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Dolna 17, Warsaw,
Poland 00-773
+48 226 0 227 03
editorial_office@rsglobal.pl

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MORE THEN A TRIAD: UNDERSTANDING THE COMPLEXITY OF PRUNE BELLY SYNDROME

Anita Krowiak *(Corresponding Author, Email: krowiakanita@gmail.com)*
Clinical Provincial Hospital No. 2 in Rzeszow, Lwowska 60, 35-301 Rzeszów, Poland
ORCID ID: 0009-0008-0077-0925

Karolina Krowiak
Clinical Provincial Hospital No. 2 in Rzeszow, Lwowska 60, 35-301 Rzeszów, Poland
ORCID ID: 0009-0001-6704-0838

Anita Warzocha
University Clinical Hospital F.Chopinin Rzeszów, Fryderyka Szopena 2,35-055 Rzeszów, Poland
ORCID ID: 0009-0006-9845-4634

Piotr Pitrus
Clinical Provincial Hospital No. 2 in Rzeszow, Lwowska 60, 35-301 Rzeszów, Poland
ORCID ID: 0009-0006-2168-9187

Karol Bednarz
Clinical Provincial Hospital No. 2 in Rzeszow, Lwowska 60, 35-301 Rzeszów, Poland
ORCID ID: 0000-0001-9196-1621

Maria Jasiewicz
Medical Center in Łańcut ul. Paderewski 5, 37-100 Łańcut, Poland
ORCID ID: 0009-0008-0718-2528

Gabriela Majka
Rzeszów University, Rzeszów, Poland, Tadeusza Rejtana 16C, 35-310 Rzeszów, Poland
ORCID ID: 0009-0002-7100-5119

Magdalena Kowalczyk
Rzeszów University, Rzeszów, Poland, Tadeusza Rejtana 16C, 35-310 Rzeszów, Poland
ORCID ID: 0009-0000-6786-8232

Wiktoria Hander
Rzeszów University, Rzeszów, Poland, Tadeusza Rejtana 16C, 35-310 Rzeszów, Poland
ORCID ID: 0009-0002-3000-9703

Aleksandra Karnaś
Rzeszów University, Rzeszów, Poland, Tadeusza Rejtana 16C, 35-310 Rzeszów, Poland
ORCID ID: 0009-0005-9708-3467

ABSTRACT

Aims: Prune Belly Syndrome (PBS), also known as Eagle-Barrett syndrome, is a rare congenital disorder defined by a triad of abdominal muscle deficiency, urinary tract anomalies and bilateral cryptorchidism. This review aims to summarize current knowledge on its etiology, clinical presentation and management.
Methodology: This review was based on an extensive search of literature related to PBS using PubMed, Google Scholar and other scientific databases. Relevant clinical studies, reviews and case reports were analyzed to summarize current knowledge on diagnosis, management and outcomes of PBS.
State of knowledge: PBS affects approximately 1 in 30,000 to 1 in 50,000 live births, with a strong male predominance. Its pathogenesis is thought to be multifactorial, involving mesodermal maldevelopment and possible genetic factors. The clinical spectrum ranges from severe cases with renal dysplasia and pulmonary hypoplasia to a milder, incomplete form. Woodard's classification is commonly used to guide prognosis and treatment. Besides the classic triad, patients may present with gastrointestinal, musculoskeletal, cardiopulmonary and developmental systems. Early prenatal diagnosis is possible through imaging. Postnatal care includes urologic reconstruction, orchiopexy, abdominoplasty, renal support and in some cases, transplantation. Assisted reproductive techniques have allowed for fertility in select male patients.
Conclusion: PBS is a complex and variable condition requiring early diagnosis and individualized, multidisciplinary care. Advances in surgical and medical treatment have improved outcomes, but long-term monitoring and supportive care remain essential for preserving function and quality of life.

