

Mr. M: Cystic Fibrosis and SVC Syndrome Complications

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Background: Cystic Fibrosis and SVC Syndrome

Cystic Fibrosis:

- Recessive, genetic disorder affecting secretory cells
- Thick mucus secretions impair digestive, pulmonary, and reproductive systems causing an increased risk of infection, decreased digestion and absorption, increased risk of diabetes/poor glycemic control

Nutrition concerns with Cystic Fibrosis:

- Increased risk of infection related to malnutrition risk
- Decreased digestion and absorption increases malnutrition risk—fat-soluble vitamins, calcium, possibly need for additional salt
- Increased risk of digestive problems: bulky stools, intestinal gas, gut distention, severe constipation
- Increased risk of diabetes, poor glycemic control related to impaired secretion from pancreas

Superior Vena Cava Syndrome:

- The superior vena cava (SVC) is a major vein leading to the heart—the syndrome occurs when SVC is partially blocked

Introduction to Mr. M

- 50 year old male
- PMH: Cystic fibrosis (CF) and SVC syndrome
 - Sternotomy (12/5/13) for SVC bypass graft
 - Positive for MDR Pseudomonas infection—treated with antibiotics
 - Hypercapneic respiratory failure and tracheostomy
 - Was weaning from vent at outside hospital
 - Admitted to HMC MICU with acute kidney failure in the setting of a 30-40 lb weight gain despite attempted diuresis

Assessment

Nutritional Concerns for Mr. M throughout admit:

- Adequate digestion and absorption of nutrients with CF
- Adequate nutrition to help prevent additional infections and maintain nutritional status for possible lung transplant
- Support continued healing from SVC graft surgery
- Long-term enteral nutrition—evaluate need for PEG
- Glycemic control
- Baseline chronic diarrhea/loose stool
- Need for strict NPO for maintaining lung health
- Fluid status and diuresis

Interventions

1. Maintain nutritional status for possible lung transplant

- Home vitamin and mineral supplements
- Importance of pancreatic enzymes timed with TF bolus
- Metabolic cart to monitor calorie needs:
3/10: AMEE 1967kcal; 4/27 AMEE 1613kcal
- Partially hydrolyzed tube feeding formulas for improved digestion/absorption: Vivonex and Vital 1.5

2. Long-term Enteral Nutrition—evaluate need for PEG

- Pt refused PEG hoping patient could eat adequately again
- PEG placement recommended per transplant discussion and GOC
- Pt consents and PEG is placed on 3/31

3. Glycemic control

- Pt with unexpected drops in blood glucose
- Worked with glycemic control team throughout admit to adjust feeds and insulin regimen
- Switched patient to bolus feeds

4. Baseline chronic diarrhea/loose stool

- Worsened with bolus tube feeding
- Bolus vs continuous tube feeding: improved glycemic control with bolus, improved stooling with continuous
- Increased pancreatic enzyme dose and improved timing with bolus feeds due to greasy stools
- Team later adds loperamide - stooling improves significantly

