The latter were fed a formula of humanized cows' milk (Similac-20). Though the lactose content of this formula is almost identical to that of human milk (7-2 vs. 7-0 g/100 ml, respectively), as is shown in Tables 1 and 2, and so this cannot account for the greater proportion of breast-fed infants who have increased levels of stool reducing substances. Some other explanation must be sought, including the contribution of gut flora, but the inter-relationships between faecal sugar, pH, and flora is likely to be very complex. Whether the primary event in this inter-relationship is unabsorbed lactose entering the colon of breast-fed infants, influencing their colonic pH and flora, or whether small intestinal flora in such infants modifies the absorption of lactose is unknown and warrants investigation.

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Haemolytic uraemic syndrome in sibs

Sir,

The occurrence of multiple cases of a disease in some families does not by itself provide irrefutable evidence for genetic predisposition for that disease. This appears to be the case in the haemolytic-uraemic syndrome (HUS), which has been reported to affect multiple sibs in some families (Kaplan et al., 1975). The reports of simultaneous occurrence of the disease in unrelated adopted sibs (Chan et al., 1969) and the presence of asymptomatic mild cases in family contacts (Tune et al., 1974), along with a well-recognized predilection of the disease to appear in endemic (Gianantionio et al., 1964) and epidemic (McLean et al., 1966) forms in various parts of the world, however, appear to implicate environmental agents, namely infections, as causative factors.

In a review of 41 sibships with multiple cases of HUS, Kaplan and associates (1975) could distinguish two groups of families on the basis of (1) the interval between onset of disease in affected sibs, (2) geographical location (endemic versus nonendemic) of the families and, (3) severity of the syndrome as indicated by mortality. The authors speculated that the milder disease occurring simultaneously in sibs in families living in endemic areas could be attributed to environmental agents, while genetic factors may have played an important role in severe form of the disease occurring in sibs after longer intervals in families living in nonendemic areas.

We have seen a New York City family in which 2 male infants were affected with HUS with an interval of 2 years between the cases. The parents were healthy non-consanguineous black Americans. Both parents and 2 sisters (7- and 1-year-old, respectively) were in good health. The first infant was born in December 1970 and died in May 1971, the second was born in February 1973 and died in August 1973. The clinical picture included gastroenteritis, haemolytic anaemia, disseminated intravascular coagulopathy, hypertension, and renal failure. Necropsy was performed on the second infant only, and showed widespread fibrin thrombi in arterioles and capillaries throughout both kidneys.

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Dr. Walker-Smith comments:

I was pleased to see the letter of Drs. Nitzan and Rosenfeld confirming our finding that breast-fed infants have increased stool reducing substances. This observation supports the view that breast-fed babies have a temporary 'physiological malabsorption' of lactose. I agree that there is not a higher concentration of lactose in breast milk as compared to the artificial feeding formula used.
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These 2 cases provide additional examples of HUS occurring in sibs after a long interval. The family resided in New York City, which is not considered as an endemic area for HUS, and the clinical course of the disease in both infants was almost identical, culminating in early death.

The aetiology of HUS may indeed be multiple, and the genetic basis for a significant proportion of cases does not seem very likely. However, the familial cases with longer intervals between onset, as presented here, cannot simply be discarded as fortuitous instances of environmental causative factors. It may be that the genetic predisposition for HUS is operative in such families, implicating an autosomal recessive trait.

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References


Aetiology of hydrops fetalis

Sir,

Drs. Radford, Izukawa, and Rowe (1976) in their article on congenital paroxysmal atrial tachycardia happen also to contribute a great deal to the literature on hydrops fetalis. Several reviews on the conditions associated with hydrops fetalis point to the varying aetiologies of the disease (Becker, 1975; Driscoll, 1966). Most of these diseases are associated with congestive heart failure, severe anaemia (usually haemolytic), and/or cellular damage to the liver or placenta. Unfortunately, no one process can explain all the cases. In erythroblastosis fetalis, hydrops fetalis is most related to hyproproteinaemia (Phibbs et al., 1974) or low oncotic pressure (Baum and Harris, 1972) rather than universal anaemia. Whether hyproproteinaemia is due to liver disease or to congestive heart failure is uncertain; however, cases of 'pure' output failure such as those reported by Radford et al. offer an ideal opportunity to test the hypothesis that hydrops results from hyproproteinaemia secondary to heart failure and not vice versa. Do they have the necessary protein levels on their patients?

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References


Dr. Radford and colleagues comment:

Regarding the interesting letter from Dr. K. L. Harkavy, we do have serum protein data on 3 of our 12 patients (Cases 8, 11, 12). Initial blood specimens were taken on the first day of life in all 3 patients and repeated later in 2. Electrophoretic pattern analyses were also done. The relevant results are in the Table.

Although the data are limited, there was definite hypalbuminaemia in Cases 8 and 11. This would fit with the hypothesis raised by Dr. Harkavy, i.e. that hyproproteinaemia was secondary to heart failure and led to hydrops.

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555, University Avenue, Toronto,
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<table>
<thead>
<tr>
<th>Patient age</th>
<th>Case 8 1d</th>
<th>Case 11 1d</th>
<th>Case 12 1d</th>
<th>Age related normal range</th>
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<tbody>
<tr>
<td>Total protein g/100 ml</td>
<td>4.7</td>
<td>4.6</td>
<td>5.5</td>
<td>4.6–7.0</td>
</tr>
<tr>
<td>Albumin g/100 ml</td>
<td>3.1</td>
<td>2.6</td>
<td>3.6</td>
<td>3.2–4.8</td>
</tr>
<tr>
<td>$\alpha_1$</td>
<td>0.3</td>
<td>0.4</td>
<td>—</td>
<td>0.1–0.3</td>
</tr>
<tr>
<td>$\alpha_2$</td>
<td>0.5</td>
<td>0.4</td>
<td>—</td>
<td>0.2–0.6</td>
</tr>
<tr>
<td>$\beta_1$</td>
<td>0.5</td>
<td>0.7</td>
<td>—</td>
<td>0.3–0.6</td>
</tr>
<tr>
<td>$\gamma$</td>
<td>0.4</td>
<td>0.5</td>
<td>—</td>
<td>0.6–1.2</td>
</tr>
</tbody>
</table>
Haemolytic uraemic syndrome in sibs.

Q H Qazi and E J Schutta

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