

# Anatomy-based treatment strategy for abdominal and retroperitoneal cystic lymphatic malformations in children

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Lymphatic malformations (LMs) are rare congenital benign malformations of the lymphatic system, consisting of fluid-filled cysts or channels with unusual growth. The low incidence of LMs leads to a challenging differential diagnosis and a complicated choice of further treatment strategies. In this study, we analyzed our experience in the treatment of abdominal and retroperitoneal cystic LMs and proposed an anatomy-based treatment strategy that could improve the treatment outcomes.

**OBJECTIVE** — to prove that the anatomical localization of LM influences treatment effectiveness and should be taken into account when making treatment decisions.

**MATERIALS AND METHODS.** Out of 240 pediatric patients who underwent treatment for cystic LMs at a single center from December 2012 to December 2020, 43 (19.1 %) were diagnosed with abdominal and retroperitoneal LMs. The follow-up period was  $3.50 \pm 2.16$  years.

**RESULTS.** Surgical resection of abdominal LMs without evident connection with cisterna chyli does not lead to a recurrence. Sclerotherapy is the best treatment option for retroperitoneal LMs. Diffuse mesentery affection can be successfully treated by sirolimus systemic therapy. According to a logistic regression model, initial choice of LM treatment without considering anatomical localization influences the risks of LM treatment failure ( $p = 0.000503$ ). All patients in our study group received the following treatments: laparoscopic resections ( $n = 10$ , 23.2 %), video-assisted resections ( $n = 11$ , 25.5 %), laparotomy resections ( $n = 9$ , 20.9 %), sclerotherapy ( $n = 4$ , 9.3 %), sclerotherapy and surgery combination ( $n = 1$ , 2.3 %), sirolimus systemic therapy ( $n = 2$ , 4.6 %), and splenectomy ( $n = 1$ , 2.3 %). Six (13.4 %) patients are under dynamic observation. The recommended technique for treating abdominal LMs produced excellent outcomes in 35 (81.4 %) patients, good outcomes in 5 (11.6 %) patients, satisfactory outcomes in 2 (4.7 %) patients, and unsatisfactory outcomes in 1 (2.3 %) patient.

**CONCLUSIONS.** Treatment strategies for abdominal and retroperitoneal LMs should be based on their anatomical localization. Retroperitoneal localization indicates a high risk of surgical treatment failure ( $p = 0.0006$ ).

## KEYWORDS

abdominal and retroperitoneal lymphatic malformations, children, sclerotherapy, minimally invasive surgery, abdominal cysts.

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Lymphatic malformations (LMs) are congenital benign malformations of the lymphatic system, consisting of disorganized spongy fluid-filled cysts or fluid-filled channels with unusual growth. According to the ISSVA 2018 classification, cystic LMs can be macrocystic, microcystic, or mixed type [1, 7]. Abdominal LMs have a relatively rare incidence [17], amounting to up to 5 % [9], or less than 10 % [14] of all LMs. Retroperitoneal LMs are even rarer, representing less

than 1 % of all cystic LMs [16]. It results in a challenging differential diagnosis and a complicated choice of further treatment strategies. Abdominal and retroperitoneal LMs treatment is preferred to be planned, that could be beneficial for following outcome prognosis. The hypotheses of our study suggest that the anatomical localization of LM and involvement of the cisterna chyli must serve as the foundation for the selection of the LM treatment approach.

**OBJECTIVE** — to prove that the anatomical localization of LM influences treatment effectiveness and should be taken into account when making treatment decisions.

## Materials and methods

The Committee on Clinical Investigation of Bogomolets National Medical University approved this study (Protocol #127; 02.12.2019). All investigations were conducted in accordance with applicable regulations in consideration of GCP-ICH and the Declaration of Helsinki [18]. All parents or legal guardians of participants provided their written informed consent.

Patient data was collected from case records and analyzed retrospectively. Out of 240 pediatric patients who underwent treatment for cystic LMs at a single center from December 2012 to December 2020, 43 (19.1 %) patients were diagnosed with abdominal and retroperitoneal LMs. A follow-up period assessment was completed by December 2021. The mean follow-up was  $3.5 \pm 2.16$ .

