

Diagnosis and Treatment of Prenatal Urogenital Anomalies: Review of Current Literature

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Abstract

With current advances in antenatal medicine it is feasible to diagnose prenatal anomalies earlier and potentially treat these anomalies before they become a significant postnatal problem. We discuss different methods and signs of urogenital anomalies and emphasize the use of telemedicine to assist patients and healthcare providers in remote healthcare facilities, in real time. We discuss prenatal urogenital anomalies that can be detected by antenatal ultrasound and fetal magnetic resonance imaging. Ultimately, we address the natural history of urogenital anomalies, which need surgical intervention, and emphasize ex-utero intrapartum treatment procedures and other common surgical techniques, which alter the natural history of urogenital anomalies.

Keywords: Prenatal anomalies; Hydronephrosis; Kidney; Ureter; Bladder; Urethra; Oligohydramnios; Genitourinary

Abbreviations: US: Ultrasound; GU: Genitourinary; MRI: Magnetic Resonance Imaging; CAH: Congenital Adrenal Hyperplasia; BRA: Bilateral Renal Agenesis; URA: Unilateral Renal Agenesis; VUR: Vesicoureteral Reflux; ARPKD: Autosomal Recessive Polycystic Kidney Disease; ADPKD: Autosomal Dominant Polycystic Kidney Disease; MDK: Multicystic Dysplastic Kidney; MCKD: Medullary Cystic Kidney Disease; UPJO: Ureteropelvic Junction Obstruction; BD: Bladder Diverticulum; MMIHS: Megacystis Microcolon Intestinal Hypoperistalsis Syndrome; HPGA: Hypothalamic-Pituitary-Gonadal Axis; Posterior Urethral Valves; PST: Penoscrotal Transposition; BFS: Bifid Scrotum; PBS: Prune-Belly Syndrome; CH: Congenital Hydrocele; EXIT Procedure: Ex-utero Intrapartum Treatment procedure

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Introduction

Advancements in medicine have made it possible to diagnose and treat antenatal anomalies. Many imaging techniques have provided methods of evaluating pregnancy, in both obstetric and fetal points of view. Detection of urological anomalies was common in the past, however the exact nature was difficult to isolate [1]. For the sake of brevity, we address antenatal anomalies diagnosed by ultrasound and fetal magnetic resonance imaging

Diagnostic Methods

Ultrasound

Ultrasound (US) uses high frequency sound waves that are used to detect objects and measure distances. Implementation of

US as a routine prenatal screening was established in the late 1970s. Studies identified that in two percent of all fetuses, lethal anomalies were present. Among the anomalies, genitourinary (GU) abnormalities were the most common [2]. The purpose of US of the GU tract is to confirm and evaluate the progression of prenatal findings. US is used as a first-line method for GU tract imaging because of its combination of excellent anatomical delineation, lack of radiation, high availability, dynamic evaluation capabilities, decreased need for sedation, and low cost [3].

Diagnostic amniocentesis

Amniocentesis is a technique using a transabdominal approach to remove amniotic fluid from the uterine cavity. The fluid is tested to determine fetal health because it contains fetal substances. Commonly the amniotic fluid is used for prenatal genetic studies

and fetal lung capacity. However, it is also useful for identifying neural tube defects [4-7].

Oligohydramnios

Oligohydramnios is characterized by three factors: amniotic fluid volume less than 500 milliliters at the third trimester, single deepest pocket of less than two centimeters, and amniotic fluid index of less than five centimeters [8-10]. Oligohydramnios is a good indicator of renal dysfunction and can be identified using US [11-17].

Fetal magnetic resonance imaging

Fetal magnetic resonance imaging (MRI) has been used for identifying prenatal anomalies, beginning in the mid-1990s [3,18,19]. For diagnosing congenital anomalies both US and fetal MRI are used. However, US remains the predominant method for evaluation of disorders in the fetus, because it is relatively inexpensive and has widespread availability for real-time imaging [3,20-22]. Due to some limitations of US, fetal MRI is used. Fetal MRI has a larger field-of-view and is not hindered by soft tissues and bone [18,21,23]. Though MRI can be used to diagnose anomalies in the fetus, it is limited [24,25].

Telemedicine

Telemedicine has improved the face of medicine, by increasing the distance at which health care can be provided. It is being used to diagnose fetal urologic disorders primarily using tele-US and maternal-fetal medicine consultations [26]. This method of diagnosis empowers patients, increases administrative efficiency, and ensures expertise is available in places where it is needed the most [26-29]. Telemedicine is used in hospitals to overcome physician [30] and nursing staff shortage [31]. This also provides improved patient safety by standardizing practice by real-time collaboration [29,32]. This technique is being used in medicine and will assist physicians when diagnosing antenatal anomalies.

Prenatal Anomalies

Adrenal

Congenital Adrenal Hyperplasia

Congenital adrenal hyperplasia (CAH) combines a set of autosomal recessive disorders. These disorders are related to a deficiency in the enzyme necessary for the synthesis of cortisol, and may or may not include secondary effects in the deficiency of aldosterone and excess of androgens [33]. CAH is characterized by the degree of cortisol deficiency [34]. The phenotype of CAH depends on the deletion or mutation of a gene corresponding to the enzymatic function. The most common form of CAH is due to 21-hydroxylase deficiency, due to mutations of CYP21A2 [35]. Another form of CAH is defined by the loss of 11-beta hydroxylase, due to a mutation or deletion of CYP11B1 [36]. In the United States, CAH is prevalent in 1 case per 12,000, related to CYP21A2 mutations, the classic form of the disorder [37]. CAH due to CYP11B1 mutations account for 1 in 100,000, 5 to 8% of cases overall [38]. Diagnosis of CAH is dependent on the

level of production of cortisol, aldosterone, and/or androgen. Deficiency of 21-hydroxylase can be identified by measuring serum concentration of 17-hydroxyprogesterone and urinary pregnanetriol; high concentrations of these are suggestive of the disease [39,40]. Deficiency of 11-beta hydroxylase can be identified by high concentrations of 11-deoxycortisol and deoxycorticosterone [41]. CAH can also be diagnosed using amniocentesis [42]. Management of CAH can be done using glucocorticoid replacement therapy [43]. A prenatal treatment option for female fetuses diagnosed with CAH, can be done using dexamethasone [42,44]. The last line of treatment for CAH, only utilized in the most severe cases, involves bilateral adrenalectomy [45,46].

