

**ACUTE ILLNESS PROTOCOL  
FATTY ACID OXIDATION DISORDERS  
VERY LONG CHAIN ACYL CoA DEHYDROGENASE (VLCADD) DEFICIENCY**

**CLINICAL PRESENTATION**

- Nausea or vomiting
- Lethargy
- Hypoglycemia with lack or only 'trace' of urinary ketones
- Seizures
- Hepatomegaly
- 'Reye' like syndrome
- cardiomyopathy, arrhythmias
- coma
- near/rescued SIDS

First presentation can occur in the neonatal period but more often when the infant is being weaned from night time feeds. The usual picture is nausea, vomiting and/or lethargy after a period of fasting. This can progress to hypoglycemic seizures or coma within 1-2 hours of ONSET of symptoms. There may, or may not, be a history of a recent viral infection associated with diminished oral intake, or of a similar episode in the past. FAODs are responsible for a small but significant proportion of sudden infant death syndrome, which may be preventable with prompt early recognition and treatment.

NOTE that in the acute crises patients can be seriously ill WITHOUT hypoglycemia although typically FAOD crises are associated with hypoglycemia. At these times the urine typically tests 'absent' or 'small' for the presence of ketones. Liver function tests may be mildly elevated; hyperammonemia and hyperuricemia are often present during acute episodes.

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