

Patient Perspective and Advocacy Tools

Perhaps you are experiencing cyclical chest, shoulder, neck, scapular pain or shortness of breath. Perhaps you are wondering if you have endometriosis on/within any structures involved with respiration. The intent of this article is to provide a better perspective of what the disease is, how it can present and, if concerned, things that you can document and track for discussion with a care provider.

Endometriosis affects 10% of the female population. Among this 10% of women, a smaller portion of them will be affected by endometriosis lesions that involve organs other than gynecological. This is referred to as 'extra-pelvic' disease. The prevalence of 'extra pelvic' endometriosis is estimated to affect 12% ^(1,2) to 20% ⁽³⁾ and was reported as high as 37% percent in an older study of 181/485 subjects with endometriosis of the GI system ⁽⁴⁾ of those with endometriosis. A conservative estimate range of 12 -20% of those with endometriosis (or 1:5), will develop lesions in non-gynecological tissues and organs. These locations of extra-pelvic disease include the digestive, excretory, urinary and respiratory systems' and other less reported areas. ^(1,2,3)

This paper will focus on one region of extra-pelvic disease: the thorax and respiratory system. There have been no large epidemiology studies conducted in regards to the disease, however recent estimates place prevalence at 2.3-5.6% of all women with Endometriosis may have TE. ⁽⁵⁾ However, it can be assumed these estimates are conservative. The fact that many women with disease are mis- and delayed diagnosis, assuming other diagnosis (ie. primary spontaneous pneumothorax, general pleurisy, chest wall musculoskeletal dysfunctions, gastrointestinal, biliary and other concerns), suggest these statistics are less than true prevalence of disease.

There are many theories in regards to the origins of Thoracic Endometriosis (TE). Based upon scientific findings, it is supported that more than one etiology and pathological process leads to TES. No single theory can explain the myriad of presentations.⁽⁶⁾ As mentioned, TES includes a vast array of manifestations. Numerous structures can be affected: diaphragm; parietal pleura lining the interior of chest wall and thoracic side of the diaphragm; visceral pleura lining the exterior surface of the lung parenchyma; lung parenchyma with its air sacs and bronchiole airway system; lymphatic and arterial vessels, lymph nodes, and pericardial sac of heart. The location of lesions determine symptoms and manifestations. This is important to understand the necessity of diligent documentation by the sufferer of where, how, when and description of any symptoms experienced. If a Pneumothorax, Pleural/Hemothorax or coughing of blood has occurred, any imaging records can be important clue to determine location of lesion(s). Very unique to TE, although there are gradients of symptoms, there are also a variety of end manifestations that occur, which differs from lesions of the abdominopelvic cavity. Uniquely, all (5) varieties of presentations (CP, CHt, CH, Nodules and Catamenial Cyclic Chest Pain) are all collectively considered 'Thoracic Endometriosis' (TE). Despite different complications there are specific attributes that consistently present across the disease spectrum.

The structures that comprise the respiratory system are located within or define the borders of the chest cavity. These structures have shared innervations. As a result, tissue(s) present with symptoms that are felt in the same location(s) as other structures with the same innervation patterns. These sensations may be felt far from the site (referred) or directly over the involved area(s). A specific example of this: gallbladder dysfunction and endometriosis lesions or adhesions of the diaphragm all refer pain to the right shoulder areas. To complicate things

further, in addition to other organs outside of the chest, musculoskeletal structures can also refer pain to the same areas and must be taken into consideration (ie gastrointestinal system). An example: Intestinal adhesions can restrict the normal peristalsis of the bowel. As a result, a person may not experience 'cramps' in a typical manor but, instead, may feel a deep ache within the chest. These sensations can be the result of irritated free nerve endings within the fascia lining the abdominal cavity and intestines. If adhesions are present, the fascia's restricted mobility and distortion causes irritation of the free nerve endings along the stress lines emitting various symptoms. Another example is complaint of heartburn and circumferential/substernal pressure which is often associated with hiatal hernia and GERD but may also be a sign of diaphragmatic endometriosis. Hence documentation noting any relation to meal times etc. As discussed later, careful documentation of symptoms characteristics is helpful to determine which tissues are involved. In regards to fascia mentioned previous, the entire abdominal-pelvic cavity is lined with this continuous tissue. It can be described as 'slimy' and covers the muscles and all the organs inside the body. Fascia plays an important role in the mobility between organs and tissues to reduce friction. The diaphragm, at the top, and the pelvic floor at the bottom, have a lot more connection that most would believe. Imagine a knitted sweater that has a 'snag' in one area. What happens when that 'snag' is pulled on? The area around the 'snag' becomes more and more distorted. The same effects occur within the body. A 'snag' along the pelvic sidewall can lead to discomfort along the ribs and up into the diaphragm due to the closed 'barrel' encased in fascia and wrapped in overlapping muscles through the torso to support and protect these structures.

