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The Enemy Within: Renal complications in 13yr old girl with Spina Bifida

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Abstract:*Neural tube defects are associated with complications related to vesicoureteric reflux, urinary incontinence; recurrent urinary infections which all lead to severe renal damage which progress to renal scarring. Renal scarring with renal failure remains life threatening among the children who have a spinal dysraphism problem. In the case of spinal anomalies which include distortion of spinal roots, spinal cord or both are associated with neurological abnormalities which can affect the lower limbs of the patient. Therefore, in order to treat these problems management strategies like pharmacotherapy, psychotherapy, surgical intervention and education can be effective. Moreover, early identification of the disease can be helpful for the treatment process and patients can avoid end-stage of the disease.*

Keywords*: Spina Bifida, Neurogenic bladder, urinary incontinence, neural tube, Meningomyelocele, Vesico-ureteric reflux*

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I. Introduction

Spina Bifida is a disease that leads to urological problems including issues with bowel functions and voiding. Congenital defects of neural tube development can be seen among 1 out of 1000 newborns. 62% of patients have reported having urinary sepsis with Spina Bifida [1]. Therefore, the aim is to reduce the chances of end-stage renal disease by developing the capacity, adequate bowel and bladder continence and low-pressure reservoir [2]. The key reason behind these issues is inappropriate interventions of the pelvic sphincter and urinary bladder. Therefore, the recurrent pyelonephritis and Vesico-ureteric reflex (VUR) has mentioned being the key reason behind renal failure [3]. The goal is to protect the deterioration of kidney function which will progress to end stage kidney disease by ensuring low pressure reservoir with good capacity with good bladder and bowel continence. The neural tube defect causes incomplete innervation of the pelvic sphincter and urinary bladder causing alteration in storage and emptying function with increase in intravesical pressures which lead to kidney damage and renal failure. The management is to keep the intermittent catheterization clean and providing antimuscarinic medication neurologicalbladder [4]. Though it is not a permanent solution as in the adult age the pressure increases in the adult age and may lead to long term complication.

II. Case Study

We present a patient from Coast provincial general hospital (C.P.G.H), 13 year old female patient with history of facial puffiness, generalized body swelling, abdominal pain, reduced urine and elevated blood pressure 180/102mmHg Pulse-90/min. There was also history of bilateral lower limb weakness since birth.

Birth history: she was born preterm via emergency cesarean at 30 weeks of gestation due to pre-eclampsia. She was admitted in New Born Unit (NBU) for 21 days. Birth Weight 1100gms with Spina Bifida Occulta (Closed Spinal Dysraphism) diagnosed at birth and was managed conservatively.

Childhood Milestones: They were delayed sitting at 8 months and delayed walking past 2 years. She was referred for occupational/gait therapy. Currently she walks with support.

Immunization: it's upto date as per the Kenya Expanded Programme on Immunization (KEPI) vaccination schedule.

Infancy and Childhood: Urine incontinence and Nocturnal enuresis reported to age of 13 years. During late childhood she has suffered for recurrent symptom of UTI with frequent abdominal pains, dysuria and occasional hematuria. However no symptoms of neurogenic bowel reported. Normal intelligence and no learning disabilities were reported.

Family Social History: Patient is 1st born of the (triplets), 2nd triplet died on day two due to Acute respiratory distress syndrome(ARDS), 3rd Triplet alive and well. The second pregnancy the child is alive and was diagnosed with Down's syndrome.

Investigations:

- Full Blood Count revealed mild granulocytosis with a microcytic hypochromic anemic picture
- UECs: elevated Creatinine and BUN
- Lipid profile: Hypoalbuminemia, Hyperlipidemia
- Urinalysis: Proteinuria
- E.C.G –Normal heart study

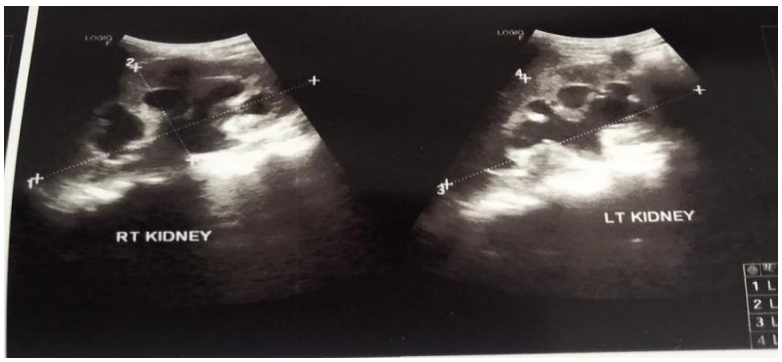

Fig1. Abdominal Pelvic ultrasound: showed bilateral grade 5 hydronephrosis and hydroureter. Urinary bladder had thickened irregular bladder wall with multiple diverticulae.

