

UPdate

Spina Bifida & Hydrocephalus Canada

Reduction in Neural-Tube Defects after Folic Acid Fortification in Canada

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In 1998, folic acid fortification of a large variety of cereal products became mandatory in Canada, a country where the prevalence of neural-tube defects was historically higher in the eastern provinces than in the western provinces. We assessed changes in the prevalence of neural-tube defects in Canada before and after food fortification with folic acid was implemented. The study population included live births, stillbirths, and terminations of pregnancies because of fetal anomalies among women residing in seven Canadian provinces from 1993 to 2002. On the basis of published results of testing of red-cell folate levels, the study period

was divided into prefortification, partial fortification, and full fortification periods. We evaluated the relationship between baseline rates of neural-tube defects in each province and the magnitude of the decrease after fortification was implemented. A total of 2446 subjects with neural-tube defects were recorded among 1.9 million births. The prevalence of neural-tube defects decreased from 1.58 per 1000 births before fortification to 0.86 per 1000 births during the full-fortification period, a 46% reduction. The magnitude of the decrease was proportional to the prefortification baseline rate in each province, and geographical differences almost disappeared after fortification began. The observed reduction in rate was greater for spina bifida (53%) than for anencephaly and encephalocele (38% and 31%, respectively). Food fortification with folic acid was associated

with a significant reduction in the rate of neural-tube defects in Canada. The decrease was greatest in areas in which the baseline rate was high.

Source Information

From Laval University, Quebec, QC (P.D.W., F.T.); the University of British Columbia, Vancouver (M.I.V.A.); British Columbia Ministry of Health, Victoria (S.-H.U.); Alberta Health and Wellness, Calgary (R.B.L., B.S.); the University of Manitoba, Winnipeg (J.A.E.); Dalhousie University, Halifax, NS (M.C.V.H.); the IWK Health Centre, Halifax, NS (P.Z.); Health Science Centre, St. John's, NL (M.C., B.F.); Food Directorate, Health Canada, Ottawa (N.S.L.) - all in Canada; and Robert Stempel School of Public Health, Florida International University, Miami (T.N.).

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SBHAC National Research Program will attempt to drive additional research in this area.

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Folic Acid Fortification Warning

Fortifying flour with folic acid to cut birth defects may lead to a range of health problems, warn scientists.

The move was approved earlier this year by the Food Standards Agency as a way to reduce defects such as spina bifida. However, an Institute of Food Research team has shown the liver could easily become saturated by folic acid. Writing in the *British Journal of Nutrition*, they warn this could lead to unmetabolized folic acid entering the blood, which could damage health. The latest study follows a letter to the Food Standards Agency from Sir Liam Donaldson, the Chief Medical Officer of England, requesting further expert consideration of two recent studies linking folic acid to bowel cancer before the government gives the final go-ahead for mandatory fortification. But the Food Standards Agency said fortification was safe.

Dramatic results

Folic acid is a synthetic form of folate, a B vitamin found in a wide variety of foods including liver and green leafy vegetables.

But while folates are broken down in the gut, the latest research shows that folic acid is metabolized in the liver. The researchers warn that the liver is an easily saturated system, and mandatory fortification could lead to significant unmetabolised folic acid entering the blood.

Researcher Dr Sian Astley said fortifying flour would undoubtedly reduce the number of neural tube defects among babies.

Mandatory fortification has already been introduced in the US, Canada and Chile, where it cut defect rates by up to half. But she said: "With doses of half the amount being proposed for fortification in the UK, the liver becomes saturated and unmetabolised folic acid floats around the blood stream.

"This can cause problems for people being treated for leukaemia and arthritis, women being treated for ectopic pregnancies, men with a family history of bowel cancer, people with blocked arteries being treated with a stent and elderly people with poor vitamin B status.

"For women undergoing in-vitro

fertilization, it can also increase the likelihood of conceiving multiple embryos, with all the associated risks for the mother and babies."

Dr Astley warned it could take 20 years for any potential harmful effects of unmetabolized folic acid to become apparent.

