Case Report

Neonatal infravesical obstruction in females

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ABSTRACT

Neonatal infravesical obstruction in females is less common than in boys and rarely reported. In this article, we report on 3 cases of infravesical obstruction in female neonates with different etiologies. We also reviewed the literature on this topic. Including our cases, 17 cases of neonatal infravesical obstruction in girls have been reported. Neonatal infravesical obstruction in females has different underlying pathology compared to boys with congenital tumors being more common in girls. A high index of suspicion is essential for early recognition and management.

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Infravesical obstruction in the neonate should be considered an emergency due to the potential long-term renal damage if left untreated. Although, we recognize and study the infravesical obstruction in male neonates, it is less common in female neonates. In boys, infravesical obstruction usually presents early during the neonatal and infancy period and is commonly caused by posterior and anterior urethral valves.[1,2] In girls, infravesical obstruction usually presents later in childhood and imperforate hymen is the main cause.[3] In this article, we report 3 cases of infravesical obstruction in female newborns and review the literature. We performed the English literature review using Medline and Cinahl databases. The search strategy used the terms urinary retention, retention of urine, bladder outlet obstruction, and infravesical obstruction in females and age group up to 23 months.

Case Report. ***Patient 1***. A 2.02 kg female was delivered by an elective cesarean section after 31 weeks of gestation. Antenatally, ultrasound scan at 18 weeks of gestation showed a cystic mass consistent with sacrococcygeal teratoma associated with urinary bladder distension and bilateral hydroureteronephrosis **(Figure 1a)**. Subsequent antenatal scans showed an increase in the size of the mass and progressive deterioration of the bilateral hydronephrosis and bladder distension. These changes were associated with oligohydramnios and 'mirror syndrome' in the mother. Postnatally, she had respiratory distress, which required ventilation. In addition, she developed bilateral pneumothoraces, which were drained. Abdominal examination revealed a pelvic mass, which was extending up to the umbilicus. Posteriorly, the tumor was extending to almost the back of the knees. Urethral catheterization produced 130 ml of clear urine. Clinically, the size of pelviabdominal mass was not affected by draining the bladder. Initially, there was a mild impairment of the renal function (creatinine: 0.12 mmol/L, urea: 8.0 mmol/L), which returned to normal by the end of the first week. At the age of 2 weeks, she underwent exploratory laparotomy. A multicystic tumor was found, which originated from the retroperitoneum just behind the vagina and rectum. It was pushing both ureters anteriorly. The tumor was extending inferiorly to the area between the coccyx and the anus and was huge enough to distort the levator ani and glutei on both sides. Approximately 200 ml of straw-colored fluid was

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aspirated intra-operatively, and the tumor was removed through a perineal approach after excising the distal coccyx. Histopathological examination revealed a mature teratoma with no evidence of malignancy. Postoperatively, she voided freely and the bladder distension disappeared. Ultrasound scan of the urinary tract and micturition cystourethrogram 6 weeks following surgery showed no abnormality. She was followed by serial serum alpha-fetoprotein, which continued to decrease over the first year postoperatively. A rise in alpha-fetoprotein occurred at 23 months of age. A CT scan suggested a recurrence of the sacrococcygeal teratoma, which was excised through a posterior approach. Histological examination revealed a yolk sac tumor, which has risen from the area of the previously excised teratoma. Postoperatively, she received chemotherapy. At 4-years follow-up, the patient continues to do well with the alpha-fetoprotein in the normal range (<10 μg/L).

Patient 2. A 4-week-old girl presented with a history of failure to thrive, lethargy and vomiting following normal vaginal delivery. Antenatal scans were normal. Physical examination revealed normal vital signs and a palpable suprapubic mass extending to above the umbilicus. Genitalia examination revealed a pinkish bulge at the introitus. Bladder catheterization produced 110 ml of urine, but the suprapubic mass did not disappear. Both serum urea (10 mmol/L [normal 2.6-6.8 mmol/L]), and creatinine (0.07 μmol/L [normal 0.050-0.11 μmol/L) were elevated. Ultrasound scan, showed an empty bladder, normal uterus and a large fluid-filled mass posterior and to the left side of the bladder consistent with obstructed vagina. Both kidneys were normal but the lower ureters were dilated. Examination under anesthesia confirmed the presence of imperforate hymen, for which hymenoplasty was performed. Colposcopy revealed no vaginal septum and the presence of a single cervix. Postoperatively, she voided well and her renal function parameters dropped back to normal levels. An ultrasound scan showed bilateral uretero- pelvic dilatation, which proved to be due to vesicoureteric reflux (right, grade VI, and left, grade V) **(Figure 1b).**