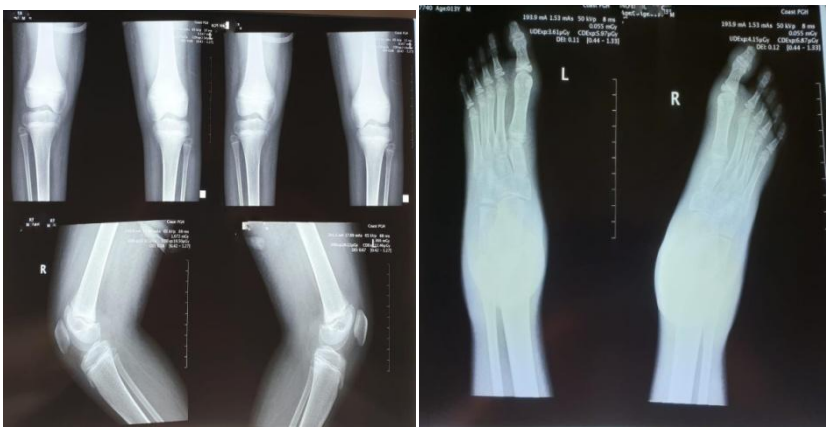

Fig 2. X-ray of the knee and foot are unremarkable normal

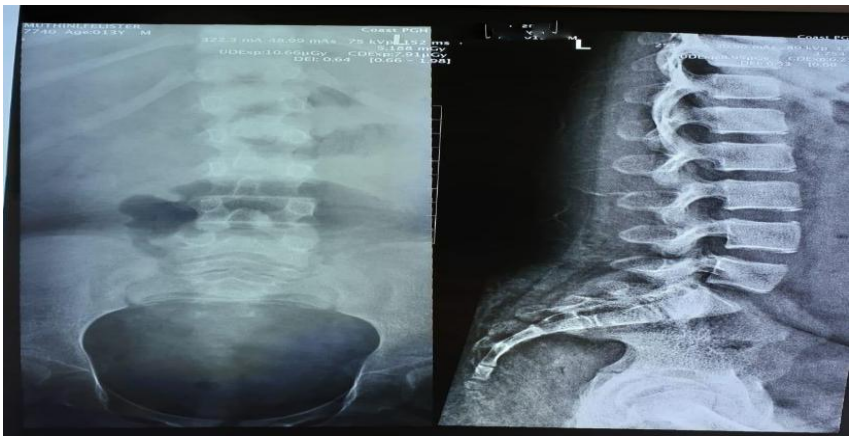

Fig 3. Lumbosacral radiograph shows a defect of the posterior elements of the sacral vertebrae

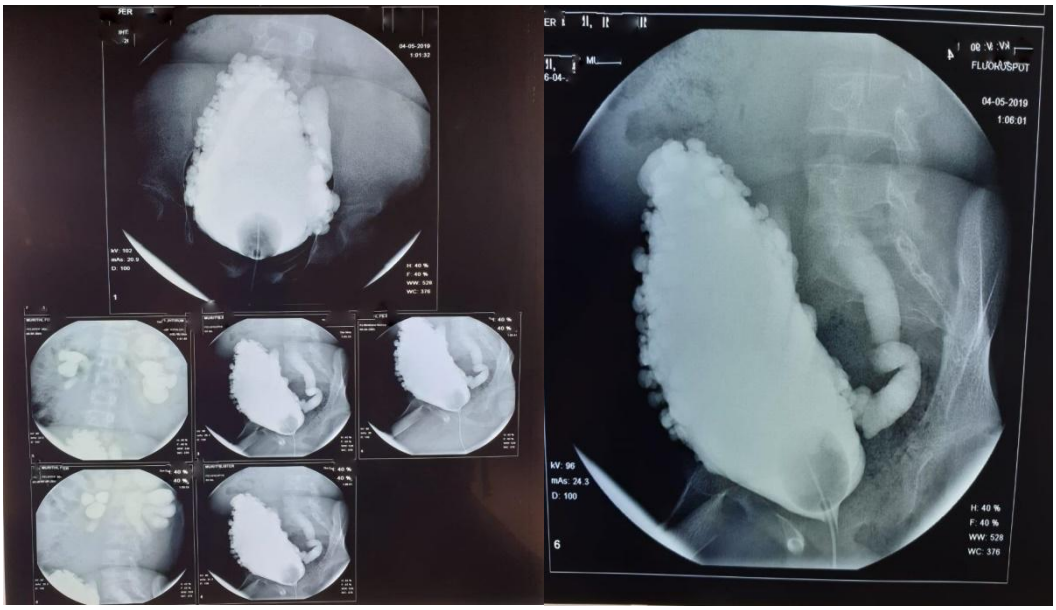

Fig 4. Spot film from MCU series demonstrates a pine tree appearance of the urinary bladder with an irregular wall and multiple diverticulae. Grade 5 vesicoureteric reflux is noted bilaterally with severe dilatation tortuous ureter.

Consent was obtained from the mother to use the photos on the investigations

Pathogenesis

Spina dysraphism cases, 30-50% are associated with Vesico-ureteric reflex (VUR) which may lead to intravesical pressure that occurs during micturition leading to intrarenal reflux which progress to renal scarring [3]. The renal scaring scarring associated with VUR is referred to as reflux nephropathy (RN). The late complications of reflux nephropathy include proteinuria, chronic renal failure and hypertension [19].

