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Achalasia Esophageal: A Narrative Literature Review

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A B S T R A C T

Esophageal achalasia is a primary esophageal motility disorder characterized by loss of esophageal peristalsis and partial or no relaxation of the lower esophageal sphincter (LES) in response to swallowing. As a result, there is a functional obstruction at the level of the gastroesophageal junction, resulting in impaired esophageal emptying. Most of the patients experienced severe dysphagia, regurgitation, and weight loss, which affected their quality of life. Achalasia is a rare disease with an estimated prevalence of 1 in 100,000 individuals. However, recent data suggest that prevalence is at least two to three times greater than previously estimated. Achalasia occurs in men and women with equal frequency, with a peak in the 30th and 60th decades of age. The etiology of esophageal achalasia is unknown, and treatment is focused on the palliation of symptoms by eliminating resistance at the level of the gastroesophageal junction.

1. Introduction

Achalasia is an esophageal motility disorder of unknown cause and occurs at all ages with symptoms, especially difficulty swallowing solid/liquid food and regurgitation. The term achalasia means "failure to relax" and refers to the inability of *the lower esophageal sphincter* (the ring of muscle between the lower esophagus and stomach) to open and let food pass into the stomach. Failure to relax in the swallowing process causes dilatation of the proximal part of the esophagus without peristalsis. People with achalasia feel the need to push or force their food down with water or drinks to complete the swallowing process. Other symptoms may include a feeling of substernal fullness, and regurgitation is common.[1-3]

Achalasia began to be recognized by Thomas Willis in 1672. At first, it was suspected that the cause was a blockage in the distal esophagus, so he dilated with whale bones and pushed food into the stomach. In 1908 Henry Plummer performed dilation with a balloon catheter. In 1913 Heller performed surgery by way of cardiomyotomy outside the mucosa, which continues to be embraced today. However, the cause of achalasia is still not known with certainty. Theories for the cause of achalasia began to emerge, such as a process involving infection, disorder, or inherited (genetic), the immune system causing the body itself to damage the esophagus (autoimmune disease), and the aging process (degenerative process).[4-6]

Achalasia is rare. The incidence of achalasia is around 1-10:100,000 population with the same distribution of males and females, which is 1:1. There is no predilection based on race. Achalasia occurs at

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all ages, with incidence from birth to 7-8 decades and a peak incidence at 30-60 years of age. In the United States, approximately 10 cases of achalasia are found in a year. Although this disease is rare, we must be able to recognize and diagnose this disease because the complications caused by this disease are very life-threatening, such as respiratory tract obstruction to sudden death. Therefore, it is very important for us to know the diagnosis of esophageal achalasia. The diagnosis of esophageal achalasia was based on clinical symptoms, radiological features, esophagoscopy, and manometry.[7-10]

Esophageal achalasia

Esophageal achalasia is also known as cardiospasm ectasia, megaesophagus, or idiopathic esophageal dilatation, is a neuromuscular disorder. Achalasia is characterized by the absence of esophageal peristalsis and the absence of relaxation of the LES. Slowly progressive dysphagia is usually followed by weight loss in patients with achalasia. People with achalasia feel the need to push or force their food down with water or other drinks to complete the swallowing process. There is a sense of substernal fullness, and regurgitation is common. Achalasia is also classified as a precancerous condition.[2,3,11]

Epidemiology

Achalasia is a rare condition. The incidence and prevalence of achalasia can be seen in table 2. The incidence of esophageal achalasia is about 1-10:100,000, with a prevalence of 10 in the 10,000 population. The distribution of men and women is equal, without any racial differences. Achalasia occurs at all ages, with incidence from birth to 7-8 decades and a peak incidence at 30-60 years of age. The incidence of esophageal achalasia in the United States is reported to be 2000 cases every day, with ages between 25-60 years, and it is rarely found in children. The Gastroenterohepatology Division of the Faculty of Medicine, University of Indonesia, reported about 48 cases over 5 years (1984-1988) at Cipto Mangunkusumo Hospital, with the majority of the age group being the same. In 2018 there were reported 3 cases of achalasia patients who came for treatment at the ENT clinic, KL RSMH, men aged 67 years, 56 years, and 32 years.[7-10]

Etiology

The etiology of achalasia is not known with certainty. However, there is evidence that degeneration of Auerbach's plexus leads to loss of neurological regulation. Histologically, abnormalities were found in the form of degeneration of Auerbach's plexus ganglion cells along the thoracic esophagus. This is thought to be the cause of esophageal peristalsis. According to the etiology, achalasia can be divided into primary achalasia and secondary achalasia. Primary achalasia is most often found. The exact cause is unknown, thought to be caused by neurotropic viruses and hereditary factors. Secondary achalasia (rarely), this disorder can be caused by infection, intraluminal tumors such as cardiac tumors, or extraluminal thrusts such as pancreatic pseudocysts. Another possibility can be caused by anticholinergic drugs or post-vagotomy. [4,6,12,13]

