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Scrotal flap phalloplasty as temporary neophallus in infants and children with penile agenesis: Multi-institutional experience and long-term follow-up

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Summary

Introduction
Aphallia is a rare congenital disorder pertaining to genotypic males. Early surgical creation of a neophallus is recommended to reinforce the child's male gender-identity, favoring proper psychosexual development. Modern microsurgical techniques used to create a neophallus in adults are not recommended in children due to the invasiveness and complexity of the procedures, along with high complication rates. Scrotal flap phalloplasty is a simple and reproducible technique to create a temporary neophallus in prepubertal boys with aphallia.

Objective
We present a multi-institutional experience, ten years after the initial description of the scrotal flap phalloplasty (SFP) technique, in which a flap from the well-developed scrotum is used to build a temporary neophallus, without obvious scars in patients with aphallia.

Study design
The records of surgical neophalloplasty for aphallia patients from 4 centers between 2011 and 2021 were reviewed. All patients had at least one year follow-up to assess for short and long-term complications. Age at initial operation, associated anomalies, and other related surgical procedures were analyzed.

Results
The post-operative aesthetic result in all patients was satisfactory and has been maintained in the long-term follow-up, with all patients presenting a cylindrical structure resembling an uncircumcised penis, without evidence of significant contraction or loss of length. (Summary Figure)

Discussion
Non-microsurgical neophalloplasty techniques in patients with penile agenesis are temporary procedures that help to establish the body image and preserve the psychosexual development of the patient with aphallia. These techniques do not involve tissue transplant from a distant region, and are simpler to perform, with less scarring at the donor sites. Due to significant donor scars and considerable morbidity and complexity associated with the definitive phalloplasty techniques, we created a simple, reproducible and straightforward procedure to serve as a temporary neophallus in young boys with aphallia. As affected patients usually have a well-formed scrotum with normal and orthotopic testicles, it is the ideal donor site for a temporary neo-phallus in childhood. Furthermore, other donor sites are preserved for a definitive phalloplasty. There are limitations to this study, as quality of life could not be assessed and psychological or gender-identity investigations have not been carried out. None of these children have reached puberty, and hence decision and outcomes of definitive neophallus reconstruction has not been considered to date.

Conclusion
Scrotal flap phalloplasty is a minimally invasive, simple and reproducible technique used to create a temporary neophallus in boys with aphallia, while waiting for definitive reconstructive surgery after puberty.

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Summary Figure Long-term results (patients 1 and 2).

Introduction

Penile agenesis, or aphallia, refers to the complete absence of a penis in a normal genotypic male due to failed development of the genital tubercle [1]. Even if no *corpus cavernosum* or *corpus spongiosum* are noted, the scrotum is well-developed and the testicles are descended. The urethra is short and usually opens at the *linea pectina* or anus. **Newborns with aphallia have karyotype 46XY and the SRY gene which leads to male gonadal development. Other than the absence of a phallus, boys with aphallia are genotypically and phenotypically males**. Supporting **a cisgender approach, early** sex reassignment to female gender is no longer recommended in these patients due to conflicting results and high incidence of gender identity disorder [2]. To favor proper psychosexual development of a **cisgender** child with aphallia, a phallus should be created in early childhood. Various techniques have been proposed to create a neophallus prior to puberty, with variable outcomes and degrees of difficulty [1,2]. We present a multi-institutional experience, ten years after our initial description of the scrotal flap phalloplasty (SFP) technique, in which we use a flap from the well-developed scrotum to build a temporary neophallus, without obvious scars in patients with aphallia.

Methods

The records of surgical neophallus reconstruction for aphallia patients from 4 centers between 2011 and 2021 were reviewed. All patients had at least one year follow-up to assess for short and long-term complications. Age at initial operation, associated anomalies, and other related surgical procedures were analyzed.

All parents signed informed consents prior to these procedures. They all received counseling preoperatively and postoperatively by social workers, psychologists, pediatric endocrinologists, pediatric surgeons, pediatric urologists, and geneticists (multidisciplinary team). There was no standardized protocol for the multidisciplinary approach of these patients since this is a retrospective study performed in different countries (different health care systems, hospital regulations, etc.) and in different languages (English, Spanish, and Portuguese). Local hospital ethics committees and parents approved this publication.

