

A RARE OPERATIVE APPROACH TO A WORSENING SCOLIOSIS IN A FACIOSCAPULAHUMERAL MUSCULAR DYSTROPHY ADOLESCENT

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Introduction: Troubled by a rare occurrence of a rare muscular dystrophy disorder, a progressive worsening Lumbar curvature scoliosis leads this patient for a corrective Scoliosis operation. Effecting both male and female on an equal scale, a Facioscapulohumeral dystrophy is a rare disorder with prevalence of less than 10 per 100,000 people. An abnormal DUX4 gene at the DAZ4 region of Chromosome 4, is the most common cause of the Facioscapulohumeral Dystrophy. This abnormality of fewer methyl group, leading to production of DUX4 protein in cells and tissue, causes a progressive worsening of power and atrophy of the muscle.

Discussion: This case reports the battle of a young adolescent girl with a progressive worsening of the Cobs Angle of her single curve Lumbar Scoliosis. Diagnosed at an earlier adolescent age clinically and confirmed by a genetic study, she had a slow progression of the muscle weakness limiting the range of motion of her bilateral upper limb. Although she can manage to initiate the flexion, extension and abductions as well as adduction of the shoulder joint, she is unable to carry them above 90 degrees. This limits her overhead activities and relies on help of family members to manage herself. An otherwise school-attending adolescent, she has been followed up by the Neuromedical Team and recently been referred for a progressive worsening lower lumbar discomfort and worsening curve. An outpatient follow-up of her Lumbar curve has brought our attention to a worsening Scoliosis condition concurrent with her growth spur. A worsening curve from a conservative 24 degrees to a worrying 50 degrees of a single level lumbar scoliosis curve.

Conclusion: A commonly unilateral weakness disease, this rare case demonstrates bilateral weakness. A hefty discussion of pro's and con's of surgery, has led to a planned correction and stabilisation of the Scoliosis.