Based on the etiological theory, there are four theories of the occurrence of achalasia, namely genetic theory, infection theory, autoimmune theory, and degenerative theory. In genetic theory, the finding of cases of achalasia in several people in the same family has supported that achalasia may be inherited genetically. This possibility ranges from 1% to 2% of the population with achalasia. Associated factors include bacteria (diphtheria pertussis, clostridia, tuberculosis, and syphilis), viruses (herpes, varicella-zoster, polio, and measles), toxic substances (gas combat), esophageal trauma, and uterine esophageal ischemia during rotation of the digestive tract. Intrauterine. The strongest evidence supports neurotrophic infectious factors as an etiology. First, the specific location of the esophagus and the fact that the esophagus is the only part of the digestive tract where smooth muscle is covered by squamous cell

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epithelium allows the infiltration of infectious factors. Second, the many pathological changes seen in achalasia could explain viral neurotrophic factors that result in lesions of the dorsal vagus nucleus in the brainstem and myenteric ganglia in the esophagus. Third, serological examination showed an association between measles and varicella-zoster in achalasia patients. The discovery of the autoimmune theory for achalasia is drawn from several sources. First, the inflammatory response in the myenteric esophageal plexus is dominated by T lymphocytes, which are known to play a role in autoimmune diseases. Second, the highest prevalence of class II antigens, which are known to be associated with other autoimmune diseases. Finally, in some cases of achalasia, autoantibodies were found from the myenteric plexus. Epidemiological studies from the US found that achalasia is associated with the aging process with neurological status or psychological diseases such as Parkinson's disease and depression.[4,6,12,13]

Pathophysiology

During the last 75 years, research on the pathology of achalasia has shown a decrease in myenteric plexus neurons. This illustration is based on a study by Goldblum in 42 achalasia patients who underwent esophagectomy, 64% had no myenteric ganglion cells, and 36% had a decrease in myenteric ganglion cells in the esophagus. The study also demonstrated a predominance of T-cell inflammatory infiltration in the myenteric plexus and fibrosis. A recent study of achalasia patients treated at an early stage showed the presence of intact ganglion cells but with reduced cell numbers. Patients at this early stage have a shorter duration of symptoms and no esophageal dilation. There is an assumption that inflammation in the myenteric is an early phase of achalasia, causing aganglionosis and fibrosis. Several theoretical hypotheses for the occurrence of achalasia include the presence of cholinergic innervation ability and loss of inhibitory innervation.[14-16]

Theoretical hypotheses of cholinergic innervation ability. An in vitro study by Troounce in 1957 showed that striated muscle contractions in achalasia patients were a combination of an acetylcholinesterase inhibitor, serine, a ganglionic agonist, and nicotine. Acetylcholinesterase activity in LES ganglion cells in achalasia patients was described by Adams in 1961. The acetylcholinesterase inhibitor edrophonium chloride was subsequently shown to significantly increase LES pressure in patients with achalasia. These findings suggest that at least some postganglionic cholinergic nerve endings remain intact. Other studies have demonstrated the effectiveness of the anticholinergic agent atropine in achalasia patients. There is a 30% to 60% reduction in LES pressure in patients with achalasia who are given atropine. A similar decrease was found in the control group of healthy volunteers. However, the residual pressure after atropine administration was significantly higher in achalasia patients (17 mmHg) compared to normal subjects (5 mmHg).[14-16]

The theoretical hypothesis of loss of inhibitory (inhibitory) innervation in achalasia is that neurons in the cholinergic innervation of the esophagus are lost upon excitation, and these neurons are selective in inhibitory neurons. In achalasia patients, cholecystokinin induces LES contraction and, conversely, induces LES relaxation in healthy subjects, suggesting a postganglionic inhibitory neuronal disorder.[14]

Evidence supporting the concept of inhibitory neuron loss comes from immunohistochemical and physiological studies. Early studies suggested an inhibitory esophageal adrenergic nerve defect in achalasia patients. Vasoactive Intestinal Polypeptide (VIP) is also considered an inhibitor of the esophageal neurotransmitter, and a decrease in VIP-containing neurons is seen in achalasia patients. Another study demonstrated the absence of nitric oxide synthase in neurons in LES specimens in achalasia patients. The study showed nitric oxide synthase increased the resting phase of the LES and almost abolished LES

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relaxation. In the esophagus, nitric oxide synthase cause contractions in the esophageal body simultaneously, so there is an opinion that achalasia is a combination of the loss of selective inhibitors and the presence of enteric cholinergic nerve function.[14-16]

Pathological studies of esophageal resection specimens in end-stage achalasia patients show aganglionosis. Aganglionosis is the end result of myenteric inflammation in the majority of achalasia patients. This supports that achalasia is caused by the presence of cholinergic nerve excitation and the absence of nitric oxide inhibitors. Under such circumstances, functional obstruction of the gastroesophageal junction is caused by myogenic remnants of the LES. The absence of esophageal peristaltic activity results from the absence of enteric neural innervation.[14,16,17]