Surgical technique

Scrotal flap phalloplasty (SFP) begins with demarcation of a wide rectangular-type mid-scrotal flap along with Glenn-Anderson lateral scrotal flaps (Fig. 1). Monopolar electrocautery as well as sharp dissection are used to incise the scrotum following the lines, incorporating generous amount of subcutaneous fat in the liberated flap to ensure blood supply, without reaching the fascia vaginalis. After medial approximation of the lateral scrotal margins, reconstructing the scrotal wall, the rectangular flap is then reconfigured into a cylindrical structure with 5—0 Monocryl interrupted sutures ventrally, beginning at the penoscrotal junction and progressing towards the distal aspect of the flap (Fig. 2). In secondary procedures, a rolled tube of AlloDerm® can be inserted within the shaft of the neophallus to improve the length, diameter, and appearance of the neophallus (Fig. 3). **Likewise, bulking agents (e.g. polyacrylate-polyalcohol copolymer) can also be later injected along the neophallus shaft with the same objective.**

Results

Six patients with aphallia underwent SFP at ages ranging from 11 months to 5 years.

Patient 1 was an infant born with imperforate anus, left-single kidney and aphallia. A colostomy and vesicostomy were performed during the first day of life. At six months of age, the imperforate anus was managed surgically and the urethra was taken down from the atretic anal canal and

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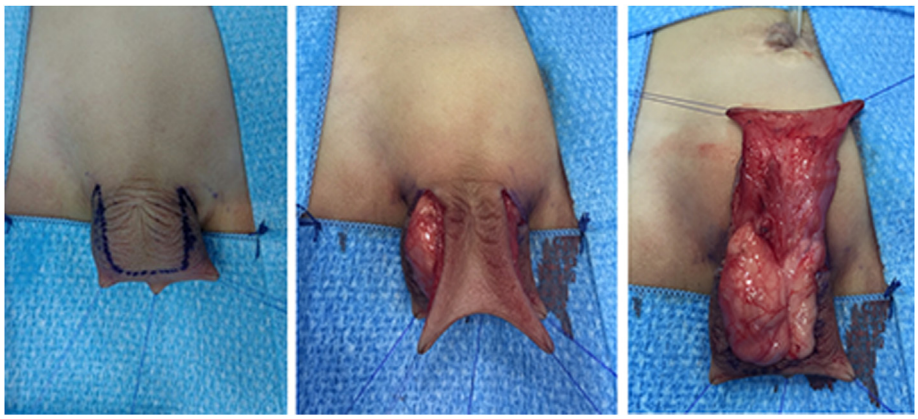


Fig. 1 Demarcation and harvesting of the scrotal flap.

relocated to the perineum (perineal urethrostomy). SFP was performed at 12 months of age.

Patient 2 was born with an anal stricture, patent urachus and aphallia. Colostomy was performed early in life, but urinary diversion was not required as the patent urachus served as an incontinent diversion. SFP was performed at 18 months of age.

Patient 3 was a boy with solitary left kidney, imperforate anus and aphallia, who underwent early colostomy and left cutaneous ureterostomy at two days of age. The ureterostomy was later taken down and a vesicostomy was fashioned. He was referred to us at 5 years-of-age when the vesicostomy was closed in exchange for a perineal urethrostomy with simultaneous SFP.

Patient 4 was an 11-month-old male with posterior urethral valves (PUV), bilateral vesico-ureteral reflux with an atrophic left kidney and aphallia, who underwent PUV ablation in addition to the SFP and urethral mobilization.

Patients 5 and 6 underwent SFP at ages 14 and 18 months, also with simultaneous urethral mobilization from anal verge to mid-scrotum or further. Secondary urethroplasty to advance the scrotal/peno-scrotal or perineal urethra distally is planned to be performed in all patients at the time of definitive phalloplasty.

Follow-up ranged from 1 to 10 years (median = 7 years). Only patient #5 developed a temporary skin dehiscence, which healed with local treatment. The aesthetic result in all patients was very satisfactory in the early post-operative period and has been maintained in the long-term follow-up, with all patients presenting a cylindrical structure resembling an uncircumcised penis, without evidence of significant contraction or loss of length (Fig. 4). According to parents' wishes, two patients underwent neophallus enhancement procedures with bulking substances, while undergoing later surgical interventions not directly related to the neophallus (with longitudinal injection of 2 cc of **bulking agent (polyacrylate-polyalcohol copolymer-brand name Vantris®)** in patient #1 and insertion of a rolled tube of AlloDerm® in patient #3 with satisfactory results (Fig. 4). Table 1 summarizes the characteristics of our patients.

Only the parents of two patients have opted to continue with ongoing psychological support. However, all of the parents consider their child is doing well and developing appropriately as boys.

