CASE REPORT
DEGENERATIVE
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SURGICAL TREATMENT OF CRANIOVERTEBRAL STENOSIS IN TWO SIBLINGS WITH TYPE VI MUCOPOLYSACCHARIDOSIS

TRATAMENTO CIRÚRGICO DE ESTENOSE CRANEO-VERTEBRAL EM DOIS IRMÃOS COM MUCOPOLISACAREDOSE TIPO VI

TRATAMIENTO QUIRÚRGICO DE ESTENOSIS CRANEO-VERTEBRAL EN DOS HERMANOS CON MUCOPOLISACARIDOSIS TIPO VI

PEREVERZEV VLADIMIR S.[1], KAZMIN ARKADII I.[1], KOLESOV SERGEY V.[1]

1. N.N Priorov National Medical Research Center, Moscow, Russia.

ABSTRACT

We describe two cases of surgical treatment of craniovertebral stenosis in preschool-aged brothers with Maroteaux-Lamy (MPS type VI) syndrome. The older brother was diagnosed with MPS during her second pregnancy. Literature describing familial cases of the disease and the treatment strategy in young children with MPS type VI and spinal canal stenosis is scarce. Based on the presented observations, indications, surgical treatment approaches, and perioperative management of patients with mucopolysaccharidosis are suggested. MPS type VI may have familial forms of the disease and the course of craniovertebral stenosis is similar in siblings. Surgical treatment of craniovertebral stenosis in these patients should be performed timely. We adhere to the point of view of early treatment of craniovertebral stenosis in patients with MPS before irreversible spinal cord dysfunction develops. ***Level of Evidence IV; Prognostic Studies - Investigating the Effect of a Patient Characteristic on the Outcome of Disease and Case series.***

Keywords: Vertebrobasilar Insufficiency; Mucopolysaccharidosis VI; Perioperative Care; Surgical Procedure.

RESUMO

Descreve-se dois casos de tratamento cirúrgico de estenose craniovertebral entre irmãos em idade pré-escolar com síndrome de Maroteaux-Lamy (MPS tipo VI). O irmão mais velho foi diagnosticado com MPS durante a segunda gravidez. A literatura que descreve casos familiares da doença e a estratégia de tratamento em crianças pequenas com MPS tipo VI e estenose do canal raquidiano é escassa. Com base nas observações apresentadas, foram sugeridas indicações, abordagens de tratamento cirúrgico e manejo perioperatório de pacientes com mucopolissacaridose. A MPS tipo VI pode apresentar formas familiares da doença e o curso da estenose craniovertebral é semelhante entre irmãos. O tratamento cirúrgico da estenose craniovertebral nesses pacientes deve ser realizado em tempo hábil. Adere-se ao conceito de tratamento precoce da estenose craniovertebral em pacientes com MPS antes que se desenvolva uma disfunção irreversível da medula espinhal. ***Nível de Evidência IV; Estudos Prognósticos - Investigando o Efeito de uma Característica de Paciente sobre o Resultado de uma Doença e de uma Série de Casos.***

Descritores: *Insuficiência Vertebrobasilar; Mucopolissacaridose VI; Assistência Perioperatória; Procedimento Cirúrgico.*

RESUMEN

Se describen dos casos de tratamiento quirúrgico de estenosis craneovertebral en hermanos de edad preescolar con síndrome de Maroteaux-Lamy (MPS tipo VI). Al hermano mayor se le diagnosticó MPS durante el segundo embarazo. La bibliografía que expone casos familiares de la enfermedad y la estrategia de tratamiento en niños pequeños con MPS tipo VI y estenosis del tubo vertebral es escasa. Sobre el fundamento de las observaciones presentadas, se sugieren indicaciones, enfoques de tratamiento quirúrgico y manejo perioperatorio de pacientes con mucopolisacaridosis. La MPS tipo VI puede presentar formas familiares de la enfermedad y el curso de la estenosis craneovertebral es semejante en los hermanos. El tratamiento quirúrgico de la estenosis craneovertebral en estos pacientes debe realizarse tempranamente. Se adhiere al planteamiento del tratamiento precoz de la estenosis craneovertebral en pacientes con MPS anticipándose al desarrollo de una disfunción irreversible de la médula espinal. ***Nivel de Evidencia IV; Estudios Pronósticos - Investigando el Efecto de una Característica del Paciente en el Resultado de la Enfermedad y Series de Casos.***

Descriptores: *Insuficiencia Vertebrobasilar; Mucopolisacaridosis VI; Atención Perioperativa; Procedimiento Quirúrgico.*

