CASE REPORT

A stepwise treatment approach to neonatal genital prolapse

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INTRODUCTION

Neonatal genital prolapse (NGP) is a rare phenomenon that is typically associated with spinal cord malformations. Several treatment methods have been described, on a case report basis, ranging from digital reduction, the intravaginal self-retaining device for pelvic support, to more invasive partial labial fusion. In this report, we describe a case of severe congenital neonatal genital prolapse and the management approach pursued.

CASE REPORT

A full term 38-week female with a known antenatal diagnosis of myelomeningocele, was born in an uneventful spontaneous vaginal delivery. Her exam demonstrated normal external genitalia, however, a 4cm interlabial mass was protruding from the introitus (Figure 1). On the second day of life, her neural tube defect (NTD) was corrected by neurosurgery.

The degree of prolapse was severe, with near complete externalization of the vaginal canal. After successful digital reduction of the prolapse, the urethral meatus was identified in an orthotopic position. A catheter was placed without difficulty. A pessary was fashioned using a 10 Fr silicone Foley catheter coiled up into a disk and secured with silk suture (Figure 2), which was then placed in the vaginal vault. Over the following days, the pessary repeatedly fell out with complete re-prolapse during strong Valsalva maneuvers.

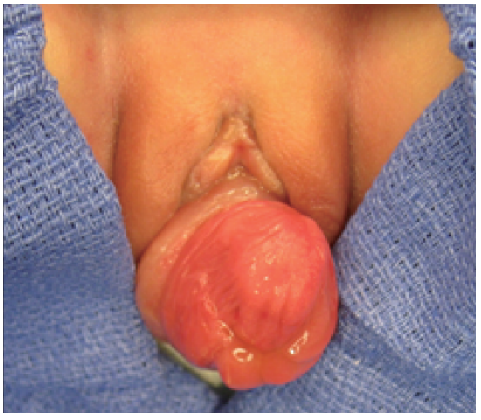

FIGURE 1 Thirty-eight-week full term neonate with genital prolapse, initial examination.

Given this unsuccessful attempt at pessary management, an 8 Fr silicone Foley catheter was inserted into the vaginal vault after reduction of the prolapse. The balloon was inflated with 10 mL of sterile water so it was seated above the ischial spines. Within an hour of placement, the patient's lower extremities became cyanotic, edematous, and pedal pulses were lost

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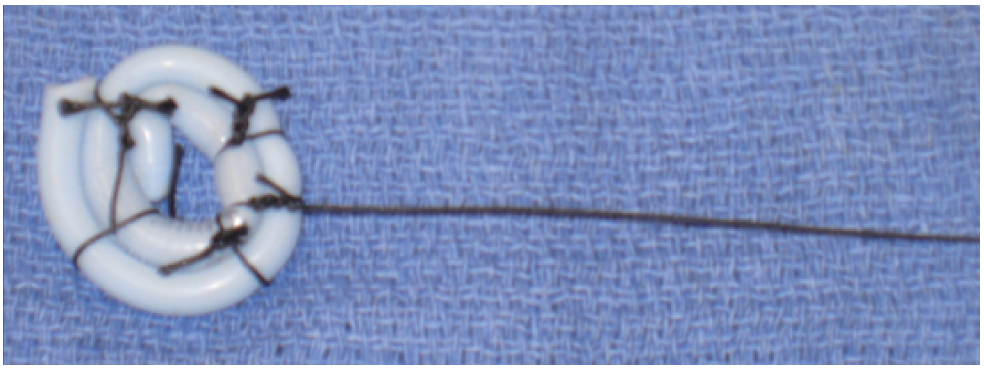

FIGURE 2 Pessary fashioned from 10Fr silicone catheter and silk suture material.

