**Review Article**

**Approach and management modalities in esophageal perforations**

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**ABSTRACT**

Esophageal perforation leads to increased morbidity and mortality. Treatment of choice is debatable. In adult cases of esophageal perforation, the important causes can be due to trauma, or due to foreign body or due to iatrogenic and sometimes it may be spontaneous. The rate of morbidity and mortality is increased among those who fall prey to delayed diagnosis and delayed treatment. Even after the proper treatment started within one day of diagnosis, the death rate can be 10-25%. And it increases to 40-60% among those who receive treatment after two days. In case the diagnosis of perforation is made within 24 hours, the treatment of choice is “primary closure of the perforation site and wide drainage of the mediastinum”. On the other hand, if the diagnosis is delayed for more than 24 hours, then the treatment of choice is questionable. Now a days conservative method of management is more or less adopted especially patients with small size perforation and minimum involvement of extra esophageal part. Mortality rate is determined by some factors like cause, the injury site, pre-existing disease of esophagus, treatment method used and the timing of the diagnosis. Hence multi-disciplinary approach is recommended for proper outcome. Timing of intervention is extremely critical in ensuring low mortality. Diagnosis is difficult, and a high degree of suspicion is required, especially in pediatric patients. As Endoscopic therapists play an increasingly “important role in the management of upper digestive disease”, a proper understanding of this complication of therapy is required by the therapist and the attending surgeons. Management of such perforations are both non-operative and operative. Nonoperative management used for limited perforations without sepsis, while operative management is reserved for more florid perforations and those in whom obstruction distal to the perforation exists. Endoscopic intervention may save some patients from being operated upon.

**Keywords:** Esophagus, Mediastinitis, Perforation, Pediatric

**INTRODUCTION**

Esophageal perforation leads to increased morbidity and mortality. Treatment of choice is debatable. In adult cases of esophageal perforation, the important causes can be due to trauma, or due to foreign body or due to iatrogenic and sometimes it may be spontaneous. The rate of morbidity and mortality is increased among those who fall prey to delayed diagnosis and delayed treatment. Even after the proper treatment started within one day of diagnosis, the death rate can be 10-25%. And it increases to 40-60% among those who receive treatment after two days. Typical structure and placement of esophagus leads to infection by bacteria easily as well as damage by digestive enzymes can occur which in turn can lead to increased mortality. But fortunately this condition of esophageal perforation is rare. Its presentation is also not typical in all the cases. These two factors combined are responsible for diagnostic delay. In adult cases of esophageal perforation, the important causes can be due
to trauma, or due to foreign body or due to iatrogenic and sometimes it may be spontaneous.7-10

In case the diagnosis of perforation is made within 24 hours, the treatment of choice is primary closure of the perforation site and wide drainage of the mediastinum.1-17 On the other hand, if the diagnosis is delayed for more than 24 hours, then the treatment of choice is questionable.18-37

Mortality rate is determined by some factors like cause, the injury site, pre-existing disease of esophagus, treatment method used and the timing of the diagnosis.1,14

Esophageal perforations are dramatic complications and if managed improperly result in mortality. Proper management requires an appreciation of the anatomy, the etiology and the pathophysiology of this entity.1,2

Anatomy

The Esophagus “extends from the level of the” 6th cervical vertebra marked externally by “the cricoids cartilage” to enter the Stomach at the level of L1 vertebra after passing through the “Right crus of the diaphragm”. It has a sinuous course situated more to the right superiorly and to the left in its lower 1/3 of the thoracic course before it takes a bend to the left to enter the stomach. It is a muscular tube lined by serosa only in its inferior intra-abdominal portion. It is related to the membranous trachea, the Recurrent laryngeal nerve in the trachea-esophageal groove, the left main bronchus, the aortic arch and then to the left mediastinal pleura. The Thoracic duct lies posterior to it within the thorax. It therefore, courses through all three regions- cervical, thoracic and Abdominal (Intra and Extra-peritoneally) and therefore any sepsis can pass from one cavity to the other increasing greatly the lethality of a perforation. It is narrowest at the Cricopharyngeal sphincter and also narrows “where it is crossed by the aortic arch and the left main bronchus” and at its entry into the abdomen. Esophageal perforations “often occur near the pharyngo-esophageal junction where the wall is weakest.” “Loose stromal connective tissue” surrounds the esophagus and hence the inflammatory response to infection can spread to nearby organs easily and this makes esophageal perforation a surgical emergency. Older age of more than 65 years, patients having pre-existing disease of esophagus, predisposes to perforation in instrumentation cases and this has been found to affect distal area of instrumentation.1,3

Etiology

The main cause of esophageal perforation are Iatrogenic (55%) and Spontaneous (15%) or (Boerhaave’s Syndrome).1,5 Iatrogenic main cause is diagnostic endoscopy like endoscopic biopsy and therapeutic endoscopy such as endoscopic dilatations, variceal sclerotherapy, endoscopic laser therapy, endoscopic photodynamic therapy, endoscopic stent, nasogastric tube placement, endotracheal intubations, transoesophageal echocardiography, echocardiography, mini-tracheostomy, operative endoscopy like aortic aneurysm, cervical disk surgery, thoracic surgery, achalasia.

