VERTEBRAL SYNDROME IN CONSEQUENCES OF SPINA BIFIDA: CLINICAL FEATURES AND TREATMENT

S.O. Ryabykh[1], A.Yu. Mushkin[2], D.M. Savin[1]

[1]*Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, Kurgan, Russia*

[2]*St. Petersburg Research Institute of Phthisiopulmonology, St. Petersburg, Russia*

The guidelines for the evaluation and treatment of vertebral pathology in patients with the consequences of spina bifida are presented for discussion among professional groups before their adoption. The guidelines are based on the literature data and the authors' own experience. The purpose of the study is algorithmization of treatment, the material is presented mainly in the form of tables.

Key Words: vertebral syndrome, spina bifida, practical guidelines.

Please cite this paper as: *Ryabykh SO, Mushkin AYu, Savin DM. Vertebral syndrome in consequences of spina bifida: clinical features and treatment. Hir. Pozvonoc. 2018;15(4):107–114. In Russian.*

DOI: http://dx.doi.org/10.14531/ss2018.4.107-114.

General methodology of guidelines

The guidelines are based on the data of modern medical literature and authors' own experience.

An analysis of publications devoted to this problem has demonstrated that almost all of the publications are based on series of clinical cases. No studies that might be attributed to an ASMOK (Association of Medical Societies for Quality of Medical Care and Education) level exceeding 2+ and to a class I or II evidence level were found. Accordingly, all guidelines in this document are of evidence level C or less.

The purpose of this work is to develop an algorithm for treatment of vertebral syndrome in patients after surgery for myelomeningocele (post-myelomeningocele syndrome (post-MMC)), which is reflected in the referential style of the paper: the material is presented mainly as tables, which, in our opinion, is convenient for practical use and making tactical decisions.

Features of analysis and material selection. These guidelines intentionally avoid consideration of *spina bifida* consequences presenting as "pure" congenital kyphosis and scoliosis in the setting of accompanying vertebral anomalies, the management of which is in detail reflected in modern literature on congenital spinal malformations; management of anterior *spina bifida* and its consequences.

The main syndromic terminology used in the guidelines for the pathology in question is described in Table 1.

General information

The prevalence rate of *spina bifida* across the world is 4.7 cases per 10,000 live births [4]. According to the Federal State Statistics Service website, 14,969 children under the age of 18 years in the Russian Federation in 2014 were for the first time registered as disabled due to congenital anomalies (malformations), deformities, and chromosomal abnormalities, with the rate of *spina bifida* being 2.1 cases per 1,000 population [5].

The rate of *spina bifida* in different spine parts varies significantly: 2–5 % in the cervical spine, 2 to 3 % in the thoracic spine, 25 % in the lumbar spine, and 65–70 % in the lumbosacral spine [6].

Clinical and radiographic signs of myelomeningocele consequences are presented in Table 2.

Clinical and radiographic features of vertebral syndrome associated with post-MMC are as follows [2, 7–11]:

– lack of posterior vertebral structures;
– wide spinal canal;
– flattening of the vertebral bodies;
– low bone density of the caudal vertebrae;
– hypoplasia of the sacrum and pelvis;
– cicatricial soft tissue changes at the deformity apex;
– soft tissue deficiency and formation of ulcer at the deformity apex;
– prevalence of combined kyphosis consisting of congenital (anomaly) and neurogenic components [12–14];
– sagittal displacement of the pelvis [15];
– rapid progression of deformity (up to 12° per year) [7, 16];
– spinal cord tethering [17–20];
– tolerance to conservative treatment [21–23].

The goals and basic principles of conservative treatment of *spina bifida* consequences in children are summarized in Table 3 [2, 13, 14, 24–29], and the basic principles of surgical treatment of *spina bifida* and its consequences are presented in Table 4.

The goals of surgical treatment are as follows [2, 13, 25–30]:

– prevention of respiratory disorders;
– ability to maintain the vertical position;
– improved upper limb manipulations;
– improved quality of life;
– longer life span.

Variants of surgical correction of spinal deformities, with allowance for vertebral syndrome features, are presented in Table 5.

