The Journal of Medical Research

***Case Report***

JMR 2015; 1(3): 81-82
May- June
ISSN: 2395-7565

www.medicinearticle.com

# Ask-Upmark kidney: Renovascular hypertension in a child; A case report

*Bandya Sahoo*[*]*, Sibabratta Patnaik*

*Department of Pediatrics, Kalinga Institute of Medical Sciences, Bhubaneswar, Odisha, India-751024*

## Abstract

Ask-Upmark Kidney is a rare disease complex which is characterised by segmental renal hypoplasia and is usually associated with hypertension. We report the clinical, laboratory and radiological features of a case, which was managed effectively with antihypertensive drugs. Pathological examination of kidney revealed features consistent with Ask-Upmark Kidney.

**Keywords:** Young Female, Hypertension, Segmental Renal Hypoplasia, Ask-Upmark Kidney.

### Introduction

Renal segmental hypoplasia also known as the Ask-Upmark kidney is a congenital kidney disorder associated with hypertension. It was first described by Eric Ask-Upmark in 1929 who reported it as congenital unilateral renal hypoplasia. It was first reported in 6 patients, 5 of whom were adolescents and presented with malignant hypertension and congenital segmental hypoplasia of the kidney [1].

### Case Report

An 11 years old female was admitted to PICU with hypertensive encephalopathy. On examination blood pressure (BP) was 198/116 mmHg. There was no family history of hypertension or past history of renal disease. Upon investigation the following results were obtained: WBC count was 19.8x $10^9$ cells/L, urinanalysis showed 3-4 WBC/HPF, no haematuria or proteinuria. The blood urea was 17 mg/ dl, creatinine was 0.57 mg/dl. Urine culture was sterile. Lipid profile was normal and antinuclear antibody was negative. Chest radiograph and electrocardiogram were normal.

On abdominal ultrasound the right kidney measured 9.7 x 5 cms, the left kidney measured 8.4 x 3.6 cms, both had a normal cortex and no evidence of hydronephrosis. A micturating cystourethrogram showed no vesicouretric reflux. Tc 99m DTPA scan (Diethylene Triamine Penta Acetate) showed normal sized right kidney with normal function and no evidence of sub-renal obstruction; small contracted left kidney with severely impaired function and atrophy that was more marked in its upper lobe. The glomerular filtration rate(ml/min/1.73 $m^2$) of right kidney was 58.77 and that of left kidney was 11.85. Differential functions (%) of right and left kidney were 83.22 and 16.78 respectively. A Di-Mercapto Succinic Acid (DMSA) scan showed a non-functioning left kidney with normal right kidney function. Selective renal angiography revealed a normal right kidney and hypoplasia of upper lobe of left kidney. The main renal artery on the right showed focal insignificant stenosis and left renal artery was not visualized, suggestive of occlusion or agenesis. Two accessory left renal arteries were seen arising from aorta and supplying middle and lower polar region of left kidney. Color Doppler flow imaging of renal arteries showed increased acceleration time in bilateral intra-renal arteries suggestive of renal artery stenosis. Renal biopsy from hypoplastic areas of left upper lobe showed atrophic glomeruli with thickened vessels and thyroidisation of the tubules. She was treated conservatively with antihypertensive drugs.

### Discussion

Segmental hypoplasia of the kidney is a particular form of renal hypoplasia which is very frequently associated with hypertension. Severe hypertension is the usual presenting symptom. Our patient presented as hypertensive encephalopathy with convulsion. This condition is seen more frequently in females below the age of 12 years [2]. Mild proteinuria is a frequent finding in some series [3, 4]. Urinary tract infection has been reported in some cases. In our case there was no evidence of proteinuria or urinary infection. The occurrence of renal dysfunction depends upon the degree of renal involvement, the severity of hypertension and duration of the disease. Our patient had unilateral left renal dysfunction.

The radiological appearance of Ask-Upmark kidney is characteristic. The intravenous pyelography shows

***Corresponding author:**
***Dr. Bandya Sahoo***
Department of Pediatrics, Kalinga Institute of Medical Sciences, Bhubaneswar, Odisha, India-751024

the contour of the affected kidney to be irregular, lobulated with one or more indentations separating the hypoplastic zones from normal or hypertrophied tissue. Beneath the depressed area, the calyces are often dilated and clubbed. Renal arteriogram done in our patient showed a normal right kidney and a small left kidney with a deep notch at its upper lobe.

Structurally the kidney is characterized by distinctive groove on the capsular surface which denotes the site of thin hypoplastic segment of the cortex overlying an elongated and dilated calyx–like recess of renal pelvis [5]. The hypoplastic segments may affect the upper or lower poles more frequently, sometimes the central portion of the kidney and very rarely the whole kidney.