Adrenal Cystic Neuroblastoma

Neuroblastoma is the most common intra-abdominal tumor in children [47]. It is derived from the neural crest ectoderm, when cells fail to respond to normal signaling. Incidence of adrenal cystic neuroblastoma is 1 in 10,000 cases [48]. Adrenal cystic neuroblastoma increases the production of catecholamines [49]. These cases can be diagnosed using US [50] or fetal MRI, both of which can clearly identify the disorder [23,51]. To treat and manage neuroblastoma, surgery is the ideal option [52,53].

Adrenal Hemorrhage

Adrenal hemorrhage frequency is increasing in both prenatal and neonatal infants. This anomaly was localized by US. Ruminska et al. reported 13 neonates with adrenal hemorrhage. They were able to identify birth trauma, infection, and perinatal asphyxia as some of the risk factors in majority of the cases that resulted in adrenal hemorrhage. However, this anomaly rarely led to adrenal insufficiency. If the neonate had bilateral adrenal hemorrhage, an extended hormonal diagnosis was necessary. All patients were required to have an US follow-up [54]. Gyurkovits et al. identified vaginal delivery, macrosomia, and fetal acidemia were the most important risk factors of adrenal hemorrhage. Also, they found adrenal glands on both sides were similarly involved [55].

Renal

Bilateral Renal Agenesis

Bilateral renal agenesis (BRA) is a congenital absence of both kidneys. This is also known as Potter's syndrome, Potter's sequence or Oligohydramnios sequence, coined by pathologist Edith Potter. She was also able to distinguish the sequence of events that leads to BRA [56-59]. Using US, it is possible to identify BRA after 16 weeks of gestation because the amount of amniotic fluid is no longer dependent on transmembrane flow, but rather due to fetal urine production [60]. If a fetus has BRA, you will be able to see a condition of oligohydramnios because the volume of amniotic fluid is less than normal in the amniotic cavity [61]. Genetic aspects of BRA are not fully understood. BRA is estimated to occur in 0.1 per 1000 births [62]. Maternal factors associated with BRA include a body mass index of greater than 30 kg/m² prior to pregnancy, smoking during the periconceptional period, and binge drinking during the second month of pregnancy [63]. Survival rate of fetuses with BRA is extremely low, as BRA

is a lethal congenital anomaly [64]. Recent research has shown success for the in utero intervention for bilateral renal agenesis using serial amnioinfusion [65].

Unilateral Renal Agenesis

Unilateral renal agenesis (URA) is a congenital absence of one kidney. This condition is not fatal, unlike BRA, and patients can have a normal life expectancy. However, urological anomalies often accompany URA and patients should be monitored to decrease the risk of renal failure. Urological anomalies found in patients with URA included ureterovesical junction obstruction, bladder dysfunction, vesicoureteral reflux (VUR), ureteropelvic junction obstruction, ureterovesical and ureteropelvic junction obstruction, duplicated collecting system plus grade IV VUR, ectopic kidney plus grade V VUR, ectopic kidney, and development of chronic renal insufficiency [66]. URA is more common than BRA and the general incidence is 1 in 2000 [67]. Patients with URA have an increased risk of hypertension [68]. US can be used to identify a fetus with URA [69-72].

Polycystic Kidney Disease

Polycystic kidney disease is a disorder that can be found in both adult and pediatric patients. It involves the development of bilateral renal cysts with dysplasia. Polycystic kidney disease is characterized into two forms: autosomal recessive polycystic kidney disease (ARPKD) and autosomal dominant polycystic kidney disease (ADPKD). Both involve the presence of renal cysts. This disease can lead to chronic kidney disease and end-stage renal disease, respectively. Patients experiencing ARPKD or ADPKD should be monitored carefully because once they develop end-stage renal disease, they would need to undergo dialysis [73]. For prenatal evaluations, the first line method to identify these disorders should be US. When US findings are suboptimal, fetal MRI serves as a useful tool in prenatal diagnosis [70,71,74].

ARPKD is characterized by cystic dilation of the collecting ducts [75]. It was first recognized in 1902, however the histology was not reported until much later. Osathanondh et al. and Potter et al. classified this disease first [76-80]. In 1994, the ARPKD gene was identified on chromosome six [81]. PKDHD1 is a gene that is expressed on the cilia of the renal and bile ducts. It is believed to be crucial in maintaining normal tubular architecture of the ducts. It is not yet completely understood but this finding strengthens the theory that ARPKD is linked to ciliary dysfunction [82]. ARPKD is reported as one case per 20,000 live births [83]. US of a fetus shows enlarged kidneys, small bladder with absence of urine, renal masses, and oligohydramnios [84,85]. Normal US findings do not exclude ARPKD, as abnormalities may not be seen until late in the second trimester [83,86].

ADPKD is the most common inherited renal cystic disease [87]. It is characterized by progressive cystic dilation of both kidneys and can have manifestations in other physiological systems. Dysfunction of PKD1 and PKD2, polycystin 1 and polycystin 2, respectively, are thought to be responsible for ADPKD, primarily by ciliary dysfunction [88]. ADPKD differs from ARPKD in that cysts of ADPKD can develop anywhere along the nephron. ADPKD is more prominent and the reported prevalence is between 1 in 400

and 1 in 1000 [87]. US findings of ADPKD can include enlarged echogenic kidneys in utero [89].

