

SWISS SOCIETY OF NEONATOLOGY

A paraurethral cyst or the  
mandatory peek into the  
diapers of newborn girls

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This girl was born at 38 1/7 weeks of gestation at our institution. Antenatal history, pregnancy and maternal serologies were unremarkable. Delivery was by primary caesarean section due to breech presentation. Apgar scores were 9, 10, 10 at 1, 5 and 10 minutes, respectively. Arterial cord pH was 7.31.

On the routine physical examination on day three of life, the external genitalia appeared normal at first (Fig. 1A), but when the major labia were pulled apart, a tumefaction was noted (Fig. 1B). The surface of the tumefaction was smooth and covered with thin vessels. It was slightly displaced to the right. The urethral meatus was identifiable at 1 o'clock, whereas the hymen was displaced laterally and inferiorly. Catheterization of the bladder was not attempted since voiding was not impaired. There was no vaginal discharge and meconium was passed normally. The physical examination was otherwise normal.

The incidence of vulvar tumefactions in newborn girls varies between 1:500 and 1:7000 (1, 2). In a retrospective review over a period of 20 years, 26 newborns with vulvar tumefactions were observed: 11 had a paraurethral cyst, 10 a hydrocolpos, 2 an ectopic ureterocele and 3 were not further described (3). 6 of these cases were associated with vaginal bleeding, urinary tract infection (UTI) and intraabdominal masses or pain.

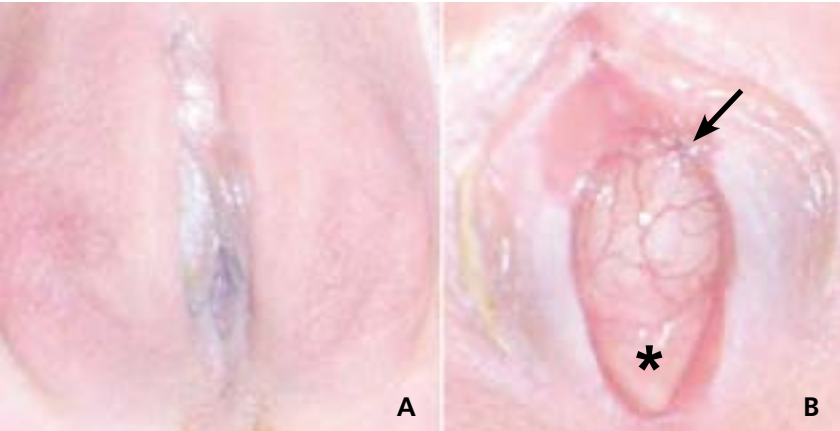
## CASE REPORT

## DISCUSSION

Although these entities are rare, a timely diagnosis is crucial, as one of the differential diagnoses is rhabdomyosarcoma of the vagina (botryoid sarcoma). The prevalence in the Intergroup Rhabdomyosarcoma Study (IRS) patients (n=3217) of neonatal rhabdomyosarcoma was 14; of these, 3 were vaginal rhabdomyosarcomas (4). The main symptoms are vaginal bleeding and/or the appearance of a grape-like (“botryoid”) mass outside of the vagina (Fig. 2A). Local metastasizing is frequent.

Urethral prolapse is rare in newborns and occurs more commonly in teenaged girls and postmenopausal women. It is more frequent in afro-american girls. Typically, the prolapsing and edematous mucosa encircles the urethra and may bleed slightly (Fig. 2B). Resection of the protruding urethra and reanastomosis with its proximal part is usually performed (5).

A prolapsed ectopic ureterocele is sometimes diagnosed prenatally by ultrasonography. The ureterocele protrudes through the urethral meatus and voiding occurs circumferentially around the mass (Fig. 3A). The majority of infants with this condition can be treated conservatively with antibiotic prophylaxis to avoid urinary tract infections since in the majority of cases the condition resolves. However, in the presence of renal outflow obstruction, vesico-ureteral reflux > III or bladder neck obstruction, surgical correction is indicated (6).