Study conducted by the N. N. Priorov National Medical Research Center, Moscow, Russia.
Correspondence: Vladimir Pereverzev. Str, 10, Priorov, Moscow, Russia. 127299. vcpereverz@gmail.com

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INTRODUCTION

Mucopolysaccharidosis type VI (MPS VI), or Maroteaux-Lamy syndrome, first described by Pierre Maroteaux and Maurice Emil Joseph Lamy in 1963, is a rare autosomal recessive disease caused by deficiency of the enzyme N-acetylgalactosamine-4-sulfatase, also called arylsulfatase B (ARSB). The pathophysiological mechanisms underlying the development of type VI MPS involve the systemic progressive accumulation of dermatan sulfate (a glycosaminoglycan) due to a secondary deficiency of the ARSB enzyme. Clinical manifestations of type VI MPS depend on the time of onset, rate of progression, and severity of the disease.[1] The rapidly progressing disease usually manifests in the first year of life. Typically, the clinical scenario involves growth retardation, delay in physical development, coarsening of facial features, systemic skeletal lesions, muscle hypotension, and motor disorders. Internal organ changes manifest in hepatosplenomegaly and frequent respiratory infections leading to respiratory failure, joint damage, cardiovascular disorders, and eye and skin damage. The cognitive function remains unaffected, but cardiopulmonary complications and movement restrictions due to joint damage and spinal cord compression usually lead to deterioration of the quality of life and patient death before the third decade of life.[2] The slowly developing form tends to progress over decades, and patients can live for as long as 40-50 years. Previously, the approach to treating such patients was limited to palliative care. However, enzyme replacement therapy (ERT) with recombinant human ARSB has led to effective pathogenic treatment as it has become widely available.[2] Neurological complications usually develop with disease progression and have been widely described in the literature.[3] The most common neurological manifestations of type VI MPS are carpal tunnel syndrome (CTS), spinal cord compression, optic nerve damage, and communicating hydrocephalus. Neurological disorders have also been observed in other types of MPS.[4] Craniovertebral stenosis develops due to C2 odontoid process hypoplasia, atlantoaxial instability, and accumulation of glycosaminoglycans in the dura mater and posterior longitudinal ligament, which subsequently leads to compression myelopathy at this level, development of bulbar disorders, and central respiratory failure.[5] Due to this pathology's complex and rare nature, there is sparse literature on the treatment strategies for these patients. Here, we describe two clinical cases of siblings with MPS type VI.

Patient I (Figure 1)

A boy, aged five years and two months at admission, presented with weakness in the lower extremities, limited movement in the upper and lower extremities, pain in the knee and elbow joints, as well as in the thoracic and lumbar spine, hearing, and visual acuity loss, and difficulty breathing. The patient was born to consanguineous parents of Ingush descent from the first pregnancy. His mother had gestational toxicosis throughout the pregnancy, with a risk of interruption at 34-35 weeks, acute respiratory viral infection in the 2nd and 3rd trimesters, edema, and arterial hypertension. Delivery occurred during the 40th week through a Caesarean section (clinically narrow pelvis). At birth, the child's condition was considered severe: weight, 4300 g; height, 57 cm; head circumference, 37 cm; chest circumference, 38 cm; Apgar score, 6. From Presented with hepatosplenomegaly, bilateral inguinal hernia, increased irritability, and a refusal to eat. The child was discharged from the maternity hospital in moderate severity with the following diagnoses: moderately severe perinatal hypoxic-ischemic encephalopathy, hyperirritability syndrome, edema, and facial paresis on the left. Subsequently, the child was treated at the Department of Neonatal Pathology and routinely hospitalized for respiratory infections.