Harmful effects

It has already been shown that folic acid fortification can cause harm to some people.

For example, studies have confirmed that unmetabolised folic acid accelerates cognitive decline in the elderly with low levels of vitamin B12.

Similarly, dietary folates have a protective effect against cancer, but folic acid supplementation may increase the incidence of bowel cancer. It may also increase the incidence of breast cancer in postmenopausal women.

However, since the 1980s a consensus formed that folic acid is metabolized in the small intestine in a similar way to naturally-occurring folates. Dr Astley said: "We challenge the underlying scientific premise behind this consensus. "This has important implications for the use of folic acid in fortification, because even at low doses it could lead to over consumption of folic acid with its inherent risks."

Response

In a statement, the Food Standards Agency said its recommendation was made after an extensive and scientifically robust assessment.

"The FSA Board would not have recommended mandatory fortification if the scientific evidence suggested that there were unacceptable health risks for some groups. "As part of the process, an expert committee of scientists considered the evidence regarding unmetabolised folic acid and they concluded that the data in humans was insufficient to assess any associated risks. "They also examined in detail the potential cancer risks to some groups from high folic acid consumption." The FSA added that it had recommended controls on voluntary fortification, and clear guidance on the use of folic acid supplements. Both were designed to ensure most people did not

exceed the recommended upper daily limit for folic acid.

Andrew Russell, of the Association for Spina Bifida & Hydrocephalus, said the risks of unmetabolised folic acid had been "thoroughly investigated and discounted" by top experts.

Professor Nicholas Wald, director of the Wolfson Institute of Preventive Medicine, said: "Fortification would prevent many cases of spina bifida and would also benefit the health of the country as a whole.

"Further delay in this public health measure will result in hundreds more babies being disabled by this serious disorder, or pregnancies being needlessly terminated due to a neural tube defect."

Story from BBC NEWS:

<http://news.bbc.co.uk/go/pr/fr/-/1/hi/health/7069077.stm>

Published: 2007/10/30

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We challenge the underlying scientific premise behind this consensus

*Dr Sian Astley
Institute of Food Research*

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Mutation Linked to Spina Bifida Birth Defect

A genetic mutation has been linked to the developmental disorder spina bifida, researchers in Montreal say. Spina bifida is the second most common birth defect in humans after cardiac abnormalities.

"This discovery won't have a major impact on the search for a cure yet, but it can have an immediate impact on diagnosis or risk assessment," said Philippe Gros, a professor of biochemistry at McGill University. "A pre-natal diagnosis would allow physicians to decide whether to follow a pregnancy more closely." Scientists have suspected that neural tube defects such as spina bifida have a genetic component.

In Thursday's issue of the New England Journal of Medicine, Gros's team said they have identified three mutations that implicate a gene as a risk factor for neural tube defects in humans. Neural tube defects, in which the central nervous system fails to develop fully, affect as many as two births in 1,000.

Incomplete closure

In the neural tube defect spina bifida, the spine does not form completely. Effects vary from severe physical deformities and developmental problems to minor bladder problems that may be corrected by surgery,

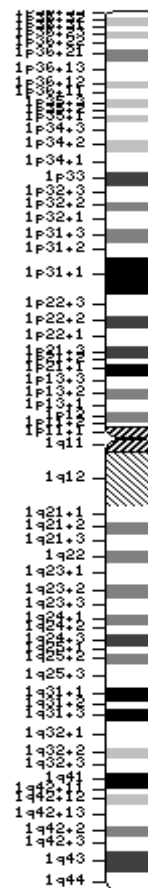
according to the Spina Bifida and Hydrocephalus Association of Canada.

The gene, called VANGL1, codes for a protein that cells need to line up properly during development, the researchers said. The mutation leads cells to lose their orientation, which is needed for neural tubes to close without gaps exposing nerve tissue. VANGL1 is the first gene shown to cause the disorder in humans, said Gros. The neural defect occurred when the team removed the gene in mice. The same mutations were found when the researchers looked at several children treated at birth defect centres in Italy and France.

