

## **Protocols and Best Practices for Treating Diaphragmatic Endometriosis**

***Keywords: Endometriosis of the lungs, chest, diaphragm, upper abdomen***

**By Camran Nezhat, M.D., Farr Nezhat, M.D., Ceana Nezhat, M.D.**

Lately there have been many discussions reminding us of just how crucial it is to eradicate endometriosis as completely as possible. As a surgical imperative, this is actually a time-honored principle, one that has been observed since at least the late 19th and early 20th centuries, when progenitor pioneers like J. Marion Sims, [Thomas Cullen](#), and [John Sampson](#) found that patients were more likely to experience relief from symptoms when as much disease as possible was removed. [Sims was so committed to this principle that he even used his own fingernails to “excise” endometriosis](#) that was otherwise difficult to remove with the standard instruments of his day! (1 , 2 )

However, implicit in some of these narratives is the flawed assumption that endometriosis returns as a result of an incomplete and/or inadequate method of treatment (i.e., ablation/excision) from a previous surgery, While it’s certainly true that inexperienced surgeons might treat endometriosis inadequately, what’s equally true, if not more so, is the fact that endometriosis can recur despite the most thorough treatment possible.

In other words, all the medical interventions in the world, by all the world’s greatest practitioners, [cannot prevent endometriosis from returning in certain cases](#). Since these and other similar misconceptions about endometriosis have become rather prevalent lately, we thought it was a good time to bring these issues into the conversations.

### **Diaphragmatic Endometriosis: Commonly misdiagnosed form of endometriosis**

Since we are on the topic of thoroughly treating endometriosis, we would like to add to the discussion the issue of [diaphragmatic endometriosis](#), a potentially debilitating extragenital form that actually is sometimes overlooked and left behind. (3 ) In fact, diaphragmatic endometriosis is likely one of the most commonly overlooked forms of the disease. There are many reasons why this is the case; gendered medicine, the rarity of this form of endometriosis, and outdated beliefs that endometriosis only occurs in the lower pelvic region are all contributing factors. However, from a surgeon’s standpoint, another reason that diaphragmatic endometriosis can be overlooked so easily is because it has a propensity for hiding behind the liver and other structures of the upper abdomen, making it difficult to visualize from the traditional surgical field. (3 )

Even though diaphragmatic endometriosis is rare relative to other forms of endometriosis, nevertheless it’s absolutely critical that health care practitioners include it in their differential diagnosis, so that it can at least be ruled out. If these atypical forms of endometriosis continue

to be left out of differential diagnostic assessments, we'll never be able to gain an accurate understanding of their true incidence. As well, another entire generation of women and girls would potentially suffer needlessly for years and years, without a proper diagnosis and without adequate medical care.

As part of our [ongoing global campaign to end the silence about endometriosis](#), we've written this article with the hope that it will help reduce diagnostic delays by encouraging the general public, as well as primary care physicians, school nurses, and other health care professionals, to heighten their level of clinical suspicion about endometriosis, including atypical manifestations of the disease like diaphragmatic endometriosis.

