CASE REPORT

Prune Belly Syndrome in Adolescence: A Case Report

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Abstract:

The Prune Belly syndrome also known as Eagle Barret syndrome is a rare disorder. We report a rare case of Prune Belly syndrome in 17 year old boy. Patient presented with complains of absence of both testis in scrotum since birth. On examination patient was found to have lax abdominal wall. Patient was further evaluated and found to have shrunken small right kidney and left hydroureteronephrosis and the diagnosis of Prune Belly Syndrome was made. Prune Belly Syndrome represents a wide spectrum of disease. Each patient must be dealt with on an individual basis. A course of watchful waiting with selective surgical intervention has also been successful.

Keywords: Eagle Barret syndrome, Prune Belly Syndrome

Introduction:

Prune Belly syndrome is an uncommon congenital anomaly which is constellation of three major findings i.e. deficiency of the abdominal musculature, bilateral intra-abdominal testes, and an anomalous urinary tract. A deficiency of abdominal musculature gives the patient's abdomen a wrinkled, prune like, appearance. In some infants, the disease is so severe that kidney development and urine production *in utero* are impaired. Subsequent oligohydramnios leads to pulmonary hypoplasia. These severely affected children may be stillborn or die shortly after birth. Children may also present with normal renal function in spite of abnormal radiographic appearance of the urinary tract. These children may grow up without much physical or physiologic impairment. Therefore, each patient must be dealt with on an individual basis.

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17 years male patient presented with absence of both testis since birth. On examination, he had distended abdomen and wrinkled lax abdominal wall (Fig.1). Penis was normal but scrotum was underdeveloped and testes were absent (Fig. 2).

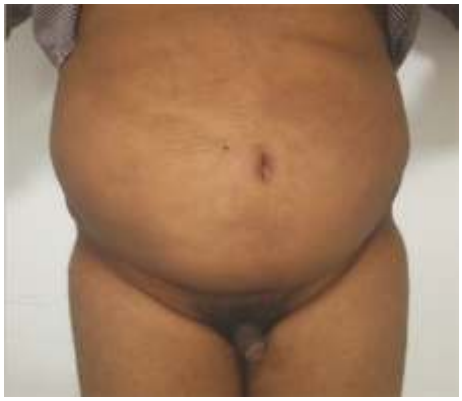

Fig. 1 Wrinkled Lax Abdominal Wall giving Prune Belly Appearance

Fig. 2 Bilateral Undescended Testis

Ultrasonography showed smooth shrunken right kidney and left moderate hydroureteronephrosis. Intravenous Urogram showed left gross hydroureteronephrosis and right kidney was not visualized (Fig. 3).

On voiding cystourethrogram, there was no evidence of vescicoureteric reflux on either side and prostatic urethra was dilated (Fig. 4).