Duplex Kidneys

Duplex kidney is a renal system containing a single renal parenchyma and drained by two pyelocaliceal systems [90]. Duplex kidneys occur in 0.8% of the general population [91]. When examining a patient with duplex kidney, the duplex kidney is often more elongated than their non-duplex kidney and may contribute more to total renal function [92]. Renal duplex anomalies can be diagnosed by prenatal US. Antenatal diagnosis and proper postnatal care may prevent urinary tract infections and renal function impairment [93]. Children with duplex kidneys can be prone to urinary tract infections due to vesico-ureteric reflux or obstruction [91].

Horseshoe Kidney

Horseshoe kidney is the most common type of renal fusion anomaly, where two separately functioning kidneys are fused at the midline. Horseshoe kidney is found in 1 in every 400 births. It is more commonly found in males than in females [94]. There are two theories on the formation of horseshoe kidney. Classical teaching of mechanical fusion states that it is formed during organogenesis, and fusion is held by fibrous isthmus. The other theory states that the fusion is due to abnormal migration of parenchymatous isthmus, resulting in a teratogenic event [95,96]. Patients with horseshoe kidney often remain asymptomatic and it is often only discovered during radiological exams. When symptoms are present they include nausea, abdominal discomfort, kidney stones, increased incidence of urinary tract infections, kidney obstruction, or kidney infections associated with vesicoureteral reflux [97,98].

Cross Fused Renal Ectopia

Cross fused renal ectopia is an anomaly wherein the kidneys fuse and are located on the same side of the midline. Cross fused renal ectopia is more likely to occur in males than females. It is more commonly found on the left side than the right. US can be used to detect the anomaly. Patients with cross fused renal ectopia are usually asymptomatic. However, complications of cross fused renal ectopia can include infection (pyelonephritis), obstruction (hydronephrosis due to pelviureteric junction obstruction), urolithiasis, and vesicoureteral reflux [99]. The exact incidence is unknown, due to the majority of patients remaining asymptomatic, however the estimated prevalence is 1 in 2000 [100].

Fused Pelvic Kidney

Fused pelvic kidney, also known as pancake kidney, is a kidney that does not ascend as it normally should during fetal development, and instead it is fixed in the pelvis. Detection is possible using radiological exams - US or MRI [101-104].

Renal Malrotation

Renal malrotation, also known as abnormal renal rotation, is an anatomical variation in the position of the kidneys, particularly the orientation of the renal hilum. Rotational variations are rare. The

exact incidence of malrotation is unknown and under reported because many patients have no clinical symptoms [105,106].

Hydronephrosis

Hydronephrosis is a common clinical condition encountered by physicians when visualizing the fetus using US [107-109]. Detection of hydronephrosis is possible as early as the 12th to 14th week of gestation [110]. Hydronephrosis is seen in 1-5% of pregnancies [109], and persists in 30-75% of infants postnatally [111]. It can result from interruption of urine flow, and obstruction can be from anywhere along the urinary tract. Obstruction is the key cause of hydronephrosis and is reported in 5-60% of cases [112]. No surgical treatment is necessary, as the condition generally resolves itself. Postnatal check-ups should be repeated every 3 to 6 months [113]. However, higher grades of hydronephrosis would require surgery to treat [114].

Multicystic Dysplastic Kidney

Multicystic dysplastic kidney (MDK) is the most common antenatally diagnosed cystic renal pathology. It is characterized by the presence of multiple, noncommunicating cysts separated by parenchyma and absent normal pelvocaliceal [115]. Unilateral and bilateral MDK are both possible. MDK is more commonly found in males. Isolated unilateral MDK that is not linked with other anomalies often has a good prognosis, whereas bilateral MDK is linked with a bad prognosis [115]. The incidence of MDK ranges from 1 in 1000 to 4300 live births. It is recommended that MDK be managed conservatively [116]. Long term prognosis is usually good. However, due to reduction in nephron mass, the early prevention of cardiovascular risk and nephrotoxicity is recommended [117].

Medullary Cystic Kidney Disease

Medullary cystic kidney disease (MCKD) is passed down in an autosomal dominant pattern and usually presents with adult-onset renal failure [118]. It is also known as autosomal dominant interstitial kidney disease to highlight the inheritance pattern and slowly progressive kidney disease due to interstitial fibrosis. There are several genetic defects that can result in MCKD. MCKD type 1 is a result of MUC1 gene mutations, which causes a buildup of mucin 1 protein in the distal nephron [119]. MCKD type 2 is a result of UMOD gene mutation. A mutant form of uromodulin protein cannot exit the endoplasmic reticulum and this results in abnormal accumulation of protein, which causes tubular cell death and chronic kidney disease [120-122].

Renal Hypoplasia

Renal hypoplasia is when part of the kidney does not develop fully in the womb, therefore it may not function as properly as a normal sized kidney. The exact definition is abnormally small kidneys with normal morphology and a reduction in the number of nephrons. Renal hypoplasia is a common cause of pediatric renal failure. Epidemiologic studies suggest an incidence of 1 in every 400 births [123].

Pyelectasis/Renal Pelvic Dilation

Pyelectasis is characterized as a dilation of the renal pelvis in utero. It can be found utilizing US, and is often detected in the second trimester. Renal pelvis dilatation is a common anomaly detected during routine second trimester scans. Treatment is often not needed for pyelectasis because most cases resolve themselves during pregnancy or in the first year after birth. While most cases of mild pyelectasis will show spontaneous resolution, persistent mild pyelectasis may lead to postnatal morbidity and should be monitored [124]. Pyelectasis can result from many factors, including an obstruction in the kidney or urethral obstruction, or duplex kidney. Fetal pyelectasis can be associated with decreased differential renal function [125]. It is recommended that all fetal renal pyelectasis greater than or equal to 5 mm and detected during second trimester US, should be followed antenatally. In addition fetuses with persistent pyelectasis should be evaluated after birth and followed until resolution of pyelectasis is achieved [126].

