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Contemporary concepts of the vertebral column additional cervical ribs anatomy

Daniil N. Strelkov[1], Elena Yu. Kalinina[1], Ekaterina V. Shcherbakova[2], Andrei M. Fedyuk[1], Andrei A. Sevruk[1], Aleksandra D. Kramarenko[1], Karina I. Lazareva[1], Anna V. Kogan[3], Lyubov M. Strelkova[4]

[1] Saint Petersburg State Pediatric Medical University, 2 Litovskaya str., Saint Petersburg, 194100, Russian Federation

[2] Central Pathological Department of General Pathology of the Leningrad Region Bureau of Forensic Medicine, 7 Santiago de Cuba str., Saint Petersburg, 194291, Russian Federation

[3] Pavlov First Saint Petersburg State Medical University, 6-8 L'va Tolstogo str., Saint Petersburg, 197022, Russian Federation

[4] City Polyclinic No. 81, 54A Kazanskaya str., Saint Petersburg, 196000, Russian Federation

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Corresponding author:
Daniil N. Strelkov.
E-mail:
cataliz123@yandex.ru

Abstract. The additional ribs of the cervical spine, or cervical ribs, are a fairly rare developmental anomaly that occurs as a result of a mutation of the *HOX* genes responsible for the formation of the bone system. This developmental feature is most often localized in the C_7 region, however, there are also possible formation variants in which it can be found in the C_4–C_6 projection. Cervical ribs are classified according to their size and the presence of a fibrous junction with the first rib. Different authors disagree on which variant of formation prevails, one-sided or two-sided. It is also believed that the cervical ribs mainly develop on the left. This feature is more common in the female population. For the most part, the anomaly is asymptomatic. However, sometimes, in the presence of certain developmental features of the body, as well as a number of provoking factors, a condition called thoracic outlet syndrome may occur. This syndrome may differ clinically depending on the effect of the cervical ribs on nearby structures: the brachial plexus, subclavian artery and/or subclavian vein. The diagnosis is made on the basis of a physical examination, instrumental diagnostic data methods and certain provocative tests. Treatment can be both conservative and surgical if medication and physiotherapy are ineffective. Surgical treatment may include resection of the first rib and scalenotomy. This article presents a literary review of scientific papers using PubMed and eLibrary resources and scientific journals over the past 5 years. The aim of the work is to summarize the available data on the prevalence, causes of formation and clinical symptoms caused by additional cervical ribs.

Keywords: additional cervical ribs, thoracic outlet syndrome, interscalene space

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Современные представления об анатомии добавочных ребер шейного отдела позвоночного столба

Даниил Николаевич Стрелков[1], Елена Юрьевна Калинина[1], Екатерина Васильевна Щербакова[2], Андрей Михайлович Федюк[1], Андрей Андреевич Севрук[1], Александра Дмитриевна Крамаренко[1], Карина Игоревна Лазарева[1], Анна Вадимовна Коган[3], Любовь Михайловна Стрелкова[4]

[1] Санкт-Петербургский государственный педиатрический медицинский университет, 194100, г. Санкт-Петербург, ул. Литовская, д. 2, Российская Федерация

[2] Бюро судебно-медицинской экспертизы, 194291, г. Санкт-Петербург, ул. Сантьяго-де-Куба, д. 7, Российская Федерация

[3] Первый Санкт-Петербургский государственный медицинский университет имени академика И.П. Павлова, 197022, г. Санкт-Петербург, ул. Льва Толстого, д. 6–8, Российская Федерация

[4] Городская поликлиника № 81, 196000, г. Санкт-Петербург, ул. Казанская, д. 54, литера А, Российская Федерация

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Автор для корреспонденции:
Даниил Николаевич Стрелков.
E-mail: cataliz123@yandex.ru