Table 1
Main syndromic terminology used in the guidelines for the pathology in question [1–3]

Vertebral syndrome	A complex of clinical and radiographic symptoms characterizing the anatomical condition (structure and shape) and functional changes of the spine, spinal canal, and spinal cord
Vertebrogenic syndrome	A complex of clinical neurological (motor, sensory, visceral, and autonomic), postural, and radiographic symptoms that are pathogenically caused by changes in the anatomy and functions of the spine and spinal cord
Post-myelomeningocele syndrome (post-MMC)	A complex of clinical (orthopedic, neurological, and adaptive) and radiographic symptoms that characterize consequences of surgical treatment for different types of spina bifida
Dysraphic syndrome (synonyms: Bremer syndrome, dysraphic complex (status), dysraphic myelodysplasia, dysraphism, Fuchs myelodysraphism)	A common term for developmental anomalies characterized by incomplete closure of any anatomical structures along the midline
Spinal dysraphism (vertebral component of Bremer syndrome)	Incomplete or no fusion of midline structures — vertebrae, spinal canal, and spinal cord

Table 2
Clinical and radiographic signs of myelomeningocele effects

Orthopedic	— spinal deformity; — lower limb deformity; — dislocations and contractures of the lower limb joints, most often the hip joint; — clubfoot, including recurrent clubfoot after treatment; — osteoporosis; — fractures of long tubular bones of the lower limbs, including recurrent fractures
Neurological and neurosurgical	— paresis and paralysis; — tethered spinal cord syndrome; — hydrosyringomyelia; — Arnold–Chiari syndrome
Radiographic	— underdevelopment, up to complete absence of vertebral arches; — vertebral anomalies; — spinal deformities: kyphosis, scoliosis, kyphoscoliosis, and lordoscoliosis; — various myelodysplasias at the spina bifida level and in the craniovertebral junction area
Other clinical signs	— skin stigmas located along the spine: hypertrichosis, pigment spots, dimpling, subcutaneous lipomas; — decubitus, usually at the kyphotic deformity apex; — pelvic organ dysfunctions; — chronic urinary tract infection; — respiratory failure in the setting of an elevated diaphragm and reduced chest volume

The advantage of pelvis fixation during correction of deformity associated with spina bifida consequences is achieving spinal stability. However, spinopelvic fixation in early childhood, on the one hand, may be associated with the anatomical features of supporting bone structures (their size, spatial position, and strength) and, on the other hand, significantly change further formation of this skeleton part. In this regard, the issue of extending fixation of the spine to the pelvis may in some cases be resolved not at the time of primary intervention but be scheduled for implementation during further follow-up of the child (Table 6).

Restrictions on the use of guidelines for surgical treatment of vertebral syndrome associated with spina bifida consequences

Given that the main goal of surgical approaches described in the guidelines is to improve the child's quality of life, social adaptation, and the possibility of caring for the child, the contraindications to application of the guidelines are as follows:

– severe decompensated concomitant, including genetic, diseases and congenital malformations that are life-threatening or have significant limitations on the expected survival period;

– concomitant cerebral pathology accompanied by deep mental retardation and/or frequent convulsive seizures;

– infectious exacerbations.

Conclusion

Spinal deformities associated with post-MMC syndrome are frequent and typical manifestations. Kyphoscoliosis often develops in patients with thoracolumbar myelodysplasia, while lordoscoliosis often occurs in patients with conus medullaris dysplasia.

Corset therapy is usually ineffective and can lead to chest deformity, a reduced tidal volume, and neuropathic ulcers.

Table 3
Goals and basic principles of conservative treatment of spina bifida consequences in children

Improvement of neurological status*	Neurotropic and vascular therapy
Prevention of urinary tract infection	Uroseptics, antibiotic therapy, intermittent or continuous catheterization
Improvement of orthopedic status	Corset therapy, massage, exercise therapy, orthoses, orthopedic correction of pathological postures, contractures, etc.
Ulcer prevention	Motor regimen, treatment of skin, especially sites of the largest contact with underlying surfaces (sacrum, greater trochanter area, heels), anti-ulcer mattresses, gel pads, etc.
Social adaptation**	Physical and functional rehabilitation, training in the use of auxiliary devices: verticalizers, splints, apparatus

*the most valid scale for assessing the neurological status of patients with post-MMC is the modified JOA (mJOA) scale proposed by Benzel;
**an integrative score of life impairment and role limitations is often obtained using the functional independence measure (FIM) scale.