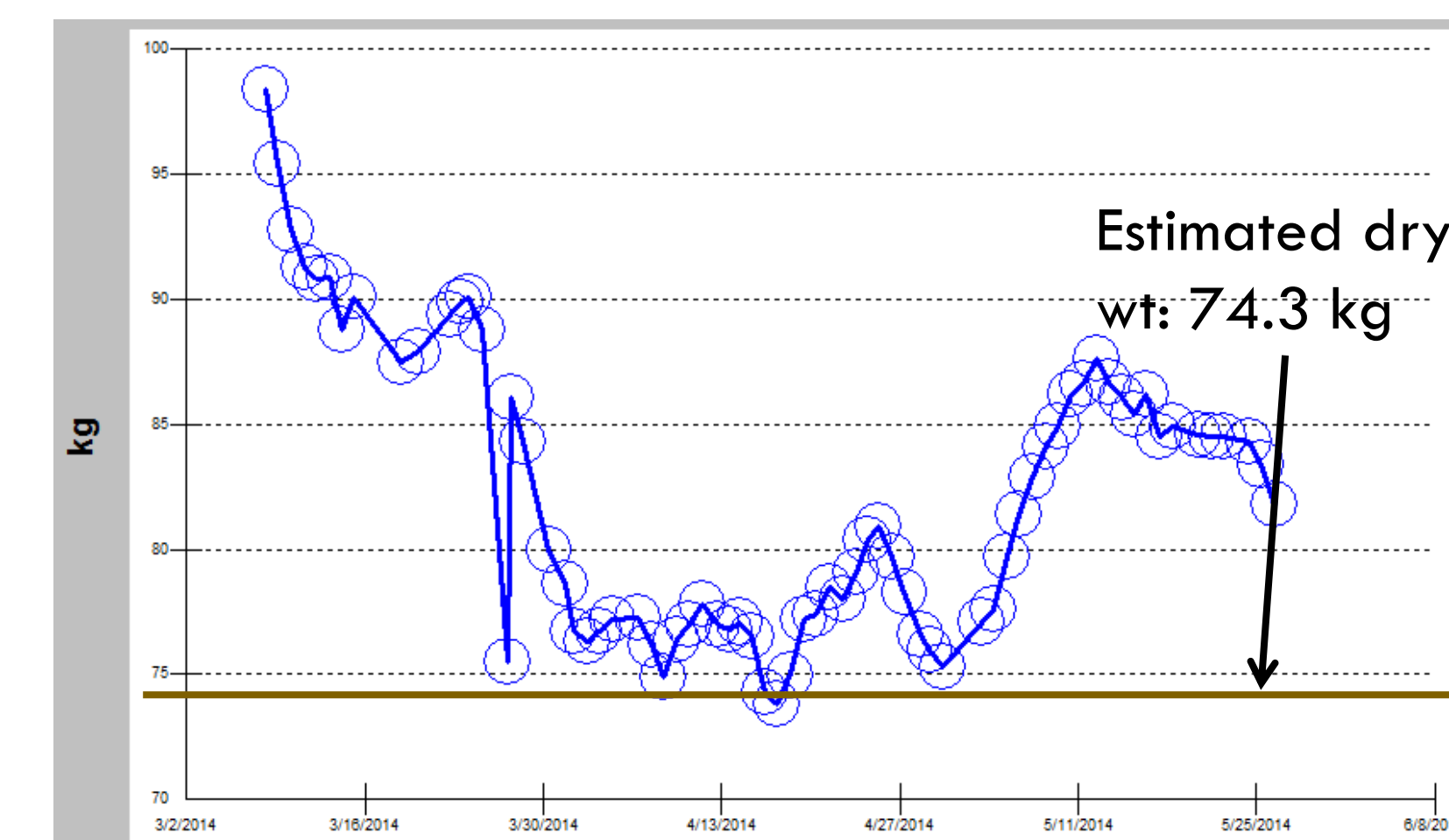


Figure 1: Daily weight—fluctuations related primarily to fluid status. Patient with 30-40 lb of fluid on admit

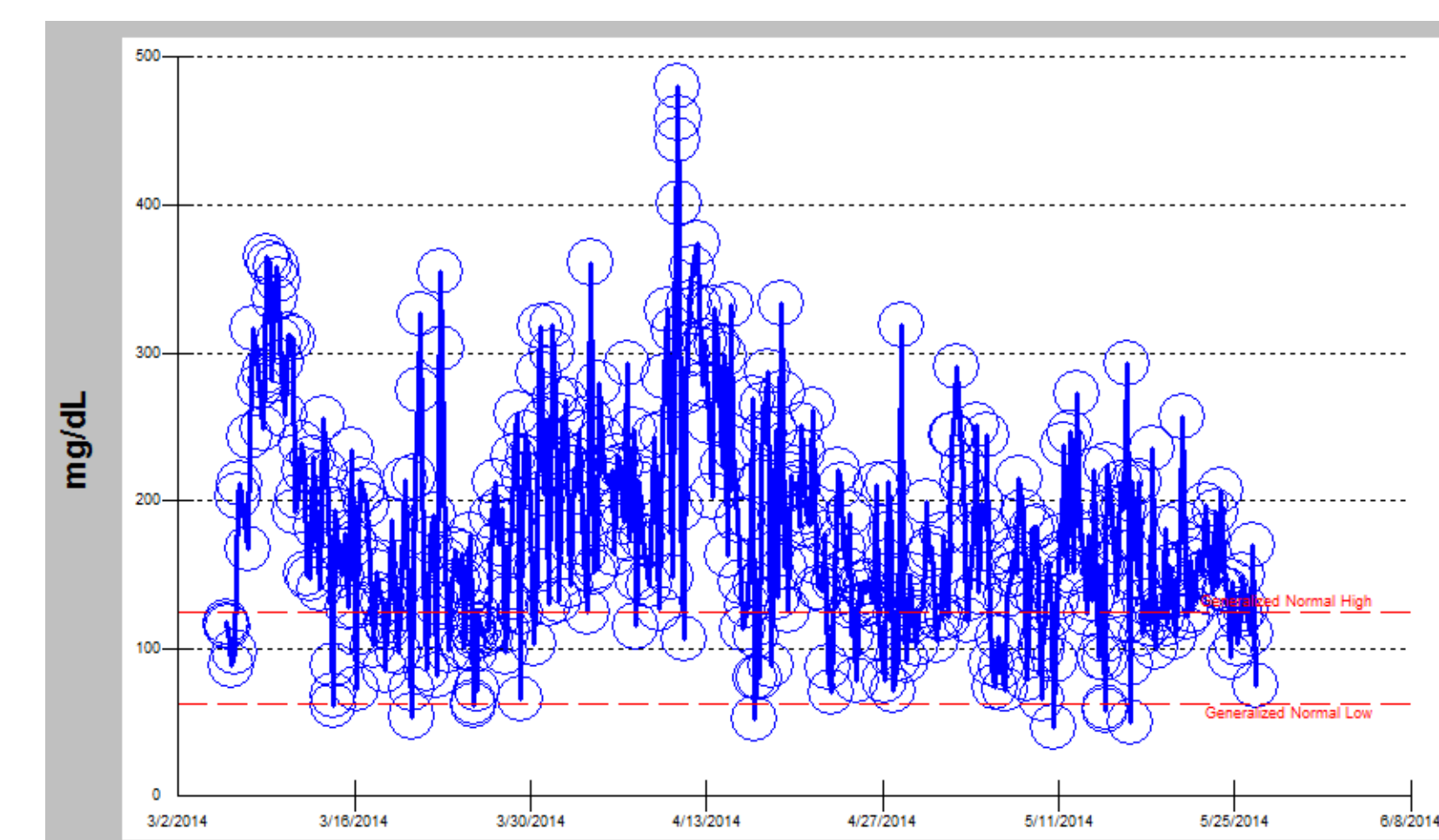


Figure 2: Blood glucose levels throughout admit: patient with highly variable levels—improved glycemic control over time

Discussion

Goal to list patient as lung transplant candidate:

- Maintain lung health—strict NPO status
- Increase stamina through daily walks and pressure support
- Maintain overall health, including nutritional status
 - Continue tube feeds, multivitamin, pancreatic enzymes
 - Monitor stooling

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