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# Spinal anesthesia for cesarean section in a patient with Ehlers-Danlos syndrome, classical subtype: a case report

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## Abstract

Ehlers-Danlos syndrome includes thirteen subtypes, with different characteristics and clinical implications. There are no guidelines to help clinicians managing patients affected by this syndrome, but from 2014 recommendations exist. These patients have an increased anesthetic risk, both for general and regional technique. We describe the choice of anesthesia in a 34 years old pregnant woman, affected by Ehlers-Danlos syndrome classical subtype, who underwent cesarean section. Literature shows case reports about pregnancy and delivery in different subtypes, but not in classical one. We chose to perform a spinal block, with success.

**Keywords:** Ehlers-Danlos syndrome, Classical subtype, Pregnancy, Cesarean section, Spinal anesthesia.

## Introduction

Ehlers-Danlos syndrome (EDS) refers to a large group of connective tissue disorders, in which thirteen subtypes, with different causative gene mutations and phenotypes, are included [1]. Genetic mutations involve fibrillar collagen proteins or enzymes for their post-translational modification. Common clinical features are joint hypermobility, skin hyperextensibility and tissue (vascular and internal organ) fragility. Inheritance pattern can be autosomal dominant or recessive. Total prevalence of EDS is between 1 in 2500 and 1 in 5000 people, even though clinical findings suggest that they may be more common [2]. Because of the wide spectrum of clinical manifestations, there are no standardized obstetric and anesthetic guidelines, but, in 2014, Wiesmann et al. [3] published a review with important recommendations for anesthesia and perioperative management in EDS patients. Until that moment, literature consisted mostly of case reports and expert opinions [3]. We describe the anesthetic management of a pregnant woman, suffering from classical subtype EDS, who underwent cesarean section. Literature shows case reports about pregnancy and delivery in different subtypes, but not in classical one. It is of great importance to differentiate between subtypes, because each one has its own characteristics and clinical implications. Major criteria for diagnosis of classical subtype are: 1) skin hyperextensibility and atrophic scarring; 2) generalized joint hypermobility. Minor criteria are: easy bruising; doughy skin; skin fragility; molluscoid pseudotumors; subcutaneous spheroids; hernia; epicanthal folds; complications of joint hypermobility (e.g., luxation/subluxation, pain); family history of a first degree relative who meets clinical criteria [1]. The first major criterion is necessary, plus either the second one or at least three minor criteria. Final diagnosis needs genetic testing, even though its absence does not exclude the diagnosis [4]. More than 90% of classical type patients have a heterozygous mutation in genes encoding for type V collagen (COL5A1 and COL5A2); the inheritance pattern is autosomal dominant.

## Case Report

A 34-year-old pregnant woman, affected by classical EDS, nulliparous. Her past medical history revealed recurring joints dislocations and effusions. She suffered from chronic multifocal joint pain and there were some persistent atrophic scars on her body surface. Moreover, she had several comorbidities: hypertension; bronchial hyperreactivity; gastroesophageal reflux; obesity (BMI 36,9); migraine; urinary incontinence; polycystic ovary syndrome, with insulin resistance. Gestational diabetes complicated her pregnancy. An echocardiogram revealed normal cardiac function and a mild mitral valve regurgitation. She underwent local anesthesia, in the past, for dental surgery and the removal of a frontal angioma, without any problem; she had no other experiences with anesthesia. The airway Mallampati classification was 3. She had a history of several temporomandibular joint dislocations, with mouth opening limitation (3 cm); moreover, she was affected by tonsillar hypertrophy. The lumbar interspaces were not well defined. Preoperative laboratory investigations, including coagulation, were normal. There was a multidisciplinary discussion when patient came to our attention at the beginning of her unexpected pregnancy. Our gynecologists stated that a vaginal delivery was to prefer. In this case, we would have performed an epidural analgesia. In the case of cesarean section, we would have

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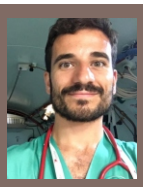

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performed a spinal block; if our spinal block had failed or if surgery had been prolonged, we would have converted from spinal to general anesthesia. At 33 weeks gestation, the patient was admitted for observation, because of uterine contractility. At 38 weeks, she had a cesarean section because of transverse fetal lie. She entered the operating room, where two peripheral venous lines (18 and 16 gauge) were established. A 1000-mL fluid coload of Ringer lactate solution was administered as prophylaxis against hypotension, while inducing a spinal block, in sitting position, with 0,5% levobupivacaine 10 mg, sufentanil 1 mcg and morphine 0,1 mg; a 25-gauge spinal needle was used. Then, we put patient in supine position, with left uterine displacement and 30° reverse-Trendelemburg. Oxygen (5 L/min) was administrated via face mask. A T4 sensory level was established. The patient gave birth to a male fetus (3630 g), with Apgar score of 8 at 5'. 5 IU of oxytocin as bolus, followed by 10 IU in 500 mL of 0.9% sodium chloride solution, were given intravenously to the mother after delivery. For post-operative pain, 1 g of paracetamol was infused at the end of surgery, repeated every 8 hours during recovery, in association with 600 mg of ibuprofen orally. Delivery and postoperative course were uneventful.