The normal ureters usually enter the bladder obliquely through muscular layer and pursue a course in submucosal layer. There is contraction of the detrusor which leads to collapse of the intravesical segments of the ureter, and serves as a valvular mechanism in preventing VUR [2]. This serves as an antireflux mechanism and is related to the length of the sub-mucosal ureter [16]. The congenital shortening of the sub-mucosal ureteral segment results valve dysfunction and VUR.

Prevalence

It has observed that the rate of prevalence is high among infants. 0.4-1.8% of newborns has observed to be prevalent of the disease. On the other hand, the prevalence rate among children is 30-50% and in the case of infants, the rate is 40-50% have Urinary tract infection secondary to VUR [5].During infancy the prevalence is common in boys but the condition is more prevalence in girls.

Consequence of reflux Nephropathy

Reflux nephropathy defines diffuse or focal area of diffuse scarring which is associated with VUR. The consequence of renal scarring in a patient may results in intrarenal reflux of infected urine with interstitial inflammation and damage, high pressure reflux which may damage kidney through immunological or mechanical mechanism and abnormal embryological development with renal dysplasia[6]. The most important risk factor for renal scaring is urinary tract infection.

Since Renal Nephropathy is an acquired condition it requires early detection to prevent the sequelae. The incidence of renal scarring is related to severity of VUR. Reflux Nephropathy seen according to International classification was 85% patients with grade V, 37-64% with grade IV, 25% with grade III and 6-14% with grade II VUR [7]. RN is common cause of sustained hypertension in children about 38% patients with RN shows hypertension [8].Proteinuria and hypertension is important indicator of progressive renal damage and poor outcome.

Clinical presentation

The most common clinical presentation of Reflux Nephropathy and Vesicoureteric Reflux is UTI along with other manifestation.

Vesicoureteric reflux	**Reflux nephropathy**
Urinary tract infection	Urinary tract infection
Hydronephrosis in fetal stage	Hypertension
Associated urinarytract constraints	Chronic renal failure
Urinary calculi screening in peers	Associated UTI problems

Diagnosis

Radio contrast micturiting cystourethrography (MCU) or radionuclide cystography (DRGG) are used in diagnoses of VUR [9]. MCU is preferred as the initial examination in both sexes and enable grading of VUR and also visualize the urethra in boys and structural anomalies such as posterior urethral valves, ureteroceles and bladder diverticula [10]

DRCG is more sensitive than MCU for diagnosis VUR. It also has lower radiation as compared to MCU which makes it ideal for follow up. Limitations include inability to precisely grade the reflux and evaluate urethra during voiding.

Ultrasonography (US), intravenous pyelography (IVP) and radionuclide scanning can be used to show renal nephropathy [11]. The main limitations in IVP are the inability to show information on renal function and failure to show early scars. Caution in patients with impaired renal function.

Antibiotic Prophylaxis

Antibiotic Prophylaxis can be considered as one of the most effective preventive measures for the reduction of colonization and ascending infections along with maintaining sterile urine [12]. Therefore, in this case broad spectrum antibiotics are used they should be able to achieve high urinary concentration with minimal bowel flora alteration.

Drug	**Dosage (mg/kg/ day)***	**Remarks**
Co-trimoxazole	1-2 trimethoprim 50 sulphamethoxazole	Fluid intake limit is maintained. Infant under 6 weeks may avoid this; contraindicated in G6PD deficiency [13]
Nitrofurantoin*(NFT)*	1-2	Considerable GI upset; resistance rare; contraindicated in G6PD deficiency, infants under 3 months, renal insufficiency;
Cephalexin	20	Can be used for young infants with approval of NFT and co-trimoxazole is restricted [14]
Methenaminemandelate	75 in 2 divided doses	Rash; GI upset, not under the medication of co-trimoxazole or in renal impairment

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Surgical Treatment

Surgery as a treatment option has no clear consensus in VUR Management. Surgery of VUR consists of ureteric reimplantation with success rates exceeding 95% with experience surgeon [14, 19, 21]. Major complication of surgery include ureteral obstruction due to kinking, torsion and devascularisation of the distal ureter which may occur up to 4% of patients and reflux may persist in 2.5%[15,20]. Grade V or bilateral grade IV reflux in these children require surgical repair because itsunlikely for it to resolve.

Management

Careful evaluation of patients with vesico-ureteric reflex who have reduced renal function or hypertension must be managed early. Proteinuria which is present is associated with renal scarring and progression to renal nephropathy. Hypertension must be controlled and urinary tract infection treated with patients continues with antibiotic prophylaxis [16]. A restricted dietary intake or protein (1-105/kg per day is advised.Angiotensin converting enzymes (ACE) are associated with reduction of proteinuria and preservation of renal function [11,17]. Patients with chronic renal failure as a result of renal nephropathy should be managed using standard guidelines of chronic kidney disease (CKD). General measures including increase fluid intake and regular complete emptying of the bladder. Constipation should be avoided and also technique to avoid faecal soiling of the periurethral area.

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