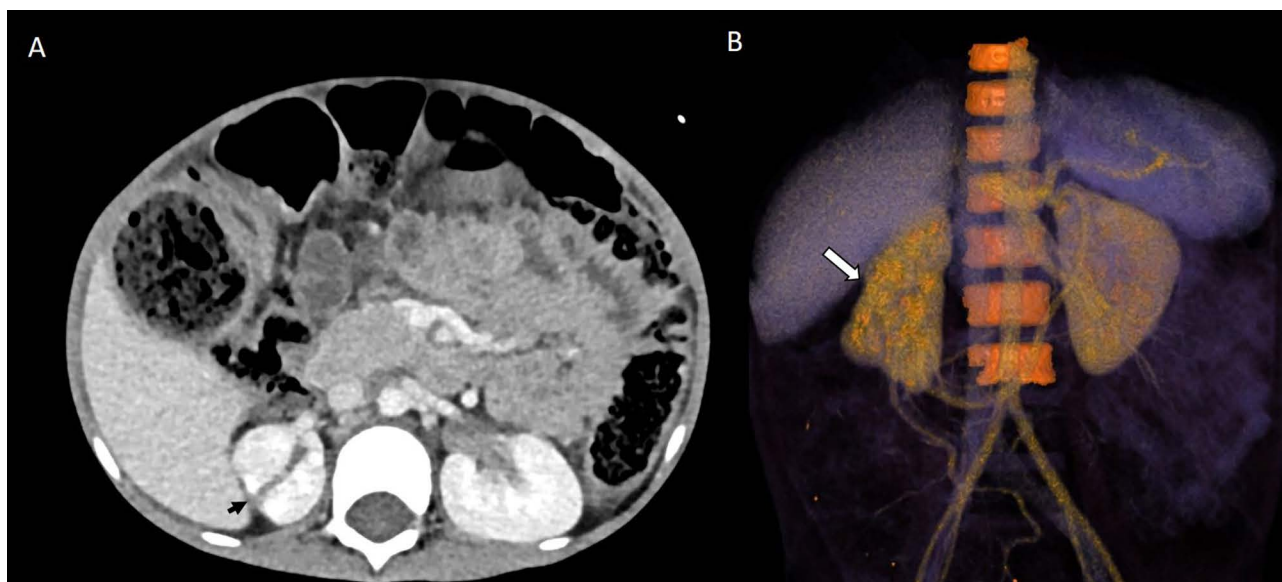


Ask-Upmark Kidney, Imaging Features

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A, axial contrast-enhanced CT image shows linear hypoattenuating scar (arrow) in the interpolar region of right kidney; B, volume-rendered CT image depicts segmental hypoplasia and scar indentation (arrowhead) in the right kidney and normal appearing left kidney.

Renovascular hypertension is known to occur due to various causes. Absence of Renal artery stenosis or changes in renal vascular flow make it difficult to establish a diagnosis of renovascular hypertension in the absence of invasive investigations. Segmental renal hypoplasia is not only a rare cause for renin mediated hypertension but also requires expertise and high index of suspicion while evaluating children with hypertension. We describe the classical imaging findings of a child with secondary hypertension which was due to hypoplasia of a renal segment.

We present classical imaging findings in a child with Ask-Upmark Kidney (segmental renal hypoplasia)

A 10-year old boy presented with hypertensive urgency (BP = 166/120 mmHg), pallor, and constipation. He had a history of primary encopresis and enuresis since childhood. On examination he had short stature and severe anaemia. An initial diagnosis of chronic kidney disease was made and he managed and investigated. However, renal function (blood urea, 15 mg/dL; creatinine, 0.21 mg/dL), serum electrolytes (serum sodium, 134 mEq/dL; serum potassium, 4.7 mEq/dL), calcium/phosphorous/alkaline phosphatase (serum calcium, 8.6 mg/dL; phosphorous, 4.4 mg/dL; ALP, 97) and PTH (24.5 pg/mL) were normal. Low serum iron (15 µg/dL) and high TIBC (567 µg/dL) were suggestive of severe nutritional anaemia. Spinal MRI revealed no cord abnormalities to explain bladder bowel dysfunction, and no reflux was noted on MCU. Renal doppler was reported normal. Pheochromocytoma as a cause of hypertension was ruled out by imaging. As his blood pressure was controlled with ACE-inhibitors (Enalapril) and Amlodipine, renal vein sampling was not done.

CECT abdomen with angiography was performed and it revealed that a scar was present in the interpolar region of right kidney (Figure 1A and 1B). This is a classical imaging finding in a case of Ask-Upmark kidney or segmental renal hypoplasia.

The first description was provided by Eric Ask-Upmark in 1929 as congenital unilateral renal hypoplasia. In the first description of this condition, 5 out of 6 patients were adolescents with malignant hypertension. A distinctive groove on the capsular surface denoting the site of thin hypoplastic segment of the cortex

characterises the kidney structurally in these patients. Either upper, lower or sometimes the entire kidney may get affected.¹ The pathogenesis of the Ask-Upmark kidney is controversial. It has been suggested that the entity originates as a result of congenital defect of renal vasculature that could lead to segmental hypoplasia, absence of glomeruli, and thick arterial wall. Some cases have been reported to be associated with vesico-uretral Reflux, however these may be all absent at the time of diagnosis. Increased Serum renin levels, or rather selective renal angiography and sampling for plasma renin activity may show increased levels and this may attribute hypertension to be renin mediated. However, there are some reported cases where normal renin levels were found. In cases with renin mediated hypertension, unilateral or partial nephrectomy may lead to resolution of symptoms.²

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