

# Pathology

## Team 435



### Lecture (3): Myopathies

#### Summary & MCQs

#### Objectives:

- Understand the structure of the various types of muscle fibers.
- Acquire a basic knowledge of the classification of myopathies and give examples of these disorders.
- Understand the meaning of the term muscular dystrophy and have a basic knowledge of the incidence and clinicopathological manifestations of Duchenne's and Becker's muscular dystrophies.
- Know the pattern of inheritance of myotonic dystrophy and its clinicopathological presentations.

## Summary:

- Skeletal muscle function can be impaired **secondarily** due to of problems with muscle innervation or by **primary** myopathy that can be inherited or acquired.
- The genetic forms of myopathy fall into several fairly distinct clinical phenotypes including:
  - **Muscular dystrophy**
  - **Congenital myopathy.**
  - **Congenital muscular dystrophy.**
- Depending on the type of mutation the disease may be severe such as Duchenne muscular dystrophy, or mild like Becker dystrophy.
- Acquired myopathies have diverse causes including **inflammation** and **toxic exposures.**

Name	Description	Causes	Characters	Histological Appearance
<b>Myopathy</b>	Diseases that affect skeletal muscle	-	Present with muscle weakness	-
<b>Muscular dystrophies</b>	A heterogeneous group of inherited disorders.	-	Often presenting in childhood. Progressive degeneration of muscle fibers	Muscle fibers are replaced by fibrofatty tissue
<b>DMD</b>	X-Linked Muscular Dystrophy	Abnormalities in the dystrophin gene	Most severe and the most common form of muscular dystrophy. Becomes clinically evident by age of 5	<ul style="list-style-type: none"> <li>-Marked variation in muscle fiber size (atrophy and hypertrophy)</li> <li>-Range of degenerative changes (fiber necrosis)</li> <li>-Regeneration, including sarcoplasmic basophilia, nuclear enlargement, and nucleolar prominence</li> </ul>
<b>BMD</b>	X-Linked Muscular Dystrophy	Abnormalities in the dystrophin gene	Less common and much less severe than DMD	<ul style="list-style-type: none"> <li>-Connective tissue is increased</li> <li>-Abnormal staining for dystrophin</li> <li>-Extensive fiber loss and adipose tissue infiltration</li> </ul>
<b>Infectious</b>	Heterogeneous group of rare disorders	<b>Causes 3 types of disorders:</b> <ul style="list-style-type: none"> <li>- Polymyositis</li> <li>- Dermatomyositis</li> <li>- Inclusion body myositis</li> </ul>	Immune-mediated muscle injury and inflammation.	-
<b>Myasthenia gravis</b>	Disorder of the neuromuscular junction	-	-	-
<b>Muscle Atrophy</b>	A non-specific response that means loss of muscle tissue	<ul style="list-style-type: none"> <li>- Simple disuse, type II fibers.</li> <li>- Exogenous glucocorticoids or endogenous hypercortisolism (proximal weakness), type II fibers</li> <li>- Myopathies</li> <li>- Neurogenic atrophy</li> </ul>	Abnormally small myofibers	-
<b>Neurogenic Atrophy</b>	Loss of a single neuron will affect all muscle fibers in a motor unit, so that the atrophy tends to be scattered over the field	<ul style="list-style-type: none"> <li>- Both fiber types are disused.</li> <li>- Clustering of myofibers into small groups</li> <li>- Deprived of their normal enervation, skeletal fibers undergo progressive atrophy</li> </ul>	-	-