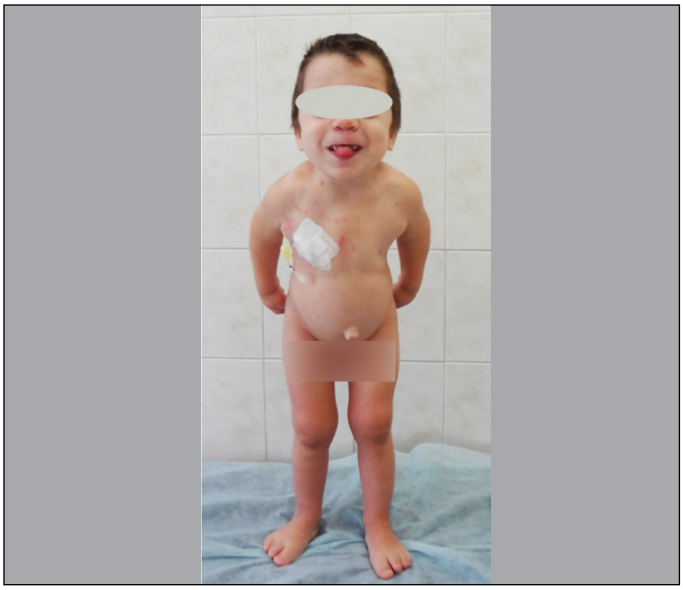

Figure 1. Appearance of the first sibling.

At 14 months, the patient underwent surgical treatment for a bilateral inguinal hernia. Owing to complaints of spinal deformities, genetic counseling is recommended. At 15 months, the patient was diagnosed with mucopolysaccharidosis. A test for arylsulfatase B, which was severely deficient, confirmed this diagnosis. At 18 months, the patient underwent an MRI and was diagnosed with spinal canal stenosis at C1-C2, in connection to which, Resection of the posterior arch of C1 was performed. No complications were observed in the postoperative period. Currently, the patient is receiving enzyme-replacement therapy with galsulfase. At the age of five years, the patient's condition deteriorated as he became lethargic and unable to walk. Magnetic resonance imaging (MRI) data demonstrated stenosis progression at the level of C0-C2 (Figure 2). The patient was hospitalized for surgical treatment in our department.

Presented with a height of 94.5 cm, weight of 14.5 kg, a short and thick neck, multiple signs of skeletal dysplasia, flexion contractures in the wrists, hypertelorism, sunken nose, corneal opacity, hypermetropia and astigmatism, bilateral otitis media and deafness, umbilical hernia, hepatomegaly, growth retardation, and delayed psycho-speech development. With congenital dermal melanocytosis, the patient's skin was dense enough to touch. During preoperative preparation, the patient was also noted to have experienced acute weakness in the upper and lower extremities, leading to him falling without losing consciousness and episodes of night apnea.

Preoperative radiography, magnetic resonance imaging (MRI), and computed tomography data demonstrated spinal canal stenosis at the level of the craniovertebral junction and kyphotic deformity of the cervicothoracic thoracolumbar regions with an apex at the L2 level (local kyphosis angle of 45°). Neurological examination revealed signs of cervical stenosis without pyramidal or myelopathy. Ultrasound examination demonstrated developmental anomalies of the median nerves at the level of the carpal canal entrance (three branches) and signs of median nerve neuropathy at the carpal joint level. The echocardiographic evaluation revealed cardiomyopathy with AV valve and aortic valve pathology.

Considering the patient's metabolic disease and progression of craniovertebral stenosis, he underwent surgical treatment: a halo was applied, six pins were embedded within the outer cortical plate of the skull, 2 in the area of the frontal tubercles, four in the parietal-occipital region. When pulling on the ring, the latter is firmly fixed. Aseptic dressings. The patient is turned over on his stomach; skeletal traction is mounted behind the halo. After treatment of the surgical field with antiseptics, a skin incision was made from the

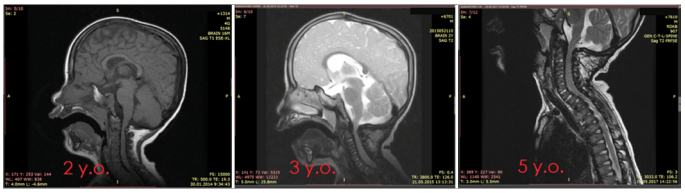

Figure 2. Progression of craniovertebral stenosis, MRI.

occipital bone to the spinous process of the Th1 vertebra. The subcutaneous tissue and fascia are dissected in layers. The posterior elements C1-Th1, occipital bone is exposed. Hemostasis. Laminectomy of the C1-C2-C3 vertebrae and resection of the posterior edge of the foramen magnum was performed, and the dural sac pulsates. Transpedicular insertion of polyaxial screws into the body of the Th1 vertebra from both sides was performed. Fluoroscopic control -the position of the screws is correct. On the right, under the arms C5 from above and C6 from below, on the left and right, hooks are installed according to the type of crab capture. Rods are fixed in the installed hooks on the left and right, stabilizing the occipital bone and the cervical spine. Hemostasis. Holes are made in the occipital bone to install rods that stabilize the occipital bone and the cervical spine. Fixed with two cortical screws on the right and left. Local autografts were placed on the decorticated posterior elements of the C2,3,4,5 vertebrae. Drainage of the wound through an additional incision. Layered suturing of the wound. Dismantling of the halo ring. The blood loss was 150 ml. The operation time was 2 hours and 45 minutes

