Parkinsons Congress 2019: Epilepsy and neurocutaneous syndromes in developmental age- Michele Roccella- University of Palermo, Italy

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Abstract

The Neurocutaneous Syndromes (N.S.) are a rarther heterogeneous group of diseases from both a clinical and genetic point of view. Most N.S. reveal themselves through convulsive crises, wich sometimes do not respond to the pharmacological treatment. The purpose of this study is to evaluate the therapeutic and clinical aspects of and adequate therapeutic procedure. 78 children have been studied, they areaffectedby: 33childrenfromneurofibromatosis; 23 children from tuberous sclerosis; 9 children from the Sturge-weber syndrome; 5 from Ito hypomelanosis; 4 from incontinentia pigmenti: | from Dubowitz syndrome; | from the Schimmelpenning-Feuerstin-mims syndrome;! from kippel- Trenaunay-weber syndrome, 1 from ataxia-telangiectasia. The anamnesis, the hystory of the cris and of the antiepileptic therapy was recorder for each case. The initial critical symptomatology was divided into groups: infantile spasms; simple focal seizures; focal focal epilepsy; generalized crises. The age of onset at the beginning of the critical symptomatology is between 15 days and 5 years of age. The neuroradiological pictures observed with brain MRI are fairly heterogeneous. The evolution and the current state of epileptic symptoms has been evaluated: 40% of cases have generalized epilepsy: 60% of cases a focal epilepsy. The most used drugs are: VPA, CBZ, PB, LTG, BDZ. In some cases cycles of cortisone therapy (cases with infantile spasms, S. di Lennox-Gastaut) were also practiced. Conclusions: The neurocutaneous syndromes constitute a group of quite heterogeneous pathologies. The use of tools such as TAC and, above all, MRI are essential for a correct diagnostic classification. EEG changes are clearly correlated with the type and extent of the malformation pattern; it seems difficult to establish a specific and pathognomonic picture of a form of S.N. It remains however difficult to establish, in these pathologies, what is the evolution, the therapeutic strategy to be implemented and the prognosis of the epileptic symptomatology based on its time of onset and the associated neuroradiological framework. It is therefore important to underline how the use of new pharmacological therapies leads to a considerable reduction of critical episodes and consequentially to an improvement in the life of these subjects. Keywords: Epilepsy, Epilepsy and neurocutaneous syndromes, Antiepileptic drugs Biography Michele Roccella is Associate Professor of Child Psychiatry at the Department of Psychology, Educational Science and Human Movement, University of Palermo, Italy.