The Prune Belly Syndrome

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I. General

A) History

The first description of Prune Belly Syndrome or abdominal wall muscle aplasia dates back to 1839. It was described as a triad in 1895 [16], and subsequently identified as a particular entity from 1901 [14].

B) Definition

This is a clinical, anatomical and radiological entity that involves mainly boys and is associated with

- Aplasia or severe hypoplasia of the anterior abdominal wall muscles
- Enlargement of the whole urinary tract, and
- Undescended testes.

The typical appearance of the wrinkled abdominal skin (Prune Belly) is due to a parietal malformation (Fig. 1).

These three findings are almost always seen together [21], but rarely one of the elements does not exist where the condition is called the "Pseudo-Prune Belly Syndrome". In the pseudo-prune belly syndrome, the aplasia of the muscles may be partial or concerning the genitalia, the patient may be a female, hence the undescended testes are no more an associated condition.

C) Epidemiology

This is a rare malformation, about 1 in 40,000 births [7], affecting mainly boys. Only 300 cases have been reported in the pediatric literature. This malformation has never been a subject in adult surgery. There is a spectrum for prune belly patients; on the one end there is the stillborn baby with dysplastic kidneys and major pulmonary problems, on the other end, a rather normal newborn.

The prune belly syndrome represents a challenge in pediatric urology since Waldbaum and Marshall's paper [22] where they presented a series of 50 patients from 1950 to 1979 of which 48 were post mortem reports.

This was approved by Wooddard [25] who reported that more than half of the 150 cases published before 1970 were autopsy results. These were either stillborn babies, or infants deceased earlier than 2 years of age without receiving any form of treatment whatsoever. Death was related with renal insufficiency or complications of urinary tract infections.

Welch [23] presented a series with 70% mortality, and Duckett [4] admits that 20% of the patients die during the first 2 months of life and 50% in the first 2 years. The causes of death varies from septic complications with failed therapeutic attempts to stillborn babies.

This data probably explains the unsettled opinions on how to handle this syndrome.

II. Etiology

The basic alteration seems to be a morphogenic abnormality of the intermediate and lateral folds of the para-axial mesoderm which occurs around the 23rd day of gestation [19]. The formation of the kidneys from the ureteric buds takes place at the same time. This synchronism probably explains the different components of the condition. There is no precise hereditary influence although 3 cases of prune belly syndrome in twins was reported. It is known from many pairs of monozygotic twins that twinning is discordant for the syndrome. There is also no evidence of the influence of maternal ingestion of chemicals during pregnancy.

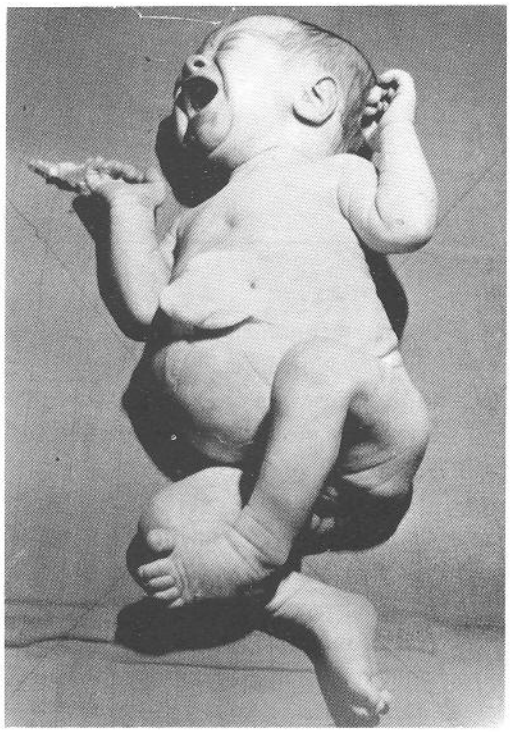

Figure 1. Newborn Prune Belly Syndrome.

III. Pathophysiology-Anatomic Pathology

A) The muscles

Muscular dystrophy, loss of coherence of Z-bands, mitochondrial anomalies and abnormal glycogene storage [10] seem to be the principal question. These disorders are also found at the ureteral and vesical muscle network. The fibrous tissue surrounding the muscle fibers may be extremely important in some cases. The clinical findings of abdominal muscular pathology is not uniform and frequently more serious at the lower end. Muscle deficiency may be so pronounced that not only sitting is impossible but major respiratory problems arise due to lack of abdominal respirations and inability to cough.

