

LAB PROCEDURES AND ANALYSIS

The test for ammonia should only take a few minutes after receipt of the sample, so clinicians can expect a rapid report from the laboratory.¹ Direct procedures, such as enzymatic methods, are more widely used than indirect procedures to measure plasma ammonia levels, as they are more easily automated.¹¹

NORMAL BLOOD AMMONIA REFERENCE LEVELS⁷

Decision limits of plasma ammonia concentrations* (to be interpreted with the clinical situation)¹:

Approximate age	µmol/L
Premature neonates	< 150
Term neonates	< 100
Infants	< 40
Adults	11-32

*Individual laboratory reference levels may vary.

INTERPRETATION OF RESULTS

The goal is to rule out hyperammonemia without delay.¹

If hyperammonemia is confirmed, other laboratory tests that may be helpful in diagnosing a urea cycle disorder (UCD) include¹:

- Blood glucose, blood gases, electrolytes, lactate, and transaminases
- Plasma amino acids and blood acylcarnitines
- Urine amino acids, organic acids, and orotic acid

Regardless of cause, any delay in recognizing or treating hyperammonemia increases the risk of irreversible brain damage or death.¹

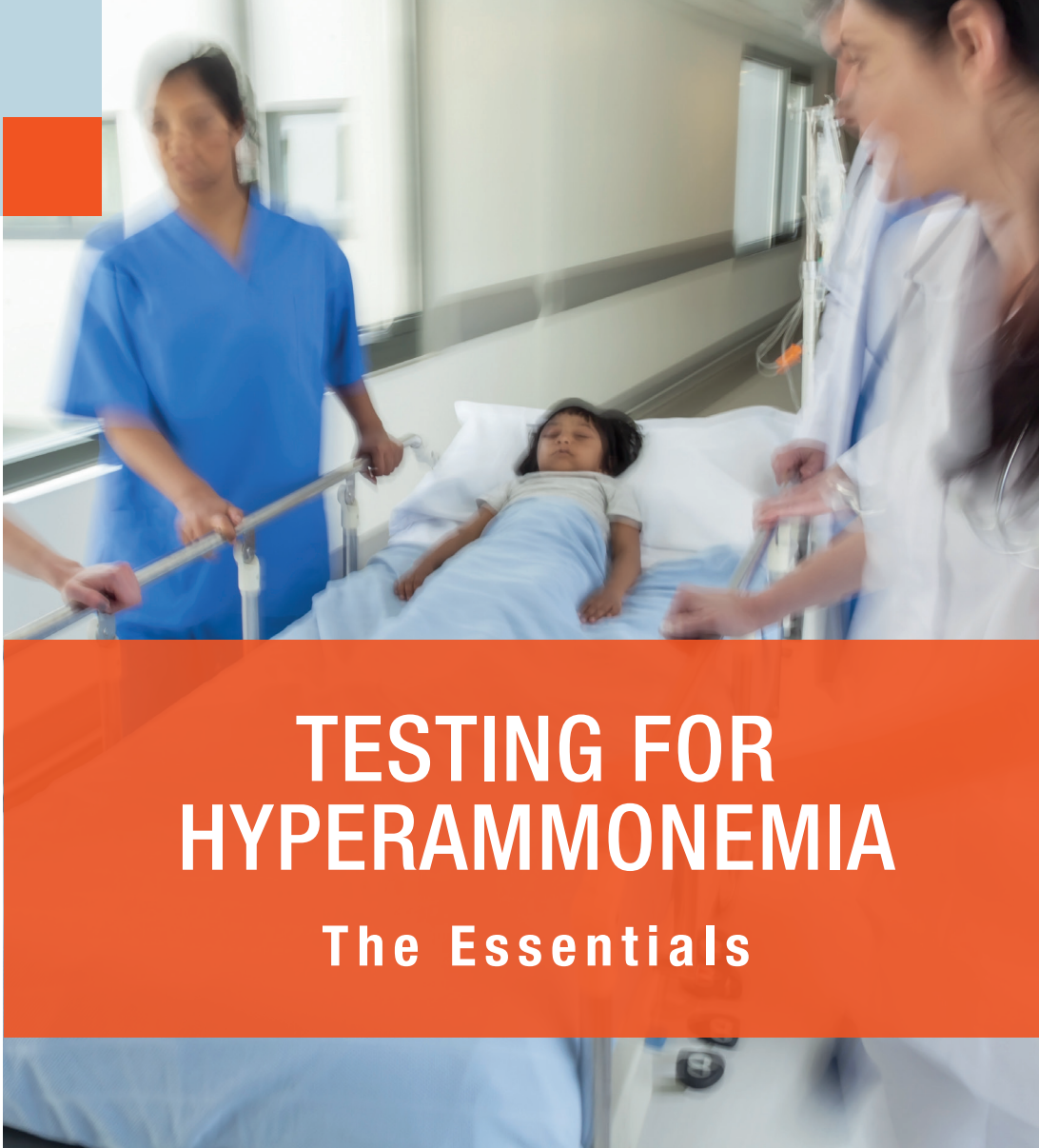
RAPID, ACCURATE BLOOD AMMONIA DRAW AND ANALYSIS: A QUICK SUMMARY

- Alert laboratory to STAT order
- Sample free-flowing venous or arterial blood – no tourniquet
- Use lithium heparin or EDTA tube
- Immediately place sample on ice, with orders to be handled STAT

