Canadian Deafblind Association (National)

Manifestations of Congenital Rubella Syndrome in Canada: A 2014 Follow-Up Study

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Table of Contents

3.0 DISCUSSION

About the Organization

CDBA National is a Canadian charitable "umbrella" organization focused on the needs of individuals who are deafblind, their families and the professionals who provide services. A volunteer Board of Directors comprised of a representative from each of our provincial chapters, and an equal number of at-large board members from across the country manage the organization.

CDBA National's mission is to support and enhance the wellbeing of people who are deafblind through awareness, education and the provision of support to our chapters and community partners.

Our vision is that **all people who are deafblind will live rich meaningful lives**.

About the Project Leader/Author

Stan Munroe graduated from the University of New Brunswick with a B.A. (Biology and Psychology, 1967) and M.Sc. (Wildlife Biology, 1969). He worked with the Ontario Government (1969-1997) in Fish and Wildlife Management. Stan is a past President of the Canadian Deafblind Association (CDBA) (National) (formerly called the Canadian Deafblind and Rubella Association (CDBRA), former Executive Director of CDBA National and currently is Information Officer for Deafblind International. He is also the project lead and author of CDBA National's:

1. A Survey of Late Emerging Manifestations of Congenital Rubella in Canada (1999)
2. Developing a National Volunteer Registry of Persons with Deafblindness in Canada (2003)

Summary

The purpose of this project was to undertake an updated investigation of the Manifestations of Congenital Rubella Syndrome in Canada to complement the study published by the Canadian Deafblind and Rubella Association in 1999.

The objectives of the study were to: a) compare the extent of the medical conditions or manifestations reported by individuals with CRS in a study over fifteen years ago; b) determine whether any changes in these average conditions had occurred over a period of time; c) Reinforce the valuable information available to family members and staff working in facilities which provide services for persons with CRS, and d) provide additional data to the bank of information that is being accumulated worldwide on the late onset phenomenon.

Sampling Method

Canadian participants known to have CRS 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.

Incidence of 'early onset manifestations' reported (n=53):

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%

Incidence of 'delayed manifestations' reported:

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 (n=51)	52.9%	Allergies to various drugs (n=51)	35.3%
Such spinal conditions as scoliosis or kyphosis (n=51)	19.6%	Diabetes (Types 1&2) (n=52)	21.2%
Thyroid dysfunction (hyper and hypo-thyroidism) (n=52)	43.2%	Various gastrointestinal issues (n=53)	50.9%

Incidence of Neurological Impairment reported (n=51):

Reporting all neurological issues	49.0%	Incidence of seizures	27.5%
Signs of motor skill degeneration	27.5%	Demonstrating at least one sign of cognitive degeneration	33.3%

Incidence of Psychosocial Behavioural Issues reported:

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/rage (n=52)	69.2%

Incidence of Signs of Depression reported (n=47):

Reporting signs of depression	20 (44.7%)	Rate of change in signs of depression in past year: Increased (10.6%); Decreased (36.8%) Remained the same (52.6%)

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.

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.

1.0 Introduction

Fifteen years ago the Canadian Deafblind Association (CDBA) National published a report titled 'A Survey of Late Emerging Manifestations of Congenital Rubella Syndrome in Canada (1999)'[1]. This project described the late manifestations of congenital rubella syndrome[2] (CRS) from a volunteer sample of 100 individuals who reported that they had CRS. Prior to the 1999 project, the only previous longitudinal investigation about this issue was conducted by the Helen Keller National Centre (Long Island, New York) in 1991 (O'Donnell, 1996)[3]. Since our study was published, several other countries (Denmark, Norway and Brazil) carried out similar investigations.

The findings from these early projects led family members and professionals working with individuals with CRS to inquire about further health changes that are occurring within this population of individuals. As a follow-up to their first investigation, the Helen Keller National Centre did a major US comparative study in 2004[4], ten years after their first project and representing 40 years since the 1962-1965 global rubella pandemic[5]. The purpose of this project was to undertake an updated investigation of the Manifestations of Congenital Rubella Syndrome in Canada to complement the study published by the Canadian Deafblind and Rubella Association in 1999.

The objectives of the study were to: a) compare the extent of the medical conditions or manifestations reported by individuals with CRS in a study over fifteen years ago; b) determine whether any changes in these average conditions had occurred over a period of time; c) Reinforce the valuable information available to family members and staff working in facilities which provide services for persons with CRS, and d) provide additional data to the bank of information that is being accumulated worldwide on the late onset phenomenon.

The project was financially supported by a $15,000 grant supplied by the Federal Minister of Human Resources and Skills Development through the Social Development Partnerships Program (SDPP).

1.2 Study Plan

CDBA National appointed Stan Munroe, the project leader of the 1998-1999 study, as project leader for this follow up study. CDBA National established a working group of professionals representing the CDBA National Office[6], the Ontario Chapter of CDBA[7], Lions McInnes House[8] and Deafblind Ontario Services[9].

Through a number of meetings, the working group provided advice regarding: the development of a new questionnaire; how and where to seek potential participants for the study; the types of introductory letters to be sent to organizations known to provide

[1] www.cdbanational.com
[2] www.hknc.org/Rubella.htm
[3] http://www.hknc.org/images/CRS_SURVEY1991.htm
[4] Email: nancy.odonnell@hknc.org
[5] www.cdc.gov/vaccines/pubs/surv-manual/chpt15-crs.html
[6] www.cdbanational.com
[7] www.cdbaontario.com
[8] http://www.brantfordexpositor.ca/2009/12/21/deaf-blind-residents-have-all-of-the-comforts-of-home
[9] www.deafblindontario.com

services to potential participants; considerations for statistical design and project time table.

1.2.1 The Questionnaire

The questionnaire was designed to gather data on individuals from two perspectives: demographic history and health issues. The demographic history section asked basic questions about date and place of birth, current address, ethnicity, CRS diagnostic information, educational and employment history, methods of communication used and the type of residence they are living in. The health section was designed to learn about the extent and age of onset of any related problem (if any) affecting the following biological systems: vision, hearing, cardiac, respiratory, musculoskeletal, endocrine, gastrointestinal and neurological. Further to the neurological system, specific questions were developed to learn more about the extent of psychosocial/behavioural issues reported, including depression. Participants were also asked to provide information on the use of prescribed medications, supplements and natural remedies.

The information requested in the questionnaire is not unlike the questionnaire from the 1999 study with the exception of increased emphasis placed on Neurological and Psychosocial issues. The format for the questionnaire was adapted from that of the Helen Keller National Rubella Survey (2004) for which CDBA National received approval to use.

1.2.2 Seeking participants for the study

The procedure used to seek participants for this current project was quite different from that undertaken for the 1999 study. In the earlier study, the project leader first canvassed a very wide array of Canadian non-profit organizations, educational and residential facilities to seek cooperation for the identification of individuals known to have CRS. A number of organizations eventually volunteered to participate with the study eventually locating 100 individuals with CRS who participated in the 1999 study. Having this old database provided an initial start for seeking participants for the 2014 project.

The project leader also had another source of potential individuals with CRS outside the original rubella study. CDBA National published in 2001 'Voluntary Registry of Deafblind Persons in Canada' project[10]. Of the almost 800 Canadians with deafblindness who volunteered for that study, an additional 49 individuals (who were not part of the 1999 Rubella Project), reported their deafblindness was the result of CRS.

The approach taken to seek volunteer participants for the current study were several-fold: a) seek support from the various Chapters of CDBA to contact family members or advocates of individuals known to have CRS; b) seek support from the various independent living facilities throughout Canada known to provide services to individuals with CRS; c) contact directly by mail those individuals or their family members/advocates (a total of 32) who participated in the previous study but not

[10] www.cdbanational.com

believed to be accessible through (a) or (b), and d) contact directly by mail those individuals or their family members/advocates (a total of 25) who reported to the Volunteer Registry Project of 2001 that their deafblindness was the result of CRS.

