Short Reports

fractions C2, C3, and C4 are required in the warm phase, and fraction C1 is probably required in the cold phase (Hinz, Picken, and Lepow, 1961).

Since the introduction of penicillin treatment for syphilis, PCH has become much less common, and an increasing proportion of cases is now seen in association with acute viral infections (Dacie, 1962). In the majority of virus-associated cases the attack of haemolysis occurs either during the prodromal stage or shortly after the clinical appearance of the infection. One patient, however, developed PCH 5 weeks after an attack of mumps (Colley, 1964): our patient developed PCH 6 weeks after measles immunization. The possibility that her mild measles-like illness 3 weeks after immunization was in fact a coincidental natural measles infection cannot be excluded, though the absence of a positive history of contact and the frequency of such reactions after immunization with live measles vaccine make this unlikely.

An important difference between syphilitic and virus-associated PCH is the acute transitory nature of the latter (Dacie, 1962). The haemoglobinuria generally persists for only a few days, and may, as in our patient, have ceased before admission. The antibody disappears from the serum after a period ranging from a few days to several weeks, at which time the direct antiglobulin and Donath-Landsteiner tests become negative. However, the attack, while it lasts, may endanger life, and usually necessitates transfusion with ABO and Rhesus compatible cells, warmed to approximately 37 °C. The great rarity of P negative cells, which would be unaffected by the antibody, makes their use impracticable. The severity of haemolysis in these cases may be due to the antibody having a higher thermal range for erythrocyte sensitization (up to 25 °C in our patient) than in syphilitic cases (rarely as high as 20 °C) (Dacie, 1962; Schubothe and Haeenle, 1961; Colley, 1964).

Summary

A case of paroxysmal cold haemoglobinuria is described in a 19-month-old girl following measles immunization.

Haemoglobinuria occurred during the sixth week after immunization, and was associated with a raised measles antibody titre. The autohaemolysin was a typical Donath-Landsteiner antibody, having specificity within the P blood group system, but it showed the higher thermal range for erythrocyte sensitization characteristic of the acute transient variety of this disorder.

We are indebted to Dr. B. S. B. Wood, consultant paediatrician, for permission to publish details of his patient, and to Dr. J. Stuart, consultant haematologist, for his helpful encouragement and advice.

REFERENCES


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Intrahepatic Gas Shadows in Neonatal Duodenal Obstruction

The presence of branching intrahepatic gas shadows extending to the liver edge in the neonate has been attributed to gas in the portal venous system, and has been considered a grave prognostic sign as it may be a feature of necrotizing enterocolitis (Touloukian et al., 1967; Wilson and Woolley, 1969), often with a Gram-negative bacteraemia.

Case Reports

Case 1. A male child, 2,835g, was born by lower caesarian section for antepartum haemorrhage. Bile-stained vomiting started on the first day. Meconium was passed from the 1st to 3rd days. Continued vomiting led to transfer from the maternity hospital. Examination on admission on the 5th day showed visible epigastric peristalsis in a scaphoid abdomen. X-ray of the abdomen (Fig. 1), showed a branching pattern of gas shadows extending out nearly to the edge of the liver shadow. A little gas was seen in the intestines. Laparotomy revealed a severe stenosis of the duodenum distal to the common bile duct opening. The jejunum and ileum were normal and there was no evidence of ischaemic gut. A posterior duodenojunostomy was performed with good recovery.

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Case 2. A male child was delivered normally at term. On the 5th day bile-stained vomiting started. The abdomen became distended and vomiting continued till admission on the 7th day. The abdomen was distended but the rectum was empty. X-rays (Fig. 2) revealed gas in the gastric wall, a branching pattern of gas shadows extending out to the liver edge, and no distal intestinal gas. Blood cultures were negative, and after electrolyte correction laparotomy revealed duodenal obstruction from bands with some malrotation.

Progress was stormy early, and a further laparotomy and pyloroplasty were performed after which a satisfactory recovery was made.

Discussion

The intrahepatic gas shadows in these 2 children extended nearly to the edge of the liver shadow, and thus fulfill the criteria of Susman and Senturia (1960) and Sisk (1961) for gas in the portal venous system. No necrotic tissue was shown on laparotomy in either of these cases and blood cultures in Case 2 were negative. Isdale (1970) reviewing interstitial emphysema of the stomach states that raised intragastric pressure may be a cause.