Because of anatomical changes in the respiratory tract, macroglossia, and enlarged adenoids, intubation was performed using a fiberoptic bronchoscope. Owing to an allergy to cephalosporins, a carbapenem antibiotic was used postoperatively. On the third day after surgery, the patient presented with fever and signs of bilateral pneumonia on CT. Antibiotic therapy was continued, supported by mucolytic inhalation, breathing exercises, and physical therapy. The patient was mobilized using a soft cervical collar. The signs of pneumonia resolved after one week of treatment. Nine days after surgery, wound dehiscence due to a hematoma was observed. Was administered anti-inflammatory and immunomodulatory medications to improve microcirculation and prevent infectious complications. The wound healed with a secondary intention. The sutures were removed 14 days after surgery. Postoperative radiographic images confirmed the optimal implant position (Figure 3). The patient was discharged 50 days after the surgery. The neurological and functional status were evaluated before and after surgery using the modified Japanese Orthopedic Association (JOA) scale, and the modified Ranawat, Lansky scale.

According to the scales, positive dynamics were noted (mJOA from 13 to 14, modified Ranawat from 1 to 0, Lansky from 80 to 90) when evaluated 12 months after the operation; these indicators were unchanged during the entire follow-up period, indicating the effectiveness of the treatment.

Patient II (Figure 4)

The younger brother was admitted to our department at the age of 3 years 10 months with complaints of increased irritability, gait imbalance, chronic constipation, restricted movements in the upper and lower extremities, hearing and visual acuity loss, difficulty breathing, and developmental delay. The patient was born in the second pregnancy, accompanied by toxicosis in the first and third trimesters and edema in the third trimester. Delivery occurred during the 37th week through Caesarean section (uterine scar, clinically narrow pelvis). At birth, the condition was considered severe. Weight – 3880 g, height: 50 cm, Apgar score: 7. The child was discharged from the maternity hospital in moderately severe condition on the 6th day. The child was diagnosed with an umbilical hernia and presented with anxiety, restless sleep, and constipation from birth. At the age of 1 month, the patient was predisposed to frequent respiratory infections. Considering his brother's diagnosis, the patient was evaluated at a genetics institution where type VI MPS was confirmed. The patient was then referred to our department. Upon evaluation, the patient presented with convergent strabismus, mitral valve prolapses, insufficiency with minimal regurgitation, patent foramen ovale, colon dyskinesia, multiple skeletal dysplasias, corneal opacity, umbilical hernia, bilateral chronic otitis media, deafness, and growth retardation. The patient's height and weight were 87 cm and 14.5 kg.

Owing to the progression of craniovertebral stenosis, the patient was hospitalized in the Department of Spinal Pathology. Radiographs and CT of the spine revealed cervicothoracolumbar kyphosis of 40°, with an apex at T10-T11 (Figure 5). Neurological examination confirmed signs of craniovertebral stenosis, without pyramidal or myelopathic symptoms. The patient underwent surgical treatment analogous to that his older brother underwent earlier: laminectomy at C1-C3, resection of the posterior edge of the foramen magnum, spinal cord decompression, and occipito-cervico-thoracic fusion (C0 to T1) using a hybrid construct (screws and hooks). Surgery was performed under intraoperative halo-traction. The operation

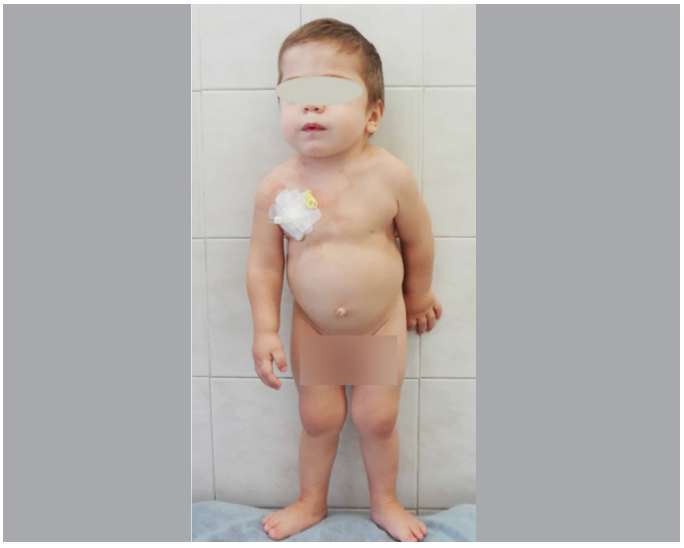

Figure 4. Appearance of the second sibling.

