

# This Case Is Nuts

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PRESENTED BY: R LOGAN JONES, MD

ACP – OREGON CHAPTER ANNUAL MEETING

SALEM, OR; NOV 8TH 2018



New Patient to Clinic

Chief Complaint : Right Flank Pain

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Generally healthy man in his 60s      Non-substance user

Proud Vegetarian      CKD 3 without proteinuria

Horseshoe kidney      Recurrent oxalate renal stones

Multiple procedures for nephrolithiasis      No history of UTI

Right CVA tenderness      Recent CT with non-obstructive stones

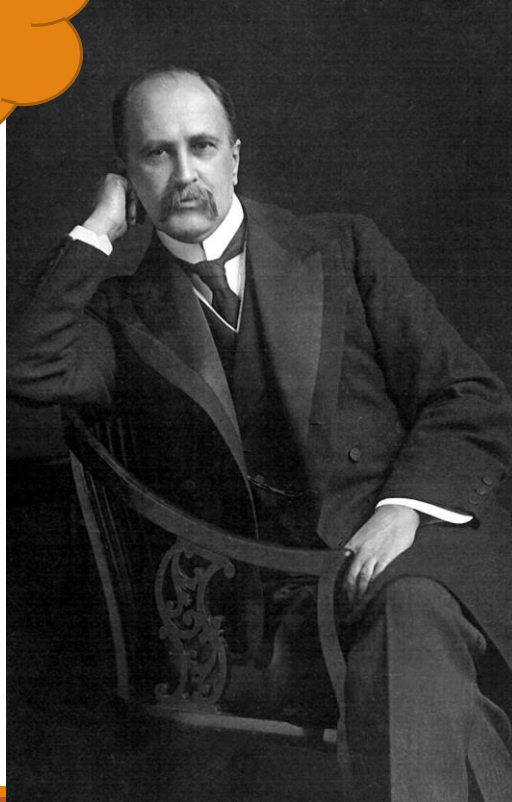


# My Assessment

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RENAL COLIC SECONDARY TO RECURRENT  
NEPHROLITHIASIS

**“Pity the  
hurried  
practitioner”**



Reviewing outside records while staffing...

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Things get interesting!

# 24-Hour Urine Collection

**EXTREME**  
**HYPEROXALURIA**

460 mg/day

## Summary Stone Risk Factors

SAMPLE ID: [REDACTED] PATIENT COLLECTION DATE: 11/16/2017

ANALYTE	← DECREASED RISK	INCREASING RISK FOR STONE FORMATION →
Urine Volume (liters/day)		● 1.84
SS CaOx		● 6.41
Urine Calcium (mg/day)	● 11	
Urine Oxalate		460 mg/day
Urine Citrate (mg/day)		● 442
SS CaP	● 0.03	
24 Hour Urine pH		● 5.677
SS Uric Acid	● 0.65	
Urine Uric Acid (g/day)	● 0.330	

## Interpretation Of Laboratory Results

**Suboptimal urine volume.** Increase urine volume above 2.5 liters.

**Low urine calcium.** Consider bowel disease or intestinal resection, renal insufficiency, very low calcium diet, thiazide diuretic.

**Extreme hyperoxaluria.** If bowel disease is not present consider primary hyperoxaluria, a rare hereditary disease that can cause progressive renal failure. Diagnosis and management of primary hyperoxaluria is beyond the scope of this report. If hyperoxaluria due to bowel disease use low oxalate, low fat diet, calcium supplements with meals Recheck at 6 weeks.

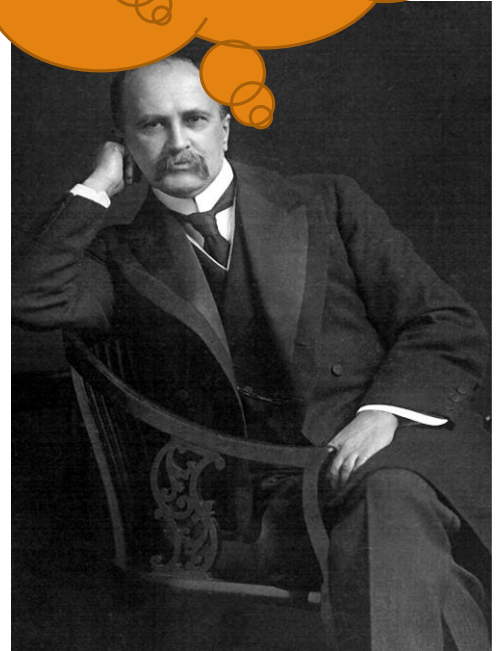
# “Extreme Hyperoxaluria”

“The Wise  
Counsel of the  
Specialist is  
Comforting”

## Urology:

- Impressed by magnitude of hyperoxaluria
- Concerns for primary genetic disease –  
**“Primary Hyperoxaluria”**

