Hydrocolpos/hydrometrocolpos

Patient Information Series – What you should know, what you should ask.

What are Hydrocolpos and Hydrometrocolpos?

Hydrometrocolpos is the technical term for a condition where there is fluid in the vagina and the uterus. The words derive from the Greek words, “hydro-“ water, “colpo-“ vagina and “metro-“ uterus.This is a condition which typically presents late in pregnancy, at around 30 weeks on average.

How does Hydrocolpos or hydrometrocolpos happen?

These conditions are relatively rare and occur only in female fetuses. During pregnancy, hormones produced in the mother cause increased secretions in the uterus, cervix and vagina of the baby. When there is a blockage to the flow of secretions out of the vagina, they accumulate in the baby’s vagina and sometimes in the uterus.

There are several places where these secretions can be blocked. The sites where these blockages can occur include:

the hymen (a normal structure at the end of the vagina), a transverse septum in the vagina (piece of tissue crossing the vagina and blocking it higher up in the vagina), vaginal atresia (a condition where the vagina doesn’t form properly) or persistence of the urogenital sinus (a normal structure in early pregnancy which should disappear) or a cloaca (an abnormal separation of the urethra, which is the canal letting the urine out of the bladder), the vagina and the rectum so that there is only one opening for urine, vaginal secretions and stool rather than the three normal openings.

There are a variety of reasons why this occurs. In some fetuses, the condition is isolated, meaning that this is the only problem that the baby has. In other cases, there is a genetic syndrome which explains the findings. Common genetic syndromes or diseases include McKusick-Kaufman syndrome (hydrocolpos, extra fingers or toes and a heart defect), Herlyn-Werner-Wunderlich syndrome (malformation of the uterus, blocked vagina and absence of kidneys), Mayer-Rokitansky-Kuster-Hauser syndrome and MURCS (absence of complete development of the vagina and uterus, absence of kidneys, spine anomalies). When the fetus is found to have additional ultrasound abnormalities, your doctor may examine for one of these genetic syndromes.

Should I have more tests done?

Your doctor may recommend further testing be performed to know more about the baby. An amniocentesis is commonly suggested to check for an abnormal number of chromosomes and to see if the baby is genetically male or female. This is especially important if the sex of the baby cannot be clearly be seen by ultrasound.
A careful ultrasound of the baby should be performed. The hands and feet should be looked at closely to see if there are extra fingers or toes, and the kidneys and heart should be thoroughly

evaluated. It is important to make sure that the baby is clearly female, that there is an anus, and that the rectum is normal in appearance.

Sometimes, a fetal MRI may be recommended to see if additional information can be obtained. MRI uses magnetic fields to create images of the inside of the body. It is safe in pregnancy and can be helpful if there is a concern for a urogenital sinus malformation or a cloaca. Since hydrocolpos and hydrometrocolpos are commonly found late in the pregnancy, MRI may not change the management of the pregnancy. It is also possible that MRI may not be available where you live or is not always performed.

What are the things to watch for during the pregnancy?

Additional ultrasound examinations will be performed to evaluate your baby's growth and development to watch for an abnormally small baby. The fluid in the pelvis will be re-evaluated at each visit. Your doctor will make sure the baby is growing well. Other things to look for include extra fluid (ascites) in the baby's abdomen, oligohydramnios (too little fluid in the sac around the baby), polyhydramnios (too much fluid in the sac around the baby) and compression of the kidneys. If these problems occur, the management of the pregnancy could change.

If no additional complications are identified during pregnancy, delivery of the baby should be at term. Fetuses with hydrocolpos or hydrometrocolpos may deliver vaginally; caesarean delivery is not necessary in most cases, unless there is another reason for it.

What does it mean for my baby after it is born?

After the baby is born, the pediatric team will evaluate the baby with a physical examination and blood tests to check to see if the kidneys are functioning normally. Most likely, the mass will be able to be felt on the abdominal examination. The baby will have a careful evaluation of the external genitalia and anus. The pediatricians may order an MRI to help evaluate the internal organs of the baby and to exclude imperforate anus and urogenital sinus. A pediatric genetics doctor may also evaluate the baby to determine if the baby has a genetic syndrome.

Based on these results, the pediatricians will reach a final diagnosis for the baby and recommend the best course of action. If the baby has obstruction at the hymen, a simple incision into the hymen may be all that the baby requires. If the vagina is absent in part, surgery may be necessary to place the vaginal opening in its normal position. If there is a vaginal septum, this may need surgery to open.

If a cloaca or a persistent urogenital sinus is found, the baby may require multiple surgeries by an experienced team at a specialized hospital. How well the baby will do and the number and type of surgeries and the timing of the surgeries recommended for the baby depends on the exact problems identified after birth. Some children with cloaca or persistent urogenital sinus will have life-long difficulties with bowel movements, incontinence or may need a tube placed in their bladder when it needs to be emptied. Some will have long term issues with kidney

function. Others will require further surgical repair of the vagina prior to sexual activity. Many children, however, have no long-term complications.

Will it happen again?

The chance that your next baby has the same problem can only be determined once the baby is examined and the final diagnosis is obtained. If the baby has a genetic syndrome, the risk of this happening again will depend on that specific syndrome. Conditions like McKusic-Kaufman syndrome, for example, have a 1 out of 4 chance of happening again. Other conditions have no additional risk. Consultation with a genetic specialist may be helpful in determining future risk.

What other questions should I ask?

- How sure are you of the diagnosis?
- Do you see any other abnormalities in my baby?
- Have you evaluated the rest of the body of the baby thoroughly?
- Is the growth of the baby fine?
- How often will I have ultrasounds performed to follow up on this issue?
- Where is the best place for my baby to be born?

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