Introductory letters explaining the project were then prepared along with consent forms (in English and French) for distribution along with the questionnaire to the various potential sources of support and participation. The questionnaire was also put online (in French and English) on the CDBA National website (www.cdbanational.com).

1.2.3 Data Analysis

For the purpose of data analysis, the data from each questionnaire was entered using the Access Program of Microsoft Office Professional Plus10[11].

2.0 Results

2.1 Section A: Demographics

2.1.1 Participation in survey

Table 1 summarizes the distribution of the 53 completed questionnaires according to province and source. Thirty-nine (73.6 %) of the questionnaires were submitted through the agency cooperators listed; the remaining fourteen or 26.4% were returned directly from individuals/family members as the result of direct mail contact by the project leader.

Table 1: Distribution of volunteer participants by Province

Province	Source of participants	No.
Ontario (35)	CDBA ON Chapter	12
	Deafblind Ontario Services[12]	12
	Lions McInnes House	6
	Rotary Cheshire Homes[13]	2
	Via Mail	3
Nova Scotia (5)	Regional Residential Services Society[14]	2
	Via Mail	3
Alberta (4)	Via Mail	4
Saskatchewan (4)	CDBA SK Chapter[15]	4
British Columbia (3)	CDBA BC Chapter[16]	1
	Via Mail	2
Manitoba (1)	Via Mail	1
Newfoundland (1)	Via Mail	1

[11] www.microsoft.com
[12] www.deafblindontario.com
[13] www.rotarycheshirehomes.org
[14] rrss.ns.ca
[15] https://en-gb.facebook.com/cdba.saskatchewanchapter
[16] www.cdbabc.ca

Total completed questionnaires		53

2.1.2 Gender and age distribution of participants

A total of 53 individuals (29 males and 24 females), born between 1951 and 1985 and ranging in ages 29 to 62, participated in the survey (Table 2).

Table 2: Gender and age distribution of the respondents

Range of Birth Years	Males	Females	Total
1950-1955 (age 62)	1	0	1
1956-1960 (ages 54-58)	1	3	4
1961-1965 (ages 49-53)	2	2	4
1966-1970 (ages 44-48)	9	7	16
1971-1975 (ages 39-43)	8	4	12
1976-1980 (ages 34-38)	5	4	9
1981-1985 (ages 29-33)	3	4	7
Total	29	24	53

The total participants in this survey represent a little over one half of the number (100) that participated in the 1999 study where the sex ratio was 58% female: 42% male. Twenty-eight (52.8%) of the same participants (15 females and 13 males) fifteen years ago participated in this survey.

It should be pointed out that the age distribution of the 1999 participants represented a much wider range than in this current study. The earliest reported year of birth was 1935 while the latest year was 1993. In that report, one participant reported being born in the 1930's, three reported being born in the 1940's, while 18 were born in the 1950's.

The 2014 study was not able to reach any of the four individuals participating in the 1999 survey born in 1989, 1990, 1992 and 1993. It was reported that one of those individuals (born in 1993) was deceased.

The report does include detailed information (compiled by family members and or care givers) for the three individuals who died prior to the survey being initiated. One male born in 1963, died at age at age 48; a female born in 1976, died at the age of 36, while another male born in 1974 died at age 30. All three died from cardiac related issues.

2.1.3 Ethnic Representation of Participants

The fifty-three participants reported their ethnic origin as follows: 42 Caucasian, 5 African-Canadian, 4 Asian (not specified), 1 Aboriginal Canadian and 1 mixed European-Asian**.**

2.1.4 Regional Distribution within Canada of Participants

The current Canadian residency of participants and the region where they contracted maternal rubella is presented in Table 3. About two thirds (35) currently reside in the Province of Ontario; while 41.5% (22) report having Ontario as their province of birth. Five individuals reported contracting rubella outside Canada: London, UK (1966), Taiwan (1976), Kenya (1980), Germany (1983) and UK (1985). Four of these individuals currently live in Ontario and one in BC.

Table 3: Comparison of number of respondents reporting current Canadian Province of residency versus province or region where born

Region	Region Currently Residing	Region Where Rubella Contracted
Atlantic Provinces	6 (NS-5, NF-1)	9 (NS-5, NB-2, NF-2)
Ontario	35	22 born in Ontario
Prairie Provinces	9 (MB -1, SK-4, AB-4)	10 (MB-2, SK-5, AB-3)
BC	3	3
Unknown Canada		4
Outside Canada		5 (Kenya, UK-2, Germany, Asia)
Total	53	53

2.1.5 Verification of CRS Diagnosis

Participants were asked about the nature of their diagnosis of CRS. Table 4 summarizes the results of the inquiry regarding the nature of the diagnosis of CRS for the 53 respondents.

Table 4: Verification of CRS diagnosis

Diagnosis of CRS	No.
Blood test	8
Physicians diagnosis or observation	34
Unknown or unreported	4
Never officially diagnosed	7
Total	53

Medical diagnoses, either through a blood test and/or consultation with a physician comprised 42 (79.2%) of the participants' responses. The remaining eleven responses were: never officially or medically diagnosed (7); unknown/unreported (4). Interestingly,

of those who indicated their rubella was not medically diagnosed, one mother was positive she had contracted the rubella virus while another parent indicated that an educational professional strongly believed their child had rubella syndrome.

2.1.6 Estimated gestational age at time of maternal infection with rubella virus

Participants were asked to report the gestational age that their mother knowingly came into contact with the rubella virus. The responses are summarized in Table 5. Seventeen of the respondents did not report or know the timing of rubella contact.

Of the 36 individuals who reported knowing their gestational age of maternal infection with rubella, all but two (or 94%) reported contracting the rubella virus during their first trimester of pregnancy. Of those 27 who specified the specific month, 40.7% (or 11) reported the first month, 37.0% (or 10) reported the second month and 22.2% (or 6) reported the third month. One each reported contacting rubella in months four and five of the second trimester.

The incidence of reporting contacting rubella during first trimester results in this survey (94.4 %) is similar to the responses reported (95 %) in the 1999 survey.

Table 5: Summary of reported gestational age of maternal Infection with rubella

Gestational Period Reported	Number
First month	11
Second month	10
Third month	6
First trimester (month not specified)	7
Second trimester (one reported month 4; one month 5)	2
Unknown or not reported	17
Total	53

2.2 Section B: Manifestations

2.2.1 Congenital Birth Anomalies

Respondents were asked to list the early manifestations of congenital rubella syndrome (congenital birth anomalies) observed or medically diagnosed at birth or in very early childhood. Further to this, respondents were also asked to provide information about the earliest known time of onset of visual, auditory, neurological, cardiac or other health problems.

Table 6 is constructed to document the broad array of congenital birth anomalies (i.e. early manifestations of congenital rubella syndrome) reported for the 53 respondents

during the first two years of their lives. Almost all of the respondents reported a vision loss (96.2%), 92.5% reported a hearing loss and 64.2% indicated a congenital heart defect. Forty-seven or 88.7% reported a combined visual and hearing loss; thirty-four or 64.1% reported a combined vision and hearing loss along with a congenital cardiac defect. This compares to the very low reported incidence of insult to vision alone (7.6%), hearing alone (0.0%) and cardiac system alone (0.0%).

In the 1999 survey the incidence of congenital defects reported by the 100 participants were as follows: congenital vision loss (86%); congenital hearing loss (94%); congenital heart defect (65.7%); combined congenital vision and hearing loss (80.2%) and combined congenital vision loss, hearing loss and heart defect (58.9%).

In addition to hearing loss, vision loss and congenital heart defects, 20 or 37.7% of respondents noted other symptoms that were diagnosed at birth. These symptoms are listed below Table 6.

