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Successful Management of Lymphangioleiomyomatosis with Sirolimus During Pregnancy and Favorable Maternal–Fetal Outcomes in Two Siblings: A Case Report

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ABSTRACT

Lymphangioleiomyomatosis (LAM) is a rare multisystem disorder predominantly effecting women of reproductive age. Pregnancy may be associated with an increased risk of pulmonary complications, including pneumothorax, chylous effusions, and decline in lung function, potentially because of hormonal influences on LAM-cell proliferation. Sirolimus is the only established disease-modifying treatment shown to stabilize lung function in LAM, whereas its use during pregnancy remains controversial because of limited human safety data.

A 28-year-old woman with LAM and impaired pulmonary function conceived unexpectedly after 8 months of treatment with sirolimus. At diagnosis, pulmonary function testing showed an FEV_1 of 2.40 L (70% predicted), an FVC of 2.70 L (66% predicted), and a DLCO/VA of 3.4 mL/min/mm Hg/L (70% predicted). Sirolimus had stabilized her pulmonary function and improved exercise tolerance before pregnancy. Treatment was discontinued during the first trimester because of concerns regarding fetal exposure and organogenesis and was later reintroduced at a reduced dose with therapeutic drug monitoring. Pulmonary function remained stable throughout pregnancy, and she delivered a healthy male infant at 39 weeks. At 31 years of age, she conceived a second pregnancy while receiving low-dose sirolimus. Treatment was again interrupted during the first trimester and subsequently continued at low dose. Maternal pulmonary function remained stable throughout the second pregnancy, and a healthy female infant was delivered at 37 weeks. Neither child had a congenital anomaly or documented sirolimus-related complication during follow-up. The mother maintained stable pulmonary function during long-term follow-up.

This case describes favorable maternal and neonatal outcomes during two pregnancies in a woman with LAM managed with individualized sirolimus therapy and close multidisciplinary surveillance. Although a single case cannot establish the safety of sirolimus during pregnancy, this experience contributes to the limited literature describing maternal and neonatal outcomes in women with LAM receiving sirolimus during gestation. Pregnancy in women with LAM requires individualized risk assessment for maternal and fetal outcomes. In selected patients with clinically significant disease, modification or continuation of low-dose sirolimus may be considered with close respiratory, obstetric, and fetal surveillance after careful assessment of the potential maternal benefits and fetal risks. Prospective multicenter registries with long-term pediatric follow-up are needed to better define the risks and benefits of sirolimus therapy during pregnancy.

Keywords
Lymphangioleiomyomatosis, LAM, Sirolimus, Pregnancy, mTOR inhibitor, Maternal outcomes, Pulmonary function, Fetal outcomes.

Introduction
Lymphangioleiomyomatosis is a rare, progressive neoplastic disorder of the lung, kidneys, and axial lymphatics caused by loss of TSC1 or TSC2 function and consequent constitutive activation of the mechanistic target of rapamycin (mTOR) pathway. The disease occurs almost exclusively in women of reproductive age, either sporadically or in association with tuberous sclerosis complex, and is characterized by diffuse thin-walled pulmonary cysts, airflow limitation, and, in a subset of patients, chylous effusions, lymphangioleiomyomas, and renal angiomyolipomas [1-3]. Untreated, LAM typically causes progressive impairment of FEV_1 and gas transfer. Estrogen has been implicated in LAM-cell proliferation and disease progression whereas pregnancy, a high-estrogen physiologic state accompanied by substantial cardiopulmonary adaptation, may lead to increased risk of complications, including accelerated lung function decline, pneumothorax, and chylous effusions attributed to estrogen-driven disease activity causing potential for rapid clinical deterioration that may expedite disease progression [4-7].In a combined United Kingdom and National Institutes of Health cohort, mean FEV_1 fell from 77% to 64% predicted across pregnancy, and pneumothorax was common [8].

