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Benign syringomyelia with the abortive type of the course

Syringomyelia is a chronic disease with progressive cavitation and clinical presentations of spinal cord injury. The paper describes clinical cases of a rare benign variant of syringomyelia with spontaneous cavity collapse. The peculiarity of the described clinical cases is childhood-onset of the disease, lack of progression and/or development of myelopathic symptoms, and signs of syringomyelia cavity collapse according to magnetic resonance imaging findings. The authors designate this childhood-onset variant of the disease as abortive. The tendency towards collapse in the cavity in these patients may be due to a single pathogenetic mechanism, which is of interest for a further investigation.

Keywords: syringomyelia; Chiari malformation; spontaneous collapse; magnetic resonance imaging; posterior cranial fossa.

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Syringomyelia is a disease characterized by formation of progressive fluid-containing cavities in the spinal cord. The emergence of neuroimaging diagnostic methods empowered the research of this disease. It has been shown that primary cavity progression occurs in the first 5–10 years after the disease onset [1]. In the majority of cases, progression of symptoms decelerates with increased duration of the disease, which is associated with a certain degree of reduction of the cavity size due to its spontaneous collapse [2]. On magnetic resonance imaging (MRI) scans the cavity phenomenon can undergo changes from the initial round-oval shape on the axial section to its collapse in a long duration of the disease. It is characterized by various degrees of cavity flattening in the anterior-posterior direction (up to a threadlike shape) with concomitant signs of spinal cord atrophy [1, 2, 3]. Long-term follow-up of unoperated patients has demonstrated that 25.8% of them had a significant decrease in the cavity size over time [1]. This phenomenon, characterized by the full-scale myelopathy clinical picture and the collapsed cavity on MRI, was defined as a stage of prolonged syringomyelia and designated as "post-syrinx" syndrome [3].

According to recent publications, besides the described typical course of the disease, there are variants that have a benign course with the absence of spinal symptoms progression. Asymptomatic syringomyelia can be regarded as one of such variants. This variant is characterized by the presence of a cavity in the spinal cord, which is detected incidentally on MRI during an examination for other diseases, and has no clinical manifestations typical of long-term syringomyelia. The absence of symptoms in

such patients does not correlate with the cavity size [4]. According to T.H. Millorat et al. (1995), symmetrical central cavities tend to have an asymptomatic course [5].

We observed a variant of syringomyelia with a spontaneous collapse of the cavity at the early stages of its development, which can lead to regression of the initial clinical symptoms or to the arrest of their progression. There are few clinical descriptions of such cases in literature [6]. We assume that they occur more frequently, but remain undiagnosed. Identification of such cases and their analysis allow us to determine the mechanisms of the reverse development of syringomyelia with childhood onset. We are presenting 4 clinical cases. The first one contains a complete evidence base for the dynamics of clinical and MRI data, while the others demonstrate a similar course of the disease.

Case 1. Patient S., 9 years old, was referred to a neurologist with complaints of dull pain in the back. Cranial-cerebral nerves (CCN) demonstrated no abnormalities. Active and passive movements in the arms and legs were in full range. Muscle tone was normal. The strength in the limbs was sufficient. Tendon reflexes were equal and brisk. There were no pathological findings. Sensitivity was not affected. Coordination tests were performed satisfactorily. The patient was stable in the Romberg position. The examination revealed kyphoscoliotic deformation of the spine..

