values (Table). The lowest concentration was found in the mother who has the highest IQ of all family members. The highest concentration was obtained in the first child who was severely mentally defective. Apart from low values of threonine and tyrosine in all family members, no uniform trend in the distribution of the other amino acids in the plasma of the mentally affected children and the two adults with normal and borderline intelligence could be detected.

Discussion

Perry et al. (1970) found a low concentration of glutamine to be the only significant disturbance in plasma amino acid pattern in untreated PKU patients with defective mental development. Two phenylketonuric adults with normal and borderline intelligence did not have a low plasma concentration of glutamine.

In the family we have studied, the glutamine levels were in the normal range in all members except in the mother who presented the highest IQ. These observations indicate that in these cases a pathogenic effect of glutamine depletion on the brain function is difficult to accept. More cases with a similar familial structure must be studied to give a definite answer as to the damaging effect of glutamine depletion in PKU. * 

Summary

Plasma glutamine analyses were done in two severely retarded phenylketonuric sibs and in their mother and her brother, both of near normal intelligence.

The concentration of glutamine lay in the normal range, except in the subject with the highest IQ. These observations cast some doubt as to the damaging effect of CNS glutamine depletion in PKU.

References


*Since this paper was submitted, McKean and Peterson (1970) reported increased concentrations of glutamine in CSF and brain tissue of untreated retarded PKU subjects. Their findings also do not support the hypososis that glutamine depletion in CNS is responsible for cerebral abnormalities in PKU.

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Cephalhaematomata with Disseminated Intravascular Coagulation

Disseminated intravascular coagulation may be associated with many illnesses in the newborn period, such as viral infections (Hathaway, Mull, and Pechet, 1969) and the respiratory distress syndrome (Du, Briggs, and Young, 1970; Stark, Abramson, and Erkan, 1968).

It also occurs with pregnancy complications in the mother, such as pre-eclamptic toxaemia (PET) (Leissring and Vorlicky, 1968) and antepartum haemorrhage (APH) (Edson et al., 1968). Few cases have survived. The case described by Berglund (1970) was treated successfully however with heparin. We have encountered two cases of disseminated intravascular coagulation in the newborn who survived without specific treatment. They were unusual in that they presented with bilateral subperiosteal cephalhaematomata within 24 hours of birth.

Case Reports

Case I. A girl, birthweight 2.8 kg, was born at term after a surgical induction of labour. The mother was a 22-year-old gravida I whose pregnancy had been uneventful. The delivery was spontaneous by the vaginal route. At 1 minute the baby had an Apgar score of 1 and at 5 minutes an Apgar score of 5. Resuscitation was performed by suction, and oxygen was given via the Cardif bag. She was admitted to the Special Care Unit at 30 minutes of age with a rectal temperature of 36 °C. She was given 2 mg vitamin K on admission. She was wasted but was otherwise healthy, not juddery, and the anterior fontanelle tension was considered normal. At 15 hours of age when bilateral subperiosteal cephalhaematomata were noted, coagulation studies were performed (Table 1). The question of heparin therapy was discussed, but decided against in view of the fact that the child was clinically very well. She was eventually discharged at 2 weeks of age. At follow-up, development seems completely normal.
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Resuscitation included nasopharyngeal suction, intubation, and positive pressure ventilation. At 20 minutes of age he was admitted to the Special Care Unit with a rectal temperature of 36 °C and given 2 mg vitamin K intramuscularly. On examination gross joint abnormalities were noted, and a diagnosis was made of arthrogryposis multiplex congenita. There were bilateral fractures of humeri with radiological evidence of osteopatathyrosis. At 12 hours of age, bilateral subperiosteal cephalhaematoma appeared and coagulation tests were performed. Initially he appeared well and no further therapy was given other than the 2 mg vitamin K given on admission to the Unit. However, at 18 hours of age he was pale and rather shocked. The haemoglobin was 7·6 g/100 ml, PCV 24%. In view of the low haemoglobin 40 ml fresh blood was transfused. The haemoglobin rose to 10·3 g/100 ml. Progress thereafter was uneventful.

**Discussion**

Cases of disseminated intravascular coagulation in the newborn period have proved fatal almost invariably. However, Berglund (1970) recently described a case in association with a marked degree of birth asphyxia which was treated with heparin and survived. Cephalhaematoma are common in the newborn, but it is unusual for them to present on the first day of life. In both our cases, gross coagulation defects were found without haemorrhage other than the cephalhaematoma.

