ureter was apparently normal; the pelvis and calices alone were dilated; the uretero-pelvic junction was well marked; and yet no cause of ureteric obstruction was found.

Jewett (1940), after careful study, showed that normally there is no absolute uretero-pelvic junction, the pelvis blending into the ureter in a funnel-shaped prolongation. He believed that when such a junction exists, as it did in the cases described here, stenosis should be considered, particularly if there is pyelectasis. The fact that he considered this stenosis often congenital, since there were no acquired factors to explain it, would make such a mechanism an exceedingly attractive explanation for the present series of familial cases. However, no evidence of stenosis was found. *Faute de mieux*, we must therefore fall back on such a conception as inadequacy of the ureter as an efficient urinary duct which has been posited as a causal factor in hydronephrosis; or we must postulate a functional defect in the neuromuscular mechanism of the renal pelvis, as the familial and inherited primary aetiological factor, though there is no positive evidence for either supposition. Consideration of the familial nature of these cases does, however, serve to show that, contrary to general opinion, a pelvic and caliceal hydronephrosis of this type can occur as a congenital condition without evidence of ureteric obstruction.

Summary

Four cases of non-calculous hydronephrosis occurring in two generations of a family are described. The literature is reviewed. The aetiology of the condition is discussed.

I am indebted to Mr. R. J. Rutherford, who was responsible for the surgical care of Case 1; to Mr. M. C. Pinkerton, who was similarly responsible for Cases 2, 3, and 4; and to Dr. C. H. Kitchen, who carried out the radiological examinations.

REFERENCES

- Aaron, G., and Robbins, M. A. (1948). *J. Urol.*, **60**, 702.
Blackwood, C. M. (1896). *Edinb. med. J.*, **41**, 919.
Jewett, H. J. (1940). *J. Urol. (Baltimore)*, **44**, 247.
Mackay, H. (1944). *Proc. roy. Soc. Med.*, **38**, 567.
Winsbury-White, H. P. (1925). *Brit. J. Surg.*, **13**, 247.
— (1951). *British Encyclopaedia of Medicine Practice*, 2nd ed., 7, 396. Butterworth, London.

UNUSUAL CASE OF JUVENILE G.P.I.

BY

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Juvenile general paralysis is commonly thought to manifest itself as a gradual dementing process similar to that seen in the adult disease, but with the differences in symptomatic conduct and behaviour disorder that the onset in childhood or the school years would produce. Thus intellectual deterioration may appear as a failure to ascend the normal educational ladder or as an unexpected loss of already acquired learning. Grandiose delusions, which are uncommon, may take the form of claiming ownership of vast stores of sweets or toys. Reduced social standards, such as minor dishonesty or lack of personal cleanliness and tidiness, often presenting symptoms in adults, are likely to be less readily noticed in the juvenile age group. This is the picture given in the standard textbooks of neurology and psychiatry, though Ford (1949) and others mention a maniacal reaction as an occasional presenting symptom.

The following case is worth recording because juvenile G.P.I. is now an uncommon disease and because the

method of presentation was unusual. The sudden development of a hemiplegia drew attention to the disease, and shortly thereafter the patient developed an acute mania.

Case History

A National Service man, aged 19, was admitted to Wheatley Military Hospital in July, 1953. On the previous day, while picking peas on leave at home, he had suddenly "collapsed" in a semicomatose state, from which he could be roused only by repeated shouted requests. He was able to obey simple commands, but his answers to questions were incoherent and unintelligible. On examination he proved to have a right hemiparesis with dysphasia and right homonymous hemianopsia. There were some continuous clonic movements of the right face and hand resembling epilepsy partialis continua. The pupils were large, irregular, and did not react to light. The optic fundi were normal, as were other bodily systems. The blood pressure was 140/90. The urine was free from albumin and sugar. Lumbar puncture revealed a clear fluid under normal pressure, with a protein of 70 mg. per 100 ml. and 25 white cells per c.mm. (90% lymphocytes, 10% polymorphs). His electroencephalogram showed high-voltage delta activity in both hemispheres but focusing on the left mid-parietal electrode.