Sirolimus has revolutionized LAM management by stabilizing FEV_1, reducing cyst progression, and alleviating the need for supplemental oxygen improving the quality of life as demonstrated in the MILES trial [9]. Sirolimus effectively stabilizes lung function and reduces disease progression with improved quality of life [10,11]. Concerns regarding fetal safety have precluded the routine use of sirolimus during pregnancy, owing to the scarcity of human safety data. Its use in pregnancy therefore remains controversial, with available evidence derived primarily from case reports and small observational series. Published reports have described pregnancies exposed to sirolimus, including cases involving low-dose therapy and therapeutic drug monitoring; however, systematic data on maternal and fetal outcomes remain scarce. Emerging case reports and small series suggest that low-dose sirolimus targeting trough 3–5 µg/L may be tolerated with favorable maternal-fetal outcomes in carefully selected cases [12,13].

We describe a female patient with LAM who underwent two consecutive pregnancies under a protocol of first-trimester sirolimus interruption followed by resumption of low-dose, trough-guided therapy, with serial pulmonary function testing, VEGF-D measurement, and neonatal follow-up. The present case contributes to the limited literature on pregnancy in LAM and provides clinically relevant insights into the potential safety and effectiveness of carefully selected sirolimus therapy during pregnancy that illustrates the complexities of balancing maternal disease control against potential fetal risks.

Case Presentation
A 28-year-old woman with LAM diagnosed 2 years earlier earlier presented with an unplanned pregnancy. Sporadic LAM had been diagnosed 2 years earlier on the basis of characteristic high-resolution computed tomography (HRCT) findings and a high serum VEGF-D concentration of 980 pg/mL, above the 800 pg/mL threshold endorsed by the American Thoracic Society/Japanese Respiratory Society guidelines for a noninvasive diagnosis in the presence of typical cystic lung disease [1,14]. At diagnosis, serum VEGF-D was 980 pg/mL. Initial pulmonary function testing showed an FVC of 2.70 L (66% predicted), an FEV_1 of 2.40 L (70% predicted), and a DLCO/VA of 3.4 mL/min/mm Hg/L (70% predicted). Before conception, she had received sirolimus at a dose of 2 mg/day for approximately 8 months, with trough concentrations reported at approximately 6–8 µg/L. During this period, pulmonary function remained stable and exercise tolerance improved. She was an industrial engineer working in an office-based setting, with no history of smoking or alcohol consumption and without any relevant environmental or occupational exposures. Her personal and family medical histories were otherwise unremarkable and lacked any history of tuberous sclerosis complex, pneumothorax, chylous effusion, or renal angiomyolipoma.

The patient was admitted due to an unexpected pregnancy of two weeks. On examination, blood pressure was 120/70 mm Hg, respiratory rate 14 breaths/min, and chest auscultation demonstrated bilateral vesicular breathing without adventitious sounds. Arterial blood gas analysis on room air showed pH: 7.42, pO_2: 74 mm Hg, pCO_2: 38 mm Hg, and oxygen saturation was 92% while breathing ambient air. ECG showed sinus rhythm at a rate of 84 beats/min with a normal QRS axis. The chest x-ray showed faint lower-zone reticular markings without pneumothorax or pleural effusion (Figure 1). HRCT revealed innumerable thin-walled, round-to-oval cysts, ranging from a few millimeters to approximately 20 mm in diameter, distributed relatively uniformly from apex to base and involving both central and peripheral lung, with several larger subpleural cysts in the right upper lobe and preserved intervening parenchyma without architectural distortion (Figure 1).

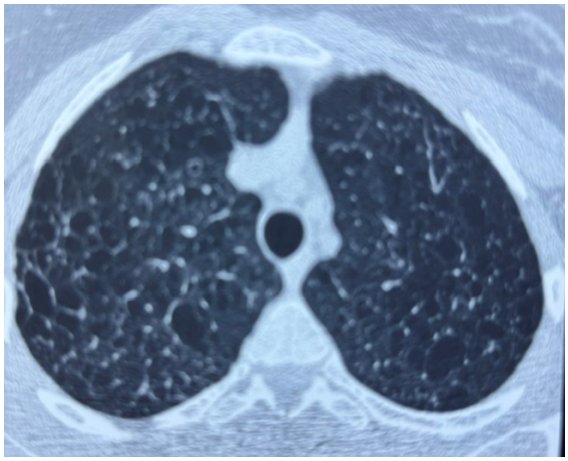


Figure 1. HRCT showing diffuse bilateral thin-walled pulmonary cysts with relatively uniform distribution and preserved intervening parenchyma,

highly characteristic of pulmonary lymphangioleiomyomatosis.