KEYWORDS

Prune Belly Syndrome, Urinary Tract Anomalies, Abdominal Wall Deficiency, Cryptorchidism, Congenital Disorder, Renal Dysplasia

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1. Introduction

Prune Belly Syndrome (PBS), also known as Eagle-Barrett syndrome, is a rare genetic disorder characterized by abnormal development of the abdominal muscles, urinary tract abnormalities and cryptorchidism [1, 2]. First described in the early 20th century, PBS remains one of the most unexplained pediatric conditions, with significant phenotypic variability and an unclear etiology [3]. Approximately 95% of individuals affected are male. The estimated incidence of live births with PBS ranges from 1 in 35, 000 to 1 in 50, 000 [4]. PBS presents a wide spectrum of severity, ranging from life-threatening complications to outcomes compatible with a normal expectancy. In some cases, individuals with PBS may retain fertility and have children [5].

This review aims to provide an up-to-date, comprehensive knowledge of PBS, with particular emphasis on the latest scientific reports on its pathogenesis, the expanding phenotypic spectrum, recent advances in prenatal and postnatal diagnosis, and evolving treatment strategies and long-term management. Promoting up-to-date knowledge of PBS as a multisystem disorder is crucial to improving the quality of diagnosis, therapeutic approaches, and patient survival outcomes.

2. Material and methods

The analysis was conducted based on a thorough review of studies and data related to Prune Belly Syndrome, using PubMed, Google Scholar and other scientific databases. Keywords such as"Prune Belly Syndrome" and"Eagle-Barrett Syndrome" were used. To broaden the scope of the search and ensure comprehensive coverage, additional terms such as"abdominal muscle deficiency syndrome, ""urinary tract anomalies, ""pediatric urology, ""congenital urinary tract malformations, ""genitourinary syndrome, ""fetal megacystis, ""bladder obstruction, ""renal dysplasia, " and"antenatal diagnosis of urinary tract anomalies" were also included. Additionally, relevant clinical studies were reviewed to provide a well-rounded perspective on the topic.

3. Epidemiology

The worldwide incidence of PBS syndrome is estimated to be between 1 in 30, 000 and 1 in 50, 000 live births [2, 4]. The disease affects approximately 95% of men, while it occurs much less frequently in women, with only about 3–5% of cases [4, 6]. In the study by Stoll et al. (2025), conducted over a 29-year period in northeastern France, the researchers analyzed 1, 946 cases of congenital anomalies of the kidney and urinary tract (CAKUT) among 387, 067 births. Within this cohort, 27 cases were identified as PBS, representing approximately 1.4% of all CAKUT cases [7]. The finding emphasizes the rarity of PBS and its association with other congenital anomalies and highlights the importance of a comprehensive evaluation of affected individuals. Moreover, in a population-based study from Finland, the total prevalence of PBS was 1 in 44, 000 births, with a live birth prevalence of 1 in 90, 000. Although rare, PBS carries a considerable disease burden, including a 20% infant mortality rate [8]. However, actual statistics may be underestimated due to early fetal loss, perinatal mortality of more severe phenotypes, or underdiagnosis of milder phenotypes of PBS syndrome [2].

4. Pathogenesis

The pathogenesis of PBS is multifactorial and remains the subject of ongoing research. One embryological hypothesis points to a primary mesenchymal defect, potentially resulting from abnormal differentiation or lateral migration of the somite mesoderm into the abdominal musculature or muscles of the urinary tract. Additionally, cases of divergent PBS symptoms in identical twins have led to the formulation of the theory of uneven embryonic division. It assumes that the affected twin may receive an insufficient amount of mesenchymal tissue necessary for the proper development of the abdominal wall and urinary tract structures [9]. In a study conducted by Boghossian et al., 34 cases of PBS identified over a 27-year period in New York were analyzed, leading to the identification of copy number variants involving the BMPR1B and NOG genes. Since these genes regulate the bone morphogenetic protein signaling pathway, which is essential for mesodermal development, their disruption reinforces the hypothesis that abnormal mesodermal differentiation may underlie the pathogenesis of PBS [10]. Furthermore, a scientifically validated association between mutations in the FLNA gene and PBS has been demonstrated, particularly in males surviving into adulthood [11]. Another widely discussed theory suggests that intrauterine urinary tract obstruction may lead to progressive distension of the bladder and ureters, resulting in secondary hypoplasia of the abdominal wall and failure of testicular descent [12]. On the other hand, based on long-term data from Finland, PBS has been observed only in male infants, suggesting a strong gender dependence of the incidence. Among the maternal characteristics assessed, a prior history of multiple miscarriages showed a significant association with the presence of a fetus burdened with PBS syndrome. No significant correlation was found with other factors such as maternal age, gestational age or gestational diabetes, indicating that the causes of PBS are still not well understood [8]. In summary, due to the lack of definitive causes and the existence of multiple hypotheses regarding the pathomechanism of the disease, further research integrating clinical, molecular and genetic data is essential for a comprehensive understanding of the etiopathogenesis of PBS. Multicenter studies are needed to identify new genes and signaling pathways as well as to develop targeted therapies that improve patient outcomes.