Table 4
Basic principles of surgical treatment for spina bifida and its consequences

Principles of surgical treatment	Indications, surgical features, and timing
Myelomeningocele plastic removal, skin defect closure	First days and weeks of life; in rachischisis —first hours of life
Removal of a bone spur in the spinal canal	Upon reaching an age of 1 year and/or weight of more than 10 kg
CSF shunt surgery (triventriculocisternostomy, VPS) [20]	Hypertension syndrome — regardless of age, progressive hydrocephalic syndrome
Elimination of caudal spinal cord tethering	Tethered spinal cord syndrome: — possibility to improve neurological symptoms; — progressive hydrosyringomyelia
Foramen magnum decompressive craniectomy	Arnold-Chiari syndrome: — presence or progression of basilar insufficiency symptoms; — progressive hydrosyromingomyelia
Deformity correction with instrumented stabilization of the spine	Recurrent ulcer at the deformity apex; progression of spinal deformity; failure of stable verticalization, in particular in sitting position

Table 5
Variants of surgical correction for spinal deformities with allowance for vertebral syndrome features

Characterization of spinal deformity	Features of orthopedic correction
Mobile deformities less than 30° (Fig. 1)	Basic treatment technique — corset therapy [2, 9, 11, 12, 21]. Spine fixation with dynamic systems (VEPTR or Shilla), including fixation of the sacrum or pelvis, should be considered as an option [14, 31–34]
Rigid deformities larger than 30° (Fig. 2)	Corrective vertebrotomy (kyphectomy or VCR), correction and posterior instrumented fixation of the spine, including fixation of the sacrum or pelvis [33, 35, 36]
Deformities complicated by n ulcer in the intended surgery site, usually at the kyphotic apex (Fig. 3)	Two-stage surgical treatment: stage 1 — external instrumented fixation (halo-pelvic, transpedicular-pelvic) with dosed correction of deformity to reduce tension of soft tissues and improve conditions for their healing; stage 2 — kyphectomy with correction and posterior instrumented fixation of the spine [14]

Early surgical correction improves body balance and quality of life, reducing aggressiveness of the surgical procedure.

The dual growing rod technique is safe and effective in cases of moderate neuromuscular spinal deformities at an early age.

Kyphectomy is a complex procedure associated with a high risk of complications, but it has no alternative in the case of severe rigid kyphosis.

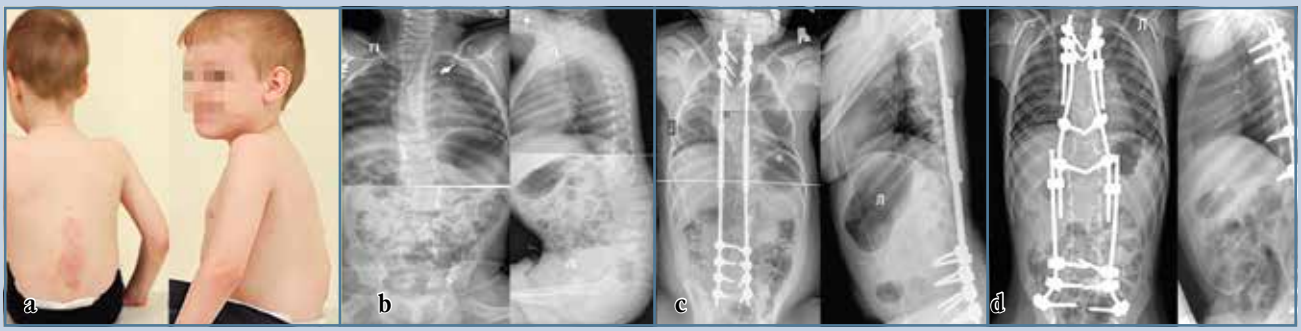

Fig. 1
Examples of surgical treatment of mobile spinal deformity using growing rods; variants of arrangement of instrumentation with longitudinal and parallel connectors: **a** – the appearance of a 5-year-old child; **b** – preoperative radiographs; **c** – radiographs after placement of growing rods with a longitudinal connector; **d** – a dynamic system with side-by-side connectors and the ability of separate control for growth of the thoracic and lumbar spine

