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Duplex Kidneys

Duplex kidney also known as duplex collecting system is the most frequent congenital anomaly of the urinary tract carrying an incidence of approximately 1%. The diagnosis is defined as a renal unit comprised of two pelvicalyceal system. Duplex kidneys are further classified as either complete or partial. The complete variety consist of two separate pelvicalyceal system arising from two ureteral buds from the mesonephric duct. The two ureters fuse separately to the mesenchyme and leads to independent draining sections of the kidney described as upper or lower moiety ureters. The upper moiety ureter tends to insert more caudally and medial into the bladder than the normally inserting lower moiety ureter (Meyer-Weigert rule). The lower moiety ureter is often affected by vesicoureteral reflux (VUR) In the partial variety there occurs a range of incomplete duplications within a kidney and ureter leading to bifid moieties and ureter, but a single ureteric insertion distally. Duplex kidneys are found to be two times more common in females and bilateral in less than 20% of children. Most duplex abnormalities are asymptomatic carrying no clinical significance. A minority of duplex kidneys can be associated with main clinical manifestations such as urinary tract infection, urinary incontinence, dysuria, and the presence of an abdominal mass. Some of the most frequent complications are associated with ectopic insertion of the upper moiety ureter in the complete duplex kidney, often occurring with an ureterocele. In females this can cause incontinence since the insertion often occurs below the urethral sphincter. In males, fluid slowly drain into an accessory sexual structure producing a pelvic cystic mass. Another complication of duplex kidneys is VUR of the lower moiety caused by lateral displacement of the ureters insertion into the bladder predisposing the patient to recurrent urinary tract infections and renal scarring with permanent functional impairment of the kidney. VUR is regarded as the most common abnormality associated with complete ureteral duplication. VUR which occurs in 30% of cases is much more likely to occur into the lower pole ureter because it inserts laterally into the bladder and has a shortened intramural segment. Still, other complications of duplex kidneys include a multicystic dysplastic moiety and pelvic-ureteric junction (PUJ) obstruction. PUJ obstruction occurs most commonly in the lower moiety. The complication that occurs in duplex kidneys is specific to the moiety involved. Upper moiety complications are the most common being ectopic ureteric insertion, with or without ureterocele and multicystic dysplastic moiety. Lower moiety complications are VUR, renal scarring and PUJ obstruction. Any ureter that does not insert onto the trigone may be considered ectopic. If the ectopic ureter is located in the proximal female urethra, the result is usually obstruction, reflux or both presenting with urinary tract infection. If the location of the ectopic ureter is distal to the external sphincter, the child presents with continuous incontinence despite a

normal voiding pattern. An ectopic ureter can also insert into the vestibule, vagina, or uterus. An ectopic ureter in male inserts most commonly into the prostatic urethra, though may insert into the ejaculatory duct, seminal vesicles, or vas deferens presenting with urinary tract infection or epididymitis. The first imaging modality to diagnose a duplex kidney is abdominal ultrasound whether prenatally or postnatally. Further imaging needed to characterize the anomaly includes nuclear medicines studies (DMSA), voiding cystourethrograms, CT-scan, MRI and cystoscopy. A MAG3 renogram may confirm the diagnosis of a duplex kidney. VUR is the most common anomaly associated with renal duplex systems and may occur in both moieties but is more frequent in the lower moiety. CT angiography can be performed to offer detailed information on renal vessels in case a partial nephrectomy is needed. The goals of management of duplex kidneys is the preservation of functional renal parenchyma, elimination of infection, obstruction and VUR with maintenance of urinary continence. Reflux is managed with antibiotics and frequent radiological follow-up with spontaneous resolution in 50% of children. Recurrent infection or renal scarring are indications for surgery. In patients with ectopic ureter treatment depends on function of the upper pole segment. With poor or absent function upper pole heminephrectomy and ureterectomy is indicated. Partial or heminephrectomy in a duplex kidney is a procedure performed for the complicated clinical course of the disease. The most common indications for surgery in children are recurrent urinary tract infections, ureterocele with hypo- or a functioning moiety and ectopic ureter causing incontinence in girls. An upper urinary tract approach (heminephroureterectomy) is described for a poorly functioning upper moiety or an ectopic upper moiety ureter. A lower approach (bladder reconstructive surgery) with excision of ureterocele and ureteric re-implantation has been described for children with good lower moiety function and high grade VUR. Ureterocele management has tended toward transurethral endoscopic incision. A more complete ureterocele excision is necessary following transurethral incision or if an ureterocele is very large or ectopic causing bladder outflow obstruction.

