OPEN ACCESS Freely available online

Andrology-Open Access

Mini Review

Prune-Belly Syndrome Fertility: Is Fertility Fruitful or Futile

Lane Shish[1*], Stanley Kogan[2]

[1]Department of Urology, University of Minnesota, Minneapolis, USA; [2]Department of Pediatrics and Regenerative Medicine, Wake Forest University, Winston-Salem, USA

ABSTRACT

Patients with Prune-Belly Syndrome (PBS) have historically been considered infertile, however significant improvements in medical care, research and reproductive technologies have greatly improved their reproductive capabilities. Our recent scoping review reviewing the extant literature over the last 70 years demonstrated favorable testicle histology, sex hormone profiles, semen analyses and successful conception potential in PBS patients. Despite their fertility potential, specialized and individualized guidance is needed to help manage their reproductive needs.

Keywords: Prune-belly syndrome; Infertility; Spermatogenesis; Artificial reproductive technologies

INTRODUCTION

Prune-Belly Syndrome (PBS), also known as Eagle-Barrett syndrome, is a rare congenital disorder with a constellation of symptoms that primarily affects the abdominal musculature, urinary tract and in males, the reproductive system. First described in the 19th century, PBS is characterized by a notable deficiency of abdominal muscles, a spectrum of urinary tract abnormalities and in many cases, undescended testes (cryptorchidism). This triad of features can present a complex challenge for those affected, raising significant questions about their overall health and quality of life. Among the various concerns is the impact of PBS on fertility, which varies widely depending on the severity of the condition and the specific complications present.

Understanding the implications of PBS on fertility requires a nuanced approach, as the syndrome's effects can differ markedly between males and females, as well as among individuals with varying degrees of severity. In males, cryptorchidism is a common feature, where one or both testes fail to descend into the scrotum. This condition not only affects the external appearance but can also compromise spermatogenesis, the process of sperm production [1]. Testicular function is highly sensitive to temperature and the warmer environment within the abdomen can impair sperm production and lead to infertility if left untreated. Surgical correction, typically through orchidopexy, is often necessary to reposition the testes and improve the chances of successful sperm production. Even with surgical intervention, however, the extent of the improvement in fertility can be variable and some individuals may still face challenges in achieving fatherhood.

For females with PBS, the direct impact on fertility is generally less well-documented. The absence of abdominal muscles, while primarily affecting physical function and health, does not inherently interfere with reproductive organs. However, the associated urinary tract abnormalities, which are common in PBS, may have indirect effects on reproductive health. These abnormalities can range from minor to severe, potentially influencing overall health and complicating pregnancy. Despite these concerns, many females with PBS are capable of having successful pregnancies with appropriate medical management and support [2].

Overall, the question of whether fertility in individuals with PBS is fruitful or futile is not straightforward and depends on a variety of factors, including the severity of the syndrome and the effectiveness of medical interventions. Advances in surgical techniques and management strategies have significantly improved the outlook for many individuals with PBS, allowing for successful fertility outcomes where once there may have been limited hope. Nonetheless, each case is unique and a comprehensive evaluation by a healthcare provider specializing in PBS is essential for tailoring treatment plans and optimizing reproductive health.

As the understanding of prune-belly syndrome continues to evolve, so too does the ability to address its challenges, including

Correspondence to: Lane Shish, Department of Urology, University of Minnesota, Minneapolis, USA; E-mail: shish020@umn.edu

Received: 13-Feb-2024, Manuscript No. ANO-24-29572; **Editor assigned:** 16-Feb-2024, PreQC No. ANO-24-29572 (PQ); **Reviewed:** 01-Mar-2024, QC No. ANO-24-29572; **Revised:** 03-Mar-2025, Manuscript No. ANO-24-29572 (R); **Published:** 10-Mar-2025, DOI: 10.35248/2167-0250.25.14.350

Citation: Shish L, Kogan S (2025) Prune-Belly Syndrome Fertility: Is Fertility Fruitful or Futile. Andrology. 14:350.

those related to fertility. With ongoing research and medical advancements, the outlook for individuals with PBS regarding their reproductive potential remains cautiously optimistic, highlighting the importance of personalized care and ongoing support.

