

## The Department of Vermont Health Access Clinical Criteria

**Subject:** Familial Adenomatous Polyposis/Associated Polyposis Conditions (APC) Genetic Testing

**Last Review:** August 30, 2022\*

**Past Revisions:** February 18, 2020, November 1, 2017, and August 30, 2016

**\*Please note: Most current content changes will be highlighted in yellow.**

### Description of Service or Procedure

Familial Adenomatous Polyposis (FAP) and Attenuated Familial Adenomatous Polyposis (AFAP) are caused by mutations of the Adenomatous Polyposis Coli (APC) gene inherited by **autosomal dominant inheritance**. FAP and AFAP are characterized by the development of numerous colonic or adenomatous polyps and frequently result in development of cancers of the large intestine and rectum.

**Turcot syndrome, Gardner syndrome, and gastric adenocarcinoma and proximal polyposis of the stomach (GAPPS), all subtypes of FAP are also associated with mutations of the APC gene and are variants of FAP. GAPPS is associated with high risk of gastric cancer.**

**MUTYH-associated polyposis (MAP) is caused by mutation of both alleles of the MUTYH gene inherited by autosomal recessive inheritance. MAP is also characterized by the development of numerous adenomatous polyps and frequently results in development of cancer of the large intestine and rectum.**

#### Presentation:

- FAP is characterized by the presence of hundreds to thousands of adenomas in the colon and rectal area.
- AFAP is characterized by the presence of less than 100 adenomatous polyps in the colon and rectal.
- Gardner syndrome is characterized by colon polyps as well as growths outside the colon such as **benign cutaneous lesions, osteomas, dental abnormalities**, and desmoid tumors.
- Turcot syndrome is the association of colonic adenomatous polyposis and CNS tumors, usually medulloblastoma.
- GAPPS is characterized by greater than 100 polyps of the gastric body and fundus with antral sparing
- **MAP is characterized by the presence of ten to a few hundred adenomas in the colon area as well as thyroid nodules, benign adrenal lesions, jawbone cysts, and congenital hypertrophy of the retinal pigment epithelium.**



In FAP, the formation of polyps begins in childhood for the majority of individuals typically in the distal colon. By adolescence, polyp formation has spread throughout the colon, increasing in size and number. Average age of colorectal cancer development in an individual with FAP is within the third decade of life and for an individual with AFAP by the fifth decade of life. Lifetime risk for colorectal cancer for individuals with FAP if untreated is 100%.

Because mutations of the APC and MUTYH genes may greatly predispose the individual to cancers including colorectal and gastric adenocarcinoma and require early intervention, genetic testing may be considered for individuals with clinical symptoms of FAP, AFAP, Gardner syndrome, Turcot syndrome, and MAP.

## **Disclaimer**

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Coverage is limited to that outlined in Medicaid Rule or Health Care Administrative Rules that pertains to the member's aid category. Prior Authorization (PA) is only valid if the member is eligible for the applicable item or service on the date of service.

## **Medicaid Rule**

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Medicaid and Health Care Administrative Rules can be found at <https://humanservices.vermont.gov/rules-policies/health-care-rules/health-care-administrative-rules-hcar/adopted-rules>

7102.2	Prior Authorization Determination
7405	Laboratory and Radiology Services
4.101	Medical Necessity for Covered Services
4.104	Medicaid Non-Covered Services

## **Coverage Position**

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APC and MUYTH genetic testing may be covered for beneficiaries:

- When the test is prescribed by a licensed medical provider, enrolled in the Vermont Medicaid program, operating within their scope of practice as on the Vermont's Office of Professional Regulation's website\*, Statute, or rule who is knowledgeable regarding APC and MUYTH genetic testing and who provides medical care to the member AND
- When the clinical criteria below are met.

\* Vermont's Office of Professional Regulation's website: <https://sos.vermont.gov/opr/>

## **Coverage Criteria**

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APC and MUYTH genetic testing may be covered for members who have clinical symptoms consistent with FAP, AFAP, Gardner's syndrome or Turcot's Syndrome, or MAP in accordance with the National Comprehensive Cancer Network® Clinical Practice Guidelines in Oncology for genetic/familial high-risk assessment of colorectal cancers.

APC and MUTYH gene testing for members suspected to have FAP, AFAP, or MAP may be considered when the following conditions are met:

- Personal history of greater than or equal to 20 cumulative adenomas

- When familial variant is known, deletion/duplication genetic testing is considered medically necessary for the specific APC or MUTYH variant. APC genetic testing is considered medically necessary for first degree relatives of an individual with FAP, etc.
- Multifocal/bilateral congenital hypertrophy of retinal pigment epithelium (CHRPE)
- Additionally testing may be considered for personal history of between 10-19 cumulative adenomas, desmoid tumor, hepatoblastoma, cribriform-morular variant of papillary thyroid cancer, unilateral CHRPE, or serrated polyposis syndrome (previously know as hyperplastic polyposis) with at least some adenomas. All clinical history should be taken into account when these findings are present.

Considerations:

Consider which gene should be tested when heritability pattern is clear. For example, if only one sibling is exhibiting colonic polyposis, consider recessive inheritance, which would point to MUTYH testing.

When there is a known familial mutation in the APC gene and familial history of FAP, genetic testing should be completed by 10 years of age to align with initiation of cancer screening. This should similarly be considered for family history of AFAP.

Prenatal diagnosis or pre-implantation genetic testing may be considered if a pathogenic variant has been identified in an affected family member.

Per CMS, Vermont Medicaid will “provide comprehensive services and furnish all Medicaid coverable, appropriate, and medically necessary services needed to correct and ameliorate health conditions, based on certain federal guidelines.”

Providers requesting this test should provide pre- and post-test genetic counseling for the member and family, if applicable.

Early and Periodic Screening, Diagnostic and Treatment (EPSDT): Vermont Medicaid will provide comprehensive services and furnish all Medicaid coverable, appropriate, and medically necessary services needed to correct and ameliorate health conditions for Medicaid members under age 21.

Please note, Vermont Medicaid Clinical Criteria is reviewed based on available literature, evidence- based guidelines/standards, Medicaid rule and policy, and Medicare coverage determinations that may be appropriate to incorporate when applicable.

### **Clinical criteria for repeat service or procedure**

Repeat complete sequencing of the same gene (APC or MUTYH) is not indicated. Repeat testing for the same pathogenic variant is not indicated.

### **Type of service or procedure covered**

Genetic testing of patients with suspected adenomatous polyposis syndromes should include APC and MUTYH-associated polyposis (MAP) gene mutation analysis if no known familial pathogenic variant/mutation.

## References

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