Mesoblastic Nephroma

Mesoblastic nephroma is a renal stromal neoplasm. It represents 3-10% of all pediatric renal tumors [127]. It is more commonly found in males than females. It is generally benign and unilateral [128]. Diagnosis can be made using US or fetal MRI, and it manifests along with oligohydramnios [129,130]. The mass can be diagnosed after 18-20 weeks of gestation. Treatment of mesoblastic nephroma has favorable outcomes; a nephrectomy is usually the simplest option [131]. Mesoblastic nephroma can be described as classic, cellular, or mixed. Cellular mesoblastic nephroma is often larger than classic, presents later, and appears more heterogeneous on imaging. Distinct from classic, cellular mesoblastic nephroma can exhibit aggressive behavior such as vascular encasement and metastasis [132].

Ureteral

Ectopic Ureter

Ectopic ureter, also known as ureteral ectopia, is a congenital renal anomaly, which results from abnormal migration of the ureteral bud during its insertion to the bladder. In females, the ureter may insert itself into the lower urinary bladder, urethra, or vagina. In males, it may insert itself into the lower urinary bladder, posterior urethra, seminal vesicle, vas deferens, or the ejaculatory duct [133-135]. It is more common in females than males. In females, the common symptom is dribbling urinary incontinence [136]. Other presenting symptoms can include urinary tract infection, abdominal pain, and renal failure [137]. US or MRI urography may provide assistance to view an ectopic ureter [138]. Treatment options involve surgical methods. Most cases of ectopic ureter can be managed by heminephrectomy, however if adequate function is demonstrated in the upper pole, ureteropyelostomy is recommended [136].

Duplication of Ureters

Ureteral duplication is the most common renal abnormality, as it manifests in 1% of the population and is estimated as the

cause in 10% of children who are diagnosed with urinary tract infections [139]. Incomplete ureteral duplication has no clinical significance. However, complete ureteral duplication, where two ureters enter the bladder, can result in vesicoureteral reflux (VUR), ectopic ureterocele, or ectopic ureteral insertion [140,141] [142]. This occurs when two separate ureteric buds arise from a single Wolffian duct. US is the first line method to investigate for ureteral duplication. For patients with ureteral duplication with a severely dilated ureter, a commonly used surgical technique is ipsilateral ureteroureterostomy for treatment of reflux and/or obstruction [143].

Congenital Megaureter

Congenital megaureter, or primary megaureter, encompasses the causes of enlarged (abnormally dilated) ureter and may result in obstruction and reflux, or unobstructed and not refluxing [144]. The underlying cause of congenital megaureter is an abnormality of the Wolffian duct and the ureteric buds [145]. The exact prevalence is unknown. Primary megaureter may also be caused by vesicoureteric reflux, obstructive disease, an increased urinary output from kidneys, or by lack of development of the ureteral muscularity [146]. Refluxing congenital megaureter is caused by short or absent intravesical ureter, or derangement of vesicoureteric junction. Obstructed congenital megaureter is due to an obstruction of the aperistaltic juxtavesical segment, preventing urine transport at acceptable rates. Primary obstructive megaureter is often symptomatic and has high complication rates, including infections, stone formation, and renal failure. Surgical treatment of obstructive megaureter is recommended [147]. Primary obstructive megaureter should be differentiated from nonobstructed, nonrefluxing megaureter, which occurs in 6% to 10% of infants with antenatal hydronephrosis [148]. This sub-class may improve or resolve as demonstrated on serial ultrasound, often beyond four years with higher grade hydronephrosis [149,150]. Hence, initial management of primary nonrefluxing megaureter is conservative [151-153].

Ureteropelvic Junction Obstruction

Ureteropelvic junction obstruction (UPJO) is defined as an obstruction of urine flow from the renal pelvis to the proximal ureter. UPJO is the most frequently observed cause of obstructive nephropathy in children [154]. UPJO is a predominant cause of obstructive hydronephrosis [155]. The incidence of UPJO is estimated at 1 in 1000-1500 [154,156]. Imaging techniques, such as US, have made it easier to detect the anomaly sooner [157]. The exact pathophysiology of UPJO remains unknown but research suggests it is multifactorial [158]. Surgical treatment is the primary method for UPJO. A highly successful technique utilized is pyeloplasty [159]. Robot assisted laparoscopic pyeloplasty has been gaining acceptance among pediatric urologists in treatment of UPJO [160,161]. UPJO is determined by many factors - pressure within the renal pelvis, diameter of UPJ, compliance of renal pelvis, and the activity of the ureter. Increased volume and pressure results in renal pelvis dilation. Over time excessively increased pressure results in hypertrophy of the renal pelvis and hinders its compliance, which in turn results in decreased renal function. Untreated UPJO often induces mild to severe impairment of renal

function, including impaired glomerular filtration and tubular exchanges of water and solutes [154].

Ureterocele

Ureterocele is a congenital cystic pouching of the distal ureter into the urinary bladder. It is a challenging urologic anomaly. Characterization of ureterocele is based on the relationship with the renal unit. It can either be orthotopic or ectopic. Incidence is reported as between 1 in 5000 and 1 in 12,000 children, with the incidence of ureterocele at autopsy as high as 1 in 500 [162,163]. Ureterocele is more commonly diagnosed in females [162]. Diagnosis of ureterocele is done by US, but in some cases MRI can clarify diagnosis [164-166]. Ureteroceles form during embryogenesis of the kidney and ureter [162]. To treat ureterocele surgical options include endoscopic ureterocele incision, ipsilateral ureteroureterostomy, and ureterocele moiety heminephrectomy. Research supports the endoscopic puncture method as a safe and effective treatment for symptomatic children with both single-system and duplex-system intravesical ureteroceles [167].

Vesicoureteral Reflux

Vesicoureteral reflux (VUR) is the retrograde flow of urine from the bladder into the upper urinary tract. This disorder has substantial morbidity, both from infection and from reflux nephropathy [168,169]. Normally, the distal ureter passes through a submucosal tunnel, after entering the bladder, and opens into the bladder lumen. However, if the length of the submucosal tunnel or the muscular backing is inadequate, the valve does not function properly and results in reflux. Diagnostic methods involve US and urodynamics [170-172].

