

## Repair of an inguinoscrotal hernia in a patient with Becker muscular dystrophy

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**SUMMARY: Repair of an inguinoscrotal hernia in a patient with Becker muscular dystrophy.**

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**Introduction.** Inguinal hernia repairs are routinely performed as outpatient procedures in most patients, whereas a few require admission due to clinical or social peculiarities. Muscular dystrophies are inherited disorders characterized by progressive muscle wasting and weakness. In case of surgery there is no definite recommendation for either general or regional anesthesia.

**Case report.** This contribution regards a 48 y. o. male patient diagnosed with Becker Muscular Dystrophy by muscle biopsy 10 years earlier. He had a left-sided sizable inguinoscrotal hernia with repeat episodes of incarceration. An elective mesh repair with suction drainage

was accomplished under selective spinal anesthesia. The post-operative course was uneventful.

**Discussion.** A few inguinal hernia repairs require admission due to peculiarities such as extensive scrotal hernias requiring suction drainage. Muscular dystrophies are inherited disorders with no cure and no two dystrophy patients are exactly alike, therefore the health issues will be different for each individual. In case of surgery there is no definite recommendation for either general or regional anesthesia. This contribution regards the successful elective mesh repair with suction drainage of a large left-sided inguino-scrotal hernia in a 48 y. o. male patient affected by Becker muscular dystrophy by selective spinal anesthesia obtained by 10 milligrams of hyperbaric bupivacaine.

**Conclusion.** Effective mesh repair with suction drainage of large inguinal hernias under spinal anesthesia can be achieved in patients affected by muscular dystrophy.

KEY WORDS: Inguinoscrotal hernia - Becker muscular dystrophy - Mesh repair - Spinal anesthesia.

### Introduction

Inguinal hernia repairs are routinely performed as outpatient procedures in most patients who can be discharged within few hours, whereas less than 5% of cases require admission due to peculiarities such as extensive scrotal hernias requiring suction drainage, severe comorbidities, or hometown more than one hour's drive away (1). Muscular dystrophies are inherited disorders characterized by progressive muscle wasting and weakness of variable distribution and severity. On the basis of distribution of predominant muscle weakness six major forms can be delineated, with the addition of congenital dystrophy where muscle weakness is more generalized. There is no cure for any of the dystrophies (2). No

two dystrophy patients are exactly alike, therefore the health issues will be different for each individual (3). In case of surgery there is no definite recommendation for either general or regional anesthesia. There are few reports of spinal, epidural and caudal anesthesia without any complication. Post-operative care needs focus on avoidance of prolonged immobilization, as the resulting muscular atrophy worsens disease. Ambulatory anesthesia should only be chosen for low risk surgery in early stages, where no cardiopulmonary involvement is demonstrated (4). This contribution regards the successful elective mesh repair with suction drainage of a large left-sided inguinoscrotal hernia in a 48 y. o. male patient diagnosed with Becker muscular dystrophy by muscle biopsy 10 years earlier.

### Case report

The patient had been noticing a soft lump in his left groin about 5 years before the neurologic diagnosis. Over the last 10 years the hernia had been descending into the

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scrotum causing chronic pain, exacerbated by his obvious difficulties in physical activities, and repeat episodes of incarceration that were resolved by bed rest and manipulation of the hernia. On physical exam the patient appeared in good general condition with macroglossia and hyperlordosis of his lumbar spine. His limbs appeared hypotrophic, with slight bilateral calf hypertrophy. The gluteal regions were thinned out. His gait was typical: toe walking, waddling and leaning his body toward the other side to balance the center of gravity. While lying down on the examination bed his lower limbs were kept slightly elevated with flexed thighs and legs due to knee retractions and clubfoot. When asked to leave the bed he first had to carefully sit up pushing down on his thighs. His chest examination revealed a II/VI proto-mesosystolic murmur on the left apical area and clear breath sounds bilaterally. His abdomen was soft and nontender. The left inguinal region was occupied by an obvious hernia descending down into the scrotum that was very tender on palpation and could not be reduced back into the abdominal cavity. His pre-operative work-up included an EKG showing first-degree block and left ventricle hypertrophy, echocardiography showing thickened aortic and mitral valve flaps, left ventricle hypertrophy, an ejection fraction of 50%, an unremarkable chest X-ray and a neurologic approval of the planned surgery by his neurologist. The following abnormal laboratory exams were noticed: creatine phosphokinase 718 U/L, hematocrit 51.8%, hemoglobin 18.1 g/dl. The operation was performed under selective spinal anesthesia obtained by 10 milligrams of hyperbaric bupivacaine through a 27 G Sprotte needle keeping the patient in a left lateral position for 10 minutes. A total of 3 milligrams of midazolam, 40 milligrams of omeprazole and 30 milligrams of ketorolac were also injected i.v. The hernia sac was carefully dissected off the spermatic cord and could not be reduced intact so it was cut open. A sizable amount of greater omentum was found partially fixed to the sac wall and descending into the scrotum (Fig. 1). After excision of the herniated omentum measuring 24x13x6 cm the sac was also partially excised, suture ligated with absorbable running suture and reduced back into the abdominal cavity. The inguinal canal was repaired in a modified Lichtenstein fashion with polypropylene mesh and sutures (Fig. 2) (5). After closing the inguinal canal a cutaneous counter-incision in the left lower quadrant was



Figure 1 - Incarcerated omentum prior to resection.