Table 6: Reported early manifestations of congenital rubella syndrome

CRS Manifestations	Number Reporting
a) Reporting a vision loss	51 (96.2%)
b) Reporting a vision loss only	4 (7.57%)
c) Reporting a hearing loss	49 (92.5%)
d) Reporting a hearing loss only	0
e) Reporting a cardiac defect	34 (64.2%)
f) Reporting a cardiac defect only	0
g) Reporting a combined hearing and visual loss	47 (88.7%)
h) Reporting a combined vision loss, hearing loss and heart defect	34 (64.1%)
Individuals reporting other early health conditions (listed below)	20 (37.7%)

Other health conditions reported:

- 7 reported microphthalmia[17] (abnormally small eyes)
- 5 reported having a low birth weight[18] (birth weight less than 2.5 kg)
- 1 reported being premature
- 1 reported having a swollen liver and spleen
- 1 reported having kyphosis or hunchback[19]
- 1 reported having postnatal anoxia and cyanosis[20] (an accumulation of carbonic and lactic acids due to lack of oxygen at birth)
- 1 reported having breathing issues and microcephaly[21] (a condition in which a person's head size is much smaller than that of others of the same age and sex)

[17] ghr.nlm.nih.gov/condition/microphthalmia
[18] www.babycentre.co.uk/a1033196/low-birth-weight-in-babies
[19] en.wikipedia.org/wiki/Kyphosis
[20] www.ncbi.nlm.nih.gov
[21] www.nlm.nih.gov/medlineplus/ency/article/003272.htm

- 1 reported having a cleft palate and hair lip[22] (birth defects that occur when a baby's lip or mouth do not form properly during pregnancy)
- 2 diagnosed as developmentally delayed[23] (diagnosed by a doctor according to strict guidelines).

2.2.2 Vision Loss

All 53 respondents reported a vision loss of various degrees. (Note that this is higher than mentioned in Table 6 (n=) as several individuals did not indicate vision loss within their first two years of age.) Most (46 or 86.8%) of the responding individuals reported their vision loss occurred at or near their birth; 6 (or 11.3%) reported first onset by age five. Only one individual (1.9%) reported a visual loss occurring beyond their early childhood and that was with the onset of cataracts at age 44.
Respondents were asked to indicate their degree of vision loss. Their responses are summarized in Table 7. There would be little debate to suggest from these results that all of these individuals are seriously visually impaired.

Table 7: Responses regarding degree of vision loss

Degree of Vision Loss	Number Reporting
Severely Visually Impaired	12
Legally Blind	18
Light Perception only	1
Totally Blind	17
Unable to test-uses vision	4
Totally blind in one eye	1
Total	**53**

Participants were asked whether they were using or had used corrective lenses in the past. Of the 52 that responded to this question, 16 (30.8%) indicated they were using or had used corrective lenses in the past.

2.2.2.1 Causes of vision loss

Participants were asked to report the various causes of their visual loss. Their responses are summarized in Table 8 (a-d). Note the sample size (n=) for each item reflects those that answered yes or no to each question. Those who did not answer were removed from the sample; hence the reason for sample size less than 53.

[22] ww.cdc.gov/ncbddd/birthdefects/CleftLip.html
[23] http://www.med.umich.edu/yourchild/topics/devdel.htm

Table 8(a) Cataracts

	Age reported of onset of cataracts/or surgery				
Cataracts n = 52	**Yes**	**Age Unknown**	**Birth to Six Months**	**6mo-2yr**	**3 years +**
Reported with cataracts	48 (92.3%	0	43	4	1 (age 44)
Cataracts reported (one eye)	8 (16.7%)				
Cataracts reported (both eyes)	40 (83.3%)				
Number reporting cataract surgery n= 48	39 (81.3%	3	11	19	6: (2 age 5; 2 age 7; 1 age 17; 1 age 44)

According to Table 8(a), congenital cataracts are the primary cause of visual loss among the respondents, representing 92.3% of those answering to the condition. The majority of those reported onset as to at or near birth; one indicated the onset being at age 44.

The incidence of cataracts in the 2014 study (92.3%) is higher than the rate reported (79.6%) in the 1999 study. This difference could be attributed to the inclusion in the earlier study of individuals who were not classified as deafblind but claimed that either their blindness or deafness was caused by rubella.

Table 8(b) Glaucoma

		Age reported of onset of glaucoma						
Glaucoma n = 47	**Yes**	**Age Unknown**	**Birth**	**Age 3-12**	**Teens**	**20's**	**30's**	**40's**
Glaucoma reported	25 (53.2%)	3	3	7	3	4	3	**2**
Glaucoma reported (one eye)	8 (32%)							
Glaucoma reported (both eyes)	17 (68%)							

Glaucoma (Table 8(b) represents the next largest cause of vision loss among the participants in this study (53.2%). The incidence is significantly higher than that reported (32.3%) in the 1999 study. In the 1999 study, only 6.5% (2 of 31) reported the onset of glaucoma during their 20's. In the 2014 survey, nine of the 25 individuals (or 36%) reported onset during their 20's, 30's, and 40's. This suggests that glaucoma is a significant late manifestation of CRS.

According to Table 8(c), twelve or 28.6% of respondents reported corneal damage. The known age of onset of corneal damage ranged over the entire age spectrum of the

participants reporting corneal damage.

Table 8(c) Corneal Damage

		Age reported of onset of corneal damage						
Corneal Damage n = 42	**Yes**	**Unknown**	**Birth**	**Age 3-12**	**Teens**	**20's**	**30's**	**40's**
Corneal Problems reported	12 (28.6%)	5	1	2	1	1	1	1
Reported (one eye)	6 (50%)							
Reported (both eyes)	6 (50%)							

Table 8(d) presents statistics on the other causes/conditions reported related to vision loss among the respondents.

Microphthalmia, nystagmus, strabismus and rubella retinopathy are recognized as congenital eye defects. Note that the sample size for Rubella Retinopathy (RR) is low (26) with prevalence reported at 19.2%. According to the footnote reference #26, RR is one of the usual symptoms of congenital rubella syndrome but was under-reported in this study.

Table 8(d) Other Reported Causes/Conditions

		Age reported onset of other conditions						
Other Causes	**Yes**	**Unknown**	**Birth**	**Age 3-12**	**Teens**	**20's**	**30's**	**40's**
Microphthalmia[24] n=42	8 (19.1%)							
Nystagmus[25] n=39	14 (35.9%)							
Strabismus[26] n = 40	10 (25%)							
Rubella Retinopathy[27] n = 26	5 (19.2%)							
Detached Retina[28] n = 46	10 (21.7%)	7	1	1				1

[24] https://www.nei.nih.gov/health/anoph/anophthalmia. (Microphthalmia is a rare disorder in which one or both eyes are abnormally small. This rare disorder develops during pregnancy and is usually associated with other birth defects.)

[25] http://www.nlm.nih.gov/medlineplus/ency/article/003037.htm. (Nystagmus is a term to describe fast, uncontrollable movements of the eyes. The involuntary eye movements of nystagmus are caused by abnormal function in the areas of the brain that control eye movements. Nystagmus may be caused by congenital diseases of the eye.)

[26] http://www.nlm.nih.gov/medlineplus/ency/article/001004.htm. (Strabismus is a disorder in which both eyes do not line up in the same direction, so they do not look at the same object at the same time. The condition is more commonly known as "crossed eyes." When the condition is present at birth it is called congenital strabismus.)

[27] http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2814186. (Rubella retinopathy (RR) is a common manifestation of congenital rubella syndrome. It is defined by diffuse mottling of the retinal pigment epithelium (RPE) and focal areas of decreased and increased pigmentation, giving the characteristic "salt-and-pepper" fundus.)

[28] http://www.healthlinkbc.ca/healthtopics/content.asp?hwid=hw187829. (Retinal detachment usually happens because there's a tear (hole) in the retina. The most common cause of a tear is posterior vitreous detachment (PVD).Vitreous gel fluid flows through

No. reporting enucleation[29] n = 53	7 (13.2%)	3	1			2	1	
No. reporting enucleation-one eye	4 (57%)							
No. reporting enucleation-both eye	3 (43%)							

The only statistic reported in Table 8(d) for comparison with the 1999 survey was for the incidence of retinal detachments. The 2014 survey reported a detached retina incidence rate of 21.7%. This is double the reported incidence of retinal detachments in the 1999 study (11.0%).