LITERATURE REVIEW

Prune-belly syndrome is a condition noted at or before birth consisting of major dysplasia of the entire urinary tract, prostate and abdominal wall and intra-abdominally undescended testes. The condition occurs with an incidence of 1 out of 30,000-40,000 births and occurs almost exclusively in male patients. Medical management of PBS patients has changed drastically over time and has resulted in longer life expectancies. Now that many of these patients are living into adulthood, questions regarding their reproductive health and the best management options are becoming more prevalent.

Historically, PBS patients have been considered infertile due to impaired spermatogenesis from their universally present intra-abdominal testes, retrograde ejaculation, anatomical prostatic abnormalities and the lack of successful reproductive interventions. However, our recent scoping review evaluating all pertinent articles published over the last 70 years demonstrates that PBS patients may now have a reasonable expectation for fathering their own offspring [3].

Our review demonstrated that orchiopexies are now being performed earlier (at or before the age of 12 months) after recognizing the technical feasibility as well as the confirmed fertility benefits of this early approach. Additionally, more recent articles demonstrate the presence of spermatogenesis in testicular biopsy samples, findings that were not noted prior. These findings suggest that 1) Early orchiopexy may be helping to preserve future reproductive function and/or; 2) PBS has a spectrum of phenotypes and genotypes, resulting in the presence or absence of spermatogonia. These findings should influence future reproductive management considerations by demonstrating the need for an individualized approach and the need for ART, given the presence of spermatogonia in testicular samples (Table 1).

Table 1: Clinical summary.

Testicular histology		
Leydig, sertoli and spermatogonia (n=36)		
% Patients without spermatogonia present (n)	50.0% (18)	
% Patients with reduced spermatogonia (n)	47.2% (17)	
% Patients with normal number of spermatogonia (n)	2.7% (1)	
% Patients with Leydig hyperplasia (n)	19.4% (7)	
% Patients patients with Sertoli only (n)	19.4% (7)	
Prostate		
% Patients with hypoplasia	93.6% (102/109)	
% Patients with abnormally dilated prostatic urethra	91.4% (106/116)	
Prostate histology (n=41)		
% Patients with decreased smooth muscle within prostatic urethra (n)	97.6% (40)	
% Patients with reduced number of prostatic glands/gland abnormalities (n)	95.1% (39)	
Seminal vesicles/epididymis (n=59)		
% Patients with abnormal seminal vesicle(s)/epididymis (n)	50.8% (30)	
Hormones		
	Before 2000	**After 2000**

% Patients with testosterone within normal limits (n)	88.9% (8)	94.7% (54)
% Patients with luteinizing hormone within normal limits (n)	37.5% (3)	94.7% (54)
% Patients with follicle-stimulating hormone within normal limits (n)	27.3% (3)	87.7% (50)
Semen		
	Before 2000	**After 2000**
% Patients with azoospermia (n)	100% (14)	63.6% (14)
% Patients with oligospermia (n)	0	36.4% (8)
% Patients with retrograde ejaculation (n)	38.8% (7)	20.0% (2)
% Patients with antegrade ejaculation (n)	50.0% (9)	80.0% (8)
% Patients with anejaculation (n)	11.1% (2)	0
ART/conception		
	Before 2000	**After 2000**
Number reports of successful sperm extraction with MESA	3	0
Number conceptions using MESA and ICSI	2	0
Number reports successful sperm extraction with TESE	0	1
Number conceptions using TESE and ICSI	0	0
Number reports of successful sperm extraction from semen	0	2
Number conceptions using sperm extracted from semen and ICSI	0	2
Number reported natural conceptions	0	2

DISCUSSION

Another finding influencing fertility is description of prostatic abnormalities, both macroscopic and microscopic, in some patients. The PBS prostate was most commonly described to be hypoplastic with a wide prostatic urethra and few functional glands histologically. These findings support further the theories describing the syndrome's phenotypic spectrum and emphasize the need for individualizing reproductive management furthermore, given the relatively high prevalence of prostatic abnormalities and retrograde ejaculation present in PBS patients, consideration should be given to obtaining a post-void bladder semen sample to assess this possibility [4].

