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Clinician Management Resource for *APC* (Familial Adenomatous Polyposis)

This overview of clinical management guidelines is based on this patient's positive test result for an *APC* gene mutation. Unless otherwise stated, medical management guidelines used here are limited to those issued by the National Comprehensive Cancer Network® (NCCN®)[1] in the U.S. Please consult the referenced guideline for complete details and further information.

Clinical correlation with the patient's past medical history, treatments, surgeries and family history may lead to changes in clinical management decisions; therefore, other management recommendations may be considered. Genetic testing results and medical society guidelines help inform medical management decisions but do not constitute formal recommendations. Discussions of medical management decisions and individualized treatment plans should be made in consultation between each patient and his or her healthcare provider, and may change over time.

SCREENING/SURGICAL CONSIDERATIONS*,[1]	AGE TO START	FREQUENCY
Colorectal Cancer		
Unaffected patients (*i.e.* no symptoms, findings, adenomas): Colonoscopy (preferred) or flexible sigmoidoscopy	10-15 years old	Every 12 months
Affected patients: Colectomy or proctocolectomy**	Individualized by polyp burden	N/A
Additional surveillance post-colectomy is recommended and varies by surgery type performed. *Please consult complete NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for additional details.*	Post-surgery	Varies by surgery type
Consider chemoprevention to facilitate management of the remaining rectum for patients who have undergone surgery.	Post-surgery	N/A
Duodenal Or Periampullary Cancer^		
Upper endoscopy including complete visualization of the ampulla of Vater *Refer to the complete NCCN Guidelines® for further details regarding type and frequency of surveillance.*	20-25 years old (consider baseline upper endoscopy earlier, if family history of aggressive duodenal adenoma burden or cancer)	Individualized
Gastric Cancer^		
Refer to the complete NCCN Guidelines for further details regarding the management of fundic gland and non-fundic gland polyps, if applicable.	Individualized	Individualized
Thyroid Cancer^		
Thyroid exam • Ultrasound	Late teenage years	If normal, consider repeating every 2-5 years. If abnormal, refer to a thyroid specialist†
Central Nervous System Cancer^		
Physical exam *No additional screening recommendations^^*	Individualized	Every 12 months
Intra-Abdominal Desmoids^		
Consider abdominal MRI (with and without contrast) or CT (with contrast), if family history of symptomatic desmoids and/or suggestive abdominal symptoms	Individualized	Annually, at minimum
Small Bowel Polyps And Cancer^		
Consider adding small bowel visualization via capsule endoscopy, especially if duodenal polyposis is advanced.^^	Individualized	Individualized

* *Some individuals have a milder phenotype of FAP, known as attenuated FAP (AFAP). Recommendations for individuals with AFAP may vary from the guidelines outlined here, including differences in the age, frequency, and type of management recommended. Please consult the complete NCCN Guidelines (as referenced on page 2) for management details.*

** *Timing of proctocolectomy in patients <18 y of age is not established since colon cancer is rare before age 18. In patients <18 y without severe polyposis and without family history of early cancer or severe genotype, the timing of proctocolectomy can be individualized. An annual colonoscopy is recommended if surgery is delayed.*

^ *Other than colorectal cancer, screening recommendations are expert opinion rather than evidence-based.*

^^ *Limited data*

† *Shorter intervals may be considered for individuals with a family history of thyroid cancer*

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SCREENING/SURGICAL CONSIDERATIONS*,[1]	AGE TO START	FREQUENCY
Hepatoblastomaˆ		
May consider • Liver palpation, abdominal ultrasound, measurement of alpha-fetoprotein (AFP)ˆˆ.	0-5 years old	Every 3-6 months
Pancreatic Cancerˆ		
Screening may be individualized based on family history of pancreatic cancerˆˆ	Individualized	Individualized

* *Some individuals have a milder phenotype of FAP, known as attenuated FAP (AFAP). Recommendations for individuals with AFAP may vary from the guidelines outlined here, including differences in the age, frequency, and type of management recommended. It is recommended that patients be managed by physicians or centers with expertise in FAP and that management be individualized to account for genotype, phenotype, and personal considerations. Please consult the complete NCCN Guidelines (as referenced below) for management details.*

ˆ *Other than colorectal cancer, screening recommendations are expert opinion rather than evidence-based.*

ˆˆ *Limited data*

1. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Genetic/Familial High-Risk Assessment: Colorectal. V1.2020. © National Comprehensive Cancer Network, Inc. 2020. All rights reserved. Accessed August 20, 2020. To view the most recent and complete version of the guideline, go online to NCCN.org. NCCN makes no warranties of any kind whatsoever regarding their content, use or application and disclaims any responsibility for their application or use in any way.

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