

## Overview

### Useful For

Detection and titering of coagulation inhibitor to the specific factor requested, primarily factor IX in patients with hemophilia B

This test is **not useful** for the detection of a lupus-like circulating anticoagulant inhibitor, a nonspecific circulating anticoagulant, or other inhibitors that are not specific for coagulation factors.

### Profile Information

Test ID	Reporting Name	Available Separately	Always Performed
9INHT	FIX Inhib Profile Tech Interp	No	Yes
F_9	Coag Factor IX Assay, P	Yes	Yes

### Reflex Tests

Test ID	Reporting Name	Available Separately	Always Performed
9AINH	FIX Inhib Profile Prof Interp	No	No
9BETH	FIX Bethesda Units, P	No	No
F9_IS	Factor IX Inhib Scrn	No	No

### Testing Algorithm

Testing begins with coagulation factor IX activity assay with dilutions to evaluate assay inhibition; if the factor IX activity assay is normal or increased, then a technical interpretation will be provided.

If the factor IX activity assay is decreased, then an inhibitor screen will be performed at an additional charge to look for specific factor IX inhibition and a professional interpretation will be provided. If specific inhibition is apparent, the titer of the inhibitor will be determined.

### Special Instructions

- [Coagulation Guidelines for Specimen Handling and Processing](#)

### Method Name

Optical Clot-Based

### NY State Available

Yes

## Specimen

### Specimen Type

Plasma Na Cit

**Ordering Guidance**

This test is for factor IX inhibitors only. If the presence or type of inhibitor is unknown, order APROL / Prolonged Clot Time Profile, Plasma, except for when screening studies are needed for patients with known hemophilia A or B.

**Shipping Instructions**

Send all vials in the same shipping container.

**Necessary Information**

If priority specimen, mark request form, give reason, and request a call-back.

**Specimen Required****Patient Preparation:**

1. Patient must not be receiving Coumadin (warfarin) or heparin therapy
2. Fasting preferred

**Specimen Type:** Platelet-poor plasma

**Collection Container/Tube:** Light-blue top (3.2% sodium citrate)

**Submission Container/Tube:** 3 Plastic vials

**Specimen Volume:** 3 mL in 3 plastic vials, each containing 1 mL

**Collection Instructions:**

1. Specimen must be collected prior to factor replacement therapy.
2. For complete instructions, see [Coagulation Guidelines for Specimen Handling and Processing](#) in Special Instructions.
3. Centrifuge, transfer all plasma into a plastic vial, and centrifuge plasma again.
4. Aliquot plasma (1-2 mL per aliquot) into 3 separate plastic vials, leaving 0.25 mL in the bottom of centrifuged vial.
5. Freeze plasma immediately (no longer than 4 hours after collection) at -20 degrees C or, ideally, < or = -40 degrees C.

**Additional Information:**

1. Double-centrifuged specimen is critical for accurate results as platelet contamination may cause spurious results.
2. Each coagulation assay requested should have its own vial.

**Forms**

If not ordering electronically, complete, print, and send a [Coagulation Test Request](#) (T753) with the specimen.

**Specimen Minimum Volume**

2 mL plasma in 2 plastic vials, 1 mL each

**Reject Due To**

Gross hemolysis	Reject
Gross lipemia	Reject
Gross icterus	Reject

**Specimen Stability Information**

Specimen Type	Temperature	Time	Special Container
Plasma Na Cit	Frozen	14 days	

**Clinical and Interpretive**
**Clinical Information**

Factor IX inhibitors arise in patients with severe hemophilia B after factor IX transfusion. Patients with factor IX inhibitors may also develop anaphylactic reactions in response to factor IX infusions. Acquired factor IX inhibitors, occurring in previously healthy people, are exceedingly rare.

**Reference Values**

FACTOR IX ACTIVITY ASSAY

Adults: 65-140%

Normal, full-term newborn infants or healthy premature infants may have decreased levels (> or =20%), which may not reach adult levels for > or =180 days postnatal.\*

\*See Pediatric Hemostasis References section in [Coagulation Guidelines for Specimen Handling and Processing](#) in Special Instructions.

**Interpretation**

Normally, there is no inhibitor (ie, negative result).

If the screening assays indicate the presence of an inhibitor, it will be quantitated and reported in Bethesda (or equivalent) units.

**Cautions**

Occasionally, a potent lupus-like anticoagulant may cause false-positive testing for a specific factor inhibitor (eg, factor VIII or IX). See Ordering Guidance.

**Clinical Reference**

1. [Hoffman R, Benz Jr EJ, Silberstein LE, et al, eds: Hematology: Basic Principles and Practice. 7th ed. Elsevier; 2018](#)

2. Chitlur M, Warriar I, Rajpurkar M, et al: Inhibitors in factor IX deficiency a report of the ISTH-SSC international FIX inhibitor registry (1997-2006). *Haemophilia*. 2009;15(5):1027-1031. doi: 10.1111/j.1365-2516.2009.02039.x

## Performance

### Method Description

This assay consists of measuring the difference in factor IX activity (partial thromboplastin time assay) before and after incubation of a mixture of normal plasma and patient's plasma for 1 hour at 37 degrees C. For optimal sensitivity, the factor IX value of the normal plasma is adjusted to approximately 20%, because the factor IX assay is more sensitive in this area of the curve. In addition, an excess of patient's plasma will make the test more sensitive to small amounts of inhibitors. (Owen CA Jr, Bowie EJW, Thompson JH Jr: The Diagnosis of Bleeding Disorders. 2nd ed. Little, Brown, and Company:1975;143-145; Meijer P, Verbruggen HW, Spannagi M: Clotting factors and inhibitors: Assays and interpretation. In: Kottke-Marchant K, ed. Laboratory Hematology Practice. Wiley Blackwell Publishing; 2012:435-446)

If the inhibitor screen is positive for an inhibitor of factor IX, the inhibitor will be quantitated by the Bethesda assay. In the Bethesda procedure, inhibitors are quantified by mixing equal volumes of serially diluted plasma with normal plasma. This mixture is incubated 2 hours at 37 degrees C, and its factor IX activity is measured and compared to a control run at the same time. The difference between the factor IX activity of the patient's incubation mixture and that of the control is used to calculate the titer. The residual factor IX activity is converted to Bethesda units: 50% residual factor IX is equal to 1 Bethesda unit. Assays using the same basic principle as the Bethesda assay are used to quantitate the inhibitors of other coagulation factors. (Kasper CK, Aldedort LM, Counts RB, et al: A more uniform measurement of factor VIII inhibitors. Thromb Diath Haemorrh. 1975;34:869-872; Meijer P, Verbruggen HW, Spannagi M: Clotting factors and inhibitors: Assays and interpretation. In: Kottke-Marchant K, ed. Laboratory Hematology Practice. Wiley Blackwell Publishing; 2012:435-446)

### PDF Report

No

### Day(s) Performed

Monday through Friday

### Report Available

1 to 3 days

### Specimen Retention Time

7 days

### 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 has been modified from the manufacturer's instructions. Its performance characteristics were determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the US Food and Drug Administration.

**CPT Code Information**

85390-Factor IX Tech Interp

85250-Factor IX activity assay

85335-Bethesda titer (if appropriate)

85335-Factor IX inhibitor screen (if appropriate)

85390-Factor IX Professional Interp (if appropriate)

**LOINC® Information**

Test ID	Test Order Name	Order LOINC Value
9INHE	Factor IX Inhib Profile, P	96459-3

Result ID	Test Result Name	Result LOINC Value
9INHT	FIX Inhib Profile Tech Interp	69049-5
F_9	Coag Factor IX Assay, P	3187-2