MATTERS ARISING

Cerebral localisations in articulatory dyspraxias

Clarke *et al*¹ reported a case of pure articulatory dyspraxia (pure anarthria, PA) without orofacial apraxia (OFA), in a patient with a predominantly cortical haemorrhagic contusion. The pure form of the phonetic disintegration syndrome (PA) of Alajouanine, Ombredane and Durand proceeds from a triple dysfunction: paretic, dystonic and dyspraxic.^{2,3} Likewise, PA is considered a very special form of OFA. Finally, OFA is interpreted as an ideomotor or a motor (melokinetic) apraxia, the latter being a motor dysfunction intermediate between "pure" palsy and "pure" (ideomotor) apraxia. Alajouanine *et al* suggested that the components of PA may be related to specific localisations.² Cardebat *et al*² reported such anatomoclinical correlations in cases of partial PA. We also recently described a case of paretic-dystonic PA without OFA³ and, like Clarke *et al*,¹ located the lesion in the inferior part of the dominant precentral gyrus.

Such cases^{2,4} of partial PA and related cases reported in the literature^{3,5} suggest precise anatomoclinical correlations: a) A predominantly cortical lesion of the inferior part of the dominant precentral gyrus responsible for paretic PA or motor apraxic PA; and orofacial palsy or a motor but not ideomotor OFA may be seen; b) A predominantly subcortical lesion of the dominant opercle responsible for dystonic PA without OFA or palsy; c) A lesion of the lower part of the dominant middle frontal gyrus F2 responsible for dyspraxic PA or ideomotor apraxic PA, with ideomotor OFA; d) A lesion of the lower part of the dominant inferior frontal gyrus F3 responsible for Broca's aphasia, without OFA. The case of Clarke *et al*¹ appears to agree with such a hypothesis.

YD DE SMET
Clinique Saint-Louis,
Hôpital Régional du Nord,
BP 103 L-9002 Etelbruck,
Luxembourg

- Clarke CE, Holder DS, Currie S, Stone J. Cerebral localisation in articulatory dyspraxia. *J Neurol Neurosurg Psychiatry* 1992; 55:168-9.
- Cardebat D, Puel M, Nespoulous JL, Rascol A. Trois observations de désintégration phonétique: étude neurolinguistique. In: Messerli P, Lavorel P, Nespoulous JL, eds. *Neuropsychologie de l'expression orale*. Paris: CNRS, 1983:37-51.
- Lecours Ar, Lhermitte F. The "pure form" of the phonetic disintegration syndrome (pure anarthria): anatomo-clinical report of a historical case. *Brain and Language* 1976;3:88-113.
- De Smet YD. Pure anarthria without simultaneous buccofacial apraxia in a Luxembourgish patient. In: Poeck K, Hacke W, Schneider R, eds. *Verhandlungen der Deutschen Gesellschaft für Neurologie 4*. Berlin: Springer-Verlag, 1987:7-8.
- Tonkonogy J, Goodglass H. Language function, foot of the third frontal gyrus, and Rolandic operculum. *Arch Neurol* 1981;38:486-90.

Non-invasive diagnosis of internal carotid artery dissections

We read the article by Mullges *et al* with interest.¹ Carotid artery dissection is prob-

ably one of the commonest causes of young stroke.² Many Centres operate a selective policy for angiography in young stroke. The presence of neck pain, Horner's syndrome or history of trauma are widely thought to be useful pointers to the diagnosis of carotid dissection. We would agree with Mullges *et al* that these signs are only present in a small number of cases, if an alternative screening test is used to detect carotid dissection. We have used continuous wave Doppler to screen all possible young strokes (age range 14-55 years).³ All dissections had either a considerably reduced internal carotid artery flow or a to and fro signal. During the period 1986-91, 22 cases of carotid dissection have been seen at this unit. A history of trauma, neck pain or Horner's syndrome was only present in five cases. All cases were confirmed by angiography. The small number of cases with these clinical parameters may have been accentuated by our policy of not performing angiograms on patients with a severe deficit who have little to lose from a further stroke. Patients with a severe completed stroke are probably more likely to have neck pain and a Horner's syndrome.

