

aEEG in Metabolic Disease

History

G1P1, both parents originally from Chechnya, no consanguinity, fetal MRI performed because of oligohydramnios showed asymmetry of lateral ventricles and confirmed oligohydramnios, otherwise uneventful pregnancy.

Caesarian section at 42+0 weeks gestation, rupture of membranes 36 h antenatally, mother on Penicillin, but without any laboratory signs of infection.

Initial assessment

Uneventful birth of a baby girl, Apgar 9/10/10 at 1/5/10 minutes of life, transferred to the nursery.

First pediatric ward round and examination showed normal results of a healthy baby.

Clinical course

The patient began having periods of cyanosis, mainly periorally, and demonstrated an extensive opisthotonus and jitteriness.

Blood gas analysis revealed normal glucose levels, raised lactate levels (7 mmol/L) and a metabolic acidosis (base deficit -6, Bicarbonate 18 mmol/L).

The infant's condition worsened and she developed apnoeic episodes requiring bag and mask ventilation, hence was transferred to the NICU.

NICU admission

The infant continued to show some extensive apnoeas. aEEG monitoring was commenced, and the aEEG showed epileptic discharges (Figure 1).

The infant was intubated and lumbar puncture performed. BGA demonstrated constantly elevated lactate levels.

Seizures were drug-resistant and ongoing despite administration of phenobarbitone, levetiracetam, midazolam and topiramate.

Investigations/results

- Lumbar puncture: microbiological and virological results negative
- Cranial MRI: general atrophy including cerebellum and brainstem, corpus callosum thinning, enlarged lateral ventricles, bilateral cystic periventricular lesions, myelination in pons only, hyperintense signal in basal ganglia (Figure 2 & 3)
- aEEG: drug-resistant saw-tooth pattern on a continuous/discontinuous background pattern showing some rudimentary sleep-wake cycling (Figure 4)
- Metabolic results: amino acids (plasma) and organic acids (urine) normal

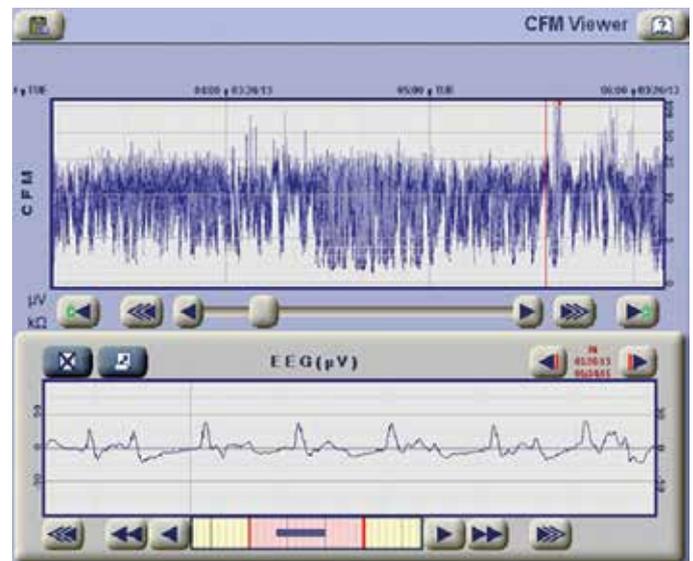


Figure 1

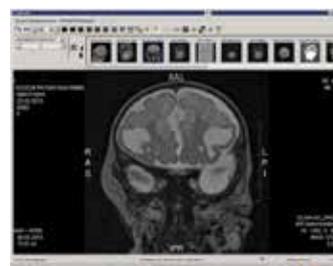


Figure 2

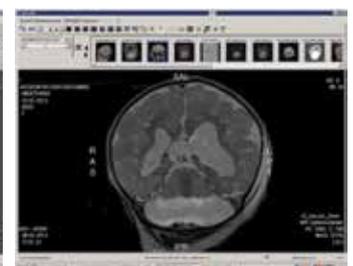


Figure 3

Day of life 13

The infant's clinical condition worsened due to urosepsis and increasing lactate acidosis, requiring triple antibiotics and inotropic support with arterenol and cortisone.

Continuous buffering and increase of respiratory support was also needed, and the aEEG demised to burst suppression and eventually to an Isoelectric flat trace (Figure 5).

The baby girl died on day of life 14 despite all efforts of neonatal intensive care in the presence of both parents.

Discussion

Even though the outcome of the infant was sadly unsuccessful, we believe that the aEEG was extremely helpful in terms of guiding our management. Seeing the atypical trace, we hypothesized a metabolic disorder being the cause of the infant's clinical state in a very early course of the disease, and could therefore prepare and counsel the parents for a very likely bad outcome at a very early stage.

As a neonatal community, more aEEG traces need to be collected in order to establish clear pictures for designated metabolic diseases.

Dr Monika Resch

Associate Professor of Neonatology
Division of Pediatric Intensive Care, Neonatology & Neurology
Medical University, Vienna, Austria

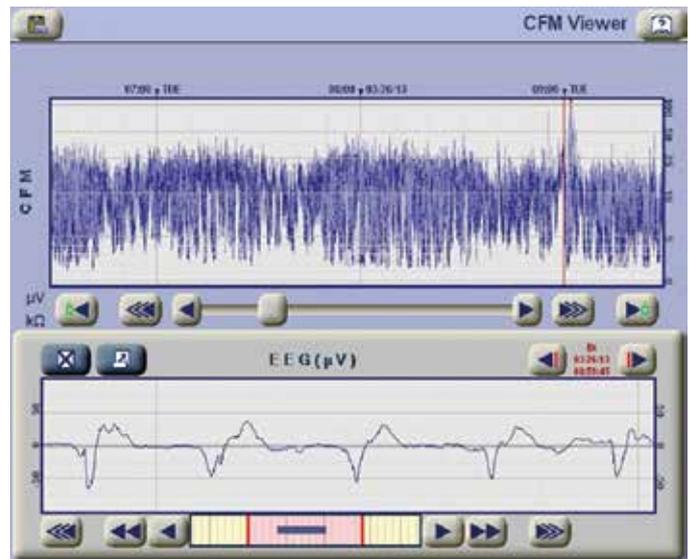


Figure 4

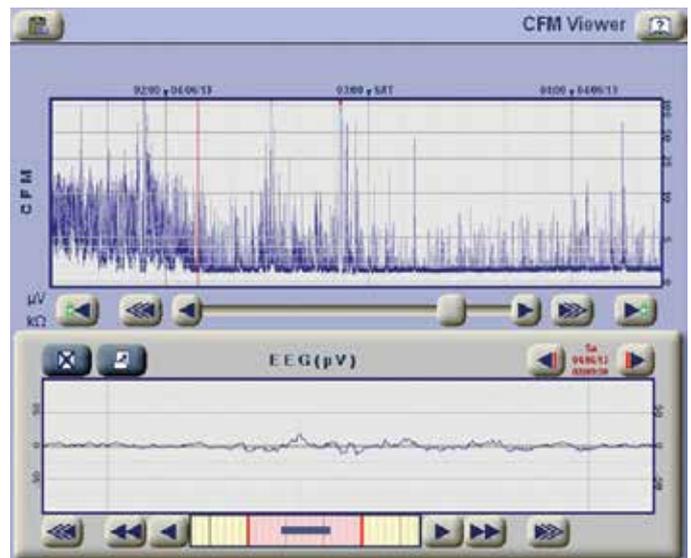


Figure 5

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