International Journal of Obstetric Anesthesia (2009) **18**, 408–412
0959-289X/$ - see front matter
doi:10.1016/j.ijoa.2009.03.012

ELSEVIER
www.obstetanesthesia.com

CASE REPORT

Difficult intubation during rapid sequence induction in a parturient with Ehlers-Danlos syndrome, hypermobility type

V. Sood, D.A. Robinson, I. Suri
Warwick Hospital, South Warwickshire General Hospitals NHS Trust, Warwick, UK

ABSTRACT
There have been several reports of resistance to local anaesthetic agents in women with Ehlers-Danlos syndrome, hypermobility type, also known as Ehlers-Danlos syndrome Type III. General anaesthesia with rapid sequence induction was performed for caesarean section due to prolonged second stage of labour, but intubation proved to be difficult. We propose that intubation difficulty probably arose from collapse of fibro-elastic tissues and adjoining C-shaped cartilages of the trachea with appropriately applied cricoid pressure. We found no other case reports of difficult intubation in patients with Ehlers-Danlos syndrome, hypermobility type. There are reports of cervical spine instability and temporomandibular joint dysfunction in patients with this syndrome suggesting a potential for difficult airway management. Additional anaesthetic problems associated with Ehlers-Danlos syndrome involve patient positioning and vascular access.

Keywords: Intubation; Caesarean section; Hypermobility; Ehlers-Danlos syndrome

Introduction

Ehlers-Danlos syndrome (EDS) is a heterogeneous group of inherited disorders of connective tissue. Ehlers-Danlos syndrome, hypermobility type (EDS-HT) is one of six types of EDS in a classification system proposed by Beighton et al.[1] in 1997, which is based on clinical, biochemical and molecular findings (Table 1). Prior to this system, EDS was classified into 11 subtypes and the EDS-HT classified as EDS type III.[2] Clinically, EDS-HT is currently divided into *major* features (skin laxity, velvet skin, generalised joint hypermobility) and *minor* features (recurrent joint dislocations and chronic limb or joint pain); it is considered one of the milder types of EDS.[1] To complicate matters further, some authors consider EDS-HT and a condition called benign joint hypermobility syndrome (BJHS) to be identical or spectrums of the same disorder because of overlapping phenotypical features.[3–8] BJHS was first defined by Kirk et al.[9] in 1967 as the occurrence of musculoskeletal symptoms in hypermobile but otherwise healthy persons. More recently it has been described as a connective tissue disorder with hypermobility in which musculoskeletal symptoms occur in the absence of systemic rheumatologic disease.[10] For the purpose of clarity, we will refer to the condition as EDS-HT, but where the literature refers to BJHS, this is made clear in the text. We describe a parturient with EDS-HT in whom we encountered a difficult intubation.

Accepted March 2009
Correspondence to: Ishwinder Suri, Consultant Anaesthetist, Anaesthetics Department, Warwick Hospital, South Warwickshire General Hospitals NHS Trust, Lakin Road, Warwick, CV34 5BW. Tel.: 01926495321.
E-mail address: ishwinder.suri@swh.nhs.uk

Case report

A 31-year-old, G1P0, 154 cm, 53 kg, woman at 32 weeks of gestation with EDS-HT was referred by her obstetrician to our obstetric anaesthetic assessment clinic (OAAC) for discussion of possible anaesthetic interventions. She gave a history of childhood asthma, which had completely resolved, needle phobia and episodes of local anaesthetic failure. Local anaesthesia had failed during dental treatment and in a chronic pain clinic; at the pain clinic she had received subcutaneous infiltration of lidocaine and facet-joint injections with a bupivacaine and steroid mixture, both of which failed to produce analgesia.

At the OAAC following a discussion of efficacy, risks and benefits of various methods of analgesia, the patient agreed to the initial use of a transcutaneous electrical nerve stimulator (TENS) during labour. A remifentanil infusion was also discussed, but depended on the presence of an anaesthetist experienced in its use. Epidural

Table 1 Villefranche classification of Ehlers-Danlos syndrome

New name	Old name	Inheritance	Mutated genes
Classic type	Gravis: type I; Mitis: type II	AD	COL5A1, COL5A2
Hypermobility type	Hypermobile: type III	AD	Unknown; ?TNXB?
Vascular type	Arterial-ecchymotic: type IV	AD	COL3A1
Kyphoscoliosis type	Ocular-scoliotic: type IV	AR	Lysyl-hydroxylase
Arthrochalasia type	Arthrochalasis multiplex congenital: type VIIA and type VIIB	AD	COL1A1, COL1A2
Dermatosparaxis type	Dermatosparaxis: type VIIC	AR	Procollagen, N-peptidase

AD: autosomal dominant; AR: autosomal recessive.

analgesia was suggested as a possibility, but the patient was advised that the local anaesthetic might not be effective and additional opioid analgesia could be provided. In the event of caesarean section, she agreed to general anaesthesia. Airway evaluation revealed a Mallampati class 2 with no features to suggest a difficult airway.

