

Overview of methods for determination of cystine and protein in white blood cells

This document summarizes the most common methods used by clinical laboratories to measure the cystine and protein content in granulocytes and mixed leucocytes for the diagnosis of cystinosis and monitoring of cysteamine therapy. Detail protocols are not included in this document but can be found in the literature references provided.

Quantification of cystine

The majority of clinical laboratories use liquid chromatography with different detection systems rather than the radioactive cystine binding protein assay. Methods using liquid chromatography with mass spectrometry detection are more specific and have lower limits of quantification.

Measurement of cystine can be performed in granulocytes or mixed leucocytes and while the use of granulocytes increases the detection of cystinosis and the discrimination between healthy controls and obligate heterozygotes, the methods require a greater volume of blood cells and a derivatization step to improve the limit of quantification.

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Quantification of protein

The clinical laboratories use spectrophotometric analytical procedures to measure the concentration of protein in white cells. The majority of methods are based on modified assays of the original methods reported more than four decades ago such as the Lowry, Bradford and BCA methods; however some laboratories may be using methods, from their routine clinical chemistry laboratories, validated to measure the protein content in cerebrospinal fluid and urine.

Bibliography for the determination of protein in white cells:

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