# What now?

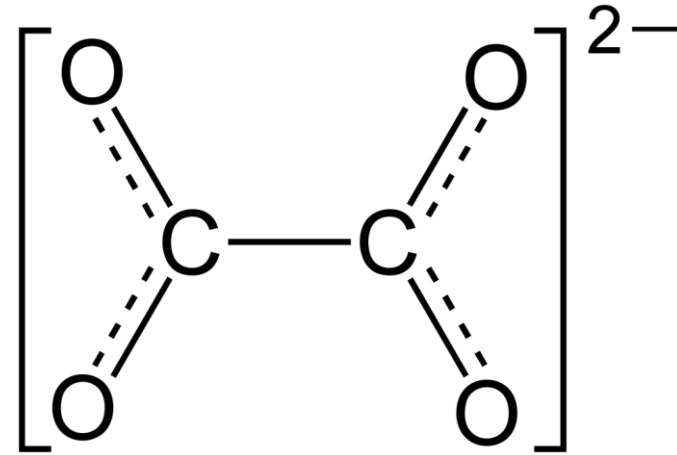


# Hyperoxaluria = Too Much Oxalate in the Urine

## Primary Hyperoxaluria

- Too much endogenous oxalate production
- Oxalate via collagen catabolism as a “dead-end” metabolic byproduct

## Secondary Hyperoxaluria



Oxalate



# Primary Hyperoxaluria (PH)

## PH 1

- Represents **80% of cases**
- Usually presents in **childhood**
- Nearly all are **ESRD by 5th decade**

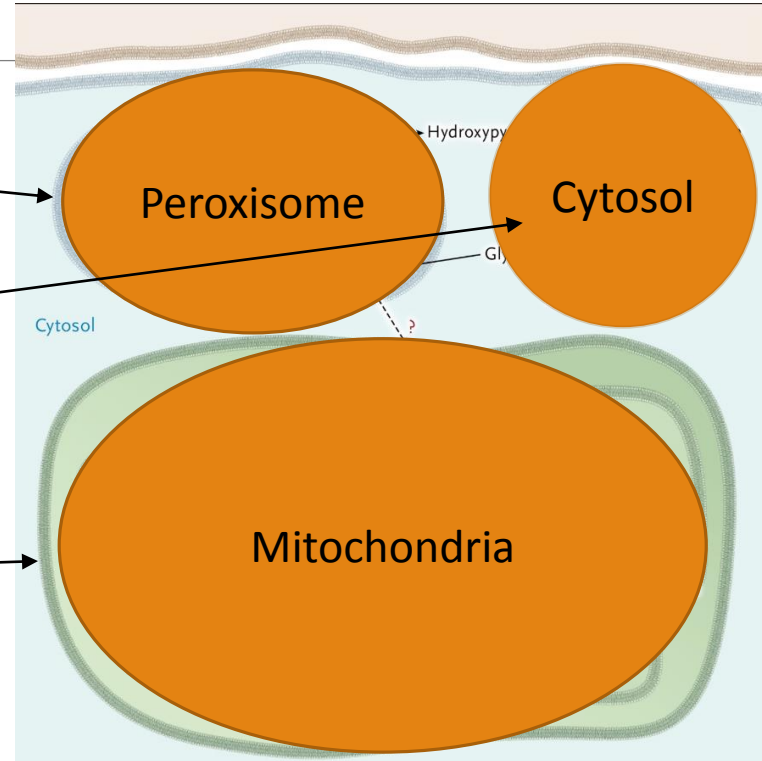
## PH 2

- Represents **~10% of cases**

## PH 3

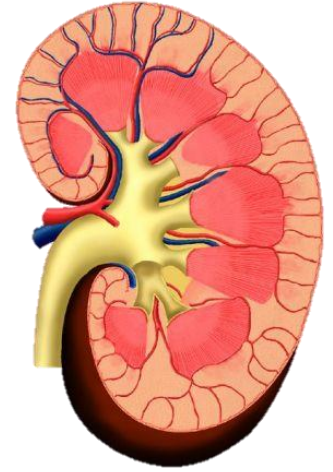
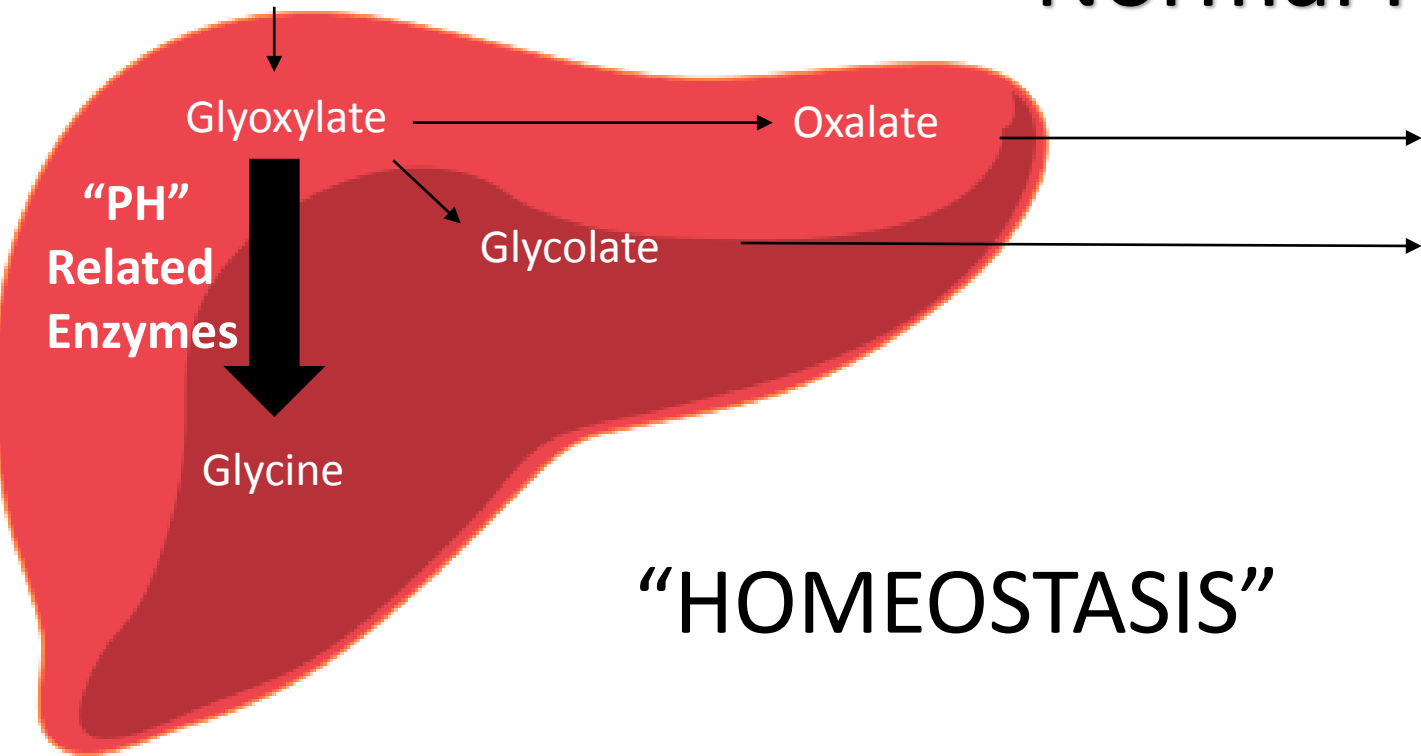
- Presents during **childhood/adolescence**

