SAŽETAK

Uvod: Lymphangioleiomyomatosis (LAM) je retka bolest i uglavnom se javlja kod žena u generativnom periodu, kao i tokom trudnoće, dok je kod muškaraca opisan samo nekoliko pojedinačnih slučajeva. Javlja se u vidu sporadičnog oblika ili je povezana sa kompleksom tuberозne skleroze. Dijagnoza se može postaviti samo nekoliko pojedinačnih slučajeva. Javlja se u vidu sporadičnog oblika

Zaključak: Pneumotoraks je česta komplikacija limfangioleiomyomatose. Zbog visokih stope recidiva, treba izvršiti definitivnu ranu hiruršku intervenciju. Prilikom lečenja pneumotoraksa u trudnoće, limfangioleiomyomatose

SAŽETAK

Uvod: Lymphangioleiomyomatosis (LAM) is a rare disease which mainly occurs in women in the generative period, as well as during pregnancy, while only a few individual cases have been described in men. It occurs in sporadic form or is associated with tuberous sclerosis complex. The diagnosis can be made on the basis of high-resolution computed tomography (HRCT) findings, or histopathological analysis is required. Clinical manifestations of the disease include the following: progressive dyspnea on exertion, recurrent pneumothorax, chylothorax, angiomyolipomas and lymphangiomyomas.

Case report: A 32-year-old female patient was admitted to our clinic, in her third trimester of pregnancy, after a left-sided pneumothorax was verified on chest X-ray. Initial treatment included needle aspiration, followed by thoracic drainage which required thoracic drainage, as well as an insufficiently reexpanded left lung. HRCT was performed and cystic bullous changes in the lungs were noted; LAM was histopathologically verified through a minimally invasive thoraco-surgical approach, first on the left and then on the right side, while bilateral pneumothorax was surgically treated.

Conclusion: Pneumothorax is a common complication of LAM. Due to the high recurrence rate, definitive early surgical intervention should be performed. Current guidelines recommend chemical pleurodesis and surgery for the first pneumothorax. When treating pneumothorax in pregnancy, the appropriate therapeutic procedure should be applied, taking into account the safety of the pregnancy and of the delivery.

Key words: pneumothorax, pregnancy, lymphangioleiomyomatosis

MINIMALLY INVASIVE THORACOSCOPIC SURGERY AS A DIAGNOSTIC AND THERAPEUTIC APPROACH IN BILATERAL PNEUMOTHORAX IN PREGNANCY CAUSED BY LYMPHANGIOLEIOMYOMATOSIS – A CASE REPORT

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ABSTRACT

Introduction: Lymphangioleiomyomatosis (LAM) is a rare disease which mainly occurs in women in the generative period, as well as during pregnancy, while only a few individual cases have been described in men. It occurs in sporadic form or is associated with tuberous sclerosis complex. The diagnosis can be made on the basis of high-resolution computed tomography (HRCT) findings, or histopathological analysis is required. Clinical manifestations of the disease include the following: progressive dyspnea on exertion, recurrent pneumothorax, chylothorax, angiomyolipomas and lymphangiomyomas.

Case report: A 32-year-old female patient was admitted to our clinic, in her third trimester of pregnancy, after a left-sided pneumothorax was verified on chest X-ray. Initial treatment included needle aspiration, followed by thoracic drainage which required thoracic drainage, as well as an insufficiently reexpanded left lung. HRCT was performed and cystic bullous changes in the lungs were noted; LAM was histopathologically verified through a minimally invasive thoraco-surgical approach, first on the left and then on the right side, while bilateral pneumothorax was surgically treated.

Conclusion: Pneumothorax is a common complication of LAM. Due to the high recurrence rate, definitive early surgical intervention should be performed. Current guidelines recommend chemical pleurodesis and surgery for the first pneumothorax. When treating pneumothorax in pregnancy, the appropriate therapeutic procedure should be applied, taking into account the safety of the pregnancy and of the delivery.

