Eight days following hyperbaric treatment, he received one right unilateral ECT IM one hour prior to the procedure. He was anaesthetised with 2 mg/kg sodium pentathol IV and 40 mg succinyl choline IV without complications. Within several hours, the patient began exhibiting prominent verbal sexual innuendos, undressing and exposing himself, and lunging at the breasts of female staff and groins of male staff. These outbursts of sexually aggressive behaviour lasted 10 to 15 minutes and were interspersed with otherwise lethargic and apathetic behaviour. The patient ate excessively, constantly complained of hunger, showed excessive handling of objects, and was easily distracted. He displayed prominent amnesia, both retrograde and anterograde, and denied any memory of his inappropriate behaviour. Disorientation to time and severe attention deficit were present. Neurological examination revealed ataxia, moderate rigidity, and brisk muscle stretch reflexes. Electroencephalography demonstrated low-to-moderate voltage 4 to 6 Hz semirhythmic theta and paroxysmal high voltage rhythmic 5 Hz theta activity. These diffuse paroxysmal theta bursts were associated clinically with blank staring followed by agitation and poor compliance with commands. The patient was placed on phenytoin. Haloperidol and lorazepam were required to control the outbursts of aggressive hypersexual behaviour that continued to alternate with long periods of somnolence. The inappropriate behaviour resolved over a course of 10 days and neuropsychiatric and benzodiazepines were gradually withdrawn.

Repeat EEG 10 days after the initial recording revealed minimal-to-moderate generalised 6 to 7 Hz theta without paroxysmal irregularities. Cranial CT scan was normal. Mental status examination progressively improved in all spheres. At discharge 4 weeks following admission, neuropsychological testing revealed only mild-to-moderate deficits in naming and recent memory. There was no evidence of inappropriate behaviour or neurologic deficits. As the encephalopathy cleared, the patient’s primary affective disorder again became more apparent.

In 1937, Klüber and Bucy described an unusual syndrome appearing in rhesus monkeys following bilateral temporal lobectomy.14 These monkeys displayed (1) “psychic blindness,” (2) hyperorality, (3) an increased tendency to touch and examine objects (“hypermetamorphosis”), (4) loss of normal anger, fear, and pleasure responses, and (5) increased and altered sexual behaviour. The human Klüber-Bucy syndrome (KBS) closely parallels the syndrome seen in monkeys, involving (1) visual agnosia (especially prosopagnosia), (2) hyperphagia, bulimia, (3) hypermetamorphosis, (4) placidity, apathy, and blunted affect, and (5) alterations in sexual behaviour.5 In addition, the human KBS is accompanied by more complex behavioural manifestations, such as aphasia, amnesia, dementia, and/or seizures.5 Partial or complete human KBS has been described in association with a variety of encephalopathic processes invariably associated with bilateral temporal lobe involvement. This paper is the first description of human KBS occurring in association with delayed carbon monoxide leucoencephalopathy.

Neuropsychiatric sequelae of carbon monoxide poisoning occur in 15–40% of surviving patients.6 These sequelae frequently develop after a period of apparent recovery from the carbon monoxide exposure. The clinical picture of delayed neuropsychiatric deterioration following carbon monoxide poisoning has been described in numerous case reports.7 These patients are usually deeply comatose following carbon monoxide inhalation but regain consciousness within 24 to 48 hours. Subsequent improvement is rapid, and they are often discharged from the hospital within 1 week. Choi found that a period of apparent recovery between the carbon monoxide insult and the onset of neuropsychiatric sequelae occurred in 11.8% of patients requiring hospital admission.8 The latent period varied from 2 to 40 days with a mean of 22.4 days. Recovery from the delayed sequelae occurred in 75% of patients within 1 year.

Similar descriptions of delayed post-anoxic encephalopathy have been reported following anaesthetic complications, cardiac arrest, hypotension, and anoxic anoxia.79 Several authors have reported that the delayed onset of neurological deterioration occurred immediately following an increase in patient activity, after discharge from the hospital, after emotional stress, and in one case, following ECT.6910 While a period of bed rest following carbon monoxide poisoning has been recommended as a means of preventing delayed post-anoxic deterioration, it has yet to be determined that this therapeutic measure is effective.

