LETTER TO THE EDITOR

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FREE TISSUE TRANSFER IN EHLERS–DANLOS SYNDROME

Dear Editor,

Ehlers–Danlos syndrome (EDS) is a spectrum of heritable connective tissue disorders leading to abnormal collagen synthesis, and a triad of skin extensibility, joint hypermobility and tissue fragility.[1] Historically, elective surgery has been avoided in EDS due to wound healing and scarring concerns.[2] We have shown successful free tissue transfer in a patient with EDS.

A 44-year-old woman presented with classical EDS and BRCA II gene mutation, requesting bilateral risk reducing mastectomies and immediate autologous reconstruction. Bilateral DIEP flaps were raised in standard fashion. Arterial anastomoses were performed to the internal mammary arteries at the third intercostal space. Venous coupler anastomoses were performed to the internal mammary veins. The vessel walls were extremely fragile, but all anastomoses were uneventful and flowed well immediately. The abdomen was closed in three layers.

The patient was discharged on postoperative day 7. At 2 weeks postoperatively, her wounds had healed well with no dehiscence. A good result was achieved, both in terms of shape and symmetry. A small periumbilical hernia was noted, which was subsequently repaired in conjunction with excision of the monitoring skin paddles.

We have shown successful autologous breast reconstruction in a patient with classical EDS. Whilst the literature suggests a higher rate of wound complications,[3] our patient had no issues with delayed wound healing and, at 6 months, had no evidence of atrophic scars.

Due to joint hypermobility and skin fragility, careful intraoperative positioning and attention to pressure areas is vital.[3] Intraoperative bleeding is also more likely.[4] The literature suggests nonabsorbable sutures should be left for twice the standard duration.[3] Our surgical practice did not deviate from that of a routine DIEP flap, with meticulous haemostasis and a tension-free multilayered wound closure. Care was taken when performing anastomoses to handle vessels delicately, and to avoid multiple passes of the needle and intimal damage.

Our patient did develop a postoperative hernia. This is a recognized complication of DIEP reconstruction,[5] but may suggest that patients with EDS benefit from routine mesh reinforcement of the rectus sheath, even when the defect is minimal as in our case.

We have shown that classical EDS is not a contraindication to microsurgical breast reconstruction. Due to the heterogeneity of the syndrome, the applicability of this case to other subtypes remains unclear. Patients should be assessed using a case-by-case approach to ensure optimal outcomes.

HENRIETTA CREASY, MBCh.B., B.Sc.,*
CHARLES BAIN, MBCh.B., B.Sc., M.Sc., and
JIAN FARHADI, M.D., P.D., FMH (PLAST) EBOPRAS
Department of Plastic and Reconstructive Surgery,
Guy's and St Thomas' NHS Foundation Trust,
London, UK

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*Correspondence to: Henrietta Creasy, MBCh.B., B.Sc., Flat 54, St George's Wharf, 6 Shad Thames, London SE1 2YS, UK. E-mail: henriettacreasy@gmail.com

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