Key words: pneumothorax, pregnancy, lymphangioleiomyomatosis
UVOD

Limfangioleiomiomatoza (LAM) je progresivna, retka, multisistemska bolest nepoznate etiologije, koja se pretežno javlja kod žena, pri čemu uzrokuje oštećenje plućne funkcije [1]. Epidemiološka studija, sprovedena u sedam zemalja, pokazala je da je incidencija limfangioleiomiomatoze 3,4 – 7,8 slučajeva na milion žena [2]. LAM se uglavnom manifestuje u vidu cistične destrukcije plućnog parenhima, kao i ekstrapulmonalne bolesti, koja se sastoji od angiomiolipoma, limfnih tumora (limfangioleiomiomatoza) i hiloznih izliva [3]. Limfangioleiomiomatoza je retka u opštoj populaciji, ali je česta kod žena sa kompleksnom tuberzuznom skleroze (engl. *tuberous sclerosis complex – TSC*). Javlja se u 30 – 40% slučajeva (TSC-LAM) [4]. Prušina je kod žena između menarhe i menopauze, prosečne starosti od 34 godine [5].

Značajan broj pacijentkinja sa LAM-om razvija svoje početne simptome tokom trudnoće, Zabeleženo je da je trudnoća kod limfangioleiomiomatoze povezana sa povećanjom incidencijom pneumotoraksa, iako neke pacijentkinje imaju nekomplikovanoću trudnoću [6]. Najčešće, pacijenti imaju kretak dah, kašalj i pneumotoraksa. Less frequently, there are chylous pleural effusions [3]. Limfangioleiomyomatosis is rare in the general population but is frequent in women with tuberosus sclerosis complex (TSC). It occurs in 30 – 40% of cases (TSC-LAM) [4]. It is present in women between menarche and menopause, of the average age of 34 years [5].

Cilj ovog rada jeste da se prikaže slučaj trudnice koja je razvila spontani bilateralni pneumotoraks tokom trećeg tromeseca trudnoće i odmah nakon po- rođaja, ali i da se ukaže na značaj minimalno invazivnoj video-assistiranoj torakohirurškoj operaciji (VATS) u dijagnostici limfangioleiomiomatoze i lečenju pneumotoraksa izazvanih limfangioleiomiomatozom.

PRIKAZ SLUČAJA

Tridesetdvogodišnja pacijentkinja je primljena na našu kliniku, u 36. nedelji gestacije dva trudnoće, dva dana nakon pojave iznenadnog bola u levom hemitorku, praćenog akutnom kratkom dana. Pacijentkinja nije imala istoriju ranijih hroničnih plućnih bolesti ili drugih komorbiditeta. Njeno prethodno trudnoće je bilo bez komplikacija. Prilikom izveštavanja o pregledu, imala je tahipneju, tahikardiju i hiperrezonantne perkusione zvuk, sa smanjenim disajnim pokretima levog hemitoraka. Pregledima kardiokavskog sistema nije bilo zabeležen patološki izraz niti je verifikovana organomegalija. Rezultati krvne slike, kao i testovi funkcije jetre i bubrega bili su normalni. Radiografski snimak grudnog koša potvrdio je prisustvo kompletog pneumotoraksa (Slika 1).

Početni tretman je zaključen u eksuflaciju levostranog pneumotoraksa, a nakon učinjene navedene intervencije i nalaza bez poboljšanja, učinjena je tara-
Kalna drenaža levog pleuralnog kavuma, sa torakalnim drenom povezanim na podvodnu aspiraciju. S obzirom da nije došlo do reekspanzije levog plućnog krila, torakalni dren je naknadno priključen na aktivnu aspiraciju. Zbog produženog gubitka vazduha kroz torakalni dren i poodmakle trudnoće (37. nedelja gestacije), urađen je carski rez. Rođena je devojčica od 3.130 grama sa dobroim Apgar skorom (9).

Nakon porođaja, radiografijom grudnog koša, utvrđen je kompletni desnostran pneumotoraks, koji je zahtevao torakalnu drenažu, uz prethodno plasirani torakalni dren, zbog levostranog pneumotoraksa (Slika 2).

Nalaz kompjuterizovane tomografije grudnog koša visoke rezolucije (engl. high-resolution computed tomography HRCT) je pokazao bilateralne višestruke difuzne cistične promene tankih zidova, rasute po plućnom parenhimu, veličine do 22 milimetra (Slika 3). Kompjuterizovana tomografija abdomena i male karlice nije pokazala abnormnalnosti.

U ovim okolnostima odlučili smo se za hirurško le-čenje pneumotoraka.

