patients did the impairment persist. In a series of necropsies on patients with porphyria in South Africa, about 80% had renal tubular necrosis histologically (Eales et al., 1971). Most of these patients died of respiratory failure and the tubular necrosis may have resulted from hypoxia. Investigation in this patient has failed to suggest any of the common renal diseases and it may be that one of the accumulated compounds is nephrotoxic.

Lees et al. (1970) found raised plasma cholesterol values in 5 out of 10 patients with acute intermittent porphyria. Plasma triglycerides were normal, as were the very low density and high density lipoproteins. However, the low density (β-) lipoproteins were increased in 9 out of 10 patients. There have been no reports to date of raised α-(high density) lipoprotein in acute intermittent porphyria, though this fraction is sometimes increased by oestrogen administration (Rifkind, 1970). This patient also had increased thyroxine-binding globulin which has been previously reported in porphyria and also in oestrogen administration (Brownlie, 1973). There may be increased synthesis of a number of different proteins in porphyria. It is of interest that oestrogens can also precipitate attacks of acute intermittent porphyria.

This patient is reported because renal damage is an unusual and unexplained complication of acute intermittent porphyria and because increased high density lipoproteins have not been previously reported in this condition.

Summary

An 11-year-old girl presented with typical clinical and biochemical findings of acute intermittent porphyria. After recovery from her third attack, she was found to have persistently raised serum cholesterol and high density (α-) lipoproteins. Her creatinine clearance was persistently reduced to below 40% of normal. Possible interpretations are discussed.

I am grateful to Professor T. E. Oppé and Drs. P. Adams and G. Tait for their help in the investigation of this patient.

References


Short reports


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Hydrocephalus related to pulsion diverticulum of lateral ventricle

The clinical syndrome of headache and vomiting together with signs of raised intracranial pressure is known to be associated in paediatrics with a variety of causes, both benign and malignant. We describe below a rare and interesting cause which we have not seen previously reported in children.

Case report

A 6-year-old boy had been seen in outpatients a year before his emergency admission in November 1972 with persistent episodic headaches for 2 to 3 years, thought to be associated with upper respiratory tract infections. One severe attack had lasted a week and had been associated with vomiting. His mother had typical attacks of migraine. On examination nothing abnormal was found and his parents were reassured.

When admitted, his attacks of headache and vomiting were said to have steadily increased in frequency and severity and he had become irritable. In the preceding 6 months he had had three particularly bad attacks in which he had woken complaining of headache which had increased in severity for about half an hour and was associated with nausea followed by vomiting which would temporarily alleviate the headache. During these attacks he was noticeably drowsy. One attack lasted for a week and during this time he had resented normal noises and shown an intense dislike for the smell of food. His mother and teacher had noticed no unsteadiness, but he had apparently developed a tendency to fall over.

Past history was noncontributory. Antenatal and neonatal progress were uneventful and his general health was excellent. The paternal grandmother had recently died of a cerebral tumour, and this had clearly contributed to the parents’ anxiety.

On examination he was fully conscious and alert, and no abnormalities were detected outside the nervous system. There was a definite cracked pot sound on percussion of his skull, though there was no clinically obvious hydrocephalus. There was minimal neck stiffness and Kernig’s sign was negative. Cranial nerves were intact. Visual fields were full. Fundoscopy showed no papilloedema. His eye movements and gaze co-ordination were normal. There was no nystagmus. Power, tone, reflexes, and sensation were all normal.

Skull x-ray showed definite evidence of raised
intracranial pressure with slight enlargement of the pituitary fossa, mild copper beating, and sutural diastasis. EEG was grossly abnormal with excess delta wave activity and possibly some lateralization suggesting a focus in the left temporal region. He was, therefore, referred to the neurosurgical service for further investigation and treatment.

