Uretero What? A Systems-level View of a Pregnancy with Medical Complications

by

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Part I— Gynecological Health Issues

Yvonne has been a student of science nearly all her life. She values her independence and loves her career in the biopharmaceutical industry. She is extremely focused, as evidenced by her three-, five-, and 10-year plans for her career. Her plans include having a child, once she has realized her career aspirations.

Yvonne's recent promotion to senior principal scientist at the young age of 30 means she now has more responsibilities at work. She feels a deep sense of security as her dreams are coming to fruition. However, she is about to learn that the best laid plans have a way of, well, not working out as planned.

Seldom needing to see a doctor for most of her life, Yvonne has spent the past several months receiving gynecological treatment for cervicitis and cervical neoplasia, a disease for which the cause is unknown. A couple of weeks ago, Dr. James Trent, her gynecologist/obstetrician, ordered several tests and performed an in-office diagnostic procedure to try to narrow down the cause of Yvonne's condition.

At her next scheduled appointment, Dr. Trent tells Yvonne that the tests for bacterial or viral causes (which would indicate the presence of a sexually transmitted disease, or STD) are negative. He explains that the procedure Yvonne had undergone last month, a colposcopy with cervical biopsy to evaluate the cervical topography and its epithelial character, is benign for cervical cancer.

"The results are promising," Dr. Trent explains, "but I am afraid that at this point we have more questions then answers. I believe our best course of action is an outpatient procedure called cervical cryosurgery."

"What is the intent of the surgery?" Yvonne asks.

"With cervical cryosurgery, we remove the superficial layers of tissue lining the cervix in hopes of permanently removing any abnormal cells, whatever their cause, thereby eliminating the precancerous condition," Dr. Trent answers.

The following week, Yvonne has the outpatient cryosurgery procedure. At her next appointment, Yvonne listens closely as Dr. Trent says, "The cervical cryosurgery didn't achieve the result we had hoped for; the cervical abnormality has in fact recurred.

"Our remaining option is a surgical procedure called cervical conization (or cone biopsy). We remove a cone-shaped section of tissue from the endocervix and send it to a pathologist to determine whether all abnormal tissue containing the precancerous cells is likely to have been removed."

Dr. Trent continues, "A cone biopsy will often cure the problem. However, in a small number of cases, it may interfere with childbearing. You could choose to have the conization surgery now before a pregnancy, or after. It's your decision.

"If you choose to have the procedure now, you will have more time to prepare for a pregnancy at a later date. However, because cervical conization involves removing endocervical tissue, the ability of the uterus to bear weight will be diminished. As the fetus' weight increases, added strain will be placed on the muscular wall. About half way through your pregnancy, you most likely would be placed on bed rest until the baby comes," Dr. Trent explains. "If you decide to become pregnant first, we can perform the surgery at a later date. I would recommend you start taking daily prenatal vitamins now to prepare for a pregnancy in case you decide to pursue such an option sooner rather than later."

Yvonne's choices are clear. She must decide between having a child in the not-too distant future with little to no risk of complications, or have a high-risk pregnancy at a more fitting time in her life. Yvonne takes some time to consider her options, turning to the people closest to her in her private life for support and advice. At age 30, although still young, Yvonne is cognizant of the medical literature concluding that a woman's ability to conceive a child decreases with advancing age.

Questions

1. What is the anatomical relationship between the cervix and the uterus?
2. What are the current statistics on the frequency of occurrence of cervical cancer? Does a women's race affect her chances of developing this cancer? What is the standard test for detecting cervical cancer?
3. Most cases of cervical cancer result from human papillomavirus (HPV) infection. What information can you find about transmission, contraction, and consequences of HPV from valid medical sources?
4. The success of the new HPV vaccine (approved in June 2006) will rely on parents' willingness to vaccinate their prepubescent daughters. Discuss your own willingness to do so were you a parent of a young girl.
5. Yvonne faces a potentially life-changing decision—to have a child now or wait until after a surgical procedure. Help her weigh the pros and cons as you see them.

Part II—Yvonne Chooses

Yvonne decides to have a child before opting for the cervical conization surgery. At Dr. Trent's suggestion, she has already begun taking prenatal vitamins, and with the assistance of an over-the-counter ovulation predictor kit, she is able to closely approximate the days of the month when she is most fertile and likely to conceive.

After the second month of trying to conceive, Yvonne's home pregnancy test shows a positive result. With deep feelings of anxiety and elation at the thought of carrying a baby, Yvonne calls Dr. Trent's office to schedule an appointment. The urine test conducted in his office confirms the results of the home pregnancy test.