Cervicothoracic spine MRI showed a syringomyelia cyst up to 0.8 cm in diameter at the C2 level and lower, the cystic index (CI) was 0.5 (Fig. 1B). MRI scans of the posterior cranial fossa (PCF) revealed Chiari malformation (CM) (2.2 cm

CLINICAL OBSERVATIONS

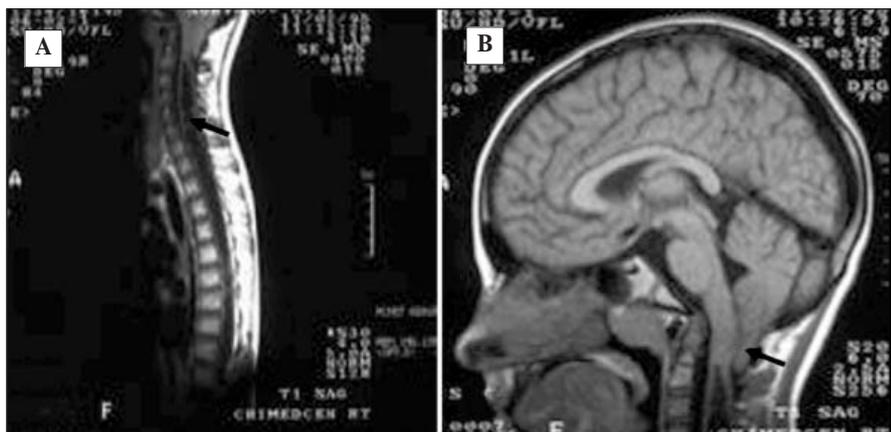


Figure 1. MRI of patient S. at the initial stages of the disease (at the age of 9 years). T1-weighted images: A – Sagittal section of the brain MRI: ectopic cerebellar tonsils – CM (arrow). B – Sagittal section of the cervicothoracic spinal cord MRI: syringomyelic cavity of the cervicothoracic spinal cord (arrow).

lower the foramen magnum), as well as the signs of narrow PCF and disturbance of bone indexes: Boogart's angle increased up to 144° . (Fig. 1A).

Given the lack of clinical manifestations other than the presence of scoliosis, the patient's parents decided not to have their child operated on by a neurosurgeon. The patient "dropped off the radar" for 10 years.

10 years later, patient S. complained of musculoskeletal pain in the interscapular area. The kyphoscoliotic deformation of the spine remained. The patient had growth delay. As for neurological status, there was CCN disease, muscle atrophy, while no disturbances of reflexes or sensitivity were revealed.

Cervical spine MRI revealed an almost complete collapse of the syringomyelic cavity (Figure 2A), the spinal cord

which gradually regressed within 1–3 years. Spine scoliotic deformation and torticollis developed at the preschool age, and later remained at the same level.

CCN showed no abnormalities. Active and passive movements in the arms and legs were in full range. Muscle tone was normal. Limb strength was sufficient – 5 points for all muscle groups. Upper limb reflexes S<D were brisk, while lower limb ones were high S<D with extension of reflexogenic zones. There were no pathological findings. Pain and thermal hyposensitivity was noted in C1–T10 dermatomes on the left. Deep sensitivity was preserved. Coordination tests were performed satisfactorily. The patient was stable in the Romberg position. Minor traces of burns and wounds were noted on the skin of the left hand. S-shaped spine scoliotic deformation, mild scapular hunchback on the left, vicar torticollis to the right and mild partial acromegaly of the left upper limb were revealed.

PCF MRI scans indicated cerebellar tonsils location at the level of the foramen magnum line. Bone indices were normal: declive length was 42 mm, occipital bone length was 39 mm and Klaus index was 39 mm. Boogart's angle was 123° , Welcker's angle was 118° (Figure 3A). Cervicothoracic spine MRI revealed an irregularly shaped syringomyelic cavity sized up to 2–3 mm at C3–T11 segment level located paracentral on axial sections with a shift to the left, CI = 0.4. The spinal cord was flattened and stretched to the sides (Fig. 3B, C).

