

# Chagas Disease Surgery

Alcino Lázaro da Silva

Departamento de Cirurgia Digestiva, Universidade Federal de Minas Gerais, Caixa Postal 340, 30130-100  
Belo Horizonte, MG, Brasil

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Chagas disease has a chronic course in various regions of the body, creating functional incompetence and dilatations known as mega formations.

For the general surgeon, the digestive forms of Chagas disease are the dilatations that occur throughout the digestive tract jeopardizing the normal transit or even impeding it completely.

## NOMENCLATURE

The dilatations of the digestive tract are named megaesophagus, megastomach (achalasia of the pylorus), megaduodenum, megacolon, megasigmoid and megarectum.

Dilated forms have not been described in the jejunum and in the ileum. This may be due to the fact that these two segments of the digestive tract, because of their essential functions, are very resistant to infection.

## CLASSIFICATION

The dilatations that occur in the body are classified according to different systems: (1) biliary: mega-gallbladder, mega-common bile duct; (2) cardiovascular: cardiomegaly; (3) urinary: megapelvis, megoureter, megabladder; (4) respiratory: bronchiectasis, megabronchus; (5) megapharynx (?); (6) megaesophagus; (7) megastomach; (8) achalasia of the pylorus or hypertrophic pyloric stenosis; (9) megaduodenum; (10) megaintestine (mega-small bowel); (11) megappendix; (12) megacolon; (13) megasigmoid; (14) megarectum; (15) hypertrophy of the salivary glands (parotid).

## MEGAESOPHAGUS

This is the most common form of mega in Chagas disease.

As for megacolon, all therapeutic attempts have some restrictions for this reason most surgeons tend to adopt more than one procedure.

## History

(Ferreira Santos 1965, Chaib 1969).

## Classification

Classification is based on the stage (grade) of the disease - grade I: dilatation of up to 3 cm; grade II: dilatation of 3 cm to 7 cm; grade III: dilatation of more than 7 cm; grade IV: dilatation of more than 7 cm with deviation of the longitudinal axis of the esophagus (Rezende 1973).

## Symptoms

The most prevalent symptoms are: dysphagia (100%), retrosternal pain, regurgitation and loss of weight.

Diagnosis is based on the symptoms, by swallowing of barium examination, manometric criteria, mecolil test, endoscopy, biopsy and radiography of the chest. Serological tests may confirm the results of these exams.

## Surgical operations

Operations to correct megaesophagus are divided as follows (Ferreira Santos 1965): operations on the esophagus (6); operations on the intrinsic obstacle (31); intra lumen (oral route) operations (3); operations on the extrinsic obstacle (4); operations on the nerves of the esophagus (4); operations on the esophagus and the cardiac opening (5): (a) partial esophagectomy followed by sub aortic esophagogastronomy; (b) resection of the esophagogastric junction followed by interposition of a jejunal segment; (c) subtotal esophagectomy followed by cervical transmediastinal anterior esophagogastronomy; (d) same as (c) followed by transmediastinal posterior esophagogastronomy; (e) esophagojejunal anastomosis with mobilization of the stomach to the neck for a cervical gastrostomy. At this level, during a second operation, an anastomosis between the stomach and the esophagus is carried out.

## Preferred operations

*Esophagocardomyotomy (Heller operation)*  
- The main steps for this operation are: upper paramedian internal pararectus laparotomy; partial mobilization, from the left side, of the esophagogastric junction preserving the vagus nerves and their main branches; exposure of the anterior aspect of the esophagus; and an 8 cm extra mucosal myo-

tomy of all muscular layers (longitudinal, oblique and transverse) of the esophagus, cardiac region and 3 cm on the stomach is completed.

This operation benefits swallowing but allows free gastroesophageal reflux and may generate esophagitis and stenosis.

*Esophagocardiamyotomy plus lateral-lateral esophagogastric fundic wrap (Heller operation associated with the Toupet-Lind procedure)* - After completing the previous procedure, the fundus and the cranial part of the greater curvature of the stomach are mobilized. Nonabsorbable sutures are placed between the elements cited and the walls of the esophagus, for an extension of approximately 10 cm.

The association of both techniques will provide a better antireflux mechanism which will prevent esophagitis.

The success of this operation may be jeopardized by a folded, sigmoid type esophagus. Dysphagia is rare, but simple dilatation may occur in the postoperative period, even if esophagitis with esophageal shortening is present.

*Longitudinal esophagocardiomyotomy plus esophagogastric fundic wrap (Girard operation associated with the Toupet-Lind procedure)* - An 8 cm incision is made on the distal esophagus, cardiac region, and extending to the stomach. The opening is sutured transversely in order to enlarge the diameter of the esophagus. Due to the possibility of free gastroesophageal reflux, an antireflux mechanism is created as previously described. The possible complications are as previously cited.

*Cardiaectomy plus interposition of a jejunal segment (Merendino procedure)* (Merendino & Dillard 1955) - The esophagogastric junction is mobilized. Both vagus nerves are transected. The cardiac region of the stomach is resected along with 3 cm of the stomach. On the esophagus, resection varies with the elongation of the organ, in order to make the esophagus straight. Transit is reconstructed by placing a 20 cm segment of the jejunum between the divided ends of the esophagus and the stomach.