Due to concerns regarding potential fetal exposure to sirolimus during organogenesis, treatment was discontinued during the first trimester. Given the perceived risk of disease exacerbation following treatment withdrawal, low-dose sirolimus was subsequently resumed at 1 mg/day with a target trough concentration of 4–6 µg/L. Serial pulmonary function testing and serum VEGF-D measurements were performed throughout pregnancy and postpartum (Table 1). Maternal pulmonary function remained stable during pregnancy. FEV_1 ranged from 2.45 to 2.54 L (71–74% predicted), while FVC remained between 2.70 and 2.80 L (66–69% predicted). DLCO/VA maintained a similar range, varying from 3.2 to 3.6 mL/min/mm Hg/L (66–75% predicted). Serum VEGF-D levels gradually decreased from 820 pg/mL at baseline to 720 pg/mL postpartum. Fetal biometric measurements on ultrasonography were appropriate for gestational age. Pregnancy progressed without any maternal respiratory complication, and the patient delivered a healthy male infant at 39 weeks' gestation. Birth weight was 3.2 kg, and the Apgar score was 9/10. Breastfeeding was not initiated owing to the excretion of sirolimus in human milk.

At age 31, the patient conceived her second pregnancy while on sirolimus 2 mg daily, through concentrations of 3.0–5.0 µg/L, that was discontinued during the first trimester concerning the adverse effects of sirolimus on fetal organogenesis. On admission, blood pressure was 110/78 mm Hg, respiratory rate was 16 breaths/min, and auscultation revealed bilateral vesicular breath sounds. Arterial blood gas analysis on room air showed pH: 7.40, pO2: 76 mm Hg, pCO2: 40 mm Hg, and oxygen saturation of 94%. ECG displayed a sinus rhythm of 82/min and normal cardiac axis. Repeat axial thorax HRCT findings did not show any significant differences compared to the initial admission findings (Figure 2). Sirolimus dose, VEGF-D levels, and pulmonary function values with the predicted reference percentages during pregnancy, and postpartum are shown in Table 2. Sirolimus was again withheld during the first trimester and then resumed at 1 mg daily without further interruption that was increased to 2 mg/day postpartum. Serial serum VEGF-D measurements and pulmonary function tests are presented in Table 2. Pulmonary function remained stable throughout the second pregnancy, with FEV_1 ranging from 2.40 to 2.54 L (70–74% predicted), FVC from 2.71 to 2.86 L (66–70% predicted), and DLCO/VA ranging from 3.4 to 3.8 mL/min/mm Hg/L (70–79% predicted). Ultrasonographic assessment demonstrated fetal growth appropriate for gestational age. Serum VEGF-D levels decreased from 740 pg/mL at baseline to 680 pg/mL postpartum. A healthy female infant was delivered at 37 weeks via cesarean section. Birth weight was 2.9 kg and Apgar score was 8/10. Breastfeeding was deferred due to the known excretion of sirolimus into human milk.

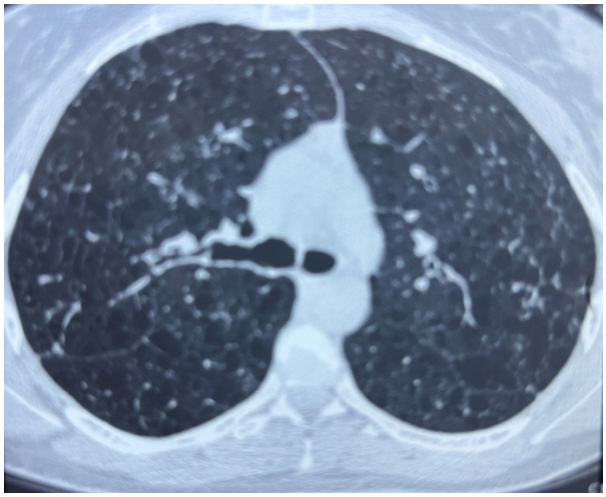


Figure 2. HRCT revealing diffuse innumerable thin-walled cystic airspaces predominantly round to mildly oval, range in diameter from a few millimeters to approximately 15–20 mm, distributed throughout all lobes without clear zonal predominance.

Throughout both pregnancies, maternal respiratory status, pulmonary function, fetal growth, and laboratory parameters were monitored serially. At the most recent follow-up, approximately 6 years after the first delivery and 3 years after the second, maternal FEV_1 remained stable at 74% of the predicted value

Table 1: Serum VEGF-D levels and PFT results during first pregnancy.