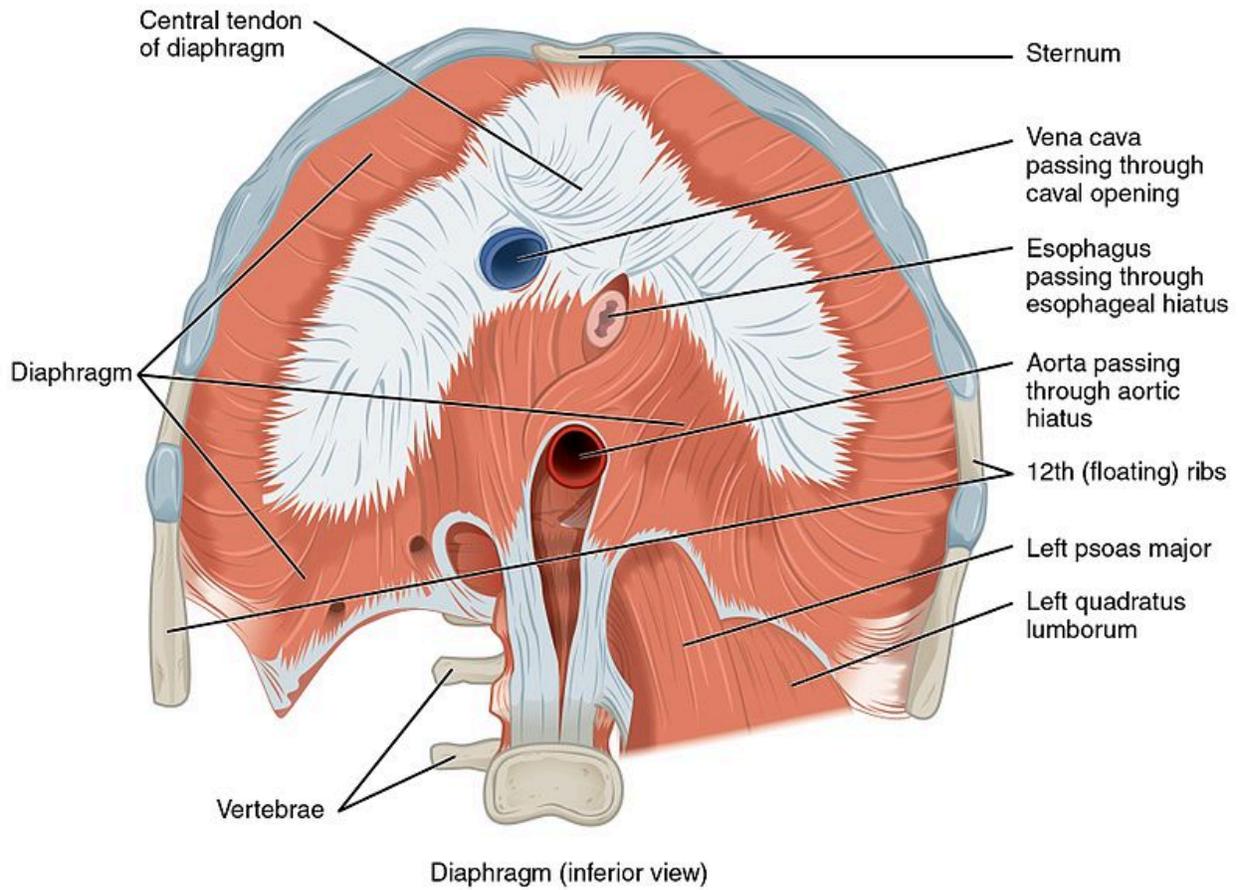
### **Quick overview of the diaphragm's anatomical features**

For those who might want a quick anatomy refresher course, we thought the following review of the [thoracic diaphragm's anatomical features](#) would be a good place to start. One of the most important muscles for human respiration (aside from the lungs), the diaphragm is a long and thin convex organ located below the heart and lungs, attaching to the lower ribs, sternum, and lumbar spine, and forming the base of the thoracic cavity. Positioned as it is, it conveniently separates the chest cavity from the abdominal cavity. Lengthwise it ranges from 6 to 12 inches, while its thickness usually measures somewhere between one-quarter to one-eighth of an inch.