**Check your understanding:**

1. **Which one of the following is considered as primary muscle disease and associated with necrosis?**
  - A. Myopathies
  - B. Neuropathic changes
  - C. Cardiac muscle atrophy
2. **Breast meat is an example of:**
  - A. Slow twitch myofibers
  - B. Fast twitch myofibers
  - C. Checkerboard appearance
  - D. Muscle atrophy
3. **The most common type of mutations in DMD and BMD:**
  - A. Deletion
  - B. Point mutation
  - C. Insertion
  - D. Duplication
4. **Abnormal levels of Creatine Kinase can provide early warning of a certain disease such as:**
  - A. Acute Renal Failure & DMD
  - B. BMD & DMD
  - C. Myotonic dystrophy & BMD
  - D. Polymyositis & Heart Attack
5. **Disuse and steroid induced atrophy primarily affect :**
  - A. type 1 fibers
  - B. type 2 fibers
  - C. both fibers
  - D. none of the above
6. **In the inflammatory myopathies, which one of the following is an autoimmune disorder:**
  - A. dermatomycosis
  - B. polymyositis
  - C. Ethanol myopathy
  - D. Inclusion body myositis
7. **A kid found himself to be clumsy as he has just turned five. His mother decided to bring to to a clinic and explained how her son couldn't keep up with the peers which made her worry. Which of the following could be the disease?**
  - A. BMD
  - B. Inflammatory disease
  - C. DMD
  - D. Fatigue

1-A 2-A 3-A 4-A 5-B 6-B 7-C

8. **Which of the following causes muscle atrophy:**
- A. Excessive exercise
  - B. Eating high sugar food
  - C. Inflammation
  - D. High steroid intake
9. **Which of the following may take the form of acute or chronic proximal muscle weakness:**
- A. Inflammatory myopathy
  - B. Ethanol myopathy
  - C. Thyrotoxic myopathy
  - D. Drug myopathy
10. **Inherited diseases that result in progressive muscle injury in patients who usually appear normal at birth:**
- A. Muscular dystrophies
  - B. Congenital muscular dystrophies
  - C. Congenital myopathies
  - D. Myopathies
11. **Which of the following is The dystrophin gene**
- A. Xp21
  - B. Xp22
  - C. Xq21
  - D. Xq22
12. **Which of the clinical features below is related to mitochondrial myopathies:**
- A. Manifest in early childhood
  - B. Manifest in early adulthood.
  - C. Distal muscle weakness
  - D. B & C
13. **Which of the following is inherited only as an autosomal dominant form:**
- A. Emery-dreifuss muscular dystrophy
  - B. Facioscapulohumeral dystrophy.
  - C. Limb-girdle muscular dystrophy
  - D. All of the above
14. **Simple disuse of type II fibers will cause:**
- A. Muscle hypertrophy
  - B. Muscle spasm
  - C. Neurogenic atrophy
  - D. Muscle atrophy
15. **Immune mediated muscle injury and inflammation are characteristics of which of the following:**
- A. Infectious myopathy
  - B. DMD
  - C. BMD
  - D. All of the above

8- D 9-C 10-A 11- A 12- B 13- B 14-D 15- A

Contact us: [Pathology435@gmail.com](mailto:Pathology435@gmail.com)

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## Team members

Nouf Altwajri  
Aljohara Almazroue  
Dalal AlHuzaimi  
Deema Alfaris  
Ghaida Aljamili  
Haifa Bin Taleb  
Lina Alshehri  
Lina Ismael  
Munerah AlOmari  
Nojood Alhaidari  
Nouf Alabdulkarim  
Ola Alnuhayer  
Raghad Almansour  
Samar AlOtaibi  
Shahad Albeshr  
Shahad Aldakhyil

Wael Al Oud  
Abdulaziz AlHammad  
Abdulaziz Redwan  
Abdullah AlGhizzi  
Abdulrahman Thekry  
Ahmad Alkhiary  
Ahmed Alrwaly  
Ahmed Al Yahya  
Ammar Almansour  
Anas Ali  
Faris Alwarhi  
Khalid Ali Alghsoon  
Mana AlMuhaideb  
Naif Alhadi  
Saleh Alkhalifa  
Rayan Almuneef

قال صلى الله عليه وسلم: من سلك طريقاً يلتمس به علماً سهل الله له به طريقاً إلى الجنة.

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