

insonism has been reported; that patient also had pyramidal signs.<sup>4</sup> A further patient exhibited dopa-responsive tremor and facial impassivity during recovery from a more typical presentation of pontine myelinolysis with stupor, abnormal eye movements and tetraparesis.<sup>5</sup> Pathological changes in the basal ganglia have been well documented in typical cases of pontine myelinolysis,<sup>6</sup> and it has been suggested that the pontine lesion masks the extra-pontine clinical features. In our case the large lesion in the pons was clinically silent. MRI is clearly the investigation of choice in patients presenting with neurological syndromes associated with hyponatraemia; subclinical or clinically atypical pontine myelinolysis may be more common than is currently realised.

We are grateful to Dr S P Kane for permission to report a patient under his care.

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### Visual hallucinations and the cholinergic system in dementia

Cholinergic deficits are associated with various forms of dementia, including Alzheimer's and Parkinson's disease, in which they relate to the degree of cognitive impairment.<sup>1</sup> New findings on the neurochemical pathology of senile dementia of Lewy body type (SDLT) suggest that particularly extensive cholinergic abnormalities in certain neocortical regions are associated with visual hallucinations. SDLT may be the second most common form of dementia in the elderly (after Alzheimer's disease).<sup>2</sup> It is characterised, clinically, by acute presentation with confusion frequently accompanied by hallucinations and, neuropathologically, by the presence of Lewy bodies, particularly in archicortical areas. The relative absence of neocortical neurofibrillary tangles in SDLT<sup>2</sup> also suggests such patients might be more amenable to transmitter replacement therapy.

In 12 SDLT cases with frozen tissue available for neurochemical analyses, visual

Table Cortical choline acetyltransferase activities<sup>a</sup> in senile dementia of Lewy body type<sup>b</sup> mean, SD

	Parietal	Temporal	Occipital
Normal	10.8, 5.2	8.7, 5.6	11.6, 3.4
SDLT	(7)	(11)	(8)
Without			
hallucinations	5.1, 2.7	4.0, 1.7	9.2, 4.0
	(5)	(5)	(6)
With			
hallucinations	1.6, 1.4	1.5, 1.6	6.0, 4.2
	(6)	(6)	(4)

<sup>a</sup>ChAT expressed as nmol/mg protein<sup>3</sup> in Brodmann areas 39 or 40 (parietal), 21 or 22 (temporal) and 17, 18 or 19 (occipital). Case numbers in parentheses.

<sup>b</sup>In parietal and temporal but not occipital cortex, ChAT was significantly different between the two subgroups ( $p = 0.009$  and  $0.015$ , respectively, Mann Whitney U test). The normal and SDLT groups were matched for age and postmortem delays mean (SD) 78 (6), 81 (6) and 76 (7) years; 27 (12), 41 (24) and 23 (15) hours, respectively).

hallucinations had been noted in six at presentation and continued throughout the course of the disease; in the remainder this symptom was not recorded. Choline acetyltransferase (ChAT) activities were reduced in both SDLT sub-groups compared with normal levels but were significantly lower in those with hallucinations compared with those without for two of three cortical areas examined (see table). Thus in hallucinating cases ChAT activities in parietal and temporal cortex were reduced by 80-85% compared with 50-55% in the non hallucinating cases. No other neurochemical or neuropathological parameter so far examined (including dopamine, serotonin, cholinergic receptors, Lewy bodies or plaques) divided the two groups.

The suggestion that extensive ChAT loss in SDLT cases with hallucinations may be related to this clinical feature is supported by psychopharmacological data. Thus, although no patient was receiving any anticholinergic medication at the time they presented, drugs such as scopolamine are known in certain cultural<sup>4</sup> or medical<sup>5</sup> situations to induce hallucinations. Severe degeneration of cholinergic neurons innervating certain cortical areas in dementing disorders such as SDLT may similarly give rise to this symptom. If confirmed, this observation is important in relation to the function of cholinergic innervation of human neocortex (innervation of other areas such as hippocampus being involved in memory) and in relation to selecting suitable cases of dementia for cholinergic therapy (those with hallucinations having a more profound cholinergic defect).