Резюме. Добавочные ребра шейного отдела позвоночного столба, или шейные ребра, довольно редкая аномалия развития, возникающая в результате мутации *HOX*-генов, ответственных за формирование костной системы. Данная особенность развития чаще всего локализуется в области VII шейного позвонка (C_7), однако возможны также варианты формирования, при которых она может обнаруживаться в проекции C_4–C_6. Шейные ребра классифицируют в зависимости от размера и наличия фиброзного соединения с I ребром. Различные авторы расходятся во мнении, какой вариант формирования преобладает, односторонний или двусторонний. Также считается, что шейные ребра преимущественно развиваются слева. Чаще данная особенность встречается у женского населения. В большинстве своем аномалия клинически не проявляется. Однако иногда, при наличии определенных особенностей развития организма, а также ряда провоцирующих факторов, может возникнуть состояние, называемое синдромом грудного выхода. Данный синдром может клинически различаться в зависимости от влияния шейных ребер на расположенные рядом структуры: плечевое сплетение, подключичную артерию и/или подключичную вену. Диагноз ставят на основании физикального обследования, а также благодаря инструментальным методам исследования и определенным провокационным пробам. Лечение может быть как консервативным, так и хирургическим при неэффективности медикаментозной и физиотерапии. Оперативное вмешательство может включать резекцию I ребра и скаленотомию. В данной статье представлен литературный обзор научных трудов с использованием таких ресурсов, как поисковые системы PubMed и eLibrary, а также научных журналов за последние 5 лет. Целью работы является обобщение данных о распространенности, причинах формирования и клинических симптомах, вызываемых добавочными шейными ребрами.

Ключевые слова: добавочные шейные ребра, синдром грудного выхода, межлестничное пространство

Источник финансирования. Авторы заявляют об отсутствии внешнего финансирования при проведении исследования.

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HISTORY

Accessory ribs of the cervical spine are supernumerary ribs that represent an elongation arising at the level of the transverse process, most commonly of the C_7 vertebra [1, 2]. This anomaly, known as the "rib of Eve," is referenced in the works of Galen from the 2nd century AD and later in the writings of Vesalius from the 16th century AD [3, 4].

EMBRYOGENESIS

The skeleton develops from the mesoderm. Mesodermal cells give rise to somites on either side of the neural tube, which further subdivide into a ventral sclerotome and a dorsal dermatome. The cells of the sclerotome transform into mesenchymal cells that will give rise to the future ribs during the fourth week of embryogenesis. These cells are guided by *HOX* genes and growth differentiation factor 11 (GDF11). Initially, 29 pairs of rudimentary ribs form from the mesenchyme, after which the 12 pairs of ribs projecting from the 1–12 thoracic vertebrae continue to develop. The remaining 17 pairs normally undergo regression; however, should this fail to occur for any reason, they may ossify and present either as elongated transverse processes or as fully formed accessory ribs [5–8]. Given that the rudiments regress in a craniocaudal direction, accessory ribs arising from C_7 are encountered most frequently in clinical practice, followed by those arising from C_6 and C_5 [1]. The rarest variant is the formation of this anomaly at the C_4 level [9, 10]. Early in development, cervical ribs are thought to arise from a mutation in *HOX* gene expression [11, 12].

ANATOMY

A cervical rib consists of a head, a neck, and a tubercle. The rib ends may terminate freely within the soft tissues or articulate with the 1st rib near the insertion of the anterior scalene muscle [1, 13] (Fig. 1). The literature contains conflicting data regarding the prevalence and incidence of the various morphological forms of this anomaly. Some sources report that unilateral cervical ribs are more common than bilateral ones (Fig. 2, 3), with a predilection for the left side [1, 13, 14]. Other sources, however, indicate that cervical ribs are bilateral in 50–80% of cases [5] (Fig. 4, 5). This discrepancy is most likely attributable to differences in the patient cohorts studied. There is also evidence that this developmental anomaly occurs twice as frequently in women as in men [5, 15]. The most authoritative work on cervical ribs is the monograph by the Russian anatomist W.L. Gruber. His classification, based on the degree of osseous development and the presence of an articulation with the 1st rib, delineates four types of cervical ribs (Table 1) [5, 16, 17].

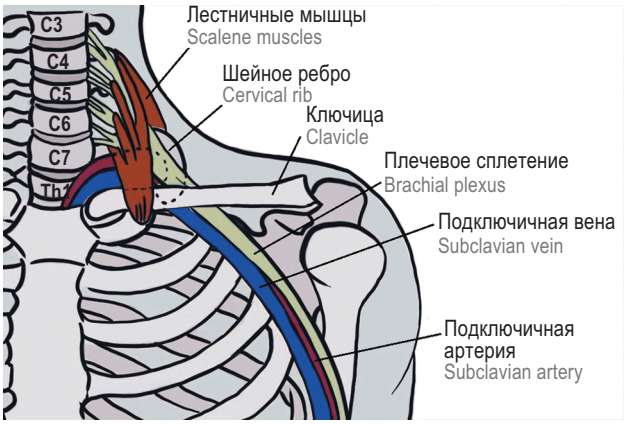


Fig. 1. **Relative position of the structures of the upper chest aperture in the presence of an accessory cervical rib (the author of the illustration is A.V. Kogan)**