B) The testes

The testes are present in the majority of the cases, but are located very high in the abdomen and it is almost impossible to bring them into the scrotum without dividing the spermatic pedicle. In this situation the arterial supply will depend only on the deferential artery. Although the testicular histology is normal, fertility in the prune belly population has never been documented. Infertility can be related to prostatic hypoplasia and lack of seminal fluid rather than an anomaly of the spermatozoids per se.

It has been suggested that the testes should be left in the abdomen in order to preserve their maximal function and testicular prostheses placed in the scrotum later at puberty. Microsurgical testicular transplantation should be kept in mind considering the risk of the neoplastic development.

C) Urinary system

1. The kidneys

Renal involvement is part of this syndrome, it presents as a focal and asymmetrical dysplasia. As the condition affects the kidneys from the intrauterine life, the urinary system is prone to complications. Scarring of the renal parenchyma is unavoidable due to stasis and/or infection. At this point, there is an ungoing debate: some authorities believe that surgery for drainage of urine is mandatory, whereas some others suggest that low pressure stasis is not dangerous and appropriate medication to prevent infection is sufficient. Pelvi-calyceal enlargement is a common finding, but dilatation is more pronounced at the lower part of the urinary tract. Manometric studies of the pelvis fail to demonstrate a distal obstruction. Although this proximal distension does not seem to be of major importance, adequate evacuation of the distal urinary system should be checked as a rule.

2. The ureters

Macroscopic appearance of the ureters in prune belly syndrome on IVP or during the operation is quite typical; the dilations are gigantic, distal portions being more prominent. Kinking of the tortuous parts act as pseudovalves (Fig. 2). It has been suggested that pathologic changes of the smooth muscle fibres and fibrosis is more intense at the lower ureteric wall [8]. This view is not shared by Nunn and Stephens [13]. They claim that similar changes are found throughout the ureteric wall. This is confirmed by Palmer and Teslik [18], and the idea is well accepted by the supporters of large ureteral resections since three quarters of the ureteric length can be discarded during surgery.

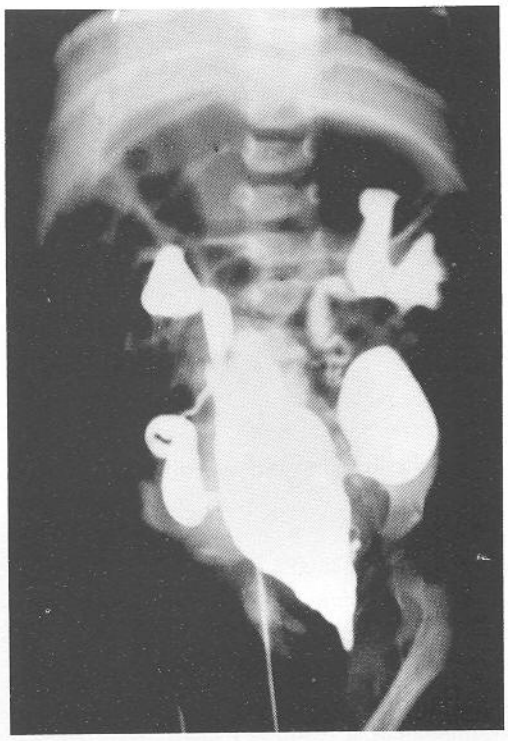

Figure 2. Prune Belly Syndrome on IVP.

3. The bladder

The bladder capacity is enormous. The dome is more enlarged and the urachus is patent. In cases with severe hypoplasia or atresia of the urethra, the patent urachus is draining urine through the umbilicus. The trigone is large, ureteric orifices placed laterally and the submucosal tunnels are absent in most of the cases. Although the intravesical pressure is low, vesico-ureteric reflux is present in 80% of the prune belly patients. Voiding is incomplete but the bladder neck is well formed and incontinence is not common in these children. As the bladder functions are inadequate, even the strongest advocates for "No Touch" approach are convinced for cystostomies in severely affected patients. Bladder functions and cervico-urethral autonomy improve with age.

4. The urethra

a) Posterior urethra

The posterior urethra is dilated in almost all patients up to the level of bulbous urethra. This dilation is associated with or secondary to the hypoplasia of the prostate.

The appearance is somewhat similar to the posterior urethral valve pathology. There are some authors who report dramatic improvement following endoscopic ablation of these pseudo-valves [2]. However another group of authors do not accept the existence of obstruction in these patients.

b) Anterior urethra

The anterior urethra is normal in almost all cases. Megaurethra is seen occasionally. A few urethral aplasias were also reported. In these patients a urachal fistula is present. Some infants with all the features of prune belly syndrome have a completely normal urethra.

