Chapter 9
Orthopaedic Management of the Child with Muscular Dystrophy

Robert K. Lark and Elizabeth W. Hubbard

Introduction

The orthopaedic care of children with muscular dystrophy (MD) is a challenging endeavor. Although many similarities exist, each subtype of MD can present differently. It is imperative that the proper diagnosis be confirmed so that treatment can be initiated based on knowledge of the natural history of the disease. Advancements in the medical management of MD are challenging historic recommendations for orthopaedic care. This chapter is intended to provide a brief, general overview of orthopaedic management of the most common childhood MDs.

Duchenne Muscular Dystrophy

Duchenne muscular dystrophy (DMD) is the most common childhood MD. As diagnostic genetic testing continues to improve, referral to the orthopaedic surgeon for consideration of muscle biopsy may be on the decline. Orthopaedic manifestations of this disease include gait abnormality, muscle weakness/imbalance, joint contractures, fractures, and scoliosis.

R.K. Lark, M.D., M.S. • E. Hubbard, M.D. (✉)
Department of Orthopaedic Surgery, Duke University Medical Center,
Lenox Baker Children's Hospital, 3000 Erwin Rd,
Durham, NC 27705, USA
e-mail: elizabeth.hubbard@duke.edu

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Non-operative Management

Although not necessarily applicable to other forms of MD, steroids (i.e. deflazacort, prednisone) are showing promise for prolonging ambulatory ability and decreasing the rate of scoliosis [1–4]. Unfortunately, side effects such as obesity, osteopenia, and fractures may negate some of the positive attributes of the drugs [5–7]. Studies are ongoing to determine the appropriate dosing regimen and duration of steroid treatment as cessation of the drug seems to lead to rapid decline of muscle strength [1, 3].

A common presentation to the orthopaedic surgeon is for evaluation of toe walking in early childhood, often in the three- to five-year age range. Careful assessment of the child's birth history and complete musculoskeletal physical exam is essential to rule out other causes such as idiopathic toe walking, cerebral palsy, etc. It is not uncommon for these children to have a mild equinus contracture at this age and physical therapy can aid in maintaining ankle joint range of motion. Serial casting may also be considered followed by maintenance of foot position with an ankle foot orthosis [8]. As the child continues to grow, hamstring and hip flexion contractures worsen as the proximal muscles weaken. Continued stretching and consideration of a knee-ankle-foot orthosis may be considered to prolong ambulation and standing ability [9–11]. Contractures also occur in the upper extremities and may require occupational therapy evaluation for stretching and assistive devices to aid in activities of daily living [12–14].

Patients not receiving steroids have a high incidence of developing scoliosis. Once confined to a wheelchair, thoracic supports may aid in maintaining sitting posture, but likely do not impact progression of the curve. Bracing is not indicated as the efficacy of brace treatment has not been shown to be beneficial in this condition [15–20].

Fractures are common once ambulatory ability is lost [5–7, 21]. Most fractures occur about the knee (most commonly the distal femur). Fracture management typically is non-operative. Care should be taken to avoid rigid immobilization with a heavy fiberglass or plaster cast as this can cause a fracture at the proximal end of the wrap. Most children are comfortable in a bulky wrap for four to six weeks until signs of radiographic union are evident. Bisphosphonates have been shown to improve back pain in patients with vertebral fractures, but have not been shown to decrease fracture risk [6].

Operative Management

When conservative measures fail, operative management of children with DMD can be beneficial. Clear goals of the surgical procedure must be discussed with and understood by the family. Shapiro et al. proposed a system of surgical approaches based on the ambulatory ability of the patient [9]. The three basic categories include

ambulatory, rehabilitative, and palliative. The ambulatory category was subdivided into early-extensive, moderate, and minimum ambulatory approaches. The early-extensive approach is intended to be performed while the patient remains ambulatory and attempts to prevent the extensive contractures around the hip, knee, and ankle. The procedure involves excising the tensor fascia, lengthening the hip flexors, hamstrings, Achilles tendon, and possibly transferring the posterior tibialis through the inter-osseus membrane to the dorsum of the foot. While early reports of this method were promising, longer follow-up studies have failed to show much benefit. The "moderate ambulatory approach" is intended to address existing contractures in hopes to maintain ambulatory ability. The approach is similar to the extensive approach by utilizing intramuscular lengthening techniques of the gastrosoleus complex as well as lengthening the hamstrings. The lengthening of hip flexors and tensor fasciae showed no increased benefit. The minimum ambulatory approach addresses only the equinus contracture. An intramuscular approach such as the Vulpius lengthening is recommended to minimize the risk of over-lengthening potential that can occur with a z-lengthening. The rehabilitative approach is intended to allow a child with recent loss of ambulation to regain the ability to walk. This involves addressing the hip and knee contractures as well as percutaneously tenotomizing the Achilles. This approach requires post-operative brace management, but has been reported to increase ambulatory ability from several months to a few years. The palliative approach addresses the severe equinus deformity that prevents the patient from achieving a plantigrade foot to rest on the plate of his wheelchair. The procedure involves tenotomies of the Achilles, flexor digitorum, and flexor hallucis longus as well as tenotomy vs. transfer of the posterior tibialis to the dorsum of the foot [22, 23].