Рис. 1. Взаимное расположение структур верхней апертуры грудной клетки при добавочном шейном ребре (автор иллюстрации А.В. Коган)

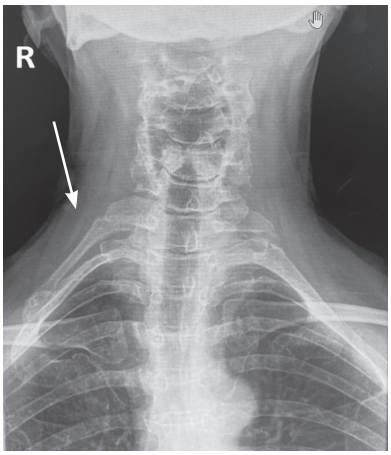


Fig. 2. **X-ray of patient F. A full-fledged accessory cervical rib is detected on the right, and hypertrophy of the transverse process of C7 on the left. The arrow points to the right-hand fully formed cervical rib (from personal photo archive of L.M. Strelkova)**

Рис. 2. Рентгенография пациента Ф. Определяются полноценное добавочное шейное ребро справа, гипертрофия поперечного отростка С7 слева. Стрелка указывает на правостороннее полностью сформированное шейное ребро (из личного фотоархива Л.М. Стрелковой)

CLINICAL MANIFESTATIONS

Despite the spectrum of developmental variations, cervical ribs serve no established physiological function in the body [18]. In most instances, cervical ribs are incidental findings on routine chest or cervical spine

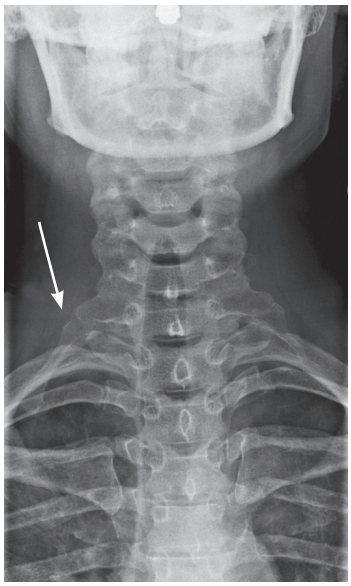


Fig. 3. **X-ray of patient M. A full-fledged accessory cervical rib is detected on the right, as well as a fragment of a rib on the left. The arrow points to the right-sided underdeveloped cervical rib) (from personal photo archive of L.M. Strelkova)**

Рис. 3. Рентгенография пациентки М. Определяется полноценное добавочное шейное ребро справа, а также фрагмент ребра слева. Стрелка указывает на недоразвитое шейное ребро с правой стороны (из личного фотоархива Л.М. Стрелковой)

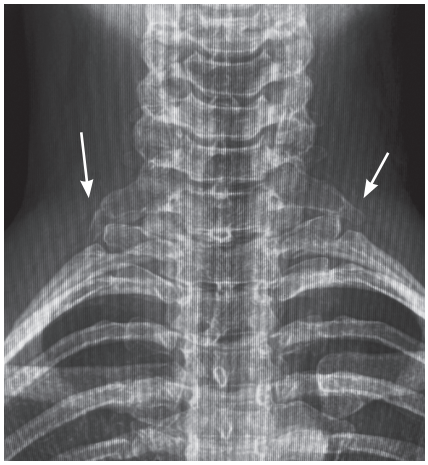


Fig. 4. **X-ray of patient G. Bilateral underdeveloped cervical ribs are detected. The arrows point to the underdeveloped bilateral cervical ribs (from personal photo archive of L.M. Strelkova)**

Рис. 4. Рентгенография пациента Г. Определяются двусторонние слаборазвитые шейные ребра. Стрелки указывают на недоразвитые двусторонние шейные ребра (из личного фотоархива Л.М. Стрелковой)

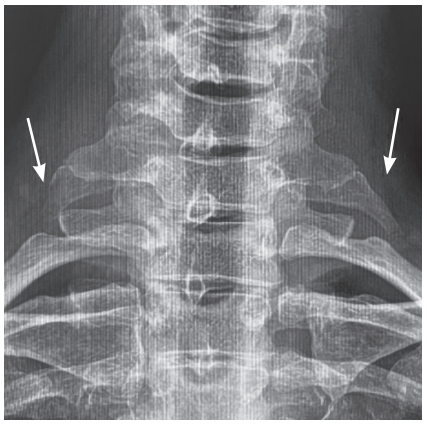


Fig. 5. **X-ray of patient T. Bilateral cervical ribs are detected with a more pronounced left rib. The arrows point to the underdeveloped bilateral cervical ribs (from personal photo archive of L.M. Strelkova)**