We would endorse the view that clinical parameters are a poor guide to the presence of carotid dissection and that carotid dissection should be considered the most likely diagnosis in all young strokes if no cardiac source of emboli is found.

The ultra sound findings act as a useful screening test and when combined with MRI scanning would seem to obviate the need for conventional angiography.

PRD HUMPHREY
MDM SHAW
Mersey Regional Department of Medical
and Surgical Neurology, Walton Hospital,
Rice Lane, Liverpool L9 1AE, UK

- Mullges W, Ringelstein EB, Leibold M. Non-invasive diagnosis of internal carotid artery dissections. *J Neurol Neurosurg Psychiatry* 1992;55:98-104.
- Bogousslavsky J. Ischaemic Strokes in adults younger than 30 years of age. *Arch Neurol* 1987;44:479-82.
- Eljamel MSM, Humphrey PRD, Shaw MDM. Dissection of the cervical internal carotid artery: the role of doppler/duplex studies and conservative management. *J Neurol Neurosurg Psychiatry* 1990;53:379-83.

Neurocysticercotic versus idiopathic epilepsy

I read with interest the letter by Arruda.¹ The conclusion drawn by the author that "Any patient with late onset epilepsy with normal neurological examination and living in an endemic area of taeniasis/cysticercosis should be considered as having neurocysticercosis regardless of seizure type and EEG findings" is erroneous. The data presented by the author and literature on the subject do not support such contention. Epilepsy has varied causes in any age group. Ahuja *et al*² in a prospective study of late onset epilepsy from India (an endemic area) found cysticercosis as a cause in only 5.1% cases, while no cause could be detected to 75.1% patients. To say that all these patients had neurocysticercosis because they had normal neurological examination and lived in an endemic area would be stretching the imagination too far. It is interesting that none of the patients in the latter group on follow up which extended to 18 months showed any evidence of neurocysticercosis.

To label all patients of late onset epilepsy

with normal examination as neuro-cysticercosis just because the authors did not find any difference in certain parameters studied, is logically incorrect. Since the prognosis and length of treatment required in neurocysticercosis and idiopathic epilepsy may be different, the practical implications of such a statement are obvious. In my opinion, the conclusion drawn by the author is not supported by facts, is unscientific and dangerous.

G K AHUJA
All India Institute of Medical Sciences,
New Delhi-110029, INDIA

- Arruda WO. Neurocysticercosis versus idiopathic epilepsy: a comparative study of 175 patients. *J Neurol Neurosurg Psychiatry* 1991;54:1015.
- Ahuja GK, Mohanta A. Late onset epilepsy. A prospective study. *Acta Neurol Scandinav* 1982;66:216-26.

Arruda replies: I appreciated the comments by Dr Ahuja and I quite agree that the conclusion as presented is misleading.¹ In fact, a single but essential word is missing in the final sentence: possibly! It should read: "Any patient with late onset epilepsy with normal neurological examination and living in an endemic area of taeniasis/cysticercosis, should be considered as possibly having neurocysticercosis regardless of seizure type and EEG findings". This conclusion is supported by previous studies²⁻⁴ showing neurocysticercosis (NC) in 25.5% to 50% of patients with late onset epilepsy and living in endemic areas of these diseases in Brazil and Mexico. Ahuja and Mohanta² report a much lower prevalence of NC in Northern India (5.1%). Nevertheless, NC should always be considered in the differential diagnosis of patients with late onset epilepsy even in these populations due to its possible therapeutic and prognostic implications.

