associated infection with antibiotics. If other measures are ineffective, acetohydroxamic acid, a urease inhibitor, can be considered, but its use is limited by significant side effects.

Cystine Stones

Cystinuria is the most common of the rare hereditary kidney stone diseases. It is caused by inherited defects of dibasic amino acid transport in the kidney and intestine. Mutations of one of the two subunits of the amino acid transporter in the kidney leads to defective renal tubular reabsorption of dibasic amino acids such as cystine, arginine, lysine, and ornithine. Cystine stones are the main complication of this defect due to the low solubility of cystine in urine. Characteristic hexagonal cystine crystals can be seen on urine sediment analysis.

The diagnosis of cystinuria is based on a family history of stones, stone formation at a young age, mildly radiopaque stones, and measurement of urinary cystine excretion. Patients with cystinuria excrete 250 to 1000 mg of cystine per day (normal is about 30 mg/day).

Treatment must be aimed at decreasing the urinary cystine concentration by increasing urine volume to more than 4 L/day, alkalization of urine (urine pH >6.5) with potassium citrate or sodium bicarbonate, reducing urine cystine excretion by reducing sodium intake and raising urinary pH by decreasing animal protein intake. Thiol derivatives such as D-penicillamine and α-mercaptopropionylglycine split cystine molecules into two cysteines and produce a highly soluble disulfide compound. However, their use may be limited by their side effects.

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