tests have improved considerably. We thus hope that we have started treatment before significant permanent damage has occurred.

Comment

The occurrence of hæmolysis in Wilson's disease seems first to have been noted by Cartwright *et al.* (1954) who described 2 sisters, one of whom was otherwise asymptomatic. Of 20 cases of Wilson's disease described by Walshe (1962), 6 presented with jaundice which was at least partly hæmolytic in 3 cases; one of his cases had acute episodes like ours. Turpin *et al.* (1962) also describe a very similar case.

Mechanism of hæmolysis: Acute hæmolytic anæmia has been described in sheep feeding on high-copper pastures in which, for unknown reasons, stored copper is suddenly liberated into the plasma in high concentration; this is followed by acute hæmolysis. This mechanism seems unlikely in Wilson's disease where wide fluctuations in copper concentration have not been observed, but it has not, of course, been possible to study these patients just before an attack. Our patient's red cells were unaffected by a moderate concentration of copper.

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Dr J M Walshe: Although it is not commonly recognized, hæmolytic episodes are not a rare complication of Wilson's disease. Unfortunately one seldom sees the patient at the time when hæmolysis is in progress so that studies of the erythrocytes are usually made on surviving cells which are resistant to the hæmolytic process. We studied this patient in Cambridge shortly after a major hæmolysis and could find no evidence of the mechanism involved. It has been suggested in the past that the red cell copper concentration rises to very high levels before hæmolysis occurs (as is known to happen in sheep on copper-rich pasture). Her red cell copper was slightly below our normal range at 87 μg/100 ml. Her red cells showed a slightly increased resistance to hypotonic saline and incubation of her cells in a copper-containing buffer (100 μg/100 ml) did not increase the fragility of the red cells. It did, however, result in slight hæmolysis during the incubation period but no more than was observed in red cells from a normal individual. During the course of *in vivo* radio-copper studies we found that her red cells bound less copper than those of our control subjects. There is, however, evidence that copper can interfere with red cell metabolism and Dr W D Stein (PhD thesis, London) was able to show that copper blocked the uptake of substrate by the red cell membrane by blocking the N terminal histidinyl group and that the mechanism could be reversed by the addition of histidine. It is therefore possible that this system in the cell membrane becomes inactivated as the cells age and that finally a stage is reached when the older cell population is destroyed by hæmolysis and the younger cells remain resistant so that the crisis is self-limiting and the patient is then apparently well until a further generation of cells have aged and become vulnerable. To get evidence that this occurs *in vivo* it is clearly necessary to see a patient either just before or at the time of hæmolysis and we hope one day this may be possible.

'Spontaneous' Rupture of the Rectum

D G Young FRCS
(for Professor A W Wilkinson ChM FRCS)

Spontaneous perforation of the alimentary tract is rare, Thelander (1939) found only 20 cases of spontaneous perforation of the large bowel in the neonate. The majority of these patients died without surgical intervention and the condition continued to be uniformly lethal until Doyle (1951) reported the first successfully treated baby.

That surgical intervention is necessary is widely accepted but it is interesting to note the variety of surgical procedures which have been adopted in the surviving infants. These are: (1) Simple closure of the perforation (Doyle 1951, Sabransky 1961). (2) Proximal colostomy (Standard 1952, Scott 1963). (3) Right hemicolectomy (Roseman 1958). (4) Closure with instillation of neomycin to the bowel through an enterostomy tube (Nienhuis 1963).

Case 1 Boy. Birth weight 4·1 kg. First-born child He had been normal for the first eighteen hours of life. On taking a rectal temperature the thermometer was broken and he subsequently developed abdominal distension and œdema of the anterior abdominal wall. He looked ill and X-ray examination of the abdomen showed a pneumoperitoneum. At laparotomy greenish yellow fluid escaped from the peritoneal cavity and a 4 mm perforation of the anterior rectal wall was found. This was considered to be due to spontaneous gangrene of bowel and was closed, proximal colostomy being performed. The baby made a good recovery, the colostomy being closed six months later. He is now fit and weighs 11·2 kg at 10 months of age.

