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Review

Achalasia During Pregnancy: Proposed Management Algorithm Based on a Thorough Literature Review

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Fewer than 40 cases of achalasia occurring in pregnant woman have been reported in the literature. Given the rarity of achalasia during pregnancy, and the numerous treatment options that are available for achalasia in general, no guidelines exist for the management of achalasia during pregnancy. Diagnosis of new cases may be difficult as symptoms and physiological changes that occur during pregnancy may obscure the clinical presentation of achalasia. The management of achalasia in pregnancy is also challenging. Treatment decisions should be individualized for each case, considering both the welfare of the mother and the fetus. Since pregnant women suffering from achalasia represent a diagnostic and therapeutic challenge with complex maternal-fetal aspects to consider, we have reviewed the available literature on the subject and summarized current diagnostic and therapeutic options. Additionally, we present a management algorithm as a means to guide treatment of future cases. We recommend that a conservative approach should be adopted with bridging therapies performed until after delivery when definitive treatment of achalasia can be more safely performed.
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Key Words
Esophageal achalasia; Disease management; Pregnancy

Introduction

Achalasia is characterized by destruction of the smooth muscle ganglion cells of the myenteric plexus of Auerbach resulting in esophageal body motor dysfunction, incomplete lower esophageal sphincter (LES) relaxation, and progressive esophageal dilation. Clinically, patients complain of dysphagia to solid foods and liquids, heartburn, regurgitation, vomiting, chest pain, and weight loss.[1] Recent data suggests that the disease process involves an interaction between autoimmune and inflammatory responses, possibly triggered by viral infection, in genetically susceptible individuals.[2]

Relationship Between Pre-existing Achalasia and Fertility

Mayberry et al[3] surveyed 36 women with achalasia and compared them to 36 age-matched controls. The study found no difference in the number of conceptions or live births between the groups, including both in the time before achalasia was diagnosed and after it was symptomatic. They concluded that achalasia presented no hindrance to becoming pregnant.[3] A study by Vogel et al[4]

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of 43 women also found no correlation between achalasia and fertility.

A recent study evaluated women that had already undergone peroral endoscopic myotomy (POEM) as treatment for achalasia. They reported that 5 women had become pregnant afterwards (for a total of 7 pregnancies), and concluded that achalasia treated by POEM did not negatively impact future conception.[5]

Relationship Between Pregnancy and the Onset of Achalasia

Recent studies have shown that an 8-residue insertion in HLA-DQβ1 is a genetic risk factor for achalasia. Interestingly, Becker et al[6] found that pregnancy itself may be a disease-triggering factor in women carriers of this insertion. Further investigation of this risk is needed especially as other studies have not found any correlation between pregnancy and the onset of achalasia.[4]

Clinical Presentation of Achalasia During Pregnancy

The diagnosis of achalasia during pregnancy is often difficult because the typical symptoms of achalasia, such as regurgitation and heartburn, can occur early in pregnancy. Frequently, pregnant women presenting with recurrent vomiting are assumed to have hyperemesis gravidarum (HG). Many patients who were eventually diagnosed with achalasia were initially mistakenly diagnosed with HG. Consequently, the diagnosis of achalasia may be delayed until patients present with more advanced disease or life-threatening complications.

Patients with known achalasia can have variable disease courses during pregnancy. A recent study including 18 women with pre-existing achalasia reported that 8 of them claimed that their symptoms worsened during pregnancy, while 7 had no change and 3 had an improvement in symptoms.[4]

An older study by Mayberry et al[3] described 16 women with pre-existing achalasia. They found that 3 women reported that their symptoms worsened during pregnancy, but 11 had no change, and 2 had an improvement.[3]

Several theories have been suggested as to how pregnancy may affect achalasia symptoms. It has been suggested that increased intra-abdominal pressure and elevation of the diaphragm may affect the esophagus. Also, increased progesterone during pregnancy may reduce smooth muscle motility and tone in the gastrointestinal tract. However, these theories have not been proven and remain speculative.[4]

