SURGICAL TREATMENT OF CRANIOVERTEBRAL STENOSIS IN PATIENTS WITH MUCOPOLYSACCHARIDOSIS TYPE I, II, AND VI

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Objective. To analyze surgical treatment of craniovertebral stenosis in patients with mucopolysaccharidosis.
Material and Methods. A total of 9 patients (4 men, 5 women) with mucopolysaccharidosis were operated on for craniovertebral stenosis in 2012—2017. All patients underwent clinical examination, standard radiography of the cervical spine with functional tests to assess atlantooccipital stability according to Rothman, and MRI to determine the degree of the spinal cord compression, stenosis of the spinal canal, and changes in soft tissues surrounding the spinal canal due to fibrosis and accumulation of glycosaminoglycans. Maximum stenosis at the C0—C2 level was noted in 5 patients, at C1—C2 in 2, at C2—C4 in 1, and at C0—C1 in 1. Myelopathy in the upper cervical spine was detected in five cases. The overall somatic and functional statuses, as well as neurological parameters were assessed. Follow-up examination and X-ray examination were performed at 3, 6 and 12 months after surgery, and further every 6 months.
Results. The average follow-up period was 2.5 years. The age of the patients varied from 1.8 to 34 years. After 3, 6, and 12 months after surgery, instrumentation was radiologically stable in all cases, there were no bone resorption signs around the elements of instrumentation. Seven patients showed a regression of neurologic symptoms and positive dynamics in the somatic status according to the assessment scales. In one patient, the neurological status remained unchanged. In one case, a temporary deterioration in the condition was noted after a failed intubation attempt, which required emergency tracheostomy, and operation was postponed to a later date to stabilize the condition. Complications after surgical treatment were observed in five patients. In one case, pseudoarthrosis, rod fracture, and instrumentation instability developed 1.5 years after the intervention, and revision surgery was required. Wound healing problems were observed in three patients and a liquorrhea in the early postoperative period, which was stopped without revision intervention — in one case.
Conclusion. Decompression and stabilization in patients with mucopolysaccharidosis should be performed in advance, before any neurological disorder development. Fixation is required to prevent the development of instability, post-laminectomy deformities, and progression of cicatricial stenosis. Timely initiation of enzyme-replacement therapy improves the results of mucopolysaccharidosis treatment.
Key Words: craniovertebral stenosis, mucopolysaccharidosis, glycosaminoglycans, Maroteaux — Lamy syndrome, cervical stenosis, spinal cord compression, surgical treatment.
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Mucopolysaccharidoses (MPS) are severe heterogeneous and progressive diseases caused by a deficiency of lysosomal enzymes involved in the cascade of glucosaminoglycan (GAG) degradation reactions [1].

Progradient accumulation of GAG in cells and tissues of the body leads to a variety of clinical manifestations, which differ among different types of MPS.

All types of MPS, with the exception of type III, are characterized by the envolvement of skeleton and joints, cardiorespiratory diseases, hepatosplenomegaly, hearing loss and visual impairment [2–5].

MPS belongs to the group of orphan diseases. The incidence of type I MPS ranges from 1:40 000 to 1:100 000, of type II, from 1:140 000 to 1:156 000, of type VI, from 1:238 000 to 1:300 000 [6–8].

Spinal stenosis is more common in patients with types I, II, IV and VI MPS at the level of the craniovertebral junction or in the region of the thoracolumbar spine [2, 9–13]. Neurological symptoms (general weakness, gait disturbance, paraparesis, pyramidal symptoms, paresthesias, gastrointestinal and bladder dysfunction, and apnea) develop due to compression of the spinal cord at the level of the craniovertebral junction [14–16]. Lack of treatment for stenosis can ultimately lead to serious complications, up to sudden death [6, 17–21]. Therefore, early diagnosis of spinal stenosis is extremely important to prevent irreversible neurological manifestations. However, patients with airway obstruction, restrictive lung diseases, diseases of the cardiovascular system or their combination have a high risk of complications during anesthesia, including airway obstruction, difficulties or failures during intubation, after extubation, or the need for emergency tracheostomy [21, 22].