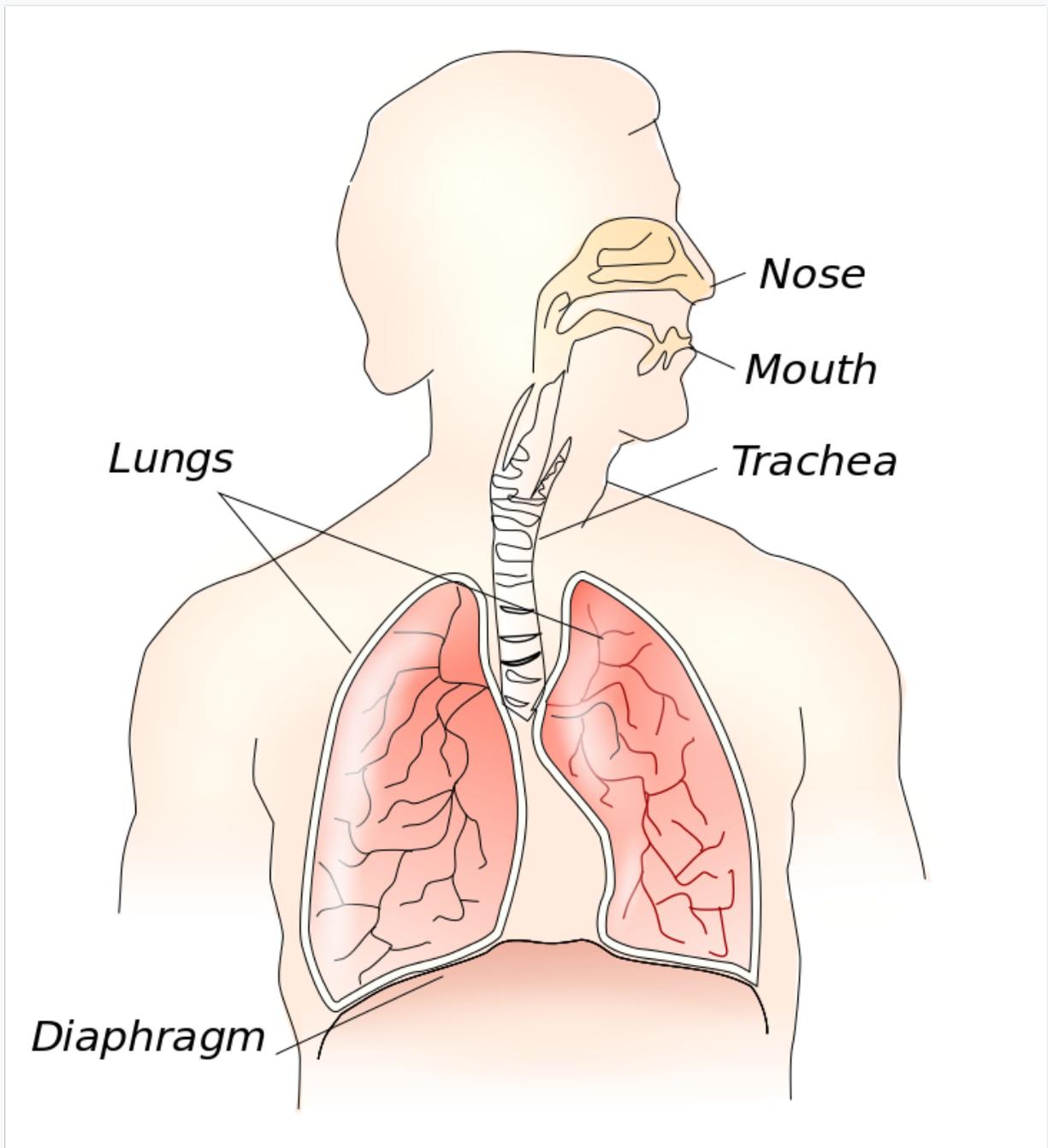
With its symmetrical, lobe-like formation, the diaphragm's right and left sides are distinctly demarcated (though still contiguous). For this reason, many physicians refer to the two lobes as the right and left hemidiaphragms. The diaphragm's main innervating source is the [phrenic nerve](#), which in turn is fed by the cervical nerves C3, C4, and C5. (Many of our physician friends will probably remember the mnemonic, "C-3, 4, 5, keep the diaphragm alive.") However, there are also two peripheral innervating sources called the intercostal and subcostal nerves, located at T5-T11 and T12, respectively.

Highly flexible, the diaphragm contracts and relaxes as breathing takes place. Specifically, when a person inhales the diaphragm and its intercostal muscles contract downward, thereby widening the thoracic cavity in which the lungs are contained. As a result, air fills the lungs through the mechanical process of negative pressure suction. When a person exhales, the diaphragm and intercostal muscles relax, thereby mechanically forcing air out of the lungs.

F



**Figure 1: Thoracic Diaphragm, [image source courtesy of Version 8.25 Textbook OpenStax Anatomy and Physiology Published May 18, 2016.](#)**



**Figure 2: Thoracic Diaphragm, [image source courtesy of Version 8.25 Textbook OpenStax Anatomy and Physiology Published May 18, 2016.](#)**

### **Most Common Symptoms of Diaphragmatic Endometriosis**

As mentioned, symptomatic diaphragmatic endometriosis can potentially cause severe and debilitating symptoms. The classic symptoms of diaphragmatic endometriosis are pain in the chest area (pleuritic, usually on the right side), [shortness of breath \(dyspnea\)](#), epigastric pain (upper GI pain), pain in the right shoulder, and upper abdominal pain (right or left side). Many patients report that these symptoms can occur suddenly, with severe and sharp stabs of pain radiating throughout the upper abdomen, chest, and shoulder. However, it's important to remind patients and health care professionals alike that diaphragmatic endometriosis can be asymptomatic; that is, some women experience essentially no symptoms at all, or only very vague ones that are difficult to pinpoint.