In the event the patient develops scoliosis, it should be addressed quickly (Cobb angle >20°) as it remains unclear which patients will progress [20, 24]. Bracing is ineffective in this condition. Delaying surgery may unintentionally cause the child to have to forego spinal fusion due to decline in pulmonary reserve [20, 24]. Surgical correction of scoliosis in patients with DMD has generated much debate. Instrumentation must be individualized to each patient and may include pedicle screw fixation, sublaminar wires, hooks, or a combination of anchors. Patients with pelvic obliquity should be fused to the pelvis, whereas those with a level pelvis may have success stopping at L5 [25–27]. The patient and family should be warned of potential risks such as bleeding, prolonged intubation, and infection. Additionally, children with moderate to severe upper extremity weakness may no longer be able to feed themselves as they lose the ability to move their trunk to their hands [28, 29].

Becker Muscular Dystrophy

Becker muscular dystrophy (BMD) is a less severe dystrophinopathy compared to Duchenne. These children ambulate for a longer duration and may never become confined to a wheelchair. Scoliosis is infrequent in this population.

Non-operative Management

As with Duchenne, corticosteroids play a role in the non-operative treatment of BMD. Johnsen [30] reported that two patients with Becker's had a significant improvement in overall strength and reduction in serum creatine kinase levels after therapeutic treatment with prednisone. Because the severity of the disease varies with the level of functional dystrophin protein that is expressed, patients with BMD can differ in the clinical manifestations of the disease. Further studies are required in this patient population to determine which subset of patient's with BMD would best benefit from prolonged corticosteroid treatment.

Patients with BMD often remain ambulatory longer and have an overall slower disease course than patients with DMD. Orthoses can be beneficial in patients who develop ankle and forefoot equinus [31]. Patients with BMD are felt to be better candidates for bracing than patients with Duchenne, both because they remain ambulatory and retain muscle strength longer.

Operative Management

Patients with BMD can develop similar orthopaedic conditions as those with Duchenne. However, the manifestations are typically delayed and less severe. The need for and timing of surgical intervention for orthopaedic manifestations of Becker's is both reduced and delayed when compared with patients who have DMD [32].

Forefoot and ankle equinus have been described in these patients. When refractory to stretching and orthoses, intramuscular lengthening of the Achilles tendon is effective for management of ankle equinus [33]. Patients should also undergo concurrent posterior tibialis tendon transfer to the dorsum of the foot if appropriate [32, 33].

Scoliosis is seen more commonly in non-ambulatory adolescents. Because most patients with BMD remain ambulatory through adolescence and into adulthood, fewer patients develop scoliosis in adolescence [32]. Patients with BMD are still at greater risk for scolisosis overall and should be monitored closely with serial exams. As with patients who have DMD, patients with BMD should be considered surgical candidates when curves progress beyond 20° and the surgical principles are the same for both conditions [34].

Facioscapulohumeral Muscular Dystrophy

Facioscapulohumeral muscular dystrophy (FSHD) is an autosomal dominant disorder resulting in progressive weakness of the shoulder girdle and facial musculature. Orthopaedic manifestations include severe shoulder weakness, hyperlordosis, and eventually gait abnormalities. Scoliosis is rare but does occur.

Non-operative Management

Non-operative approaches to the management of FSHD have evolved over recent years. Early literature reported that muscle strengthening placed patients at risk for disease progression due to destruction of muscle fibers. However, more recent literature has shown that strength training and physical therapy can have positive effects. Andersen et al. showed that creatine kinase levels normalize within 24 h compared with pre-exercise levels, suggesting that irreversible or prolonged muscle damage is not an effect of exercise in these patients [35]. Olsen et al. found that a three-month low-intensity aerobic training program both improved oxygen uptake and caused no evidence of muscle damage among patients with FSHD [36]. Bakhtiary et al. attempted to optimize muscle function and found that simple motor learning programs could help FSHD patients adopt more effective muscle performance during basic tasks that require shoulder abduction and elbow flexion [37].