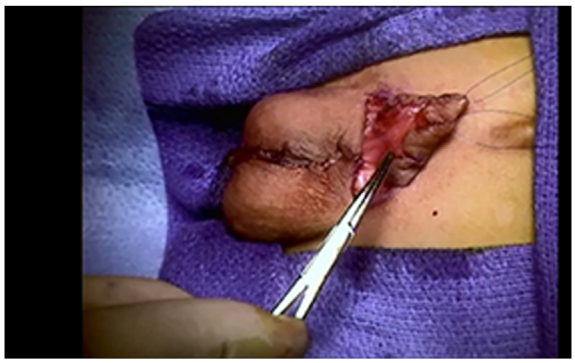


Fig. 2 Closure of the scrotum and tubularization of the flap, forming the neophallus.

Discussion

Penile agenesis is a rare disorder with an estimated incidence of 1 in 10—30 million births [1]. **This condition is associated with impairment in the formation of the genital tubercle in 46XY individuals. Accompanied** genitourinary anomalies are present in approximately 54% of the patients. **To** support **a cisgender approach of male identity in aphallia patients**, significant efforts have been undertaken to create a definitive neo-phallus.

The first penile reconstruction was performed in 1936 by Bogoraz consisting of a tubularized abdominal flap with autologous rib fascia [3]. This technique was advanced to free-flap and pedicle-flap phalloplasties in the 1970s and 1980s until the single-stage microsurgical urethral-phalloplasty tube-within-a-tube design using the free radial forearm flap was introduced [4—9]. The microsurgical techniques used today include extended pedicle island groin flap [10,11], innervated anterolateral thigh flap [12], lower abdominal skin flap [10], parascrotal flap [13], and scrotal flap [14]. All are complex procedures with long operative times, usually undertaken in adulthood as definitive procedures. They are accepted as the gold standard for phalloplasty in transsexual patients or those who lost their penis due to malignancy or accident, usually providing an adult-sized, functional neophallus, despite the high

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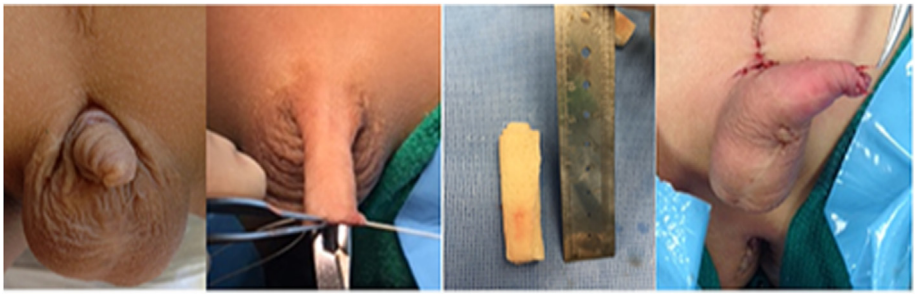


Fig. 3 Bulking procedure with insertion of Alloderm tube.

incidence of urethral strictures and urethrocutaneous fistulae.

Non-microsurgical neophalloplasty techniques in patients with penile agenesis are considered "temporary procedures" that help to establish and maintain body image and preserve the psychosexual development of the patient with aphallia. These techniques, most commonly employed in infants or children, do not involve tissue transplant from a distant region and are simpler to perform, with less scarring at the donor sites [15]. Oliveira et al. [15] published a systematic review combining outcomes of neophalloplasty using various techniques performed during infancy, favoring a two-stage approach: initial simple non-microsurgical neophalloplasty as early as possible, and later, after puberty, a more complex microsurgical reconstruction to restore sexual and urinary function.

In 2007, De Castro published the phalloplasty with abdominal flap technique (PAF), which utilizes a quadrangular lower abdominal skin flap [10]. He has performed PAF in infants younger than 2 years of age but reported complications when performing neophalloplasty and urethroplasty at the same time. Complications have also been associated with the urethroplasty only, suggesting that the phalloplasty alone is a more simple procedure and an eventual two-stage surgical plan is desirable [10]. A penile prosthesis could be inserted during adolescence or adulthood in a further stage, as reported by Gouvea [16].

Goyal et al. reported a three-stage neophallus reconstruction process, which includes urethroplasty at the second and third procedures [13]. After three years of follow-up, his patient was voiding in standing position with no apparent complications. However, hair growth in the urethra may cause future complications with the potential for additional procedures.



Fig. 4 Long-term results (patients 1 and 2).

