ICD-10-CM Code Refinement:
for Duchenne/Becker MD & FSHD

More specific ICD-10 codes for
Duchenne/Becker and Facioscapulohumeral MD
will be included in the
CMS FY 19 Coding Addenda — effective October 1, 2018
Several major types of MD with dozens of sub-types (most sub-types are extremely rare).

4 most common types of muscular dystrophy are:

<table>
<thead>
<tr>
<th>Muscular dystrophy</th>
<th>Prevalence</th>
<th>Mode of inheritance</th>
<th>Age at onset / Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Becker</td>
<td>1.5 per 100,000 males</td>
<td>X-linked</td>
<td>Adolescence / Up to middle age</td>
</tr>
<tr>
<td>Duchenne</td>
<td>4.8 per 100,000 males</td>
<td>X-linked</td>
<td>2–6 years / Mid 20s</td>
</tr>
<tr>
<td>Facioscapulohumeral</td>
<td>4.0 per 100,000 population</td>
<td>Autosomal dominant</td>
<td>Childhood / Decades after onset</td>
</tr>
<tr>
<td>Myotonic</td>
<td>8.3 per 100,000 population</td>
<td>Autosomal dominant</td>
<td>20–40 years / Decades after onset</td>
</tr>
</tbody>
</table>

- Myotonic muscular dystrophy already has its own ICD 10 code.

- We proposed new codes for Becker, Duchenne, and Facioscapulohumeral muscular dystrophies.

- Duchene and Becker are caused by mutations in the same gene and they share symptoms, except that the symptoms in Duchenne start earlier and are more severe than in Becker; therefore, both muscular dystrophies could have a common code.
Relevance to Duchenne


MD STARnet Surveillance – CDC (2002 – present)

The CDC Care Considerations for DBMD - Lancet, 2010
CINRG’s natural history study and international data collection
The establishment of the CINRG Clinical Trial Network
PPMD’s Duchenne Connect Registry
national Certified Duchenne Care Centers
Medicaid state demonstrations projects (Duchenne utilization)
OneFlorida consortium
DOD’s Congressionally Directed Medical Research Program
Burden of Disease studies in Duchenne
DBMD Care Considerations Update – Lancet, 2018
Broad ICD code has proven a barrier to diagnosis, care, surveillance, research, and access for DBMD community:

- For our surveillance programs, we have also had to develop **costly and time consuming** ‘calculated variables’ to discern Duchenne cases from those abstracted using the 359.1 (ICD-9) and G71.0 (ICD-10) code.

- **Payer decisions are being impacted** as over-estimates of economic impacts are made when applying the calculations to their datasets.

- Care standards have been established but CDC has been **unable to assess whether those standards are being implemented**.
Assigning individual ICD 10 codes to the most common types of muscular dystrophies will:

• facilitate *surveillance*;
• allow *more accurate estimates* of incidence, prevalence, survivorship, mortality and its causes, injuries, symptoms, and health visits;
• help to identify factors that *influence health status and secondary conditions*
• *compare health information* across hospitals, regions, clinical settings, countries, and even across time in a given location;
• serve to keep *track of reimbursements*
• facilitate the *evaluation of guidelines* (implementation, adherence, and impact)
• **G70-G73** Diseases of myoneural junction and muscle
  o **G70** Myasthenia gravis and other myoneural disorders
  o **G71** Primary disorders of muscles
    ▪ **G71.0** Muscular dystrophy
    ▪ **G71.1** Myotonic disorders
      • **G71.11** Myotonic muscular dystrophy
      • **G71.12** Myotonia congenita
G71.00 Muscular dystrophy, unspecified
G71.01 Duchenne or Becker muscular dystrophy
  Autosomal recessive, childhood type, muscular dystrophy
  Benign [Becker] muscular dystrophy
  Severe [Duchenne] muscular dystrophy
G71.02 Facioscapulohumeral muscular dystrophy
  Scapulohumeral muscular dystrophy
G71.09 Other specified muscular dystrophies
  Benign scapuloperoneal muscular dystrophy with early contractures [Emery-Dreifuss]
  Congenital muscular dystrophy NOS
  Congenital muscular dystrophy with specific morphological abnormalities of the muscle fiber
  Distal muscular dystrophy
  Limb-girdle muscular dystrophy
  Ocular muscular dystrophy
  Oculopharyngeal muscular dystrophy
  Scapuloperoneal muscular dystrophy

**Oct 1, 2018**
PPMD Wishes to Acknowledge:

Dr. Christina Westfield, New York State Department of Health
Dr. Kathryn Wagner, Kennedy Krieger Institute
Dr. Katherine Mathews, University of Iowa
Dr. Emma Ciafaloni, University of Rochester
Centers for Disease Control and Prevention, NCBDD –
Dr. Julie Bolen, Natalie Street, Dr. Rudolfo Valdez
American Academy of Pediatrics
Global Partners Supporting our ICD Nomination

AMIS FSH (France)
Charley's Fund
Chris Carrino Foundation for FSHD
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Dutch FSHD Foundation
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FSH Society
Hope For Gus
Hope for Javier
JB's Keys for DMD
Jett Foundation
Little Steps Association (Israel)
Muscular Dystrophy Association (MDA)
Muscular Dystrophy Queensland
Muscular Dystrophy UK (MDUK)
Parent Project Muscular Dystrophy (PPMD)
Save Our Sons (Australia)
Suneel's Light
United Parent Project Muscular Dystrophy (UPPMD)
Walking Strong
Neuromuscular Clinical Community Overwhelmingly Supports this Nomination

** Please see Letter of Support provided by Clinical & Scientific Leaders

Thank You