Diagnosis of Achalasia During Pregnancy

Manometry

An assessment of esophageal motor function by high-resolution manometry (HRM) is recommended to confirm the diagnosis of achalasia.[7] It is the gold standard test for confirming the diagnosis of achalasia.[1] Further, HRM can classify achalasia into 3 subtypes (Type 1: classic achalasia with aperistalsis, Type 2: achalasia with panesophageal pressurization, and Type 3: achalasia with spasm) which have prognostic value as outcomes differ with regards to various therapies.[1] While it may be uncomfortable, HRM is a safe diagnostic test during pregnancy, with no risk of teratogenicity. Unfortunately, there are no HRM values specifically for pregnant woman. The only study assessing manometric values in pregnant women is from 1978. This study, using conventional line-tracing manometry, revealed that esophageal peristalsis in pregnant women has lower wave speed and lower amplitude compared to non-pregnant women.[8]

Radiology

The preferred radiological evaluation for achalasia is a barium esophagram. The classic finding on an esophagram is a dilated esophagus with a tapering at the esophagogastric junction ("bird's beak" sign).[1] A timed barium esophagram, in which the height of barium that remains in the esophagus is assessed at various time intervals, may provide evidence as to the severity of the case and predict response to treatment.[1] Other radiographic exams, such as a chest X-ray of CT scan, may show a dilated or tortuous esophagus, or residual food in the esophagus.

The choice of radiological modality in a pregnant woman has to take 2 factors into consideration; the accuracy of the test and minimizing least radiation exposure to the fetus. According to the American College of Obstetricians and Gynecologists' Committee on Obstetric Practice, with few exceptions, radiation exposure through radiography and CT scans is at a dose much lower than the exposure associated with fetal harm. They conclude that if these tests are necessary, they should not be withheld from a pregnant patient.[9]

Endoscopy

The primary role of endoscopy in the workup of achalasia is to exclude a mechanical obstruction causing pseudoachalasia.[2] Endo-

scopic findings in achalasia may range from a seemingly normal examination to a tortuous dilated sigmoid-shaped esophagus, retained food and secretions, and ulcerations secondary to stasis or candida infection. Endoscopy is relatively safe for the fetus and may be performed when strongly indicated during pregnancy.[10,11] Fetal risks from endoscopic medications can be minimized by avoiding Food and Drug Administration category D drugs and anesthesiologist attendance at endoscopy.

Management of Achalasia

The main goal of all existing therapies in achalasia is to improve esophageal emptying, relieve symptoms, and prevent long-term complications. In addition, many patients with achalasia present with malnutrition due to their symptoms. In pregnant patients, where fetal development is negatively impacted by malnutrition, ensuring adequate nutritional support is especially important. Therapeutic options for non-pregnant patients can be divided into nutritional support, medical treatment, endoscopic treatment, and surgical treatment. Case reports using these various treatment options are summarized in Table 1,[12-39] while these modalities themselves are summarized in Table 2.

Nutritional Support Treatments

Parenteral Nutrition

Parenteral nutrition (PN) is a means of maintaining or restoring nutrition via an intravenous route when oral enteral routes are not feasible.[40] PN during pregnancy has been used most often to provide adequate nutrition for those who suffer from prolonged hyperemesis or who have difficulty absorbing adequate nutrients because of such conditions such as Crohn's disease.[41]

Sufficient clinical experience suggests that PN is a relatively safe and effective method for reversing maternal malnutrition and promoting normal fetal growth and development. Pregnancy outcomes among 122 women diagnosed with HG who received total parenteral nutrition support during early pregnancy showed a decreased risk for perinatal morbidity.[42] PN has proved to be helpful in malnourished pregnant women and promotes fetal growth.[43] It can be used as long as needed throughout the pregnancy, until delivery of the baby.

Potential complications of PN include line complications (occlusion, sepsis, venous thrombosis, and pulmonary embolism) and cholestasis, which may be associated with sepsis.[44] Given the risks, there should be a clear need for PN before using this modality.[45]

Nasogastric Tube Feedings

Nasogastric tube (NGT) intubation for feeding during pregnancy is generally well tolerated by the mother, with only rare and mild maternal complications, and with mostly favorable fetal outcomes.[46] Insertion of the NGT may be more difficult in a patient with achalasia, and endoscopic assistance may be needed.

Percutaneous Endoscopic Gastrostomy Feedings

As an alternative, Godil and Chen[47] described 2 cases of percutaneous endoscopic gastrostomy (PEG) placement in 2 conscious pregnant women with HG. However, PEG tubes do not reduce the symptoms or risks of aspiration of salivary retention. PEG placement also requires the use medications including sedatives, painkillers, and antibiotics, all of which may have risks to the mother or fetus. For all of these reasons, PEG does not appear to be a promising option in pregnant women, and there are no reports of its use in pregnant women with achalasia.

