

**Primary Hyperoxaluria Registry**  
**Date of Summary: April 2009**  
**Patients Enrolled: 203**  
**PH Type**

Type	Number	Percent
PH Type 1	172	85
PH Type 2	16	8
Non PH1/PH2	11	5
Unknown	4	2

**Of the 203 patients in the registry as of April 2009:**

- **172 patients (85%) have Primary Hyperoxaluria Type I**
- **16 patients (8%) have Primary Hyperoxaluria Type II**
- **15 patients (7%) have Primary Hyperoxaluria of an unknown type**
- **There are at least 2 forms of Primary Hyperoxaluria.**
- **Type I is caused by a deficiency of the enzyme Alanine Glyoxylate Transferase (AGT) which is found only in the liver.**
- **Type II is caused by a deficiency of the enzyme Glyoxylate Reductase/Hydroxypyruvate Reductase (GR/HPR) found in the liver and other tissues.**

**Primary Hyperoxaluria Registry**  
**Date of Summary: April 2009**  
**Patients Enrolled: 203**  
**Country**

<b>Country</b>	<b>Number</b>	<b>Percent</b>
ALGERIA	1	1
ARGENTINA	1	1
BRAZIL	1	1
CANADA	9	5
CHINA	1	1
COLUMBIA	1	1
DENMARK	1	1
GERMANY	12	6
GREECE	2	1
INDIA	4	2
JORDAN	3	2
KAZAKHSTAN	2	1
LEBANON	1	1
MEXICO	1	1
NETHERLANDS	1	1
PAKISTAN	1	1
POLAND	1	1
SOMALIA	1	1
SOUTH AFRICA	1	1
SPAIN	5	3
USA	135	73
VENEZUELA	1	1

**Primary Hyperoxaluria Registry**  
**Date of Summary: April 2009**  
**Patients Enrolled: 203**  
**Decade of Diagnosis**

<b>Decade</b>	<b>Number</b>	<b>Percent</b>
1960's	8	4
1970's	18	9
1980's	20	10
1990's	50	25
2000's	107	53

**Of the 203 patients in the registry as of April 2009:**

- **8 patients (4%) were diagnosed in the 1960's**
- **18 patients (9%) were diagnosed in the 1970's**
- **20 patients (10%) were diagnosed in the 1980's**
- **50 patients (25%) were diagnosed in the 1990's**
- **107 patients (53%) have been diagnosed since the year 2000**

**Primary Hyperoxaluria Registry**  
**Date of Summary: April 2009**  
**Patients Enrolled: 203**  
**Symptoms and Findings at Presentation**

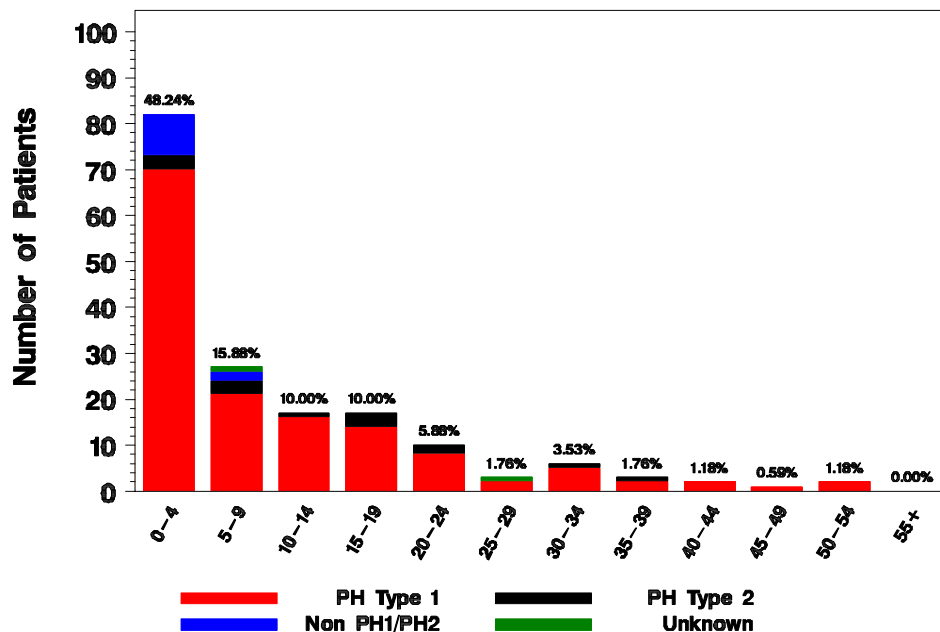
<b>Symptoms and Findings</b>	<b>Percent Present</b>
Urolithiasis	84
Hematuria	36
Abdominal pain	53
Asymptomatic	15
Genetics/family history	80
End stage renal failure	25
Nephrocalcinosis	41

**Of the 203 patients in the registry as of April 2009:**

- **84% of the patients had a history of urolithiasis (kidney stones)**
- **36% of the patients had a history of hematuria (blood in the urine)**
- **53% of the patients had a history of abdominal pain**
- **15% of the patients were asymptomatic (had no symptoms at all)**
- **80% of the patients had a family history of relatives affected by PH**
- **25% of the patients were already in end stage renal failure**  
**(End stage renal failure means the patient's kidneys no longer work and they need dialysis or kidney transplantation.)**
- **41% of the patients had nephrocalcinosis**  
**(Nephrocalcinosis is when the kidneys are extensively filled with calcium oxalate crystals that can often cause loss of the kidney.)**