The coagulation changes found were those of a consumption coagulopathy with greatly depressed levels of platelets, fibrinogen, and plasminogen and raised levels of fibrin degradation products. The thrombin clotting times were prolonged, suggesting either low levels of fibrinogen or raised levels of fibrin degradation products in the circulation acting as an anticoagulant by interfering with polymerization of the fibrin clot (Bang, 1963).

Since clinically both babies were well and had no other manifestation of haemorrhage, heparin was not given. However, Case 2 did require a blood transfusion, in view of the fall in haemoglobin value over several hours.

Hypoxia as a result of birth asphyxia is presumed to have been the initiating cause of disseminated intravascular coagulation in both cases. This has been reported by Abildgaard (1969). The condition may have been maintained by acidosis (Case 1) which is known to potentiate disseminated intravascular coagulation (Von Kaula and Swan, 1958).

It is possible that many cases of cephalhaematoma are manifestations of disseminated intravascular coagulation, since they are found without undue trauma but often after a hypoxic episode at birth.

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**TABLE I**

Coagulation Studies on Case 1

<table>
<thead>
<tr>
<th>Coagulation Values for Normal Newborn in this Laboratory Given in Brackets</th>
<th>15 Hours Age</th>
<th>36 Hours Age</th>
<th>54 Hours Age</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>pH</strong></td>
<td>7·18</td>
<td>7·3</td>
<td>—</td>
</tr>
<tr>
<td><strong>Base deficit</strong></td>
<td>+13</td>
<td>+6</td>
<td>—</td>
</tr>
<tr>
<td><strong>Thrombotest (5-20%)</strong></td>
<td>22,000</td>
<td>35,000</td>
<td>50,000</td>
</tr>
<tr>
<td><strong>Platelets/mm³</strong></td>
<td>10%</td>
<td>80%</td>
<td>80%</td>
</tr>
<tr>
<td><strong>Fibrin degradation products (Burroughs Wellcome method)</strong></td>
<td>40·32 μg/ml</td>
<td>20·16 μg/ml</td>
<td>10·08 μg/ml</td>
</tr>
<tr>
<td><strong>Thrombin clotting time; as ratio (1·0-1·3)</strong></td>
<td>360·15/24</td>
<td>35·12 = 2·9</td>
<td>28·12 = 2·3</td>
</tr>
<tr>
<td><strong>Kaolin cephalin clotting time; as ratio (0·9-1·3)</strong></td>
<td>15/33 = 0·4</td>
<td>63/53 = 1·1</td>
<td>55/33 = 1·66</td>
</tr>
<tr>
<td><strong>Plasminogen (immunological method (3-38 units) 1-3 units)</strong></td>
<td>0-66 units</td>
<td>0-66 units</td>
<td>0-66 units</td>
</tr>
<tr>
<td><strong>Fibrinogen (immunological method) (90-240 mg/100 ml)</strong></td>
<td>27·1 mg/100 ml</td>
<td>27·1 mg/100 ml</td>
<td>—</td>
</tr>
</tbody>
</table>

**TABLE II**

Coagulation Studies on Case 2

<table>
<thead>
<tr>
<th>Coagulation Values for Normal Newborn in this Laboratory Given in Brackets</th>
<th>12 Hours Age</th>
<th>18 Hours Age</th>
<th>36 Hours Age</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>pH</strong></td>
<td>7·3</td>
<td>—</td>
<td>7·39</td>
</tr>
<tr>
<td><strong>Base deficit</strong></td>
<td>—2</td>
<td>—2</td>
<td>—2</td>
</tr>
<tr>
<td><strong>Haemoglobin</strong></td>
<td>15·39 g</td>
<td>7·6 g</td>
<td>10·39 g</td>
</tr>
<tr>
<td><strong>PCV</strong></td>
<td>49%</td>
<td>24%</td>
<td>70%</td>
</tr>
<tr>
<td><strong>Thrombotest (5-20%)</strong></td>
<td>42%</td>
<td>14%</td>
<td>—</td>
</tr>
<tr>
<td><strong>Platelets (phase)</strong></td>
<td>22,000</td>
<td>70,000</td>
<td>90,000</td>
</tr>
<tr>
<td><strong>Fibrin degradation products (Burroughs Wellcome method)</strong></td>
<td>40·32 μg/ml</td>
<td>40·32 μg/ml</td>
<td>20·16 μg/ml</td>
</tr>
<tr>
<td><strong>Thrombin clotting time; as ratio (1·0-1·3)</strong></td>
<td>29·15 = 2·06</td>
<td>29·15 = 2·06</td>
<td>29·15 = 2·06</td>
</tr>
<tr>
<td><strong>Kaolin cephalin clotting time; as ratio (0·9-1·3)</strong></td>
<td>180·51</td>
<td>104·51</td>
<td>40·35 = 1·1</td>
</tr>
<tr>
<td><strong>Plasminogen (immunological method) 3·38-11·3 units</strong></td>
<td>3·38 units</td>
<td>3·38 units</td>
<td>5·1 units</td>
</tr>
<tr>
<td><strong>Fibrinogen (immunological method) (90-240 mg/100 ml)</strong></td>
<td>21·1 mg/100 ml</td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>
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Our experience demonstrates that a conservative policy of management of disseminated intravascular coagulation can result in a satisfactory outcome and thereby avoid the hazards of heparin therapy.

