

## Achalasia mimicking large paraesophageal hiatal hernia

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**Section:** Chest imaging

**Area of Interest:** Gastrointestinal tract Oesophagus

**Procedure:** Contrast agent-oral

**Procedure:** Dynamic swallowing studies

**Procedure:** Barium meal

**Procedure:** Contrast agent-other

**Imaging Technique:** Conventional radiography

**Imaging Technique:** Fluoroscopy

**Imaging Technique:** CT

**Special Focus:** Dilatation Motility Swallowing disorders

Case Type: Clinical Cases

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**Patient:** 67 years, male

### Clinical History:

An outpatient was referred for an oesophagram (barium swallow) because of worsening symptoms including reflux, daily vomiting, meal-related pain, and weight loss. Relevant medical history with onset ten years ago included reflux and aspiration pneumonias. Dysphagia had only recently been described in his medical chart.

### Imaging Findings:

The oesophagram shows a straight, centrally located and moderately dilated oesophagus with tertiary contractions (Fig. 1-3). There is unimpeded contrast passage into a spherical intrathoracic cavity with an air/fluid/contrast level (Fig. 2-5), located above the diaphragm to the right. Contrast moves swiftly to the intra-abdominally located ventricle (Fig. 2-4). The measurements of the cavity are stationary compared to the only previous oesophagram five years ago. Due to slow passage from ventricle to duodenum, an image was recorded 1 hour post contrast (HPC, Fig. 6). This shows contrast retention within the intrathoracic cavity, and normal passage from ventricle to duodenum. The description concluded with para-oesophageal hiatal hernia with reduced passage to the ventricle, severe passive reflux in recumbent position (Fig. 7), and tertiary oesophageal contractions.

CT-examinations and chest X-rays (Fig. 8-10) also concluded with hiatal hernia. Manometry showed mixed peristalsis, and gastroscopy at symptom onset found undigested food in an intrathoracic cavity.

### Discussion:

Based on symptoms and imaging findings, the patient was referred to subacute laparoscopic operation for para-oesophageal hernia. Surprisingly, during surgery the entire ventricle was found intra-abdominally, thus refuting the diagnosis. Instead, the lower oesophageal sphincter (LES) was found severely stenotic in the midst of the cavity. A diverticulum was considered, but disproved by a simultaneously performed gastroscopy. Hence, the diagnosis of achalasia was reached perioperatively. A thorough read through the patient chart revealed that biopsies were taken from abnormal mucosa in what was believed to be a para-oesophageal hiatal hernia during onset of symptoms. The biopsies consisted of oesophageal mucosal and muscular layers, and specified the absence of cylindrical epithelium

otherwise expected to be found in ventricular mucosa.

Primary achalasia is a progressive motility disorder of the oesophagus, characterised by abnormal or absent peristalsis, and insufficient relaxation of the LES due to a loss of inhibitory innervation of smooth muscle [1]. The incidence is 1 per 100 000, and the prevalence is 10 per 100 000 [2]. The aetiology remains unknown, although auto-antibodies have been detected which supports the hypothesis of an autoimmune component [3].

Clinically, the cardinal symptom of achalasia is dysphagia for solids and liquids. It also presents with regurgitation or vomiting of undigested food, and occasionally chest discomfort and weight loss. Symptoms are initially mild and intermittent, but progress in severity and frequency [1, 4]. Complications include aspiration pneumonia and the feared albeit rare oesophageal squamous cell carcinoma. The latter has a low absolute risk with a yearly incidence rate of 0.34%, but a high relative risk with a hazard ratio of 28 [5].

Manometry is the golden standard among diagnostic tests, and is used to detect increased pressure over LES. It usually relies on support from an oesophagram where typical imaging findings include "bird beak"-configuration of the distal oesophagus, dilated oesophagus, tertiary contractions and failure of normal peristalsis in supine position, resulting in contrast retention. Gastroscopy can show a dilated oesophagus with retention of content [1, 4].

There is no cure for achalasia, only symptomatic treatment which often needs to be repeated [1]. This patient awaits a laparoscopic Heller myotomy. Meanwhile, botox injections give immediate but temporary relief of symptoms [4].

Clinical signs of achalasia can overlap with reflux and hiatal hernia [1]. The purpose of this clinical case is to focus on the atypical presentations of achalasia on oesophagrams. Food regurgitation, dilated oesophagus with contrast retention and tertiary contractions should always lead us to consider achalasia.