Medical Treatments

Oral Pharmacological Therapies

Pharmacological treatment for achalasia is aimed at reducing LES pressure, thereby facilitating passage of esophageal contents into the stomach. The 2 most commonly employed medications are calcium channel blockers and long-acting nitrates, both category C pregnancy risk.[48] Sildenafil, a phosphodiesterase-5-inhibitor (Category B), is another option.[49] Less commonly used medications include anticholinergics (Category B and C), β-adrenergic agonists (Category C), and theophylline (Category C). In addition to risks to the fetus, the efficacy of these medications is generally poor and there are potential side effects for the patient. Use of these medications for treatment of achalasia is rare nowadays.

Endoscopic Treatments

Botulinum Toxin Injection

Botulinum toxin type A (BTX-A) is a purified neurotoxin from the bacterium *Clostridium botulinum*. It is used to treat a variety of conditions of increased muscle tone, including achalasia, where it is injected into the LES.[50] Since BTX-A is a large protein with a high molecular weight (150 kDa), when injected locally in recom-

Table 1. Case Reports of Achalasia in Pregnant Women With the Type of Treatment and Pregnancy Outcome

Publication (Author and year)	Patient age (yr)/ gestation (wk)	How achalasia was diagnosed	Type of achalasia treatment	Delivery and outcome
Roques, 1932[12]	25	BE	BD	Preterm labor with fetal and maternal deaths
Roques, 1932[12]	37	BE, EGD	BD	Termination at 16 wk.
Lindert, 1956[13]	32	BE	No treatment	Cesarean section
Stroup, 1961[14]	27	EGD, BE	Medical treatment	Normal spontaneous delivery
Bloomfield, 1963[15]	35	EGD, BE	Medical treatment	Cesarean section
Karjalainen, 1964[16]	23 (1st pregnancy)	EGD, BE	Medical treatment	Termination
Karjalainen, 1964[16]	23 (2nd pregnancy)	EGD	Medical treatment	Premature labor, spontaneous birth
Clemendor et al, 1969[17]	34 (1st pregnancy)	BE, Manometry	BD	Premature labor with fetal death
Clemendor et al, 1969[17]	34 (2nd pregnancy)	As above	Medical treatment	Normal spontaneous delivery
Clemendor et al, 1969[17]	22	BE, Manometry	BD, PD	Normal spontaneous delivery
Satin et al, 1992[18]	28/38	BE, EGD, Manometry	PD	Induced vaginal delivery at 38 wk. Healthy baby
Fiest et al, 1993[19]	24/8	BE, Manometry	PD	Spontaneous at 35 wk. Healthy baby
Faloon, 1993[20]	26/prenatal	BE	BD	Cesarean section at 36 wk. Healthy baby
Fassina Osculati, 1995[21]	23/24	Autopsy findings	No treatment	Unexplained sudden maternal death, megaesophagus
Aggarwal et al, 1997[22]	20/18	BE, EGD, Manometry	PD	Spontaneous abortion in 7th month
Kalish et al, 1999[23]	42/31	EGD	PN	Spontaneous at 38 wk. Healthy baby
Ohno et al, 2000[24]	34/27	BE, Manometry	No treatment	Intrauterine fetal death
Ghoshal and Davies, 2007[25]	19y/33	Manometry	NGT (1500 kcal/day)	Induced labor at 37 wk. Healthy baby
Pulanic et al, 2008[26]	30/26	Not described in report	PD	Spontaneous at 38 wk
Palanivelu et al, 2008[27]	24/2nd trimester	EGD, BE	LHM	Spontaneous. Healthy baby
Díaz Roca et al, 2009[28]	36/26	Not described in report	SEMS	Uneventful delivery
Wataganara et al, 2009[29]	39y/33	EGD	BTI	Cesarean section at 35 wk
Paulsen et al, 2010[30]	34/33	CT scan, EGD, Manometry	PD	Uncomplicated birth
Khandelwal and Krueger, 2011[31]	22/15	BE, Manometry, EGD	PN, nifedipine	Cesarean section at 34 wk. Twins Intrauterine fetal death of 1 twin; healthy 2nd twin
Spiliopoulos et al, 2013[32]	38/29	EGD, Manometry	PN (1215 kcal/day)	Cesarean section at 37 wk. Healthy baby
Hooft et al, 2015[33]	23/14	Manometry	BTI	Spontaneous. Healthy baby
Orth, 2015[34]	30/34	CT scan	BTI	Spontaneous at 38 wk. Healthy baby
Holliday and Baker, 2016[35]	17/31	MRI scan, BE	BTI	Spontaneous at 37 wk
O'Leary et al, 2016[36]	28/32	Not described in report	NJT	Cesarean section at 37 wk
Neubert and Stickle, 2019[37]	28/22	Manometry	BTI	Healthy baby at term
Lora Acuña et al, 2019[38]	26/8	EGD, BE, Manometry	NGT, PD	Cesarean section at term
Narang and Narang, 2019[39]	35/11	EGD, BE	NGT	Intrauterine fetal death
Vosko et al, 2021 (the present study)	28/29	EGD, BE	PN	Induced vaginal delivery at 34 wk. Healthy baby

