

### Learning points

- Ehlers-Danlos syndrome type IV is caused by deficiency of type III collagen.
- spontaneous arterial and bowel rupture in a young individual should prompt urgent investigations and genetic assessment of the patient and close family members.
- trauma, pregnancy, invasive procedures and elective surgery are associated with very high morbidity and mortality.
- prenatal diagnosis and prevention of the disease is now possible

an experienced surgeon who is aware of the complications of the condition. This applies especially to varicose vein ED Grech surgery<sup>7</sup> which is frequently carried out in these patients. One of the authors has personal experience of a patient with Ehlers-Danlos syndrome type IV who eventually required above knee amputation following an elective varicose vein surgery.

### PROGNOSIS

With autosomal dominant inheritance there is a 50% risk of transmission to the offspring of

affected individuals. Genetic counselling and examination of other family members is therefore important. Females also have a high mortality risk, up to 25% during pregnancy.<sup>8</sup> Affected individuals are recommended to modify their life style to avoid physically strenuous jobs and sports which involve body contact to lessen the chances of traumatic vascular and bowel catastrophes.

In conclusion we emphasize that spontaneous rupture of the infrarenal abdominal aorta in a young healthy, adult male should prompt urgent and thorough clinical survey, laboratory investigations and genetic counselling of the patient and close relatives to rule out Ehlers-Danlos syndrome type IV. This rare disorder should now be considered in the differential diagnosis of the haemoperitoneum in a young individual with or without previous history of connective tissue disorder.

### Final diagnosis

Spontaneous rupture of an infrarenal abdominal aorta in Ehlers-Danlos syndrome type IV

**Keywords:** Ehlers-Danlos syndrome type IV, infrarenal aorta

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## THE PATHOLOGY MUSEUMS GROUP

Presents a special one-day event on Friday 31 May 1996 hosted by  
The Royal College of Surgeons

The morning session will consist of a series of 30 minute lectures on the following topics:

- the effects of boxing on the brain. New research has shown advanced brain-damage in a professional boxer aged 23
- the 'Piltown Man'. A fraudulent discovery of the 'missing link' which lasted 40 years
- forensic odontology – both victims and criminals may be identified by their teeth – even if they are false!
- an overview of pathology museums – their use for teaching and some of the unique specimens they contain

The afternoon session will be dedicated to the splendid collections housed at the Royal College of Surgeons which contain a wealth of material. The famous Hunterian Museum contains an unusual mixture of both human and animal anatomy and pathology dating back to the 18th century. Human anatomy is also represented in a separate dedicated museum as is pathology and dentistry. The College also features a fine collection of surgical instruments

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