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Childhood organic neurological disease presenting as psychiatric disorder

T. M. RIVINUS,* D. L. JAMISON,† and P. J. GRAHAM

From the Departments of Neurology and Psychological Medicine, The Hospital for Sick Children, London

Rivinus, T. M., Jamison, D. L., and Graham, P. J. (1975). *Archives of Disease in Childhood*, **50**, 115. **Childhood organic neurological disease presenting as psychiatric disorder.** Over a period of one year 12 children with complaints which had been diagnosed as due to a psychiatric disorder presented to a paediatric neurological unit where neurological disease was diagnosed. The group was characterized by behavioural symptoms such as deteriorating school performance, visual loss, and postural disturbance, which are unusual in children attending child psychiatric departments. It is suggested that where there is diagnostic uncertainty the presence of these physical symptoms calls for periodic neurological reassessment, and attention is drawn to the rare but serious disorders which may thus be diagnosed. Making an organic diagnosis, however, should not preclude psychosocial management of emotional reactions in these families.

There are many situations in which, though a child complains of bodily symptoms, it would be inappropriate after adequate investigation to regard the problem as anything other than a disorder of emotional life. Environmental stress leads certain individuals to become unusually aware of normal body sensations. The physiological concomitants of anxiety may be experienced as uncomfortable bodily sensations. Loss of sensory or motor function, especially in the limbs, may occur as a morbid response to an otherwise intolerable internal conflict or external stress. A child's complaint of physical discomfort, especially pain, may represent a communication where other methods of calling for attention have failed. In all these situations a psychiatric approach aimed at understanding the life situation of the child and family will be more rewarding than any attempt to deal with the problem as a physical one with organic aetiology and needing physical treatment.

Yet the knowledge that physical symptoms occur, indeed occur frequently, as a response to stress without any sinister somatic implications may clearly lead us mistakenly to miss physical illness where it is present. This paper provides data on a number of children with neurological disorders whose complaints had previously been thought to be purely of a psychiatric nature.

The aims in reporting this study are first to prevent or at least reduce the likelihood of similar diagnostic errors in the future, and secondly to draw attention to a number of rare neurological conditions which general practitioners, school health doctors, child psychiatrists, and paediatricians might overlook.

Method

In the 12 months between July 1972 and June 1973, 12 children were admitted to the neurological wards of The Hospital for Sick Children who had previously been diagnosed as suffering from psychiatric disorder, but who on investigation were found to have neurological disorder. All of these were seen personally by either T.M.R. or D.L.J. In none of these children was there any reasonable doubt that the original symptoms of cognitive disability or behaviour and emotional disorder were due to the neurological disorder ultimately diagnosed.

Information on these children was obtained from hospital notes and from reports obtained from child psychiatrists and paediatricians who had seen them previously. The information was tabulated in terms of presenting symptoms, age at which these appeared, psychiatric diagnosis, the professional group of doctors making the psychiatric diagnosis, final neurological

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Present addresses: *The Children's Hospital Medical Center, 300 Longwood Avenue, Boston, Massachusetts, U.S.A.

†105 Barrack Road, Auckland 6, New Zealand.

diagnosis, and period of time between initial psychiatric and final neurological diagnoses.

Patients. Among the 12 children there were 7 girls and 5 boys. There are approximately 550 admissions per year to these wards, so that this group represented about 2% of the total. The age range of the 12 children at the time of presenting symptoms was between 4 years 5 months and 10 years, with a mean age of 6 years 9 months. A diagnosis of childhood schizophrenia or childhood psychosis had previously been made in 2 children, conduct or behaviour disorder in 2 children, emotional reaction or anxiety state in 4 children, and hysterical reaction in 2 children. A psychiatrist, usually a child psychiatrist, had been involved in making the psychiatric diagnosis in 10 cases, and an orthopaedic surgeon in 2 cases. Nearly all the children had seen other consultants, usually paediatricians who had either concurred in the psychiatric diagnosis or been responsible for making it in the first place.

The behavioural symptoms which the children showed and which led to a psychiatric diagnosis being made were various, but the two main types of symptoms present were deteriorating school performance in 6 of the 12 children, and disturbances of posture in 5. The remaining child had difficulty in seeing the blackboard at school. In a number of the children these symptoms (which are unusual among children seen in psychiatric departments) were not prominent and in many were overshadowed at the time of initial presentation by commoner psychiatric symptoms such as aggressive behaviour, temper tantrums, and symptoms of anxiety. Psychological testing had been carried out in only a few of the children and no characteristic pattern of test results had been noted. The psychiatric symptoms are given in Table I in relation to final neurological diagnosis.