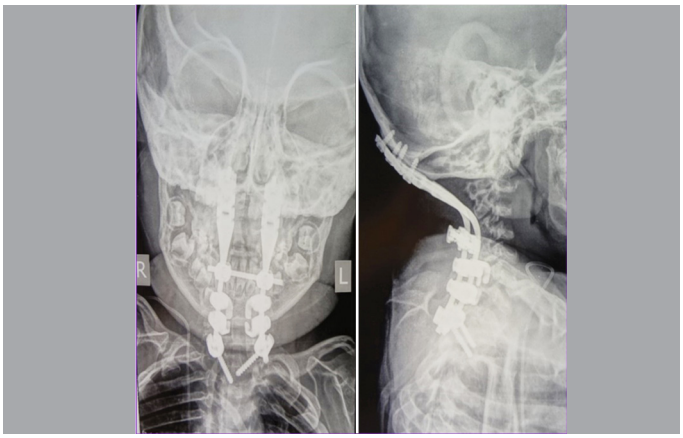

Figure 3. Postop X-rays. The position of the implants. First sibling.

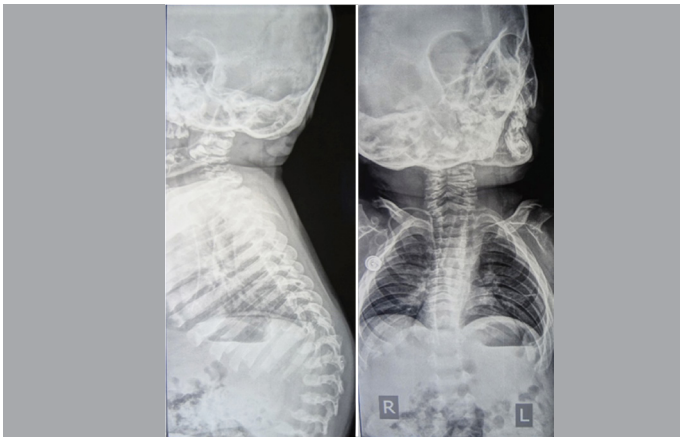

Figure 5. Thoracolumbar kyphosis in patients with MPS, second sibling.

time was 2 hours 30 minutes, blood loss 450 ml. No complications were observed in the early postoperative period. The patient was mobilized using a soft cervical collar. The wound healed with primary intention, and the patient was discharged ten days after surgery. Radiographs at two days and CT one week after surgery demonstrated satisfactory positioning of the implants. (Figure 6)

During hospitalization, the patients underwent regular weekly galsulfase infusions through infusion ports within the framework of continuing enzyme replacement therapy, with antihistamine administration for allergy prophylaxis.

According to the evaluation scales, improvement (mJOA from 13 to 15, modified Ranawat from 1 to 0, Lansky from 70 to 100) 12 months after the surgery. The indicators did not change significantly during further observation.

Ethical Compliance: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki Declaration and its later amendments.

Data Access Statement: Research data supporting this publication are available from the N. N. Priorov National Medical Research Center repository at located at https://www.sechenov.ru/upload/medialibrary/7d8/Dissertatsiya-Pereverzev.pdf

DISCUSSION

Stenosis of the spinal canal at the upper cervical spine level and craniovertebral pathology is more common in patients with type IV and VI MPS and is least common in type I MPS.[6] Cervical spine pathology is the most common indication for surgical intervention in patients with type IV MPS.[2] Pathoanatomical changes at this level include hypoplasia of the odontoid process, atlantoaxial instability, and extradural soft-tissue thickening. These pathological changes occur relatively early, and the manifestations of cervical spinal cord compression become apparent before the age of two in patients with type VI MPS.[7] Moderate stenosis of the spinal canal can be identified radiologically in the absence of significant clinical manifestations.[8] One of the approaches to surgical treatment of such patients is based on the notion of a critical spinal cord compression threshold, suggesting that in patients with MPS, spinal cord decompression and cervical stabilization are best performed before the development of gross neurological disorders.[9]