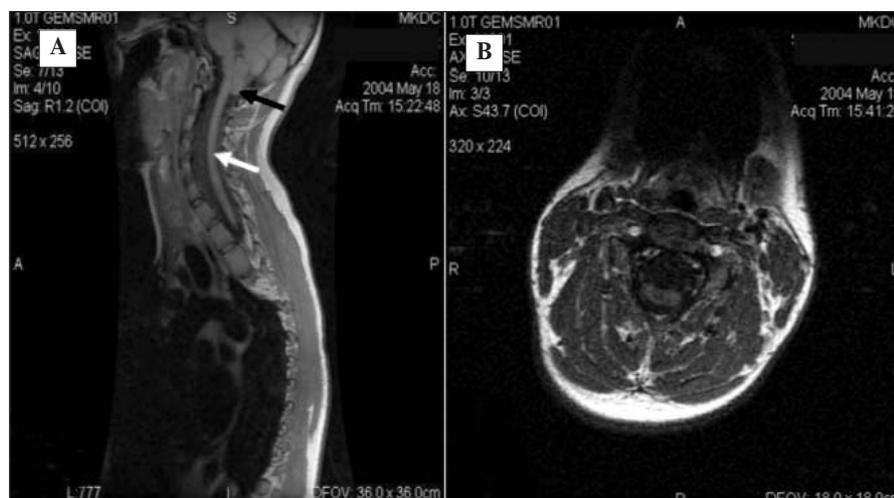


Figure 2. MRI of patient S. (aged 19 years). T1-weighted images:

A – Sagittal section of the cervical-thoracic spine and craniovertebral junction (CVJ): ectopic cerebellar tonsils (black arrow), collapse of the cavity at the cervicothoracic level of the spinal cord (white arrow), pronounced spinal cord atrophy. B – Axial section at the cervical spinal cord level – minimal signs of collapsed cavity, spinal cord atrophy with "flattening" in the anterior-posterior direction.

Case 3. Patient M., 69 years old.

Spine scoliotic deformation development has been noted since the age of 14. It was managed by wearing a hard

corset and exercise therapy. No further progression of the symptoms was marked. She sought neurological care at the age of 69 for the first time because of pain in the neck of a musculoskeletal nature.

CCN showed no abnormalities. Active and passive movements in the arms and legs were in full range. Muscle tone was normal. Limb strength was sufficient. Tendon reflexes were brisk and symmetric. There were no pathological findings. Sensitivity impairment was not noted. Coordination tests were performed satisfactorily. The patient was stable in the Romberg position. Paravertebral neck muscle palpation did not show any tenderness or tension. Spine scoliotic deformation to the right and a mild scapular hunchback on the right were revealed.

MRI performed for the first time at the age of 69 years showed that the cerebellar tonsils were 3 mm lower the foramen magnum line, their edges were round. No cerebrospinal fluid (CSF) circulation disturbance at the foramen magnum level was revealed. The study showed signs of PCF depth reduction: Klaus index was reduced to 34 mm and occipital bone shortened up to 37 mm ($N=39-47$ mm). Declive length, Boogart's angle and Welcker's angle were within the normal range (Fig. 4.A). Spinal cord atrophy and syringomyelic cavity were revealed on the cervical spine MRI sagittal section at C2 level and lower with anterior-posterior size up to 2 mm, $CI = 0.3$ (Fig. 4A, B). The signs of spinal cord atrophy and collapsed cavity (elongated-threadlike shape in anterior-posterior direction) were seen on the axial section.

Case 4. Patient P. visited a neurologist at the age of 54 years with complaints of megalgia episodes in occipital region of a bursting nature, accompanied by balance problem, shaky walking, which have been disturbing her since childhood. There were no other complaints in her medical history. MRI performed for the first time revealed a syringomyelic cavity.

CCN showed no abnormalities. Active and passive movements in the arms and legs were in full range. Muscle tone was normal. Limb strength was sufficient – 5 points for all muscle groups. Tendon reflexes were brisk with mild asymmetry ($S < D$).

