonal
  - similar in size
  - Fit together like a puzzle
- Inflamed skeletal muscles:
  - Round, irregular
  - areas of muscle atrophy
  - Lots of surrounding inflammatory cells surrounding muscles and cuffing around blood vessels

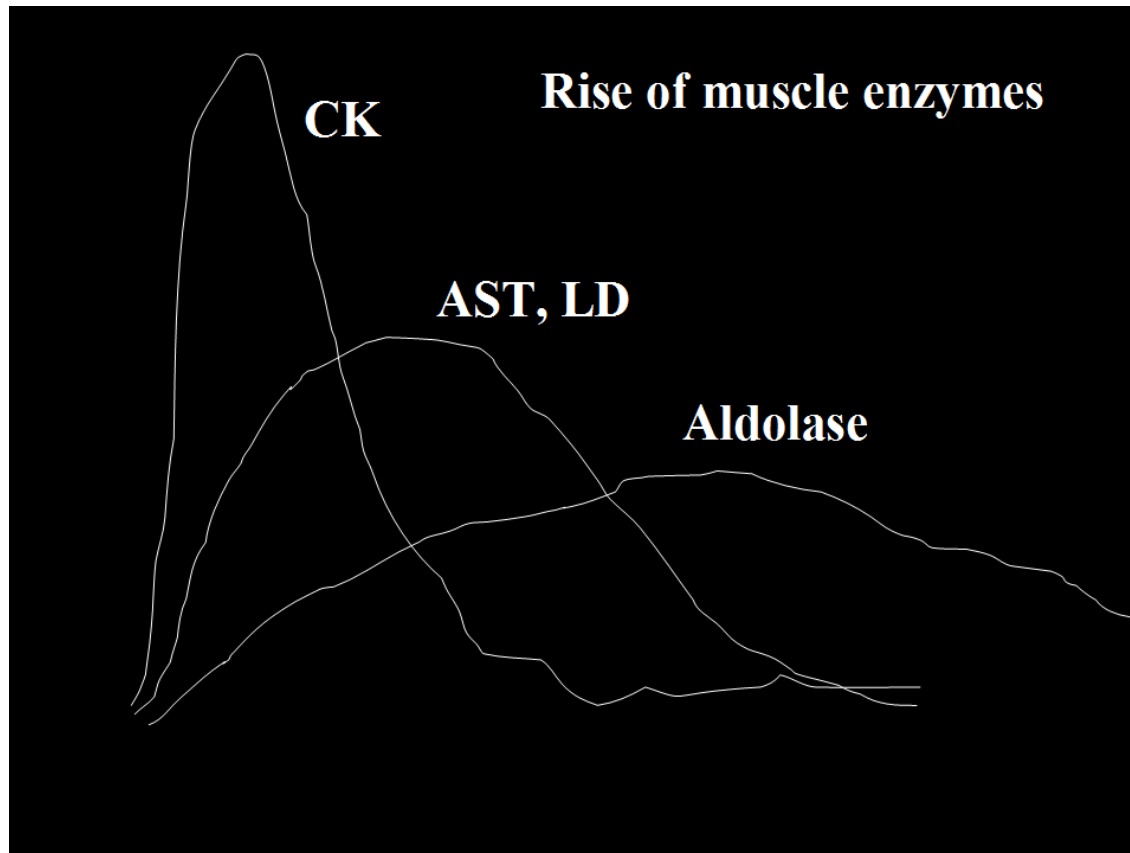
# Most Commonly Checked Labs: Muscle Enzymes



Most common enzymes followed to monitor disease activity:

- ▶ Creatine Kinase (CK)
- ▶ Aldolase
- ▶ Lactate Dehydrogenase (LDH)
- ▶ Aspartate Aminotransferase (AST)

# Pattern of Muscle Enzyme Elevation



# Less Specific Markers

## Sometimes Checked via Labs:



- Inflammation markers
  - Sed rate (Erythrocyte Sedimentation Rate), CRP (C-reactive Protein)
    - Can be increased in anything that causes inflammation, including viral infections
- Vascular Damage markers
  - Von Willebrand Factor Ag
    - Can be increased in other conditions such as exercise/stress, estrogen use, liver disease, or vasculitis.

# Myositis Markers Used for Prognosis (usually just checked at diagnosis):

## New Juvenile Myositis Autoantibody Phenotypes Differ in Risks, Presentations and Prognosis

*Anti-p155/140  
(Transcriptional  
Intermediary factor 1)*



JDM, JCTM, CAJDM  
Mod to severe weakness, widespread  
photosensitive rashes, lipodystrophy,  
chronic course

*Anti-MJ  
(Nuclear Matrix  
Protein 2)*



JDM > JPM, JCTM  
Muscle cramps, atrophy,  
contractures, dysphonia,  
calcinosis, GI ulcers

*Anti-CADM-140  
(Melanoma differentiation-  
associated protein 5)*



JDM, CAJDM;  
Fever, weight loss, fatigue,  
adenopathy, arthritis, ulcers,  
ILD; RPLD with high mortality

