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Date: 13 July 2017

Sources Searched: Medline, Embase, PubMed.

## Motor Neuron Disease and Pregnancy

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### **1. Total intravenous anesthesia without muscle relaxant in a parturient with amyotrophic lateral sclerosis undergoing cesarean section: A case report.**

**Author(s):** Xiao W; Zhao L; Wang F; Sun H; Wang T; Zhao G

**Source:** Journal of clinical anesthesia; Feb 2017; vol. 36 ; p. 107-109

**Publication Date:** Feb 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28183545

Available in full text at [Journal of Clinical Anesthesia](#) - from ProQuest

**Abstract:**Cases of amyotrophic lateral sclerosis with pregnancy are quite rare. The aim of this case report is to present the successful use of total intravenous anesthesia without muscle relaxant for cesarean section in a parturient with amyotrophic lateral sclerosis at 35 weeks' gestation. The parturient presented with impaired neuromuscular function. Titrated general anesthesia with short-acting anesthetics was applied. During the procedure, no muscle relaxant was used. This strategy helped avoid prolonged ventilation and prevent maternal respiratory complications.

**Database:** PubMed

### **2. Motor neurone disease in pregnancy: A case report**

**Author(s):** Beebeejaun Y.; Gao J.; Krishna A.; Veerareddy S.

**Source:** BJOG: An International Journal of Obstetrics and Gynaecology; Dec 2016; vol. 123 ; p. 26

**Publication Date:** Dec 2016

**Publication Type(s):** Conference Abstract

Available in full text at [BJOG: An International Journal of Obstetrics and Gynaecology](#) - from John Wiley and Sons

**Abstract:**Background Amyotrophic Lateral Sclerosis (ALS), is especially rare in obstetrics with only 6 case reports having appeared in the medical literature since 1977. Consequently, the management of an obstetrics patient suffering motor neurone disease (MND), including ALS, can be particularly challenging for both physician and patient. Little is known about the risks of ALS in pregnancy but with its progressive and degenerative nature, a combination of upper and lower motor neurone deficits can be expected. Even though ALS is not particularly associated with poorer neonatal outcomes, maternal disease does not regress and may in fact worsen throughout the pregnancy due to increased respiratory and weight bearing demands. Case We describe the case of a previously healthy 41-year-old who presented to us complaining of slurred speech and generalised weakness in

her fingers at 25 weeks of gestation. A complete neurological assessment further revealed evidence of tongue atrophy and fasciculation, and mild bilateral lower limb spastic hypertonia. The diagnosis of Motor Neurone Disease was made via Nerve Conduction Studies and a multidisciplinary approach was adopted for the ongoing management of this patient. Respiratory efforts became increasingly laborious for our patient and an elective caesareans section was performed at 38 weeks of gestation. The operation was uneventful with no neonatal concerns. Conclusion Pregnancy in women with ALS is rare and is generally considered a potentially dangerous event. Complications of this disease mainly affect the respiratory system and labour management should be tailored to the patient's need and severity of the disease. MND does not tend to involve the uterine sensory and motor nerves and therefore pregnancy and the delivery may be normal, but respiratory function should be carefully monitored. Generally, ALS does not have harmful consequences on fetal development but careful assessment of disease in the mother is vital.

**Database:** EMBASE

### **3. Acute motor and sensory axonal neuropathy in pregnancy: A rare case**

**Author(s):** Nikath Nasreen N.; Saraswathi K.

**Source:** Research Journal of Pharmaceutical, Biological and Chemical Sciences; 2015; vol. 6 (no. 6); p. 192-194

**Publication Date:** 2015

**Publication Type(s):** Article

**Abstract:**This is a rare case of AMSAN with good prognosis. Guillain-Barre syndrome (GBS) was first described in 1916 (Guillain G, 1916) and is approaching its 100th anniversary. Guillain barre syndrome is rare in pregnancy. incidence is between 1.2 to 1.9 per 100000 persons annually. It is an autoimmune neurological disorder. There are many subtypes acute inflammatory demyelinating polyradiculoneuropathy (AIDP) being most common followed by acute motor axonal neuropathy (AMAN) and much rarer is acute motor and sensory axonal neuropathy (AMSAN). IVIG and plasmapheresis is the mainstay of treatment.

**Database:** EMBASE

### **4. Total intravenous anesthesia for cesarean section in a pregnant woman with spinal muscular atrophy.**

**Author(s):** Fang, Qiwu; Gao, Guolan; An, Jianxiong; Liu, Caicai; Qian, Xiaoyan; Wen, Hui; Wu, Jianping; Wang, Yong; Cope, Doris K; Williams, John P

**Source:** Chinese medical journal; 2014; vol. 127 (no. 18); p. 3350-3351

**Publication Date:** 2014

**Publication Type(s):** Research Support, Non-u.s. Gov't Case Reports Journal Article

**PubMedID:** 25266539

**Database:** Medline

## **5. Acute non-invasive ventilation during pregnancy in a patient with amyotrophic lateral sclerosis**

**Author(s):** Kock-Cordeiro D.B.M.; Eggink A.J.; Van Der Marel C.D.; Van Den Biggelaar R.J.M.

**Source:** European Respiratory Journal; Sep 2014; vol. 44

**Publication Date:** Sep 2014

**Publication Type(s):** Conference Abstract

Available in full text at [European Respiratory Journal](#) - from Free Access Content

Available in full text at [European Respiratory Journal](#) - from Highwire Press