On admission to the Neurosurgical Unit, Frenchay Hospital, Bristol, a right frontal burr hole was performed under local anaesthesia. An umbilical catheter was passed into the frontal horn of the right lateral ventricle, which contained clear CSF at high pressure. A myodil ventriculogram showed large lateral ventricles and an enlarged 3rd ventricle with an elongated and raised suprapineal recess, suggestive of a lesion in the region of the pineal gland. The myodil flowed through the normally placed 4th ventricle into a capacious cisterna magna (Fig.). Throughout this procedure the child remained cheerful and alert.

On 3 December 1972, after prolonged ventricular drainage, a right carotid angiogram under general anaesthesia excluded an aneurysm of the vein of Galen. A right posterior craniotomy was performed immediately (Mr. D. G. Phillips). On retracting the right occipital lobe laterally a greyish cyst covered with arachnoid mater was seen at the free margin of the tentorium. The tentorium was incised sagittally 1 cm from the midline and the grey-walled cyst was displayed, spreading the cerebellar hemispheres apart and displacing the vein of Galen upwards and to the left. When the cyst wall was excised a brain cannula was passed into the right lateral ventricle and its blunt tip was found in the cavity of the cyst. The cyst contained fluid similar to CSF, but with a raised protein content (138 mg/100 ml compared with the ventricular CSF protein of 20 mg/100 ml), implying partial but not complete loculation of the diverticulum. No solid neoplastic lesion was seen and it was assumed that the cyst was a diverticulum of the right lateral ventricle which had extruded beneath the tentorium. Since adequate removal of the cyst wall had been effected, no shunting or bypass procedure was considered necessary.

The patient made a good recovery with a transient left homonymous hemianopia. At outpatient examination 1 month later, he was putting on weight, was active and alert, and was normal on clinical examination.

**Discussion**

A pulsion diverticulum of the ventricular system complicating obstructive hydrocephalus was first described by Penfield (1929). In his case, a cholesteatoma of the 3rd ventricle had blocked the foramen of Monro causing massive ventricular dilatation.

Pennybacker and Russell (1943) recorded 3 cases where the diverticulum had occurred at the weakest point of the lateral ventricular wall, in the area of the collateral trigone between the fornix and the forceps major. At this site, diverticula caused by high intraventricular pressure may enlarge to extend across the midline, pushing the vein of Galen upwards, and may protrude down through the tentorial hiatus, ballooning out beneath the tentorium on top of the vermis of the cerebellum. In this situation obstructive hydrocephalus is compounded by blocking of the passage of CSF through the cisterna ambiens. A characteristic bubble of air beneath the tentorium may be seen on air ventriculography and may give the clue to the presence of a diverticulum. However, it is now unusual to fill the ventricles with air because the complications of this investigation are many, particularly in the presence of raised intracranial pressure. The first 6 cases recorded died, 3 soon after operation. Myodil ventriculography is perhaps less informative but safer.

The only case to survive operation successfully was recorded by MacFarlane and Falconer (1947). As recommended by Pennybacker and Russell (1943), a posterior fossa craniotomy was performed on a 23-year-old carpenter. The diverticulum was dissected and found to be composed of two layers, both of which were excised in part to open the lateral ventricle into the cisterna ambiens. It seems more logical, as in our case, to approach the neck of the sac above the tentorium, splitting the tentorium to remove the fundus.

Other sites of pulsion diverticula are possible, both into the hemisphere substance or more commonly as pouching out of the lamina terminalis of the 3rd ventricle anteriorly, or the suprapineal recess posteriorly.

Six cases have been reported (Kanjilal, 1972) of spontaneous rupture of diverticula into the subarachnoid space, affording 'nature's cure'. It seems likely that this occurs more frequently than is recorded, and may account for some of the spontaneous remissions of hydrocephalus seen in infants. It is said to happen in a fair proportion of mice with a recessive gene for hydrocephalus (McLone, 1972).

Our case is puzzling because no cause has been found for his hydrocephalus. The myodil flowed freely through the ventricular system and out into a capacious cisterna magna. Perhaps, like McLone's mice, it began with a communicating hydrocephalus which converted by pressure on the aqueduct to an obstructive form, which itself was relieved by the diverticulum. No sign or symptom at present points to any recurrence of hydrocephalus.

