

INDICATIONS FOR SURGICAL TREATMENT OF THORACOLUMBAR KYPHOSIS IN PATIENTS WITH MUCOPOLYSACCHARIDOSIS

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Introduction. Circumferential fusion of kyphosis in patients with MPS is currently accepted as the most effective surgical approach. However, long-term results remain debatable. This study assesses the effectiveness of posterior-only compared to circumferential fusion.

Methods. Eleven patients (7 male, 4 female) with MPS and thoracolumbar kyphosis underwent surgical treatment. Hurler Syndrome (type I) was diagnosed in 5 patients, Morquio Syndrome (type IV) in 2, and Maroto-Lamy (type VI) in 4 patients.

Indications for surgical treatment included more than 40° kyphosis, sagittal spinal imbalance, progressive neurological symptoms and severe pain. In 3 cases, patients underwent circumferential arthrodesis combining anterior and posterior approaches. In 8 cases, instrumentation included hooks and/or pedicular screws, placed two levels above and two levels below the deformity apex. The follow-up period ranged from 2 to 5 years.

Results. In 8 cases solid spinal fusion was achieved. Complications after surgical treatment were observed in 4 patients (36%). PJK developed in one case 2 years after surgery, pseudarthrosis was observed in one case, wound supuration was observed in one case, and a broken metal rod in one case.

Conclusions. Surgical treatment of MPS patients with thoracolumbar kyphosis is accompanied by a high risk of complications when circumferential stabilization is performed. Most authors and our data show that the most optimal method of surgical treatment of thoracolumbar deformation is dorsal correction and fixation in combination with a wide laminectomy at the level of stenosis. The second stage includes the anterior decompression and interbody fusion. However, if the patient's lung function is dramatically compromised, and a high risk of respiratory complications exists, surgery may be limited to only posterior correction and fixation in conjunction with a wide laminectomy, which allows to achieve a comparable level of fixation with a lower risk of complications.

SURGICAL TREATMENT OF SEVERE DEFORMITIES OF CERVICO-THORACIC JUNCTION

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In this study, the outcomes of surgical treatment of severe spinal cervico-thoracic deformities are evaluated. An analysis of 8 patients who underwent surgery between 2012 and 2015 is presented. Mean age was 11,2 years (range from 2 to 18 y.o.). In 6 patients KFS with cervico-thoracic kyphosis was observed. In 2 patients, both 2-year-old girls, there was anterior displacement at the Th1 vertebra, most likely due to congenital dislocation. In 2 cases type I neurofibromatosis was observed. All the patients with KFS presented with neurological deficit: four patients presented with inferior paraparesis and two patients – with tetraparesis due to cervical myelopathy. In all cases staged surgical treatment was performed: halo-traction for 10-14 days, then occipito-cervico-thoracic instrumented fixation as the 2nd stage. In one case, anterior cervical multilevel fusion with autografting was performed as the 3rd stage. In all but one patient full-scale 3D models of the vertebral column at the deformity level was manufactured based on CT-scans. Mean follow-up time was 18,8 months (range 12-36). In all cases, sufficient correction was achieved. In two cases, there was improvement in neurological status. In two cases fractures of one of the 2,5 mm rods in occipito-cervical instrumentation were observed. This condition requires reoperation and additional reinforcing occipito-cervical fixation using cortical peroneal autograft.

Conclusion. Due to the severity and complexity of congenital deformities of the cervico-thoracic junction, full-scale 3-D models are indispensable for understanding anatomical relationships and for surgery planning. Halo-traction is recommended for preoperative correction and neurological complication prevention.