Agenda

- Cystic Fibrosis Foundation (CFF) Guidelines
- Annual tests include:
  - Oral Glucose Tolerance Test (OGTT)
  - Dual Energy X-ray Absortiometry (DXA)
  - Fat-soluble vitamin labs
- Improving patient outcomes
Cystic Fibrosis Foundation Guidelines
• Recommendations and statements created by a multi-disciplinary board of CF practitioners
  ▫ CFRD Guidelines 2010 developed by CFF in collaboration with ADA and PES

• Labs and procedures ordered annual as part of health maintenance and monitoring
  ▫ Are the tests always done annually?
National Statistics

• Results are entered into PortCF database on an annual basis
• National statistics are published yearly by CFF:
  ▫ Compare our CF center’s progress year-to-year
  ▫ Compare CF centers nationally
  ▫ May be used for research and education purposes
Oral Glucose Tolerance Test
What’s the big deal?

- CF-related diabetes (CFRD) is often clinically silent
- Concern regarding nutrition and pulmonary consequences:
  - Unintentional weight loss
  - Protein catabolism
  - Decline in lung function
  - Increase in mortality
  - Increased infection risk
  - Increased risk of developing microvascular complications
The test

• OGTT is the gold standard for screening
• Must be done fasting
  ▫ No food or drink for 8-12 hrs before test
• At the lab:
  ▫ Fasting blood glucose (BG) is taken
  ▫ Drink ~ 75 grams glucose
  ▫ Check blood glucose trend after 30 mins, 1 hr, and 2 hrs
• Plan ahead → do on day of clinic visit.
How often do I need to get it done?

- Recommended for all patients $\geq 10$ yrs old
- The test **must** be done annually
  - Unless you meet the criteria for CFRD
- Exocrine pancreatic insufficiency vs. sufficiency
  - Does it matter?
- Pregnancy
  - Routinely performed in 2\textsuperscript{nd} trimester
  - If had Gestational DM, must complete OGTT 6-12 weeks post-partum
Interpreting the results

The results are interpreted by the CF team.

You can be diagnosed with CFRD if:

- Fasting BG ≥ 126 mg/dL
- 2 hr BG ≥ 200 mg/dL
- A1c > 6.5% at time of OGTT
  - With 2 hr BG ≥ 200 mg/dL
- Casual BG ≥ 200 mg/dL + symptoms of polyuria, polydipsia

*It is possible to be diagnosed with CFRD during acute illness, while on steroids, and/or with nutrition support.
Interventions

- If the results are normal:
  - Repeat the test annually
- Impaired glucose tolerance [IGT] and Indeterminate glycemia [INDET]
  - May be given a glucometer
  - Increased risk for developing CFRD
- If diagnosed with diabetes:
  - Prescribed a glucometer
  - Check your BS 3-4 x/day or as recommended
  - Referral to an endocrinologist
  - Ongoing education and counseling
Can I prevent diabetes?

- Not really ....
  - CFRD is not like Type I or Type II DM
  - Insulin deficiency vs. insufficiency vs. resistance

- Role of diet and exercise
  - Need for high calorie, high fat, & protein diet
    - Balanced meals
    - Role of fats and protein
  - Goal: > 150 min moderate aerobic exercise/week
Dual Energy X-ray Absorptiometry (DXA)
Bone Disease in Cystic Fibrosis

DXA Procedure

- Performed in the Nuclear Medicine department
- Lay flat on x-ray table
- A scan is completed of hip and spine to check for osteoporosis
How to interpret the results

• Compares your bone density to the bone density of:
  ▫ A healthy young adult (T score)
  ▫ Someone of your same age, gender, and ethnicity (Z score)
    • Looked at more often in adult population
• Results are given in the radiologist’s report
  ▫ Interpreted by the CF Attending and CF NP
Interventions