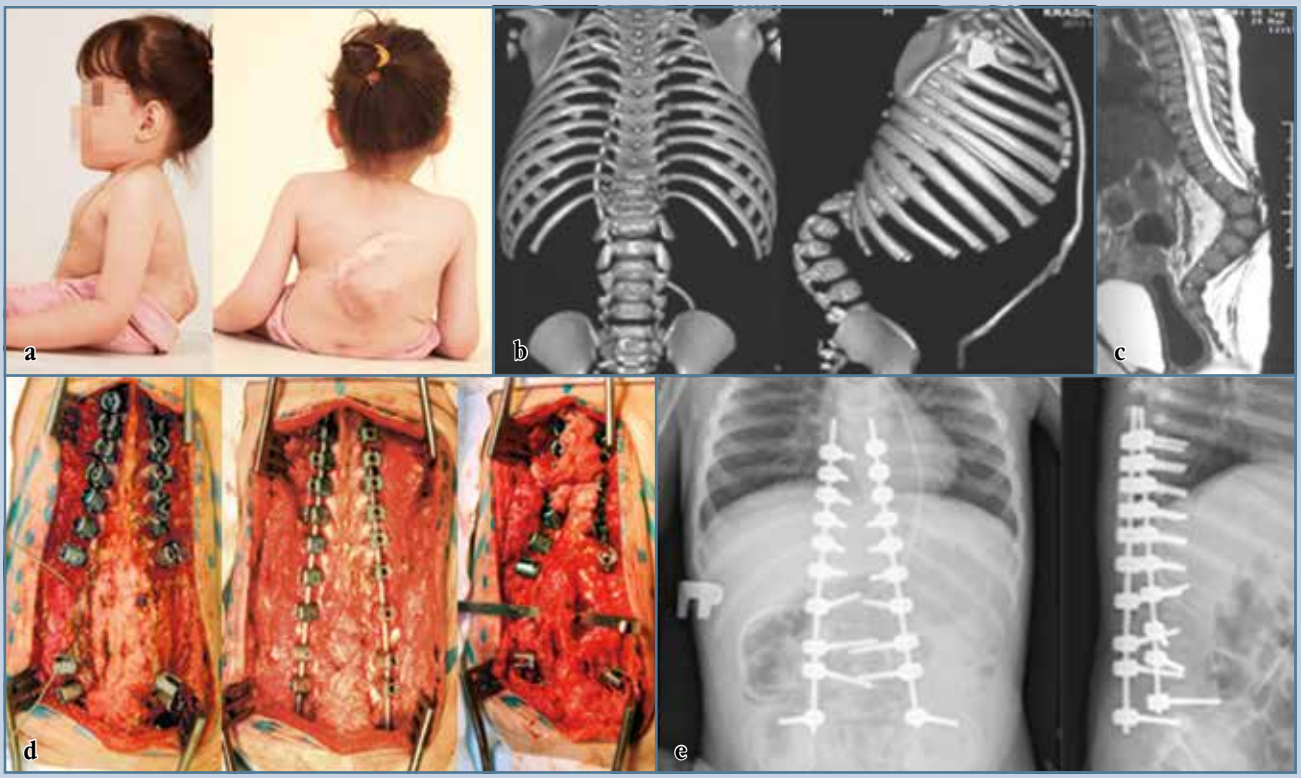

Fig. 2
One-stage correction of rigid spinal deformity in an 18-month-child using polysegmental transpedicular and pelvic fixation: a child's appearance **(a)**, a CT scan **(b)**, and a MRI scan **(c)** before surgery; intraoperative photographs of instrumentation and kyphectomy stages **(d)**; postoperative radiographs of the spine **(e)**

The use of external fixation systems should be considered as a "parachute" technology in patients with ulcers.

