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What is the Potter sequence?

Potter sequence results when there is decreased urine output in utero from the kidneys resulting in oligohydramnios which causes potentially fatal hypoplastic lungs, intrauterine growth retardation, characteristic facial abnormalities including a flattened nose and limb abnormalities to include clubbed foot. Although this can occur from any cause of prolonged oligohydramnios, the most classic etiology is bilateral renal agenesis but this could also result from posterior urethral valves or autosomal recessive polycystic kidney disease.

What is VACTERL?

VACTERL is an association and not a syndrome. Basically, VACTERL describes common congenital anomalies that are all associated with one another. This stands for Vertebral anomalies, Anal anomaly (typically imperforate anus), Cardiac anomalies, TracheaEsophageal fistula or Esophageal atresia, Renal anomalies and Limb anomalies. An interesting fact is that if both limbs are abnormal you can also assume both kidneys are abnormal and if 1 limb is involved, usually only 1 kidney will be involved. VACTERL associations are very high yield for the ABR core exam.

What renal abnormality is associated with Caroli syndrome?

Caroli syndrome is an autosomal recessive abnormality with classic cystic intrahepatic biliary ductal dilatation consistent with Todani 5 classification. Caroli syndrome is associated with polycystic kidney disease (autosomal dominant or autosomal recessive) as well as medullary sponge kidney. Remember that the central dot sign is classic for Caroli syndrome in which the portal vein is surrounded by the cystic dilated bile ducts.

Does autosomal recessive polycystic kidney disease (ARPCKD) classically present with large or small kidneys?

ARPCKD classically presents with enlarged kidneys. ARPCKD consists of renal microcysts and renal dysfunction with up to 1/3 of affected neonates not surviving the neonatal period due to pulmonary hypoplasia. ARPCKD is also associated with hepatic fibrosis and degree of hepatic fibrosis is often inversely correlated with degree of renal involvement (more hepatic fibrosis means less renal disease and vice versa). If hepatic fibrosis is severe this can lead to portal hypertension and risk of death. Look for history of hypertension and renal failure with smoothly enlarged and echogenic kidneys.

What common associations are found with unilateral renal agenesis?

As a reminder bilateral renal agenesis results in potter sequence whereas unilateral renal agenesis may be asymptomatic initially. There are very common gynecological associations and the majority of genetic females with unilateral renal agenesis will have genital anomalies. In genetic males, anomalies can also be present that include unilateral absence of the ipsilateral epididymis and vas deferens, an ipsilateral seminal vesicle cyst. Unilateral renal agenesis is also associated with an absent ipsilateral ureter and absent ipsilateral hemitrogone of the bladder.

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How on imaging can you potentially differentiate a surgically absent vs a congenitally absent kidney?

One key feature is the "pancake adrenal sign" in which the ipsilateral adrenal gland appears elongated and pancake like. Surgical clips would obviously point you towards surgical absence and the clinical history would obviously also be informative.

What is Mayer-Rokitansky-Küster-Hauser syndrome?

MRKH syndrome is associated with Mullerian duct anomalies to include absence of the uterus vs an atretic uicornuate uterus, absence of the upper 2/3 of the vagina, normal external genitalia and unilateral renal agenesis or other renal anomalies such as horseshoe kidney and possible vertebral anomalies.

What are classic potential complications of horseshoe kidney?

Classic potential complications of horseshoe kidney include renal trauma due to an abdominal force causing compression of the horseshoe kidney centrally over the underlying vertebral body, ureteropelvic junction obstruction, recurrent UTI/pyelonephritis, and renal stones. Patients with horseshoe kidney also have a much higher risk of developing Wilms tumor and/or transitional cell carcinoma as well as renal carcinoid. Horseshoe kidney is also associated with Turner syndrome.

What is crossed fused renal ectopia and which kidney is typically ectopic?