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### Type 1 Arnold-Chiari malformation in a 77 year old woman

Chiari malformations may present in a variety of ways, most commonly between the ages of 20 and 60 years.<sup>1-3</sup> Onset of symptoms after the seventh decade is distinctly unusual.

A 77 year old woman had fallen and was found unconscious by her general practitioner. She was thought to have aspirated. She stopped breathing before reaching hospital but was successfully resuscitated and needed artificial ventilation for a week. As she recovered, frequent choking and aspiration of fluids were noted. On closer questioning she admitted having dysphagia with nasal regurgitation over a two year period. During the preceding six months she had become unsteady and dizzy, with a tendency to fall, and there had been fleeting episodes of diplopia. Some years before she had had bilateral total hip replacement and had experienced several serious falls.

The right hip prosthesis had loosened in a recent fall and she was unable to walk. Her eye movements were full and there was no nystagmus in the primary position. There was down beating nystagmus on downgaze and obliquely down beating nystagmus on lateral gaze to either side. Palatal movement and gag reflexes were absent although pharyngeal sensation and tongue movements were preserved and speech was normal. Fluid aspiration occurred consistently and she was fed through a nasogastric tube. Her limbs were ataxic, but there were no pyramidal features.

A sleep study demonstrated considerable nocturnal hypoxia ( $\text{SaO}_2 < 80\%$ ). MR imaging showed cerebellar hypoplasia and tonsillar herniation down to the level of the second cervical vertebral body (fig). The medulla was elongated and kinked over the odontoid peg. There was mild associated hydrocephalus. Her foramen magnum was decompressed, the dura incised and a fascial graft was inserted. Although there was no change in the physical signs following operation, she was able to swallow fluids normally without choking, aspirating or regurgitating. The nasogastric tube was successfully removed and she was discharged from hospital.

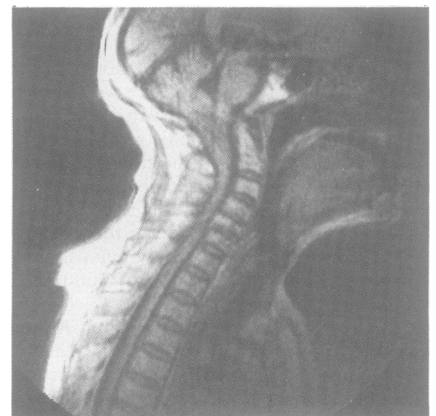


Figure Sagittal MR image.

This case illustrates how Chiari malformations may present at any age and should be considered even in septuagenarians. The onset of symptoms in the eighth decade is extremely unusual, but presentation at the age of 74 has been reported in one case.<sup>3</sup> Symptoms may precede diagnosis by several years,<sup>1</sup> as in this patient. However, it is unclear why a congenital malformation should suddenly become symptomatic. Several reports have highlighted the role of trauma,<sup>4</sup> degenerative changes in the cervical spine and cerebrovascular disease that may also contribute to neurological deterioration.

Palatal paresis, dysphagia, diplopia and ataxia are well recognised symptoms of type 1 Arnold-Chiari malformations,<sup>1,2</sup> and recent reports have highlighted the respiratory problems that these patients may encounter.<sup>5,6</sup> The presence of downbeat nystagmus strongly implied an abnormality at the cranio-cervical junction and this was confirmed, non-invasively, by MR imaging.

Despite their ages both our patient and the one reported by Hosford and Spector<sup>3</sup> made a good recovery following surgical decompression showing that age alone should not be regarded as a barrier to active management.

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as Ramsay Hunt syndrome. Review and restudy of this material established the diagnosis of mitochondrial encephalomyopathy (MERRF) in 11 of the 13 cases; the remaining two cases were not available for reexamination.<sup>3-5</sup> Those who wish to preserve the Ramsay Hunt syndrome differ widely in their concept of the disorder,<sup>6,7</sup> which only reinforces our view that the term should be buried.