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

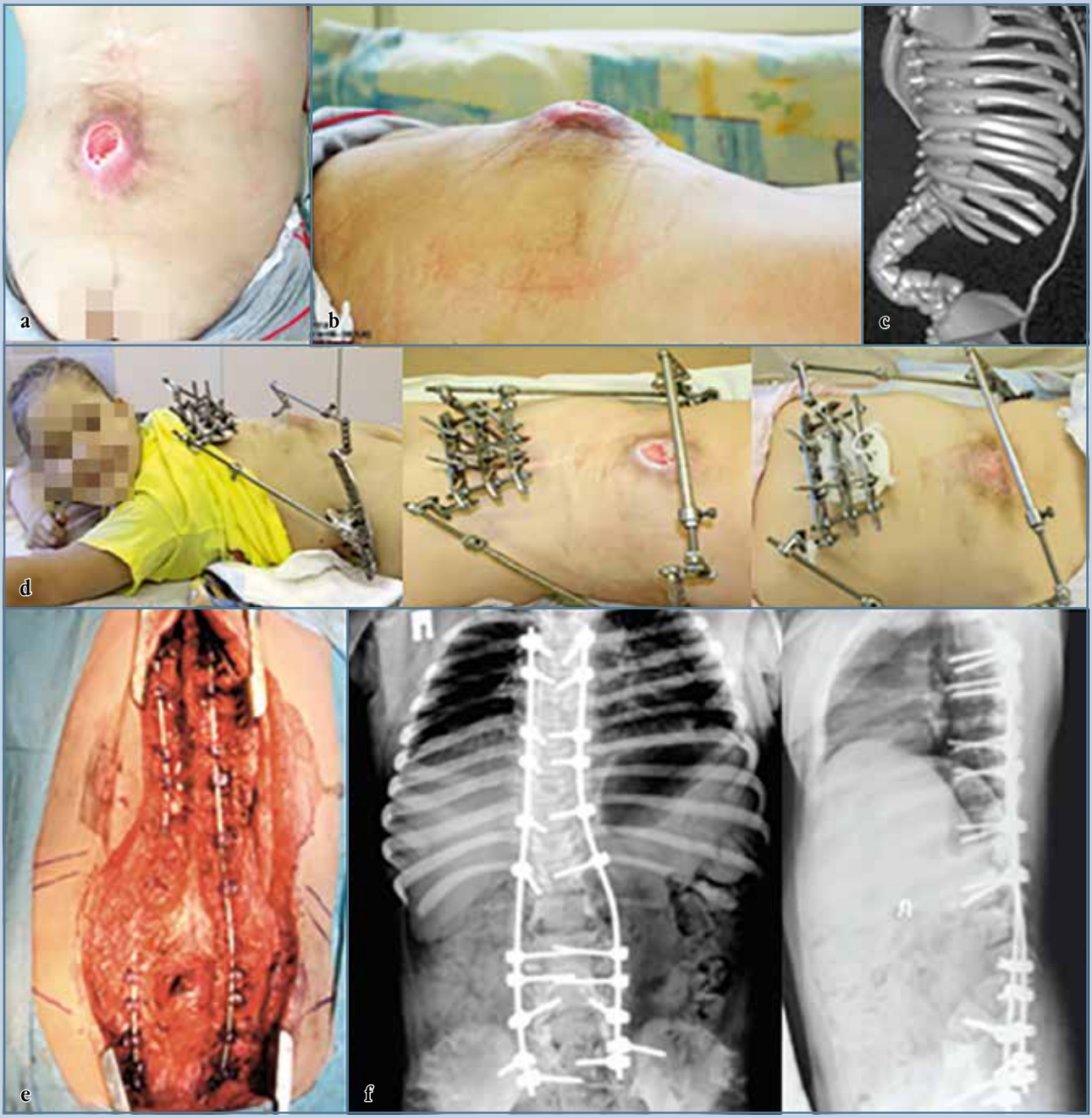

Fig. 3
Staged surgical treatment of post-MMC spinal deformity complicated by a recurrent ulcer in a 9-year-old child: an appearance **(a, b)** and a spinal CT scan **(c)** before starting surgical treatment; an appearance at treatment stages with an external fixation device **(d)**; intraoperative photograph **(e)**; radiographs of the spine after kyphectomy with polysegmental vertebropelvic fixation **(f)**

Table 6

Complications of surgical treatment for spinal deformities associated with spina bifida consequences and methods for their prevention [30, 37–39]