Crossed fused renal ectopia occurs when one kidney crosses midline and fuses with the other kidney. The ectopic kidney is usually the kidney that is more inferior, most commonly the left kidney that crosses and fuses with the right kidney. Associations include hydronephrosis, recurrent infection, and renal stones.

What are the classic features of prune belly syndrome?

Prune belly syndrome consists of absent abdominal musculature, hydronephrosis/hydroureter and cryptorchidism. Cause of cryptorchidism is massive urinary bladder distention that prevents normal testicular descent. This is also sometimes termed Eagle Barrett syndrome. Has VACTERL associations.

What is the most common congenital anomaly of the genitourinary tract?

Congenital ureteropelvic junction obstruction. This may be bilateral in about 20% of cases. Etiology may be intrinsic defects in the renal pelvis muscular bundles and treatment is pyeloplasty.

True or false: neonatal renal vein thrombosis is associated with maternal diabetes.

True. Left renal vein thrombosis is most common. If acute, expect renal enlargement. If chronic, expect renal atrophy.

What is the most common cause of neonatal renal artery thrombosis?

Umbilical artery catheterization is the most common cause of neonatal renal artery thrombosis.

What are top causes of congenital megaureter aka primary megaureter?

Top causes of congenital megaureter include an adynamic segment of the distal ureter, ureterovescicular junction reflux or idiopathic in etiology. Hydronephrosis may or may not be present but is most common with the distal adynamic type of congenital megaureter.

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A "reverse J" or "fishhook" appearance of the ureter in an intravenous pyelogram may be a sign of what entity?

Retrocaval ureter which results from a developmental anomaly of the inferior vena cava which causes partial obstruction of the ureter and may present with recurrent urinary tract infections.

What is the Weigert-Meyer rule?

The Weigert-Meyer rule is a must-know rule for the ABR core exam and describes the anatomy of a duplicated renal collecting system. The rule states that the upper pole has an ectopic insertion inferior and medial on the bladder and the upper pole is associated with a ureterocele and obstruction. The inferior pole inserts superior and laterally compared to the upper pole moiety and is prone to reflux. (Hint: A mental trick I use is "IR" as this is a familiar abbreviation for radiologists (Inferior pole Refluxes; I also remember that the Upper pole has a Ureterocele (UU) and this helps me remember that the upper pole is also prone to obstruction as manifest by the ureterocele.

What is a ureterocele?

A ureterocele is a cystic ballooning of the intravescicular ureter. Cause is thought to be obstruction of the ureter at the ureteral orifice.

What is the top cause of urethral obstruction in newborn males?

Posterior urethral valve which is a congenital obstruction near the posterior urethra. On a VCUG look for an abrupt change of caliber between the dilated posterior urethra and the normal sized anterior urethra. Can show hydronephrosis on ultrasound/MRI as well as a "key hole" appearance of the bladder due to the dilated posterior urethra. If you see a perirenal fluid collection this is likely from forniceal rupture due to the high pressure in the urinary system with resultant perirenal urinoma. If severe, this may be diagnosed in utero with findings of oligohydramnios and Potter sequence manifestations. If less severe may manifest initially with recurrent UTIs due to vesicoureteral reflux.

Are posterior urethral valves seen in newborn genetic females?

No, this is a disease exclusively of anatomic males. The obstructive tissue on the posterior urethra derives from Wolffian duct tissue and is typically sporadic though can be associated with some genetic syndromes including Down syndrome.

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Episode 2

What are key features differentiating grade 1 through 5 vesicoureteral reflux?