Taking things a step further, Pasotti et al. designed a six-month exercise and nutrition supplement program for a 43-year-old patient with FSHD [38]. At the time of the initiation of the program, the patient was noted to have severe proximal muscle weakness, hyperlordosis, and was no longer ambulatory. Pulmonary function tests revealed mild restrictive lung disease. The patient began a regimen of both endurance and strength training. Her diet was supplemented with branched chain amino acids, creatinine, and conjugated linoleic acid, based on previously published data that these agents can limit exercise-induced injury [39–41]. The patient developed a modest increase in shoulder abduction strength, with improvement in body mass composition and stabilization of pulmonary function tests, with no evidence of muscle soreness or muscle destruction [38].

There is also growing literature on the effects of albuterol as a potential adjunct in the treatment regimen for patients with facioscapulohumeral dystrophy. A pilot trial of 15 patients with FSHD shows that daily treatments of sustained-release albuterol over a three-month period significantly improved patients' lean body mass and strength [42]. A follow-up randomized clinical trial involving 90 FSHD patients showed that daily treatment with long-acting albuterol resulted in improved grip strength and significantly improved muscle mass, although global measures of muscle strength did not significantly improve [43]. Albuterol was relatively well-tolerated by the patients in the study and the authors suggested that combining albuterol with other treatment modalities, such as strength training programs, might result in more significant anabolic effects.

Operative Management

Surgical management of facioscapulohumeral dystrophy is largely directed at scapular stabilization. The manifestation of FSHD that mostly limits daily activities is the patient's inability to abduct his or her shoulders. Weakness of the trapezius,

rhomboids, levator scapulae, and subscapularis causes significant scapular winging with any attempt at shoulder abduction. Stabilizing the scapula against the thoracic wall allows the deltoid and supraspinatus to abduct and forward flex the upper extremity. The shoulder range of motion achieved with stabilization is still less compared to that of unaffected patients, but the motion and strength is significantly improved post-operatively for patients with FSHD. Stabilization can be done either through scapulopexy or scapulothoracic arthrodesis.

Scapulopexy involves stabilizing the scapula against the thoracic cavity without attempting to achieve an arthrodesis. This can be achieved by using autograft, such as fascia lata graft, or other materials such as merseline tape, dacron, or looped wire [44, 45]. The procedure requires minimal post-operative immobilization. After surgery, patients can begin immediate shoulder range of motion. Because immediate range of motion is encouraged, patients can undergo contemporaneous bilateral scapulopexy or briefly stage the procedure so that both shoulders can be addressed. Ketenjian suggested that scapulopexy should be the preferred treatment in patients with FSHD because it does not interfere with rib excursion and therefore would not have significant negative effects on pulmonary function [45]. He reported improved shoulder abduction of an average 33°, as well as improvement in strength, endurance, pain, and cosmesis in five patients who underwent scapulopexy. Average patient follow-up in this study was 34 months. Giannini et al. reported significant improvements in both shoulder abduction and forward flexion in 10 patients who underwent scapulopexy for FSHD. The scapulae were fixed to the underlying fourth through seventh ribs using wires passed through bone tunnels. Although initial results were good, average forward flexion and muscle strength declined during long-term follow-up [44].

Scapulothoracic arthrodesis is a more technically demanding procedure than scapulopexy. The goal is to fuse the scapula to the underlying ribs. Numerous techniques have been described, although many follow a similar surgical plan [46–55]. Authors recommend making a posterior incision and elevating the rhomboids and trapezius off the medial border of the scapula, followed by a subperiosteal exposure of the underlying ribs [46, 48, 50, 55]. While most techniques recommend using iliac crest autograft, there are reports of successful arthrodesis using allograft [51]. Wires or multifilament cables are then threaded around the underlying ribs and attached to the scapula through bone tunnels, with more recently reported techniques recommending using a reconstruction or LCP plate to supplement scapular fixation and prevent scapular fracture and wire cut-out [47, 49, 54]. Most methods involve a post-operative immobilization period of 6–12 weeks to allow time for a successful fusion, although some authors encourage immediate range of motion post-operatively [50, 55]. Once shoulder range of motion is initiated, the rehabilitation protocol typically involves gentle range of motion passively, then actively, and then range of motion with weight bearing. The recovery time from this procedure is approximately six months. Simultaneous bilateral scapulothoracic arthrodesis is not encouraged because of the prolonged immobilization and weight-bearing restrictions post-operatively.