Table 1. **Characteristics of study group patients**

Characteristic	Total
Male	25 (58.1 %)
Female	18 (41.9 %)
Age at diagnosis, years	
0–1	13 (30.2 %)
2–5	16 (37.3 %)
6–9	7 (16.3 %)
10–11	2 (4.6 %)
≥ 12	5 (11.6 %)
Anatomic location	
Omentum	7 (16.3 %)
Intestines	18 (41.9 %)
Retroperitoneum	10 (23.4 %)
Retroperitoneum+cisterna chyli connection	3 (6.9 %)
Mesentery	2 (4.6 %)
Spleen	3 (6.9 %)
Treatment option	
Surgery	34 (79.1 %)
Sclerotherapy	4 (9.3 %)
Surgery+sclerotherapy	1 (2.3 %)
m-Tor inhibitors systemic therapy	2 (4.6 %)

The following criteria were analyzed for each patient: gender, age at disease onset, common clinical symptoms and signs, complications, treatment options, and treatment outcomes.

As part of the examination process, a CBC with platelet count and a coagulation panel with D-dimer and fibrinogen were done. Each patient underwent initial screening using gray-scale ultrasonography (US), color Doppler, and spectral Doppler tracings. These diagnostic procedures were also prescribed at later stages of treatment and after the follow-up period. In most cases, MRI was typically used to confirm the diagnosis ( $n = 26$ ) and to evaluate the effectiveness of the treatment ( $n = 28$ ) during the follow-up period. Nine patients underwent a CT scan before treatment.

Treatment outcomes were considered «excellent» when LM's reduction reached 90–100 %, «good» when it reached 60–89 %, «satisfactory» when it reached 20–59 %, and «unsatisfactory» when it reached 0–19 %.

Data distributions were compared using the paired Student's t-test or Wilcoxon criteria. The logistic regression model was built to identify factors that could influence the risk of poor treatment outcomes. Statistical analysis was performed using IBM SPSS for Windows version 24.0 (IBM Corp., Armonk, NY) and EZR (R-statistics). A P-value of  $< 0.05$  was considered statistically significant.

## Results and discussion

Table 1 shows patient data. Male predominance is seen in the study group. At routine pregnancy examinations, LMs were detected prenatally in 5 (11.6 %) patients. LMs were discovered at various ages after birth, mostly at young ages. 29 (67.4 %) patients had their LMs identified when they were under 5 years of age. 37 (86 %) patients had solitary lesions, 3 (7.0 %) patients had them in combination with mediastinal LMs, and the other 3 (7 %) patients had them in combination with axillar and neck masses as part of the GLA-syndrome.

5 (11.6 %) patients had previously undergone surgeries for LMs that ultimately led to recurrence.

In 14 (34.2 %) cases, children were admitted to the emergency department with acute pain and required urgent surgery. A surgical procedure was planned for the remaining 27 (65.8 %) patients.

According to our hypothesis, all patients were divided into subgroups based on the anatomical location of cystic LMs.

Subgroup 1. Omental LMs. Omental LMs are the most favorable in terms of treatment effectiveness. They are represented by omental cysts, formed by

ectopic lymphatic vessel proliferation, without connection with major lymphatic channels (Fig. 1).

Among seven children with omental LMs, only one child was asymptomatic, and the lesion was revealed accidentally during a routine US examination. All the others presented abdominal distention, abdominal mass, pain, vomiting, fever, and mild signs of deterioration.

According to our hypothesis, omental LMs should be resected surgically using laparotomy, laparoscopic, or video-assisted procedures. It is safe and effective. All children with omental LMs underwent surgical treatment using various techniques. All of

them had an uneventful postoperative recovery, and no recurrences were observed during follow-up.

#### Subgroup 2. Intestinal LMs.

Intestinal LMs can arise from mesentery with or without affection of the bowel in its different parts. If lesions do not affect the mesentery root, or if there is no total mesentery affection, which is discussed later in subgroup 5, intestinal LMs are also favorable in terms of treatment effectiveness. However, when the bowel wall is involved, resection of the affected segment with subsequent anastomosis is required as an additional operation stage (Fig. 2).