REFERENCES

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HYPERAMMONEMIA CAN BE A LIFE-THREATENING CONDITION THAT MAY AFFECT PATIENTS AT ANY AGE.¹

■ A LIFE-THREATENING CONDITION

Hyperammonemia is a metabolic condition characterized by excess ammonia in the blood that can affect patients at any age. **If left untreated, hyperammonemia may lead to irreversible neurological damage or death.**¹ Thus, particular care should be taken when sampling and handling plasma specimens to increase the positive predictive value of ammonia test results.²

■ SIGNS AND SYMPTOMS OF ACUTE HYPERAMMONEMIA

Signs and symptoms of acute hyperammonemia are nonspecific and are mostly neurological in origin.³ Symptoms may include:

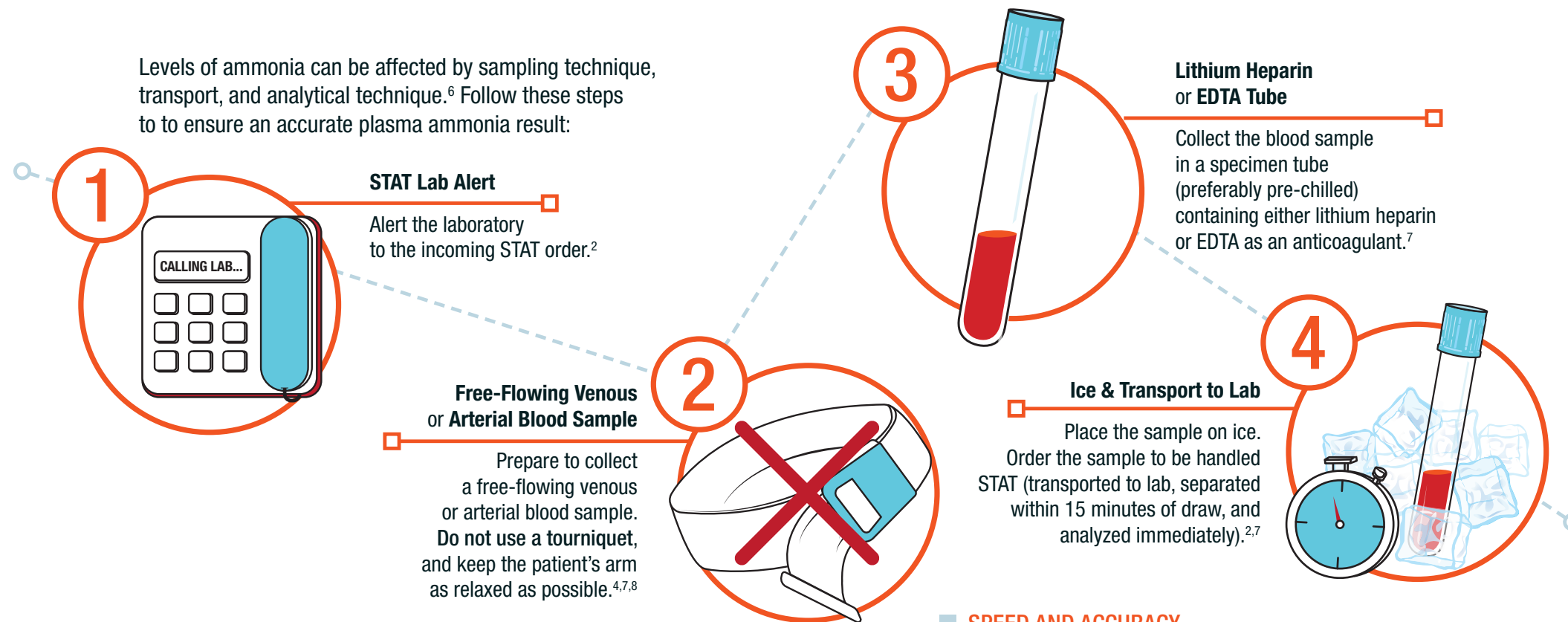
- Somnolence and lethargy progressing to coma³
- Vomiting (metabolic acidosis)¹
- Seizures¹
- Peripheral circulatory failure¹
- Cerebral edema (respiratory alkalosis)¹
- Liver failure¹
- Multiorgan failure¹
- Postpartum psychosis¹
- In neonates: sepsis-like picture, respiratory distress, hypo/hyperthermia¹

■ COMMON CAUSES OF HYPERAMMONEMIA

Common causes of hyperammonemia include⁴:

- Liver failure
- Reactions to drugs (e.g., valproic acid)
- Hemolytic disease
- Gastrointestinal bleeds
- Urea cycle disorders (UCDs)
- or other inborn errors of metabolism (IEMs)^{2,5}

ACCURATE BLOOD AMMONIA DRAW AND ANALYSIS: THE ESSENTIAL STEPS



< **Newborns** who develop severe hyperammonemia after 24 hours of age usually have a UCD or an organic acidemia—both IEMs.⁵

< In **older patients**, liver disease is a likely cause of hyperammonemia. However, UCDs should be considered since 69% of UCD patients present with symptoms of hyperammonemia later in life.^{9,10}

■ SPEED AND ACCURACY

- The management of a hyperammonemic crisis not only demands a rapid plasma ammonia reading but also an accurate one.¹
- Critical clinical decisions depend on having the right diagnostic information.
- Hyperammonemic crises can develop quickly, increasing the risk of neurological damage and death.¹
- If hyperammonemia is confirmed, treatment should not be delayed.¹
- Consider calling a metabolic geneticist.