

## Case Report

### Spontaneous Chylothorax in Pregnancy

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#### Abstract

A left spontaneous chylothorax was detected at 22 weeks in a pregnant woman. Initial chest MRI revealed no mediastinal or pulmonary abnormality. To avoid fetal malnutrition, an appropriate low-fat diet was first established associated with tube thoracostomy. Then, because of major persistent chyle leaks, intravenous nutrition was added while maintaining adapted oral feeding. Associating both feeding ways was atypical in such a situation, but the goal was to maintain a correct level of blood proteins, particularly albumin. Despite preterm labor at 25 weeks, the evolution was favorable after 10 days of drainage. The patient delivered a healthy baby at 35 weeks while second Lymphatic MRI revealed lymphangiomyomatosis, which had caused chylothorax. Multidisciplinary management of chylothorax helped ensure favorable outcomes for the mother and the fetus.

**Keywords:** Chylothorax; Fetal malnutrition; Lymphangiomyomatosis; Lymphatic MRI, Multidisciplinary management, Pregnancy

**Abbreviations:** LAM: Lymphangiomyomatosis; MRI: Magnetic Resonance Imaging

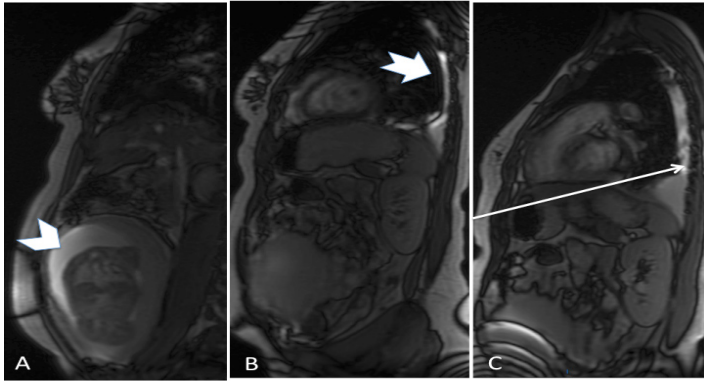
#### Introduction

Spontaneous chylothorax during pregnancy is unusual and can be related to rare lymphatic diseases. Its adapted management, based on clinical experience and a few reports, must take the nutritional needs into consideration to absolutely avoid dietary deficiencies for the mother and her baby. Chest Lymphatic MRI during and after pregnancy may be the most suitable exam to identify underlying causes of chylothorax. Reporting such a complex case may help clinicians in their therapeutic approach to this rare disease.

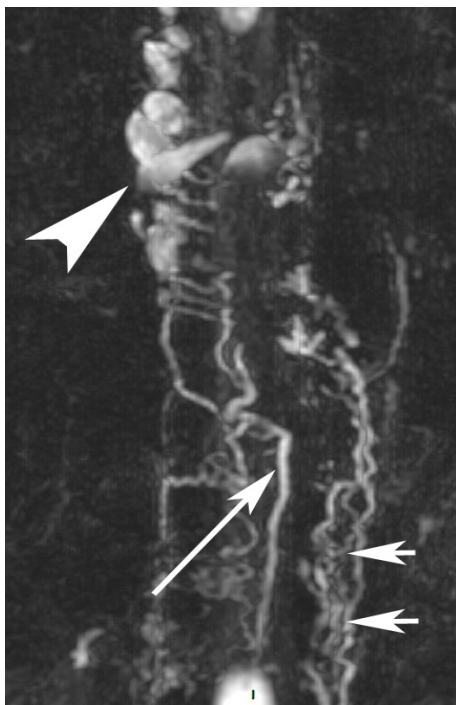
#### Case report

A 31-year-old woman in her 22<sup>nd</sup> week of pregnancy came for urgent consultation because of sudden shortness of breath. Chest X-ray revealed a left pleural effusion. Two liters of chyle were drawn out through thoracocentesis. An oral low-fat diet was started and several thoracocenteses were required. The patient was transferred to our department of thoracic surgery. The chest MRI showed a left pleural effusion (Figure 1A) without mediastinal abnormality (Figure 1B,1C). A soft tube thoracostomy was placed to improve the drainage. The daily chyle flow - initially 1 liter per day – progressively decreased allowing a widened oral regimen to avoid fetal malnutrition. At 25 weeks, a threat of preterm labor occurred justifying specific treatment associated with high-protein intravenous feeding with the oral diet. The chylothorax stopped. The low-fat diet was maintained and a healthy baby was born at 35 weeks through cesarean because of placenta previa. Then another lymphatic MRI showed multiple small mediastinal cystic