Case 2 Boy. Birth weight 2·8 kg. First-born child Admitted with a similar lesion. He was well for seven days and then refused feeds, vomited, and

the abdomen became distended. The following morning he required resuscitation with intravenous fluids and was given hydrocortisone before he was considered fit for transfer. On admission he was gravely ill. X-ray examination showed a pneumoperitoneum. He collapsed before going to theatre and aspiration of 45 ml gas and 3 ml thick straw-coloured fluid from the peritoneal cavity relieved him sufficiently to allow cardiac and respiratory function to return. At laparotomy gross fæcal staining of the peritoneal cavity was present and a 3 cm tear in the anterior rectal wall was found. This was closed and transverse colostomy performed, but despite all resuscitative measures the baby died eight hours post-operatively.

In each of these cases cultures from the peritoneal cavity yielded *Esch. coli;* in Case 2 proteus and *Strep. fæcalis* were also grown. Blood culture was not done, but it is interesting to note that Sabransky's successful case yielded *Esch. coli* in cultures of peritoneal fluid and blood and he used chloramphenicol. Case 1 was treated with penicillin and streptomycin. Case 2 was treated with ampicillin to which all the organisms were subsequently shown to be sensitive.

Comment

The ætiology of spontaneous rupture of the bowel remains obscure but in Case 1 there was a history of a thermometer being broken while a rectal temperature was being taken. This perforation may have been traumatic. No history of trauma could be elicited in Case 2 and the habit in the nursing home in which this child was being nursed was simply to take a rectal temperature if it was thought to be indicated by the child's general condition.

The site of these perforations on the anterior rectal wall just above the peritoneal reflection corresponds with the site of perforation in cases reported following barium enema (Hartman & Hills 1957) and in perforations of the rectum by a thermometer (Segnitz 1957, Warwick & Gikas 1959, Miller 1962, Canby 1963). I suspect that in these two cases perforation of the rectum was produced by a thermometer and was not spontaneous, as considered at operation.

Two points emerge: Instruction to nurses on the anatomy of the rectum must be clear and to take a rectal temperature the thermometer should be inserted into the anal canal and then advanced at an angle of 30 degrees backwards, not straight into the rectum parallel to the cot as one so often sees. Secondly, one should be aware of this rare but dangerous complication as early recognition is vital if treatment is to be successful. Straight X-ray of the abdomen in the erect position is the most useful aid in diagnosis.

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Iatrogenic Vitamin B Deficiency

Douglas Pickering MB DCH
(for John Lorber MD FRCP)

K W, girl, born 9.12.63. Birth weight 6½ lb

History: Presented 28.5.64 with failure to gain weight for one month, vomiting and constipation. Physical examination showed a grossly wasted child (weight 9 lb 4 oz). She had sunken eyes, her liver was enlarged one fingerbreadth, her blood pressure was 65 systolic. There were no other abnormal physical signs.

Relevant investigations: Urine: albumin 10 mg/100 ml; WBC 15–20 pus cells per high-power field; heavy growth of *Esch. coli* and proteus repeatedly; specific gravity 1010. Electrolytes: sodium 146, chloride 113, potassium 4·3, CO_2 23 mEq/l., urea 94, calcium 11·7, phosphate 6·2 mg/100 ml, alkaline phosphatase 5 K-A units. IVP: no stones, no nephrocalcinosis, no osteosclerosis.

An initial diagnosis of urinary tract infection was made and the following treatment given: sulphadimidine 0·5 g t.d.s. (1.6.64–4.6.64), nalidixic acid 12·5 mg t.d.s. (4.6.64–1.7.64), nitrofurantoin 25 mg t.d.s. (19.6.64–1.7.64). The bacteria and white cells disappeared from the urine, but there was no clinical improvement. The vomiting continued and required treatment with intravenous fluids. Serum calcium showed persistently elevated levels, reaching 13·4 mg/100 ml on 18.6.64. A diagnosis of hypercalcæmia was made. Treatment was begun 18.6.64 with low calcium milk. By 3.7.64 her general condition had much improved: weight 10 lb. 4 oz. She was discharged home on a low calcium diet and sulphadimidine 0·5 g t.d.s.

27.7.64: Seen at Outpatients; continuing to gain weight.

13.8.64: Readmitted; vomiting for twenty-four hours. Bowels opened normally. She had lost 1 lb 3 oz in twenty-four hours. Physical examination showed marked dehydration; no other