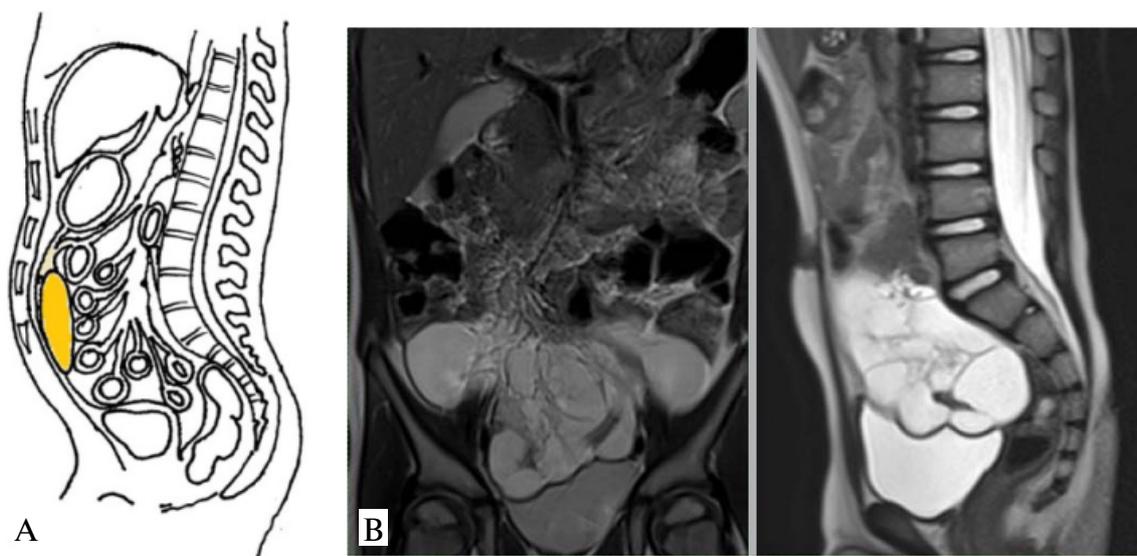


Figure 1. **Omental LMs:** A – schematic representation of the location of the omental LM; B – initial MRI of a patient with the omental LM. On frontal and sagittal T2 soft scans, a mass can be seen in the pelvis, which was initially identified as partial bowel affection. Intraoperatively, an omental cyst was confirmed and resected successfully

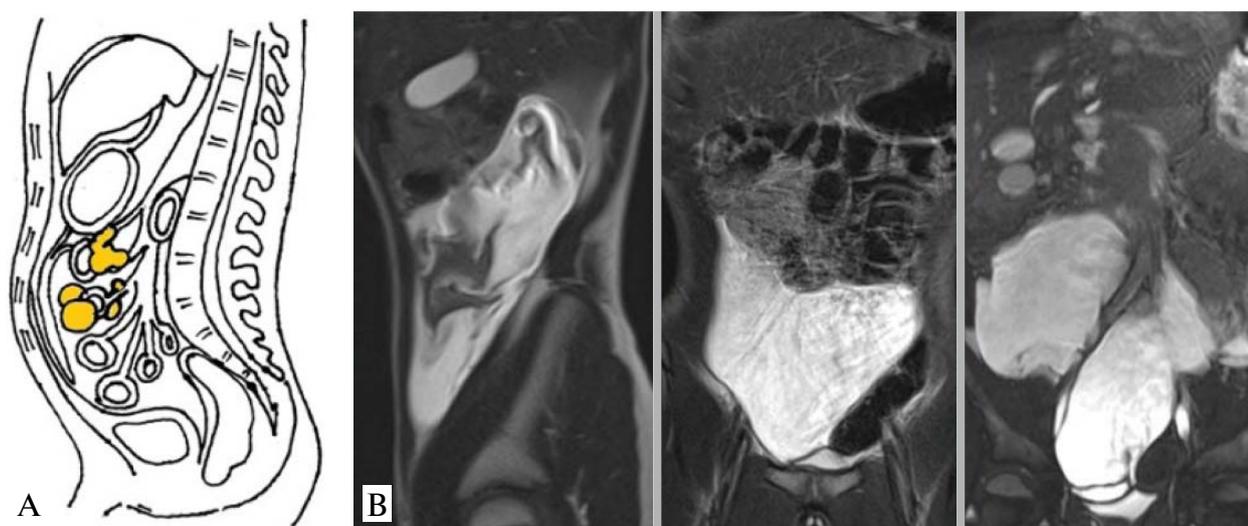


Figure 2. **Intestinal LMs:** A – schematic representation of the possible location of the intestinal LM; B – initial MRI of a patient with the intestinal LM. On sagittal and two frontal T2 soft scans, a mass can be seen, which was identified as an ileocecal angle

Intestinal LMs were discovered in 18 patients who represented the largest subgroup. Five of these patients were asymptomatic. The remaining 13 patients presented clinical signs similar to omental LMs. A two-month-old female developed intestinal obstruction due to a twisted bowel. According to our hypothesis, intestinal LMs should be resected surgically using laparotomy, laparoscopic, or video-assisted procedures. It is safe and effective. All our patients underwent surgical treatment, with bowel segment resection in five of them, uneventfully.