Due to significant donor scars and considerable morbidity and complexity associated with the "definitive" phalloplasty techniques, we opted to create a simple, reproducible and straightforward procedure to serve as temporary neophallus in young boys with aphallia. The scrotum is the ideal donor site for a temporary neophallus in childhood, since boys with the disorder generally have a well-formed scrotum with normal testicles positioned inside. Furthermore, other donor sites are preserved for a definitive phalloplasty after puberty. A year after performing our first case of SFP [17], a similar scrotal phalloplasty technique was published by Bajpai et al., in 2012 [14]. In our opinion, the scrotal phalloplasty appearance achieved with their technique is suboptimal due to the lack of creation of rotational Glenn-Anderson scrotal flaps. Furthermore, contraction of their neo-phallus was documented at one year of follow up. Venkatesh et al. also performed a scrotal phalloplasty in a two-year-old patient with aphallia [18]. They opted to create a neo-urethra using a grafted tube of buccal mucosa, which eventually contracted. **If desired by the patient, scrotal flap phalloplasty (SFP) can be replaced after puberty by a definitive neophallus using any of the microsurgical phalloplasty techniques.**

Chaudhry et al. reported the use of AlloDerm graft for additional girth and vascularity of the neophallus during scrotal phalloplasty [19]. They used a second layer of tunica vaginalis as a flap to cover the matrix. After short-term follow-up (12 months) they observed appropriate cosmetic appearance without complications. Before Chaudry's report, we also used an AlloDerm® graft inserted as a rolled tube inside the neophallus in one of our patients, with the same goal of enhancing the diameter of the neophallus. At last follow-up, the phallus has remained appropriate in diameter and satisfactory to the patient and family. **We have also used a bulking agent (e.g. polyacrylate-polyalcohol copolymer-brand name Vantris®) which is a non-reabsorbable material commonly used to treat vesico-ureteral reflux [20], by injecting it along the neophallus shaft of one patient with satisfactory long-term results and no complications.**

There are obvious limitations to this multicenter, multi-surgeon study, as quality of life could not be assessed and most importantly, psychological and gender-identity investigations have not been carried out. None of these children have reached puberty, and hence decision and

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Table 1 Patients' characteristics.

Patient	Age at surgery	Associated malformations	Associated procedures	Complications	Follow-up (years)
#1	12 mo	Single kidney, congenital anal stricture	Colostomy, vesicostomy, anal cutback, scrotal urethrostomy	no	6
#2	18 mo	Imperforated anus, patent urachus	colostomy	no	9
#3	60 mo	Single kidney	Vesicostomy closure, perineal urethrostomy	no	10
#4	11 mo	Single functional kidney, vesicoureteral reflux, PUV	PUV ablation	no	1
#5	14 mo	Left solitary kidney	no	Skin dehiscence	1.5
#6	18 mo	no	no	no	1

outcomes of "permanent" neophallus reconstruction has not been considered to date. Regardless, our follow-up to date supports the adoption of 1-stage scrotal-based neophallus early in life, as a suitable temporizing procedure that appears to be adequate through pre-pubertal years.

Despite the best intentions of the parents and physicians to provide optimal treatment for these children with aphallia, it is likely that implicit and cognitive biases may have played a role in the surgical decision and the time for the intervention. In order to mitigate these biases, all authors involved in this study took into consideration evidence-based literature as well as the opinions of other members of the multispecialty team, in order to present up to date, unbiased, and assessable literature to the parents. This comprehensive information guided the parents to opt for early temporary phalloplasty while waiting for the child to participate in the decision to proceed with a definitive phalloplasty after puberty.

The challenges faced by pediatric urologists and surgeons who encounter a child with aphallia include both the lack of experience with the entity itself and the complexity of traditional phalloplasty techniques used to correct it. Since aphallia is extremely rare, efforts must be focused to create a simple and reproducible temporary phalloplasty technique with good outcomes. Non-experienced surgeons, leaving more complex procedures to be performed after puberty by experienced surgeons in these rare cases, can perform SFP satisfactorily. Considering this, we recommend performing urethroplasty at the time of the definitive reconstruction, since complications with early urethroplasty may affect the outcome of a temporary neophallus, and even compromise the possibility of a penile prosthesis in the future.

It is paramount to stress the relevance of a multidisciplinary team approach to support the creation of a temporary neo-phallus in early childhood, and subsequent definitive phalloplasty after puberty if there is evidence of congruence of male gender identity with male phenotype/genotype [21].

Conclusions

Scrotal flap phalloplasty is a minimally invasive, simple and reproducible technique used to create a temporary neophallus in boys with aphallia, **while waiting for definitive reconstruction if desired.** Additionally, SFP preserves other donor sites required by the microsurgical phalloplasty techniques.

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Conflict of Interest

None.

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