2.2.3 Hearing Loss

Participants were asked to report the evaluation of their hearing loss. Their responses are summarized in Table 9. All 53 of the respondents reported a hearing loss. (Note that this is higher than mentioned in Table 6 (n=) as some individuals did not indicate hearing loss within their first two years of age). Most or 90.6% reported their hearing loss occurring at birth or in the very early childhood period. Almost three-quarters reported hearing loss affecting both ears. The major degree of loss reported was 'profound loss', accounting for almost half or 47.2% of all respondents. Together with the category 'severe to profound' hearing loss, 62.3% of respondents would be considered in the profound range of loss.

Comparing these results with the 1999 survey, the hearing loss reported in the severe to profound range in 1999 was 83.2% versus the 62.3% reported in the current survey. What would account for the difference between these two statistics is unknown; other than the different individuals completing the surveys and the current information that the individuals completing the questionnaires had at hand. It is probably fair to say that the hearing loss evaluations are not that precise due to the difficulty with testing as well as imprecise nature of the evaluation description.

Table 9: Summary of responses regarding evaluation of hearing loss

		Reported age of onset of hearing loss		
Evaluation of Hearing Loss	**Yes**	**Birth to 2nd year**	**3-4**	**other**
Reported Hearing Loss n=53	53 (100%)	49	3	1 (age 8)
Reported Hearing Loss (Both Ears)	38 (73.7%)	n/a	n/a	n/a
Mild Hearing Loss	0	n/a	n/a	n/a

the tear, pools beneath the retina, and lifts the retina off the back of the eye. Some of the reasons that make a person more likely to get a retinal detachment are an eye or head injury, nearsightedness, eye disease, and diabetes.)

[29] en.wikipedia.org/wiki/ Enucleation_of_the_eye. (Enucleation is the removal of the eye that leaves the eye muscles and remaining orbital contents intact. This type of ocular surgery is indicated for a number of ocular tumors, in eyes that have suffered severe trauma, and in eyes that are otherwise blind and painful.)

Moderate Hearing Loss	1 (1.9%)	n/a	n/a	n/a
Moderate to Severe Hearing Loss	1 (1.9%)	n/a	n/a	n/a
Severe Hearing Loss	3 (5.7%)	n/a	n/a	n/a
Severe to Profound Hearing Loss	8 (15.1%)	n/a	n/a	n/a
Profound Hearing Loss	25 (47.2%)	n/a	n/a	n/a
Unable to test- uses hearing	8 (15.1)	n/a	n/a	n/a
Unable to test- Does not use hearing	5 (9.4%)	n/a	n/a	n/a
Not reported	2 (3.7%)			

Participants were asked if there was any noticeable change in their hearing ability since childhood. Responses are found summarized in Table 10. This question was not answered by about one-third of the participants which generally reflects their lack of understanding of ability to measure changes. For those responding, about one quarter or 25.0% indicated there had been a change. Most causes (5) were unknown, but two reported an infection, one reported the cause was the result of self-abuse and one reported that their loss was a slow and steady decline.

Compared again to the previous survey, 30.1% of the 1999 participants reported a change in hearing loss compared to the 25.0% of participants in the 2014 survey. Again, the reasons for this difference between both surveys are unknown other that what was stated in the above section about the difference between the evaluation in hearing loss in the severe to profound category.

Table 10: Responses regarding change in hearing ability

		Reported age of onset				
	Yes	**Unknown Age**	**Childhood**	**Post Puberty**	**20's**	**40's**
Reported Change in Hearing Ability N=36	9 (25.0%)	3	1	1	3	1
Cause of Change of Hearing Ability	9	1-Infection 2-Unknown cause	1 Infection	1 Unknown	1 self-abuse; 1 slow decline; 1 Unk	**1** Unknown

The use of hearing devises by participants is summarized in Table 11. The responses were such that it was unclear what proportion of the 21 (or 39.6%) individuals were currently using hearing devises. In most instances, the individuals had used a hearing devise, either hearing aids or phonic ears during their formal school years, but were not using them in their adult years. For the majority reporting the profound nature of their

hearing loss, it was reported that their hearing devises did not seem to improve their hearing ability. Responses included: “prefers sign language over the hearing aids”; “used to wear hearing aids but not tolerant of them any longer”; ‘hearing aids cause headaches”; “used hear aids when younger but they don’t appear functional any more”; used hearing aids and FM when younger but not any longer”, etc.

The proportion (39.6%) ‘using hearing devises’ is much reduced from the number reporting (65%) using hearing devises in the 1999 survey. There may be several reasons for this: the current sample size is much lower and not totally representative of the group sampled in the 1999 study; the current participants are older now and prefer not to use hearing devises that were expected of them when in an educational setting; and more likely their hearing has deteriorated further and like some individuals have reported, the aids are not as functional as when they were younger.

Table 11: Summary of responses about the use of hearing devises

		Reported age of onset				
		Unknown Age	**Childhood**	**Post-puberty**	**20’s**	**30’s +**
Used or Using Hearing aids n = 53	21 (39.6%)	10 reported in one ear only	1-Unknown if still using			
FM system n = 53	5 (9.4%)	Used as a child in school				
Cochlear Implant n = 53	1 (1.9%)				Age 27	

2.2.4 Circulatory System Defects

Table 12 summarizes the responses by the study participants to the inquiry about the incidence of defects to their circulatory system. Of the 52 who responded, 35 or 67.3% of the participants reported an insult to their circulatory system. The three major circulatory system affects were reported in order of occurrence: heart murmur (31.4%); patent ductus arteriosis or PDA (28.6%) and pulmonary stenosis or hole in the heart (28.6%). Four individuals (7.8%) reported an unspecified congenital heart defect. As the foot note (#31) indicates, a heart murmur is not necessarily a disease but can indicate an underlying heart problem. The rate of incidence for patent ductus arteriosis (28.6%) and pulmonary stenosis (28.6%) compare well to results in Cherry (1987)[30] for patent ductus arteriosis (30%) and the various types of stenosis (25%).

The incidence of reported heart defects in the current survey (67.3%) is similar to the rate reported (65.7%) in the 1999 survey. There are, however, large differences between the two surveys regarding the proportions reported in 199 for such reported defects as: heart murmur (11.3%), patent pulmonary stenosis (17.2%) and septal defects (9.7%). In my opinion, the differences between the two surveys for these conditions reflect the accuracy of the initial identification of the defect at the time of birth,

[30] Cherry, J.D. (1987). *Rubella.* In: Feigin, R.D. and Cherry, J.D. (Eds). *Textbook of Pediatric Infectious Diseases.* 2nd Ed. W.B. Saunders, Philadelphia, pp. 1810-1841.

the memory of the family member/advocates and possibly the understanding of the caregivers preparing or assisting with the surveys in both 1999 and 2014.

Table 12: Extent and type of circulatory system defect

Reporting Heart Defects **n = 52**	**35 (67.3%)**
Heart Murmur[31]	11 (31.4%)
Patent Ductus Arteriosus[32]	10 (28.6%)
Pulmonary Stenosis[33]	10 (28.6%)
Unspecified congenital heart defect	4 (7.8%)
Congestive Heart failure[34] (1 identified at birth; 1 died age 30; 1 died age 48)	3 (5.9%)
Septal defect (unspecified)[35]	1 (2.9%
Mitral Valve Disease[36]	1 (2.9%
Born with small arteries (diagnosis not confirmed)	1 (2.9%
Pericarditis[37] (Onset at age 39)	1 (2.9%
Hypertrophic Cardiomyapathy[38]	1 (2.9%
Reporting more than one heart condition	6 (17.1%)
Patent Ductus Arteriosis + Heart Murmur	1
Patent Ductus Arteriosis + Pulmonary Stenosis	3

[31] http://www.mayoclinic.org (Heart murmurs are abnormal sounds during your heartbeat cycle. Heart murmurs can be present at birth (congenital) or develop later in life. A heart murmur isn't a disease — but murmurs may indicate an underlying heart problem.)