**Differential Diagnosis List:** Achalasia, Hiatal hernia, Epiphrenic diverticulum, Oesophageal cancer, Ventricular cancer

**Final Diagnosis:** Achalasia

#### **References:**

- Vaezi MF (2016) Achalasia: from diagnosis to management. Annals of the New York Academy of Sciences (PMID: [27571581](#))
- Sadowski DC (2010) Achalasia: incidence, prevalence and survival. A population-based study. Neurogastroenterology and motility (PMID: [20465592](#))
- Furuzawa-Carballeda J (2016) New insights into the pathophysiology of achalasia and implications for future treatment. World Journal of Gastroenterology (PMID: [27672286](#))
- Stavropoulos SN (2016) Diagnosis and management of esophageal achalasia. BMJ (PMID: [27625387](#))
- Leeuwenburgh I (2010) Long-term esophageal cancer risk in patients with primary achalasia: a prospective study. The American Journal of Gastroenterology (PMID: [20588263](#))

**Figure 1**

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**Description:** Supine position, AP view, 1 HPC. **Origin:** Department of Radiology, Hvidovre University Hospital, Denmark.

**Figure 2**

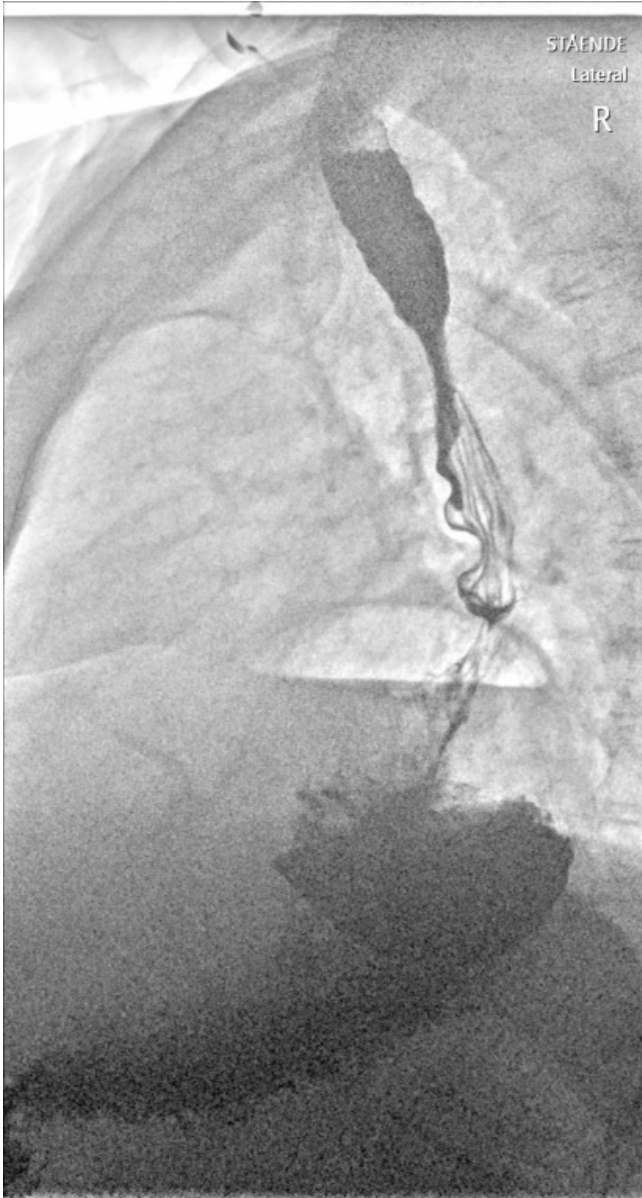
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**Description:** Right recumbent position, lateral view, 10 MPC. **Origin:** Department of Radiology, Hvidovre University Hospital, Denmark.

**Figure 3**

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**Description:** Standing position, lateral view. **Origin:** Department of Radiology, Hvidovre University Hospital, Denmark.

**Figure 4**

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**Description:** Standing position, AP view. **Origin:** Department of Radiology, Hvidovre University Hospital, Denmark.

