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 Hour, Urine

Urine oxalate > 0.7 mmol/1.73 m²/24 hours or 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.

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

Perform genetic testing as indicated in the HYOX interpretive report**
- AGXTZ / AGXT Gene, Full Gene Analysis
- GRHPZ / GRHPR Gene, Full Gene Analysis
- HOGA1 gene analysis

Homozygosity or compound heterozygosity for known mutations of AGXT, GRHPR, or HOGA1

**Kidney Insufficiency or Kidney Failure**
- Increased serum creatinine with calcium oxalate stones
- Calcium oxalate tissue deposits
- Nephrocalcinosis

Perform urine and plasma oxalate OXU / Oxalate, 24 Hour, Urine
POXA / Oxalate, Plasma

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

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.

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

Evaluate and treat accordingly

Based on clinical presentation, consider clinical management of hyperoxaluria
OR
Consider liver biopsy for alanine:glyoxylate aminotransferase (AGT) and glyoxylate reductase/hydroxypyruvate reductase (GRHPR) enzyme activities

- Decreased AGT activity
- Decreased GRHPR activity
- Normal AGT and GRHPR activities

Oxalate disorder of undetermined type

*Random oxalate/creatinine ratios vary significantly by age. Consult pediatric reference range tables for interpretation.
**Interpretive report includes an overview of results and of their significance along with a recommendation for confirmatory molecular testing for either AGXT, GRHPR, or HOGA1.


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