

Overview

Useful For

Diagnosing and monitoring patients with lactic acidosis

Monitoring lactate-to-pyruvate ratios

Testing Algorithm

For more information see [Epilepsy: Unexplained Refractory and/or Familial Testing Algorithm](#)

Special Instructions

- [Epilepsy: Unexplained Refractory and/or Familial Testing Algorithm](#)

Highlights

This test provides results for both lactate and pyruvate on a single collection

Method Name

Gas Chromatography Mass Spectrometry (GC-MS)

NY State Available

Yes

Specimen

Specimen Type

Plasma

Ordering Guidance

This test does not measure D-lactate, an uncommon, often undiagnosed cause of lactic acidosis. If D-lactate testing is needed, order DLAU / D-Lactate, Urine (preferred) or DLAC / D-Lactate, Plasma.

Analytes from this test are included in test MMPP / Mitochondrial Metabolites, Plasma. If ordered together, this test may be canceled.

Specimen Required

Collection Container/Tube:

Preferred: Green top (Sodium heparin)

Acceptable: Green top (Lithium heparin)

Submission Container/Tube: Plastic vial

Specimen Volume: 0.5 mL

Collection Instructions: Centrifuge and aliquot plasma into a plastic vial.

Forms

If not ordering electronically, complete, print, and send a [Biochemical Genetics Test Request](#) (T798) with the specimen.

Specimen Minimum Volume

0.1 mL

Reject Due To

| | |
|-----------------|--------|
| Gross hemolysis | Reject |
| Gross lipemia | OK |
| Gross icterus | OK |

Specimen Stability Information

| Specimen Type | Temperature | Time | Special Container |
|---------------|--------------------|---------|-------------------|
| Plasma | Frozen (preferred) | 90 days | |
| | Ambient | 7 days | |
| | Refrigerated | 7 days | |

Clinical & Interpretive

Clinical Information

Lactic acid (lactate) is primarily produced from glucose metabolism via the glycolytic pathway. Although primarily metabolized by the liver, other tissues also use small amounts of lactate. Typically, the amount of lactate produced parallels the amount utilized. Both the rates of lactate production and liver clearance impact the lactate concentration in blood. Lactic acidosis, or the accumulation of excess lactate, results from a combination of increased lactate production with decreased utilization.

Patients experiencing lactic acidosis present with tachypnea, weakness, and fatigue. Untreated, patients may develop confusion and progress to coma. Lactic acidosis may be associated with hypoxic conditions (eg, shock, hypovolemia, heart failure, pulmonary insufficiency), metabolic disorders (eg, diabetic ketoacidosis, malignancies, inborn errors of metabolism), and toxin exposures (eg, ethanol, methanol, salicylates).

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. However, pyruvic acid levels alone have little clinical utility.

Combined analysis of lactate and pyruvate may suggest an inborn error of metabolism when elevations of both analytes are observed or when there is an abnormal lactate-to-pyruvate (L:P) ratio. For example, several mitochondrial respiratory chain disorders exhibit elevated L:P ratios. Mitochondrial disorders vary widely in both clinical presentation and age of onset. Patients commonly present with neurologic and myopathic features. In addition, patients may experience involvement of multiple organ systems with features such as myopathy, ophthalmoplegia, ptosis,

cardiomyopathy, sensorineural hearing loss, optic atrophy, pigmentary retinopathy, diabetes mellitus, encephalomyopathy, seizures, and stroke-like episodes.

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 observed in infants and children with PDHC deficiency are developmental delay, hypotonia, seizures, and ataxia. Other manifestations may include congenital brain malformations, degenerative changes including Leigh disease, and facial dysmorphism.

Reference Values

Lactic Acid

< or = 4000.0 nmol/mL

Pyruvic Acid

< or = 350.0 nmol/mL

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 artifactually 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

No significant cautionary statements

Clinical Reference

1. Munnich A, Rotig A, Cormier-Daire V, Rustin P. Clinical presentation of respiratory chain deficiency. In: Valle DL, Antonarakis S, Ballabio A, Beaudet AL, Mitchell GA, eds. The Online Metabolic and Molecular Bases of Inherited Disease. McGraw-Hill; 2019. Accessed July 5, 2024. Available at <http://ommbid.mhmedical.com/content.aspx?bookid=2709§ionid=225086827>
2. Robinson BH. Lactic acidemia: Disorders of pyruvate carboxylase and pyruvate dehydrogenase. In: Valle DL, Antonarakis S, Ballabio A, Beaudet AL, Mitchell GA, eds. The Online Metabolic and Molecular Bases of Inherited Disease. McGraw-Hill; 2019. Accessed July 5, 2024. Available at <http://ommbid.mhmedical.com/content.aspx?bookid=2709§ionid=225087140>
3. Shoffner JM. Oxidative phosphorylation diseases. In: Valle DL, Antonarakis S, Ballabio A, Beaudet AL, Mitchell GA, eds. Online Metabolic and Molecular Bases of Inherited Disease. McGraw-Hill; 2019 Accessed July 5, 2024. Available at <http://ommbid.mhmedical.com/content.aspx?bookid=2709§ionid=225088339>

Performance

Method Description

A mixture of labeled internal standards is added to patient plasma following oximation of keto acids. The samples are acidified and extracted. After evaporation, the dry residue is silylated analyzed by capillary gas chromatography mass spectrometry using selected ion monitoring with positive electron impact ionization and stable isotope dilution.(Unpublished Mayo method)

PDF Report

No

Day(s) Performed

Wednesday

Report Available

3 to 9 days

Specimen Retention Time

2 months

Performing Laboratory Location

Mayo Clinic Laboratories - Rochester Main Campus

Fees & 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 account representative. 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. It has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information

83605

84210

LOINC® Information

| Test ID | Test Order Name | Order LOINC® Value |
|---------|---------------------------|--------------------|
| LAPYP | Lactate Pyruvate Panel, P | 101656-7 |

| Result ID | Test Result Name | Result LOINC® Value |
|-----------|------------------|---------------------|
| 616797 | Interpretation | 59462-2 |
| 616794 | Lactic acid | 2524-7 |
| 616795 | Pyruvic acid | 32338-6 |
| 616796 | Reviewed by | 18771-6 |