Patient 3. After 36 weeks of gestation, a female was born by an uneventful vaginal delivery. Antenatal scans showed left hydro-ureteronephrosis and right cystic kidney **(Figure 1c).** Apgar scores were 8, 8 and 9 at 1, 5 and 10 minutes. Subsequently, she developed respiratory distress syndrome and required continuous positive airway pressure after which she was intubated and ventilated for one day. Ultrasound examination on day 2 showed left hydro-ureteronephrosis, right small multi-cystic dysplastic kidney with no ureteric dilation, right-sided ureterocele and a distended urinary bladder. On urethral catheterization, the bladder drained 120 ml. At the age of 3 days,

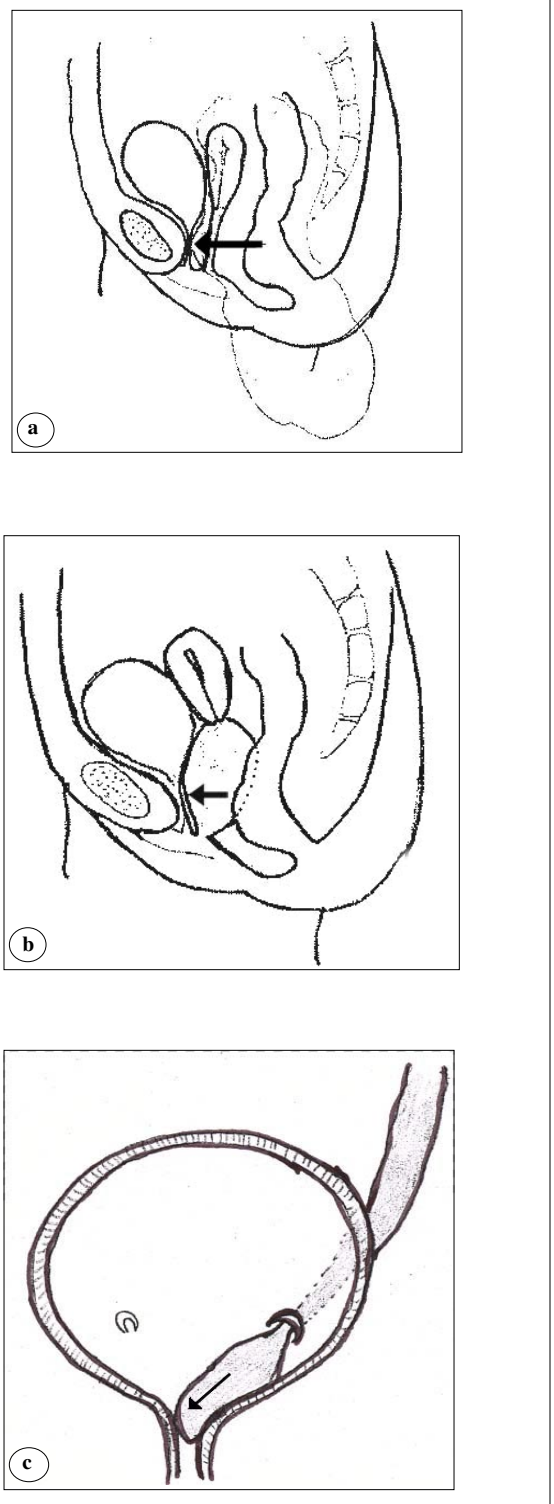

Figure 1 - Diagrammatic presentation of the 3 case reports, illustrating the mechanism of neonatal infravesical obstruction in females. The arrows indicate the direction of the obstructing mechanism **(a)** Sacrococcygeal teratoma **(b)** Hydrocolpos and **(c)** Ureterocele.

micturition cystourethrogram revealed right ureterocele and no vesicoureteric reflux. Ultrasound examination at the same day showed resolution of the earlier left sided hydro-ureteronephrosis; hence, the left side renal tract dilatation was probably due to bladder outlet obstruction caused by the prolapsing ureterocele. At the age of 5 days, cystourethroscopy was performed and the right ureterocele was incised after which she was voiding freely with no evidence of bladder distension.