### Age in Years at First Symptoms

N=203

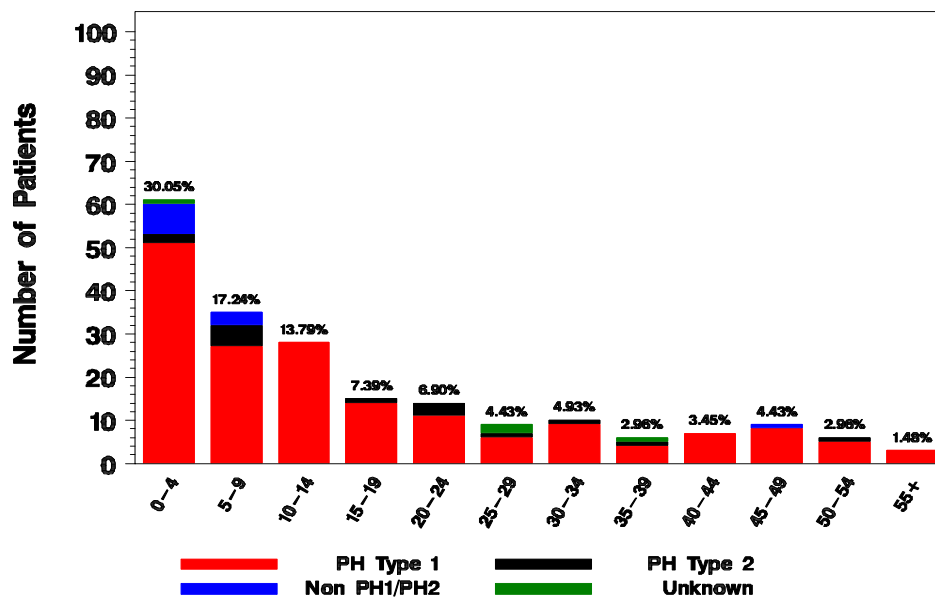


#### Of the 203 patients in the registry as of April 2009:

- More than 80 patients had their first symptom between the ages of 0-4
- More than 20 patients had their first symptom between the ages of 5-9
- Approximately 20 patients had their first symptom between the ages of 10-19
- Most patients have symptoms before the age of 25
- Type of PH is indicated by color

### Age in Years at Diagnosis

N = 203

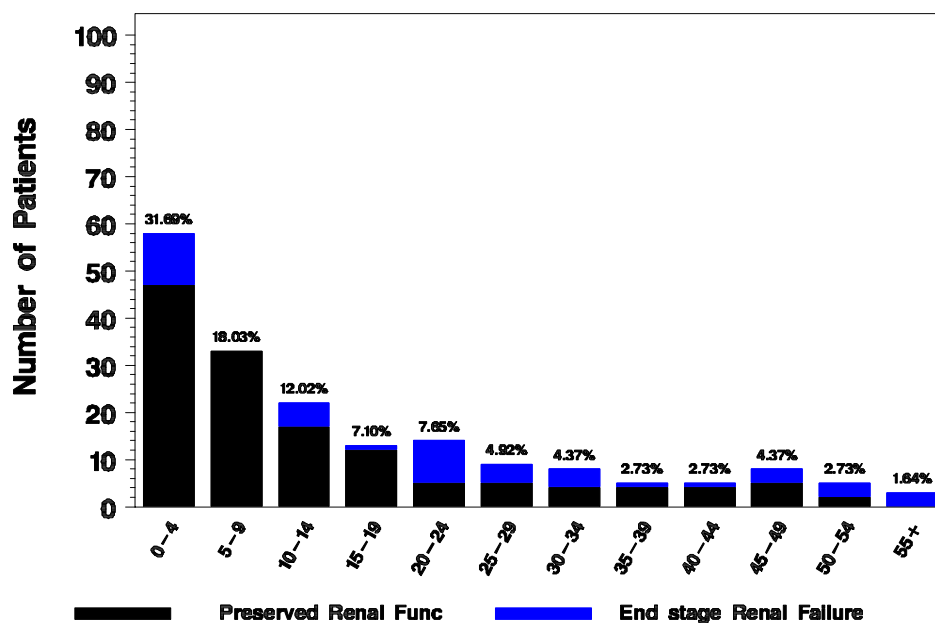


#### Of the 203 patients in the registry as of April 2009:

- 61 patients were diagnosed between the ages of 0-4
- 35 patients were diagnosed between the ages of 5-9
- 54 patients were diagnosed between the age span of 10-39
- The chart shows that most cases of PH are diagnosed before the age of 50 with a large percentage of diagnoses made before a patient's 25th birthday
- Many patients are diagnosed before age 10
- Type of PH is indicated by color

## Renal Status by age at Diagnosis

N = 203



### Of the 203 patients in the registry as of April 2009:

- The majority of patients have functioning kidneys at the time of diagnosis
- However, if a patient is not diagnosed until after age 25, they are more likely to have kidney failure at the time of diagnosis.