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Short Communication
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Resolution of a Large Cysteine Stone with Medical Therapy

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Recently an 8-month-old baby presented with irritability, an ultrasonography (US) of abdomen revealed a large Size renal stone (19 mm) located in lower pole of left kidney. He was consulted with a pediatric urologist and meanwhile some investigations for metabolic causes of renal stone were requested. His older brother was a case of renal stone due to cystinuria. The laboratory evaluation revealed a positive sodium-nitroprusside test for cysteine on his urine. The urologist replied that since the stone is non-obstructive we can wait and give an opportunity for medical therapy. I started poly citrate potassium (poly-citra K), D-penicillamine, captopril and vitamin B6 for him with an appointment for an US 6 weeks later.

Six weeks later US revealed that the stone size has decreased to 12 mm and the irritability had decreased. His medications were continued and he was asked for another US for 3 months later. Surprisingly in this time no stone was detected in US. No side effects of D-penicillamine or other medications were observed during these 4.5 months and thereafter in continuation of treatments in his one year follow up.

Cystinuria is a defect in amino acid transport in proximal renal tubules that is characterized by excessive excretion of cysteine and other dibasic amino acids lysine, arginine, and ornithine. Only excessive excretion of cysteine causes renal stone in affected individuals [1]. Cystinuria is inherited as autosomal recessive and accounts about 8% of renal stones in children and 1% of renal stone in adults [2]. More than 50% of Homozygote individuals are subject to symptomatic or asymptomatic stone formation which is bilateral in 75% of the cases. Symptoms of stone are not different from other causes (renal colic, infection and obstruction). Cases of acute renal failure due to bilateral uretero pelvic junction obstruction by stone have been described in infants with cystinuria [3].

Regarding the treatment of cysteine stones, surgical and interventional modalities are the same as other causes of renal stone disease. Modalities other than hydration includes low sodium and low methionine diet, urinary alkalinization to have a urine PH between 7-7.5 and < 8 (at PH 8 and higher risk of calcium phosphate stone formation), chemolysis either direct chemolysis or medical chemolysis by thiol compounds [4-6]. Thiol compounds includes: D-penicillamine, alpha-mercaptopropionylglycine (MPG), and captopril, they combine with cystine and forms a disulfide bond with cystine (cysteine complex) which is more soluble than cystine. Bucillamine (2-mercapto-2-methylpropanoyl-L-cysteine) is a di-thiol compound available and used in Japan and South Korea and proposed to be more effective than Mono-thiol compounds [7]. The more recent advances in prevention of L-cystine kidney stones are use of Crystal growth inhibitors [8].

In this infant with the mentioned large renal stone and the difficulty of interventional approaches for this age from the beginning all the available medications started simultaneously and fortunately the success was unpredictably amazing.

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