Discussion. Early recognition and proper management of urinary retention in the neonate are essential in order to avoid short and long-term sequelae. Either mechanical bladder outlet obstruction or non-mechanical factors such as the effect of some medications, caused urinary retention in the neonate.[4] Bladder outlet obstruction is the main cause of urinary retention in newborn boys.[1,2] In female neonates, this condition is uncommon with only a few cases reported in the literature. Here, we report on 3 cases of mechanical infravesical obstruction in female neonates and review the literature on this topic. We found 14 cases of neonatal infravesical obstruction in females in 8 articles as shown in **Table 1**. Including our 3 cases, the reported causes of infravesical obstruction in female neonates include ureteroceles in a relative frequency of 23%, hydrometrocolpos (23%), neuroblastoma (23%), sacrococcygeal teratoma (18%), and rectal duplication (13%). Thirty-nine percent of cases presented at birth as a result of prenatal ultrasound diagnosis of pelvic pathology. With the wider use of prenatal ultrasound and the development achieved recently in this technique, we expect all cases of infravesical obstruction especially those caused by pelvic tumors to be detected prenatally.[5,6] Sacrococcygeal teratoma accounts for 18% of infravesical obstructions in female neonates. Although rare, it is the most common tumor of the sacrococcygeal region seen in the neonatal period.[7] There is a female predominance of 3:1.[7] Most of the tumors are benign and consist of fully differentiated mature tissues with less than 5% of the tumors being malignant at birth.[8] In our patient, the teratoma was mainly internal and therefore, a Altman type III tumor.[9] The internal or pelvic part of the teratoma resulted in bladder outlet obstruction and bilateral hydronephrosis due to pressure effects. Delay of treatment could have caused further renal obstruction and damage. Consequently, it is essential to look for signs of bladder outlet obstruction in such tumors and to relieve the obstruction by bladder catheterization until the patient is stable enough to undergo surgery.

Ureteroceles are well-recognized causes of bladder outlet obstruction. The incidence of ureteroceles at autopsy can be as high as 1 in 500 and occur most frequently in females.[10] Ureteroceles are increasingly diagnosed by prenatal ultrasound; however, we still diagnose many ureteroceles after causing symptoms. The most common presentation is that of an infant who has a urinary tract infection or sepsis.[10] If the ureterocele is large enough, it can obstruct the contralateral ureteric orifice or even the bladder neck as occurred in our patient. The antenatal ultrasound scan in our patient showed a cystic contralateral kidney with no ureterocele.

As shown in **Table 1**, hydrocolpos accounts for 23% of neonatal infravesical obstruction in females.

Table 1 - Reported cases of neonatal infravesical obstruction in females.

Author(s) and references	Year	Etiology of infravesical obstruction	Age at presentation	Volume of retained urine (ml)
Mann CM, et al[12]	1974	Neuroblastoma	birth	unknown
Touloukian RJ, et al[2]	1976	Ureterocele	birth	150 ml
Touloukian RJ, et al[2]	1976	Ureterocele	6 hours	50 ml
Touloukian RJ, et al[2]	1976	Hydrocolpos	24 hours	250 ml
Touloukian RJ, et al[2]	1976	Hydrocolpos	24 hours	150 ml
Touloukian RJ, et al[2]	1976	Neuroblastoma	16 days	250 ml
Pinter A[1]	1984	Ureterocele	6 days	unknown
Pinter A[1]	1984	Neuroblastoma	27 days	unknown
Pinter A[1]	1984	Neuroblastoma	21 days	unknown
Zaninovic AC, et al[6]	1992	Sacrococcygeal teratoma	birth	350 ml
Rauch MK, et al[13]	1993	Rectal duplication	birth	300 ml
Elchalal U[5]	1995	Sacrococcygeal teratoma	birth	180 ml
Emir H, et al[14]	2001	Hydrometrocolpos	birth	unknown
Park WH, et al[15]	2001	Rectal duplication	7 days	unknown