• Normal ↔ Osteopenia ↔ Osteoporosis
• Depending on trend of bone density score:
  ▫ Calcium supplementation
  ▫ Additional vitamin D (and vitamin K) supplement
  ▫ Weight bearing activity
  ▫ Sun exposure
  ▫ Antiresorptive agents prescribed
    • Bisphosphates (Aredia, Fosamax)
Screening (DXA) and Treatment Protocol

Screen all adults and children > 8 years old if <90% Ideal body weight, FEV1 < 50% predicted, glucocorticoids of ≥ 5 mg/day for ≥ 90 days/yr, delayed puberty, or a history of fractures

Baseline DXA

T/Z score\(^*\) ≥ -1.0

-1.0 > T/Z score\(^*\) > -2.0

T/Z score\(^*\) ≤ -2.0

Nutrition

Vitamin D Supplementation
See text (evidence grade III).
Calcium Supplementation
0.3-0.5 mg (1.7-2.9 nmol)/day (2 ADEKS)
Target BMI >25th percentile (both evidence grade II-I).
Encourage, outdoor weight bearing exercise (evidence grade I)

Repeat DXA every 5 years

Pulmonary & Endocrine Topics

Aggressive pulmonary infection treatment (evidence grade III).
Minimize steroid dosing (evidence grades I-II).
Treat CF Diabetes, delayed puberty or hypogonadism (evidence grades I-II).
Endocrine referral.
If fragility fractures have occurred, patient is awaiting transplant or BMD loss is >3-5%/yr, start bisphosphonate (evidence grades I-III)

Repeat DXA 2-4 years

Consider Bisphosphonates

Oral
Alendronate 70 mg weekly (or 10 mg daily)
Risedronate: 35 mg weekly (or 5 mg daily)

IV
Pamidronate 30 mg in 500 ml saline infused over 3 hrs every 3 months (evidence grade I).
Zoledronic acid: 4-5 mg infused over 15-20 minutes yearly (evidence grade III).

Annual DXA

Fat Soluble Vitamin Labs
- Need fat in the diet for vitamin absorption in the intestine
- With EPI and malabsorption certain vitamins may not be absorbed well
- Fat soluble vitamins:
  - Vitamin A, D, E, and K
Vitamin A

• Role
  ▫ Vision, bone growth, reproduction, helps regulate the immune system, promotes healthy surface linings
• Lab ordered → serum free retinol level
• Sources
  ▫ Preformed vitamin A – liver, whole milk, eggs, some fortified foods
  ▫ Proformed vitamin A – fruits and vegetables, oatmeal
Vitamin E

• Role
  ▫ Antioxidant, metabolic processes, regulating gene expression, role in immune function,
• Lab ordered serum alpha-tocopherol level
• Sources
  ▫ Nuts, seeds, nut oils, dark leafy green vegetables
Vitamin D

- **Role**
  - Help with calcium absorption, bone health, immune function, reduce inflammation,
- **Lab ordered** 25-hydroxy vitamin D
- **Sources**
  - Dairy, fortified juices, some fish, fish liver oils
  - Sunlight
  - Supplements (cholecalciferol or ergocalciferol)
Vitamin K

• Role
  ▫ *Blood coagulation, bone metabolism*
• Lab monitored ➔ prothrombin time (PT)
• Sources
  ▫ *Leafy green vegetables, avocado, kiwi fruits*
Interpreting the lab results

- Results are reviewed by CF team
- Check compliance with taking CF-specific MVI
- Dietary Intake
- Sunlight exposure
  - Vitamin D levels
- Additional vitamin supplements
- Closer monitoring of lab trends
Game Plan For Our Center
Improving Patient Outcomes

- All patients are asked to complete labs annually
  - 1st quarter of the year
  - Some tests are not required annually
- Letters mailed to patient
  - If OGTT or DEXA not completed within required time frame
- Lab location
  - Do it at SUH lab for faster results!
  - Must discuss with RN Coordinator about doing labs locally
Questions?