Clinical manifestations

Patients with achalasia, regardless of the primary or secondary cause, have almost the same clinical symptoms. Symptoms include swallowing abnormalities or progressive dysphagia, odynophagia, regurgitation, chest pain, and weight loss. In any patient who has complaints of dysphagia with solid and liquid food accompanied by regurgitation of food and saliva, achalasia should be suspected. The onset of dysphagia is usually gradual, initially described as a "fullness in the chest" or "sticking sensation" and occurring daily or with every meal. Initially, dysphagia is mainly in solid foods, but over time there is dysphagia in solid and liquid foods, especially cold drinks. The presence of a "power swallow" and carbonated drinks increases intraesophageal pressure and may increase esophageal emptying.[15,16]

Regurgitation becomes a problem as the disease progresses, especially when the esophagus is dilated. Regurgitation of retained food and accumulation of saliva is sometimes misdiagnosed as post-nasal phlegm or bronchitis. Usually occurs after eating at night, when the patient often wakes up coughing and choking. Aspiration pneumonia is a rare problem. Chest pain occurs in some patients, especially at night, and is seen in patients with mild disease or minimally dilated esophagus. The mechanism of chest pain is unknown, but this symptom is not only a simultaneous contraction of repeated episodes but can lead to obstruction of the esophageal lumen. Pneumatic or surgical dilation can reduce dysphagia and regurgitation. Heartburn is a common complaint in achalasia, despite the fact that achalasia is not associated with increased episodes of acid reflux. The cause of these symptoms is speculative, possibly related to the retention of acidic beverages such as soda or fruit drinks and, in some cases, due to the production of lactic acid from food retained in the dilated esophagus. Most patients with achalasia have some degree of weight loss but usually over a period of months to years.[15,16]

Diagnosis

The diagnosis of achalasia is established based on anamnesis, clinical symptoms, radiological examination, esophagoscopy, and manometry. From the anamnesis, the patient complained of difficulty swallowing, swallowing disorders, progressive dysphagia, odynophagia, regurgitation, chest pain, and weight loss. The diagnosis of achalasia should be suspected in any patient who has complaints of dysphagia with solid and liquid foods with regurgitation of food and saliva. The onset of dysphagia is usually gradual, initially described as a "fullness in the chest" or "sticking sensation" and occurring daily or with every meal. Initially, dysphagia mainly occurs in solid foods, but over time dysphagia also occurs in liquid foods. Regurgitation becomes a problem as the disease progresses, especially when the esophagus is dilated. Regurgitation of retained food and accumulation of saliva is sometimes misdiagnosed as post-nasal phlegm or bronchitis. Usually occurs after eating at night, when the patient often wakes up coughing and choking. Aspiration pneumonia is a rare problem. Chest pain occurs in some patients, especially at night, and is seen in patients with mild

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disease or a minimally dilated esophagus. Heartburn or a burning feeling in the chest is a common complaint in achalasia, despite the fact that achalasia is not associated with an increased incidence of gastric acid reflux. In achalasia patients, this complaint is usually due to retention of gastric acid or toxins produced by lactate which is fermented by bacteria in the esophagus. Most patients with achalasia have symptoms of weight loss but usually over a long period of months to years.[15,16]

The supporting examination carried out is in the form of a radiological examination. A radiological examination of the chest X-ray of achalasia patients did not reveal any air bubbles in the upper part of the stomach. It could also show an air-fluid level picture in the posterior mediastinum. Esophageal barium examination in Chagas disease and primary achalasia is almost the same. The feature that helps distinguish primary achalasia and Chagas disease is the presence of accompanying diseases in primary achalasia, such as megacolon and cardiomegaly, which are visible on fluoroscopy examination.[6,10,18]

Examination of the barium esophagram with fluoroscopy showed radiological features in the form of normal peristaltic waves only in the proximal third of the esophagus, while dilatation in the distal two-thirds of the esophagus showed abnormal peristalsis or completely disappeared and narrowing of the distal esophagus. Resembling a mouse tail (mouse tail appearance) or resembling a bird's beak (bird's beak appearance).[6.18]