[32] http://www.mayoclinic.org (Patent ductus arteriosus (PDA) is a persistent opening between two major blood vessels leading from the heart. The opening, called the ductus arteriosus, is a normal part of a baby's circulatory system before birth that usually closes shortly after birth. If it remains open, however, it's called a patent ductus arteriosus.)

[33] http://www.mayoclinic.org (Pulmonary valve stenosis is a condition in which a deformity on or near your pulmonary valve, the valve that influences the blood flow from your heart to your lungs, slows the blood flow.)

[34] http://www.mayoclinic.org (Heart failure, sometimes known as congestive heart failure, occurs when your heart muscle doesn't pump blood as well as it should. Certain conditions, such as narrowed arteries in your heart (coronary artery disease) or high blood pressure, gradually leave your heart too weak or stiff to fill and pump efficiently)

[35] http://www.mayoclinic.org (A ventricular septal defect (VSD), a hole in the heart, is a common heart defect that's present at birth (congenital). The hole occurs in the wall that separates the heart's lower chambers (septum) and allows blood to pass from the left to the right side of the heart. The oxygen-rich blood then gets pumped back to the lungs instead of out to the body, causing the heart to work harder.)

[36] http://www.healthline.com (Mitral valve disease refers to conditions of the mitral valve. Located between the left chambers of your heart, this valve works to keep blood flowing properly. It allows blood to pass from your left atrium to your left ventricle but prevents it from flowing backward.)

[37] http://www.mayoclinic.org (Pericarditis is swelling and irritation of the pericardium, the thin sac-like membrane surrounding your heart. Pericarditis often causes chest pain and sometimes other symptoms. The sharp chest pain associated with pericarditis occurs when the irritated layers of the pericardium rub against each other.)

[38] http://www.mayoclinic.org (Hypertrophic cardiomyopathy (HCM) is a disease in which the heart muscle (myocardium) becomes abnormally thick (hypertrophied). The thickened heart muscle can make it harder for the heart to pump blood.)

Pulmonary Stenosis + ventricular septal defect	1
Heart murmur + unreported defect	1

Note that three of the individuals identified as adults died related to cardiac failure: two with congestive heart failure (one aged 36; one age 48); the other died at age 39 diagnosed as heart failure.

Fifteen or 30.0% of individuals reported having a history of heart related surgeries. The types of surgeries reported and the individuals' age is found in Table 13. The respondents in the 1999 survey reported having a cardiac surgery incidence rate of 24.5%.

Table 13: Reported history of cardiac surgeries

Reporting Cardiac Surgeries n = 50	**15 (30.0%**	
Type of surgery reported:		**Age of individual at time of surgery**
Heart valve (unspecified) (2)		1 age 2 and 1 age unknown
Patent ductus arteriosis ligation (3)		1 at infancy; 1 age 4 months; 1 age 3
Cardiac Catheterization[39] & Patent ductus arteriosis (1)		1st procedure at age 1; 2nd age 2
Patent ductus arteriosus and Pulmonary stenosis procedures together (2)		1 as infant; 1 - 1st procedure at 3 wks, 2nd procedure at age 1
Septal defect procedure (1)		Very young child
Open Heart surgery (1)		Aged 1
Pulmonary Stenosis & Septal defect procedure (1)		1st procedure at age 2; 2nd age 10
Pericardium drainage (1)		Age 43
Mitral Valve repair (1)		Age 22
Unknown (2)		Age unknown

Two adult-related circulatory issues were identified during the 2014 survey – hypertension and high cholesterol. See Table 14. Of the participants responding (n = 50) to inquiries about these conditions, 18.0% and 20.0% reported these two conditions, respectively. Table 14 also indicates the age of onset for those reporting these conditions.

For those reporting an age of onset for hypertension, 3 reported the onset during their 20's and one each during their 30's and early 40's. In fact, the one individual acquiring hypertension at age 35 died of this condition at age 36.

[39] http://www.nhlbi.nih.gov. (Cardiac catheterization (KATH-eh-ter-ih-ZA-shun) is a medical procedure used to diagnose and treat some heart conditions. A long, thin, flexible tube called a catheter is put into a blood vessel in your arm, groin (upper thigh), or neck and threaded to your heart. Through the catheter, your doctor can do diagnostic tests and treatments on your heart.)

For comparison with the 1999 survey, high blood pressure was reported by 7.2% of respondents. The 2014 survey shows more than double (18.0%) the rate of incidence of this condition.

For those reporting an age of onset of high cholesterol (seven of the ten), two each reported onset in their 20's, 30's and 40's while one reported onset of the condition at age 57. There are no comparable results for cholesterol with the 1999 survey.

Table 14: Reporting of incidence of hypertension and high cholesterol

			Reported age of onset of condition			
	# and %	**Unk. age**	**20's**	**30's**	**40's**	**50's**
Hypertension (High Blood Pressure (n = 50)	9 (18.0%)	4	3 (1 at 21; 2 during late 20s)	1 (age 35)	1 (age 41)	
High cholesterol (n = 50)	10 (20.0%)	3	2 (1-age 21, 1-during late 20s')	2 (age 30+, age 39)	2 (age 41, age 42)	1 (age 57)

2.2.5 Respiratory System

Participants were asked to report any problems related to their respiratory system according to three categories; allergies, asthma and pneumonia. Their responses are summarized in Table 15. Incidentally, in the question about allergies, respondents also reported whether they experienced any drug allergies. These responses were separated out from allergies related to environmental agents.

Table 15: Summary of responses to respiratory system problems

			Age of onset				
	Yes	**Unknown Age**	**0-5**	**6-12**	**Teen**	**20's**	**30's**
Respiratory Problems (n = 51)	**27 (52.9%)**						
Reporting allergies to environmental related agents (dust, molds, etc.) n = 51	16 (31.4%)						
Reporting Asthma or other lung problems including bronchitis n = 51	8 (15.7%)	6		1		1	
Reporting Pneumonia n = 51	15 (29.4%)	5	4[40]	1[41]		2	3

[40] One individual reported pneumonia first at age 7 months then several times later as an adult
[41] Individual reported pneumonia at ages 11, 39 & 40

Reporting various allergies, asthma and pneumonia n = 51	3 (5.9%)						
Reporting various allergies and pneumonia n = 51	7 (13.7%)						
Reporting various allergies and asthma n = 51	3 (5.9%)						
Reporting asthma and pneumonia n = 51	1 (2.0%)						
Reported allergies to various drugs (antibiotics) n = 51	13 (25.5%)						

According to Table 15, the overall incidence rate of reporting various respiratory issues such as environmental allergies, pneumonia and asthma is 52.9%. The percentage distribution of those three issues is as follows: 31.4% allergic to such environmental agents as dust, dander, pollen, molds, weeds, scents; 29.4% reported having pneumonia at some point during their lives and 15.7% reported asthma and other lung problems including bronchitis. Table 15 also depicts numbers of individuals reporting combinations of these issues.

Thirteen or 25.5% of respondents reported allergies to various types of antibiotics.

2.2.6 Musculoskeletal Conditions

Participants were asked whether they had experienced any musculoskeletal conditions in their lifetime, including (a) such spinal conditions as scoliosis, lordosis and kyphosis, (b) the presence of osteoporosis, (c) arthritis and d) whether they experienced multiple conditions. Their responses are summarized in Table 16.