Complication	Rate	Prevention methods	Treatment
Suture failure	10 %	Thorough wound closure with adaptation of edges without tension and, if possible, with excision of scars	Re-closure of the wound, closure of defects by plastic movement of flaps at excessive tension of edges; upon lack of tissue for defect closure (pronounced rough scars) – preventive placement of subcutaneous expanders
Failure of metal devices usually, pulling out lower support elements)	15 %	Clear preoperative planning; adequate assessment of sizes of supporting bones and corresponding elements of devices, strength of supporting bones, primarily caudal ones (sacrum and pelvis), and planning of staged surgery	Replacement of instrumentation; if necessary, changing fixation points of support elements
Surgical site infection	5 %	Preoperative sanitation of chronic infection foci (especially urinary infection), active wound drainage, hematoma evacuation and serous drainage, adequate antibiotic therapy, and ulcer prevention	Early revision and sanitation surgery to prevent infection spread

References

1. **Siegel MJ.** Ultrassonografia Pediatrica. 3rd ed. Rio de Janeiro, RJ: Guanabara Koogan, 2003:37–108.
2. **Lindseth RE.** Myelomeningocele spine. In: Weinstein SL (ed.), The Pediatric Spine: Principles and Practice. Vol. 2. New York: Raven Press, 1994:1043–1067.
3. **Ulrikh EV, Mushkin AYu, Gubin AV.** Vertebral Pathology in Syndromes. Novosibirsk, 2016. In Russian.
4. **Rogala EJ, Drummond DS, Gurr J.** Scoliosis: incidence and natural history. A prospective epidemiological study. J Bone Joint Surg Am. 1978;60:173–176. DOI: 10.2106/00004623-197860020-00005.
5. **Federal Se**rvice of State Statistics. Official statistics/Population/Healthcare. [Electronic resource]. URL: http://www.gks.ru/wps/wcm/connect/rosstat_main/rosstat/ru/statistics/population/healthcare. In Russian.
6. **Vakkasov NY, Akhmediev MM.** The role of the diagnostic algorithm and treatment of children with congenital spina bifida. Pediatric Neurosurgery and Neurology. 2015;(2):55–60. In Russian.
7. **Parisini P, Greggi T, Di Silvestre M, Giardina F, Bakaloudis G.** Surgical treatment of scoliosis in myelomeningocele. Stud Health Technol Inform. 2002;91:442–447. DOI: 10.3233/978-1-60750-935-6-442.
8. **Park TS, Cail WS, Maggio WM, Mitchell DC.** Progressive spasticity and scoliosis in children with myelomeningocele: Radiological investigation and surgical treatment. J Neurosurg. 1985;62:367–375. DOI: 10.3171/jns.1985.62.3.0367.
9. **Tachdijian MO.** Myelomeningocele/scoliosis. In: Pediatric Orthopaedics. Philadelphia: WB Saunders, 1990:1843–1848.
10. **Trivedi J, Thomson JD, Slakey JB, Banta JV, Jones PW.** Clinical and radiographic predictors of scoliosis in patients with myelomeningocele. J Bone Joint Surg Am. 2002;84:1389–1394. DOI: 10.2106/00004623-200208000-00014.
11. **Eysel P, Hopf C, Schwarz M, Voth D.** Development of scoliosis in myelomeningocele. Differences in the history caused by idiopathic pattern. Neurosurg Rev. 1993;16:301–306. DOI: 10.1007/BF00383841.
12. **Guille JT, Sarwark JF, Sherk HH, Kumar SJ.** Congenital and developmental deformities of the spine in children with myelomeningocele. J Am Acad Orthop Surg. 2006;14:294-302. DOI: 10.5435/00124635-200605000-00005.
13. **Carstens C, Koch H, Brocai DR, Niethard FU.** Development of pathological lumbar kyphosis in myelomeningocele. J Bone Joint Surg Br. 1996;78:945–950.
14. **Ryabykh SO, Pavlova OM, Savin DM, Burtsev AV, Gubin AV.** Surgical management of myelomeningocele-related spinal deformities. World Neurosurg. 2018;112:e431–e441. DOI: 10.1016/j.wneu.2018.01.058.
15. **Kahanovitz N, Duncan JW.** The role of scoliosis and pelvic obliquity on functional disability in myelomeningocele. Spine. 1981;6:494–497. DOI: 10.1097/00007632-198109000-00012.
16. **Muller EB, Nordwall A, Oden A.** Progression of scoliosis in children with myelomeningocele. Spine. 1994;19:147–150.
17. **Hudgins RJ, Gilreath CL.** Tethered spinal cord following repair of myelomeningocele. Neurosurg Focus. 2004;16:E7. DOI: 10.3171/foc.2004.16.2.8.
18. **Herman JM, McLone DG, Storrs BB, Dauser RC.** Analysis of 153 patients with myelomeningocele or spinal lipoma reoperated upon for a tethered cord. Pediatr Neurosurg. 1993;19:243–249. DOI: 10.1159/000120739.
