

PREGNANCY AND HHT: WHAT YOU NEED TO KNOW

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Pregnancy, labor and delivery may be a risky time for a woman with HHT, but this doesn't need to be. Most women with HHT can have a normal pregnancy and delivery with no more risk for complications than women in the general population. HHT pregnancies are sometimes considered "high-risk", but this is not necessarily always the case...some women with HHT can benefit from a "high-risk" clinic approach, but others don't really need this. **The key is information and consultation with an HHT expert**, to help the pregnant woman make the best decisions for herself and her baby.

Information is the key to helping women and their physicians make the best decisions about their care. This means being aware of the extent of the HHT in the Mother (her "HHT footprint") and planning accordingly, rather than simply assuming that HHT is not a problem, or conversely assuming that the pregnancy is high-risk due to the simple presence of HHT. By the "HHT footprint", we mean a conceptual map of the abnormal vessels in someone with HHT, including everything from telangiectasia in the nose to AVMs in the organs. The HHT footprint is personalized, different in each woman with HHT, and can change over her lifetime. **Knowing the specific HHT footprint in a woman should help guide pregnancy planning and care.** For example, if a woman's HHT footprint includes only telangiectasia in the nose and she has no abnormal blood vessels in the organs, then there is no reason to think that her pregnancy or delivery is high-risk. On the other hand, if a woman's HHT footprint includes lung AVMs, then several steps and precautions need to be taken to minimize risk, and in some cases this may be considered a high-risk pregnancy.

What does the term high-risk pregnancy really mean? First, it doesn't necessarily mean that the risks are extraordinary, but rather that they are higher than in the general population. A high-risk pregnancy is one in which some condition puts the mother or the developing fetus, or both, at an increased risk for complications during or after pregnancy and birth. These

women may benefit from additional monitoring and specialized care during pregnancy and delivery, to reduce the risk of complications. This can be also very reassuring for the mother and the family, when there is higher risk.

As typical for rare diseases, there are not many studies published about pregnancy and delivery in HHT. As such, there are no clear guidelines for physicians to follow for care of pregnant women with HHT. However, the published experience in the last few years has been reassuring and can help dispel some of the myths and concerns about risks for pregnancy in women with HHT.

Myth #1: Women with HHT are at risk of excessive bleeding (from the uterus) during or after delivery.

NO, there is no evidence of this. In fact, the experience is that women with HHT are no more likely to bleed excessively at delivery or after delivery than women in the general population (ref Wain, Shovlin, Els).

Here's why: People with HHT don't have a clotting disorder. So, they are not more likely to bleed with an injury, trauma or cut. People with HHT bleed from parts of the body where they have abnormal blood vessels (AVMs and telangiectasia). The uterus is not a typical place for AVMs to occur in HHT, so there is no reason to expect women with HHT to bleed with delivery.

Myth #2: Women with HHT can't have an epidural.

NO, this is not the case. In fact most women with HHT decide to have an epidural, or at least to have the option of having one. There is no evidence of increased risk of complications.

Why is there concern? Many anesthetists and other doctors are worried about the risks of epidural anesthesia in women with HHT. Specifically, they are worried about puncturing a spinal AVM when they insert the epidural needle into the women's lower spine. This worry stems from the knowledge that approximately 1% of HHT patients can have AVMs in the spinal canal. BUT, the vast majority of spinal AVMs in HHT are much higher in the spinal canal than where the epidural needle is usually inserted. ALSO, complications of spinal AVMs are exceedingly rare in adults (most have come to attention during childhood). In fact screening for spinal AVMs hasn't been routinely recommended in adults with HHT (unlike screening for lung and brain AVMs) given how very rare they are in adults. FINALLY, no case has ever been reported in the medical literature of an epidural causing spinal bleeding in HHT. In other words, if there is a risk, it is so low that no one has been able to document a case or quantify the risk. On the other hand, there is no study that has "ruled out" any risk of epidural in HHT. For all of these reasons, we call it a "theoretical" risk.

If a pregnant woman with HHT wants the option of an epidural, she should meet with the anesthetist prior to delivery and have

a discussion about all the risks, including the “theoretical” risk related to HHT. Many anesthetists will agree to proceed if the woman understands the risks.

Myth #3: Women with HHT should have a cesarean section to avoid bleeding and other HHT-related complications with labor and delivery.

There is no evidence to support this as a routine approach. There are three main concerns that have led some doctors to recommend cesarean section for women with HHT.

The first concern is that women with HHT might be likely to bleed during or after delivery. This is addressed in Myth #1.

The second concern is that a brain AVM might be more likely to rupture when “pushing” at delivery. There is no evidence that this is the case, or that cesarean section is a safer alternative to normal vaginal delivery in people with brain AVMs. This situation needs to be handled on an individual basis. Screening for brain AVMs can be performed in early third trimester with an unenhanced MRI of the brain. If the MRI is negative (i.e. no brain AVMs), then this is no longer a concern (90% of women with HHT will have a negative MRI of the brain). If the woman does have a brain AVM then her case needs to be discussed in detail with a brain AVM expert and an obstetrician, and likely should be considered a high risk pregnancy and delivery. There may be some discussion about assisting the delivery to reduced pushing or about a cesarean section, though there is no good evidence that this is the safer approach. These decisions will need to be made on a case by case basis, with all the experts involved. If a brain AVM is diagnosed, the usual recommendation is to treat within two or three months after delivery. As such, since treatment will be delayed anyway, some women opt to postpone the brain MRI till after delivery. This is not unreasonable, but may leave the woman and her physicians unnecessarily worrying about the possibility of brain AVMs, and therefore considering it a “high-risk” pregnancy, whereas an MRI would dispel this worry in 90% of women.

The third concern is that lung AVMs might be more likely to rupture and bleed when “pushing” at delivery. There is no evidence of this. However, lung AVMs do seem to be more likely to rupture during pregnancy if untreated. Screening and preventative treatment for lung AVMs should be done prior to pregnancy, when possible. If lung screening is negative, then lung AVMs are no longer a concern for pregnancy and delivery. If the woman has lung AVMs and these have been successfully treated (and recently reassessed), then lung AVMs are not a concern for pregnancy and delivery, and the pregnancy need not be considered high risk, though some precautions* should be followed. If a pregnant woman has untreated lung AVMs, treatment during T2 or T3, with embolization should be considered, but only at an expert HHT centre. Once treated, the lung AVMs are no longer a significant concern for pregnancy and delivery. If lung AVMs are present but not treated, then the pregnancy should be considered high-risk.

Summary

For most women with HHT, the main problem is nosebleeds, and though bothersome, this is not often a major concern during pregnancy. Some women with HHT have a more extensive HHT footprint, with AVMs in the brain or lungs, which may be more concerning. However, with the right screening, treatment and surveillance, most women with HHT can have normal pregnancy and delivery, with no more risk than women without HHT.

**Any patient with lung AVMs (treated or untreated) should follow pulmonary AVM precautions, including antibiotics before any procedures that can cause bacteria in the blood as well as an air filter (bubble trap) any time they have an intravenous access.*

References

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