# HEPATOCTYTE



# Normal Physiology

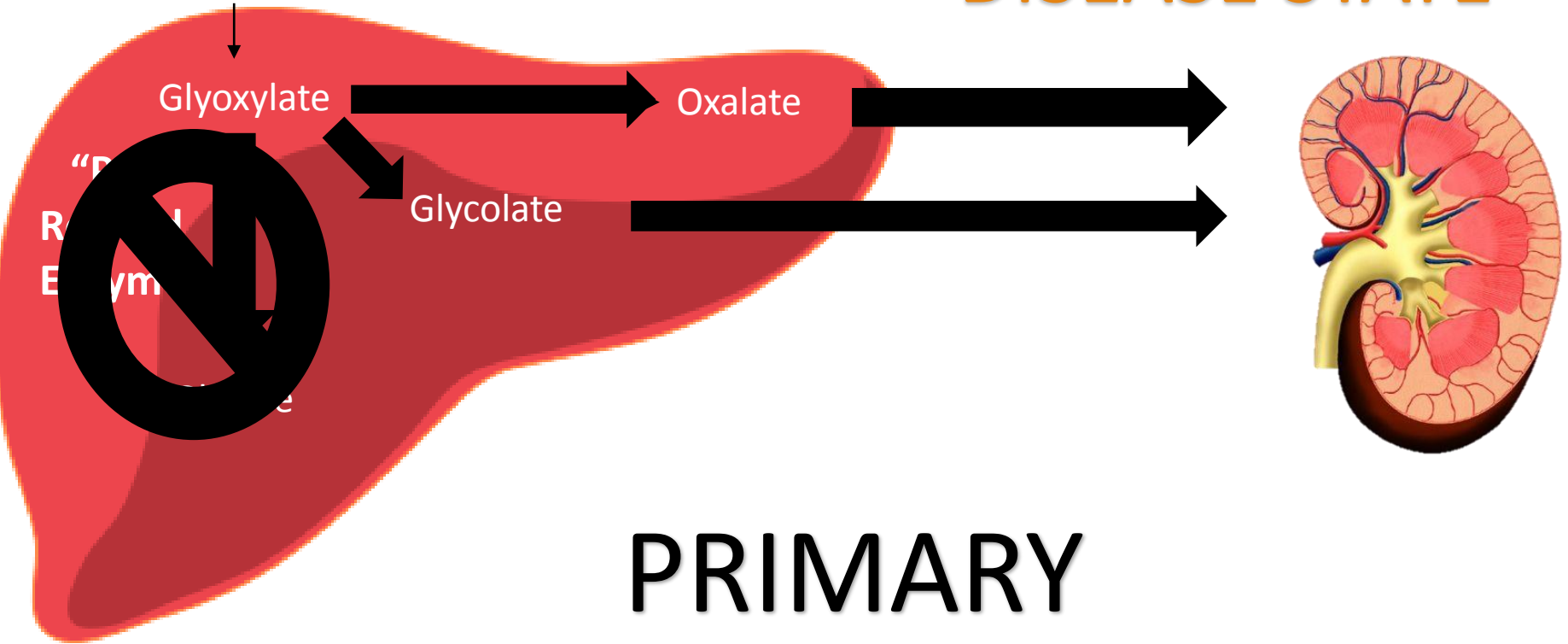
Collagen → Hydroxyproline



"HOMEOSTASIS"

