

Test Definition: HEX4

Glucotetrasaccharides, Random, Urine

Reporting Title: Glucotetrasaccharides, U

Performing Location: Rochester

Ordering Guidance:

Additional Testing Requirements:

When requested for the diagnosis of Pompe disease (glycogen storage disorder type II), urine glucotetrasaccharide concentrations need to be interpreted in light of the clinical presentation and other laboratory test results, such as blood creatine kinase, alpha-glucosidase (GAA) activity, and GAA genotype.

Necessary Information:

- 1. Patient's age is required.
- 2. Reason for testing is required.

Specimen Requirements:

Supplies: Sarstedt Aliquot Tube, 5 mL (T914) **Container/Tube:** Plastic, 5-mL urine tube

Specimen Volume: 3 mL **Collection Instructions:**

- 1. Collect a random urine specimen.
- 2. No preservative.

Forms:

If not ordering electronically, complete, print, and send a Biochemical Genetics Test Request (T798) with the specimen.

Specimen Type	Temperature	Time	Special Container
Urine	Frozen (preferred)	87 days	
	Refrigerated	28 days	
	Ambient	14 days	

Ask at Order Entry (AOE) Questions:

Test ID	Question ID	Description	Туре	Reportable
HEX4	BG710	Reason for Referral:	Answer List	Yes
		Rule out Pompe disease		
		Rule out GSDs other than Pompe disease		
		Follow-up of abnormal newborn screening		
		Follow-up of known Pompe patient		
		Not provided		

Result Codes:

Result ID	Reporting Name	Туре	Unit	LOINC®
64174	Glucotetrasaccharides, U	Numeric	mmol/mol Cr	53868-6



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BG710	Reason for Referral	Alphanumeric	42349-1
BA2896	Intepretation (HEX4)	Alphanumeric	59462-2
BA2897	Reviewed By	Alphanumeric	18771-6

LOINC® and CPT codes are provided by the performing laboratory.

Supplemental Report:

No

CPT Code Information:

82542 82570

Reference Values:

< or =14 months: < or =14.9 mmol/mol Cr > or =15 months: < or =4.0 mmol/mol Cr