Table 1- The Mean Age of Infants in Total and According to Gender

|  |  |  |  |
| --- | --- | --- | --- |
| Gender groups | Males | Females | Total |
| Mean age(day) | 4.21 ± 1.82 | 4.45 ± 1.90 | 4.33 ± 1.45 |

Table 2- Demographic Data for Maple Syrup Urine Disease Patients

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
|  | Mean age/ day | Mean birth weight / gram | Positive family history of MSUD | Positive familial marriage  | Gestational age/ weeks |
| Males(N=3) | 7.3 ± 2.1 | 3230 ± 404 | 0(0%) | 2(66.6%) | 39.2 ± 2.1 |
| Females(N=8) | 5.6 ± 2.3 | 3050 ± 379 | 3(37.5%) | 7(87.5%) | 37.3 ± 1.8 |
| Total(N=11) | 6 ± 2.8 | 3100 ± 375 | 3(27.2%) | 9(81.8%) | 38.3 ± 2.1 |
| P value | 0.137 | 0.277 | 0.21 | 0.42 | 0.365 |

Table 3 - Distribution of the Chief Complaints of Patients

|  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
|  | Poor feeding | Fever | Lethargy | Hypo-hypertonia | Cyanosis | Jaundice | Loud cry | Seizures | S.B.U |
| N.P | 8 | 1 | 4 | 3 | 2 | 1 | 2 | 1 | 4 |

**N.P= Numbers of patients S.B.U= Smell of burnt sugar in the urine**

Table 4- MSUD Data

**Male Female Total P Value**

**Life status (live/ dead)** 1/ 2 1/ 7 2/ 9 0.42

**MSUD subtypes**

Classic 2 6 8

Intermediate 0 1 1 0.33

Intermittent 0 1 1

Thiamine – Responsive 1 0 1

 **Diagnosis**

Random 1 0 1 0.08

After symptoms 2 8 10

**Neurological involvement** 2 4 6 0.62

**Treatment**

 MSUD special diets 1 8 9

 Peritoneal dialysis 1 0 1 0.08

 Blood transfusion 0 1 1

 Liver transplantation 0 1 1

**Mean age at diagnosis/ days** 20.2 ± 2.41 25.1 ± 2.30 23.3 ± 2.82 0.18

Table 5 - The Mean Serum Levels of Branched Amino Acids

|  |  |  |  |
| --- | --- | --- | --- |
|  | Leucine | Isoleucine | Valine |
| Males(N=3) | 3107 ± 937µg | 583 ± 549µg | 578 ± 663µg |
| Females(N=8) | 2983 ± 695µg | 373 ± 205µg | 473 ± 244µg |
| Total(N=11) | 2984 ± 721µg | 478 ± 315µg | 525 ± 364µg |
| P Value | 0.30 | 0.06 | 0.01 |