The usual neuropathological substrate of delayed neuropsychiatric deterioration following carbon monoxide poisoning is believed to be selective injury to the cerebral white matter.711 Cases with a delayed onset
Letters

of neuropsychiatric deterioration demonstrate focal or confluent plaques in the deep central white matter and the periventricular zones. There is diffuse, spongy demyelination with almost complete sparing of the axons.

In 1965, Geschwind discussed the behaviour seen in rhesus monkeys with KBS in relation to visual-limbic disconnections resulting from fibre tract lesions in the temporal lobe. Because temporal lobectomy monkeys demonstrate agnosias in modalities other than vision, a visual-limbic disconnection would not be expected to account for the entire Klüver-Bucy syndrome. However, experimental studies have shown that features of KBS can be produced in monkeys by removing the visual influences of the striate cortex from the temporal lobe. This was accomplished by destroying the temporal lobe on one side, and then destroying the contralateral visual cortex and the crossing visual fibres in the splenium of the corpus callosum. Features of KBS have also been produced both by interrupting the white matter tracts connecting the occipital and temporal lobes, and by severing the fibre tracts entering and exiting the temporal lobes medially.

We have described the case of a 32 year old man who developed KBS as a delayed post-anoxic syndrome following carbon monoxide poisoning. That the onset of this syndrome occurred suddenly several hours following an ECT treatment suggests that ECT may have exacerbated or precipitated the appearance of delayed neuropsychiatric deterioration. The transient nature of the KBS seen in our patient is in keeping with other descriptions of delayed post-anoxic leukoencephalopathy and suggests that the proposed disconnections are reversible.

Geschwind’s hypothesis that disconnections resulting from fibre tract lesions may be associated with KBS in monkeys is supported by the experimental investigations discussed above. The neuropathological substrate of delayed neuropsychiatric deterioration following carbon monoxide poisoning is believed to be selective white matter injury. This case of KBS as a delayed clinical syndrome following carbon monoxide poisoning suggests that disconnections resulting from white matter lesions may also underlie the human KBS.

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Matters arising

Local autonomic failure affecting a limb

Sir: I refer to the article Local autonomic failure affecting a limb, by R H Johnson and B J Robinson. The authors reported three cases where sudomotor and vasomotor dysfunction occurred together in one limb. They state that, while segmental anhidrosis has been reported in association with Holmes-Adie syndrome (HAS), their patients showed no features of that disorder. They did not mention an important difference between their cases and cases reported in association with HAS: viz that while their patients showed evidence of a preganglionic lesion (preservation of sweating around intradermal injection of acetyl choline), cases reported in association with HAS in the main have shown evidence of ganglionic or post-ganglionic lesions (lack of sweating in response to subcutaneous methacholine or pilocarpine). This notwithstanding, it seems premature to dismiss a connection with HAS completely. There has been one report of a case where a patient with normal pupils presented with segmental anhidrosis, but 6 years later developed a typical Adie pupil. Furthermore Johnson & Robinson do not mention the results of pupillometry or the results of instillation of methacholine in their cases. The observation that one of Johnson & Robinson’s patients had some ill-defined loss of sensation to pinprick in the area of sudomotor loss is of further interest in this connection. I have recently seen a case of HAS (confirmed by pupillometry and sensitivity to 2.5% methacholine) in whom, although there were no features of autonomic dysfunction, there was an area of sensorial loss to pinprick on the medial border of the right forearm, from wrist to elbow. Extensive investigation, including electrophysiological studies, myelography and spinal MRI scan showed no lesion to account for this. The possibility of a relationship between the cases of Johnson & Robinson and HAS (or the syndrome of HAS-plus, so to speak) remains to be discounted.

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Kluver-Bucy syndrome associated with delayed post-anoxic leucoencephalopathy following carbon monoxide poisoning.

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