



**NEONATAL Biotinidase**

**Newborn screening of Biotinidase Deficiency**

- Enzymatic determination of biotinidase activity from blood specimens dried on filter paper
- Fluorescence detection for high sensitivity
- No centrifuge or filtration step
- Compatible with Labsystems Diagnostics NS2400 automation system

## About

Biotinidase deficiency, the major cause of late-onset multiple carboxylase deficiency, is an autosomal recessively inherited disorder. It is caused by absent or deficient activity of biotinidase, an enzyme releasing biotin (also known as vitamin B7 or H) from dietary proteins. The disorder can lead to a decrease in biotin availability and if untreated, it is associated with the neurologic and cutaneous consequences.

Newborn screening of Biotinidase activity from dried blood spots can identify affected patients shortly after the birth. Screening is done with a direct enzyme assay, where according to literature dietary protein or biotin intake is not influencing. The Labsystems Diagnostics' Neonatal Biotinidase kit is based on monitoring of fluorescent signal of the 6-Aminoquinoline (6-AQ), product formed during the reaction as a result of the biotinidase activity.

## Assay procedure:

### STEP I

Punch out 3.2 mm disks from blood calibrators, controls and patient samples into the microplate wells.

### STEP II

Add reconstituted substrate solution and cover the plate.

### STEP III

Incubate 4h with heating.

### STEP IV

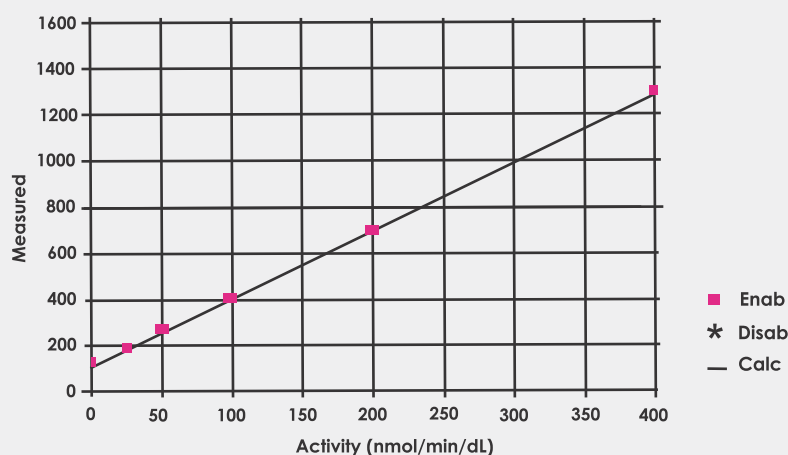
Add EtOH to each well immediately after the incubation.

### STEP V

Incubate the plate for 30 min at room temperature.

Measure immediately fluorescence at ex. 355, em. 460 nm

## Typical calibration curve:



## Product Info:

| Cat.no. | Product name         | Plate type     | Packing size | Regulatory status |
|---------|----------------------|----------------|--------------|-------------------|
| 6199821 | Neonatal Biotinidase | 96 wells Solid | 960 wells    | CE marked         |

Type 903 filter paper is used in all products.

### Labsystems Diagnostics Oy

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