worth (1970) has also been reported by Chadd (1970).

The cause of the intravascular coagulation occurring with hypothermia was suggested by Johansson and Nilsson (1964) to be stagnation of blood, disintegration of red blood cells, and platelets releasing thromboplastin and initiating coagulation. In the present series of hypothermic infants there was a marked thrombocytopenia and coagulation changes of a consumption coagulopathy which was not seen in the control infants whose rectal temperature did not fall below 34 °C. This suggests that the babies may react in a similar way to the animals of Johansson and Nilsson (1964) on exposure to cold. Though the admission rate to the Special Care Unit of our hospital has increased, the number of babies admitted who have a rectal temperature of 34 °C or less during the first day of life has dropped by more than 70% over the past 5 years. Deaths of babies with a rectal temperature of 34 °C or less are similarly reduced. These findings are attributed to the introduction of specific measures to maintain babies' body temperature during procedures such as resuscitation and exchange transfusion.

There are two explanations for the association of cerebral haemorrhage and hypothermia. It is possible that the cerebral haemorrhage is responsible for shock and hypothermia or that the haemorrhage follows a consumption coagulopathy induced by hypothermia. We favour the latter explanation, for hypothermia occurred shortly after birth, usually within 6 hours, and the work of Dyer *et al.* (1971) suggests that intraventricular haemorrhage is likely to occur after 6 hours.

Summary

Nineteen infants with a rectal temperature of 34 °C or less during the first hours of life were studied. They have been matched for age, gestation, and birthweight with 19 healthy babies whose rectal temperatures never fell below 35 °C on the first days of life. Comparison of the coagulation status of the two groups of infants shows a marked deleterious effect of cold. The aetiology of the coagulation defect has been ascribed to disseminated intravascular coagulation. Such a series is likely to be unique since there are widespread measures throughout this country to prevent the occurrence of hypothermia in the newborn baby.

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Purpura Necrotica as a Complication of Ventriculoatrial Shunts in Hydrocephalus

Purpura necrotica (or fulminans) is a rare complication of many acute infectious diseases of children, and has been reported after such conditions as vaccination, varicella, measles, streptococcal infection, and neisserial infection. It is characterized by the sudden appearance of symmetrical areas of cutaneous haemorrhage and superficial necrosis most pronounced on the lower extremities, anaemia, and severe systemic symptoms. Until a decade ago mortality in this condition was of the order of 90% (Charkes, 1961), but recently with better understanding of some of the underlying vascular and coagulation abnormalities, cures have

FIG. 1.—Case 2, (a) symmetrical facial lesions, and (b) the lesion above left knee.

been reported using heparin (Little, 1959), dextran (Patterson *et al.*, 1965), aminocaproic acid (Morse, Rowe, and Hartigan, 1966), and steroids.

Death usually occurs within 72 hours and many survivors have required amputation of gangrenous extremities. Paravertebral block has been advocated to improve peripheral blood flow and has avoided the necessity for amputation (Benoit, Raymond, and Glorieux, 1969). Changes similar to the Schwartzman phenomenon have been described, and the pathological stages seen in this reaction now form the basis for treatment (Bouhasin, 1964).

This report describes 2 children with typical lesions and infected ventriculoatrial shunts, in whom the course was much less acute than is usual, and who recovered fully.

Case Reports

Case 1. A boy aged 21 months. He had been born in December 1966, after a normal pregnancy. He had a lumbar meningocele which was closed shortly after birth. A Spitz-Holter valve was inserted for progressive hydrocephalus at the age of 2 weeks and thereafter his course was fairly satisfactory until admission to this hospital in July 1968. Four days before admission he developed pinpoint lesions on the left shoulder, left hip, and right thigh, which increased in size to 2 cm purpuric areas with central necrosis and induration and surrounding erythema. He was pyrexial (37.5°C), with enlarged lymph nodes in the neck and groin, and signs of an upper respiratory infection. The valve appeared to be functioning well and there was no neck rigidity. Hb was $8.4\text{ g}/100\text{ ml}$ with a slightly hypochromic picture; leucocytes $4380/\text{mm}^3$; platelets $53,000/\text{mm}^3$.

Clotting studies revealed a whole blood clotting time of 24 min (Lee and White method), with poor clot

retraction, a euglobulin lysis time of $>180\text{ min}$ (normal $90\text{--}180\text{ min}$), and a plasma fibrinogen of $200\text{ mg}/100\text{ ml}$.