Tassinari *et al* recently reported a series of 13 patients with "Ramsay Hunt syndrome" who had onset of myoclonic or tonic-clonic seizures at ages 6 to 15 years with a mild cerebellar syndrome. Family studies suggested autosomal recessive inheritance and muscle biopsies failed to show evidence of mitochondrial disease.<sup>7</sup> Tassinari's patients are different from the cases that we reclassified as MERRF and we agree that they do not have mitochondrial disease. The clinical, electroencephalographic and genetic features of their patients are, however, identical to those of Unverricht-Lundborg disease (Baltic myoclonus) as described by the original authors and more recently in the definitive studies of Koskiniemi.<sup>8-10</sup> There is no doubt that this disorder occurs outside the Baltic region.<sup>2,11</sup> Although there is as yet no diagnostic laboratory marker for Unverricht-Lundborg disease, the clinical picture is distinctive and a clinical diagnosis can be made with considerable certainty.<sup>2,10,11</sup> We find the use of the term "Ramsay Hunt syndrome" for such patients is historically inaccurate and diagnostically misleading.

Tassinari *et al* have emphasised the electroencephalographic features of their patients with normal or mildly slow waking background activity, fast spike-wave discharges, photosensitivity and lack of activation during slow wave sleep.<sup>7</sup> These findings are no different from those of Unverricht-Lundborg disease.<sup>12</sup> Indeed, critical study of the EEG patterns in all the PME, including MERRF and Unverricht-Lundborg disease, reveals more similarities than differences.<sup>13</sup> Tassinari *et al*, have also described vertex spikes during REM sleep.<sup>7,14</sup> Unfortunately, REM studies of Unverricht-Lundborg disease have not been reported. These spikes are, however, also seen in MERRF<sup>15</sup> and we suspect they may be common to all the PMEs, much like giant evoked potentials, which they may in fact represent.

Unlike the situation previously, the vast majority of patients with PME can now be accurately diagnosed during life.<sup>2</sup> Whilst occasional undiagnosed patients remain, there is no residual homogeneous group of cases for which the term Ramsay Hunt syndrome is appropriate. Radermecker's frustration at attempting to define the Ramsay Hunt syndrome<sup>1</sup> can at last be put to rest. In retrospect it was a true syndrome, with many causes, although we suspect that most reported cases were probably examples of MERRF. Its value as a clinical category has been overtaken by clinical, genetic, biochemical and pathological advances in specific diagnosis.<sup>2</sup>

(We) come to bury . . . , not to praise . . .

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#### Tassinari *et al* reply:

We thank Drs Berkovic and Andermann for the kind comments they made on our recent paper.<sup>1</sup> We are glad to know that the patients previously referred to as Ramsay Hunt syndrome (RHS) by these authors and subsequently found to have a mitochondrial encephalopathy (MERRF) were significantly different from those we have described under the eponym of RHS. This fact supports our statement that RHS and MERRF exhibit different clinical, EEG and evolutive features.

Drs Berkovic and Andermann however criticise the term RHS applied to our cases. In these patients the main clinical complaint was action myoclonus combined with rare generalised epileptic seizures: indeed this association was described by Ramsay Hunt under the heading of "Dyssynergia Cerebellaris Myoclonica".<sup>2</sup> Ramsay Hunt also emphasised the coexistence of "cerebellar ataxia" but, in such cases, the cerebellar component is difficult to define because of the presence of severe intention myoclonus, as recently pointed out by Harding.<sup>3</sup> Thus, in our opinion, the use of the term RHS for our patients is justified,

## MATTERS ARISING

### Ramsay Hunt syndrome: to bury or to praise

"If there is one disease in the neurological literature that is difficult to define and demarcate, it is the cerebellar dyssynergia of Ramsay Hunt." Radermecker, 1974.<sup>1</sup>

Our recent suggestion that the Ramsay Hunt syndrome (dyssynergia cerebellaris myoclonica) is no longer a useful diagnostic category<sup>2-5</sup> has provoked considerable controversy.<sup>6,7</sup> We reached this conclusion following a study of 84 cases of progressive myoclonus epilepsy (PME), of which 13 were previously regarded