



Surgical Decision-Making in Spinal Instability in Facioscapulohumeral Muscular Dystrophy Related with a Spinal Muscle Atrophy

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The indication and treatment of degenerative spinal pathology are specific to the preparation of the neurosurgeon specialist. However, there is a general lack of information on the possible surgical indication in patients suffering from rare genetic pathologies, such as myopathies, which can determine spinal deformities in adulthood. Facioscapulohumeral muscular dystrophy (FSHD) represents one of the most common genetic myopathies characterized by weakness of the facial and periscapular musculature¹.

FSHD is characterized by the adipose infiltration of the face, shoulder, trunk, and limb muscles.^{2,3} While scapular and facial muscular degeneration is well-documented, few authors described the adipose degeneration that occurs in the paraspinal muscles of patients with FSHD,⁴ resulting in a pelvic imbalance that can cause different spinal deformities³ like coronal and sagittal plane scoliosis and severe spondylolisthesis. Pure sagittal deformities such as lumbar hyperlordosis or flat back are relatively rare.⁵ In such a case, surgical correction is often considered. However, extensive traditional open spinal approaches may promote further trauma to the posterior spinal structures, and posterior spinal muscle, already impaired by atrophic degenerative changes. While several cases reported corrective surgical results on patients with Duchenne muscular dystrophy, cerebral palsy, and spinal muscular atrophy, few authors investigated the surgical results of milder but potentially inadequate, degenerative forms in terms of outcomes in

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the case of FSHD patients. Minimally invasive spinal surgery, especially the extreme lateral interbody fusion (XLIF) approach, could minimize the surgical trauma, providing fair radiological and clinical outcomes, but was nevertheless never previously reported for the management of FSHD patients.

Whenever possible, a lateral fusion approach was combined with percutaneous pedicle screw fixation,^{4,5} which not only minimizes unnecessary trauma to the atrophic spinal muscles but also provides excellent radiological results and adequate indirect decompression. In our experience, surgically-treated FSHD patients showed improvement in posture and quality of life.

Before proceeding to a surgical procedure, a complete understanding of the etiology of the focal or diffuse deformities and the compensatory mechanism is mandatory.⁵ In the case of spinal muscle involvement in the natural history of a FSHD patient, spinal complications ranging from a single-level degenerative disc disease to a severe global spinal deformity can occur, and surgery should be seriously pondered for radiologically confirmed, progressively worsening spinal disorders, even in earlier stages, before deterioration of heart and lung function would possibly increase the probability of major postoperative complications.

Conflict of Interest

None declared.

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