5. Clinical Presentation

5.1 Clinical Triad and Classification

PBS is classically defined by a clinical triad consisting of deficient or absent abdominal musculature, bilateral cryptorchidism and urinary tract anomalies [1, 2, 13]. A classification system proposed by Woodard categorizes PBS into three phenotypic severity levels to guide neonatal management. Category I includes the most severe cases with significant renal dysplasia, functional bladder obstruction and marked oligohydramnios, which often leads to pulmonary hypoplasia and characteristic Potter face with a flattened nose, recessed chin, low-set ears, epicanthal folds and hypertelorism. Most infants in this group who survive birth die within days from pulmonary failure or later from renal failure. Category II involves the full triad with variable renal involvement and risk of progressive renal impairment due to urinary tract dysfunction and recurrent infections. Category III represents milder or incomplete forms with relatively preserved renal and bladder function and favorable long-term outcomes [14, 15].

5.2 Abdominal Wall Abnormalities

The hallmark of the classic symptomatic triad is hypoplasia or atrophy of the anterior abdominal muscle wall, manifesting as a markedly flabby, thin and wrinkled abdominal skin. This distinctive appearance led to the naming of syndrome [1]. Hypoplasia of the abdominal musculature is a risk factor for the development of abdominal and inguinal hernias. These may require surgical repair using a combination of conventional and laparoscopic techniques, tailored to the individual anatomy and degree of abdominal wall deficiency [2].

5.3 Genitourinary Manifestations

In a cohort of 16 patients described by Zugor et al., common genitourinary findings included cryptorchidism in nearly 70%, megaureter in over 85% and large-capacity bladder (megacystis) in more than 80%. Renal abnormalities such as dysplasia and hydronephrosis were also commonly reported [16]. Beyond these structural anomalies, patients with Prune Belly Syndrome often exhibit significant functional abnormalities of the urinary tract. Poor bladder compliance, detrusor areflexia, and bladder outlet obstruction are frequently encountered and can lead to progressive upper urinary tract damage if not addressed appropriately [17].

Vesicoureteral reflux (VUR) is another common finding, especially in patients with markedly dilated ureters. The coexistence of VUR and bladder dysfunction significantly increases the risk of recurrent urinary infections (UTIs), which in turn contributes to renal parenchymal scarring and long-term functional decline [15]. Recurrent UTIs remain a common cause of morbidity and may require prophylactic antibiotic use, catheterization strategies or surgical correction to prevent upper tract deterioration [18].

Renal involvement in PBS varies in severity, but renal dysplasia and chronic obstructive uropathy frequently lead to progressive renal insufficiency. Studies have shown that a significant proportion of patients with moderate to severe phenotypes progress to end-stage renal disease (ESRD) by adolescence or early adulthood. In such cases, renal replacement therapy, including dialysis and eventual transplantation becomes necessary [19].

5.4 Fertility and Reproductive Function

Men with PBS face multiple fertility challenges, including undescended and ectopic testes, intrinsic testicular abnormalities, as well as prostatic and urethral dysfunction. Nevertheless, assisted reproductive technologies have made biological paternity possible. Successful sperm retrieval using microdissection testicular sperm extraction (micro-TESE), followed by in vitro fertilization (IVF) or intracytoplasmic sperm injection (ICSI), has led to live births in several reported cases [20]. On the other hand, a systematic analysis of 83 studies found that while azoospermia and impaired spermatogenesis are common, hormonal profiles in most PBS patients remain within normal ranges. Despite significant anatomical and functional barriers, preserved endocrine function and advanced reproductive techniques offer hope for selected PBS patients desiring biological parenthood [21]. These findings underscore the importance of early urological evaluation and long-term follow-up regarding reproductive potential.