BE, barium esophagram; BD, Bougie dilation; EGD, esophagogastroduodenoscopy; PD, pneumatic dilation; PN, parenteral nutrition; NGT, nasogastric tube; LHM, laparoscopic Heller myotomy; SEMS, self-expanding metal stent; BTI, botilinum toxin injection; CT, computerized topography; MRI, magnetic resonance imaging; NJT, nasojejunal tube.

Table 2. Summary of the Benefits and Complications of the Different Modalities for Achalasia Treatment in Pregnancy

Type of treatment	Benefits	Possible complications to the mother and technical difficulties unique for pregnant patient	Possible complication to the fetus	Best optional gestational time for procedure
PN	Safe for baby	Line sepsis (~17%) Deep vein thrombosis (~8%) Catheter occlusion Catheter dislodgment Pneumothorax Electrolyte and trace elements disturbances High patient compliance needed	No reported complications	Any trimester
NGT	Safe for baby and mostly safe for mother	Tubes are easily misplaced or dislodged Perforation of nasopharynx, esophagus, and stomach Transbronchial insertion Aspiration Electrolytes and trace elements disturbances	No reported complications	Any trimester
BTI	High response rate in the 1st month (80-90%) Short procedure time	Miscarriage (category C medication) Mediastinitis Allergic reaction to an egg-based protein Repeated BTIs—subsequent submucosal fibrosis that might make invasive therapies more difficult	Possible abortion or fetal malformations which have been observed in rabbits	Any trimester Therapeutic effect wanes rapidly over time Best results are at the 2nd and 3rd trimester of pregnancy to avoid repeating treatments
PD	Effective nonsurgical option Success rate (50-93%) Short procedure time Short recovery time	Perforation (0-5%) GERD (15-35%)	In case of perforation all the possible surgical complications including general anesthesia complications	Any trimester
SEMS	Safe and effective (83-100%)	Migration (5.3%) Chest pain (38.7%) GERD (20%) LES fibrosis that might make invasive therapies more difficult	No reported complications	Any trimester
POEM	Highly effective with possibly the best sustained success rates (short-term 90-100%, 3 years 88.5%, 5 years 83%)	Perforation Bleeding Risk of anesthesia GERD	Unclear	Unclear
LHM	Success rate (88- 98%)	Poor visualization due to gravid uterus Uterine injury Technical difficulties Decreased uterine blood flow Risk of general anesthesia GERD (14.9%)	Premature labor from the increased intra-abdominal pressure Increased fetal acidosis or other unknown effects of CO_2 pneumoperitoneum	2nd trimester —lowest risk of teratogenesis, preterm delivery, or miscarriage

PN, parental nutrition; NGT, nasogastric tube; BTI, botulinum toxin injection; PD, pneumatic dilation; SEMS, self-expanding metal stents; POEM, peroral endoscopic myotomy; LHM, laparoscopic Heller myotomy.

mended doses, it is not expected to enter the systemic circulation or reach the maternal-fetal interface.[51] Endoscopic ultrasound may facilitate localization with direct intramuscular injection, avoiding spillage into blood vessels.[52]

Newman et al[53] were the first to report 4 full-term uncomplicated pregnancies in a patient with severe cervical dystonia who

received BTX-A treatment, without any effect on the pregnancy outcome. Literature reviews have concluded that exposure to botulinum toxin injection (BTI) during pregnancy does not appear to increase the risk of adverse outcome in the fetus.[54,55]

In non-pregnant patients with achalasia, treatment with BTI is often reserved for patients with high surgical risk, short life expectancy, and those who are not candidates for pneumatic dilation (PD), POEM, or surgical myotomy.[56] However, in pregnant women with achalasia, BTI may serve as a bridge by temporarily improving symptoms until a more permanent treatment can be performed after delivery of the baby. There are several case reports of BTI being used in this way in the literature (Table 1).

