



# Empty Sella Syndrome

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## Synonyms of Empty Sella Syndrome

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- empty sella turcica
- ESS

## Subdivisions of Empty Sella Syndrome

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- primary empty sella syndrome
- secondary empty sella syndrome

## General Discussion

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Empty sella syndrome is a rare disorder characterized by enlargement or malformation of a structure in the skull known as the sella turcica. The sella turcica is a saddle-shaped depression located in the bone at the base of skull (sphenoid bone), in which resides the pituitary gland. In empty sella syndrome, the sella turcica is either partially filled with cerebrospinal fluid and a very small associated pituitary gland lying in the floor of the sella (partially empty sella) or completely filled with cerebrospinal fluid with no visualized pituitary gland (completely empty sella). Most individuals with empty sella syndrome do not have any associated symptoms, but the finding raises concerns about hormone deficiencies. Empty sella syndrome may occur as a primary disorder, for which the cause is unknown (idiopathic), or as a secondary disorder, in which it occurs due to an underlying condition or disorder such as a treated pituitary tumor, head trauma, or a condition known as idiopathic intracranial hypertension (also called pseudotumor cerebri) during which elevated intracranial pressure causes empty sella syndrome.

## Signs & Symptoms

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The symptoms of empty sella syndrome may vary from one person to another and depends on the underlying cause. In most cases, especially in individuals with primary empty sella syndrome, there are no associated symptoms (asymptomatic). Often, empty sella syndrome is discovered incidentally on CT or MRI examination when individuals are being evaluated for other reasons.

The most common symptom potentially associated with empty sella syndrome is chronic headaches. However, it is unknown whether headaches develop because of empty sella syndrome or are simply a coincidental finding. Many individuals with empty sella syndrome have high blood

pressure (hypertension), which can itself cause headaches if severe.

In rare cases, individuals with empty sella syndrome have developed increased pressure within the skull (benign intracranial pressure), leakage of cerebrospinal fluid from the nose (cerebrospinal rhinorrhea), swelling of the optic disc due to increased cranial pressure (papilledema), and abnormalities affecting vision such as loss of clarity of vision (visual acuity).

In the empty sella syndrome, the function of the pituitary gland is usually not affected. It is often not well seen on imaging, but is otherwise perfectly functional. The pituitary is a small gland located near the base of the skull that stores several critical hormones and releases them into the bloodstream as needed by the body. These hormones regulate many different bodily functions. Although a rare occurrence, some abnormal or decreased pituitary function can occur (hypopituitarism) in the setting of empty sella. A specific finding in some individuals with empty sella, including children, has been isolated growth hormone deficiency.

Individuals with secondary empty sella syndrome are more likely to develop abnormalities affecting vision and decreased function of the pituitary because the underlying cause of their empty sella (e.g. treated pituitary tumor or trauma) result in these other associated problems.

## Causes

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The exact, underlying cause of primary empty sella syndrome is unknown (idiopathic).

Researchers believe that a defect in the diaphragma sellae that is present at birth (congenital defect) plays a role in the development of primary empty sella syndrome. The diaphragma sellae is a fold of dura mater (the outermost layer of the membranes that line the brain and spinal cord). The diaphragma sellae covers the sphenoid bone where the sella turcica and the pituitary are located. In some affected individuals a tear in the diaphragma sellae allows the underlying membranes to push through (herniate), which allows cerebrospinal fluid to leak out and accumulate in the sella turcica. The pressure exerted by the fluid can flatten or enlarge the sella turcica. Consequently, the pituitary becomes compressed and flattened as well. In some individuals with primary empty sella syndrome the diaphragma sellae is absent at birth. The exact role that defects in the diaphragma sella play in the development of primary empty sella syndrome is unknown. Whether it causes primary empty sella syndrome directly, occurs as part of a larger disease process or is only a predisposing factor to the development of the disorder is unresolved.

Secondary empty sella syndrome is caused by a variety of different conditions including injury or trauma to the head, treated pituitary tumors, infection, radiation therapy, surgery on the pituitary region, or rare disorders such as Sheehan syndrome.

## Affected Populations

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Primary empty sella syndrome affects approximately 4 times more women than men. Most cases occur in middle-aged women who are obese and have high blood pressure (hypertension). Because most people with empty sella syndrome do not have symptoms and may go undiagnosed, determining the disorder's true frequency in the general population is difficult. Some researchers have estimated that less than 1 percent of individuals with empty sella syndrome ultimately develop symptoms associated with the disorder, although this may be higher in men compared to women.

## Diagnosis

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A diagnosis of empty sella syndrome is made based upon identification of characteristic symptoms, a detailed patient history, a thorough clinical evaluation and specialized imaging techniques. Imaging may include computerized tomography (CT) scanning and magnetic resonance imaging (MRI). During CT scanning, a computer and x-rays are used to create a film showing cross-sectional images of certain tissue structures. An MRI uses a magnetic field and radio waves to produce cross-sectional images of particular organs, tissues and structures such as the sella turcica.

## Standard Therapies

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### Treatment

Most individuals with empty sella syndrome do not have any symptoms and do not require treatment. When symptoms do occur, treatment is directed toward the specific symptoms that are apparent in each individual. If the pituitary is affected, then replacement therapy for specific hormones should be administered as needed. Surgery may be necessary when cerebrospinal fluid leaks from the nose (CSF rhinorrhea).

## Investigational Therapies

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Information on current clinical trials is posted on the Internet at [www.clinicaltrials.gov](http://www.clinicaltrials.gov). All studies receiving U.S. government funding, and some supported by private industry, are posted on this government web site.

For information about clinical trials being conducted at the NIH Clinical Center in Bethesda, MD, contact the NIH Patient Recruitment Office:

Toll-free: (800) 411-1222

TTY: (866) 411-1010

Email: [prpl@cc.nih.gov](mailto:prpl@cc.nih.gov)

Some current clinical trials also are posted on the following page on the NORD website:

<https://rarediseases.org/for-patients-and-families/information-resources/news-patient-recruitment/>

For information about clinical trials sponsored by private sources, in the main, contact:

[www.centerwatch.com](http://www.centerwatch.com)

For more information about clinical trials conducted in Europe, contact:

<https://www.clinicaltrialsregister.eu/>

## NORD Member Organizations

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- [Pituitary Network Association](#)
  - P.O. Box 1958
  - Thousand Oaks, CA 91358 USA
  - Phone: (805) 499-9973
  - Email: [info@pituitary.org](mailto:info@pituitary.org)
  - Website: <http://www.pituitary.org>

## Other Organizations

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- Genetic and Rare Diseases (GARD) Information Center
    - PO Box 8126
    - Gaithersburg, MD 20898-8126
    - Phone: (301) 251-4925
    - Toll-free: (888) 205-2311
    - Website: <http://rarediseases.info.nih.gov/GARD/>
  - NIH/National Institute of Neurological Disorders and Stroke
    - P.O. Box 5801
    - Bethesda, MD 20824
    - Phone: (301) 496-5751
    - Toll-free: (800) 352-9424
    - Website: <http://www.ninds.nih.gov/>

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**INTERNET**

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