Treatment of neurological manifestations involves surgical removal of spinal cord compression in combination with enzyme replacement therapy, if it is available for this type, or prior transplantation of hematopoietic stem cells to patients with MPS IH [23–25]. High anesthetic risk in patients with MPS requires careful assessment of the risk factors for anesthesia prior to the surgery. These

include the type of MPS, airway patency, and cardiorespiratory function prior to the anesthesia.

There are no reports on the results of surgical interventions and features of the development of cervical stenosis in patients with MPS in Russian literature. This can be attributed to the rarity and complexity of the pathology, as well as to the severity of the patient's condition caused by the manifestations of the underlying disease.

The aim of the study is to analyze surgical treatment of craniovertebral stenosis in patients with MPS.

Material and Methods

A total of 9 patients with MPS (4 men, 5 women) were operated for craniovertebral stenosis at the N.N. Pirogov National Medical Research Center of Traumatology and Orthopedics in 2012–2017. Of them, 7 patients had Maroteaux – Lamy syndrome (MPS VI), one had Hurler–Scheie syndrome (MPS IHS) and one had Hunter syndrome (MPS II).

All patients underwent clinical examination, standard radiography of the cervical spine with functional tests to assess atlantooccipital stability according to Rothman, and MRI to determine the degree of the spinal cord compression, stenosis of the spinal canal, and changes in soft tissues surrounding the spinal canal due to fibrosis and accumulation of GAG. Maximum stenosis at the C0–C2 level was noted in 5 patients, at C1–C2 in 2, at C2–C4 in 1, and at C0–C1 in 1. Myelopathy in the upper cervical spine was detected in five cases (Fig. 1).

The overall somatic and functional statuses (6-min walking test, Karnofsky and Lansky scales), as well as neurological parameters (modified JOA Score, Ranawat scale) were assessed.

Seven patients have received enzyme replacement therapy for at least 6 months prior to the surgery; in two cases this type of treatment was not available at the time of the surgery.

Due to the anatomical features of the respiratory tract (laryngeal stenosis, deformity and abnormal development of the trachea, enlarged tonsils and adenoids, thickening of the epiglottis and vocal cords, large tongue, stiffness in the temporomandibular joints), this category of patients has difficulty with intubation. In patients with MPS, multifactorial stenosis in the upper and lower respiratory tract due to the deposition of GAG can be observed even at an early age. Challenges with intubation were resolved with the help of navigation equipment (fibrobronchoscope).

Decompression at appropriate levels was performed in all cases, occipitospondylodesis, in eight patients, and fixation according to the Harms method, in one patient (Fig. 2). One patient underwent repeated decompression due to progression of stenosis; the primary surgery without stabilization was performed in another medical institution. Screw fixation was used in four patients, hook fixation, in two, and hybrid instrumentation, in three. Pronounced neurological symptoms were observed in seven cases; two patients experienced a decrease in motor activity during a year, one of them had episodes of loss of motor function without loss of consciousness.

Follow-up examination and X-ray examination were performed at 3, 6 and 12 months after surgery, and further every 6 months.

Results

The average follow-up period was 2.5 years (from 6 months to 6 years). The age of the patients varied from 1.8 to 34 years. All patients had characteristic clinical manifestations of multiple dysostosis caused by MPS: short stature; coarsening of facial features; shoulder, elbow, wrist, hip and knee joints contractures; flattening of the feet; chest and spinal deformities. X-ray data did not reveal the instability of the cervical region before the surgery.

After 3, 6, and 12 months after surgery, instrumentation was radiologically stable in all cases, there were no bone resorption signs around the elements of instrumentation (Fig. 3). Seven patients reported regression of neurological symptoms and a positive dynamic in somatic status; for one patient the neurological status was unchanged (Table 1). In one case, there was a temporary deterioration after an unsuccessful attempt at intubation; emergency tracheostomy was required, and the surgery was postponed to a later date to stabilize the condition [26].

Complications after surgical treatment were observed in five patients. In one case, pseudoarthrosis, rod fracture, and instrumentation instability were observed 1.5 years after the intervention, and revision surgery was required. Wound healing problems were observed in three patients and a liquorrhea in the early postoperative period, which was stopped without revision intervention – in one case. Detailed information on the nature of the surgery is presented in Table 2.

