



# A pregnant woman with amyotrophic lateral sclerosis from Iran: a case report

Nooshin Amjadi, MD<sup>a</sup>, Sotoudeh Mohammadi, MD<sup>b,\*</sup>, Sepideh Paybast, MD<sup>c</sup>, Peyman Dadkhah, MD<sup>d</sup>, Maryam Talayeh, MD<sup>b,\*</sup>, Zatollah Asemi, PhD<sup>e,\*</sup>

**Introduction and importance:** Amyotrophic lateral sclerosis (ALS) is a progressive motor neuron disease, which is extremely rare during pregnancy. The severity of the disease affects the pregnancy outcome. The present study reports the first Iranian case of a woman with ALS overlapping pregnancy.

**Case presentation:** The 27-year-old lady in her second pregnancy was admitted to the emergency department with labor pain at the 37th gestation week. Following a multidisciplinary team meeting, including a neurologist, maternal-fetal medicine specialist, and anesthesiologist, a decision was made for an emergent cesarean section under spinal anesthesia. The delivery was successful without any maternal or fetal complications. A 5-month follow-up revealed the stable neurologic status of the mother.

**Clinical discussion:** The combination of ALS and pregnancy is very rare because the disease is more common in elderly men. ALS management involves a multidisciplinary approach. Riluzole is a drug that can increase the survival of the patients. ALS does not affect on motor and sensory nerves of the uterus, so vaginal delivery might be possible. The main cause of cesarean section in patients with ALS is respiratory compromise, but four patients with uncomplicated vaginal deliveries have been reported. The neonatal outcome of most cases resulted in normal healthy infants.

**Conclusion:** Management of ALS in pregnancy is challenging because of respiratory concerns, so multidisciplinary team management is important.

**Keywords:** amyotrophic lateral sclerosis, anesthesiologist, outcome, pregnancy

## Introduction

Amyotrophic lateral sclerosis (ALS) is the most common and progressive motor neuron disease, which results in progressive muscle weakness and atrophy, and eventually death over time. ALS has an annual incidence of 2–4/100 000 people worldwide. Before the age of 65–70, the incidence of ALS is higher in men than women, while the incidence is equal afterward<sup>[1,2]</sup>. The explanation for changes in clinical manifestations of patients with

## HIGHLIGHTS

- Amyotrophic lateral sclerosis (ALS) is a progressive motor neuron disease.
- The combination of ALS and pregnancy is very rare because the disease is more common in elderly men.
- Management of ALS in pregnancy is challenging because of respiratory concerns.

<sup>a</sup>Preventative Gynecology Research Center, <sup>b</sup>Department of Obstetrics and Gynecology, <sup>c</sup>Neurology Department, <sup>d</sup>Anesthesiology Department of, Imam Hossein Educational Hospital, School of Medicine and <sup>e</sup>Research Center for Biochemistry and Nutrition in Metabolic Diseases, Institute for Basic Sciences, Kashan University of Medical Sciences, Kashan, Islamic Republic of Iran

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\*Corresponding author. Address: Kashan University of Medical Sciences, Islamic Republic of Iran. E-mail: aseml\_r@yahoo.com (Z. Asemi), Tel.: +98 315 546 3378. and Postal code: 3759153111 Tehran, Iran Tel.: +98 215 631 0619. E-mail: sotoudehmohammadi1993@gmail.com (S. Mohammadi); E-mail: m\_talayeh@yahoo.com (M. Talayeh).

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ALS phenotypes may contribute to the variation of transcriptional adaptation<sup>[3]</sup>. While ALS is a rare disease, it might overlap with pregnancy, posing a significant challenge. The symptoms of ALS may worsen during pregnancy, of which respiratory compromise is of great importance. In this regard, the pregnancy outcome might also be affected by the severity of the disease<sup>[4]</sup>. Given limited reports of ALS overlapping pregnancy in the literature, we aimed to present a successful delivery in a 27-year-old woman with a 3-year history of ALS. This case has been reported in line with the (Surgical Case Report) SCARE 2023 criteria<sup>[5]</sup>.

## Case report

In April 2023, a 27-year-old lady in her second pregnancy came to the emergency department due to labor pain. Her past medical history was positive for ALS for the last 3 years. Her obstetric history included one previous vaginal delivery 3 years prior. In the postpartum period of her first vaginal delivery 3 years ago, 4 days after the delivery, her body's left side lost movement, but the sensation of warmth, cold, and pain remained. Then, her left side became involved, and her speaking became dysarthria. She

reported progressive muscle weakness and dysarthria 2 months after delivery, leading to a diagnosis of ALS 6 months later. Notably, according to the El Escorial criteria, she was diagnosed with definite ALS. Afterward, she started using Riluzole. However, her symptoms gradually worsened. The current pregnancy was unplanned, and her main problem during pregnancy was restricted mobility, which worsened in the third trimester. The patient was still on Riluzole during pregnancy. The patient was still on Riluzole. On her first examination at the prenatal ward, the patient was alert, oriented, and wheelchair-bound. She was hemodynamically stable, and no significant finding was detected in her systemic examinations. She was in the 37th week of pregnancy. An emergent neurologic consult was requested. The neurological examination was notable for four-limb paralysis (3/5) regarding the medical research council score accompanied by generalized hyperreflexia, severe spasticity, atrophy, and fasciculation. No sensory abnormality was found. Cranial nerve examination showed significant wasting of tongue muscles with fasciculation. However, the bulbar muscles were intact. Moreover, the speech was severely spastic dysarthric. During her hospitalization, she had no swallowing or breathing disorders. She made eye and verbal contact; her limbs had diffuse fasciculation. Neurological examination showed hyperreflexia in the four limbs. Four limbs were spastic, but the lower limb spasm was more significant. Her tongue muscle was atrophied. Based on the Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS), her clinical stage was estimated at 21. After a multidisciplinary team meeting of a neurologist and maternal-fetal medicine specialist, a decision was made for an emergent cesarean section. Since it was an emergency, the anesthesiologist, after consultation with the neurologist and obstetrician about her problem and probable respiratory compromise after general anesthesia, decided to select spinal anesthesia for her operation.

