acute onset following exercise, the tight puckered skin of the limbs and trunk sparing the face and joint contractures are typical features. Laboratory findings have shown a raised ESR, transient eosinophilia and hypergammaglobulinaemia. The striking feature of all previously reported cases has been the marked thickening of the deep fascia between the fat and muscle with perivascular lymphohistiocytic infiltration in the superficial muscle layer and the skin. Scattered eosinophils in the dermis and muscle have been seen by some observers (Rodnan *et al.* 1975, Caperton & Hathaway 1975). A full thickness biopsy was not carried out in this patient on account of the severe thrombocytopenia.

The patients previously described have improved remarkably on steroid treatment. The histology available in this patient together with lack of elevation of muscle enzymes and normal EMG findings would make a diagnosis of scleroderma or polymyositis appear unlikely and spontaneous remission in scleroderma is rare.

Thrombocytopenia has been described in scleroderma (Carcassonne & Gastaut 1976), but haematological abnormalities other than those described have not been a feature of this disease and it would appear that this patient had eosinophilic fasciitis with a coincidental megakaryocyte aplasia.

Addendum

Since presentation of this case, a report of a patient with diffuse fasciitis and aplastic anaemia has been published (Hoffman *et al.* 1979).

References

Bennett R M, Herron A & Keogh L (1977) *Annals of the Rheumatic Diseases* **36**, 345
Caperton E M & Hathaway D E (1975) *Arthritis and Rheumatism* **18**, 391
Carcassonne Y & Gastaut J A (1976) *British Medical Journal* i, 1446
Hoffman R, Dainiak N, Sibrack L, Pober J & Waldron J (1979) *New England Journal of Medicine* **300**, 718–721.
Rodnan G P, Di Bartolomeo A G, Medsger T A & Barnes E L (1975) *Arthritis and Rheumatism* **18**, 422
Shulman L E (1974) *Journal of Rheumatology* **1**, Suppl; p 46

Ehlers-Danlos syndrome with surgical repair of eventration of diaphragm and torsion of stomach[1]

J G Phadke MD
(for **V W Johnson** MB MRCP and **H B Young** FRCS)
Departments of Medicine and Surgery
New Cross Hospital, Wolverhampton WV10 0QP

A 63-year-old female, who presented in 1970 with vomiting, was found to have type I Ehlers-Danlos syndrome which is the classic severe type inherited as an autosomal dominant trait. When she was first reported to the Royal Society of Medicine in 1974 (Linnemann 1975), there was a history of previous poor wound healing and life-long kyphoscoliosis (Figure 1) with the development of premature osteoarthrosis. She had skin hyperextensibility, calcified subcutaneous spheroids, kyphoscoliosis (with later development of C4–5 subluxation), genu valgum, eventration of the left diaphragm (Figures 1 & 2) and organo-axial torsion of the stomach (Figure 3).

Despite radiologically demonstrable persistence of the gastric torsion, she recovered from a total of nine episodes of vomiting treated conservatively over a seven-year period, but in 1977, when aged 71 years, operative intervention became imperative. In view of the tissue

[1] Case presented to Clinical Section, 12 May 1978. Accepted 27 June 1979
Requests for reprints to Dr V W Johnson, New Cross Hospital, Wolverhampton WV10 0QP