Another critical point to remember is that, while symptoms usually occur cyclically with the onset of menses, there are many patients who experience symptoms acyclically. In fact, studies have shown that up to 30% of women with pelvic endometriosis experience acyclical symptoms that do not neatly coincide with menstruation. It is these atypical clinical presentations like acyclic symptomology that can really throw some health care providers off the trail of a correct diagnosis. That's why we urge the medical community to maintain a high level of clinical suspicion for thoracic forms of endometriosis, especially for patients who have already been diagnosed with pelvic endometriosis and even if symptoms are atypical.

### **Collapsed lung and other potentially serious co-morbidities**

The rarer but more serious and potentially life-threatening conditions of [catamenial pneumothorax](#) (collapsed lung occurring cyclically with menstruation), [hemopneumothorax](#) (air and blood in the chest cavity), chest wall lesions, and [lung parenchyma](#) can also cause similar symptoms. These potentially serious conditions may occur concomitantly with diaphragmatic endometriosis, due to the development of fenestrations (perforations or holes) in the diaphragm, caused by the necrotizing effects of the endometriotic lesions. ( 4 , 5 ) In terms of risk factors, studies show that Fifty to 84% of women diagnosed with thoracic endometriosis have concomitant pelvic endometriosis.

Conversely, the congenital presence of right hemidiaphragm fenestrations may contribute to the development of diaphragmatic endometriosis, as well as in surrounding areas of the chest, lung, and pleural cavity ([thoracic endometriosis syndrome or TES](#)). And, although rarer still, with fewer than 20 cases reported in the literature to date, [liver \(hepatic\) endometriosis](#) should also be a part of differential diagnoses, as it too can cause cyclic upper abdominal pain. ( 6 , 7 )

### **Appearance**

In terms of macroscopic appearance, lesions can present with various colors and morphologies. However, lesions that have a bluish cast appear to be among the most commonly reported in the literature (see Figure 3 below), followed by lesions with a reddish-purple appearance.



**Figure 3: Bluish pigmented lesions most commonly reported form of diaphragmatic endometriosis.**

#### **Other common features**

As you may already know, most diaphragmatic lesions – approximately 95% - occur on the right side of the diaphragm, a propensity which many of today's experts cite as the central supporting evidence for Sampson's retrograde menstruation theory of pathogenesis. However, lesions have been found on all parts of the diaphragm, including on the left side only, on both sides (bilaterally), and/or on surrounding structures like the [phrenic nerve](#). In other cases, the majority of lesions may even present anteriorly, where they lie hidden behind the liver. Given such a wide variation in locations, the entire diaphragm and adjacent areas must therefore be thoroughly investigated.

#### **Complete excision reduces recurrence**

The good news about diaphragmatic endometriosis is that, after thorough surgical treatment, studies have found that it is far less likely to recur than other forms of the disease. (4 ) While

most endometriotic implants that do occur on the diaphragm turn out to be superficial and cause no discomfort, others can be deeply infiltrating, including cases of full thickness infiltration. In one of our studies, 29% of our patients had deeply infiltrating implants. (8 ) In more serious cases, such as full thickness infiltration, as mentioned, the endometriotic growths can cause multiple small perforations which would need to be surgically repaired.

### **Preliminary steps for determining a treatment plan**

For those who are not symptomatic but who have been diagnosed with any of these extragenital forms of endometriosis incidentally, some physicians recommend to take an expectant approach with no further intervention, unless symptoms later occur. (9 ) However, for those patients who are symptomatic, [surgery has proven beneficial](#) when other medical interventions have otherwise failed. (7 , 10 , 11)

If proceeding with surgery, however, one must take care to counsel patients well about their other options because injury can occur to the diaphragm, phrenic nerve, lungs, vessels, or heart. And, as with any other procedure, care should be individualized, taking into consideration the patient's goals, age, medical condition, etc, as well the practitioner's experience and availability of appropriate inter-disciplinary consultants.

### **A few key protocols in surgical management**

While diaphragmatic endometriosis presents many unique challenges, we thought we would focus on just a few key protocols that we've relied on over the years to help us navigate through these especially tricky anatomical landscapes.

