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Two consecutive cases of probable sporadic Creutzfeldt-Jakob disease (CJD) were diagnosed in year 2008. They both presented with progressive dementia with moderate-to-severe cognitive impairment at the time of presentation to our institution. The Mini-Mental State Examination scores were 12 and 13 out of 30. Magnetic resonance imaging (MRI) and magnetic resonance angiography was performed for both patients. Both patients showed cortical grey matter lesions without any involvement of the deep grey nuclei, particularly the caudate nuclei. The cortical grey matter showed subtle increase in signals on T2-weighted MRIs with restricted diffusion. No particular MR signal change could be seen on the T1-weighted images, fluid-attenuated inversion recovery (FLAIR) images or the post-gadolinium contrast enhanced images.

Subsequent electroencephalogram showed characteristic periodic synchronous discharges. Clinically, myoclonus and extra-pyramidal signs were more obvious in subsequent follow-up of the patients.

Early recognition of this deadly condition is crucial as human-to-human transmission is possible and should be prevented by all means; especially when we are in an era of organ transplant.

Paediatric Epilepsy Surgery Programme in Hong Kong—Experience in Queen Mary Hospital/Duchess of Kent Children's Hospital

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Background: Surgery is a well-established treatment for adults with intractable seizures. Increasingly, infants and children are being considered for epilepsy surgery. In a growing child, epilepsy surgery has the additional benefit of aborting cognitive decline and improving development and behaviour.

Methods: The paediatric epilepsy surgery programme as well as paediatric video telemetry service were set up in Queen Mary Hospital since the early 1990s. From 1998 to 2006, a total of 10 patients were sent overseas for resective surgery. One patient suffered from Rasmussen's syndrome, three patients with tuberous sclerosis, three with focal cortical dysplasia, three with hypothalamic hamartoma. Since 2001, regular epilepsy surgery joint clinic was set up. From 2002 till now, 14 patients underwent curative epilepsy surgery at our centre. One patient suffered from Rasmussen's syndrome, three patients with multi-lobar cortical dysplasia, four patients with focal cortical dysplasia, one with tuberous sclerosis, two with dysembryoplastic neuroepithelial tumour, one with gliosis, two with mesial temporal sclerosis.

Results: Age at operation ranged from 3 months to 19 years. Age from seizure onset to surgery ranged from 2 months to 16 years. Seizure outcome at a mean of 3.5 years long-term follow-up (3 months-7 years), using Engel's classification, was class 1 in five (36%) patients, class II in 5 (43%) patients, and class III in 3 (21%) patients.

Conclusion: With careful case selection, early surgical intervention in paediatric patients with intractable epilepsy is associated with favourable outcome and provides an important opportunity in preventing irreversible decline in intelligence and disability.