ACP Oral Presentation: Congenital Lipodystrophy: Can Fat Harm the Lungs?

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Background Info

- Lipodystrophy refers to abnormal lipid distribution in the body
- Congenital versus acquired
- Clinical diagnosis based on:

  -PE/Body dysmorphia

  -Lipid profiles

  -Associated Comorbidities

  -Genetic testing
- Here we present a case of a patient with suspected congenital lipodystrophy who was admitted for symptomatic hypertriglyceridemia without pancreatitis.
- She subsequently developed ARDS after treatment with insulin drip for rapid reduction of serum triglyceride levels

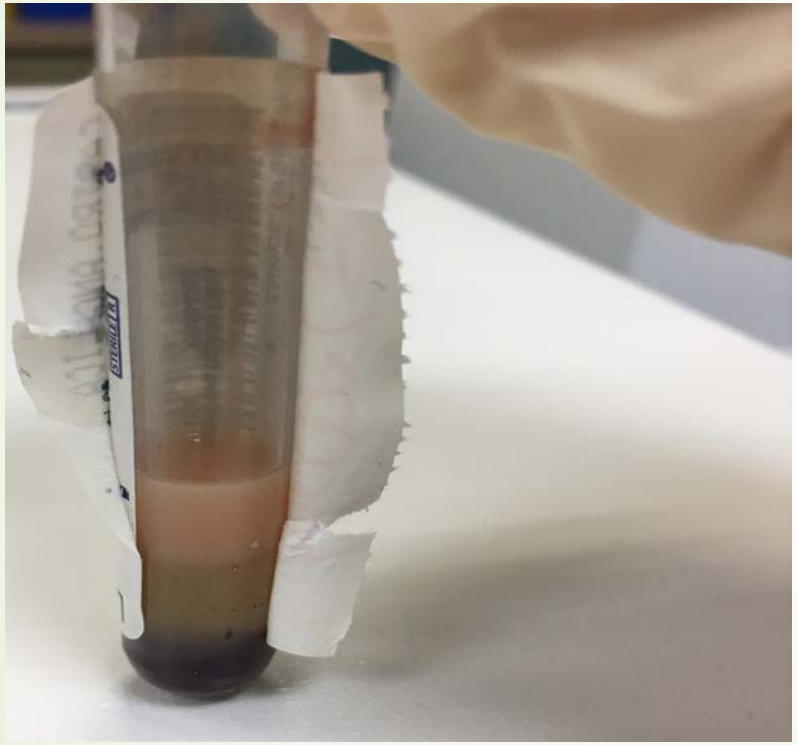
Case Presentation

- 34 y/o Hispanic female w/ a hx of congenital lipodystrophy of unknown type
- Major complications- uncontrolled DM with severe insulin resistance, DM nephropathy, neuropathy, retinopathy, gastroparesis, hepatosteatosis significant vascular atherosclerosis, hypertriglyceridemia and nephrotic syndrome

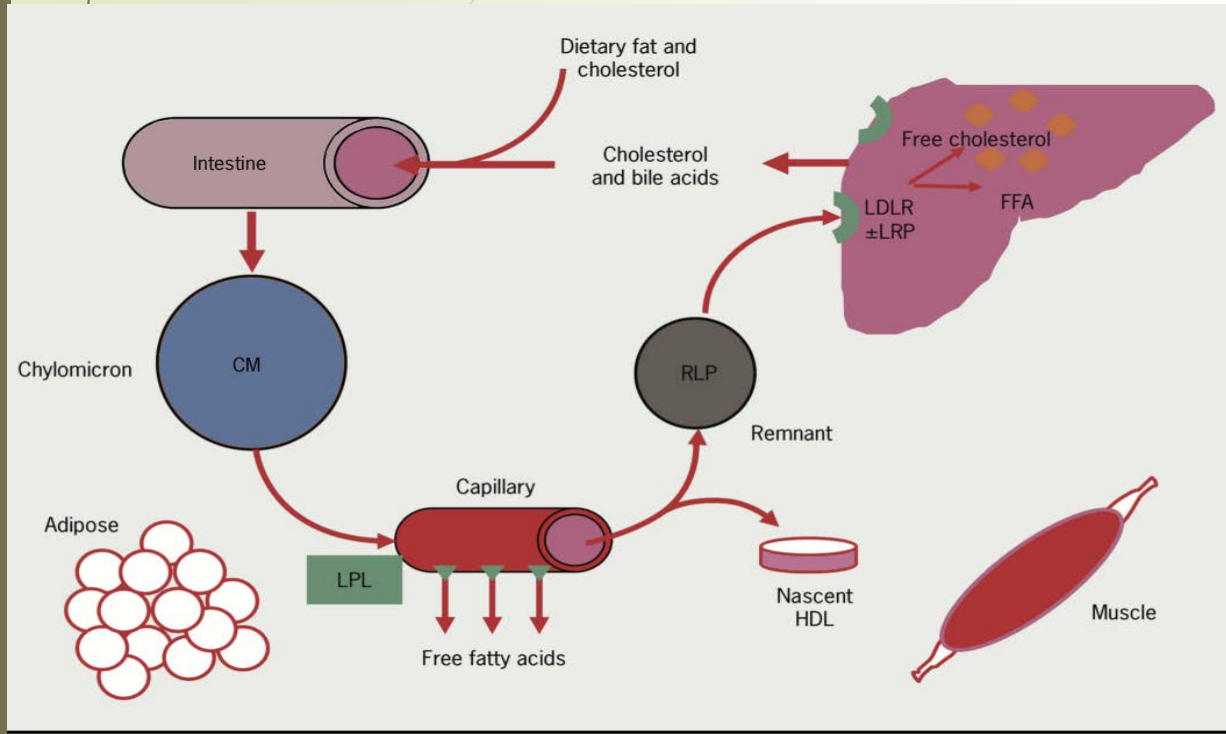
- In the ED, severe epigastric pain radiation to the back.
- Triglyceride level of 3,184 mg/dL (her baseline of 300mg/dL). Glucose of 432 mg/dL but urine with negative ketones. Lipase of 16u/L and amylase of 29u/L. LFTs were also WNL. A RUQ ultrasound and CT abdomen showed no acute pancreatic abnormalities. Started on insulin drip admitted to medical wards.