Using under water sealed drainage (UWSD). Since re-expansion of the left lung did not occur, the thoracic drain was subsequently connected to active aspiration. Due to prolonged air loss via the thoracic drain and the advanced stage of pregnancy (37th week of gestation), a C-section was performed and a girl weighing 3,130 grams with a good Apgar score (9) was born.

After the delivery, an X-ray of the patient’s chest revealed complete right-sided pneumothorax, which required thoracic drainage, in addition to the thoracic drain previously placed due to left-sided pneumothorax (Figure 2).

The high-resolution computed tomography (HRCT) chest scan showed bilateral multiple diffuse thin-walled cystic lesions, scattered throughout the lung parenchyma, which measured up to 22 mm in diameter (Figure 3). Computed tomography (CT) of the abdomen and the lesser pelvis showed no abnormalities.

In such circumstances, we decided on surgical treatment of the pneumothorax.
Inicijalno, sedmog dana nakon porođaja, izveli smo levostrani VATS pristup. Intraoperativno su otkrivene pleuralne adhezije u predelu levog gornjeg režnja. Nakon razdvajanja pleuralnih adhezija, u gornjem i donjem režnju je nađeno više mehurića i bula, predominantno u lingularnom segmentu gornjeg režnja i apikalnom segmentu donjeg režnja (Slika 4). Endostaplerom smo učinili klinastu resekciju apikalnog segmenta donjeg režnja i lingularnog segmenta gornjeg režnja. Urađena je hemijska pleurodeza sa 30 ml pedesetoprocentnog rastvora glukoze i mehanička pleurodeza sa abrazijom kostalne parijetalne pleure.

Histopatološki nalaz je pokazao proširene alveolarni prostor sa glatkim mišićnim tkivotom u njegovim zidovima, perivaskularnu proliferaciju glatkih mišića, agregate limfoidnih čelija i proširene limfne prostore. Nije pronađen granulom ili malignitet. Utvrđeno je da su kultivisane čelije glatkih mišića pozitivne na human melanomski blok-45 (HMB-45). Dakle, dijagnoza limfangioleiomiomatoze je potvrđena.

Hirurško lečenje desnostranog pneumotoraksa VATS pristupom izvedeno je 15. dana nakon porođaja. Intraoperativno smo primetili više mehurića i bula, predominantno u srednjem režnju. Učinjena je klinaster resekcija srednjeg režnja endostaplerom, a zatim hemijska i mehanička pleurodeza, kao i u prvoj operaciji. Histopatološki nalaz odgovarao je nalazu prethodne operacije.

Postoperativni tok, nakon oba hirurška zahvata, protekao je uređeno, uz potpunu reekspanziju desnog i levog plućnog krila, koja se održavala nakon uklanjanja torakalskih drenova.

Pacijentkinja je otpuštena iz bolnice 20. dana nakon porođaja. Na dan otpusta iz bolnice, rendgenski snimak grudnog koša je pokazao potpuno reekspandirano levo i desno plućno krilo.

Testiranje plućne funkcije tri, šest i dvanaest meseci nakon porođaja pokazalo je evidentne vrednosti. Ponovljani HRC test snimci abdomena i karlice, šest meseci i godinu nakon porođaja, nisu pokazali abnormalnosti. Rendgenski snimak grudnog koša, mesec dana, tri meseca, šest meseci i godinu nakon porođaja, pokazali su potpuno reekspandirano levo i desno plućno krilo. Ponovljeni HRC test snimci grudnog koša, šest meseci i godinu nakon porođaja, nisu pokazali progresiju bolesti. I pacijentkinja i njeno dete ostali su zdravi nakon godinu dana praćenja.

DISKUSIJA
Limfangioleiomiomatoza je progresivna, cistična bolest pluća, koja se javlja pretežno kod žena u tridesetim godinama života [1,5]. LAM se retko javlja u opštoj populaciji. Obično se javlja kod žena sa TSC-om (30 – 40% slučajeva) i na dve trećine žena sa TSC-om na dve trećine žena sa TSC-om, LAM je progresivna, cistična bolest, koja se javlja kod žena sa TSC-om. LAM se retko javlja u opštoj populaciji.
Pneumotoraks je čest kod pacijenata sa limfangioleiomiomatozom. Studija registra Nacionalnog instituta za srce, pluća i krv (engl. *National Heart, Lung, and Blood Institute - NHLBI*) u Sjedinjenim Američkim Državama, koja je obuhvatila 230 pacijenata sa limfangioleiomiomatozom, zabeležila je pneumotoraks u 55,5% slučajeva [10]. Druge studije su pokazale da je incidenca pneumotoraksa kod limfangioleiomiomatoze 1.000 puta veća nego u opštoj ženskoj populaciji [11]. Štaviše, 50 – 80% pacijenata će razviti pneumotoraks tokom bolesti. Pojava recidiva pneumotoraksa je česta, sa prosečnim brojem pneumotoraksa od oko četiri [10–14].

