### Hyperoxaluria Diagnostic Algorithm

**Normal Kidney Function**
- Stones or nephrocalcinosis in childhood
- Recurrent calcium oxalate stones or nephrocalcinosis in adults
- Family history of primary hyperoxaluria

Perform urine oxalate \( OXU / Oxalate, 24\text{ Hour, Urine} \)

- Urine oxalate >0.7 mmol/1.73 m²/24 hours
- Urine oxalate/urine creatinine > normal for age*

- Confirm urine oxalate with repeat testing
- Perform HYOX / Hyperoxaluria Panel, Urine

Hyperoxaluria confirmed:
- Urine oxalate >0.7 mmol/1.73 m²/24 hours
- Urine oxalate/urine creatinine > normal for age
- Urine glycolate, L-glycerate or 4-hydroxy-2-oxoglutarate > normal for age.

**Kidney Insufficiency or Kidney Failure**
- (eGFR <30 mL/min/1.73m²)
- Increased serum creatinine with calcium oxalate stones
- Calcium oxalate tissue deposits
- Nephrocalcinosis

Perform plasma oxalate: \( POXA1 / Oxalate, Plasma \)
And
If possible, perform urine oxalate: \( OXU / Oxalate, 24\text{ Hour, Urine} \)

- Urine oxalate >0.5 mmol/1.73 m²/24 hours
- Plasma oxalate >20 mcmol/L

**Secondary causes present?**
- Malabsorption
- Gastrointestinal disease
- Very high oxalate and low calcium diet
- Premature infant

YES / NO

Perform genetic testing for the primary hyperoxalurias**
- \( AGXTZ / AGXT \) Gene, Full Gene Analysis, Varies
- \( GRHPZ / GRHPR \) Gene, Full Gene Analysis, Varies
- \( HOGA1 \) gene analysis

**Homozygosity or compound heterozygosity for known variants of \( AGXT, GRHPR, \) or \( HOGA1 \)**

YES / NO

Based on clinical presentation, consider clinical management of hyperoxaluria OR
Consider research testing for novel genetic causes of primary hyperoxaluria***

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* Random oxalate/creatinine ratios vary significantly by age. Consult pediatric reference range tables for interpretation.

** If available, guided by the HYOX interpretive report which includes recommendations for molecular testing.

*** Please contact Mayo Hyperoxaluria Center at 800-270-4637 or email hyperoxaluriacenter@mayo.edu