Psychiatric treatment was initiated in most cases, and included the use of psychotropic medication, hypnosuggestion, and analytically orientated psychotherapy. Many of the children were living in stressful circumstances and various measures including parental guidance, casework with parents, and environmental alterations were undertaken in an attempt to improve the symptoms.

Reconsideration of the psychiatric diagnosis occurred

TABLE I

Psychiatric features of the 12 patients studied

Case no.	Age at psychiatric diagnosis (yr)	(m)	Sex	Age at organic diagnosis (yr)	(m)	Psychiatric symptoms	Psychiatric diagnosis	Final neurological diagnosis
1	6	1	F	12		Walking with hunched shoulders and posturing of neck	Anxiety state	Grade II astrocytoma of spinal cord
2	7		M	9	7	Destructive; uninhibited behaviour; flight of ideas; deteriorating school performance; inappropriate affect	Child psychosis	Sex-linked diffuse cortical sclerosis with Addison's disease
3	10		F	14		Provocative behaviour with considerable variability; progressive loss of cognitive skills	Behaviour disorder	Congenital syphilis (tertiary phase)
4	5	7	M	6	5	Irritable; easily frustrated; tantrums; deteriorating school performance	Child psychosis	Subacute sclerosing panencephalitis
5	8	6	F	9		Destructive; aggressive; deteriorating school performance	Conduct disorder	Metachromatic leucodystrophy
6	8	5	M	9		Anorexia; weight loss; pain in the back	Hysterical reaction	Cystic astrocytoma of spinal cord
7	7		M	8	4	Aggressive and excitable behaviour; difficulty in seeing blackboard	Behaviour disorder	Batten's disease
8	7	9	F	8	8	Dragging left leg; hunching shoulders	Anxiety state	Osteoid-osteoma of L5
9	10	7	F	14	10	Aggressive, destructive behaviour; loss of cognitive skills	Behaviour disorder	Degenerative CNS disease (idiopathic)
10	6	10	F	7		Pain in elbow, knees, hands; difficulty climbing stairs	Emotional disorder	Polymyositis
11	7	3	M	10	1	Unsteady gait; inco-ordination; tremor of hand	Anxiety state	Friedreich's ataxia
12	9	1	F	9	4	Limp left leg; fidgetiness	Hysterical reaction	Dystonia musculorum deformans

for a number of reasons. The onset of epileptic seizures was important in 3 children, and increased loss of power in the limbs and loss of visual function prompted reconsideration in 4 cases. In some of the remaining children, though the symptoms had not changed, parental pressure for a further opinion, or diagnostic doubts in the psychiatric team were mainly responsible for re-evaluation of the situation. A paediatrician requested re-evaluation in 6 children, a general practitioner in 2, a psychiatrist in 2, an orthopaedic surgeon in one. In Case 12 a paediatric neurologist who had originally concurred in the psychiatric diagnosis kept the case under review and eventually altered his diagnosis in the light of findings on examination.

The age range of the children at the time of final neurological diagnosis was between 6 years 5 months and 14 years 10 months. The period of time elapsing between the original psychiatric diagnosis and final neurological diagnosis ranged from 2 to 71 months with a mean of 21 months, so that the diagnostic delay involved cannot be regarded as trivial.

The neurological diagnoses are listed in Table II indi ating the circumstances leading to neurological referral, the examination and investigation findings, and subsequent management and outcome.

Although 5 or possibly 6 of the 12 conditions are genetically determined, only in the family with Addison-Schilder's disease was there a positive family history, there having been unrecognized affected cases among the mother's male sibs. A history of repeated miscarriages affecting the mother of the girl with congenital syphilis was in retrospect relevant to the diagnosis.

It will be seen that the final neurological diagnoses, though not in general uncommon in specialist paediatric neurological practice, would not be encountered except occasionally by general practitioners, child psychiatrists, or even consultant paediatricians. Most of the conditions diagnosed were not treatable except in a palliative sense, and 8 of the 12 children were found to be suffering from diseases which will, in all probability, result in death before they reach adult life. In the hereditary conditions genetic counselling was obviously highly relevant.

Discussion

Published reports of diagnostic error appear infrequently for obvious reasons. Yet the study of mistaken diagnosis is important, for it should enable us to avoid pitfalls into which others have fallen.