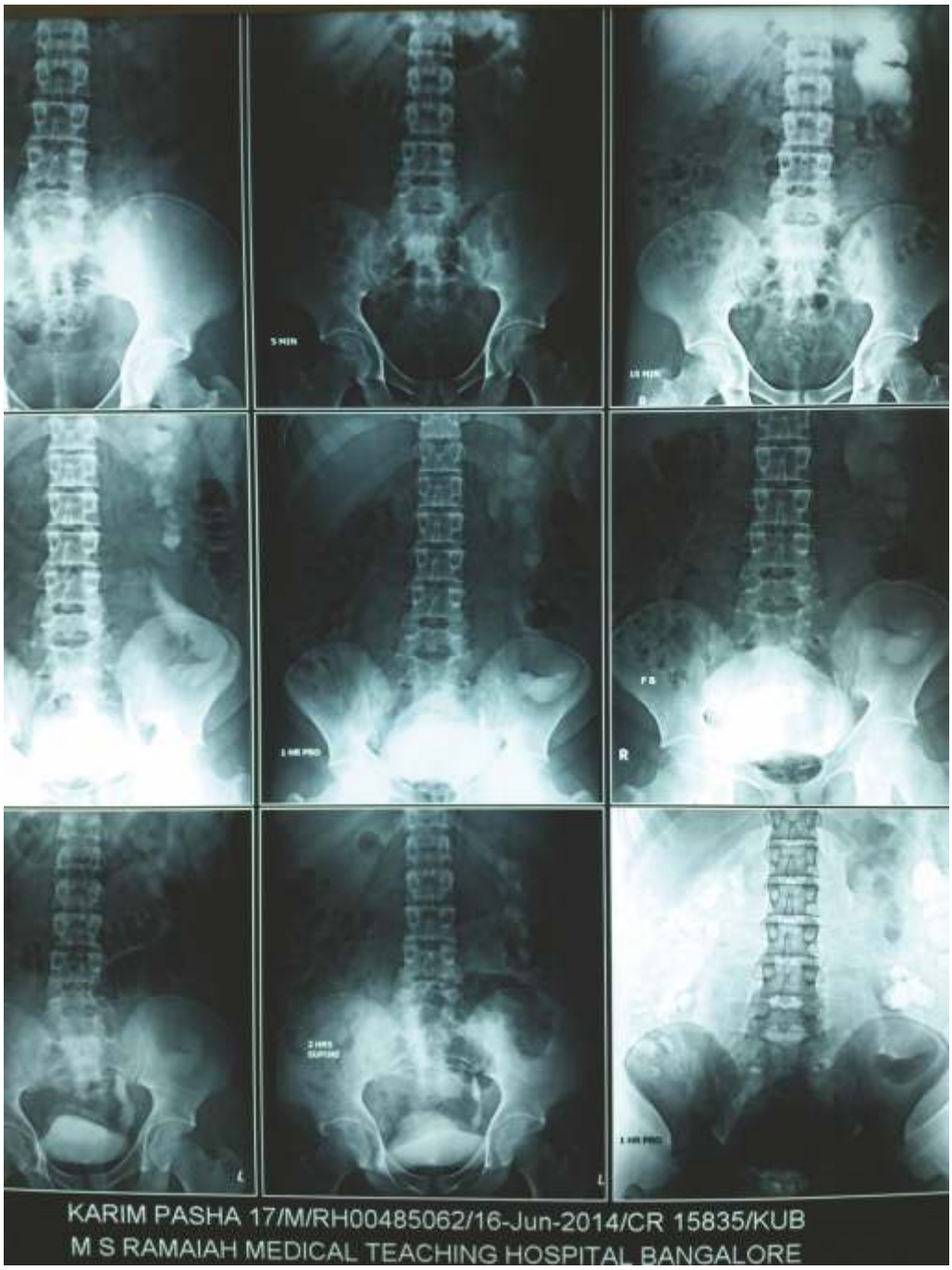

Fig. 3 IVP Showing Left Gross Hydroureteronephrosis and Non-visualized Right Kidney

