**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)\*** |

\*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).

|  |
| --- |
| **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 |