sponds to the level of the lesion. A radio-opaque marker (made of fine catheter, 1.5 cm long) is then taped with “Micropore” to the outside of the bandage in the vertical plane, centred on the gantry light (fig B). The head is replaced 5 cm deeper and the cut is repeated with the marker. This allows one to see the intracranial lesion and the vertical marker on the surface of the skull (fig C). The patient is then moved to another room with bandage “A” and the marker undisturbed, whereas the second bandage “B” is secured to the underlying bandage “A” with the aid of “Velcro” strips. The grid marks are arranged to lie on the same side as the lesion. A radiograph is then obtained in the lateral projection with the film placed on the side of the lesion (fig D) to avoid magnification. The horizontal lines of the grid marks, being parallel to the edges of both bandages “A” and “B” indicate the plane of the scan. The information provided by the skull radiograph can be transferred to the patient’s scalp either by measuring from a recognisable landmark or, when the patient is anaesthetised, by placing the film against the patient’s head.

The difficulty of localisation of small intracranial lesions on CT has been widely recognised by neurosurgeons and radiologists. Experience may minimise the errors but the precise localisation of high convexity lesions still poses a considerable challenge. Review of the world literature shows that there have been various attempts made to solve this problem. Of these attempts, those using simple techniques have proved insufficiently accurate. Furthermore, the methods which required fixed external skull markers produced scan artefacts. Computer assisted systems, although highly effective, require the help of an expert radiologist to perform, and the information gained can only be transferred to the patient using a number of highly complicated measurements. It seems that all the previously published methods are time consuming and on occasions may be rejected for that reason. The method here described works well in this department, but we shall have to await confirmation of its value, if and when it is taken up elsewhere. The main source of error in the method arises from the fact that one may not place the marker on bandage “A” to overlie the centre of the lesion in the antero-posterior plane. This problem can be overcome by observing the marker-lesion relationship on the scan and adjusting the projection on to the scalp accordingly. Localisation in the rostral-caudal plane is guaranteed by the gantry light and the placing of the markers thereon. Because of this potential inaccuracy, we do not recommend this method for precise stereotactic localisation but the technique has proved itself of value for the purpose of biopsy and planning of osteoplastic flaps. We have also found it helpful in indicating tumour-bearing territory to the radiotherapists. The method of localisation adds less than five minutes to the scanning time and takes on average only twelve minutes to complete. Construction of the bandage is easy and cheap.

I am grateful to Mr AA Jefferson for his kind help and advice in the preparation of this letter.

NI AZZAM
Department of Neurological Surgery, Royal Hallamshire Hospital, Glossop Road, Sheffield S10 2JF, UK

References


Chiari (type 1) malformation and syringomyelia in a patient with Noonan’s syndrome

SIR: Noonan’s syndrome is an inherited disorder affecting both males and females. The genetic mechanism is uncertain. Cases have been described in which the inheritance is autosomal dominant, recessive or multifactorial, and in some cases relatives have shown only isolated features. The syndrome is named after Jacqueline Noonan who performed a large survey of children with congenital heart disease and described a group of patients with valvular pulmonary stenosis who were short, mentally retarded and showed considerable physical resemblance. Such patients have been described as resembling the Turner phenotype with respect to short stature, webbed neck and cubitus valgus. However, patients with Noonan’s syndrome have normal chromosomes, mental retardation and different systemic abnormalities. The facial features of Noonan’s syndrome are hypertelorism, antimongoloid slant of the eyes, small mandible, low set ears and a relatively short neck. Deformities of the sternum and vertebral abnormalities are common and include scoliosis and spina bifida occulta. Other deformities which have been described are hepatosplenomegaly, ocular abnormalities, cardiovascular lesions in addition to pulmonary stenosis, undescended testes, delayed puberty, and a case of hypopituitarism. Neurological defects have rarely been reported. We describe a patient with Noonan’s syndrome who had a type 1 Chiari malformation and a syrinx. These malformations have not previously been reported in this syndrome.

Mr AH is a 33-year-old Caucasian male with features of Noonan’s syndrome. He is mentally retarded with a verbal IQ of 68 on the Wechsler Adult Intelligence Scale, and is registered as partially sighted. He has five siblings who are between 1.6 metres and 1.7 metres in height but no family members have obvious features of this syndrome. He was initially referred to hospital as a teenager with symptomatic pulmonary stenosis, and in 1964 he underwent pulmonary valvotomy and infundibular resection. This resulted in symptomatic relief though cardiac catheterisation ten years later showed residual stenosis of the pulmonary valve ring. In 1981 he presented to the Department of Neurology with a two year history of discomfort in his neck, difficulty in performing fine movements with his left hand and a stiff left leg which impaired his mobility. On examination he was 1.5 metres in height with characteristic facies, short neck, pectus excavatum and cubitus valgus (fig). Body hair was reduced and his testes were descended. There were no signs of cardiac failure, but an ejection systolic murmur was heard, maximal in the second left interspace. The liver was palp-
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 spinthalamic 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 are a case of arrested hydrocephalus in Noonan's original series, and a patient described by Gorke8 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 Birth injury has been suggested as a causal factor in syringomyelia10 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.

Dr John G Graham has kindly allowed us to publish this case. We thank Dr Graham, Dr A Bligh and Dr CEC Wells for their advice.

MJ BALL, A PEIRS
Department of Neurology, University Hospital of Wales, Heath Park, Cardiff, CF4 4XW, UK

References

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

Sir: The clinical manifestations of cerebral arteriovenous malformations are well documented.1–4 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.5 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-
Chiari (type 1) malformation and syringomyelia in a patient with Noonan's syndrome.

A Peiris and M J Ball

*J Neurol Neurosurg Psychiatry* 1982 45: 753-754
doi: 10.1136/jnnp.45.8.753