

# Landry Guillain-Barré Strohl Syndrome in Twin Pregnancy: A Case Report

## İKİZ GEBELİKTE ORTAYA ÇIKAN LANDRY GUİLLAIN-BARRE STROHL SENDROMU: OLGU SUNUMU

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### Summary

**Introduction:** Landry Guillain-Barré Strohl Syndrome complicating pregnancy is a rare occurrence. In mild cases the course of the pregnancy is unaffected.

**Case report:** We report a case of a twin pregnancy with Landry Guillain-Barré Strohl Syndrome and rheumatoid arthritis. Her symptoms including weakness, vague numbness, unsteady gait and muscle wasting in extremities were initially mild and mimicked many of the changes seen in pregnancy. When symptoms become severe electromyography was done, showing a severe demyelinating motor-sensory neuropathy. She gave birth to premature twins at the 34th gestational week. The mother and the infants survived.

**Conclusion:** The management of the gravid patient with Landry Guillain-Barré Strohl Syndrome does not differ much from that of non-pregnant patients with the disease.

**Key Words:** Landry Guillain-Barré Strohl Syndrome, Pregnancy

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### Özet

**Giriş:** Gebelik ile Landry Guillain-Barre Strohl Sendromu birlikteliği oldukça nadir görülen bir durumdur. İlimli olgularda gebelik herhangi bir şekilde etkilenmez.

**Olgu sunumu:** Bu çalışmada Landry Guillain Barre Strohl sendromu ve romatoid artrit ile birlikte görülen ikiz gebelik olgusu sunulmuştur. Olguda güçsüzlük, geçici uyuşma, dengelessiz yürüme ve kas atrofisi gibi yakınmalar ortaya çıkmıştır. Hastanın yakınmaları şiddetlendiğinde yapılan elektromyografisinde motor ve duyuusal demyelizan nöropati tespit edilmiştir. Hastanın gebeliği 34. gestasyonel haftada prematür ikizlerin doğumu ile sonlanmıştır. Postpartum anne ve bebekte herhangi bir komplikasyon gelişmemiştir.

**Tartışma:** Landry Guillain Barre Strohl sendromu olan gebelerde yönetim gebe olmayan hastalardaki yönetimden farklı olmamaktadır.

**Anahtar Kelimeler:** Landry Guillain Barre Strohl sendromu, Gebelik

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Landry Guillain-Barré Strohl Syndrome, or acute inflammatory demyelinating polyradiculoneuropathy, is a rare disease that occurs with an incidence of approximately one to 1.5 cases per 100,000 population per year (1). This syndrome affects both sexes and occurs in people of all ages. It is a subacute demyelinating paralytic disease with an unestablished etiology. The exact incidence of Landry Guillain-Barré Strohl Syndrome complicating pregnancy is unknown, as is the effect of pregnancy on the course and severity of the syndrome. The diagnosis of Landry Guillain-Barré Strohl Syndrome is based on descriptive clinical, laboratory, and electrodiagnostic criteria. The clinical features required for diagnosis include progressive motor weakness of more than one limb associated with some degree of areflexia. Sensory and autonomic involvement is fairly common but is usually of a mild degree. Recovery usually begins 2-4 weeks after progression stops and is virtually complete in 80% of

patients by 6 months. Cerebrospinal fluid analysis showing albuminocytologic dissociation strongly supports the diagnosis but is absent in 20% of cases. Slowing or complete block of nerve conduction, if present also supports the diagnosis but is similarly absent in 20% of patients. Disorders that may have similar clinical presentations and therefore must be excluded include polyneuritis secondary to vitamin B<sub>12</sub> deficiency, abnormal porphyrin metabolism suggestive of acute intermittent porphyria, and heavy-metal intoxications including lead and arsenic poisoning. One must also exclude the possibility of a toxic neuritis secondary to agents such as nitrofurantoin or insecticides (2, 3).

