Achalasia

Basics

Achalasia is a functional disorder of the esophagus with impaired relaxation of the lower esophageal sphincter and aperistalsis (=absence of motility) of the esophageal body. As a consequence the transport of food is incomplete or absent. In addition the lower esophageal sphincter (LES) fails to open for emptying the food into the stomach (greek achalasein = fails to relax). A functional outflow obstruction occurs. Due to the inability to eat and swallow the patients loose weight. Achalasia is a rare condition and occurs in 0.5-1.0 cases per 100.000 persons a year.

Causes

Swallowing and esophageal transport depend upon the outbalanced function of the nerves and muscles. Achalasia patients have an impaired function of the nerves responsible for the coordinated activation of transport and the relaxation of the lower esophageal sphincter (LES). As a consequence the esophagus looses its ability for adequate transport. Due to the lack of swallow-induced relaxation of the lower esophageal sphincter (LES), the food gets stuck in the esophagus. The cause for the destruction of the esophageal nerves is not known. In south America achalasia associates with an infectious disease (Chaga’s disease, caused by Trypanosoma cruzi). The infection destroys the nerves responsible for esophageal motility. Otherwise the cause for achalasia is suggested to represent an auto immune reaction against the esophageal nerves.

Symptoms

Symptoms of functional outflow obstruction define achalasia: difficulty at swallowing (dysphagia), heartburn and chest pain. Dysphagia starts for liquids and overtime also affects solid food. As a consequence of the eating disorder the patients loose weight (5-10 kg per month). Sometimes stuck food can be provoked to reach the stomach by drinking water or using the power of gravity. In some cases jumping helps to bring the food down into the stomach.

Diagnosis and Tests

Diagnosis of achalasia is assessed by esophageal manometry. The test reveals the absence of coordinated motility (contractions of the esophagus) and impaired or absent relaxation of the lower esophageal sphincter: normally the pressure of the lower esophageal sphincter decreases upon swallowing. In achalasia the pressure does not decrease or only slightly decreases upon a swallow. The impedance part of the manometry records the absence of esophageal transport. Endoscopy shows a dilated esophagus without any movement and signs of motility (looks like a sac). High-resolution manometry lists 3 types of achalasia: types I, II and III. All types show a high-pressure band at the level of the lower esophageal sphincter. The pressure does not adequately decrease upon swallowing. In addition to that type I achalasia shows absence of any motility and contraction within the body of the esophagus. Type II shows simultaneous contractions. Type III reveals a simultaneous high-pressure spasm.

Treatment
Treatment of achalasia includes endoscopic Botox injection, endoscopic balloon dilatation and laparoscopic myotomy (+anti reflux surgery; Dor fundoplication). The treatment of achalasia is not causative (since the cause is not known). The therapy aims to eliminate the functional outflow obstruction of the esophagus.

Botox injection takes 10 min and blocks the spasm of the lower esophageal sphincter. The effect lasts for 3 months, consequently the therapy has to be repeated.

During balloon dilatation the surgeon places a balloon (filled with air or water) at the level of the lower esophageal sphincter. Under endoscopic vision the balloon is inflated. Thus balloon dilatation disrupts the lower esophageal sphincter and eliminates the outflow obstruction. Prognosis: 30% success after 1 dilatation, 60% after 2-3 dilatations, the remaining patients should be offered laparoscopic myotomy. Complication: perforation in 4% of the cases.

During laparoscopic myotomy the surgeon dissects the muscle fibers of the lower esophageal sphincter. In addition the operation creates an anterior anti reflux fundoplication (Dor fundoplication) to prevent postoperative reflux. The operation is minimally invasive and lasts 60 min. Prognosis: in experienced hands dysphagia is eliminated in > 80% of the cases after 5-10 years. Complication: perforation in less than 4% of the cases.

Comparison of balloon dilatation vs. laparoscopic myotomy: Both treatments are equally effective. However up to 4 dilatations are required to obtain an effect comparable to surgery. Dilatation should be offered to patients > 70 years of age and comorbidities. Myotomy should be considered for patients with high pressure of the lower esophageal sphincter (> 20mm Hg), high swallow induced residual pressure of the lower esophageal sphincter (> 10 mm Hg), absence of motility (dilated and curbed esophagus), younger age (During the course of the disease less than 5% of the patients develop end stage achalasia with complete inability to eat. In these rare cases we have to perform the removal of the esophagus (esophagectomy).

Advantages of surgery: the surgeon dissects the muscle fibers under the laparoscopic vision. Dilatation is mainly “blind” and therefore harbors insecurities including perforation and ineffective disruption of the lower esophageal sphincter.

Outlook: The endo flip test measures the distensibility and enables us to immediately assess the effect of dilatation and myotomy. Currently studies examine the feasibility and clinical relevance of this novel approach for the treatment of achalasia.
Heller's myotomy for achalasia: the operation dissects the lower esophageal sphincter to eliminate the outflow obstruction. Anti reflux surgery (Dor fundoplication) is performed to prevent reflux.

**Prevention**

One cannot prevent the development of achalasia.

**Self test**

Swallowing difficulties may indicate the development of achalasia. We recommend gastroscopy and esophageal manometry for adequate diagnosis of achalasia. Differential diagnoses include esophageal inflammation (eosinophil esophagitis), rings, webs, strictures and cancer.

**Expert opinion**

**Johannes Lenglinger (Physiologist, Vienna):**

The diagnosis of achalasia includes gastroscopy and esophageal manometry. Gastroscopy aims to exclude other causes for dysphagia: rings, webs, stenosis, structures or even cancer. Esophageal manometry is the gold standard for establishing the diagnosis of achalasia. Modern high-resolution manometry lists 3 types of achalasia: type I, type II and type III. Type I shows absence of any motility in the esophagus and a high-pressure band at the level of the lower esophageal sphincter. Type II reveals the non-relaxing lower esophageal sphincter and so-called simultaneous contractions during swallowing. A simultaneous high-pressure spam defines Type III achalasia. This type frequently causes chest pain and heartburn.

**Martin Riegler (Surgeon, Vienna):**

Treatment of achalasia does not eliminate the cause of the disease, which is simply not known. However, we can improve the life quality of the patient by decreasing the outflow obstruction. This occurs by Botox injection, balloon dilatation and laparoscopic myotomy (=dissection of the muscle of the lower esophageal sphincter). Patients with severe comorbidities should go for Botox: that improves dysphagia for at least 3 months. Young patients with high lower esophageal sphincter pressure should go for myotomy. All others could be offered either surgery or dilatation. Basically both options are equally effective to improve the dysphagia. Personally I prefer surgery due to the following reasons: you clearly see what you dissect, you see important anatomical layer: the muscle and the mucosa. This contributes to avoid perforation. And you add the anti reflux surgery (Dor fundoplication) against reflux. Basically achalasia types I and II are good candidates for surgery, type III achalasia does not adequately respond to any treatment.

**Sebastian Schoppmann (Surgeon, Vienna):**

POEM (peroral endoscopic myotomy) is a fascinating, new method for the treatment of achalasia. Under endoscopic vision the surgeon perforates the most inner layer (mucosa) of the esophagus. Via this route one reaches the muscle layer and dissects the muscle down to the lower end of the esophagus. Initial data are promising, but outcome data of these studies are to be awaited. Currently POEM cannot be recommended for routine therapy of achalasia.

**Literature**