After an uneventful pregnancy, the patient arrived on the labour ward at 38^{+6} weeks of gestation, with spontaneous rupture of membranes and regular uterine contractions. Initially she used a TENS device, inhaled Entonox and i.v. paracetamol 1 g for analgesia. As labour progressed, she became increasingly uncomfortable, but refused i.m. pethidine because of her needle phobia, and epidural analgesia since it might not work. The patient continued inhaling Entonox until rectal pressure was felt and examination revealed a cervical dilatation of 10 cm. The patient pushed for one hour with minimal head descent, and ventouse delivery failed after three attempts. Despite her past history, the obstetrician infiltrated 1% lidocaine in hopes of doing an episiotomy but it failed and the procedure was not performed. A decision was made to proceed to caesarean section under general anaesthesia.

In theatre, to minimise aortocaval compression, a wedge was inserted under the right hip. After 3 min of preoxygenation, cricoid pressure was applied and rapid sequence induction performed with i.v. thiopental 300 mg and suxamethonium 100 mg. Laryngoscopy revealed a grade-2 view with a pendulous epiglottis; a bougie was passed between the vocal cords but appeared to meet an obstruction beyond the vocal cords and would not advance further. After a second attempt with the bougie followed by an 8-mm endotracheal tube, both met with obstruction beyond the cords. No further attempts were made because the oxygen saturation decreased to 80%. Mask ventilation with 100% oxygen while maintaining cricoid pressure increased the saturation to 99%. Cricoid pressure was released, the laryngoscopic view remained at grade 2, and a 7.5-mm endotracheal tube was easily passed beyond the vocal cords. Anaesthesia and surgery were uneventful and a live male baby was delivered with Apgar scores of 5 and 8 at 1 and 5 min, respectively.

Discussion

Obtaining specific information regarding the anaesthetic implications of EDS-HT and BJHS in the parturient was difficult for several reasons. Firstly, there are numerous names for EDS-HT and BJHS in the literature. EDS-HT is also known as EDS-familial hypermobility type and BJHS has been described as joint hypermobility syndrome, hypermobile joint syndrome, benign hypermobile joint syndrome, hypermobility syndrome and familial articular hypermobility syndrome.[4,11] In addition, diagnostic criteria have evolved over time, but consensus does not exist on their use.[5] Finally, most literature reports generalise the anaesthetic implications for all types of EDS despite there being a well-documented spectrum of clinical features and severity for the different types.

In an attempt to broaden our search, we conducted a MEDLINE® literature search of the numerous terms for EDS-HT, BJHS alone and in conjunction with the following search terms: anaesthesia, intubation, cricoid pressure, airway collapse, cervical spine, temporomandibular joint, parturient, pregnancy, labour and caesarean section or delivery. Where relevant case reports were obtained, we further researched the references provided.

The overall prevalence of EDS is estimated to be 1 in 5000.[1] The prevalence of EDS-HT is 1:10 000-15 000; it is one of the more common types.[1] The inheritance pattern for EDS-HT is autosomal dominant, whereas for BJHS the inheritance is described as being a gender-influenced autosomal dominant trait, since it is more common in females.[1,4,11] BJHS is also more common in Asians and Africans than Caucasians.[4,11] The basic defect of EDS-HT is unknown but 5-10% of individuals with the disorder have a mutation in one copy of the TNXB gene, which encodes for the tenascin-X protein in collagen; tenascin-X provides structure, strength and flexibility to connective tissue.[12] BJHS appears to be due to an abnormality in collagen or the ratio of collagen subtypes; mutations in the fibrillin gene have also been identified.[13]

The diagnosis of both EDS-HT and BJHS is based on clinical features. EDS-HT is divided into *major features:* (skin laxity, velvet skin, generalised joint hypermobility) and *minor features:* (recurrent joint dislocations and