RISK FACTORS FOR THE PRESENCE OF SYRINGOMYELIA IN IDIOPATHIC SCOLIOSIS: ANALYSIS OF 3,285 CASES AND BRIEF LITERATURE REVIEW

V.V. Belozerov, M.V. Mikhaylovskiy
Novosibirsk Research Institute of Traumatology and Orthopaedics n.a. Ya.L. Tsivyan, Novosibirsk, Russia

Objective. To analyze possible radiological and clinical risk factors for syringomyelia in patients with scoliotic deformity.
Material and Methods. An analysis of data from 3,285 patients with idiopathic scoliosis treated from 1997 to 2020 was performed. Syringomyelia was detected in 38 (1.16 %) cases. In 26 cases, syringomyelia was combined with Chiari malformation. In order to search for information on the topic under discussion, the international databases (Scopus, Medline, and GoogleScholar) were used. Additional search for publications listed in the article references was carried out. According to the literature data, possible risk factors for the presence of syringomyelia have been identified.
Results. Among patients with confirmed syringomyelia, a left-sided thoracic scoliotic curve was found in 23.7 % of cases, a double thoracic curve in 18.4 %, an upper thoracic scoliotic curve in 7.9 %, an increase in thoracic kyphosis in 60.5 %, and initial neurological deficit in 47.4 %, which is significantly higher than the same indicators in the entire group of patients. The ratio of male and female patients in the entire group was 1.0 : 6.7, in the group with syringomyelia — 1.00 : 1.71.
Conclusion. According to the literature review and statistically significant factors identified in the analysis of the monocentric group, it can be said that the left-sided thoracic scoliotic curve, upper thoracic curve, double thoracic curve, increased thoracic kyphosis, initial pyramidal insufficiency in the lower extremities, male sex, and a decrease in the initial somatosensory evoked potentials are risk factors for syringomyelia. If the above factors are present in a patient, the MRI study may be recommended to determine further treatment tactics. Before surgical correction of scoliotic deformity, MRI is recommended for all patients.
Key Words: syringomyelia, scoliosis, spinal deformity, risk factors.
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Syringomyelia is one of the most common comorbidities in patients with idiopathic scoliosis. The incidence rate of syringomyelia is 1.2–9.7 % in adolescents and adults and up to 21.7 % in children under the age of 10 years [1–4]. Clinical manifestations may include sensitive ataxia, dissociated sensory deficits, and paresis in the limbs leading to their atrophy [5]. Patients undergoing surgical intervention for scoliosis correction typically have no complaints of syringomyelia symptoms [6]. MRI is considered as the modern standard for preoperative examination of patients with scoliosis. It allows diagnosing both spinal malformations and various types of maldevelopment of other organs and systems [7]. Some authors even recommend performing MRI of the entire spinal cord in order to more comprehensively assess the patient's condition [4]. Up to 13 % of the patients requiring surgical correction of scoliosis and diagnosed with asymptomatic syringomyelia have indications for neurosurgery [8, 9]. Considering this fact, it is reasonable to question whether scoliotic patients who have no indications for surgical correction of the deformity require an MRI. This applies especially to patients under the age of 10 years, since they have a high risk of both scoliotic deformity progression and development of neurological symptoms [10]. There are many studies aimed to identify radiological and clinical risk factors for diagnosing intraspinal abnormalities before conducting MRI. Some of these works are multicenter studies. However, the results vary significantly. Some authors [11, 12] deny a relationship between radiological parameters and MRI findings, while others [13–16] find some correlation, such as side and localization of the primary scoliotic curve, kyphotic component, as well as the patient's age and gender.

The aim of the study is to analyze possible radiological and clinical risk factors for syringomyelia in patients with scoliosis.

This is a retrospective monocentric study.