Role of diet

Other researchers have looked for mutations linked to neural tube defects without success, which suggests a combination of different genes and environmental factors affect the incidence and severity of the defect, the study's authors wrote. Neural tube defects occur during the first four weeks of pregnancy when the brain and spinal cord develop. The early onset makes it difficult to detect and treat, since the first signs generally do not appear until the first ultrasound at 18 weeks. At that point, the damage has already occurred.

Given the early onset, women who want to conceive are often advised to take 0.4 milligrams a day of folic acid supplements. Folic acid is the synthetic form of folate, also called vitamin B-9, which is found in citrus fruits, nuts, liver and dark green, leafy vegetables. Since the Canadian government introduced mandatory fortification of whole-grain breads, cereals, flour, cornmeal and pasta with folic acid in 1998, the rate of birth defects such as spina bifida has declined.



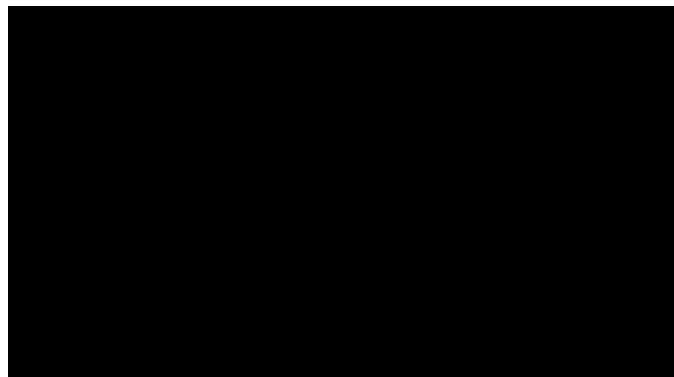
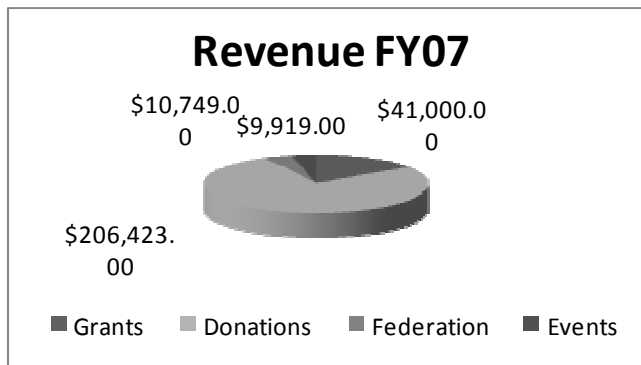
CEP Funds Reproductive Health Study

The Communications, Energy and Paperworkers Union of Canada (CEP) will fund a historic research project into the reproductive health of working men and women. The initial goals of the project are to gather facts on reproductive outcomes in CEP members (stillbirth/miscarriage, sterility, birth defects, premature births) and their exposure to toxic substances (including endocrine disruptors), stressors

(including shiftwork), physical and biological agents. Dr. Gideon Koren, a prominent expert on reproductive hazards, will lead the research team. Dr. Koren is the founder and director of "Motherisk", a world-renowned research, counseling and education program on reproductive hazards at Toronto's Hospital for Sick Children and affiliated with the University of Toronto.

"IF IT'S TOXIC OR DANGEROUS WORK, OUR MEMBERS DO IT," SAID CEP PRESIDENT DAVE COLES WHEN SPEAKING ABOUT THE PROJECT TO CEP'S NATIONAL EXECUTIVE BOARD,

Financial Highlights



Taken from the annual report. For questions or to receive a copy (English or French) please call 800-565-9488



**Did you know that John Mellencamp
has spina bifida?**

John Mellencamp has signed on for our corporate campaign this year, lending his image and words to our efforts to raise awareness and funds.