Both scapulopexy and scapulothoracic arthrodesis are complex procedures and a full pre-operative evaluation of the patient must be performed. Pre-operatively, the patient's passive and active shoulder abduction and forward flexion should be evaluated and recorded. The patient's active shoulder abduction and forward flexion should then be re-evaluated with the examiner stabilizing the patient's scapula. This is known as the Horwitz maneuver. With the examiner holding the patient's scapula, the deltoid and supraspinatus can contract against a stable scapula, allowing the muscles to abduct and forward flex the shoulder [33, 56]. Patients who will most benefit from a scapulopexy or scapulothoracic arthrodesis have active abduction and forward flexion from 90° to 120° when their scapulae are stabilized during the Horwitz maneuver. This represents the range of abduction required to carry out most activities of daily living [45]. Repeating the Horwitz maneuver with the scapula fixed at varying degrees of rotation against the thoracic wall can help determine the ideal position for scapular fixation intra-operatively by allowing the surgeon to see which scapula position gives the patient the optimal amount of shoulder motion.

When deciding between scapulopexy and scapulothoracic arthrodesis, the patient's pulmonary function and rate of disease progression should both be considered. Scapulopexy is a less invasive procedure and has been shown to have a less significant effect on a patient's pulmonary function, specifically forced vital capacity and overall vital capacity. This is even more important if the patient is considering bilateral procedures, as bilateral scapulothoracic arthrodesis has been shown to significantly affect pulmonary function both in the early recovery period and over the long-term follow-up. Also, patients with a rapidly progressive disease are likely better candidates for scapulopexy, as they would require less immobilization and would gain immediate function after the procedure. Most patients with FSHD have little to no decreased pulmonary function compared to unaffected patients and disease course is generally slow, with a normal life-expectancy [31–33, 56]. Therefore, most patients would have a greater overall benefit from a scapulothoracic arthrodesis as the range of motion and daily function is retained over time. Patients should be warned that both cross-body adduction and internal rotation behind the body will be limited post-operatively.

Intra-operatively, important considerations include how to position and prep the patient and where to anchor the scapula on the thoracic wall. Prepping and draping the entire upper extremity into the surgical field allows the surgeon to check pulses in the extremity to confirm that scapular stabilization has not affected the vascular supply to the upper extremity. Mackenzie et al. reported taking a patient back to the operating room for immediate revision after the patient was noted to have a cold upper extremity in the post-anesthesia recovery area [57]. Repositioning the scapula on the ribs resulted in immediate return of the radial pulse [57]. Including the extremity in the sterile field also allows the surgeon to range the arm after scapular stabilization to test the strength of the fixation. Most authors recommend attaching the scapula to the third through sixth or fourth through seventh ribs, in no more than 30° of external rotation. Preoperative evaluation can help the surgeon plan how to position the scapula on the thoracic wall to achieve optimal range of motion for the patient. Once the scapula is fixed to the ribs, it is recommended that the surgical field

be filled with normal saline and that the anesthesia team initiate positive pressure ventilation to check for any leak in the pleura. If a leak is detected, a chest tube should be placed intra-operatively to prevent a pneumothorax post-operatively [49].

Although early reports of scapulopexy and scapulothoracic arthrodesis indicated that there were minimal complications with these procedures, more recent reports have shown that complication rates are higher than previously indicated. Goel et al. reported a 50% complication rate overall after scapulothoracic arthrodesis in 12 shoulders [49]. Intra-operative complications include pleural tear, pneumothorax, and hemothorax [49, 52, 53, 55, 58]. Brachial plexus palsy has been reported infrequently, but can be a devastating complication [47, 57, 59]. In scapulothoracic arthrodesis, non-union rates have been reported as frequently as 15–17% [47–49, 53, 55]. Most authors do not obtain routine post-operative CT scans on patients to evaluate for bony fusion, so the reported non-union rates are reflective of painful pseudoarthroses. It is possible that routine CT evaluation of patients at 6 or 12 months post-operatively would demonstrate a slightly higher nonunion rate than what is currently recorded in the literature. Rib and scapula fractures intra-operatively as well as rib stress fractures post-operatively have also been reported [53, 55, 58]. Post-operatively, painful hardware has been reported and studies have shown up to a 50% return to surgery rate for removal of hardware after arthrodesis is confirmed through imaging [49].