Material and Methods

A total of 3,285 patients with idiopathic scoliosis underwent surgical correction of spinal deformity at the Clinic for Pediatric and Adolescent Vertebrology of Novosibirsk Research Institute of

Traumatology and Orthopaedics n.a. Ya.L. Tsivyan in the period of 1997–2020. The minimum scoliotic curve angle among the patients was 33°. The mean age at the time of admission was 16.2 (range, 2–61) years. There were 427 males and 2,858 females (ratio, 1.0 : 6.7). Patients with epilepsy, various types of encephalopathy, and delayed psychomotor development were excluded from the study. All patients underwent conventional radiography, MRI, and examination by a neurologist. Traction test with full body weight was performed in cases with the scoliotic curve of >80°.

Syringomyelia was diagnosed in 38 (1.16 %) cases: 14 males and 24 females (ratio, 1.00 : 1.72). Syringomyelia was combined with Chiari malformation in 26 cases.

The following pattern of patient distribution based on the location of syringomyelic cysts was observed: seven cases had cysts in the cervical spine, 20 patients had cervicothoracic cysts, nine individuals were diagnosed with thoracic syringomyelia, and two patients developed lumbar pathology. The mean age of the patients was 15 (range, 10–36) years. Twenty (52.6 %) patients had neither symptoms nor clinical manifestations of syringomyelia.

In order to search for relevant information in the literature, Scopus, Medline, and GoogleScholar international databases were used. Additional search for publications listed in the article references was carried out. According to the literature data, the following risk factors were identified: left thoracic curve, double thoracic curve, upper thoracic curve, right lumbar curve, increased thoracic kyphosis, male gender, age of ⩽10 years, and absence of abdominal reflexes [14–18].

The criteria were analyzed using the methods of descriptive statistics. The χ^2 test, McNemar's test, and Fisher's exact test for independent samples were applied to compare the qualitative characteristics of dependent groups. The strength of correlation coefficient relationships was evaluated using the following scale of ρ value intervals: <0.19 – very weak relationship, 0.20–0.29 – weak relationship, 0.30–0.49 – moderate relationship, 0.50–0.69 – medium relationship, and >0.70 – strong relationship. The relationship was considered significant for value ⩾0.3.

Results and Discussion

A total of 124 (3.80 %) patients with left thoracic curve were identified in the total group, of which nine (7.30 %) patients had syringomyelia.

There were 21 (0.92 %) cases of syringomyelia among 2,281 patients with right thoracic curve (69.40 %).

Double thoracic curve was diagnosed in 274 (8.3 %) individuals; seven (2.6 %) of them had syringomyelia (Fig. 1).

Upper thoracic curves were noted in 23 (0.7 %) patients, with three cases (13.0 %) of syringomyelia among them.

Right lumbar/thoracolumbar scoliotic curve was detected in 234 (7.1 %) patients. Among these, two (0.86 %) individuals had syringomyelia.

Increased thoracic kyphosis was observed in 699 (21.3 %) cases, 23 (3.2 %) of which were diagnosed with syringomyelia (Fig. 2).

The ratio of male and female patients was 1.0 : 6.7 in the total group and 1.00 : 1.71 among individuals with syringomyelia. The mean age at the time of admission did not differ significantly between genders.

The mobility of the primary scoliotic curve was determined during functional radiography of the spine in the inclined position. Rigid scoliosis with <25 % mobility was noted in 1,431 (43.5 %) cases of the total group and in 19 (1.3 %) individuals with syringomyelia.

The initial neurological deficit, which manifested itself mainly as pyramidal insufficiency in the lower limbs, including the traction-induced one in the group with idiopathic scoliosis, was observed in 85 (2.6 %) cases. Various initial neurological abnormalities were diagnosed in 18 patients of the group with syringomyelia, which accounted for 21.2 % of the total number of patients with neurological deficit.

The patients with confirmed syringomyelia had the following distribution of curve patterns: left thoracic curve (23.7 %), double thoracic curve (18.4 %), upper thoracic curve (7.9 %), right lumbar/thoracolumbar curve (5.3 %), increased thoracic kyphosis (60.5 %), and initial neurological deficit (47.4 %; Table).