Discussion

Spinal stenoses at the level of the upper cervical spine and craniovertebral pathology are more common in MPS types IV and VI, and less frequent in MPS type I [25]. The lesion of the cervical spine is the most common indication for surgery in MPS type IV [27]. Changes in this area include several components: hypoplasia of the odontoid process, atlantoaxial instability and thickening of extradural soft tissues. In patients with type VI MPS, pathological transformation occurs quite early and manifestations of spinal cord compression at the cervical level are diagnosed before the age of two years [28, 29]. Mild spinal stenosis can be determined radiographically in the absence of significant clinical manifestations [30–33]. Some people believe, based on the principle of critical compression of the spinal cord, that decompression and stabilization in patients with MPS are best done in advance, prior to development of any severe neurological disorder [11].

Hypoplasia of the odontoid process is the result of a disruption of its ossification, and the presence of *os odontoideum* can be considered as an evidence of instability [31]. Stabilization of the cervical spine leads to ossification of the odontoid process and restoration of the structure of the surrounding soft tis-

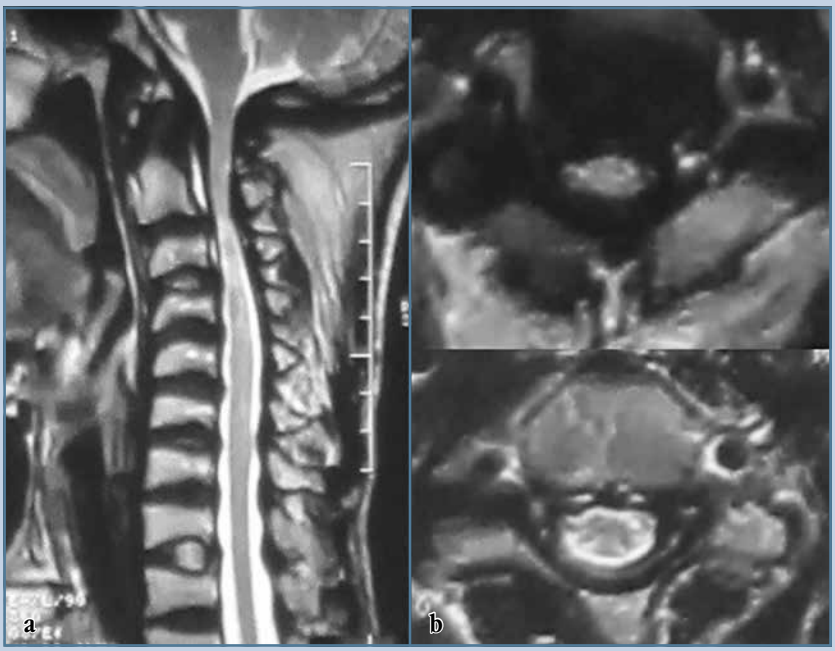

Fig. 1
MRI of the cervical spine of a 34-year-old patient with type VI mucopolysaccharidosis, before surgery: **a** – T2-weighted sagittal image, myelopathy locus at the level of C2–C4; **b** – T2-weighted axial image, myelopathy locus