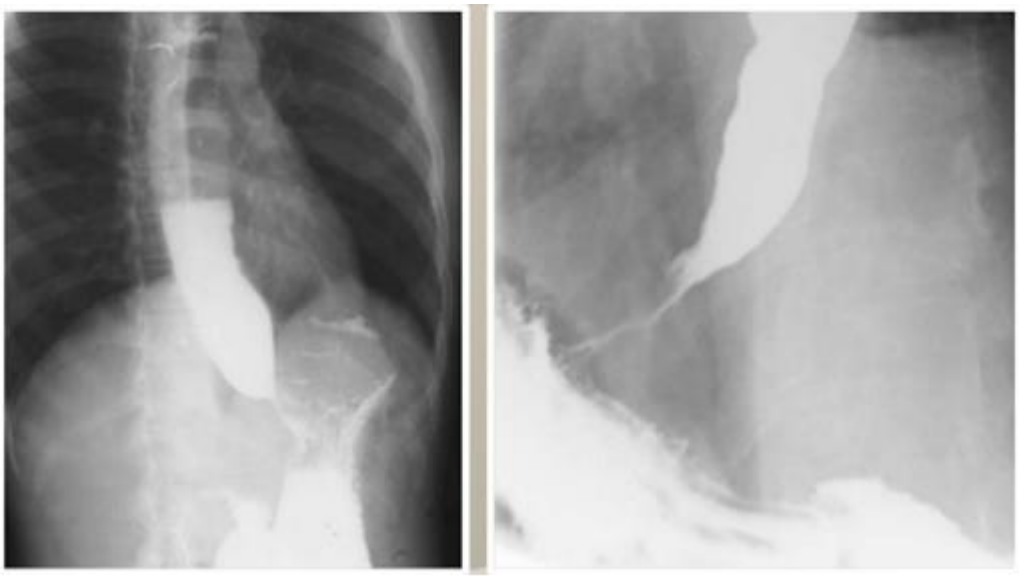

Figure 1. Esophagogram in achalasia[6]

On esophagoscopy examination, the lumen of the esophagus is widened with a narrowed distal part. There are food and fluid residues proximal to the narrowing area. The esophageal mucosa looks pale and edematous, and sometimes, there are signs of esophagitis due to food retention. The lower esophageal sphincter opens with a little pressure, and the esophagoscope can easily enter the stomach. Endoscopy in patients with achalasia is not a diagnostic tool unless other disease entities are present and complications are known. Endoscopic findings in primary achalasia are normal esophageal mucosa with a degree of endoscopic pressure resistance as it passes through the esophageal and gastric junction of mild to moderate degrees.[19-21]

In manometry examination, the function of manometry examination is to assess the motor function of the esophagus by examining the pressure

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in the lumen and the esophageal sphincter. This examination is to show motility abnormalities quantitatively and qualitatively. A manometry examination is done by inserting a tube through the mouth or nose. In achalasia, the motor function of the body of the esophagus and the lower esophageal sphincter is assessed. In the body of the esophagus that is assessed is the pressure at rest and its peristaltic activity. The lower esophageal sphincter was assessed for resting pressure and relaxation mechanism.[19-21]

Esophageal manometry is the gold standard for the diagnosis of achalasia. The three classic features of achalasia on manometric examination are aperistalsis of the esophageal body, an increase in LES pressure greater than 45 mmHg (normal 15-30 mmHg), and impaired LES relaxation during swallowing. The typical manometric picture is that as the resting pressure of the esophageal body increases, there is no peristaltic movement along the esophagus as a reaction to the swallowing process. Lower esophageal sphincter pressure is normal or elevated, and there is no relaxation of the sphincter during swallowing.[19-21]

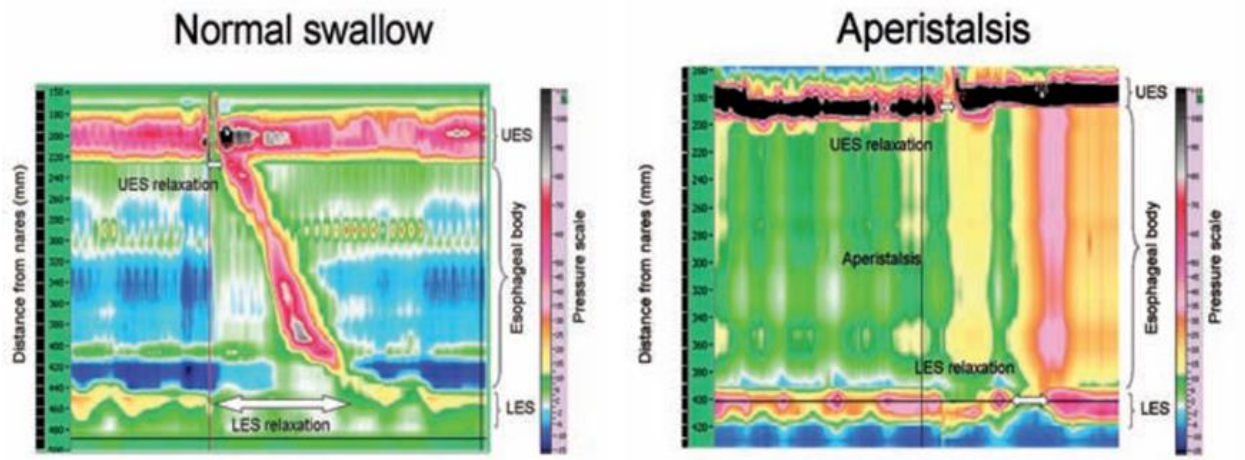

Figure 2. High-resolution manometry in normal patients (a) and in patients with achalasia (b). The image above is a temporospatial representation of the pressure data recorded in 32 pressure channels in the esophagus. Different colors are associated with a range of pressures, as shown on the scale. At rest, the upper and lower esophageal sphincters are identified as two distinct pressure bands. During normal swallowing (a), the upper esophageal sphincter relaxes and opens. This is followed by esophageal peristalsis as the LES relaxes. In a patient with achalasia (b), there is only slight peristaltic contraction under the upper esophageal sphincter (achalasia does not affect skeletal muscle peristalsis), and complete aperistalsis in the distal esophagus. The LES is often poorly relaxed (e.g., residual LES pressure > 8mmHg) and possibly overpressured (e.g., resting LES pressure >45 mmHg).[12]

