EM0215: The Globe and Mail, March 22, 2002, page A11

Down syndrome cuts cancer risk

New study says extra chromosome protects the body, cells seem slower to replicate

BY ANDRÉ PICARD
PUBLIC HEALTH REPORTER

People with Down syndrome are one-tenth as likely to get most types of cancer as those in the general population, according to a study.

Researchers say the dramatic difference likely is due to a combination of factors but that the finding could provide clues in how to fight a broad range of cancers.

Dr. Quanhe Yang of the National Center on Birth Defects and Development Disabilities at the U.S. Centers for Disease Control and Prevention said the finding is unexpected and exciting but that more research is needed. He said there are three factors that could explain lower cancer rates in people with Down syndrome:

- They have an extra copy of chromosome 21. While that causes a number of physical and mental problems, it means there are extra tumour-suppressor genes;
- The cells of people with the syndrome seem to replicate more slowly than do cells of people without the disorder, creating less opportunity for mutations, often at the root of cancers;
- Environmental influences. Very few people with Down syndrome smoke or drink, and because they tend to have intestinal problems, they often eat very healthy diets.

The study, published in today's edition of *The Lancet*, is the most extensive look ever at the causes of death among people with Down syndrome, a common but little-studied chromosomal disorder.

Dr. Yang's study reveals a stunning life-expectancy increase in one generation. From 1982 to 1997, the average age at death of people with the syndrome jumped to 49 years from 25. In the general population, life expectancy is about 80.

Sue Fletcher, chairwoman of the Canadian Down Syndrome Society, said that the data reflect what she observes in the community: "People are living longer and living better." She attributed the gains to parental advocacy, which has led to better medical care and to the integration of people with Down syndrome into the community.

"I know I'm raising my son [Marc, aged 15] to be a productive member of society. I know that inclusion is essential to his health," Ms. Fletcher said.

She said that longer life expectancy highlights glaring gaps in the opportunities and services for people with developmental disabilities – and their caregivers – that urgently need to be addressed.

While the life expectancy of people with Down syndrome compares poorly to that of the general population, it was unheard of for people with a severe developmental disability to live into mature adulthood.

Fifty years ago, more than half of babies born with the syndrome died before they reached one year of age, and often infants were left to die instead of being treated for their medical conditions.

About one in every 800 babies is born with Down syndrome. It is the leading cause of developmental disabilities. An estimated 42,000 Canadians live with the condition.

A person with Down syndrome has 47 chromosomes in each cell instead of 46. This leads to a plethora of health problems, including cardiovascular disease, thyroid problems, gastrointestinal disorders, vision and hearing problems and Alzheimer's disease.

The vast majority of people with the syndrome die of heart failure because of congenital malformations of the heart. The research found that the death rate from this cause is as much as 80 times greater than in the general population.

While people with Down syndrome have much lower rates of most types of cancers, the study found that the likelihood of their getting testicular cancer or leukemia is far higher than in the general population.

Lower risk of cancer

People with Down syndrome have dramatically lower rates of cancer, in part because the condition seems to give them extra cancer-fighting genes. The following table lists types of cancer and the odds a person with the syndrome will die of the cancer compared to a person without the chromosomal disorder.

Cancer type	Odds of death
Oral	5%
Stomach	13%
Colo-rectal	8%
Liver	41%
Pancreatic	14%
Lung	2%
Skin	6%
Breast	4%
Cervical	N/A
Uterine	22%
Ovarian	7%
Prostate	8%
Testicular	323%
Bladder	20%
Kidney	8%
Brain	9%
Lymphatic	17%

SOURCE: THE LANCET THE GLOBE AND MAIL

REFERENCE: Yang, Q., Rasmussen, S.A. and Friedman, J.M.: Mortality associated with Down's syndrome in the USA from 1983 to 1997: a population-based study. *The Lancet* **359**(#9311): 1,019-1,025 (2002). [DC Library call number: PER R31.L3]

The article EM0215 reprinted above is used in Chapter 11 of the STAT 231 Course Materials.