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Introduction & Objectives: Cystic fibrosis (CF) is characterized by chronic pulmonary disease, insufficient pancreatic and digestive function, and abnormal sweat concentration. Patients with cystic fibrosis also have an increased incidence of nephrolithiasis. Furthermore, there is controversy about predisposing factors of nephrolithiasis. Here we report our experience.

Materials & Methods: A 19 yrs boy with destructive airway with chronic *Pseudomonas Aeruginosa* infection and liver and pancreas failure was in follow up at Cystic Fibrosis Unit of our Hospital . He was admitted to our department for fever and right flank pain. Non contrast-enhanced computed tomography (NCCT) showed bilateral urolithiasis: right upper ureteral stone (diameter 2 cm) with hydronephrosis and left lower calix stone (diameter 1.6 cm). Right tube nephrostomy was quickly placed, followed by ureteroscopy (URS) with ureteral stent Double J placement. Stone free of right ureteral stone was achieved after a second ureteroscopy: ureteral stent Mono J was placed and removed 24 hours later. Besides, a metabolic evaluation performed on 24-hour urine showed hyperoxaluria (45 mg), hypocitraturia (123 mg) and hypericosuria (485 mg); urinary pH was 5.3. The patient started allopurinol and potassium citrate therapy and was discharged with planned retrograde intrarenal surgery (RIRS) for left lower calix stone. After 15 days he was re-admitted to our department for left flank pain. NCCT showed left calix stone migrated in the upper ureter. Laser ureteroscopy for stone removal was performed; ureteral stent DoubleJ was placed and removed 2 weeks later in the clinic.

Results: At 3 months of follow-up sonogram showed small fragments in the right lower calix.

Conclusions: Endourological procedures are safety and effectiveness for the treatment of urolithiasis. Urinary abnormalities promoting stone formation are common in patients with cystic fibrosis and a metabolic evaluation is necessary. Hyperoxaluria could be multifactorial (fat malabsorption, multivitamin supplementation, the absence of *Oxalobacter formigenes* induced by prolonged widespread used of antibiotics). Correcting detected stone risk factors may decrease stone recurrence.