**Subgroup 3. Retroperitoneal LMs.** Retroperitoneal LMs are considered to be rare lesions that arise from retroperitoneal lymphatics (Fig. 3). These lesions can be both right-sided and left-sided. This subgroup is characterized by a comparatively high recurrence rate when repeated lesion growth is caused by incomplete initial resection.

In this subgroup, nine children underwent treatment in our clinic. Five of them were initially asymptomatic. Others manifested repeated pain attacks and signs of deterioration. According to our hypothesis, retroperitoneal LMs should be treated using laparoscopically-guided sclerotherapy to deliver and monitor the injection. This method is safe and reduces overall operational trauma and risks. Five patients in this subgroup underwent surgical treatment, the other four had laparoscopically-guided sclerotherapy. In three patients, surgical resection was complete according to the follow-up visualization. Others had residual mass, the size of which was constantly monitored; treatment outcomes in these patients were classified as «good» or «satisfactory».

**Subgroup 4. Retroperitoneal LMs involving the cisterna chyli.** This type of cystic LM is extremely rare, and it carries a high risk of complications, such as uncontrolled chylothorax, chyloperitoneum, or both, due to the involvement of a major lymphatic collector (Fig. 4).

In this subgroup, three children underwent treatment in our clinic. All patients were symptomatic, with a varied spectrum of deterioration symptoms and pain. One child had respiratory failure with a chylothorax. According to our hypothesis, retroperitoneal LMs, involving the cisterna chyli, should be treated using laparoscopically-guided sclerotherapy that ensures safe delivery and monitoring of the injection. Two patients underwent aforesaid treatment, with satisfactory and subtotal lesion regression, respectively, and with symptom resolution. One patient underwent laparotomy for a huge abdominal lesion in the primary hospital, with possible injury to major lymphatic vessels in the retroperitoneal space. At admission to our clinic, the patient manifested treatment-resistant chylothorax, which eventually caused sepsis, multiorgan dysfunction syndrome and death.

**Subgroup 5. Total mesentery affection.** This type of cystic LM is rare. According to a widely accepted theory, total mesentery affection develops due to the failure of numerous mesentery vessels to connect to the major venous system (Fig. 5).

Two children were included in this subgroup. Both patients had undergone laparotomies for suspected acute abdomen at primary hospitals before initial admission to our center. One surgery was finalized by revision only, but later provoked intestinal obstruction



**Figure 3. Retroperitoneal LMs: A – schematic representation of the LM's retroperitoneal location; B – initial MRI of a patient with the retroperitoneal LM. On axial, sagittal, and two frontal T2 soft scans, a mass can be seen in the right retroperitoneum, which is descending to the pelvis and scrotum**

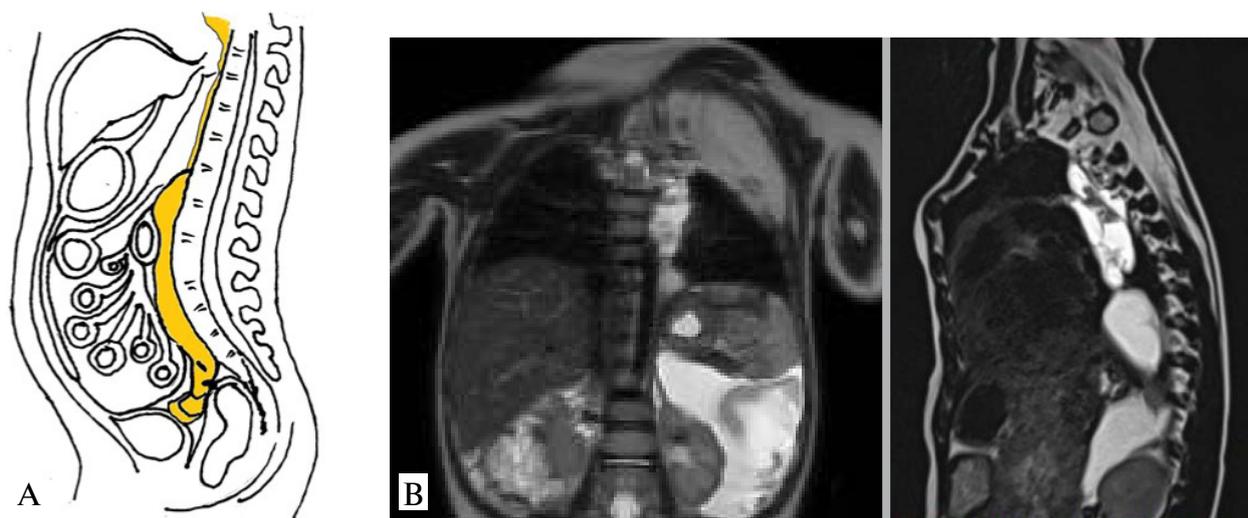


Figure 4. **Retroperitoneal LMs involving the cisterna chyli: A – schematic representation of the retroperitoneal LM involving the cisterna chyli; B – a patient’s initial MRI. On a frontal scan, a large mass, compressing the kidney and dislocating the intestines to the left, can be seen. On a sagittal scan, the enlarged cisterna chyli and dilated thoracic duct are visualized**