Case 2 showed air in the stomach wall, and it is suggested that in both these cases of duodenal obstruction, gas had been forced into the stomach wall and then passed into the portal venous system. It is concluded that portal venous gas is not always associated with necrotic tissue or overwhelming infection in the neonate.
Summary

Descriptions are given of two cases of gas in the portal venous system in association with duodenal obstruction and also one case with interstitial emphysema of the stomach.

I am grateful to Mr. D. G. Young and Mr. J. A. S. Dickson for their collaboration and for the co-operation of Messrs. Harvey Miller and Medcalf, Aylesbury.

REFERENCES


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Incidence of Salt-losing Form of Congenital Virilizing Adrenal Hyperplasia

The C-21 hydroxylase defect in the biosynthesis of adrenocortical steroids is the most common cause of congenital virilizing adrenal hyperplasia (CAH). Clinically and biochemically, two distinct forms of this syndrome are recognized: the non-salt-losing form and the salt-losing form. Since salt-losers and non-salt-losers generally occur in different families, it is believed that the two forms of CAH are genetically independent (Childs, Grumbach, and Van Wyk, 1956; Prader, Anders, and Habich, 1962).

Estimates for both forms of CAH in populations have ranged from 1 in 5,041 births in the canton of Zurich, Switzerland (Prader, 1958), to 1 in 67,000 births in Maryland (Childs et al., 1956). Assuming equal incidence for the two forms, Prader estimated the incidence of 1 in 12,500 births for the salt-losers and the non-salt-losers separately. The incidence of the salt-losing form alone has been reported to be 1 in 1,481 livebirths in Alaskan natives and 1 in 490 livebirths in Alaskan Yupik Eskimos (Hirschfeld and Fleishman, 1969). The present communication reports the incidence of the salt-losing form of CAH in Toronto, Canada.

Patients and Methods

Patients included in this report were born in metropolitan Toronto, a well-defined municipal area, between January 1960 and December 1967. The diagnosis of the salt-losing form of CAH was made on the basis of symptoms typical of adrenal insufficiency (vomiting, diarrhoea, failure to thrive, dehydration, electrolyte imbalance), ambiguous genitalia in female infants, and abnormal urinary steroid excretion.

Twelve patients (7 male and 5 female) with the salt-losing form of the syndrome were born during the 8-year period. These were all referred to The Hospital for Sick Children. Only one patient, a male, was diagnosed after necropsy. No additional cases were detected in other hospitals in the city. During the same period, there were 315,509 livebirths in the area.

Results

The incidence of the salt-losing form of CAH in Toronto was calculated to be 1 in 26,292 livebirths, with an estimated gene frequency of 0·00617 and heterozygote incidence of 1 in 82 livebirths (Table I).

Since only proven cases were used, it is not possible that these figures could overestimate the true incidence in the period under consideration. The results of the present study are compared with those from other series in Table II.

<table>
<thead>
<tr>
<th>Year</th>
<th>Total No. of Livebirths</th>
<th>No. of Cases Ascertained</th>
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</tr>
<tr>
<td>1961</td>
<td>40,157</td>
<td>1</td>
</tr>
<tr>
<td>1962</td>
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</tr>
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<td>1963</td>
<td>40,693</td>
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</tr>
<tr>
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<td>41,210</td>
<td>2</td>
</tr>
<tr>
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<td>39,126</td>
<td>2</td>
</tr>
<tr>
<td>1966</td>
<td>37,394</td>
<td>2</td>
</tr>
<tr>
<td>1967</td>
<td>36,430</td>
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</tr>
<tr>
<td></td>
<td>315,509</td>
<td>7</td>
</tr>
</tbody>
</table>

Note: Minimum incidence of salt-losing form = q² = 1 per 26,292 livebirths.
Frequency of salt-losing gene q = 0·00617.
Incidence of heterozygotes, 2pq = 1 in 82 livebirths.

Discussion

It is difficult to obtain complete ascertainment of patients with the non-salt-losing form of CAH in a population, since the diagnosis is either delayed, or not made at all. On the other hand, because of the life-threatening nature of the disease, patients with the salt-losing form of CAH usually present for treatment soon after birth (Iversen, 1955). Therefore, it is unlikely that any salt-losers born during the study period would be as yet undiagnosed. Limiting the study to a definite geographical area.
Intrahepatic gas shadows in neonatal duodenal obstruction.

D G Shaw

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