METABOLITE TEST REQUISITION FORM

PATIENT INFORMATION

Last Name: __________________________________________
First Name: __________________________________________
DOB/Age: ___________________________________________
Gender: □ Male □ Female
Medical Record #: Patient ID #: ________________________

SPECIMEN INFORMATION

Accession/Lab ID #: ___________________________________________________________________________
Specimen Type: __________________________________ Time: ___________________
Specimen Date: __________________

ADDITIONAL INFORMATION

Primary presenting symptoms: ________________________________________________________________
Abnormal labs: _____________________________________________________________________________
Suspected diagnosis: _________________________________________________________________________
Diet or infant formula: ______________________________________________________________________
Medication(s): _____________________________________________________________________________

If this space is not sufficient please attach clinical summary or patient history.

TEST(S) REQUESTED

☐ 5- Methyltetrahydrofolate - CSF
☐ Acylcarnitine profile - DBS, plasma, serum or whole blood
☐ Amino acids - plasma, serum or CSF
☐ Carnitine levels - DBS, plasma, or serum
☐ Everolimus - whole blood
☐ GABA (free and total) - CSF
☐ Homocysteine (total) - plasma or serum (CSF is a research test)
☐ Lactate - CSF
☐ Monoamine neurotransmitter metabolites - CSF*
☐ Neopterin - CSF
☐ Organic acids - urine
☐ Sirolimus (rapamycin) - whole blood
☐ Tertrahydrobiopterin and neopterin - CSF*

*Must be collected in Baylor CSF collection tubes

RESULTS INFORMATION

Name: __________________________________________
Results Address: __________________________________
Phone: __________________________________________
Fax: ____________________________________________

BILLING INFORMATION

Billing Address: __________________________________
Phone: __________________________________________
Fax: ____________________________________________

The IMD does not bill patients, Medicare, Medicaid, or insurance.

Baylor Use Only

Demographic Entry Quality Check

Sample Process: _________________________________
Testing Department: ___________________________
Client Services: ________________________________

UPDATE: 6/28/17
<table>
<thead>
<tr>
<th><strong>TEST NAME</strong></th>
<th><strong>SPECIMEN REQUIREMENTS</strong></th>
<th><strong>SHIPPING</strong></th>
<th><strong>TURNAROUND TIME</strong></th>
<th><strong>CPT CODE</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>5-Methyldihydrofolate</td>
<td>• CSF- 0.5 mL; minimum 250 µL; store at -70°C until transport Can be collected in a large sterile tube or in a Baylor provided CSF collection kit.</td>
<td>• 3-4 pounds dry ice</td>
<td>12 business days</td>
<td>82542</td>
</tr>
<tr>
<td>Acylcarnitine profile</td>
<td>• DBS (preferred*)- 3 completely filled spots; minimum is 1 spot; air-dry for 4-6 hours and then store individually in glassine envelope at 2-8°C until transport • Plasma/Serum- 0.2 mL of heparinized/EDTA plasma/serum; minimum is 0.1 mL; store at -20°C until transport • Whole blood- 3 mL; collected in heparin (green-top) or EDTA (purple-top) tube; minimum is 2 mL; store at room temperature until transport</td>
<td>• DBS-/Whole blood-room temperature • Plasma/Serum- 3-4 pounds dry ice</td>
<td>1-3 business days</td>
<td>82017</td>
</tr>
<tr>
<td>Amino acids</td>
<td>• CSF- 0.5 mL; minimum 250 µL; store at -20°C until transport Can be collected in a large sterile tube or in a Baylor provided CSF collection kit. • Plasma (preferred)/Serum- 1 mL of plasma (green or purple-top)/serum (pink or tiger-top); minimum is 250 µL; separate within 1 hour of collection and store at -20°C until transport</td>
<td>• 3-4 pounds dry ice (all specimen types)</td>
<td>7 business days</td>
<td>82139</td>
</tr>
<tr>
<td>Carnitine levels</td>
<td>• DBS- 3 completely filled spots; minimum is 1 spot; air-dry for 4-6 hours and then store individually in glassine envelope at 2-8°C until transport. • Plasma (preferred**)/Serum- 0.2 mL of heparinized/EDTA plasma/serum; minimum is 0.1 mL; store at -20°C until transport • Whole blood- 3 mL; collected in heparin (green-top) or EDTA (purple-top) tube; minimum is 2 mL; store at room temperature until transport</td>
<td>• DBS-/Whole blood-room temperature • Plasma/Serum- 3-4 pounds dry ice</td>
<td>1-3 business days</td>
<td>82379</td>
</tr>
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</table>

