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VACTERL

The VACTERL association was described by Quan and Smith in 1973 as a group of congenital anomalies with a nonrandom tendency for concurrence. V = vertebral anomalies (hemi- and bifid vertebrae), A = anorectal malformation (imperforate anus), C = congenital cardiac defects (VSD, ASD, Tetralogy of Fallot), TE = trachea-esophageal anomalies (esophageal atresia), R = renal-urinary defects (absent kidney, hypospadias) and L = limb defects (radial dysplasia). The mesodermic defect has been traced to the third week of intrauterine life. Other associated problems are a single umbilical artery, duodenal atresia, Meckel's diverticulum. Cases are preferentially males, with higher perinatal mortality rates, higher frequency of fetal loss in previous pregnancies and lower mean birth weights. Most patients have normal brain function. VACTERL is generally described whenever an infant born with esophageal atresia has two or more of this associated defects. Those associated with vertebral, ribs or sternal anomalies may harbor a high lying upper esophageal pouch. Heart failure is the major cause of mortality. Infants with VACTERL that included both renal anomalies and anorectal atresia is more likely to have genital defects. Preaxial but not other limb defects are associated with any combination of the four nonlimb anomalies. Growth retardation can be seen during the first three years of life. Despite multiple operative procedures infants with VACTERL association may lead a reasonably normal life.

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Vaginal Rhabdomyosarcoma

Vaginal rhabdomyosarcoma presents during the first two years of life with vaginal bleeding, discharge and prolapse associated with a vaginal mass. The diagnosis is made by vaginoscopy and biopsy of the lesion. Most cases are of the embryonal histologic subtype (botryoid). The lesion usually arises from the anterior vaginal wall around the embryonic vesico-vaginal septum (urogenital sinus). This means that structure such as bladder, prostate and lower vagina might be involved with tumor extension. Initial work-up

must include biopsy, cystoscopy, vaginoscopy and pelvic/chest CT-Scan. Since the tumor is very chemosensitive most patients are managed with primary combination chemotherapy (pulse VAC plus adriamycin and cisplatinum). Repeated biopsy and chemotherapy without resection may be adequate for many patients. Radiotherapy and surgical resection are reserved for cases with residual disease. Sequelae of irradiation (colorectal, vaginal, urethral, and ureteral stenosis) can be seen in these children. Tumors with diameters of five cm or more have a less favorable outcome compared with smaller tumors. Although late relapses do occur, they can usually be salvaged by a combination of chemotherapy, radiotherapy and surgery.

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Cystic Neuroblastoma

Suprarenal masses in the newborn period include a differential of adrenal hemorrhage, simple cyst, abscess, neuroblastoma, renal duplication, hydronephrosis, renal cyst, nephroblastoma, pancreatic cyst, hepatic cyst, choledochal cyst or duplication cyst. Hemorrhagic pseudocyst (adrenal hemorrhage) accounts for more than 80% of all adrenal cystic lesions. Cystic neuroblastoma (NB) is a rare form of neuroblastoma. Development of cysts may be related to a prominent microcytic arrangement of tumor nests. US characteristics of NB are usually of a complex or echogenic mass with a thick complex wall. Solid tissue within the mass suggests the possibility of a tumor. Serial US exam and urinary collection of VMA and HVA is warranted. In situ NB can be an incidental finding in a small percentage of autopsies done for other reason. Therapeutic decisions are dictated by symptoms, size and potential complications. Given the good prognosis of cystic NB in the newborn period it seems feasible to avoid prompt surgery after birth for patients with an adrenal cyst without any evidence of metastasis, reserving indications for surgery until they fail to resolve after several weeks of follow-up.

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