**Fig. 1**

*A) External female genitalia without traction on the labia majora and B) paraurethral cyst visible under traction on the labia majora with laterally displaced meatus urethrae (arrow head) and inferiorly displaced hymen (asterisk).*

Hydro(metro)colpos is caused by an imperforated hymen. Accumulation of retained secretions or blood shines through the hymen and appears as a grayish or bluish mass (Fig. 3B). The presence of an imperforated hymen without retention of secretions or blood can easily be missed. Once the diagnosis is made, ultrasonography is indicated to rule out ascites, renal obstruction and abdominal masses (7). Incision of the imperforated hymen may be necessary.

The key feature in paraurethral cysts is the lateral displacement of the urethral meatus (Fig. 4A). The pathogenesis of paraurethral cysts is not completely understood. Obstruction of the Skene's duct draining the paraurethral glands is postulated. Alternatively, cystic degeneration of involuting embryological remnants such as the urogenital sinus and Müller's or Gartner's duct may lead to cyst formation. The diagnosis is usually made by clinical examination. Further imaging is only required if voiding is impaired. In our case, a gynecological consultant confirmed this diagnosis and recommended conservative management. At a follow-up visit 2 weeks later, the cyst had disappeared. In a literature review of 41 cases, 14 cysts ruptured spontaneously within the first weeks of life. Spontaneous rupture becomes less likely after 8 weeks of age. Persistence of the cyst beyond this time may be an indication for surgical treatment (excision, marsupialisation or aspiration) (8).

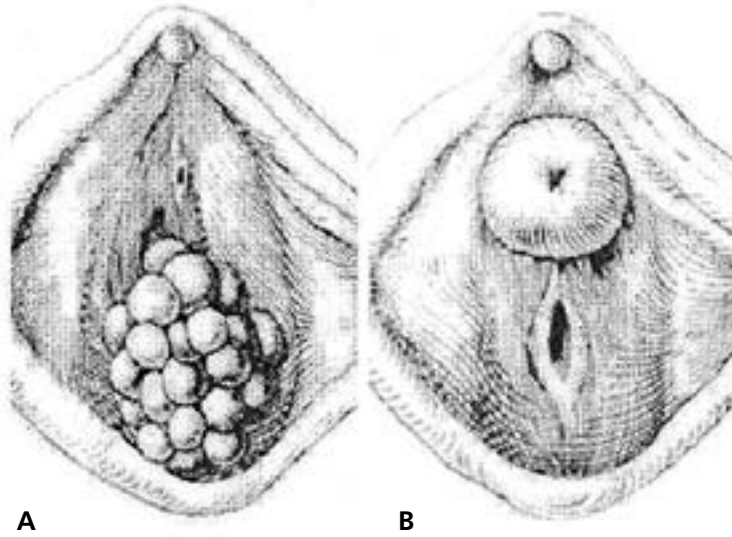


Fig. 2

*A) Rhabdomyosarcoma of the vagina (botryoid sarcoma), B) urethral prolapse.*

*A) Prolapsed ectopic ureterocele, B) hydro-(metro)colpos.*

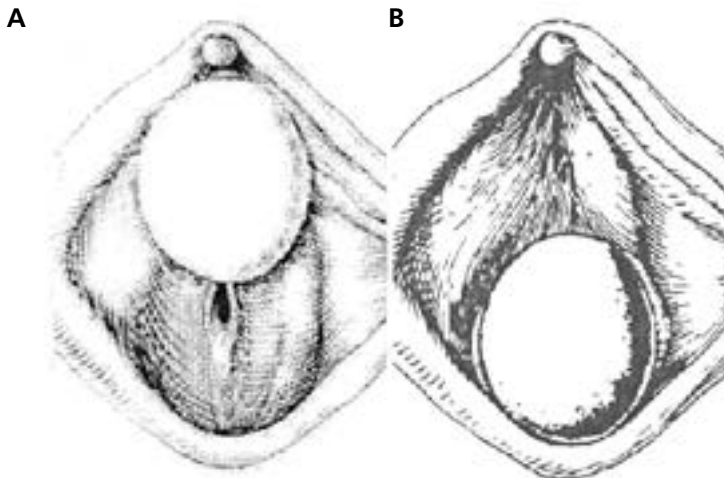


Fig. 3



Fig. 4

*Paraurethral cyst.*



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