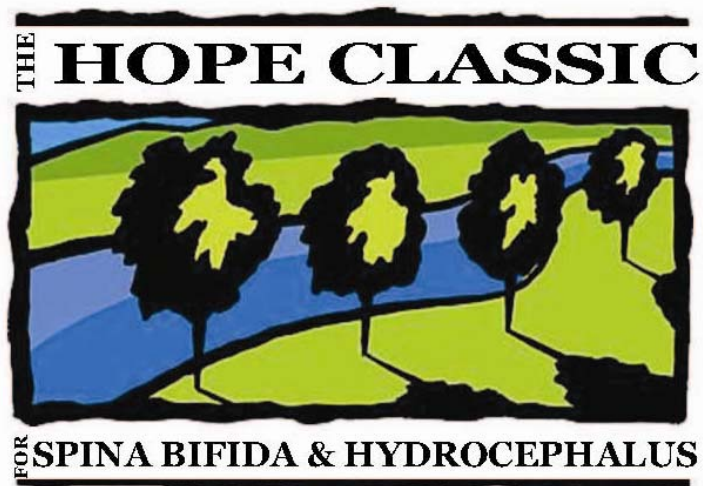
As he tours across Canada in 2008 take the time to say thank you!

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Our law firm is recognized across Canada as a leader in the field of Medical Malpractice. We have successfully represented children with Spina Bifida and their families for many years. We understand that most people have very limited or no financial resources to contribute to the cost of litigation. **If we do not win a monetary award for our client, we do not charge for our services.**

For **FREE** consultation call 1-800-961-7356; (416) 961-1212;
Fax (416) 961-2827 or visit our website at: www.sommersandroth.com



SAVE THE DATE
August 16, 2008
www.hopeclassic.ca
for cities and details
First National
Walk Run and Wheel

The Hope Classic is a nationally branded run/walk/wheel event to be held on August 16, 2008 to raise funds for the spina bifida and hydrocephalus associations across Canada. This is a family-friendly and non-competitive event where "Finishing is Winning". We encourage those in wheelchairs and strollers to participate.

The BC association has held two successful annual events in Chilliwack, BC under the Hope River Classic banner. With the information shared by them at the Annual General Meeting in September and the enthusiasm of other member associations, we are now looking forward to getting everyone involved in the Hope Classic.

Brad Hagkull, a youth pastor and father of 8 year old Benjamin who was born with spina bifida and hydrocephalus, has been instrumental in the success of the Chilliwack event. Brad and Benjamin participate in triathlons as Team Hagkull. Check out their website: www.teamhagkull.com. Benjamin's story will be featured in the pledge/registration forms for the Hope Classic.

We have secured a website www.hopeclassic.ca which will have all the information on event

sites, contacts, sponsors etc. and will be continually updated.

We have a Hope Classic sponsor who has made a verbal commitment to sponsor the event. Pledge/registration forms, banners, posters, t-shirts, water bottles, and other giveaways will be supplied to participating associations through the national office at no cost.

A planning kit is being sent to all associations with step by step instructions on getting started and keeping on track.

So what do we need from you?? Your commitment to get involved and share in the success of this event.

If you have any questions regarding the Hope Classic contact:

Bonnie Hidlebaugh 1-800-565-9488

bonnie.hidlebaugh@sbhac.ca



Severe Hydrocephalus Associated with Congenital Varicella Syndrome

Infants born to mothers who become infected with varicella during the first half of pregnancy are at risk for a very serious condition known as congenital varicella syndrome, characterized by low birth weight, cutaneous scarring, limb hypoplasia, microcephaly, cortical atrophy, chorioretinitis, cataracts, and other anomalies. In a prospective study conducted in Europe from 1980-1993 involving

nearly 1400 mothers who had varicella during pregnancy, the highest risk (2%) for congenital varicella syndrome was observed when maternal infection occurred during 13-20 weeks' gestation.

If a pregnant woman is vaccinated or becomes pregnant within 1 month of vaccination, she should be counseled about potential effects on the fetus. Wild-type varicella poses a small risk to the fetus, and because the

virulence of the attenuated virus used in the vaccine is less than that of the wild-type virus, the risk to the fetus should be lower or absent.