The children reported here were originally diagnosed as having a psychiatric disorder but later were found to have neurological disease. Most doctors would probably regard missing a physical problem in this way as somehow more blameworthy than misdiagnosing, and therefore mistreating, a psychiatric disorder. Such an attitude has little justification, for psychiatric disability is just as 'real' as physical disability, and indeed as a cause of handicap in childhood is also more frequently encountered in the general population (Rutter, Tizard, and Whitmore, 1970b). Nevertheless, there can be no doubt that inappropriate psychiatric labelling can result in unnecessary distress, and the question arises how much of the diagnostic confusion reflected in the cases described above could have been avoided.

Perhaps the most important point to make is that the symptoms with which these children presented were unusual in child psychiatric practice. Although failure to make progress in school is common enough, actual *deterioration* in academic performance—reading, writing, and number work—is infrequently encountered. Deterioration of these skills can occur for purely psychiatric reasons, but such an explanation should not be readily accepted. Regression in other areas of behaviour, encopresis, enuresis, and general reversion to an immature pattern of social and emotional behaviour with clinging and reduced frustration tolerance are, by contrast, common in child psychiatric practice, and after appropriate physical examination can more readily be accepted purely as stress reactions. The fact that apart from these unusual symptoms the children with brain disorders in this group showed a variety of conduct and emotional symptoms is in line with the findings of others that there is no characteristic behavioural syndrome diagnostic of brain damage (Rutter, Graham, and Yule, 1970a). Symptom variability is often taken to be suggestive of psychiatric illness at the diagnostic stage. Variability was in fact very common in these cases and therefore should not be taken as a sign that neurological disorder is absent.

Loss of function of the limbs or reduction in visual acuity are not frequently seen by child psychiatrists, and though they may represent manifestations of hysterical reactions, caution is again necessary in making this diagnosis. Caplan (1970) followed up 28 children diagnosed in a child psychiatric department as suffering from 'conversion hysteria', 13 (46%) of whom were later found to have proven organic illnesses to explain their symptoms. Similarly, Harcourt and Hopkins (1972) recently described a number of children in whom tapetoretinal degeneration, possibly representing Batten's disease (Wilson, 1972a), presented as a disturbance of behaviour reactive to frustration occurring with diminished visual acuity. Our own observations add weight to the clinical impression that hysterical reaction is rare in prepubertal children.

Although abnormal neurological or other physical signs may not have been present at the time of

TABLE II

Neurological features

Case no.	Reason for reconsideration of case	Signs at time of neurological diagnosis	Relevant investigations	Neurological diagnosis and outcome
1	Progression of postural symptoms	Kyphoscoliosis; upper motor neurone signs in legs; lower motor neurone and sensory signs in left arm	Spine *x*-rays: widened canal from C2–T8; air myelogram: expanded cord; biopsy	Grade II astrocytoma of cord; laminectomy; decompression of cord; radiotherapy; improvement of symptoms; discharged
2	Onset of uncontrollable focal seizures, loss of speech; incoherence of thinking	Focal seizures; speech loss; papilloedema; bilateral upper motor neurone signs; dry skin	Serum sodium: low; Synacthen test: impaired (2 younger 'normal' brothers had impaired response to Synacthen at this time)	Sex-linked diffuse cortical sclerosis with Addison's disease; rapid deterioration; died 2 m later.
3	Progressive deterioration of school performance; bizarre behaviour; admitted with drowsiness, dysarthria, ataxia, and focal fits	Disorientated; hyperactive; lethargic; dilated left fixed pupil; left homonymous hemianopia; bilateral upper motor neurone signs; pathologically brisk reflexes; extensor plantar responses; bilateral corneal opacities; iritis, uveitis	Positive serum, CSF, Wassermann, VDRL; paretic Lange CSF; *Treponema pallidum* identified from diagnostic tap of anterior chamber of eye	Congenital syphilis (tertiary phase); penicillin therapy; neurological signs and behaviour improved; continuing iritis with cataracts and corneal scarring
4	Progressive loss of skills, school performance; loss of use of left hand, ataxia, and dysarthria; ?papilloedema	Inattentive; ataxic; mild left hemiparesis with left homonymous hemianopia	EEG: large periodic stereotyped complexes at 8–12 s intervals; paretic Lange CSF; raised serum; measles antibody titres	Subacute sclerosing panencephalitis; progressive deterioration
5	Progressive loss of skills; aggressive, destructive behaviour; left-sided ataxia; weakness; double incontinence	Labile personality; facial grimacing; dysarthria; ataxic gait; increased tone and tendon reflexes	Urine for intracellular metachromatic granules (+); leucocyte arylsulphatase A activity: low	Metachromatic leucodystrophy; progressive deterioration; no response to vitamin A-deficient diet
6	Parental pressure for second opinion	Weight loss, back stiffness; torticollis; no neurological deficit	Spine *x*-rays: eroded pedicles; myelogram: space-occupying lesion of spinal cord; exploratory laminectomy	Cystic astrocytoma of spinal cord; radiotherapy; torticollis improved, regained weight
7	Deteriorating vision	Drop attacks; reduced visual acuity; attenuated retinal arteries; abnormal retinal pigmentation; optic atrophy	Electroretinogram: reduced responses; rectal biopsy: neuronal storage	Batten's disease; registered as blind; now in mental subnormality hospital
8	Persisting disability	Kyphoscoliosis; limp; limitation raising left leg; depressed left knee jerk	Further spine films: sclerotic lesion L5 lamina	Osteoid-osteoma; surgical resection of lesion; improvement resulted
9	Onset focal fits aged 13 yr 3 m; slow progress at school; persisting unusual behaviour	Focal fitting; nystagmus; dysmetria; pathologically brisk reflexes; bizarre behaviour	Electroretinogram: abnormal	Degenerative CNS disease, type unknown; anticonvulsants; day admission to training centre
10	Referral to exclude organic condition before psychiatric referral	Generalized lympho-adenopathy; waddling gait; papular rash; reduced power of proximal muscles; depressed tendon reflexes	Serum creatine phosphokinase: raised; electromyogram: myopathic changes; muscle biopsy; myopathic	Acute polymyositis; steroid therapy; slow improvement
11	Finding of heart murmur by GP	Cerebellar ataxia; positive Romberg; depressed knee and ankle reflexes; extensor plantars; kyphoscoliosis; cardiac murmur	Electrocardiogram: right ventricular hypertrophy; electromyogram: dennervation; nerve conduction: slowed	Friedreich's ataxia; change to school for physically handicapped; deterioration
12	Kept under review by paediatric neurologist	Dystonic posturing left arm and leg; wasted left leg muscles	None	Dystonia musculorum deformans; stereotaxic surgery with short-lived success

