Test Definition: CHOU

Carbohydrate, Urine

Overview

Useful For

Screening for conditions associated with increased excretion of carbohydrates, including inborn errors of fructose and galactose metabolism.

This test is **not recommended** as a follow up test for abnormal newborn screening for galactosemia.

Testing Algorithm

Qualitative testing for the presence of reducing substances is performed followed by quantitative analysis of carbohydrates including fructose, galactose, xylose, glucose, sucrose, lactose, maltose, and raffinose.

Special Instructions

- Biochemical Genetics Patient Information

Method Name

Gas Chromatography Mass Spectrometry (GC-MS)

NY State Available

Yes

Specimen

Specimen Type

Urine

Ordering Guidance

This test is a quantitative analysis of carbohydrate excretion in the urine. It is **not appropriate** for evaluation of an abnormal newborn screen for galactosemia. For those cases, order GCT / Galactosemia Reflex, Blood and consider GAL1P / Galactose-1-Phosphate, Erythrocytes and GATOL / Galactitol, Quantitative, Urine.

Necessary Information

Patient's age is required.

Specimen Required

Supplies: Urine Tubes, 10 mL (T068)
Container/Tube: Plastic, 10-mL urine tube
Specimen Volume: 5 mL
Collection Instructions:
1. Collect an early-morning (preferred) random urine specimen.
2. No preservative.

Forms

Biochemical Genetics Patient Information (T602)

Specimen Minimum Volume

1 mL

Reject Due To

Urine containing preservatives	Reject

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Urine	Frozen	42 days	

Clinical & Interpretive

Clinical Information

Carbohydrates are a group of mono-, di-, and oligosaccharides of endogenous and exogenous sources. Their presence frequently reflects dietary consumption but can indicate specific pathology if either a particular saccharide or a particular excretory pattern is present. Most saccharides (except glucose) have low renal thresholds and are readily excreted in the urine.

The identification and quantitation of carbohydrates, in particular galactose and fructose, is useful to screen for inborn errors of galactose and fructose metabolism such as galactosemia and hereditary fructose intolerance. Additionally, xylose may also be detected in individuals with hereditary pentosuria, a benign trait with high frequency among individuals with Ashkenazi Jewish ancestry.

This test is useful as an initial screen. To establish any diagnosis, abnormal results require confirmation by enzyme assay, molecular genetic analysis, or correlation with other laboratory testing.

Reference Values

Reducing Substances: Negative

Quantitative results are reported as mmol/mol creatinine.

Age range	<12 months	12 months-18 years	> or =19 years
Xylose	< or =13	< or =38	< or =9
Fructose	< or =32	< or =31	< or =16
Galactose	< or =117	< or =32	< or =5
Glucose	< or =139	< or =15	< or =22
Sucrose	< or =27	< or =46	< or =20

Lactose	< or =160	< or =18	< or =5
Maltose	< or =5	< or =1	< or =2
Raffinose	< or =1	< or =1	< or =1

Interpretation

When abnormal results are detected, a detailed interpretation is given, including an overview of the results and of their significance, a correlation to available clinical information, elements of differential diagnosis, recommendations, if any, for additional biochemical testing, and a phone number to reach one of the laboratory directors in case the referring physician has additional questions.

Cautions

Ingestion of sorbose, a naturally occurring ketohexose,(1) will interfere with the quantitation of fructose and an interference comment will be included in the report. Retesting is recommended in these cases.

In some instances, normal carbohydrate values for infants (<12 months) and pediatric patients can result in a positive result for reducing substances. This is due to higher normal excretion of carbohydrates in younger age groups.

Clinical Reference

1. Hastings J, Owen G, Dekker A, et al. ChEBI in 2016: Improved services and an expanding collection of metabolites. Nucleic Acids Res. 2016;44(D1):D1214-D1219. doi:10.1093/nar/gkv1031
2. Steinmann B, Gitzelmann R, Van den Berghe G. Disorders of fructose metabolism. In: Valle DL, Antonarakis S, Ballabio A, Beaudet AL, Mitchell GA. eds. The Online Metabolic and Molecular Bases of Inherited Disease. McGraw-Hill Education; 2019. Accessed February 21, 2025. https://ommbid.mhmedical.com/content.aspx?bookid=2709&sectionid=225080452
3. OMIM. #260800 Pentosuria; PNTSU. Johns Hopkins University; 1986. Updated July 9, 2016. Accessed February 21, 2025. Available at https://omim.org/entry/260800
4. Gaughan S, Ayres L, Baker P II. Hereditary fructose intolerance. In: Adam MP, Feldman J, Mirzaa GM, et al, eds. GeneReviews [Internet]. University of Washington, Seattle; 2015. Updated February 18, 2021. Accessed February 21, 2025. Available at www.ncbi.nlm.nih.gov/books/NBK333439

Performance

Method Description

The urine is evaluated for reducing substances using the AimTab tablet (Benedict's test). The tablet is a standardized self-heating method for the determination of the reducing substances by copper reduction.

Urine is spiked with a mixture of labeled internal standards and evaporated. The dry residue is oximated, derivatized, then extracted. Specimens are analyzed by gas chromatography mass spectrometry, selected ion monitoring using positive ammonia chemical ionization and stable isotope dilution.(Unpublished Mayo method)

PDF Report

No

Day(s) Performed

Tuesday

Report Available
3 to 9 days

Specimen Retention Time
2 months

Performing Laboratory Location
Mayo Clinic Laboratories - Rochester Main Campus

Fees & Codes

Fees
- Authorized users can sign in to Test Prices for detailed fee information.
- Clients without access to Test Prices can contact Customer Service 24 hours a day, seven days a week.
- Prospective clients should contact their account representative. For assistance, contact Customer Service.

Test Classification
This test was developed and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. It has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information
84379

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
CHOU	Carbohydrate, U	74447-4

Result ID	Test Result Name	Result LOINC® Value
622709	Reducing Substances	5809-9
622710	Xylose	75051-3
622711	Fructose	34309-5
622712	Galactose	25102-5
622713	Glucose	34312-9
622714	Sucrose	76132-0
622715	Lactose	74438-3
622716	Maltose	74895-4
622717	Raffinose	48147-3
622744	Reviewed By	18771-6
622708	Interpretation	59462-2