Dr. Trent, with calendar in hand, tells Yvonne, "Your estimated due date is March 7th. I want you to continue to take the prenatal vitamins once daily for the extra iron and folic acid you need, and add four glasses of milk or milk equivalents daily. Second," he continues, "moderate caffeine is okay, but no smoking or drinking alcohol. Continue your normal exercise routines, but listen to your body in terms of fatigue or nausea. Third, at about 14 weeks gestation, we'll schedule routine blood work to screen for spina bifida and Down syndrome. Then, between weeks 24 and 28, we'll do the routine screen for gestational diabetes using an oral glucose tolerance test. Now, I know that neither you nor anyone in your family has a history of genetic conditions or diabetes, but such screening is routine.

"Finally, Yvonne, I would like you to schedule prenatal appointments with me once every four weeks. Also, as with all my expectant mothers, schedule an appointment with Dr. Costa at 20 weeks for a fetal medicine survey. Dr. Jean Costa is a top fetal medicine specialist. She will analyze the condition of the baby's major body organs and alert me of any abnormalities at the midpoint of prenatal development."

Yvonne leaves the doctor's office that day with her next appointment scheduled in four weeks. That time passes by uneventfully, except for the morning sickness during the whole first trimester that actually occurs anytime, day or night. She tries to make good food choices, and understands that gaining some weight during her pregnancy is natural. In fact, as she learns from Dr. Trent, the Institute of Medicine recommends that for women like Yvonne, who was of normal weight before her pregnancy, total pregnancy weight gain should be 25–35 pounds.

It is a Tuesday morning when Yvonne, now 20 weeks pregnant, has her planned ultrasound performed at the office of the fetal medicine specialist. Gloria, the ultrasound technician, asks Yvonne if she wishes to have the sex of the unborn child determined. "Absolutely," Yvonne replies emphatically.

Gloria begins the imaging procedure, and Yvonne sees a clear picture showing the body image, four heart chambers, blood in circulation, and individual body organs of the fetus.

"It's a girl!" Gloria exclaims. Yvonne is delighted. Gloria then captures several sonogram images of the unborn child for Yvonne and for Dr. Costa's examination.

Dr. Costa begins her consultation with Yvonne on a somber note. "Yvonne, unfortunately, I have some bad news; there is a problem with the baby's kidney. There is an abnormal accumulation of urine in one kidney, a condition called hydronephrosis. This situation could be an indication of a more severe and widespread genetic disorder, or it could be limited to a birth defect of a single organ."

"How can we be sure if it is limited to the kidney?" Yvonne manages to ask.

Dr. Costa responds, "I want to do amniocentesis in order to determine the extent of the abnormality. Although the procedure poses some risk—about one percent of miscarriages are related to the procedure—I would recommend it so that we can better understand the fetal anomaly."

Yvonne agrees to the amniocentesis, but is in utter shock. The words "genetic disorder," "hydronephrosis," and "fetal anomaly" echo in her head as she waits for Dr. Costa to finish the day's patients and perform the procedure.

Within an hour or so, it is underway. Yvonne sees the needle penetrate her abdomen, and watches on the sonogram screen as the needle enters the amniotic sac. In a matter of minutes, it is over and she is again seated in the doctor's consultation chair. Dr. Costa explains that Yvonne needs to go home and rest for the remainder of the day. When Yvonne returns for her appointment next week, Dr. Costa will have the results of the genetic testing.

Over the next several days, Yvonne thinks about the results of the genetic tests and her options. The joy of knowing she is having a little girl is shadowed by the range of possibilities in her case: from all being well with her baby, to the child needing some sort of corrective surgery, to severe developmental problems.

Questions

1. What is the range of normal gestation for humans? What are the current guidelines for good prenatal care beyond those mentioned here? For example, why do pregnant women need extra calcium, iron, and folic acid?
2. In cases of severe morning sickness, are prescription medicines advised? If so, what is the mechanism of action of these drug(s)?
3. What hormone(s) in the urine do over-the-counter ovulation predictor kits, fertility monitors, and early pregnancy tests (EPTs) measure? How soon after conception can EPTs be performed?
4. How does ultrasound technology allow image capture of the fetus in utero? When is an ultrasound performed during pregnancy? When is an amniocentesis warranted and how is it analyzed?
5. Up to 5% of all pregnant women are diagnosed with gestational diabetes, and women who have had gestational diabetes have an increased risk of developing Type 2 diabetes later on in life. Distinguish between gestational diabetes and Type 2 diabetes in terms of cause, management, and consequences/outcomes.

Part III—Andrea

The week spent waiting for the amniocentesis results is one of the most mentally draining of Yvonne's life. Her insatiable curiosity leads her to investigate on the web what the doctor called "hydronephrosis." Her research reveals reports of infant mortality in some cases, while many cases require that the infant undergo corrective surgery.

