Surgical Management of Pelvic Organ Prolapse in a Woman With Achondroplasia

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BACKGROUND: Achondroplasia is an autosomal-dominant disorder resulting in short-limbed dwarfism. Limited data exist regarding the management of pelvic organ prolapse in women with achondroplasia.

CASE: A young nulligravid woman with stage IV uterovaginal prolapse desired surgical correction of her prolapse with uterine preservation. The severity of her prolapse, profound cervical elongation, and distortion of the bony pelvis presented surgical challenges. She underwent abdominal lumbohysteropexy with polypropylene mesh. At 1 year postoperative, she has marked improvement in symptoms and objective findings.

CONCLUSION: Abdominal lumbohysteropexy appears to be an effective surgical approach in women with achondroplasia desiring uterine preservation.

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Achondroplasia results from a gain-of-function mutation in the fibroblast growth factor receptor-3 gene on chromosome 4p16.3. Inheritance is autosomal dominant, and affected individuals display short-limbed dwarfism.[1] Although numerous medical conditions are associated with this disorder, pelvic organ prolapse has not been widely reported. Combining MEDLINE search terms "achondroplasia" and "prolapsed," one case report exists in the current medical literature.[2] We describe the management of stage IV uterovaginal prolapse in a woman with achondroplasia who desired uterine preservation.

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CASE

A 30-year-old nulligravida achondroplastic dwarf presented for surgical correction of symptomatic uterovaginal prolapse. She was sexually active and prioritized a surgical plan incorporating uterine preservation. She denied symptoms of urinary incontinence.

On examination, the patient was of short stature (48 inches) and had foreshortened limbs consistent with the achondroplastic phenotype. Pelvic organ prolapse quantification examination demonstrated stage IV uterovaginal prolapse (Aa=+3, Ba=+7, C=+7, GH=5, PB=3.5, TVL=9, Ap=–3, Bp=+5, D=–5). Bimanual examination was significant for palpation of the sacral promontory 4 cm proximal to the hymen. Magnetic resonance imaging showed the aortic bifurcation above L4–L5 vertebra, the sacrum at a 90-degree angle to the lumbar spine resulting in a pelvic inlet of 4.4 cm, and profound cervical elongation (Fig. 1).

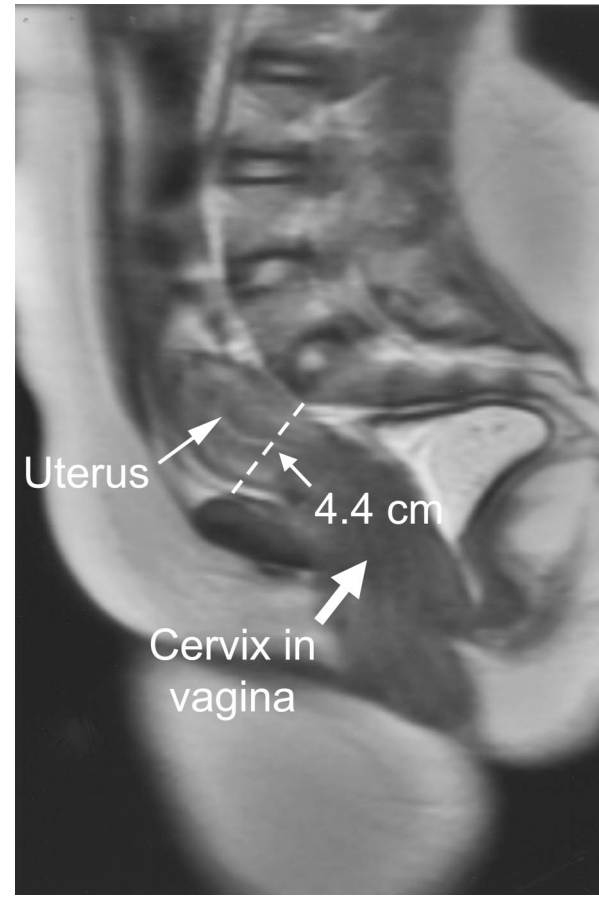

Fig. 1. Magnetic resonance imaging demonstrating the uterus (*thin arrow*), elongated cervix (*thick arrow*), and short distance between the pubic symphysis and sacral promontory (4.4 cm, *dotted line*).

