

**ACUTE ILLNESS PROTOCOL  
ORGANIC ACIDEMIAS  
MAPLE SYRUP URINE DISEASE (MSUD)**

**PRESENTATION**

Well at birth but, symptoms may develop quite rapidly within 4 -7 days. Later in childhood similar symptoms develop during acute illness.

- poor suck, disinterest in feeding
- lethargy
- weight loss
- neurological deterioration with alternating hypertonia and hypotonia
- dystonic “decerebrate-like” extension of the arms
- seizures
- coma
- pseudotumor cerebri and/or bulging fontanelles occasionally seen
- maple syrup/burnt sugar like odor (from urine, bodily secretions & ear wax)

Presentation varies depending on the severity of the metabolic defect. The neonatal form presents in the first week of life with a life-threatening illness. Survivors of severe neonatal episodes are often neurologically devastated, when treatment was delayed. The infantile or late-onset form has a more insidious presentation with failure to thrive, developmental delay, and perhaps other neurologic features. Intermittent forms of MSD are also described. All forms can have acute severe life-threatening decompensatory episodes.

**Parents of children with diagnosed metabolic disorders know the early signs of decompensation in THEIR children. Listen to them !!!**