

LIVING WELL WITH HAE

HAE & Gynaecology

Dr. Andrew Pesce

Watch the video: <https://haeaustralasia.org.au/resources/video-resources/>

HOW DO HORMONES AFFECT FEMALE HAE PATIENTS? (VIDEO REF: 0:18)

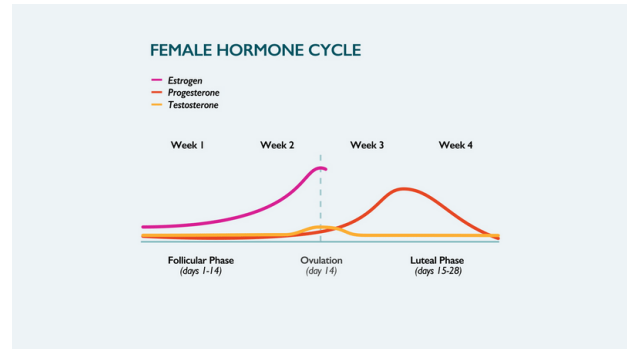
Hormones are very important in HAE and there are triggers for HAE attacks that are related to changes in the hormonal cycle. It's thought that estrogen has an effect on the bradykinin pathway, which is the ultimate trigger for the angioedema episodes because of fluid leaking out of blood vessels. It's also thought that estrogens are a significant factor in the background, baseline condition. By preventing fluctuations of estrogens levels, it may be that the background activity of the disease can be lessened.

WHAT HORMONAL TREATMENTS ARE AVAILABLE TO HAE PATIENTS? (VIDEO REF: 1:08)

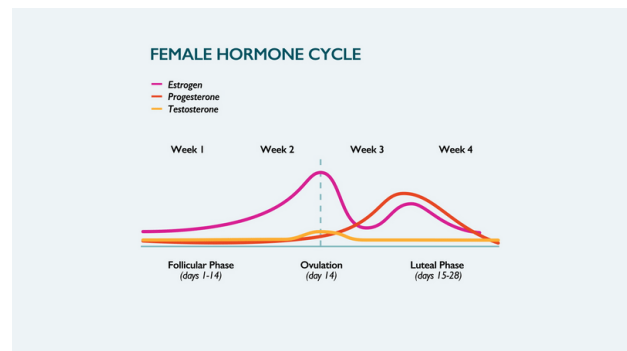
A Gynaecologist would always work in collaboration with the primary treating physician who has specialist knowledge of immunology. Often a patients' basic treatment plan manages symptoms. If, despite those preventative strategies the patient still gets breakthrough episodes due to hormonal changes, she will be referred to a Gynaecologist who will try to optimise the hormonal status of that woman and minimise those events.



The treatments that are available rely on suppressing ovulation and avoidance of estrogens. That's important because those two things are associated with increasing estrogen levels. Ovulation is associated with a surge in estrogen.



The decrease in estrogen levels before periods can trigger events as well. So it's not just an increase, it's a decrease as well, a change in estrogen levels.



Adding estrogen and giving women estrogen containing medications can also trigger HAE. So the Gynaecologists expertise is in finding how to achieve what a woman needs, at the various times in her life where hormones are necessary, and minimise the impact and worsening of the HAE.

IS ESTROGEN A TRIGGER ALL THE TIME OR JUST WHEN PREGNANT OR USING ORAL CONTRACEPTIVES? (VIDEO REF: 2:46)

Estrogen levels can trigger HAE episodes at any time in a woman's life. Many women have described that they never had HAE attacks until they hit puberty.

Hormonal triggers

- Puberty – increase in estrogen
- Going on the oral contraceptive pill
- Pregnancy
- Hormone treatments for menstrual disorders
- Menopause – decrease in estrogen

HOW CAN PATIENTS MANAGE HORMONAL AND CYCLE RELATED CHANGES? (VIDEO REF: 3:56)

The management will depend on what symptoms they're experiencing. If a woman is lucky and doesn't get exacerbations, nothing has to happen. If a woman after puberty has gynaecological problems associated with hormonal levels that need management, her HAE needs to be taken into account when organising treatment. It's important to understand that individual responses are variable, with or without HAE. The reason there are so many types of contraceptive pills on the market is because there isn't one that's good for everyone.