"Have a seat," says Dr. Costa, as Yvonne enters her office. "I want you to know that the results of the genetic tests came back normal for your baby girl. What we are dealing with is most likely limited to a congenital condition—in other words, a developmental birth defect—with the left kidney. To be certain, I would like you to see Dr. Steven Solfvin, a pediatric cardiologist. Dr. Solfvin will check to see if there is normal cardiac development *in utero*. I would also recommend that you see a pediatric nephrologist, Dr. Mark Mackey, on a bimonthly basis for ultrasounds and examinations pre and post birth."

Yvonne meets with Dr. Solfvin, who confirms by more ultrasonography that the birth defect most likely is limited to the kidney, as the baby's heart is developing normally. Dr. Mackey confirms the hydronephrosis diagnosis, and suggests that the condition stems from a problem with the baby's bladder. Dr. Mackey also mentions that until direct imaging is conducted on the child after the birth, he cannot accurately predict the extent of the urinary tract abnormalities.

Yvonne's preparation for her daughter, whom she names Andrea, intensifies now that she is more than half way through her pregnancy. She is thankful and aware of how fortunate she is that her employer's medical coverage plan affords nearly full coverage of all medical expenses accrued by her regular visits to various medical specialists for monitoring her unborn child's health.

One day short of the 35-week mark for the pregnancy, Yvonne is giving an important oral presentation to members of her company's executive board. As she introduces herself and the topic of her presentation, without any warning signs, Yvonne's water breaks. While on her way to the hospital, Yvonne can not help but feel that the best laid plans …

Five hours later, baby girl Andrea is born. She weighs 2,925 grams and is 47 centimeters long. Having earned a normal score on the Apgar scale, and making the minimum weight of six pounds, Andrea is admitted to the regular nursery rather than to the neonate ICU. Twenty-four hours after birth, renal and bladder ultrasonography is performed on Andrea in the hospital. The next day, a stress test in her car seat in the hospital (required for all preterm babies) confirms that Andrea is ready to go home. Yvonne is a very happy mother.

During the first few weeks of her life, little Andrea begins daily antibiotic prophylaxis treatment to lessen her risk of urinary tract or kidney infections. She undergoes outpatient trips to the hospital for diagnostic imaging procedures (nuclear renal scans, CT scans, and more ultrasonography) and functional studies such as a voiding cystourethrography to determine the extent of her urinary tract abnormality.

Dr. Mackey makes the diagnosis of ureterocele associated with a duplicated ureteral system. This is a condition where the terminal segment of the ureter is dilated and is ballooning into the bladder. This fluid-filled structure within the bladder causes vesicoureteral reflux to the kidneys. If left untreated, there is a risk for urinary tract infections and obstructive uropathy, which may progress to renal scarring and eventual renal failure.

Dr. Mackey tells Yvonne that this can be corrected by a surgical procedure called transurethral puncture. Although elective, the procedure is strongly recommended by the medical profession. Yvonne is once again faced with a difficult decision: to have Andrea undergo the transurethral puncture now, or delay the surgery until she is older and perhaps has a urinary tract infection.

Yvonne elects for Andrea to have the outpatient procedure. At three months old, Andrea undergoes the transurethral puncture with general anesthesia. Although Andrea's procedure does decompress the ureterocele, it leads to a complication called kidney reflux that often is seen with this type of corrective surgery. When she urinates, a percentage of the urine goes back up the ureter into the kidney rather than all voiding via the urethra. The severity of kidney reflux will be determined by diagnostic and functional outpatient evaluations every six months to assess the need for secondary surgical procedures.

Medical Update Three Years Later . . .

Yvonne, in consultation with her gynecologist, underwent a successful cervical conization surgery when Andrea was about two years old, and remains in good health. The extent of kidney reflux in Andrea continued to worsen to the point where at three years of age function in her left kidney was compromised. Now, Yvonne and her family are preparing for Andrea's upcoming surgery to repair the bladder abnormality and stop the kidney reflux. This surgery will require an open abdominal incision, a few days in hospital, and recovery at home for one to two weeks. Dr. Mackey, the pediatric urologist, explains to Yvonne that this surgical intervention is 98% likely to be all that will be required to permanently correct Andrea's condition.

Questions

1. When a newborn is delivered, an Apgar score is determined. What is the scale for such a score and what information does it provide the medical practitioner?
2. Diagram the anatomy of the urinary tract from kidney to urethra.
3. What are the common causes of premature births? What sort of medical care in the hospital immediately post birth is necessary for a premature baby, one born much earlier than Andrea?
4. Research the incidence rates of the more common birth defects. How common are ureteroceles? What is the long-term outcome for surgical treatment to repair a ureterocele?
5. Yvonne is faced with making a decision on behalf of her infant that would involve surgical treatment. There are inherent risks with surgery, regardless of age. Research the condition malignant hyperthermia, the most common cause of death related to general anesthesia.