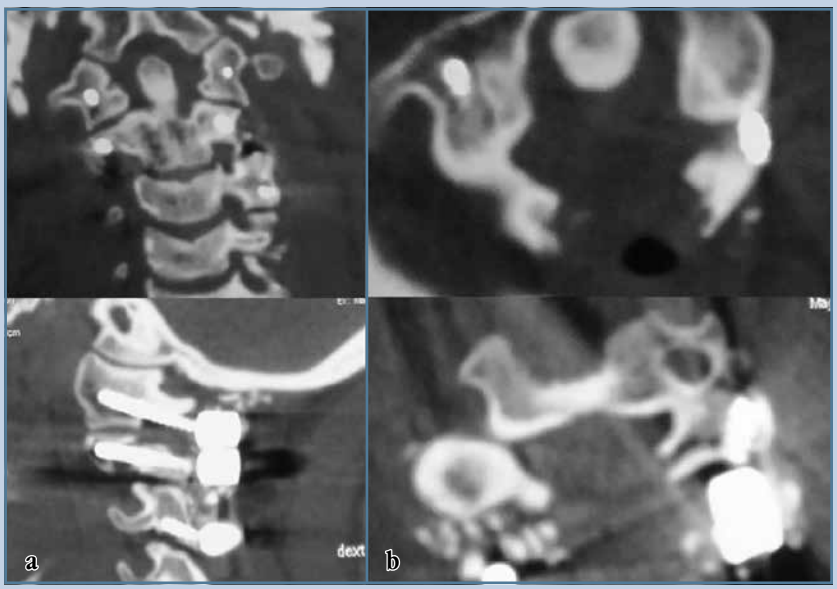

Fig. 2
CT scan of the cervical spine of a 34-year-old patient, with type VI mucopolysaccharidosis, after surgery: **a** – Harms fixation, the position of the metal structure is stable; **b** – decompression of the spinal cord at C1–C3, the lument of the spinal canal is clear, the position of the screws is satisfactory

sues [31]. Early successful hematopoietic stem cell transplantation reduces odontoid dysplasia in MPS IH [34]. Enzyme replacement therapy does not prevent the development of cervical myelopathy in patients with MPS IS [35]. Deformity of the subaxial spine (kyphosis or kyphoscoliosis), which is common in patients with MPS IV and IH, contributes to the development of the compression [2, 11, 16].

Preventive occipitospondylodesis before the development of myelopathy is recommended for all patients with MPS type IV to prevent persistent neurological manifestations [32].

Surgical treatment based only on the data of X-ray study in patients with type IV MPS is confirmed by reports that the main goal of the surgery is stabilization, rather than improvement of neurological symptoms [18, 30, 36]. Full recovery of neurological functions is rare [28].

Some authors [30, 31] advocate the upper cervical stabilization or occipitospondylodesis with or without decompression. Satisfactory clinical results are described in patients with MPS of types IV [30] and VI [36] following the decompression without stabilization although this tactic seems to be risky due to the likelihood of instability in other departments [17, 19, 37]. There are reports on development of progressive spinal deformity in children from 16 to 100% after its primary surgical destabilization [20]. The continued accumulation of GAG products in this area can lead to repeated surgical interventions [38]. Krenzlin et al. [39] presented the results of treatment of 15 patients with type I, IVA and VI MPS who underwent craniocervical decompression from minimal posterior approach without any fixation with an average follow-up period of 6 years. However, reoperation due to continued progression of stenosis was required in more than half of the cases [39]. Instrumentation significantly reduces the development of cicatricial stenosis. Successful outcomes of operations that consisted only of decompression may be attributed to the fact that atlantoaxial instability is not always identified in dynamic X-ray studies in patients

with type IV MPS, even in the presence of neurological disorders [27, 29]. It is not known what causes these difficulties in diagnosing instability based on X-ray data: spasticity of the patient's muscles or fibrous hypertrophy [28].

Involvement of the respiratory tract in the pathological process is most characteristic of severe forms of MPS such as I, II and VI. There are reports [22, 26, 40] of various complications during anesthesia, in up to 60 % of cases. According to Muhlebach et al. [21] and based on our experience it is advisable to use a flexible bronchoscope to reduce the risk of complications during intubation [41].

Surgery in the upper cervical region can be challenging in small patients due to changes in anatomy and poor bone quality. We described 9 patients, 5 of whom are children. In these patients, stabilization techniques included bilateral C2 fixation with laminar screws [42], C1–C2 transarticular screws or wire, and external fixation in the halo device [18]. Hybrid instrumentation is actively used in children. There are reports on a combination of rigid rods with sublaminar suture fixation at the level of the cervical spine, which takes into account the fragility and immaturity of the bone tissue. Ha et al. [43] used nonabsorbable sutures for severe distraction damage and did not report any complications during the postoperative follow-up. The postulated advantages of this technique are associated with preservation of movements and prevention of iatrogenic damage of the ossification zone and the vertebral arch, which can be observed with the use of titanium wire or screws [43]. Occipital and laminar hooks with pedicle screws in C2 can provide greater structural stability in the immature occipital bone, preventing complications from bicortical screws. In addition, Odent et al. [44] reported significant instability of screw fixation in children and the effectiveness of using hook structures, including four patients with MPS of various types. Literature data concerning the surgical treatment of cervical stenosis and instrumentation in pediatric patients with MPS are limited due to the rarity of the pathology. Most surgical methods, results and complications are extrapolated to studies involving older children or adults [44]. In our opinion, it is reasonable to use of hybrid constructions with laminar hooks in children with MPS. Additional bone graft from the femur or tibia [29], the iliac crest [38], ribs [18], and bone morphogenetic proteins are often necessary to achieve the bone block with a small bone surface area. In most of our cases, local autograft obtained as a result of decompression was sufficient, sometimes in combination with a biocomposite (bioplastic) material.