Imperforate hymen occur in 1 in 1,000 female newborns. It is caused by failure of the vaginal plate to completely canalize. This typically occurs by the 8th week of gestation. Westerhout et al,[11] reported hydrocolpos with an overall incidence of 1 in 16,000 female neonate. The cervical and vaginal glands secrete the accumulated mucinous fluid. Imperforate hymen usually presents at puberty with primary amenorrhea, cyclic abdominal pain and hematocolpos. In cases presenting in neonates and infancy, the hymen appears pink and bulging. Typically, the fluid collection is sterile. Elective drainage and hymenectomy are indicated to prevent urinary retention and secondary infection. Our patients presented at 4 weeks with symptoms of urinary obstruction and sepsis and therefore, the need for urgent drainage. This could have been avoided by a thorough physical examination of the genitalia at birth. Congenital pelvic tumor or mass due to pressure effect can also cause infravesical obstruction in female neonates. Examples of such disorders, reported in the literature, include neuroblastomas and rectal duplication. As stated earlier, congenital pelvic tumors cause more than 40% of infravesical obstruction in female neonates. Congenital cystic disorders such as ureteroceles cause the remaining cases.

In conclusion, neonatal infravesical obstruction in females is less common compared to male neonates. The etiology in both genders is different with congenital tumors being more common in girls. Therefore, a high index of suspicion and early management is essential to avoid the long-term sequelae. The principles of management include early decompression of the urinary obstruction followed by definitive treatment of the underlying lesion.

References

1. Pinter A. Bladder outlet obstruction in the neonate. ***Acta Paediatr Hung*** 1984; 25: 355-362.
2. Touloukian RJ, Weiss RM, Burke WR, Schiff M. The obstructed bladder syndrome in the neonate. ***Surg Gynecol Obstet*** 1976; 143: 965-969.
3. Hammad FT, Alawadhi N. Acute urinary retention associated with unilateral hematocolpos and ipsilateral renal agenesis. ***Saudi Med J*** 1997; 18: 523-524.
4. Das UG, Sasidharan P. Bladder retention of urine as a result of continuous intravenous infusion of fentanyl: 2 case reports. ***Pediatrics*** 2001; 108: 1012-1015.
5. Elchalal U, Ben-Schachar I, Nadjari M, Gross E, Appleman Z, Caspi B. Prenatal diagnosis of acute bladder distension associated with fetal sacrococcygeal teratoma: a case report. ***Prenat Diagn*** 1995; 15: 1160-1164.
6. Zaninovic AC, Westra SJ, Hall TR, Sherman MP, Wong L, Boechat MI. Congenital bladder rupture and urine ascites secondary to a sacrococcygeal teratoma. ***Pediatr Radiol*** 1992; 22: 509-511.
7. Salaymeh MT. Giant sacrococcygeal teratoma in the newborn. Case report and review of the literature. ***Int Surg*** 1971; 56: 56-60.
8. Grosfeld JL, Ballantine TV, Lowe D, Baehner RL. Benign and malignant teratomas in children: analysis of 85 patients. ***Surgery*** 1976; 80: 297-305.
9. Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American Academy of Pediatrics Surgical Section Survey-1973. ***J Pediatr Surg*** 1974; 9: 389-398.
10. Coplen DE, Duckett JW. The modern approach to ureteroceles. ***J Urol*** 1995; 153: 166-171.
11. Westerhout FC Jr, Hodgman JE, Anderson GV, Sack RA. Congenital Hydrocolpos. ***Am J Obstet Gynecol*** 1964; 89: 957-961.
12. Mann CM, Leape LL, Holder TM. Neonatal urinary ascites: a report of 2 cases of unusual etiology and a review of the literature. ***J Urol*** 1974; 111: 124-128.
13. Rauch MK, Martin EL, Cromie WJ. Rectal duplication as a cause of neonatal bladder outlet obstruction and hydronephrosis. ***J Urol*** 1993; 149: 1085-1086.
14. Emir H, Yesildag E, Soylet Y, Senyuz O. Association of biliary atresia and urogenital sinus. ***J Pediatr Surg*** 2001; 36: 635-637.
15. Park WH, Choi SO, Park KK. Cystic rectal duplication: a rare cause of neonatal bladder-outlet obstruction and hydronephrosis. ***Pediatr Surg Int*** 2001; 17: 221-223.