*Anti-HMG CoA  
Reductase (HMGCR)*

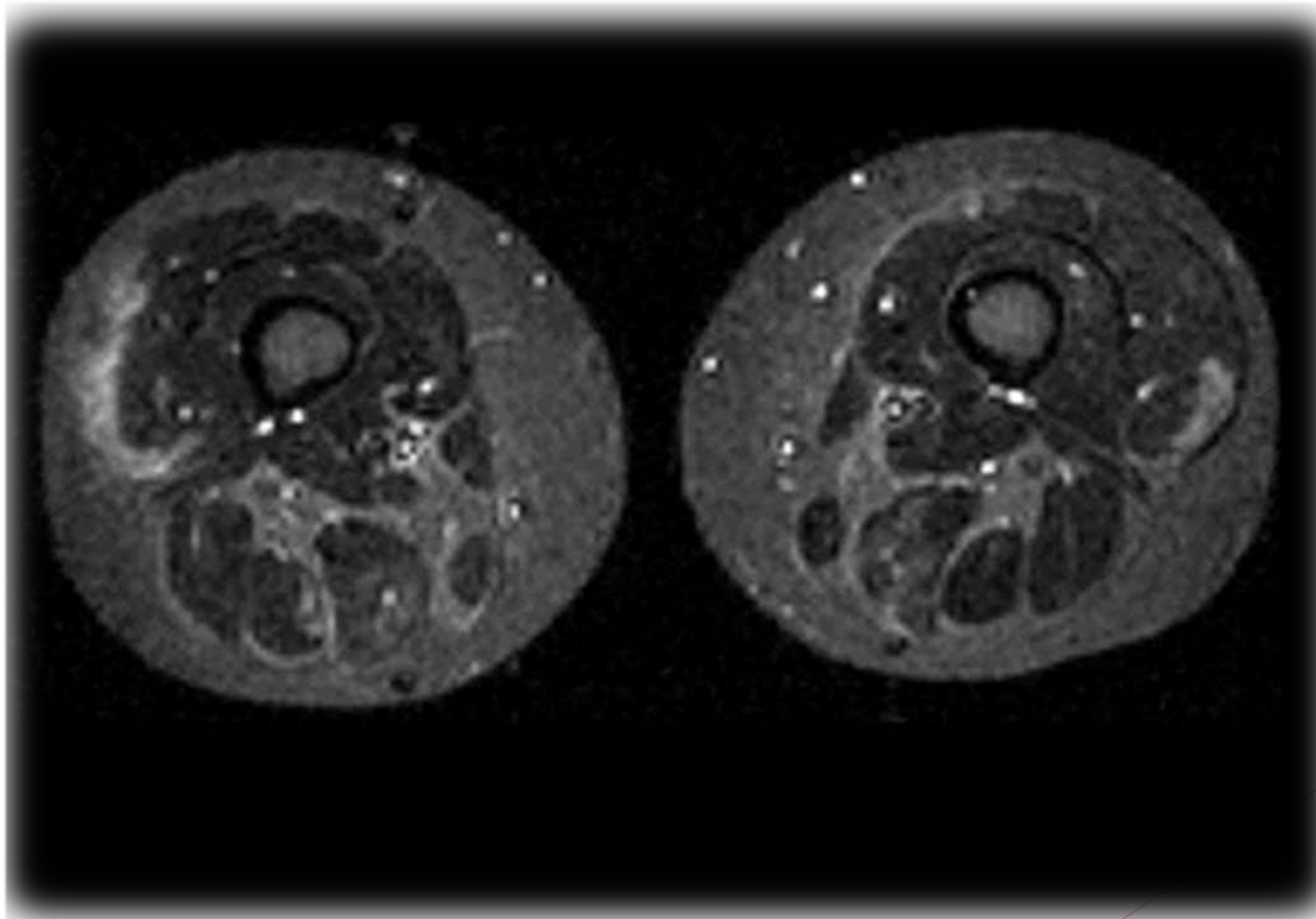


JPM, JDM  
Severe weakness, atrophy,  
contractures, arthralgias,  
High CK, necrotizing myopathy,  
chronic course, No statins

# Other Tests Used to Determine Extent of Disease

- ▶ Magnetic Resonance Imaging (MRI)
- ▶ Electromyography (EMG)
- ▶ Video Fluoroscopy Swallowing Study (VFSS)
- ▶ Pulmonary Function test (PFT)
- ▶ Xrays/CT/Ultrasound

