

Our patient differs from most others described in that the symptoms and E.C.G. changes were readily reproduced on swallowing, without resort to balloon dilatation of the oesophagus. In addition his syncope was a presenting symptom of oesophageal carcinoma. Since an oesophageal lesion is commonly found in patients with "swallow syncope" we suggest that a barium swallow should be performed in all patients with this syndrome, even in the absence of dysphagia.

We are grateful to Dr. R. W. Portal for permission to report on one of his patients and for his help with the preparation of the manuscript, and for the help of Mr. K. Moghissi who performed the operation.

References

- Lichstein, E., and Chadda, K. D., *American Journal of Cardiology*, 1972, 29, 561.
- Kopald, H. H., et al., *New England Journal of Medicine*, 1964, 271, 1238.
- Sapru, R. P., et al., *British Heart Journal*, 1971, 33, 617.
- Levin, B., and Posner, J. B., *Neurology*, 1972, 22, 1086.
- Alstrup, P., and Pedersen, S. A., *Acta Medica Scandinavica*, 1973, 193, 365.

The Cardiac Department, Kingston General Hospital, Hull HU3 1UR
I. W. TOMLINSON, M.B., B.S., Registrar
K. M. FOX, M.B., M.R.C.P., Registrar

Treatment of Fractures of the Long Bones in Brain Stem Injury

The partial maintenance of the position of a reduced fracture by the stabilizing action of the muscles with constant normal tone is well known. If the muscle tone becomes abnormally increased the muscles will displace the fracture by anteroposterior displacement or overlapping. Traction will only increase the abnormal tone of the muscles through the stretching reflex and displacement will increase. This is true in the conservative treatment of fractures of long bones associated with decerebrate rigidity due to brain stem injury, and this paper emphasizes the danger of such treatment.

Case Report

An 8-year-old boy, knocked unconscious by a car, had brisk reflexes with a positive Babinski sign and some generalized spasticity. There was a fracture deformity of the right femur. An x-ray examination of the skull and cervical spine showed no bony injury but that of the right femur showed oblique fracture at the junction of the middle third to the distal third. The diagnosis was a closed oblique fracture of the right femoral shaft associated with brain stem injury.

He was kept under observation and traction with a 1.8 kg weight in a Thomas splint was applied to the femur. An x-ray examination showed a 2.5 cm overlapping, and an anteroposterior displacement. The traction weight was increased by 1.8 kg. Two days later blood was noticed on the crêpe bandage around the thigh, and there was a 3.75-cm wound with a spike of bone protruding. This was treated by suturing the wound and applying skeletal traction. There was still persistent overlapping and anteroposterior displacement (see figure). Clinical examination at the time showed generalized rigidity with elbows in flexion, wrists in palmar flexion, and feet in equinus.

Two days later the thigh bandage was again wet with blood, and another wound, 2.5 cm long, was found in a different position from the previous one, with the spike of the proximal fragment of the fractured femur protruding through it. There was some redness of the skin around the wound and some skin crepitation. Gas gangrene was suspected. The patient had a total wound excision, muscle biopsy, and swab for culture. In addition, an open reduction and internal fixation of the fracture using a six hole plate was performed. Antibiotics were started after operation. The culture proved negative.



The displacement of the fracture persists. The skeletal traction did not improve the position of the fracture.

Discussion

The complications of fractures after electric convulsion treatment have been reported.^{1,2} In a patient with a fracture any coexisting injury to the central nervous system with increased muscle tone can produce severe displacement of the fracture. Any lesion which cuts off the descending tract from the red nucleus and leaves Deiter's nucleus intact leads to decerebrate rigidity,³ and this happens in brain stem injury. This results in full extension of the hips and knees and plantar flexion of the ankles. The arms may be extended or flexed and they are held firmly in this position.

The fluctuation in the level of consciousness determines the severity of the brain stem injury. If, after a head injury, consciousness is not lost or is rapidly gained no serious degree of injury to the brain stem will occur. The importance of this observation is that the original trace of resistance present on testing joint movement in the patient will not be followed by decerebrate rigidity and the fracture may be treated by conservative methods provided that the slightly increased muscle tone is not displacing the fracture.

I should like to thank Mr. D. F. Paton, F.R.C.S., for permitting me to report the details of his patient.

- Kelly, J. P., *Journal of Bone and Joint Surgery*, 1954, 36B, 70.
- Newbold, H. I., *Diseases of the Nervous System*, 1958, 19, 385.
- Rowbotham, G. F., *Acute Injuries of the Head*, 4th edn. Baltimore, Williams and Wilkins, 1964.

Whittington Hospital, London N19 5NF
P. MERIANOS, M.D., Orthopaedic Registrar

Reversible Infertility in Male Coeliac Patients

Though infertility reversed by treatment with a gluten-free diet has been reported in women with coeliac disease¹ it has not been reported in men. In our group of 40 men with coeliac disease, we encountered two in whom infertility was corrected after treatment for three and five years with a gluten-free diet.

