

## Conclusions

Children with headache may not present as the typical headache pattern in adults. While the IHS criteria was developed primarily for headache disorders in adults, the number of tension headache and migraine may be underestimated in our study. A longer follow-up period is required for establishing a definite diagnosis for the type of headache in children. Male is more affected than girls. This was consistent with Caucasian's data. Co-occurrence of other pain symptoms was commonly seen in our children.

## References

1. Lewis et al. Practice parameter: evaluation of children and adolescents with recurrent headaches. *Neurology* 2002;59(4):490-8.
2. Headache Classification Committee of the International Headache Society: The international Classification of Headache Disorders 2<sup>nd</sup> edition. *Cephalalgia* Volume 24 Suppl 1 2004.

## Spinal Muscular Atrophy (SMA) – Natural History and Functional Status in Hong Kong Children P 21

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### Objective

To study the natural history and current functional status of children with SMA in Hong Kong.

### Methods

A SMA Registry had been collected since 1984 in Duchess of Kent Children's Hospital. Families of SMA (FSMA) has been established in 2000. A total 102 SMA cases had been collected. We analyzed 81 SMA cases (34 males; 47 females) with clinical information available in September 2002. SMA cases were classified according to classification of the International SMA Consortium. SMA type III was further classified into IIIa (onset <3 years) and IIIb (>3 years). The validated Chinese Functional Independence Measure for Children (WeeFIM) was administered to 39 active SMA cases to assess their performance in daily functional skills in 3 domains (self care, mobility and cognition).

### Results

Our SMA Registry consisted of type I (severe) = 20 (24.7%); type II (intermediate) = 26 (32.1%) and type III (mild) = 35 (43.2%).

The survival probability for SMA I was 55%, 40%, 30%, 30%, 30%; for type II was 100%, 100%, 100%, 92%, 88% at 1, 2, 4, 10, and 20 years of age respectively.

The probability of being ambulatory (defined as walking with or without assistance) at 2, 4, 10, 20 years of age for SMA II was 64%, 59%, 32%, 5%; and 100%, 100%, 100%, 82%, 71% for type IIIa; and 100%, 100%, 89%, 67% for type IIIb.

WeeFIM score: The total scores were 30 (N=4; mean total quotient = 24%), 72 (N=15; mean total quotient = 57%), 94 (N=9; mean total quotient = 75%) and 97 (N=11; mean total quotient = 78%) for types I, II IIIa and IIIb respectively.

For the domain of mobility, more than 90% of type I, II, IIIa and 63% of those with type IIIb required assistance. Around 55% SMA type IIIa and b cases attained functional independence in both self-care and cognition domains.

### Conclusion

The natural history of progression for Chinese SMA patients was similar to the Caucasians. Knowledge on the prognosis such as long term survival and functional status is important for counseling.

Survival probability in SMA types I and II and probability of being ambulatory for SMA types II, IIIa and IIIb

SMA type	Age (y)					
	1	2	4	10	20	40
Survival Probability at a certain age (%)						
I (N=20)	55	40	30	30	30	—
II (N=26)	100	100	100	92	88	85
Probability of being ambulatory at a certain age (%)						
II (N=26)	64	64	59	32	5	—
IIIa (N=17)	100	100	100	82	71	59
IIIb (N=18)	100	100	100	89	67	67

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## Valproate-induced Hyperammonemic Encephalopathy P 22

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### Background

Valproate-induced hyperammonemic encephalopathy is uncommon with less than 30 cases reported on Medline Search. We review the clinical, EEG and magnetic resonance spectroscopy findings in a patient with this condition.

### Case

A 35-year-old man had left parietal oligodendroglioma with debulking surgery done in 1997. He later underwent a second operation with postoperative radiotherapy in 2001 because of tumour progression. Because of a generalized tonic-clonic convulsion, he was put on valproate 800 mg Q8H intravenously. After two days on this regime he developed impaired consciousness. Urgent CT brain did not show evidence of tumour relapse but the EEG revealed diffuse slow waves at 3-4 Hz. Valproate level was 680 umol/L, phenobarbitone 175 umol/L and ammonia level was 85 umol/L (upper limit 47 umol), rising to 113 umol/L on the next day, along elevated liver enzymes. The valproate dosage was reduced to 300 mg Q8H, after which the patient's improved markedly over two days with normalization of both liver function and EEG. MRI and MR spectroscopy later both showed no evidence of tumour relapse or congenital urea cycle disorders.

### Conclusion

Asymptomatic hyperammonemia is a recognized feature of chronic use of valproate. The mechanism is not completely understood but is probably related to the inhibition effect of ammonia on glutamate metabolism of astrocytes; this leads to intracellular accumulation of glutamate and subsequent osmotic cellular swelling. In cases sufficiently severe to cause encephalopathy, the ammonia level is usually >100 umol as in our patient. However, our patient is particularly susceptible to the development of encephalopathy because of multiple factors underlying including history of brain surgery and concomitant phenobarbitone administration and valproate induced deranged liver function which delayed the clearance of ammonia. But the rapid improvement with reduction of valproate in this case highlights the importance of recognising this complication.

## Case Report: The First Reported Case of Intracranial Vertebral Artery Angioplasty in Hong Kong P 23

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