Nespecifične kliničke karakteristike sa neupadljivim rendgenskim snimcima grudnog koša na prezentaciji dovode do kašnjenja u dijagnozi ili netačne dijagnoze [7,8].

Komplikacije povezane sa limfangioleiomiomatozom, uključujući pneumotoraks i hilotoraks, mogu se javiti tokom trudnoće, a izražena je zabrinutost zbog moguće povezanosti trudnoće sa progresijom limfangioleiomiomatoze. Takođe je zabeleženo da žene sa LAM-om imaju lošiju ishod trudnoće, uključujući i višu prevremenih porođaja i abortusa [6,15,16].

U literaturi je opisan pneumotoraks izazavan LAM-om, u poslednjem tromesečju trudnoće i ubrzo nakon porođaja. U ovaj fenomen se objašnjava činjenicom da hormonska stimulacija tokom trudnoće dovodi do povećanja cističnih struktura u plućima, te potom, na kraju trudnoće ili ubrzo nakon porođaja, dolazi do opadanja hormonske stimulacije i posledično do rupture neke od tankozidnih cističnih struktura i pojave pneumotoraksa. Ovo takođe objašnjava činjenicu da je LAM, skoro tipično, ženska bolest [17,18].

Ranije se pokazalo da je dispneja pri naporu, kao prisutni simptom, povezana sa povećanim mortalitetom kod pneumotoraksa [19]. Nasuprot tome, prethodno se smatrao da pacijenti sa pneumotoraksom imaju povoljniju prognozu [1]. Kasnije studije nisu pokazale vezu između pneumotoraka i dugoročnog ishoda [20,21].

Ako pacijent sa potvrđenom LAM-om ima pneumotoraks, treba ga lečiti u skladu sa standardnim smernicama za lečenje sekundarnog pneumotoraksa [22]. Lečenje pneumotoraksa u trudnoći zavisi od stepena i faze progresije. Obično se koristi torakalna drenaža, sa ili bez aktivne aspiracije [16]. U slučaju naše pacijentkinje, stimulacije limfangioleiomiomatoze nije bila pozna, a to prilikom donošenja odluke o inicijalnom zbrinjavanju pneumotoraksa, pa smo koristili preporuke zvanica za lečenje pneumotoraksa u trudnoći [23].

DISCUSSION

Lymphangioleiomyomatosis is a progressive cystic pulmonary disease, occurring mainly in women in their thirties [1,5]. LAM rarely occurs in the general population. It usually develops in women with TSC (30 – 40% of cases), or it may occur as a sporadic form. There are specific criteria for diagnosing TSC [9]. In the case of our patient, the TSC criteria were not met.

Pneumothorax is frequent in patients with lymphangioleiomyomatosis. The study of the National Heart, Lung, and Blood Institute (NHLBI) registry (USA), involving 230 patients with LAM, registered pneumothorax in 55.5% of cases [10]. Other studies have shown that the incidence of pneumothorax in LAM is 1,000 greater than in the general female population [11]. Moreover, 50 – 80% of patients will develop pneumothorax during the illness. The recurrence of pneumothorax is frequent, with the average number of pneumothorax episodes being around four [10–14].

Nonspecific clinical characteristics with unremarkable chest X-rays on presentation lead to delayed diagnosis or misdiagnosis [7,8].

Complications connected to LAM, including pneumothorax and chylothorax, may occur in pregnancy, and the possible connection between pregnancy and the progression of LAM is cause for serious concern. Also, it has been recorded that women with LAM have a poorer outcome of pregnancy, including a greater number of premature births and miscarriages [6,15,16].