WO ARRUDA
Rua Gonçalves Dias 713,
80-240 Curitiba PR, BRAZIL

- Arruda WO. Neurocysticercotic versus idiopathic epilepsy: a comparative study of 175 patients. *J Neurol Neurosurg Psychiatry* 1991;54:1015.
- Arruda WO. Etiology of epilepsy - a prospective study of 210 cases. *Arq Neuro-psiquiatria* 1991;49:251-54.
- Vasconcelos D, Lombardo L. Crisis epilepticas del adulto: estudio prospectivo de 65 casos. *Gac Med Mex* 1983; 119:23.
- Medina MT, Rosas E, Rubio-Donnadieu F, Sotelo J. Neurocysticercosis as the main cause of late epilepsy in Mexico. *Arch Intern Med* 1990;150:325.
- Ahuja GK, Mohanta A. Epilepsy of late onset - a prospective study. *Acta Neurol Scand* 1982;66:216-26.

Isolated muscle hypertrophy as a sign of radicular or peripheral nerve injury

The article by Mattle *et al*¹ showed that hypertrophy of a single muscle can be a sign of partial nerve or ventral root lesion, and this diagnosis is confirmed by the presence of "profuse spontaneous activity" in the hypertrophic muscle. However, I am concerned about the proposed diagnostic value of this somewhat arbitrary EMG sign and the authors' suggestion that knowledge of this clinical sign might help to avoid "unnecessary, costly, and sometimes invasive investigations", in search of for example, "focal myositis or muscle neoplasms".

I have seen two cases with lipomatous tumours of the calf with profuse spontaneous activity in the "hypertrophic" muscle. Case 1

Figure A) Case 1. CT scan of the calves just below the fibular head. B) Case 2. Ventral aspect of the calves.

was a 48 year old man who presented with the history and clinical signs of L5 and S1 radiculopathy including fibrillations, positive sharp waves, and bizarre low-frequency discharges in all peroneal, and some in the gluteal muscles, associated with unilateral calf enlargement. Distal motor latencies to the peroneal muscles were mildly prolonged. CT scans showed a $9 \times 3 \times 3$ cm lipoma, which was excised, (fig A) compressing both the deep and superficial peroneal nerves, and a homolateral S1 root compression by a slipped disc. Case 2, a 33 year old housewife, noted progressive painless calf enlargement over several years. Eventually, the circumference of the right calf exceeded that of the left one by 9 cm (fig B). Her neurological status was normal. Electromyography revealed dense fasciculations, bizarre low-frequency discharges, and chronic neurogenic alteration of motor unit potentials exclusively in the right triceps surae muscle. On surgery, diffuse lipomatosis was found and confirmed by microscopical examination. In both cases intramuscular areas with "silent" EMG—close to areas with profuse spontaneous activity—were indicative of a non-myogenic nature of the calf enlargements.

Large series suggest that intramuscular tumours are far more common than denerva-

tion hypertrophy.²⁻⁴ Focal myositis or muscle neoplasms, if invading the terminal nerve branches, might induce "profuse spontaneous activity".

HM MEINCK

Section of Clinical Neurophysiology,
Ruprecht-Karls-Universität Heidelberg,
Heidelberg, Germany

- 1 Mattle HP, Hess CW, Ludin H-P, Mumenthaler M. Isolated muscle hypertrophy as sign radicular or peripheral nerve injury. *J Neurol Neurosurg Psychiatry* 1991;54:325-9.
- 2 Kindblom L-G, Angervall L, Stener B, Wickbom I. Intermuscular and intramuscular lipomas and hibernomas. *Cancer* 1974;33:754-62.
- 3 Booher RJ. Lipoblastic tumors of the hands and feet. *J Bone Joint Surg.* 1965;47:727-40.
- 4 Fettel MR, Duffy PE, Rowland PE. Infiltrating lipoma: a cause of monomelic hypertrophy. *Muscle Nerve* 1978;1:75-80.

