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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REFERENCES

- Benoit, P., Raymond, R., and Glorieux, F. H. (1969). Purpura fulminans. *Canadian Medical Association Journal*, **101**, 42.
- Bouhasin, J. D. (1964). Purpura fulminans. *Pediatrics*, **34**, 264.
- Charles, N. D. (1961). Purpuric chickenpox. *Annals of Internal Medicine*, **54**, 745.
- Emery, J. L. (1964). Fibrin and thrombosis in the central nervous system in children with particular reference to congenital hydrocephalus. *Journal of Clinical Pathology*, **17**, 348.
- Little, J. R. (1959). Purpura fulminans treated successfully with anticoagulation: report of a case. *Journal of the American Medical Association*, **169**, 36.
- Morse, T. S., Rowe, M. I., and Hartigan, M. (1966). Purpura fulminans. *Archives of Surgery*, **93**, 268.
- Patterson, J. H., Pierce, R. B., Amerson, J. R., and Watkins, W. L. (1965). Dextran therapy of purpura fulminans. *New England Journal of Medicine*, **273**, 734.

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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

FIG. 1.—The flow diagram of the two-subject sensitive floor, showing the encoder, the data punch, and the tape processing sequence.

coeliac child and his dizygotic twin, over the initial period of therapeutic withdrawal of gluten.

Case Report

Our subjects were dizygotic twins of different sex, who were initially bottle fed and weaned onto cereals at 6 weeks of age. They continued to develop normally, but at 8 months the male twin (M.C.) was admitted to hospital with an acute diarrhoeal illness. After recovery he developed recurrent upper respiratory tract infections and failed to thrive. At 1 year he was admitted to the Birmingham Children's Hospital with a history of passing frequent stools. He weighed 6.5 kg compared to his normal twin (W.C.) who weighed 9.5 kg. On examination he was miserable and hypotonic, with a distended abdomen and wasting of the glutei.

The diagnosis of coeliac disease was confirmed by jejunal biopsy which showed flat villi. Treatment with a gluten-free diet was begun and he responded well, gaining 1 kg in 3 weeks. He has been followed up at regular intervals for a year and continues to make excellent progress on his diet.

Materials and Methods

The instrument (MacCulloch, Birtles, and Bond, 1969), a free space-time traversal data logging system, was installed in a normal child-observation playroom. It consisted of 156 plywood tiles, each 1 foot square, giving a floor area of approximately 17 ft by 11 ft (Fig. 1). The tiles were covered by a vinyl carpet and insulated from below by a rubberfoam underlay. Each plywood tile had weight and magnetic field sensitive

switches, arranged so that the movement of two subjects, one wearing magnetic shoes and the other wearing ordinary shoes, could be recorded and differentiated by an electronic encoder. This encoder was designed to sample signals from the floor every $\frac{1}{8}$ th second and to encode this data onto an 8-channel punched paper tape. The tape was later decoded by an ICLKD F9 computer and X/Y co-ordinates determined for the two subjects every $\frac{1}{8}$ th second. It was possible to compute a two-dimensional position on the floor at any given instant for each subject, and also the distance each subject moved along a given path.

The experimental procedure was carefully explained to the patient's mother who wore magnetic plastic sandals and sat in a corner of the playroom. Each child was placed in turn in the centre of the floor for 15 minutes, during which time the other child was held by the mother. On each occasion the initial position of the child, and of two toys on the floor, was kept constant. Lighting conditions, temperature, and time of day were also controlled during the test period. This procedure was repeated daily for 4 days before the withdrawal of gluten in the coeliac child and on 16 occasions thereafter over the next 100 days. Whenever possible the normal female twin was used as a control. The children's behaviour during the test was observed through one-way glass and a detailed description dictated into a tape recorder.

Results

The computer printout showed the average distance travelled during consecutive 10-second

FIG. 2.—The distance covered on separate occasions by the recovering coeliac child and his twin.

intervals and the cumulative distance travelled by each subject.

Each subject was studied for 15 minutes at each session. Because of the large amount of data produced, we decided to limit our preliminary analyses to the average distance travelled during consecutive 10-second intervals for the first 50 seconds only. Our results are expressed as the average distance travelled by each subject in feet per second. The results are shown (Fig. 2).

The hypothesis to be tested is that withdrawal of gluten increases exploration by the coeliac child and therefore reduces the difference in exploratory behaviour between the normal and the coeliac subjects, as indexed by distance travelled per session. That is to say, that time after gluten withdrawal and the difference between the movements of the two subjects are negatively correlated. To test this hypothesis, Spearman's method of rank correlation was applied to the data in Fig. 1 and indicated that the hypothesis might be accepted at the 0.05 level of significance.

Discussion

Clinical observation suggests that altered motor activity plays an important part in the clinical picture of the children with untreated coeliac disease. In our experiment the retardation of motor activity was shown to be severe in the untreated patient (M.C.) and very little movement was recorded initially on three out of four occasions. 6 days after the start of a gluten-free diet an increase in motor activity was noted and this continued until our last recording. Even after 102 days of treatment the coeliac child had still failed to catch

up with his normal twin, indicating that recovery in this condition may be prolonged, in spite of an initially rapid response. Though the analysis of each child's movements reveal considerable day-to-day variation, the trend of our data is in line with clinical experience.

