

The impact of nurse-led clinics in the management of patients with Cystinuria

Mrs Emily Cooper¹, Dr Graham Lipkin¹, Dr Lavanya Kamesh¹

¹University Hospitals Birmingham Nhs Foundation Trust, Birmingham, United Kingdom

Introduction:

Cystinuria is an autosomal recessive disorder that affects reabsorption of cystine in the proximal convoluted tubules and lead to renal calculi. Therapy to reduce stone formation is directed towards maintaining high fluid intake and alkalinisation of urine. Addition of cystine binding chelating compounds is reserved for recurrent stone formers for whom conservative therapies are insufficient. No curative treatment exists for this condition and patients quality of life can be affected due to lifelong risk of stone formation, multiple operative procedures and impaired renal function. This often leads to poor concordance. We present our experience of specialist nurse-led monitoring clinics that focus on patient education and safe monitoring of patients when chelating agents are used.

Methods:

We retrospectively reviewed the data of all patients with cystinuria attending the renal metabolic clinic (n=37). Patient demographics, renal function, stone burden, number of interventions and urine cysteine levels were noted. In addition, the side effects of the medications were also recorded. All patients attending the monitoring clinic were given advice on high fluid intake. Urinary alkalinisation (pH 7-7.5) was achieved with the use of potassium citrate or sodium bicarbonate.

Results:

18 of 37 patients were male (age 17-68 years) and had a follow up between 6 months to 10 years. Renal function, as measured by eGFR was decreased to <30mls/min in 2 patients. Majority of the patients (n=33) used potassium citrate and the rest used sodium bicarbonate (n=3) for urinary alkalinisation. Fifteen patients used chelating agents (D-penicillamine n=13; Tiopronin=2). These drugs were generally well-tolerated. Only 1 patient could not tolerate either of the chelating agents due to gastro-intestinal side effects. The side effect profile included skin rash (n=1) and proteinuria (n=1) which settled with conversion from D-penicillamine to Tiopronin. The hematological and liver parameters checked in the monitoring clinic were stable. In this group of patients on chelating agents, the urinary cysteine levels reduced to solubility range in 11/15 patients (73%) and the overall mean number of surgical procedures/patient/year was 0.33.

Conclusion:

Nurse-led clinics for patients with cystinuria allow patient education and safe monitoring for chelating agents. It encourages compliance and could potentially reduce morbidity related to recurrent stone formation. Chelating agents are well-tolerated and reduce the risk of stone formation.