Pneumothorax caused by LAM in the third trimester of pregnancy and shortly after delivery has been described in literature. This phenomenon is explained by the fact that hormone stimulation during pregnancy leads to the increase in cystic structures in the lungs, and subsequently, at the end of pregnancy or soon after delivery, hormone stimulation drops, and consequently, some of the thin-walled cystic structures rupture, leading to pneumothorax. This also explains the fact that LAM is, almost typically, a female disease [17,18].
Smernice Američkog torakalnog društva/Japan- skog respiratornog društva (engl. American Thoracic Society/Japanese Respiratory Society Clinical Practice Guidelines) podržavaju klinički dijagnozu limfangioleiomatozo asanzovanu na HRCT nalazima tipičnim za limfangioleiomatozo (npr. ciste/mehurići difuzni, tankih zidova, okrugli) koji su praćeni bilo kojom od sledećih kliničkih karakteristika: TSC, angiomiolipom bubrega, cistični limfangioleiom, ili hilozni izliz u grudnom košu i ili abdomenu. Smernice daju snažnu preporuku za korišćenje testa VEGF-D (vaskularni endotelni faktor rasta D; engl. vascular endothelial growth factor D test) za postavljanje dijagnoze limfangioleiomatozo, pre razmatranja biopsije pluća, kod pacijenata sa cističnim abnormalnostima na HRCT nalazu, karakterističnim za LAM, ali bez drugih potvrdnih kliničkih karakteristika [24].

Histopatološka potvrda limfangioleiomatozo može se dobiti na različite načine, uključujući transbronhijalnu, perkutanu ili VATS biopsiju. Rizik od pneumotoraksa nakon transbronhijalne biopsije je 1 – 6%, a čak i veći nakon perkutane biopsije. Prednost VATS-a je minimalna hirurška trauma, kao i odgovarajući uzorak za patološku potvrdu, kada se radi bulektomija i pleurektomija [25]. Neke retrospektivne serije sugeriru da transbronhijalna biopsija pluća može biti bezbedna i efikasna u delu pacijenata sa sumnjom na limfangioleiomatozu. U studiji koju su objavili Meraj et al., među 63 pacijenata koji su bili podvrgnuti transbronhijalnoj biopsiji pluća, kada se prvobitno sumnjalo na limfangioleiomatozu, kod 35 pacijenata (56%) je ovim postupkom potvrđena LAM. Stopa prijavljenih komplikacija prilikom transbronhijalne biopsije bila je približno 14% (6% sa pneumotorakom, 4% sa krvarenjem, 2% sa bolom u grudima i 2% sa pneumonijom) [26].

Međutim, hirurška biopsija pluća primenom VATS pristupa se smatra zlatnim standardom za dobjavanje histopatološke potvrde limfangioleiomatozo. Kada je potrebna definitivna dijagnoza kod pacijenata sa parenhimskim cistama specifičnim za limfangioleiomatozu prisutnim na HRCT nalazu, ali bez dodatnih potvrdnih karakteristika limfangioleiomatozo, Smernice Američkog torakalnog društva/Japanskog respiratornog društva predlažu dijagnostički pristup koji uključuje transbronhijalan biopsiju pluća, pre hirurške biopsije pluća [27]. U slučaju naše pacijentkinje, dijagnoza limfangioleiomatozo je postavljena VATS pristupom, koji je ujedno predstavljao i terapijski postupak za rešavanje pneumotoraksa.

Pošto je stopa recidiva visoka, trenutne smernice preporučuju pleurektomiju u vreme prve epizode pneumotoraksa, kod pacijenata sa potvrđenom LAM-om. It has been found earlier that dyspnea on exertion, as a symptom, is connected with increased mortality in LAM [19]. Conversely, it is used to be believed that patients with pneumothorax had a more favorable prognosis [1]. More recent studies have not shown a link between pneumothorax and the long-term outcome [20,21].

If a patient with confirmed LAM has pneumothorax, this patient should be treated in keeping with the standard guidelines for treating secondary pneumothorax [22]. Treatment of pneumothorax in pregnancy depends on the degree and phase of progression. Usually, thoracic drainage is used, but without active aspiration [16]. In the case of our patient, the diagnosis of LAM was not known at the time when the decision was made on initial pneumothorax treatment, which is why we applied the recommendations of the official guidelines for treating pneumothorax in pregnancy [23].