For megaesophagus grade III or higher this operation provides a definitive solution for the dysphagia and safely prevents reflux. Lack of success may occur due to a high incidence of immediate postoperative complications which may happen when one is not familiarized with the technique; in very advanced forms of megaesophagus, specially those sigmoid types (folded esophagus) an experienced surgeon should do the small bowel interposition.

In this operation, the gastroesophageal reflux does not occur because the interposed jejunal seg-

ment (approximately 20 cm) always presents distal peristalsis, acting as an excellent sphincter (Rezende 1973, Lázaro et al. 1987, Lázaro 1991).

#### MEGASTOMACH

Isolated gastric dilatation is rare. It occurs more frequently in association with dilatations of the esophagus and the free part of the duodenum, although they are also rare.

Significant gastric stasis may occur due to the dilated stomach and the dyskinesia of the gastroduodenal segment, similarly to pyloric stenosis. Two alternatives are available to overcome the obstruction.

The first one is a gastrojejunostomy which creates a large passage, but has the inconvenience of a marginal ulcer or the late recurrence of the stenosis (narrowing of the gastrojejunostomy due to the decrease of the gastric dilatation).

The second alternative is the distal resection of the stomach and a funnel shaped reconstruction. The upper part of this funnel is located at the level of the major gastric curvature favoring gravitational drainage to the duodenum through the gastroduodenal anastomosis.

#### MEGADUODENUM

Isolated megaduodenum is the most infrequent form of mega; but more frequent is its association with megaesophagus and megastomach.

The duodenum becomes elongated, widened, and has evident symptomatic stasis.

Little is known about the causes of megaduodenum: if the problem is incoordination along the duodenum or at the level of the duodenal jejunal junction, like in achalasia of the lower esophageal sphincter (Lázaro 1969).

Supposing the latter is true, we may resect the duodenal jejunal junction and then construct an end to end anastomosis recreating the transit, with satisfactory results. This is a more delicate operation than the simple side to side duodenal jejunostomy, but provides material for histopathological and motility studies which will allow further research in this subject (Lázaro & Pereira 1977).

#### MEGA-SMALL BOWEL

This phenomenon, if it really exists, it is extremely rare. If a patient presents this abnormality in all the extension of the small bowel, a surgical solution would be probably impossible because this segment of the digestive tract is vital. From a physiopathological point of view, one may suggest the displaying of the bowel loops in an organized way by means of a partial thickening of the tract, suturing one another, in order to better orient the bowel stasis.

**MEGAPPENDIX**

This form is rare and not very significant because it is usually associated with dilatation of the colon. The chosen procedure is resection, and it can be associated or not with portions of the colon.

**MEGACOLON**

This form may appear alone or with dilatation of other parts of the digestive tract, it frequently occurs in association with megaesophagus.

Stasis is the most common symptom, it may lead to chronic or acute obstruction with faecal impaction, or even torsion of the sigmoid colon.

Although the pathophysiology is the same, differences occur due to individual physiological and anatomical characteristics of the colon in relation to the esophagus. Motility disorder however is observed as follows.

In megas caused by Chagas disease, the sigmoid colon and the rectum present increased motility with or without associated megaesophagus. The movements are independent in the healthy individual and synchronic in the patient with Chagas disease. The use of pentagastrin in megacolon did not present any response because the neural communication between the hormone and the muscle cell is lost.

The intra luminal stimuli in the esophagus determine, in the healthy individual, rectal contraction through the mioenteric plexus (Rezende et al. 1985).

Despite its frequency in megacolon, faecal impaction rarely causes acute colonic obstruction due to the colon characteristic of retaining large amounts of feces (Lázaro da Silva 1983).

Association with megaesophagus in the advanced phases of the disease, may cause difficulty to prioritize treatment in severe cases. If we correct the megaesophagus first, we may, due to the post-operative ileus, facilitate acute obstruction caused by faecal impaction and normal feeding. Operating on the megacolon first, the upper tract obstruction (esophageal) and the malnutrition remains. We prefer the second option.

**Classification**

Megacolon may be empirically divided as: total megacolon, partial megacolon, megasigmoid, dolichosigmoid, and megadolichosigmoid.

**Surgical operations**

The history of megacolon surgery is long and harsh. The attempted surgical solutions for the problem ranged from curious to exuberant. Among the various authors, 46 methods have been executed.

