Twin Pregnancy Normalizes Triglyceride Level in Familial Chylomicronemia Syndrome
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Objectives:
- Role of lipoprotein lipase in triglycerides processing
- Discuss severe complications of elevated triglycerides in pregnant person with FCS
- Role of heterozygous twin in normalizing triglycerides

Introduction:
Familial chylomicronemia syndrome (FCS) is a rare autosomal recessive disorder characterized by abnormal high level of triglycerides (>11.3 mmol/L) and massive accumulation of chylomicrons in the blood stream leading to potentially life-threatening complication of acute pancreatitis requiring hospitalizations.1,5

This condition is caused by gene mutation of the enzyme called lipoprotein lipase (or Apoprotein C-II). Lipoprotein Lipase removes triglycerides (TG) from triglyceride-rich lipoproteins (TRL) and breaks them down into free fatty acids for energy use. It is estimated to occur 1 in 1-2 million people, can be diagnosed at any age and affect gender, race and ethnicity equally.2

During pregnancy estrogen increases liver synthesis of triglycerides to support fetal processing. It is estimated to occur 1 in 1-2 million people, can be diagnosed at any age and affect gender, race and ethnicity equally.2

We present a very rare case of pregnant patient affected by FCS and how twin pregnancy normalizes triglyceride level and prevents life threatening complication of acute pancreatitis.1,3,6

Case Report
A 37-year-old female gravida 2, para 2 diagnosed with FCS at the age of 3 and managed with a strict low-fat diet. She started developing recurrent pancreatitis in her early 20s, requiring multiple hospitalizations per year. She was started on TG lowering medications and lifestyle management but with limited response. During one of her hospitalizations for pancreatitis, she was found to be 20 weeks pregnant with a TG level of 3500 mg/dL. Due to significant concerns for potentially life-threatening pancreatitis and central nervous system complications, she was started on weekly total plasma exchange (TPE) with average TG levels of 1340mg/dL and peaked to 6000mg/dL. She had no subsequent hospitalizations for recurrent pancreatitis and delivered a healthy baby boy. Her TG levels remained <1000 mg/dL after discontinuing TPE.

At her one-year follow-up, she was found to be 12 weeks pregnant with fraternal twins and weekly plasma exchanges were re-initiated but surprisingly the patient’s TG levels normalized to 150 mg/dL and the patient did not require any further plasma exchange, nor did she have any further hospitalizations for pancreatitis during the remainder of her pregnancy. She delivered one boy and one girl, both were heterozygous for LPL p.G236Gfs*15 deletion.

We hypothesize that the twin gestations functioned with a completely normal LPL enzyme processing the mother’s elevated TG and hence prevents complications and hospitalization.

Discussion:
1. LPL removes triglycerides from triglycerides rich lipoprotein hence prevents chylomicron accumulation in the blood stream.
2. In FCS, there is missense/deletion mutation in gene coding for LPL resulting in non-functional LPL hence chylomicron accumulates causing complications of acute pancreatitis.
3. Pregnant person with FCS, their triglycerides are markedly elevated compared to only pregnant person resulting in recurrent complication of acute pancreatitis and hospitalization.
4. Heterozygous twins function as complete LPL that process mother’s elevated TG and hence prevents complications and hospitalization.

References: