

**ACUTE ILLNESS PROTOCOL
FATTY ACID OXIDATION DISORDERS
SHORT CHAIN Acyl-CoA DEHYDROGENASE (SCAD) DEFICIENCY**

PRESENTATION

- Asymptomatic
- Vomiting
- metabolic acidosis but USUALLY HAVE ketosis (unlike other FAODs)
- failure to thrive
- developmental delay
- hypotonia,
- chronic skeletal myopathy this is seen in some older patients.
- seizures
- encephalopathy
- 'Reye like' syndrome *of liver failure, hyperlacticacidemia and coma*
- Sudden death

When ill, patients with SCADD are at risk for developing metabolic acidosis and hypoglycemia. Carnitine levels are typically low. Hyperammonemia has been described. Whereas most other FAODs may be associated with hypoketotic hypoglycemia, metabolic crises due to SCADD are associated with significant ketosis.

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