Successful vaginal birth after caesarean section in patient with Ehler-Danlos syndrome type 2

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Summary: We present the case of a 31-year-old woman with Ehler-Danlos syndrome (EDS) type 2. She had a previous caesarean section and went on to have an uncomplicated vaginal birth in her last pregnancy. To our knowledge, this is the first case of a successful vaginal birth after caesarean section in a patient with EDS. EDS is a multisystem disorder involving a genetic defect in collagen and connective-tissue synthesis and structure. It is a heterogeneous group of 11 different inherited disorders. Obstetric complications in these patients include miscarriages, stillbirths, premature rupture of the membranes, preterm labour, uterine prolapse, uterine rupture and severe postpartum haemorrhage. There has been much controversy over the appropriate mode of delivery. Abdominal deliveries are complicated by delayed wound healing and increased perioperative blood loss. Vaginal deliveries may be complicated by tissue friability causing extensive perineal tears, pelvic floor and bladder lesions. Our case highlights that in specific, controlled situations it is possible to have a vaginal delivery even after previous caesarean section in patients with EDS.

Keywords: high-risk pregnancy, rheumatology, clinical genetics, complications

CASE REPORT

A 31-year-old woman booked at 14 weeks gestation in her seventh pregnancy. She had a past medical history of Ehler-Danlos syndrome (EDS) type 2, diagnosed at age 9 with characteristic features of easy bruising, scarring, loose-jointedness and dislocations. She had a left shoulder dislocation at age 19 and two left hip dislocations by the age of 24. Interestingly, she also suffered recurrent spontaneous haemorrhages that included two severe mesenteric bleeds requiring blood transfusions and a laparotomy in one instance. Since these are atypical of EDS type 2 and recognized features of EDS type 4, a skin biopsy for culture and biochemical studies was performed. The results yielded a normal type 3 collagen level in cultured skin fibroblasts, thus making EDS type 4 an unlikely diagnosis.

The patient had two first trimester miscarriages at age 16 and 17. At 22, her third pregnancy proceeded uneventfully to term. She laboured spontaneously and delivered through a large central perineal tear between the fourchette and anus.¹ This was repaired in layers. The posterior vaginal wall was repaired in one continuous layer. The perineal muscles were repaired with interrupted sutures. The perineal skin was repaired with interrupted sutures to allow for drainage of any postoperative haematoma. Number 1 vicryl was used to allow for delayed wound healing. Healing was good and she was discharged home well on day 5 postrepair. In her next pregnancy, she had a normal term spontaneous vaginal delivery with an elective episiotomy. Her fifth pregnancy was a

missed miscarriage and an evacuation of retained products of conception and elective laparoscopic sterilization was performed. Three years later, she met a new partner and moved from the region. She presented to another hospital requesting reversal of sterilization which was performed. She then conceived and the baby delivered via a term elective lower segment caesarean section.

She booked with our unit in her seventh pregnancy. Because of her significant past medical and obstetric history, we were faced with a dilemma regarding the mode of delivery. The antenatal period was uneventful until 37 weeks when a biometric ultrasound scan showed the abdominal circumference to be on the third centile. There was normal liquor volume and end diastolic flow. Unstable lie was also diagnosed at this time. Successful external cephalic version for breech presentation was performed at 39 weeks. She presented at 40 weeks and two days in spontaneous labour. The first stage lasted three hours and 50 minutes and the second stage four minutes. She had a normal vaginal delivery of a live 2.28 kg male infant. She sustained a small vulval haematoma which was drained. She also sustained a second-degree perineal tear which was repaired. Apart from this, she had a normal postnatal period and was discharged well. To our knowledge, this is the first reported case of successful vaginal birth after caesarean section in a patient with EDS type 2.

DISCUSSION

EDS is a heterogeneous group of 11 different inherited disorders, all involving a genetic defect in collagen and connective-tissue synthesis and structure. All three major inheritance patterns (autosomal dominant, autosomal recessive

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and X-linked) have been identified. The underlying collagen abnormality is different for each type and has been identified in six of the 11 types.

EDS is a multisystem disorder. Clinical recognition of the type of EDS is important. The classical variants (types 1 and 2) present with cutaneous fragility, poor wound healing, joint hypermobility and easy dislocations. Of particular importance is identifying the vascular form (type 4) which accounts for approximately 6% of cases, but this is the only form associated with a high risk of early death (median life expectancy of 50 years) due to spontaneous arterial, intestinal and uterine ruptures.²

Our patient was diagnosed clinically with type 2 disease. It is possible she may actually have type 4 EDS. Her skin biopsy for culture and biochemical studies yielded a normal type 3 collagen level in cultured skin fibroblasts. This does not exclude a diagnosis of type 4 as skin biopsies can be normal in some cases. We suggest further studies to confidently exclude the diagnosis of type 4 EDS. To our knowledge, this patient has not had genomic sequences studies to date.

Patients with EDS who are considering conceiving or who have become pregnant need genetic counselling and must be made aware of the risks of pregnancy. These patients are at increased risk for various obstetric complications, which include miscarriages, stillbirths, premature rupture of the membranes, preterm labour, uterine prolapse, uterine rupture and severe postpartum haemorrhage.^{3,4} This increased miscarriage rate and preterm deliveries have been attributed to cervical incompetence.⁴ However, a general recommendation regarding the use of a prophylactic cervical cerclage to treat cervical incompetence in women with EDS cannot be made. The maternal mortality in type 4 EDS is quoted as high as 25%.⁵

There has been much controversy over the appropriate mode of delivery for these patients. In the 1960s primary caesarean section was indicated for most cases of EDS.⁶ This view has changed considerably with some authors recommending caesarean delivery only in patients with EDS type 4 because of the significant risk of spontaneous uterine rupture. In most other cases, the mode of delivery will vary depending on individual circumstances. Abdominal deliveries are complicated by delayed wound healing and increased perioperative blood loss. The use of non-absorbable sutures has been advocated and tranexamic acid has been used postoperatively to reduce episodes of re-bleeding. There are reports of correction of the bleeding time or improvement in symptoms in patients treated with desmopressin.⁷ There is also a case report of the use of prophylactic desmopressin in labour to prevent haemorrhage in EDS.⁸

Vaginal deliveries may be complicated by tissue friability causing extensive perineal tears, pelvic floor and bladder lesions.⁵ An elective episiotomy was used during our patient's

third labour. This was performed with a view to prevent a recurrence of her previous obstetric trauma (perineal tear). There is no evidence for its use for this indication in a patient with EDS. Any theoretical benefit needs to be balanced against known risks of increased bleeding and delayed healing of episiotomies in patients with EDS. Our case highlights that in specific, controlled situations it is possible to have a vaginal delivery even after previous caesarean section in patients with EDS.

There are few reports on fetal outcome. In our case, there was evidence of intrauterine growth restriction at 37 weeks. Growth restriction, particularly in late pregnancy, appears to be a recognized complication.⁹ Hernia and luxation may occur, and the floppy infant syndrome is observed in about 13% of patients.¹⁰

Owing to the multiorgan involvement and varied presentation of this disease, no uniform obstetric recommendations can be made. A multidisciplinary approach is required seeking the assistance of geneticists, anaesthetists and paediatricians. Guidance and management must be tailored individually and according to the clinical subtype.

DECLARATIONS

The authors have no conflicts of interest to declare.

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