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**Lab Dept:** Chemistry

**Test Name:** BIOTINIDASE

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***General Information***

**Lab Order Codes:** BIT

**Synonyms:** Biotin-amide amidohydrolase; Biotinidase deficiency; Biotinidase, serum

**CPT Codes:** 82261 – Biotinidase, each specimen

**Test Includes:** Biotinidase level reported in U/L.

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***Logistics***

**Test Indications:** Useful for screening for biotinidase deficiency (preferred method) and for follow-up testing for certain organic acidurias.

**Lab Testing Sections:** Chemistry - Sendouts

**Referred to:** Mayo Clinic Laboratories (MML Test: BIOTS)

**Phone Numbers:** MIN Lab: 612-813-6280

STP Lab: 651-220-6550

**Test Availability:** Daily, 24 hours

**Turnaround Time:** 4 - 8 days, test set up Monday and Thursday

**Special Instructions:** N/A

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***Specimen***

**Specimen Type:** Blood

**Container:** SST (Gold, marble or red) tube

**Draw Volume:** 3 mL (Minimum: 1.5 mL) blood

**Processed Volume:** 1 mL (Minimum: 0.5 mL) serum

**Collection:** Routine venipuncture

**Special Processing:** Lab Staff: Centrifuge specimen, remove serum aliquot immediately into a plastic screw-capped vial. Store and ship at frozen temperatures. Forward promptly.

Specimen stable frozen (preferred) for 21 days, refrigerated for 5 days.

**Patient Preparation:** None

**Sample Rejection:** Mislabeled or unlabeled specimens; gross hemolysis

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### ***Interpretive***

**Reference Range:** 3.5 - 13.8 U/L

Partial deficiencies and carriers may occur at the low end of the reference range. Suggest maintenance on a biotin supplementation and retesting in 1 month.

Values <3.5 U/L are occasionally seen in specimens from unaffected patients.

**Critical Values:** N/A

**Limitations:** A diet high in biotin may result in normal clinical presentation even when the biotinidase level is low.

Assay interference may occur for specimens collected when the patient is being treated with a sulfa drug.

**Methodology:** Colorimetric

**References:** [Mayo Clinic Laboratories](#) (January 2024)

**Updates:**  
7/13/2006: Specimen type change, previously listed as frozen serum.  
6/17/2012: Specimen type change, previously listed as whole blood.  
11/8/2017: Collection container update  
12/5/2019: Draw volume update  
1/24/2024: Added specimen stability and additional limitation/interference.