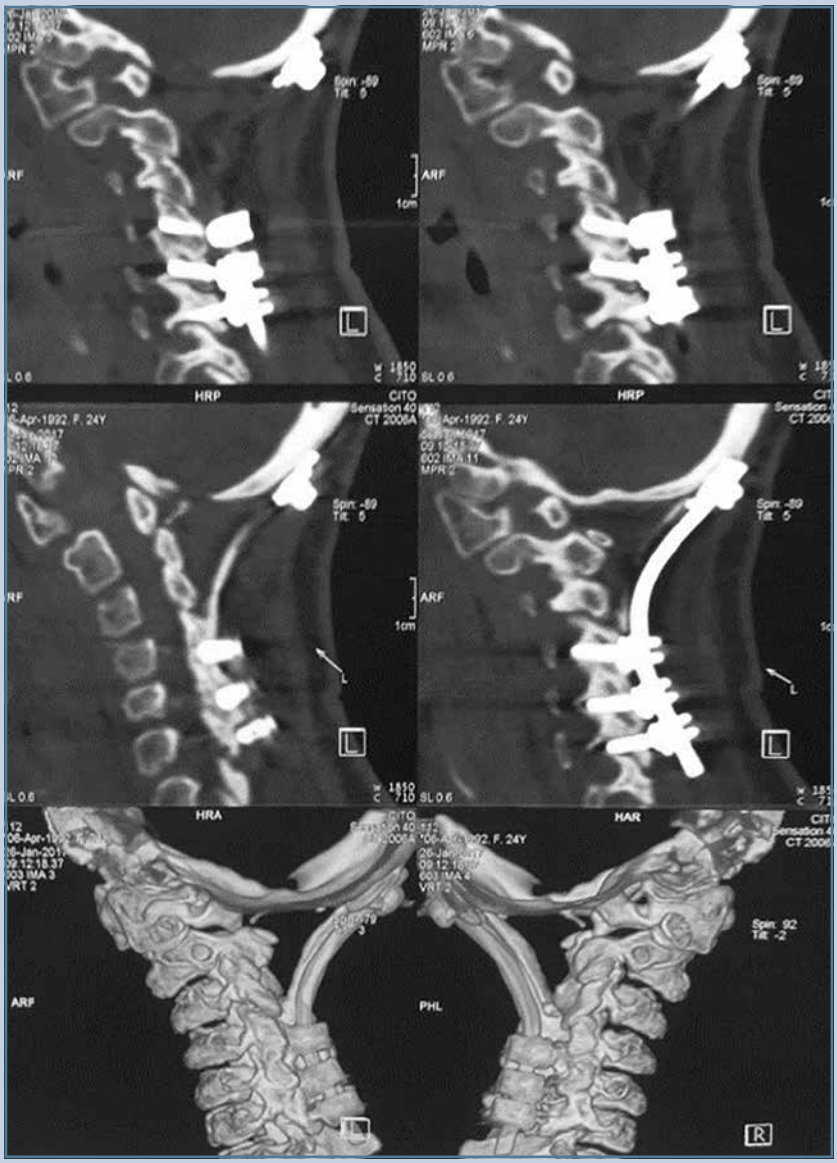

Fig. 3
CT scan of the cervical spine of a 25-year-old patient with type VI mucopolysaccharidosis, 3 years after occipital spondylodesis: metal construction is stable, no signs of osteoresorption around the metal construction

Table 1

Changes in the neurological status of patients with mucopolysaccharidosis (MPS) 6 months after the surgery