# MRI Findings: Muscle Edema/Atrophy

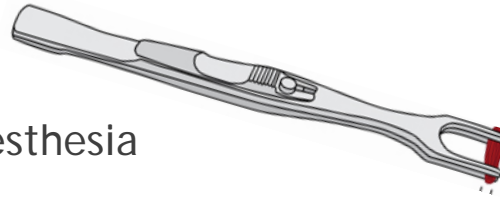




# Biopsy: Sometimes Used to Confirm the Diagnosis

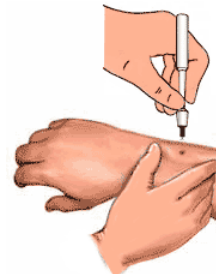
## ▶ Muscle biopsy

- ▶ Usually done under general anesthesia
- ▶ Guided by sites of inflammation seen on MRI
- ▶ Can help differentiate between JDM and other types of myositis/neuromuscular diseases if JDM presentation is not classic

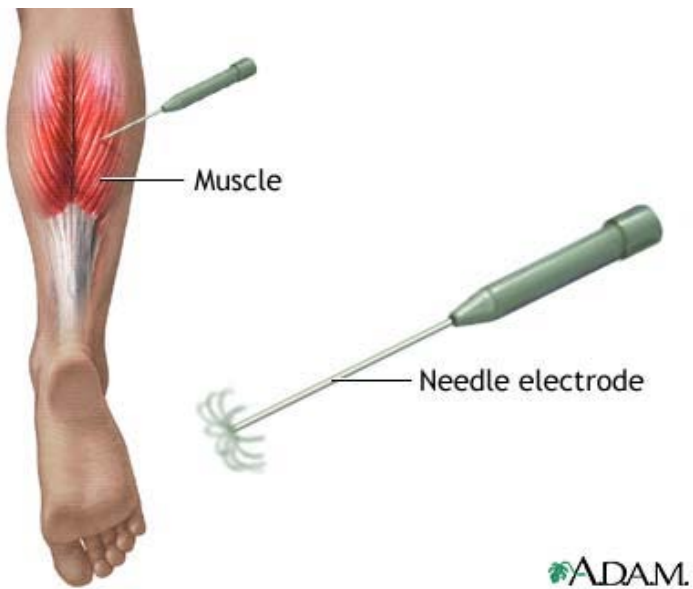


## ▶ Skin Biopsy

- ▶ Usually done under local anesthesia
- ▶ Helps distinguish between gottron's papules and other skin conditions if the skin findings are not classic



# Electromyography (EMG)



- ▶ Not commonly used in pediatrics as it is painful and operator-dependent.
- ▶ Measures how fast nerves can send electrical signals in the muscle.
- ▶ Sometimes used to differentiate neuromuscular diseases if JDM presentation is not typical.

# Video Fluoroscopy Swallowing Study

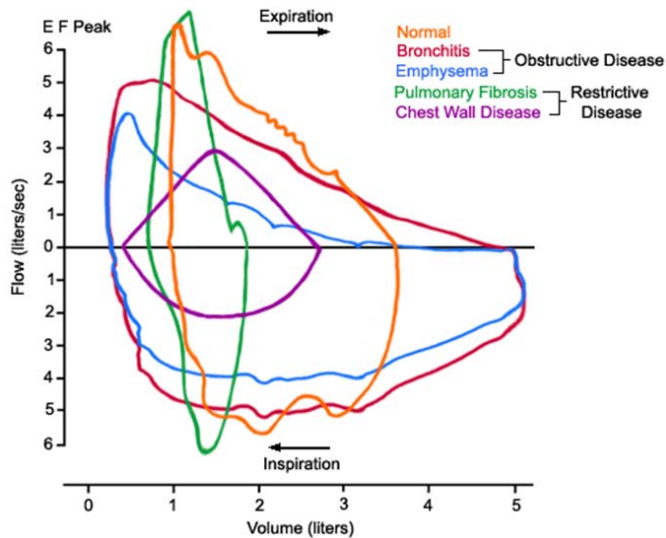


- ▶ In a radiology suite under fluoroscopy (which creates moving rather than still x-ray images), the patient is asked to swallow barium in thin liquid and paste consistencies, and then in paste on a cookie or cracker. The barium bolus is followed radiographically through the mouth, throat, and into the esophagus.



# Pulmonary Function Test

- ▶ Can check for “Restrictive Disease” - how well your muscles work in allowing you to inhale and exhale deeply.
  - ▶ Lung Volumes
  - ▶ Maximum inspiratory and expiratory pressures
  - ▶ Diffusion Capacity



# Other Studies, Sometimes Used to Monitor for Organ Dysfunction

- ▶ CT- if lung or abdominal involvement suspected
- ▶ Ultrasound-if abdominal involvement or arthritis suspected
- ▶ ECHO/EKG- if cardiac involvement is suspected
- ▶ Xrays- if calcinosis is suspected





# Why is Laboratory and Procedural Testing Important?

- ▶ Gives us information about how extensive a patient's JDM is, to decide level of aggressiveness in therapy
- ▶ May give us prognostic information to help us follow various organ systems more closely
- ▶ Gives us markers to follow, to allow us to taper the evil steroids, or add therapy if needed
- ▶ Gives us a way to monitor a patient in remission, to pick up on flares and treat them quickly