Spontaneous esophageal perforation caused by traumatic such as foreign body such as bones, dentures, caustic agents such as acids and alkali, blunt and penetrating thoracic injury, malignant and miscellaneous like rupture of aortic aneurysm, erosion by aortic prosthesis.

Presenting features

They depend upon factors like etiology, time of patient presenting to hospital, and the perforation site.1,2,6,8,12 80% of the patients have pain at the perforation site.1,2,16 Other presenting symptoms are dysphagia, cough, vomiting, fever, hematemesis, and tachypnea.2,8,12

Iatrogenic rupture is “the most common” group and may follow dilatation of a benign stricture, balloon dilatation of Achalasia, dilatation/lumenisation of carcinoma esophagus, during endoscopic mucosal resection of Barret’s esophagus, Endoscopic submucosal dissection of early gastric cancers with 4% chances of early or delayed presentation of perforation, diagnostic trans esophageal echocardiography of cardiac myopathies 1 in 10,000 esophageal perforation can be seen or intra-operatively. These diseases have the primary symptomatology, in addition to severe chest, cervical or abdominal pain following the procedure. Suspicion should be raised when pain persists more than 15 minutes. Esophageal perforation is suspected if the patient complains of pain in chest and related parts. The dilator may be stained with blood. In patients with relumination for malignancy, using laser, cautery or alcohol, perforation may be delayed and may present late with abdominal pain, fever and features of sepsis or fistulisation into the trachea or bronchus.1,2,4,5

Spontaneous perforation

Up to 50% are not diagnosed in time. Vomiting of severe nature associated with acute pain suggest spontaneous.13,15 Spontaneous rupture may follow any straining effort-labour, asthma, blunt trauma etc. When in addition, there is cervical subcutaneous emphysema, the term Mackler’s triad is used. Cervical emphysema takes at least one hour to develop. Patients may also present with epigastric or cervical pain with sweating, and dyspnoea is almost always present. Physical signs are not remarkable early in the disease course. Signs of fluid in the left pleural cavity maybe present due to rupture of the esophagus into the left thoracic cavity. A mediastinal crunch with each heartbeat may be present and is called Hamman’s sign.1,2,5

Foreign bodies as cause of perforation are common in children and they commonly have dysphagia, pain and, if
recognition is delayed- sepsis. Batteries, pins especially safety pins are commonly associated with perforation whether primarily or during their extraction. In elderly dental prosthesis as in artificial dentures are quite frequently ingested without notice, one reason is that tactical sense of the palate is lost in carriers of dental prosthesis, when not removed early these can cause esophageal erosions, perforation, abscess and mediastinitis.6

Corrosive perforations

Ingestion of caustic material causes corrosive perforations. The authors have recommended that these lesions staging should be done by endoscopy. They experienced that diagnosis of this condition on war foot basis and aggressive therapy are needed. Acute bleeding and complications due to perforation can be prevented by early operation.38

In pediatric age group accidental ingestion of corrosives are rare and need a brisk diagnostic and therapeutic approach. “Management of caustic ingestion in children” is still controversial. Minimally invasive procedures are well accepted because at younger age, tissues have high healing capacity.

Investigations

The surgeon must bear the high degree of suspicion for appropriate diagnosis. lateral X-ray of the neck is recommended in cervical esophageal perforation. It shows air in the pre-vertebral facial planes. Posterior and lateral chest radiographs are recommended in thoracic or intra-abdominal esophageal perforation.18

A plain radiograph of the Chest shows emphysema either subcutaneous or mediastinal, an effusion, a Pneumothorax (77% of cases) and this pneumothorax is due to injury to the mediastinal pleura. 70% of the cases have it on the left side, some 20% on the right side and remaining 10% can be on both the sides.18 Hydropneumothorax on left side is common in patients with distal third esophageal perforations.18-24 A mediastinal widening, Impacted dentures are difficult to diagnose by radio imaging if there is no wire in then unless there is a complication.

A contrast radiograph, using Gastrograffin is required for both diagnosis and decision making. It is important to take a supine swallow in both left and right decubitus position since inadequate filling of the esophagus may not reveal the perforation. A barium esophagogram increases the yield by about 22% and is indicated if the Gastrograffin swallow fails to show the perforation.12,5

Findings to be noted are location of the perforation, size, whether it is contained, or contrast is leaking freely, whether it is self-emptying if contained, associated pathology, distal obstruction and presence of emphysema, pneumothorax, hydropneumothorax, pleural effusion, sub-phrenic collection etc.5

Treatment1-39

Treatment depends on whether the patient has clinical sepsis, the etiology of perforation and the existence of distal obstruction.

Spontaneous perforations of the Esophagus are often large and are best treated by Primary closure of the perforation within 24 hours of occurrence, after freshening the edge; buttressing the repair with either a pleural patch or the fundus of the stomach. Initial stabilization of the patient with fluids, antibiotics, pain killers and drainage of the chest must be done while investigations are being performed.

