

## Sheehan's Syndrome and Lymphocytic Hypophysitis Fact sheet for patients

Sheehan's Syndrome and Lymphocytic Hypophysitis (LH) can present after childbirth, in similar ways. However, in Sheehan's there is a history of profound blood loss and imaging of the pituitary will not show a mass lesion. In Lymphocytic Hypophysitis, there is normal delivery and post-partum, and it can be a month or more after delivery that symptoms start. An MRI in this instance may show a pituitary mass and thickened stalk.

Management of Sheehan's is appropriate replacement of hormones, in LH - replacement hormones and in some circumstances, steroids and surgical biopsy. The key of course is being seen by an endocrinologist with expertise in pituitary and not accepting the overwhelming features of hypopituitarism as just 'normal'. If features of Diabetes Insipidus are present the diagnosis is usually easier, as severe thirst and passing copious amounts of urine will be present.

### Sheehan's syndrome

Sheehan's syndrome is a rare condition in which severe bleeding during childbirth causes damage to the pituitary gland. The damage to pituitary tissue may result in pituitary hormone deficiencies (hypopituitarism), which can mean lifelong hormone replacement.

#### What causes Sheehan's syndrome?

During pregnancy, an increased amount of the hormone oestrogen in the body causes an increase in the size of the pituitary gland and the volume of blood flowing through it. This makes the pituitary gland more vulnerable to damage from loss of blood. If heavy bleeding occurs during or immediately after childbirth, there will be a sudden decrease in the blood supply to the already vulnerable pituitary gland. This can cause tissue death and subsequent loss of pituitary function. Usually only the anterior (front) part of the pituitary gland is affected. The following hormones are released from the anterior pituitary and may be deficient in patients with Sheehan's syndrome:

- Adrenocorticotrophic hormone (stimulates the adrenal glands to secrete steroid hormones, such as cortisol)
- Growth hormone (regulates growth, metabolism and body composition)
- Luteinising hormone and follicle stimulating hormone (also known as the gonadotrophins, these act on the ovaries to stimulate sex hormone production and ovulation)
- Prolactin (stimulates milk production)
- Thyroid stimulating hormone (stimulates the thyroid gland to secrete thyroid hormones)



**Sheehan's Syndrome  
 and Lymphocytic  
 Hypophysitis**  
*awareness*

## What are the symptoms of Sheehan's Syndrome?

In some women, Sheehan's Syndrome may cause few, if any, symptoms. In others, symptoms may be non-specific and may not be diagnosed unless specifically tested for. Most symptoms will not be apparent straight away and may take months or even years to develop. Most commonly, women have difficulty with or are not able to breastfeed (due to lack of prolactin) and have infrequent or no periods after childbirth (due to lack of gonadotrophins).

The patient may also feel tired and experience weight loss or weight gain (due to lack of thyroid hormones), loss of pubic or underarm hair (due to lack of sex hormones) and have low blood pressure which can make them feel light-headed and dizzy (due to lack of adrenocorticotrophic hormone).

## How common is Sheehan's Syndrome?

Sheehan's syndrome is rare in developed countries where improved maternal care usually prevents extreme blood loss during delivery. The condition is still common in developing countries where women may still bleed heavily during childbirth. Sheehan's syndrome is not inherited; it is related exclusively to pregnancy.

Although Sheehan's Syndrome is rare, the results of our recent survey with patients diagnosed, was that awareness and education amongst health care professionals (midwives, GPs, Health Visitors, A&E and endocrinologist in local district hospitals) is vital. This awareness is not only towards appropriate and speedy diagnosis but to actually save lives.

Also mentioned were factors of isolation, distress and the issues of caring for a baby/toddler whilst being unwell, seeking diagnosis and going through testing, which has a huge impact on the mother and her family. The lack of information or how to access the correct information was a feature described by the majority in this survey. Linking up the symptoms displayed and pituitary hormones was not evident to all of our respondents. Improved links and access to online information is important with the mention of Sheehan's being clear, re hypopituitarism.

## How is Sheehan's Syndrome diagnosed?

The diagnosis of Sheehan's Syndrome will partly be based on the patient's medical history - in particular, whether any blood loss during delivery occurred, or any other complications relating to childbirth. It is also important to assess whether there were difficulties with breastfeeding or a lack of periods after delivery, which are two important signs of Sheehan's syndrome. The first step is to see your GP, discuss your symptoms and ask for basic pituitary function blood tests. A referral to an endocrinologist at a specialist centre is recommended for further testing if basic bloods taken are not in normal range.

Your history and blood tests would be repeated with an endocrinologist to check pituitary hormone levels. A simple blood test may be sufficient or a more detailed type of testing called 'dynamic' testing may be carried out. This type of testing involves checking hormone levels before and after stimulation of the pituitary gland to assess how the pituitary is functioning. Scans such as computerised tomography (CT) or magnetic resonance imaging (MRI) may also be carried out to investigate the size and structure of the pituitary gland.