Collagen → Hydroxyproline

DISEASE STATE



# PRIMARY HYPEROXALURIA

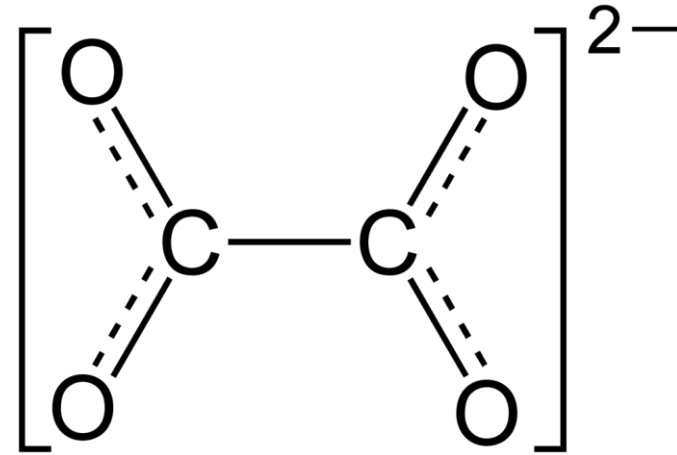
# Hyperoxaluria = Too Much Oxalate in the Urine

## Primary Hyperoxaluria

- Too much endogenous oxalate production
- Oxalate via collagen catabolism as a “dead-end” metabolic byproduct

## Secondary Hyperoxaluria

- Too much absorbed oxalate from exogenous sources
- Mostly from plant-based foods



Oxalate

# Secondary Hyperoxaluria

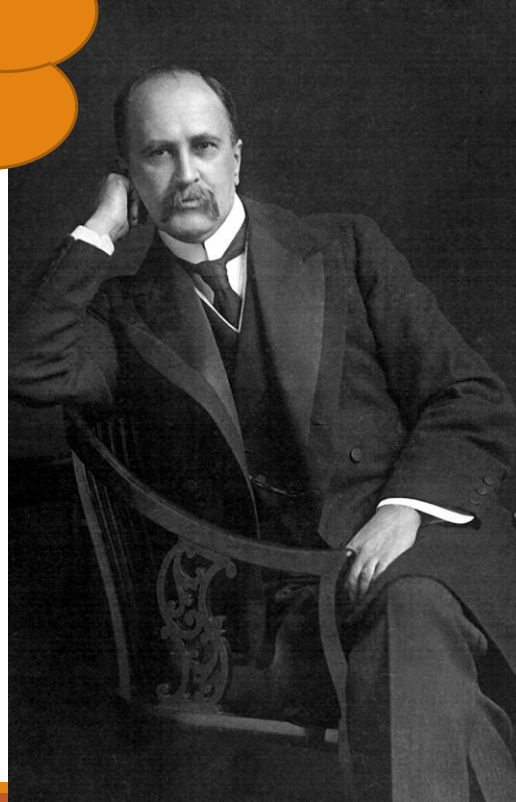
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Enteric Hyperoxaluria - *Decreased Calcium & Increased Bile Acid*

Increased Total Oxalate Content - *Oxalate Overload*



**“Methodical  
examination leads to  
safe inductions”**



# Recurrent Renal Stones: American Urologic Association Guidelines

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1. **Obtain a detailed medical and dietary history, serum chemistries and urinalysis**
2. **When a stone is available, clinicians should obtain a stone analysis at least once.**
3. **Clinicians should obtain or review available imaging studies to quantify stone burden.**

## Approach to Hyperoxaluria

Clinical feature	Primary Hyperoxaluria	Secondary Hyperoxaluria
24-hr Urinary Excretion		
Clinical Presentation		
Presence of Systemic Oxalosis		
History		
Composition of Renal Stones		



## Approach to Hyperoxaluria

Clinical feature	Primary Hyperoxaluria	Secondary Hyperoxaluria
24-hr Urinary Excretion	> 90 mg per day 	< 90 mg per day 
Clinical Presentation		
Presence of Systemic Oxalosis		
History		
Composition of Renal Stones		

# Progression of Primary Hyperoxaluria

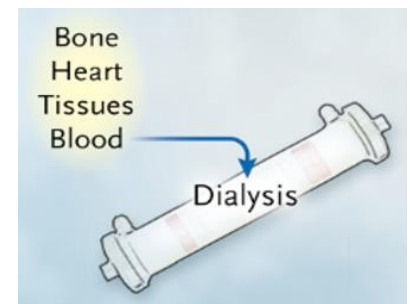
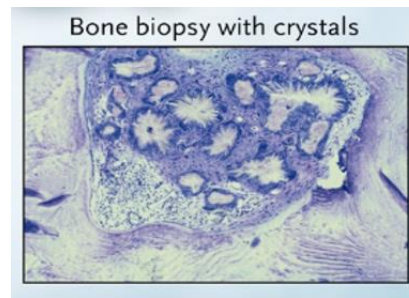
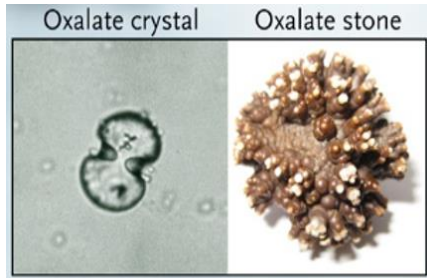
## Stages of CKD by eGFR

