

A photograph of a large, modern Stanford University building with a prominent palm tree in the foreground. The building has a dark, angular roof and large glass windows. The scene is dimly lit, suggesting dusk or dawn. The text is overlaid on the left side of the image.

COLON CANCER SCREENING IN CYSTIC FIBROSIS

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Department
of Medicine

Outline

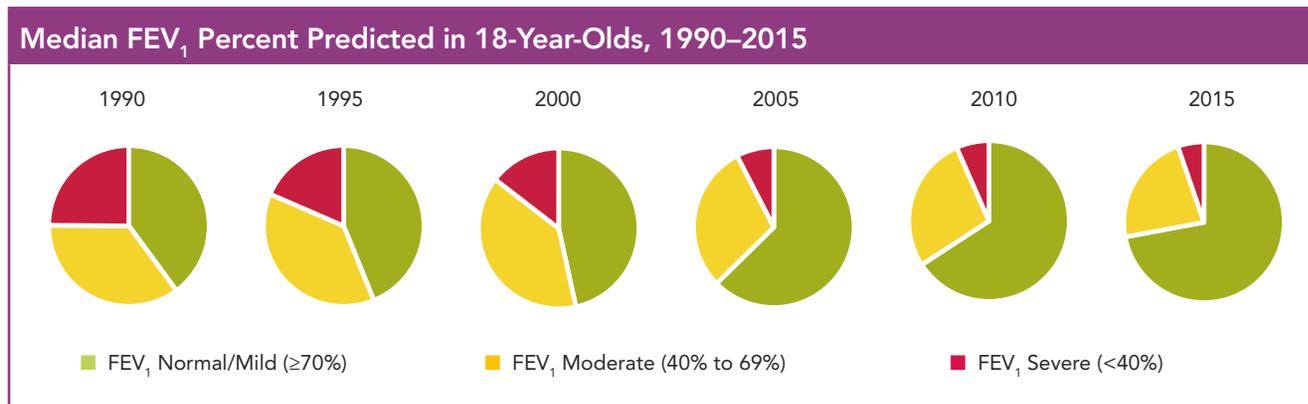
- Background
- Risk of colorectal cancer in cystic fibrosis
- Screening Recommendations
- Challenges and Opportunities
- Summary

Background

- Colorectal cancer (CRC):
 - 2nd leading cause of cancer deaths
 - 134000 new cases in 2016
 - Estimated 49000 deaths in 2016

Background

- Improving pulmonary function in adults with Cystic Fibrosis
 - FEV₁ is the single best predictor of mortality in CF patients
 - US CF Foundation patient registry 2015



Colorectal Cancer Risk in Cystic Fibrosis

- Cystic Fibrosis (CF)
 - Concern: cases of CRC in CF population will rise
 - Increased life expectancy + large age-related increased risk of CRC
- Adults with CF: **5 to 10 times risk** compared to general population
- CF patients after organ transplantation: **25 to 30 times** greater risk compared to general population

Hadjiliadis et al. *Gastroenterology* 2018; 154: 736 - 745

Colon Cancer in Cystic Fibrosis

- Nearly all CRC in CF patients observed in patients **below age 50 years**
- **Average onset** of CRC in CF population is **40 years**, or about 20 to 30 years younger than in the non-CF population
- SEER: incidence of CRC in CF population at age 40 to 49 years is similar to incidence of CRC in 65 to 69 year old persons in the non CF population

Why the increased risk?

- Inflammatory and non-inflammatory risk factors
- CFTR gene is a tumor suppressor gene in the intestinal tract in mice
 - Loss of CFTR is associated with intestinal tumor formation
- Loss of CFTR expression in early stage human CRC in non-CF patients is associated with markedly decreased disease-free survival
- Cystic fibrosis: a colon cancer syndrome

Than BL et al *Oncogene* 2016;35(32):4179-4187

Starr TK et al *Science* 2009;323(5922):1747-1750

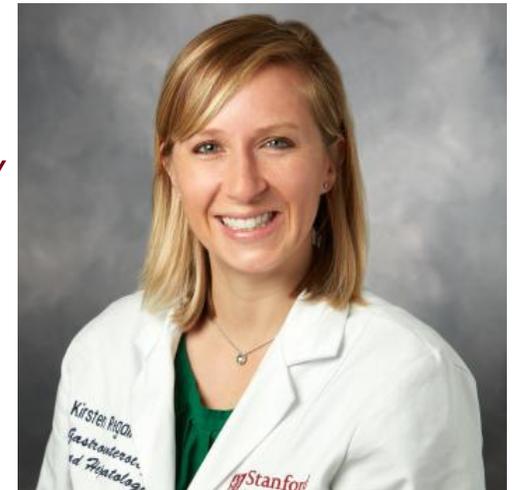
Screening and Prevention: Consensus Recommendations

- All decisions on colorectal cancer screening and surveillance in individuals with cystic fibrosis be based on *shared decisions* between the provider and individual with CF about treatment, comorbidities, safety and quality of life
 - *Reviewing guidelines at CF Education Day*
 - *Reviewing guidelines with CF team members*
 - *Discussion at clinic visits*

Hadjiiladis et al. Gastroenterology 2018; 154: 736 - 745

Screening and Prevention: Consensus Recommendations

- All colorectal cancer screening and surveillance for individuals with cystic fibrosis are jointly managed by cystic fibrosis health care professionals and an endoscopist: *dedicated CF trained endoscopist familiar with CF GI issues*
- *Collaboration with Stanford Gastroenterology*
 - *Dr Kirsten Regalia*