19. **Sarwark JF, Weber DT, Gabrieli AP, McLone DG, Dias L.** Tethered cord syndrome in low motor level children with myelomeningocele. Pediatr Neurosurg. 1996;25:295–301. DOI: 10.1159/000121143.
20. **Bowman RM, Mohan A, Ito J, Seibly JM, McLone DG.** Tethered cord release: a long-term study in 114 patients: Clinical article. J Neurosurg Pediatr. 2009;3:181–187. DOI: 10.3171/2008.12.PEDS0874.
21. **Dias MS.** Neurosurgical causes of scoliosis in patients with myelomeningocele: an evidence-based literature review. J Neurosurg Pediatr. 2005;103:24–35. DOI: 10.3171/ped.2005.103.1.0024.
22. **Samuelsson L, Skoog M.** Ambulation in patients with myelomeningocele: a multivariate statistical analysis. J Pediatr Orthop. 1988;8:569–575.
23. **Sharma S, Wu C, Andersen T, Wang Y, Hansen ES, Bunger CE.** Prevalence of complications in neuromuscular scoliosis surgery: a literature meta-analysis from the past 15 years. Eur Spine J. 2013;22:1230–1249. DOI: 10.1007/s00586-012-2542-2.
24. **Samdani AF, Fine AL, Sagoo SS, Shah SC, Cahill PJ, Clements DH, Betz RR.** A patient with myelomeningocele: is untethering necessary prior to scoliosis correction? Neurosurg Focus.2010;29:E8. DOI: 10.3171/2010.3.FOCUS1072.
25. **Yuan N, Fraire JA, Margetis MM, Skaggs DL, Tolo VT, Keens TG.** The effect of scoliosis surgery on lung function in the immediate postoperative period. Spine. 2005;30:2182–2185. DOI: 10.1097/01.brs.0000181060.49993.4a.
26. **Wai EK, Owen J, Fehlings D, Wright JG.** Assessing physical disability in children with spina bifida and scoliosis. J Pediatr Orthop. 2000;20:765–770. DOI: 10.1097/00004694-200011000-00013.
27. **Wai EK, Young NL, Feldman BM, Badley EM, Wright JG.** The relationship between function, self-perception, and spinal deformity: implications for treatment of scoliosis in children with spina bifida. J Pediatr Orthop. 2005;25:64–69. DOI: 10.1097/00004694-200501000-00015.
28. **Sharma S, Wu C, Andersen T, Wang Y, Hansen ES, Bunger CE.** Prevalence of complications in neuromuscular scoliosis surgery: a literature meta-analysis from the past 15 years. Eur Spine J. 2013;22:1230–1249. DOI: 10.1007/s00586-012-2542-2.
29. **Gubin AV, Koryukov AA, Reznik A., Ryabykh SO, Savin DM, Mikhaylov AG, Mukhtyaev SV, Pukhov SP.** Complex treatment of a female patient with multiple developmental abnormalities using surgical intervention and prosthetics/orthotics. Genij Ortopedii. 2016;(1):40–43. In Russian.
30. **Boemers TM, Soorani-Lunsing IJ, de Jong TP, Pruijs HE.** Urological problems after surgical treatment of scoliosis in children with myelomeningocele. J Urol. 1996;155:1066–1069.
31. **Wild A, Haak H, Kumar M, Krauspe R.** Is sacral instrumentation mandatory to address pelvic obliquity in neuromuscular thoracolumbar scoliosis due to myelomeningocele? Spine. 2001;26:E325–E329. DOI: 10.1097/00007632-200107150-00019.
32. **Maloney WJ, Rinsky LA, Gamble JG.** Simultaneous correction of pelvic obliquity, frontal plane, and sagittal plane deformities in neuromuscular scoliosis using a unit rod with segmental sublaminar wires: a preliminary report. J Pediatr Orthop. 1990;10:742–749.
33. **Rodgers WB, Williams MS, Schwend RM, Emans JB.** Spinal deformity in myelodysplasia. Correction with posterior pedicle screw instrumentation. Spine. 1997;22:2435–2443.
34. **Smith JT, Novais E.** Treatment of Gibbus deformity associated with myelomeningocele in the young child with use of the vertical expandable prosthetic titanium rib (VEPTR): a case report. J Bone Joint Surg Am. 2010;92:2211-2215. DOI: 10.2106/JBJS.I.00856.
35. **Guille JT, Betz RR, Balsara RK, Mulcahey MJ, D'Andrea LP, Clements DH.** The feasibility, safety, and utility of vertebral wedge osteotomies for the fusionless treatment of paralytic scoliosis. Spine. 2003;28:S266–S274. DOI: 10.1097/01.BRS.0000092485.40061.ED.
36. **Cardoso M, Keating RF.** Neurosurgical management of spinal dysraphism and neurogenic scoliosis. Spine. 2009;34:1775–1782. DOI: 10.1097/BRS.0b013e3181b07914.
37. **Drummond DS, Moreau M, Cruess RL.** The results and complications of surgery for the paralytic hip and spine in myelomeningocele. J Bone Joint Surg Br. 1980;62:49–53.