Рис. 5. Рентгенография пациентки Т. Определяются двусторонние шейные ребра с более выраженным левым ребром. стрелки указывают на недоразвитые двусторонние шейные ребра (из личного фотоархива Л.М. Стрелковой)

Table 1

Classification table of cervical ribs based on their length according to W.L. Gruber [5, 16]

Таблица 1

Таблица классификации шейных ребер в зависимости от их длины по В.Л. Груберу [5, 16]

Типы шейных ребер Cervical ribs types	Степень развития костных структур Degree of bone structures development
I тип Type I	Ребро чуть больше поперечного отростка C_7 длиной менее 2,5 см Rib less than 2.5 cm
II тип Type II	Ребро длиной более 2,5 см Slightly over 2.5 cm
III тип Type III	Полное шейное ребро, которое соединяется с I ребром фиброзной связкой A complete cervical rib connected to rib 1 by a fibrose ligament
IV степень 4th degree	Полное шейное ребро, которое соединяется с I ребром посредством хряща A complete cervical rib connected to rib 1 by a cartilage

radiographs. The anomaly is predominantly clinically asymptomatic [13, 14, 19, 20]. In a subset of cases, however, thoracic outlet syndrome (also termed superior thoracic aperture syndrome in some publications) may arise, particularly in the presence of specific additional anatomical variants and secondary changes in the adjacent structures [14]. A congenital cervical rib constitutes one of the predisposing factors and is implicated in up to 5% of cases of this condition [21].

Superior thoracic aperture syndrome was characterized as a distinct clinical entity in 1958 [22]. The superior thoracic aperture is the space between the thorax and the axilla, transmitting the subclavian vein, subclavian artery, and brachial plexus [23]. The space is bounded by the scalene muscles, the 1st rib, the clavicle, and, in some individuals, a cervical rib [24]. Thoracic outlet syndrome is an uncommon condition (1–3 cases per 100,000 population) caused by compression of the neurovascular bundle at the superior thoracic aperture. In some cases, the condition is exacerbated by hypertrophy of the anterior scalene muscles, which may impinge upon the brachial plexus and/or the subclavian vessels [19, 25, 26]. Thoracic outlet syndrome is categorized as neurogenic, arterial, or venous, according to the compromised structure [27, 28]. It is postulated that short, incomplete ribs tend to produce neurogenic compression, whereas long or complete ribs predominantly contribute to vascular manifestations [29]. The neurogenic form is the most common in both sexes, representing up to 95% of all diagnosed cases [30]. In the pediatric population, the venous type is the most frequent presentation of thoracic outlet syndrome [1, 31]. While anatomical and mechanical factors, together with sex-based differences in pain perception, are thought to contribute, the definitive reasons for the disparity in incidence between men and women remain incompletely elucidated [32]. Compression of the neurovascular bundle occurs most frequently at the following anatomical sites. These include: 1) the superior pleural sulcus; 2) the interscalene triangle (between the anterior and middle scalene muscles); 3) the costoclavicular space (between the clavicle and the 1st rib); and 4) the subpectoral space (between the pectoralis minor and subscapularis muscles) [33]. A cervical rib is regarded as the second most prevalent anatomical anomaly giving rise to neurovascular bundle compression [34, 35].

The pathogenesis of thoracic outlet syndrome is form-dependent. In the neurogenic form, pectoralis minor hyperactivity predominates, resulting in altered scapular position and narrowing of the retropectoral space. As a result, arm elevation leads to compression of the brachial plexus. In the vascular form (both arterial and venous), contraction of the scalene muscles elevates the 1st rib, thereby narrowing the costoclavicular space. The subclavian vein is the primary structure compressed, while the subclavian artery is affected to a lesser degree. Cervical ribs exacerbate these changes by reducing the interscalene space. Persistent pulsation of the subclavian artery generates friction between the vessel wall and the adjacent osseous structures, which may lead to fibrosis of the arterial wall, as well as stenosis or aneurysm formation, potentially complicated by intraluminal thrombus development [36–39]. This may be further complicated by subclavian artery thrombosis and, in rare instances, by stroke resulting from retrograde thromboembolism [40].