IV. Diagnosis

Clinical findings of a newborn with prune belly syndrome will reveal all the features of this entity. Gross dilation of the urinary system is easily recognized with ultrasonography either prenatally or at birth. Prune Belly patients are classified in 3 categories:

Stage 1- This is the biggest group. The babies are in good shape at birth, they have no renal insufficiency but their radiologic findings are surprising. Urinary tract infections and dysuria are threatening these patients, and the discussion on conservative treatment or surgery is continuing.

Stage 2- In this group the patients are mildly affected but still represent one of the serious urologic conditions where the evolution is not clear. They always need a surgical drainage procedure to prevent their urinary retention. Longterm follow up of these patients show a survival rate of less than 50%.

Stage 3- Diagnosis is straightforward in these severely affected babies with multiple malformations. They are often oliguric and easily infected and show rapid signs of renal deterioration. There is no treatment modality for this neonates who succumb shortly.

V. Treatment

There are different opinions on treatment policy. Some authors advise early reconstructive surgery for the genitourinary system and abdominal wall [5,8,11,25].

Another group performs high urinary diversion in every patient [17] and some authors suggest that no particular treatment other than regular surveillance is needed [4,20].

A) Methods

1- Conservative treatment

The advocates of "No Touch" approach claim that stasis without infection and low pressure is not harmful for the kidneys. In fact, if there is no obstruction that can be demonstrated, its treatment is illogical and may be dangerous. Low pressure vesico-ureteric reflux without urinary infection is also acceptable as it is not endangering renal functions.

This group of authors base their thesis on the fact that in most of the patients urodynamic parameters tend to improve with time, hence there should be no need for surgery unless complications occur.

2- External diversions (Cutaneostomies)

There is a certain number of patients with electrolyte imbalance or infectious problems who need urinary diversion. Bilateral cutaneous ureterostomies and vesicostomies are frequently used in these patients. Diversion in the form of end ureterostomy or the "Sober" technique may improve the patient's condition dramatically in a short time. But in the long term follow up, infections, stoma stenoses, technical difficulties with external appliances and tedious stoma revisions are the reasons why this kind of surgery is not commonly used.

The pediatric surgeons who insist on diversions, prefer to do a total or subtotal resection of the ureters and high ureterostomies or cutaneous pyelostomies with an intestinal segment [17].

3- Reconstructive surgery

Single-stage surgery for treatment of malformations in prune belly syndrome is a difficult task and subject to controversy. This technique aims early correction of all the malformations [5,8,11,25].

Surgery is done in the first few weeks of life. The infant should be in good condition; electrolyte balance and infection under control.

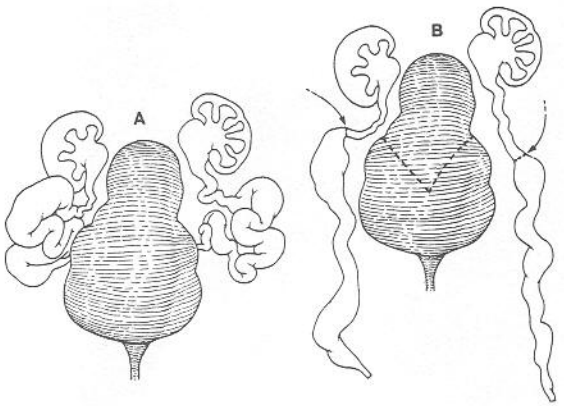

Figure 3. Reconstructive surgery in Prune Belly Syndrome.

- The whole pelvic ureter is removed in order to find a better functioning upper ureter which is reimplanted to the bladder. This portion of the ureter may need a longitudinal tapering according to Hendren [6] (Fig. 3).
- The reimplantation is either Cohen's transtrigonal advancement [3] or , if the trigone is too large, a modification of Leadbetter-Politano [15] operation where a decent submucosal tunnel is achieved by intra and extra-vesical approach. This reimplantation should resolve the vesico-ureteric reflux problem.
- A partial cystectomy removing the dome of the bladder which gives a smaller capacity and a better tonus in order to obtain a normal bladder emptying.
- Endoscopic sectioning of the pseudo-valve.
- Bilateral orchidopexies with high ligation of the spermatic pedicles. The pedicle should be clamped first to assess the viability of the testis depending on the deferential artery.
- Finally, the abdominal wall is repaired either with a simple anterior resection or a double-breasted-plasty in order to obtain a better cosmetic and functional result [1]. Spontaneous improvement of the abdominal wall is also observed. We have described [11] a technique for reconstruction of the abdominal wall called "Calisson" which preserves the umbilicus for a much better cosmetic result (Fig. 4).

B) Results

There are very few publications on longterm results of prune belly series and none on adult patients.