## How is Sheehan's Syndrome treated?

Once the diagnosis of Sheehan's Syndrome is confirmed, the treatment is lifelong replacement of the hormones that are found to be deficient. This may take the form of hydrocortisone, thyroxine, oestrogen and rarely, growth hormone. The exact form of treatment will vary between patients depending on the hormones that need to be replaced. *See our booklet called **The Pituitary Gland: Its conditions and hormones explained.***

### Are there any side-effects to the treatment?

If blood levels of hormones are monitored carefully, there should not be any side-effects. Side-effects may rarely occur when the dosage of hormone treatment is too high or too low and should be easily corrected by dose adjustment of the relevant medication. Patients should discuss any concerns with their endocrinologist or GP.

### What are the longer-term implications of Sheehan's Syndrome?

Hormone treatment is usually long-term and patients will require regular check-ups with their endocrinologist or GP. If the patient is on steroids, they must be aware of sick-day rules and have an emergency hydrocortisone injection, which they know how to use. They should also wear Medic-Alert jewellery stating that they are on steroid medication. See our **Hydrocortisone advice leaflet for patients**.

### Lymphocytic Hypophysitis (LH)

This is a condition in which lymphocytes (type of white blood cell that is part of the immune system) enter the pituitary gland, resulting in pituitary enlargement and impaired function. It most often occurs in women in late pregnancy or the postpartum period, but can also occur in pre-pubertal or post-menopausal women, and in men. Symptoms of LH may include headache, visual field impairment and more rarely, double vision (diplopia). The exact cause is unknown but is thought to be autoimmune-related. Although some cases resolve on their own or after a short course of steroids, other cases cause persistent problems even with aggressive medical or surgical treatment.

### What are the symptoms of LH?

Individuals affected with Lymphocytic Hypophysitis typically have headaches as their first symptom. This usually precedes or occurs with visual field impairment; rarely, double vision may be present. Hyperprolactinemia affects approximately one third of individuals, causing amenorrhea (absence of menstruation) or galactorrhea (production of breast milk) in women and sexual dysfunction in men.

Other features that occur more rarely and are related to alterations in pituitary secreted hormones may include:

- Hypo-prolactinaemia (deficiency of prolactin)
- Subclinical hypopituitarism (decreased production of pituitary hormones with no symptoms)
- ACTH deficiency
- Acute hypoadrenalism
- Hypogonadotropic hypogonadism (usually diagnosed only in males)
- Isolated growth hormone (GH) deficiency

### What causes LH?

The exact cause of Lymphocytic Hypophysitis (LH) remains unclear. It has been suggested that LH has an autoimmune cause. This has generally been supported by the frequent postpartum occurrence and the lymphocytic infiltration, as well as several laboratory and clinical findings.

### Diagnosis

If you think you are affected by LH, see your GP, taking a list of your symptoms, when they started and how they affect you. Asking you about the symptoms, their relation with a pregnancy and, if you have any history of other autoimmune diseases, the GP can order blood tests. The specialist

recommended for referral by your GP, would be an endocrinologist experienced in pituitary (within a centre of excellence). Further tests and scans would be done to help confirm diagnosis but as below, in some cases surgery may be carried out for a diagnosis and treatment.

### How is LH treated?

Lymphocytic Hypophysitis can be different among affected individuals, so different treatments may be required. Some individuals experience spontaneous remission (improvement or reversal of the condition without any formal treatment). Careful follow-up is typically recommended.

Many affected individuals only have headaches as an apparent symptom, and there have been multiple reports of these individuals improving with steroids alone. Although the use of steroids or other anti-inflammatory drugs have been suggested as medical treatment, their long-term efficacy still needs to be confirmed. High-dose methylprednisolone pulse therapy (short, intensive administration given at set intervals) seems to be effective in about 30% of treated patients.

Transsphenoidal surgery (accessed through the nose) to confirm diagnosis and save viable pituitary tissue may be required in individuals with symptoms or signs of severe compression. Visual improvement following decompression with this type of surgery has been reported. In some cases, pituitary biopsy is both diagnostic and therapeutic, because after this procedure a progressive recovery of pituitary function may occur.

After treatment, you will need to be followed-up. The timing can vary by individual case. For example, in those patients with a complete (anatomical and functional) recovery it is necessary for only a periodical endocrine consultation. If the recovery is partial, the patient needs to have regular check-ups to monitor pituitary function (by hormone blood test measurements for example) and the pituitary gland (by MRI imaging or optician examination). In some patients, especially in those with a history of autoimmune diseases, we suggest also to perform a general autoimmune evaluation, checking the most common markers of autoimmune disease.

.....

### Further support and information:

[www.pituitary.org.uk](http://www.pituitary.org.uk)

See specific webpages for this awareness campaign and read patient stories by visiting: <https://pituitary.org.uk/get-involved/campaigns-and-raising-awareness/current-campaigns/>

Our Support & Information Helpline - 0117 370 1320 Mondays to Fridays 10am to 4pm

Our Endocrine Nurse Helpline - 0117 370 1317 Mondays 10am to 1pm & 6pm to 9pm, Thursdays 9am to 1pm



**Sheehan's Syndrome  
and Lymphocytic  
Hypophysitis**  
*awareness*

The Pituitary Foundation  
Registered charity number 1058968