Patients (age/gender)	MPS type	Enzyme replacement therapy at the time of the surgery	Karnofsky and Lansky scales, points		JOA Score, points		Ranawat scale, points	
			prior to the surgery	6 months after the surgery	prior to the surgery	6 months after the surgery	prior to the surgery	6 months after the surgery
1 (31/F)	IHS	Aldurazyme for 7 years	60	80	9	12	3	2
2 (34/F)	VI	Naglazyme for 3 years	70	90	8	13	3	1
3 (21/F)	VI	Naglazyme for 3 years	60	80	8	14	3	1
4 (15/F)	VI	Not available at the time of the surgery	40	60	4	11	4	3
5 (22/M)	VI	Naglazyme for 3 years	30	50	9	13	4	2
6 (11/M)	VI	Not available at the time of the surgery	40	/40	2	4	4	4
7 (1.8/M)	II	Elaprase for 1 year	40	70	–	–	4	2
8 (3.8/M)	VI	Naglazyme for 11 months, a break of 10 months, renewal for 4 months before the surgery	70	100	13	15	1	0
9 (5.1/M)	VI	Naglazyme for 11 months, a break of 10 months, renewal for 4 months before the surgery	80	90	13	14	1	0

Conclusion

Decompression and stabilization in patients with MPS should be used well in advance, before the development of any neurological disorder. Fixation is required to prevent instability, postlaminectomy deformities and the continuation of cicatricial stenosis. The timely initiation of enzyme replacement therapy improves the prognosis of this disease.

The study did not have sponsorship. The authors declare no conflict of interest.

Table 2

Stenosis level, type of surgery, complications in patients with mucopolysaccharidosis

Patients (age/ gender)	Level of craniovertebral stenosis	Surgery	Complications	Follow-up period
1 (31/F)	C1–C3	Resection of the posterior edge of the large occipital foramen; laminectomy C1–C3; occipitosponylodesis C0–C5 (trans-articular screws) with autobone	None	3 years
2 (34/F)	C2–C4, myelopathy	C1–C3 posterior decompression of the spinal cord, C1–C3 posterior fixation using Harms technique; autograft posterior spinal fusion	None	2 years
3 (21/F)	C1–C3, myelopathy	Resection of the posterior edge of the large occipital foramen; laminectomy C1–C3; occipitosponylodesis C0–C5 (trans-articular screws), allo- (spongy chips) and autobone	None	3 years
4 (15/F)	C0–C2, myelopathy	Resection of the posterior edge of the large occipital foramen; laminectomy C1–C3; occipitosponylodesis C0–C6 (screws + hooks); correction of kyphotic deformity of the thoracolumbar region	None	6 years
5 (22/M)	C0–C2	Two-stage correction of kyphotic deformity of the thoraco-lumbar spine; resection of the posterior edge of the large occipital foramen; laminectomy C1–C3; occipital spondylodesis C0–C6 (trans-articular screws); the imposition of secondary sutures; reassembly of instrumentation; posterior spinal fusion with autorib (after 1.5 years)	The dihescence of the wound edges within the subcutaneous tissue; pseudarthrosis, fracture of metal rods	4 years
6 (11/M)	C0–C2, inclination of C1 into the large occipital foramen	Halo-device; laminectomy C1; occipitosponylodesis C0–C5 (hooks) with autobone	Failed intubation attempt, deterioration	1 year
7 (1.8/M)	C0–C1, myelopathy, odontoid hypoplasia C2	Halo-device; spinal cord decompression at C0–C1 level; occipitosponylodesis C0–C5 (hooks) osteoinductive crumb + autobone, secondary suture	The dihescence of the wound edges	2 years
8 (3.8/M)	C0–C2, lack of C2 odontoid union	Laminectomy C1–C3, resection of the posterior edge of the large occipital foramen; C0–T1 occipito spondylodesis (hooks + pedicle screws in T1) + autobone	Liquorrhea	6 months
9 (5.1/M)	C0–C2, lack of C2 odontoid union	Resection of the C1 vertebra arch without fixation; C1, C2, C3 laminectomy, resection of the posterior edge of the large occipital foramen; C0–T1 occipital spondylodesis (hooks + pedicle screws in T2) + autobone	The dihescence of the wound edges within the subcutaneous tissue	6 months

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