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- B. : e 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. e purpose of this study is to evaluate the therapeutic and clinical aspects of and adequate therapeutic procedure.
- M : :78 children have been studied, they are a ected by: 33 children from neuro bromatosis; 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. e anamnesis, the hystory of the cris and of the anti-epileptic therapy was recorder for each case.
- e initial critical symptomatology was divided into groups: infantile spasms; simple focal seizures; focal focal epilepsy; generalized crises. e age of onset at the beginning of the critical symptomatology is between 15 days and 5 years of age. e neuroradiological pictures observed with brain MRI are fairly heterogeneous. e