Grade 1 vesicoureteral reflux: reflux to ureter only (on boards will probably show reflux half way up ureter)

Grade 2 reflux: reflex to level of the non-dilated renal collecting system

Grade 3 reflux: Reflux to any level with a mildly dilated ureter and renal pelvis

Grade 4 reflux: Reflux to any level with moderate tortuosity of the ureter and renal pelvis blunting of renal fornices/papillary projections

Grade 5 reflux: Reflux to any level with marked tortuosity of ureter and renal pelvis with loss of renal fornices/papillary projections

Note that the most common cause of vesicoureteral reflux is abnormal insertion of the ureter at the ureterovescicular junction. The normal insertion of the ureter in the bladder needs to be at an oblique angle to create a valve mechanism. About half of kids with recurrent UTIs will have an abnormal insertion of the ureter into the bladder. Chronic reflux can lead to impaired renal function (reflux nephropathy) and hypertension.

What are the approximate odds that different grades of reflux will resolve without intervention?

Grade 1 and 2 reflux will typically resolve without intervention something like 80-90% of time. Grade 3 reflux may resolve about half of the time without intervention. Grade 4 and grade 5 reflux will resolve in the minority of cases (approximately 25% self-resolution). Antibiotic prophylaxis may be considered for all grades of reflux and surgical intervention may be considered primarily for grade 4 or 5 reflux or patients who experience breakthrough infections and/or progressive reflux nephropathy.

What structure does the urachus become after undergoing physiologic atrophy?

The urachus becomes the median umbilical ligament.

If the urachus fails to obliterate what are the spectrum of findings that can result from a fully or partially patent urachus?

Complete failure of the urachus to atrophy results in a patent urachus from bladder to the umbilicus, presents with urine leakage from the umbilicus in a neonate. A partially patent urachus can end up as a urachal sinus if the patent aspect as at the umbilicus end or vesicourachal diverticulum if the patent aspect is at the bladder. If the patent portion is isolated mostly to the middle of the urachus this is a urachal cyst.

What cancer does a patent urachal tract have risk of later in life?

Adenocarcinoma is the top cancer that can develop in a patent urachal tract later in life.

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What are key features of nephroblastomatosis?

Nephroblastomatosis occurs when nephrogenic rests persist beyond 36 weeks. Nephrogenic rests are persistent embryonal cells in the kidneys that normally regress by birth. On imaging nephroblastomatosis looks like homogeneous round mass(es) and/or a kidney with a hypodense rind.

Nephroblastomatosis can classically be a precursor for which tumor?

Nephroblastomatosis is commonly considered a precursor for Wilms tumor. If bilateral Wilms tumor is found this essentially always started as nephroblastomatosis. To screen for Wilms tumor in a patient with nephroblastomatosis perform serial ultrasound or MRI screening. Importantly, if you see any necrosis in an area of nephroblastomatosis this is concerning for Wilms tumor and not regression of the nephroblastomatosis.

What is the most common solid renal tumor of infancy and what is the most common solid renal tumor of childhood?

The most common solid renal tumor of infancy is a mesoblastic nephroma and nearly all of these present within the 1st month of life. This is considered to be a generally benign tumor similar to a fetal hamartoma. The top solid renal tumor of childhood is a Wilms tumor. Remember that Wilms tumors typically do not present in infancy. On board exams, if you see a large solid renal tumor in a neonate think mesoblastic nephroma.

What are key features of multicystic dysplastic kidney?

Key for board exams is to remember that multicystic dysplastic kidney has no functioning renal tissue which means that the involved kidney cannot excrete urine. So one way to diagnose this entity if you are having a hard time telling between hydronephrosis versus the dilated cystic spaces of multicystic dysplastic kidney is to do renal scintigraphy such as a MAG3 study and if there is excretion of radiotracer this is not multicystic dysplastic kidney. With multicystic dysplastic kidney multiple cysts form in utero instead of the normal renal tissue and the kidney will have a multicystic appearance. Remember that there is about a 50% rate of contralateral renal anomalies with the top contralateral renal anomaly being a ureteropelvic junction obstruction.

What are key features of Wilms tumor in terms of age of presentation and top sites of metastatic disease?