Table 16: Summary of responses regarding musculoskeletal conditions

			Age First Reported					
	Yes	**Unknown Age**	**0-5**	**6-12**	**Teen**	**20's**	**30's**	**40+**
Physical Conditions (n=51)	**18 (35.3%)**							
a) Spinal condition: (10)	**19.6%**							
Reporting Scoliosis[42](only)	4	1		1		1		**1**
Reporting Kyphosis[43] (only)	3	1	1 (birth)	1				
Reporting Scoliosis & Kyphosis	3				2			**1**
b) **Osteoporosis**[44] (7)	**13.7%**	1			1	2	3	

[42] www.mayoclinic.org. (Scoliosis is a sideways curvature of the spine that occurs most often during the growth spurt just before puberty)

[43] http://www.spine-health.com. (Kyphosis, or "dowager's hump," is an unnatural curving of the upper back that creates a hunchback appearance in the posture, often associated with osteoporosis.)

[44] http://www.osteoporosis.ca (Osteoporosis is a disease characterized by low bone mass and deterioration of bone tissue. This leads to increased bone fragility and risk of fracture (broken bones), particularly of the hip, spine, wrist and shoulder.)

c) **Arthritis**[45] (7)	**13.7%**	2		1		2	1	**1**
d) **Reporting multiple physical conditions** (4)	**7.8%**							
Reporting scoliosis + osteoporosis + arthritis	1							
Reporting scoliosis + arthritis	1							
Reporting Kyphosis + osteoporosis	1							
Reporting osteoporosis + arthritis	1							

Just over one-third (35.3%) of respondents reported one or more musculoskeletal condition. Of those conditions, 10 or 19% reported a spinal condition, either scoliosis (4), kyphosis (3) or both (3) together. One individual reporting kyphosis also indicated having Reynaud's Disease.[46]

In the 1999 survey, 26% of the respondents reported a spinal condition.

Seven or 13.7% reported having osteoporosis and seven (13.7%) experiencing arthritis. Four or 8% reported having more than one of the physical conditions. The 1999 survey reported 7.1% of the respondents having osteoporosis, one half of the incidence reported in this survey. The presence of arthritis was not a question in the 1999 survey.

What is interesting about the incidence of osteoporosis and arthritis is the relatively early reported age of onset reported by some of the individuals. While early incidence of osteoporosis may be connected to the individuals' lack of physical activity, one has to wonder about the auto-immune connection to the early age of onset of arthritis.

2.2.7 Neurological Impairment

Participants were asked a series of questions to ascertain any indications of neurological problems. The questions were organized into three categories: a) whether the individual had a history of seizures, b) was there any noticeable degeneration of motor skills, and c) was there any noticeable degeneration of cognitive skills, as evidenced by indications of poor attention and distraction (i.e. forgets instructions easily, has trouble concentrating on tasks); disorientation (i.e. regarding time or place); impaired short-term memory (i.e. evidence of difficulty forming new memories, misplacing objects) and impaired long term memory (i.e. well learned memories appear lost). The responses to the questions are summarized in Table 17.

[45] http://www.medicalnewstoday.com (Arthritis is an inflammation of the musculoskeletal system, specifically the joints. It is the main cause of disability among people over fifty-five years of age in industrialized countries. Arthritis may be caused by autoimmune disease)

[46] http://www.mayoclinic.org (Raynaud's (ray-NOHZ) disease causes some areas of your body — such as your fingers and toes — to feel numb and cold in response to cold temperatures or stress. In Raynaud's disease, smaller arteries that supply blood to your skin narrow, limiting blood circulation to affected areas (vasospasm).

Table 17: Summary of responses regarding possible neurological impairment

		Age first reported						
	Yes	**Unk age**	**Age 0-5**	**6-12**	**teen**	**20's**	**30's**	**40+**
Neurological Problems Reported N=51	25 (49%)							
(a) Seizures	14 (27.5%)	1	4	1	2	3	1	2
(b) Motor Skill Degeneration	10 (19.6%)	1	2			5	2	
(c) Cognitive Degeneration								
Demonstrating 1 of the following	17 (33.3%)							
• Demonstrating poor attention and distraction	15 (88.2%)	11		1		1		2
• Experiencing disorientation	4 (23.5%)	1				1	1	1
• Evidence of impaired short-term memory	3 (17.7%)	3						
• Experiencing impaired long-term memory	0							
• Demonstrating poor attention, distraction, & disorientation	4 (23.5%)							
• Demonstrating poor attention, distraction and short term memory loss	2 (11.8%)							
• Demonstrating poor attention, distraction, disorientation & short term memory loss	1 5.9%							

According to Table 17, twenty-five or 49% of the respondents reported one or more neurological related problems. Fourteen or 27.5% reported experiencing seizures during some time in their lives. As per Table 17, age of first onset ranged over all the age groups through to the 40's. Of the fourteen reporting seizures, eight (or 57%) reported their condition remained the same, while 6 or (43%) indicated the problem with their seizures had decreased.

The proportion of respondents experiencing seizures (27.5%) compares closely to the proportion reporting seizures (30%) in the 1999 survey. Similarly, the age of first onset

ranged over all the age classes (except during their 40's) reporting fifteen years ago. According to Dr. Jude Nicholas (Personal Communication)[47], this unchanged pattern of reported seizures may suggest that the underlying cause for the seizures could be congenital. When the cause is congenital, the seizures tend to be present in early childhood through adolescence. Furthermore, the reported seizures may indicate signs for a diffuse brain dysfunction[48].

The rate of incidence for those reporting a degeneration of motor skills was 19.6% (ten individuals). Of those reporting their known age of onset, five individuals were in their 30's while two were in their 40's. The 1999 survey asked participants to report any deterioration in walking, balance or fine motor skills. The incidence rate for the 100 individuals responding to those inquiries was 21.2%, 23.2% and 4.1%, respectively. Except for the fine motor skills, the current incidence rate for degeneration of motor skills is quite comparable to the 1999 results.

For the third indicator of neurological impairment (cognitive degeneration) shown in Table 17, seventeen or 33.3% of respondents reported one indicator of cognitive degeneration. Fifteen of the individuals demonstrated poor attention and distraction, four experienced disorientation, while three showed evidence of impaired short-term memory. No one showed any evidence of experiencing impaired long term memory loss. Seven of the individuals showed various combinations of those three indicators of cognitive degeneration.

The only data from the 1999 survey for any comparison was that 12.6% demonstrated some indications of memory loss while 14.3% reporting various neurological conditions (disorientation, wandering, forgetfulness, etc.). If we considered that the 1999 list of neurological conditions was similar to what was referred to as 'degeneration of cognitive skills' in current survey, then we could safely say that over twice as many individuals in the 2014 survey are demonstrating some degree of degenerating cognitive skills.

According to Dr. Jude Nicholas (Personal Communication), this pattern of cognitive disturbances is often the hallmark of a diffuse encephalopathy. Encephalopathy is the term used to describe a general alteration in brain function, manifesting as an attentional disorder with less or no progressive loss of long-term memory. The encephalopathy may be due to the prenatal rubella infection.

2.2.8 Psychosocial and Behavioral Issues

It is with respect to these issues that family members and Intervenors have the most concerns regarding the 'mental health' of individuals who are deafblind in their care. To help evaluate this issue, family members and/or caregivers were asked to report on whether the individuals demonstrated specific behaviors described as stereotypical,

[47] Dr. Jude Nicholas, Neuropsychologist, Vestlandet Resource Centre, Bergen, Norway

[48] www.merckmanuals.com/.../brain...disorders/brain-dysfunction/overview...

self-injurious, compulsive, ritualistic, sameness, restrictive, aggressive or classed as angry outbursts or rage. Definitions were provided (see the footnotes #'s 49-54) to assist those completing the questionnaires.
The questionnaire was designed to seek information about the age of first onset of these behaviors (if prevalent) and an evaluation of whether the behavior increased or decreased or stayed the same during the past year.

Table 18 summarizes the results of the inquiries about the prevalence of each behavior while Table 19, summarizes the behavior's status in the past year.