**Abstract:**One of the most common progressive neurodegenerative diseases is amyotrophic lateral sclerosis (ALS). It is diagnosed in the 5th to 7th decade and is common in males. ALS in women of childbearing age and coexisting pregnancy is uncommon. We would like to describe the first reported case in which non-invasive ventilation (NIV) was acutely initiated in a pregnant patient with ALS. Case report A 25-year-old patient, G2P1, had complaints of speech, swallowing difficulty and tiredness that she credited to her pregnancy. She later developed weakness in her right arm and sought consultation. Physical exam revealed myopathic facies, dysarthria, tongue atrophy and fasciculation, weakness in neck flexor and right arm muscles. After two months ALS was diagnosed. Fetal development was uncomplicated. At her first visit to the respiratory department at 32 weeks of gestation, she had no pulmonary complaints. Examination revealed weak cough capacity, peripheral saturation 98% (room air) and decreased forced vital capacity (42% of predicted). Transcutaneous CO<sub>2</sub> and O<sub>2</sub> measurement was normal during sleep. Four days later she was admitted with progressive dyspnea, orthopnea and respiratory exhaustion. NIV with oronasal mask was acutely initiated and well tolerated despite poor bulbar function. Due to rapid deterioration a caesarian section was performed under NIV and spinal anesthesia. Intubation and total anesthesia with was averted. A healthy male baby was born. Postoperative NIV could be weaned slowly to only nighttime use and she was discharged with domiciliary NIV. Conclusion Acute NIV in pregnant ALS patient with respiratory distress could be safely initiated.

**Database:** EMBASE

## **6. Amyotrophic lateral sclerosis in pregnancy is associated with a vascular endothelial growth factor promoter genotype.**

**Author(s):** Lunetta, C; Sansone, V A; Penco, S; Mosca, L; Tarlarini, C; Avemaria, F; Maestri, E; Melazzini, M G; Meola, G; Corbo, M

**Source:** European journal of neurology; Apr 2014; vol. 21 (no. 4); p. 594-598

**Publication Date:** Apr 2014

**Publication Type(s):** Research Support, Non-u.s. Gov't Journal Article

**PubMedID:** 24471417

Available in full text at [European Journal of Neurology](#) - from John Wiley and Sons

**Abstract:**BACKGROUND AND PURPOSEThe occurrence of amyotrophic lateral sclerosis (ALS) during pregnancy is uncommon and the effect of one on the other is not well described.METHODThe clinical and genetic features of five cases of ALS are reported with an onset during pregnancy or within 1 month from delivery. Charts from 239 women with a diagnosis of ALS attending the neuromuscular clinics at the Neuromuscular Omnicentre (NEMO) and at IRCCS Policlinico San Donato from 2008 to 2011 were reviewed.RESULTSO f these, 12.8% of the women in child-bearing age had a diagnosis of ALS during pregnancy or immediately after delivery. Genetic screening of the major causative genes revealed two mutations in superoxide dismutase 1 (SOD1) gene; the analysis of vascular endothelial growth factor (VEGF) promoter variation showed a segregation of the haplotype CA/AG (-2578C/A; -1190A/G) in patients developing ALS related to pregnancy. No effects

on foetal development or neonatal course were observed. **CONCLUSIONS** Pregnancy may unmask ALS but whether this is coincidental is unclear. Hormonal and inflammatory modifications might trigger ALS in subjects with increased susceptibility to oxidative stress related to the toxic function of SOD1 or in subjects with a reduction of neuroprotective molecules such as VEGF.

**Database:** Medline

### **7. Amyotrophic lateral sclerosis in pregnancy: clinical outcome during the post-partum period after stem cell transplantation into the frontal motor cortex.**

**Author(s):** Martínez, Hector R; Marioni, Sergio Salazar; Escamilla Ocañas, César E; Gonzalez Garza, María Teresa; Moreno-Cuevas, Jorge E

**Source:** Cytotherapy; Mar 2014; vol. 16 (no. 3); p. 402-405

**Publication Date:** Mar 2014

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 24418405

**Abstract:** **BACKGROUND** AIMS Amyotrophic lateral sclerosis (ALS) is rare in pregnant patients. Stem cell therapy has been proposed as a potential therapeutic strategy for ALS. **METHODS** We describe a young woman with sporadic ALS that started during the second trimester of pregnancy with a rapid progression after delivery and severe motor impairment. Several drugs and stem cell injection by lumbar puncture were performed without changes before the patient was referred to our institution. **RESULTS** After bilateral autologous stem cell transplantation into the frontal motor cortices, we observed stabilization in ALS functional rating scale, significant delay of ALS progression and an extension in her life expectancy. **CONCLUSIONS** Stem cell transplantation may alter the clinical course of ALS and improve quality of life in pre-menopausal women.

**Database:** Medline

### **8. Case report - Anesthetic management for cesarean section in a patient with amyotrophic lateral sclerosis**

**Author(s):** Faria J.; Pereira S.; Castro P.

**Source:** Regional Anesthesia and Pain Medicine; 2012; vol. 37 (no. 5)

**Publication Date:** 2012

**Publication Type(s):** Conference Abstract

Available in full text at [Regional Anesthesia and Pain Medicine](#) - from Ovid

**Abstract:** **Introduction:** Amyotrophic Lateral Sclerosis (ALS) causes progressive degeneration and death of upper and lower motor neurons. Anesthetic management has been controversial and can be challenging. Occurrence in women of reproductive age is exceedingly rare. We describe a case in which epidural anesthesia was successfully performed in an elective cesarean section (CS). **Case report:** A 38 year-old parturient with ALS, progressive bulbar palsy and no other relevant medical records was scheduled for a CS at 34 weeks pregnancy. She had dysarthria, dysphagia, fasciculations, muscle weakness, atrophy of upper and lower extremities as well as intercostal and pectoralis muscles. No sensory loss or autonomic dysfunction were detected. Anesthetic management was performed under epidural block with standard monitoring. A total volume of 12ml ropivacaine 0.75% and 5Kg sufentanil were administered and a sensitive blockade up to T7 was determined. A healthy female infant was delivered. Procedure lasted 40 minutes and was uneventful except for transient hypotension treated with 10mg ephedrine. Postoperative analgesia: morphine epidural bolus and acetaminophen. Patient was discharged on day 4 post CS without complications.