	2 weeks	3 months	6 months	9 months	Postpartum
Serum VEGF-D	820 (pg/ml)	800 (pg/ml)	790 (pg/ml)	780 (pg/ml)	720 (pg/ml)
FEV_1	2.450 (71%)	2.500 (73%)	2.450 (71%)	2.480 (72%)	2.540 (74%)
FVC	2.700 (66%)	2.800 (69%)	2.750 (67%)	2.700 (67%)	2.800 (69%)
DLCO/VA	3.2 (66%)	3.6 (75%)	3.5 (72%)	3.4 (70%)	3.6 (75%)
Sirolimus	(-)	(-)	1 mg/d	1 mg/d	2 mg/d

Table 2: Serum VEGF-D levels and PFT results during second pregnancy.

	2 weeks	3 months	6 months	9 months	Postpartum
Serum VEGF-D	740 (pg/ml)	720 (pg/ml)	700 (pg/ml)	690 (pg/ml)	680 (pg/ml)
FEV_1	2.400 (70%)	2.520 (73%)	2.440 (71%)	2.480 (72%)	2.540 (74%)
FVC	2.750 (67%)	2.820 (69%)	2.780 (69%)	2.710 (66%)	2.860 (70%)
DLCO/VA	3.4 (70%)	3.5 (72%)	3.7 (77%)	3.6 (75%)	3.8 (79)
Sirolimus	(-)	(-)	1 mg/d	1 mg/d	2 mg/d

while receiving continuous sirolimus therapy. Neither pregnancy nor the postpartum period was complicated by pneumothorax, chylous complications, respiratory failure, angiomyolipomas, or other major LAM-related events.The only adverse effect attributed to sirolimus was mild hyperlipidemia, which was managed conservatively without lipid-lowering therapy. Both children exhibited normal growth and development throughout follow-up, without any documented congenital anomalies, immune-mediated disorders, or other complications attributed in utero sirolimus exposure. Maternal pulmonary function remained stable during long-term follow-up, and over approximately 8 years of subsequent observation, no clinically significant LAM-related complications or adverse effects attributable to sirolimus were identified. Breastfeeding was avoided after both pregnancies because of concerns regarding sirolimus transfer into breast milk and potential infant exposure. Follow-up HRCT did not reveal complications associated with LAM (Figure 3) and the overall cystic burden appeared radiologically stable, without evident progression or disease related complications.

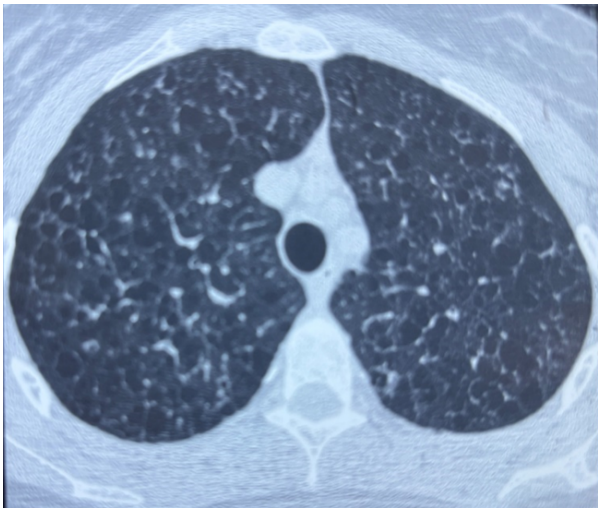


Figure 3. Follow-up HRCT demonstrates diffuse bilateral cystic lung disease without evidence of LAM-related complications.

