Matters arising

Aqueduct stenosis and manic depressive psychosis

Sir: Reveley and Reveley¹ have drawn attention to an association between aqueduct stenosis, hydrocephalus and schizophrenia. We present a case of bipolar affective psychosis occurring with internal hydrocephalus due to aqueduct obstruction by an ependymal cyst.

The patient, a male and only child, was born normally in December 1952 into a family without known history of mental illness or epilepsy. His motor activity and speech developed normally but he displayed a number of childhood behavioural disorders which may have been related to the loss of his father in a flying accident. The patient attended a normal school but his progress at a College of Further Education proved disappointing and was followed by unsettled employment. The patient was first admitted to a psychiatric hospital in February 1974 after he had created a disturbance while looking for his girl friend. Although alert and fully orientated on admission, he was hyperactive, garrulous, aggressive and voicing grandiose delusions. There was a history of well-controlled major epileptic seizures since the age of 13 but no evidence of recent fits or anticonvulsant intoxication. Physical examination and routine haematological and biochemical screening revealed nothing untoward. An EEG, however, showed a mild disturbance in the fronto-temporal areas which was more marked on the left side. His full scale Intelligence Quotient was 81 (Verbal 80, Performance 86). Since admission the patient has spent most of his time in hospital. At times there were spells of hyperactivity associated with flight of ideas and grandiose delusions. On other occasions he was apathetic, complained of depression, slept poorly with early waking, and he lost his appetite. In June 1981 a neurological opinion was sought because of persistent frontal headaches, ataxia and intermittent diplopia. A CT scan of the brain revealed moderate internal hydrocephalus and a large cyst-like lesion in the posterior fossa. On 28 July 1981 the patient underwent sub-occipital craniectomy. An ependymal cyst was found and the cyst was shunted into the subarachnoid space. Subsequent radiographic studies have shown that the cyst remains collapsed and there has been some diminution in the size of the lateral

ventricles. Two weeks after operation the patient had his first post-operative major seizure and afterwards was transiently disorientated and deluded. He has been free of other psychotic phenomena and his mood has been stable. He is now away from the hospital and the dosage of his neuroleptic medication is being gradually reduced.

We believe that this case history suggests that manic depressive psychosis, as well as schizophrenia as in the patients reported by Reveley and Reveley, may occur in association with internal hydrocephalus due to aqueduct obstruction. Furthermore, surgical intervention resulted in a noticeable improvement in the patient's mental state. It is tempting, therefore, to conclude that the psychotic illness was related to the brain disturbance. Presumably only time will tell.

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Reference

- 1 Reveley AM, Reveley MA. Aqueduct stenosis and schizophrenia. *J Neurol Neurosurg Psychiatry* 1983;46:18-22.

Reveley and Reveley reply

We are pleased that our case reports have aroused comment. Our purpose was to highlight the probable complexity of the relationship between ventricular enlargement and schizophrenia; we do not believe that the pathological processes leading to aqueduct stenosis and hydrocephalus are necessarily integral to the development of psychosis, nor specific to schizophrenia. From a genetic standpoint, our reasoning is as follows: schizophrenia has a known and substantial genetic component, which can be theoretically explained as a continuum of liability with a threshold beyond which the disorder becomes manifest. Individuals with a major genetic predisposition may then require little else to become psychotic; others may do so only after additional environmental insult, and a few may develop a psychosis secondary to environmental factors alone. We suggest that our patients may fall into one of the latter two categories; similarly the patient reported

by Bhanji, Gardner-Thorpe and Rahavard. The aqueductal stenosis may serve as a stress factor which "uncovers" an underlying and variable predisposition, rather than acting as a specific aetiological circumstance.

We are grateful to Dewan and Bick for bringing the reports of Lying-Tunell¹ and Price and Tucker² to our attention. In the light of these reports, we should revise our statement to say "we could find no reports linking schizophrenia specifically with aqueduct stenosis and hydrocephalus." For it is clear from the radiological and clinical description that Lying-Tunell does not report cases of aqueduct stenosis, just as we do not report cases of normal pressure hydrocephalus (NPH). For example, Case 1 of Lying-Tunell actually had "marked dilatation of Sylvian aqueduct" on pneumoencephalogram, while Case 2 had a "highly dilated ventricular system". Price and Tucker gave no details of the pneumoencephalogram, other than to say that it is "diagnostic for NPH". The size of the fourth ventricle was not specifically noted.

NPH characteristically develops in adulthood³ and is associated with dementia, ataxia, and incontinence, as well as other symptoms less characteristic. It is noted in the cases of Lying-Tunell and Price and Tucker. In contrast our cases 2 and 3 had onset of hydrocephalus in infancy, within a few weeks of birth, and developed skull enlargement. Case 1 had onset in adolescence, but did not develop any clinical signs of NPH. She had a difficult forceps delivery, which may have contributed to the development of aqueduct stenosis.

The questions of CSF flow disturbance and progression of ventricular enlargement in schizophrenia have yet to be fully explored. Weinberger and Wyatt's⁴ finding of non-progression was based on only five cases after 2-3 year follow up. Disturbances in CSF flow in schizophrenics have been found by Sedvall and associates⁵ using isotope cisternography.

Hydrocephalus is a condition with multiple aetiologies. It is unknown if any of these processes are linked to the ventricular enlargement occasionally occurring in functional psychoses, and this should be the subject of further research.

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