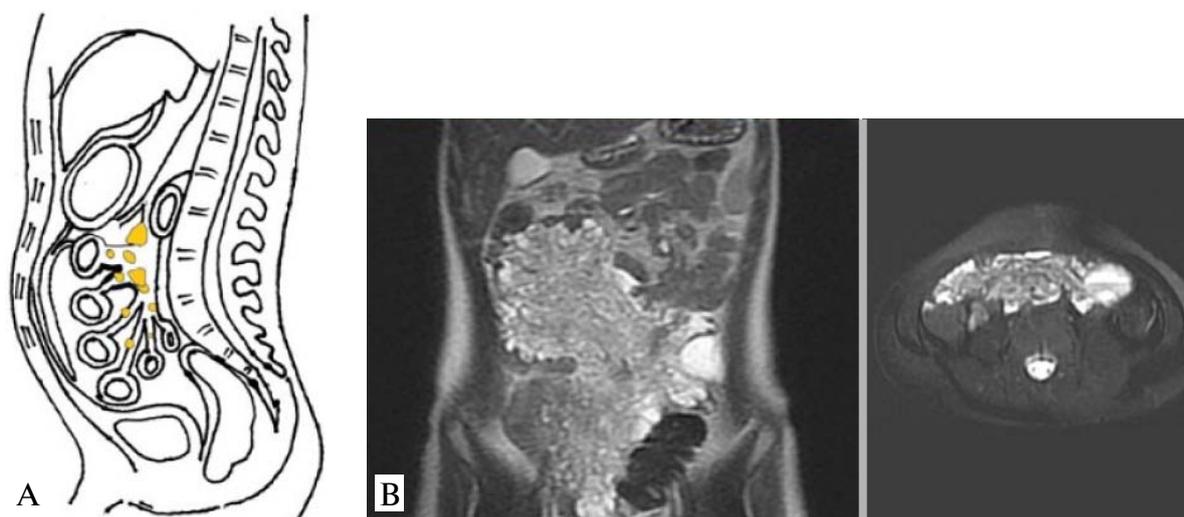


Figure 5. **Total mesentery affection: A – schematic representation of LM affecting the mesentery; B – a patient’s initial MRI. On frontal and axial scans, numerous microcystic lesions, affecting all the mesentery, are visualized**

that required repetitive laparotomy with adhesiolysis. The other child had undergone appendectomy, despite the appendix presenting no inflammatory changes. No active actions had been taken against lesions. According to our hypothesis and our limited successful experience, LMs with total mesentery affection can be effectively treated by sirolimus systemic therapy. The first patient received a one-year sirolimus systemic therapy. That resulted in a significant reduction in LM quantity and symptom resolution. The other one was asymptomatic. Therefore, it was decided to perform dynamic observation.

Subgroup 6. Splenic LMs. There are numerous causes of splenic cysts in the pediatric population,

so differential diagnosis is challenging. This type of cystic LM is extremely rare. Our study group included 3 patients with the GLA-syndrome (Fig. 6).

All three patients had from 7 to 18 cystic LMs in the spleen and were under thorough dynamic observation while asymptomatic. In adolescence, one child developed significant pain syndrome that required splenectomy with an uneventful postoperative period and resolution of pain in the follow-up period.