Self-expanding Metal Stents

The temporary use of self-expanding metal stents (SEMS) has been reported in the management of achalasia and other benign esophageal diseases. Complications include migration (5.3%), chest pain (38.7%), and reflux (20.0%), but no perforations or 30-day mortality were reported in achalasia cases.[57-59] Only 1 case report exists of SEMS use in a pregnant woman with achalasia. Díaz Roca et al[28] described a pregnant 36-year-old woman diagnosed with achalasia during her 28th week of gestation. They chose to treat her with SEMS placement and achieved successful relief of symptoms, weight gain, and spontaneous childbirth. The stent was then removed without complication.[28] Despite this case, the lack of corroborating data, concern over stent migration, and need for fluoroscopic guidance make SEMS placement unattractive for use in pregnancy.

Pneumatic Dilation

PD is an effective non-surgical procedure for the treatment of achalasia.[1,60] The only randomized comparative study between PD and surgery, carried out by the European Achalasia Trial Investigators Group in 2011, showed similar results for both techniques over a follow-up period of 2 years.[61] A meta-analysis found that both techniques, PD and laparoscopic Heller myotomy (LHM), were effective in the treatment of achalasia.[62] Candidates for PD should be those for whom surgery is not contraindicated taking into account that the most severe complication of this technique is esophageal perforation, which occurs in approximately 1.9% (range 0.0-16.0%).[63,64]

In 1969, Clemendor et al[17] were the first to report a successful case of PD for achalasia during pregnancy, which was performed at 24 weeks gestation, followed by an uneventful delivery of a healthy baby at 36 weeks. Prior to that, several cases of Bougie dilation had been reported. Since then, 6 additional cases of PD have been reported (Table 1). In all of these cases healthy babies were delivered suggesting that PD is an effective and safe therapeutic option. With 7 cases reports, there is more evidence to support the use of PD than any other treatment modality.

No studies have clearly shown what the optimal balloon size for PD is in achalasia patients overall, let alone in pregnant ones; nor are there studies comparing balloon sizes used in PD for non-pregnant versus pregnant achalasia patients. Therefore it is not clear what size balloon should be used for PD in pregnant women. Using the smallest size (30 mm diameter) balloon may decrease the risk of perforation in these high-risk patients, although this may also limit the therapeutic benefit.

Peroral Endoscopic Myotomy

POEM is quickly becoming a safe and effective treatment option for patients with achalasia with better efficacy rates compared to surgery.[65,66] As the effectiveness of POEM has been established over the past few years, and its complications are comparable to or better than that of surgery, it may be an effective treatment for achalasia during pregnancy. However, at the current time, we are hesitant to comment on its safety or efficacy during pregnancy as there are no case reports of its use in pregnant women with achalasia. As POEM continues to become more widely practiced, it may only be a matter of time until it is used in such a patient. The risks of POEM include perforation, as well as the need for intubation and anesthesia, all of which add to the risk of performing it in a pregnant woman.[65]

Surgical Myotomy

The timing and the type of surgery are 2 important issues to consider before any surgery during pregnancy due to concerns over fetal maturity and maternal safety. In general, surgery is safest in the second trimester because the risk of spontaneous abortion or preterm delivery is less than 5% at this time. During the first trimester, there is a risk that anesthetic medications may impair fetal organogenesis and lead to abortions in up to 12%. During the third trimester, surgery may precipitate labor in up to 40% of patients.[67]

Palanivelu et al[68] reported on 19 different laparoscopic surgical procedures performed in pregnant patients during the second trimester, including one case of LHM after the patient had no response to PD. All the laparoscopic procedures were successful without mortality, morbidity, or complications for either mother or child.[68] However, others emphasize that pregnant women remain at risk for perinatal

complications, including fetal loss, and that surgeries should be postponed until after delivery if at all possible.[69] The case by Palanivelu et al[27] remains the only case report of LHM during pregnancy. Although the patient did well and eventually delivered a healthy baby, there remains insufficient evidence to recommend LHM during pregnancy at the present time.

