

Overview

Useful For

Identifying fumarate hydratase-deficient neoplasms

Supporting the diagnosis of an atypical smooth muscle tumor over leiomyosarcoma

Reflex Tests

Test ID	Reporting Name	Available Separately	Always Performed
IHTOI	IHC Initial, Tech Only	No	No
IHTOA	IHC Additional, Tech Only	No	No

Testing Algorithm

For the initial technical component only immunohistochemical (IHC) stain performed, the appropriate bill-only test ID will be reflexed and charged (IHTOI). For each additional technical component only IHC stain performed, an additional bill-only test ID will be reflexed and charged (IHTOA).

Method Name

Immunohistochemistry

NY State Available

Yes

Specimen

Specimen Type

TECHONLY

Advisory Information

[This test includes only technical performance of the stain \(no pathologist interpretation is performed\). If diagnostic consultation by a pathologist is required order PATHC / Pathology Consultation.](#)

Shipping Instructions

Attach the green pathology address label and the pink Immunostain Technical Only label included in the kit to the outside of the transport container.

Specimen Required

Specimen Type: Tissue

Supplies: Immunostain Technical Only Envelope (T693)

Container/Tube: Immunostain Technical Only Envelope

Preferred: 2 unstained positively charged glass slide (25- x 75- x 1-mm) per test ordered; sections 4-microns thick

Acceptable: Formalin-fixed, paraffin-embedded (FFPE) tissue block

Digital Image Access

1. Information on accessing digital images of IHC stains and the manual requisition form can be accessed through this website: www.mayocliniclabs.com/test-info/ihc/index.html

2. Clients ordering stains using a manual requisition form will not have access to digital images.

3. Clients wishing to access digital images must place the order for IHC stains electronically. Information regarding digital imaging can be accessed through this website: www.mayocliniclabs.com/test-info/ihc/faq.html

Reject Due To

Wet/frozen tissue Cytology smears Nonformalin fixed tissue Nonparaffin embedded tissue Noncharged slides ProbeOn slides	Reject
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Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
TECHONLY	Ambient (preferred)		
	Refrigerated		

Clinical and Interpretive

Clinical Information

A ubiquitously expressed mitochondrial enzyme fumarate hydratase (FH) catalyzes the reversible hydration of fumaric acid to yield L-malic acid during the Krebs cycle. Germline mutations in the *FH* gene cause a predisposition to renal defects like hereditary leiomyomatosis and renal cell carcinoma (HLRCC). While morphologic features are characteristic enough that they can be suspected, FH deficiency or germline DNA testing are necessary for its diagnosis. HLRCC can be associated with multiple cutaneous leiomyomas, uterine leiomyomas, and an aggressive variant of renal cell carcinoma (RCC) frequently occurring in young patients.

Reference Values

NA

Interpretation

The positive and negative controls are verified as showing appropriate immunoreactivity. If a control tissue is not included on the slide, a scanned image of the relevant quality control tissue is available upon request. Contact 855-516-8404.

Interpretation of this test should be performed in the context of the patient's clinical history and other diagnostic tests by a qualified pathologist.

Cautions

Age of a cut paraffin section can affect immunoreactivity. Stability thresholds vary widely among published literature and are antigen-dependent. Best practice is for paraffin sections to be cut fresh.

Clinical Reference

1. Carter CS, Skala SL, Chinnaiyan AM, et al: Immunohistochemical Characterization of Fumarate Hydratase (FH) and Succinate Dehydrogenase (SDH) in Cutaneous Leiomyomas for Detection of Familial Cancer Syndromes. Am J Surg Pathol 2017 Jun;41(6):801-809
2. Trpkov K, Hes O, Agaimy A, et al: Fumarate Hydratase-deficient Renal Cell Carcinoma Is Strongly Correlated With Fumarate Hydratase Mutation and Hereditary Leiomyomatosis and Renal Cell Carcinoma Syndrome. Am J Surg Pathol 2016 Jun;40(7):865-875
3. Harrison WJ, Andrici J, Maclean F, et al: Fumarate Hydratase-deficient Uterine Leiomyomas Occur in Both the Syndromic and Sporadic Settings. AM J Surg Pathol 2016 May;40(5):599-607
4. Llamas-Velasco M, Requena L, Kutzner H, et al: Fumarate hydratase immunohistochemical staining may help to identify patients with multiple cutaneous and uterine leiomyomatosis (MCUL) and hereditary leiomyomatosis and renal cell cancer (HLRCC) syndrome. J Cutan Pathol 2014 Nov;41(11):859-865

Performance**Method Description**

Immunohistochemistry on sections of paraffin-embedded tissue.(Unpublished Mayo method)

PDF Report

No

Day(s) and Time(s) Test Performed

Monday through Friday

Analytic Time

1 day

Maximum Laboratory Time

3 days

Specimen Retention Time

Until staining is complete

Performing Laboratory Location

Rochester

Fees and Codes**Fees**

- Authorized users can sign in to [Test Prices](#) for detailed fee information.
- Clients without access to Test Prices can contact [Customer Service](#) 24 hours a day, seven days a week.
- Prospective clients should contact their Regional Manager. For assistance, contact [Customer Service](#).

Test Classification

This test was developed and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the U.S. Food and Drug Administration.

CPT Code Information

88342-TC, primary

88341-TC, if additional IHC

LOINC® Information

Test ID	Test Order Name	Order LOINC Value
IHCFH	FH IHC, Tech Only	Order only;no result

Result ID	Test Result Name	Result LOINC Value
606335	FH IHC, Tech Only	Bill only; no result