The incidence of biotinidase deficiency has been reported as follows: profound biotinidase deficiency (<10% activity) 1 in 112,000, partial deficiency (10%–30% activity) 1 in 129,000, and profound and partial deficiency together 1 in 60,000\(^1\). Symptoms include seizure and possible skin disorders, followed by developmental delays, speech problems and possible vision and hearing difficulties.

The GSP Neonatal Biotinidase assay is intended for the quantitative determination of biotinidase activity using dried blood spot specimens.

Screening for biotinidase deficiency with GSP Neonatal Biotinidase assay

- Improved performance due to fully automated plate processing and optimized assay concept
- Combination of enzymatic and immunoassay
- Ready to use reagents, no reconstitution required
- No ethanol precipitation in the assay
- 14 days on-board reagent stability
- Calibrator and control medium special processing to reduce lot-to-lot variation
- New unit in use; U/\(\text{dL}\) in blood
GSP Neonatal Biotinidase assay

The GSP Neonatal Biotinidase assay combines an enzyme reaction with a solid phase time-resolved immunofluorescence assay. The enzyme reaction is the cleavage by biotinidase of the amide bond in Eu-labeled biotin. The enzyme reaction is stopped by addition of streptavidin which has high affinity for biotin (either Eu-labeled or free biotin). The streptavidin-biotin complexes are captured by the solid phase monoclonal antibody directed against streptavidin. DELFIA® Inducer dissociates the molecules into the solution where the europium fluorescence is measured. The measured fluorescence is inversely proportional to the biotinidase activity of the sample. Assay processing time is 4h15min.

For use with the GSP® instrument

GSP is the new generation automated neonatal screening instrument from PerkinElmer. It has multi-technology capability, which means that both DELFIA® and prompt fluorescence assays can be run on a common platform. Its versatility and speed help it to accommodate present and future screening needs.

ORDERING INFORMATION

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References


This brochure is intended for distribution in the USA and describes products offered for sale in this country. For information on the availability of PerkinElmer neonatal screening products in other countries, please contact your local representative.