

Adult Metabolic Diseases Clinic

4th Floor – 2775 Laurel Street

Vancouver, BC V5Z 1M9

Tel: 604-875-5965

Fax: 604-875-5967

**PROTOCOL FOR CSF COLLECTION FOR NEUROTRANSMITTERS
WITH OR WITHOUT AMINO ACIDS**

Indications:

- Neurological presentation with movement disorder (dystonia, tremor, myoclonus, spasticity), seizures, autonomic dysfunction
- Mild elevation of plasma phenylalanine
- Elevation of prolactin that are not known to be secondary side effects of medications

Preparation:

Location

LP to be performed by physician in the metabolic clinic, in medical day care, in radiology (for ultrasound guided procedures) or on the ward

- If LP will be done away from the metabolic clinic:
 - o consider taking the rolling cart so that there is a working surface for the LP
 - o Take gowns, sterile gloves and masks
 - o Take extra labels
 - o Take the requisitions
- orders will be written by the Adult Metabolic Physician and will indicate whether or not CSF and plasma amino acids need to be collected
- sedation is not required unless the neurological/behavioural condition of the patient warrants it (uncooperative, anxious, with severe cognitive delay)
- should be done in the morning as patient is required to fast

Booking

two AMDC RNs need to assist with this procedure. Book on a day when all RNs are working so there is backup as needed. **Procedure must be booked on a Monday, Tuesday, Wednesday, Thursday, NOT Friday*

VGH Radiology/ultrasound: phone 604-875-4340 push #1

VGH MRI: phone 54684

Situation	Paperwork details
<p>ultrasound-guided LPs in radiology without sedation</p>	<ul style="list-style-type: none"> □ medical imaging requisition: <ul style="list-style-type: none"> ○ check off “ultrasound” ○ under “Exam requested”: LP for CSF neurotransmitters with ultrasound guidance ○ under “Pertinent history/medications”: ___ year old male/female with underlying history of _____. LP for CSF: glucose, lactate, protein, amino acids, neurotransmitters. *This needs special sample collection and handling. PLEASE CALL AMDC WITH THE APPOINTMENT so that the RN can assist with sample handling and collection. Needs blood glucose collection prior to LP for CSF/blood glucose ratio calculation. ○ Fax it to 604-875-4061 with our contact information who will give it to the neuroradiologist who protocols the request. The clerks will then call us with an appointment.
<p>for ultrasound-guided LPs in radiology with sedation (ie. patient uncooperative, anxious, or has severe cognitive delay)</p>	<ul style="list-style-type: none"> □ medical imaging requisition: <ul style="list-style-type: none"> ○ check off “Angiogram/Interventional” ○ under “Exam requested”, indicate “LP for CSF neurotransmitters under sedation with ultrasound guidance” ○ under “Pertinent history/medications”: ___ year old male/female with underlying history of _____. LP for CSF: glucose, lactate, protein, amino acids, neurotransmitters. *This needs special sample collection and handling. PLEASE CALL AMDC WITH THE APPOINTMENT so that the RN can assist with sample handling and collection. Needs blood glucose collection prior to LP for CSF/blood glucose ratio calculation. ○ Fax it to 604-875-4061 with our contact information who will give it to the neuroradiologist who protocols the request. The clerks will then call us with an appointment.
<p>LP is being done in conjunction with an MRI under sedation:</p>	<ul style="list-style-type: none"> ○ complete an MRI requisition indicating the patient requires sedation ○ fax it to MRI at 604-875-4175 ○ need to send patient/family/caregiver the MRI checklist and questionnaire ○ MRI will not set an appointment date until they have all the paperwork ○ once you have the date for the MRI, complete a medical imaging requisition: <ul style="list-style-type: none"> ▪ indicate the date of the appointment at the top and that the LP needs to be done AFTER the MRI so that the MRI is not affected (may look like meningitis otherwise) ▪ Check off “Angiogram/Interventional” ▪ under “Exam requested”: LP for CSF neurotransmitters under general anesthetic post MRI ▪ under “Pertinent history/medications”: ___ year old male/female with underlying history of _____. LP for CSF: glucose, lactate, protein, amino acids, neurotransmitters. *This needs special sample collection and handling. PLEASE CALL AMDC WITH THE APPOINTMENT so that

	<p>the RN can assist with sample handling and collection. Needs blood glucose collection prior to LP for CSF/blood glucose ratio calculation.</p> <ul style="list-style-type: none">○ Fax the medical imaging requisition to 604-875-4061 with our contact information who will give it to the neuroradiologist who protocols the request. The clerks will then call us with an appointment.
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Equipment