Management

Treatment for achalasia is palliative because the peristaltic function of the esophagus cannot be restored. There is no treatment that can restore muscle activity and innervation in the esophagus in cases of achalasia. The treatment of achalasia is to reduce the level of pressure in the lower esophageal sphincter (LES). The goals of therapy include relieving the patient's symptoms, particularly dysphagia and regurgitation, increasing esophageal emptying by correcting impaired LES relaxation and preventing the development of megaesophagus. Therapy can be done

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by giving a high-calorie diet, medication, dilation, psychotherapy, and esophagus cardiotomy surgery (Heller's surgery).[19-21]

Administration of medication can only relieve symptoms for a short time, but the results are unsatisfactory. The drugs used are nitrite preparations, anticholinergics, and adrenergic blockers. Calcium channel blockers (nifedipine 10-30 mg sublingually) and nitrates (nitroglycerin 5 mg sublingual or 10 mg orally) are the two most commonly used drugs for reducing LES pressure and esophageal emptying. In addition, phosphodiesterase-5-inhibitors and sildenafil can also be used. Other drugs that are rarely used include anticholinergics (atropine, dicyclomine, cimetropium bromide), -adrenergic agonists (terbutaline), and theophylline. However, only about 10% of patients had success with this therapy.[19-21]

Calcium channel blockers and nitrates are effective in reducing lower esophageal sphincter pressure and temporarily reducing dysphagia but do not improve lower esophageal sphincter relaxation or peristalsis. The clinical response of this drug is short-term, does not reduce overall symptoms, and the drug's efficacy decreases over time. Because of these limitations, the use of pharmacologic therapy is recommended only in patients who cannot undergo pneumatic dilatation or myotomy or who fail to administer botulinum toxin injections.[13,18]

Treatment of achalasia with botox injection. Botox is a potent presynaptic inhibitor of acetylcholine from nerve endings that can be used to inhibit the release of acetylcholine in the LES by blocking LES cholinergic stimulation, which then interferes with the neurogenic component of the sphincter causing esophageal emptying. Using an endoscope, the toxin is injected using a sclerotherapy needle which is inserted into the esophageal wall at an angle of 45°. The needle is inserted until the mucosa is approximately 1-2 cm above the squamocolumnar junction. The injection site of this needle is located just above the proximal border of the LES, and the toxin is injected caudally into the sphincter. The effective dose used is 80-100 units/mL, which is divided into 20-25 units/mL to be injected in each quadrant of the LES. Repeat treatments at 6 – 24 months intervals. The side effect of this therapy often causes an inflammatory reaction at the gastroesophageal junction, which can make myotomy more difficult. This therapy should be applied to elderly patients who have contraindications to pneumatic dilation or surgery.[19-21]

Pneumatic dilatation (PD) is the most effective nonsurgical option. Pneumatic dilation is performed endoscopically using air pressure to dilate and destroy the circular muscle fibers of the lower esophageal sphincter. Balloon dilators there are 3 diameters, namely 3; 3.5; and 4 cm, placed on the end of the endoscope. Of particular importance in effective pneumatic, dilatation is the placement of a balloon dilator in the lower esophageal sphincter (LES), followed by fluoroscopy visualization. After pneumatic dilatation, a barium examination should be performed to exclude esophageal perforation. The predictor for the pneumatic dilatation response was a decrease in lower esophageal sphincter pressure after dilatation (<10 mmHg).[13,18]

A balloon is inflated at the gastroesophageal junction with the aim of rupturing the muscle fibers and leaving the mucosa intact. The initial success rate is between 70% and 80%, but it will decrease to 50% in the next 10 years, even after several dilations. The ratio of the occurrence of perforation is about 5%. If a perforation occurs, the patient is immediately taken to the operating room for closure of the perforation and myotomy, which is done by means of thoracotomy. The incidence of abnormal gastroesophageal reflux is about 25%. Patients who fail to treat pneumatic dilation are usually treated with Heller myotomy.[19-21]

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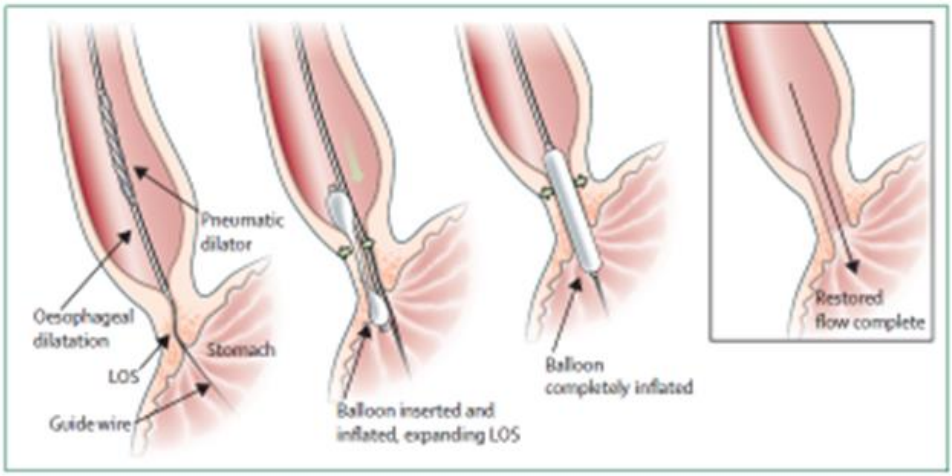

