

**PREGNANCY MANAGEMENT OF WOMEN WITH  
SUSPECTED OR KNOWN  
FATTY ACID OXIDATION DISORDERS**

**BACKGROUND INFORMATION**

Fatty Acid Oxidation Disorders (FAODs) are genetic metabolic deficiencies in which the body is unable to oxidize fatty acids to make energy due to an enzyme deficiency. Normally when glucose runs out, fat is broken down into energy. However, that energy is not readily available to people with an FAOD.

**RISK MANAGEMENT**

-patients with FAODs are at risk of the following:

- HELLP syndrome (hemolytic anemia, elevated liver enzymes, low platelets). HELLP syndrome can cause maternal and fetal death
- Rhabdomyolysis
- Acute cardiomyopathies
- Hypoglycemia (low but not zero)

**INTERVENTIONS**

**Carnitine supplementation**

-500 mg L-carnitine QID throughout pregnancy except in patients with LCHAD

-carnitine supplementation is contraindicated in LCHAD deficiency as it leads to an increase in hydroxy long chains, leading to cardiac arrhythmias and cardiomyopathy.

If free carnitine is low, a small dose can be given, but avoid routine supplementation

-carnitine may be contraindicated in VLCAD deficiency. If the patient has VLCAD deficiency, please consult with the metabolic physician before prescribing carnitine

**Monitoring**

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- monitor serum and blood spot acyl carnitine profiles on a monthly basis
- monitor for essential fatty acid deficiency
- closely monitor fetal growth and weight gain; patient needs to be referred to maternal/fetal medicine

### Dietary Management

- Low fat, high carbohydrate
- avoidance of fasting.
- consume regular meals and snacks with a low fat, high carbohydrate snack before bed.

### LABOUR AND DELIVERY CONSIDERATIONS

- if a C-section is elective, order echocardiogram and ECG to be done prior to surgery
- if patient is fasting, run an IV with Dextrose (see guidelines below)
- for C-sections, run a high carbohydrate infusion when patient starts fasting, during C-section and recovery period (see guidelines below)

### ANESTHETIC CONSIDERATIONS

Consideration	Rationale
Avoid propofol	Propofol, a high fat anesthetic agent, can cause decompensation in FAOD, possibly leading to Propofol Infusion Syndrome
Maintain an adequate carbohydrate supply of 4 mg/kg/minute of D10W <ul style="list-style-type: none"><li>• Glucose infusion not to exceed 100 mL/hr</li><li>• Monitor blood sugars</li></ul>	Provides adequate glucose so that fat is not used as an energy source and fatty acid oxidation is suppressed

Weight (kg)	Rate (mL/hour)
<50	50
50-75	75
>75	100

For more information, please contact the Adult Metabolic Diseases Clinic, 604-875-5965.