**Figure 5**

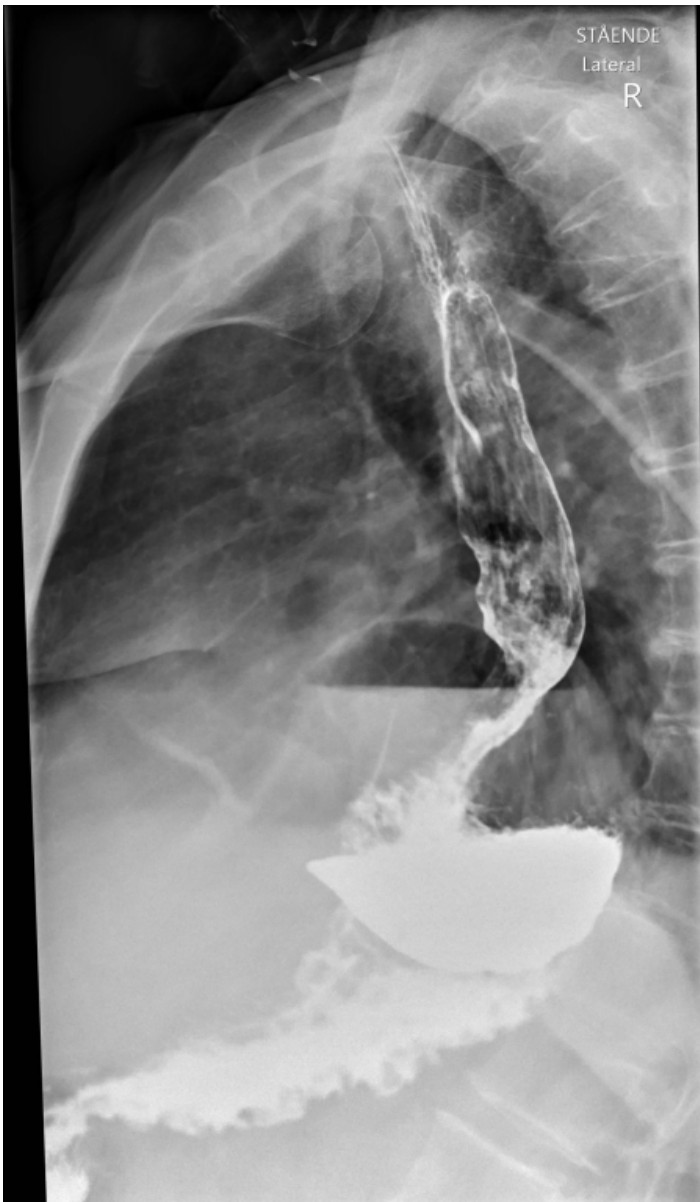
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**Description:** Standing position, AP view, 1 minute past contrast (MPC). **Origin:** Department of Radiology, Hvidovre University Hospital, Denmark.

**Figure 6**

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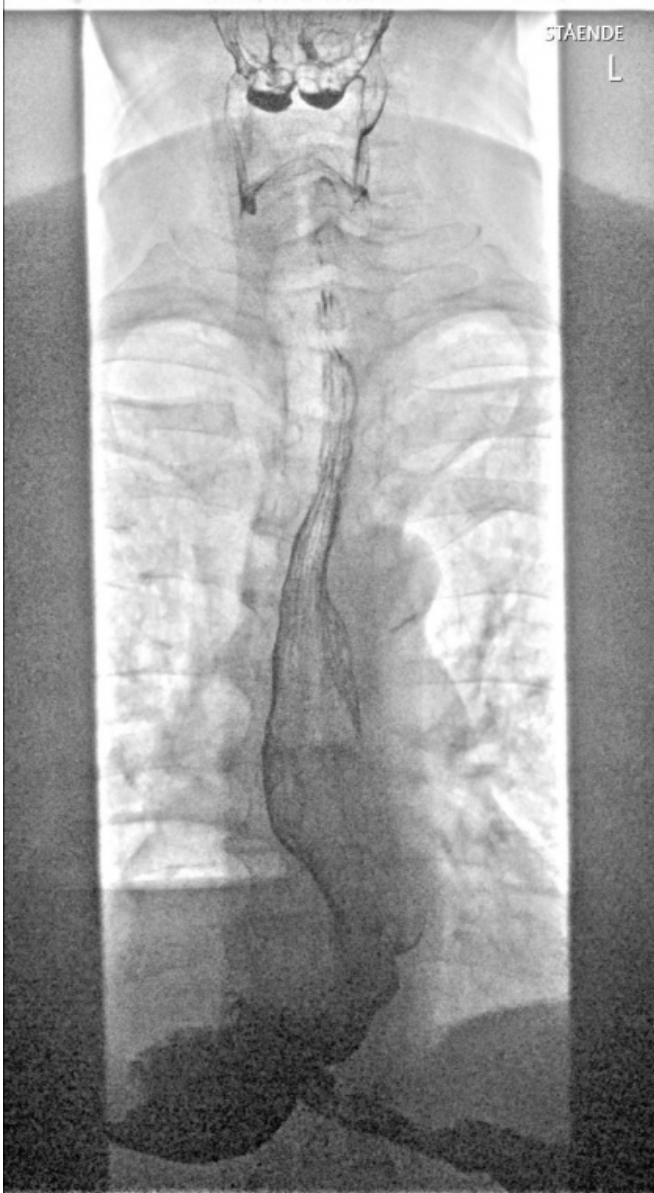