Discussion

LAM is a rare cystic lung disorder influenced by hormonal factors that predominantly effects women reproductive age. Pregnancy presents a particular clinical challenge in LAM, as increased estrogen exposure may accelerate cystic lung disease, increase the risks of pneumothorax, chylothorax, and respiratory deterioration that may lead to a worse prognosis. Cohen and colleagues first reported a high rate of pneumothorax and premature birth in this population [14]. Subsequently, Taveira-DaSilva et al. documented clinically significant declines in FEV_1 and diffusing capacity for carbon monoxide (DLCO) during pregnancy, together with frequent pneumothorax, in 16 women from two national cohorts [8]. In a series of 34 pregnancies occurring after a diagnosis of LAM, Shen et al. reported live birth in only 10 pregnancies, while LAM-related complications, including dyspnea, pneumothorax, and angiomyolipoma hemorrhage, occurred in 29% [15]. Nevertheless, pregnancy may be tolerated without substantial deterioration in carefully selected women with stable disease and appropriate clinical surveillance [1,2,11-13]. Sirolimus is an established treatment option for LAM that stabilizes pulmonary function, reduces serum vascular endothelial growth factor-D (VEGF-D) concentrations, and improves quality of life through inhibition of mammalian target of rapamycin (mTOR) signaling and abnormal LAM-cell proliferation. Importantly, the therapeutic effect is not considered disease-modifying after treatment withdrawal; lung function decline generally resumes following discontinuation of therapy [9,16-18]. Consequently, complete cessation of sirolimus throughout pregnancy may expose women with clinically significant disease to a period of renewed disease activity at a time of increased hormonal stimulation. The central therapeutic dilemma is therefore whether the potential maternal benefit of continued mTOR inhibition justifies the uncertain fetal risks associated with sirolimus exposure.

This case provides a rare longitudinal observation of two sequential pregnancies in a woman with LAM managed with an individualized sirolimus strategy involving interruption during the first trimester followed by low-dose, concentration-targeted treatment during the second, and third trimesters. Maternal pulmonary function remained stable during both pregnancies, neither of which was complicated by pneumothorax, chylous effusion, clinically significant respiratory deterioration, or other major LAM-related events. Both infants were born without documented congenital anomalies, subsequently demonstrated normal growth and development. Although the favorable maternal and neonatal course cannot establish treatment efficacy or safety, the experience is clinically relevant because pregnancy remains one of the least evidence-based aspects of LAM management. Longitudinal observations of women receiving sirolimus during pregnancy may help refine individualized treatment strategies and generate hypotheses regarding the interaction between hormonal exposure, mTOR signaling, and disease activity. Such an observation is clinically relevant because pregnancy among women with LAM who wish to conceive is not uncommon, yet it remains one of the least evidence-based aspects of disease management which may also yield useful data to elucidate disease pathogenesis. Against this background, the absence of LAM related complications and decline in pulmonary function is noteworthy while a causal relationship with sirolimus therapy cannot be established from a single case.

The available literature provides limited but increasingly informative context. Faehling et al. described two pregnancies in a woman with LAM in whom different sirolimus strategies were used [10]. During the first pregnancy, sirolimus was interrupted early in gestation, followed by pneumothorax and a decline in pulmonary function. Through second pregnancy, low-dose sirolimus was continued, with a concentration of approximately 3.5 µg/L; the pregnancy was uncomplicated from a pulmonary perspective, although delivery occurred at 32 weeks. More recently, Krishna et al. reported 12 pregnancies in nine women with sporadic LAM, most of whom had moderately impaired baseline lung function (median pre-pregnancy FEV_1 56% predicted). Sirolimus was

maintained throughout most pregnancies, generally at low doses, with a median trough concentration of 2.95 ng/mL. Pulmonary function remained stable, no pneumothorax occurred during sirolimus exposure, and 10 pregnancies resulted in live births. At a median pediatric age of 3.5 years, no adverse child-health outcomes were reported. Seven of the 10 infants were breastfed while their mothers continued sirolimus, without apparent adverse effects [12]. Although reassuring, these observations remain insufficient to establish the safety of sirolimus during pregnancy or lactation. The sirolimus exposure strategy used in our patient falls within the low-exposure range reported in the limited pregnancy literature. In both pregnancies, treatment was interrupted during the first trimester and subsequently reintroduced at a reduced dose with therapeutic drug monitoring. Serial FEV_1 measurements remained within a narrow range throughout gestation, without clinically meaningful deterioration. The absence of pneumothorax, chylous complications, angiomyolipoma-related hemorrhage, or respiratory compromise is noteworthy, particularly given the potential for pregnancy-associated disease progression in LAM. However, the favorable course should not be interpreted as evidence that sirolimus prevented pregnancy-related complications. The patient's baseline disease characteristics, natural disease heterogeneity, and close multidisciplinary monitoring may all have contributed to the observed outcome.

