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Müllerian Duct Syndrome

Müllerian Duct Syndrome (MDS) refers to a genetic disorder of male pseudohermaphroditism (46 XY karyotype) characterized by normal masculinization of the external genitalia and the presence of uterus and fallopian tubes. Most cases are discovered during surgery for undescended testis, inguinal hernia or transverse testicular ectopia. MDS is caused by a deficient activity (most cases) or receptor insensitivity of antimüllerian hormone (also known as Müllerian inhibitor factor). This hormone is produced by testicular Sertoli cells and is responsible of producing fetal regression of Müllerian structures (uterus & fallopian tubes) in genetic males. Most cases are transmitted as autosomal recessive restricted to males (sex-linked). Anatomic variants include fallopian tube or uterus within the inguinal canal, testis and tubes in a hernia sac or bilateral cryptorchidism with the testes embedded in the broad ligaments. The vas deferens is intimately adhered to the uterus lateral wall. Initial procedure consists of hernia repair, replacement of structure within pelvis and karyotype. After diagnosis follow-up management has been controversial. A few suggest partial removal of the uterus (leaving vas deferens intact on a thin pedicle of myometrium) and fallopian tubes with testicular fixation. Most content that surgical excision of persistent MDS structure may result in ischemic or traumatic damage to the vasa deferentia and testes and optimal management is orchiopexy leaving the uterus and fallopian tubes in situ. The testis in MDS are at risk of malignant degeneration.

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Bleeding Nipple

Breast enlargement is commonly seen in newborns babies, a condition associated with clear or milky nipple discharge. Maternal hormones are considered the culprit. On rare occasions the mother of an infant will bring to you the attention that the child is having intermittent episodes of bleeding through the nipple. In infants it is a benign, self-limited condition that should be managed conservatively (it could take six months to go away).

The main reason of bloody nipple discharge is mammary ductal ectasia, which extends down to the collecting tubules. Mammary duct ectasia was first reported in 1983, characterized by dilatation of the subareolar duct system, and by inflammatory reaction and fibrosis. The infant's own endocrine system is responsible for breast enlargement and mammary duct ectasia, though infection has also been postulated as etiologic factor. Surgical procedures should be avoided, because injury to the breast bud may cause permanent damage. Stimulation or massages to the breast should also be avoided. When associated with hypertrophied mammary glands, prepubertal gynecomastia should be sought.

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Pilonidal Sinus

Pilonidal sinus disease (PSD) is the result of a chronic inflammatory reaction of the subcutaneous tissue of the low back from midline skin orifices that ingest hair shafts in hirsute buttocks. PSD arises from hair follicles in the skin, appears after puberty and affects mostly males adolescents. Male sex, adolescence and a familial disposition are associated with the development of PSD. Minor local trauma ('Jeep Disease' in military personnel) and overweight are the most important factors for development of symptomatic PSD. Primary closure after excision of PSD has frequently been complicated by wound breakdowns. Sepsis and hematoma formation are the main causes of wound breakdowns. PSD is best treated in the acute situation with follicle removal and lateral drainage. The method is suited to outpatient management and gives minimal disability and good long-term control. In the chronic situation conservative therapy (meticulous hair control by natal cleft shaving, perineal hygiene, and limited lateral incision/drainage for abscess) effectively controls PSD in the nonoperative outpatient setting while promoting near-normal work status preferred over excisional operations.

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