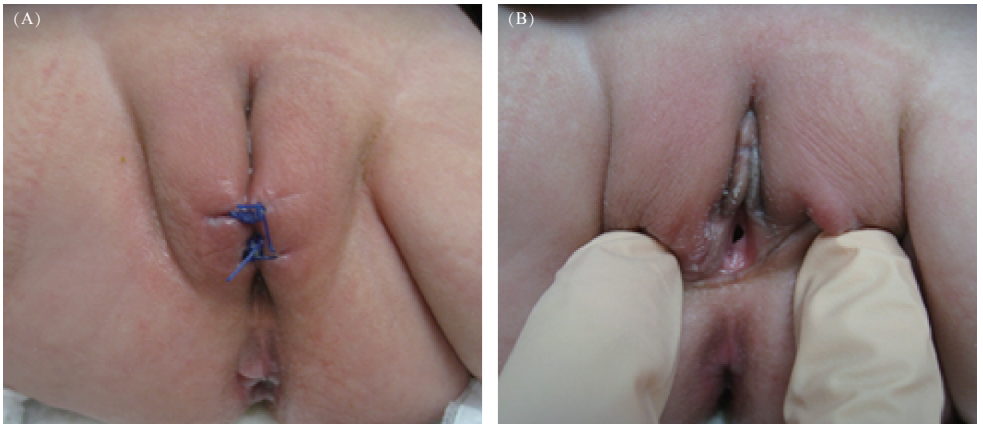

FIGURE 3 Partial labial fusion (A) and after removal of sutures (B).

bilaterally. The balloon was deflated with prompt return of color and pulses. The balloon was re-inflated with careful monitoring of pedal pulses and lower extremity perfusion, ultimately stopping at 7 mL. Despite proper positioning of the inflated balloon, the catheter was still repeatedly expelled.

The decision was made to pursue partial labial fusion. Two interrupted stitches of 0 nylon suture were placed across the labia majora at bedside (Figure 3). The sutures were secured tight enough to prevent prolapse, but with enough spacing to maintain access to the urethra for intermittent catheterization. Labial fusion was successful at maintaining continuous reduction. This was left in place for six weeks, and after suture removal, she had no further recurrence. Over a 4.5-year period thereafter, the patient's neurogenic bladder was managed on an outpatient basis. Her physical exam at each visit demonstrated normal Tanner 1 female external genitalia, without any recurrence of prolapse.

DISCUSSION

NGP, or procidentia, is concomitant with spina bifida in 86% of cases.[1] The association of NGP with neural tube defects is explained by dysfunctional sacral innervation to the levator ani muscles, leading to weakened support of pelvic organs.[2] As in adult pelvic organ prolapse, NGP is the extrusion of the vaginal canal, with or without the uterus, beyond the vaginal introitus. The urethral meatus remains orthotopic.

NGP is part of a differential diagnosis that includes vaginal polyps, urethral prolapse, paraurethral cysts, prolapsing ureterocele and rhabdomyosarcoma. NGP can be differentiated from conditions of urethral origin as the urethra should be cephalad to the prolapsed mass and easily catheterizable. Prolapsing ureteroceles are associated with ectopic ureters and duplicated systems, which can be identified with ultrasonography. Vaginal or uterine rhabdomyosarcoma presents with a bleeding interlabial mass that often has a classic "cluster of grapes" appearance.

There are a wide range of treatment modalities for NGP. The most conservative intervention is digital reduction, which can be effective for mild NGP. This has a substantial risk of recurrence. For moderate prolapse, intravaginal support can be utilized while pelvic structures mature during the neonatal period. Intravaginal self-retaining devices include pessaries, Foley catheter balloons and even inverted rubber nipples.[3,4] While these devices can achieve satisfactory results, they are limited by their ability to maintain position within the vagina, as reflected in this

case report. A unique complication of large intravaginal devices, which we unexpectedly encountered, is occlusion of vasculature within the pelvis causing interrupted perfusion to the lower extremities.

Beyond vaginal support, partial labial fusion and purse-string sutures of the vaginal wall have also been utilized. Partial labial fusion is a simple procedure that is well tolerated in patients with NTD because of decreased lower body sensation.[5] In the most severe of cases, surgical intervention is warranted. These range from cervical amputation and hysterectomy, now of historical interest, to uterine ventrosuspension, vaginal slings or sacral cervicopexy. While effective, surgery in the neonate is not without complications such as infertility.

This case report illustrates that partial labial fusion for NGP is a feasible option that avoids the pitfalls of intravaginal support, and the risks of surgery. A step-wise progression from conservative to more invasive interventions is a safe approach to managing NGP.

CONFLICT OF INTEREST

The authors have no conflicts of interest relevant to this article to disclose.

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