Update from the New Zealand Familial GI Cancer Service

Dear Colleagues

In view of some of the changes at the NZ Familial GI Cancer Service and in the world of familial GI cancer over the past few years, we thought a letter outlining these may be helpful.

Firstly we changed our name from Registry to Service. The rationale for this change was to reflect the fact that now, in addition to assessing and “registering” families (for whom we co-ordinate surveillance) we are increasingly being asked to provide management advice for individuals and families, registered or otherwise.

In addition we now have branches in Auckland, Wellington and Christchurch. A/Prof Susan Parry, Gastroenterologist, continues as our National Clinical Director/ Medical Advisor in Auckland with Mr John Keating, Colorectal Surgeon our Medical Advisor in Wellington and Dr Teresa Chalmers-Watson, Gastroenterologist and Mr Chris Wakeman, Colorectal Surgeon, our Medical Advisors in Christchurch.

Lynch Syndrome

Aspirin as a chemopreventative agent—We are now recommending aspirin, if appropriate, to all those who carry a Lynch Syndrome mismatch repair gene mutation. This follows the Lancet 2011 publication (Vol. 378:2081-87) of the first randomised trial of aspirin as a chemoprevention agent with cancer as the primary end point (CAPP2 study). The long-term data showed no significant difference in time to first colorectal cancer in the intention to treat analysis. However, the per-protocol analysis revealed a significant difference with a hazard ratio of 0.41 (p=0.02) providing clear evidence of the effectiveness of aspirin as a chemo-preventative agent in this group via a delayed effect on CRC. We have opted to recommend low dose Aspirin (100-300mg) rather than the high dose aspirin used in the trial because of the potential side effects. The CAPP 3 trial which will help to answer this question is due to start shortly.

Interval cancers—Despite frequent colonoscopic surveillance, interval colorectal cancers are documented in Lynch Syndrome patients. Unfortunately in NZ over the last year we have had three such CRC’s in Lynch Syndrome patients undergoing annual colonoscopy – these procedures have been performed in both large and small centres and by highly regarded endoscopists.

A paper by Vasen H et al in Gastroenterology 2010;138:2300 -2306 documented a cumulative CRC risk of 6% after 10 year follow-up of 205 families with Lynch Syndrome undergoing 1-2 yearly colonoscopy. Parry S et al in Gut. 2011 Jul;60(7):950-7 reported the cumulative metachronous CRC risk for 332 MMR gene mutation carriers undergoing regular colonoscopy, following a segmental resection for CRC, to be 16% at 10 years, and 41% at 20 years. This compared with no metachronous CRC in those who had extensive colectomy as the initial cancer operation.
This highlights the need for great care when undertaking surveillance colonoscopy in patients with proven or presumed Lynch Syndrome but it also highlights the need to discuss the role of more extensive surgical resection when an MMR carrier is diagnosed with their first CRC.

**Extra-colonic cancers**—For all new patients diagnosed with Lynch Syndrome we are now writing to their General Practitioners and specialists involved in their care including Gynae-Oncologists and Urologists to make them aware of this diagnosis and the need for awareness of the increased risk of extra-colonic cancers/surveillance as per the National Guidelines for Gynaecological and Urological malignancy.

**Familial Adenomatous Polyposis (FAP)/MutYH Associated Polyposis (MAP)/Serrated Polyposis Syndrome (SPS)**—There is now more of an emphasis on cumulative polyp count for both adenomatous and serrated/ hyperplastic polyps. An increasing adenomatous polyp count over years may lead to genetic testing for Attenuated FAP and MYH associated polyposis.

New guidelines and criteria are being developed in conjunction with the Genetics Service. For hyperplastic/ serrated polyps an increasing cumulative count may also lead to a diagnosis of the Serrated Polyposis Syndrome (SPS) and a change in the recommended surveillance intervals for the patient and their first degree relatives. We are happy to provide advice on surveillance intervals or engage in discussion re the appropriateness, timing and optimal extent of resection for patients with polyposis.

We are also recommending that all FAP and MAP patients have annual thyroid examinations. There is a slightly increased risk of thyroid cancer in these patients but there is no worldwide consensus on thyroid screening. The Cleveland Clinic recommend annual ultra sound but the British favour annual manual examination. Screening ultrasound potentially picks up non-significant lesions leading to a cascade of investigations and treatment which may not be needed and may cause complications.

**Patient/family education**—Our Christchurch branch is planning patient & family information sessions this year. Our Auckland branch ran sessions for both Lynch Syndrome and FAP/MAP patients/families at the end of last year and these were well attended and well received. We hope to have a separate Doctors education session in the next year.

The National Guidelines relating to colorectal cancer risk and recommended surveillance are on the Ministry of Health website [www.health.govt.nz](http://www.health.govt.nz) and our Service website [www.nzfgcs.govt.nz](http://www.nzfgcs.govt.nz) contains patient information on Lynch Syndrome, FAP/MAP and SPS along with our contact details.

Please feel free to contact us at anytime.

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