Conclusions: Respiratory failure represents the main risk of general anesthesia due to abnormal responses to neuromuscular blockers. Although neuraxial anesthesia is not contraindicated, still carries the fear of neurologic exacerbation. However, evidence demonstrates that it seems to be safe. Besides the advantages of neuraxial anesthesia in obstetrics, epidural block in particular was a prudent approach, allowing titration of block height, gradual onset, smaller concentration of local anesthetic in cerebrospinal fluid and good postoperative pain control.

**Database:** EMBASE

### **9. Anesthetic management of a parturient with spinal muscular atrophy type II.**

**Author(s):** Maruotti, Giuseppe Maria; Anfora, Rita; Scanni, Emilio; Rispoli, Marco; Mazzairelli, Laura Letizia; Napolitano, Raffaele; Morlando, Maddalena; Sarno, Laura; Milanes, Giovanna Mallia; Simioli, Stefania; Migliucci, Annalisa; Martinelli, Pasquale; Mastronardi, Pasquale

**Source:** Journal of clinical anesthesia; Nov 2012; vol. 24 (no. 7); p. 573-577

**Publication Date:** Nov 2012

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 22999982

Available in full text at [Journal of Clinical Anesthesia](#) - from ProQuest

**Abstract:**In the past, pregnancy was contraindicated in patients with spinal muscular atrophy. Recently, more cases are occurring because of improvement in survival and functional status. The goals for anesthetic management of these patients include satisfactory anesthesia during surgery and excellent postoperative analgesia with minimal compromise of respiratory function. Spinal anesthesia may be considered contraindicated due to spinal deformities, but successful spinal anesthesia was performed in a 37 year old parturient following magnetic resonance imaging of the spine.

**Database:** Medline

### **10. Amyotrophic lateral sclerosis and riluzole use during pregnancy: a case report.**

**Author(s):** Scalco, Renata Siciliani; Vieira, Matias Costa; da Cunha Filho, Edson Vieira; Lago, Eleonor Gastal; da Silva, Irenio Gomes; Becker, Jefferson

**Source:** Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases; Sep 2012; vol. 13 (no. 5); p. 471-472

**Publication Date:** Sep 2012

**Publication Type(s):** Letter Case Reports

**PubMedID:** 22670879

**Database:** Medline

## **11. Neuromuscular Disorders in Pregnancy**

**Author(s):** Guidon A.C.; Massey E.W.

**Source:** Neurologic Clinics; Aug 2012; vol. 30 (no. 3); p. 889-911

**Publication Date:** Aug 2012

**Publication Type(s):** Review

**PubMedID:** 22840795

**Abstract:**Preexisting and coincident neuromuscular disorders in pregnancy are challenging for clinicians because of the heterogeneity of disease and the limited data in the literature. Many questions arise regarding the effect of disease on the pregnancy, delivery, and newborn in addition to the effect of pregnancy on the course of disease. Each disorder has particular considerations and possible complications. An interdisciplinary team of physicians is essential. This article discusses the most recent literature on neuromuscular disorders in pregnancy including acquired root, plexus, and peripheral nerve lesions; acquired and inherited neuropathies and myopathies; disorders of the neuromuscular junction; and motor neuron diseases. © 2012 Elsevier Inc.

**Database:** EMBASE

## **12. Postpartal onset of amyotrophic lateral sclerosis (ALS): A case report**

**Author(s):** Kuppelich N.; Tettenborn B.; Felbecker A.

**Source:** Schweizer Archiv fur Neurologie und Psychiatrie; 2011; vol. 162

**Publication Date:** 2011

**Publication Type(s):** Conference Abstract

**Abstract:**Introduction: ALS is a rare neurological disorder leading to degeneration of upper and lower motor neurons. Mean age of onset ranges from 45 to 65 years and males are affected more often than females. There are only a few reports of ALS presenting during pregnancy. We report the case of a 25-year-old woman who developed first symptoms of ALS in childbed immediately after delivery of her second child. Case presentation: One day after uncomplicated spontaneous vaginal delivery of her second child the 25-year-old woman noticed muscle weakness of her right hand. The weakness eventually spread to the right foot and finally to the left hand within four months. Moreover, the patient developed dysphagia, dysarthria and forced laughing and crying. Results: Neurological examination five months after symptom onset revealed tetraspasticity with hyperreflexia in all limbs, atrophy of the small hand and foot muscles, fibrillation of the tongue as well as severe dysarthrophonia. Electromyography showed signs of widespread denervation in three out of four regions of the body. Motor evoked potentials (MEP) showed signs of pyramidal tract lesions to all limbs, whereas somatosensory evoked potentials (SEP) and sensory nerve conduction studies revealed normal results. Further examinations including MRI of brain and spine and extensive laboratory and CSF testing were performed to rule out other causes of motor neuron degeneration, all with negative results. Particular attention was given to exclude immune-mediated diseases like paraneoplastic disorders. At last, the diagnosis of ALS was definite according to El Escorial and Awaji criteria. Treatment with riluzole was started. Conclusion: To our knowledge, this is the first report of ALS onset in childbed. Whether this coincidence is a matter of chance or does reflect a causal link remains unclear to date. It might be possible that the disease already began during pregnancy without overt clinical signs. As mentioned above, some reports of ALS onset in pregnancy exist. Thus, a relationship between hormonal changes during pregnancy and an increased susceptibility to ALS may be discussed. Finally, the number of patients with ALS onset during pregnancy or in childbed is too small to draw definite conclusions.

**Database:** EMBASE

### **13. Management of a pregnancy complicated by type III spinal muscular atrophy.**

**Author(s):** Howarth, L; Glanville, T

**Source:** BMJ case reports; Feb 2011; vol. 2011

**Publication Date:** Feb 2011

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 22707496

Available in full text at [BMJ Case Reports](#) - from Highwire Press

Available in full text at [BMJ Case Reports](#) - from National Library of Medicine

**Abstract:**The authors report the successful management of a pregnancy in a patient with spinal muscular atrophy (SMA) type III. It is a genetically inherited condition causing increasing weakness of the skeletal muscle. The patient in our case was confined to a wheelchair due to marked weakness in her lower limbs. A review of the available literature identified potential risk factors for the antenatal, intrapartum and postpartum period. These include increased risk of thromboembolism, urinary tract infections, intrauterine growth restriction and preterm delivery, anaesthetic problems and increased risk of uterine atony with subsequent postpartum haemorrhage. The authors report the management of these risk factors and the reasons for delivery by Caesarean section at 32 weeks. Apart from a decline in muscle function postdelivery requiring physiotherapy, there were no adverse outcomes for mother or baby. In both this case and on review of the literature, it is proven that a successful pregnancy is possible with SMA.