## Discussion

Management of these patients is controversial. Ehlers-Danlos syndrome is a systemic disease, with multi-organ involvement and an increased anesthetic risk both for general and regional technique. There is no clear recommendation for either general or regional anesthesia: the benefits of neuraxial block should be balanced against the increased risks in specific subtypes [3]. There is no agreement on optimal delivery method: uterine rupture, defective wound recovery and severe bleeding can complicate both vaginal and caesarean delivery [3]. Our patient underwent cesarean section because of transverse fetal lie; in this case, our plan was to perform a spinal block, and now we explain the reasons of our decision. The latest Obstetric Anesthesia Workforce Survey states that regional anesthesia continues to be used more frequently than general, for either elective or emergency cesarean section [5]. It is true that incidents related to general anesthesia decreased in latest years, thanks to the development and widespread use of difficult airway algorithms, but literature still reveals a higher mortality rate for general anesthesia delivery [6]. There can be also fetal effects for general anesthesia. These considerations add to the specific characteristics of EDS patients, in which there is an increased risk, during intubation, of gingival bleeding and oropharyngeal tissues damage due to their fragility. Repeated intubation attempts may cause bleeding, and this is especially true in such kind of patients; also cuff pressure should be kept as low as possible [3]. Furthermore, EDS patients can have an unstable cervical spine and occipital-atlanto-axial laxity [7], and some of them report muscular weakness after extubation [8]. Sood and colleagues [9], in 2009, reported a case of difficult intubation in a parturient with hypermobility type EDS; they hypothesized that the difficulty probably arose from collapse of fibro-elastic tissues and C-shaped cartilages of the trachea with the application of cricoid pressure. So, our patient met criteria for difficult airway, considering her Mallampati classification, her reduced mouth opening, the presence of tonsillar hypertrophy and the laxity of her temporomandibular joint; she had also a bronchial hyperreactivity, that could complicate the ventilation problems due to her disease (risk of pneumothorax). Furthermore, she was obese: obese parturients are at higher risk for difficult intubation and airway complications [10]; literature shows that the increased use of regional block in obese population has helped in reducing maternal morbidity and mortality, even though there can be more technical difficulties during the maneuver [10]. Neuraxial anesthesia is not immune to complications. In Ehlers-Danlos patients, there is a higher risk of spinal hematoma after needle placement, due to tissue and vessel fragility. Anyway, literature describes some case reports, even though not in classical subtype, in which neuraxial anesthesia was performed safely in EDS pregnant patients [11-13]. Fernàndez-Garcìa R. et al. [13] affirm that a subarachnoid block is an appropriate, safe alternative to general anesthesia for cesarean section in women with EDS, who have normal coagulation tests and no history of bleeding. However, there have been several reports of resistance to local anesthetics with Ehlers-Danlos syndrome [14, 15]; the underlying mechanism has not been explained. Actually, reports of block failure are often with respect to local anesthesia for dental surgery and peripheral nerve blocks; no reports for failure are published with regard to neuraxial blocks [8]. Wiesmann and colleagues [3] affirm that an ineffectiveness of local anesthetics or a block failure in the past of a patient should be noted; the same authors assert that neuraxial blocks in EDS subtypes other than vascular are feasible. Sood et al. [9] suggest that, in case of history of local anesthetics failure, the anesthetist should take seriously this information while deciding the best anesthetic technique for a EDS patient; if no history of local anesthesia exists, patients should be aware of its possible failure and need for general anesthesia should be discussed. Our patient reported a history of a frontal angioma removal in local anesthesia, with no problems. She had not well defined lumbar interspaces, but she had no coagulation impairment, no history of hemorrhage and was hemodynamically normal. For these reasons, according to the 2014 recommendations [3], we took a decision based on the individual risk, considering our patient characteristics: she had a possible airway difficulty, a relatively reduced risk for neuraxial anesthesia complications, so our first choice for cesarean section was, in agreement with our patient, spinal block. This was successful.

## Conclusion and clinical message

We want to underline the importance of individualizing anesthetic management, for a syndrome that can have different expressions and clinical implications. There were no reports about cesarean section in classical subtype EDS pregnant women. Case-reports are of great importance, considering the lack of a consistent evidence-based knowledge. In our experience, spinal anesthesia has been safe for cesarean section in a patient affected by Ehlers-Danlos syndrome, classical subtype.

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