Several aspects of this case warrant consideration. First, pulmonary function remained stable across two pregnancies despite the physiological demands of gestation along with LAM presence. Second, the patient did not experience any LAM-related pulmonary or other organ complications during either pregnancy. Third, both pregnancies resulted in infants without documented congenital abnormalities, followed by apparently normal growth and development. Finally, maternal respiratory status remained stable during subsequent long-term treatment with sirolimus. Collectively, the longitudinal course supports the feasibility of an individualized treatment strategy in selected women with LAM but does not establish a causal relationship between sirolimus exposure and the favorable outcomes. Our findings should not, however, be interpreted as evidence that sirolimus is safe during pregnancy. The absence of adverse outcomes in a single patient cannot exclude uncommon congenital, developmental, or other fetal effects. In addition, circulating VEGF-D concentrations and pulmonary function provide measures of disease activity but do not directly assess tissue-level effects of sirolimus or fetal exposure. The clinical course may also have been influenced by the baseline disease characteristics. The decision to continue, interrupt, or modify sirolimus during pregnancy should therefore remain individualized. In patients with clinically significant LAM, discontinuation of effective treatment may carry a potential risk of disease progression, whereas continued exposure may carry uncertain fetal risks. Breastfeeding was avoided in both pregnancies because of concern regarding sirolimus exposure through breast milk. This approach reflects the uncertainty regarding the effects of postnatal sirolimus exposure on the infant and should likewise be considered in individualized maternal–infant risk assessment. While the symptoms and radiologic manifestations at admission revealed a severe LAM profile, the explicit improvement in clinical profile following sirolimus treatment, significantly demonstrated the effectiveness of sirolimus treatment in pregnant patients suggesting that ongoing mTOR inhibition has contributed to disease stabilization.

Several limitations should be acknowledged. The single-patient design precludes definitive conclusions regarding the maternal or fetal safety and efficacy of sirolimus during pregnancy. The observational nature of the report also limits causal inference and does not permit separation of treatment effects from the underlying natural history of LAM. Pediatric follow-up, although reassuring, remains insufficient to exclude subtle developmental or neurodevelopmental effects that could emerge later in childhood. In addition, formal 6-minute-walk testing and a recorded glomerular filtration rate were unavailable, and protocolized neurodevelopmental assessments were not performed in the children. Potential mild or delayed effects of sirolimus on immune function or cognition may not yet be clinically apparent. The absence of adverse outcomes in this case cannot exclude uncommon congenital or developmental effects associated with fetal exposure. The clinical course of the patient may also have been influenced by or associated with the baseline disease characteristics. Finally, the clinical course of the patient may also have been influenced by or associated with the baseline disease characteristics.

Conclusions

Pregnancy in women with LAM remains a complex clinical scenario requiring individualized assessment of maternal disease severity, treatment requirements, and potential fetal risks. This case illustrates the feasibility of an individualized low dose sirolimus treatment strategy, incorporating first-trimester interruption followed by concentration-targeted treatment later in gestation, in a woman who underwent two sequential pregnancies. Maintenance of low-dose, concentration-targeted sirolimus after the first trimester was associated with stable maternal pulmonary function and the successful delivery of two healthy infants without major maternal or fetal complications. Maternal pulmonary function remained stable, and both pregnancies resulted in infants without documented congenital abnormalities or subsequent developmental concerns during follow-up. The favorable course observed in this patient should not be interpreted as evidence that sirolimus is safe during pregnancy or as establishing a protective effect against LAM-related complications. Nevertheless, the experience adds longitudinal clinical data to the limited literature on sirolimus exposure during pregnancy and supports consideration of individualized treatment rather than uniform discontinuation in selected women with clinically significant disease. Avoidance of breastfeeding during sirolimus therapy also remains a reasonable precaution when the potential benefits of treatment are judged to outweigh the benefits of lactation. These observations challenge traditional recommendations for complete discontinuation and underscore the need for updated evidence-based guidance. Given the inherent limitations of a single case, the clinical implications must be interpreted with caution. Prospective multicenter registries and larger observational studies are warranted to

define maternal and fetal safety more precisely, establish optimal dosing and monitoring strategies, and inform evidence-based recommendations for sirolimus use during pregnancy in women with LAM.

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