Continued…

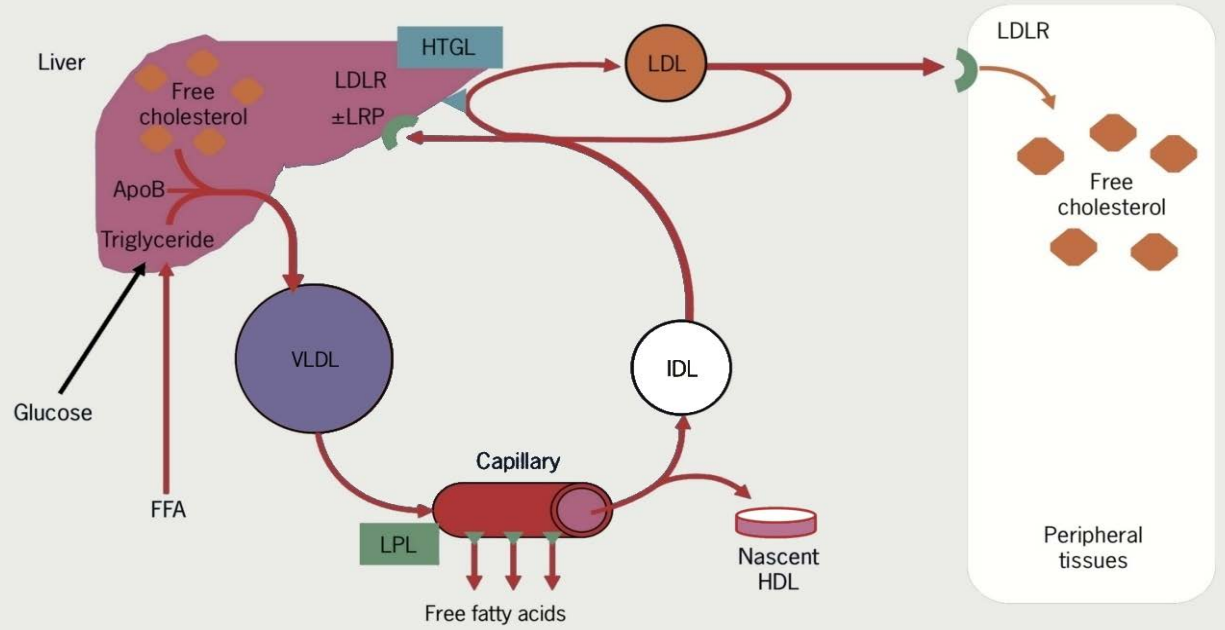
- Second day lipid profile study interpretation as evidence of Type V hyperlipoproteinemia.
- Triglyceride came down to 1500mg/dL.
- On the third day, tachycardia, leukocytosis, pleuritic chest pain. Intubated for ARDS.
- MICU course was uncomplicated received daily IV Lasix with adequate diuresis
- Triglycerides continues to come down and was at 800 before transitioning to subcu insulin.
- Extubated on the 5th day and transferred to the medical floors.



Direct Low Density Lipoprotein	
Lipoprotein, Cholesterol	641 * H
Lipoprotein, Triglyceride	3,379 * H
Lipoprotein, HDL	10 * L
Lipoprotein, LDL	68 *
APO-Lipoprotein B	183 * H
LDL Triglycerides	278 * H
VLDL Cholesterol	504 * H
VLDL Triglycerides	1,883 * H
Beta-VLDL Cholesterol	Not Detected *
Beta-VLDL Triglycerides	Not Detected *
Chylomicron Cholesterol	59 * H
Chylomicron Triglycerides	1,178 * H
Lp(a) Cholesterol	<3 *
LpX	Not detected *
Lipoprotein Interpretation	Type V Hyperlip



Key: CM = chylomicron; FFA = free fatty acids; HDL = high-density lipoprotein cholesterol; LDLR = low density lipoprotein receptor; LPL = lipoprotein lipase; LRP = LDL receptor-related protein; RLP = remnant lipoprotein