The majority of patients present with neurological symptoms: numbness and paresthesia in the little and ring fingers within the ulnar nerve distribution (C_8 and Th_1 nerve roots). Compression of the subclavian vein causes the affected limb to become swollen and cyanotic due to impaired venous return to the heart [13]. When an accessory rib lies in close proximity to the arterial vessel (the subclavian artery), compression may occur, up to and including critical impairment of blood supply. This condition has been termed Sistine Chapel syndrome, which develops secondary to an abrupt reduction in vertebral artery blood flow and consequent brainstem ischemia [16]. In some cases, secondary Raynaud's phenomenon — a peripheral circulatory disorder attributable to vasospasm — is identified [6].

DIAGNOSIS

Superior thoracic aperture syndrome may be diagnosed on the basis of clinical symptoms and physical examination findings; however, the diagnosis must be corroborated by instrumental investigations [27]. The diagnostic workup includes radiography, magnetic resonance imaging, multislice computed tomography angiography, electroneuromyography, and ultrasound [5, 42, 43]. Ultrasound has a sensitivity of merely 60–70% and is operator-dependent. The principal limitations of this modality relate to the difficulty of visualizing structures and the inability to adequately assess osseous anomalies. Moreover, challenges are encountered during provocative maneuvers: upon arm abduction, the acoustic window becomes obscured by the clavicle [44]. A range of clinical tests are also employed to elicit compression, including the Adson test, the abduction test, the hyperabduction test, the military posture test, and the three-minute stress test [5, 45].

Thoracic outlet syndrome may result from various types of trauma, repetitive movements, anatomical anomalies within the space bounded by the first thoracic vertebra, the 1st rib, and the manubrium of the sternum, as well as postural abnormalities and an excessively developed chest [33, 46] (Fig. 6). Sustained occupational loads may provoke fibrosis of the scalene muscles or displacement of a cervical rib,



Fig. 6. **3D volumetric reconstruction obtained by performing a CT scan of the cervical spine. Bilateral accessory ribs are visualized at the C7 level (https://radiopaedia.org/cases/cervical-rib-12?lang=us)**

Рис. 6. Объемная 3D-реконструкция, полученная при проведении компьютерной томографии шейного отдела позвоночного столба. Визуализируются двусторонние добавочные ребра на уровне C7 (https://radiopaedia.org/cases/cervical-rib-12?lang=us)

which, in turn, may lead to compression of the brachial plexus [27, 47, 48].

TREATMENT

Most authors agree that treatment of thoracic outlet syndrome should be initiated with conservative measures: pharmacotherapy, physiotherapy, therapeutic exercise, and massage [5]. Surgical treatment is also recommended following unsuccessful conservative therapy of a total duration ranging from 6 months to 2 years, as well as in cases where arterial complications arise [49, 50]. The primary surgical procedure is complete rib resection. The operative approach may vary; the most commonly performed procedures are brachial plexus decompression and scalenotomy (division of the scalene muscles with or without resection of the 1st rib) [13, 27, 51]. The history of thoracic outlet syndrome surgery dates back to 1927, when scalene muscle division was first performed. In 1962, the first resection of the 1st rib via a posterior approach was undertaken, and in 1966, a less invasive transaxillary approach was introduced. At present, no consensus exists regarding the choice of surgical technique; however, the majority of surgeons favor the supraclavicular approach, which permits scalenotomy, exploration of the brachial plexus branches, resection of the 1st rib, and decompression of the subclavian artery [5, 52].

CONCLUSION

Thus, accessory cervical ribs are a relatively uncommon developmental anomaly of osseous structures in the general population (>1% of the population) and are frequently asymptomatic. In most instances, they represent an incidental finding on routine radiographic examinations. Accessory ribs per se do not require treatment or ongoing surveillance; therapy is prescribed only when complications develop, such as thoracic outlet syndrome. Thoracic outlet syndrome, depending on the structure affected, may be neurogenic, arterial, or venous and may be attended by a range of neurological and/or vascular disturbances. The choice between conservative management and surgical intervention is determined by the severity and intensity of the presenting symptoms.

ADDITIONAL INFORMATION

Author contribution. D.N. Strelkov — development of the concept, collection of material, writing, formatting of the article; E.Yu. Kalinina — development of the concept, approval of the final version of the work; E.V. Shcherbakova — preparation and editing of the work; A.M. Fedyuk — collection of material, editing of the work; A.A. Sevruk — development of the concept; A.M. Kramarenko — editing of the work; K.I. Lazareva — collection of materials; A.V. Kogan, L.M. Strelkova — registration and provision of materials from the personal archive.

