Figure The patient, Mr AH

able 2 cm below the costal margin on inspiration. Neurological examination revealed poor vision due to myopia. Corrected acuities were 3/24 on the right and 3/36 on the left. In addition, he showed pendular nystagmus. Other cranial nerves were intact. There was a spastic tetraparesis, more pronounced on the left and some spinothalamic sensory loss on the right side of the body, but a precise demarcation was not possible owing to the patient's reduced intellect. Investigations showed a normal full blood count, thyroid profile and XY chromosome pattern. The cerebrospinal fluid was clear, under normal pressure, and acellular with a protein content of 0.35 g/l. Cervical spinal radiographs showed minor degenerative changes and the canal was of normal sagittal diameter (16 mm at the level of the fourth cervical vertebra). Computed tomogram of the brain was normal. The EEG showed theta activity in all leads intermittently occurring in increased voltage sharp bursts. A myodil myelogram revealed a widened cord from the craniovertebral junction to the level of the seventh cervical vertebra, and a prolapse of the cerebellar tonsil to the level of the second cervical vertebra. Cervical laminectomy revealed right cerebellar tonsillar prolapse, a vestigial left tonsil and a syrinx at the level of the fourth cervical vertebra. Decompression of the syrinx revealed a clear fluid with a protein content of 0.76

g/l. There was symptomatic improvement post operatively.

Chiari malformation and syringomyelia have not previously been reported in Noonan's syndrome. Reported neurological defects are a case of arrested hydrocephalus in Noonan's original series, and a patient described by Gorke⁶ who had an abnormal brain CT showing large basal cisterns and ventricles and a defect in the left temporal region. Gorke considered that neurological defects may be part of Noonan's syndrome. It is not known whether the association between the Chiari malformation, the syrinx and Noonan's syndrome is causal or a chance occurrence. The association between Chiari malformation and syringomyelia is well known.^{9,10} Birth injury has been suggested as a causative factor in syringomyelia¹¹ but there is no accurate information available about our patient's birth. A study of patients with syringomyelia performed at the Midland Centre for Neurosurgery and Neurology showed that 22 of 122 patients had other obvious developmental defects.¹¹ Many patients with Noonan's syndrome are mentally retarded. This may result in suppression of neurological symptoms and signs, and less investigation. In addition, the cardiovascular complications of the syndrome may result in death before the neurological problems are apparent.

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Unusual presentation of cerebral arteriovenous malformation: report of a case with visual failure

Sir: The clinical manifestations of cerebral arteriovenous malformations are well documented.¹⁻⁶ Parenchymal, subarachnoid or intraventricular haemorrhage occurs in approximately 50% of the lesions and can be recurrent. Another common presentation is with a focal or generalised seizure. Vascular headache is often mentioned as a common symptom but in a large series of patients with proven arteriovenous malformation, only 5% were found to have a history of migraine.⁷ Other features include audible bruits and progressive mental deficit. Focal neurological signs associated with arteriovenous malformations may be the result of cerebral ischaemia due to so called "stealing" of blood from adjacent normal brain tissue. We report here an unusual presentation of a cerebral arteriovenous malformation in a patient whose physical signs and investigations suggested a pituitary tumour. We believe such a case has not been reported before.

A 42-year-old woman was admitted for investigation of blindness. She was known to be mentally subnormal and had suffered grand mal seizures since the age of 6 years. Two years prior to this admission, she had been seen in the neurology clinic because of poor seizure control. There were no visual symptoms at this time. She continued on sodium valproate, sulthiame and phenytoin as anticonvulsant medication. She then attended with a history of gradual loss of vision for the past year. On examination, visual acuity was reduced to finger move-

ments and fundoscopy showed bilateral optic atrophy. Visual field assessment was not possible. Eye movements were full; the pupils were dilated and reacted sluggishly to light. The left leg was weak and wasted with diminished reflexes as a result of poliomyelitis. Plantar responses were flexor. No vascular bruit could be heard and there were no other neurological signs. A skull radiograph showed gross expansion of the sella turcica with erosion of the dorsum and posterior clinoid processes. This appearance, along with the clinical observation of bilateral optic atrophy, suggested the presence of a pituitary adenoma. A CT scan with contrast failed to identify any sellar lesion. Bilateral carotid angiography was performed to assess the degree of suprasellar extension of any pituitary lesion. The right side was normal apart from dilated veins over the lateral surface of the right hemisphere. On the left side, the cerebral circulation was extremely rapid and the whole of the arterial tree was not visualised, but a large arteriovenous malformation was demonstrated. A posterior branch of the left middle cerebral artery was grossly dilated and tortuous and drained into a tortuous loop of veins. There was early visualisation of the inferior sagittal, straight and transverse sinuses (fig 1).

Fig 1 Left carotid angiogram showing a posterior branch of the middle cerebral artery feeding the malformation with early visualisation of the venous sinuses. The destruction of the sella is also obvious.

The cortical veins on the lateral surface of the hemisphere were also visualised early and were grossly dilated and tortuous. A leash of abnormal vessels were seen in the anterior temporal and parasellar regions. The appearance was that of an arteriovenous malformation in the left temporal region fed by a posterior branch of the middle cerebral artery and shunting blood into the straight and transverse sinuses. Following angiography, she had a series of major

Fig 2 Coronal section of the brain at the level of the optic chiasma showing a large arteriovenous malformation in left temporal region involving the chiasmatic region.

seizures and remained unconscious. She developed neck stiffness and the CSF was uniformly blood-stained with supernatant xanthochromia suggesting subarachnoid bleeding from the malformation. She remained in coma and died three days later.

Postmortem examination revealed a large arteriovenous malformation arising in the region of the left middle cerebral artery, which was hard to define precisely owing to a terminal subarachnoid haemorrhage. The mass of blood and malformation closely surrounded the optic chiasma from which it could not be separated (fig 2). Both lateral walls of the sella turcica were absent, the malformation eroded the left wall of the fossa, displacing the pituitary gland towards the right side and partly through the deficit in the wall on this side. The anterior one half of the left temporal lobe was compressed and haemorrhagic; further spotty haemorrhagic infarction extended to involve the white matter in the posterior portion of the left temporal lobe, and more acutely in the midbrain. The cortical veins over the parietal lobes were engorged but no vascular or tumourous anomaly was present in the midline posteriorly, or below the tentorium. Sections from the malformation consisted of groups of abnormal arteries and veins, the elastica being mainly monolayered and often fragmented in some of the channels. Several widely expanded cavernous vessels were present in places. The pituitary gland was histologically normal.

The clinical presentation with bilateral optic atrophy and destruction of sella turcica was initially thought to be due to a pituitary adenoma but there was no evidence of any suprasellar extension on the angiogram and the gland itself was found to be normal at necropsy. Apparently, the clinical picture was due to direct involvement of the optic chiasma and the sella by the large arteriovenous malformation in

the left temporal lobe. The history of long-standing epilepsy and mental retardation were possibly related to this large malformation shunting blood directly into the venous sinuses. Unusual presentation of arteriovenous malformation previously described include extrapyramidal dysfunction,⁸ hydrocephalus and thalamic syndrome⁹ and cardiac failure in children.¹⁰ Moody and Poppen³ mentioned visual deficit in 30 of their 105 cases with arteriovenous malformation but no details of the visual loss was described. The development of severe visual loss in the present case must be a very unusual clinical manifestation and we can find no similar case in the literature.

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