The American Thoracic Society/Japanese Respiratory Society Clinical Practice Guidelines support the clinical diagnosis of LAM based on HRCT findings that are typical for lymphangioleiomyomatosis (e.g., diffuse, thin-walled, spheric cysts/blisters) accompanied by any of the following characteristics: TSC, renal angiomylipoma, cystic lymphangioleiomyoma, or chylous effusion in the thorax and/or abdomen. The guidelines strongly recommend the use of the VEGF-D test (vascular endothelial growth factor D test) for establishing the diagnosis of LAM, before considering lung biopsy in patients with cystic abnormalities characteristic of LAM present on the HRCT scan, but without other positive clinical characteristics [24].

Histopathological confirmation of LAM may be obtained in different ways, including transbronchial, percutaneous or VATS biopsy. The risk from pneumothorax after transbronchial biopsy is 1 – 6%, and it is even higher after percutaneous biopsy. The advantages of VATS are reflected in minimal surgical trauma, as well as in the fact that it enables the obtaining of an adequate sample for pathological confirmation, when bullectomy and pleurectomy or pleurectomy are performed [25]. Some retrospective studies suggest that transbronchial lung biopsy may be safe and efficient in some of the patients with suspected LAM. In a study by Meraj et al., amongst 63 patients who underwent transbronchial lung biopsy at the time when LAM was initially suspected, in 35 (56%) patients LAM was confirmed in this way. The rate of reported complications of transbronchial biopsy was approximately 14% (6% with pneumothorax, 4% with bleeding, 2% with chest pain, and 2% with pneumonija) [26].

However, surgical biopsy of the lungs with the application of VATS is believed to be the golden standard for obtaining histopathological confirmation of LAM.
Međutim, pleurodeza kod pacijenata sa LAM-om ima ograničenu efikasnost, a zabeleženo je da se stope recidiva pneumotoraksa kreću između 18% i 32% [27,28]. Nedostaju jasne smernice o tome da li je hemijska ili hirurška pleurodeza optimalna, ali je važno rano uključiti grudne hirurge, jer će nekim pacijentima biti potrebna transplantacija pluća, u dužem vremenskom roku. Smernice za dijagnozu i lečenje limfangioleiomatoze (LAM) Evropskog respiratornog društva (engl. European Respiratory Society Guidelines for the Diagnosis and Management of Lymphangioleiomyomatosis – LAM) i Smernice Američkog torakalnog društva/Japanskog respiratornog društva preporučuju hemijsku pleurodezu i operaciju, za prvi pneumotoraks [22,26]. U slučaju naše pacijentkinje, učinili smo hemijsku i mehaničku pleurodezu i operaciju, za prvi pneumotoraks [22,26]. Prethodne pleuralne procedure nisu kontraindikacija za buduću transplantaciju pluća [29].

Sukob interesa: Nije prijavljen.

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When a definitive diagnosis is necessary in patients with parenchymal cysts characteristic of LAM present on the HRCT finding, but without additional positive characteristics of LAM, the American Thoracic Society/Japanese Respiratory Society Clinical Practice Guidelines suggest a diagnostic approach which includes transbronchial lung biopsy, before surgical lung biopsy [27]. In our patient, the diagnosis of LAM was established with the VATS approach, which was also the treatment procedure for pneumothorax.

Since the recurrence rate is high, current guidelines recommend pleurodesis at the time of the first episode of pneumothorax, in patients with confirmed LAM [22,27]. However, pleurodesis in patients with LAM has limited efficiency, and it has been recorded that recurrence rates for pneumothorax range from 18% to 32% [27,28]. Clear guidelines are lacking as to whether chemical or mechanical pleurodesis is optimal, but it is definitively important to include thoracic surgeons into the process early on, as some of the patients will need a lung transplant, in the long-term. European Respiratory Society Guidelines for the Diagnosis and Management of Lymphangioleiomyomatosis (LAM) and the American Thoracic Society/Japanese Respiratory Society Clinical Practice Guidelines recommend chemical pleurodesis and surgery, for the first pneumothorax [22,26]. In the case of our patient, we performed chemical and mechanical pleurodesis. Previous pleural procedures are not a contraindication for a future lung transplant [29].

Conflict of interest: None declared.

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Minimalno invazivna torakoskopska hirurgija kao dijagnostički i terapijski pristup kod bilateralnog pneumоторакса у трудноći uzrokovanog limfangioleiomyomatozom – prikaz slučaja.