Case Histories

The first patient was investigated for infertility at the age of 29 in 1964. He had been married for nine years. A physical examination was normal, as was his sexual function. His wife had been investigated a year earlier and no abnormalities found. In 1970 his G.P. referred him for investigation of aphthous ulcers. Tests showed a low serum folate level, faecal fats 19 g/dy, and subtotal villous atrophy on jejunal biopsy. Coeliac disease was diagnosed and a gluten-free diet started in September 1970. In 1974 repeat jejunal biopsy showed mucosal return to normal, and his wife gave birth to a healthy 7 lb (3.175 kg) male infant.

Seminal Fluid Analyses

Date	Vol (ml)	Count (Millions/ml)	% Motility
Case 1			
<i>Before gluten-free diet</i>			
13.10.64	2.0	4.0	20
20.10.64	2.0	7.5	5
16.3.65	2.0	3.0	5
30.3.65	2.5	1.0	50
20.4.65	1.0	2.0	5
18.5.65	2.0	5.0	30
1.6.66	1.5	4.0	10
<i>After gluten-free diet</i>			
20.9.71	2.0	1.0	5
28.9.71	2.0	1.0	5
5.10.71	3.5	5.0	50
10.11.71	1.5	32.0	20
17.1.74	2.0	10.0	50
Case 2			
<i>Before gluten-free diet</i>			
1.7.52	2.0	1.0	5
8.7.52	2.0	1.0	5
<i>After gluten-free diet</i>			
4.2.74	4.0	20	42
11.2.74	4.0	10	60

The second patient was investigated by his G.P. at the age of 34 in 1952. He had been married for 14 years with no children. A physical examination was normal, as was his sexual function. No abnormalities had been found when his wife was investigated in 1946. In 1968 he was referred from his G.P. with a three-week history of diarrhoea. Faecal fats were 22 g/dy, and jejunal biopsy showed subtotal villous atrophy. He was diagnosed as having coeliac disease and started on a gluten-free diet. In 1973 a repeat jejunal biopsy showed normal histology.

Discussion

The beneficial effect of gluten withdrawal was judged in both cases by the return to normal of seminal fluid analyses which had been sub-normal before treatment according to established criteria,² in which sperm counts below 10 million/ml, motility less than 40%, and total volume below 1.5 ml may be regarded as "suboptimal for conception." Further proof of fertility was provided by successful pregnancy in one wife, but the second patient's wife had reached the menopause before he began his gluten-free diet.

The mechanism of this reversible infertility in both men and women remains obscure. Malnutrition, wasting, and vitamin B₁₂ deficiency may cause infertility in both sexes³ but were not present in either our two men or the three women previously reported with reversible infertility. Folate deficiency, iron deficiency, and anaemia were noted in some of these patients, but do not cause infertility by themselves.⁴

Despite the prolonged failure of conception in the marriages, these husbands were examined for infertility no less than six years and one and a half years after their wives had been investigated and pronounced normal. Coeliac disease in adults is often completely asymptomatic and should, therefore, always be considered as a possible cause in cases of infertility with oligospermia in men.

¹ Morris, J. S., Ajdukiewicz, A. B., and Read, A. E., *Lancet*, 1970, 1, 213.

² Santomauro, A. G., Sciarra, J. J., and Varma, A. O., *Fertility and Sterility*, 1972, 23, 245.

³ *Lancet*, 1945, 1, 281.

⁴ Hall, M., and Davidson, R. T. L., *Journal of Clinical Pathology*, 1968, 21, 599.

Department of Medicine, University of Bristol

P. G. BAKER, M.B., D.OBST.R.C.O.G., Senior House Officer
A. E. READ, M.D., F.R.C.P., Professor of Medicine

Late Recurrence of Thrombotic Thrombocytopenic Purpura after Splenectomy

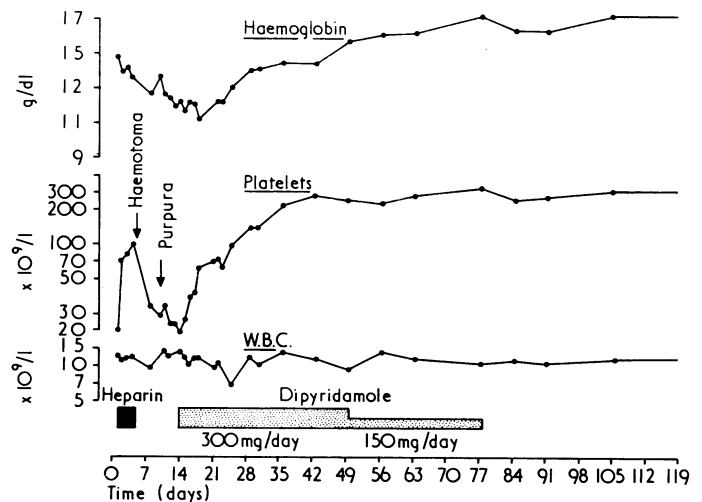
A unique case of recurrence of thrombotic thrombocytopenic purpura (T.T.P.) is described; surprisingly, this occurred 12 years after splenectomy.