Summary

Two babies who developed bilateral cephalo-haematoma within the first 24 hours of life had gross abnormalities of their coagulation mechanisms. The changes were those of a consumption coagulopathy.

Both infants survived and neither showed any other manifestation of haemorrhage.

We wish to acknowledge the technical assistance of Miss S. Muxworthy and Miss L. James and the considerable secretarial help given by Miss E. Morgan.

This work was carried out while one of us (M.A.C.) was in receipt of a grant from the Wellcome Trust.

References


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Intramural Jejunal Haematoma
After Peroral Mucosal Biopsy in a Child With Intestinal Malrotation

Intramural haematoma results from localized extravasation of blood into the wall of the oesophagus, duodenum, and small or large bowel. The lumen of the viscus is obliterated and most patients present with symptoms of partial or complete obstruction. The duodenum is most commonly affected and children, mostly boys, are affected more frequently than adults (Bailey and Akers, 1965; Stewart, Byrd, and Schuster, 1970). In the majority of cases there is a history of blunt external abdominal trauma which may often be quite trivial. The present case is recorded because of the unusual nature of the trauma from within by a peroral jejunal mucosal biopsy capsule. The case illustrates a hitherto unreported hazard of mucosal biopsy. An additional fact was the presence of intestinal malrotation.

Case Report

A girl, born at term weighing 3 kg, was first admitted to hospital aged 2 months with vomiting and failure to thrive. After investigation she was discharged home but was admitted frequently until the latest occasion, when she was 14 months old and weighed only 6-25 kg. There was now no vomiting but she had intermittent diarrhoea. On examination, apart from her overall poor development, the only positive findings were signs of pulmonary stenosis with facies suggestive of Noonan's (1968) syndrome (depressed nasal bridge, hypertelorism, micrognathia, and mild epicanticth folds).

Barium meal had been performed when she was 4 months old and showed evidence of malrotation of the intestines. There was, however, no hold-up of the barium and, as she never had any symptoms or signs of obstruction, surgical opinion was not sought. All other investigations, including those for malabsorption were negative.

It was decided to perform a jejunal mucosal biopsy, despite the lack of laboratory evidence of malabsorption. The paediatric modification of the Crosby capsule was used (Read et al., 1962). The capsule passed rapidly through the pylorus but there was a delay of about 15 minutes before it passed from the duodenum into the jejunum, its position being checked radiologically.

Biopsy was taken using 20 ml syringe suction and the capsule was withdrawn successfully.

About 48 hours after biopsy she became pyrexial and started vomiting copious amounts of bile. On abdominal examination there were numerous visible intestinal loops, and a mass in the right upper abdomen.

On opening the abdomen through a right upper paramedian incision, the proximal 12 cm of the jejunum was found to be the site of a large intramural haematoma which extended up to the duodenojejunal flexure. There was also intestinal malrotation with Ladd's bands and a large duodenum. There was a small Meckel's diverticulum. The jejunal haematoma ruptured during handling with complete dehiscence of the bowel and profuse bleeding, amounting to about 400 ml. Ladd's operation was performed and the involved segment of the jejunum was resected with end-to-end duodenojejunal anastomosis. Postoperative recovery was uneventful but there has been little improvement in the child's general condition since the operation. Examination of the biopsy and the resected specimens did not reveal any abnormality or reason for the complicating haematoma.