Tetrabenazine for Huntington's Chorea

SIR,—I read with interest Dr. M. A. Dalby's article¹ and Dr. R. Thavasoathy's letter (25 April, p. 237). I would like to report a further two patients suffering from Huntington's chorea who were treated with tetrabenazine (Nitoman). In both patients treatment resulted in marked diminution of involuntary movements. Both patients became depressed, and this responded to treatment with a monoamine oxidase inhibitor. During the period of observation the dementia continued to worsen.

A man aged 61 was first seen in January 1967 when he complained of jerky involuntary movements of all limbs for the past nine months (his wife said they had been present for three years) and some impairment of concentration. He had had a partial gastrectomy in 1947 for duodenal ulcer, and a left nephrectomy in 1961 for renal calculus. A sister who died of pneumonia at the age of 59 was said to have had Huntington's chorea. Its presence was not detected in either of his parents, but his father died at the age of 53.

On examination he had choreiform movements of the limbs and face and slight spasticity of the legs. A diagnosis of Huntington's chorea was made. The spasticity was attributed to cervical spondylosis. He was given supportive therapy.

In May 1969 he was admitted to hospital suffering from lower bowel obstruction due to faecal impaction. At this time the involuntary movements were noticeably worse, he was unable to walk unaided, had difficulty in swallowing, and his speech was unintelligible. A course of enemas relieved the faecal impaction, and treatment with tetrabenazine 50 mg. t.d.s. was started. Within 48 hours medical and nursing staff were convinced that the involuntary movements were diminished, although the patient did not accept this. When seen in outpatients two weeks later the movements had diminished considerably. He was able to pick up a cup for the first time in six months and could light his cigarette, but he had become very depressed. Tetrabenazine was continued and isocarboxazid (Marplan) 10 mg. t.d.s. was added. He continued to improve and when last seen, ten months after the start of treatment, he was able to dress himself, although with difficulty.

The second patient was observed by his wife to have abnormal movements in 1964, when he was 48 years old. There was no relevant family history. A sister died at the age of 26 from diabetes. Both parents died in old age. However, on the basis of choreiform movements and mild intellectual impairment a diagnosis of Huntington's chorea was made. Over the next five years the abnormal movements and mental impairment progressed relentlessly. By June 1969 he was making frequent flinging movements of the limbs and writhing movements of the trunk, his writing and speech were unintelligible, and he could walk only a few paces unaided. Treatment was started with tetrabenazine initially in a dose of 25 mg. t.d.s., then 50 mg t.d.s. Improvement was striking. Sixteen days after the start of treatment he could sign his name perfectly and the abnormal movements were diminished. He complained of feeling tired, and appeared depressed and more mentally retarded than usual. Treatment with tet-

rabenzazine was continued and isocarboxazid 10 mg. t.d.s. added. Two months after the start of treatment he no longer had to sit on his hands to keep them still, and appeared in reasonable spirits. He was able to walk unaided and his speech was clearer. This improvement was maintained ten months after the start of treatment, but subsequently some deterioration occurred. His dementia continued to progress.

He also suffered from late onset diabetes, which had been controlled since 1963 with diet and chlorpropamide. Control had begun to deteriorate before the tetrabenazine was commenced, and after failure of other oral hypoglycaemic agents, he was finally stabilized in February 1970 on lente insulin 30 units daily.

I would like to thank Dr. Neville Southwell for his advice and permission to study his patients, and also to Roche Products Ltd. for supplies of the drug Nitoman.—I am, etc.,

COLIN A. SOUTAR.

Orpington Hospital,
Orpington, Kent.

REFERENCE

- ¹ Dalby, M. A., *British Medical Journal*, 1969, 2, 422.

Benign Intracranial Hypertension

SIR,—I am struck by the disparity between the advice given in your leading article (5 September, p. 536) upon the radiological investigation of benign intracranial hypertension and the remarks made by Drs. B. G. R. Neville and J. Wilson in their article in the same issue (p. 554).

