



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.

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

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



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Patient Information		Sample Report	
Patient Name:	Test, Joe	Control #:	
Patient ID:	A04218	Date Collected:	05/14/2020 10:40 AM
Date of Birth:	05/02/1980	Age at Collection:	40yr 1mo 13da
Patient Sex:	M	Date Received:	05/15/2020
Specimen #:	17949	Certified:	05/29/2020 3:19 PM
Sample Type:		Certified By:	zq
Referring Diagnosis:			
Referring Clinician/Institution:			

Clinical Chemistry Results-Cystinuria Management Program			
	Results	Normal	Note
Urine Cystine (by LC-MS)†	350 mg/L	≤328.0	High
pH-High Resolution ¹	7.15	5.0 - 8.0	
24-Hour Urine Total Volume	2.8 L		
24-Hour Urine Cystine Total ¹	980 mg/24hr		
24-Hour Urine Urea Nitrogen Total ¹	19 g/24hr	7.0 - 16.0	High
24-Hour Urine Sodium Total ¹	336 mmol/24hr	0.0 - 286.0	High
24-Hour Urine Creatinine Total ¹	3220 mg/24hr	955.0 - 2936.0	High
24-Hour Urine Calcium Total ¹	448 mg/24hr	0.0 - 250.0	High

Patient Results History								
	Urine Cystine (by LC-MS) (mg/L)	pH-High Resolution	24-Hour Urine Total Volume (L)	24-Hour Urine Cystine Total (mg)	24-Hour Urine Urea Nitrogen Total (g)	24-Hour Urine Sodium Total (mmol)	24-Hour Urine Creatinine Total (mg)	24-Hour Urine Calcium Total (mg)
05/12/2020	320.0	7.04	2.5	800	12	248	3500	325
05/11/2020	280.0	7.00	3.0	840	15	750	4200	630
05/07/2020	288.0	7.40	2.6	749	12	260	3900	468
05/06/2020	380.0	7.01	3.5	1330	13	210	4200	350

Abnormal results (either high or low) are flagged in red color.

Comments
<p>†In this sample, cystine levels in this urine sample were ABOVE the saturation limit, based on a calculation including pH [1]. Based on recommendations of the American Urological Association for medical management of kidney stones, the urinary pH was ABOVE the recommended levels (>7.0), and the reported total urine volume was ABOVE the recommended minimum levels (> 2.5 L / day) [2].</p> <p>1. Dent and Senior, Br J Urol. 1955; 27(4):317-32. 2. Pearle et al., J Urol. 2014; 192(2):316-24.</p> <p><small>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.</small></p>

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 Retrophin 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. Retrophin, Inc.