5.5 Multisystem Comorbidities

In addition to the classic triad, PBS patients frequently exhibit anomalies in other organ systems, which may contribute significantly to morbidity and impact long-term prognosis.

5.5.1 Gastrointestinal System

In addition to chronic constipation and malrotation, mesenteric malfixation, intestinal volvulus, and gastroesophageal reflux disease (GERD) have been described. These complications may necessitate surgical intervention or long-term nutritional support.

5.5.2 Musculoskeletal System

Beyond scoliosis and clubfoot, joint laxity, abnormal gait, and delayed motor development are frequently reported, often attributed to the deficiency in core muscle strength and hypotonia. Physical therapy is often required to support motor milestones and musculoskeletal alignment.

5.5.3 Respiratory System

Pulmonary hypoplasia in infancy may predispose to long-term pulmonary sequelae, including restrictive lung disease, chronic hypoxia, and recurrent bronchopulmonary infections. Some patients develop chronic respiratory insufficiency requiring long-term monitoring or even ventilatory support in severe cases.

5.5.4 Cardiovascular System

Although less common, congenital heart defects such as patent ductus arteriosus (PDA) or ventricular septal defect (VSD) have been reported in association with PBS, particularly in syndromic or chromosomal variants. Routine cardiac evaluation may be warranted.

5.5.5 Neurological and Developmental Aspects

Some PBS patients experience delayed developmental milestones, likely secondary to prolonged hospitalizations, hypotonia, or comorbidities. Though not typically associated with intrinsic central nervous system abnormalities, cognitive development may be affected in severe cases, emphasizing the need for multidisciplinary follow-up.

5.5.6 Psychosocial Impact

Children and adolescents with PBS may suffer from body image issues, social withdrawal, or emotional distress, particularly due to visible abdominal deformity, urinary incontinence, or infertility concerns. Early involvement of psychological support services can significantly improve quality of life [18, 22, 23].

6. Prenatal Diagnosis

Advances in prenatal screening have enabled the early diagnosis of PBS, and in selected cases, the initiation of fetal treatment prior to birth. Suspicion of PBS can arise as early as the 12th week of gestation, primarily based on assessment of renal development. However, optimal conditions for reliably detecting anatomical abnormalities associated with PBS can be diagnosed as early as between 16 and 18 weeks of fetal life [24]. Sonographic diagnosis of PBS is based on a characteristic constellation of symptoms, including significant bladder distension, bilateral hydronephrosis and hydroureter, as well as diffuse urethral dilatation [25]. Additionally, in the article by Tonni et al., early prenatal diagnosis of PBS was reported based on the detection of megacystis, often accompanied by increased renal echogenicity. The review revealed further associated anomalies such as oligohydramnios, anhydramnios, pulmonary hypoplasia, urinary ascites, bilateral hydronephrosis, generalized fetal edema and congenital cardiac defects, such as Tetralogy of Fallot [9]. These results emphasize the importance of accurate early fetal assessment, considering the risk of serious multi-organ pathology in advanced forms of PBS.

7. Multidisciplinary management

7.1 Principles of Interdisciplinary Care

The care of patients with PBS syndrome requires an integrated, multispecialty approach. A pediatric urologist plays a central role in coordinating multidisciplinary treatment, leading the collaboration with physiotherapists, nephrologists, pulmonologists, gastroenterologists and nutritionists. Interdisciplinary care enables more effective treatment of urinary and gastrointestinal complications, supports psychomotor development, and improves patients' overall quality of life [26]. Early intervention is crucial in the management of PBS. This is particularly important during the neonatal period, when survival largely depends on the extent of pulmonary and renal involvement. Beyond this stage, long-term outcomes are significantly influenced by close urological monitoring, preservation of renal function and prevention of urinary tract infections. Equally important is the targeted treatment of disease-related complications according to the individual needs of the patient, which facilitates stabilization of the general condition and enables further psychophysical development [27].