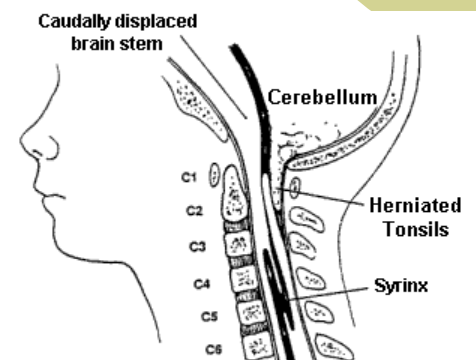
What is Syringomyelia?

Syringomyelia (syr-ING-oh-my-EE-ya) is a disorder in which a cyst forms within the spinal cord. This cyst, called a syrinx, expands and elongates over time, destroying a portion of the spinal cord from its center and expanding outward. When a syrinx widens enough to affect nerve fibers that carry information from the brain to the extremities, this damage results in pain, weakness, and stiffness in the back, shoulders, arms, or legs. Other symptoms may include headaches and a loss of the ability to feel extremes of hot or cold, especially in the hands. Each patient experiences a different combination of symptoms depending on where in the spinal cord the syrinx forms

and how far it expands. Other, more common disorders share the early symptoms of syringomyelia. In the past, this has made diagnosis difficult. But the widespread availability of an outpatient imaging procedure called magnetic resonance imaging (MRI) has significantly increased the number of syringomyelia cases diagnosed in the beginning stages of the disorder.

Because syringomyelia can occur in association with other conditions, estimates of the number of Americans with the disorder vary widely, but a conservative estimate is that about 40,000 people in the United States are affected, with symptoms usually beginning

in young adulthood. Signs of the disorder tend to develop slowly, although sudden onset may occur with coughing or straining. If not treated surgically, syringomyelia often leads to progressive weakness in the arms and legs, loss of hand sensation, loss of bladder and other functions, and chronic, severe pain.



Unlocking the Mystery of Hydrocephalus

Hydrocephalus can be defined broadly as a disturbance of formation, flow, or absorption of cerebrospinal fluid (CSF) that leads to an increase in volume in the central nervous system (CNS). This condition can be termed a hydrodynamic disorder of CSF. Acute hydrocephalus occurs over days, sub acute over weeks, and chronic over months or years. Conditions such as cerebral atrophy and focal destructive lesions also lead to an abnormal increase of CSF in CNS. In these situations, loss of cerebral tissue leaves a vacant space that is filled passively with CSF. Such conditions are not the result of a hydrodynamic disorder and therefore are not classified as hydrocephalus. An older misnomer used to describe these conditions was hydrocephalus ex vacuo.

Normal pressure hydrocephalus (NPH) describes a condition that rarely occurs in patients younger than 60 years. Enlarged ventricles and normal CSF pressure at lumbar puncture (LP) in the absence of papilledema led to the term NPH. However, intermittent intracranial hypertension has been noted during monitoring of patients in whom NPH is suspected, usually at night. The classic symptoms includes gait apraxia, incontinence, and dementia. Headache is not a typical symptom in NPH.

Benign external hydrocephalus is a self-limiting absorption deficiency of infancy and early childhood with raised intracranial pressure (ICP) and enlarged subarachnoid spaces. The ventricles usually are not enlarged significantly, and resolution within 1 year is the rule.

Communicating hydrocephalus occurs when full communication exists between the ventricles and

subarachnoid space. It is caused by overproduction of CSF (rarely), defective absorption of CSF (most often), or venous drainage insufficiency (occasionally). Non communicating hydrocephalus occurs when CSF flow is obstructed within the ventricular system or in its outlets to the arachnoid space, resulting in ventricular/subarachnoid space non communication.

Obstructive hydrocephalus results from obstruction of the flow of CSF (intraventricular or extraventricular). Most hydrocephalus is obstructive, and the term is used to contrast the hydrocephalus caused by overproduction of CSF.