Frankman. Prolapse in a Woman With Achondroplasia. Obstet Gynecol 2010.

We performed a laparotomy through Pfannenstiel incision. After intraoperative reduction of the prolapse, the lower uterine segment was at the level of L5–S1 vertebral bodies with the distal cervix at the level of the hymen. The location and orientation of the sacrum confirmed that it was inaccessible as an attachment point for hysteropexy. After identifying the ureters, great vessels, and their relationship to the lumbar-sacral region of the spine, we dissected over L4–L5 and placed three Gore-Tex (W. L. Gore & Associates, Inc., Elkton, MD) sutures into the anterior longitudinal ligament. The limited space between the pubic symphysis and sacral promontory (4.4 cm, Fig. 1) restricted access to the anterior lower uterine segment. Consequently, polypropylene mesh was only sutured to the posterior lower uterine segment. Before attaching the posterior lower uterine segment mesh to the lumbar sutures, the uterus was elevated to the level of the lumbar sutures to approximate the final suspension, and a vaginal examination was performed. Despite elevation of the uterus, the distal cervix remained near the level of the hymen. To reduce the bulk of the cervix within the vagina, a transvaginal partial trachelectomy, removing approximately 2 cm of cervical length, was performed. The posterior lower uterine segment mesh was subsequently attached to the lumbar sutures, and vaginal examination confirmed the distal cervix above the level of the hymen. A unilateral left paravaginal defect repair was performed for supplementary anterior vaginal support. We were unable to gain access to the right lateral anterior vaginal wall via the space of Retzius. Cystoscopy confirmed the absence of bladder injury and bilateral ureteral patency.

Postoperatively, the patient stated that her symptoms were markedly improved. She denied de novo urinary incontinence or defecatory difficulties. A 5-month postoperative pelvic organ prolapse quantification examination demonstrated good support of the anterior and posterior vaginal walls (Aa=–3, Ba=–2, Ap=–3, Bp=–3) and improvement in the central defect (C=–4, TVL=6.5, D=–6). Her vagina remained well supported at the 12-month postoperative visit, with the cervix palpable above the sacral promontory. She reported being sexually active without dyspareunia.

COMMENT

Achondroplasia is the most common nonlethal skeletal dysplasia, with an incidence of 0.5–1.5 per 10,000 births.[1] Orthopedic conditions, including spinal stenosis and craniocervical junction problems, are common in children and adults with achondroplasia. Compensatory lumbar lordosis often develops as a result of thoracolumbar kyphosis.[1] The pathogenesis of pelvic organ prolapse in women with achondroplasia is unknown. We hypothesize that profound lumbar lordosis may contribute to altered mechanical forces and increased intraabominal pressure, leading to progressive weakening of the connective tissue and supporting structures of the pelvic floor, ultimately resulting in prolapse.

Abdominal hysteropexy is one of several surgical approaches for the treatment of uterine and vaginal prolapse in women who request uterine preservation. Alternate approaches include laparoscopic or vaginal native tissue repair, including uterosacral or sacrospinous ligament suspension, and transvaginal mesh procedures.[3–6] A transvaginal native tissue approach was utilized in the prior case report.[2] We selected an abdominal procedure with mesh reinforcement for our patient owing to her young age, need for durability, advanced prolapse (including an elongated cervix that filled the vagina), and the bony abnormalities of her pelvis. We felt that access to the native ligaments was limited, and the patient's pelvic configuration made her an inappropriate candidate for a vaginal mesh procedure. Although attachment of mesh to the sacral portion of the anterior longitudinal ligament is advocated for restoration of the normal vaginal axis, our preoperative evaluation indicated that it was unlikely that we would have access to the sacrum and needed to consider the most cephalad attachment point while avoiding vascular injury.[6,7] Preoperative magnetic resonance imaging was helpful in predicting the relationship among the bifurcation of the aorta, the inferior vena cava, and possible attachment points for the mesh, as well as intraoperative challenges related to visualization and the shallow anterior-posterior diameter of the pelvis. In our patient, partial trachelectomy was necessary to elevate the distal cervix above the hymen. The current literature shows that prolapse becomes symptomatic in many women when the leading edge is at or beyond the hymen.[8]