We can conclude from the obtained data that male gender, initial neurological deficit, including pyramidal insufficiency in the lower limbs, left scoliotic curve, double thoracic curve, upper thoracic curve, and increased thoracic kyphosis significantly increase the risk of syringomyelia in scoliotic patients. Moderate, medium, and strong relationship was found for these parameters ($p > 0.3$). No statistically significant relationship between cyst localization, scoliotic curve angle, and patient's age was noted. The relationship between these parameters is either weak or very weak ($p < 0.3$).

Immediate correction of scoliosis in the presence of syringomyelia does not differ from that in idiopathic scoliosis without intraspinal anomalies. Intraoperative neurophysiological monitoring is a prerequisite for these surgeries. The outcome of scoliosis correction in syringomyelia is reported to be comparable to the results of correcting rigid spinal deformities [19, 20]. Neurosurgical intervention including formation of a large occipital cistern and/or syringomyelic cyst drainage was shown to reduce the possible risk of complications in spinal deformity surgery [21, 22]. Taking into account that not all patients have clinical manifestations of syringomyelia, while the above-mentioned radiological risk factors are not sufficiently reliable, all patients are recommended to undergo MRI examination prior to scoliosis correction. In our opinion, examination of the entire spine is not always required. Analyzing the regions of the primary scoliotic curve and the cervicothoracic junction, with paying particular attention to the most frequent location of syringomyelic cysts, would be enough for diagnosis [23]. Several issues arise when managing pediatric patients. Firstly, surgeons tend to delay spinal fusion as much as possible till the moment when active bone growth is completed. Secondly, there is

a high risk of progression of both scoliosis and neurological deficit, which is a manifestation of syringomyelia [24]. Of particular interest are patients under the age of 10 years with a scoliotic curvature of >20°, since timely neurosurgery may not only positively influence the disease course but also decrease the need for surgical correction of scoliosis. Our study includes patients with the mean age of 16.2 years. This is due to the fact that it is preferable to conduct surgical correction of scoliosis as late as possible, with involvement of all methods of conservative therapy. The analysis of patients of this age group provides the basis for further multicenter studies with a more detailed assessment of the results of neurosurgical treatment for syringomyelia and changes in the scoliotic curvature. Intraspinal anomalies at this age account for at least 5.6 % according to MRI data [25]. The most important aspect is that a neurosurgery performed at the age of 8–10 years based on indications may not only prevent the development of neurological symptoms but also positively influence scoliosis, while significantly reducing the need for scoliosis correction [8, 9, 25, 26]. There are also reports of asymptomatic disease course, even in the presence of large cysts detected by MRI [27]. A rather promising method for diagnosing syringomyelia in patients with scoliosis is the analysis of somatosensory evoked potentials [28]. The study is often carried out with recording of potentials of the lower extremities, since the curve apex in idiopathic scoliosis is rarely located at ⩾T5. Analysis of somatosensory evoked potentials in the upper extremities contributes to the clinical picture and allows for detection of subtle deviations that may be left unnoticed during clinical examination. The presence of detected deviations in potentials provides more indications for MRI.

Conclusion

According to the literature data and statistically significant factors identified by analyzing the monocentric group, one can conclude that left thoracic curve, upper thoracic curve, double thoracic