7.2 Surgical Management of the Urinary Tract

Management of urinary tract issues in PBS varies by severity. Patients classified as category I typically receive conservative care focused on bladder drainage and supportive measures. In contrast, category III patients require minimal intervention aside from routine monitoring. The intermediate group presents the greatest clinical challenges and necessitates individualized treatment plans. Early surgical reconstruction is recommended between six and eighteen months of age for those with significant hydroureteronephrosis, vesicoureteral reflux, recurrent urinary tract infections or declining renal function. Surgical techniques include excision of refluxing or redundant distal ureteral segments with vesicoureteral reimplantation, nephrectomy for a non-functioning kidney and reduction cystoplasty for large urachal diverticula. Cryptorchidism correction is recommended within the first year of life, typically via transabdominal orchidopexy [26].

7.3 Long-Term Urological Complications

Patients with PBS may develop complex urological complications in adulthood, including urinary tract infections, upper tract deterioration and potential bladder malignancy after enterocystoplasty. The case reported by Singla and Lotan describes a 48-year-old male with PBS, a solitary kidney and prior enterocystoplasty who was diagnosed with invasive bladder cancer with extensive squamous differentiation, highlighting the importance of long-term urological monitoring in this population [28]. Moreover, an important case report highlighting a long-term complication of PBS was presented by Kondo et al. A 38-year-old male,

survivor of PBS was diagnosed with invasive urothelial carcinoma with glandular differentiation of the bladder. His medical history included multiple childhood urological surgeries, such as vesicostomy and ureterostomy, performed to manage severe urinary tract malformations. The authors suggested that chronic urinary stasis, recurrent infections and persistent inflammation may have contributed to malignant transformation in the bladder epithelium [29].

7.4 Kidney Disease and Transplantation

Importantly, patients with PBS are frequently affected by kidney disease, which in some cases may progress to end-stage renal disease. As reported by Khondker et al. and also by Chhabra et al. In separate studies, renal transplantation is a possible therapeutic option, with outcomes comparable to other pediatric transplant populations when preceded by proper urological management [19, 23, 30].

7.5 Abdominal Wall Reconstruction

An essential aspect of managing patients with PBS involves addressing the abdominal wall and its surgical reconstruction. Abdominoplasty aims to restore the integrity and function of the abdominal muscles. Reinforcement of the abdominal wall improves core stability, enhances respiratory function and reduces characteristic abdominal distension. Various surgical techniques have been described, typically performed in conjunction with urological procedures such as orchidopexy and urinary tract reconstruction. One commonly used method involves excising redundant skin and subcutaneous tissue while preserving the musculofascial layer and umbilicus [31]. A notable case involved a 2-year-old male who underwent a modified Monfort abdominoplasty. In this technique, the supraumbilical abdominal wall was preserved for its better tensile strength, while the lax infraumbilical skin was mostly excised. The result was a stronger abdominal wall and a lower, less visible scar, improving both function and cosmetic appearance [32]. A novel technique combining fascial plication with complete fascial overlap and neoumbilicoplasty using a self-rotating island flap has also been proposed to optimize both structural support and cosmetic outcomes [33]. Furthermore, another modification of the Monfort technique was described in a series involving 17 patients, which included the use of diagnostic laparoscopy to assess the extent and topography of the abdominal wall defect. Additional refinements, such as a midline skin incision, plication of the central fascial plate, correction of lateral asymmetry and umbilical repositioning were applied [34]. On the other hand, the technique described by Lopes et al. presents that laparoscopic abdominoplasty using a one-way running suture with non-absorbable material can effectively repair localized abdominal wall bulging in PBS. Importantly, the procedure was successfully performed in conjunction with laparoscopic nephroureterectomy and bilateral orchiopexies, highlighting the feasibility of combining abdominal wall reconstruction with other urological interventions in a single operative course [35]. In a separate case, a 19-year-old male underwent abdominoplasty combined with bilateral rectus femoris and sartorius muscle transfers to address severe abdominal wall deficiency. Active abdominal muscle tone was observed during physical exertion and there was no impairment in lower limb mobility, balance or ambulation. This case highlights the potential long-term benefits of incorporating muscle transfer techniques in selected patients with extensive abdominal wall defects [36].

