

CINCINNATI CHILDREN'S CLINICAL LABORATORIES

For test inquiries please see page 3 for assay specific contact information. www.cincinnatichildrens.org/tma

**ADAMTS13 Activity STAT testing ONLY: Weekend & Holiday shipping: CCHMC Dock #5, 3333 Burnet Ave, Clinical Lab B4, Cincinnati, OH 45229-3039 Ship to:
CCHMC Division of Nephrology
Clinical Laboratory, T.6-325 Dock 1
240 Albert Sabin Way
Cincinnati Ohio 45229
MONDAY—FRIDAY DELIVERY ONLY**

THROMBOTIC MICROANGIOPATHY (aHUS and TTP) TEST REQUISITION

All Information Must Be Completed Before Sample Can Be Processed

| PATIENT INFORMATION | ETHNIC/RACIAL BACKGROUND (Choose All) | | |
|---|---|-----------------------------|--|
| | | | |
| Patient Name:,,,, | ☐ European American (White) | ☐ African-American (Black) | |
| MR#// | ☐ Native American or Alaskan | ☐ Asian-American | |
| Gender: □ Male □ Female | ☐ Pacific Islander | ☐ Ashkenazi Jewish ancestry | |
| | ☐ Latino-Hispanic (specify country/region of origin) | | |
| | □ Other | | |
| | (specify country/region of origin) | | |
| SAMPLE/SPECIME | EN INFORMATION | | |
| Collection Date:// | | | |
| Collection Time: | | | |
| Hereaffect and the comment of the Children of | | | |
| Has patient received a bone marrow transplant? ☐ Yes ☐ No If yes, date of bone marrow transplant P | ercent engraftment | | |
| Note: For post-transplant patients, we accept pre-transplant samples or post-tra | | | |
| accepted). Culturing of skin fibroblasts is done at an additional charge. | | | |
| | ADMATION . | | |
| BILLING INF | ORMATION | | |
| ☐ REFERRING INSTITUTION | | | |
| Institution: | | | |
| Address: C | ity/State/Zip: | | |
| Accounts Payable Contact Name: | | | |
| Phone: Fa | эх: | | |
| Email: | | | |
| * PLEASE NOTE: | | | |
| We do not third-party bill patient insurance. | | | |
| • For international billing or other billing questions, please call the laboratory rel | () | | |
| Genetics Lab Billers: 1-866-450-4198, Nephrology Lab: 513-636-4530, CBDI La | b: 513-636-4685 | | |
| REFERRING | PHYSICIAN | | |
| Physician Name (print): | | | |
| Address: | | | |
| Phone: () Fax: () | | | |
| | | | |
| Genetic Counselor/Lab Contact Name: | | | |
| Phone: () Fax: () | Email: | | |
| Date:/ | | | |
| Referring Physician Signature | | | |

Medical Necessity Regulations: At the government's request, the Molecular Genetics Laboratories would like to remind all physicians that when ordering tests that will be paid under federal health care programs, including Medicare and Medicaid programs, that these programs will pay only for those tests the relevant program deems to be (1) included as covered services, (2) reasonable, (3) medically necessary for the treatment and diagnosis of the patient, and (4) not for screening purposes.



| Patient Name: | Date of Birth: |
|---------------|----------------|

TEST(S) REQUESTED

TESTING PANELS

☐ TMA Profile aHUS/TTP — Neph

(Includes C3, C4, Factor H, Factor I, Factor B, Factor H autoantibody, and ADAMTS13 activity)

- 1 mL SER
- 1 mL PPP⁺ (no EDTA)
- ☐ TMA Complement Panel Neph

(Includes C3, C4, Factor H, Factor I, Factor B, Factor H autoantibody)

• 1 mL SER

☐ Eculizumab Pharmacokinetic Panel — Neph

(Includes Eculizumab level and CH50. For assessing complement activation and to assist in monitoring patients on eculizumab therapy)

• 1 mL SER

☐ ADAMTS13 Activity (STAT available**) — Neph

If ADAMTS13 Activity is <30%, ADAMTS13 Inhibition Assay is added. If the Inhibition test is >30%, ADAMTS13 Inhibitor Antibody test is added.

