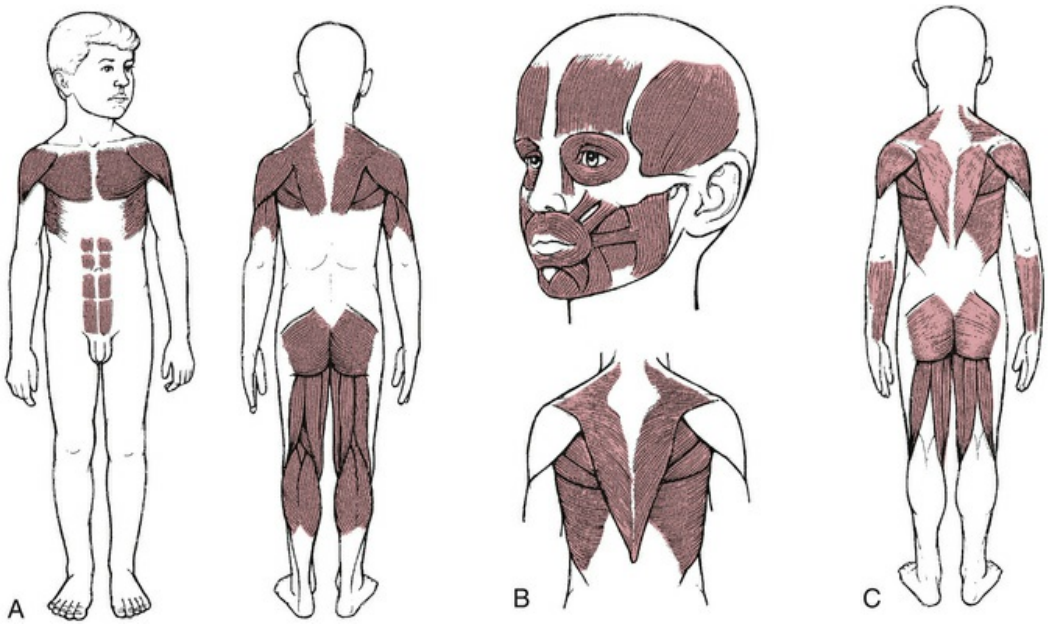

FIG 30-7 Initial muscle groups involved in muscular dystrophies (MDs). **A,** Pseudohypertrophic. **B,** Fascioscapulohumeral. **C,** Limb girdle.

Facioscapulohumeral (Landouzy-Dejerine) muscular dystrophy is inherited as an autosomal dominant disorder with onset in early adolescence. It is characterized by difficulty in raising the arms over the head, lack of facial mobility, and a forward slope of the shoulders. The progression is slow, and the life span is usually unaffected.

Limb-girdle muscular dystrophy (LGMD) is a heterogenous group of disorders with autosomal dominant and recessive inheritance whose clinical manifestations often appear in later childhood, adolescence, or early adulthood with variable but usually slow progression (Quan, 2011). All types of LGMD are characterized by weakness of proximal muscles of the pelvic and shoulder girdles. Other forms of MD include myotonic dystrophy, scapulohumeral MD (Emery-Dreifuss MD), fascioscapulohumeral MD (Landouzy-Dejerine disease), and congenital MD; these forms consist of subtypes of MD and are discussed at length elsewhere (see Sarnat, 2016b).

Treatment of the MDs consists mainly of supportive measures, including physical therapy, orthopedic procedures to minimize deformity, ventilation support, and assistance for the affected child in meeting the demands of daily living.