For final confirmation of diagnosis of Ask-Upmark kidney histological examination is considered mandatory. The tissue separating the capsule and calyx is fibrotic, either aglomerular or shows paucity of glomeruli and contains epithelial tissue lined thyroid-like tubules with thick-walled hyperplastic vessels [2]. Our case showed very few atrophic glomeruli and atrophic dilated tubules with thyroid like appearance. The blood vessels showed marked thickening of the walls and narrowing of their lumens.

The pathogenesis of the Ask-Upmark kidney is rather controversial. It is thought to be congenital or the consequence of vesicoureteral reflux [6]. Some authors attribute the principal cause to vesicoureteric reflux (VUR) with intra-renal reflux. However, not all patients demonstrate a VUR at the time of diagnosis [6, 7]. Our patient did not have VUR. The possibility of localised developmental arrest was also suggested in earlier descriptions with the presence of renal dysplasia in the abnormal segments. However, this alteration in metanephric development could probably be a consequence of an intrauterine VUR [6,7].

The deep scars seen in the Ask-Upmark kidney represent intrarenal reflux into a single lobe. The scarring can be either due to reflux of infected urine or due to Tamm-Horsfall protein in case of sterile intra-renal reflux. The protein may induce obstruction as well as its influx into the renal interstitium may set up a chain of reflux nephropathy [8].

It has been suggested that the renin-angiotensin system plays a role in pathophysiology of hypertension [9]. Increased renin levels have been reported in some patients [10, 11]. Segmental hypoplasia with scarring of the renal parenchyma leads to renin release, secondary hyperaldosteronism and arterial hypertension. It is a curable cause of secondary hypertension [6]. During selective renal angiography blood can be drawn from both renal veins for differential renin assays,which can be done under expert guidance. This procedure was not possible in our institute.

The unilateral Ask-Upmark kidney can be treated with unilateral nephrectomy. For bilateral involvement, medical treatment of arterial hypertension and renal insufficiency is the only option. Our patient currently is being managed conservatively despite the condition being unilateral, since hypertension was controlled effectively with antihypertensive drugs. She is on follow-up for regular monitoring of blood pressure. Nephrectomy may be considered at a later date, if the BP becomes uncontrolled despite medications or if there is deterioration in right sided renal functions.

**Conclusion**

The association of a unilateral small kidney with hypertension is usually attributed to pyelonephritis, primary hypoplasia or unilateral renal artery stenosis, but in such conditions the possibility of Ask-Upmark should also be considered, particularly in young females [9]. The diagnosis is crucial in view of the fact that this is another cause of potentially curable secondary hypertension in the young.

**Conflict of interest** – Nil

**Funding** - Nil

**References**

1. Sobel JD, Hampel N, Kursbaum A, Adler O, Gellet B. Hypertension due to Ask-Upmark Kidney. Br J Urol. 1977 Nov; 49(6):477-80.
2. Rosenfeld JB, Cohen L, Garty I, Ben-Bassat M. Unilateral renal hypoplasia with hypertension (Ask-Upmark kidney). BMJ.1973; 2:217–9.
3. Royer P., Habib R., Broyer M., Novaille Y. : Segmental hypoplasia of kidney in children.AdvNephrol Necker Hosp. 1971;1:145-59.
4. Benz G., willich E., Scbarer K.: Segmental renal hypoplasia in childhood. PediatrRadiol. 1976 Dec 15;5(2):86-92.
5. Bengtsson C.,Falkenbeden T., Hansson L., Hood B. Renal hypoplasia, clinical features and survival. Scand J UrolNephrol. 1970;4(2):117-24.
6. Babin J, Sackett M, Delage C, Lebel M. The Ask-Upmark kidney: a curable cause of hypertension in young patients. J Hum Hypertens 2005 Apr;19(4):315-6.
7. Marwali MR, Rossi NF. Ask-Upmark kidney associated with renal and extrarenal arterial aneurysms. Am J Kidney Dis.1999;33(4):e4.
8. Hodson CJ. Reflux Nephropathy: A Personal Historical Review. Am J Roentgenol. 1981;137(3): 451-62.
9. Prasad S, Kaler AK, Shariff S. Case Report Ask-Upmark kidney: A report of 2 cases. Int J Health Sci Res. 2013;3(1):61-4.
10. Royer P, Habib R, Broyer M &Nouaille Y. Segmental hypoplasia of the kidney in children. AdvNephrol Necker Hosp 1971; 1: 145–59.
11. Arant BS, Jr, Sotelo-Avila C & Bernstein J. Segmental 'hypoplasia' of the kidney (Ask-Upmark). J Pediatr 1979; 95: 931–9.