

# PROTECTING THE TINIEST BRAINS

**When babies are born several weeks before their due date, the focus is usually on keeping them alive. But what about after their survival has been ensured? Can a baby be born extremely early and still lead a healthy, happy life?**

**“**One of the adverse outcomes of great concern for children who have been preterm is cerebral palsy,” says Dr. Charlene Robertson, a pediatric consultant at Glenrose Rehabilitation Hospital in Edmonton. *“The term cerebral palsy (CP) includes a group of permanent disorders of movement and posture that cause activity limitation.”* As better care of very preterm infants led to higher survival rates starting in the 1970s, rates of CP also increased.

Robertson and her team looked at the CP rates over the past 30 years among infant survivors born at 20 to 27 weeks, weighing only 500 to 1,249 grams at birth, from a specific catchment area in which a great deal was known about base population, birth rates, prematurity rates, and the nature of care of premature infants. They found that rates of CP peaked in the early 1990s and then steadily

dropped off in the last decade. The change is substantial: a child born very preterm in the early 1990s had about a 13% chance of having CP, this probability dropped to under 2% in the early 2000s. Particularly encouraging was a significant drop in rates of severe CP, the type that prevents children from walking. This is indeed heartening news, both for the parents of extremely preterm infants and the professionals who care for them.

The reasons for this change are complex, reflecting changes in overall systems of care. Dr. Peter Rosenbaum, an expert in CP from McMaster University who was not involved in this research, says, *“There have probably been many small but cumulative changes in the care of premature infants that together aggregate to make huge differences to both survival and to intact survival over the period of this report.”*



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While these findings are undoubtedly positive, Dr. Robertson believes they should not obscure the fact that children born extremely preterm often have special needs and are likely to benefit from early interventions to help maximize their overall potential. 🦸

**BY ALISON PALKHIVALA**

Ref.: Robertson CMT, Watt MJ, Yasui Y. Changes in the prevalence of cerebral palsy for children born very prematurely within a population-based program over 30 years. *Jama-Journal of the American Medical Association* 2007;297(24):2733-2740.

# EARLY IDENTIFICATION DIRECTS REHABILITATION CHOICES

**Both deafness and blindness are significant disabilities. Imagine what it's like to have both! This is what occurs in a genetic condition known as Usher syndrome.**

**“**In Usher syndrome type I, the hearing loss is congenital and the blindness starts before puberty, while in Usher syndrome type II, the deafness is in early childhood and the blindness starts after puberty,” says Dr. Robert K. Koenekoop, of the McGill University Health Centre. Short of a cure, the best thing we can do for these children is identify the condition early so that appropriate rehabilitation can be provided without delay.

The 6 million French Canadians alive today are all descendants of about 8,500 French settlers, making them a relatively homogeneous

group, genetically speaking. Dr Koenekoop and his colleagues conducted a genetic analysis of 15 Usher syndrome patients from different parts of Quebec. They found mutations of a gene implicated in Usher syndrome type I, known as USH1C, in 9 of these patients. This mutation has rarely been found outside the Acadian population.

The researchers discovered what's known as a “founder mutation” for both types of Usher syndrome, so now children born either deaf or blind can easily be tested for the presence of the condition, giving them, their fam-

ilies and their physicians an idea of what's in store and how best to manage their disability.

*“These findings highlight the need for early screening of deaf infants in Quebec as well as the Acadian population in the Maritime region of Canada,”* says Dr. James C. MacDougall, an expert on deafblindness who is also at McGill University but was not involved in this study. *“Highly specialized counselling is necessary for the parents of a child who has been identified with Usher syndrome. Parents need genetic counselling concerning the risks for future children they may have and, as well, they need information on the impact of early deafness and progressive hearing loss. Unfortunately, the rehabilitation process for all deaf children remains a controversial topic with one group of professionals favouring an exclusive oral approach while another group favours the addition of some form of sign language to facilitate communication.”*

Predicting future disability, however, is crucial to choosing the best rehabilitative option. 🦸

**BY ALISON PALKHIVALA**

Ref.: Ebermann I, Lopez I, Bitner-Glindzicz M, Brown C, Koenekoop RK, Bolz HJ. Deafblindness in French Canadians from Quebec: A predominant founder mutation in the USH1C gene provides the first genetic link with the Acadian population. *Genome Biology* 2007;8(4):R47.