

# Researchers hit the headlines

*It's been a busy winter for Sick Kids scientists and the media: in recent weeks, a number of scientific discoveries and findings made the news. Here's a summary of the stories that ran.*

## Second-hand smoke affects the unborn

The first biological proof that second-hand cigarette smoke can affect a fetus was described by Dr. Gideon Koren, head of the Division of Clinical Pharmacology. His team found evidence of nicotine in the hair of fetuses carried by women who did not smoke, but who were exposed to second-hand smoke, during pregnancy. The amount of the toxin measured was as if the mothers "had smoked an average of several cigarettes a day."

It's been known for some time that fetuses and newborns are adversely affected by nicotine when the mother smokes

during pregnancy. Prematurity and spontaneous abortion are higher in fetuses; newborns are at risk for sudden infant death syndrome, low birth weight, and a number of health and behavioural problems.

"Passive smoking may also put a fetus at risk for similar health problems," concludes Dr. Koren. It may not be enough just to be a non-smoker: the research has implications for pregnant women who spend time in a smoke-filled environment either at home or in the workplace.

## Learning more about leukemia

Bone marrow is yielding more of its secrets to geneticist Dr. John Dick and his collaborators

from Montreal, Buffalo, NY, and Princess Margaret Hospital in Toronto. The latest achievement is to find the unique cells, called stem cells, found in bone marrow. These are the cells which give rise to the cells of the blood system. When they go wrong, they can cause leukemia.

In this research, Dick and his colleagues are studying acute myeloid leukemia (AML), a common form of leukemia in adults.

"Our next step is to distinguish between leukemic stem cells and healthy ones," says Dr. Dick. If such a test were now to be developed, it would open the door to future treatments.

One possibility is that, if healthy stem cells could be selected and removed from an AML patient's bone marrow and stored frozen, they could be re-transplanted into the patient after treatment to destroy the diseased bone marrow.

Dr. Dick studies the human blood system by using mice which have been transplanted with human bone marrow. In this case, mice were transplanted with cells from patients with AML, providing a model of the human disease.

## Understanding Wilson disease

Copper poisoning of the liver and brain underlie Wilson disease, whose clinical symptoms include liver failure and neurological problems. It can occur either in childhood or in adulthood.

Excess copper is usually removed from liver cells into the bile by a protein pump in the cell walls: poisoning occurs when the pump fails. Now the work of geneticist Diane Cox and her team has led to the discovery of the gene that causes Wilson disease. They have also learned how gene defects cause the disease: the key protein is missing and the pump functions poorly or not at all.

Dr. Cox notes that Wilson disease is difficult to diagnose, and is frequently not diagnosed until too late. Treatment can be effective, but only if begun in time.

Present tests take several weeks and even then are not always conclusive. The research is a key step to the eventual development of a test that's simple and conclusive. Not only that, such a test could be used to screen families with a history of the disease so that those affected could be treated before problems arise.

## Early treatment for Menkes

Menkes disease is rare. If it's not detected, a child's neurological functions will deteriorate rapidly, leading to death before age three. If it can be detected early, then effective treatment can begin: the world's oldest surviving patient with Menkes disease is now 17.

Menkes disease is caused by a copper deficiency; treatment consists of raising the body's copper levels by using a drug called copper-histidine.

"We need to raise physician awareness about the disease," explains Dr. Bibudhendra Sarkar, head of Biochemistry Research. He and his collaborators published a summary of 17 years of experience on Menkes disease in order to promote the availability of effective treatment.

"We know that copper-histidine must be given within the first month of life in order for it to prevent the neurological deterioration."

Sick Kids is considered a world leader in Menkes disease: the first Canadian patient with the disease was diagnosed here.