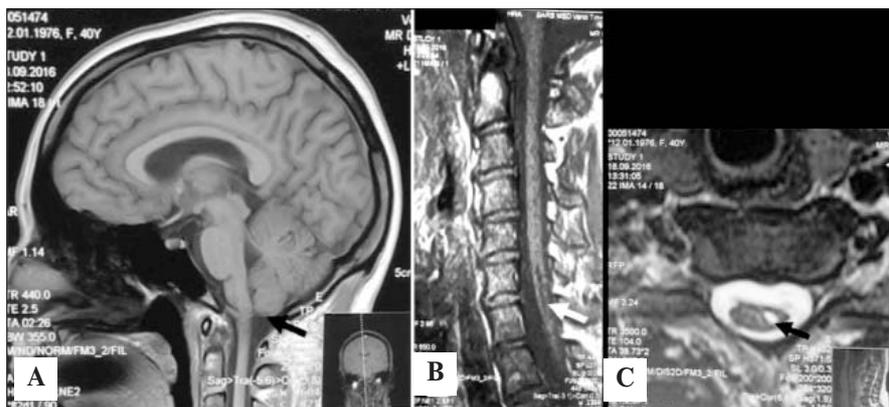


Figure 3. MRI of patient K., 40 years old. A – Sagittal section of the brain in T1-mode: physiological position of the cerebellum (arrow). B – Sagittal section of the cervical spine in T1-mode: a narrow syringomyelic cavity (arrow). C – Axial section of the spinal cord at the cervical region level in T2-mode: an eccentrically located cavity of irregular shape (arrow).

There were no pathological findings. Mild mosaic pain sensitivity impairment was revealed in C4–T3 dermatomes on the right. Thermal and deep sensitivity remained normal. Coordination tests were performed satisfactorily. The patient was stable in the Romberg position.

Brain and cervicothoracic spine MRI showed pronounced ectopia of the cerebellar tonsils 1.7 cm lower the foramen magnum, anterior and posterior subarachnoid space narrowing at the foramen magnum level, distention of III ventricle up to 1.0 cm and the absence of the lower cerebellar cistern. Craniovertebral junction bone indexes were within the normal range. Cervicothoracic spine MRI revealed central syringomyelic cavity at the C4–T7 level sized 2 mm, as well as mild spinal cord atrophy. Signs of cavity collapse and

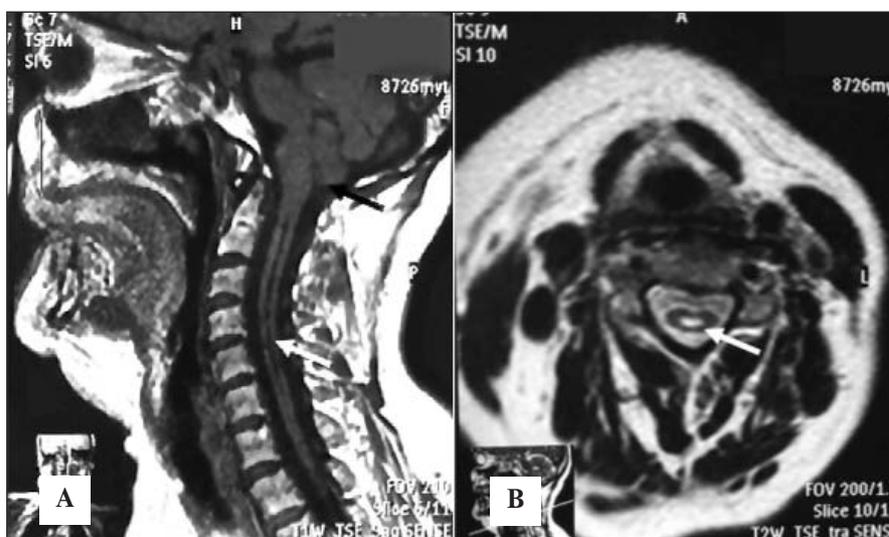


Figure 4. MRI of the patient K., 69 years old. A – Sagittal section of the cervical spine MRI including craniocervical junction in T1-mode: cerebellar tonsils are located 3 mm below the foramen magnum level (black arrow), narrow cavity at the cervical spinal cord level (white arrow) and spinal cord atrophy. B – Axial section of the spinal cord at the cervical region level in T2-mode: collapsed cavity (arrow), spinal cord is "flattened" in the anterior-posterior direction.

