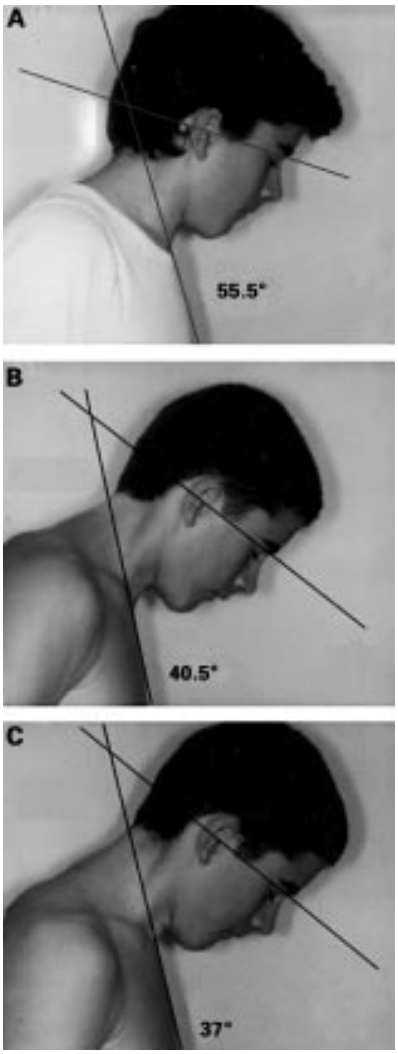

Figure 1 Three different photographs in which progressive amelioration in neck flexion is seen. To make this change objective, we measured the angle between a line joining nasion and tragus and another prolonging the sternum surface. (A) Photograph taken on 22 January 1997. (B) Photograph taken on 24 September 1997. (C) Photograph taken on 15 January 1998.

no abnormalities in nerve conduction. A biopsy of paraspinal musculature was performed and the neuropathological study of the muscle showed non-specific myopathic changes, including slight variability in muscle fibre size, occasional moth eaten fibres, and dilatations of the smooth endoplasmic reticulum, which were filled with fine granular material. The distribution and percentage of fibre types was preserved and no abnormalities in mitochondria and myofibres were seen. Immunohistochemistry to dystrophins, utrophin, and spectrin was normal. A diagnosis of rigid spine syndrome was made and botulinum toxin therapy was begun; with the aim of diminishing the imbalance between the neck flexor and extensor muscular groups to avoid fixed neck extension; for this reason we targeted muscles involved in head extension, principally the trapezium and secondly the esternocleiodomastoid and the paracervical musculature (table 1). During BOTOX therapy continuous improvement in neck flexion was seen, which was also perceived by the patient (fig 1). The radiological and functional measurements confirmed this assertion: in January 1996 the distance from chin to sternum was 10 cm in maximum neck flexion, in December 1997 this distance was 3 cm and in June 1999 the patient was able to touch his sternum with his chin. In march 1999 the patient developed myocarditis, with acute thoracic pain two weeks after a sore throat, increased creatine kinase concentrations, and electrocardiographic changes, with good recovery in 1 week. An echocardiogram performed 3 months later was normal and serology for Coxsackie virus was positive.

BOTOX is the trade mark of the commercialised typeA toxin of *Clostridium botulinum*; BOTOX causes muscle paralysis by acting at nerve endings and blocking presinaptically the release of quanta of acetylcholine[5]; this muscular paralysis is reversible and can ameliorate symptoms in patients with muscle spasms appearing as a manifestation of multiple neurological disturbances,[6–8] including myopathies.[9 10] In some situations this amelioration may become long lasting, and patients will not require further injections.[11] The American Academy of Neurology recommends its therapeutic use in blepharospasm as a primary form of therapy; its use is accepted in cervical dystonia, adductor spasmodic dysphonia, jaw closing dystonia, and in hyperkinesis of hemifacial spasm; its use is considered promising in jaw opening and deviation dystonia, abductor spasmodic dysphonia, and in other focal dystonias.

