"Hereditary epilepsy" and whole exome sequencing are more preferable.

doi:10.1016/j.yebeh.2019.08.018

Epilepsy & Behavior 101 (2019) 106744

Characterisation of an infantile rat model of de novo status epilepticus: long-term outcomes

Geatano Terrone[a,b], Rossella Di Sapia[a], Alessia Salamone[a], Ilaria Craparotta[c], Nosaibeh R. Zaniani[a], Daniele Tolomeo[a], Edoardo Micotti[a], Sergio Marchini[c], Teresa Ravizza[a], Annamaria Vezzani[a]
[a]Departments of Neuroscience, Istituto di Ricerche Farmacologiche Mario Negri IRCCS, Milano, Italy
[b]Department of Translational Medicine, Federico II University, Naples, Italy
[c]Departments of Oncology, Istituto di Ricerche Farmacologiche Mario Negri IRCCS, Milano, Italy

Background: Paediatric status epilepticus (SE) may result from acquired, metabolic, immune, genetic or unknown causes. We characterized an infantile rat model of *de novo* SE to study the pathologic sequelae ignited by unremitting seizures in the immature brain that include atrophy, cognitive deficits and epilepsy.

Methods: SE was induced by unilateral intra-amygdala injection of 2 μg kainic acid (KA) in cortical electrode-implanted postnatal day (P)13 male rat pups. Controls were injected with saline. Astrocytes and microglia activation and Fluoro-Jade-positive degenerating neurons were analyzed by immunohistochemistry and confocal microscopy; neuroinflammation and oxidative stress markers were measured by RTqPCR. Different cohorts of SE-exposed P13 rats were longitudinally video-EEG monitored, exposed to the Morris Water Maze to test learning and memory, and to T2-weighted MRI sequence to determine brain atrophy.

Results: EEG monitored convulsive SE was defined by the appearance of continuous spikes with a frequency >1.0 Hz and an amplitude at least 2.5-fold higher than the standard deviation of the baseline tracing. SE occurred 31.0 ± 2.3 min after KA injection and lasted for 3.5 ± 0.5 h (mean ± SEM, n = 9). Epileptiform events of higher amplitude were recorded in the cortex ipsilateral to injected amygdala vs the contralateral homotypic area. During SE pups displayed masticatory movements, salivation, forelimb myoclonus, loss of posture. Glia activation, induction of the ictogenic cytokines IL-1β and TNF-α and HMGB1, oxidative stress markers were measured in rats (n = 6-7 rats each group) from 2 h to 1 week post-SE. Degenerating neurons were detected in cortex, hippocampus, amydgala, striatum and reticular thalamic nucleus. Spontaneous recurrent seizures (3-5/week) developed around 1 month after SE in about 60% of rats as assessed by video-EEG recording for at least 5 months (n = 19). SE was similar in onset, severity and duration in all animals. MRI showed progressive atrophy in cortical and subcortical regions starting before epilepsy onset. Rats displayed cognitive impairment after epilepsy onset denoting an encephalopathic effect of spontaneous seizures.

Conclusions: This infantile SE rat model can be exploited for mechanistic studies, to test novel drugs and for developing biomarkers of disease onset and progression.

Acknowledgments: Ring14 International Onlus; Epitarget (grant agreement n. 602102).

doi:10.1016/j.yebeh.2019.08.019

Epilepsy & Behavior 101 (2019) 106745

Paediatric Status Epilepticus: identification of prognostic factors using the new ILAE classification

Nicola Pietrafusa, Marina Trivisano, Luca De Palma, Marcello Bellusci, Lucia Fusco, Simona Cappelletti, Federico Vigevano, Nicola Specchio
"Bambino Gesù" Children's Hospital, Rome, Italy

Background: Status Epilepticus (SE) is the commonest neurological emergency in childhood. Aim of this study is to report the characteristics of paediatric patients suffering from Status Epilepticus (SE) and their outcome with some considerations to the new classification issued by ILAE.

Methods: We included 173 children treated at "Bambino Gesù" Children's Hospital in Rome (4.35 ± 4.85 years old; follow up 2.74 ± 1.9 years). Multivariate model was constructed to predict neurocognitive outcome, recurrence of SE, development of epilepsy and mortality. Adjusted ORs were calculated with 95% Confidence interval (OR[95%CI]).

Results: We observed a different prevalence of aetiologies for the different semiologies ($p < 0.05$) and for each age-group ($p < 0.05$), overlapping only in part with the recent ILAE classification. After SE, patients developed: 70% epilepsy (drug-resistant in half of them); 20% worsening of neurological exam; 16% cognitive deficit; 16% recurrent SE. At multivariate analysis: SE lasting more than 24 hours have increased risk to develop cognitive (OR = 6.00[2.0-17.1]) or neurologic sequelae (OR = 8.58[2.7-27.1]); the same finding was observed for patient younger than 1 months (cognitive OR = 4.84[1.13-17.3] and neurologic sequelae OR 6.7[1.17-27.1]). The recurrence of SE was associated with genetic (OR = 8.87[2.46-42.63]) and cryptogenic aetiology (OR = 11.5 [2.2-61.8]), as like myoclonic semiology (OR = 6.1[1.1-29.4]). Febrile SE (OR = 0.06[0.008-0.40]) and acute symptomatic aetiology (OR = 0.12 [0.04-0.40]) have a diminished risk to develop epilepsy. Drug-resistant epilepsy post SE was less frequent in focal non-convulsive SE (OR = 0.18 [0.32-0.97]) and acute symptomatic SE (OR = 0.04[0.007-0.26]).

Conclusion: Age at onset and duration of SE are critical independent variables associated to worst neurocognitive outcome. The risk to develop epilepsy is lower after acute symptomatic and febrile SE. Semiology and age of onset are useful to predict aetiology of SE. For this reason, ILAE classification respect the 4 axes seems to be a good step forward.

doi:10.1016/j.yebeh.2019.08.020

Epilepsy & Behavior 101 (2019) 106746

Some epidemiological aspects of status epilepticus in the female epilepsy

Svetlana Kravtsova, Michael Aleksandrov, Galina Odintsova, Alexey Ulitin
Russian A.L. Polenov Neurosurgical Institute - A Branch of The Almazov National Medical Research Centre, St. Petersburg, Russian Federation

Background: Status epilepticus (SE) is a formidable manifestation of epilepsy. The study of the clinical features of polymorphism of