### Case Report

A 27-year-old woman, gravida 4, para 0, at 15 weeks' estimated twin gestation, who presented with a 15-day history of paraesthesia in the hands and feet, and a 20-day

history of muscle weakness of upper and lower extremities. She was administered low-dose cortisone therapy because of rheumatoid arthritis of 12 years duration. On initial examination she had no acute distress. Neurologic examination revealed mild weakness of the upper and lower extremities and facial muscle weakness. Creatine phosphokinase, thyroid functions, vitamin B<sub>12</sub> and folate were all within normal limits. Obstetric ultrasound confirmed her dates and showed active fetuses with no gross anomalies. Over the next days, the patient developed progressive motor weakness of all extremities, but sensory manifestations of her disease remained relatively constant. She demonstrated muscle wasting of all extremities and difficulty in walking. Ten days after admission she complained of dysphagia and nausea and vomiting. Total parenteral nutrition and vitamin B<sub>12</sub> and folate supplementations were instituted. Electromyography nerve conduction velocity studies demonstrated slowed conduction consistent with demyelinating polyneuropathy.

At 34 weeks, the patient presented to the labor and delivery unit with 4 cm dilatation and 80% effacement of the cervix and a high blood pressure with associated proteinuria. A diagnosis of severe preeclampsia was made. A primary low transverse cesarean delivery was performed under epidural anaesthesia, producing a 1800 gm female infant and a 1550 gm male infant with Apgar scores of 9 and 10; 3 and 10 at 1 and 5 minutes, respectively. The patient's postpartum course was uncomplicated, and at the time of discharge she was ambulatory with excellent motor function in both the upper and lower extremities. The infants were discharged without any medical problem.

### Discussion

Landry Guillain-Barré Strohl Syndrome is relatively uncommon, especially during pregnancy. The exact aetiology of this subacutely evolving paralytic disease is not established. However, evidence is mounting to suggest that it represents an aberrant immune response. Approximately two-thirds of patients have a history of antecedent acute infectious illness with rubella (4) or cytomegalovirus (5). The most common acute illness is the glandular fever-like syndrome caused by cytomegalovirus; approximately half of these patients develop abnormal liver enzymes (2). The main complaint is of weakness that varies widely in severity in different patients, is more often more marked proximally than distally, and is often symmetric in distribution. It usually begins in the legs, frequently comes to involve the arms, and often affects one side or both sides of the face. Weakness may progress to total paralysis and may be life-threatening if the muscles of respiration or deglutition are involved. Sensory symptoms are common but are usually less conspicuous than motor symptoms. Autonomic dysfunction may be manifested by tachycardia, cardiac

irregularities, hypotension or hypertension, facial flushing, disturbances of sweating and disturbed pulmonary function (6,7).

Management of Landry Guillain-Barré Strohl Syndrome continues to be predominantly symptomatic. The major complications relate to respiratory failure, infection, and/or vascular collapse. In the nonpregnant state, the disease is associated with a 5% mortality rate; approximately 16% of patients require ventilatory support. Review of the obstetric literature suggests that pregnant women with this disease have a slightly increased mortality rate (13%), and approximately 35% require ventilatory support (5). Plasmapheresis is used successfully in some selected pregnant patients on the premise that the basic underlying pathophysiology of this demyelinating polyradiculoneuropathy involves either a myelinotoxic antibody or other myelinotoxic substance circulating in the plasma (8). Recently, some authors reported that successful treatment in pregnancy complicated by Landry Guillain-Barré Strohl Syndrome with high-dose intravenous immunoglobulin (9-11).

There is no convincing evidence that the Landry Guillain-Barré Strohl Syndrome occurs more commonly during gestation than at other times, and the course of the disorder does not seem to be influenced by pregnancy. Improvement in neurologic status may certainly occur before delivery and is not necessarily delayed until the baby is born. Approximately 3% of patients with the Landry Guillain-Barré Strohl Syndrome have one or more relapses, sometimes several years after the initial illness. Such relapses, which are clinically similar to the original illness, occasionally occur in relation to pregnancy (12).

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