Case Report

This man's initial episode¹ fulfilled the criteria of Amorosi *et al.*² for a diagnosis of T.T.P. He was treated with massive doses of steroids, blood transfusion, and, ultimately, splenectomy. Clinical and haematological remission was maintained for 18 months before he was lost to follow up. In 1974 he presented with dark urine and slight jaundice, which had developed one week after an influenza-like illness comprising sore throat, cough, anorexia, and vomiting. Examination confirmed the icterus but the only additional finding was an initial temperature of 37.4°C, which did not recur.

Laboratory investigation showed microangiopathic haemolysis in the absence of a detectable clotting abnormality: there was red-cell fragmentation; bilirubin was 34.2 µmol/l (2.0 mg/100 ml), lactic dehydrogenase 1720 U/ml, and platelets $22 \times 10^9/l$; fibrinogen titre was 1/128 and fibrin degradation products in blood and urine <2 g/l. Renal function was impaired, with blood urea level 11.8 µmol/l (71 mg/100 ml), creatinine clearance 82 and 89 ml/min successively, and urine protein 370 mg/l; granular casts were seen. The only bacteriological abnormality was an antistreptolysin O (A.S.O) titre of 625 U/ml.

Treatment was begun with intravenous heparin, 10 000 units six hourly, and he was given penicillin because of the history of sore throat. The platelet count rose immediately (see fig.) but heparin was stopped four days later because of a large haematoma at a venepuncture site. The platelet count fell during the next week to $20 \times 10^9/l$ and purpura appeared on the legs and chest. Dipyridamole 300 mg/day was begun and the platelet count rose



Treatment and course of patient with recurrence of thrombotic thrombocytopenic purpura.

immediately. Over the next two weeks platelet levels became normal and no new purpura appeared. Subsequently dipyridamole was withdrawn by steps (fig.) without deterioration clinically or biochemically.

In contrast to the initial episode fever was brief and the patient never seemed seriously ill; there was no apparent mental or neurological impairment; there was laboratory evidence of microangiopathic haemolysis, but this was mild (as judged by a reticulocyte count of 2% or less); and there was never any evidence of excessive fibrin consumption or fibrinolysis. After five days the blood urea and serum bilirubin levels returned to normal and urobilinogen was no longer detectable in the urine. Six months after his illness he remained in remission.

Discussion

Late recurrence of T.T.P. has not been described. The condition was probably not chronically active in this patient in view of his full haematological remission and the 12 symptom-free years which intervened. The relapse may have been precipitated by an upper respiratory tract infection, and the raised A.S.O. titre raised a possible streptococcal aetiology. Heparin alone produced a pronounced rise in the platelet count. Giromini *et al.*,³ however, found that full heparin and prednisone treatment produced no response, though dipyridamole and aspirin therapy produced good results.

Dipyridamole is a potent inhibitor of platelet aggregation. Its rational use in T.T.P. is based on T.T.P.'s inclusion⁴ among thrombotic conditions in which the lesion seems restricted to platelet aggregation and consumption. Importantly for possibly debilitated patients, T.T.P. has been treated successfully with aspirin, dipyridamole, and steroids without recourse to splenectomy.⁵ Dipyridamole seems to play the key part in restoring the life span of platelets to normal. Aspirin, though ineffective alone, allows lower doses of dipyridamole to be used.

Our patient might have had only mild disease because of his earlier splenectomy, and this might account for the dramatic effectiveness of heparin and then dipyridamole alone. Certainly, dipyridamole should not be used alone in the patient who retains a spleen, but together with agents such as aspirin, steroids, and dextran 70 it may be useful for treating this serious condition.

We thank Dr. Ian Fraser, consultant haematologist, for his personal involvement and organization of haematological support in the care of this patient and Boehringer Ingelheim for 100-mg tablets of dipyridamole.

¹ Moorhead, J. F., *Archives of Internal Medicine*, 1966, 117, 284.

² Amorosi, E. L., and Ultman, J. E., *Medicine*, 1966, 45, 139.

³ Giromini, M., *et al.*, *British Medical Journal*, 1972, 1, 545.

⁴ Harker, L. A., and Slichter, S. J., *New England Journal of Medicine*, 1972, 287, 999.

⁵ Rossi, E. C., Redondo, D., and Borges, W., *Journal of the American Medical Association*, 1974, 228, 1141.

Southmead General Hospital, Bristol BS10 5NB

D. J. HOWARD, B.Sc., M.R.C.P., Medical Registrar
A. B. ROBERTS, D.C.H., M.R.C.P., Medical Registrar
F. T. PAGE, M.D., M.R.C.P., Consultant Physician