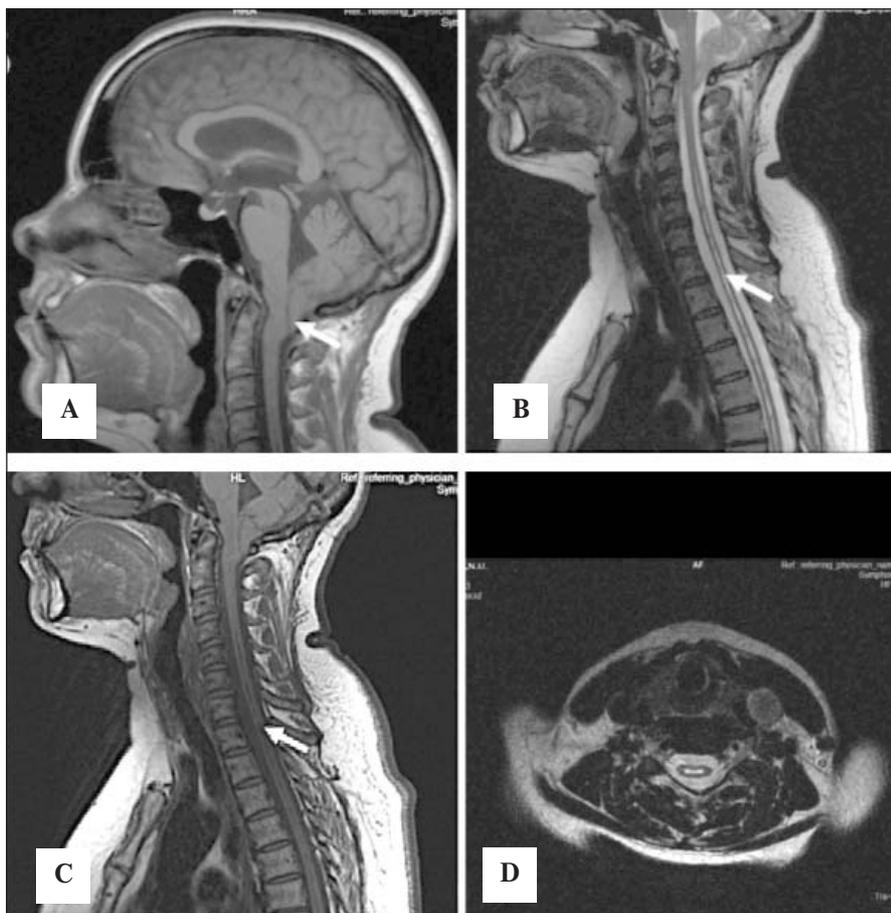


Figure 5. MRI of patient P., 54 years old. A – Sagittal section of the brain in T1 mode: pronounced ectopia of the cerebellar tonsils. B, C – Sagittal section of the cervical spine MRI in T2 (B.) and T1 (C.) modes: narrow syringomyelic cavity (arrow), signs of spinal cord atrophy. D – Axial section of the spinal cord at the cervical region level in T2 mode: signs of flattened (threadlike shaped) collapsed cavity and spinal cord atrophy (spinal cord deformation with "flattening" in the anterior-posterior direction).

spinal cord atrophy (threadlike, flattened shape of the spinal cord) were seen on the axial section, CI was 0.4.