38. **Hatlen T, Song K, Shurtleff D, Duguay S.** Contributory factors to postoperative spinal fusion complications for children with myelomeningocele. Spine. 2010;35:1294–1299. DOI: 10.1097/BRS.0b013e3181bf8efe.
39. **Sponseller PD, LaPorte DM, Hungerford MW, Eck K, Bridwell KH, Lenke LG.** Deep wound infections after neuromuscular scoliosis surgery: a multicenter study of risk factors and treatment outcomes. Spine. 2000;25:2461–2466. DOI: 10.1097/00007632-200010010-00007.

Address correspondence to:
Ryabykh Sergey Olegovich
Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics,
Marii Ulyanovoy str., 6, Kurgan 640014, Russia,
rso_@mail.ru

Received 11.09.2018
Review completed 22.11.2018
Passed for printing 22.12.2018

Sergey Olegovich Ryabykh, DMSc, Head of the Clinic of Spine Pathology and Rare Diseases, Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, Marii Ulyanovoy str., 6, Kurgan, 640014, Russia, rso@mail.ru;
Aleksandr Yuryevich Mushkin, DMSc, Prof., chief researcher, St. Petersburg Research Institute of Phthisiopulmonology, Politekhnicheskaya str., 32, St. Petersburg, 194064, Russia, aymushkin@mail.ru;
Dmitry Mikhailovich Savin, MD, PhD, head of traumatologic-orthopedic department No. 9 of the Clinic of Spine Pathology and Rare Diseases, Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, Marii Ulyanovoy str., 6, Kurgan, 640014, Russia, savindm81@mail.ru.

Invitation for discussion

At symposium "Orthopedic and Neurosurgical Problems in Children with Spina Bifida" that was held in Moscow on October 20–21, 2015 and was devoted to medical and social care of children who underwent surgical treatment for spina bifida, specialists in various fields, such as children neurologists, urologists, neurosurgeons, orthopedists, rehabilitologists, as well as children's parents decided to create a comprehensive rehabilitation program aimed at improving social adaptation and quality of life as well as facilitating care for these patients.

One of the directions of comprehensive rehabilitation is the management of vertebral post-MMC – a complex of pathological manifestations of spinal lesions in the form of spinal deformities that are characterized by various shapes, severity, complications, and, accordingly, types of social care, have a different prognosis, and, if necessary, require differentiated surgical treatment.

Surgeons with certain experience in treating spinal deformities in children have prepared recommendations that, in our opinion, should simplify and unify approaches to the management of patient. However, we consider it to be reasonable to bring up the guidelines for discussion by a wide range of specialists before approval of the guidelines by associations of orthopedists, neurosurgeons, and spinal surgeons. The journal of Spine Surgery (Hirurgia Pozvonocnika) is objectively the optimal place for this discussion that will be useful before the guidelines become a normative document.

The authors will gratefully accept additions and comments.