It has been suggested that part of the symptomatology in coeliac disease could be due to the absorption of various substances, such as alkaloids (Gibbons, 1889), peptides (Sheldon, 1959), or amines (Challacombe, Sandler, and Southgate, 1971), having a specific effect on the central nervous system. As it is known that regeneration of intestinal epithelium begins within a few days of gluten withdrawal (Yardley *et al.*, 1962), our measurements of clinical improvement could be temporally related to the reconstitution of a protective enzymatic barrier in the small bowel. As epithelial recovery is reversible with the reintroduction of gluten, this method could be useful for the elucidation of pharmacological factors concerned with the control of motor activity.

Our results are as yet unsophisticated in terms of analysis of movement. However, we are producing programmes that will analyse the data from future cases in greater depth. It will then be possible to investigate more thoroughly the relation between altered motor function and coeliac disease and also to study other disorders of motor function in childhood.

Summary

A free space-time traversal data logging system has been used to measure total body movement during exploratory play in a child with coeliac

disease and his normal dizygotic twin. This study records the retardation of motor activity in untreated coeliac disease and the progressive increase in activity starting 6 days after gluten withdrawal. It is suggested that this improvement in motor activity could be directly related to the regeneration of intestinal epithelial cells. The use of this instrument in the study of other disorders of motor activity of childhood is also proposed.

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REFERENCES

- Challacombe, D. N., Sandler, M., and Southgate, J. (1971). Decreased duodenal monoamine oxidase activity in coeliac disease. *Archives of Disease in Childhood*, **46**, 213.
- Gee, S. (1888). On the coeliac affection. *St. Bartholomew's Hospital Reports*, **24**, 17.
- Gibbons, R. A. (1889). The coeliac affection in children. *Edinburgh Medical Journal*, **35**, 321 and 420.
- MacCulloch, M. J., Birtles, C. J., and Bond, S. (1969). A free space-time traversal data-logging system for two human subjects. *Medical and Biological Engineering*, **7**, 593.
- Sheldon, W. (1959). Celiac disease. *Pediatrics*, **23**, 132.
- Yardley, J. H., Bayless, T. M., Norton, J. H., and Hendrix, T. R. (1962). Celiac disease: a study of the jejunal epithelium before and after a gluten-free diet. *New England Journal of Medicine*, **267**, 1173.

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Spinal Cord Compression as a Complication of Haemophilia*

Neurological complications of haemophilia are more common than once thought. Because of the greater life expectancy with modern treatment methods, functional deficits of these complications assume increasing importance. Most previous reports have been concerned with intracerebral complications. Silverstein (1960) reviewed 174 cases of haemophilia and found 6 proven cases of intracranial bleeding and 5 probable cases. He found 28 peripheral nerve lesions in 206 haemophiliacs (Silverstein, 1964). There have been few reports of spinal cord bleed as a complication of

haemophilia. Aggeler and Lucia (1944) reviewed the literature up to 1944 regarding neurological complications of haemophilia. They found 45 cases reported with bleeding involving the central and peripheral nervous system. 11 patients with spinal cord involvement, including 3 quadriplegics, were reviewed. 6 cases had spinal cord bleed with involvement of the spinal meninges, including epidural, subdural, and subarachnoid bleeds. 5 cases had bleeds into the spinal cord. 4 died within several weeks of onset, 2 had partial cauda equina lesions with improvement or recovery, while the others had residual neurological deficits, or the course was unknown.

Three cases of spinal cord compression in haemophilia unassociated with cervical fracture or trauma have been reported in more detail. Schiller, Neligan, and Budtz-Olsen (1948) reported a case of subdural haematoma causing paraplegia in a 16-month-old haemophiliac. Laminectomy with clot removal was performed. Haemostasis was a problem postoperatively, but the neurological status improved significantly. Schenk (1963) studied the pathological findings in a 21-year-old haemophilic paraplegic who died from wound bleeding after laminectomy. Syringomyelia with glial proliferation was present from cervical to lumbar region, and the author speculated that this might have been a reaction to repeated bleeds. Fessey and Meynell (1966) reported complete C-6 quadriplegia in a patient with mild factor IX deficiency. The patient improved immediately after administration of fresh frozen plasma and went on to complete recovery within one week.

Case Report

A 6-year-old white boy with haemophilia (factor VIII less than 1%), relatively resistant to cryoprecipitate, had factor VIII levels of 11 to 14% after receiving 10 units of cryoprecipitate (normally this would approach 50% level). He was seen on numerous occasions for treatment of haemarthrosis and soft tissue bleeds. In August 1970, he fell, hitting the lumbosacral region of his back without immediate apparent injury. During the next several days he was febrile, lethargic, and remained in bed. He presented to the hospital with abdominal distension and obstipation. On examination on admission to the hospital, the patient had no response to pin below T2-3 area, but touch was appreciated. Abdominal breathing was present. No voluntary motion was observed in the lower extremities. Kneejerks were absent and ankle-jerks hypoactive. Plantar stimulation caused flexor withdrawal of the lower extremities. There was a question of weakness of the distal upper extremities. Admission haematocrit was 15, BUN 138, arterial P_{O_2} 46, P_{CO_2} 38. Spine and skull series were negative. Catheterization of the bladder

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