Assessing Risk for Relapse Among Children with Infantile Spasms Using the Based Score After ACTH treatment: A Retrospective Study

Authors: Lin Wan^{1,2,3,†}, Yan-Qin Lei^{4,†}, Xin-Ting Liu^{1,2,3†}, Jian Chen^{1,2}, Chien-Hung Yeh⁵, Chu-Ting Zhang⁵, Xiao-An Wang⁴, Xiu-Yu Shi^{1,2,6}, Jing Wang^{1,2}, Bo Zhang⁷, Li-Ping Zou^{1,2,3,6}, Guang Yang^{1,2,3,6,*}

Affiliations:

¹Senior Department of Pediatrics, the Seventh Medical Center of PLA General Hospital, Beijing, China
²Department of Pediatrics, the First Medical Center, Chinese PLA General Hospital, Beijing, China
³Medical School of Chinese People's Liberation Army, Beijing, China
⁴Brainup institute of Science and Technology, Chongqing, China
⁵Beijing Institute of Technology, Beijing, China
⁶The Second School of Clinical Medicine, Southern Medical University, Guangzhou, China
⁷Department of Neurology and ICCTR Biostatistics and Research Design Center, Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

[†] These Authors contributed equally to the research article.

Corresponding author:

Dr. Guang Yang, Senior Department of Pediatrics, the Seventh Medical Center of PLA General Hospital, Beijing 100000, China; Email: yangg301@126.com

Supplementary Materials

The supplementary materials include the following figures and tables: Figure S1, ...

Table S1. Cox regression analysis of relapse risk factors in children with infantile spasms (72 cases) who achieved short-term response.

Model 1: Univariate cox regression analysis.

		95% Confidence interval for hazard ratio		
	HR			Р
		Lower	Upper	
Gender (Male/Female)	0.73	0.37	1.46	0.38
Presence of hypsarrhythmia before ACTH	0.54	0.26	1 15	0.11
treatment	0.34	0.20	1.15	0.11
Pathogenic structural abnormalities on MRI	0.85	0.39	1.85	0.68
Definitive etiology	1.5	0.78	2.86	0.22
Number of ASMs	1.47	0.93	2.31	0.098
VPA exposure history	2.18	1.14	4.17	0.018
TPM exposure history	0.84	0.3	2.38	0.75
VGB exposure history	0.46	0.06	3.33	0.44
Hormonal therapy history	1.09	0.43	2.81	0.85
Presence of hypsarrhythmia after ACTH treatment	3.09	1.27	7.53	0.013
Interval from onset to receive ACTH treatment	1.1	1.01	1.2	0.026
Age at spasms onset	0.39	0.99	1	1.04
Frequency of spasms	1	0.99	1	0.39
Dosage of ACTH	0.76	0.35	1.68	0.5
BASED score	1.54	1.27	1.88	<0.001
Model 2: Multivariate cox regression analysis				
Presence of hypsarrhythmia after ACTH	2.09	0.76	5.73	0.2
treatment				
VPA exposure history	1.85	0.87	3.94	0.11
Interval from onset to receive ACTH treatment	0.97	0.87	1.09	0.7
Presence of hypsarrhythmia before ACTH	0.66	0.25	1.71	0.4
treatment				
Number of ASMs	1.41	0.84	2.38	0.2
BASED score	1.5	1.2	1.87	<0.001

Note: Bold P value is statistically significant; data are expressed as number, mean standard deviation, or median (range).

Abbreviations: ACTH, Adrenocorticotropic Hormone; ASM, anti-seizure medication; BASED, Burden of AmplitudeS and Epileptiform Discharges; TPM, topiramate; VGB, vigabatrin; VPA, valproate.

Figure S1. Flow chart of screening of the subjects.



Abbreviations: ASMs, anti-seizure medicines; IS, infantile spasms; ACTH, Adrenocorticotropic hormone

Figure S2. Multivariate Cox regression analysis of relapse risk factors in children with infantile spasms (64 cases) who achieve a short-term response.



Abbreviations: DE, Definitive etiology; NOA, Number of anti-seizure medication; POHAA, Presence of hypsarrhythmia after ACTH; VEH, VPA exposure history=VEH

Figure S3. Multivariate Cox regression analysis of relapse risk factors in children with infantile spasms(72 cases) who achieve a short-term response.



Abbreviations: IFORAT, Interval from onset to receive ACTH treatment; NOA, Number of anti-seizure medication : POHAA, Presence of hypsarrhythmia after ACTH; VEH,VPA exposure history=VEH; POHBA, Presence of hypsarrhythmia before ACTH

Figure S4. Kaplan–Meier (KM) survival curves for the patients with a short-term response after ACTH treatment (n=72).



- **A.** Survival curves for the patents with short-term response after ACTH treatment.
- **B.** b.Survival curves with a cutoff value of BASED score (≥3 or ≤2)for the patents with shorterm-response after ACTH treatment.



Figure S5. Pipeline of the EEG-based functional connectivity states analysis