Case Presentation

Some of the diagnostic and therapeutic dilemmas discussed above are illustrated in the following case report. A 28-year old woman was referred to our institution at 29 weeks of gestation due to frequent vomiting since the start of her pregnancy. Endoscopy revealed a dilated esophagus containing food without any strictures or masses (Fig. 1). She was treated conservatively for achalasia, but was re-admitted 2 weeks later due to inability to eat and recurrent hematemesis. After a discussion of the therapeutic options in pregnancy, she opted for total parenteral nutrition until delivery. She was admitted again at 34 weeks of gestation with pre-eclampsia. Labor was induced, and she delivered a healthy 1.75 kg baby by vaginal delivery.

At 5 weeks postpartum, the patient underwent LHM with Dor fundoplication. However, the treatment provided only a few weeks of relief. A subsequent barium esophagram revealed significant delay in esophageal emptying and a tapered lower esophagus ("bird's beak" sign). Upper endoscopy showed a dilated sigmoid-shaped esophagus with retained food and secretions. She had continued worsening of her symptoms with a 17-kg weight loss. Approximately 6 months later, she was admitted urgently with severe hematemesis. Computed tomographic scan revealed food extending from the esophagogastric junction upwards to the proximal esophagus (Fig. 2). Upper endoscopy revealed a dilated esophagus with food and bleeding from stasis ulcerations. She underwent esophagectomy (Fig. 3) with a gastric pull-through, and a jejunal feeding tube was placed. Eight weeks later, she had good oral intake and the tube was removed.

Figure 1. Esophagogastroduodenoscopy revealed a dilated esophagus containing food, but no mass or strictures.

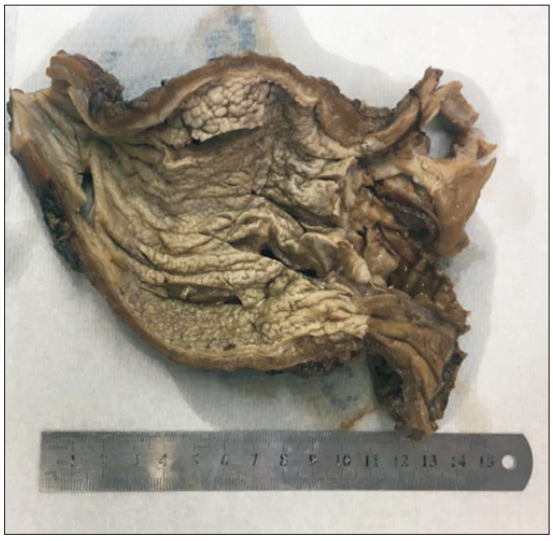

Figure 3. Esophageal resection specimen showing megaesophagus.

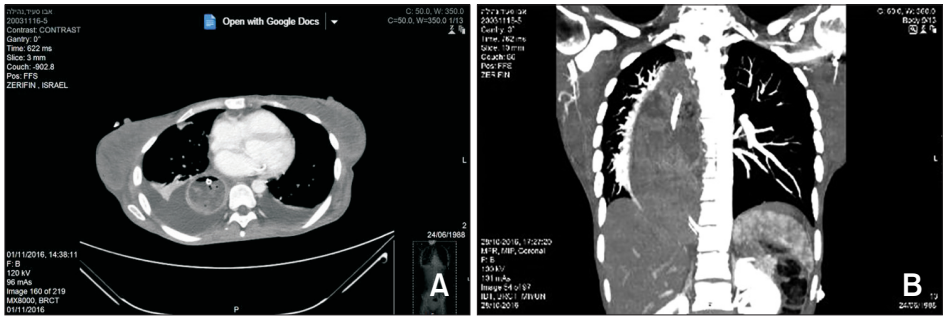

Figure 2. Computed tomographic imaging revealed megaesophagus with food extending from the proximal esophagus to the esophagogastric junction. (A) Cross-sectional view. (B) Coronal view.

Proposed Therapeutic Management Algorithm

Given the rarity of achalasia in pregnancy, and the various treatments available for achalasia in general, no guidelines exist for the management of achalasia in pregnancy. Therefore, we recommend that the decision should be individualized for each case, considering both the welfare of mother and fetus. Individual factors to be considered include the age of the patient, nutritional status, gestational age, surgical risk, comorbidities, subtype of achalasia, patient preference, esophageal anatomy, and the local expertise available. Most of the recent case reports describe good outcomes for both mother and baby, although this may reflect an element of publication bias.

