INTRODUCTION: Primary hyperoxaluria type I is due to enzyme deficit of alanine-glyoxylate aminotransferase (PH type I) specific to hepatic peroxisome, causing excessive production of oxalate. Because oxalate is eliminated by the kidneys, the kidney is the first affected organ and manifests with the appearance of lithiasis, nephrocalcinosis and early renal failure. CASE PRESENTATION: Son of consanguineous parents, with a maternal and paternal family history of renal lithiasis and renal failure and sister who died at 3 months of age due to “cystic kidney disease”. At three months of age he presented urinary tract infection and at three years of age, was found nephrocalcinosis. Extramurally, he was misdiagnosed as renal tubular acidosis type I. At 4 years old, primary hyperoxaluria type 1 was diagnosed after levels of oxalate in urine of 24 hours were found in 216.2 mg of 1.73 m2 and confirmed by the finding of the homozygous mutation c.731T>C (p.Ile244Thr) in the AGXT gene. At the moment, the patient is treated with citrate solution, hydrochlorothiazide and pyridoxine. He does not present cardiac or ophthalmological alterations. He has had hearing loss and he has image diagnostic consistent with nephrocalcinosis associated with stage III chronic kidney disease. CONCLUSION: Hyperoxaluria type I requires a high diagnostic suspicion, in order to initiate early the conservative treatment of the disease, slowing kidney damage and offer the only curative option than is the double hepatic-renal transplant. In Colombia, no cases have been published in children. The presence of lithiasis and / or nephrocalcinosis by oxalates, requires discarding primary hyperoxaluria.