Diligent documentation of your symptom characteristic(s) and behavior(s) with corresponding timeline can be an asset when approaching your provider with concerns. This is further detailed below.

Thoracic Endometriosis (TE) involves any of the following: Diaphragm (lots of pain fibers), Parietal Pleura (the inside lining of chest wall - lots of pain fibers), Visceral Pleura (a continuation of the same tissue (parietal) that lines the chest wall but no pain fibers and 'coats' the outer lung surface; Lung Parenchyma (spongy lung tissue that includes the air sacs-no pain receptors) and Bronchi airways (no pain receptors).

The diaphragm is most commonly involved tissue followed by the visceral and pleural surfaces, lung parenchyma and bronchi with endometriosis. Lesions are more commonly documented on the right side. However, left and bilateral involvement does occur in all structures. With increased awareness and more studies reported worldwide, there are recent trends of left and bilateral involvement recorded. However, the right side remains predominant. The tissue(s) involved in the disease determine the clinical manifestation(s) collectively referred to as Thoracic Endometriosis Syndrome (TES). These include: Catamenial Pneumothorax (CP-73%), Catamenial Hemo (pneumo) thorax (CHt-14%), Catamenial Hemoptysis (CH-7%) Isolated Chest Pain, Lung Nodules^(7,8) and even smaller percentage of Catamenial Pericardial Effusions.⁽⁹⁾ However, it must be clarified that not all lesions will progress to these end manifestations. A 2016 review study "revealed catamenial pneumothorax as the most common (manifestation) of TES, however, when the catamenial chest pain/pleurisy is distinguished from documented pneumothorax, pneumothorax was responsible for only 40% of symptoms, while chest pain was

seen in 80% of cases".⁽¹⁰⁾ This demonstrates that although the majority of women present with chest pain, arising from numerous structures, the development of CP still remains very low. This also reiterates that Catamenial Cyclical Chest Pain (CCCP) is the most common of the (5) listed manifestations. (CCCP) is now considered a precursor to CP. Not all persons with diaphragm disease will suffer from progressive, deep disease however, some who do may develop full thickness disease and, with sloughing, lead to compromised integrity of the diaphragm that creates an open airspace between the peritoneal and thoracic cavities. With onset of menses and cervical plug dissolving, air is theorized to enter the cervix and contribute to spontaneous pneumothorax. This is one of multiple ways a pneumothorax can develop with this disease.

Although the odds are very low to develop catamenial pneumothorax (CP), hemothorax (CHt) or hemoptysis (CH), presence of symptoms justifies pursuit of a skilled surgical team who can identify and excise all abnormal tissues observed to further reduce the risk of progression.

The location of lesion(s) present with different 'clusters' of signs and symptoms. Some areas of involvement have overlapping, similar symptoms. It is important to document the days of your symptoms in relation to your menstrual cycle. If you do not experience a menstrual cycle due to menopause, HRT, OCP or other more aggressive hormonal use it is still important to document the dates. Notable things for documentation: **Where** you feel the symptoms, **What** the symptoms feel like (short breath, hard to breathe in, versus out, burning, deep ache, sharp, throb, stab, pressure, bubbles etc. Can you **Hear** anything unusual? Crackling, gurgles, clicking? Are you able to **Relieve** the symptoms by change of positions, time of day, antacids, change of diet, over the counter medications, use of warm or cold etc.? Does anything **Aggravate** your

symptoms: cool air, specific movements, foods, specific time after meals or change of positions influence the location, pain level and type of sensations you experience. The use of a 'key' with symbols placed on the body diagram is a great tool to give visual representation of what you experience. This is an easy way for providers to read and interpret information rapidly.

Example symbols to consider include: X-throb; circles –stab; dots-bubbles;

thunderbolts=electric shock/nerve pain; compressed lines-pressure; shading-deep ache; flame-hot/burning, etc. Other symptoms to report may include: heartburn, blood-tinged sputum with coughing, shortness of breath or difficulty breathing, choking, swallowing difficulty,

lightheadedness, fatigue, nausea etc. Body Diagrams can be downloaded from Google Images.

However, www.endostats.com provides an excellent template with diagram and calendar. It can be accessed in the submenu 'Cyclic Symptoms'. It is ideal to track symptoms for 3 months.

This will help you determine if patterns are present. Consider providing copies of your calendars and body images with key. They are a great self-advocacy tool. Make sure to put your name and date of birth on all papers and request them to be included in your medical record. This may even precipitate providers to listen and pursue your concerns in greater detail without being sent away, downplayed or completely dismissed of your concerns. From the same website you can download a pdf that highlights all the major areas of extra-pelvic disease: www.endostats.com/drs.notes.html

If you have a spontaneous pneumothorax, a recurrence ALWAYS justifies investigation.