The treatment requires some fine tuning to find what's best for that individual woman. The general principles, however, are to avoid estrogens, and use non estrogen hormones to achieve the same results as the conventional estrogen containing preparations whether it's for contraception, hormone replacement therapy, or cycle control.

Gynaecologists are experienced at finding ways to get around the need for estrogen. And normally, it's by picking a progesterone analogue hormone. Progesterone is a naturally occurring hormone. Gestagen's are other hormones that have progesterone effects, but aren't actually progesterone, which is orally absorbable.

The use of medications such as Provera, or Primolut which contain those other hormones to suppress the cycle and achieve the desired result - whether it's contraception, hormonal control, cycle control or even relief of menopausal symptoms without the use of estrogen.

HOW CAN PATIENTS MANAGE BIRTH CONTROL? (VIDEO REF: 5:52)

There are two aspects to think about, apart from the contraception. Because of the HAE, we also want to suppress estrogen levels. For example, you might get satisfactory birth control by using various methods like condoms. But that's not going to suppress ovulation, so it may not achieve the therapeutic goal. We want to avoid estrogen containing oral contraceptive pills. So there are progesterone or progestogen alternatives.

There are also inter uterine devices, although they don't necessarily suppress ovulation. Therefore devices like the Mirena which might be good for contraception and cycle controls, may not be ideal.

The Gynaecologist may need some alterations to their treatment plan depending on the patients' responses. Don't be disheartened, there are always other alternatives that can be considered.

IS THERE ANYTHING A PATIENT NEEDS TO DO IF SHE'S TRYING TO GET PREGNANT? (VIDEO REF: 7:07)

Preparation for pregnancy is a very important part of a woman's life. And for all women, there are certain things that are advised - avoidance of alcohol, smoking, getting any underlying health issue as stable as it can be, and avoid taking medications which might cause problems in pregnancy.

If you are on medications, which are not desirable in pregnancy, see if you can stabilise your condition using the alternatives available. In terms of HAE, there's no specific preparation other than talking to your immunologist, and getting a management plan for what should happen in pregnancy. If you've been relying on hormonal suppression to help control the symptoms, you will need to come off that to get pregnant, so will need to have a transition plan in place. The plan should also include what to do if you do have an attack in pregnancy, you can use the preventive non hormonal therapies which are normally used.

It's very important to make sure that the obstetrician or midwife or whoever is providing care for you is in contact with the immunologist, and has an understanding of your condition, has collaborative communication and a plan of what to do.

From the pregnancy specific point of view, there's no reason that a patient can't hope for a natural birth. There's no reason that the obstetrician needs to make any changes to the plan and management of the pregnancy because of HAE. If you do require a caesarean, or there's going to be an instrument assisted delivery, you will need to be treated with C1 Inhibitors prophylactically to minimise the chance of an HAE attack triggered by that sort of event.

WHAT HAPPENS AT MENOPAUSE? (VIDEO REF: 9:12)

Some women do describe worsening of symptoms with menopause, presumably because at menopause, there's a significant drop in estrogen levels. So it's not just the estrogen level, it's the relative change in estrogen level, which is a trigger.

We know that by giving estrogen, you can trigger HAE, so unfortunately, symptoms due to menopause, such as hot flashes, vaginal dryness and bladder instability, become more problematic in being treated. If, they do require treatment, there are some non hormonal options available for treatment of menopausal symptoms.

WHAT TREATMENTS ARE AVAILABLE FOR BREAST CANCER? (VIDEO REF: 9:55)

Breast cancer has many treatments. Anyone requiring surgery or radiotherapy will need preventive C1 inhibitor therapy to minimise the chance of triggering an HAE event. Often estrogen sensitive breast cancers are treated with Tamoxifen, which is a weak estrogen that blocks estrogen receptors but can also trigger HAE, so is usually considered not desirable for use in HAE patients.

Fortunately, there are different treatments which have the same effect by blocking another pathway they call the aromatase inhibitors such as Remidex, and they can be used in breast cancer sufferers.

HOW DOES SOMEONE GO ABOUT CHOOSING THE RIGHT GYNAECOLOGIST? (VIDEO REF: 10:51)

The most important thing is to find someone who's willing to collaborate with the immunologist, be in good communication, be able to agree on plans, and also be available. There's no point having someone who you can't talk to until the next appointment in three weeks time. In short, if someone's willing to give you their mobile phone number, they're a good starting point.