

## Clinician Management Resource for *STK11* (Peutz-Jeghers syndrome)

This overview of clinical management guidelines is based on this patient's positive test result for a *STK11* gene mutation. Unless otherwise stated, medical management guidelines used here are limited to those issued by the National Comprehensive Cancer Network® (NCCN®)<sup>1,2</sup> 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*	AGE TO START	FREQUENCY
<b>Breast Cancer<sup>1,2</sup></b>		
Clinical Breast Exam	30 years old	Every 6 months
Breast Screening ▪ Mammogram and breast MRI	30 years old	Every 12 months
For consideration of risk-reducing mastectomy, manage based on family history <sup>2</sup>	Individualized	N/A
<b>Colorectal Cancer<sup>1</sup></b>		
Colonoscopy	Late teens	Every 2-3 years
<b>Gastric Cancer<sup>1</sup></b>		
Upper endoscopy	Late teens	Every 2-3 years
<b>Intestinal Cancer<sup>1</sup></b>		
Small bowel visualization via CT or MRI enterography or video capsule endoscopy	8-10 years old (baseline) but at least by age 18 y with follow-up interval based on findings	Every 2-3 years (this may be individualized, or increased with symptoms)
<b>Pancreatic Cancer<sup>1,2</sup></b>		
Consider screening using contrast-enhanced MRI/MRCP and/or EUS <sup>†</sup>	30-35 years old, or 10 years younger than the earliest exocrine pancreatic cancer diagnosis in the family, whichever is earlier <sup>2</sup>	Every 12 months. May consider shorter screening intervals based on clinical judgment for individuals found to have worrisome abnormalities on screening.
<b>Ovarian**, Cervical<sup>^</sup>, and Uterine Cancer<sup>1</sup></b>		
Physical exam for observation of precocious puberty	8 years old	Annual
Pelvic exam and Pap smear	18-20 years old	Every 12 months
<b>Testicular Cancer<sup>1,^^</sup></b>		
Testicular exam and observation for feminizing changes	10 years old	Every 12 months
<b>Lung Cancer<sup>1</sup></b>		
Provide education about symptoms and smoking cessation	Individualized	N/A

\* Due to the rarity of the syndrome and complexities of diagnosing and managing individuals with Peutz-Jeghers syndrome, referral to a specialized team is recommended.

\*\* Typically benign sex cord/Sertoli cell tumors    ^ Typically cervical adenoma malignum    ^^ Typically sex cord/Sertoli cell tumors

† For individuals considering pancreatic cancer screening, the panel recommends that screening be performed in experienced high-volume centers, ideally under research conditions. The panel recommends that such screening only take place after an in-depth discussion about the potential limitations to screening, including cost, the high incidence of pancreatic abnormalities, and uncertainties about the potential benefits of pancreatic cancer screening.

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