**Database:** Medline

### **14. Riluzole use during pregnancy in a patient with amyotrophic lateral sclerosis: a case report.**

**Author(s):** Kawamichi, Y; Makino, Y; Matsuda, Y; Miyazaki, K; Uchiyama, S; Ohta, H

**Source:** The Journal of international medical research; 2010; vol. 38 (no. 2); p. 720-726

**Publication Date:** 2010

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 20515588

Available in print at [Patricia Bowen Library and Knowledge Service West Middlesex university Hospital](#) - from Journal of International Medical Research

Available in full text at [Journal of International Medical Research](#) - from Free Access Content

**Abstract:**Pregnancy with amyotrophic lateral sclerosis (ALS) is rare and generally considered dangerous. Riluzole is the only drug approved for use in ALS, but the effect on maternal and fetal health is unknown. We describe the case of an ALS patient taking riluzole throughout pregnancy. A 34-year old Japanese woman, who had been diagnosed with probable ALS 4 years earlier, visited our hospital for abdominal distension, without knowing that she was pregnant. The patient had been taking riluzole for 2 years, inclusive of her gestational months, and we decided to continue administration of the medication. The patient delivered a normal female infant transvaginally at 38 weeks gestation. The patient's neurological status was stable 1 year after delivery and the baby had developed normally. We found that, in this case, riluzole did not cause any side-effects to the pregnant woman or her fetus.

**Database:** Medline

### **15. Two consecutive pregnancies in early and late stage of amyotrophic lateral sclerosis.**

**Author(s):** Sarafov, Stayko; Doitchinova, Maryana; Karagiozova, Zhvka; Slancheva, Boriana; Dengler, Reinhard; Petri, Susanne; Kollewe, Katja

**Source:** Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases; 2009; vol. 10 (no. 5-6); p. 483-486

**Publication Date:** 2009

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 19922145

**Abstract:**There are few reports on pregnancies in sporadic and familial amyotrophic lateral sclerosis (ALS). We report on a young woman with sporadic ALS who gave birth twice during the course of her disease. The first pregnancy occurred 13 months after the onset of symptoms, and one month after diagnosis. The pregnancy was uncomplicated and resulted in vaginal delivery of a healthy boy. Fifteen months later, when she was already bed-ridden, she became pregnant again. She received a percutaneous endoscopic gastrostomy in the 21st gestational week and underwent early Caesarean section in the 34th week of gestation. The child was ventilated for 72 h in a neonatological unit. The patient was tracheotomized and ventilated two months later, i.e. 47 months after symptom onset, and died nine months later from gastrointestinal haemorrhage. Her two children have developed without abnormalities to date. This case confirms that pregnancies in early-stage ALS can develop normally and may result in uncomplicated vaginal delivery. Pregnancies in late stages may be critical for mother and child, and early delivery by Caesarean section may become necessary although neonatal outcome can be good.

**Database:** Medline

### **16. Dexmedetomidine for awake fiberoptic intubation in a parturient with spinal muscular atrophy type III for cesarean delivery.**

**Author(s):** Neumann, M M; Davio, M B; Macknet, M R; Applegate, R L

**Source:** International journal of obstetric anesthesia; Oct 2009; vol. 18 (no. 4); p. 403-407

**Publication Date:** Oct 2009

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 19733055

**Abstract:**Spinal muscular atrophy in pregnancy is rare and poses multiple problems for the anesthesiologist. The effects of dexmedetomidine on a parturient with spinal muscular atrophy have not previously been reported. There are also no in vivo data on placental transfer of dexmedetomidine and its effects on a human neonate. We report the hemodynamic, respiratory and sedative effects of dexmedetomidine on a parturient and neonate when used for awake fiberoptic intubation before cesarean section. A 35-year-old, gravida 4 para 0 aborta 3, 41-kg parturient at 35 weeks of gestation with spinal muscular atrophy presented for cesarean section. Dexmedetomidine was administered intravenously, total dose 1.84 microg/kg over 38 minutes, followed by fiberoptic endotracheal intubation. Dexmedetomidine was then discontinued and general anesthesia was induced. The baby was delivered 68 minutes after the dexmedetomidine infusion was discontinued at which time blood samples were obtained for measurement of dexmedetomidine. During administration of dexmedetomidine, maternal heart rate, blood pressure and oxygen saturation remained stable. Apgar scores at 1 and 5 min were 6 and 8. The fetal concentration of dexmedetomidine (540 pg/mL) indicates significant placental transfer, but significant adverse neonatal effects were not observed. Dexmedetomidine alone provided adequate sedation for awake intubation without respiratory compromise in this patient.