The first issues to address are concerns about access and visualization of the upper abdomen and the diaphragm's entire surface. We would like to urge our community to not let these particular concerns be reasons to resort to laparotomy, which is known to most likely cause the painful and potentially intestinal-obstructing thick vascular sorts of adhesions in more than 93% of cases. (12 , 13) [By resorting to laparotomy](#), one treatable problem would be exchanged for a potentially disabling one.

To avoid all of the potentially serious complications that laparotomies may come with, the first step is to consult with an endoscopic surgeon experienced in upper abdomen procedures. Surgeons with sufficient experience will be able to utilize several different techniques and devices to help obtain a clear view of the diaphragm and adjacent areas. For example, the patient's position can be changed during surgery, so that the diaphragm's surrounding organs fall away from it enough so that one can see parts that would be otherwise obscured. As well, surgeons can add more port holes (the tiny incisions in which the laparoscopes are inserted) in

the upper abdomen so that better visualization can be obtained.

More advanced surgeons can even rely on highly specialized techniques, such as resecting the [falciform ligament](#) or using the laparoscopes to gently move the liver and stomach away from the diaphragm so that it can be better visualized. In fact, we routinely deploy these and other minimally invasive strategies in oncological gynecology, when it's often necessary to debulk diaphragmatic metastatic disease, which sometimes requires partial resections of the diaphragm and mesh placements. (14)

As for [excisional techniques](#), it depends on many factors, including the nature of the lesion itself. However, the most essential consideration is complete removal of endometriosis. Randomized controlled studies have shown that thorough eradication of the disease is the only variable consistently associated with any measureable reductions in pain and recurrence rates. (15 , 16) And, contrary to common misconceptions, it makes no difference which technique is utilized; vaporization, ablation, hydro-dissection, and surgical scissors excision have all proven to be effective methods capable of complete excision.

There are also many devices that can be used for complete excision, all of which have their own advantages and disadvantages. For example, compared to monopolar instruments, the CO2 laser has proven especially safe and useful for excising or otherwise eradicating both superficial and deeply infiltrating endometriosis, since it poses no danger of having its energy heat arc out in erratic ways. As a result, laser surgeries (performed by experienced surgeons) are associated with substantially fewer incidence of inadvertent injury to surrounding healthy tissue or organs. (17 , 18 , 19)

[This video of a laparoscopic diaphragmatic stripping](#) being performed for ovarian cancer demonstrates several techniques which can also be used for treating diaphragmatic endometriosis.

### **Outcomes dependent on skill of surgeon and proper instrumentation**

Of course, surgical outcomes ultimately depend entirely on the skill and experience of the surgeon. This is especially true for those who specialize in treating endometriosis, a field of such complexity and ever-changing paradigms that the only hope we have for staying ahead of such a fast-moving curve is to continuously advance our knowledge in the latest surgical tools and techniques. [The availability of proper instrumentation](#) has also proven to be critical, for no matter how great of a pianist you are or want to be, you can't play if you don't have a piano or if the one you've got is utterly out of tune.

### **Postoperative care**

Postoperative medical care depends on whether patients are planning to conceive. For those wishing to become pregnant, there are no further postoperative medical interventions, other than the routine postoperative care and perhaps [Assisted Reproductive Technologies \(ART\) treatments](#). For patients who are not planning to conceive, they may continue with some sort of hormonal suppressive treatment.

### **Collaboration with other sub-specialists is crucial**

All of these protocols will prove nearly meaningless, however, without the element of multi-discipline collaboration, especially critical for treating such a multi-organ, systemic disorder as endometriosis. For example, even though we were the first to report on the [laparoscopic treatment of liver \(hepatic\) endometriosis](#) and have, to our knowledge, performed more of these surgeries than any other surgeon or institute, we still have had only three such cases after over three decades of experience. For this reason, we've always advocated seeking out collaboration with experts from other fields.

The same goes for [endometriosis of the lungs, chest wall, and diaphragm](#), which collectively have accounted for no more than 40 of our cases over the years. Again, even though these 40 cases represent the first and largest number ever treated laparoscopically, it's still a comparatively small volume relative to the other types of surgeries we routinely perform. Given the rarity of such cases, collaboration with other sub-specialists is therefore an absolute imperative for the safety and well-being of patients.

### **Final thoughts**

We hope we've been able to provide some helpful perspectives on some of gynecology's most complex and contested issues. For further reading on these subjects, please see our list of references below.

