of labour and normal delivery occurred at 38 weeks. The birthweight of the first twin was 2720 g, and the Apgar score 9 at one minute. The second twin was severely macerated and autolysed. There was a single placenta. The cord to the first twin contained three vessels and was centrally inserted. The cord to the macerated twin was inserted marginally.

Intestinal obstruction on the first day of life in the surviving twin led to the discovery of ileal atresia which was treated surgically. Poor feeding, irritability, and grossly delayed development led to admission to hospital at 4 months of age. The occipito-frontal head circumference was 43.8 cm, on the 97th centile. The fundi were normal. Skull transillumination showed left hydranencephaly. Computed tomography showed that the left cerebral hemisphere had been replaced by fluid, with disorganised and abnormal development of the ventricular system on the right. The lymphocyte karyotype was normal and metabolic screening and a search for evidence of intrauterine infection were negative. At the age of 5 years he is severely handicapped and functioning at the level of a 6 week old child.

Discussion

The unusual feature of this child is the ability to be almost certain about the pathogenesis of his unique combination of hydranencephaly and ileal atresia. It is likely that death of a monozygotic co-twin caused release of thrombotic debris into the circulation of the surviving twin, leading to both the cerebral and intestinal defects.

Even excluding conjoined twins, there is an excess of congenital malformations in monozygotic twins compared with dizygotic twins or singletons.⁵ Either

the monozygotic twinning and the malformation share a common aetiology, or the malformation is a consequence of the twinning process. Death of a twin causing embolisation and vascular disruption of a co-twin is thought to cause hydranencephaly, porencephaly, microcephaly, intestinal atresia, congenital skin defects, and limb amputation.⁵ Vascular disruption due to various reasons is increasingly being recognised as a cause of congenital malformations.⁶ If a macerated or autolysed twin is found at birth, the paediatrician should be alerted to the possibility of serious and not immediately obvious defects in the surviving co-twin.

This child was shown at a clinical meeting of the Section of Paediatrics, Royal Society of Medicine on 28 November 1980. I thank Dr D Donnai and Dr R W Newton for their help, and Miss Michelle Turner for typing the manuscript.

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Treatment of severe coagulation disturbances as a condition of improved prognosis in fulminant liver failure

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SUMMARY A favourable outcome of fulminant liver failure by replacement treatment with antithrombin III, prothrombin concentrate, fresh frozen plasma, and plasminogen is reported. Haemostatic balance was maintained and no severe bleeding complications occurred during the 15 days of disturbed liver function.

The mortality rate for patients with coma after hepatic necrosis is still between 65 and 90%.^{1 2} There is some evidence that the coagulation disturbances observed in the course of fulminant hepatic failure may act as an intermediary in the pathogenesis of the hepatic necrosis and that rapid correction of haemorrhagic diathesis may lead to an improved prognosis.^{3 4} We report our treatment of a child with acute hepatic coma of unknown origin.

Case report

A 14 year old boy was admitted to hospital after a 24 hour illness characterised by spontaneous vomiting and drowsiness. There was no history for drug abuse or toxins. Physical examination showed a moderately ill boy with deep jaundice. The liver was palpable 4 cm below the right costal margin, no foetor hepaticus was noted. His mental state alternated between stupor, disorientation, and wild excitation. Laboratory studies yielded the following results: total bilirubin 131.6 $\mu\text{mol/l}$ (7.7 mg/100 ml); direct bilirubin 95.7 $\mu\text{mol/l}$ (5.6 mg/100 ml); serum glutamic oxaloacetic transaminase 3872 U/l; serum glutamic pyruvic transaminase 2160 U/l; γ glutamyltransferase 35 U/l, alkaline phosphatase 872 U/l; total serum protein 49 g/l; cholinesterase 4.7 KAU/l; and ammonia 259 $\mu\text{mol/l}$.

The results of initial coagulation studies were: prothrombin time 14%, partial thromboplastin time 66 seconds, thrombin time 29 seconds, fibrinogen 320 mg/100 ml; factor II 88%, factor V 54%, factor VII 1%, factor VIII 100%, factor IX 20%, antithrombin III 47%, plasminogen 30%, α_2 -antiplasmin 43%, α_2 -antiplasmin-plasmin-complex positive, fibrinogen split products substantively increased to greater than 100 $\mu\text{g/ml}$; platelet count decreased to a minimum of $88 \times 10^9/\text{l}$ on the second day after admission to hospital.

Urinary output was reduced to 53 ml/24 hours and his serum creatinine concentration was initially raised at 450 $\mu\text{mol/l}$ (5.1 mg/100 ml) with a subsequent rise to a maximum of 1260 $\mu\text{mol/l}$ (14.3 mg/100 ml).

Antibody studies, including hepatitis-, cytomegalo-, herpes-, coxsackie-, adeno-, reo-, and Epstein-Barr virus infections were entirely negative on repeated determination.

The clinical course and selected laboratory data are summarised in the Figure. Within 12 hours of hospital admission, the patient became comatose and required mechanical ventilation. Replacement of antithrombin III, plasminogen, and procoagulants was begun immediately after diagnosis and heparin was given simultaneously. In spite of the initial fluctuation in prothrombin time, haemostatic balance was maintained during the subsequent 15 days of disturbed liver function and no severe bleeding complications occurred.