**Database:** Medline

**17. Amyotrophic lateral sclerosis presenting during pregnancy: Report of clinical and genetic features of three cases**

**Author(s):** Lunetta C.; Sansone V.; Penco S.; Panzeri M.C.; Maestri E.; Meola G.; Corbo M.

**Source:** Journal of Neurology; Jun 2009; vol. 256

**Publication Date:** Jun 2009

**Publication Type(s):** Conference Abstract

Available in full text at [Journal of Neurology](#) - from ProQuest

Available in full text at [Journal of Neurology](#) - from Springer Link Journals

**Abstract:** Objectives: A few previous reports have described patients with ALS presenting during pregnancy, but the association is rare and pathogenic relationship is still to be demonstrated. In this study we wished to evaluate the frequency, clinical and genetic aspects of ALS cases, associated with pregnancy, in a cohort of patients attending our neuromuscular centre. Methods: We retrospectively analysed charts from female patients with a diagnosis of ALS attending the Muscle Clinics at Nemo Center and at IRCCS Policlinico San Donato during the year 2008. Of 76 female patients, 3 had the diagnosis of ALS during the first pregnancy (4%). Results: Case 1: a previously healthy 33-year-old woman presented with subacute, severe, left lower limb pain after delivery, followed by progressive weakness and wasting of the left leg, mimicking a lumbo-sacral radiculopathy. Electrophysiological studies showed widespread denervation involving more than one district. Genetic studies demonstrated an aspartate for glycine substitution at position 93 (G93D) in the Cu/Zn superoxide dismutase 1 (SOD1). Case 2: A 43-year-old woman started complaining of diffuse fasciculations in the right leg, followed by progressive weakness and wasting, immediately after delivery. Family history and electrophysiological studies confirmed the diagnosis of ALS. Genetic studies demonstrated a phenylalanine for leucine substitution at position 84 (L84F) in the SOD1. Case 3: A 32-year-old pregnant woman presented with progressive weakness and oedema of the left hand during her first pregnancy (at 6-months). A cesarean delivery at week 43 was performed because of gestosis. Weakness rapidly got worse and progressed to the 4 limbs. Severe muscle atrophy and widespread fasciculations became evident in the months following delivery. Electrophysiological studies showed fasciculation and denervation in several districts. Genetic studies ruled out mutations in the SOD1 gene. Conclusion: Our data confirm that, although rare, pregnancy may reveal an already present but not yet clinically overt ALS through pathological mechanisms yet to be determined. Further studies are needed to explore the possibility that, amongst others, hormonal modifications during pregnancy may increase the susceptibility to oxidative stress related, for instance, to mutations in SOD1.

**Database:** EMBASE

**18. ACOG committee opinion No. 432: spinal muscular atrophy.**

**Author(s):** ACOG Committee on Genetics

**Source:** Obstetrics and gynecology; May 2009; vol. 113 (no. 5); p. 1194-1196

**Publication Date:** May 2009

**Publication Type(s):** Practice Guideline Journal Article

**PubMedID:** 19384151

Available in print at [Patricia Bowen Library and Knowledge Service West Middlesex university Hospital](#) - from Obstetrics and Gynecology

Available in full text at [Obstetrics and Gynecology](#) - from Ovid

**Database:** Medline

**19. Rapid progression of amyotrophic lateral sclerosis presenting during pregnancy: a case report.**

**Author(s):** Leveck, David E; Davies, Gregory A

**Source:** Journal of obstetrics and gynaecology Canada : JOGC = Journal d'obstetrique et gynecologie du Canada : JOGC; Apr 2005; vol. 27 (no. 4); p. 360-362

**Publication Date:** Apr 2005

**Publication Type(s):** Case Reports Journal Article Review

**PubMedID:** 15937610

**Abstract:**BACKGROUND Amyotrophic lateral sclerosis (ALS) is a progressive neurologic disease that is rare within the obstetric population. Only 5 reports of ALS in pregnancy have appeared in the medical literature since 1977. CASEA previously healthy 25-year-old pregnant woman with 2 previous births presented with generalized weakness at 22 weeks' gestation. Initial laboratory values were all normal. She was admitted to hospital at 29 weeks' gestation and intubated because of respiratory compromise. Daily biophysical profiles and amniotic fluid indices were normal. Labour was induced at 34.5 weeks' gestation, and delivery was uncomplicated. She died at 9 months postpartum. CONCLUSION Pregnancy complicated by the onset of ALS is a rare event. Complications of this disease mainly affect the respiratory system. Death in this case occurred more rapidly than in other previously reported cases.

**Database:** Medline

**20. Caring for a laboring woman with amyotrophic lateral sclerosis: a case report.**

**Author(s):** Sobrino-Bonilla, Yusimi

**Source:** MCN. The American journal of maternal child nursing; 2004; vol. 29 (no. 4); p. 243-247

**Publication Date:** 2004

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 15238751

Available in full text at [MCN, American Journal of Maternal Child Nursing](#) - from Ovid

**Abstract:**A case presentation of a pregnant 32-year-old woman with advanced amyotrophic lateral sclerosis who presented in labor at 38+6 weeks gestation is described. Amyotrophic lateral sclerosis is a progressive neuromuscular disease that attacks nerve cells in the brain and spinal cord. It is usually diagnosed after the fourth decade of life, and is more commonly seen in men than women; thus, it is rarely found in the obstetric population. When the perinatal team is confronted with a pregnant woman with ALS, patient management can be a challenge.

**Database:** Medline

**21. Anaesthesia for caesarean section in spinal muscular atrophy type III.**

**Author(s):** McLoughlin, L; Bhagvat, P

**Source:** International journal of obstetric anesthesia; Jul 2004; vol. 13 (no. 3); p. 192-195

**Publication Date:** Jul 2004

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 15321401

**Abstract:**We describe the conduct of general anaesthesia for a patient with spinal muscular atrophy Type III (Kugelberg-Welander disease) undergoing elective caesarean section. Apart from a delayed return of skeletal muscle power following non-depolarising neuromuscular blockade the procedure was uneventful. We found no previously published reports of general anaesthesia for caesarean section in this condition in the English language literature. We review the available literature and discuss the potential anaesthetic problems in the management of obstetric patients with this degenerative neuromuscular disorder.

**Database:** Medline

**22. Anesthesia for cesarean section in a patient with spinal muscular atrophy.**

**Author(s):** Habib, Ashraf S; Helsley, Scott E; Millar, Simon; Deballi, Pete; Muir, Holly A

**Source:** Journal of clinical anesthesia; May 2004; vol. 16 (no. 3); p. 217-219

**Publication Date:** May 2004

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 15217664

Available in full text at [Journal of Clinical Anesthesia](#) - from ProQuest

**Abstract:**We describe the anesthetic management for cesarean section and tubal ligation of a 23-year-old primipara with type II spinal muscular atrophy (benign Werdnig Hoffmann). She was wheelchair-bound, had severe restrictive lung disease, and severe kyphoscoliosis, with Harrington rods extending from the thoracic to the sacral spines. A general anesthetic was given. We used propofol and alfentanil for rapid-sequence induction of anesthesia. We did not use any muscle relaxants intraoperatively. Postoperative care was provided in the intensive care unit. The patient made a good recovery.