* Acylcarnitine profile- The preferred specimen is dried blood spots (DBS) because the long-chain acylcarnitines are absorbed on the surface of the red cells so that the normal levels are much higher for DBS than in plasma. Therefore, the elevations of these in some milder forms of long-chain fatty acid oxidation disorders may not be as reliably detected in the plasma as they are in the DBS. Serum levels are acceptable.

** Carnitine levels- The preferred specimen is plasma because the free carnitine levels in plasma reflect the circulating available free carnitine and physicians are more familiar with the normal ranges for plasma free carnitine. Serum specimens are acceptable. The reference ranges for DBS free carnitine is lower than for plasma due to lower levels in red cells.
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<tbody>
<tr>
<td>Everolimus</td>
<td>• Whole blood- 3 mL; minimum 0.5 mL; collected in EDTA (purple-top); Trough levels should be obtained: 12-18 hours after oral dose, 12 hours after intravenous, or immediately prior to next dose; store at room temperature for up to 24 hours</td>
<td>• Room temperature</td>
<td>Same day if received by 9 AM</td>
<td>80195</td>
</tr>
</tbody>
</table>
| GABA (free and total)         | • CSF- 1 mL; minimum 250 µL; store at -20°C until transport  
  Can be collected in a large sterile tube or in a Baylor provided CSF collection kit. | • 3-4 pounds dry ice       | 12 business days                 | 83789    |
| Homocysteine (total)          | • Plasma /Serum- 1 mL of plasma (green or purple-top)/serum (pink or tiger-top); minimum is 250 µL; separate within 30 minutes of collection and store at 2-8°C for (up to 7 days) or at -20°C (1 month) until transport | • 3-4 pounds dry ice, or cold packs, or wet-ice | 12 business days                 | 83090    |
| Lactate                       | • CSF- 1 mL; minimum 250 µL; store at -20°C until transport  
  Can be collected in a large sterile tube or in a Baylor provided CSF collection kit. | • 3-4 pounds dry ice       | 12 business days                 | 83605    |
| Monamine neurotransmitter metabolites (HVA, 3-OMD and 5-HIAA) | • CSF^- 0.5 mL; minimum 250 µl; store at -70°C until transport  
  Must be collected in a Baylor provided CSF collection kit. | • 3-4 pounds dry ice       | 12 business days                 | 82542 x3 |
| Neopterin                     | • CSF- 1 mL; minimum 0.2 mL; store at -70°C until transport  
  Can be collected in a large sterile tube or in a Baylor provided CSF collection kit. | • 3-4 pounds dry ice       | 12 business days                 | 82542    |
| Organic acids                 | • Urine- 3 mL; minimum is 1 mL; store at 2-8°C immediately and freeze at -20°C within 4 hours | • 3-4 pounds dry ice, if local, ice packs may be used | 3-7 business days                | 83918    |
| Sirolimus (rapamycin)         | • Whole blood- 3 mL; minimum 0.5 mL; collected in EDTA (purple-top); Trough levels should be obtained: 12-18 hours after oral dose, 12 hours after intravenous, or immediately prior to next dose; store at room temperature for up to 24 hours | • Room temperature        | Same day if received by 9 AM     | 80195    |
| Tertrahydrobiopterin and neopterin | • CSF^- 1 mL; minimum 0.2 mL; store at -70°C until transport  
  Must be collected in a Baylor provided CSF collection kit. | • 3-4 pounds dry ice       | 12 business days                 | 82542 x2 |