Studies regarding the success of hysteropexy are limited, and the ideal candidate is unknown.[3,4] Based on the 12-month subjective and objective success in our patient, abdominal lumbohysteropexy appears to be an effective surgical approach in women with achondroplasia desiring uterine preservation.

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Pelvic Fracture With Complete Urethrovaginal Complex Avulsion

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BACKGROUND: The anatomic support of the urethra is controversial. We present a case of an intact urethra and vagina avulsing from the symphysis pubis after pelvic fracture.

CASE: An 18-year-old female pedestrian was struck by a motor vehicle. Operative reevaluation revealed an intact vagina and urethra; however, they were avulsed from the symphysis pubis. The avulsed urethra and vagina were reapproximated to the periosteum and surrounding soft tissue circumferentially with no urogenital sequelae.

CONCLUSION: Complete anterior vaginal wall avulsion may occur with sparing of the lower urinary tract in the setting of pelvic fracture. This case supports the current anatomic understanding of a strong and dense attachment between the urethra and the anterior vaginal wall.
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Pelvic ring fractures from high-energy blunt trauma are associated with significant organ injuries in more than 50% of cases.[1] Injury to the lower urinary tract in males is common with pelvic fracture probably because the male urethra is rigidly fixed to the pubic bone. However, in females these injuries rarely occur likely due to their more mobile urethra.[2] When traumatic female urethral injuries do occur, they usually disrupt the anterior vagina and urethral lumen. Gynecologic providers may be asked to evaluate and manage these urogenital lacerations and should be capable of identifying the variety of presentations. It should be known that the vagina and urethra can detach from the symphysis without any disruption to the vaginal epithelium or urothelium. The repair requires careful recognition of the avulsion and an anatomic reapproximation.

CASE

An 18-year-old morbidly obese nulligravid woman presented to our trauma service after a motor vehicle pinned her against a street lamp. Her injuries included a superior ramus fracture, avulsion of her knee cruciate ligaments, large left inner thigh laceration to the level of the femur, and an ankle fracture. In the operating room, the patient's habitus, orthopedic injuries, and heavy bleeding produced suboptimal visualization of the vagina and urethra. Packing was placed in what was presumed to be the vagina, and a suprapubic catheter was placed under ultrasound guidance. Subsequently, her right internal iliac and bilateral internal pudendal arteries were embolized secondary to persistent pelvic hemorrhage.

On hospital day 4, the patient returned to the operating room for external fixation of her pelvic fracture. After careful positioning by the surgical team, the urogynecology service removed the packs, which were found in the retropubic space. The service diagnosed a complete avulsion of the anterior vaginal wall from the pubic bone and surrounding soft tissue (Fig. 1). The posterior aspect of the symphysis pubis could be palpated. The presumed urethral meatus was identified on the detached vaginal tissue, and a catheter was passed easily. Cystoscopy confirmed an intact urethra and bladder. A transurethral foley catheter was inserted. Vaginal, cervical, and rectal exams were normal. The vagina and urethra were avulsed from nine to three o'clock positions. Circumferential interrupted 0 polyglycolic acid sutures were placed reapproximating the vagina to the avulsion site: laterally to soft tissue (connective tissue) and medially to periosteum on the underside of the pubic bone. The vagina was then packed for 48 hours to promote scarification and hemostasis. Reexamination on postoperative day 6 revealed an intact repair. The patient remained hospitalized for approximately 4 months for management of her orthopedic injuries and unrelated complications.

At 5 months, the patient had no interval genitourinary complaints. She denied prolapse symptoms, incontinence, or voiding difficulty. Menses resumed, and a comprehensive genitourinary examination was unremarkable. The repair appeared intact and well approximated (Fig. 2). She

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