psychiatric diagnosis, the need for periodic reassessment is raised by this group of cases. By the time of neurological diagnosis only one child had no abnormal neurological signs apart from torticollis and abnormal back stiffness. Investigations of relative simplicity, e.g. blood electrolytes (Case 2), urine examination for intracellular metachromatic material (Case 5), syphilis serology (Case 3), neurophysiological studies (Cases 4, 7, 9), serum creatine phosphokinase (Case 10), a blood film for vacuolated lymphocytes (Case 7), and repeated spinal *x*-rays (Cases 1, 6, 8), were omitted which

were of diagnostic importance. The relative rarity of the conditions listed together with the welter of complex and expensive laboratory investigations available (reviewed by Wilson, 1972b) emphasizes the necessity for referring children who present with unusual psychiatric symptoms to centres where a full paediatric neurological assessment can be undertaken.

Periodic re-evaluation is often not a straightforward procedure, as in many of the children we have described the psychiatric diagnosis was a tentative one made only after the child had been seen by a number of consultants in different specialities who had failed to make a physical diagnosis. Eventually, a child psychiatrist's opinion would be sought and often he would be in doubt. He might then suggest treating the child psychiatrically without any great conviction of the presence of a psychiatric disorder. Such diagnostic uncertainty must obviously be very stressful for the child and family involved, and, where psychiatric treatment is in progress, a periodic physical examination must inevitably affect the motivation of the family to co-operate in such treatment. Nevertheless, in these unusual situations where diagnostic doubt exists, it seems important to bear in mind and periodically test for the possibility of an organic disorder.

Helping parents and children to cope with diagnostic uncertainty is one of the main responsibilities of medical and social work staff involved in managing cases such as those described. One danger is that at the time the neurological diagnosis is made, the parents, having seen an organic problem treated as an emotional one, deny all their own and their other children's emotional reactions to the new situation. This is not an inevitable consequence. A brother of one of these children refused to go to school partly as an emotional reaction to his sib's fatal illness, and it was possible to treat this problem psychiatrically despite the previous misdiagnosis.

Finally, it should be mentioned that these cases illustrate the need for close working co-operation between general practitioners, school medical officers, paediatricians, and child psychiatrists. Misdiagnosis, with its attendant consequences of increased family distress, is less likely where doctors in different disciplines are not working in isolation, but are able to share their expertise and learn respect for each other's skills.

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Correspondence to Professor P. J. Graham, Department of Psychological Medicine, The Hospital for Sick Children, Great Ormond Street, London WC1N 3JH.