Surgical Management of Pelvic Organ Prolapse in an Achondroplastic Dwarf

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Abstract

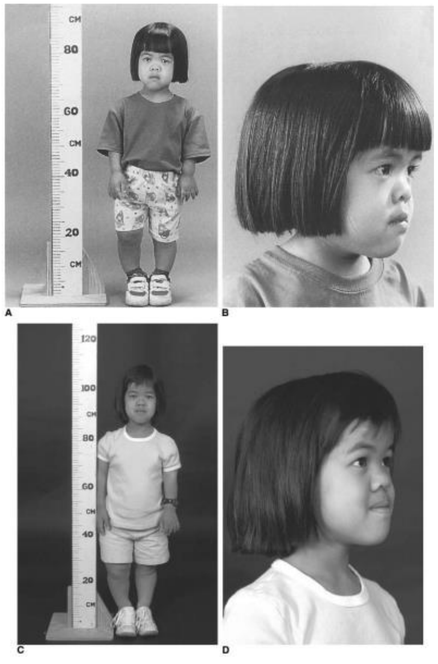
We report a case of stage IV uterovaginal prolapse in a woman with achondroplasia. The patient desired surgical correction of her prolapse with uterine preservation. She underwent abdominal sacrohysteropexy with polypropylene mesh. The postoperative visit was significant for marked improvement in symptoms and objective findings.

Introduction

Achondroplasia most common nonlethal skeletal dysplasia

- Gain-of-function mutation in the fibroblast growth factor receptor-3 gene
- Inheritance autosomal dominant
- Affected individuals display short-limbed dwarfism (Figure 1)
- Pelvic organ prolapse not widely reported as a disease-associated condition

Figure 1.



Child with achondroplasia at 6 years of age (A, B) and 12 years of age (C, D). Characteristic short stature and foreshortening of limbs.

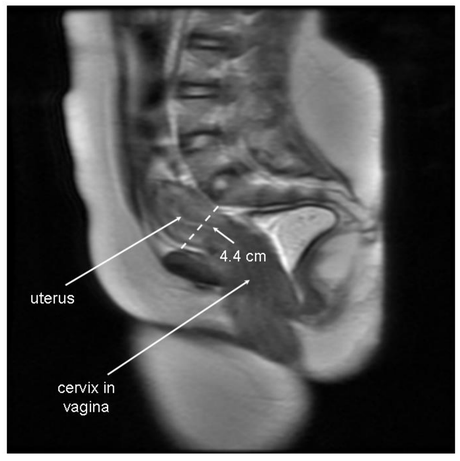
Cummings, et al. Otolaryngology: Head & Neck Surgery, 4th Ed. Mosby 2005.

Case Report

30 year old nulligravida with achondroplasia

- Stage IV uterovaginal prolapse
- Unable to retain pessary
- No urinary incontinence symptoms
- Sexually active
- Desired uterine preservation

Figure 2.



MRI demonstrating short distance between the pubic symphysis and sacral promontory (4.4 cm) and profound cervical elongation

POPQ Examination

Preop

Aa +3	Ba +7	C +7
GH 5	PB 3.5	TVL 9
Ap -3	Bp +5	D -5

5-Month Postop

Aa -3	Ba -2	C -4
GH 3	PB 4	TVL 6.5
Ap -3	Bp -3	D -6

- Exam unchanged at 12 month postoperative visit
- Sexually active without dyspareunia

Procedure

Laparotomy via Pfannenstiel incision

- Intraoperative challenges (Figure 2):
  - Lower uterine segment at level of L5-S1 with distal cervix at hymen after reducing prolapse
  - Shallow A-P diameter of pelvic inlet (4.4 cm) limited exposure and access to anterior vagina
- Polypropylene mesh sutured to posterior lower uterine segment
  - Mesh attached at L4-L5
- Paravaginal defect repair performed to address anterior vagina
- Partial trachelectomy to reduce bulk of cervix in vagina

Comment

Hypothesis:

Profound lumbar lordosis

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Altered mechanical forces

Increased intraabdominal pressure

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Progressive weakening of pelvic floor

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Symptomatic pelvic organ prolapse