**Tables**

Table 1. Clinical diagnosis and known aetiologies of the 29 patients.

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| * **Lennox-Gastaut syndrome (9)**

Postinfectious (3) del 22q11.21 (1)\* Focal cortical dysplasia type II (1) Multifocal cortical dysplasia (1) *SYNGAP1* mutation (1) *No defined aetiology* (2)* **West syndrome (7)**

 Perinatal stroke (2)MCD (2)\*PEHO syndrome (1)\* *MAGI 2* mutation (1)\* Tetrasomy 15q (1)* **Epilepsy with myoclonic-atonic seizures (4)**
* **Ohtahara syndrome (2)**

Classic lissencephaly due to mutation (1) *No defined aetiology* (1)* **Myoclonic epilepsy in infancy** **(2)**
* **Landau-Kleffner syndrome (1)**
* **Dravet syndrome (1)**
* **Focal epilepsy (1)**

Tuberous sclerosiscomplex (1)* **Generalized absence epilepsy (1)**
* **FIRES (1)\***
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\*del – deletion; FIRES – Febrile Infection-Related Epilepsy Syndrome; MAGI 2 - Membrane-Associated Guanylate kinase Inverted 2; MCD - Malformations ofCortical Development; PEHO - Progressive encephalopathy with Edema, Hypsarrhythmia and Optic atrophy**.**

Table 2. Efficacy by electroclinical diagnosis (ILAE classification), in the 29 patients on KD for more than three months.

|  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Outcomes of Efficacy | Lennox-Gastaut syndrome (n=9) | West syndrome (n=7) | Epilepsy with myoclonic-atonic seizures (n=4) | Ohtahara syndrome (n=2) | Myoclonic epilepsy in infancy (n=2) | Landau-Kleffner syndrome (n=1) | Dravet syndrome (n=1) | Tuberous sclerosis complex (n=1) | Generalized absence epilepsy (n=1) | FIRES\* (n=1) |
| Seizure control | 100% seizure reduction rate | 3 | 0 | 1 | 0 | 0 | 0 | 0 | 0 | 0 | 0 |
| >90% seizure reduction rate | 1 | 1 | 1 | 0 | 0 | 1 | 0 | 0 | 0 | 0 |
| 50-90% seizure reduction rate | 3 | 2 | 2 | 1 | 0 | 0 | 1 | 0 | 0 | 1 |
| <50% seizure reduction rate | 1 | 2 | 0 | 0 | 1 | 0 | 0 | 1 | 0 | 0 |
| 0% seizure reduction rate | 1 | 2 | 0 | 1 | 1 | 0 | 0 | 0 | 1 | 0 |
| Improvement in awareness | Marked | 7 | 5 | 3 | 1 | 0 | 1 | 1 | 1 | 0 | 0 |
| Moderate | 1 | 1 | 1 | 0 | 2 | 0 | 0 | 0 | 1 | 1 |
| None | 1 | 1 | 0 | 1 | 0 | 0 | 0 | 0 | 0 | 0 |
| Improvement in behaviour | Marked | 6 | 5 | 4 | 1 | 0 | 1 | 1 | 0 | 0 | 0 |
| Moderate | 2 | 0 | 0 | 0 | 2 | 0 | 0 | 1 | 1 | 1 |
| None | 1 | 2 | 0 | 1 | 0 | 0 | 0 | 0 | 0 | 0 |

\*FIRES – Febrile Infection-Related Epilepsy Syndrome

Table 3. Efficacy by pathogenic mutations and chromosome abnormalities identified in seven of the 29 patients.

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| Outcomes of Efficacy | SYNGAP1 gene(n=1) | SCN9A gene\*\* (n=1) | TSC1 gene\*\* (n=1) | MAGI2 gene\*\* (n=1) | LIS gene\*\* (n=1) | Tetrasomy 15q (n=1) | del 22q11.21 (n=1) |
| Seizure control | 100% seizure reduction rate |  |  |  |  |  |  | X |
| >90% seizure reduction rate | X |  |  |  |  |  |  |
| 50-90% seizure reduction rate |  | X |  |  | X | X |  |
| <50% seizure reduction rate |  |  | X | X |  |  |  |
| 0% seizure reduction rate |  |  |  |  |  |  |  |
| Improvement in awareness | Marked | X |  | X |  | X | X | X |
| Moderate |  | X |  | X |  |  |  |
| None |  |  |  |  |  |  |  |
| Improvement in behaviour | Marked | X |  |  |  | X | X | X |
| Moderate |  | X | X |  |  |  |  |
| None |  |  |  | X |  |  |  |

\*\*LIS - Lissencephaly; MAGI2 - Membrane-Associated Guanylate kinase Inverted 2; SCN9A - Sodium Voltage-Gated Channel Alpha Subunit 9 (FIRES); TSC1 - Tuberous Sclerosis 1.

Table 4. Acute and chronic secondary effects of the KD (n=29).

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| --- |
| **Acute secondary effects (n=26)**  |
| **Hypoglycemia** 6 **Vomits/ Nausea** 5 **Lethargy** 4**Acidosis** 3**Food refusal** 2 |
|
|
| **Chronic secondary effects (n=29)** |
| **Hypercholesterolemia** 23 **Hypertrigleceridemia** 21**Hypercalciuria** 14 **Hyperurecimia** 12 **Hyperphosphatemia** 10**Constipation** 9 - **de novo** 1 **Urine crystals** 9 **Hypercalcemia** 7**Carnitine deficit** 6**Hypomagnesemia** 5 **Zinc deficit** 3 **Vitamin D deficit** 2 **Calcium deficit** 2 **Hypophosphatemia** 2 **Anemia** 1  |