ICD-10-CM Documentation and Coding Best Practices

Guidance on Documentation and Coding for Muscular Dystrophy

Muscular dystrophy overview

Muscular dystrophies are a group of muscle diseases caused by mutations in a person's genes. Over time, muscle weakness decreases mobility, making everyday tasks difficult.

There are many kinds of muscular dystrophy, each affecting specific muscle groups, with signs and symptoms appearing at different ages and varying in severity. Muscular dystrophy can run in families, or a person can be the first in their family to have muscular dystrophy. There may be several different genetic types within each kind of muscular dystrophy, and people with the same kind of muscular dystrophy may experience different symptoms.

Types of muscular dystrophy

Duchenne/Becker Muscular Dystrophy (DMD/BMD): Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD) have the same symptoms and are caused by mutations in the same gene. BMD symptoms can begin later in life and be less severe than DMD. However, because these two kinds are very similar, they are often studied and referred to together (DBMD). Parts of the body that can be affected:

- Upper legs/arms, heart, lungs, throat, stomach, intestines, and spine.

Myotonic Muscular Dystrophy is a common multisystem disorder that affects the skeletal muscles (the muscles that move the limbs and trunk) as well as smooth muscles (the muscles that control the digestive system) and cardiac muscles of the heart. Parts of the body that can be affected:

- Face, neck, arms, hands, hips, lower legs, heart, lungs, stomach, intestines, brain, eyes, and hormone-producing organs.

Limb-Girdle Muscular Dystrophy (LGMD) is a term for a group of diseases that cause weakness and wasting of the muscles in the arms and legs. The muscles most affected are those closest to the body (proximal muscles), specifically the muscles of the shoulders, upper arms, pelvic area, and thighs. Parts of the body that can be affected:

- Upper arms/legs, heart, spine, hips, calves, and trunk.

Facioscapulohumeral Muscular Dystrophy (FSHD) is a genetic muscle disorder in which the muscles of the face, shoulder blades, and upper arms are among the most affected. Parts of the body that can be affected:

- Face, shoulders, upper arms, eyes, ears, and lower legs.

Congenital Muscular Dystrophy (CMD) refers to a group of muscular dystrophies that become apparent at or near birth. Parts of the body that can be affected:

- Neck, upper arms/legs, lungs, brain, heart, and spine.

Distal Muscular Dystrophy (DD) is a group of rare diseases that affect your muscles (genetic myopathies). DD causes weakness that starts in the lower arms and legs (the distal muscles). It then may gradually spread to affect other parts of your body. The muscles shrink (atrophy). Parts of the body that can be affected:

- Feet, hands, lower legs/arms, and heart.

Oculopharyngeal Muscular Dystrophy (OPMD) is a rare genetic condition. It causes weakness in the muscles around the upper eyelids and part of the throat called the pharynx. Parts of the body that can be affected:

- Eyes, throat, shoulders, upper legs, and hips.

Emery-Dreifuss Muscular Dystrophy (EDMD) is a condition that primarily affects muscles used for movement (skeletal muscles) and the heart (cardiac muscles). Among the earliest features of this disorder are joint deformities called contractures. Parts of the body that can be affected:

- Arms, legs, heart, joints, throat, shoulders and hips.

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Signs and symptoms may appear as:

- Waddling gait.
- Walking on the toes.
- Muscle pain and stiffness.
- Difficulty rising from a lying or sitting position.
- Mild to moderate degrees of non-progressive intellectual impairment and learning disabilities.

Complications can include:

- Weakness and deterioration of the heart muscles (cardiomyopathy).
- Contractures and scoliosis.
- Reduced bone density.
- Breathing, coughing, and swallowing difficulties.

Treatment for muscular dystrophy may consist of:

- Physical, respiratory, speech, and occupational therapy.
- Surgery to have a pacemaker installed or to help correct any issues of the spine such as scoliosis.
- Drug therapy including glucocorticoids, anticonvulsants, immunosuppressants, and beta blockers.
- Gene-based therapy.

ICD-10-CM Information

G71 Primary disorders of muscles
- Excludes arthrogryposis multiplex congenita (Q74.3)
- Metabolic disorders (E70-E88)
- Myositis (M60.-)
- Excludes: "Not coded here" An excludes note indicates that the condition excluded is not part of the condition represented by the code, but a patient may have both conditions at the same time.

G71.00 Muscular dystrophy, unspecified

G71.01 Duchenne or Becker muscular dystrophy

G70.02 Facioscapulohumeral muscular dystrophy

G71.031 Autosomal dominant limb girdle muscular dystrophy

G71.032 Autosomal recessive limb girdle muscular dystrophy due to calpain-3 dysfunction

G71.033 Limb girdle muscular dystrophy due to dysferlin dysfunction

G71.0340 Limb girdle muscular dystrophy due to sarcoglycan dysfunction, unspecified

G71.0341 Limb girdle muscular dystrophy due to alpha sarcoglycan dysfunction

G71.0342 Limb girdle muscular dystrophy due to beta sarcoglycan dysfunction

G71.0349 Limb girdle muscular dystrophy due other sarcoglycan dysfunction

G71.035 Limb girdle muscular dystrophy due to anoctamin-5 dysfunction

G71.038 Other limb girdle muscular dystrophy

G71.039 Limb girdle muscular dystrophy, unspecified

G71.09 Other specified muscular dystrophies

G71.11 Myotonic muscular dystrophy

G71.20 Congenital myopathy, unspecified

G71.21 Nemaline myopathy

G71.220 X-linked myotubular myopathy

G71.228 Other centronuclear myopathy

G71.29 Other congenital myopathy

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Documentation best practices

The codes for muscular dystrophy identify the specific form of the disease. However, in some cases this does not identify the severity of the patients' manifestations, and these should be reported in addition to the code for the type of muscular dystrophy.

Document all manifestations of muscular dystrophy when they occur and identify causal effects such as osteoporosis as an adverse effect of long-term prednisone use.

Describe orthopedic symptoms, respiratory complications, developmental delay, wheelchair dependence, dysphagia, dysphasia or cardiomyopathy. Document any intellectual disability as mild, moderate or severe.

For contractures in muscular dystrophy, specify whether the contracture is a muscle contracture or a joint contracture and identify the site. There are specific codes in ICD-10-CM for joint contracture (M24.5-) and for muscle contractures (M62.4-). Abbreviation use with muscular dystrophy should be avoided on first reference. Myotonic dystrophy type 1 and myotonic dystrophy type 2 are often abbreviated as DM1 and DM2, common abbreviations for type 1 and type 2 diabetes mellitus. Spell out the first reference to avoid misinterpretation of the medical record.

References

- Centers for Medicare & Medicaid Services. (2020). ICD-10-CM official guidelines for coding and reporting: FY 2024. https://www.cms.gov/files/document/2021-coding-guidelines-updated-12162020.pdf