literally hundreds of patients. In specific human terms, Cooper devoted himself wholeheartedly in the Home for Incurables to amelioration of symptoms, relief of suffering and cure of patients whom the pundits ex cathedrah said could not be helped. He focused his considerable personal attention on each of his patients; his concern and care for each was obvious to all. He was indeed a remarkable person. As one example, on several occasions when he was invited to lecture outside the country, he learned the language of his hosts so as to be able to lecture to them more efficiently.

To quote Professor Adrian RM Upton, "Cooper... excited... strong, emotional, irrational and destructive responses from his peers." The reasons for those responses are, of course, complex but Professor Upton suggests that Cooper's youth and enthusiasm (as well as the truth of his demonstrations in the face of preconceived notions to the contrary) produced unscientific, unintellectual, if not academically, responses from many senior neurosurgeons and neurologists. Cooper's beliefs that his treatment would "provide important benefits for mankind and his attempts to widen the use of the procedure were seen as self-promotion." A most interesting and enlightening exposition of these aspects of Irving Cooper's life is presented in his own words in the volume entitled The Vital Probe—My Life as a Brain Surgeon. I recommend it most strongly to those interested in neurology, neurosurgery and psychiatry.

To return to Dr Bucy's excellent review, I am unable to avoid replying to two statements which he emphasizes perhaps somewhat dogmatically. The first, "With the development of more accurate" radiological imaging "methods of diagnosis, electroencephalography gradually disappeared" is clearly not true. As we reported several years ago, "neurological/neurosurgical centres such as ours and, I believe, the Mayo Clinic, continue to perform as many EEGs as ever. In addition, EEGs are being done in new and more useful circumstances such as monitoring cerebral function during carotid endarterectomy or in the neuro-intensive care unit.

The second has to do with Dr Bucy's contention that glioblastomas are now amenable to total removal and cure if only they were diagnosed earlier, when the first symptoms appear. Cerebellar astrocytomas in children are often well circumscribed and can be removed so that the patient is cured; however, that is not the case with the overwhelming majority of glial tumours such as glioblastomas. To make the point most strongly, I would like to know if a single well-documented example of a 5-year cure of a patient with glioblastoma has even been published? I believe not but even if an exhaustive review of the literature could produce several, one's failure to find such cases is clearly not due to lack of therapeutic enthusiasm on the part of neurosurgeons or failure of tens or hundreds of thousands of patients with glioblastoma to have received operative therapy. For reasons that are yet unclear, it has not been possible to remove all the malignant tissue; glioblastomas appear not to be cured by surgery. Most clinical neuroscientists would look to the future in molecular biology, immunology or chemotherapy rather than to earlier or better ablative neurosurgery for eventual cures of these dreadful neoplasms.6

Robert R Young
Laboratory of Clinical Neurophysiology, Department of Neurology, Harvard Medical School, Massachusetts General Hospital, Boston, MA 02114, USA

References

Familial syringomyelia

Sir: We were interested to read the report by Busis and Hochberg, in the September 1985 issue of this Journal,1 of two sisters of whom one had proven and the other had possible syringomyelia. We would like to report the cases of a father and son, both of whom had a documented Arnold-Chiari Type I malformation and syringomyelia.

Case 1 A 32-year-old man presented in 1963 with pain in the right arm, reduced spinothalamic sensation in dermatomes C8 to T5 on the right and reduced tendon reflexes in the right arm. There was gradual progression of his symptoms and in 1972 spinothalamic sensation was reduced over C4 to T3 on the right and C3 to T4 on the left and there was MRC Grade 4/5 weakness of right triceps, wrist and finger extensors with absent right biceps and triceps reflexes. Myelography in 1972 indicated an Arnold-Chiari Type I malformation and a widened cervical canal and he underwent posterior fossa decompression and C1 laminectomy with some improvement. In 1984, aged 53 years, he returned with gradually worsening pain in the right shoulder; weakness of the right hand and more severe sensory loss, for 10 years. There was loss of spinothalamic sensation in dermatomes C1 to C2 on the right, MRC Grade 4 weakness in the right arm, absent right arm tendinous reflexes and an absent left triceps reflex. Myelography and subsequent CT scanning showed the previous posterior fossa decompression and laminectomy. A CT scan 24 hours after myelography showed a flattened and atrophic cord and contrast medium within the cord, indicating a syringomyelia. Case 2 The son of Case 1 presented at age 18, in 1977, with pain in the right upper arm and paraesthesiae and numbness in the whole right arm. Examination revealed reduced spinothalamic sensation in the distribution of the right trigeminal nerve and dermatomes C2 to T6 on the right. A wasted right biceps, MRC Grade 4 weakness of muscles supplied by segments C5 and C6, C8 on the right, a reduced right biceps reflex, and exaggerated lower limb tendon reflexes were noted. A myelogram demonstrated the previous posterior fossa decompression and laminectomy, tonsillar ectopia and a swollen cervical cord. A subsequent CT scan also showed the enlarged cervical cord and a large syrinx. At operation a syringomyelic peritoneal shunt was inserted and on follow-up the patient has been pain free and had no progression of neurological deficit.

