



### What is Congenital Rubella Syndrome

Congenital rubella syndrome (CRS) is a series of symptoms resulting from rubella virus infection during pregnancy. When rubella infection occurs primarily during the first trimester of pregnancy, serious consequences such as miscarriages, stillbirths, and a constellation of severe birth defects in infants can result (Centre for Disease Control).

### Study Rationale

The rationale for conducting this project was to describe (from a sample of volunteer participants) the manifestations of their congenital rubella syndrome or CRS, observed in two recognized stages: a) **early onset** - manifestations such as deafness, congenital cataracts and congenital heart defects observed in newborns or very young children; b) **delayed or later onset** – those developing during later childhood and progressively into adulthood, including glaucoma and corneal conditions affecting vision, epilepsy, endocrine dysfunction issues (diabetes and thyroid issues), advanced vascular problems, to name a few.

This 2014 Canadian study is a follow up to the 1999 study, intended to confirm the previous results and observe any changes that might be occurring over the fifteen year period since the last project.

### Sampling Method

Volunteers known to have CRS in Canada were sought through contact with various Independent Living Residences for Individuals with Deafblindness and others known to have CRS and living elsewhere in Canada. Participants were asked to report (via a questionnaire) the incidence (including an approximate age of the onset) of observed manifestations affecting each individual's sensory, neurological, vascular, musculoskeletal, endocrine, respiratory and gastrointestinal system. In addition, a sociological snapshot of the participants was provided through answers to questions related to their level of communication and education, employment and accommodation history.

### General Demographics of the Sample

Fifty-three individuals (29 males and 24 females) ranging in ages 29 to 62 years participated in this project. Seventy percent of individuals were born during the period 1966 - 1980, corresponding to the period before rubella in Canada was largely contained through immunization programs.

### Discussion

The findings reported in this follow-up study not only validated the results from the previous study but demonstrated higher rates of incidence of later manifestations for many of the medical conditions examined. Examples include: the incidence of glaucoma (54.2% in 2014 versus 32.3% reported in 1999); the incidence of hypertension (18% in 2014 versus 7.2% in 1999); and the incidence of endocrine dysfunction (42.3% in 2014 vs 21.2% in 1999).

With respect to the neurological data, over twice as many individuals showed indications of cognitive skills degeneration, between 1999 and 2015.

Difficulties with behavioural or psychosocial control were similar between the two surveys, confirming the link of these behaviours to emotional disorders observed in individuals who are congenitally deafblind. (Dr. Jude Nicholas – Neuropsychologist- Personal Communication).

This observation would suggest that, like the similar incidence of seizures (27.5% in 2014 survey versus 30% in 1999), the difficulties with behavioural control are established congenital issues that appear as late onset manifestations.

### Reported incidence rate of 'early onset manifestations' from a sample of 53 individuals with CRS

Vision Loss (Congenital cataracts)	96.2%	Hearing Loss	92.5%
Combined Vision & Hearing Loss	88.7%	Cardiac Defect	64.0%
Combined Vision loss, Hearing Loss & congenital cardiac defect	64.1%	Cardiac surgery	27.0%

Note: Cardiac defects reported included: heart murmur (31.4%), patent ductus arteriosus (28.6%), pulmonary stenosis (28.6%).

### Reported incidence rate of 'delayed manifestations' from a sample of individuals with CRS

Glaucoma (n=47)	53.2%	Corneal damage (n=42)	28.6%
Detached retina (n=42)	21.7%	Late onset change in hearing (n=36)	25.0%
Hypertension (n=50)	18.0%	High cholesterol (n=50)	20.0%
Respiratory problems (environmental allergies, lung problems and pneumonia) (n=51)	52.9%	Allergies to various drugs	35.3%
Such spinal conditions as scoliosis or kyphosis (n=51)	19.6%	Diabetes	21.2%
Thyroid dysfunction (hyper and hypo-thyroidism) (n=52)	43.2%	Various gastrointestinal issues (n=53)	50.9%

Note: Sample sizes (n=) vary according to the number of respondents answering the question either yes or no.

### Reported incidence rate of Neurological Impairment from a sample of 51 individuals with CRS

Total reporting all neurological issues	49.0%	Incidence of seizures	27.5%
Showing signs of motor skill degeneration	27.5%	Demonstrating at least one sign of cognitive degeneration	33.3%

Note: Signs of cognitive degeneration reported were: poor attention and distraction, disorientation, impaired short term memory loss.

### Reported incidence rate of Psychosocial Behavioural Issues from a sample individuals with CRS

Stereotypical behavior (n=47)	72.3%	Self-injurious behavior (n=52)	71.2%
Compulsive behavior (n=50)	42.0%	Ritualistic behavior (n=51)	56.9%
Sameness behavior (n=49)	44.9%	Restricted behavior (n=47)	34.0%
Aggression to others (n=52)	51.9%	Episodes of anger or rage (n=52)	69.2%

Note: Sample sizes (n=) vary according to the number of respondents answering the question either yes or no.

### Reported incidence rate of Signs of Depression from a sample of 47 individuals with CRS

Reporting signs of depression (n=47)	20 (44.7%)	Rate of change in signs of depression in past year: Increased (10.6%); Decreased (36.8%) Remained the same (52.6%)
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Note: Signs of depression reported were: appearing depressed; avoiding social contact, crying frequently, not taking part in activities once of interest; appearing restless or fidgety; complaining of headaches or other aches and pains; eating too much or too little.

### Final comments

The data contained in this study confirms that congenital rubella syndrome is indeed a devastating disease that continues to manifest itself throughout the life of the individual.

Based on the extent of the medical conditions reported in this sample of the CRS population, it is important that individuals with CRS be monitored closely for changes in their physical and emotional health.

While mandatory rubella immunization programs adopted throughout the world has sharply limited its impact, it's critical that these efforts continue unabated. Further, mandatory immunization initiatives must be expanded to blanket all of Eastern Europe, Africa and Asia if there is any hope of ensuring that children and eventually adults will not face the medical crises that this virus creates.

According to Dr. Jude Nicholas (Personal Communication), the reported signs of decreased mood (or depression) of individuals in this study could be associated with the reported Psychosocial behavioural issues. These emotional problems related to regulation issues may affect the deafblind person's capacity to regulate their emotions or emotional responses. Emotion dysregulation in turn may lead to increased social isolation, escalation of aggressive-disruptive behaviors and high levels of negative affect such as depression.