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

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; 4/27AMEE 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

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

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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