Summary for Cystinuria and Hyperkalemia/adrenal insufficiency

1. Prevention and treatment for cystine stones: low salt diet, avoidance of high protein diet, high UOP (over 3 liters/day), urine alkalization goal pH above 7 with potassium citrate or sodium citrate (be cautious of high sodium load) or other citrate supplements, chelating agents。
2. The most commonly used chelating agent is Tiopronin (brand name: Thiola). Dosage: 1-3gm/day in divided doses. Side effects: abnormal LFTs and proteinuria. The mechanism of proteinuria is unclear with case reports of membranous GN. Consider ACEi/ARB to treat mild proteinuria and continue the medication for stone prevention.
3. Pseudohyperkalemia can occur in severe thrombocytosis or leukocytosis (CLL). In thrombocytosis, serum K is high but plasma K is normal. In CLL, both serum and plasma K are high. Details illustrated in Uptodate (Causes and evaluation of hyperkalemia in adults).
4. Major determinants of renal potassium excretion depends on distal sodium delivery, urine flow rate, plasma K concentration, aldosterone level, arginine vasopressin level and acid base. The details are illustrated in attached reviews.
5. TTKG is used in hyperkalemia evaluation to assess urinary potassium excretion. However, it is fallacious because the formula did not take into account the urea recycling which is thought to play a role in distal K secretion. The details are illustrated in attached article.
6. Causes of adrenal insufficiency: autoimmune addrenalitis, infectious addrenalitis, hemorrhagic infarction, metastatic disease, drugs, severe inflammatory disease.
7. Heparin has direct toxic effect on adrenal zona glomerulosa cells, causing decrease in aldosterone level and subsequent hyperkalemia.
8. Pseudohypoaldosteronism type 2 (Gordon's syndrome): hypertension, hyperkalemia, metabolic acidosis, normal renal function, and low or low-normal plasma renin activity and aldosterone concentrations. It is caused by abnormalities in two serine/threonine kinases, WNK1 and WNK4. It is also considered as a mirror image of Gitelman’s syndrome. Thiazide diuretic is the treatment of choice to inhibit sodium reabsorption in DCT.