Bladder

Bladder Diverticulum

Bladder diverticulum (BD) is a congenital disorder that presents itself as an outpouching from the bladder wall. BD occurs almost exclusively in males and is seen in approximately 1.7% of cases [138,173]. BD is believed to occur due to weakness of the ureterovesical junction or posterior urethral valve causing high intravesical pressure with voiding. Using US and MRI, detection is possible and can help dictate the next plan of action regarding BD. Next plan of action is surgical treatment. Diverticulectomy is a common procedure used once BD is identified using prenatal radiological methods. Due to the potential for development of carcinoma in bladder diverticula, immediate prophylactic diverticular excision is recommended [138,174].

Exstrophy

Bladder exstrophy, also known as ectopia vesicae, is a herniation of the urinary bladder through an anterior abdominal wall defect [175]. Exstrophy is a rare congenital anomaly; occurring at a rate of approximately 1 in 10,000-50,000 [176]. It is more common in males than females [177]. It is caused by a developmental defect of the cloacal membrane, which results in the protrusion of the bladder mucosa, as a mass-like lesion [178]. Prenatal diagnosis

is often made through incidental findings during routine US. Treatment for exstrophy involves surgical options including complete primary repair of exstrophy and urinary diversion for exstrophy. MRI is a valuable tool in planning and evaluating the optimal surgical techniques for closure of bladder exstrophy [179].

Megacystis Microcolon Intestinal Hypoperistalsis Syndrome

Megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS) also known as Berdon syndrome, after the physician to first describe it in 1976 [180]. MMIHS is a rare congenital disorder characterized by a largely dilated non-obstructed urinary bladder, microcolon, and decreased or absent intestinal peristalsis. MMIHS leads to neonatal intestinal obstruction. There have been approximately 230 cases reported in the literature [180-183]. It occurs more frequently in female fetuses than in males [181,183]. In many cases in which antenatal US diagnoses a fetus with MMIHS, the pregnancy is terminated, due to its association with a fatal outcome. It is recommended that when fetal megacystis is detected by US, particularly when it is a female fetus and associated with polyhydramnios, MMIHS should be considered [184]. Pathogenesis for MMIHS is not clear. It is thought to have an autosomal recessive inheritance [181,183]. A hypothesis states that an ACTG2 mutation is the cause of MMIHS, resulting from a smooth muscle myopathy. Recent research has indicated de novo mutations in ACTG2 as a cause of fetal megacystis in MMIHS and gonadal mosaicism may be present in certain cases [185]. Surgical options are available; however patients often have a poor outcome and life expectancy is low [181,183].

Patent Urachus

Patent urachus is an opening between the bladder and the belly button/umbilical cord. It is diagnosed using US, often seen as a tubular connection between the bladder and umbilical cord. Patent urachus arises when the allantois fails to breakdown and results in an opening between the bladder and umbilical cord. If the urachal tract maintains a lumen during embryonic development it may lead to patent urachus. Treatment involves a surgical procedure to remove the patent urachus and may also involve surgical repair to the bladder [186-189].

Menkes Syndrome

Menkes syndrome, or Menkes disease, is an X-chromosome linked neurodegenerative disease, leading to impairment of copper transport. First described by Menkes et al. in 1962 [190]. Copper is a crucial metal for the proper functioning of many enzymes. Menkes syndrome reduces the function of ATP7A, which decreases transport of copper [191-193]. Menkes syndrome results in BD and VUR, along with neurological problems [194]. An extremely rare disorder with an incidence estimated as 1 in 100,000 [190,192,195]. Treatment options include injecting copper histidine in utero to avoid neurological symptoms. An early diagnosis and treatment with copper supplementation has been shown to be beneficial [195].

Penile

Chordee

Chordee is commonly associated with hypospadias and it involves the ventral shortening and curvature of the penis. Congenital chordee can also be seen with the meatus in the orthotopic position [196]. Chordee may be an end result of growth disparity between dorsal tissues of the corporal bodies and attenuated ventral urethra. It may also be due to a deficiency of ventral penile skin. In the United States it is the second most common congenital penile anomaly [197,198]. Surgery is the best option using orthoplasty to straighten the penis [199]. Both US and fetal MRI can be used to diagnose chordee in the fetus [200,201].

Epispadias

Epispadias is a rare congenital defect involving the opening of the urethra on the dorsum of the penis. This results when the urethra fails to develop into a full tube. It tends to occur alongside bladder exstrophy. Prevalence of complete male epispadias, characterized by the failed closure of the penopubic dorsal urethra, is found in approximately 1 in 120,000 births [202]. Treating epispadias requires surgery, depending on the characteristics of the penis and whether the penis is curved or straight. Based on the characteristics, a multistaged reconstruction procedure or a single-stage procedure may be used [203].

Hypospadias

Hypospadias is an abnormality of anterior urethral and penile development. The urethral opening is ectopically located at a ventral location of the penis, proximal to the tip of the glans penis, which is splayed open. The urethral opening may be located as far down as the scrotum or perineum. It is a congenital defect, occurring during urethral development, between weeks 8 and 20 of gestation. Baskin et al. proposed a modified theory stating: the urethral folds fuse to form a seam of epithelium, which turns into the mesenchyme, and is canalized by apoptosis or programmed cell resorption [204-209]. Hypospadias occurs when the urethral folds fail to fuse. Genetic predisposition can increase the incidence of hypospadias [210-213]. This is the most common penile anomaly, diagnosed in 68.3% of all cases of congenital penile anomalies [198]. Orthoplasty is the best method of treatment for hypospadias. Both US and fetal MRI, can be used to diagnosis the fetus [200,201,214,215].

