ACQUIRED PITUITARY DEFICIENCY OR ACQUIRED HYPOPITUITARISM

1. What are the pituitary and the hypothalamus?

The pituitary is a small endocrine gland that looks like a grape, is connected to the hypothalamus that resides right above it, with the pituitary stalk and secretes many hormones that orchestrate body's function and homeostasis. The anterior lobe of the pituitary secretes growth hormone (GH) that stimulates growth, the thyroid stimulating hormone (TSH) controlling thyroid function, the gonadotrophins (LH and FSH) that orchestrate puberty, controlling ovaries and testes respectively, adrenocorticotropic hormone (ACTH), that controls the adrenals and cortisol production and prolactin. The posterior lobe secretes the antidiuretic hormone (ADH) that controls body's water balance and oxytocin that has an important role during delivery and breastfeeding. The hypothalamus acts like the computer's processor and plays an important role in vital functions of the body like thirst and appetite, and controls hormone secretion from the pituitary gland. The pituitary gland is often called the master gland because it controls several other hormone glands in the body, including the thyroid and adrenals, the ovaries and testicles.

2. What is pituitary deficiency?

Pituitary deficiency or hypopituitarism is the state when pituitary cannot produce and secrete one or more of the pituitary hormones. It can be congenital or acquired, partial or complete.

3. What can cause acquired pituitary deficiency?

- Tumors of the pituitary and hypothalamic region.
- Irradiation of the pituitary and hypothalamic region (i.e. radiotherapy of the central nervous system (CNS) tumors)
- Brain trauma or hemorrhage (after neurosurgery, traumatic brain injury, vascular problems)
- CNS Infections (i.e. meningitis)
- Infiltrative diseases of CNS (Langerhans cell histiocytosis, haemosiderosis in transfused thalassemic patients)

4. How frequent is acquired pituitary deficiency?

The frequency of acquired pituitary deficiency depends on the frequency of the disease that is the underlying cause, as it is always secondary (see etiology). The majority of the children that are cancer survivors (especially those who had tumors of the hypothalamic-pituitary region) suffer from acquired hypopituitarism as a result of neurosurgical operations and cranial irradiation.

5. What are the symptoms of acquired pituitary deficiency?

Since acquired pituitary deficiency is secondary to another disorder, the symptoms are related to the primary disease (for example optic disturbances secondary to tumors of the hypothalamic-pituitary region). Furthermore, symptoms depend on the affected hormonal axis and may include the following:

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- Growth hormone deficiency: Growth failure, short stature, hypoglycaemia, relative obesity
- Secondary hypothyroidism (TSH deficiency): constipation, poor growth, hair loss, weight gain)
- Secondary adrenal deficiency (ACTH deficiency): ACTH deficiency can lead to secondary adrenal insufficiency, which means insufficient production of cortisol from the adrenal glands. Patients may have symptoms such as: fatigue, episodes of vomiting and hypoglycaemia. Cortisol is the hormone responsible for the body's response to stress, hence when it is deficient there is a risk of the body collapsing during relatively common situations such as an infection or trauma. The condition is very serious, it is called adrenal crisis and needs immediate medical treatment.
- LH and FSH deficiency: pubertal failure, infertility
- Central diabetes insipidus (ADH deficiency): Polyuria, polydipsia, dehydration.

6. How is the diagnosis made?

There is usually a strong clinical suspicion based upon the past medical history supported by the findings of clinical examination. The diagnosis is made after thorough endocrinological workup either with single measurements of hormones or after specific stimulation tests. In most cases imaging of the brain or especially the hypothalamic and pituitary region is also required (i.e. with magnetic resonance imaging, MRI)

7. How is acquired pituitary deficiency treated?

After the initial management of the primary disease, hormone replacement treatment is commenced. The exact medication given depends on the specific pituitary deficiency. Additionally, fertility issues are discussed and fertility reservation may be planned, as many of the agents used to treat oncology patients are gonadotoxic (affect irreversibly the function of testes and ovaries). This issue is being intensively studied over the last years, since we have an increasing number of cancer survivors after successful oncologic treatment.

8. What kind of medications is used for hormone replacement?

Hormone replacement medications may include:

- **Corticosteroids:** These drugs, such as hydrocortisone or prednisone replace the adrenal hormones that aren't being produced because of the adrenocorticotropic hormone (ACTH) deficiency. They are taken by the mouth.
- **Levothyroxine:** This medication treats the low thyroid hormone levels (hypothyroidism) that a thyroid-stimulating hormone (TSH) deficiency can cause.
- **Sex hormones:** These include testosterone in men and estrogens or a combination of estrogen and progesterone in women. Testosterone is administered either by injection or through the skin with a gel. Female hormone replacement can be administered with pills, gels or patches.

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- **Growth hormone:** Also called somatotropin, growth hormone is administered through daily injections. It promotes growth, which helps achieve a closer to normal height in children. Adults with symptoms of growth hormone deficiency also may benefit from growth hormone replacement, but they won't grow taller.

9. How are patients followed after diagnosis?

After the initial diagnostic workup and management, there must be a frequent follow-up period from a multidisciplinary team consisting of a Pediatric Endocrinologist, a Pediatric Oncologist, a Neurosurgeon and a Pediatrician. The team is carefully monitoring growth, progression of puberty and medical treatment until the care is transitioned to a similar adult team. For patients with hypopituitarism, it is important to carry with them a special card notifying others of their condition (for example, in emergency situations). This is especially important if they are taking corticosteroids for an ACTH deficiency.

10. Useful links for parents

https://www.pituitary.org.uk/