Birth Control and Hormonal Therapy in PAH

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The risk of pregnancy-related death in women with PAH is substantial, with risk of dying reported to be 30 to 50%. For this reason alone, pregnancy is contraindicated in patients with PAH. In addition to the hemodynamic load of pregnancy, which means that your heart has to work extra, medications used for PAH are teratogenic (harmful) to the developing fetus, including warfarin (Coumadin); endothelin receptor antoagonists (Bosentan and ambrisentan) and aldactone (which may be used by PAH patients), and further add to the risk of pregnancy. Because of the risk to both the patient and the fetus, use of some form of birth control and avoidance of pregnancy is strongly advised in women of childbearing age with PAH.

There are no published guidelines for birth control use in PAH, and there is no consensus regarding the best form of birth control. The two safest methods of birth control are 1) the barrier method, which may include condoms in men and/or a diaphragm with spermacide in women, and 2) a vasectomy in the male partner for a woman with PAH in a monogamous (one partner) relationship. The failure rate in preventing pregnancy with barrier methods, when used properly, is quite low. Tubal ligation was felt by many PH specialists to be an acceptable option in patients who were not in severe heart failure. In the sickest patients, consideration of other birth control methods is recommended.

In a survey of 23 PH specialists from North America and Europe, the majority felt that use of estrogen-containing birth control pills (BCP) was acceptable as long as patients were anticoagulated with warfarin. Birth control pills containing the lowest amount of estrogen are recommended. However, nearly half of the specialists did not advocate using BCP for their patients, and some actively discouraged patients from doing so because of concern over the possible role of estrogen in worsening PAH. Sixty-five percent of specialists indicated that use of hormonal therapy in post-menopausal women with PAH was acceptable. Progesterone alone, taken orally (by mouth), can be as effective as estrogen in combination with progesterone in preventing pregnancy. However, when using progesterone alone, the pills must be taken at the same time every day to be effective. Intramuscular injections of progesterone (Depo-Provera) are another option; however, this is often accompanied by fluid retention, which may be harmful to patients with right heart failure. There is also a 20 to 30% incidence of irregular, heavy menstrual bleeding associated with the use of progesterone injections, that may be worsened by the use of warfarin. Furthermore, intramuscular injections may be associated with localized bleeding in patients receiving warfarin.

A potential interaction exists between endothelin receptor antagonists such as bosentan (Tracleer) and hormonal birth control, although no studies have demonstrated this to date. Women should not rely on hormonal birth control alone when taking bosentan. Also, many antibiotics can decrease the effectiveness of the hormonal forms of birth control. Thus, if a woman with PAH requires antibiotics, an additional method of contraception should be used during the entire menstrual cycle until the next period occurs.

Intrauterine devices (IUDs) can be used for birth control, although this is not recommended as the first choice for birth control. Traditional IUDs may be associated with excessive bleeding at the time of menses, that may be exacerbated by the use of warfarin. The device should be removed if excessive bleeding occurs. A newer IUD that releases progesterone (Mirena) has a lower risk of bleeding (and has been used to treat excessive menstrual bleeding) and may be an option for some patients. Prophylactic antibiotic use should be used at the time of insertion and removal in most patients with PAH related to congenital heart disease.

The use of surrogate mothers (women who have fertilized eggs inserted into their uterus to carry a fetus) is an option that some PH specialists have considered for selected patients, although this has been done in only a few instances. There are many ethical, practical and medical issues associated with the use of surrogate mothers for PAH patients, that should be discussed not only with treating physicians but also with experienced reproductive counselors and often with a patient's religious leader.