These findings were considered to indicate consumptive coagulopathy. Blood culture produced a growth of *Staph. albus*, a course of penicillin was instituted, and the temperature fell to normal within 36 hours. Three days after admission he developed further similar lesions on the right cheek and right shin, and was given dextrose solution intravenously with 8000 U heparin/24 hours modified subsequently to 5000 U after estimation of clotting time. Steroids were also given in view of the low initial platelet count. A fresh blood transfusion raised his Hb to $13.5\text{ g}/100\text{ ml}$. His general condition improved despite a further crop of lesions and the heparin drip was discontinued after 3 weeks. The steroids were gradually replaced with ACTH which was finally stopped after 10 weeks, by which time his platelet count and clotting mechanism had been normal for 4 weeks. He was discharged and had no recurrence of purpura up to his death from meningitis and septicaemia at the age of 28 months.

Case 2. A girl aged 15 months. She had been born in March 1970, at 29 weeks' gestation, and had had an uneventful neonatal course. At the age of 6 months a presumptive diagnosis of hydrocephalus secondary to aqueductal stenosis was made, and a Spitz-Holter valve inserted. Apart from delay in gross motor development her progress was good until 6 weeks before admission to hospital in June 1971, when she developed purpuric lesions of both cheeks and the left knee. These had initially been attributed to trauma and had apparently varied in size over this period. There was no constitutional upset.

On admission the lesions were irregular in shape with haemorrhagic indurated centres and surrounding erythema (Fig.). The centre of the area on the right cheek subsequently underwent necrosis and sloughing, while that on the left knee became bullous.

Hb $9.5\text{ g}/100\text{ ml}$, leucocytes $9380/\text{mm}^3$, platelets

152,000/mm³, whole blood clotting time 4½ min, clot retraction absent. One-stage prothrombin time 16.3 sec, control 15.5 sec; euglobulin lysis time 58 min (normal <180 min). Thrombin clotting time 24.0 sec (normal 17–23 sec). Plasma fibrinogen: 275 mg/100 ml. Plasminogen: 1.17 Sherry units/ml per hr (normal 2–5 units).

These findings were interpreted as indicative of a chronic intermittent type of disseminated intravascular coagulation with secondary activation of the fibrinolytic enzyme system. The clot retraction remained absent on two further occasions despite normal platelet counts and low normal plasma fibrinogen levels.

Blood cultures revealed infection with *Staph. albus*, and penicillin in high dosage was started, being later changed to erythromycin after a possible hypersensitivity reaction. On the 6th day after admission she developed a further purpuric area on the right knee but as repeat clotting studies showed no deterioration it was decided to remove the Spitz-Holter valve rather than institute anticoagulant therapy. Removal was carried out 3 days later and the valve showed no evidence of fibrin deposition, but the same organism as that obtained on the blood culture was grown from it.

Thereafter all the lesions disappeared, the necrotic area on the right cheek healed with minimal scarring, and she remains well with no excessive increase in head circumference.

Discussion

The term *purpura necrotica* has been used in preference to the more usual *purpura fulminans* as the latter implies the rapidly advancing, highly fatal complication of infectious disease which neither case showed.

The aetiology of the increased coagulation and defibrination which must have been present at some stage is not clear, and the absence of clot retraction in the face of normal platelets and plasma fibrinogen in the second case is unexplained at present. (Estimation of fibrin degradation products would have provided more information about the underlying mechanisms responsible for these lesions.)

Emery (1964) has suggested that 5 to 10% of children may produce small clots by a reaction between CSF and blood, but the valve removed from Case 2 showed no evidence of fibrin deposition and Case 1 had no further episodes suggestive of defibrination.

The rather low-grade type of infection produced by *Staph. albus* may have accounted for the protracted, remittent course seen in the second child. I have been unable to find any reports in the literature of typical lesions of purpura necrotica occurring in association with either ventriculoatrial shunts or *Staph. albus* infection.

Summary

Two cases of purpura necrotica occurring in association with *Staph. albus* infection of Spitz-Holter valves are described. Cure was effected in both children by means of anticoagulants in one and removal of the infected valve in the other.

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Controlled Measures of Exploratory Movement in a Coeliac Child During Gluten Withdrawal

The clinical picture of untreated coeliac disease in young children is well defined, yet a feature of this disorder omitted in contemporary reports, but noted by earlier authors (Gee, 1888; Gibbons, 1889), is a marked retardation of normal motor activity. Though reduced motor activity may be a common and nonspecific manifestation of ill health in childhood, we believe that a reduction of exploratory movement, responding after a few days to gluten withdrawal, is a characteristic feature of coeliac disease.

As an overall improvement in the clinical picture occurs a few days after gluten withdrawal, we were interested in studying the relation of dietary gluten to motor activity in coeliac disease.

This paper reports the results of measuring total body movement during exploratory play in a