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Congenital Cystic Lung Lesions

A lung cyst is a cystic parenchymal airspace measuring one centimeter or more in diameter with a well-defined thin wall usually less than 2 to 4 mm thick. The term cavity is used for air-containing lesion with a relatively thick wall greater than 4 mm. Cystic lung lesions may contain either air or fluid or a combination of both, often with an air-fluid level. Most children with congenital cystic lung (CCL) lesions do not present complications such as respiratory failure, pneumonia, or lung abscess, immediately after birth or for the first few months of life. Indications for lung resection in patients with CCL lesions should be determined by the risk of developing infectious complications, respiratory distress, pneumothorax, hemorrhage, or malignant transformation. The cystic congenital lung lesions encountered in children include bronchogenic cyst, congenital pulmonary airway malformation (CPAM), congenital lobar emphysema, bronchial atresia, and pulmonary sequestration. Bronchial atresia and congenital lobar emphysema are not truly cystic lesions but have a focal overinflated lung that mimics cystic lung changes. Most CCL lesions are thought to be caused by in utero airway obstruction, often with an aberrant, stenotic, or atretic supplying airway. Most CCL lesions are initially discovered by prenatal US which can be further evaluated by prenatal MRI. CCL lesions are largest in size in the second trimester and tend to decrease in size or remain stable in the third trimester. Large or rapidly expanding lesions may cause hydrops fetalis and demise related to central venous compression by the lesion. When imaging is performed soon after birth, CCL lesions can be fluid filled, dense opacities that mimic a solid mass and there is delayed clearance of fetal lung fluid and replacement with air due to an obstructed or abnormal airway connection. Postnatally, CT angiography should be used to further characterized CCL lesions including any infradiaphragmatic feeding artery, as seen in pulmonary sequestration. Differential diagnosis for lesions discovered postnatally include diaphragmatic hernia, infection, lymphatic, vascular or neoplastic cystic lesions. Bronchogenic cysts arise due to abnormal budding of the foregut at 26-40 days gestation, content is serous fluid to thick mucus or blood. Imaging reveals mediastinal (most common) or less commonly intrapulmonary mass with smooth or lobulated borders and smooth enhancing wall. Air trapping or atelectasis distal to the lesion with airway compression can result in recurrent pneumonia. The most frequent symptoms is pain, cough, fever, respiratory distress or dyspnea. They are found in the mediastinum or pulmonary parenchyma, neck, pericardium, pleura, diaphragm, or abdominal cavity. Intrapulmonary cysts are 20% of all bronchogenic cyst occurring in the lower lobes. CPAM accounts for 25% cases of lung cyst and consist of a group of macrocystic or microcystic pulmonary malformations due to early lung maldevelopment. They are divided into large-cyst type 1 (2 to 10 cm cysts), small-cyst type 2 (cysts 5mm to 2cm), and microcystic or solid type 3 (cysts < 5 mm). CPAM occurs equally in either lung, usually unilobar, very rarely multilobar or bilateral. Congenital lobar emphysema (CLE) is characterized by over-distension and air-trapping in the affected lobe, concomitant compression of the remaining lung tissue, and displacement of the mediastinum by herniation of the emphysematous lobe across the anterior mediastinum into the opposite side of the chest. Deficiency of the cartilage wall of the affected bronchi suggest a developmental cause. Hyperlucency of the affected lobe and herniation to the contralateral

side are the most common radiographic findings. Pulmonary sequestration is defined as lung tissue with no normal connection to the tracheobronchial tree associated with aberrant systemic arterial blood supply from the lower thoracic or lower abdominal aorta reaching the lung via the inferior pulmonary ligament. Is divided into intralobar or extralobar types. Venous drainage is via the pulmonary veins in the intralobar and the systemic veins in the extralobar sequestration. Extralobar sequestration accounts for 25% cases of pulmonary sequestration, left lower lobe involvement is more common, has its own pleural layer and may occur in ectopic locations including mediastinum, pericardium and upper abdomen, almost invariably left suprarenal, and is more often associated with other malformations such as diaphragmatic hernia or scimitar syndrome. Intralobar sequestration has the highest risk for infection and is an indication for removal. Congenital bronchial atresia results from local obliteration or stenosis of a segmental, subsegmental or lobar bronchus at or near its origin. Usually involve the left lobe and segment bronchi of right lobe, middle lobe, and occasionally lower lobe. Imaging features include a hilar mass and overinflation of the peripheral lung often associated with an opaque round mucocele in the bronchial tree just distal to the obstruction. Cystic lung neoplasms include lymphangioma, histiocytosis and pleuro-pulmonary blastoma. Pleuropulmonary blastoma has three pathological features: type 1 is entirely cystic lesion, type 2 is characterized by cystic lesion and solid mass, while type 3 is a purely solid high-grade sarcoma. Less than 25% of children with CCL lesion develop symptoms early in life defined as respiratory compromise (stridor, excessive crying, coughing, failure to thrive, need of supplemental oxygen) or infection. Recurrent chest infection occurs in one-third of all patients. Symptomatic neonates require urgent surgery after diagnostic imaging and cardiac assessment. Symptoms of respiratory distress or episodic infection of the abnormal lung will need lung resection. Late symptoms can occur any time after life. Asymptomatic children with CCL lesions are managed expectantly and should undergo lung resection after six months of age as the surgery is less invasive, safer, and more beneficial. Most small asymptomatic lung lesions present sometime during life with intractable infective complications or pneumonia. In stratifying lesions from higher to lower risk of infection and symptoms, CPAM and bronchogenic cysts are categorized as medium to higher risk, while bronchial atresia, microcystic CPAM and extralobar sequestration are considered lower risk. Bilateral lesions signify a higher risk for malignancy and further workup is required.