Hypoplasia of the odontoid process results from incomplete ossification and the presence of the *os odontoideum* should be viewed as evidence of instability at the atlantoaxial level.[10] In support of this theory, ossification of the odontoid process is observed, and the structure of the surrounding soft tissues is restored as soon as stability is achieved.[10] Successful early transplantation of hematopoietic stem cells reduces the manifestations of dysplasia of the dens in patients with type 1H MPS.[11] In contrast, enzyme replacement therapy does not prevent the development of cervical myelopathy in patients with type 1S MPS.[12]

Preventive occipito-cervico-thoracic fusion before the development of myelopathy has been recommended for all patients with type IV MPS to prevent permanent neurological impairment.[10]

Surgical treatment based solely on radiological findings in patients with type IV MPS is supported by clinical studies, grounding such an approach on the basis that the main purpose of surgical intervention in these patients is cervical stabilization and not neurological symptom improvement.[8,13,14] Complete recovery of neurological function is rare.[7]

The published guidelines for craniovertebral pathology screening in patients with MPS differ significantly, especially with regard to enhanced dynamic visualization. There is consensus on the need for neurological examination in patients with type I, II, and VI MPS at least annually[15,16] and every six months for patients with type IV MPS.[1] Evaluation of the cervical spine is usually recommended before and during intubation. Still, there is no consensus on the timing or type of further visualization.[2,15,16]

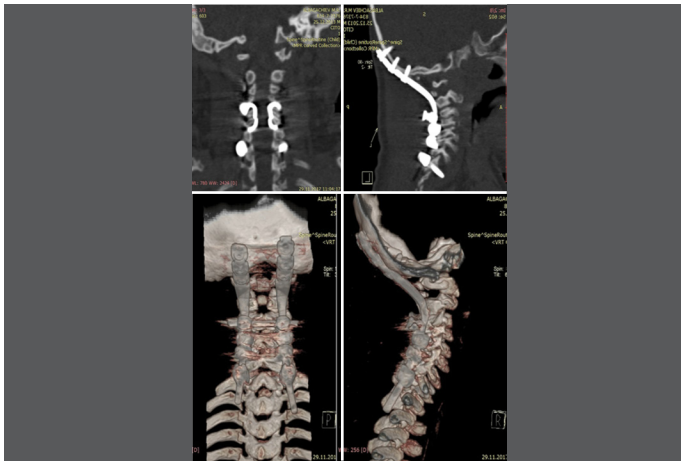

Figure 5. Postop CT. The position of the implants. Second sibling.

Most authors advocate stabilization through occipito-cervico-thoracic fusion with or without spinal cord decompression.[8,10] Satisfactory clinical results after decompression without stabilization have been described in patients with type IV[14] and VI[7] MPS. However, this approach seems unreliable due to the high likelihood of instability developing in other spine regions, as described in the literature.[17-19] Successful surgical outcomes after decompression without stabilization may be related to the fact that atlantoaxial stability is not always assessed through dynamic radiographic studies in patients with type IV MPS, even in the presence of neurological disorders.[2,8] Evaluation of spinal stability can be complicated by muscle spasticity or compression caused by dural thickening or fibrous hypertrophy.[7]

Surgery in the upper cervical region can be difficult in young patients due to changes in anatomy and poor bone quality. The stabilization techniques described in these patients included C2 bilateral laminar screws,[20] C-C2 transarticular screws or wires,[13] and external halo fixation.[13] In our opinion, it is advisable to use hybrid constructs with screws and laminar hooks in children with MPS. Additional bone grafts from the femur or fibula,[10] iliac crest,[21] ribs,[13] and bone morphogenetic proteins are often necessary to achieve adequate bony fusion with a limited bone surface area. Based on our experience, local bone autografts remaining after decompression are sufficient in most cases, sometimes requiring additional synthetic bone graft use. The continued accumulation of GAG products in the occipitocervical area can lead to repeated surgical intervention.[7]

CONCLUSION

In families raising children with Maroteaux-Lamy syndrome and planning another pregnancy, thorough medical and genetic counseling is paramount. After the diagnosis is established, patients should be observed by a pediatric orthopedic surgeon with regular screening for craniovertebral stenosis. When determining the optimal timing of surgical intervention, one should be guided by the critical spinal cord compression threshold principle, which advocates for preemptive decompression and stabilization in advance of the development of neurological disorders. The perioperative period is associated with a high risk of complications; therefore, a multidisciplinary team of specialists should consider the general condition of the patient before and after surgery and surgical wound care. Unquestionably, the early initiation of enzyme replacement therapy improves the prognosis of surgical outcomes in patients with MPS.

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