During the next 48 hours his comatose condition gave place to an acute mania. He would laugh and talk continually, and as he was still dysphasic a stream of jargon, often shouted at the top of his voice, resulted. Despite his hemiparesis he made continual attempts to get out of bed, and would keep on neither bedclothes nor pyjamas. He would try to attack anyone approaching him, and when restrained in bed made continual restless movements of his non-paretic limbs. He was incontinent of urine and faeces, and had to be tube-fed. In view of a possible left frontal abscess or tumour, a carotid angiogram was done, but this showed no convincing abnormality. By this time, some five days after admission, a strongly positive W.R. in the blood and C.S.F. had been returned, and the Lange gold curve was 4555555321. The diagnosis of juvenile G.P.I. was thus suggested, and he was given penicillin, and in view of a possible gummatous process this was combined with potassium iodide. During three weeks he had 22 mega units of penicillin and 30 gr. (2 g.) of potassium iodide three times a day. On the mental side the response was dramatic. Within a week his acute mania had subsided and he would remain in bed and feed himself. He was continent of faeces, though he still occasionally wet himself.

Four weeks later he was up and helping with ward duties, though still rather emotionally labile. His neurological condition had also improved. The involuntary movements lessened gradually and ceased after some seven days, the hemianopsia and hemiparesis also cleared, though pyramidal signs—increased tendon-jerks and an extensor plantar response—remained. Some dysphasia also persisted, and there was slight proprioceptive sensory loss in the right arm and leg. The pupils remained large and irregular. There were no other signs of congenital syphilis. His electroencephalogram became more normal, with an 11 c.p.s. alpha rhythm in the right hemisphere, and some low-voltage theta activity from the anterior leads on the left. He proved to be entirely right-handed. Three months after the onset of his illness he was well enough to return home and was invalided from the Army.

As his serology remained positive he was treated with two further courses of 20 mega units of penicillin at the Queen Elizabeth Hospital, Birmingham. In March, 1954, his C.S.F. showed a protein of 42 mg. per 100 ml., with 1 lymphocyte per c.mm., Lange 3211000000, and a positive W.R. He was still emotionally labile, had occasional dysphasia and also evidence of left parietal lobe dysfunction, such as visual and tactile inattention to the right half of the body and a disinclination to use these limbs. However, he was leading a normal social life at home and was hoping to restart work in his previous trade.

Subsequent inquiries revealed that the patient had been to an ordinary school, and, although considered backward, had attended a junior technical college for a year, leaving at the age of 15. He had been employed as a factory hand and later as a storeman before his national service. On entering the Army at the age of 18, he had been regarded as rather dull and mentally immature, but no physical abnormalities were found. Three months before the illness here reported he had "collapsed" during a battle course done as part of his Army training, and a weakness of the right hand had been noted in examination. This had cleared in a few days, and he had been returned to duty but excused further battle courses.

His father, mother, and only sister each proved to have a positive W.R.

Discussion

The rarity of juvenile G.P.I. was emphasized by Worster-Drought (1940). This case also showed several uncommon features. The clinical onset was abrupt, and, although some degree of dementia may have been present for a time before the acute breakdown, it was not sufficient to prevent his wage-earning employment or his acceptance for national service. The presentation with focal neurological signs is unusual. However, such a syndrome in G.P.I. was recognized by Lissauer and Storch (1901), and these cases are sometimes referred to as "Lissauer's paralysis." Lissauer himself explained them as being due to an intense localized atrophy of a group of convolutions, but of the same pathogenesis as the more diffuse cerebral atrophy of G.P.I. More recently, however, Stewart (1933), in reviewing the pathology of juvenile cases, could find little to differentiate histologically between ordinary G.P.I. and Lissauer's type, as localized atrophy with status spongiosus occurred in both. In our case the abrupt onset of hemiparesis, the focal clonic movements, the localized electroencephalographic abnormality, and the rapid improvement with potassium iodide and penicillin, with slight residual focal signs, all suggest that a meningovascular process with gumma formation may have been present in addition to the G.P.I.; and, indeed, this may be the explanation for other cases of "Lissauer's paralysis."

A predominantly manic psychiatric picture is also unusual, though recorded in this condition. This patient's initial response to penicillin was very striking. His subsequent improvement has continued, though more slowly, and it can certainly be said that already, during the course of a year since the diagnosis was made, progress has been much better than with older remedies such as malaria, arsenic, and bismuth. Worster-Drought (1951) has used a combination of penicillin and malaria in treating 10 cases of juvenile G.P.I., with improvement in all cases. However, it is still a matter of debate whether such a combination has any advantages over penicillin alone, used, as in the acquired form of G.P.I., in large and repeated doses until the serology shows a definite response. Either way, treatment should be continued until such a response does occur.