If there is evidence of uncontrolled sepsis or if the primary disease warrants esophagectomy- an early esophagectomy with mediastinal drainage will be the apt choice. When the patient is very sick and in a bid to preserve the esophagus a t-tube has been placed in the esophagus after thorough debridement of the mediastinum and placement of tubes for irrigation of the mediastinum and pleural cavity.

Iatrogenic perforations can be managed conservatively if they are contained with no evidence of clinical sepsis, only mild to moderate fever or leukocytosis, mild pain rapidly controlled with narcotics, self-emptying (Cameron Criteria); there is no distal obstruction or the perforation is not through a malignancy- treatment involves antibiotics, prohibition of oral intake and nutrition being maintained with total parental nutrition (or if scarce socioeconomic resources with Jejunostomy feeds). Perforations in achalasia are small and can often be managed conservatively.

In patients with unresectable esophageal cancers of the middle third placement of a stent may also cover the perforated area and is used when appropriate. Role of stents in todays practice has increased with use of self-expanding metal stents (SEMS) which are important to manage carcinogenic as well as lesions of the obstructive nature. Polyflex stent is a flexible stent which can be removed, and it is also self-expanding can be used for benign cases.

Disadvantages of stent which are migration, foreign body sensation, vomiting and bleeding have been overcome by Anchoring of the upper flare of the fully covered SEMS with an endoscopic clip is feasible and significantly reduces stent migration. There are also reports in literature of tissue glues being used to plug iatrogenic perforations.

Pediatric perforations

Pediatric esophageal perforations present with diagnostic
difficulty and high index of suspicion and temporal relationship with esophageal instrumentation or trauma is to be considered.9

Etiopathogenesis

Esophageal perforation whether traumatic, iatrogenic, corrosive is an acute medical emergency where death rate can be as high as 20-50%.5,7 If the perforation gets contaminated due to oral or gastro-intestinal contents then it can result into multi-organ dysfunction and death.10 Among pediatric cases, diagnosis delay is common. This is due to its presentation is similar to other diseases. Prognosis is worst in such cases.11 With the development of more flexible and less traumatic instruments, iatrogenic causes of perforation have come down.1

Corrosive perforation “minimally invasive management of” caustic ingestion consisting of flexible endoscopy, guide wire-assisted esophageal balloon dilatation and intra-lesional TAC injection without any gastrostomy or esophagi stent/placement is effective and leads to relief of dysphagia in almost all patients. This method of dilatation is also safe and iatrogenic esophageal perforation is very unlikely.39

Management

The treatment of choice for esophageal perforation used to be surgical repair among patients presenting within 24 hours. Otherwise the death rate was 70% or more among those who were not operated.12-17 In the present era, conservative management has been considered in cases due to iatrogenic causes, patients who are found to be stable hemodynamically, and patients showing lesser degree of contamination of the perforation.2

Of late, non-operative management is being advocated, more so in pediatric population, because at younger age, tissues have high healing capacity.22 A recent study advocates conservative management among patients who are found to be stable hemodynamically, and small size perforations who present to the health facility as early as 48 hours of injury.21

Conservative management dates back to 1965 where the death rate was 6%. All cases were iatrogenic in nature following some diagnostic procedures. They used the technique of inter costal drainage, suctions from naso-esophageal places etc.18-20

It was found that death rate in conservative approach was 17% compared to only 12% in surgical approach.20

It was found in case series among pediatric patients observed that there was no death among 18 cases which were managed conservatively. Only one case developed complication. Thus, they proved that even among children, conservative management can be adopted.13

In corrosive perforations among children conservative management with or without cervical esophagostomy and gastrostomy works nicely with relatively lesser mortality and morbidity.23

It was found in review that esophagectomy is better than conservative approach and even better that the primary repair.21

Prognosis

A review from India suggest that there was 18% mortality among patients with esophageal perforation.45 Important factors that affect prognosis are timing of diagnosis, site of perforation, any disease of esophagus, and the choice of therapy. The highest death rate is commonly seen among patients having spontaneous perforation i.e. 36%. This is followed by iatrogenic perforation in 19% of the cases. Traumatic perforations contribute to 7% of mortality.1,4,8,16-43

Prognosis also depends on the site of perforation, cervical esophageal perforations carry best outcome with only 6% mortality (range of 0-16%). Thoracic and abdominal perforations have little bit of bad prognosis with a mortality rate of 27% and 21% (range of 0-44%).1,4,8,16-43

Early diagnosis reduced death rate and complications.44 Eroglu et al also gave similar results.40

CONCLUSION

Thus, author concludes that perforation of esophagus among adults carry high death rate and is associated with dreaded complications. Death rates are influenced by factors like patient reporting to hospital, timing of the diagnosis, cause of perforation, and treatment of choice. The reported mortality from treated esophageal perforation is 10% to 25% when therapy is initiated within 24 hours of perforation, but it could rise up to 40% to 60% when the treatment is delayed beyond 48 hours.

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