- 1 mL SER
- 1 mL PPP+ (Li Hep/Cit plasma, no EDTA)
- ☐ Complement System Screen Neph

Test for the function of the complement system via Classical, Alternative and Lectin pathways

• 0.5 mL serum (separate aliiquot)

INDIVIDUAL TESTS

| \square Alternative Pathway Functional Assay — Neph | 0.5 mL SER | |
|---|--------------------------------|--|
| ☐ ADAMTS13 Activity (STAT available**) — Neph | 1 mL PPP† (no EDTA) | |
| \square ADAMTS13 Inhibitor Ab Test — Neph | 1 mL SER | |
| □ C3a — CBDI | 0.5 mL EDTA P separate aliquot | |
| □ C5a — CBDI | 0.5 mL EDTA P separate aliquot | |
| \square CH50 Complement Total — Neph | 0.5 mL SER | |
| \square Complement Bb Level — Neph | | |
| indicate specimen: □ serum | 0.5 mL SER / EDTA P | |
| □ plasma | | |
| \square Eculizumab Level — Neph | 0.5 mL SER | |
| ☐ Factor B — Neph | 0.5 mL SER | |
| ☐ Factor H Auto-Ab — Neph | 0.5 mL SER | |
| ☐ Factor H — Neph | 0.5 mL SER | |
| ☐ Factor I — Neph | 0.5 mL SER | |
| \square Lectin Pathway Functional Assay — Neph | 0.5 mL SER | |
| ☐ SC5b-9 Level (MAC) — CBDI | 0.5 mL EDTA P separate aliquot | |

SER = serum P = plasma

PPP† = platelet poor plasma; See page 3 for instructions.

All serum, plasma, and PPP samples should be processed within 2 hours of collection, frozen, and shipped frozen on dry ice.

CELLULAR PROTEIN EXPRESSION

☐ CD46 Expression/Membrane Cofactor Protein (MCP) — CBDI by Flow Cytometry

• 3 mL ACD A/B whole blood room temp.*

Note: If ordered, sample must be sent by next-day shipping for Monday – Friday delivery only.

GENETIC TESTING

- ☐ ADAMTS13 Full gene sequencing Genetics
 - 3 mL EDTA whole blood, room temp*
- \square aHUS Genetic Susceptibility Panel Genetics

(Includes sequence analysis of ADAMTS13, C3, C4BPA, CD46 (MCP), CD59, CFB, CFH, CFHR1, CFHR2, CFHR3, CFHR4, CFHR5, CFI, DGK, MMACHC, PLG, THBD and deletion/duplication analysis of CFHR1 and CFHR3 via MLPA. Also includes analysis of variants c.2653C>T and c.2654G>A in the C5 gene, which are associated with poor response to eculizumab.)

- 3 mL EDTA whole blood, room temp*
- ☐ CFHR1/CFHR3 deletion analysis by MLPA
- ☐ Reflex to del/dup of ADAMTS13, C3, C4BPA, CD59, CFB, CFI, DGKE, PLG, and THBD
- ☐ Reflex to del/dup of single gene(s)¹ (Specify): _____

¹Deletion/duplication analysis of C5, CD46 (MCP), CFH, CFHR2, CFHR4, CFHR5, and MMACHC is not available at this time

Each gene listed above is also available for order as an individual test

- \bullet 3 mL EDTA whole blood room temp*
 - ☐ Custom Gene Sequencing Genetics
 - Full Gene Sequencing for ______ gene
 - Targeted (family-specific) variant analysis for _______

Proband's name:___

Proband's DOB: _

Proband's variant:_

Please call 513-636-4474 to discuss any family specific mutation analysis with genetic counselor prior to shipment.

*Do not spin or freeze samples for Flow Cytometry or Genetic Testing. For post-transplant patients, we accept pre-transplant samples or post-transplant skin fibroblasts ONLY (blood, saliva, and cytobrushes are not accepted). Culturing of skin fibroblasts is done at an additional charge.

Cincinnati Children's Clinical Laboratories

For test inquiries please contact the lab listed next to each test name:

 ${\sf CBDI-Cancer\ and\ Blood\ Disease\ Institute:\ 513-636-4685}$

Neph — Nephrology: 513-636-4530

Genetics — Molecular Genetics: 513-636-4474



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TMA TESTING INFORMATION SHEET

| Test Name | Performing Lab | Specimen Requirements | TAT/ Days Performed | CPT Codes |
|---|---|--|--|---------------------------|
| ADAMTS13 Activity | Nephrology 513-636-4530 | 1 mL platelet poor plasma Na Cit/Li Hep only (no EDTA) — spun, separated, frozen; ship on dry ice. If you have STAT/critical requests for ADAMSTS13 Activity, call 513-636-4530. | 24 hours— available on weekends and holidays | 85397 |
| ADAMTS13 Antibody Quant | Nephrology 513-636-4530 | 1 mL red top serum spun, separated, frozen; ship on dry ice* | 1 week | 85320 |
| ADAMTS13 Panel | Nephrology 513-636-4530 | 1 mL red top serum — spun, separated, frozen; ship on dry ice | 24 hours | 85397 +85335 +85320 |
| Alternative Pathway Functional Assay | Nephrology 513-636-4530 | 0.5 ml serum-spun, separated, frozen within 2 hours of collection, ship on dry ice | 1 week | 86161 |
| CH50 | Nephrology 513-636-4530 | 0.5 mL red top serum — spun, separated, frozen within 2 hrs of collection; ship on dry ice | Monday, Wednesday, Friday | 86162 |
| Complement System Screen | Nephrology 513-636-4530 | 1.5 ml serum 3 aliquots — spun separated, frozen within 2 hours of collection, ship on dry ice | 1 week | 86161x2 +86162 |
| Eculizumab Level | Nephrology 513-636-4530 | 0.5 mL red top serum — spun, separated, frozen within 2 hour of collection; ship on dry ice | Wednesday and Friday | 80299 |
| Factor B, Factor H, Factor I | Nephrology 513-636-4530 | 0.5 mL red top serum — spun, separated, frozen; ship on dry ice | 3 days | 86160 |
| Factor H Auto-Antibody | Nephrology 513-636-4530 | 0.5 mL red top serum — spun, separated, frozen; ship on dry ice | Thursday | 83516 |
| Bb | Nephrology 513-636-4530 | 0.5 mL EDTA plasma — spun, separated, frozen within 2 hours of collection, separate aliquot each test; ship on dry ice | 1 week | 86160 |
| Lectin Pathway Functional Assay | Nephrology 513-636-4530 | 0.5 mL EDTA plasma — spun, separated, frozen within 2 hours of collection, ship on dry ice | 1 week | 86161 |
| Membrane Cofactor Protein (MCP)/CD46 by Flow | Cancer and Blood Disease Institute 513-636-4685 | 3mL ACD (A or B) whole blood — room temperature, MUST be delivered within 24 hours of collection Monday — Friday only | 24 hours | 86356x3 |
| SC5b-9 (MAC Complex) | Cancer and Blood Disease Institute 513-803-3503 | 0.5 mL EDTA plasma — spun, separated, frozen within 2 hours of collection, separate aliquot each test; ship on dry ice | 1 week | 86160 |
| C3a | Cancer and Blood Disease Institute 513-803-3503 | 0.5 mL EDTA plasma — spun, separated, frozen within 2 hours of collection, separate aliquot each test; ship on dry ice | 2 weeks | 86160 |
| C5a | Cancer and Blood Disease Institute 513-803-3503 | 0.5 mL EDTA plasma — spun, separated, frozen within 2 hours of collection, separate aliquot each test; ship on dry ice | 2 weeks | 86160 |
| ADAMTS13 Gene Sequencing | Molecular Genetics 513-636-4474 | 3mL EDTA — whole blood; room temperature* | 28 days | 81479 |
| aHUS Genetic Susceptibility Panel | Molecular Genetics 513-636-4474 | 3 mL EDTA — whole blood- room temperature* | 28–42 days | 81443 |
| Custom Gene Sequencing (full or targeted) | Molecular Genetics 513-636-4474 | 3 mL EDTA — whole blood— room temperature* | 28 days | Call lab |

DO NOT FREEZE SAMPLES FOR GENETIC or CD46 TESTING.