The origin of spine stiffness in rigid spine syndrome is not well understood. Shortening of paraspinal ligaments or shortening of muscle fibres due to myofibrilar disorganisation have been invoked as possible origins of stiffness[12]; weakness of neck flexors can make this group of muscles incapable of counteracting extensor strength, finally causing spinal rigidity and cervical lordosis. Botulinum toxin may have an important part to play in preventing development of contractures and avoiding stiffness, not only in a symptomatic way, but also in a curative manner, as in our patient.

We thank Ms Julie Myers and Mr Josep Graells for linguistic assessment.

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1 Dubowitz V. "Pseudo" muscular dystrophy. In: Research Committe of the Muscular Dystrophy Group of Great Britain, eds. *Research in muscular dystrophy: Proceedings of the Third Symposium*. London: JB Lippincot, 1965:57–73.
2 Poewe W, Willeit H, Sluga E, *et al*. The rigid spine syndrome—a myopathy of uncertain nosological position. *J Neurol Neurosurg Psychiatry* 1985;**19**:105–8.
3 Moghadaszadeh B, Desguerre I, Topaloglu H, *et al*. Identification of a new locus for a peculiar form of congenital muscular dystrophy with early rigidity of the spine, on chromosome 1p35–36. *Am J Hum Genet* 1998;**62**:1439–45.
4 Giannini S, Ceccarelli F, Granata C, *et al*. Surgical correction of cervical hyperextension in rigid spine syndrome. *Neuropediatrics* 1988;**19**: 105–8.
5 Brin MF. Botulinum toxin: chemistry, pharmacology, toxicity, and immunology. *Muscle Nerve* 1997;**20**(suppl 6):146–68.
6 Davis D, Jabbari B. Significant improvement of stiff-person syndrome after paraspinal injection of botulinum toxin A. *Mov Disord* 1993;**8**:371–3.
7 Giladi N, Honigman S. Botulinum toxin injections to one leg alleviate freezing of gait in a patient with Parkinson's disease. *Mov Disord* 1997;**12**:1085–106.
8 Grazko MA, Polo KB, Jabbari B. Botulinum toxin A for spasticity, muscle spasms and rigidity. *Neurology* 1995;**45**:712–17.
9 Doyle M, Jabbari B. Hypertrophic branchial myopathy treated with botulinum toxin type A. *Neurology* 1994;**44**:1765–6.
10 Nix WA, Butler IJ, Roontga S, *et al*. Persistent unilateral tibialis anterior hypertrophy with complex repetitive discharges and myalgia: report of two unique cases and response to botulinum toxin. *Neurology* 1992;**42**:602–6.
11 Heinen F, Wissel J, Philipsen A, *et al*. Interventional neuropediatrics: treatment of dystonic and spastic muscular hyperactivity with botulinum toxin A. *Neuropediatrics* 1997;**28**:307–13
12 Lotz BP, Stügben JP. The rigid spine syndrome: a vacuolar variant. *Muscle Nerve* 1993;**16**:530–6.

"Hot cross bun" sign in a patient with parkinsonism secondary to presumed vasculitis

Brain MRI is an important tool in the investigation of patients with unusual parkinsonian syndromes. The "hot cross bun" sign is a radiological sign which has been said to be highly specific for multiple system atrophy.[1] However, we now report on a patient with the hot cross bun sign who presented with parkinsonism secondary to presumed vasculitis.

Our patient was a 31 year old woman who was referred with an 18 month history of double vision, balance problems, and deafness. Brain MRI performed 9 months before this admission had demonstrated a non-enhancing swelling of the pons (fig 1 A). She had not responded to a 4 week course of oral adrenocorticotropic hormone treatment at that time. On admission to our unit there had been no change in her symptoms. On examination she had mild cognitive impairment (mini mental state score 24/30) and a labile affect. She had a bilateral horizontal supranuclear gaze palsy. In addition she had a right upper motor neuron facial palsy and bilateral sensorineural deafness (confirmed by audiometry). Examination of her limbs showed axial and bilateral limb rigidity. She exhibited bradykinesia but did not have a resting limb tremor. She had signs of cerebellar ataxia in all her limbs and walked with a broad based gait requiring the assistance of another person. Limb power and sensation were normal and her plantars were flexor. There was no evidence of dysautonomia or rheumatological disease.