According to our hypothesis, 5 (11.6%) patients received treatment based on the initial tactics of the primary hospital that did not take into account the LM anatomical location. That resulted in worsening of clinical presentation, symptom progression, and an

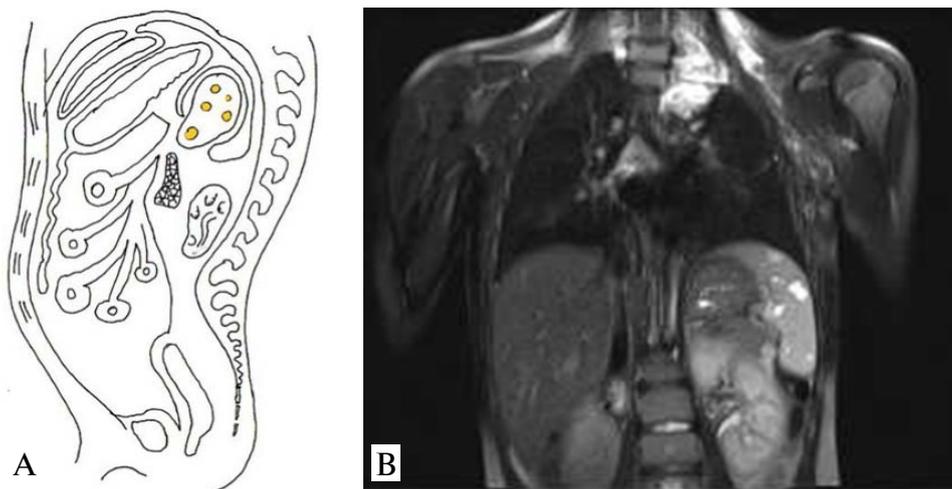


Figure 6. **Splenic LMs: A – schematic representation of LM, affecting the mesentery; B – a patient’s initial MRI. On a frontal scan, numerous microcystic lesions can be seen in the spleen. In addition, axillar and neck masses can be visualized**

increase in LM size. Consequently, it was necessary to identify the factors that influence the outcomes of surgical treatment. For this purpose, a multifactorial analysis was performed, where the anatomical localization of LMs was chosen as a factor. A logistic regression model was constructed (Fig. 7A) to identify factors that could predict the risk of surgical treatment failure. The Stepwise method revealed one factor that is associated with the risks of LMs’ surgical treatment failure: «location in the retroperitoneum». A logistic predictive model was constructed based on this factor (Fig. 7B).

Table 2. shows the factors under investigation.

According to a logistic regression model, initial choice of LM treatment without considering anatomical localization influences the risk of LM surgical treatment failure, as the factor identified is «location in the retroperitoneum» ( $p=0.000607$ ). We consider that «involvement of the cisterna chyli» and «mesentery affection» have not shown any significance for the small number of patients, 2 and 3 patients, respectively. However, it does not change our opinion on the impermissibility of surgical treatment in the mentioned group of patients due to all the complication risks.

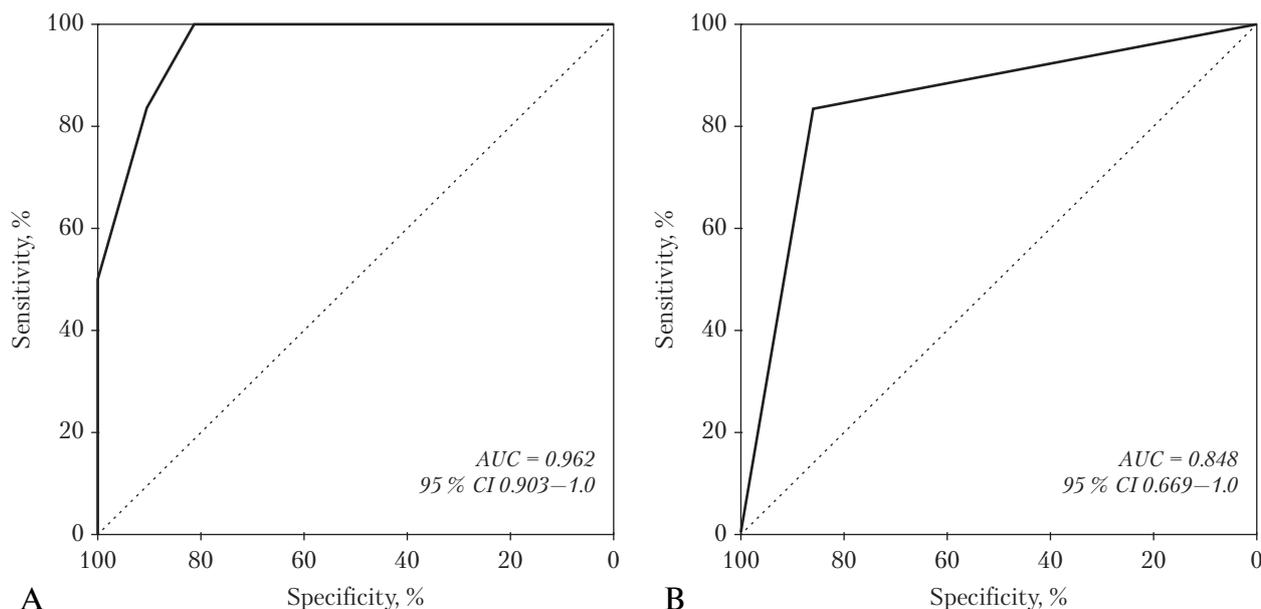


Figure 7. **ROC-curve of a 4-factor logistic regression model of LMs surgical treatment failure risks (A); a final 1-factor logistic regression model of LMs surgical treatment failure risks (B)**