CKD 1-3

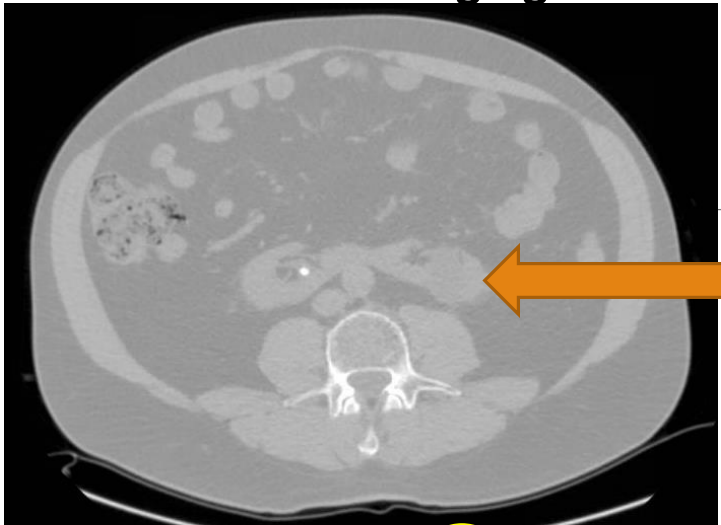


CKD 4-5

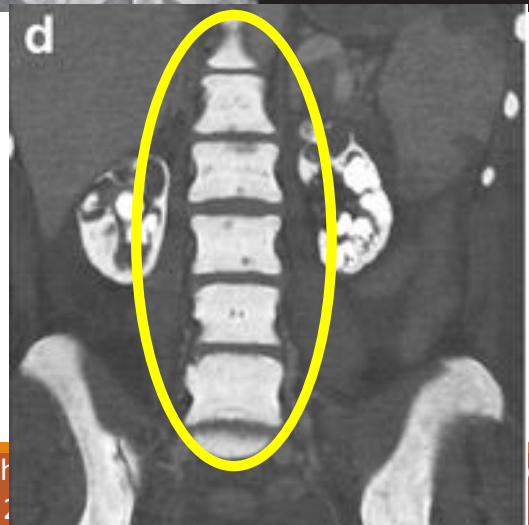
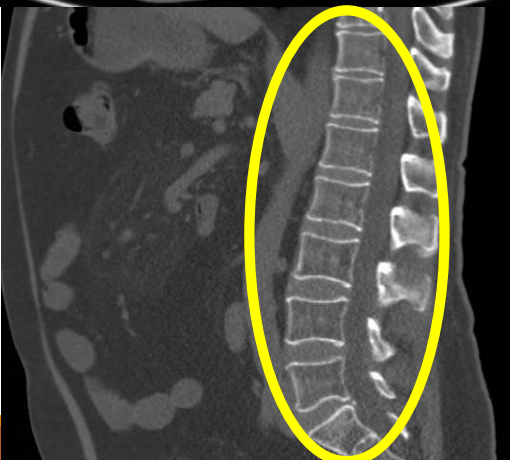
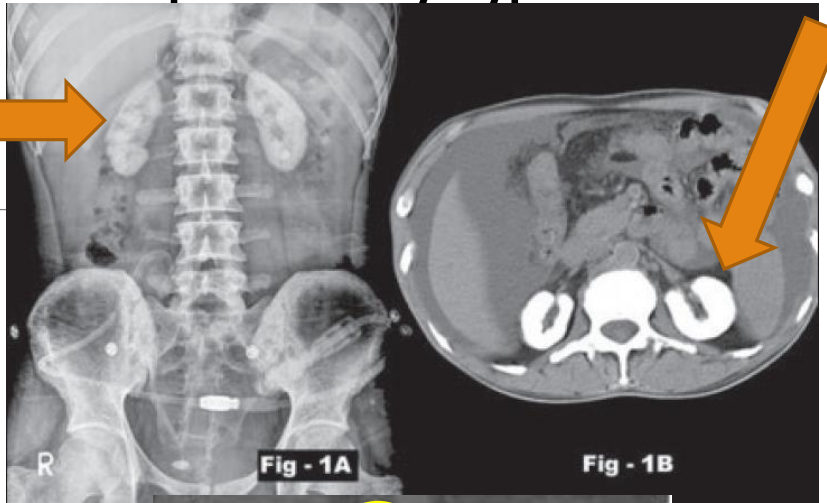
ESRD