Blood investigations showed a raised erythrocyte sedimentation rate at 36 mm/hour, raised serum IgG at 21.6 g/l (with a polyclonal pattern on electrophoresis), a positive rheumatoid factor titre (>1:320), a positive speckled ANA titre (>1:640), and positive anti-Ro antibodies (33 units). Schirmer's test, thyroid function tests, copper studies, and manganese were all negative or normal. Brain MRI showed severe atrophy of the medulla, pons, cerebellum, and middle cerebellar peduncles with cross shaped T2 signal hyperintensity within the pons (hot cross bun sign) and high signal change in the middle cerebellar peduncles (fig 1 B). There were no supratentorial lesions. Phase contrast MR angiography of the brain was normal. Examination of CSF showed no increase in cells

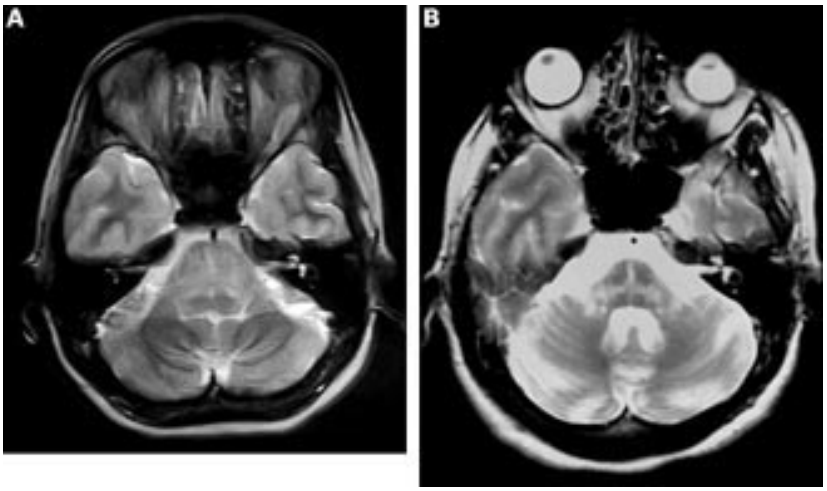

Figure 1 (A) Initial T2 weighted brain MRI of the patient disclosed non-enhancing pontine swelling. (B) Nine months later T2 weighted MRI showed severe atrophy of the medulla, pons, cerebellum, and middle cerebellar peduncles with cross shaped T2 signal hyperintensity within the pons (hot cross bun sign) and high signal change in the middle cerebellar peduncles.

and normal protein, lactate, and glucose; however, CSF electrophoresis demonstrated intrathecal oligoclonal IgG production. The patient was treated with pulsed intravenous cyclophoshamide and a reducing course of steroids but did not improve significantly. There has been no further deterioration since treatment.

The hot cross bun appearance in multiple system atrophy is due to loss of pontine neurons and myelinated transverse pontocerebellar fibres with preservation of the corticospinal tracts which run craniocaudally.[2] Our patient presented with a severe parkinsonian syndrome associated with cerebellar and brain stem dysfunction. The absence of dysautonomia together with the initial MRI appearance of swelling of the pons made the diagnosis of multiple system atrophy extremely unlikely. Although she had a supranuclear gaze palsy her scans were not typical of progressive supranuclear palsy.[1] The serological and CSF findings together with initial pontine swelling suggested probable vasculitis, a recognised cause of parkinsonism.[3] Wallerian degeneration secondary to vasculitic infarction results in hyperintensity on T2 weighted MRI.[4] The hot cross bun sign in our patient may reflect selective wallerian degeneration of transverse pontocerebellar fibres. Thus, the clinical findings of this case highlight the need to consider alternative diagnoses to multiple system atrophy in patients with the hot cross bun sign.

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1 Schrag A, Good CD, Miszkiel K, *et al*. Differentiation of atypical parkinsonism syndromes with routine MRI. *Neurology* 2000;**54**:697–702.
2 Schrag A, Kingsley D, Phatouros C, *et al*. Clinical usefulness of magnetic resonance imaging in MSA. *J Neurol Neurosurg Psychiatry* 1998;**65**: 65–71.
3 Walker RH, Spiera H, Brin MF, *et al*. Parkinsonism associated with Sjogren's syndrome: three cases and a review of the literature. *Mov Disord* 1999;**14**:262–8.
4 Ramsey R, ed. *Neuroradiology*. 3rd ed. Philadelphia: WB Saunders, 1994:514.

