Porphyria (Acute) Testing Algorithm*

**Symptoms:**
- Neurovisceral attacks (abdominal pain, neuropathy, psychiatric symptoms)
- Tachycardia and hypertension

**Possible acute porphyria:**
- Acute intermittent porphyria (AIP)
- Variegate porphyria (VP)*
- Hereditary coproporphyria (HCP)*
- Aminolevulinic acid dehydratase deficiency porphyria (ADP)

**Increased coproporphyrin III/I ratio (<10) and protoporphyrin**

**Increased coproporphyrin III/I ratio (>10) and coproporphyrin III**

**Normal PBGD activity and fecal porphyrin profile**

**Normal results—were samples collected during an acute episode?**
- **YES**
  - Excludes acute porphyrias
- **NO**
  - Retest during acute episode

**To differentiate ADP from tyrosinemia type I and heavy metal intoxication order:**
- ALAD / Aminolevulinic Acid Dehydratase (ALAD), Whole Blood*
- OAUI / Organic Acids Screen, Urine
- HMCRIU / Heavy Metal/Creatinine Ratio, with Reflex, Urine OR
- HMDB / Heavy Metals Screen with Demographics, Blood

**Decreased ALAD activity**
- Confirms ADP

**Normal ALAD activity**
- Excludes ADP

**Isolated ALA increase**

**To perform:**
- PBGD / Porphobilinogen Deaminase (PBGD), Whole Blood*
- OAU / Organic Acids Screen, Urine
- HMCRU / Heavy Metal/Creatinine Ratio, with Reflex, Urine OR
- HMBF / Heavy Metals Screen with Demographics, Blood

**Specimens collected during symptomatic period will be most informative**

**a. 80% of patients with VP have cutaneous symptoms**
**b. 20% of patients with HCP have cutaneous symptoms**
**c. Specimens collected during symptomatic period will be most informative**
**d. Plasma specimen for those unable to give urine**
**e. ALAD test is not useful for lead intoxication cases**
**f. 5% of AIP patients have normal PBGD activity in erythrocytes**
**g. Specimens collected during asymptomatic period will be most informative.**

**Interpretive report provided for all tests in this algorithm**

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