Figure 3. Pneumatic dilatation technique on akalasia[20]

Laparoscopic Heller myotomy and partial fundoplication are the procedures of choice for esophageal achalasia. This operation consists of a separation of the muscle fibers (myotomy) of the lower esophageal sphincter (5cm) and the proximal part of the stomach (2cm), followed by partial fundoplication to prevent reflux. Patients were hospitalized for 24-48 hours and returned to their activities after ±2 weeks. Effectively, this surgical treatment reduces symptoms by about 85-95%, and the incidence of postoperative reflux is between 10% and 15%. Because of its excellent efficacy, short hospital stay, and fast recovery time, this therapy is considered the mainstay of therapy in the treatment of esophageal achalasia. Patients who fail to undergo this therapy may require dilatation, a second operation, or removal of the esophagus (esophagectomy).[19-21]

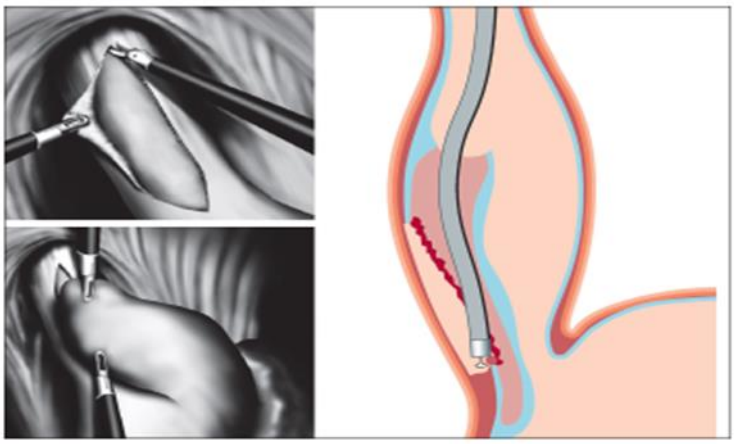

Figure 4. Laparoscopic Heller myotomy with dor fundoplication (left) and peroral endoscopic myotomy (right)[20]

Operative myotomy in achalasia, i.e., performing an anterior myotomy through the lower esophageal sphincter (Heller's myotomy) is usually associated with an antireflux procedure. Myotomy can be done orally by endoscopy, with a success rate of 80-94%. The oral endoscopic myotomy procedure was started by inserting the endoscope into the submucosal cavity after the submucosal injection with 0.3% saline and

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carmine indigo, then performing a mucosal incision in the mid-esophagus. Then make a submucosal canal (submucosal tunneling) 2-3 cm from the distal esophagus, with circular muscle fibers perpendicular to the longitudinal direction of the canal. Perform endoscopic myotomy of the deep, circular muscle using the tip of the endoscope blade, starting from 2-3 cm distal to the mucosal entry to 2-3 cm distal to the esophagogastric junction, leaving the outer longitudinal muscle layer intact. Close the mucosal incision using a hemostat clamp.[20.21]

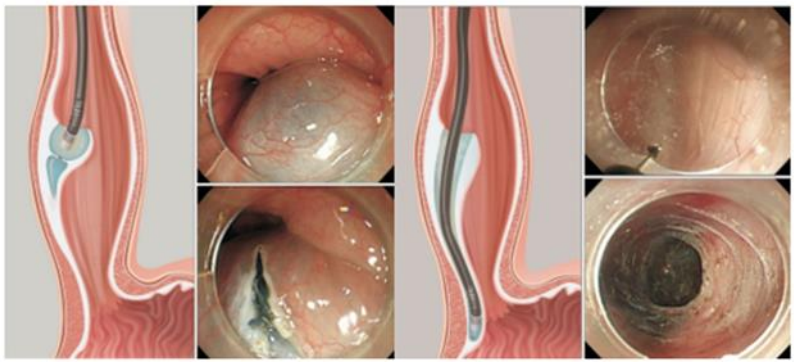

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Figure 5. a. Insert the endoscope into the submucosal cavity. b. Submucosal tunneling[21]

