COGNITIVE DISORDER IN CHILDREN WITH EPILEPSY

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Evaluation of different factors influence on cognitive function (CF) in children with epilepsy. 20 children with epilepsy: 13 girls (75%) and 5 boys (25%), from 6 to 17 years old (mean age 11 years 9 months). The investigation included: standard neurology status, neuroimaging by CT or MRI, EEG, cognitive function examination by Epi Track Junior it was test. Anamnesis of Life and disease (duration and debut age of epilepsy, type and frequency of seizures, antiseizures therapy), delivery anamnesis and early motor and speech development.

There were several groups in Epi Track Junior results testing: good results 0%, mild disorder of CF – 25% (5 children), average disorder of CF – 20% (4 children), severe disorder of CF – 55% (11 children).

In light asphyxia (Apgar less than 7 degree) delivered 5 children (25%), in the rest cases (75%) Apgar was normal. Early development was normal in 19 children (95%) and 1 child (5%) with average speech retardation.

Unknown epilepsy etiology has 12 patients (60%), genetic – 3 (15%) and structural – 5 (25%). Focal seizures without consciousness impairment were in 2 cases (10%), generalized in 14 (70%) and focal with secondary generalization in 4 (20%). In remission were 3 children (15%), with rare seizures were 9 (45%), 1 (5%) were with frequent seizures and 7 children were (35%) with very frequent seizures.

All patients (3 (15%) in polytherapy had severe CF disorder. Children with frequent seizures (8 patients (40%)) had mild CF disorder in 2 cases (10%), average CF disorder in 2 cases (10%) and severe CF disorder in 4 cases (20%).

Debut of seizures up to 7 years was in 9 children (45%) and 7 (35%) had severe CF disorder.

Duration of epilepsy from 1 to 3 years was in 4 children (20%) and 2 children (10%) of them had severe CF disorder. Duration epilepsy from 3 to 7 years were in 7 children (35%) and all of them had severe CF disorder.

Severe and average CF disorder were showed in children with frequent seizures, early (up to 7 years) debut of seizures, duration of disease more than 3 years and polytherapy cases.

There was no influence on CF: Apgar score, early development, neurology deficit, structure changes in neuroimaging, type of epilepsy and seizures, EEG changes.

Aromatic L-amino acid decarboxylase (AADC) deficiency is a rare autosomal recessive disorder resulting in marked or complete loss of dopamine, impeding normal motor development. Eladocagene exparvovec, a recombinant adeno-associated virus containing the human cDNA encoding the AADC enzyme, is in clinical development for treatment of AADC deficiency.

Eladocagene exparvovec was administered via bilateral infusion into the putamen of 28 children with AADC deficiency in 3 clinical trials (AADC-CU/1601 [8 patients, completed], AADC-010 [10 patients, ongoing], and AADC-011 [10 patients data cutoff of 26 February 2020, ongoing]). Patients received a total dose of 1.8 × 1011 vg (n=21) or 2.4 × 1011 vg (n=7; AADC-011). Motor milestone achievement was assessed using the Peabody Developmental Motor Scales, 2nd Edition. Improvements in motor function from treated patients in the full analysis set were compared to 49 subjects from a comprehensive Natural History Database (NHDB). This database was derived from a systematic review of literature reporting data from patients with AADC deficiency. From an original pool of 237 patients, 49 were chosen as matched natural history controls. These natural history controls were chosen because they met the criteria of unique patients with confirmed AADC deficiency who had not participated in clinical trials of eladocagene exparvovec, had documented lack of motor milestone achievement, and had a similar disease phenotype to patients in clinical trials.

As early as 12 months after receiving eladocagene exparvovec, 44% of patients achieved head control and 20% of...