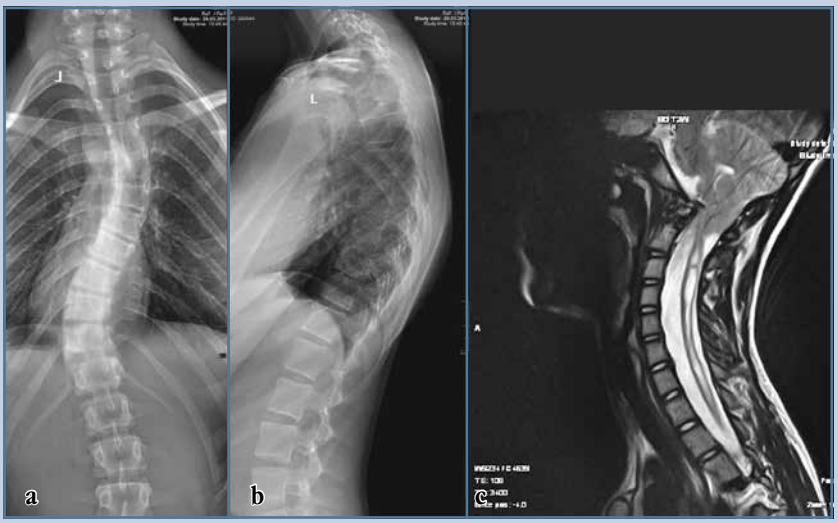

Fig. 1
Spine radiographs and MRI scan of a 14-year-old patient: **a** – spine radiography in the frontal projection: double thoracic scoliotic curve of 40° in the middle thoracic region and of 38° in the lower thoracic region; b – spine radiography in the lateral projection: increased thoracic kyphosis (56°); **b** – MRI scan of the cervical spine, sagittal view: the cerebellar tonsils are elongated, prolapse into the foramen magnum, and located about 20 mm below the Chamberlain line; the spinal canal is sharply expanded, the sagittal diameter is up to 17 mm; the spinal cord is flattened; an intramedullary cyst is detected along the entire cervical region, its dimensions reach up to 5 × 8 mm in the axial projection, the inner contour of the cyst is clear and even, a septum is observed in the cyst at C3 vertebra; initial neurological symptoms are absent

Fig. 2
Spine radiographs and MRI scan of a 28-year-old patient: **a** – spine radiography in the frontal projection: left scoliotic curve of 62°, lumbar anticurvature of 40°; **b** – spine radiography in the lateral projection: increased thoracic kyphosis of 54°; **c** – MRI scan of the cervical spine, sagittal view: a syringomyelic cyst is observed at C2–T2 (total length, ≈98.0 mm; maximum width, ≈6.5 mm), Arnold – Chiari malformation

Table

Distribution of patients of the total group and the group with confirmed syringomyelia by potential risk factors

Parameter	All patients, n (%)	Patients with syringomyelia	
		n (% of the number of identified parameters)	% of the number of patients with syringomyelia
Left thoracic scoliotic curve	124 (3.8)	9 (7.30)	23.7
Right thoracic scoliotic curve	2281 (69.4)	21 (0.92)	55.3
Double thoracic scoliotic curves	274 (8.3)	7 (2.60)	18.4
Upper thoracic scoliotic curve	23 (0.7)	3 (13.00)	7.9
Right lumbar/thoracolumbar scoliotic curve	234 (7.1)	2 (0.86)	5.3
Increased thoracic kyphosis	699 (21.3)	23 (3.20)	60.5
Rigid scoliosis	1431 (43.5)	19 (1.30).	50.0
Initial neurological deficit	85 (2.6)	18 (21.20)	47.4

curve, increased thoracic kyphosis, initial pyramidal insufficiency in the lower limbs, male gender, and decreased somatosensory evoked potentials are risk factors for the presence of syringomyelia.

MRI is recommended for patients falling under these criteria in order to determine further treatment strategy. MRI is preferable for all patients prior to surgical correction of scoliosis.

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Address correspondence to:
Belozerov Vadim Vasilyevich
Novosibirsk Research Institute of Traumatology and Orthopaedics n.a. Ya.L. Tsivyan,
17 Frunze str., Novosibirsk, 630091, Russia,
vad-belozerov@yandex.ru

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Vadim Vasilyevich Belozerov, junior researche, of Research Department of Children and Adolescent Spine Surgery, Novosibirsk Research Institute of Traumatology and Orthopaedics n.a. Ya.L. Tsivyan, 17 Frunze str., Novosibirsk, 630091, Russia, ORCID: 0000-0003-2441-2686, vad-belozerov@yandex.ru;
Mikhail Vitalyevich Mikhaylovskiy, DMSc, Prof., chief researcher of Department of Pediatric Vertebrology, Novosibirsk Research Institute of Traumatology and Orthopaeducs n.a. Ya.L. Tsivyan, 17 Frunze str., Novosibirsk, 630091, Russia, ORCID: 0000-0002-4847-100X, MMihailovsky@niito.ru.