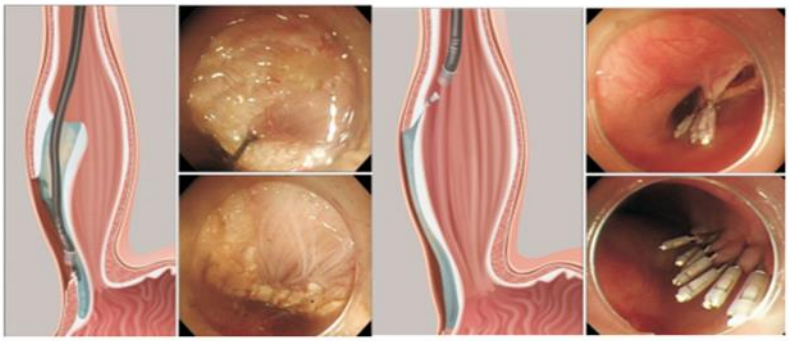

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Figure 6. a. Endoscopic myotomy with a knife tip on the circular muscle. b. Closure of the mucosal incision[21]

Complications

Some of the complications of achalasia associated with the progression of the disease and its treatment intervention are aspiration pneumonia, megaesophagus, in 10% of untreated cases, squamous cell carcinoma due to high levels of nitrosamines produced by bacterial growth due to stasis in the esophagus leading to chronic inflammation and dysplasia, esophageal adenocarcinoma, respiratory tract obstruction, bronchitis, lung abscess, diverticulum, esophageal perforation, sudden death.[19-21]

Prognosis

There is no cure for achalasia. Treatment is symptomatic, aiming to increase esophageal emptying and reduce symptoms of dysphagia. Esophageal peristalsis remains absent, swallowing does not completely return to normal, and the patient can only swallow in a straight/upright position. The prognosis for achalasia depends on the duration of the disease and the extent of the motility disorder. The shorter the

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duration of the disease and the less the motility disorder, the better the prognosis for returning to normal esophageal size after surgery ('s myotomy) is very good. When a surgeon is available, surgery results in better symptom relief in the majority of patients and gives better results than pneumatic dilation. Drugs and botulinum toxin should be used only in patients unable to undergo pneumatic dilation and laparoscopic Heller myotomy.[19-21]

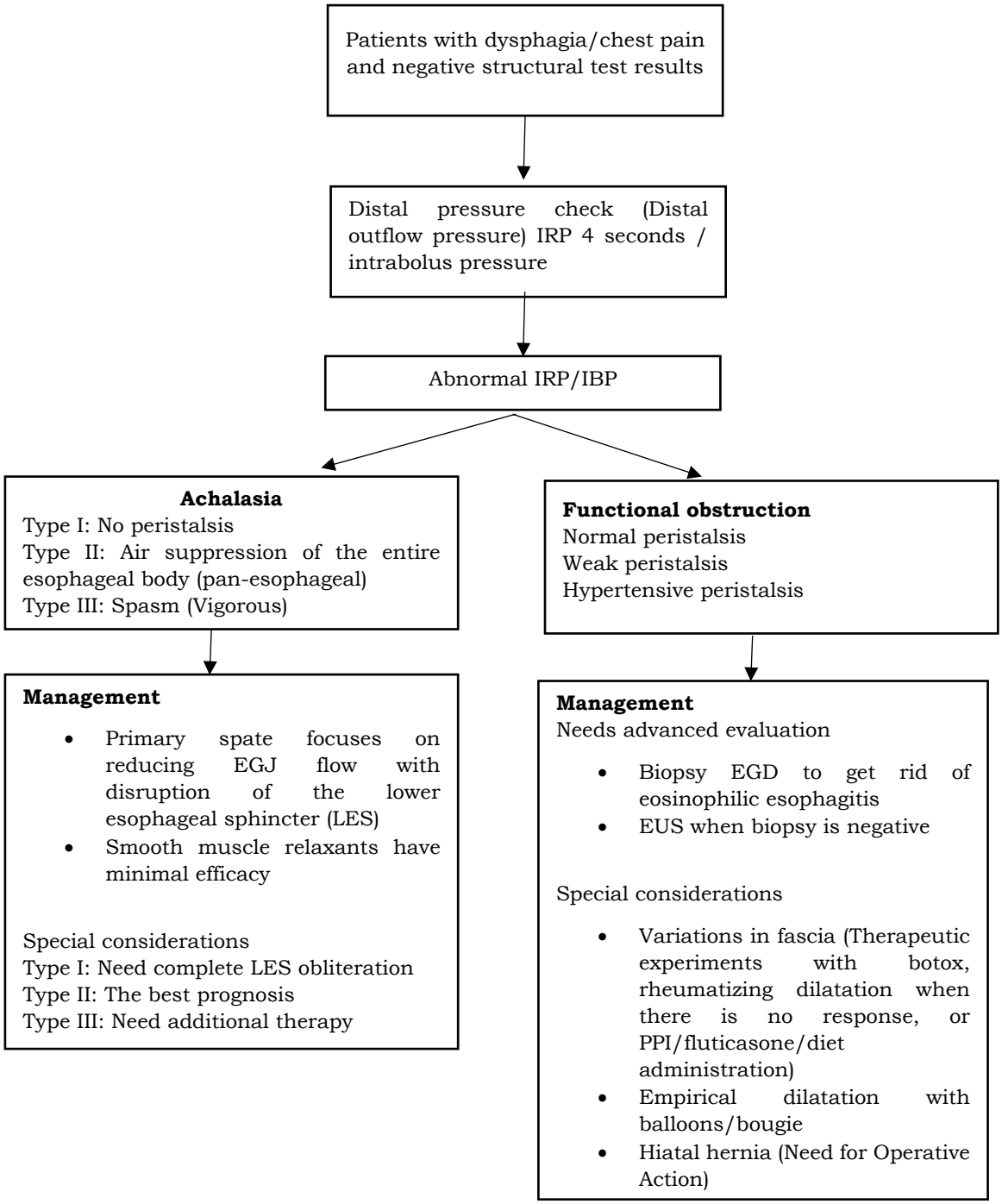

Figure 7. Algorithm for treatment of achalasia[10]

