

Hamilton General Hospital Core Lab, Level 1 237 Barton Street East Hamilton, Ontario L8L 2X2 Telephone (905) 577-1477 Fax No. (905) 528-1464

TO:

REFERRING CLIENTS

FROM:

Barb Baltzer, Coordinator Laboratory Reference Centre

Hamilton Regional Laboratory Medicine Program

DATE:

February 22, 2012

SUBJECT:

Laboratory Test Information Guide Updates

Bayer, the manufacturer of the reagent for determining "reducing substances" (Clinitest) has discontinued production of this reagent. We have not been able to find a commercial replacement reagent, therefore **effective March 31, 2012**, we will no longer be able to provide "reducing substances, urine or feces" as a stand-alone test or provide 'reducing substances, urine' as part of the Urine Metabolic Screen.

This will impact the "sugar chromatography" analysis which was done as part of the Urine Metabolic Screen. This is a labour intensive manual assay that cannot be offered on all Urine Metabolic Screening samples. Instead, this test was automatically performed when a reducing substance test was positive. With the loss of the "reducing substances" screen, effective March 31, 2012, we will no longer be able to provide "sugar chromatography" as part of the Urine Metabolic Screen.

All samples collected on or after March 31, 2012 will be affected by these changes. This will include minor changes in the reporting format to remove lines reporting "reducing substances" and "sugar chromatography".

The primary metabolic indication for urine reducing substances is to screen for elevation of fructose or galactose in the urine. To reduce the clinical impact of the above changes, we will offer specific testing for these conditions:

Effective March 31, 2012:

1. For clinical suspicion of fructose metabolism defects, urine "sugar chromatography" will now be offered as a stand-alone test. The main fructose metabolism defect that is of clinical concern is Hereditary Fructose Intolerance (aldolase B deficiency), which has symptoms of hypoglycemia and lactic acidosis on ingestion of fructose, as well as renal Fanconi syndrome, hepatomegaly and eventual hepatic failure. It typically presents only after sucrose or fructose is introduced into the diet, and patients tend to develop an aversion to these foods. Increased fructose in the urine is only seen in this disorder upon ingestion of sucrose or fructose, therefore, the urine test may be negative if there is no

recent ingestion. Fructosuria can also be seen in the benign condition Essential Fructosuria (fructokinase deficiency), which is asymptomatic.

Test Name: Sugar Chromatography

Alternate Names: Fructose, Galactose, Glucose, Lactose, Sucrose

Specimen Requirements: 1 mL, Random Urine. Specimen Handling: Store and ship frozen.

TAT: 15 days

Reference Interval: Descriptive report.

Price: \$25.00

Effective immediately:

2. For clinical suspicion of galactosemia (early onset cataracts with or without liver failure, hyperbilirubinemia, renal Fanconi syndrome, primary ovarian failure), urine "sugar chromatography" can be requested as detailed above. This test can detect all types of galactosemia, but the urine test <u>may be negative</u> if there is no recent ingestion of lactose or galactose. For classical galactosemia, i.e. galactose-1-phosphate uridyltransferase (GALT) deficiency, the more specific and sensitive "galactosemia screen" will now be offered to screen for GALT activity in the blood.

Test Name: Galactosemia Screen Specimen Requirements: ADULT:

4 mL Green Heparinized Tube, Blood

NEONATAL VOLUME:

800 µL (0.8 mL) Green Microtainer, Blood

Specimen Handling: Store and ship whole blood at 4° - 8°C. Do NOT freeze.

Specimen must be analyzed within 5 days of collection.

TAT: 7 days Price: \$75.00

If you have further questions, please contact:

Dr. Murray Potter, MD, FRCPC, FCCMG

Head – Biochemical Genetics Laboratory Hamilton Regional Laboratory Medicine Program Hamilton Health Sciences

Tel (905)521-2100 ext 73722 Email: <u>mpotter@hhsc.ca</u>