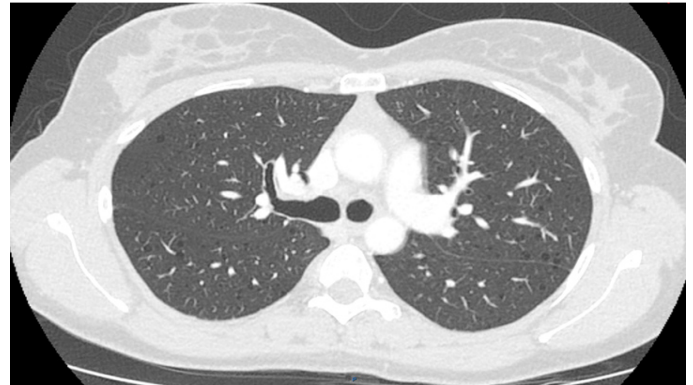
lymphangiomas (Figure 2) and Chest CT scan revealed pulmonary cysts allowing the diagnosis of lymphangioliomyomatosis (LAM) (Figure 3). At 30-month follow-up the woman was in good physical condition with normal daily food intake.



**Figure 1:** Sagittal views by T2-weighted Magnetic Resonance Imaging in the pregnant woman. The fetus is shown by an arrowhead in part A. The left pleural effusion is shown by a short arrow in part B, and a long arrow in part C. There is no mediastinal lymphatic abnormality.



**Figure 2:** Reconstruction views by T2-weighted Magnetic Resonance Imaging performed after delivery. Maximum Intensity Projection reconstruction demonstrates the main thoracic duct (long arrow) and the left lymphatic thoracic plexus (short arrows) replaced by several small cystic lymphangiomas (arrowhead).



**Figure 3:** Chest CT scan performed after delivery demonstrates pulmonary cystic lesions signing the diagnosis of lymphangioliomyomatosis.

## Discussion

Chylothorax complicating pregnancy is rare. Most cases are associated with delivery due to modified intrathoracic and intra-abdominal pressure during labor [1,2]. Other rare cases are related to the exacerbation of LAM or Gorham-Stout disease [1-3] during pregnancy. In our case, the chylothorax was spontaneous and occurred in the second trimester of pregnancy. The first chest MRI did not reveal mediastinal or respiratory disease, so no diagnosis was possible. We limited imaging exploration to minimize radiation and ensure pregnancy progress. Only clinical evaluation was done to follow the evolution of chylothorax. The aim of the treatment was to make the fetus mature and stop the chylothorax, avoiding malnutrition or respiratory distress [1]. The main biological goal was to maintain a normal blood-protein level. A minimal low-fat diet, containing medium chain fatty acids, combined with well tolerated drainage, was effective at the beginning. Then, the persistence of chyle leaks at time of gynecological complications justified associating a partial oral regimen with intravenous nutrition. In fact, exclusive parenteral nutrition is usually required to decrease major chyle leaks but it probably represents an increased risk of malnutrition during pregnancy. Surgical options such as pleurodesis or thoracic duct ligation were discussed, but they were felt to be too risky during the second trimester of pregnancy. Finally, this combined care strategy associating a thoracic surgeon, a gynecologist and a nutritionist allowed a favorable outcome.

Lymphatic MRI is an interesting but probably underused exam to locate the lymphatic abnormalities [4]. In our case it showed multiple small mediastinal cysts suggestive of cystic lymphangiomas. The CT scan done later after delivery revealed multiple pulmonary cysts of varying sizes with a 1mm-thick wall which was pathognomonic of LAM [5]. LAM is a rare systemic

disorder which almost exclusively affects women of childbearing age [5]. It is characterized by an abnormal smooth muscle proliferation in the peribronchial, perivascular and perilymphatic areas of the lung. It occurs sporadically or can be associated with tuberous sclerosis complex [6]. The prevalence of sporadic LAM widely varies from 3.3 to 7.4 per million women [4] Chylothorax has been reported in 0 to 14% of patients with LAM at the beginning of care and 22% to 39% during the course of the disease [7]. Mechanisms of chylothorax formation in LAM include chyle leaks secondary to proximal lymphatic obstruction or direct involvement of the thoracic duct or its tributaries. General oozing comes from pleural lymphatic or collateral vessels and transdiaphragmatic flow of chylous ascites [7]. LAM is a hormonal-dependent disease and may be intensified by the high level of estrogens during pregnancy [2].