Microphallus

Microphallus, or micropenis, is defined by the length of the penis, stretched, 2.5 or more standard deviations below the mean; for an infant age 0 to 5 months, the lower limit is 1.9 cm [216]. Normal male development is controlled by testosterone and dihydrotestosterone. Microphallus can be caused by a defect in the hypothalamic-pituitary-gonadal axis (HPGA), and deficiency in androgen. Adequate penile growth requires normal production of testosterone towards the end of gestation. The fetus has normal levels early on, before 14 weeks of gestation. However, if testosterone levels drop after 14 weeks of gestation, due to an issue in the HPGA, this will result in inadequate penile growth.

Microphallus can also occur in fetuses with LH-receptor defects and defects in testosterone biosynthesis. Surgical options such as genitoplasty can be performed to treat patients with microphallus [216,217]. Another option is testosterone therapy. However, only a short course of testosterone therapy is recommended (not more than three months). Additional testosterone is not recommended during childhood as to avoid unwanted virilization and bone maturation [218,219].

Urethral

Urethral Meatal Stenosis

Meatal Stenosis is the abnormal narrowing of the urethral meatus. The opening where the urine passes becomes blocked. This can occur after the newborn is circumcised, and is very rarely seen prenatally. In rare cases it can result in urinary tract obstruction [220-222]. Of males circumcised during the neonatal period, incidence of meatal stenosis can be as high as 20.4% [220]. Moderate to severe cases can be treated by topical ointments and dilation, while severe cases may require surgical intervention [220,223].

Posterior Urethral Valves

Posterior urethral valves (PUV) are the most common congenital obstructive lesions of the urethra, causing bladder obstruction. First described by Young et al. in 1919, PUV is believed to develop 8 weeks into gestation, as the Wolffian duct fuses with the developing cloaca. PUV obstruction during the critical organogenesis period can result in lifelong damage to renal, ureteral, and bladder function [224]. Antenatal US can help identify PUV during early pregnancy if clinical presentations occur. Primary valve ablation is the standard treatment for PUV [225].

Megalourethra

Megalourethra is a rare congenital anomaly involving nonobstructive urethral dilation that affects the anterior urethra. Developmental abnormalities are seen of the corpus spongiosum and corpora cavernosa. It is characterized by two forms, scaphoid and fusiform [226-228]. It is commonly seen associated with prune-belly syndrome (PBS). Scaphoid megalourethra is identified due to the lack of corpus spongiosum. Fusiform megalourethra lack both spongiosum and corpora cavernosa. Fusiform megalourethras are associated with lethal congenital anomalies and commonly present in stillborns. Fusiform megalourethra can be caused by temporary obstruction during early development. Scaphoid megalourethra occurs due to the failed development of corpus spongiosum [229-231]. US findings can help identify and diagnose the type of megalourethra [232-234].

Vaginal

Vaginal Obstruction

Vaginal obstruction is a blockage of the vaginal opening, preventing outflow. This is an extremely rare defect in neonates and infants. However when present, a missed diagnosis can lead to poor outcomes, such as death from infection. Vaginal obstruction can result from the vaginal canal failing to develop,

also known as high transverse septum, or from the vaginal opening being completely covered by the imperforated hymen. Surgical intervention can be used to correct vaginal obstruction. Causes of vaginal obstruction can include low transverse vaginal septum, imperforate hymen, or high transverse vaginal septum. Low vaginal septum can be treated by incision and drainage of hydrometrocolpos, imperforate hymen can be treated by hymenotomy and drainage of hydrometrocolpos, and high vaginal septum can be treated by surgical excision [235]. US and MRI can be used to diagnose antenatal vaginal obstruction [236,237].

Hydrocolpos/Hydrometrocolpos

Hydrocolpos is characterized by an expanding fluid filled distention of the vaginal cavity. If this is associated with distention of the uterine cavity, it is named hydrometrocolpos. It is frequently caused by imperforate hymen, and also, less commonly, transverse vaginal septum. US can be used to diagnosis hydrocolpos. The fluid filled distended vaginal canal may be seen as a mass between the bladder and rectum [236,238-242]. Treatment for hydrocolpos involves a hymenotomy [242].

Testicular and Scrotal

Congenital Hydrocele

Congenital hydrocele (CH) is a collection of fluid within the processus vaginalis, leading to swelling of the inguinal region or scrotum. CH shares similar etiology and pathophysiology as inguinal hernia. During fetal development, the testicle is found in the peritoneal cavity, under the kidney. It descends through the inguinal canal, into the scrotum. Normally the inguinal region and scrotum do not connect. During development if the processus vaginalis fails to close, due to the lack of adequate smooth muscle in the patent processus vaginalis, this allows the peritoneal fluid to enter into the scrotum, resulting in CH [243-246]. Hydroceles can be diagnosed prenatally through US [247,248]. Surgical options may be used to treat CH but in many cases may not be necessary [249].

Cryptorchidism

Cryptorchidism is the most common genital anomaly. It is characterized by obscure or absent testis (either one or both testes) from the scrotum. It is also referred to as undescended or maldescended testis, as the testis fails to descend into a scrotal position [250]. Research shows early diagnosis and surgery are important interventions to reduce the negative impact of cryptorchidism [251]. There are many theories as to the onset of cryptorchidism. Ongoing research suggests cryptorchidism could be caused by loss of HOXA10 and HOXA11, suggested by experiments in knockout mice. HOXA10 polymorphisms are seen in human cryptorchid populations [252-254].

Testicular Torsion

Pre-natal (in utero) torsion is most commonly an extravaginal event. Whereas post-natal torsion is almost invariably extravaginal [255]. It is likely that most cases of "vanishing testis" occur secondary to vascular compromise during descent of

the testis [256,257]. If torsion were to occur during canalicular descent, one would surgically identify blind-ending spermatic cord remnants and/or nubbin remnants in the scrotum [257]. The “acute” hemiscrotum encountered at birth is almost always the end result of extravaginal torsion during the final phase of descent into the scrotum [258]. Testicular salvage is rare as the vast majority have occurred not within hours, but rather days or weeks before birth [259].

