Prune Belly Syndrome in under Developed Countries: What Perspective in Management

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Prune Belly syndrome

Prune Belly syndrome (PBS) or Eagle-Barrett syndrome, is an anatomo-radiologic entity combining aplasia or hypoplasia of the large muscles of the anterior abdominal wall, urinary tract dilatation and cryptorchidism [1-3]. With the complexity of these malformations and its complications what are the perspective in the management of Prune Belly Syndrome in under developed countries?

The incidence of PBS is estimated at approximately 1/40000 birth and it's mostly reported in male [1]. Antenatal period is ideal for it's through obstetrical ultrasound in the second or third trimester of pregnancy [2,3]. In this context of polymalformations, prenatal diagnosis has advantages to organize an early multidisciplinary management in under developed countries antenatal diagnosis is still extremely difficult. These problems are closely related to both the low socioeconomic level and lack of specialists.

At birth even if the clinical presentation may be polymorphic variant between complete and incomplete forms, the diagnosis sometimes remains easy and patients are often referred to hospitals where a physical examination by a doctor is enough to guide the diagnosis to a syndrome Belly plum.

Paraclinical explorations usually do not cause problems and specially designed to find the exact position of the testicles, look for abnormalities of urinary tract. In practice an ultrasound an intravenous urography plus cytobacteriological urinalysis help for the diagnosis.

Although the post natal diagnosis of prune belly syndrome seems easy in under developed countries, the management of this syndrome is a real problem due to the mains complications in these children. Indeed this complex management requires several surgeries. Cryptorchidism management is done in two stages using the technique of Fowler-Stephens [1,2]. In these countries qualified human resources including anesthesiologists and pediatric surgeons is a real problem. This shortage of qualified staff partly explains the high morbidity and mortality of Prune Belly syndrome.

The urinary system malformations combine several elements in case of prune belly syndrome. Thus it may be lower urinary tract malformations with posterior urethral valves associated with impaired quality of the detrusor and the terminal portion of the urethra. Upper urinary tract malformations are resuming in vesico-ureteral reflux [1-4]. Urinary stasis cause repeated infections. These infections pose a real threat both the kidney (risk of kidney failure) and the organism (risk of infection with acute pyelonephritis).

In addition to urinary tract malformations, respiratory system disorders are another entity complicating prune belly syndrome management [3,4]. In this disadvantaged environment endoscopic resection is impossible because of lack of resources. Posterior urethral valve management consists on urethral valves lamination with a Foley catheter increasing size. It is important to note that this treatment has its own complications.

Regarding the achievement of the upper urinary tract in front of repeated infections and almost inevitable progression to kidney failure renal transplantation can be the perfect solution [3,4]. Some authors recommend an ureterovesical reimplantation associated with a bladder volume reduction surgery [1]. But in underdeveloped countries renal transplantation is still at the experimental stage. Aesthetically abdominoplasty techniques are known but there is the real problem of their feasibility in these areas because of human resources.

The management of urinary system disorders is very important for the survival of these patients. Presence of the respiratory malformation is a bad prognostic factor. These findings make PBS a multidisciplinary pathology and patients are poly-operated. What solution can be proposed to pediatrics surgeons in under developed countries for PBS management?

References