**Table I. Clinical features of ZSD by age of onset**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| Features | Neonate | 1- 6 mos | 6 mos- 4 yrs | > 4 yrs |
| Hypotonia and poor feeding | ZS, NALD | NALD, IRD | NALD, IRD | IRD |
| Craniofacial dysmorphism | ZS, NALD | NALD, IRD |  |  |
| Seizures | ZS, NALD | NALD, IRD | NALD, IRD | IRD |
| Neuronal migration disorders | ZS |  |  |  |
| Chondrodysplasia punctata | ZS (limited to knees and hips) |  |  |  |
| Direct hyperbilirubinemia | ZS, NALD, IRD | NALD, IRD |  |  |
| Liver dysfunction | ZS | NALD, IRD |  |  |
| Hepatomegaly | ZS | NALD, IRD |  |  |
| Calcium oxalate renal stones |  |  | IRD | IRD |
| Cataracts | ZS | NALD, IRD | NALD, IRD | IRD, atypical |
| Retinal abnormalities | ZS | NALD, IRD | NALD, IRD | IRD, atypical |
| Sensorineural hearing loss | ZS | NALD, IRD | NALD, IRD | IRD |
| Failure to thrive | ZS | NALD, IRD | NALD, IRD |  |
| Psychomotor retardation |  | ZS, NALD, IRD | ZS, NALD, IRD | IRD |
| Leukodystrophy |  | NALD | NALD | IRD, atypical |
| Peripheral neuropathy |  |  | IRD, atypical | IRD, atypical |
| Cerebellar ataxia |  |  | IRD, atypical | IRD, atypical |