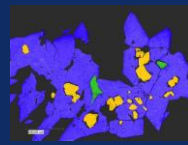


Drug treatment of PH





Specific Treatment

Daily fluid intake > 3 Liter

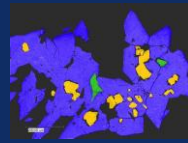
–Via (naso)gastral tube in small infants and children

Pyridoxine in PH type I

Alkaline citrate or orthophosphate

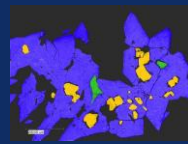
Magnesium

Avoid diet with high oxalate content



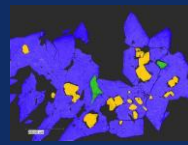
Treatment in normal kidney function

1. Fluids



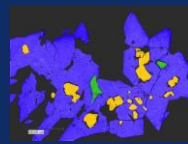
Drink as much as you can!

**All other treatment efforts
are „worthless“, if daily
fluid intake is
insufficient!**



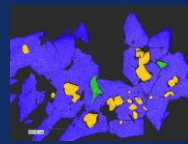
Drink, drink, drink.....

- | Repeated, means continuous daily fluid intake of at least $> 1 - 1.5$ Liter/per m^2 BSA**
- | the earlier young patients accept the necessity to drink the better they get used to it!**
- | Older kids, adolescents and adults should drink $> 2-3$ liter per day.**



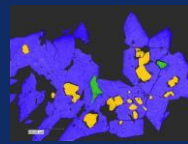
...drink...

- | Increase fluid intake at great heat, fever, fluid loss, e.g. severe diarrhea**
- | If oral fluid intake is no longer sufficiently possible, prompt i.v. (re-)hydration is necessary!**
- | A gastrostomy tube may sometimes be helpful in small infants and children**



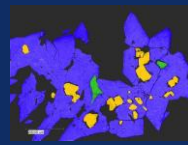
Diet?

Food	Ox content mg/100 g	Food	Ox content mg/100 g
Beans (fresh)	43.7	Cacao powder	623
Spinach (cooked)	356-780	Coffee	1.0
Rhubarb	537	Coffee powder	57-230
Potatoes (cooked)	14.5	Beer	1.7
Apple	1.5	Wine	3.1
Orange	6.2	Tea (2 Min.)	7.0-10.8
Strawberry	15.8	Tea leaves	375-1450



Treatment in normal kidney function

2. Pyridoxine in primary hyperoxaluria type I



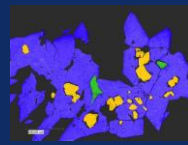
Dosage?

Vitamin B6 should be administered in every patient with PH I

Result of liver biopsy, or molecular genetic testing may give evidence on therapeutic efficiency

Dosage: 5-20 (30) mg/kg body weight/day

Attention: side effects



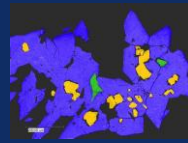
Treatment control?

| 24 h urine analysis before/under treatment

| How much reduction of urinary oxalate excretion is achieved?

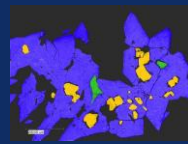
| Stop of treatment when no success?

| In end stage renal failure => control of plasma oxalate levels?!



Treatment in normal kidney function

3. Alkaline citrate/Orthophosphate



Dosage?

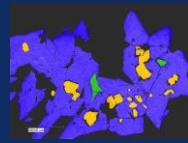
- | Alkaline citrate

- | 0.1-0.15g/kg body weight/d (or 1-1.5 meq/kg/d)

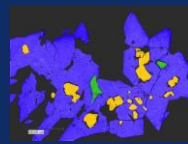
- | Preparations

- | Bicitra

- | Shol's solution



Future treatment options



Oxalobacter formigenes

New phase III study starting 05/2013

Small molecules (chaperones)

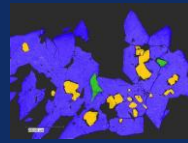
Studies/Research ongoing

Hepatocyte transplantation

Performed in other metabolic diseases

Gene/stem cell therapy

Performed in other metabolic diseases



- | Drink as much as possible
- | (Diet)
- | Increase urine solubility
- | Pyridoxine (in PH I)
- | Hope for new treatment options