**Database:** Medline

**23. Caesarean section in a parturient with type III spinal muscular atrophy and pre-eclampsia.**

**Author(s):** Kitson, R; Williams, V; Howell, C

**Source:** Anaesthesia; Jan 2004; vol. 59 (no. 1); p. 94-95

**Publication Date:** Jan 2004

**Publication Type(s):** Letter Case Reports

**PubMedID:** 14687114

Available in full text at [Anaesthesia](#) - from John Wiley and Sons

**Database:** Medline

#### **24. Amyotrophic lateral sclerosis associated with pregnancy: report of four new cases and review of the literature.**

**Author(s):** Chiò, Adriano; Calvo, Andrea; Di Vito, Nicoletta; Vercellino, Marco; Ghiglione, Paolo; Terreni, Anna; Mutani, Roberto; Mora, Gabriele

**Source:** Amyotrophic lateral sclerosis and other motor neuron disorders : official publication of the World Federation of Neurology, Research Group on Motor Neuron Diseases; Apr 2003; vol. 4 (no. 1); p. 45-48

**Publication Date:** Apr 2003

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 12745618

**Abstract:** Pregnancy in women with ALS is rare and is generally considered a potentially dangerous event. We describe four ALS cases associated with pregnancy, together with a review of the literature. Three of the four women described developed ALS during pregnancy. In three cases a normal delivery was performed, with a healthy child. One patient, with severe respiratory failure, underwent an interruption of pregnancy. Seven other cases are reported in the literature, featuring a total of 11 pregnancies. The association between pregnancy and ALS is quite rare, and a pathogenic relationship cannot be excluded. The pregnancy and the delivery may be normal, but respiratory function should be carefully monitored. Generally, ALS does not have deleterious effects on fetal development. However, pregnancy in a woman with severe respiratory failure may precipitate the disease.

**Database:** Medline

#### **25. Successful pregnancy in a patient with spinal muscular atrophy and severe kyphoscoliosis.**

**Author(s):** Yim, Roger; Kirschner, Kristi; Murphy, Eileen; Parson, John; Winslow, Christopher

**Source:** American journal of physical medicine & rehabilitation; Mar 2003; vol. 82 (no. 3); p. 222-225

**Publication Date:** Mar 2003

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 12595774

Available in full text at [American Journal of Physical Medicine and Rehabilitation](#) - from Ovid

**Abstract:** Pregnancy imposes a load on the respiratory system that is usually easily assumed because of alterations in the thoracoabdominal architecture. It is presumed that the respiratory mechanical disadvantage of severe kyphoscoliosis and the muscle weakness of spinal muscular atrophy impede these adaptations sufficiently to preclude a successful gestation. We report the case of a successful pregnancy in a woman with spinal muscular atrophy, severe uncorrected scoliosis, and the lowest spirometric values reported in the literature without the use of ventilatory support. This patient demonstrates that women with severe kyphoscoliosis and a profound ventilatory limitation can carry a successful pregnancy well into the third trimester without requiring full ventilatory support.

**Database:** Medline

**26. Anaesthesia for caesarean section in a patient with spinal muscular atrophy.**

**Author(s):** Buettner, A U

**Source:** Anaesthesia and intensive care; Feb 2003; vol. 31 (no. 1); p. 92-94

**Publication Date:** Feb 2003

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 12635403

Available in full text at [Anaesthesia and Intensive Care](#) - from ProQuest

**Abstract:** This report describes a 33-year-old primigravid woman with spinal muscular atrophy Type III (Kugelberg-Welander syndrome). Elective caesarean section was performed at 38 weeks gestation under spinal anaesthesia. The implications of spinal muscular atrophy for anaesthesia for caesarean section are described.

**Database:** Medline

**27. December 2001: Rapidly progressive motor weakness, starting in pregnancy**

**Author(s):** Hilton D.A.; McLean B.

**Source:** Brain Pathology; 2002; vol. 12 (no. 2); p. 267-268

**Publication Date:** 2002

**Publication Type(s):** Article

**PubMedID:** 11958383

Available in full text at [Brain Pathology](#) - from John Wiley and Sons

**Database:** EMBASE

**28. Worsening of multifocal motor neuropathy during pregnancy.**

**Author(s):** Chaudhry, Vinay; Escolar, Diana M; Cornblath, David R

**Source:** Neurology; Jul 2002; vol. 59 (no. 1); p. 139-141

**Publication Date:** Jul 2002

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 12105326

Available in full text at [Neurology](#) - from Ovid

**Abstract:** Three women with multifocal motor neuropathy (MMN) were treated during pregnancy. Compared with their pregestation strength, the women became weaker in previously involved muscles and showed new weakness in previously unaffected muscles. All were treated with IV immunoglobulin during pregnancy and improved in strength. After pregnancy, strength in all patients returned to the prepregnancy state. The authors conclude that pregnancy may worsen MMN.

**Database:** Medline

**29. Stable motor and lung function throughout pregnancy in a patient with infantile spinal muscular atrophy type II.**

**Author(s):** Rudnik-Schöneborn, Sabine; Breuer, Christian; Zerres, Klaus

**Source:** Neuromuscular disorders : NMD; Feb 2002; vol. 12 (no. 2); p. 137-140

**Publication Date:** Feb 2002

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 11738355

**Abstract:**Patients with infantile spinal muscular atrophy rarely decide to have their own children especially if there is major respiratory impairment. We studied prospectively the pregnancy course and outcome of a 34-year-old woman with spinal muscular atrophy type II who delivered a healthy boy. Pregnancy was entirely uneventful, except that for 1-2 weeks after the caesarean section, the patient was extremely weak with dyspnoea and bulbar involvement. Several weeks after delivery her motor function had returned to pre-pregnancy levels. Pulmonary function remained stable throughout pregnancy, and thereafter, at approximately 70% predicted levels for forced vital capacity and for forced expiratory volume in 1 s. Blood gases after midgestation revealed low normal PaO<sub>2</sub> values around 85 mmHg and concomitant hyperventilation resulting in PaCO<sub>2</sub> levels below 30 mmHg.

