



Arthroplasty in patients with rare conditions

Total Hip Arthroplasty in a Patient With Fascioscapulohumeral Dystrophy

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ABSTRACT

Fascioscapulohumeral dystrophy is an autosomal dominant disorder that results in progressive muscle weakness. Patients most commonly present with facial and periscapular muscle weakness, which progresses to involve their upper and lower extremities as well as truncal muscles. We present a patient with fascioscapulohumeral dystrophy who underwent staged bilateral total hip arthroplasties but developed late prosthetic joint infection. This case also reports the management of periprosthetic joint infection after total hip arthroplasties through explant and placement of an articulating spacer as well as both neuraxial and general anesthetic management for this uncommon neuromuscular disorder.

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Introduction

Fascioscapulohumeral dystrophy (FSHD) is the third most common form of muscular dystrophy [1] and usually presents in late adolescence or adulthood. FSHD typically presents with facial and periscapular muscle weakness, progressing caudally to upper arms, trunk muscles, and lower extremities [2]. Life expectancy is generally unaffected since life-threatening bulbar, respiratory, and cardiac involvement is rare.

FSHD is most commonly transmitted in an autosomal dominant manner. The pathophysiology relates to contraction of the usual number of D4Z4 repeats in a section of the 4q35 chromosome. Each D4Z4 repeat codes for a gene called *DUX4* which is important in fetal development but condenses and inactivates once fetal development is completed. The contraction in the number of D4Z4 repeats decreases the inactivation of the *DUX4* gene and leads to aberrant production of the *DUX4* protein, which can cause muscle toxicity. Due to increased cell apoptosis, serum creatinine kinase can be 3 to 5 times the upper limit of normal [1]. Muscle biopsy shows nonspecific chronic myopathic changes with mononuclear

inflammatory reaction. FSHD is diagnosed via genetic testing, assessing the number of D4Z4 repeats [1].

While respiratory involvement occurs in less than 10% of the FSHD population, surgery in these patients is challenging given their atypical muscle weakness. There is often marked weakness in abdominal and paraspinal musculature leading to significant lumbar lordosis or camptocormia (bent spine syndrome). Scapulothoracic arthrodesis has been described to improve shoulder function [3].

This case report aims to portray the experience and outcome of treating a patient with FSHD who underwent bilateral asynchronous total hip arthroplasty (THA) through a direct anterior (DA) approach, complicated by periprosthetic joint infection (PJI). To the best of our knowledge, this is the first report of a THA in a patient with FSHD.

Case history

Informed, written consent was obtained from the patient for inclusion in this case report.

A 57-year-old female, diagnosed with FSHD at age 53 via genetic testing with 8 D4Z4 repeats, presented to the joint reconstruction clinic at a tertiary care academic medical center with complaints of bilateral, left greater than right, hip pain. Her surgical history was significant for multiple failed spinal surgeries, complicated by hardware failure and delayed wound healing. The patient's prior

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spinal hardware failure was believed to be the result of significant fatty infiltration and atrophy of the patients paraspinal muscles [4]. At initial presentation, she had limited household ambulatory ability with a rolling walker and was using a wheelchair for longer-distance ambulation. She was noted to have 4/5 strength diffusely in her left lower extremity and had limited range of motion in the left hip. Radiographs upon presentation showed severe left hip osteoarthritis. She underwent a course of physical therapy and was treated with oral anti-inflammatory medication and several intra-articular guided steroid injections to the left hip which led to significant, but transient, pain relief.

Approximately 18 months after the initial evaluation, she returned to clinic for reevaluation of her hip pain, which was now refractory to conservative measures. Her ambulatory status remained stable with ability to ambulate short distances in her house utilizing a walker despite camptocormia. She weighed 93 kgs with a height of 165.1 cm and body mass index of 32. She reported persistent low back pain at the time of evaluation, but the pain in her hips was more limiting. Radiographs showed progression of her bilateral hip arthritis (Fig. 1). The patient elected to undergo left THA. Preoperative workup, including nutritional screening, was within normal limits. The treating surgeon performs primary THA through both DA and posterolateral surgical approaches; in this case, the DA approach was felt to be advantageous for this patient given concerns about muscle weakness and abnormal spinopelvic kinematics.

