Medical Memoranda

Fragilitas Ossium: A Report of a Family

"Needless writing may be avoided by the statement that we know practically nothing of the aetiology of the condition (fragilitas ossium) beyond the fact that there is a strong element of heredity involved." It was nearly 50 years ago that Lovett and Nichols (1906) wrote these words, but they may still be considered true to-day. Spurway (1896) was the first to recognize the triad of blue sclerotics, liability to fractures, and familial occurrence. Bell (1928) in an exhaustive paper surveyed all previously reported family trees. Included were 25 families with 241 affected individuals, all occurring in Great Britain. Since then 13 families with 129 affected individuals have been reported in British literature. Thus, in all, 38 families totalling 370 affected individuals

have been reported in this country. This familial condition is therefore uncommon, which perhaps justifies the recording of a family tree hitherto unpublished.

CASE REPORT

On July 23, 1953, a woman aged 34 was admitted to hospital with suspected skull fracture. She gave a history of multiple fractures in her childhood. It was noted that her sclerotics were of a slate-blue colour and that she was partially deaf. Inquiry revealed that others in her family were similarly affected.

In order to define the family history three of the members were interviewed and others gave information by personal correspondence. As a result of this information the family tree (see diagram) was compiled.

COMMENT

Thirteen members of this family have blue sclerotics. Of these, 11 have suffered multiple fractures and 5 are partially deaf. The varying severity of the disease is indicated by comparison of Case 5, who has had only three rib fractures, with Case 13, who has suffered 41 fractures of limbs.

The liability to fractures ceases at the time of onset of otosclerotic deafness, at the end of the second decade. These findings are in accord with previous reports.

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Traumatic Diaphragmatic Hernia

Numerous historical cases of traumatic diaphragmatic hernia were reported by J. Reid in 1840, and were mostly due to injuries, penetrating and otherwise, received in warfare or in private quarrels. The following case is of interest in view of its relative rarity in civilian practice.

CASE HISTORY

On May 5, 1952, a previously healthy man of 48 accidentally left his car in gear while using the starting-handle. The car came forward and the front bumper crushed him across the lower chest against a nearby lorry.

He was admitted to hospital in a shocked condition and in great pain. There was no evidence of internal haemorrhage, but orthopnoea was a prominent feature. The most striking symptom at this stage was severe dysphagia. Water and saliva could be swallowed only with the greatest difficulty. The bruise caused by the bumper could be seen across the lower chest and epigastrium. Breath sounds were absent over the left base, but no adventitious sounds or mediastinal shift could be elicited. In the abdomen no distension or free fluid was detected. A straight x-ray film showed an irregularity in the left dome of the diaphragm, and diaphragmatic injury was suspected.

On the following day his condition was much improved and he could swallow fluids without difficulty. This change may have been due to a widening of the hernial orifice, leading to relaxation of constriction at the cardiac end of the stomach. On May 7, however, his abdomen was distended and he reported that he had passed neither flatus nor faeces since the accident. He was very short of breath and had a constant desire to eructate, but it hurt him to do so. A barium swallow showed the fundus of the stomach above the diaphragm and distended with gas, and that as the stomach ascended into the chest the greater curve had revolved upwards and the lesser curve downwards owing to its fixation by the left gastric vessels. No diaphragmatic movements could be detected. A barium enema showed an obstruction at the site of the splenic flexure, and this flexure was in an abnormally high position. A diagnosis was then made of ruptured diaphragm with herniation of the stomach and colon.

At operation the combined thoraco-abdominal approach recommended by Carter and Giuseffi (1948) was used. The eighth rib was resected as far back as its angle. By this means the distended colon and bulky omentum were more easily displaced downwards, though it was realized that, had the hernia been smaller, the thoracic approach recommended by Blackburn and D'Abreu (1945), Robertson (1947), Dunhill (1948), and Dugan and Samson (1948) would have been perfectly adequate. A transverse tear 5 in. (12.5 cm.) long was found in the left dome, extending as far as the oesophageal hiatus, and through it the stomach, left lobe of the liver, splenic flexure, and greater omentum had passed. The edge of the opening was frayed and of normal thickness, except at the medial end, where it became smooth and thin, suggestive of a *pre-existing* hiatal defect. The stomach was emptied through an oesophageal tube and returned to the abdomen without difficulty. The splenic flexure and omentum were wrapped in moist towels and delivered through the abdominal end of the incision. The tear was then repaired with mattress sutures of silk reinforced by a second row of interrupted sutures in a manner similar to the Mayo repair for para-umbilical hernia.

No attempt was made to close the hiatus completely, as what was left of it after repair was protected by the left lobe of the liver and, according to Robertson (1947), there is always a risk of placing a suture too close to the oesophagus and so causing a stricture. Crushing of the phrenic nerve is recommended if the diaphragmatic movements make repair difficult (Lam, 1950), or if there is a gross diaphragmatic defect (Dunhill, 1948). In this case, however, it was