**Database:** Medline

**30. Amyotrophic lateral sclerosis associated with pregnancy.**

**Author(s):** Tyagi, A; Sweeney, B J; Connolly, S

**Source:** Neurology India; Dec 2001; vol. 49 (no. 4); p. 413-414

**Publication Date:** Dec 2001

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 11799421

Available in full text at [Neurology India](#) - from Free Access Content

**Abstract:**Amyotrophic lateral sclerosis (ALS) is the most common, progressive motor neurone disease but is rare in the obstetric population. Only 4 cases have been described in the English literature since 1975. We describe a 29 year old woman who presented with ataxia, lower limb weakness and dysarthria 4 weeks after the birth of her first child. The symptoms had onset during the pregnancy but had not been considered remarkable. There were clinical features of upper and lower motor neurone involvement without any sensory loss. MRI of brain and spine was normal. CSF analysis was negative. EMG studies confirmed the presence of widespread anterior horn cell dysfunction compatible with ALS. The patient was commenced on Riluzole and has progressed clinically, at 12 months post diagnosis.

**Database:** Medline

**31. Successful pregnancy and spinal muscular atrophy.**

**Author(s):** Pugh, C P; Healey, S K; Crane, J M; Young, D

**Source:** Obstetrics and gynecology; Jun 2000; vol. 95 (no. 6); p. 1034

**Publication Date:** Jun 2000

**Publication Type(s):** Journal Article

**PubMedID:** 10808021

Available in print at [Patricia Bowen Library and Knowledge Service West Middlesex university Hospital](#) - from Obstetrics and Gynecology

Available in full text at [Obstetrics and Gynecology](#) - from Ovid

**Database:** Medline

**32. Amyotrophic lateral sclerosis presenting during pregnancy.**

**Author(s):** Jacka, M J; Sanderson, F

**Source:** Anesthesia and analgesia; Mar 1998; vol. 86 (no. 3); p. 542-543

**Publication Date:** Mar 1998

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 9495410

Available in full text at [Anesthesia and Analgesia](#) - from Ovid

**Database:** Medline

**33. Labor analgesia and anesthesia in a patient with spinal muscular atrophy and vocal cord paralysis.**

**Author(s):** Golden, S

**Source:** Regional anesthesia; 1997; vol. 22 (no. 6); p. 595-596

**Publication Date:** 1997

**Publication Type(s):** Letter Comment

**PubMedID:** 9425992

Available in full text at [Regional Anesthesia](#) - from Ovid

**Database:** Medline

### 34. Peripartum management of a patient with Isaacs' syndrome

**Author(s):** Morgan P.J.

**Source:** Canadian Journal of Anaesthesia; Nov 1997; vol. 44 (no. 11); p. 1174-1177

**Publication Date:** Nov 1997

**Publication Type(s):** Article

**PubMedID:** 9398957

Available in full text at [Canadian Journal of Anesthesia/Journal canadien d'anesthésie](#) - from Springer Link Journals

Available in full text at [Canadian Journal of Anesthesia](#) - from Free Access Content

Available in full text at [Canadian Journal of Anesthesia](#) - from ProQuest

**Abstract:** Purpose: To describe the peripartum management of a patient with Isaacs' syndrome with specific reference to the anaesthetic implications of the disease process. Associated medical problems included obesity, pregnancy induced hypertension and a difficult airway. Clinical features: This 30-yr-old gravida V para 0 woman presented to the anaesthesia consultation clinic at 37-wk gestation to discuss pain relief options for labour and delivery. She had a history of Isaacs' syndrome (a peripheral motor neuron disorder), congenital heart disease (ASD and VSD), treated Hashimotos thyroiditis, obesity and a family history of haemachromatosis. On the day of consultation, she was hypertensive and peripheral oedema was noted. Her urine showed trace protein. Four days later, she presented to the labour suite and her cervix was 9 cm dilated. An epidural anaesthetic was given without difficulty and she had an uneventful labour and delivery course. There were no subsequent neurological complications. Conclusion: Isaacs' syndrome is an extremely rare peripheral motor neuron disorder. This patient was successfully managed with epidural analgesia for labour and delivered a healthy child with no congenital anomalies.

**Database:** EMBASE

### 35. Labor analgesia and anesthesia in a patient with spinal muscular atrophy and vocal cord paralysis. A rare and unusual case report.

**Author(s):** Weston, L A; DiFazio, C A

**Source:** Regional anesthesia; 1996; vol. 21 (no. 4); p. 350-354

**Publication Date:** 1996

**Publication Type(s):** Case Reports Journal Article Review

**PubMedID:** 8837195

Available in full text at [Regional Anesthesia](#) - from Ovid

**Abstract:** BACKGROUND AND OBJECTIVES A case of labor analgesia and anesthesia in a 23-year-old woman with spinal muscular atrophy and vocal cord paralysis is reported. As spinal muscular atrophy is a progressive degenerative disorder of spinal anterior horn cells, with generalized neuromuscular weakness as a common sequela, the goal of anesthetic management is to provide satisfactory labor analgesia and anesthesia with minimal compromise of respiratory function. METHODS A lumbar epidural anesthetic technique was used to provide satisfactory labor analgesia and anesthesia for a low forceps delivery. RESULTS The anesthetic technique provided a safe delivery. As the patient was awake, she was able to assist with the expulsion phase of labor. The rare published reports of spinal muscular atrophy and obstetric management are reviewed, the known pertinent physiologic derangements of the syndrome in concert with pregnancy being detailed, along with any information provided regarding anesthetic techniques. CONCLUSIONS It is believed that labor analgesia and anesthesia can be provided adequately with lumbar epidural



techniques. An understanding of the physiology underlying spinal muscular atrophy is essential to safe anesthetic management of the laboring parturient.