In Figure 4, we propose an algorithm for the management of achalasia based on the available literature. In general, treatment should be aimed at maintaining the patient's nutritional status throughout pregnancy with the goal of delivering a healthy baby at term. This should be achieved through the use of bridging treatments to reach this goal. Definitive treatment of achalasia should ideally be performed after completion of a successful pregnancy.

For patients with inadequate nutritional intake during pregnancy, NGT feedings seem to be the safest option and is recommended as an initial therapy for nutritional replacement and support. The main concern with NGT use is its tolerability, especially for prolonged periods. For those who do not want or cannot tolerate a NGT, PN can be a reasonable treatment option. PN is a relatively safe and effective method for reversing maternal malnutrition and promoting normal fetal growth and development, although it is not free of complications.

If significant achalasia symptoms persist, BTI appears to be a safe bridging modality until delivery of the baby. In patients who refuse BTI or for whom BTI has failed, PD is an alternative as it has more successful cases reported in the literature than any other modality. Given the risk of perforation, it should be carried out in hospitals with experienced gastroenterologists and surgeons.

Finally, if bridging modalities do not provide adequate relief, LHM can be considered, especially during the 2nd trimester. However, since it is an elective procedure with all of the risks of surgery, we recommend that LHM should be delayed until after delivery if at all possible. Given the growing evidence that POEM is as effective and safe as LHR, it may also be an option in pregnant achalasia patients requiring definitive treatment. However, as there have been no reports of its use in pregnant women to date, we cannot definitively recommend its use at this time.

Adequate monitoring of the therapeutic response to achalasia treatment is also limited by pregnancy. Performing additional invasive procedures, such as post-treatment barium esophagram, endoscopy, or manometry, should be avoided if possible. Instead, therapeutic success should be based on clinical parameters such as symptom improvement, weight gain, and fetal development.

Conclusions

Achalasia during pregnancy is a rare clinical situation. Pregnant women suffering from achalasia represent a diagnostic and

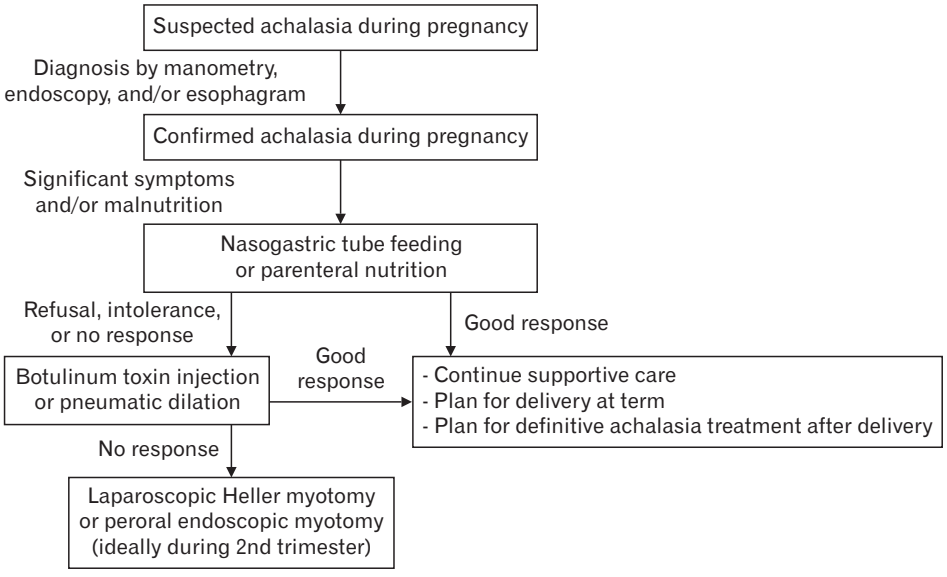

Figure 4. Proposed algorithm for the management of achalasia during pregnancy based on a thorough review of the literature.

therapeutic challenge as a delay in the diagnosis and treatment can have serious implications for maternal health and fetal viability. A multidisciplinary team including gastroenterologist, obstetrician, dietician, radiologist, and surgeon is needed for decision making. A thorough discussion with the patient of the expected risks and benefits for each treatment modality is required, with special concern for her wishes regarding fetal safety. In general, a conservative approach should be adopted with bridging therapies performed until after delivery when definitive treatment will be safest.

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