Penoscrotal Transposition

Penoscrotal transposition (PST) is an uncommon congenital abnormality where the scrotum is located towards the cephalad position with respect to the penis. Most cases are sporadic, but a genetic factor may be involved. PST results from abnormal genital tubercle development during the 6th week of gestation [260-262]. Surgery can be used to correct certain cases of PST [263].

Bifid Scrotum

Bifid scrotum (BFS) is the less severe form of PST, in which two halves of the scrotum are separated by a raphe fused to underlying subcutaneous tissue. BFS is caused by incomplete fusion of the labioscrotal folds. US and fetal MRI can give an early diagnosis for BFS [214,264]. There is evidence that 5-alpha-reductase type 2 deficiency may be involved in BFS. 5-alpha-reductase type 2 deficiency is an autosomal recessive sex-limited condition that prevents the conversion of testosterone to dihydrotestosterone [265-267].

Prune-Belly Syndrome

Prune-belly syndrome (PBS) was first identified by Frolich et al., also known as Eagle-Barret syndrome. It is referred to as an abdominal muscle deficiency syndrome that often is associated with other anomalies [268]. The exact cause of PBS is unknown [138]. Incidence of PBS is rare and it occurs mainly in males [269]. The incidence of PBS for males is reported as 3.76 cases per 100,000 live births [270]. There are many theories for PBS; the prevailing theory is urethral obstruction and mesodermal arrest. It takes place between the 6th and 10th week of gestation. Histologic findings help support this theory [271]. US can diagnose PBS antenatally [272,273]. Children with PBS generally have severe comorbidities and require frequent surgical intervention. Early end-stage renal disease is also common and approximately 15% of children require kidney transplantation [274].

Allantoic Cysts

Allantoic cysts are a type of cyst of the umbilical cord. Allantois forms from the fetal yolk sac, and connects to the urogenital sinus and base of the umbilical cord. Allantois usually regresses on its own but it may persist and present itself as a cystic mass [275-279]. Antenatal US can identify allantoic cysts [280].

Spinal Dysraphism

Rachischisis

Rachischisis is the most severe form of spina bifida, wherein a cleft is found through the entire spine, and is sometimes referred to as complete spina bifida. Spina bifida is a result of a teratogenic

process, resulting in abnormal differentiation of the embryonic neural tube and failure of the neural tube to close. Rachischisis is almost always fatal. This can be identified using US or fetal MRI, at 18 weeks of gestation [281-283].

Myelomeningocele

Myelomeningocele is a major congenital neural tube defect and is the most common defect among neonates with spina bifida. The problem resides in the formation of a meningeal cyst, which includes the cord tissue. Failure to close at the caudal end of the neural tube causes myelomeningocele, resulting in an open lesion containing dysplastic spinal cord, nerve roots, meninges, vertebral bodies, and skin. Risk reduction is possible with the administration of folic acid [284,285]. At 18 weeks of gestation, US or fetal MRI can be used to diagnose myelomeningocele [281,286]. Reconstructive urological surgeries play a role in protecting the upper urinary tract and achieving continence in patients with myelomeningocele [287].

Myeloschisis/myelocele

Myeloschisis, or myelocele, is the most severe form of myelomeningocele occurring due to a complete opening of the neural tube and exposure of the ependymal layer. US and fetal MRI are diagnostic tools to help identify myeloschisis prenatally [288,289].

Diastematomyelia

Diastematomyelia is a congenital disorder in which the spinal cord is split, at the level of the upper lumbar vertebra. It is characterized by a sagittal cleft. Diastematomyelia is more common in females. It is caused by an osseous, cartilaginous, or fibrous septum, resulting in a complete or incomplete sagittal division into two hemicords [290,291]. It is classified into two types: type I, duplicated dural sac, or type II, single dural sac. Type I, duplicated dural sac, shares a common midline, separating the two hemicords, and type II, contains both hemicords in a single sac [292]. Antenatal US, focused at the midline between the fetal spine posterior, is a reliable tool [293,294].

Treatments

Surgical options to treat antenatal anomalies should only be used when the situation is life threatening urogenital anomalies or anomalies that threaten proper renal function.

Ex-utero intrapartum treatment (EXIT) Procedure

Ex-utero intrapartum treatment (EXIT) procedure, was developed in order to secure the neonatal airway while the fetus is on placental support during an elective procedure by partial delivery. Airway compression can result from many congenital disorders. EXIT procedure is an extension of the classical caesarean section. The fetus is partially delivered while remaining attached to the umbilical cord to the placenta. Then a multidisciplinary team works together to correct the airway compression. Lastly the fetus is fully delivered. Challenges of the EXIT procedure include preservation of enough blood flow through the umbilical cord, protecting the placenta, and preventing uterine contractions.

Maternal morbidity is also of concern with the EXIT procedure [295]. However in many cases, the benefits outweigh the risks [296].

Fetoscopy

Fetoscopy is another surgical procedure that is used for medical interventions. This procedure allows access to the fetus, through a small incision made in the abdomen, and enables access to the amniotic cavity, umbilical cord, and fetal side of the placenta [297,298]. Currently, many anomalies that are amenable to intrauterine surgical treatment are rare [299]. Fetoscopic laser occlusion is also used to treat certain congenital anomalies [300,301].

Myelomeningocele Repair

Myelomeningocele leads to many neurological complications in infant. Brown et al. conducted in utero repair of myelomeningocele using a fetal lamb model. Seven fetal lambs underwent myelomeningocele corrections, using an autologous amniotic membranes patch. This study showed an increased protect of spinal cord, although the overlying skin failed to closed. This is a potential technique but further study is required [302]. Management of Myelomeningocele Study conducted between 1998 and 2003 have shown patients develop no urological complications and decreased the need for ventriculoperitoneal shunting [303].

Fetal Cystoscopy

Fetal cystoscopy is a technique used to treat patients with obstructive uropathy. Sananes et al. collected data of 40 fetal cystoscopies. Twenty-three cases involved in treating PUV. Survival rate of the technique was 61% and resulted in normal renal function in 85% of successful cases. A major complication of this technique is urological fistulas. However, can be avoided if PUV were diagnosed at an earlier gestational age [304].

