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 detecting a nonspecific circulating anticoagulant or the presence of inhibitors directed against other clotting factors.

This test is not useful for the detection of lupus anticoagulants.

Profile Information

<table>
<thead>
<tr>
<th>Test ID</th>
<th>Reporting Name</th>
<th>Available Separately</th>
<th>Always Performed</th>
</tr>
</thead>
<tbody>
<tr>
<td>F_9</td>
<td>Coag Factor IX Assay, P</td>
<td>Yes</td>
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Reflex Tests

<table>
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<th>Test ID</th>
<th>Reporting Name</th>
<th>Available Separately</th>
<th>Always Performed</th>
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<tbody>
<tr>
<td>IBETH</td>
<td>Bethesda Units</td>
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<tr>
<td>F9_IS</td>
<td>Factor IX Inhib Scrn</td>
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<td>No</td>
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<tr>
<td>CCCR</td>
<td>Special Coagulation</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td></td>
<td>Interpretation</td>
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</table>

Testing Algorithm
Testing begins with coagulation factor IX activity assay with dilutions to evaluate assay inhibition; if the factor IX activity assay is decreased, an inhibitor will be performed to look for specific factor IX inhibition. If specific inhibition is apparent, it will be titered.

Special Instructions
- Coagulation Guidelines for Specimen Handling and Processing
- Coagulation Profile Comparison

Method Name
Optical Clot-Based

NY State Available
Yes

Specimen

Specimen Type
Plasma Na Cit
Advisory Information
This test is for factor IX inhibitors only. If the presence or type of inhibitor is unknown, either APROL / Prolonged Clot Time Profile, Plasma or ALUPP / Lupus Anticoagulant Profile, Plasma should be ordered first.

Multiple coagulation profile tests are available. See Coagulation Profile Comparison in Special Instructions for testing that is performed with each profile.

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
See Coagulation Guidelines for Specimen Handling and Processing in Special Instructions.

Patient Preparation: Fasting preferred

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

Submission Container/Tube: 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. Centrifuge, transfer all plasma into a plastic vial, and centrifuge plasma again.
3. Aliquot plasma into 3 separate plastic vials (1 mL in each) leaving 0.25 mL in the bottom of centrifuged vial.
4. Freeze plasma immediately (no longer than 4 hours after collection) at-20 degrees C, or ideally <=-40 degreesC.

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

<table>
<thead>
<tr>
<th>Gross hemolysis</th>
<th>Reject</th>
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</thead>
<tbody>
<tr>
<td>Gross lipemia</td>
<td>Reject</td>
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Test Definition: F9INH
Factor IX Inhib Profile

Specimen Stability Information

<table>
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<tr>
<th>Specimen Type</th>
<th>Temperature</th>
<th>Time</th>
<th>Special Container</th>
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<tbody>
<tr>
<td>Plasma Na Cit</td>
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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.

FACTOR IX INHIBITOR SCREEN

Negative

BETHESDA TITER

0 Units

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 Advisory Information.

Clinical Reference

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. Second edition. Little, Brown, and Company, Boston, MA, 1975, pp 143-145; Meijer P, Verbruggen and Spannagi M: Chapter 33: Clotting factors and inhibitors: Assays and Interpretation. In Laboratory Hematology Practice. Edited by K Kottke-Marchant. Wiley Blackwell Publishing, 2012, pp 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 and Spannagi M: Chapter 33: Clotting factors and inhibitors: Assays and Interpretation. In Laboratory Hematology Practice. Edited by K Kottke-Marchant. Wiley Blackwell Publishing, 2012, pp 435-446)

PDF Report
No

Day(s) and Time(s) Test Performed
Monday through Friday; Varies

Analytic Time
1 day

Maximum Laboratory Time
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 Definition: F9INH
Factor IX Inhib Profile

Test Classification
See Individual Test IDs

CPT Code Information
85250-Factor IX activity assay
85335-Bethesda titer (if appropriate)
85335-Factor IX inhibitor screen (if appropriate)
85390-26-Special coagulation interpretation (if appropriate)

LOINC® Information

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<td>F9INH</td>
<td>Factor IX Inhib Profile</td>
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<table>
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