What is Hyperoxaluria?

Hyperoxaluria occurs when your body produces too much oxalate in your urine. High levels of oxalate are toxic because oxalate cannot be broken down by the body and accumulates in the kidneys, causing kidney stones.

The disease spectrum extends from recurrent kidney stones, nephrocalcinosis, and urinary tract infections to chronic kidney disease and can lead to kidney failure.

Hyper (too much)

oxal (oxalate)

uria (in the urine)

What Causes Hyperoxaluria?

There are several causes of hyperoxaluria:

PRIMARY HYPEROXALURIA (PH) *(comes from within)-* which means that you have a genetic disorder

ENTERIC HYPEROXALURIA (EH) *(entering the body)-* is characterized by elevated urinary oxalate excretion due to increased gastrointestinal oxalate absorption:

IDIOPATHIC HYPEROXALURIA *(unknown)-* which means that we are still unsure of what exactly causes this form of hyperoxaluria

Types of Primary Hyperoxaluria (PH)

In addition to enteric hyperoxaluria and idiopathic PH, there are 3 known types of PH. PH is a family of rare, genetic liver disorders that can damage the kidneys. The liver normally makes proteins, called enzymes, that prevent the body from making too much oxalate. In PH, the liver doesn't create enough of this enzyme, or the enzyme doesn't work properly.

Primary hyperoxaluria can present at any age and at any time. Historically, PH has been underdiagnosed. Recurrent kidney stones in adults or any kidney stone in a child is usually the most common sign that you might have PH.

PH1	PH2	PH3
AGXT genetic mutation ↓ **AGXT enzyme deficieny**	**GRHPR genetic mutation** ↓ **GR/HPR enzyme deficiency**	**HOGA1 genetic mutation** ↓ **HOGA1 enzyme deficiency**

Types of Enteric Hyperoxaluria (EH)

1. Fat malabsorption – several intestinal diseases, such as Crohn's disease or short bowel syndrome following bariatric and gastric bypass surgical procedures and/or complications, increase the absorption of oxalate in foods, leading to increased levels of oxalate

2. Dietary – this means eating large amounts of foods high in oxalate can increase your risk of hyperoxaluria

Prevalence of Hyperoxaluria

Approximately 1 to 3 of every million people have PH. Type 1 is the most common form, accounting for approximately 80% of cases. Types 2 and 3 each account for about 10% of cases

The prevalence of hyperoxaluria has been estimated at 5-24% of all patients with gastrointestinal diseases associated with malabsorption. EH is a frequent complication of inflammatory bowel diseases (IBD), ileal resection and Roux-en-Y gastric bypass (RYGB) and is well-known to cause kidney stones, and nephrocalcinosis. EH can also contribute to chronic kidney disease (CKD) and kidney failure.

For more information about hyperoxaluria, visit **www.ohf.org**.

Oxalosis & Hyperoxaluria Foundation
STEPPING STONES TO A CURE

Accelerating research, providing hope empowering patients.

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