### **REFERENCES**

- 1) [Nezhat C. Nezhat's History of Endoscopy: A Historical Analysis of Endoscopy's Ascension since Antiquity. Tuttlingen, Germany: Endo-Press; 2011.](#)
- 2) [Nezhat Camran; Nezhat, Farr; Nezhat, Ceana. Endometriosis: ancient disease, ancient treatments. Fertil Steril. 2012 Dec;98\(6 Suppl\):S1-62.](#)
- 3) [Nezhat, Camran; Nicoll, Linda M; Bhagan, Lisa; Huang, Jian Qun; Bosev, Dorian; Hajhosseini, Babak; Beygui, Ramin E. Endometriosis of the diaphragm: four cases treated with a combination](#)

[of laparoscopy and thoracoscopy. Journal of Minimally Invasive Gynecology. 2009; 16\(5\):573-80.](#)

4) [Nezhat C, et al. Bilateral Thoracic Endometriosis Affecting the Lung and Diaphragm - Case Report. JSLS. 2012; 16\(1\):140-142.](#)

5) [Endometriosis: Science and Practice. Linda C. Giudice, MD, Johannes L. H. Evers, David L. Healy. John Wiley & Sons. 2012.](#)

6) [Nezhat, C. et al. Laparoscopic management of hepatic endometriosis: report of two cases and review of the literature. Journal of Minimally Invasive Gynecology. 2005; 12, 196-200.](#)

7) [Nezhat F, Nezhat C, Levy JS. Laparoscopic Treatment of Symptomatic Diaphragmatic Endometriosis: A Case Report. Fertil Steril. 1992; 58\(3\): 614-616.](#)

8) [Nezhat's Operative Laparoscopy, 3rd ed. Cambridge Press. 2005. p 289-291.](#)

9) [Falcone T, Lebovic DL. Clinical Management of Endometriosis. Obstet Gynecol. 2011;118\(3\):691-705.](#)

10) [Nezhat, C. C., Seidman, D. D. S., Nezhat, F. F. & Nezhat, C. C. Laparoscopic surgical management of diaphragmatic endometriosis. Fertil Steril. 1998; 69: 1048-1055.](#)

11) [Mangal, R. R., Taskin, O. O., Nezhat, C. C. & Franklin, R. R. Laparoscopic vaporization of diaphragmatic endometriosis in a woman with epigastric pain: a case report. Journal of Reproductive Medicine. 1996; 41: 64-66.](#)

12) [Parker MC, Ellis H, Moran BJ, Thompson JN, Wilson MS, Menzies D, McGuire A, Lower AM, Hawthorn RJS, O'Brien F, et al. Postoperative adhesions: Ten-year follow-up of 12,584 patients undergoing lower abdominal surgery. Diseases of the Colon & Rectum. 2001; 44\(6\):822-9.](#)

13) [Redwine DB. Diaphragmatic endometriosis: diagnosis, surgical management, and long-term results of treatment. Fertil Steril. 2002; 77\(2\):288-96.](#)

- 14) [Nezhat FR, DeNoble SM, Liu CS, Cho JE, Brown DN, Chuang L, Gretz H, Saharia P. The safety and efficacy of laparoscopic surgical staging and debulking of apparent advanced stage ovarian, fallopian tube, and primary peritoneal cancers. JSLS. 2010 Apr-Jun;14\(2\):155-68.](#)
  
- 15) [Healey M, Ang C, Cheng C. Surgical treatment of endometriosis: a prospective randomized doubleblinded trial comparing excision and ablation. Fertil Steril 2010; 94:2536-40.](#)
  
- 16) [Wright J, Lotfallah H, Jones K, Lovell D. A randomized trial of excision versus ablation for mild endometriosis. Fertil Steril. 2005; 83\(6\):1830-6.](#)
  
- 17) [Sutton CJ, Pooley AS, Ewen SP, Haines P. Follow-up report on a randomised controlled trial of laser laparoscopy in the treatment of pelvic pain associated with minimal to moderate endometriosis. Fertil Steril 1997; 68: 1070-4.](#)
  
- 18) [Luciano AA, Whitman GF, Maier DB, Randolph JF, Maenza RM: A comparison of thermal injury, healing patterns and postoperative adhesion formation following CO2 laser and electromicrosurgery. Fertil Steril.1987;48:1025-1029.](#)
  
- 19) [T, Vilos GA: A comparison between laser surgery and electrosurgery for bilateral hydrosalpinx. Fertil Steril. 1985; 44: 846.](#)