**Description:** Standing position, lateral view, 2 MPC. **Origin:** Department of Radiology, Hvidovre University Hospital, Denmark.



**Figure 7**

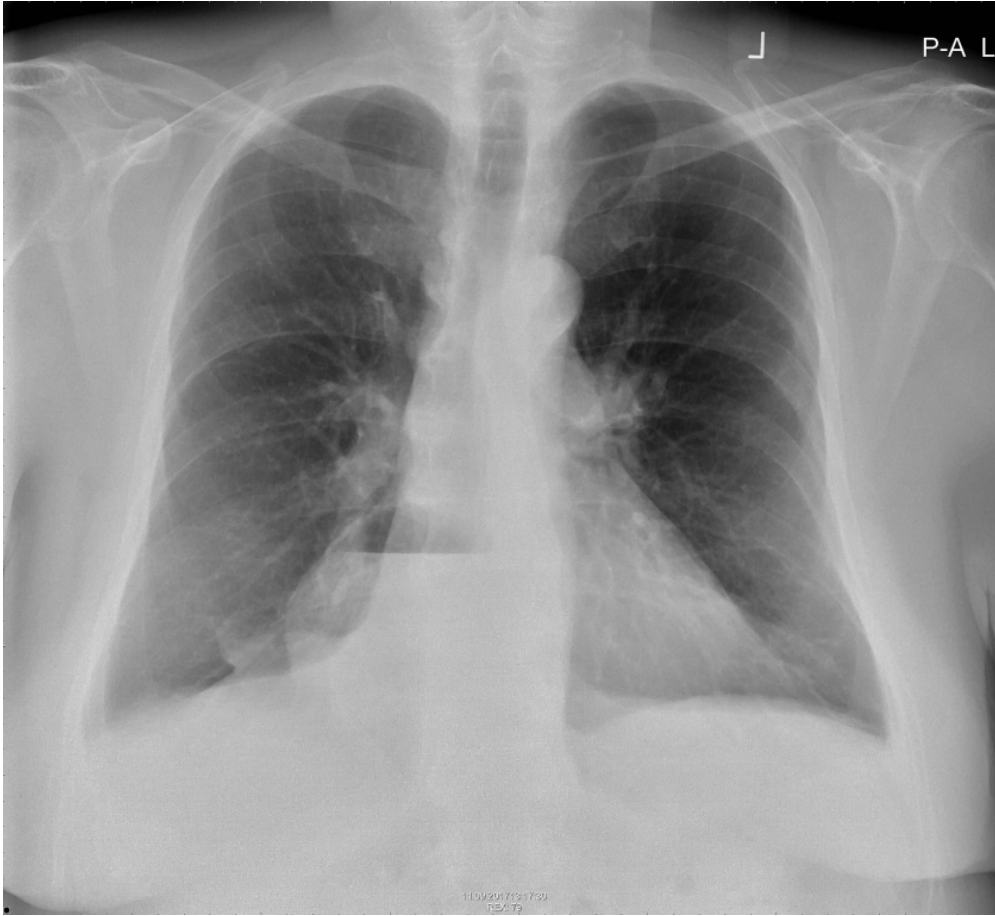
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**Description:** Standing position, AP view. **Origin:** Department of Radiology, Hvidovre University Hospital, Denmark.

**Figure 8**

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**Description:** Standing position, AP view, showing intrathoracic cavity with air/fluid level, interpreted as hiatal hernia. **Origin:** Department of Radiology, Hvidovre University Hospital, Denmark.

**Figure 9**

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**Description:** Dilated oesophagus (arrows) and intrathoracic cavity to the right. **Origin:** Department of Radiology, Hvidovre University Hospital, Denmark.

**Figure 10**

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**Description:** Intrathoracic cavity (arrows), believed to be hiatal hernia, but proved to be dilated oesophagus due to achalasia. **Origin:** Department of Radiology, Hvidovre University Hospital, Denmark.