**Summary**

A 6-year-old boy presented with a chronic history of episodic headaches and vomiting, together with
FIG.—Myodil and air introduced via the catheter in the right lateral ventricle. 1. Dilated lateral ventricle, with air in the frontal horn and myodil in the posterior horn. 2. Elongated and raised suprapineal recess of the 3rd ventricle lying along the superior border of the cyst. 3. Position and approximate size of the cyst. 4. Depressed 4th ventricle. 5. Normal cisterna magna.
hydrocephalus. The cause of his symptoms was a pulsion diverticulum of the lateral ventricle, a rarity which does not usually figure in the differential diagnosis of this syndrome, though the history is characteristic of the few recorded cases.

We thank the consultant staff of the Department of Child Health for permission to publish this case.

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Thyroxine levels in normal newborn infants

It is well known that thyroxine (T₄) levels in the blood are higher in infancy than at any other time of life. This was shown in the papers of Danowski et al. (1951), Durham et al. (1954), and Pickering et al. (1958). These papers were based on the measurement of protein-bound iodine and butanol-extracted iodine in infants. O’Halloran and Webster (1972) measured thyroid function in Australian Caucasian babies during the first year of life. They used a technique involving a resin column, ¹²³I-labelled thyroxine, and a γ counter. This method of T₄ measurement is marketed in the form of Tetrason Biochemistry laboratories, Tetrakits (Ames Company), and Thyropac 4 (Amersham Radio Chemical Centre). It is finding increasing favour in biochemistry laboratories as a method of estimating T₄ levels in the blood, for it is quick, easy, accurate, and uses only 0.1 ml serum. The method is attractive to paediatricians because it can be performed on heel prick samples of blood.

In view of the probable increased use of this test, a study was done to define the normal range of T₄ levels in Caucasian neonates in Britain.

Methods

T₄ measurement was carried out on a series of venous and some capillary samples of blood. The estimations were carried out in the Fazakerley Hospital Biochemistry Department by one of us (D.W.) using a Tetrakits. This system is based on the printed instructions. 30 samples of cord blood, 30 blood samples from 6-hour to 44-hour-old babies, and 30 samples from 4- to 7-day-old babies were taken for T₄ estimations.

The infants in the trial were normal Caucasian term babies. Any complicating factor such as prematurity, small-for-dates status, jaundice, asphyxia, sepsis, etc., excluded the infant from the study. The only maternal complication allowed was an elective caesarean section for disproportion or because of a previous caesarean section. The samples were not taken serially from individual infants.

Results

The values of serum thyroxine in μg/100 ml blood were as follows. In 30 samples of cord blood the range was from 7·2 to 13·5 μg/100 ml. The mean value of the cord samples was 9·9 μg/100 ml and the normal range (mean ± 2 SD) was 6·1 to 13·7 μg/100 ml.

In 30 samples of blood from infants 6 to 44 hours old, T₄ values ranged from 13·2 to 19·6 μg/100 ml. The mean value was 16·6 μg/100 ml and normal range 13·4 to 19·8 μg/100 ml.

In 30 samples of blood from infants 4 to 7 days old, T₄ values ranged from 8·6 to 18·5 μg/100 ml. The mean value was 14·4 μg/100 ml and normal range 10·4 to 18·4 μg/100 ml.

Discussion

The cord blood values are similar to those found by O’Halloran and Webster (1972). The mean cord blood T₄ in our series was 9·9 μg/100 ml, which was lower than their mean value of 11·3 μg/100 ml.

The mean T₄ value of O’Halloran and Webster’s 20 babies aged 0 to 13 days was 13·2 μg/100 ml. This is exceeded by the mean value of 14·4 μg of our 30 4- to 7-day-old babies. The mean T₄ levels of our 30 6- to 44-hour-old infants was even higher at 16·6 μg/100 ml.

Our findings are in keeping with the findings of Danowski et al. (1951) and Fisher and Odell (1969) who found that peak thyroxine levels occurred between days 1 to 4 of life. Fisher and Odell showed a marked increase in thyroxine stimulating hormone at this time. It can be seen that our
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