Epidural anesthesia provides more hemodynamic stability than spinal anesthesia and is a suitable method for these patients, but it takes time to perform. As mentioned, it was an emergency, and time was limited for doing it. The needle used for spinal anesthesia was spinal needle G25 and the level of needle insertion was at L4-L5. Bupivacaine 0.5% in the amount of 12 mg was used for spinal anesthesia. The level of sensory block was allowed to rise to about T6 for this patient for not involving more intercostal muscles and causing further respiratory distress. The level of the block was checked by pinprick test. Standard monitoring, such as NIBP, Spo<sub>2</sub>, ECG, and HR, was used during surgery. After delivery, 1 mg of midazolam was administered to sedate the patient.

The cesarean section was performed without any complications, and the patient was transferred to the ICU for close vital monitoring. The baby's birth weight was 3140 g. The Apgar score, neurological, and systemic examinations of the neonate were normal. After 2 days, she was transferred to the ward. Both the mother and baby were discharged home 5 days later. The mother received a prophylactic dose of enoxaparin low molecular weight heparin 40 mg once a day until 6 weeks after delivery. At follow-up 5 months later, she remained unchanged, and the baby was in good health.

## Discussion

The coexistence of ALS and Pregnancy is extremely rare because the disease is more prevalent in elderly men. The management of

ALS involves a multidisciplinary approach that aims to manage symptoms, address complications, and provide supportive care to improve the quality of life of the patient<sup>[2]</sup>. Managing ALS during pregnancy can be particularly challenging for both the obstetrician and the patient. Minute ventilation increases by 40% during pregnancy. In the third trimester of pregnancy, diaphragmatic upward elevation due to an enlarging uterus causes the functional residual capacity to decrease, which might lead to ventilation failure if costal and diaphragmatic muscles are involved<sup>[4,6]</sup>. Moreover, pregnancy, delivery, and the immediate postpartum period require an increase in respiratory work, which is normally achieved through diaphragmatic breathing<sup>[7]</sup>. Along with rehabilitation therapy, Riluzole, a class C drug by the Food and Drug Administration (FDA), could be used in pregnancy<sup>[8]</sup>. Still, the association of fetal growth restriction and neonatal malformation with Riluzole is unclear. It defers ventilator dependency and increases the survival of the patients. Being aware of hepatotoxicity, liver function tests are evaluated every 3 months.

There are also major concerns about the mode of delivery. Since ALS does not affect the motor and sensory nerves of the uterus, a vaginal delivery might be possible. However, respiratory compromise is the main cause of cesarean section. The oxygen demand increases during vaginal delivery which might lead to a respiratory crisis in uncontrolled patients<sup>[8]</sup>. Chio *et al.*<sup>[9]</sup> reported four patients with uncomplicated vaginal deliveries. In another report, the patient was initially planned for vaginal delivery. However, the deterioration of symptoms led to the caesarian section<sup>[8]</sup>. In our case, as the patient was not under periodic assessment, and it was not possible to perform spirometry to assess her respiratory function, it was decided to have a cesarean section. Notably, the choice of anesthesia is also challenging. Given the limited reports of ALS overlapping pregnancy, there is no guideline to choose regional or general anesthesia. While regional anesthesia is related to less lung volume decline, it affects intercostal muscles and prevents expiratory flow, leading to a decision-making challenge. In our case, we performed spinal anesthesia leading to successful delivery. Similarly, Kock-Cordeiro *et al.*<sup>[10]</sup> recommended spinal anesthesia to avoid unnecessary intubation in patients with fast-worsening ALS. Eventually, pregnancy and neonatal outcomes are other important issues in all pregnancies. In a recent systematic review of 38 pregnancies overlapping ALS, 95% of patients gave live birth. However, only 33% of these completed pregnancies reached 38 weeks. In terms of neonatal outcome, most cases resulted in normal healthy infants. Only three infants showed transient complications after delivery, while one infant had a small atrial communication and a small patent ductus arteriosus<sup>[7]</sup>. In line with previous reports, we showed no complications in the neonate and mother. Taking all considerations, there is little information on pregnancy in women with ALS. To the best of our knowledge, there is no report of pregnancy overlapping ALS in Iran. In our case, a successful pregnancy was achieved 3 years after ALS onset. Our patient's symptoms worsened, especially around the third trimester, as has been reported in previous cases. However, the delivery was successful, and a 5-month follow-up revealed the neurologic stability of both the mother and the baby. The present report highlights the importance of appropriate counseling for women with ALS.

## Conclusion

ALS in pregnancy is an important condition because of respiratory impairment. Riluzole is a drug that can be used during pregnancy. A multidisciplinary team is necessary for the care of patients with ALS in pregnancy to achieve the best fetal and maternal outcomes.

## Ethical approval

Given the nature of the article, a case report, no ethical approval was required.

## Consent to participate

Written informed consent was obtained from the patient for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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## Author contribution

All authors contributed to this manuscript. N.A., S.M., N.P., and P.D.: the case treatment and writing original draft; M.T.: original draft, reviewing, and editing; Z.A.: supervision, reviewing, and editing.

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