0141–0768/79/100781–03/$01.00/0 © 1979 The Royal Society of Medicine

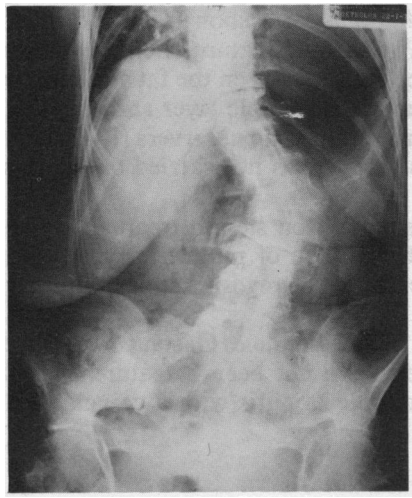

Figure 1. Kyphoscoliosis and diaphragmatic eventration

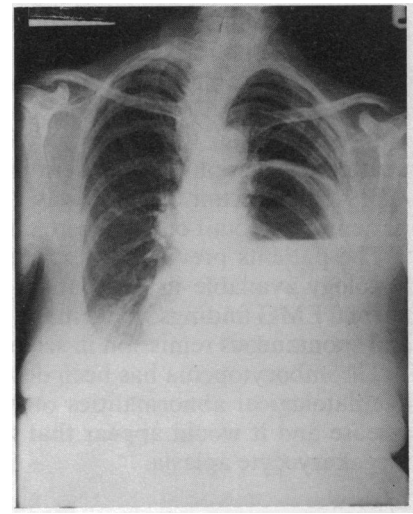

Figure 2. Eventration of the left diaphragm

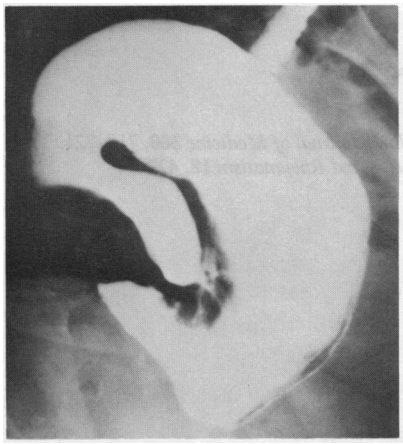

Figure 3. Organo-axial gastric torsion

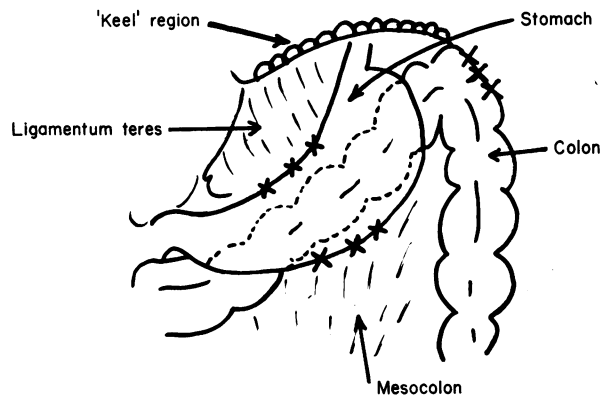

Figure 4. Repair of gastric torsion

fragility it was decided to use a combination of Tanner's methods of 'keeling' of the diaphragm followed by colonic displacement into the subphrenic space and gastropexy (Tanner 1968). The procedure was restricted to the transabdominal route because of her poor respiratory reserve and the operative risks.

At operation the ribs and lung markings were clearly visible through the tissue-paper-thin diaphragm and the stomach was mobilized easily due to its extensibility and the paucity of adhesions. The diaphragm was retracted into two subdiaphragmatic keel-like folds in a radial direction which were then sutured together with nylon resulting in correction of the eventration. The subphrenic space was filled with colon, which was sutured to the diaphragm, the greater curvature of the stomach was sutured to the mesocolon and the lesser curve to the ligamentum teres (Figure 4), and the abdomen was closed with reinforcing sutures. Postoperatively her asthenia and poor diaphragmatic movements necessitated assisted ventilation for two days. There was no dehiscence and although the wound took nine weeks to heal, partly because of infection from previous intertrigo, there has been no subsequent

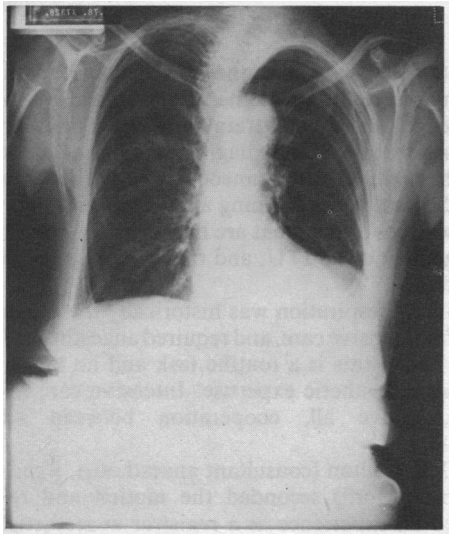

Figure 5. Chest X-ray a year postoperatively shows diaphragm in the corrected position

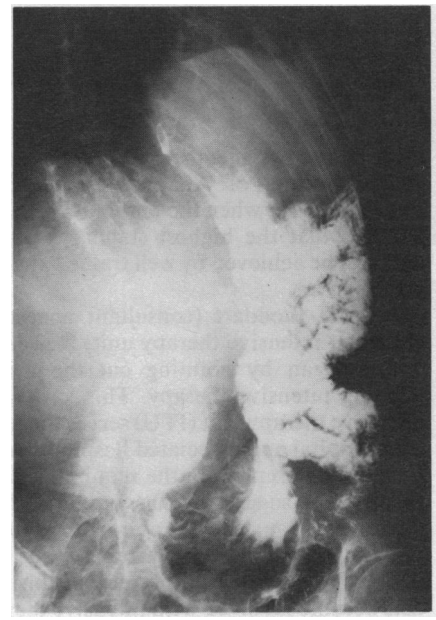

Figure 6. Barium meal a year postoperatively shows elongated stomach in a normal position

herniation. During the year since the operation she has remained well except for relatively minor chest infections controlled by antibiotics. A year postoperatively X-rays show that the diaphragm has remained in the corrected position (Figure 5) and the stomach is in its normal position but is elongated (Figure 6).

Discussion

Factors of importance in the production of gastric torsion are eventration of the diaphragm producing a potential increase in the subphrenic space, negative intrathoracic pressure, kyphoscoliosis, ligamentous laxity, paradoxical diaphragmatic movement and intra-abdominal adhesions in the axis of rotation, but the last two factors were not demonstrated in our patient.

In 1968 Tanner reviewed 21 of his patients who had been treated for gastric volvulus with eventration of the diaphragm. Gastropexy with colonic displacement was performed in 15 cases and partial gastrectomy in 4. In one patient he achieved transthoracic repair of the eventration by inverting folds of the diaphragm into the abdomen and suturing the edges together to produce keel-like projections in both circumferential and radial directions into the peritoneal cavity. Satisfactory progress was demonstrated in 14 patients (age range 35–74 years) who were followed up 2–12 years after colonic displacement.

Our patient is the first in whom gastric torsion has been described in the Ehlers-Danlos syndrome and this is the first report of the use of this combination of Tanner's techniques for the correction of diaphragmatic eventration and organo-axial gastric torsion.

References

Linnemann M P (1975) *Proceedings of the Royal Society of Medicine* **68**, 330
Tanner N C (1968) *American Journal of Surgery* **115**, 505