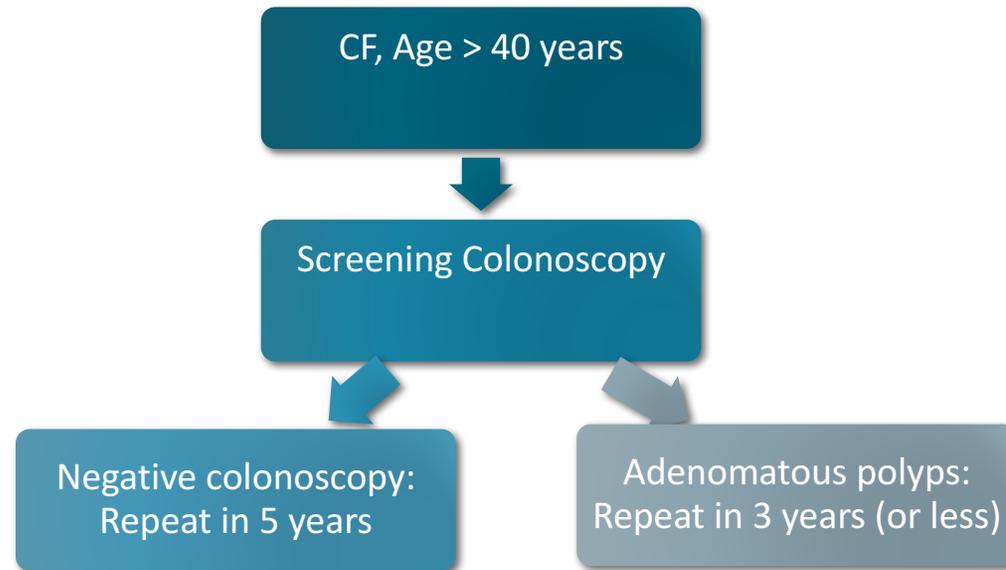
Screening and Prevention: Consensus Recommendations

- Colonoscopy as the screening exam
- Evidence is insufficient to recommend use of CT colonography, stool based tests or flexible sigmoidoscopy
- Intensive bowel preparation for optimal exam

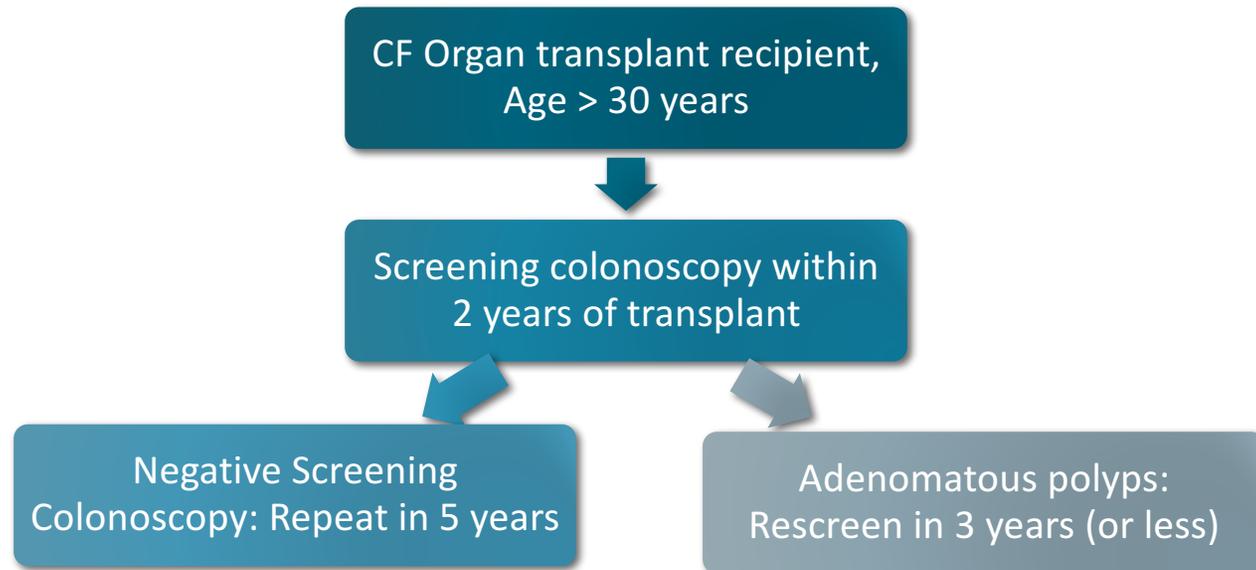
Screening and Prevention: Consensus Recommendations

- Colorectal cancer screening begin at age 40 years in individuals with CF
- After solid organ transplant, patients who are 30 years or older should begin colon cancer screening within 2 years of transplant

Screening and Prevention: Consensus Recommendations



Screening and Prevention: Consensus Recommendations



Colon Cancer Prevention in CF

- Pharmacologic agents (investigational)
- Effect of CFTR Modulators?

Colon Cancer Screening

- Challenges:
 - Colonoscopy
 - Nature of procedure and intensive bowel preparation
 - Time away from work/ life
 - Patient population with underlying lung disease
 - Test findings may provoke anxiety
- Team-based approach to individualize decisions to the patient and in keeping with their values and preferences

Summary

- Let's discuss colon cancer screening at your next Cystic Fibrosis Clinic Visit!!