EEG revealed generalised encephalopathy and epileptic seizure predisposition on the right hemisphere, consistent with the clinical and radiological findings. Lumbar puncture was not immediately performed in view of the intracranial bleed and anti-coagulant treatment. Due to her poor clinical response, steroid treatment was started for suspicion of an autoimmune or inflammatory condition. A CT head done after 48 hours didn’t show progression of the haemorrhage. We proceeded with lumbar puncture after withholding a dose of LMWH, screening for infection, metabolic and autoimmune panels. HSV-1 was detected in the CSF, despite no cutaneous or serological evidence of HSV infection. Concurrent cell counts showed pleocytosis confirming the clinical suspicion of encephalitis. She remained on aciclovir for 21 days as a definitive treatment.

**Discussion** Ischemic stroke and haemorrhage are increasingly recognised CNS manifestations of HSV infection. The brain ischemia is mostly related to multifocal cerebral large vessel vasculitis. HSV-related infarction is a rare but potentially treatable cause of stroke. Steroid treatment may be considered even in the absence of confirmation of vasculitis on neuroimaging. A systematic review showed up to 50% of cases with HSV ischemic manifestations presented with encephalitis while 30% presented with stroke-like symptoms.

Performing lumbar puncture is crucial to differentiate encephalitis from other forms of encephalopathy. Ideally it should be undertaken prior to initiation of aciclovir treatment, however this is usually delayed by other investigations, due to the nature of presentation. Therefore, high index of suspicion is needed to start aciclovir empirically as early treatment could reduce mortality and morbidity significantly.

**Conclusion** Cerebrovascular events in children should be recognised as a possible manifestation of HSV encephalitis. Aciclovir should be commenced early and CSF sampling should be undertaken at the earliest opportunity once safe to perform.

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**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%), generalised 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.

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**EFFICACY OF GENE THERAPY WITH ELADOCAGENE EXUPARVOVEC IN PATIENTS WITH AADC DEFICIENCY COMPARED WITH NATURAL HISTORY CONTROLS**


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 at data cutoff of 26 February 2020, ongoing]).

Patients received a total dose of $1.8 \times 10^{11}$ vg (n=21) or $2.4 \times 10^{11}$ 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.

The 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