from the patient's home and that of a neighbour whose hot water system also used copper piping. The results were compared with samples from the hospital (mixed copper and other piping) and North London (no copper piping) (Table).

<table>
<thead>
<tr>
<th>Source of Water</th>
<th>Cold Water Tap (Cu µg/100 ml)</th>
<th>Hot Water Tap (Cu µg/100 ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient's home</td>
<td>35</td>
<td>79</td>
</tr>
<tr>
<td>Neighbour's home</td>
<td>30</td>
<td>68</td>
</tr>
<tr>
<td>Hospital</td>
<td>4</td>
<td>30</td>
</tr>
<tr>
<td>North London</td>
<td>8</td>
<td>16</td>
</tr>
</tbody>
</table>

From these results it is seen that the copper content of 'hot' water is significantly higher than water from the cold tap. The presence of copper piping apparently leads to a significant increase in the copper content of the water.

As was mentioned earlier the copper oxidase activity was only 0.10 units, and because of this the caeruloplasmin (α-copper binding protein) level was estimated at 22.5 mg/100 ml (Dr. J. Walsh). The determination was based on the enzyme assay on whole plasma and compared with results obtained from similar assays using purified caeruloplasmin. This result effectively eliminates hepato-lenticular degeneration from the differential diagnosis. Low levels of caeruloplasmin have also been reported in portal cirrhosis (Gubler et al., 1957) and in nephrosis (Cartwright, Gubler, and Wintrobe, 1954), but in this case these conditions can be ignored. During the convalescent phase liver biopsy was performed and histology revealed a normal architecture. Copper assay on biopsy material gave levels of 4.7 and 5.1 µg/g wet weight, which are at the lower end of the normal range. It thus appears that he had cleared his stored copper very well. It is disappointing not to have the results of liver biopsy before treatment was started as this would have made the diagnosis of copper poisoning wholly tenable. The infant's condition on first admission was such that biopsy was never considered. The critical change in the clinical status after starting D-penicillamine was thought to be due to mobilization and excretion of stored copper. In fact it constituted acute copper toxicity superimposed on a more chronic picture.

In conclusion it is suggested that this child was subjected to a high copper intake over a period of three months and showed abnormal sensitivity to the metal. The clinical picture was that of pink disease. From the progress following chelation with D-penicillamine and the fact that mercury was not found, we put forward the suggestion that in some children excess copper can cause a clinical picture that cannot be distinguished from pink disease. The long-term prognosis for this child is probably good.

Summary

A case of chronic copper poisoning in a male infant of 15 months is reported. It is believed to be the first case report. The infant presented with a picture indistinguishable from that of classical pink disease (acrodynia). The copper had been ingested over a period of 3 months from contaminated water. Treatment consisted of D-penicillamine and prednisolone. Recovery was slow and at one time the patient's condition became critical.

We thank our Ward Sister, Miss B. M. Barchard.

REFERENCES


Michael A. Salmon and Trevor Wright

Park Hospital for Children, Oxford, and the Children's Hospital, Sheffield.

Some Problems of Gastrointestinal Bleeding in Children

Gastrointestinal haemorrhage is not an uncommon condition in children, yet little has been written about it from Britain. This is surprising since assessment of the severity of the bleed, its site of origin, and the subsequent management may be difficult, especially when no obvious cause is found. A retrospective study was therefore, made of all
cases of bleeding from the gut admitted to the children's ward of a general provincial hospital with a view to finding any useful guides to management. All children who had gastrointestinal bleeding as a principal complaint were included in the study, which was of a period of just over 8 years.

Reactive or secondary bleeds after tonsillectomy were not included. Children thought to have gastroenteritis were sent direct to another hospital with facilities for isolation, but inevitably a few cases slipped through the net and so have been included. The ward accepted admissions varying in age from birth to puberty. The study was undertaken in a general provincial hospital and did not attract cases from outside its immediate catchment area because of a special interest in the condition.

Results of Survey
The Table shows the breakdown of causes of the bleed with the age of the patients.

<table>
<thead>
<tr>
<th>Cause</th>
<th>No. of Cases</th>
<th>Ages of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Undiagnosed</td>
<td>16</td>
<td>1 mth, 1 mth, 2 mth, 5 mth, 6 mth, 9 mth, 10 mth, 10 mth, 19 mth, 21 mth, 2 yr, 2½ yr, 4 yr, 5 yr, 7 yr, 7 yr</td>
</tr>
<tr>
<td>Haemorrhagic disease of the newborn</td>
<td>8</td>
<td>7 hr, 1 dy (4), 2 dy (2), 3 dy 2½, 3, 4, 7, 10 yr</td>
</tr>
<tr>
<td>Rectal polyp</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Trauma to pharynx and foreign body</td>
<td>3</td>
<td>3 mth, 11 mth, 3 yr</td>
</tr>
<tr>
<td>Henoch-Schönlein purpura</td>
<td>3</td>
<td>3, 6, and 8 yr</td>
</tr>
<tr>
<td>Anal fissure</td>
<td>3</td>
<td>5 wk, 3, and 9 yr</td>
</tr>
<tr>
<td>Hiatus hernia</td>
<td>2</td>
<td>7 wk, 18 mth</td>
</tr>
<tr>
<td>Gastroenteritis</td>
<td>2</td>
<td>1 mth, 8 yr</td>
</tr>
<tr>
<td>Haemolytic uraemic syndrome</td>
<td>2</td>
<td>20 mth</td>
</tr>
<tr>
<td>Total bowel volvulus</td>
<td>1</td>
<td>1 dy</td>
</tr>
<tr>
<td>Inflamed pharynx with ulcers</td>
<td>1</td>
<td>10 yr</td>
</tr>
<tr>
<td>Meckel's diverticulum</td>
<td>1</td>
<td>9 mth</td>
</tr>
<tr>
<td>Acute infection elsewhere (encephalitis)</td>
<td>1</td>
<td>2 yr</td>
</tr>
<tr>
<td>Intussusception</td>
<td>1</td>
<td>4 mth</td>
</tr>
<tr>
<td>'Haemangiomatosia'</td>
<td>1</td>
<td>3 yr</td>
</tr>
<tr>
<td>Oesophageal varices</td>
<td>1</td>
<td>5 yr</td>
</tr>
<tr>
<td>Haemophilia</td>
<td>1</td>
<td>Repeated bleeds 6 mth to 5 yr</td>
</tr>
<tr>
<td>Total</td>
<td>51</td>
<td></td>
</tr>
</tbody>
</table>

