Tailor-made management of thoracic scoliosis with cervical hyperextension in muscular dystrophy.

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Abstract

PURPOSE: We report the case of a 13-year-old boy managed for fixed cervical hyperextension due to congenital muscular dystrophy with partial merosin deficiency. He presented a right decompensated thoracic scoliosis (T6-L1 Cobb angle 72°) associated with cervical and lumbar lordosis. The spinal extension was accompanied by major flexion of the hip resulting in the trunk being bent forward. This posture caused daily severe back pain responsible for significant loss of quality of life. This led to the decision to perform surgery.

METHODS: Initially, the surgery was limited to the thoraco-lumbo-sacral area. An anterior release was done, followed by posterior T1-pelvis vertebral fusion using a modified Luque-Galveston technique. The correction achieved was satisfactory in the coronal plane, but the correction of the thoracic kyphosis was insufficient to compensate for the cervical hyperextension. Cervical spine was fixed at 52° of lordosis, and associated with a left 50° rotation and a right 45° inclination of the head. We performed a posterior and lateral release of the cervical muscles followed by positioning of the halo, itself connected to a made-for-measure thoracic corset. A daily adjustment of the threaded rods was done daily for 3 months to correct the cervical position. Then, we performed a spinal fusion without instrumentation, by posterior articular abrasion and grafting from the occiput to T1. Following that, the halo-corset was kept in place for 4 months.

RESULTS: At the end of 8 month treatment, the clinical result was satisfactory with a balanced spine both face on, and sideways, allowing for comfortable painless positioning. At 5 year follow-up, he showed stable spinal fusion without any loss of correction.

CONCLUSION: There is no gold standard treatment for cervical hyperextension, but approaches have to be tailor-made to the patient's needs and the team's experience.

KEYWORDS: Cervical hyperextension; Muscular dystrophy; Neuromuscular scoliosis; Surgery

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