Given the potential for underlying respiratory involvement as well as her small stature and camptocormia, spinal anesthesia was selected to avoid possible use of intravenous neuromuscular blockade, further limiting respiratory mechanics. Due to prior spinal fusion at L4-L5, the L3-L4 level was selected, and 2.5 mL of 0.5% isobaric bupivacaine was administered. Standard antibiotic prophylaxis as well as 1 g of intravenous tranexamic acid were also administered prior to incision. The patient's bone quality and muscle tissue did not appear compromised during the procedure. However, given her limited baseline mobility, there was significant lymphedema in the subcutaneous adipose tissue. Bone quality appeared appropriate for cementless components. 2-Octyl cyanoacrylate skin closure system (Prineo; Ethicon, Somerville, NJ) was used to reinforce skin closure. There were no intraoperative complications; the patient was discharged home on postoperative day 2.



Figure 1. Anteroposterior (AP) pelvis radiograph demonstrating bilateral hip osteoarthritis.

At her initial postoperative visit from the left THA, she was noted to have a small area of delayed wound healing at the proximal incision near the fold in the pannus without active drainage; the incision ultimately healed without additional intervention by 10 weeks postoperatively, at which time her pain was mostly resolved. Inflammatory markers were normal, and she elected to move forward with right THA. Five months after her left THA, she underwent an uncomplicated right THA through identical conditions; a DA approach under spinal anesthesia using 2.5 mL of 0.5% isobaric bupivacaine. Her hospital course this time was complicated by development of allergic contact dermatitis around the right hip incision attributed to the 2-octyl cyanoacrylate dressing system, treated with removal of the covering and topical application of corticosteroid cream. Following the patient's right hip procedure, she experienced global progressive weakness and difficulty mobilizing postoperatively but ultimately was discharged home on postoperative day 5. Postoperatively she again developed delayed wound closure which eventually healed without additional surgical intervention.

Seven months after her left THA and 2 months after her right THA, she developed increasing pain with motion in the left thigh region attributed to her muscular dystrophy; radiographs showed unchanged component positioning (Fig. 2), and the patient completed a course of physical therapy.

The patient was temporarily lost to follow-up but represented with continued left hip and thigh pain, especially with ambulation, and right hip pain in the region of the greater trochanter. Radiographs demonstrated gross loosening of the left acetabular component (Fig. 3); inflammatory markers showed a normal sedimentation rate (25; normal range 0-30) and slightly elevated C-reactive protein (0.85 mg/dL; normal range <0.5 mg/dL). Hip aspiration was completed, notable for synovial fluid cell count of 9983 cells composed of 95% neutrophils without growth on aerobic or anaerobic culture medium. Based off criteria from the 2018 Musculoskeletal Infection Society, the patient had possible PJI, and left revision THA was recommended.

Despite the presumed elevated risk of pulmonary complications, general endotracheal anesthesia was selected due to the anticipated prolonged length of surgery. The hip was approached this time using a standard posterolateral incision with the patient in the right lateral decubitus position. Positioning of upper extremities was challenging due to shoulder girdle weakness. Proper positioning required the use



Figure 2. AP pelvis radiograph obtained 7 months after left THA and 2 months after right THA demonstrating unchanged component positioning.



Figure 3. AP pelvis radiograph demonstrating loosening of the left acetabular component.

of additional pillows, pegs in the peg board, and tape. These supplementary items were used to support the shoulder girdle and prevent anterior collapse of the left shoulder. Intraoperatively, at the time of revision left THA, purulent fluid was noted in the left hip. Two of 5 histology sections were positive for acute inflammation with repeat fluid cell count showing 46,100 nucleated cells and 87% neutrophils. Therefore, diagnosis of chronic PJI was confirmed, and we proceeded with explant of all components. The acetabular component was grossly loose with Paprosky 2B bone loss; the femoral component was well-fixed circumferentially and required extended trochanteric osteotomy for removal. A lateralized polyethylene liner was cemented into the acetabulum using medium-viscosity cement containing 2 g of vancomycin and 2.4 g of tobramycin. After placement of cerclage cables around the extended trochanteric osteotomy site, a 200-mm Prostalac hip stem (DePuy, Warsaw, IN) coated with the same concentration of antibiotics in medium-viscosity cement was implanted (Fig. 4). Postoperatively, a minimally displaced fracture of the greater trochanter was noted on radiographs.

Intraoperative cultures were positive for *Staphylococcus hominis*. After consultation with the infectious disease team, the patient was treated with a 6-week course of intravenous cefazolin. Postoperatively her weight-bearing was restricted to 30 pounds on the left lower extremity with posterior hip precautions for 6 weeks. She was discharged to inpatient rehabilitation and then ultimately to home. Following her intravenous antibiotic therapy, inflammatory markers were downtrending; therefore, she was transitioned to oral cephalexin at the recommendation of the infectious disease consultant and remains on cephalexin twice daily for chronic suppression.