Discussion. The analysis of the presented cases showed almost complete cavity regression and spinal atrophy on sagittal and axial sections in Case 1. Spinal cord atrophy is the result of brain tissue compression by a cavity of considerable size and integration of remaining parenchyma volume after cavity regression (complete or partial). Clinical and MRI dynamics of Case 1 indicate the presence of a wide cavity at the initial stages of the disease, accompanied by scoliosis, which is the most frequent debuting symptom of syringomyelia in children. At a distant stage of the patient's illness, cavity collapse and cerebellar tonsils elevation were not associated with any new symptoms. We have designated such type of interruption of the further development of syringomyelia as abortive type of the disease. The other three cases are similar. In our opinion, they also represent an abortive type of the disease. In all these cases the disease began in childhood (with initial symptoms of scoliosis in 2 cases and symptoms of CM in 4 cases). Clinical manifestations of spinal cord damage

were minimal in cases 2 and 3 and had no progression or were absent in case 4. In cases 2–4 the cavities were revealed at a mature or elderly age during an examination for other reasons. MRI data in all cases (2–4) revealed narrow cavities (2 mm) that had the sign of collapse on axial sections (a "flattened" threadlike cavity). There were also signs of spinal cord atrophy on sagittal and axial sections. All these data allow us to assume that the arrest of the disease progression at the early stage in childhood is associated with spontaneous collapse of the cavity. A significant factor is the cavity collapse at the early stages of the disease before the development of irreversible damage to the spinal cord or severe myelopathic symptoms due to prolonged pressure on the spinal cord parenchyma by the cavity. The influence of cavity long existence on the course of syringomyelia was confirmed by the results of surgical treatment of syringomyelia at the early and late stages of the disease. It is known that in long-lasting syringomyelia the possibility of significant neurologic regression after surgery is minimal [7].

To date, dozens of cases have been described along with the assumption of the mechanism of cavity spontaneous collapse in syringomyelia [6]. Most often, collapse of the cavity was verified in childhood and was accompanied by cerebellar tonsils elevation [8–12]. This is supposed to be related to the skeleton growth in children, to the increase of the PCF volume and, as a consequence, tonsil elevation, restoration of normal CSF circulation in the foramen magnum area and subsequent disappearance of the syringomyelic cavity [8]. Unlike children, spontaneous cavity collapse was accompanied by cerebellar tonsils elevation only in 1/3 of cases in adult patients, indicating different mechanisms of cavity resolution in adults and children. In 1991 C.R Jack et al. explained the adult case of a cavity collapse without regression of CM degree by an increase in intracavitary pressure and rupture of the spinal cord tissue, which led to the development of drainage between the syringomyelic cavity and spinal subarachnoid space resulting in the leak of the cavity contents [13]. Spontaneous cavity collapse, accompanied by cerebellar tonsil elevation in adults, could not have occurred because of the PCF growth. In this connection, J.

Klekamp et al. (2001) suggested the presence of arachnoid adhesions at the level of the foramen magnum and foramen of Magendie, which could be the cause of subarachnoid space obstruction. Their rupture led to the restoration of liquor outflow [14]. As for the cases of spontaneous isolated CM regression in adults, a hypothesis has been put forward on the change in the ratio between the PCF and cerebellum volume due to the age-related cerebellum atrophy [15]. This theory is applicable to elderly patients with syringomyelia associated with CM. In 1998 F. Fukutake at T. Hattori [16] published an observation that exclusion of physical exertion and other Valsalva-like maneuvers resulting in increased piston movement of the cerebellar tonsils [17] may lead to regression of syringomyelia associated with CM.

Taking into account the mechanisms proposed in literature, it is possible to distinguish 2 main groups of theories of spontaneous cavity collapse: 1) CSF flow restoration through the foramen magnum (cerebellar tonsil elevation in response to the PCF growth [8–12]; arachnoid adhesion rupture at the foramen magnum level [14, 18]; Valsalva-like maneuvers elimination [16, 19–21], cerebellar tonsils elevation due to the brain atrophy [15]); 2) development of drainage between the cavity and the spinal subarachnoid space [13, 22, 23].

It is important to note that it is not always cerebellar tonsils elevation in connection with the PCF growth that leads to spontaneous cavity collapse in childhood. In addition to this mechanism, syringomyelic cavities may collapse in children, as in adults, as a result of drainage into the spinal subarachnoid space, as well as after elimination of Valsalva-like maneuvers provoking syringomyelia development in pathological conditions at the foramen magnum level [24, 25].