Key: ApoB = apolipoprotein B; FFA = free fatty acids; HDL = high-density lipoprotein cholesterol; HTGL = hepatic triglyceride lipase; IDL = intermediate-density lipoprotein cholesterol; LDLR = low-density lipoprotein receptor; LPL = lipoprotein lipase; LRP = LDL receptor-related protein; VLDL = very low-density lipoprotein cholesterol

Discussion

- In this case it was evident that the patient did not develop pancreatitis based on lipase levels and abdominal imaging.
- This is despite extremely elevated triglycerides in the blood
- Treatment of hypertriglyceridemia with insulin drip is standard to reduce the amount of triglycerides in the blood in hypertriglyceridemic pancreatitis. Has also been reported to be safe.
- In research on pancreatitis causing ARDS, FFA found to cause oxidative damage in addition to damage from pancreatic enzymes

Discussion

- We believe she likely developed ARDS from FFA mediated pulmonary endothelial damage and surfactant disruption with rapid treatment with insulin drip
- Presented with ARDS on second day of insulin infusion with decreasing triglycerides from 3,000 to 1,500 accompanied by high free fatty acid.
- The process likely predisposed by her comorbidities
- Future consideration in such patient may indicate a slower treatment of hypertriglyceridemia with insulin or to directly treat with plasmapharesis

Questions?

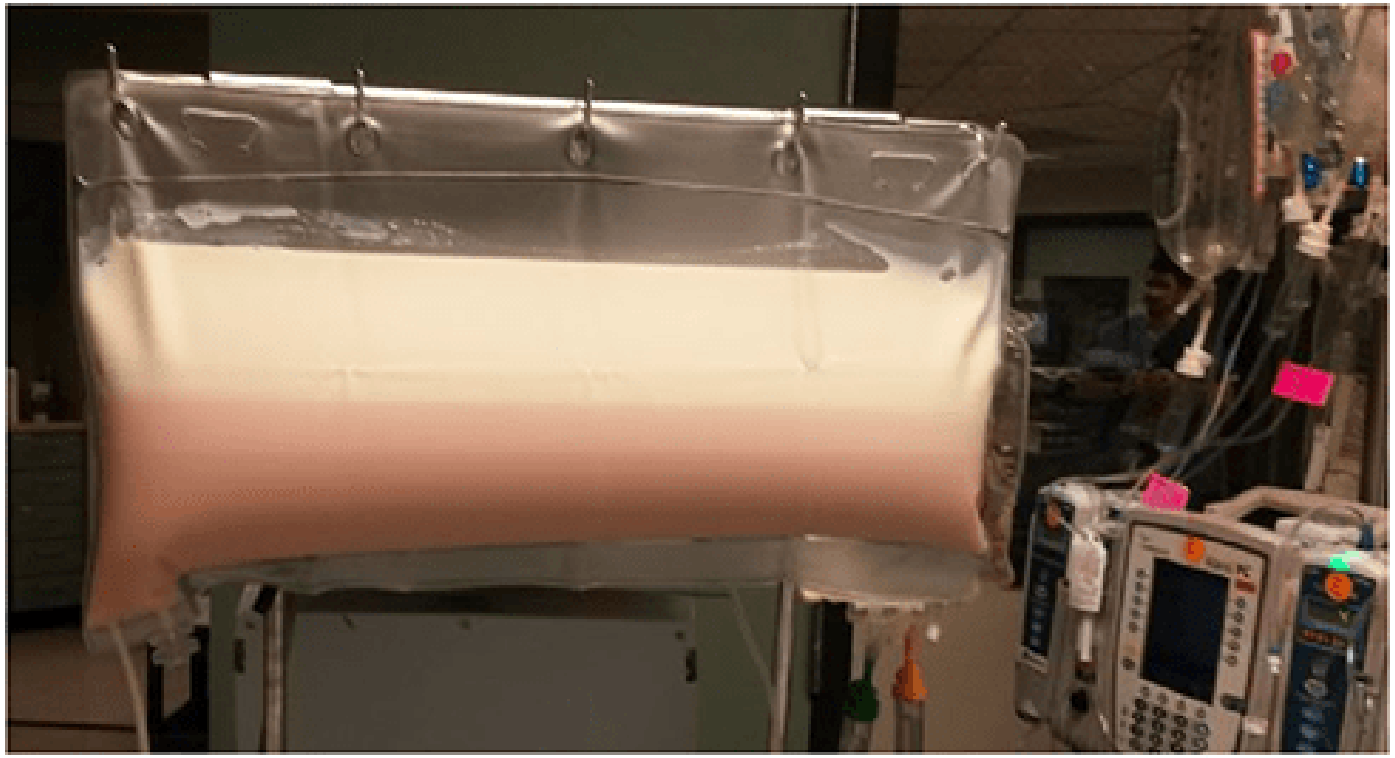


Figure 2 Milky ultrafiltrate obtained after a single cycle of plasmapheresis with drop in serum triglyceride levels from > 4000 mg/dL to < 500 mg/dL.

Gupta A et al. *J Gen Intern Med* 2015; 31:252

Thank You!

References

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