Mattle et al reply:

We thank Dr Meinck for his comments and the presentation of two additional cases of isolated muscle hypertrophy. However, comparing his and our cases, the EMG results were substantially different. As Dr Meinck states, both his patients showed intramuscular areas which were silent on EMG close to areas with profuse spontaneous activity. Silent EMG areas were not present in our patients. We intended our cases to make a contribution to the understanding of the pathophysiology of isolated muscle hypertrophy after peripheral nerve or root lesions. We showed that this phenomenon is not restricted to the calves and may affect other muscles as well, where the stretch hypothesis for generation of muscle hypertrophy might not be true and profuse spontaneous activity of the muscle fibres might have caused hypertrophy. We agree that tumours are much more common causes of muscle masses and might cause spontaneous activity on EMG when invading terminal nerve branches. However, as stated, there are silent EMG areas within tumours. In these and doubtful cases we also perform MRI.

H MATTLE

Department of Neurology,
Inselspital, CH-3010 Berne,
Switzerland

Iron and Akathisia

The report by O'Loughlin *et al*¹ on the serum iron status of patients with acute neuroleptic-induced akathisia is of interest to us as our own similar investigation revealed no association between the two.² Since the publication of our findings, we have repeated the

study on a further 50 patients recently started on neuroleptic medication, with further negative results (unpublished). Studies of serum iron status in "chronic akathisia" have reported similarly conflicting findings.³⁻⁶ It would therefore be worthwhile at this stage to re-examine the rationale and the methodology of the studies. We commend Dr O'Loughlin and colleagues for their attention to detail, except for the following points: 1) The definition of akathisia used in their study was based on scale scores.³ Not only have the psychometric properties of this scale not been published, it was also designed to rate and not "diagnose" akathisia. As we point out elsewhere,⁷ the issue of appropriately defining akathisia is of paramount importance; 2) The small number of patients with akathisia involved; 3) The failure of the authors to report the differential neuroleptic doses of the two groups. The dose can be an overriding factor that can make the contribution of any other factors difficult to determine; 4) The lack of any correlation between Simpson and Angus scores and akathisia scores is surprising,⁷ suggesting that their patients may not have typical acute akathisia.

There are some important theoretical considerations. What constitutes iron deficiency, and whether serum biochemical measures accurately reflect the state of brain iron, is debatable.⁸ While transferrin is important for the exchange of iron between tissues, its saturation in the serum fluctuates between 20-50% in normal subjects (due to mono- and diferric forms), and small variations in saturation as seen in the above study are unlikely to affect iron delivery. The most important determinant of tissue iron delivery is the total number of transferrin receptors,⁸ and an efficient homeostatic mechanism exists to maintain the rate of iron transported to the tissues. The turnover of iron in the brain is low, and it has a tendency to resist iron depletion,⁹ raising further doubts about the pathophysiological significance of the reported findings. It is also possible that neuroleptic drugs either mobilise iron from peripheral tissues into the brain, or decrease iron turnover in the brain,¹⁰ making peripheral measures of iron status even more suspect in revealing the iron status in the brain. MRI may be one method of overcoming this problem,¹¹ but only good animal models¹² are likely to provide the final answer.

Despite these objections, we feel that there are compelling reasons to study the interaction of iron and neuroleptic drugs. The fall in serum iron and transferrin in the patients with akathisia reported by O'Loughlin *et al* is intriguing, even though its significance for the development of akathisia is questionable. Rosebush and Mazurek¹³ reported somewhat similar findings for patients who developed neuroleptic malignant syndrome. Little is known about the effect of neuroleptics on ferrokinetics. Clearly this area needs further informed investigation.

PERMINDER SACHDEV
Neuropsychiatric Institute,
The Prince Henry Hospital,
Anzac Parade, Little Bay,
NSW 2036, Australia.

- 1 O'Loughlin V, Dickie AC, Ebmeier KP. Serum iron and transferrin in acute neuroleptic induced akathisia. *J Neurol Neurosurg Psychiatry* 1991;54:363-4.
- 2 Sachdev P, Loneragan C. Acute drug-induced akathisia is not associated with low serum iron status. *Psychopharmacol* 1991;103:138-9.
- 3 Brown KW, Glen SE, White T. Low serum iron status and akathisia. *Lancet* 1987;i:1234-6.