

Highlights from our 4th Annual DFW Hereditary Colon Cancer Conference
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The 4th annual DFW Hereditary Colon Cancer Conference was held in early March. We were pleased that several families from the DFW area participated. A few of the enlightening and well-presented lectures are summarized below:

Overview of Hereditary Colon Cancer Syndromes-

C. Richard Boland, MD –*Chief of Gastroenterology at Baylor University Medical Center Dallas*

About 32% of patients with colon cancer have a family history of colon cancer. About 3% have Lynch syndrome (LS) and 1/7000 have Familial Adenomatous Polyposis (FAP). The remaining colon cancer syndromes make up an even smaller fraction of patients with colon cancer.

Familial Adenomatous Polyposis (FAP)- Patients with FAP have 100's-1000's of adenomatous polyps. The average age of onset is 16 and the median age for colorectal cancer is 40 years of age. The chance to develop colorectal cancer is nearly 100%. FAP is caused by mutations in the APC gene and is inherited in an autosomal dominant manner. There is a subset of mutations in the APC gene that cause attenuated FAP. These individuals develop on average 25 adenomas and have a 70% lifetime risk to develop colon cancer.

MYH-Associated Polyposis- MYH mutations are similar to APC mutations in that they cause adenomatous polyposis. Generally, patients with MYH mutations develop 50-200 and develop colorectal cancer at younger ages than the general population. About 1.3% of the general population has one MYH mutation. However, MYH-associated polyposis is one of the few cancer syndromes that is inherited in an autosomal recessive manner. Therefore, patients will not develop MYH-associated polyposis unless they have two MYH mutations.

Lynch Syndrome (LS)- Unlike the cancer syndromes described above, patients with LS generally do not have polyposis, rather they may have up to a couple dozen polyps. LS is inherited in an autosomal dominant manner and is caused by mutations in the DNA mismatch repair genes. A recent article described the method used to determine which MMR genes to test for. However, patients who meet Amsterdam criteria are considered to have LS, regardless of whether a genetic mutation can be found.

Amsterdam Criteria:

1. At least 3 relatives with LS associated cancers (colorectal, endometrial, ovarian, stomach, small bowel, and ureter cancers)
2. One must be a first degree relative of two others
3. At least two successive generations
4. At least one <50 with cancer
5. FAP is excluded

Families meeting Amsterdam criteria in which there are no extra-colonic cancers present, and in which no germline genetic mutation can be found, are considered to have Familial Colorectal Cancer type X. These families are not thought to be at high risk for extra-colonic cancers. For families meeting Amsterdam criteria, it is preferable when a genetic mutation can be found. Finding a genetic mutation in

patients makes it possible for cancer-free family members to be tested to determine whether they have LS as well.

Interestingly, colorectal cancer patients who have LS tend to have a better overall survival compared to colorectal patients without LS.

Surgical Role for Hereditary Colorectal Cancer Patients-

Glen Balch, MD –*Division of Surgical Oncology, UT Southwestern Medical Center*

To better prepare for life after surgery, it helps to understand bowel function: The small bowel absorbs nutrients whereas the colon is used to absorb water. The rectum absorbs water in addition to storing stool.

For patients who are at increased risk of colorectal cancer, surgery can provide therapeutic benefits and/or prophylactic benefits. The following are procedures commonly used:

Segmental colectomy or hemicolectomy (partial removal of colon)

- Advantages- preserves function
- Disadvantages- potential for second cancer; potential for second operation; still requires colonoscopy with risk of missing polyps.

Total colectomy with ileorectal anastomosis (fusing of the ileum to the rectum):

- Advantages- removes risk of colon cancer
- Disadvantages- rectum remains; including risk of cancer; more frequent bowel movements; loose stools

Total proctocolectomy with ileostomy (rectum is removed and small bowel has opening on abdominal wall):

- Advantages- removes risk of colon and rectal cancer
- Disadvantages- higher output of waste; potential for dehydration and electrolyte imbalance

Total proctocolectomy with J-pouch (a pouch is created from the ileum, forming a J-shape, and fused with the internal anal sphincter):

- Advantages- removes risk of colon and rectal cancer
- Disadvantages- 5-6 bowel movements per day; possible passive incontinence; higher complication rate; requires two operations.

Psychosocial Aspects of Colon Surgery-

Laura Howe-Martin, PhD –*Assistant Professor, Department of Psychiatry, UT Southwestern Medical Center*

In order to help minimize stress both in the short and long term, it is important to address the following early in the process:

RELATIONSHIP WITH PROVIDER:

- *Knowledgeable*- Determine whether the physician is knowledgeable about the condition

- *Shared Perspective*- Does the physician share the same perspective as the patient?
- *Improve Health Literacy*- Address the problem, what needs to be done to fix the problem, and why it is important to address the problem

SURGERY PREPARATION:

- *Consider nicotine cessation program*
- *Update list of medications*
- *Know the risks, benefits, and alternatives*
- *Prepare to be a patient*- there will be some loss of dignity and privacy. Be aware of how you will look and feel when you wake up after surgery

ACUTE PAIN MANAGEMENT:

- *Sleep is important*
- *Discuss nausea treatment with health care providers*

POST-OPERATIVE SUPPORT:

- *Possible stigma and embarrassment, and reluctance to ask questions or self-disclose*
- *Why and how*- continue to ask 'why' and 'how' problems should be solved
- *Fear*- anticipate feelings of fear when nearing discharge

Generally patients experience a sharp mental and physical decline after surgery, followed by steady improvement over the next year. Most experience 'total recovery' in all realms. However, complaints about social restrictions and/or sexual intimacy concerns tend to persist.