Overview

Useful For
Screening for possible disorders of mitochondrial metabolism, when used in conjunction with blood lactate collected at the same time to determine the lactate-to-pyruvate ratio

Genetics Test Information
The lactate:pyruvate (L:P) ratio is considered a helpful (not diagnostic) tool in the evaluation of patients with possible disorders of mitochondrial metabolism, especially in patients with neurologic dysfunction and normal blood L:P ratios. Pyruvic acid levels alone have little clinical utility.

Testing Algorithm
See Epilepsy: Unexplained Refractory and/or Familial Testing Algorithm in Special Instruction.

Special Instructions
- Biochemical Genetics Patient Information
- Epilepsy: Unexplained Refractory and/or Familial Testing Algorithm

Method Name
Spectrophotometry (SP)

NY State Available
Yes

Specimen

Specimen Type
Whole blood

Additional Testing Requirements
This test does not calculate the lactate:pyruvate ratio. To obtain this information, LACS1 / Lactate, Plasma and PYR / Pyruvic Acid, Blood must be ordered. The ratio can be calculated from the results of those tests.

Specimen Required
Call 800-533-1710 or 507-266-5700 to order special collection tube.

Patient Preparation: Fasting (at least 4 hours)

Supplies: Perchloric Acid–Pyruvate Tube (T012)

Container/Tube: Special collection tube containing 2.5 mL of 6% perchloric acid (T012)

Specimen Volume: Exactly 1 mL

Collection Instructions:
1. Special collection tube must be prechilled prior to draw.
2. Draw enough blood directly into syringe to add exactly 1 mL of blood to the prechilled special collection tube.
3. Once drawn, immediately transfer blood to the prechilled, special collection tube and shake vigorously to mix.

Additional Information:

1. **Check expiration date before using.** Supplied collection tube expires 12 months after preparation.

2. If perchloric acid spills, obtain new, prechilled tube.

**Forms**

1. [Biochemical Genetics Patient Information](#) (T602) in Special Instructions.

2. If not ordering electronically, complete, print, and send an [Inborn Errors of Metabolism Test Request](#) (T798) with the specimen.

**Specimen Minimum Volume**

See Specimen Required

**Reject Due To**

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<table>
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<tbody>
<tr>
<td>Hemolysis</td>
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<tr>
<td>Lipemia</td>
<td>Mild OK; Gross OK</td>
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<tr>
<td>Icterus</td>
<td>NA</td>
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<tr>
<td>Other</td>
<td>Use of any anticoagulant other than special tube</td>
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**Specimen Stability Information**

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<tr>
<th>Specimen Type</th>
<th>Temperature</th>
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<tr>
<td>Whole blood</td>
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**Clinical and Interpretive**

**Clinical Information**

Pyruvic acid, an intermediate metabolite, plays an important role in linking carbohydrate and amino acid metabolism to the tricarboxylic acid cycle, the fatty acid beta-oxidation pathway, and the mitochondrial respiratory chain complex. Though isolated elevated pyruvate is not diagnostic of any inborn error of metabolism, analysis with lactate may suggest an inborn error of metabolism as some present with lactic acidosis or a high lactate-to-pyruvate (L:P) ratio.

The L:P ratio is elevated in several, but not all, mitochondrial respiratory chain disorders. Mitochondrial disorders vary widely in presentation and age of onset. Many mitochondrial disorders have neurologic and myopathic features and may involve multiple organ systems. Determination of lactate, pyruvate, and L:P ratio in cerebrospinal fluid is helpful in directing attention toward a possible mitochondrial disorder in cases with predominantly neurologic dysfunction and normal blood lactate levels, though further confirmatory testing will be required to establish a diagnosis.

A low L:P ratio is observed in inherited disorders of pyruvate metabolism including pyruvate dehydrogenase complex (PDHC) deficiency. Clinical presentation of PDHC deficiency can range from fatal congenital lactic acidosis to relatively mild ataxia or neuropathy. The most common features in infants and children with PDHC deficiency are...
delayed development and hypotonia. Seizures and ataxia are also frequent features. Other manifestations can include congenital brain malformations, degenerative changes including Leigh disease, and facial dysmorphism.

**Reference Values**

0.08-0.16 mmol/L

NIH Unit

0.7-1.4 mg/dL

**Interpretation**

An elevated lactate-to-pyruvate (L:P) ratio may indicate inherited disorders of the respiratory chain complex, tricarboxylic acid cycle disorders and pyruvate carboxylase deficiency. Respiratory chain defects usually result in L:P ratios above 20.

A low L:P ratio (disproportionately elevated pyruvic acid) may indicate an inherited disorder of pyruvate metabolism. Defects of the pyruvate dehydrogenase complex result in L:P ratios below 10.

The L:P ratio is characteristically normal in other patients. An artificially high ratio can be found if the patient is acutely ill.

Cerebrospinal fluid (CSF) L:P ratio may assist in evaluation of patients with neurologic dysfunction and normal blood L:P ratios. Blood and CSF specimens should be collected at the same time.

**Cautions**

Correct specimen collection and handling is crucial to achieve reliable results.

Pyruvic acid levels alone have little clinical utility. Abnormal concentrations of pyruvic acid, and lactate-to-pyruvate (L:P) ratios, are not diagnostic for a particular disorder but must be interpreted in the context of the patient’s clinical presentation and other laboratory studies. The determination of pyruvic acid is of diagnostic value when lactic acid is measured and the L:P ratio is established in the same specimen.

When comparing blood and cerebrospinal fluid (CSF) L:P ratios, blood and CSF specimens should be collected at the same time.

**Clinical Reference**


**Method Description**

Pyruvate, in the presence of excess NADH, H+, and lactic dehydrogenase, is reduced to lactate. The reaction is stoichiometric; the decrease in absorbance at 340 nm is directly proportional to the concentration of pyruvate. (Standard Methods of Clinical Chemistry, 1979;6:245-259; Huckabee WE: Relationships of pyruvate and

PDF Report
No

Day(s) and Time(s) Test Performed
Tuesday, Friday; Varies

Analytic Time
6 days

Maximum Laboratory Time
8 days

Specimen Retention Time
3 weeks

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
84210

LOINC® Information

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