We assume that the basis for spontaneous collapse described in Cases 1–3 was the cerebellar tonsils elevation leading to the reversal of CSF circulation block which was related to the patients' growth and PCF distention, as evidenced by the presence of small PCF size on the initial MRI in Case 1, as well as preservation of minimal signs of PCF shallowness in Case 3. Cerebellar tonsils elevation was verified in the first case. In Cases 2 and 3 cerebellum was already in a physiological position at the time of the examination, which suggests the possibility of cerebellar tonsils elevation in childhood. In the last case, the cerebellar tonsils had a pronounced ectopia with preserved CSF circulation blockage at craniovertebral junction level. Therefore, the most likely mechanism of cavity collapse in this case is development of spontaneous drainage between the cavity and the spinal subarachnoid space.

The natural course of syringomyelia is poorly understood to date. It raises multiple questions for clinicians in choosing further patient management strategy in children, especially in those having asymptomatic course or minimal manifestations. The cases of spontaneous resolution of syringomyelia in childhood provide the opportunity to consider the ambiguity of surgical intervention in patients with minor symptoms and a cavity of small or medium size on MRI. The choice of conservative strategy involves a thorough neurologic examination and repeated MRI with a frequency of at least once in every 6 months in order to monitor neurological symptom progression and MRI indices to the age of skeleton and PCF full growth.

Despite the existence of numerous hypotheses, the specific mechanism leading to spontaneous syringomyelia resolution remains unknown. Further research is needed to explain the mechanism of spontaneous syringomyelia resolution and to discover new directions of treatment.