Ten months after removal of her left THA and placement of an articulating spacer, she is advancing her activity as tolerated and ambulating with the use of a walker around her home. Given her high risk of anesthesia complications, due to generalized weakness combined with low metabolic equivalents, the articulating spacer is planned to be used as definitive fixation unless loosening were to develop.

Patient perspective

In discussion with the patient, her preoperative hip pain improved significantly postoperatively. Following each procedure, she expressed that her weakness diffusely worsened, although temporarily. She continues to use a wheelchair for mobility except for short distances where she will ambulate with the use of a walker. Her use of a walker continues to be limited by her bilateral upper-extremity

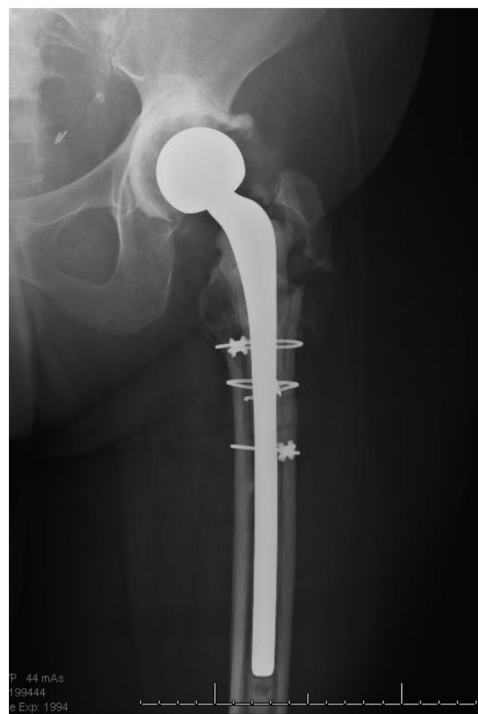


Figure 4. AP radiographs of the left hip after removal of components and placement of a cemented acetabular liner and Prostalac hip stem. Interval healing of extended greater trochanter osteotomy and interval displacement of greater trochanter fracture.

weakness; thus, she hopes to maintain the spacer as a definitive management of her left hip. She reports that her quality of life has improved postoperatively through reduction in pain but wishes that she would be able to ambulate without the use of mobility aids.

Discussion

This study reports a case of FSHD who underwent bilateral, asynchronous THA for severe hip degenerative joint disease. To our knowledge this is the first report of THA in a patient with FSHD. Indications for THA in patients with neuromuscular imbalance are primarily for pain, followed by functional impairment [5]. Good outcomes after THA have been reported in the literature in both ambulators and wheelchair users [6].

In this case, a DA approach was selected due to its application of true internervous and intermuscular planes [7,8]. The use of internervous intervals may allow for earlier restoration of gait and decreased risk of dislocation [6]. The DA approach also avoids abductor insufficiency that can occur following a lateral approach for THA [9]. Analysis of myofibrils is mostly limited to Duchenne-type dystrophy, but patients with muscular dystrophy have decreased capacity for myocyte recovery as the breakdown of myofibrils cannot be fully compensated for by satellite cell proliferation [10]. As a result, muscle inflammation leads to necrosis with fibrotic remodeling and fatty cell replacement within muscle tissues [4,10], something that had been reportedly noted at one of the patient's lumbar spine procedures. In cases where a DA approach was used, creatinine kinase levels have been found to be lower than those in cases where a posterior approach was used [7]. In FSHD, the development of camptocormia is common, which may result in abnormal spinopelvic kinematics; this patient also had a prior attempt at lumbar spinal fusion. Anterior-based surgical approaches to the hip may mitigate some risk in patients with spinopelvic stiffness [11].