In their article Drs. Neville and Wilson state that "the risk of special neuroradiological studies outweighed the advantages." The leading article states that after skull radiographs "Contrast radiography is then needed. . ."

I am in full agreement with Drs. Neville and Wilson. This diagnosis, as stated in your leader, is one of exclusion of the causes of raised intracranial pressure. Angiography can do damage to an already oedematous brain. Needling what may be classically small and pinched ventricles is far from easy and again may be a traumatic procedure. Lumbar encephalography carries all the attendant risks in the presence of raised pressure and should not, in my view, be attempted without prior preparation with mannitol or urea. This will enhance the chances of ventricular filling. Isotope studies can be of great value in the differential diagnosis of raised intracranial pressure and should be the investigation of first choice.

It is of interest to note that in several cases seen in this hospital and at least one of Drs. Neville and Wilson's (Case 3) there has been no sutural diastasis in the presence of markedly raised intracranial pressure. The only reference I know to this fact is recorded by Kalbag and Woolf.¹—I am, etc.,

A. S. BLIGH.

Department of Radiology,
Royal Infirmary, Cardiff.

REFERENCE

- ¹ Kalbag, R. M., and Woolf, A. L., *Cerebral Venous Thrombosis*, p. 241. London, Oxford University Press, 1967.

Malignancy and Dermatitis

SIR,—Having read the reports of two cases of dermatitis herpetiformis, steatorrhea, and malignancy by Drs. E. Gjone and A. Nordoy (7 March, p. 610), I would like to report a case of dermatitis herpetiformis associated with carcinoma of the lung.

The patient, a man, was first diagnosed as suffering from dermatitis herpetiformis in 1948 when he was aged 34 and therapy with sulphapyridine was then commenced and continued until his death in 1969. In 1963, he suffered an uncomplicated myocardial infarction and was further investigated in 1965 for a vague complaint of generalized weakness, with a negative result.

He was re-admitted in November 1969 for investigation for a clinically detected pleural effusion. Haemoglobin 12.8 g./100 ml., M.C.H.C. 32, W.B.C. 10,800 (normal differential), urinalysis revealed glycosuria, but a subsequent glucose tolerance test was within normal limits; serum LDH 310 units. Chest x-ray confirmed the presence of rib metastases. The patient had not described gastrointestinal symptoms on any of his admissions. A small bowel biopsy was not performed.

Post-mortem examination confirmed the diagnosis of carcinoma of lung, which microscopically was seen to be a poorly differentiated round cell carcinoma of bronchus.

It may well be that dermatitis herpetiformis can be added to the list of skin lesions known to coexist with or precede lung cancer.—I am, etc.,

J. H. HORGAN.

Department of Medicine,
St. Laurence's Hospital,
Dublin.

Induction of Labour with Synthetic Oxytocin

SIR.—The article by Dr. M. E. Pawson and Mr. S. C. Simmons (25 July, p. 191) on the induction of labour prompts us to record a method of delivering intravenous synthetic oxytocin which we have been using for some time and have found most satisfactory.

The equipment needed is shown in the diagram. A 1-litre bottle of 5% dextrose, an 18 in. (45 cm.) long FG 6 suction catheter, and a 20 ml. syringe.

The catheter is fed well into the bottle down the glass airway. The solution of synthetic oxytocin is then mixed in the syringe (20 units with 18 ml. of dextrose), the syringe is connected to the catheter, and in turn taped to the side of the bottle (transparent tape has the advantage that the marks on the syringe can be seen through it). The infusion is then set up intravenously in the conventional manner and set running at 40 drops per minute.

Two units (2 ml.) are then injected from the charging syringe into the infusion bottle and the dose is increased by an increment of two units every 15 minutes until either satisfactory labour pains are induced or the full 20 units (20 ml.) have been added. Occasionally the syringe is recharged with 20 units of syntocinon in 20 ml. and the infusion strength increased even further. The advantages of this method are: