Atypical HUS

Lab Tests and What They Mean

Diagnosing Atypical HUS

- **Atypical HUS is primarily a clinical diagnosis coupled with consistent laboratory findings.**
- **It produces a microangiopathic hemolytic anemia. This is the hallmark finding and is necessary to establish the diagnosis.**
    - **Schistocytes** (red blood cell fragments that result from membrane damage encountered during passage through vessels) **are found in the peripheral smear. They reflect the fragmentation of RBCs that occurs as the RBCs traverse vessels partially occluded by platelet and hyaline microthrombi. The peripheral smear also may contain giant platelets. This is a reflection of the reduced platelet survival time resulting from the peripheral consumption/destruction of platelets.**
    - **Thrombocytopenia** (refers to any disorder in which there are not enough platelets) **is present but is mild to moderate in severity, typically less than 60,000 per mL.**
- **Elevation of lactate dehydrogenase (LDH) and indirect bilirubin reflects intravascular hemolysis. The bilirubin rarely exceeds 2-3 mg/dL.**
- **BUN and creatinine are markedly elevated.**
- **Urine, if present, may contain protein and RBCs.**

- **The reticulocyte count (**A reticulocyte count is a blood test performed to assess the body's production of immature red blood cells) **is elevated.**

- **Coombs test results are negative, indicating that the anemia is not immunologically mediated.**

- **Plasma contains free hemoglobin that often can be seen with the naked eye. The degree correlates with the severity of the anemia.**

- **The urine contains hemoglobin, hemosiderin, albumin, RBCs, WBCs, and casts.**

Emergency Department Care should focus on supportive management, treatment of blood pressure elevation, blood transfusions, and admission with arrangement for prompt dialysis.

- **Plasma exchange (plasmapheresis combined with fresh-frozen plasma replacement) is currently the treatment of choice. Plasma exchange is performed daily until remission is obtained.**

Below are lab values that are monitored in those with atypical HUS. Values might differ from lab to lab but in general the values below approximate normal.

Renal Related Lab Values

B.U.N. (Blood Urea Nitrogen) - **Urea is the end product of protein metabolism. It is what is left over after your body uses the proteins in meat, fish, fowl and dairy products. It is important to your overall health and healing ability to eat enough protein.**

Increases can be caused by excessive protein intake, kidney damage, certain drugs, low fluid intake, intestinal bleeding, exercise or heart failure. Decreased levels may be due to a poor diet, malabsorption, liver damage or low nitrogen intake.

Normal Adult Range: 7 - 25 mg/dl
Optimal Adult Reading: 16
Pediatric Range 5-18 mg/dl

CREATININE - **Creatinine is a waste product of muscle metabolism. Low levels are sometimes seen in kidney damage. Elevated levels are sometimes seen in kidney disease due to the kidneys job of excreting creatinine, muscle degeneration, and some drugs involved in impairment of kidney function.**

Normal Adult Range: .7 - 1.4 mg/dl
Optimal Adult Reading: 1.05
Pediatric under .5 mg/dl

Hematology Values

HEMATOCRIT (HCT) **Red blood cells (RBCs) are produced in the bone marrow, in response to a decrease in their number (thus oxygen carrying capacity). Usually, the decrease is caused by the normal removal of aged RBCs by your spleen but, of course, bleeding will result in the loss of RBCs, too. Special cells that line the smallest renal blood vessels (i.e., renal capillaries) sense any significant loss of oxygen and secrete the hormone, erythropoietin (EPO). EPO travels through the bloodstream and when it reaches the bone marrow, the marrow reacts by producing RBCs. When there are enough RBCs to enable the oxygen level in the blood to return to normal, the secretion of EPO stops.**

Most renal diseases interfere with the normal secretion of EPO and thus cause severe anemia. Fortunately, human EPO is now available as an injection to prevent the severe form of the anemia, and thus improve the quality of your life. In case you wondered, most doctors are reluctant to administer enough EPO to return the Hct to absolutely normal levels, as there is evidence that the thicker blood could jeopardize your vascular access and perhaps cause other problems.

Normal Adult Female Range: 37 - 47%
Optimal Adult Female Reading: 42%
Normal Adult Male Range 40 - 54%
Optimal Adult Male Reading: 47
Normal Newborn Range: 50 - 62%
Optimal Newborn Reading: 56
Pediatric Range 30-40%

HEMOGLOBIN (HGB) **Hemoglobin is the iron-containing pigment of red blood cells (RBCs); its function is to carry oxygen from your lungs to all the tissues throughout your body. Hemoglobin is decreased in uremia because the number of RBCs is decreased. Taking erythropoietin (EPO) improves the RBC count and, thus, the Hb level. Improved oxygen-carrying capacity markedly improves your exercise capacity, brain function (clarity of thinking) and overall quality of life.**

Normal Adult Female Range: 12 - 16 g/dl
Optimal Adult Female Reading: 14 g/dl
Normal Adult Male Range: 14 - 18 g/dl
Optimal Adult Male Reading: 16 g/dl
Normal Newborn Range: 14 - 20 g/dl
Optimal Newborn Reading: 17 g/dl
Pediatric Range 10-13 gm/dl

R.B.C. (Red Blood Cell Count) **Responsible for carrying oxygen and carbon dioxide throughout the body. Iron deficiency will lower RBC count. In more reduced**

count, it may indicate hemorrhage or hemolysis. RBC lives for 120 days so an anemia of any kind other than hemorrhage indicates a long standing problem.

Normal Adult Female Range: 3.9 - 5.2 mill/mcl
Optimal Adult Female Reading: 4.55
Normal Adult Male Range: 4.2 - 5.6 mill/mcl
Optimal Adult Male Reading: 4.9
Lower ranges are found in Children, newborns and infants

W.B.C. (White Blood Cell Count) The body's primary means of fighting infection. Decreased levels may indicate overwhelming infections (viruses). Increased levels indicate bacterial infection, emotional upsets and blood disorders.

Normal Adult Range: 3.8 - 10.8 thous/mcl
Optimal Adult Reading: 7.3
Children, newborns and infants. 3,000 – 10,000 cells

PLATELET COUNT Platelets (PLT) - Play an important role in blood clotting. Decrease in number occurs in hemolytic anemia and by monitoring them disease activity can be monitored and platelets replaced.

Normal Adult Range: 130 - 400 thous/mcl
Optimal Adult Reading: 265
Children 170-380 thous/mcl

Reticulocyte count is a blood test performed to assess the body's production of immature red blood cells (reticulocytes). A reticulocyte count is usually performed when patients are evaluated for anemia and response to its treatment. It is sometimes called a retic count.

Normal Range Adults 0.5-2.5%. Women and children usually have higher reticulocyte counts than men.

LDH (lactate dehydrogenase) The main use for LDH is as a general indicator of the existence and severity of acute or chronic tissue damage and, sometimes, as a monitor of progressive conditions. LDH isoenzymes, there are 5, may also be used in differential diagnosis to help determine which organs are likely to be involved. LDH-1 is indicative of tissue damage in the renal cortex.

Normal Range: 105 to 333 IU/L