## Conclusion

The present case illustrates the difficulty in the diagnosis and management of chylothorax during pregnancy. Therapeutic options combining nutritional, thoracic and gynecological care can be appropriate and discussed with the multidisciplinary team. Indeed, this combined approach can ensure a favorable outcome for the mother and the fetus.

## References

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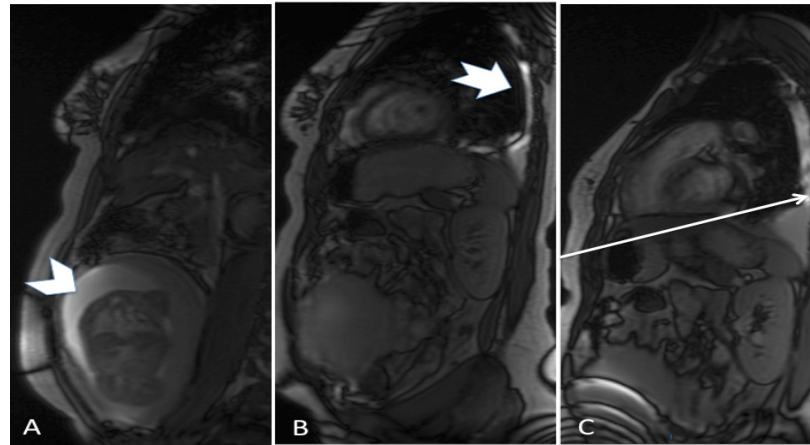
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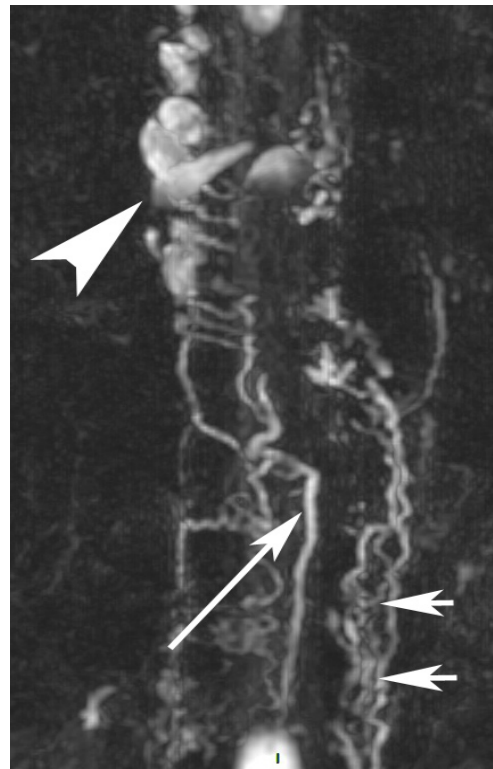
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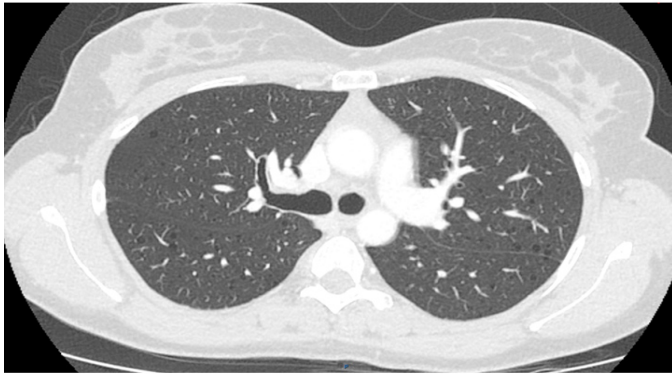


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