HEREDITARY COLORECTAL CANCER REGISTRY: A CLEVELAND CLINIC FOUNDATION EXPERIENCE

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INTRODUCTION

The Hereditary Colorectal Cancer registry is one of the oldest and largest registries of its kind. It cares for patients with all hereditary syndromes of colorectal cancer using the three basic approaches of patient care, education and research. This article summarizes the structure and function of the registry, and gives examples of its contributions to the management of affected patients.

ACTIVITY

In 2016 the registry served over 1000 families with FAP, 224 families with Lynch syndrome, 61 with MYH associated polyposis and 146 with one of the hamartomatous polyposes. In 2016 there were 1009 patient visits with 80 new patients and 879 endoscopies. Over 60 surgeries were performed.

SUMMARY

the Cleveland Clinic approach to hereditary colorectal cancer is described. This is multidisciplinary, involving several specialties and both genetic counseling and mental health services within the registry.

Key words: Hereditary colorectal cancer, registry.

INTRODUCTION

Registries have a very important role in the management of patients and families with syndromes of hereditary colorectal cancer. They literally save lives through effective surveillance and expert clinical care (1,2). Patients with these syndromes deserve to be cared for at a registry, or at least by experts with experience and expertise in the management of these syndromes.

The Hereditary Colorectal Cancer Registry at the Cleveland Clinic began in 1979, when Dr. David G. Jagelman established a registry for patients and families with Familial Adenomatous Polyposis (FAP). In the subsequent 38 years the registry has grown to become the largest single institution Hereditary Colorectal Cancer registry in the world. Making use of the high number of patients and families, the Clinic registry has been a leader in developing clinical practice guidelines for managing patients with hereditary colorectal cancer syndromes. During its existence, the Registry has undergone many changes; in knowledge, technology and personnel. In 1988 Dr. Jagelman moved to the new Cleveland Clinic satellite hospital in Florida. Dr Church took over direction of the registry when he arrived at the Clinic in 1989. In 2017 the leadership was transferred to

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another colorectal surgeon, Dr Matthew Kalady. Over the years the genotypes of most of the major syndromes of hereditary colorectal cancer have been discovered. While some syndromes remain a puzzle, we are now able to offer genetic testing to families with suggestive phenotypes. The technology of DNA sequencing has changed, leading to the introduction of Next Generation Sequencing (3). This allows Multigene Panel Testing, using large panels of genes that cover all the known syndromes and some peripheral genes as well. Multigene Panels are quicker and cheaper than the old single gene Sanger sequencing, and can lead to surprising results. This has heightened the need for genetic counseling and for an understanding of the biology of colorectal carcinogenesis. There are at least 10 known hereditary syndromes of colorectal cancer, where we can offer genetic testing and sound clinical management. Because these syndromes are rare, multigenerational and familial, and because affected patients need complex multidisciplinary care for life, a central repository of information is essential so that care can be organized. This central repository of information is **the registry.**

DEFINITION

In simple terms a registry is a list; in the medical world this is often a list of patients. A **Hereditary Colorectal Cancer Registry** is a list of names; names of patients with a hereditary colorectal cancer syndrome and the names of their relatives. Other facts about these patients and relatives can be stored to facilitate clinical care, to allow education of the families, and to enable clinical research. As information technology advances, databases have been designed for use in this setting. The Cleveland Clinic designed its own database, Cologene™, for use as a translational tool to store family pedigrees as well as clinical information, and to use the information to generate patient appointments letters and to perform research. The database is in use at several Hereditary Colorectal Cancer Centers around the world.

STRUCTURE

Most Hereditary Colorectal Cancer Registries arise around one or two key individuals, with special interest and expertise in the area. The St Marks Polyposis Registry, the original registry founded in 1929, arose around the surgical skills of John Percy Lockhart-Mummery and the pathology expertise of Cuthbert Dukes (4). The Johns Hopkins Registry was founded in 1973 by Victor McKusick, the "father" of American Medical genetics and the Creighton Registry in Omaha, Nebraska by a medical oncologist named Henry Lynch. There is evidence of the founder in many registries and there is usually a special expertise or interest, such as surgery, or medical genetics, or gastroenterology. In 2004 we surveyed all the 18 registries for Hereditary Colorectal Cancer in the USA, and described a wide range of definitions and methods. The most striking finding was the small number of registries available to the potentially large number of affected patients and families. The effects of this lack of registries can be seen in the unfortunate outcomes of treatments performed by those with little experience. In Cleveland the registry was based on the interest of David Jagelman, a colorectal surgeon who had trained at St. Marks Hospital and had noticed the benefits of the registry there. The Cleveland registry was therefore centered on colorectal surgical expertise, but over the years expert Gastroenterology, Pathology, Genetics, General Surgery, Endocrinology, Urology, Psychology, and Gynecology services have been added.

A Registry therefore starts with an interested clinician who sets about organizing a registry coordinator. The Coordinator is the "go between" between patients/families and the registry staff. A Coordinator receives referrals, gathers records, arranges appointments, and is the contact between the registry and the patient/family. Registries need a database, an office, and some organizational support. Above all, they need funding. Registries can run on a tight budget but some support needs to come from the host institution. Registries generate income by attracting referrals, seeing patients and families on a regular basis, and need appointments for their patients with multiple specialties. To this extent they generate their own support. The current structure of the Cleveland Clinic Registry is shown in figure 1. Because of the large number of patients and families the syndromes are divided into those involving polyposis and those without polyposis. The "Florida branch "of the Registry shares the Cologene database but patients are identified according to Institution so that duplicate records are not created.

PROCESS

The mission of the Hereditary Colorectal Cancer Registry at the Cleveland Clinic is to prevent death from syndrome-related cancer while maintaining quality of life. The essence of fulfilling this mission is excellence in patient care. This involves timely and accurate diagnosis, effective surveillance, and appropriate endoscopy and surgery. In addition to patient care, the fulfillment of the mission involves education of patients, families and healthcare providers, and the performance of relevant research. These three components of the registry process are detailed in table 1.