However, research is attempting to establish screening questionnaires to those with catamenial/ovulation related cyclical chest pain BEFORE the first incident or after first occurrence designed to identify and make 'probable clinical' diagnosis of disease. ⁽¹¹⁾ The

purpose is to aid justify for earlier inspection of the chest cavity with minimal invasive techniques before disease progresses. As awareness and clinical investigative techniques into TES has increased, the earlier prevalence estimates that endometriosis contributes to 3-6% of all pneumothoraxes among females is considered a great underestimate. ^(11,12) Most recent reports estimate 1 of every 3 females with a pneumothorax are the result of endometriosis. ^(7,11,14) Approximately 25% of all lung collapses in females occur during the 'catamenial' time frame. A number of time periods have been used to define the 'catamenial' period, however the most universally recognized time frame is: '24 hours prior-to and up-to 72 hours after menses onset'. ^(5,11,16) In addition to the 25%, another 9-10% of women who suffer pneumothorax(es), outside of the 'catamenial' time period is estimated to be the result of endometriosis. These estimates are based upon results of multiple large scale studies. This brings the average estimates of thoracic endometriosis-related pneumothoraxes (TERP) to 1 of every 3 occurrences. ⁽¹⁶⁾

As awareness and interest in TES has increased and minimal invasive surgery (MIS) to the chest cavity have emerged, the population of females with signs or symptoms suggestive of TE has expanded. It was initially presumed that any spontaneous pneumothorax occurring outside of the 'catamenial' time frame was not likely a result of endometriosis. However, studies are presenting objective findings to refute this claim. As a result, it is estimated that a smaller portion of women (9-10%) who suffer a pneumothorax outside of the 'catamenial' time period, are considered thoracic-endometriosis-related-pneumothoraxes (TERP).⁽¹¹⁾ Another evolution into identification of women affected by endometriosis is occurring. Some investigators are not only questioning the existence of TE among women with signs and/or symptoms suggestive of

its presence, who initially present with pelvic disease; Some investigators have followed up more closely with women whose initially presents with manifestations of TES (CP, CH, CHT) whom may not have been previously identified with pelvic disease present. Subsequent interviews with these women revealed other health complaints previously overlooked prior to their collapsed lung, coughing blood or hemothorax.

The early and increased frequency of investigation through advent of MIS and Video-Assisted Thoracic Surgery (VATS) has also increased the number of women identified with the disease that would go un/misdiagnosed. With an expanding population under investigation, the patient profile of women who present with TES continues to adjust. Among these profile adjustments include the rate of co-morbid abdominopelvic endometriosis among women with TE. Co-morbid prevalence ranges have been reported to range between 20% to 100% of subjects in larger sample studies. ^(11,17-21) The evolution of invasive procedures and awareness has also improved recognition of TES in adolescent women. Many adolescent females are misdiagnosed with a Primary Spontaneous Pneumothorax (PSPT). This often occurs due to lack of awareness and that the most common profile of PSP occurs in the younger population with pneumothorax while pneumothoraxes associated with endometriosis occurs at a slightly older age (mean age 30-34 yrs). Awareness, progressive understanding of the disease and MIS has expanded the age range of females with TERP to include adolescent females. ⁽²¹⁻²³⁾ and reported frequency of left and bilateral thoracic involvement has increased with greater awareness of the disease, however the right chest cavity remains most frequently side of involvement.

Lastly, it is important to communicate any history of pelvic surgery or other invasive gynecological procedures. These may include: Dilation and Curettage for miscarriage (D&C), abortion or abnormal bleeding secondary to polyps or endometrial hypertrophy and extraction of a retained placenta. There is an abnormally high frequency of women who experience Catamenial Pneumothoraxes or Catamenial Hemoptysis with a history of pelvic surgery or uterine scraping. ⁽¹¹⁾

Each case is unique. Establishment of a care team that understands and respects the various theories of pathogenesis and can perform the appropriate procedures to remove all visible disease and implement preventive measures to reduce manifestations of any 'invisible' disease is vital. 'Invisible' disease is not 'missed' disease in this regard. Similar to all other areas where endometriosis occurs, conservative intervention with hormone manipulation has a very low rate of resolution when discontinued. Discontinued treatment of hormone therapy has consistently reported high rates of CP, CHt, or CH recur.^(17,20,23,24) Excision of all visible lesions is the best intervention available. However, the lungs and chest cavity contain a vast amount of surface area that, even with advanced imaging, still 'hides' lesions with timing, motion and composition still variables that make the disease evasive. With its large surface areas, minute lymphatic/vascular structures and minute airways that are smaller than high resolution MRI, surgeons often use preventive measures to reduce risk of future manifestations. Some of these include peeling the pleural linings off (pleurectomy) and/or adhering the inner and outer layers of the lung to the chest wall (mechanical or chemical pleurodesis).

This paper presented an overview of Thoracic Endometriosis Syndrome, its prevalence and manifestations. A few examples of other conditions which can create symptoms within the upper torso were also mentioned. The importance of medical history and symptom documentation, with attention to specific details as supportive information to share with your care provider has also been provided.

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