Table 18: Demonstrating Difficulties with Behavioral Control

			Age of onset of specific behaviours					
Specific Behavior	**Reported**	**Unk. age**	**Age 0-5 yrs**	**Age 6-12**	**Teens**	**20's**	**30's**	**40's**
Stereotyped behavior[49] (n=47)	34 (72.3%)	18	10	1	2	2	1	
Self-injurious behavior[50] (n=52)	37 (71.2%)	11	14	6	2	2	1	1
Compulsive behavior[51] (n=50)	21 (42.0%)	14	3		1	3		
Ritualistic behavior[52] (n=51)	29 (56.9%)	16	7	2	1	2		1
Sameness behavior[53] (n=49)	22 (44.9%)	13	5			2		
Restricted behavior[54] (n=47)	16 (34.0%)	11	1	2			2	
Aggression to others (n=52)	27 (51.9%)	6	6	10	2	2	1	
Episodes of angry outbursts or rage (n=52)	36 (69.2%)	9	13	4	5	2	3	

According to the results in Table 18, the rates of observation of these eight behaviors in descending order are: Stereotyped behavior (72.3%), Self-injurious behavior (71.2%), Episodes of angry outbursts or rage (69.2%), Ritualistic behavior (56.9%), Aggression to others (51.9%), Sameness behavior (44.9%) and Restricted behavior (34.0%).

Regarding the age of onset, most respondents were not able to provide an accurate

[49] Stereotyped behavior definition: apparent purposeless movements or actions i.e. with the whole body, head, hands, using objects, eyes and ears, etc. that are repeated in a similar manner.
[50] Self-injurious behavior definition: movement or actions i.e. hitting self with hands or objects, biting, pulling, rubbing, picking etc., that have the potential to cause redness, bruising, or other injury to the body, and that are repeated in a similar manner
[51] Compulsive behavior definition: behavior that is repeated and is performed according to a rule, or involves things being done "just so", i.e. arranging, washing, cleaning, counting, hoarding, repeating, touching, etc.
[52] Ritualistic behavior definition: performing activities of daily living in a similar manner, i.e. eating routines, bedtime routines, travel details, leisure and communication activities, etc.
[53] Sameness behavior definition: resistance to change, insisting that things stay the same, i.e. items remain in same place, objects to new places, insists on same walking patterns, sitting locations and activities, etc.
[54] Restricted behavior definition**:** Limited range of focus, interest or activity, i.e. fascination with one object or activity and part thereof.

age. For those that were able to provide an age, the onset was primarily reported during the years up to and into their teenage period.

The 1999 survey reported results about some of these types of behaviors, such as self-injurious behavior (61%), aggressive behavior (65%), angry outburst (73.7%) and ritualistic behaviors (30.6%).

According to Dr. Jude Nicholas (Personal Communication), the high incidence of reported difficulties with behavioral control such as the stereotyped behavior (72.3%), self-injurious behavior (71.2%), episodes of angry outbursts or rage (69.2%), ritualistic behavior (56.9%) and aggression to others (51.9) could be linked to the emotional problems or mood disorder observed in individuals who are deafblind.

Averaging out the rate prevalence of the eight behaviors over the past year (Table 19), most behaviors 'remained the same' (64.5%), 23.0% of 'the behaviors decreased', while 12.5% 'reported an increase'. According to the 1999 survey, 30.3% of the respondents indicated that behaviors had increased over the past years while 53.5% reported a decrease.

Table 19: Prevalence in the past year regarding difficulties with behavioral control

	Prevalence of the behavior in the past year		
	Increased	**Decreased**	**Remained the same**
Stereotyped behavior n=34 of 34 reporting	2 (5.9%)	7 (20.6%)	25 (73.5%)
Self-injurious behavior n=33 of 37 reporting	4 (12.1%)	12(36.4%)	17 (51.5%)
Compulsive behavior n=22 or 22 reporting	4 (18.2%)	0.0%	18 (81.8%)
Ritualistic behavior n=29 of 29 reporting	0.0%	1 (3.5%)	28 (96.5%)
Sameness behavior n=22 of 22 reporting	3 (13.6%)	5 (22.7%)	14 (63.7%)
Restricted behavior n=15 of 16 reporting	4 (26.7%)	0	11(73.3%)
Aggression to others n=27 of 27 reporting	4 (14.8%)	13 (48.2%)	10 (37.0%)
Episodes of angry outbursts or rage n=36 of 36 reporting	3 (8.3%)	19 (52.8%)	14 (38.8%)

2.2.9 Depression[55]

Further to gathering information on the evaluation/observation of various psychosocial behavior tendencies of the respondents, the survey inquired whether the participants exhibited signs of depression. According to Table 20(a), 43.5% of those reporting indicated they exhibited signs of depression. For those indicating a known age of onset, the age of onset ranged almost similarly over the entire age spectrum, from infancy through the 40's. Interestingly, in the 1999 Canadian survey, 44.4% showed indications of depression.

Table 20(a): Individuals Demonstrating Signs of depression

			Age of onset of signs of depression					
	Yes	**Unk. age**	**0-5 yrs**	**6-12**	**Teens**	**20's**	**30's**	**40's**
Respondents showing signs of depression (n=47)	21 (44.7%)	10	2	1	3	2	1	2

Those completing the questionnaires were asked to base their response according to a number of determinants of depression. Table 20(b) shows the breakdown of the signs of depression determinants expressed by the twenty (of the 21) individuals demonstrating signs of depression.

Table 20(b):

			Reporting signs of depression determinants				
Reporting Depression (20 of 21)	Appearing depressed	Avoiding social contact	Cries often	Not taking part in activities once of interest	Appears restless or fidgety	Complains of headaches or other aches and pains	Eating too much or two little
	6 (30%)	4 (20%)	8 (40%)	8 (40%)	10 (50%)	6 (30%)	8 (40%)

Along with indicating 'the signs of depression', the respondents were then asked to evaluate whether the signs of depression behavior increased, decreased or stayed the same over the past year. The results of this evaluation are presented in Table 20 (c).

The majority (52.6%) indicated their signs of depression had remained the same, 36.8% indicated the signs had decreased and while only 10.6% showed an increase in their 'signs of depression'.

[55] According to Wikipedia (http://en.wikipedia.org), Depression is a state of low mood and aversion to activity that can affect a person's thoughts, behavior, feelings and sense of well-being. People with depressed mood can feel sad, anxious, empty, hopeless, helpless, worthless, guilty, irritable, ashamed or restless. They may lose interest in activities that were once pleasurable, experience loss of appetite or overeating, have problems concentrating, remembering details or making decisions, and may contemplate, attempt or commit suicide. Insomnia, excessive sleeping, fatigue, aches, pains, digestive problems or reduced energy may also be

Table 20 (c): Rate of change of signs of depression

Number reporting Signs of depression during past year	Increased	Decreased	Remained the same
n=19	2 (10.6%)	7 (36.8%)	10 (52.6%)

For those that reported a change in signs of depression (19 of 20), 52.6% reported no change, 36.8% reported the signs decreased, while 10.6% reported an increase in the signs**.**

According to Dr. Jude Nicholas (Personal Communication), the reported signs of decreased mood of individuals in this study could be associated with the reported difficulties with behavioral control. 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.

2.2.10 Sleeping disorders

Sleeping is recognized as a common concern reported within the population of individuals who are deafblind. This issue is also considered as another recognized sign or indicator of depression. The survey inquired of the participants whether there was an indication of a sleeping disorder. According to Table 21(a), fifty-eight percent reported there was a sleeping disorder. The incidence rate of 58% of respondents reporting sleeping disorders is much higher than the 39.4% reported in the 1999 survey.

Table 21 (a): Individuals reporting sleeping disorders

Respondents reporting sleeping disorders	Yes
n=51	30 (58.8%)

Table 21(b) summarizes the reporting of the various sleeping disorder signs among the 30 individuals reporting a disorder with their sleeping. With the exception of 'sleeping for excessive periods of time' at 36.7%, just over half of the others reported the other four signs. Two or 6.9% reported having sleep apnea.