Further relevant details of this series are as follows.

Mortality. Zero.

Morbidity. The most severe complication was in the case of the 1-day-old baby with a total intestinal volvulus. At operation a considerable length of small intestine was removed. In the immediate postoperative period all was well, but six months later the baby had a severe subarachnoid haemorrhage. Investigations suggested that this was because of a clotting deficiency due to failure of absorption of vitamin K as a result of removal of the ileum. The subarachnoid haemorrhage left the child with a severe left hemiparesis.

The case of oesophageal varices has had several bleeds in spite of a portacaval anastomosis. The prognosis in this child appears poor.

The haemophiliac is admitted to hospital frequently for bleeding from a variety of sites (not always the gut). He is now aged 5 and has been under review virtually since birth.

The case of haematemesis as a sequel of encephalitis has subsequently become mentally retarded, presumably secondary to the encephalitis.

Other than these cases there is no significant morbidity.

Comment and Discussion
Haematemesis is unusual in children when compared with bleeding per rectum. In this series there was a total of 13 patients who vomited blood, all referable to lesions of the stomach or higher, except single cases of haemophilia, encephalitis, and haemorrhagic disease of the newborn. All other cases presented with bleeding per rectum.

Of the 9 cases presenting in the first week of life, 8 had haemorrhagic disease of the newborn and 1 had total bowel volvulus. Concerning particular causes of rectal bleeding after the first week, there was only one case of Meckel's diverticulum in this series. The view of Hutchison (1964) that, 'The commonest cause of unheralded gastrointestinal bleed in infancy is peptic ulceration of a Meckel's diverticulum', is held by others, notably Abrams and Lynn (1962). Likewise, only one case of intussusception occurred here, but it must be pointed out that several cases of this condition did occur in the period covered, but did not present with haemorrhage.

No one condition stands out as being a common cause for paediatric gastrointestinal bleeding, but a large number of relatively uncommon potential causes exist. The trio of careful history, examination, and special investigations where indicated will be a good guide in the majority of cases. As Brayton (1964) has pointed out, the details of vomiting of blood, and whether rectal bleeding is fresh or melaena in nature, can sometimes sway the balance when considering laparotomy for possible conditions such as bleeding Meckel's diverticulum.
Short Reports

Peripheral Neuropathy and Ichthysis in Krabbe’s Leucodystrophy

The association of peripheral neuropathy and ichthysis is well recognized in Refsum’s syndrome but has not previously been recorded in Krabbe’s leucodystrophy.

Case Report

N.C. was born to West Indian parents at full term weighing 3409 g. She was first referred at the age of 10 months because of generalized ichthysis which had been present since early infancy.

Her early development was normal and she could sit unaided at 6 months. After the age of 7 months, however, she lost the ability to sit. She became irritable, cried constantly, and refused her feeds. She made no attempt to pull herself to a standing position.

On examination at 12 months, she lay with her arms and legs extended and feet plantar-flexed. She was unable to sit or stand and would not take any weight on her legs. There was increased tone round her elbow and knee joints but marked hypotonia round the wrist joints. Marked head lag with traction on her arms was present. Extensor spasms were frequent and distressed crying resulted from the slightest handling of her limbs. The hands and feet were cold and cyanosed. All the tendon reflexes were absent. The fundi were normal.

The CSF contained 70 mg/100 ml protein with a normal cell count and Lange curve. The EEG showed a preponderance of slow wave activity. The bone age was advanced to a 2-year level at 10 months.

Motor nerve conduction velocities were very slow; that for the right ulnar nerve was 13-0 m/sec; the left ulnar 13-3 m/sec, and the right posterior tibial nerve 10-3 m/sec (normal at one year = 46·1 ± 3·0 m/sec for the ulnar nerve and 38·2 ± 3·3 m/sec for the posterior tibial nerve).

The slow nerve conduction velocities confirmed the presence of a demyelinating peripheral neuropathy. This together with the progressive nature of the central nervous system disturbance suggested the diagnosis of either Krabbe’s leucodystrophy or metachromatic leucodystrophy. The former was thought more likely because of the early onset and rapid progression of the disease.

Urine examination for intracellular metachromatic material was repeatedly negative. Blood leucocyte arylsulphatase A levels were normal. The combination of ichthysis and peripheral neuropathy suggested the diagnosis of Refsum’s syndrome. Other features of this syndrome, such as ataxia and retinitis pigmentosa, were absent. There was no increase of phytic acid in the patient’s serum.

The patient’s condition rapidly deteriorated and she died at 14 months. Histological examination of the nervous system revealed the characteristic globoid cells confirming the diagnosis of Krabbe’s leucodystrophy.
Some problems of gastrointestinal bleeding in children.
R E Collins

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