**Table 2. Risk factors that were included in the logistic regression model of LMs surgical treatment failure risks**

Risk factors	OR	p
Omental localization	9.23e+06	1.0
Intestinal localization	2.80e-09	0.998
Retroperitoneal localization (95 % CI)	31.700 (2.680–374.000)	0.007
Retroperitoneal + cisterna localization*	1.27e+08	0.997
Mesenteric localization*	4.68e+08	0.996

\* Small number of patients in the group.

All patients in our study group received the following treatments: laparoscopic resections (n = 10, 23.2%); video-assisted transumbilical resections (n = 11, 25.5%); laparotomy resections (n = 9, 20.9%); ultrasound-guided and laparoscopically-guided sclerotherapy (n = 4, 9.3%); a combination of sclerotherapy and surgery (n = 1, 2.3%); sirolimus systemic therapy (n = 2, 4.6%); and splenectomy (n = 1, 2.3%). Six (13.4%) patients with small asymptomatic cystic lesions were under continuous dynamic observation.

The recommended technique for treating abdominal LMs produced excellent outcomes in 35 (81.4%) patients, good outcomes in 5 (11.6%) patients, satisfactory outcomes in 2 (4.7%) patients, and unsatisfactory outcomes in 1 (2.3%) patient.

In most studies, the incidence of abdominal LMs is assessed to be detected at 1 per 20000 admissions to pediatric hospitals [11]. Retroperitoneal LMs are even less common [15]. Earlier studies [5,13] stated that LMs in children affected the bowel and mesentery only, while later publications included the omentum, mesocolon [1, 11, 12], mesentery root [3], and retroperitoneum [1, 12]. 19.1% of patients in our study group had abdominal LMs, as opposed to previous studies claiming that they account for no more than 10% [14]. It can be attributed to the concentration of these patients in our center.

In some studies, all children from study groups were treated surgically [3, 5, 11, 13], and surgery was described as the gold standard for treating abdominal LMs. We partially agree with this approach, as all mentioned study groups presented only mesenteric, bowel, and omentum LMs, which, according to our hypothesis, should be treated surgically. On the other hand, there are LMs that are located in regions of the body where surgical treatment can be ineffective or even harmful. E.g., S. Nam et al. [12]

mention three partial surgical resections of retroperitoneal LMs, during which one child died. The same authors used OK-432 injection into residual masses after incomplete resection and during a biopsy without resection. A recent Korean study [10] of a small group that involved 12 patients with abdominal and retroperitoneal LMs showed that symptomatic children required urgent surgery. On the other hand, all children in their group underwent surgical treatment without considering anatomical localization of LMs, including retroperitoneal LMs and LMs that affected the mesentery root and resulted in incomplete resections. Most publications dedicated to retroperitoneal LMs are case-reports. Patients with retroperitoneal LMs were treated with sclerotherapy to reduce cyst size and surgery with incomplete resection, as described by R. Cryer et al. [4].

In their research, D. Gilony et al. [6] studied LMs in all locations and demonstrated that all treatment options have a place in the management of lymphatic malformations. They suggested observation for some time before sclerotherapy or surgery was necessary, noting that surgical options should be reserved in case of microcystic disease, sclerotherapy failure, or the need for biopsy. L. Jiao-ling et al. [8] in their study of a large group of 80 patients with fetal lymphangiomas note that surgery, as one of the options, has both advantages and disadvantages. Five children with abdominal and retroperitoneal LMs were included in the large study group. Two of these children underwent interventional sclerotherapy; the other three underwent surgical resection of the LM that involved the mesenteric root. During surgery, a lymphangioma that was located in the mesenteric root and that was close to neurovascular structures presented a risk for damage to blood vessels and nerves. In addition, incomplete resection is still considered to be the main cause of recurrence. In many other studies, the surgical option is used in cases of acute abdomen, sometimes without visualization. In their study, G. Chaudry et al. [2] demonstrated a group of 10 patients with abdominal, mesenteric, or retroperitoneal LMs who received doxycycline sclerotherapy, including those who were asymptomatic. Seven patients had complete resolution, two had partial resolution, and it was still unclear whether any of the lesions could be surgically removed without risk.