All authors made a substantial contribution to the conception of the study, acquisition, analysis, interpretation of data for the work, drafting and revising the article, final approval of the version to be published and agree to be accountable for all aspects of the study.

Competing interests. The authors declare that they have no competing interests.

ДОПОЛНИТЕЛЬНАЯ ИНФОРМАЦИЯ

Вклад авторов. Д.Н. Стрелков — разработка концепции, сбор материала, написание, оформление статьи; Е.Ю. Калинина — разработка концепции, утверждение итоговой версии работы; Е.В. Щербакова — подготовка и редактирование работы; А.М. Федюк — сбор материала, редактирование работы; А.А. Севрук — разработка концепции; А.М. Крамаренко — редактирование работы; К.И. Лазарева — сбор материала; А.В. Коган, Л.М. Стрелкова — оформление и предоставление материалов из личного архива.

Все авторы внесли существенный вклад в разработку концепции, проведение исследования и подготовку статьи, прочли и одобрили финальную версию перед публикацией.

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Authors' info

*** Daniil N. Strelkov** — Assistant, Department of Human Anatomy. E-mail: cataliz123@yandex.ru; https://orcid.org/0009-0004-1928-9753
Elena Yu. Kalinina — Can. Sci. (Med.), Associate Professor, Department of Human Anatomy. E-mail: drkalinina@yandex.ru; https://orcid.org/0000-0001-7077-3584; SPIN: 1176-5739
Ekaterina V. Shcherbakova — Pathologist. E-mail: maestrovody@mail.ru; https://orcid.org/0000-0002-3818-1535; SPIN: 7685-0130
Andrei M. Fedyuk — Lecturer, Department of Human Anatomy. E-mail: andrej.fedyuk@gmail.com; https://orcid.org/0000-0002-2378-2813; SPIN: 3477-0908
Andrei A. Sevruk — Lecturer, Department of Human Anatomy. E-mail: sevruka@mail.ru; https://orcid.org/0009-0008-6949-3241; SPIN: 1976-5178
Aleksandra D. Kramarenko — Assistant, Department of Human Anatomy. E-mail: al.kramarenko97@mail.ru; https://orcid.org/0009-0008-8351-9233
Karina I. Lazareva — Assistant, Department of Human Anatomy. E-mail: dr.lki@mail.ru; https://orcid.org/0009-0007-1655-8630
Anna V. Kogan — student. E-mail: anakogan6@gmail.com; https://orcid.org/0009-0006-5802-1922
Lyubov M. Strelkova — radiologist. E-mail: zvereva.lm@yandex.ru; https://orcid.org/0009-0006-0766-7440

* Corresponding author.
Автор, ответственный за переписку.

Об авторах

*** Даниил Николаевич Стрелков** — ассистент, кафедра анатомии человека. E-mail: cataliz123@yandex.ru; https://orcid.org/0009-0004-1928-9753
Елена Юрьевна Калинина — к.м.н., доцент, заведующий кафедрой анатомии человека. E-mail: drkalinina@yandex.ru; https://orcid.org/0000-0001-7077-3584; SPIN: 1176-5739
Екатерина Васильевна Щербакова — врач-патологоанатом. E-mail: maestrovody@mail.ru; https://orcid.org/0000-0002-3818-1535; SPIN: 7685-0130
Андрей Михайлович Федюк — преподаватель, кафедра анатомии человека. E-mail: andrej.fedyuk@gmail.com; https://orcid.org/0000-0002-2378-2813; SPIN: 3477-0908
Андрей Андреевич Севрук — преподаватель, кафедра анатомии человека. E-mail: sevruka@mail.ru; https://orcid.org/0009-0008-6949-3241; SPIN: 1976-5178
Александра Дмитриевна Крамаренко — ассистент, кафедра анатомии человека. E-mail: al.kramarenko97@mail.ru; https://orcid.org/0009-0008-8351-9233
Карина Игоревна Лазарева — ассистент, кафедра анатомии человека. E-mail: dr.lki@mail.ru; https://orcid.org/0009-0007-1655-8630
Анна Вадимовна Коган — студент. E-mail: anakogan6@gmail.com; https://orcid.org/0009-0006-5802-1922
Любовь Михайловна Стрелкова — врач-рентгенолог. E-mail: zvereva.lm@yandex.ru; https://orcid.org/0009-0006-0766-7440