Vesicoamniotic Shunting

Vesicoamniotic shunting is currently the most common method of treating fetal lower urinary tract obstructions. Involves the placement of a catheter using US guidance. Distal end of the catheter is placed into the bladder and the proximal end into the amniotic cavity to allow drainage [305] **Table 1** and **Table 2**.

Summary

Life threatening urogenital anomalies are detected and treatment options are discussed.

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Compliance with Ethical Standards

The authors declare they have no conflict of interest.

Table 1 Methods of Prenatal Anomaly Diagnosis.

Diagnostic Tools for Prenatal Anomalies			
	Ultrasound	Amniocentesis	Fetal MRI
Adrenal	Adrenal cystic neuroblastoma [23,306,307]	Congenital Adrenal Hyperplasia (CAH) [308-310]	Adrenal cystic neuroblastoma [23,311,312] CAH [312]
Renal	Bilateral renal agenesis [14,313] Unilateral renal agenesis [69,72] Autosomal recessive polycystic kidney disease [314-316] Autosomal dominant polycystic kidney disease [89,317,318] Duplex Kidneys [93,319] Horseshoe kidney [320,321] Cross fused renal ectopia [322] Fused Pelvic kidney [323] Hydronephrosis [324-326] Multicystic dysplastic kidney [313] Renal hypoplasia [16] Pyelectasis/renal pelvic dilation [327,328] Mesoblastic Nephroma [130,329]	Autosomal recessive polycystic kidney disease [330] Autosomal dominant polycystic kidney disease [331]	Bilateral renal agenesis [71,332-334] Unilateral renal agenesis [70,71] Polycystic kidney disease [70,71,74] Autosomal recessive polycystic kidney disease [316,335-337] Autosomal dominant polycystic kidney disease [318] Hydronephrosis [326,338,339] Multicystic dysplastic kidney disease [71] Mesoblastic Nephroma [129,130]
Ureteral Anomalies	Duplication of one or both ureters [340,341] Ectopic ureter [341] Primary megaureter [172,342,343] Ureteropelvic junction obstruction [344,345] Ureterocele [164,165,172,340]		Ectopic ureter [346] Ureteropelvic junction obstruction [166] Ureterocele [166,347]
Vesicoureteral Reflux	Vesicoureteral Reflux [170-172]		
Bladder Anomalies	Bladder diverticulum [348] Exstrophy [349,350] Patent Urachus [351-354] MMIHS [14,171,354]	Menkes Syndrome [355,356]	Neurogenic bladder [286]
Penile	Chordee [201,214] Epispadias [357] Hypospadias [200,201] Microphallus [358]		Chordee [214] Hypospadias [215]
Urethral Anomalies	Urethral stenosis [359,360] Posterior urethral valve [172,361] Megalourethra [232-234]		Posterior urethral valve [166]
Vaginal Anomalies	Vaginal obstruction [237] Hydrocolpos/hydrometrocolpos [238,239,241]		Hydrocolpos/hydrometrocolpos [236]
Testicular and Scrotal Anomalies	Congenital hydrocele [247,248] Cryptorchidism [362,363] Testicular torsion [247,364] Penoscrotal transposition [214,365] Bifid scrotum [214,264]		Cryptorchidism [366] Penoscrotal transposition [214] Bifid scrotum [214]
Prune-belly syndrome	Prune-belly syndrome [171,272,273]		
Allantois	Allantoic cysts [280,353]		
Spinal Dysraphism	Rachischisis [281] Myelomeningocele [281] Myeloschisis (myelocele) [289] Diastematomyelia [293,294]		Rachischisis [281] Myelomeningocele [281,286] Myeloschisis (myelocele) [288] Diastematomyelia [293]

Table 2 Earliest trimester in which the anomaly was detected.

Trimester of Prenatal Anomaly Identification			
	1 st Trimester (1-12 weeks)	2 nd Trimester (13-27 weeks)	3 rd Trimester (28-40 weeks)
Adrenal	CAH [308,309,367,368]		Adrenal cystic neuroblastoma [307,369,370]
Renal		Bilateral renal agenesis [332,333,371] Unilateral renal agenesis [69,372] Autosomal recessive polycystic kidney disease [315,373,374] Autosomal dominant polycystic kidney disease [89,331] Duplex Kidneys [319] Horseshoe Kidney [320,321,375] Cross fused renal ectopia [322] Fused Pelvic kidney [323] Hydronephrosis [326,376] Multicystic dysplastic kidney [14,115,377,378] Renal hypoplasia [16] Pyelectasis/renal pelvic dilation [14,379,380] Mesoblastic Nephroma [381]	
Ureteral Anomalies		Ureterocele [165]	Ureter duplication [340] Ectopic ureter [346] Megaureter [382,383] Ureteropelvic junction obstruction [336,384,385]
Vesicoureteral Reflux			Vesicoureteral reflux [171,386]
Bladder Anomalies	MMIHS [14,171,354,387] Patent Urachus [354] Menkes disease [388-390]	Exstrophy [313]	Bladder diverticulum [348]
Penile		Hypospadias [200,391] Microphallus [392]	
Urethral Anomalies		Posterior urethral valve [393] Megalourethra [232-234]	Urethral stenosis [360]
Vaginal Anomalies		Hydrocolpos/hydrometrocolpos [239]	Vaginal obstruction [237] Hydrocolpos/hydrometrocolpos [238,241,394]
Testicular and Scrotal Anomalies		Bifid scrotum [395]	Congenital hydrocele [248,396] Cryptorchidism [362,363] Testicular torsion [364,397] Penoscrotal transposition [365]
Prune-belly syndrome	Prune-belly syndrome [171,272,273]		
Allantois		Allantoic cysts [280]	
Spinal Dysraphism		Rachischisis [281,282] Myelomeningocele [286]	Diastematomyelia [293,398]

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