IMPERATIVES FOR DUCHENNE MD

A Guide for Providers

D — DIAGNOSIS

- If developmental delay or elevated liver enzymes, do a creatine kinase (CK) (ChildMuscleWeakness.org)
- If male patients have a high CK (CK>800), order full genetic testing for Duchenne Muscular Dystrophy
- Discuss carrier testing/reproductive options for mother and testing for other family members

U — UNDERSTAND BREATHING PROBLEMS

- Pulmonary function test at least every year starting as young as possible
- Discuss cough assist when cough peak flow is < 270 liters per minute or if cough becomes weaker (use during respiratory illnesses while ambulatory and daily and as needed after loss of ambulation)
- Discuss nighttime Bi-PAP as needed or when forced vital capacity (FVC) < 30%
- Keep immunizations (including pneumonia and annual flu) up to date
- Treat respiratory infections promptly and aggressively
- Do NOT give supplemental oxygen without monitoring CO2

C — CORTICOSTEROIDS

- Start early!
- Discuss the benefits and possible side effects of corticosteroids by age 3 years, or as young as possible
- Evaluate efficacy and manage side effects of corticosteroids at each neuromuscular visit
- Discuss the rationale for lifelong steroid management
- Never stop taking steroids abruptly
- Discuss the need for stress dosing of steroids for illnesses or surgeries

H — HEART

- Cardiology visit with imaging (cardiac MRI preferred; echocardiogram if cardiac MRI not available) every year from diagnosis or more often if needed
- Discuss cardiac medications if fibrosis is seen on cardiac MRI, for any decrease in cardiac function decreases from baseline, or for heart failure (SF or shortening fraction <28% or ejection fraction <55%) or by age 10 even if findings are normal

E — ENDOCRINE

- If taking steroids, check 25-OH vitamin D prior to starting steroids, then annually
- Supplement vitamin D as needed
- Encourage sun exposure
- Nutrition discussions of adequate calcium and vitamin D intake
- Discuss measurement of bone density and use of bisphosphonates
- Assess spine for scoliosis at each visit
- Monitor for puberty starting at 9 years of age for need for testosterone therapy

N — NEVER FORGET PHYSICAL & OCCUPATIONAL THERAPY

- Specialized PT evaluations every 4-6 months
- Stretching every day
- Discuss and encourage contracture prevention (splints, stretches), appropriate exercise, assistive mobility devices (strollers, scooters, wheelchairs) and other assistive devices (beds, arm assistance, lifts, etc.) from the time of diagnosis

N — NUTRITION & GASTROINTESTINAL

- Monitor weight
- Assess/discuss diet (healthy eating, calcium, vitamin D)
- Evaluate swallowing/need for intervention
- Treat GERD and constipation as necessary
- See your dentist every 6 months

E — EMERGENCY

- Have patients/parents carry a copy of their last visit/note summary (including medications and neuromuscular contact information) and a Duchenne emergency card with them at all times
- Use caution with all anesthesia; avoid inhaled anesthesia
- Never use succinylcholine

M — MENTAL HEALTH

- Assess adjustment, coping, behavioral and emotional disorder and social isolation for the patient and family at each visit
- Screen for learning disability (reading and math), language problems, attention deficit disorder (ADD), attention deficit and hyperactivity disorder (ADHD), autism and obsessive compulsive disorder (OCD)
- Neurocognitive evaluation done at diagnosis and prior to formal schooling; screening/management as needed
- Discuss the need for individualized/special educational plan

D — DON'T DO IT ALONE

- Direct to trustworthy, reliable online resources
- Organize follow up via a comprehensive neuromuscular center with expertise in caring for people living with Duchenne
- Offer contact with organizations (**ParentProjectMD.org**, **TREAT-NMD.eu**, **WorldDuchenne.org**)

Care for Duchenne
parentprojectmd.org/care

Emergency Care Considerations
parentprojectmd.org/emergencycare

Center for Disease Control and Prevention Care Considerations
parentprojectmd.org/careguidelines

Parent Project Muscular Dystrophy

TREAT-NMD Neuromuscular Network

WORLD DUCHENNE ORGANIZATION UPPMD