

## CJON WRITING MENTORSHIP PROGRAM ARTICLE

# Understanding Lynch Syndrome: Implications for Nursing

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Colorectal cancer (CRC) is the second-leading cause of cancer-related death in the United States. Approximately 10% of CRC is hereditary, and hereditary nonpolyposis CRC (HNPCC), or Lynch syndrome I, is the most common form. Lynch syndrome I is characterized by onset at an early age, poor differentiation, predominance of proximal tumors, and an excess of synchronous and metachronous tumors. In Lynch syndrome II, patients exhibit Lynch syndrome I features and also have extracolonic cancers. Lynch syndrome is an inherited autosomal dominant disorder caused by a germline mutation in one of several genes responsible for DNA mismatched repair. Amsterdam I criteria, Amsterdam II criteria, and Bethesda guidelines are the international diagnostic criteria for Lynch syndrome. Nursing care for patients with Lynch syndrome includes identifying patients who would benefit from genetic counseling, providing education, and assessing and meeting patient psychosocial needs.

**S**.C. was a Caucasian, married woman, who was 62 years old at the time of her death. She underwent genetic testing in a large university setting in response to a personal and family history of cancer developing at a young age. Her genetic results showed high microsatellite instability (MSI) and the expression of the wild-type hMSH2 protein. She was diagnosed with hereditary nonpolyposis colorectal cancer (HNPCC) (i.e., Lynch syndrome). Her personal cancer history includes colon cancer in the cecal region and ascending colon, which was diagnosed at age 55. At age 60, S.C. developed transitional cell carcinoma of the renal pelvis. She had a history of a prophylactic hysterectomy and oophorectomy at age 44. The family history included multiple family members over five generations who developed cancer at a young age. Some family members had multiple cancers (see Table 1).

S.C. was assisted and supported by a caring family, which included a spouse, son, daughter, and sister. They were knowledgeable regarding Lynch syndrome, particularly the family's manifestations of the illness. The family viewed the nursing staff as a source of support and education when new and recurring issues surfaced. They were concerned particularly about how specific treatments would affect the disease process and how genetic forms of the cancers compared to sporadic cases. The family expressed concerns and worries about other family members who had Lynch syndrome or who chose not to be tested. They did not express blame or anger toward the woman's maternal ancestors, although they were aware that this was the lineage by which the syndrome initiated. When S.C. developed her second hereditary cancer, she was hopeful that she would survive as she had with her first cancer. She also was encouraged because her sister had survived multiple cancers. However, when she and her family became aware that she

### At a Glance

- ◆ Hereditary nonpolyposis colorectal cancer (HNPCC) is an inherited autosomal dominant disorder caused by germline mutations in one or several genes responsible for DNA mismatched repair.
- ◆ Patients with HNPCC may have Lynch syndrome I, which is characterized by a susceptibility to colorectal cancer or Lynch syndrome II. Lynch syndrome II exhibits Lynch syndrome I features and extracolonic cancers.
- ◆ Nurses have a significant role in providing assessment, education, and counseling to patients with HNPCC and their families.

was entering the final stages of her life, they chose to accept palliative care and were cared for and supported by hospice nurses. The woman died in a graceful, dignified manner.

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