- Obtain **2 sets** of special tubes from Biochemical Genetics Lab at BC Children's Hospital (one set for collection and one set in case the CSF needs to be centrifuged and transferred into a clean tube)
- obtain wet ice (ice chips with a bit of water) from VGH lab the morning of the procedure for the Styrofoam box and ice in the urine collection container
- obtain orange or apple juice to give to patient post procedure as needed
- Gabriella's tube holder to collect CSF in the non-sterile micro tubes from MNG (one in LP bag)

Lab work

- Patient needs to have a platelet and INR level from at least one month prior to the procedure
- Patient will require a fasting glucose and lactate level (***IF CSF AMINO ACIDS ARE BEING COLLECTED**, add plasma amino acids) done the day of the procedure. If the procedure is first thing in the morning, advise an overnight fast. If the procedure is in the afternoon, advise fasting after breakfast. This can be ordered as STAT bloodwork in the outpatient lab so the patient doesn't have to wait.

Paper work

- Lab requisitions:
 - CSF neurotransmitters to be sent out by BGL (Biochemical Genetics Lab)
 - CSF glucose and proteins, cell count and differential, lactate
 - And as needed, CSF amino acids, to be sent to BGL (Biochemical Genetics Lab)
- 10 S cards to label the tubes (ask clerks to print out), D cards for requisitions
- Medical Neurogenetics Neurochemistry Req
- patient needs to sign Bill 73 and a hospital consent for the procedure
- LP patient teaching pamphlet

Collection and sample handling:

- 8 vials of CSF will be collected in total and in this order:
 - o 5 tubes for neurotransmitters
 - o 1 tube for glucose and proteins (from tray)
 - o 1 tube for cell count and differential (from tray)
 - o 1 tube for lactate
 - o 1 tube for CSF amino acids (as ordered)
- For CSF Neurotransmitters:
 - Use special collection tubes from Biochemical Genetics Lab at BC Children's Hospital (5 micro tubes with marked CSF volumes to be collected without losing any drop of CSF from tube #1 to #5, starting from the first drop)
 - if the physician performing the LP is using a tube with a clamp, the tube may be clamped in between specimen collections
 - **specimens need to be placed immediately in a labeled, ice-filled urine sample container with a label over the top of all tubes and immediately placed on wet ice after collection.**
 - **Samples must be delivered to Lab on wet ice, within 10 mins of collection.**

Contact staff at VGH lab: Bev Kisby and Anna Lew within 10 minutes of collection

Sample processing and shipping instructions for Lab:

- *For CSF Neurotransmitters, if CSF is bloody, each tube (1 – 5) must be centrifuged at 1800 g, at 4 °C, for 10 mins immediately. Collect supernatant (CSF) and transferred to a clean equivalently labeled tube (1 – 5).
- Each CSF tube (free of blood contamination), is frozen immediately (within 1 hr of collection) at – 70 ° C.
- ***FOR CSF amino acids**, if CSF is blood stained, spin at 1800 g, at 4 ° C for 10 mins. Collect supernatant (CSF) and freeze at – 70 °C. Note on requisition that CSF was blood stained and CSF was collected after spinning.
- Package and send on Dry Ice by Dynamex courier. Include all paperwork (Lab requisition, Medical Neurogenetics Requisition, Bill 73 and signed patient consent form).

Send to:

Attention Dr.Hilary Vallance
4480 Oak Street, Rm 2F22
Biochemical Genetics Lab, C&W Hospital,
Vancouver , BC V6H 3V4
Ph: 6048752307

Phone Biochemical Genetics lab to alert them of the shipment:

Bee Toh 604-875-2345 ext 7394; email btoh@cw.bc.ca

Results:

- samples will be sent out for measurements of HVA (dopamine metabolite), 5HIAA (serotonin metabolite), 3OMD (dopamine accessory pathway metabolite), and 5MTHF (folate metabolite)
- pterins (biopterin and dihydrobiopterin) will be measured upon request, if initial results are suggestive of a biopterin deficiency
- metabolites diagnostic of pyridoxine dependent epilepsy are also reported

Diagnosed metabolic conditions:

- Biopterin disorders: GTP Cyclohydrolase deficiency (including some cases of AD dopa responsive dystonia), PTPS deficiency
- AADC deficiency, tyrosine hydroxylase deficiency, cerebral folate deficiency, Pyridoxine dependent epilepsy, pattern may be suggestive for Aicardi-Goutiers syndrome