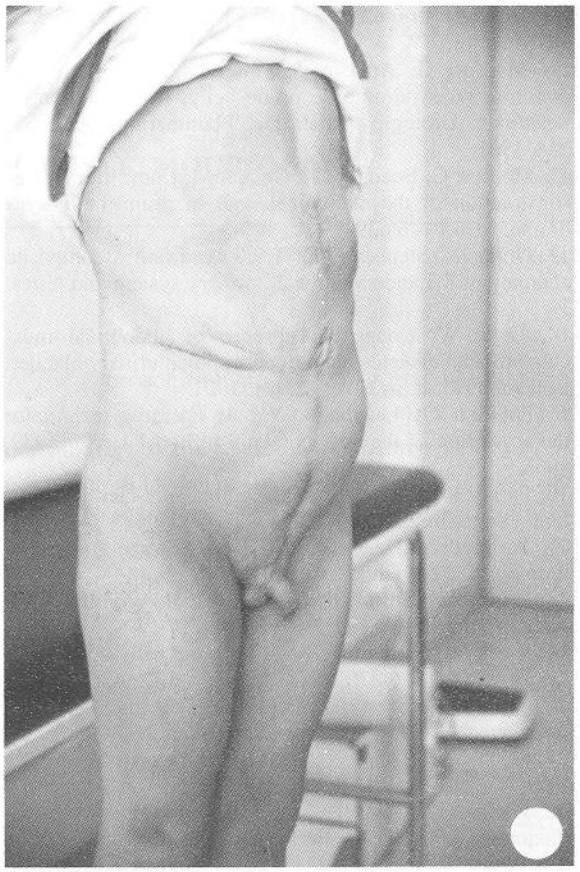

Figure 4. Results of the "Calisson" technique in prune belly syndrome.

• Duckett [4] presented 30 cases with 6 deaths. 7 patients had vesicostomies. 4 of these patients had their vesicostomies closed eventually and 3 left with permanent diversion.

• Williams [24] presented 12 adolescents. Only 3 of them were suffering from renal insufficiency. The radiologic appearance which is always shocking does not necessarily mean poor function. (The endocrine function of the testes is normal but the exocrine function is doubtful.)

• The same series were reviewed by Woodhouse and Ransley [26] with the following results:

5 children in Stage 3: No survival.

13 children in Stage 2: 12 were treated with cutaneous ureterostomies (6 secondary reconstructions and 1 death after refusal to treatment).

29 children in Stage 1: No deaths. 20 patients in this group had endoscopic section of the pseudovalves.

In total, 25 children are doing well; 8 children had diversions; 4 are well, 4 in renal insufficiency (3 on dialysis).

Other problems include:

2 Arterial hypertension, 1 stone formation, 1 cerebral lesion, 1 testicular teratoma and 1 uremic patient who deceased at 14 years of age.

• Woodard [25] published 16 cases. Total reconstruction was performed in 10 patients with 10 success. The remaining 6 patients:

1 Refusal to treatment: died

2 Infants deceased within 48 hours.

2 Infants were not given any treatment.

1 Trans-intestinal cutaneous ureterostomy ended with renal insufficiency.

• Welch [23] presented 43 patients followed up from 1941 to 1973. Only 14 of 43 were alive. 12 of them were treated with diversions, and 9 of this 12 had ultimate reconstructions. 2 patients did not need any treatment.

• The series of Cendron and Valayer [7] consisted of 34 cases. 41% of the total number had deceased. The Stage 3 group was the worst with 100% death rate (8/8), Stage 2 group 22% (6/23), and 20 patients are all well in Stage 1. Only 6 of 20 patients had surgical treatment.

• We have published statistical analysis of 18 patients [12] that we have treated since 1969. Our policy on treatment of urinary system complications was rather aggressive:

Material: 18 patients (2 females) followed at the Children's Hospital of Timone, Marseille.

Method: Urinary diversion performed on 4 cases (3 of them were definitives) and reconstructive surgery on 10 cases.

Results: No mortality and negligible morbidity (All postoperative courses were under control and there has been no serious complication).

C) Conclusions

It seems that no emergency treatment is needed in Stage 1 patients. But one should make sure that spontaneous emptying of the bladder is satisfactory. If a pseudovalve prevents adequate bladder emptying, endoscopic resection is helpful.

Surgery for vesico-ureteric reflux remains open for discussion.

Stage 3 patients are helpless, there is no medical approach to cure these babies. In Stage 2, reconstructive surgery and cutaneous diversions are two surgical options. It is our feeling that cutaneous di-

versions are helpful to solve most of the problems in short term, but they may create as many problems in the longrun. Therefore total reconstructive surgery offer better chances for these patients.

The mortality rate for this condition still remains between 20 and 50%, according to various published series.

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