**Database:** Medline

### **36. Amyotrophic lateral sclerosis and pregnancy.**

**Author(s):** Vincent, O; Rodríguez-Ithurralde, D

**Source:** Journal of the neurological sciences; May 1995; vol. 129

**Publication Date:** May 1995

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 7595617

**Abstract:**We describe the first case of pregnancy in a patient with amyotrophic lateral sclerosis (ALS) reported in Uruguay. The 27-year-old white woman who came to our clinic complaining of general weakness and gait symptoms was diagnosed as being pregnant and met the El Escorial criteria of probable ALS. The advised abortion caused her to abandon medical treatment, which was only resumed after a period of severe deterioration, at 32 weeks of gestation. After several days of serious vital risk, as evaluated by a multidisciplinary team, a normal male baby was born. Four months later, the patient had gradually improved and reached a stable condition, but presented with restrictive ventilatory distress.

**Database:** Medline

### **37. Successful pregnancies in the presence of spinal muscular atrophy: two case reports.**

**Author(s):** Carter, G T; Bonekat, H W; Milio, L

**Source:** Archives of physical medicine and rehabilitation; Feb 1994; vol. 75 (no. 2); p. 229-231

**Publication Date:** Feb 1994

**Publication Type(s):** Research Support, U.s. Gov't, Non-p.h.s. Case Reports Journal Article

**PubMedID:** 8311683

**Abstract:**We report two cases of successful pregnancy in women with chronic, infantile onset, or type II spinal muscular atrophy, both of whom delivered healthy, unaffected babies. The patients required concurrent management by a physiatrist, pulmonologist, and perinatologist throughout the pregnancy. Complications included recurrent urinary tract infections, dyspnea and worsening of pulmonary function, wheelchair seating and positioning problems, and musculoskeletal and low back pain. These problems resolved postpartum. One woman had vaginal delivery, the other had caesarean section, both of which were well-tolerated. Because of severe musculoskeletal deformity, pelvic assessment is necessary to determine the mode of delivery. The uterus has normal contractility and effective labor patterns can be established. Spinal/epidural anesthesia may be contraindicated because of spine deformity. The pregnancies had no deleterious effect on the progression of the disease in our patients, both of whom reported a positive experience with great personal fulfillment.

**Database:** Medline

### **38. Spinal muscular atrophy and pregnancy.**

**Author(s):** Wilson, R D; Williams, K P

**Source:** British journal of obstetrics and gynaecology; Jun 1992; vol. 99 (no. 6); p. 516-517

**Publication Date:** Jun 1992

**Publication Type(s):** Case Reports Journal Article

**PubMedID:** 1637771

**Database:** Medline

### **39. Pregnancy and spinal muscular atrophy.**

**Author(s):** Rudnik-Schöneborn, S; Zerres, K; Ignatius, J; Rietschel, M

**Source:** Journal of neurology; Jan 1992; vol. 239 (no. 1); p. 26-30

**Publication Date:** Jan 1992

**Publication Type(s):** Research Support, Non-u.s. Gov't Journal Article Review

**PubMedID:** 1541965

Available in full text at [Journal of Neurology](#) - from Springer Link Journals

**Abstract:**We investigated the course and outcome of pregnancy and its influence on muscle weakness in 12 females with proximal spinal muscular atrophy (SMA) who delivered a total of 17 infants when aged 18-32 years. In 4 females the SMA clearly followed an autosomal recessive mode of inheritance. The disease was autosomal dominantly inherited in 2 patients; the other 6 were sporadic cases. Ages of onset of SMA ranged from 8 months to 29 years; all the females learned to walk, and 10 out of 12 are still ambulatory aged 30-60 years. Pregnancy and delivery were complicated in 10 out of 12 patients by premature labour (4), prolonged labour (3) and delayed postpartum recovery (6). Caesarean section was performed in 3 cases. No deleterious effects on fetal outcome could be detected. Exacerbation of muscle weakness after the second trimester of pregnancy was experienced by 8 females: 5 noticed a persistent deterioration of SMA; in 3 muscle weakness worsened temporarily during pregnancy and was followed by marked improvement in the puerperium. The psychological perceptions, in retrospect, of 10 females concerning their decision to have children were evaluated.

**Database:** Medline

## Strategy 239204

#	Database	Search term	Results
1	Medline	("motor neuron disease" OR MND).ti,ab	4635
2	Medline	exp "MOTOR NEURON DISEASE"/	23082
3	Medline	(1 OR 2)	24980
4	Medline	(pregn*).ti	190993
5	Medline	exp PREGNANCY/	812139
6	Medline	(4 OR 5)	831166
7	Medline	(3 AND 6)	275
8	Medline	("amyotrophic lateral sclerosis").ti,ab	18131
9	Medline	(6 AND 8)	64
10	EMBASE	exp "MOTOR NEURON DISEASE"/	37411
11	EMBASE	("motor neuron disease" OR MND).ti,ab	6910
12	EMBASE	("amyotrophic lateral sclerosis").ti,ab	24506
13	EMBASE	(10 OR 11 OR 12)	41632
14	EMBASE	(pregn*).ti	238256
15	EMBASE	exp PREGNANCY/	670072
16	EMBASE	exp "PREGNANCY COMPLICATION"/	121609
17	EMBASE	(14 OR 15 OR 16)	745463
18	EMBASE	(13 AND 17)	141

19	EMBASE	18 [English language]	131
20	PubMed	("motor neuron disease" OR MND).ti,ab	7403
21	PubMed	("amyotrophic lateral sclerosis").ti,ab	21964
22	PubMed	(pregn*).ti,ab	914584
23	PubMed	(20 OR 21)	26656
24	PubMed	(22 AND 23)	155