



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 NS2400/1200 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. According to the literature, because the screening is done with a direct enzyme assay, it is not influenced by dietary protein or biotin intake. 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 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

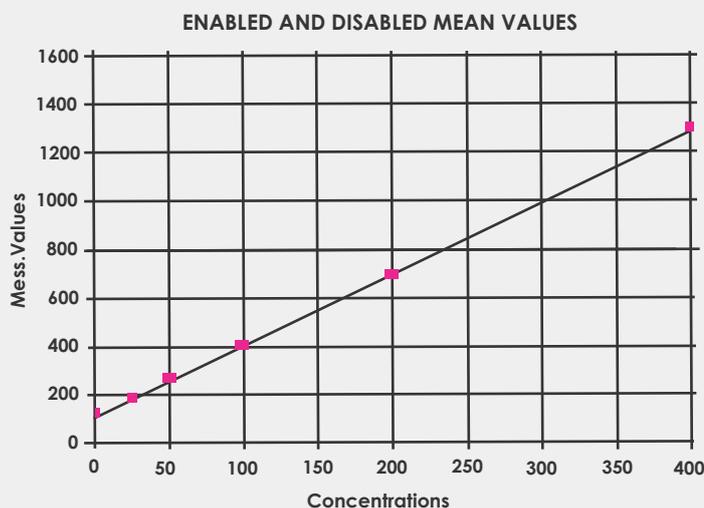
Add EtOH to each well immediately after the incubation.

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:

- 6199 820, Grade 903, 5 strip plates (12x8), 480 wells
- 6199 821, Grade 903, 10 solid plates (1x96), 960 wells
- 6199 822, Grade 903, 10 strip plates (12x8), 960 wells

Labsystems Diagnostics Oy

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