

Top Tips for Muscle Disease

1. Muscle weakness may result from pathology initiating anywhere throughout the neuromuscular system, from cerebral cortex to muscle cells and their surrounding metabolic environment.
2. Differential diagnosis varies widely, across the age groups, with timing of onset and predominant muscles groups involved.
3. History and examination will narrow the differential diagnosis and indicate appropriate initial investigations.
4. Examination of muscles for weakness, must include a neurological examination, to help identify level of neuromuscular system involvement, which will narrow the differentials for a diagnosis.
5. Be familiar with normal motor milestones and investigate early with creatine kinase levels if there are any sign of delayed development.
6. The Child Muscle Assessment Score tool was developed for Juvenile Dermatomyositis (JDM) assessment and monitoring in the over 5 year olds; a modified version can be used in pre-schoolers, but assessment tends to be more functionally based at this age.
7. Inherited myopathies are more likely with delay in development, whereas inflammatory myopathies are more likely with regression of abilities.
8. The cardinal features of JDM are proximal muscle weakness, characteristic rash predominately over extensor surfaces and raised muscle enzymes.
9. EMGs and muscle biopsies are generally not required for the diagnosis of JDM – diagnosis is made by genetic testing.
10. Acute weakness may be life threatening (cardio-respiratory failure, risk of aspiration).

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