Sex hormone profiles were another pertinent topic evaluated. While earlier articles showed that LH and FSH were commonly elevated in PBS men, modern literature shows that FSH, LH and testosterone are often within normal limits. Though reasons for this divergence are unclear, we speculate that since orchiopexies are occurring earlier, this is likely better preserving testicular function. Though we may expect these measurements to be normal, hormone assessment may be helpful in contemporary overall fertility management to exclude other additional potentially affecting morbidities [5].

Semen and ejaculate were also commonly evaluated and generally abnormal in PBS patients. Earlier studies showed that about half of patients had retrograde or anejaculation and all were azoospermic [6]. However, importantly, newer articles published after 2000 noted 80% of patients had antegrade ejaculation and 36.4% had sperm present in their semen. We hypothesize that the improvement in both ejaculatory and

semen metrics in modern literature is again, likely due to earlier orchiopexies in PBS patients and possibly due to the phenotypic and genotypic spectrum of the syndrome [7]. These data illustrate the potential need for differing reproductive management strategies and that a semen analysis and post void semen analysis can be useful in evaluation and treatment planning [8].

When considering these promising changes noted in more recent publications the main question remaining is: Can PBS patients conceive? Our review demonstrated that ART was successful in extracting sperm in 6 patients (3 with MESA and 3 with TESE) which led to 4 conceptions (2 with MESA and 2 with TESE) [9]. Surprisingly, there were also two reports of natural conception in modern literature. Our review demonstrates that PBS men can have a reasonable expectation to father a child, especially with the use of ART. Current fertility management for PBS patients should highly consider the use of assisted reproductive techniques in an individualized manner to increase the likelihood of conception [10].

CONCLUSION

The historical acceptance of infertility as a given for PBS men is being rightfully challenged. Recent data highlights the progress made utilizing contemporary management approaches and also the need for consistent and individualized reproductive strategies. Strides made in understanding and managing fertility in PBS men should inspire a collective commitment to continued research and tailored approaches. PBS men, once resigned to the notion of infertility, now have a reason to be hopeful with early collaboration with a fertility specialist. Further investigations are required to confirm the most effective reproductive management strategies and further enhance the fertility prospects for these individuals with PBS.

While PBS can present significant hurdles to fertility, both medical and surgical advancements offer hope for many individuals affected by the syndrome. The outlook for fertility in PBS is not uniformly futile but rather contingent upon personalized care and intervention. With ongoing advancements in medical treatment and a comprehensive approach to managing the syndrome's diverse aspects, many individuals with PBS can achieve fruitful fertility outcomes. Each case is unique, underscoring the importance of tailored medical evaluation and support to navigate the challenges and maximize reproductive potential.

REFERENCES

1. Straub E, Spranger J. Etiology and pathogenesis of the prune belly syndrome. Kidney Int. 1981;20(6):695-699.
2. Arlen AM, Nawaf C, Kirsch AJ. Prune belly syndrome: Current perspectives. Pediatric Health Med Ther. 2019;10:75-81.
3. Lopes RI, Baker LA, Denes FT. Modern management of and update on prune belly syndrome. J Pediatr Urol. 2021;17(4):548-554.
4. Jennings RW. Prune belly syndrome. Semin Pediatr Surg. 2000;9(3):115-120.
5. Hutson JM. Prune belly syndrome. Rickham's neonatal surgery. 2018:1211-1216.
6. Kolettis PN, Ross JH, Kay R, Thomas Jr AJ. Sperm retrieval and intracytoplasmic sperm injection in patients with prune-belly syndrome. Fertil Steril. 1999;72(5):948-949.
7. Halpern JA, Das A, Brannigan RE. Successful sperm retrieval in prune belly syndrome. Asian J Urol. 2020;7(4):376-378.
8. Wheatley JM, Stephens FD, Hutson JM. Prune-belly syndrome: Ongoing controversies regarding pathogenesis and management. Semin Pediatr Surg. 1996;5(2):95-106.
9. Greskovich, Nyberg LM. The prune belly syndrome: A review of its etiology, defects, treatment and prognosis. J Urol. 1988;140(4):707-712.
10. Uehling DT, Zadina SP, Gilbert E. Testicular histology in triad syndrome. Urology. 1984;23(4):364-366.