

The application of fibrin sealant in a case of Ehlers–Danlos type IV

Rishi Shonpal · A. N. Griffiths

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Introduction

This case demonstrates how to control haemorrhage in patients with Ehlers–Danlos syndrome with the use of a fibrin sealant.

Case report

A 24-year-old woman, gravida 3 para 0, was tertiary referred to the antenatal clinic at 17 weeks gestation with Ehlers–Danlos type IV. In addition, she was known to have a degree of pulmonary stenosis, mild aortic regurgitation, repaired congenital pyloric stenosis, previous pulmonary valvotomy, previous carotid cavernous fistula embolisation, clipped left internal carotid artery aneurysm, false femoral artery aneurysm after invasive radiology procedure and extensive keloid scarring

She had previously been advised pre-conceptually of the substantial risks of mortality. The patient's mother, who

was also known to have Ehler–Danlos Type IV, had died of an aortic aneurysm.

Multidisciplinary antenatal care was instigated including transthoracic echocardiography which suggested satisfactory pressure gradients and a low risk of developing cardiac failure.

From 34 weeks gestation, she was admitted with raised BP. She was treated with labetalol (Trandate®; Allen & Hanbury's, NC, USA). Her biochemistry tests were normal. After an extensively literature review, a plan was made for an elective caesarean section at 38 weeks gestation. The spinal anaesthetic was commenced with meticulous control of BP.

At 37+6 weeks, she was admitted with a sudden onset of abdominal pain and a blood pressure of 180/108. A caesarean section was performed according to NICE guidelines, and a transverse incision was made in the uterine lower segment which was formed but not thin. The uterus was of normal appearance with no abnormalities evident.

The baby was in a cephalic presentation and was delivered with ease with minimal fundal pressure. The placenta was removed manually; the liquor was clear and of normal volume.

It was noted that there was generalised ooze from the lower segment; yet, suturing proved difficult with friable uterine tissue. Due to the high-risk nature of the case and in order to minimise the risk of bleeding, it was decided to use a haemostatic aid on the uterine incision.

The surgeon used fibrin sealant (QUIXIL®; Omrix Biopharmaceuticals SA, Brussels, Belgium) with a spray technique over the uterine wound after suturing the incisions in order to achieve secure haemostasis. There was no further bleeding from the uterus, and post-operative complications were uneventful.

R. Shonpal (✉)
18 Green Lane,
Purley, Surrey CR8 3PG, UK
e-mail: rishi108@gmail.com

A. N. Griffiths
University Hospital of Wales,
Heath Park,
Cardiff CF14 4XN, UK

Discussion

Women with Ehlers–Danlos syndrome are at increased risk of complications during pregnancy and delivery, particularly the vascular type, formerly type IV, which is one of the most dangerous clinical subtypes [1]. Complications in childhood are rare, but around 80% of patients experience at least one complication by 40 years of age [2]. Maternal mortality rates have been reported to be between 10% [3] and 25% [4] which are highest during labour, intra and immediate post-partum.

Gynaecology complications can be divided into pre-pregnancy which include: anovulation, sexual dysfunction, recurrent vaginal infections, irregular menses, abnormal cytologic smears and endometriosis [5]. Pregnancy complications include spontaneous rupture of great vessels, perforation of bowel, vascular and uterine malformations and spontaneous rupture of the foetal membrane leading to premature delivery. This necessitates careful counselling before pregnancy and regular antenatal screening tests and BP monitoring. Intra-partum complications include damage to the vagina and perineum, and postpartum complications include frequent reports of severe haemorrhage which are sometimes only resolved by hysterectomy [6].

A fibrin sealant (QUIXIL®; Omrix Biopharmaceuticals SA, Brussels, Belgium) is anecdotally a well-known haemostatic derived from human blood plasma. It takes 10 min to prepare due to its frozen storage and has been successfully used in the past for other surgical specialties [7] including its use in general [8], reconstructive [9] and endonasal surgery. In one study, postoperative haemorrhage occurred in 22.9–25% of 494 patients with nasal packing vs. 3.12–4.65% in the fibrin sealant groups [10]. However, its use in gynaecology surgery, particularly in patients with Ehlers–Danlos syndrome, has not been evaluated. In this case, the use of Quixil® was successful in providing a quick and effective intra-operative haemostasis, and its efficacy should be more formally evaluated.

Conclusion

In females with a high risk of bleeding, it is important to consider all haemostatic procedures.

Conflict of interest There is no conflict of interest. There have been no financial interest/arrangements with one or more organisations that could be perceived as a real or apparent conflict of interest in the context of the subject of this article.

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