### Table 1. Patient Characteristics

|                  | P1           | P2         | P3         | P4           | P5            | P6            |
|------------------|--------------|------------|------------|--------------|---------------|---------------|
| **Origin**       | Turkish      | United Kingdom | French       | French (West Indies) | Turkish | Turkish |
| **Current age**  | 6 y          | 2 y        | 22 y       | 10.5 y       | died 93rd day | 3 m          |
| **Onset age**    | 3 weeks      | 1 week     | 1 week     | 4 days       | 1 day         | 5 days       |
| **Severe diarrhea** | yes, with hypovolemic shock | yes | yes, intermittent until 10 y | yes | yes | yes |
| **Duodenal villous atrophy** | yes | yes | yes | yes | ND | ND |
| **Sub apical PAS accumulation** | yes | yes | yes | yes | ND | ND |
| **Sub apical CD10 accumulation** | yes | yes | ND | yes | ND | ND |
| **Electron microscopy** | Vesicles or tubulovesicular structures MVI Basolateral microvilli large lysosomes/autophagosomes Defective microvilli anchored deep in the cytoplasm thickened terminal web region | Vesicles or tubulovesicular structures | Defective microvilli Basolateral microvilli large lysosomes/autophagosomes | ND | ND | ND |
| **Treatment**    | TPN          | TPN        | Exclusive enteral nutrition | TPN | TPN | TPN |
| **Cholestasis**  | yes          | no         | yes        | no           | yes           | yes           |
| **Bone fragility** | Fracture of the left inferior part of the femur at 7th months and at 2 y | Fracture of the right wrist at 1 y | Fracture of the scapula at 7 y Other fracture at 10 y | no | ND | One bone fracture upon delivery |
| **Deafness**     | no           | no         | yes        | no           | no            | no            | ND |
| Other manifestations | no | no | blind: right anophthalmia, Peters anomaly & left pyramidal cataract / severe ID & behavioral disorder | ID | ND | ND |

ND: not determined; ID: intellectual disability; TPN: total parenteral nutrition; MVI, microvillus inclusion.