We should like to thank Professor P. C. Cloake for the follow-up notes of the case, and the Director-General, Army Medical Services, for permission to publish.

REFERENCES

- Ford, F. R. (1949). *Diseases of the Nervous System in Infancy, Childhood, and Adolescence*. Thomas, Springfield, Illinois.
Lissauer, H., and Storch, W. (1901). *Msschr. Psychiat. Neurol.*, 9, 401.
Stewart, R. M. (1933). *J. ment. Sci.*, 79, 602.
Worster-Drought, C. (1940). *Neurosyphilis*. John Bale, Sons and Staples, London.
— (1951). *Modern Trends in Neurology*, edited by A. Feiling. Butterworth, London.

Spastic and other disabled children in hospitals and special schools are finding the use of magnetic letters both entertaining and instructive. Clinging to the surface of special blackboards, which can be held in any position, these letters are particularly helpful in the education of children who have to lie on their backs.

ANAPHYLACTIC SHOCK AS A COMPLICATION OF INSULIN COMA THERAPY

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Changes in insulin sensitivity are fairly common during insulin coma therapy and are shown by changes in the time and depth of coma, usually in the direction of increased sensitivity, requiring reduction of the original coma-producing dose. Insulin hypersensitivity (or allergy) is far less common and actual anaphylactic shock is extremely rare.

Case Report

A 40-year-old male paranoid schizophrenic was admitted to hospital on October 15, 1953. He had been in hospital in 1950 (when he had a three-weeks course of modified insulin) and in 1952. There was no personal or family history of allergy. He was in satisfactory physical health, and routine blood investigations were normal. After preliminary electroplexy, modified by relaxants, he began deep insulin therapy on October 22. This was administered by the routine technique modified by the use of intravenous insulin. Daily injections of insulin were given for six days a week, at first intramuscularly, beginning with 10 units, then 20 units, then increasing daily by 20 units, changing to intravenous injection at 60 units. Various brands of soluble insulin of 20 units per ml. strength were used in the treatment unit, but only one batch was in use at any particular time.

November 1 was a rest day, and on the 2nd an intravenous injection of 160 units of soluble insulin was given at the usual slow speed at 7.50 a.m. This was the tenth treatment, and the dose was the same as that administered on October 31, when only sweating had resulted, with termination of the hypoglycaemia by oral glucose at 10.30 a.m. No untoward reactions had previously been noted. On this occasion, however, five minutes after injection coughing and retching occurred. The manifestation of anaphylaxis was immediately suspected, and during the preparation of an adrenaline injection severe bronchospasm occurred with startling rapidity and severity, and the patient became pallid and cold. Intravenous adrenaline tartrate, 2 ml. of 1 in 1,000 solution, was given, and general treatment for shock was begun. Bronchospasm and vasomotor collapse remained very pronounced, and consecutive intravenous injections of 60 ml. of 33½% dextrose solution and 2 ml. of nikethamide were given, accompanied by continuous insufflation of oxygen (later changed to oxygen with 5% carbon dioxide). Slow intermittent intravenous injection of adrenaline was maintained, and between 8 and 8.35 a.m. 8 ml. of 1 in 1,000 solution was administered. The case proceeded as follows.

At 8.40 a.m. he was given atropine sulphate, 0.6 mg. (1/100 gr.), and aminophylline, 0.5 g., intramuscularly and 33½% dextrose, 60 ml., intravenously. He was now sufficiently recovered to ingest chlorcyclizine hydrochloride tablets ("histanin"), 100 mg., crushed in a little glucose solution. 9.5 a.m.: Blood sugar, 143 mg. per 100 ml. 9.10 a.m.: I.V. 33½% dextrose 60 ml., and normal saline 40 ml. 9.30 a.m.: B.P. 114/76; pulse rate, 130 a minute. 10.5 a.m.: B.P. 94/62; I.M. nikethamide 2 ml., and I.V. injection aminophylline 0.25 g.; general condition unchanged. 11 a.m.: B.P. 80/50; pulse rate, 80 a minute; blood sugar, 43 mg. per 100 ml. 11.15 a.m.: Patient suddenly became very collapsed, with dyspnoea and very weak pulse volume; I.V.