^ Follow the steps located on the CSF collection protocol sheet (click here).
ADDITIONAL INFORMATION

- All specimens must be labeled with at least two patient identifiers that match the test requisition. All specimens will be rejected if they are received without two matching patient identifiers.
- Use indelible ink or gummed labels to label specimens.
- As per CLIA and CAP regulations, all specimens must be submitted with a complete test requisition.
- Place specimens inside a specimen transport bag and the associated documents inside the pouch in the specimen transport bag. Do NOT place the documentation inside the specimen transport bag with the specimen.
- Laboratory Hours: Monday through Friday, 8:30 am – 5:00 pm (CST).
- Always ship Monday-Thursday using an overnight trackable courier.
- No Saturday deliveries accepted.
- For STAT analysis, please contact the Program Director, Teodoro Bottiglieri, Ph.D. (Teodoro.Bottiglieri@BSWHealth.org) or the laboratory manager Erland Arning, Ph.D. (Erland.Arning@BSWHealth.org)
- Only critical results are reported immediately by telephone and fax.
- Results are available for a verbal report (or if possible, a preliminary fax on request) within the turnaround time specified.
- Result reports are faxed and mailed to the submitter and physician (if provided).
- The IMD does not bill patient, Medicare, Medicaid or insurance.
- Please contact client services at (214)820-4533 with question about test price, CPT codes, billing or invoicing.

SHIPPING ADDRESS

Institute of Metabolic Disease
ATTN: Sample Processing
3812 Elm St.
Dallas, TX 75226

Updated: 6/28/17
CSF COLLECTION PROTOCOL

REQUIREMENTS
- The CSF must be collected in our sample collection tubes for the measurement of Monoamine Neurotransmitter Metabolites and Tetrahydrobiopterin and Neopterin Profile metabolite assays (these specimens may be used for 5-MTHF, Amino Acids, Lactate, and GABA as well, if requested).
- Call Institute of Metabolic Disease (214-820-4533) to obtain appropriate sample collection tubes.
- Each sample collection set consists of 5 microcentrifuge tubes in a cardboard holder. Tube #3 contains antioxidants necessary to protect the sample integrity. One set of tubes is required per patient.
- Please contact us at 214-820-4533 if you have any additional questions.
- If the sample has been already been collected without using our Special Collection Kit, please contact us at 214-820-4533 to discuss testing options.

COLLECTION INSTRUCTIONS
1. The CSF must be collected from the first drop into the designated tubes in the order indicate in the following table. **DO NOT COLLECT THE CSF IN ONE LARGE TUBE AND ALIQUOT INTO THE TUBE SET.**
2. Fill each tube to the marked line with the following volumes, indicate in the following table.

<table>
<thead>
<tr>
<th>Tube Number</th>
<th>Required volume</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>0.5 mL</td>
</tr>
<tr>
<td>2</td>
<td>0.5 mL</td>
</tr>
<tr>
<td>3</td>
<td>1.0 mL</td>
</tr>
<tr>
<td>4</td>
<td>1.0 mL</td>
</tr>
<tr>
<td>5</td>
<td>0.5 mL</td>
</tr>
</tbody>
</table>

The total CSF volume required is at least 3.0 mL

3. If the samples are not blood contaminated, place the tubes on ice (or dry ice if available) at the bedside. Transfer the samples to a -80°C freezer ASAP. If the samples are blood contaminated, the tubes should immediately be centrifuged (prior to freezing) and the clear CSF transferred to new similarly labeled tubes then frozen and stored at -80°C ASAP. **BLOOD CONTAMINATED SAMPLES WILL BE REJECTED!**
4. Store all samples at -80°C until transport.