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Circumcision

Male circumcision (CCC) represents the surgical removal of some or all the foreskin (prepuce) from the penis. Extremely common surgical procedure around the world mostly performed during the newborn period. Health benefits of newborn male CCC outweigh the risks justifying access of this procedure for families who chose it. Benefits of CCC include prevention of urinary tract infections, acquisition of HIV, transmission of some sexually transmitted disease, and penile cancer. Male CCC does not adversely affect penile sexual function, sensitivity, or sexual satisfaction. Elective CCC should be performed only if the infant's condition is stable and well, and it should be performed by trained and competent physicians using sterile technique and effective pain management. During newborn CCC the three most common operative methods consist of the Gomco clamp, the Plastibell device, and the Mogen clamp. Later in life the dorsal slit method is preferred. Parents of newborn boys should be instructed in the care of the penis at the time of discharge from the newborn hospital stay, regardless of whether the child has been circumcised or not. The no circumcised penis should be washed with soap and water. And the foreskin should not be forcibly retracted. CCC reduces the bacteria that accumulate under the prepuce which can cause a urinary tract infection (UTI). Periurethral flora contains fewer pathogens in circumcised penis. There is a protective effect for male with CCC in reducing the risk of HIV acquisition among heterosexual partners. CCC before puberty is more protective for HIV than circumcision occurring at a later age. Male CCC is also associated with a lower prevalence of human papillomavirus infection and herpes simplex virus type 2 transmission. CCC is not associated with a lower risk of gonorrhea, syphilis, or chlamydial infection. The incidence of UTI among boys under age 2 years is reduced threefold to 10-fold in all studies after CCC. It is estimated 7-14 of 1000 uncircumcised male infants will develop a UTI during the first year of life, compared to 1-2 infants among 1000 circumcised male infants. The absence of CCC increase the risk of developing penile invasive squamous cell carcinoma but not for carcinoma in situ. Indications for CCC in children include religious and cultural reasons, recurrent balanoposthitis and UTI, phimosis, and balanitis xerotica obliterans. CCC is also done during corrective hypospadia. Emergency dorsal slit CCC may be necessary for late presenting paraphimosis. Preputial retraction can occur at variables age with the mean age being approximately 10.4 years. CCC in boys with VUR has been shown to decrease the incidence of UTI. Phimosis is a condition in which the foreskin cannot be full retracted from the penis. Ballooning of foreskin is a sign of pathological phimosis. Phimosis alone is a risk factor for developing invasive cancer. Phimosis and lichen sclerosus (balanitis xerotica obliterans) are common indications for medical CCC after the newborn period. Topical steroid therapy significantly increased complete or partial clinical resolution of phimosis. Complications after CCC are rare, occurring approximately in 1 of 500 newborns circumcised male. Acute complications occur one-third of the time, while later complications two-third. Acute complications are usually minor and involve bleeding, infection, or an imperfect amount of tissue removed. Bleeding is the most common acute

complication, followed by infection and penile injury. Bleeding is significantly more common in boys undergoing CCC for balanitis xerotica obliterans (Lichen sclerosus) compared with boys undergoing CCC for other reasons. Late complications include excessive residual skin (incomplete CCC), excessive skin removal, adhesions (natural and vascularized skin bridges), phimosis, epithelial inclusion cysts, penile torsion, and meatal stenosis. The incidence of postoperative bleeding and hematoma formation in children CCC can be reduced with the use of tissue glue. Tissue glue reduces operative time, reduce postop pain, less overall cost, and have superior cosmetic results. Contraindications to newborn CCC include significantly prematurity, blood dyscrasias, babies with a family history of bleeding disorders and those with congenital anomalies such as hypospadia, congenital chordee or deficient shaft skin such as penoscrotal fusion or congenital buried penis. CCC in premature infants and newborns with prominent suprapubic fat pad or penoscrotal webbing has a higher risk for long-term complications described. Major complications are very infrequent and include glans or penile amputation, transmission of herpes simplex, methicillin resistant staphylococcal aureus infection, urethral cutaneous fistula, glans ischemia, and death. The preventive and public health advantages associated with newborn male CCC warrant third-party reimbursement of the procedure. Arguments opposing male CCC are supported mostly by low-quality evidence and opinion and are contradicted by strong scientific evidence.

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