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Figure 2 - Mesh repair.

done and a suction drainage was placed on top of the sutured external oblique aponeurosis with its tip lying down in the scrotum. The operation took 95 minutes. A Foley catheter was positioned before the patient left the operating room. The post-operative course was uneventful. The Foley catheter was removed on post-operative day 1 when ambulation was allowed, the drainage on day 2 (total output 100 cc of sero-sanguineous fluid). The patient was discharged to home on the 3<sup>rd</sup> post-operative day. 30 days after discharge the surgical wound was healing well, a small non tender solid mass was palpable at the root of the left side of his scrotum compatible with regular post-operative changes, the patient was back to his job as hospital nurse in another facility.

## Discussion

Muscular dystrophies are inherited disorders characterized by progressive muscle wasting and weakness of variable distribution and severity. On the basis of distribution of predominant muscle weakness six major forms can be delineated, with the addition of congenital dystrophy where muscle weakness is more generalized: Duchenne and Becker muscular dystrophy (DMD and BMD), Emery-Dreifuss, distal, facioscapulohumeral, oculopharyngeal, limb-girdle. DMD's onset is in early childhood, with difficulties in running and, later, climbing stairs. Weakness is mainly proximal and progressive and results in Gower's maneuver due to weak knee and hip extensors: the patient climbs up his thighs, pushing down on them, to extend the hips and trunk. Most patients have enlarged calves, "pseudohypertrophy". Ultimately a wheelchair becomes necessary in most cases by age 12 and pneumonia compounded by cardiac involvement is the most frequent cause of death in the late teens or early 20s. In BMD the distribution of muscle wasting is closely similar to that in DMD, but the course of the disease is more benign. The Duchenne gene is located at Xp21, which affects the sarcolemmal protein dystrophin, and is allelic with BMD. Dystrophin is usually absent in DMD but is reduced in amount or abnormal in size in BMD. Diagnosis is obtained by elevated creatine kinase levels, electromyography that demonstrates the myopathic nature of dystrophy and rules out neurogenic causes, and most importantly by muscle histology, that shows variations in fiber size, fiber necrosis, invasion by macrophages and replacement by fatty and connective tissue. The pathophysiology of these disorders is related to altered interaction between the sarcolemma, sarcolemma-associated proteins such as dystrophin, and calcium channels. There is no cure for any of the dystrophies, emphasis is on respiratory care and treatment of cardiologic complications. Surgery may be indicated

in later stages for correction of contractures in order to prolong ambulation or for scoliosis correction to preserve lung function but is not universally accepted because of anesthetic risks and the post-operative bed-rest that might be detrimental (2). No two dystrophy patients are exactly alike, therefore the health issues will be different for each individual (3). In case of surgery there is no definite recommendation for either general or regional anesthesia. Succinylcholine and volatile anesthetics are best avoided because there is a risk of hyperkalemic cardiac arrest or severe rhabdomyolysis. General anesthesia has to be performed as total intravenous anesthesia. Nitrous oxide can be used, but should be avoided in case of manifest cardiac involvement. Opiates and propofol, at times with higher doses, and local anesthetics have been used without complications. There is one report of higher toxicity of paracetamol in patients with DMD. There are few reports of spinal, epidural and caudal anesthesia without complications. In young patients with an early stage of the disease there is no contraindication for sedation, always considering the possible onset of respiratory failure. In patients with advanced stages of the disease (cardiopulmonary involvement, pharyngeal muscle weakness, loss of ambulation) sedation should only be done after carefully calculating the individual risks, especially with respect to respiratory failure and risk of aspiration. Body temperature must be monitored to avoid shivering and increased oxygen demand. Pre-operative cardiomyopathy must be searched by electrocardiography and echocardiography. Lung function test including lung volumes and blood gas analysis should be done to evaluate grade of pulmonary involvement, even though no clear correlation has been shown between lung function and post-operative respiratory complications. Creatine kinase level is usually very high, shows no correlation with disease severity, and is checked pre-operatively only to obtain a baseline level in case of post-operative rhabdomyolysis. If muscular weakness is present and regional anesthesia is planned neurological consultation is helpful for judicial reasons. Effective airway management must be carefully planned because macroglossia is frequent and there may be swallowing difficulties with higher risk of aspiration. Invasive surgery may be associated with higher blood transfusion requirement. A higher risk of post-operative thrombosis may be suspected in advanced stage patients. Post-operative care needs focus on avoidance of prolonged immobilization, as the resulting muscular atrophy worsens disease. Ambulatory anesthesia should only be chosen for low risk surgery in early stages, where no cardiopulmonary involvement is demonstrated (4). The successful repair of our patient's large inguino-scrotal hernia could be accomplished by a few well planned steps. Careful pre-operative work-up was carried out, intra-operative scrupulous surgical and anesthetic te-

chniques were adopted, early post-operative mobilization was stimulated.

## **Conclusion**

It is generally considered that muscular dystrophy pa-

tients usually present health issues that will be different for each individual and when surgery is planned no definite recommendation for either general or regional anesthesia exists. This contribution shows that effective mesh repair with suction drainage of large inguinal hernias under spinal anesthesia can be achieved in patients affected by muscular dystrophy.

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