1. Менделевич ЕГ, Давлетшина РИ, Валиева ЛК. Клинические и нейровизуальные варианты течения сирингомиелии, манифестировавшей в различные возрастные периоды. *Неврологический вестник*. 2012;(4):45-50. [Mendelevich EG, Davletshina RI, Valieva LK. Clinical and neurovisual variants of the course of syringomyelia manifested in different age periods. *Neurologicheskii vestnik*. 2012;(4):45-50. (In Russ.)].
2. Bogdanov EI, Mendelevich EG. Syrinx size and duration of symptoms predict the pace of progressive myelopathy: retrospective analysis of 103 unoperated cases with craniocervical junction malformations and syringomyelia. *Clin Neurol Neurosurg*. 2002 May;104(2):90-7.
3. Bogdanov EI, Heiss JD, Mendelevich EG. The post-syrinx syndrome: stable central myelopathy and collapsed or absent syrinx. *J Neurol*. 2006 Jun;253(6):707-13. Epub 2006 Mar 6.
4. Nishizawa S, Yokoyama T, Yokota N, et al. Incidentally identified syringomyelia associated with Chiari I malformations: is early interventional surgery necessary? *Neurosurgery*. 2001 Sep;49(3):637-40; discussion 640-1.
5. Milhorat TH, Johnson RV, Milhorat RN, et al. Clinicopathological correlations in syringomyelia using axial magnetic resonance imaging. *Neurosurgery*. 1995 Aug;37(2):206-13.
6. Менделевич ЕГ, Нуруллина ЧС. Спонтанный регресс сирингомиелии – редкий вариант течения заболевания: анализ клинико-нейровизуальных описаний. *Неврологический вестник*. 2018;(1):54-60. [Mendelevich EG, Nurullina ChS. Spontaneous regression of syringomyelia is a rare variant of the disease course: analysis of clinical and neuroimaging descriptions. *Neurologicheskii vestnik*. 2018;(1):54-60. (In Russ.)].
7. Alfieri A, Pinna G. Long-term results after posterior fossa decompression in syringomyelia with adult Chiari type I malformation. *J Neurosurg Spine*. 2012 Nov;17(5):381-7. doi: 10.3171/2012.7.SPINE12272. Epub 2012 Aug 31.
8. Sudo K, Doi S, Maruo Y, et al. Syringomyelia with spontaneous resolution. *J Neurol Neurosurg Psychiatry*. 1990 May;53(5):437-8.
9. Tokunaga M, Minami S, Isobe K, et al. Natural history of scoliosis in children with syringomyelia. *J Bone Joint Surg Br*. 2001 Apr;83(3):371-6.
10. Mallinger B, Marson F, Sevely A, et al. Spontaneous resolution of syringomyelia in a child with Chiari I malformation: a case report. *J Radiol*. 2004 Nov;85(11):1943-6.
11. Mazumder AK, Das S, Krishnan P. Spontaneous resolution of Chiari malformation and associated syringomyelia. *Neurol India*. 2016 Nov-Dec;64(6):1335-1336. doi: 10.4103/0028-3886.193819.
12. Ramnarayan R, Ganesh CVS, Kumar R. Spontaneous Resolution of Chiari I-Associated Syringomyelia: A Report of Two Cases. *Pediatr Neurosurg*. 2018;53(4):238-242. doi: 10.1159/000488461. Epub 2018 May 7.
13. Jack CR, Kokmen E, Onofrio B. Resonance imaging findings. *J Neurosurg*. 1991 Feb;74(2):283-6.
14. Klekamp J, Iaconetta G, Samii M. Spontaneous resolution of Chiari I malformation and syringomyelia: case report and review of the literature. *Neurosurgery*. 2001 Mar;48(3):664-7.
15. Castillo M, Wilson JD. Spontaneous resolution of a Chiari I malformation: MR demonstration. *AJNR Am J Neuroradiol*. 1995 May;16(5):1158-60.
16. Fukutake T, Hattori T. Reversible hydromyelia in a synchronised swimmer with recurrent girdle pains. *J Neurol Neurosurg Psychiatry*. 1998 Oct;65(4):606.
17. Oldfield EH, Muraszko K, Shawker TH, et al. Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils: Implications for diagnosis and treatment. *J Neurosurg*. 1994 Jan;80(1):3-15.
18. Muthukumar N, Christopher J. Spontaneous resolution of Chiari I malformation and associated syringomyelia following parturition. *Acta Neurochir (Wien)*. 2013 May;155(5):817-8. doi: 10.1007/s00701-013-1620-5. Epub 2013 Jan 26.
19. Khanna AR, Coumans JV. Spontaneous Improvement of Chiari I Malformation and Syringomyelia in a Patient with Cystic Fibrosis: Case Report. *Neurosurgery*. 2016 Feb;78(2):E305-8. doi: 10.1227/NEU.0000000000000980.
20. Kyoshima K, Bogdanov EI. Spontaneous resolution of syringomyelia: report of two cases and review of the literature. *Neurosurgery*. 2003 Sep;53(3):762-8; discussion 768-9.
21. Perrini P. Spontaneous resolution of syringomyelia in an adult patient with tight cisterna magna. *Neurol Sci*. 2012 Dec;33(6):1463-7. doi: 10.1007/s10072-012-0946-8. Epub 2012 Jan 19.
22. Sung WS, Chen YY, Dubey A, Hunn A. Spontaneous regression of syringomyelia — review of the current aetiological theories and implications for surgery. *J Clin Neurosci*. 2008 Oct;15(10):1185-8. doi: 10.1016/j.jocn.2007.08.017. Epub 2008 Aug 16.
23. Vaquero JS, Ferreira E, Parajon A. Spontaneous resolution of syrinx: report of two cases in adults with Chiari malformation. *Neurol Sci*. 2012 Apr;33(2):339-41. doi: 10.1007/s10072-011-0670-9. Epub 2011 Jun 28.
24. Coloma-Valverde G. Spontaneous resolution of the syrinx. A case report and survey of the literature. *Rev Neurol*. 2003 Jun 16-30;36(12):1156-8.
25. Rafia S, Pascual-Castroviejo I. Syringohydromyelia: Report of a case, which resolved spontaneously. *Rev Neurol*. 2001 Apr 1-15;32(7):635-7.

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