There may be a large number of other examples of publications that focus on a single method of treatment for abdominal cystic LMs, regardless of where they are anatomically located. Those studies, which focus on a variety of therapeutic approaches, switch from one to the next in case of failure. It is obvious

that anatomical location must be a determining factor when developing a treatment plan for cystic abdominal and retroperitoneal LMs. When surgery is applied inappropriately, it results in a recurrence, or a worsening of the patient's condition. According to our hypothesis, when a treatment strategy is chosen based on the anatomical location of cystic LMs, it ensures treatment effectiveness and, therefore, lowers the risks of treatment failure.

## Conclusions

Treatment strategies for abdominal and retroperitoneal LMs should be based on their anatomical localization. Retroperitoneal localization indicates a high risk of surgical treatment failure ( $p = 0.0006$ ). Surgical resection of abdominal LMs without evident involvement of the cisterna chyli does not lead to a recurrence. Sclerotherapy is the best treatment option for retroperitoneal LMs. Diffuse mesentery affection can be successfully treated by means of sirolimus systemic therapy. Spleen cysts should be carefully differentiated. Splenectomy is most effective when it is clinically substantial; otherwise, regular US control is helpful.

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## DECLARATION OF INTERESTS

The author has no competing interests to declare that are relevant to the content of this article.

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# Анатомічно-обґрунтована стратегія лікування лімфатичних мальформацій черевної порожнини та заочеревинного простору у дітей

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Лімфатичні мальформації (ЛМ) є рідкісними вродженими доброякісними мальформаціями лімфатичної системи. Вони представлені кістами та каналами, що містять рідину і мають нетиповий характер росту. Низька частота патології в популяції призводить до труднощів при диференційній діагностиці та виборі тактики лікування. Проаналізовано власний досвід лікування ЛМ черевної порожнини та заочеревинного простору. Запропоновано стратегію лікування, що ґрунтується на анатомічній локалізації ЛМ, яка може поліпшити результати лікування.

**Мета** — показати, що анатомічна локалізація впливає на результати лікування та повинна бути врахована при виборі методу лікування.

**Матеріали та методи.** Із 240 пацієнтів дитячого віку, які отримували лікування кістозних ЛМ на базі клініки з грудня 2012 р. до грудня 2020 р., 43 (19,1 %) встановлено діагноз «ЛМ черевної порожнини та заочеревинного простору». Період спостереження становив у середньому  $(3,50 \pm 2,16)$  року.

**Результати.** Лімфатичні мальформації черевної порожнини з доведеною відсутністю зв'язку з cisterna шулі підлягають хірургічному видаленню без подальших рецидивів. Склеротерапія є лікуванням вибору для ЛМ заочеревинного простору. Тотальне ураження брижі успішно лікується системною терапією сіролімусом. Модель логістичної регресії виявила вплив анатомічної локалізації на ризик виникнення незадовільних результатів лікування ЛМ, первинний вибір лікування без урахування анатомічної локалізації впливає на ризик невдачі лікування ( $p = 0,000503$ ). Використано такі способи лікування: лапароскопічні резекції ( $n = 10, 23,2\%$ ), відеоасистовані резекції ( $n = 11, 25,5\%$ ), лапаротомні резекції ( $n = 9, 20,9\%$ ), склеротерапія ( $n = 4, 9,3\%$ ), комбінація склеротерапії та хірургічного лікування ( $n = 1, 2,3\%$ ), системна терапія сіролімусом ( $n = 2, 4,6\%$ ), спленектомія ( $n = 1, 2,3\%$ ). Шість (13,4%) пацієнтів перебувають під динамічним спостереженням. Відмінних результатів лікування досягнуто у 35 (81,4%) випадках, добрих — у 5 (11,6%), задовільних — у 2 (4,7%), незадовільних — в 1 (2,3%).

**Висновки.** При визначенні стратегії лікування кістозних ЛМ черевної порожнини та заочеревинного простору слід урахувати їх анатомічну локалізацію. Лімфатичні мальформації заочеревинного простору пов'язані з високим ризиком неефективності хірургічного лікування ( $p = 0,0006$ ).

**Ключові слова:** лімфатичні мальформації черевної порожнини та заочеревинного простору, діти, склеротерапія, малоінвазивна хірургія, кісти черевної порожнини.

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