Early reports of both scapulopexy and scapulothoracic arthrodesis did not routinely analyze the effects that these procedures had on a patient's pulmonary function. However, more recent studies include the effects that these surgeries have on patients' pulmonary function tests immediately after the procedure and during follow-up. Studies show that there are usually mild losses in FEV1 and forced vital capacity after unilateral scapulothoracic arthrodesis, but more significant decline can be seen after bilateral surgeries [44, 47, 48, 50, 54, 55, 58]. However, there are isolated reports of significant decline in pulmonary function after unilateral surgeries [58].

Patients with FSHD do not generally require operative intervention for the lower extremities or the spine. If tibialis anterior and/or peroneal weakness develops, patients may develop a flexible equinus or equinovarus foot position. This usually responds well to bracing with ankle-foot orthoses. If the deformity becomes rigid, the patient could undergo an intramuscular lengthening [33, 56]. Scoliosis is rare in patients with facioscapulohumeral dystrophy. If a curve greater than 40–50° develops, it should be managed following the same principles that one would use to manage adolescent idiopathic scoliosis [56]. Patients often develop hyperlordosis and, if severe, this can be managed with an orthosis. However, an orthosis may interfere with the patient's ability to ambulate, as hyperlordosis allows the patient to compensate for progressive hip extensor weakness [56].

Infantile Facioscapulohumeral Muscular Dystrophy

Infantile facioscapulohumeral muscular dystrophy (iFSHD) has become increasingly recognized as a related but distinct disease process over the past several decades [31, 32, 56, 60–63]. Although molecular evaluation has shown that the same gene is

affected as in adolescent facioscapulohumeral dystrophy, the disease course is more rapid and clinically this mirrors more severe MDs, such as Duchenne [61]. Infants usually develop facial diplegia [62]. Children begin walking at a normal age, but they rapidly develop pelvic girdle weakness. Children can have a positive Gower's sign and demonstrate significant hip extensor weakness on exam. They develop marked, severe hyperlordosis and use their hands to help stabilize their hip extensors while standing and walking, which is a near pathognomonic sign of this disease [63]. An equinus foot position develops, usually as a compensatory measure for quadriceps and tibialis anterior weakness. Patients develop shoulder girdle weakness, but unlike their adolescent and adult counterparts, it is the pelvic girdle weakness that dictates treatment in these patients.

The overall treatment goal for patients with iFSHD is to preserve function. Although hyperlordosis is severe, these patients do not respond well to bracing and use of orthoses should be limited. The hyperlordosis is a compensatory development, meant to counter the severe hip extensor weakness in these patients. Correcting the hyperlordosis with a brace or with surgery can actually inhibit ambulation because the patient can no longer compensate for their pelvic girdle weakness [33, 63]. Similarly, the equinus foot position is also a compensatory measure. Most children respond well to ankle-foot orthoses or knee-ankle-foot orthoses, but on the rare occasion when a patient develops a rigid equinovarus foot position, an intramuscular lengthening could be considered [33, 63].

Most patients lose the ability to ambulate by the second or third decade [60–62]. If the hyperlordosis is severe at that point, one could consider either bracing or surgical intervention to improve the patient's ability to sit in a wheel chair [33]. The shoulder girdle weakness is typically not a limiting condition for these patients because their spine and lower extremity conditions tend to be more severe. If the shoulder girdle weakness does interfere significantly with daily function, scapular stabilization can be considered. Scapulopexy is favored over scapulorthoracic arthrodesis in these settings because it allows immediate shoulder range of motion, there is no post-operative immobilization, and the procedure has less of an effect on pulmonary function, which is significantly more limited in patients with infantile facioscapulohumeral dystrophy. Scapular stabilization is not routinely performed in these patients because their disease course is so rapid, and the patients lose function in their upper extremities so quickly that the surgical benefits would not outweigh surgical risks to make the procedure worthwhile [32].

Limb-Girdle Muscular Dystrophy

Limb-girdle muscular dystrophy (LGMD) encompasses an increasing number of diseases that cause proximal muscle weakness, especially of the shoulders, pelvic region, and thighs. Due to the heterogeneity of this diagnosis, specific recommendations should be tailored to the genetically diagnosed disease. This section's brevity reflects the difficulty in recommending orthopaedic management for this group of diseases as they all present varying clinical phenotypes.