# Patient's Imaging

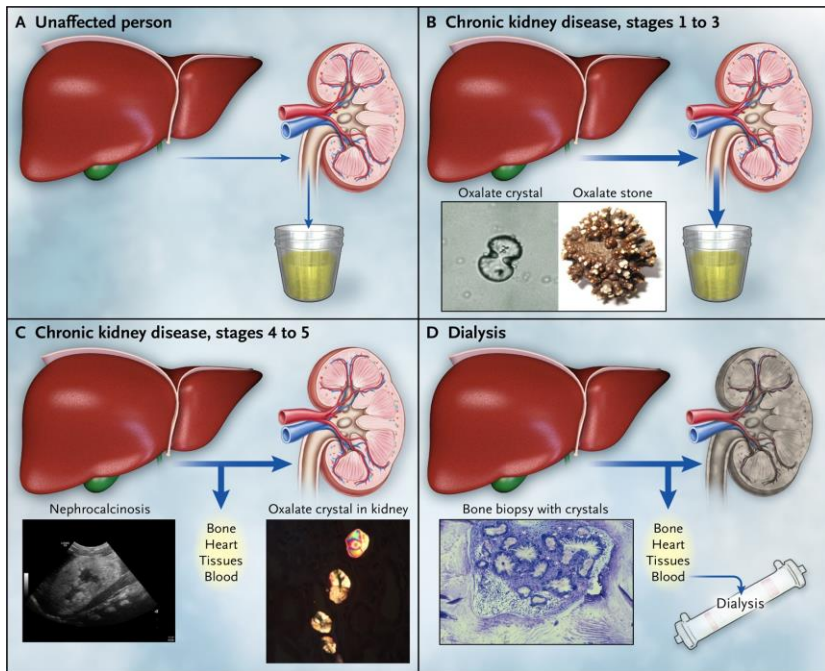


# Example: Primary Hyperoxaluria



## 1:800,000 live births

Risk of our patient developing adult onset  
Primary Hyperoxaluria type 1

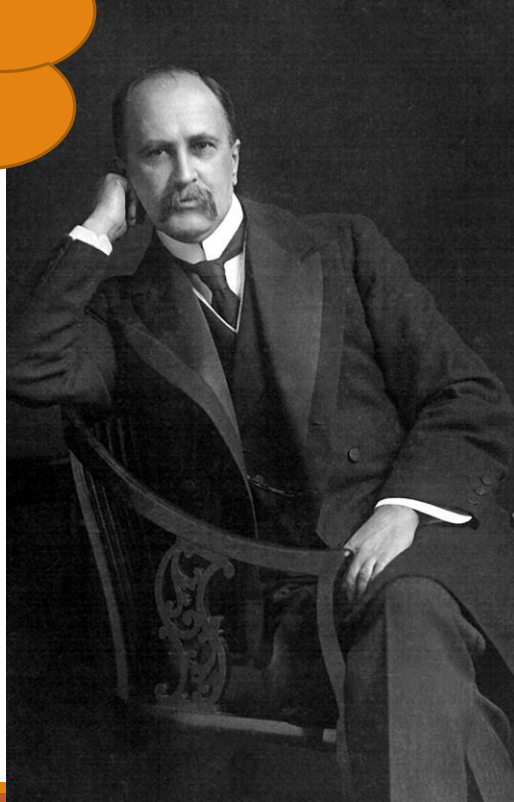


## 1:700,000 lifetime odds







Lifetime risk of being killed by an  
earth/space object collision  
(Alan Harris – Astronomer)









**“Be satisfied with  
probabilities in  
diagnosis”**



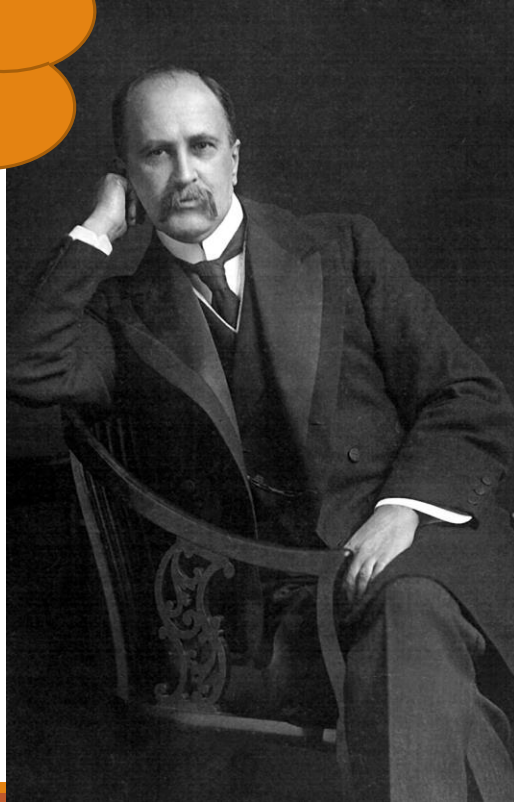
## Approach to Hyperoxaluria

Clinical feature	Primary Hyperoxaluria	Secondary Hyperoxaluria
<b>24-hr Urinary Excretion</b>	> 90 mg per day 	< 90 mg per day 
<b>Clinical Presentation</b>	<ul style="list-style-type: none"> <li>-Usually before 5<sup>th</sup> decade</li> <li>-Recurrent stones </li> <li>-Nephrocalcinosis</li> <li>-ESRD or ESLD common</li> </ul>	<ul style="list-style-type: none"> <li>-Recurrent renal stones </li> <li>-Occasional Nephrocalcinosis</li> <li>-Occasional CKD and ESRD</li> </ul>
<b>Presence of Systemic Oxalosis</b>	Frequent part of the presentation 	Less common 
<b>History</b>		
<b>Composition of Renal Stones</b>		

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<b>Presence of Systemic Oxalosis</b>	Frequent part of the presentation 	Less common 
<b>History</b>	Family history is often suggestive with other affected relatives	History of GI tract disease or dietary history may suggest cause
<b>Composition of Renal Stones</b>		

**“Listen to the patient,  
he is telling you the  
diagnosis”**





# Detailed Pati

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## Family:

- Has a horse
- No history of liver, or

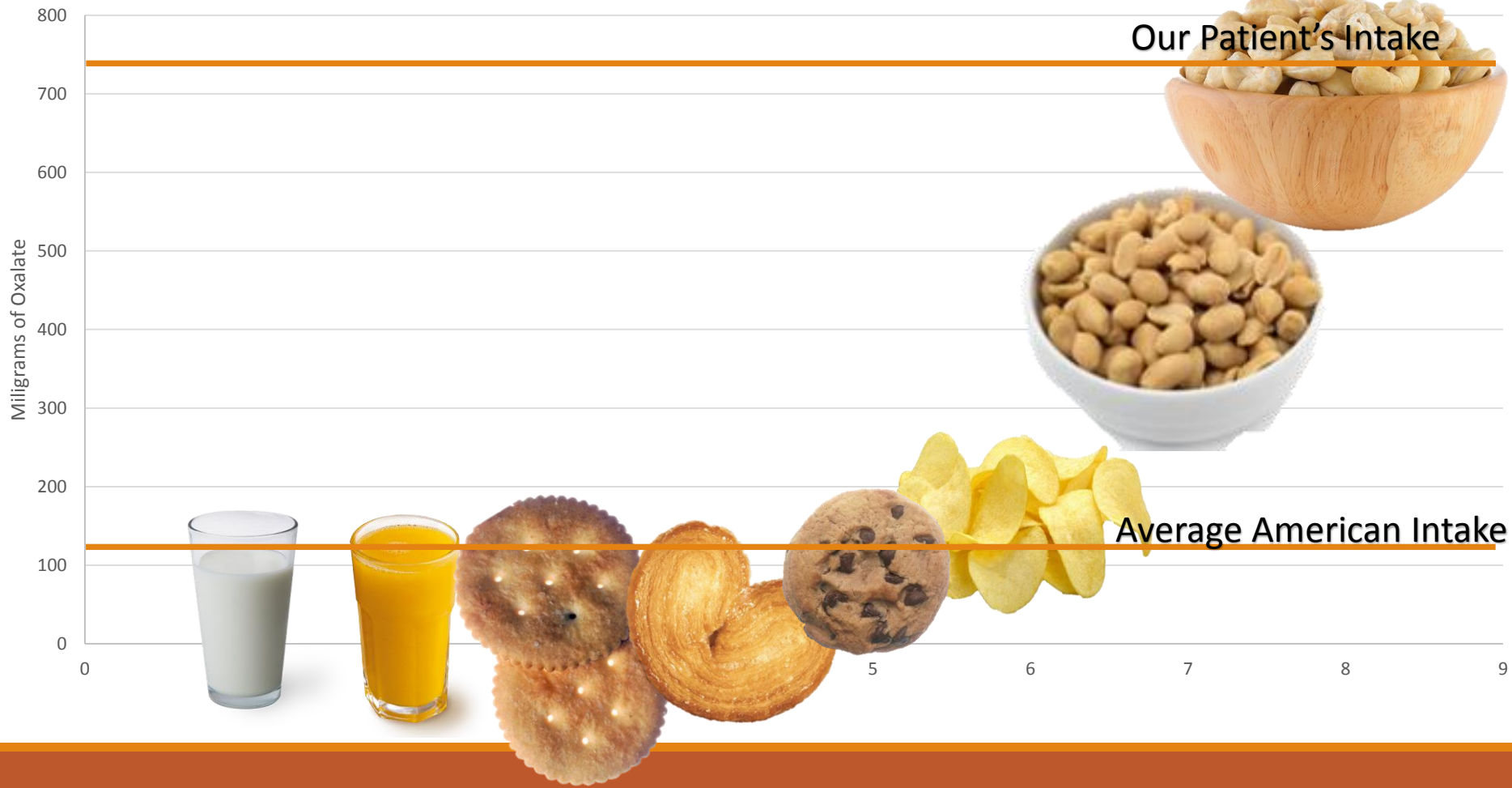
- I haven't eaten meat in decades
- I eat a bowl of cashews and a bowl of peanuts everyday for protein
- I will also eat potato chips, and sometimes some veggies

and



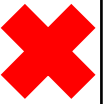

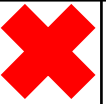



is mostly

and cigarettes, consumed alcohol, or used drugs.



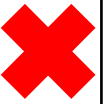

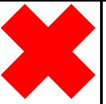



# Our Patient's Dietary Journal: Cumulative Daily Oxalate in milligrams



## Approach to Hyperoxaluria

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<b>Composition of Renal Stones</b>		