7.6 Dental and Oral Health Considerations

Additionally, the multi-morbidity in PBS syndrome also affects the oral cavity. The most common problems observed include enamel hypoplasia, increased caries susceptibility, delayed or abnormal tooth eruption and xerostomia, often intensified by the medications used. As noted by Quilici et al, close cooperation between the dentist and the patient's general practitioner is recommended in order to tailor oral health care to the specific needs of each patient [37].

7.7 Prenatal and Perinatal Management

Moreover, prenatal diagnosis of PBS requires careful evaluation, as coexisting structural anomalies may suggest termination of pregnancy. In isolated cases with normal karyotype, a vesicoamniotic shunt may be considered to decompress the bladder and improve perinatal outcomes [38].

7.8 Psychosocial and Family Support

Significantly, due to multi-morbidity and the potential need for continuous care, it is essential to support families with nursing, psychological and social services to help manage the complex needs of our patients [24]. These findings underscore the importance of close collaboration among specialists to ensure effective and coordinated care for individuals with PBS.

7.8 Prognostic

This condition is associated with high neonatal mortality, particularly in cases with severe anomalies. The outcome for patients depends mainly on how severely the kidneys and urinary tract are affected, as well as whether pulmonary hypoplasia is present. These factors have the greatest impact on survival and clinical severity [12].

8. Quality of life

Arlen and colleagues found that 84% of children with PBS scored significantly below the typical range seen in healthy children, falling more than half a standard deviation below the norm. These findings clearly highlight that children with PBS face complex and multifaceted challenges in their daily lives. These include not only physical limitations, but also emotional, social, and educational problems. Caregivers of these children confirm that the need for constant, demanding care significantly affects the quality of life of the entire family. The results of the study underscore the need for a holistic approach in the care of PBS patients, one that takes into account not only the treatment of somatic symptoms, but also psychological and social support, aimed at both children and their loved ones [39].

9. Pseudoprune-belly syndrome

The incomplete form of PBS represents a rarer and more clinically variable manifestation of the classic PBS. In the literature, it is also referred to as Pseudoprune-belly Syndrome (PPBS). Furthermore, the phenotypic variability often complicates early diagnosis and requires careful clinical evaluation. In incomplete PBS, abnormalities of the abdominal wall may be subtle or even absent, while genitourinary malformations range from mild to severe and can include renal dysplasia, hydronephrosis, hydroureter, vesicoureteral reflux and urethral dilatation. Due to this variability, the condition often remains undiagnosed during the neonatal period, especially when symptoms are limited to isolated urological findings such as unilateral cryptorchidism or mild urinary tract anomalies [40 - 45].

This variability is illustrated by several case reports. For instance, Olori et al. described an 8-day-old male infant with PPBS associated with microcolon and rectal atresia, emphasizing the potential for gastrointestinal anomalies [46]. Similarly, Fishman and Franco reported two male patients exhibiting symmetrical bilateral hypoplasia or absence of the internal and external oblique muscles without genitourinary abnormalities, underscoring the phenotypic heterogeneity of PPBS and the importance of differential diagnosis. This rare congenital abdominal wall defect is often misdiagnosed as an incomplete form of PBS [47]. Patients with PPBS face significant clinical challenges, primarily due to the high incidence of serious urinary complications such as urinary septicemia and progressive renal failure. Surgical reconstruction addressing both abdominal wall defects and urinary tract abnormalities is often necessary to manage these risks. However, a significant proportion of patients still require complex procedures, including ureteral reimplantation or urinary diversion procedures, such as cutaneous vesicostomy or ileal conduit. Therefore, continuous multidisciplinary monitoring by urology and nephrology specialists is essential for the early detection and treatment of emerging complications [42].

10.Conclusion

PBS remains a rare complex condition that requires an individualized diagnostic and therapeutic approach. Advances in modern medicine now allow for earlier diagnosis and more precise assessment of disease severity, leading to improved outcomes. The development of high-resolution imaging technologies has refined surgical techniques, and multidisciplinary care has enabled more effective treatment while minimizing the risk of complications. However, due to the rarity of PBS, raising awareness among healthcare professionals remains essential. Increasing clinical awareness is key to early detection, appropriate management planning and improving patients' quality of life. Continued education, interdisciplinary collaboration and ongoing research are fundamental to meeting the evolving needs of this unique patient population.

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