Non-operative Management

LGMD is a general diagnosis that encompasses at least 16 genetically distinct disease processes [32]. As such, the manifestations of the disease can vary significantly from one patient to another, depending on the underlying genetic abnormality. Overall, symptoms and disease progression mimic that of BMD. Most treatment is supportive and includes physical therapy and orthoses to maximize muscle strength and prevent contracture formation [64, 65].

Operative Management

Surgical indications are similar to those utilized for BMD [31]. Unlike patients with DMD, scoliosis does not develop in most patients with LGMD. Patients who do develop scoliosis also generally have mild curves that do not require surgical intervention [32, 33].

References

1. Biggar WD, et al. Long-term benefits of deflazacort treatment for boys with Duchenne muscular dystrophy in their second decade. Neuromuscul Disord. 2006;16(4):249–55.
2. Bushby K, et al. Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. Lancet Neurol. 2010;9(1):77–93.
3. King WM, et al. Orthopedic outcomes of long-term daily corticosteroid treatment in Duchenne muscular dystrophy. Neurology. 2007;68(19):1607–13.
4. Lebel DE, et al. Glucocorticoid treatment for the prevention of scoliosis in children with Duchenne muscular dystrophy: long-term follow-up. J Bone Joint Surg Am. 2013;95(12):1057–61.
5. Bothwell JE, et al. Vertebral fractures in boys with Duchenne muscular dystrophy. Clin Pediatr. 2003;42(4):353–6.
6. James KA, et al. Risk factors for first fractures among males with Duchenne or Becker muscular dystrophy. J Pediatr Orthop. 2014.
7. Pouwels S, et al. Risk of fracture in patients with muscular dystrophies. Osteoporos Int. 2014;25(2):509–18.
8. Main M, et al. Serial casting of the ankles in Duchenne muscular dystrophy: can it be an alternative to surgery? Neuromuscul Disord. 2007;17(3):227–30.
9. Bushby K, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: implementation of multidisciplinary care. Lancet Neurol. 2010;9(2):177–89.
10. Garralda ME, et al. Knee-ankle-foot orthosis in children with Duchenne muscular dystrophy: user views and adjustment. Eur J Paediatr Neurol. 2006;10(4):186–91.
11. Siegel IM. Maintenance of ambulation in Duchenne muscular dystrophy. The role of the orthopedic surgeon. Clin Pediatr. 1980;19(6):383–8.
12. Alemdaroglu I, et al. Different types of upper extremity exercise training in Duchenne muscular dystrophy: effects on functional performance, strength, endurance, and ambulation. Muscle Nerve. 2014.