**Preferred choice**

Duhamel-Grob procedure (transanal rectal pull-through of the colon): (a) lower left paramedian internal pararectus laparotomy; (b) exposure of the descending colon, sigmoid and the cranial portion of the rectum by opening the mesentery anteriorly and laterally; (c) ligation and division of the sigmoid and superior rectal vessels, preserving the vascular arcade parallel to the isolated segment; (d) lacing the sigmoid with a gauze tape occluding the lumen above and below; (e) blunt dissection of the rectum through the rectosacral fascia up to the levator ani muscle; (f) division of the rectum at the level, or slightly below, of the peritoneal fold. Care is taken not to inflict damage to the medial rectal vessels. Sigmoidectomy is carried out protecting the proximal portion of the colon by investing it in a sterile surgical glove; (g) the rectal stump should be sutured with 00 chromic catgut interrupted by two or three layers of invaginating stitches; (h) from the perineum, the rectal sphincter is delicately dilated exposing the pectinate line. Halfway along the circumference of the posterior mucosa is transversely divided at the level of or slightly above the pectinate line; (i) submucosal dissection beyond the level of the levator ani; (j) opening of the posterior rectal wall reaching the previously dissected rectosacral fascia; (k) through this opening the proximal stump is exteriorized; (l) the posterior wall of the colon is sutured, with interrupted sutures of catgut, to the posterior edges of the perineal skin, halfway along the total circumference of the perineal wound; (m) two hemostats are placed in such a way that an inverted "V" is created. These hemostats crush the septum (spur) without going beyond the rectal stump. The referred spur is created by the anterior wall of the lowered colon and the posterior wall of the rectum; (n) if there happens to be only one team doing the operation, at this time surgical gowns and instruments are changed to new sterile ones; (o) the opened peritoneal fold is now reconstructed around the lowered colon; (p) the abdominal wall is sutured in layers.

There are four ways to crush the spur created by the walls of the lowered colon and the rectum: (1) with two long curved hemostats as those used in gynecologic procedures (Lázaro da Silva 1991); (2) two hemostats attached to one median handle only. The objective here is to be the least uncomfortable to the patient as possible during the post-operative period (Zachary-Lister); (3) single wide hemostat of which the external part is taken out (Léger); (4) single wide oval hemostat (Ikeda). In order to prevent the uncomfortable situation brought up for the patient by the placement of the hemostats, Haddad suggests the following modifi-

cation: (5) proceed as in the Duhamel technique up to the phase of the exteriorization of the colon; (6) the colon is lowered through the retro-rectal space and then exteriorized for about 6 cm to 8 cm transanally as if it were a perineal colostomy; (7) suture of the rectal mucosal edge to the serosal layer of the colon; (8) after the 7th postoperative day, the inverted "V" spur corresponding to the "crushed septum" of the original operation, is resected. Interrupted sutures are placed between the rectal ampulla and the divided colon (Lázaro & Tafuri 1975, Lázaro 1991).

#### MEGASIGMOID

Megasigmoid as an isolated form is rare. It occurs at the beginning of the formation of the mega. It is more frequently associated with rectal and left colon dilatations.

The procedures for the acute and the chronic phases of the disease have been previously described.

#### MEGARECTUM

Megarectum isolatedly is also rare. It is usually associated with sigmoidal dilatations. In the hypothesis of its isolated occurrence, the surgical procedure to control the faecal stasis is to defunct the rectum, and among other procedures referred to before, is the transanal retro-rectal lowering of the colon.

#### HYPERTROPHY OF THE SALIVARY GLANDS

Hypertrophy of the salivary glands, specially the parotid, is observed in the digestive forms of Chagas disease, more frequently in the cases of megaesophagus. Apparently, what happens is a denervation of the gland leading to stasis and ductal dilatation. However, there is still some discussion over this topic. For example, the sialographic study conducted by us in patients with megaesophagus from Chagas disease, revealed no alteration in the ductal pattern of the gland. What possibly occurs is a compensatory hyperplasia or hypertrophy of the gland in order to permit better swallowing (Lázaro et al. 1977).

#### MEGA-GALLBLADDER

From a practical point of view, mega-gallbladder as an isolated form does not exist. There can be bile duct dilatations in association with dilatations of other parts of the digestive tract. If it happens that stasis occurs due to dyskinesia of the gallbladder or its association with biliary stones; a cholecystectomy would solve the problem.

If the mega-gallbladder occurs associated to a long peritoneal attachment, torsion and necrosis can happen, leading to an acute abdomen.

This can be more or less severe, depending on the occurrence or not of biliary perforation and peritonitis.

#### MEGA-COMMON BILE DUCT

Dilatation of the biliary tree as an isolated form of mega is also extremely rare. It occurs in association with other dilatations.

Because of the risk of cholangitis and its complications, biliary stasis, due to dilatations from Chagas disease, is very severe.

A bilioenteric bypass is the best solution if the symptoms of biliary stasis (pain, jaundice, infection) exist. This is specially true if a dilatation of the intra or extra-hepatic biliary tree is present. The ideal bypass is an hepaticojejunostomy with an excluded Roux-en-Y loop of jejunum.

For initial cases, a cholecystectomy associated with an ample papilla sphincteroplasty is performed.

#### PERSPECTIVES

There are no promising perspectives for Chagas disease when the stasis and the dilatation present themselves in the form of megas.

Surgeons' knowledge in this subject has already accomplished what can be achieved.

We have observed that the best method is also the most fragile if applied before time. When the prolonged stasis does not persist, the patient resumes the same pattern.

Prevention is, therefore, the only perspective we can propose.

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