There was no history of birth difficulty or significant past history in either patient. The parents of Case 1 died in their eighties with no history of neurological disorder. Case 1 has four siblings and Case 2 has two siblings, all of whom are well.

Our cases had documented syringomyelia and Arnold-Chiari Type I malformations, adding to the cases reported and reviewed by Busis and Hochberg. In our cases syringomyelia
dysarthria may have been secondary to the abnormality at the craniocervical junction which may prevent the normal egress of CSF from the fourth ventricle. There was no history of any difficulty in pregnancy or birth, infection or trauma that may have resulted in adhesions in this area. As discussed by Buis and Hockberg, the possibility of this condition being inherited must be entertained.

Matters arising

JOHN S DUNCAN
NIGEL M HYMAN
CHRISTOPHER BT ADAMS
Departments of Neurology and Neurosurgery, Radcliffe Infirmary, Oxford OX2 6HE, UK

Address for reprint requests: Dr John Duncan, Institute of Neurology, Queen Square, London WC1N 3BG.

Accepted 23 November 1985

Isolated central facial palsy

Sir: With great interest we read Drs Huang and Broe’s article Isolated facial palsy: a new lacunar syndrome.\(^1\) They reported three cases of isolated upper motor neuron facial palsy without significant limb weakness. Two of their cases showed some degree of dysarthria and dysphagia. Their CT scans were consistent with small infarcts in the region of the corona radiata bilaterally in one case, the right corona radiata in another case, and the anterior corona radiata bilaterally in the third case. Recently we encountered such a case of central facial palsy with mild dysarthria and clumsiness of the hand. The CT scan showed a small infarct in the region of the left corona radiata as in their cases. We believe the symptoms of our case and their cases are identical to those of Fisher’s dysarthria-clumsy hand syndrome,\(^2\) as shown in our case reported below.

A 58-year-old right-handed man developed sudden dysarthria on 20 July 1984. A right central facial palsy with dysarthria was noted and treated as a cerebral infarction by his doctor. Five days later, he visited our hospital for CT scan. On examination he showed right central facial palsy and mild dysarthria and showed normal tongue and soft palate movement. Tendon reflexes and strength of extremities were normal. He complained of difficulty writing, although his writing appeared normal. CT scan was consistent with a small infarct in the region of the left corona radiata (fig). On 5 September, his symptoms improved. CT scan was unchanged.

In 1967, Fisher reported 20 cases of the dysarthria-clumsy hand syndrome.\(^2\) He stated, “The cardinal features of our cases have been: moderate to severe dysarthria; central facial weakness of one side of the face; deviating of the tongue on protrusion; impairment of la-la-la; a trace of dysphagia; clumsiness, awkwardness, and slowness of the manipulations of the affected hand; questionable weakness of the hand; difficulty in writing…” One pathologically studied case revealed a pontine lacune, which was responsible for the symptoms, he said. He speculated that a similar syndrome might also result from a lesion in the internal capsule or corona radiata. Spertell and Ransom reported a case of the dysarthria-clumsy hand syndrome associated with acute infarction of the genu of the internal capsule.\(^3\) Although Huang and Broe did not mention Fisher’s report, we consider their cases and our case to be examples of his speculation.

KEN NISHIMOTO
HIROSHI ISHIMITSU
YASUHIRO HARADA
ICHIRO MIYATA
NOBUHIKO MATSUMI
Department of Neurosurgery, Iwakuni National Hospital, 2-5-1 Kuroiso-cho, Iwakuni 740, Japan

References