13. Bartels B, et al. Upper limb function in adults with Duchenne muscular dystrophy. J Rehabil Med. 2011;43(9):770–5.
14. Mattar FL, Sobreira C. Hand weakness in Duchenne muscular dystrophy and its relation to physical disability. Neuromuscul Disord. 2008;18(3):193–8.
15. Arun R, Srinivas S, Mehdian SM. Scoliosis in Duchenne's muscular dystrophy: a changing trend in surgical management: a historical surgical outcome study comparing sublaminar, hybrid and pedicle screw instrumentation systems. Eur Spine J. 2010;19(3):376–83.
16. Hsu JD. The natural history of spine curvature progression in the nonambulatory Duchenne muscular dystrophy patient. Spine. 1983;8(7):771–5.
17. Karol LA. Scoliosis in patients with Duchenne muscular dystrophy. J Bone Joint Surg Am. 2007. 89 Suppl. 1:155–62.
18. Kurz LT, et al. Correlation of scoliosis and pulmonary function in Duchenne muscular dystrophy. J Pediatr Orthop. 1983;3(3):347–53.
19. Miller F, et al. Pulmonary function and scoliosis in Duchenne dystrophy. J Pediatr Orthop. 1988;8(2):133–7.
20. Shapiro F, et al. Progression of spinal deformity in wheelchair-dependent patients with Duchenne muscular dystrophy who are not treated with steroids: coronal plane (scoliosis) and sagittal plane (kyphosis, lordosis) deformity. Bone Joint J. 2014;96-B(1):100–5.
21. Gray B, Hsu JD, Furumasu J. Fractures caused by falling from a wheelchair in patients with neuromuscular disease. Dev Med Child Neurol. 1992;34(7):589–92.
22. Leitch KK, et al. Should foot surgery be performed for children with Duchenne muscular dystrophy? J Pediatr Orthop. 2005;25(1):95–7.
23. Scher DM, Mubarak SJ. Surgical prevention of foot deformity in patients with Duchenne muscular dystrophy. J Pediatr Orthop. 2002;22(3):384–91.
24. Yamashita T, et al. Prediction of progression of spinal deformity in Duchenne muscular dystrophy: a preliminary report. Spine. 2001;26(11):E223–6.
25. Alman BA, Kim HK. Pelvic obliquity after fusion of the spine in Duchenne muscular dystrophy. J Bone Joint Surg Br. 1999;81(5):821–4.
26. Bui T, Shapiro F. Posterior spinal fusion to sacrum in non-ambulatory hypotonic neuromuscular patients: sacral rod/bone graft onlay method. J Child Orthop. 2014;8(3):229–36.
27. Sengupta DK, et al. Pelvic or lumbar fixation for the surgical management of scoliosis in Duchenne muscular dystrophy. Spine. 2002;27(18):2072–9.
28. Duckworth AD, Mitchell MJ, Tsirikos AI. Incidence and risk factors for post-operative complications after scoliosis surgery in patients with Duchenne muscular dystrophy: a comparison with other neuromuscular conditions. Bone Joint J. 2014;96-B(7):943–9.
29. Ramirez N, et al. Complications after posterior spinal fusion in Duchenne's muscular dystrophy. J Pediatr Orthop. 1997;17(1):109–14.
30. Johnsen SD. Prednisone therapy in Becker's muscular dystrophy. J Child Neurol. 2001; 16(11):870–1.
31. Lovell WW, Weinstein SL, Flynn JM. Lovell and Winter's pediatric orthopaedics. 7th ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2014.
32. Herring JA, Tachdjian MO. Texas Scottish Rite Hospital for Children. Tachdjian's pediatric orthopaedics. 4th ed. Philadelphia: Saunders/Elsevier; 2008.
33. Shapiro F, Specht L. The diagnosis and orthopaedic treatment of inherited muscular diseases of childhood. J Bone Joint Surg Am. 1993;75(3):439–54.
34. Daher YH, Lonstein JE, Winter RB, Bradford DS. Spinal deformities in patients with muscular dystrophy other than Duchenne. A review of 11 patients having surgical treatment. Spine. 1985;10(7):614–7.
35. Andersen SP, Sveen ML, Hansen RS, et al. Creatine kinase response to high-intensity aerobic exercise in adult-onset muscular dystrophy. Muscle Nerve. 2013;48(6):897–901.
36. Olsen DB, Orngreen MC, Vissing J. Aerobic training improves exercise performance in facioscapulohumeral muscular dystrophy. Neurology. 2005;64(6):1064–6.