Arrested hydrocephalus is defined as stabilization of known ventricular enlargement, probably secondary to compensatory mechanisms. These patients may decompensate, especially following minor head injuries.

Incidence

In the US: Incidence of congenital hydrocephalus is 3 per 1,000 live births, while the incidence of acquired hydrocephalus is not known.

There are no current numbers for Canada, however based on incidence it is a modest assumption to liken the numbers.

Internationally: Incidence of acquired hydrocephalus is unknown. About 100,000 shunts are implanted each year in developed countries, but little information is available for other countries.

FACTS

In untreated hydrocephalus, death may occur by tonsillar herniation secondary to raised ICP with

compression of the brain stem and subsequent respiratory arrest.

Shunt dependence occurs in 75% of all cases of treated hydrocephalus and in 50% of children with communicating hydrocephalus.

Patients are hospitalized for scheduled shunt revisions or for treatment of shunt complications or shunt failure.

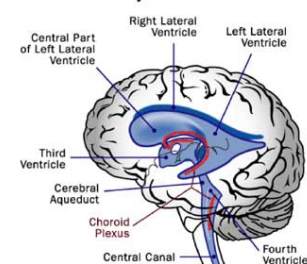
Poor development of cognitive function in infants and children, or loss of cognitive function in adults, can complicate untreated hydrocephalus. It may persist after treatment.

Visual loss can complicate untreated hydrocephalus and may persist after treatment.

Generally, incidence is equal in males and females. The exception is Bickers-Adams syndrome, an X-linked hydrocephalus transmitted by females and manifested in males. NPH has a slight male preponderance.

Hydrocephalus presents a bimodal age curve. One peak occurs in infancy and is related to the various forms of congenital malformations. Another peak occurs in adulthood, mostly resulting from NPH. Adult hydrocephalus represents approximately 40% of total cases of hydrocephalus.

The Ventricular System of the Human Brain



Cephalic disorders are congenital conditions that stem from damage to, or abnormal development of, the budding nervous system. Cephalic is a term that means "head" or "head end of the body." Congenital means the disorder is present at, and usually before, birth. Although there are many congenital developmental disorders.

Our mission is to improve the quality of life of people living with Spina Bifida and/or hydrocephalus and their families through awareness, education, advocacy and research, and to reduce neural tube defects.”



La mission de l'ASBHC consiste à améliorer la qualité de vie tous les individus vivant avec le spina-bifida ou l'hycrocéphalie et de leurs familles par l'information, l'éducation, la recherche et les références en plus de réduire l'incidence des malformations du tube neural (MTN) par la prévention.

President's Message *Lorelei Fletcher*

2007 has been a year of change.

The Communications, Energy and Paperworkers Union of Canada (CEP) continue to be the backbone of the Association. CEP is approaching the 1,000,000 mark in contributions since the beginning of our relationship. We look forward to celebrating this amazing milestone together.

Our organization is undertaking some exciting and challenging directions in the coming years, as we focus on growth and development towards our goal of becoming a strong, collaborative national health charity in Canada.

Our new communications strategy will focus our branding (reputation) and visual identity to create a unified look and feel. We will build stronger and more effective awareness

campaigns using national opportunities to tell our story.

We will focus on our internal structure using the federation framework to pull all provinces together toward our common goal. How we commit to interacting with each other will go a long way toward furthering our growth and development, both together and individually.

Internationally we are developing key relationships with SBA (America) and the International Federation. Canada has a reputation for being a leader in the global market and we see opportunities for SBHAC to demonstrate this in our field as well. We have much to learn as well as much to offer to our international colleagues.

We continue to build our national team with professionals in their

respective fields. The key areas include Communications & Development as well as Education & Information. It is vital to our success as an organization that our ability to generate the funds needed to carry out our programs are secured and our products are considered to be of high quality and credible in the field of SB&H.

I see positive outcomes that will continue to strengthen our National identity, creating stronger relationships across Canada.

Together we can achieve!

I wish to express my thanks to our staff and volunteers across Canada who make SBHAC the success that it is.

