

CYSTINURIA MANAGEMENT PROGRAM

Interpreting test results

The free, no cost to patients, **Cystinuria Management Program** test is designed to help you proactively monitor your patients' urinary cystine levels to determine their risks of forming stones.

As part of the program, you will receive detailed and accurate results from your patients' 24-hour cystine urine tests. Cystinuria Management Program test results provide valuable information so you can make optimal management decisions for your patients with cystinuria.

Urinary Cystine Concentration

Cystine concentration is calculated by dividing the total daily urinary cystine by the total daily urine volume. The management goal is to reduce urinary cystine concentration to below the solubility limit, generally <250 mg/L at a pH of 7.0.1,2 When urinary cystine levels are above

the solubility limit, stones are more likely to form.¹

Reducing cystine concentration involves multiple adjustments including maintaining high fluid intake, a low-sodium and low-animal-protein diet, increased urine pH, and taking cystine-binding thiol medications.¹⁻⁴

Urinary pH

The solubility of cystine is dependent on urinary pH. Patients should aim to maintain a urinary pH of 7.0 as part of a conservative treatment plan.²

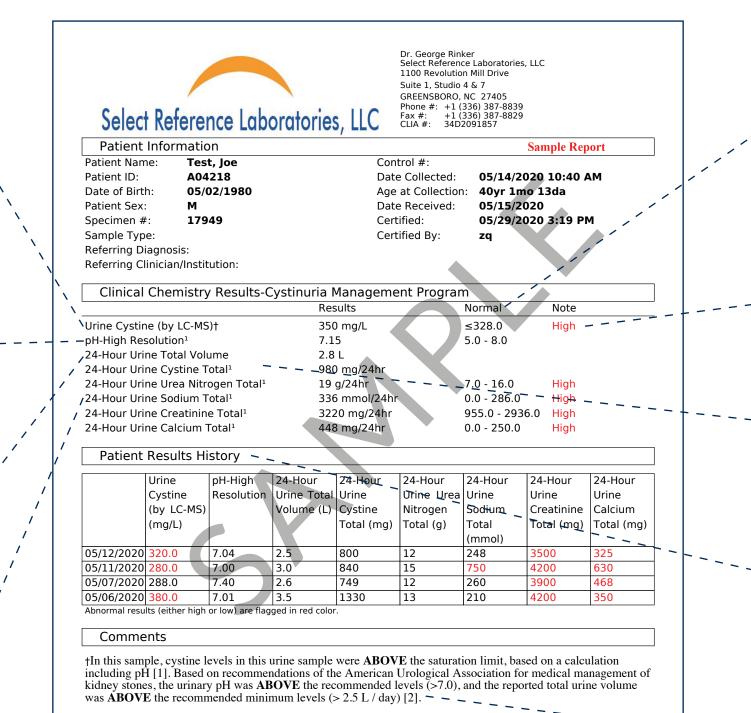
Reported Urine Volume

Targeted urine volume is typically higher for patients with cystinuria compared with other stone formers in order to reduce urinary cystine below the solubility limit. A minimum urine output of 2.5 L/day on a consistent basis is recommended.²

Urinary Sodium

A high urine sodium level might indicate a diet high in sodium. Patients should follow a diet low in sodium, as low sodium intake has been shown to reduce cystine excretion.²

RESULTS FROM THE CYSTINURIA MANAGEMENT PROGRAM TEST CAN HELP INFORM HOW TO ADJUST AND OPTIMIZE YOUR PATIENTS' MANAGEMENT PLANS



1. Dent and Senior, Br J Urol. 1955; 27(4):317-32. 2. Pearle et al., | Urol. 2014; 192(2):316-24.

¹ This test was developed and its performance characteristics determined by Select Reference Laboratory. It has not been cleared or approved by the U.S. Food and Drug Administration.

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Reference Range

The reference range is provided as a quick and easy way to determine if the patient is on track. Note that the reference range for cystine concentration will be adjusted based on the patient's pH.

Flag

Flags are included to identify and draw attention to analytes that are outside the reference ranges provided.

Additional Analytes

Additional analytes listed are provided to help guide treatment decisions.

Previous Lab Result

Results of any previous CMP cystine urine collection will be provided to compare over time

Interpretation

The interpretation section provides additional information and context for the physician based on the patient's test results. These results can help indicate if the patient is at risk of stone development.

ENGAGE YOUR PATIENTS BY SHARING AND EXPLAINING THEIR RESULTS

Review **Cystinuria Management Program** test results with your patients so they can take an active role in managing their cystinuria. You can use this tool as a guide for what topics to cover when reviewing test results with them.

The more knowledgeable your patients are about what causes their urinary cystine levels to become elevated, the more empowered they will be to better manage and take control of their cystinuria.

Make ongoing urinary cystine monitoring a regular part of your patients' management plans

The Cystinuria Management Program is an essential tool to help routinely monitor your patients' treatment plans and inform your cystinuria management decisions.

For more information, contact your Travere Therapeutics representative.

References: 1. Pareek G, Steele TH, Nakada SY. Urological intervention in patients with cystinuria is decreased with medical compliance. *J Urol.* 2005;174(6):2250-2252. doi:10.1097/01.ju.0000181817.89703.66. **2.** Pearle MS, Goldfarb DS, Assimos DG, et al. Medical management of kidney stones: AUA guideline. *J Urol.* 2014;192(2):316-324. doi:10.1016/j.juro.2014.05.006. **3.** Knoll T, Zöllner A, Wendt-Nordahl G, Michel MS, Alken P. Cystinuria in childhood and adolescence: recommendations for diagnosis, treatment, and follow-up. *Pediatr Nephrol.* 2005;20(1):19-24. doi:10.1007/s00467-004-1663-1. **4.** Data on file. Travere Therapeutics, Inc.