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Author(s)	Wong, VCN; Yung, AWY
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Risk Factors Associated with Refractory Epilepsy in Children—The University of Hong Kong Experience

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Virginia CN Wong, Ada WY Yung

Division of Child Neurology/Developmental Paediatrics/Neurohabilitation, Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital/Duchess of Kent Children's Hospital, The University of Hong Kong, Hong Kong

Background: There is a lack of consensus about the definition of intractable or refractory epilepsy in children. Medically intractable epilepsy occurred in 10 to 20% of epilepsy with childhood onset. Patients with medical intractability had immense resource implication and lifelong disability/disabilities. Early identification of risk factors for refractory epilepsy offers a chance of appropriate and timely treatment thus affecting prognosis.

Methods: A retrospective study was performed for our cohort of 505 children aged below 18 years with new-onset epilepsy, diagnosed between 1979 and 2006, and actively managed at the Comprehensive Epilepsy Clinic, Department of Paediatrics and Adolescent Medicine of the University of Hong Kong. We arbitrarily defined refractory epilepsy as those who had never been seizure-free for more than 12 months despite receiving anti-epileptic drug (AED) treatment. Responders were arbitrarily defined as those who had at least been seizure-free for consecutive 12 months. All patients had been on one or more AEDs and were followed up for at least 24 months after AED initiation. The demographic, clinical, diagnostic, investigative, management and seizure outcome at 2 years were analysed.

Results: At 2 years' follow up, 42% (n=212) had refractory epilepsy. Risk factors significantly correlated with refractory epilepsy included history of status epilepticus ($P<0.001$), symptomatic aetiology ($P<0.001$), use of two or more AEDs ($P=0.001$), abnormal neurological co-morbidities including mental retardation ($IQ<70$) [$P<0.001$], learning disabilities ($IQ=70-90$) [$P=0.009$], cerebral palsy ($P=0.011$), abnormalities in EEG ($P<0.001$) and neuroimaging ($P<0.001$).

Conclusions: Early identification of risk factors to predict possible medical intractability is important in improving treatment strategies especially in the selection of traditional versus newer AEDs, mono- versus poly-pharmacy or even earlier alternative epilepsy management decision plans including evaluation for possible surgical therapies.

Posterior Reversible Encephalopathy Syndrome: Paediatric Heart Transplant with Cyclosporine Neurotoxicity

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W Tso¹, AWY Yung¹, KS Lun², TC Yung², K Fan³, VCN Wong¹

¹ Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital, The University of Hong Kong, Hong Kong

² Department of Paediatric Cardiology, Queen Mary Hospital, The University of Hong Kong, Hong Kong

³ Department of Cardiology, Grantham Hospital, Hong Kong

Posterior reversible encephalopathy syndrome (PRES) is associated with a specific disorder of cerebrovascular autoregulation. Clinical features of PRES consisted of headache, decreased consciousness, altered mental functioning, seizures, visual loss or cortical blindness. Characteristic findings on neuroimaging included high signal intensity on T2-weighted as well as diffusion-weighted imaging MRI in the posterior cerebral hemispheres, indicative of vasogenic subcortical oedema without infarction. Cyclosporine neurotoxicity had been described following bone marrow and organ transplantation; however, there are few reports of PRES in children especially post-paediatric heart transplantation. We report a case of cyclosporine-related PRES in a paediatric heart transplant recipient. She made a good recovery with no residual neurological deficits after withdrawal of cyclosporine, control of possible risk factors as well as symptomatic control of seizure.