37. Bakhtiary AH, Phoenix J, Edwards RH, Frostick SP. The effect of motor learning in facioscapulohumeral muscular dystrophy patients. Eur J Appl Physiol. 2000;83(6):551–8.
38. Pasotti S, Magnani B, Longa E, et al. An integrated approach in a case of facioscapulohumeral dystrophy. BMC Musculoskelet Disord. 2014;15:155.
39. D'Antona G, Ragni M, Cardile A, et al. Branched-chain amino acid supplementation promotes survival and supports cardiac and skeletal muscle mitochondrial biogenesis in middle-aged mice. Cell Metab. 2010;12(4):362–72.
40. Tarnopolsky M, Zimmer A, Paikin J, et al. Creatine monohydrate and conjugated linoleic acid improve strength and body composition following resistance exercise in older adults. PLoS One. 2007;2(10):e991.
41. Tarnopolsky MA. Creatine as a therapeutic strategy for myopathies. Amino Acids. 2011;40(5):1397–407.
42. Kissel JT, McDermott MP, Natarajan R, et al. Pilot trial of albuterol in facioscapulohumeral muscular dystrophy. FSH-DY Group. Neurology. 1998;50(5):1402–6.
43. Kissel JT, McDermott MP, Mendell JR, et al. Randomized, double-blind, placebo-controlled trial of albuterol in facioscapulohumeral dystrophy. Neurology. 2001;57(8):1434–40.
44. Giannini S, Ceccarelli F, Faldini C, Pagkrati S, Merlini L. Scapulopexy of winged scapula secondary to facioscapulohumeral muscular dystrophy. Clin Orthop Relat Res. 2006;
449:288–94.
45. Ketenjian AY. Scapulocostal stabilization for scapular winging in facioscapulohumeral muscular dystrophy. J Bone Joint Surg Am. 1978;60(4):476–80.
46. Bunch WH, Siegel IM. Scapulothoracic arthrodesis in facioscapulohumeral muscular dystrophy. Review of seventeen procedures with three to twenty-one-year follow-up. J Bone Joint Surg Am. 1993;75(3):372–6.
47. Cooney AD, Gill I, Stuart PR. The outcome of scapulothoracic arthrodesis using cerclage wires, plates, and allograft for facioscapulohumeral dystrophy. J Shoulder Elbow Surg. 2014; 23(1):e8–13.
48. Demirhan M, Uysal O, Atalar AC, Kilicoglu O, Serdaroglu P. Scapulothoracic arthrodesis in facioscapulohumeral dystrophy with multifilament cable. Clin Orthop Relat Res. 2009;467(8):2090–7.
49. Goel DP, Romanowski JR, Shi LL, Warner JJ. Scapulothoracic fusion: outcomes and complications. J Shoulder Elbow Surg. 2014;23(4):542–7.
50. Jakab E, Gledhill RB. Simplified technique for scapulocostal fusion in facioscapulohumeral dystrophy. J Pediatr Orthop. 1993;13(6):749–51.
51. Kocialkowski A, Frostick SP, Wallace WA. One-stage bilateral thoracoscapular fusion using allografts. A case report. Clin Orthop Relat Res. 1991;273:264–7.
52. Krishnan SG, Hawkins RJ, Michelotti JD, Litchfield R, Willis RB, Kim YK. Scapulothoracic arthrodesis: indications, technique, and results. Clin Orthop Relat Res. 2005;435:126–33.
53. Letournel E, Fardeau M, Lytle JO, Serrault M, Gosselin RA. Scapulothoracic arthrodesis for patients who have facioscapulohumeral muscular dystrophy. J Bone Joint Surg Am. 1990; 72(1):78–84.
54. Rhee YG, Ha JH. Long-term results of scapulothoracic arthrodesis of facioscapulohumeral muscular dystrophy. J Shoulder Elbow Surg. 2006;15(4):445–50.
55. Twyman RS, Harper GD, Edgar MA. Thoracoscapular fusion in facioscapulohumeral dystrophy: clinical review of a new surgical method. J Shoulder Elbow Surg. 1996;5(3):201–5.
56. Birch JG. Orthopedic management of neuromuscular disorders in children. Semin Pediatr Neurol. 1998;5(2):78–91.
57. Mackenzie WG, Riddle EC, Earley JL, Sawatzky BJ. A neurovascular complication after scapulothoracic arthrodesis. Clin Orthop Relat Res. 2003;408:157–61.
58. Berne D, Laude F, Laporte C, Fardeau M, Saillant G. Scapulothoracic arthrodesis in facioscapulohumeral muscular dystrophy. Clin Orthop Relat Res. 2003;409:106–13.
59. Wolfe GI, Young PK, Nations SP, Burkhead WZ, McVey AL, Barohn RJ. Brachial plexopathy following thoracoscapular fusion in facioscapulohumeral muscular dystrophy. Neurology. 2005;64(3):572–3.

60. Bailey RO, Marzulo DC, Hans MB. Infantile facioscapulohumeral muscular dystrophy: new observations. Acta Neurol Scand. 1986;74(1):51–8.
61. Brouwer OF, Padberg GW, Wijmenga C, Frants RR. Facioscapulohumeral muscular dystrophy in early childhood. Arch Neurol. 1994;51(4):387–94.
62. Korf BR, Bresnan MJ, Shapiro F, Sotrel A, Abroms IF. Facioscapulohumeral dystrophy presenting in infancy with facial diplegia and sensorineural deafness. Ann Neurol. 1985;17(5): 513–6.
63. Shapiro F, Specht L, Korf BR. Locomotor problems in infantile facioscapulohumeral muscular dystrophy. Retrospective study of 9 patients. Acta Orthop Scand. 1991;62(4):367–71.
64. Narayanaswami P, Weiss M, Selcen D, et al. Evidence-based guideline summary: diagnosis and treatment of limb-girdle and distal dystrophies: report of the guideline development subcommittee of the American Academy of Neurology and the practice issues review panel of the American Association of Neuromuscular & Electrodiagnostic Medicine. Neurology. 2014; 83(16):1453–63.
65. Wicklund MP, Kissel JT. The limb-girdle muscular dystrophies. Neurol Clin. 2014;32(3):729–49. 3.