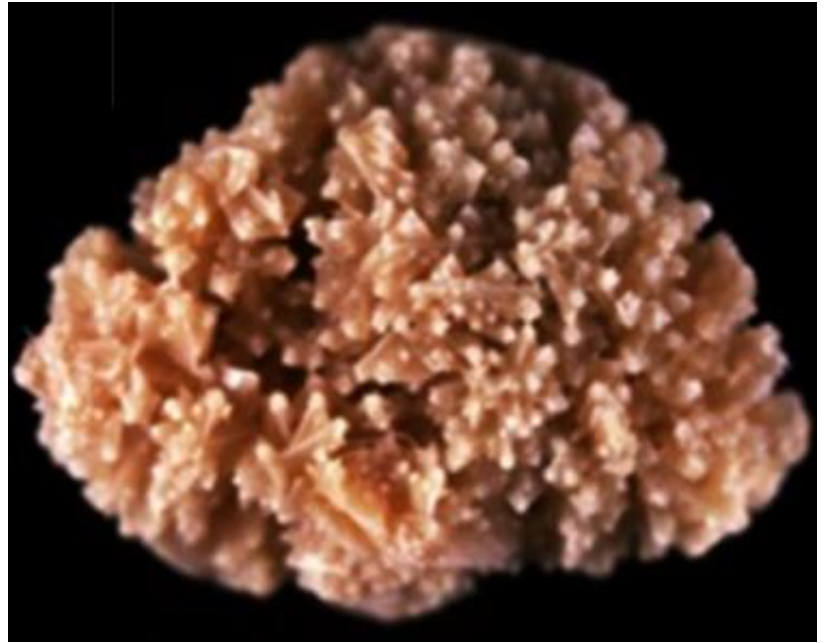
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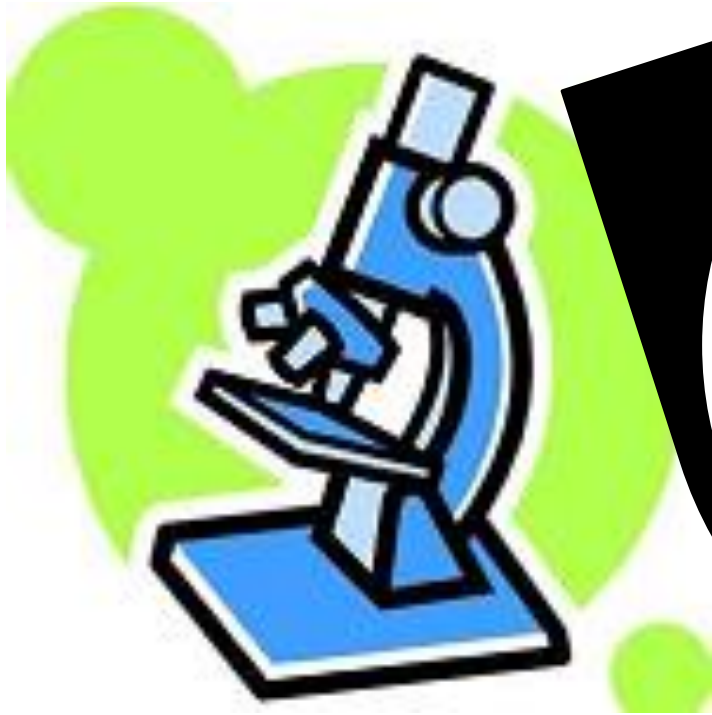
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<b>Composition of Renal Stones</b>	95% calcium oxalate monohydrate (whewellite)	Mixed stones (whewellite and weddellite)

**Calcium Oxalate Monohydrate**  
(Whewellite)





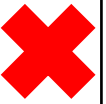

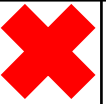



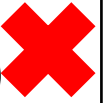

**Calcium Oxalate Dihydrate**  
(Weddelite)





**Mixed stone:**  
-80% calcium  
oxalate  
monohydrate  
- 20% calcium  
oxalate dihydrate.

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# Hypothesis & Interventions

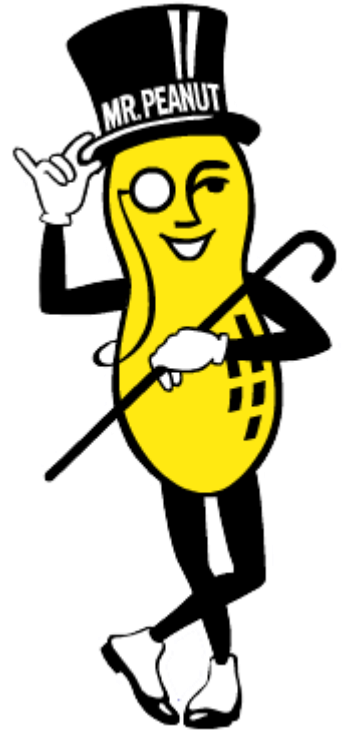
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- Very-high lifelong oxalate intake
- Horseshoe kidney
- Progressive CKD
  - Anatomic disease
  - Possible oxalate crystal nephropathy
- Inability to handle oxalate secretion thus leading to stone formations
- Oxalate restricted diet
  - Stopped cashews, cut down on peanuts
- Recommended Calcium Carbonate 1-2 tabs with meals
  - Oxalate binder
- Re-check 24-hour urine studies



<b>DATE</b>	<b>24-Hour Oxalate Level</b>
Pre-Interventions → 11/16/17	460 mg
Post-Interventions → 8/28/18	80 mg
<b>REFERENCE RANGE</b>	20-40

That's  
Nuts!



# Take Home Points

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- A systematic approach can make sense of diagnosis in unfamiliar disease processes
- Patients with recurrent episodes of nephrolithiasis should be evaluated by:
  - 24-hour metabolic urine studies
  - Stone analysis
  - Dietary History
- The majority adult-onset hyperoxaluria will be due to secondary causes
- Patients with oxalate stones should be counseled to avoid high oxalate containing foods such as leafy greens, chocolate, potatoes, and nuts.