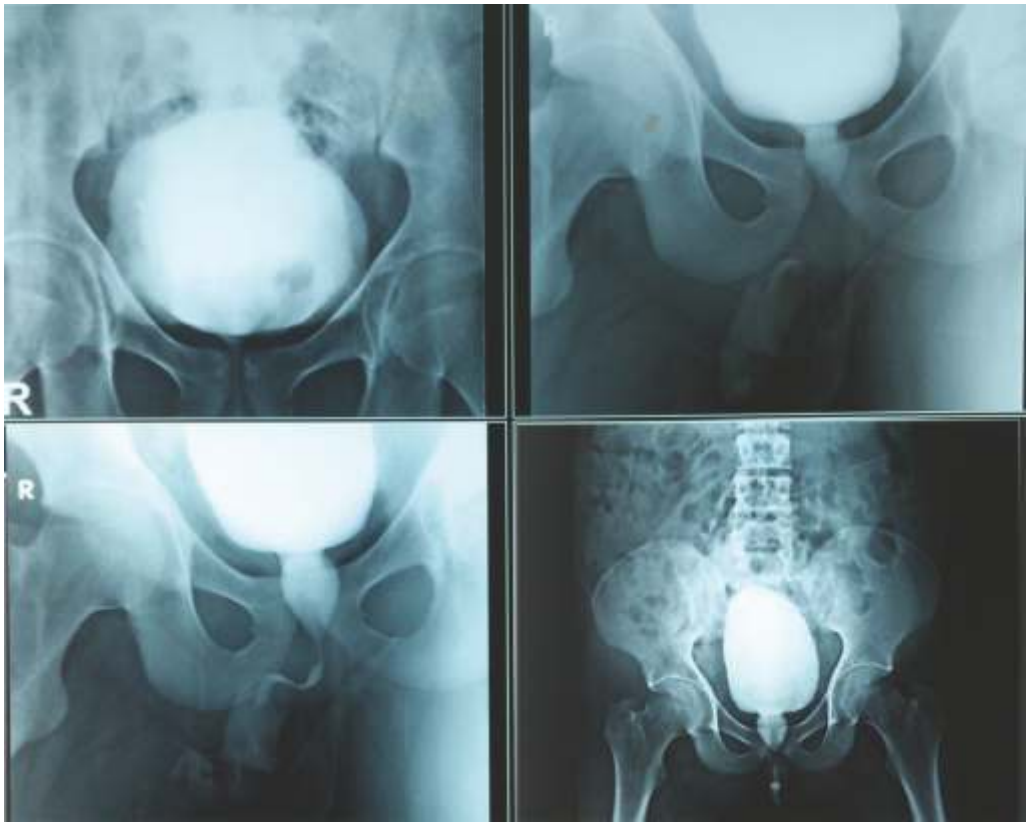

Fig. 4 MCU Showing Dilated Prostatic Urethra with No Evidence of Vesicoureteral Reflux

Diethylene Triamine Pentaacetic Acid (DTPA) renal scan was suggestive of well perfused normal functioning left kidney. Right kidney was non perfused and not visualised. Urodynamic study showed normal pressure bladder without any evidence of neurogenic component or bladder outlet obstruction. On cystoscopy, urethra, bladder and both ureteric orifices were normal. Thus patient was diagnosed to have nonobstructive nonrefluxing megaureter. Now patient is on oral tablet of 100 mg trimethoprim once daily and he is on regular follow up.

Discussion:

The incidence of Prune Belly syndrome has been reported to be 1 in 29,000 to 1 in 40,000 live births [1]. Parker in 1895 first described the association of abdominal muscle defects with urogenital anomalies [2]. This syndrome has been called by many other names, including Eagle-Barrett syndrome[3], absence of abdominal musculature [4-6], triad syndrome [7], and mesenchymal dysplasia syndrome [8]. Prune Belly syndrome is rare in females with fewer than 30 cases reported in literature [9]. The disease has wide spectrum of clinical presentations. Woodard in 1985 described three major categories of presentation [10]. Our patient falls in Category III as patient has stable renal function in spite of nonobstructive nonrefluxing megaureter (Table 1).

The urinary tract anomalies are characterized by variable degrees of hydronephrosis, renal dysplasia, dilated tortuous ureters, an enlarged bladder, and a dilated prostatic urethra. The dilatation of the posterior urethra is due to prostatic hypoplasia, probably related to abnormal mesenchymal-epithelial development [11]. Our patient was diagnosed to have right nonfunctioning

Table 1: Woodard's Categories of Presentation

Category	Characteristics
I	Renal dysplasia
	Oligohydramnios
	Pulmonary hypoplasia
	Potter features
	Urethral atresia
II	Full triad features
	Minimal or unilateral renal dysplasia
	No pulmonary hypoplasia
	May progress to renal failure
III	Incomplete or mild triad features
	Mild to moderate uropathy
	No renal dysplasia
	Stable renal function
	No pulmonary hypoplasia

contracted kidney with left nonobstructive nonrefluxing megaureter with dilated prostatic urethra. Urethral obstruction was ruled out by USG and by cystoscopy. Urologic intervention is reserved for patients who demonstrate repeated urinary tract infections, probably related to urinary stasis or vesicoureteral reflux, or development of upper tract deterioration [12].

Conclusion:

Prune Belly syndrome represents a wide spectrum of disease. The outcome greatly depends on the amount of renal dysplasia and renal function and the degree of respiratory embarrassment. In some patients disease is so severe that patient dies within first few weeks of life. Rarely some patients with mild disease grow up without much physical or physiologic impairment. Each patient has to be managed with on an individual basis. The urinary tract is usually dilated but rarely obstructed. In patients with stable renal function, watchful waiting with selective surgical intervention has also been successful.

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