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2. Conclusion

Achalasia is an esophageal motility disorder of unknown cause and occurs at any age with symptoms, especially difficulty swallowing solid/liquid food and regurgitation. The diagnosis of esophageal achalasia was based on clinical symptoms, radiological features, esophagoscopy, and manometry. The treatment of achalasia is to reduce the level of pressure in the lower esophageal sphincter (LES). Therapy can be done by giving a high-calorie diet, medication, dilation, psychotherapy, and esophagus cardiotomy surgery (Heller's surgery).

3. References

1. Hedayanti Nor, Supriono. Achalasia: A review of etiology, pathophysiology, and treatment. The Indonesian journal of gastroenterology, hepatology and digestive endoscopy, 2013; 24(3): 243-251.
2. Siegel G. Leighton. Diseases of the lower airway, esophagus, and mediastinum endoscopic considerations. In: Adams, GL, Boies, Lawrence R., Higler, PA BOIES Textbook of ENT Diseases, 6th eds. Jakarta: EGC, 1998: 4-462
3. Pressman A, & Behar J. Etiology and pathogenesis of idiopathic achalasia. Journal of Clinical Gastroenterology. 2016; 13(3): 201-210.
4. Sjamsuhidajat. Wim de Jong textbook of surgery. Jakarta: EGC, 2010: 9-67
5. O'Neill, OM Achalasia: A review of clinical diagnosis, epidemiology, treatment, and outcomes. World Journal of Gastroenterology, 2015; 19(35): 58-66.
6. Soepardi EA. dysphagia. Textbook of Ear Nose Throat Head & Neck Health Sciences. 7th eds. Jakarta: Balai Penerbit FKUI, 2012: 244-248.
7. Krill, Joseph T, Naik, Rishi D. Clinical management of achalasia: current state of the art. Clinical and Experimental Gastroenterology. 2016; 9(3): 71–82.
8. Vaezi MF, Richter JE. Diagnosis and management of achalasia. American College of Gastroenterology Practice Parameters Committee. Am J Gastroenterol, 1999; 94(12): 3406–3412.
9. Francis DL, Katzka DA. Achalasia: update on the disease and its treatment. Gastroenterology. 2010; 139(2): 369–374.
10. Patel, DA, Kim, HP, Zifodya, JS, & Vaezi, MF Idiopathic (primary) achalasia: a review. Orphanet Journal of Rare Diseases, 2015;10(1):121-131.
11. El Kafsi J, Foliaki A, Dehn TCB, and Maynard, ND. Management of achalasia in the UK, do we need new guidelines? Annals of Medicine and Surgery. 2016; 12(3): 32–36.
12. Pohl, Daniel, Tutuian, Radu. Achalasia: an overview of diagnosis and treatment. J Gastrointestin Liver Dis. 2007; 16(3): 297-303.
13. Farrokhi F, Vaezi MF. Idiphaic (primary) achalasia. Orphanet Journal of Rare Disease, 2007; 2(38): 1-9.
14. Ritcher JE. Achalasia - an update. J Neurogastroenterol Motile. 2010; 16(3): 232-242.
15. Alawiyah AB. Achalasia: Distinguishing Primary and Secondary. Yogyakarta: Fakultas Kedokteran Universitas Gadjah Mada. 2014.
16. Vaezi MF, Pandolfino JE, and Vela MF. ACG clinical guideline: diagnosis and management of achalasia. The American Journal of Gastroenterology. 2013; 23(4): 55-67.
17. Boeckxstaens GE, Zaninotto G, Richter JE. Achalasia. Lancet. 2014; 38(3): 86-97.
18. O'Neill OM, Johnston BT, Coleman HG. Achalasia: A review of clinical diagnosis, epidemiology, treatment, and outcomes. World Journal of Gastroenterology. 2013; 19(4): 5809-5810.

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19. Vela MF, Richter JE, Wachsberger D. Complexities of managing achalasia at a tertiary referral center: use of pneumatic dilatation, Heller myotomy, and botulinum toxin injection. Am J Gastroenterol, 2004; 99(2): 1029-1036.
20. Luckey, AE, DeMeester, SR. Complications of Achalasia Surgery. Thoracic Surgery Clinics, 2006; 16(1): 95–98.
21. Youn Y. Peroral endoscopic myotomy for treating achalasia and esophageal motility disorders. J Neurogastroenterol Motil, 2016; 22(1): 14-24.

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