The patient in this report developed several postoperative complications. Most significantly, she developed a chronic left-hip PJI and had delayed wound healing on both hips. The source of the patient's deep prosthetic hip joint infection was unknown; delayed wound healing after THA may be a risk factor for PJI [12] as it can provide a route for infection; the patient had a history of delayed wound healing after previous spinal surgery. The patient did not have other risk factors for infection, but given the lack of bony ingrowth to the acetabular component, the infection may have been present since the index operation. In the setting of infection, component loosening results from the release of osteolytic factors by macrophages leading to bone resorption mediated by RANKL [12]. Delayed wound healing has been reported to be a higher risk condition with DA surgical approaches in some papers, [13,14] but others show equivalent or lower rates [13,15] than the posterior approach. In this patient, the DA approach was chosen despite concern for slow wound healing given concerns about the patient's risk of instability from her abnormal spinopelvic kinematics and concern about the potential for muscle healing in approaches that do not utilize an intramuscular plane. Poor wound healing and increased risk of infection are not typical features of FSHD. In the thymus, Duxbl, another DUX4 homolog seen in mice, plays a role in elimination of T cells that fail to rearrange beta-chains although the impact on maturation of the immune system is unknown [16]. While 2-stage exchange arthroplasty remains the gold standard for management of PJI, Wang et al. reported that nearly 20% of patients never received their intended replantation as part of a 2-stage revision [17]. Of the 20%, 26% of patients declined reimplantation because they did well with their retained spacer and were unwilling to proceed with reimplantation, as in the case of this patient. The final notable postoperative complication, allergic contact dermatitis to 2-octyl cyanoacrylate, has not been previously reported [18] in the FSHD population but has been previously noted after exposure; it is unclear whether FSHD is associated with increased risk of allergic contact dermatitis.

The literature remains limited regarding anesthetic management of patients with FSHD. In 2 case reports of anesthetic management of patients with FSHD, both case presentations had patients portray similar sensitivity to the general population to nondepolarizing neuromuscular blocking agents; however, each experienced a much faster recovery [19,20]. Recovery time in both studies was measured by time to recovery of the first twitch of the train-of-four ratio. Patients with FSHD who are wheelchair-

dependent and have severe proximal weakness are, however, at increased risk of hypoventilation [1]. Solakovic compared the height of sensory block with both hyperbaric and isobaric bupivacaine [21] and recorded each respective peak height after spinal anesthesia administration. The most frequent highest sensory level with hyperbaric bupivacaine was at the T5 level while isobaric bupivacaine remained at the T10 level. They concluded that hyperbaric bupivacaine can give an unnecessarily high block that can influence hemodynamic status. Our initial spinal anesthetic approaches using isobaric bupivacaine in this patient were to obtain an effective low(er) thoracic level in an attempt to avoid interfering further with the patient's respiratory mechanics. In this patient, both primary THAs using spinal anesthesia with isobaric bupivacaine as well as a prolonged revision THA under general endotracheal anesthesia with intravenous nondepolarizing neuromuscular agents were demonstrated successfully.

Current controversies and future considerations

Further considerations in preoperative and intraoperative decision-making for THA in patients with FSHD include the use of constrained implants. In patients with severe muscle weakness, intraoperative assessment of stability is essential. Some studies have reported increased risk of dislocation in patients with neuromuscular disorders [22]. With the introduction of highly crosslinked polyethylene liners, larger femoral heads show reduced risk of dislocation [23]. Dual-mobility acetabular component or constrained acetabular liner can reduce instability. For patients where muscle weakness is so pronounced that the arthroplasty is unstable, a constrained device may be necessary to provide stability.

Summary

This case report describes a, now 62-year-old, female patient with FSHD who underwent bilateral, asynchronous THAs. Although this report is limited to 1 patient with FSHD, we demonstrate that THA may lead to a reduction in pain symptoms; however, improvements in ability to ambulate may not be realized, and these patients may be at higher risk of surgical complications. This case also reports management of PJI after THA in a patient with FSHD, through explant and placement of an articulating spacer, as well as successful anesthetic management of both neuraxial and general endotracheal anesthesia with paralyzing agents. Further studies are needed to show the long-term outcomes of primary and revision THA in this patient population.

Conflicts of interest

For full disclosure statements refer to <https://doi.org/10.1016/j.artd.2022.08.014>.

Informed patient consent

The author(s) confirm that written informed consent has been obtained from the involved patient(s) or if appropriate from the parent, guardian, power of attorney of the involved patient(s); and, they have given approval for this information to be published in this case report (series).

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KEY POINTS

1. Facioscapulohumeral dystrophy is a form of muscular dystrophy that most commonly presents with facial and periscapular muscle weakness and then progresses to involve upper arms, trunk muscles, and lower extremities.
2. Total hip arthroplasty (THA) is a viable option for facioscapulohumeral dystrophy patients with hip osteoarthritis.
3. Indications for THA in patients with muscular dystrophy is primarily for pain, followed by functional impairment.
4. In patients with muscular dystrophy, spinal anesthesia provides adequate neuraxial anesthesia for THA avoiding potential use of intravenous neuromuscular blocking agents.
5. Low-demand patients who develop periprosthetic joint infection can be managed with explant, placement of an articulating spacer, and targeted antibiotics.

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