First of all, remember again that on board exams Wilms tumor will not present in neonates but is a tumor of childhood with an average age of about 3 years old. Wilms tumor is a solid renal tumor that can spread by direct invasion or metastasize with the #1 site of metastasis being the lungs and the #2 site of metastasis being the liver.

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For the ABR core exam what are some key associations with Wilms tumor?

Beckwith Wiedemann syndrome is key to remember for the ABR core exam. Beckwith Wiedemann syndrome has A LOT of associations and key features but these include Wilms tumor, omphalocele, hepatoblastoma, macroglossia, hemihypertrophy, cardiac anomalies and enlargement of the liver, spleen and kidney. In terms of tumors Beckwith Wiedemann syndrome has a common association with both Wilms tumor and neuroblastoma.

Also remember WAGR syndrome: Wilms tumor, aniridia, GU anomalies and mental retardation/impairment.

Also, Drash or Denys Drash syndrome: Wilms tumor, male pseudohermaphroditism (46 XY karyotype with partial androgen insensitivity causing female phenotypic features) and progressive glomerulonephritis.

What are key features of clear cell sarcoma of the kidney?

First of all, this is a very rare tumor but is still the #2 most common malignant tumor of the kidney in childhood. If you are being tested on clear cell sarcoma it is important to remember that this generally has a worse prognosis than a Wilms tumor, can present with a palpable abdominal mass, and has a high propensity to metastasize to the bones.

What are key features of cystic nephromas on imaging and at which age is this entity most likely to present?

Cystic nephromas, previously termed multilocular cystic nephroma, classically presents with fluid filled locules surrounded by a thick fibrous capsule the protrude into the renal pelvis. Cystic nephromas characteristically do not have solid components, calcifications, or necrosis. These were previously termed multilocular cystic nephroma and were taught to have a bimodal age presentation consisting of young boys and middle aged women. We now know from genetic and histologic testing that these are actually different entities but on imaging they still appear similar. Pediatric cystic nephroma is most common in young childhood in boys. Adult cystic nephroma is more common in women around 40-50 years of age.

What is the key imaging appearance of the botryoid variant of rhabdomyosarcoma of the bladder?

The botryoid variant of bladder rhabdomyosarcoma is highly polypoid and looks like a bunch of grapes. Classic rhabdomyosarcoma of the bladder, on the other hand, is highly infiltrative and can present as a paratesticular mass. Rhabdomyosarcoma is the top bladder cancer in young children and often metastasizes to lung, lymph nodes and bone.

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What are key features that can help you differentiate between neuroblastoma and Wilms tumor?

Neuroblastoma is the most common extracranial solid malignancy of childhood and can present at birth or otherwise in very young children. Wilms classically presents in slightly older kids and is said to almost never present before age 2 months so if you are presented with a question asking about a solid renal mass in a neonate, the answer should be neuroblastoma. Calcification is very common for neuroblastoma and very rare for Wilms tumors. Wilms tumors are circumscribed whereas neuroblastomas are infiltrative lesions. If you see vascular invasion this favors Wilms tumor. Neuroblastoma more commonly metastasizes to bones than Wilms tumor (excepting the clear cell variant) and remember that MIBG is often the imaging modality of choice to evaluate for bone metastases in the setting of neuroblastoma.

A few additional features of neuroblastoma need to be mentioned as they are high-yield for board exams: Raccoon eyes can be a sign of orbital neuroblastoma and you need to know what this looks like including proptosis and periorbital bruising. You also should be aware of blueberry muffin syndrome with multiple blue/purple lesions of the skin related to extramedullary hematopoiesis in the setting of neuroblastoma and other entities including CMV infection and AML. Additionally, you need to be aware of stage 4s neuroblastoma because this actually has a good prognosis, despite presenting in a child often less than 1 year of age with a thoracic primary neuroblastoma lesion with skin, liver, and bone marrow metastases. Neuroblastomas can arise most commonly from the adrenal glands but can arise anywhere along the sympathetic chain.