Due to rapid deterioration of renal function, haemodialysis was necessary between days 4 and 15. As liver function recovered diuresis increased gradually. The patient was awake and responsive for the first time on day 19 and was discharged home three months after admission without any obvious hepatic, renal, or neurological disturbances.

Percutaneous liver biopsy performed after hepatic function and coagulation disturbances resolved showed focally accentuated portal round cell infiltration without fibrosis. No fatty degeneration, but moderate regenerative activity of liver cells was noted. Hepatitis B antigen was absent in immunopathological examination.

Discussion

Efforts to reduce the high mortality from fulminant liver failure have included procedures such as peritoneal dialysis, haemodialysis, plasmapheresis haemoperfusion, exchange transfusion, heterologous liver perfusion, and cross circulation. In paediatric patients there is wide experience only with exchange transfusion. The data on survival rates are conflicting,^{1 5} however, in a controlled study in adults, exchange transfusion was found to be of no benefit.⁶

Various reports emphasise the importance of supportive treatment in hepatic failure, and there is some evidence in adults that the prevention of bleeding complications, in particular, may improve the prognosis.²⁻⁴

In fulminant liver failure, a rapidly decreasing activity of coagulation factors and inhibitors, particularly of antithrombin III, has been suggested by two mechanisms: increased consumption in intravascular coagulation, stimulated by the necrosis of hepatocytes and impaired hepatic synthesis.³ The developing thrombotic haemorrhagic syndrome results in an ischaemic damage of liver tissue after thrombotic occlusion of the hepatic microcirculation.

Similar mechanisms—increased turnover and impaired hepatic synthesis—have been suggested concerning the lack of factors and inhibitors of the fibrinolytic system.³ In agreement with this pathophysiological concept, our coagulation studies showed simultaneously features of intravascular coagulation, hyperfibrinolysis, and impaired production of factors and inhibitors of coagulation and fibrinolysis.

Encouraging results in the treatment of acute liver failure have been reported since 1979 by Egbring and Vogel.^{3 4} Similar to our treatment, the regimen of these authors consisted of a rapid elevation of inhibitor values by replacement of antithrombin III-concentrate and subsequent low dose heparin treatment. Replacement of the coagulation factors II, VII, IX, and X was achieved by prothrombin concentrate, factor V, and other inhibitors like α_2 -antiplasmin were replaced by fresh frozen plasma. Impaired production and increased consumption of fibrinolytic activity were corrected by plasminogen concentrate. Five adult patients with fulminant

hepatic necrosis treated consecutively according to this regimen at the Centre of Internal Medicine, University of Marburg, survived, and complete functional recovery was achieved.

It is concluded that the effective control of coagulation disturbances in fulminant hepatic failure may improve the prognosis. The rapid and sustained replacement treatment may prevent secondary damage of hepatic function as a result of thrombotic occlusion of the microcirculation affecting the regenerative capacity of hepatocytes. The nearly normal liver biopsy in our patient seems to confirm the efficacy of the treatment.

It must be stressed that only patients treated early in the course of the disease may benefit from replacement treatment; at a certain stage damage of the liver and of other organs as well as the associated coagulation disturbances may become irreversible.

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Ultrasound monitoring of diaphragm activity in bilateral diaphragmatic paralysis

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SUMMARY Recovery of diaphragm activity after bilateral diaphragmatic paralysis was monitored in a term infant using a mechanical sector scanner fitted with a 5 MHz transducer. The ratio of diaphragmatic excursion during spontaneous breathing and ventilator assistance was used as an objective measure for comparison of diaphragmatic activity during recovery. Ultrasound assessment of diaphragm contraction may be used to study progress in diaphragmatic paralysis.

Bilateral diaphragmatic paralysis secondary to birth trauma is a rare occurrence¹ and most of these babies require ventilatory assistance soon after birth.² Assessment of diaphragmatic recovery clinically or by fluoroscopy becomes difficult once ventilatory assistance has begun. We describe an infant with bilateral diaphragmatic paralysis whose recovery was monitored ultrasonically.

Case report

A boy weighing 2650 g was born at term to a 25 year old mother after an uneventful pregnancy and the spontaneous onset of labour. Fetal bradycardia was

noted during the second stage of labour and delivery was expedited using Keilland's forceps. Apgar scores were 3 and 4 at one and five minutes respectively. He was intubated at one minute and ventilated with 100% oxygen. Soon after ventilation began his pH was 7.04 with a base deficit of 19 mmol/l. On examination, the infant was hypotonic with no spontaneous movements below the neck. Spontaneous movement of the legs was seen on day 2 and this gradually extended cranially, partial recovery of the deltoid having occurred by day 7. Inadequate, jerky, inspiratory efforts were first seen on day 12. Diaphragmatic activity was studied on several occasions using real time ultrasound.

Ultrasound images of the right hemidiaphragm were obtained using a mechanical sector scanner fitted with a 5 MHz transducer (Honeywell Ultramager). The transducer was placed below the right costal margin, and with the liver providing an acoustic window, sagittal views were obtained of the posterior portion of the diaphragm until maximum diaphragmatic excursion was observed (Fig. 1). The transducer was held in this position and the movement of the right hemidiaphragm was recorded as an M mode tracing during intermittent positive pressure ventilation and spontaneous breathing. A positive end expiratory pressure of 5 cm was provided at