BOOK REVIEWS

Charcot-Marie-Tooth disease. A practical guide. Compiled by CMT INTERNATIONAL UK (Pp 113, £10.00). Published by CMT International UK, Penarth, 2000. ISBN 0 9533883 0 1.

Charcot-Marie-Tooth disease. A practical guide, is a book compiled by CMT International UK with the aim of providing an overview of Charcot-Marie-Tooth disease (CMT) with a particular emphasis on providing practical day to day advice for living with the disease. It is aimed at doctors and patients and other people involved with CMT . It is well written and excellently presented and provides a range of information that the intended audience will find invaluable.

The book is divided into three main sections. The first section deals with genetic and medical issues. The known genetic variants of the disease are well described and accurate except for one mistake stating that the gene duplication that causes CMT1a is on chromosome 22 when it is actually on chromosome 17. I thought the section covering CMT inheritance was particularly well presented and illustrated using simple diagrams to explain the various inheritance mechanisms. There was also a very useful glossary of scientific and medical terms in this section.

The second section deals with living with CMT. This covers many important areas for the patient including coming to terms with the diagnosis, care of the feet, pain control, and secondary complications. Foot deformities and their surgical correction were particularly well covered.

The third section deals with practical issues including finding work, having a baby, driving, and CMT and aids to daily living. This section is particularly useful in providing contact details for many different organisations who will help patients. All three sections are supported by informative appendices.

This book is an excellent patient oriented guide, full of useful information and contacts. It will be a particularly useful book to recommend to newly diagnosed patients.

MARY REILLY

Conduct disorders in childhood and adolescence. Edited by JONATHAN HILL and BARBARA MAUGHAN (Pp 569, £39.95). Published by Cambridge University Press, Cambridge, 2000. ISBN 0 521 78639 8.

This is an extremely interesting and informative book that does justice to the complexity of perspectives on child and adolescent conduct problems. It is evident that considerable attention was given to shaping this book, which succeeds in being more than a collection of papers on conduct problems. Individual authors have been careful to introduce their particular area of interest to readers unfamiliar with their field. For example, Herbert and Martinez's chapter on "Neural mechanisms underlying aggressive behaviour" is a lucid account available to a novice reader. Throughout the book there are discussions that refer to other theoretical perspectives, thus illuminating the theoretical, methodological, and clinical issues. Reading the book is rather like a mental brass rubbing in that the reader's patience is rewarded by the emergence of an increasingly complex but fascinating pattern of relations between biological, genetic, neuropsychological, social, interactional and psychological stand points.

The book moves back and forth between chapters that contextualise, for example the historical perspective offered by Costello and Angold's chapter, to consideration of very specific mechanisms such as Lynham and Henry's chapter on the role of neuropsychological deficits and Petit, Pohlaha, and Mize's chapter on perceptual and attributional processes. Each chapter gives a critical view of relevant research and raises methodological concerns. The spirit of the book is captured in Hill's chapter on biosocial influences, in which he conveys a sense of curiosity about the interaction between biological and social phenomena and how that might be further investigated.

Kazdin gives careful attention to treatment of conduct disorders in an excellent chapter. Le Marquand, Tremblay, and Vitaro consider issues of prevention and Knapp's chapter brings forward the economic costs of conduct disorder.

In conclusion I return to the subjects of this work, the children and young people, and their families who experience great emotional distress and difficulty, very often in the context of socioeconomic hardship. Inclusion of qualitative research would have further enriched this book, by bringing their voices more directly into the important debates so elegantly presented. It deserves to become a standard work, available widely to all clinicians and researchers interested in this field.

MOIRA DOOLAN

Half a brain is enough: the story of Nico. Edited by ANTONIO M BATTRO (Pp 118, £12.95). Published by Cambridge University Press, Cambridge, 2000. ISBN 0 521 78307 0.

This short book describes the fascinating recovery and remarkable neurocognitive compensation of Nico, a little boy who at the