Table 21(b): Reporting various signs of sleeping disorders

Reporting Sleeping disorders n=30	Difficulty sleeping when going to bed	Waking in the middle of the night with difficulty returning to sleep	Waking very early in the morning with difficulty returning to sleep	Staying awake for excessive periods of time	Sleeping for excessive periods of time	Sleep Apnea
	16 (53.3%)	**16 (53.3%)**	**16 (53.3%)**	**15 (50.0%)**	**11 (36.7)**	**2 (6.7)**

The participants were asked whether their reported symptoms of sleep disorder were increasing, decreasing or staying the same. The results for the 30 individuals reporting

a disorder with their sleeping are shown in Table 21 (c).

Table 21(c): Rate of change of symptoms of sleeping disorders

For those reporting sleeping disorders, were the symptoms:	Increasing	Decreasing	Staying the same
n=30	13 (43.3%)	3 (10.0%)	14 (46.7%)

As Table 21(c) indicates, only 10.0% of the individuals (reporting sleeping disorders) indicated that their symptoms were decreasing. This compares to 46.7% who report their symptoms staying the same and 43.3% reporting a worsening of their symptoms. While the survey did not ask about the age of onset of sleeping disorders, we can assume that this is now becoming an issue with ageing.

According to Dr. Jude Nicholas (Personal Communication), the high incidence of sleeping disorders (58.8 %) reported in Table 21(a) could also be linked to the reported signs of depression (Table 20(b)). Sleep and mood affect each other. Disturbed sleep patterns or lack of sleep can alter mood significantly. On the other hand, being depressed puts a person in high risk for chronic sleeping disorders.

2.2.11 Endocrine System

Family members and Intervenors of individuals with congenital rubella syndrome are usually curious whether the increasing dysfunction of CRS individuals' endocrine system is a late manifestation of CRS. According to Table 22(a), 42.3% of all individuals reported an endocrine dysfunction. Just over twenty one percent (21.2%) had either diabetes or thyroid dysfunction. One of the individuals (a male) had both diabetes (Type 2) and hyperthyroidism. For this person, the age of onset of both conditions was during his 20's. He eventually died at age 36.

The incidence of both diabetes and thyroid condition reported in this survey are almost double what was reported in the 1999 study when 10% were diagnosed with a thyroid condition and 12.1% diagnosed with diabetes.

In this regard, it's worth repeating the following statement made in the 1999 report from Remington and Klein (1990)[56]: "for example, if diabetes is 100 to 200 times more prevalent (for an individual with congenital rubella than the general population), this is all the more reason to be monitored regularly for this serious health condition. The same can be said for thyroid condition."

[56] Remington, J.S and Klein, J.O. (Editors) 1990. *Infectious Diseases of the Fetus and Newborn Infant.* 3rd Ed. W.B Saunders, Philadelphia.

Table 22 (a): Incidence of reported endocrinal conditions

		Age of onset of diabetes or thyroid						
	Yes	**Age unk**	**Age 0-5**	**Age 6-12**	**Teens**	**20's**	**30's**	**40+**
Reporting diabetes/thyroid issues (n=52)	22 (42.3%)							
Total reporting Diabetes (8 males; 3 females)	11 (21.2%)							
Reporting Type 1 Diabetes (3 males; 2 females)	5	1	1		1	1	1	
Reporting Type 2 Diabetes (5 males; 1 female)	6	1				2	1	2
Total reporting Thyroid issues (5 males; 6 females)	11 (21.2%)							
Reporting Hypothyroidism (3 males; 5 females)	8	3			1	1	2	1
Reporting Hyperthyroidism (2 males; 1 female)	3	2				1		
Reporting Diabetes and hyper-thyroidism (male)	1							

In the questionnaire section related to the endocrine system, males and females were asked specific questions related to their reproductive systems. Only females reported any gender specific endocrine related issue other than the ones previously discussed. Table 22(b) outlines the results regarding the reports about the prevalence of hirsutism[57] and polycystic ovarian disease[58] which are endocrine gland related issues.

Table 22(b) Other Female Endocrine Related Issues

	Age of onset of hirsutism or polycystic ovarian disease							
	Yes	**Age unk**	**Age 0-5**	**Age 6-12**	**Teens**	**20's**	**30's**	**40's +**
No. of females reporting Hirsutism N=21	7 (33.3%)	3				1	2	1
No. of Females reporting polycystic ovarian disease n=20	1 (5%)	1						

Seven or 33.3% of the females reporting (21) indicated the presence of hirsutism. According to the Canadian Women's Health Network (http://www.cwhn.ca) this condition affects 6% to 10% of women. The results in this survey far exceed the

[57] http://www.mayoclinic.org. (Hirsutism (HUR-soot-iz-um) is a condition of unwanted, male-pattern hair growth in women. Hirsutism results in excessive amounts of stiff and pigmented hair on body areas where men typically grow hair — face, chest and back. Hirsutism may arise from excess male hormones called androgens, primarily testosterone.

[58] http://www.mayoclinic.org. (Polycystic ovary syndrome (PCOS) is a common endocrine system disorder among women of reproductive age. Women with PCOS may have enlarged ovaries that contain small collections of fluid — called follicles — located in each ovary as seen during an ultrasound exam.)

CWHN's statistics. Two of the individuals reporting hirsutism also reported diabetes, one with Type 1, the other with Type 2.

Only one (5%) of the twenty females indicated having polycystic ovarian disease. That individual female also reported hirsutism.

2.2.12 Urogenital Tract

Participants were asked to report any unusual issues with respect to their urogenital tracts. Table 23 lists the specific issues that males and females reported about personal related problems.

Table 23: Listing of reported male and female urogenital tract issues

Females (2)

- Fibroids
- Born with a double uterus

Males (10)

- Testicular cancer
- Orchidectomy[59] (age 14)
- Enlarged prostate and kidney stones (age 48)
- Individual reporting difficulties from birth with urogenital tract (bladder) issues including incontinence and a urethra implant. The bladder surgeries occurred in early childhood; the urethra implant happened during later childhood.
- Undescended testes with hormone shots (age 6)
- One testicle removed (age 19)
- Recurring torsion (twisting of spermatic cord) of testes
- Hydrocele[60] operation (pre-teen)
- Both testes removed (age 6)
- Double hernia operation (age 5 months) resulting in loss of 1 testicle

It appears from Table 23 that urogenital tract abnormalities are more a male related issue in this study group. Of the 24 males where any information was reported, 10 or 41.7% reported an issue. In the 1999 study, 12 (of 42 males) or 28.6% reported a problem with their urogenital tract, and the issues were largely related to undescended testes. This issue was reported once among those who provided an answer in the current survey.

2.2.13 Gastrointestinal

Questions were posed to participants to learn about any difficulties they experienced with eating and digestion. A summary of their responses is outlined in Table 24. Just over fifty percent of the participants (50.9%) reported at least one of the problem areas listed. Compared to the 1999 survey, 40% of the respondents reported problems with

[59] medical-dictionary.thefreedictionary.com/orchidectomy
[60] www.mayoclinic.org/diseases-conditions/hydrocele

one of more issues related to chewing, swallowing, gagging and vomiting. The new issues added to the 2014 survey included questions about esophageal reflux[61] and reporting a special diet.

Table 24: Incidence of gastrointestinal issues reported

	Yes	Unk Age	Age 0-5	6-12	Teens	20's	30's	40+
Reporting Gastrointestinal Problems n=53	27 (50.9%)							
Reporting Eating Difficulties	11 (20.8%)	5	5					1
Reporting Cyclic Vomiting[62]	3 (5.7%)	1				1	1	
Reporting Gagging	8 (15.1%)	6					1	1
Reporting Swallowing Difficulties	11 (20.8%)	7	2				1	1
Reporting Esophageal Stricture[63]	3 (5.7%)	1					1	1
Reporting Esophageal Reflux	12 (22.6)	11					1	
Reporting being on a special diet	11 (21.8%)	11						
Special diets included: • Pureed foods (5 individuals) • Special carbohydrate diet (2) • Halal foods only (1) • Gluten free diet (1) • Special diet for diverticulosis[64] (1) • Special diet for diabetes (1)								

In addition to reporting on the set questions, some of the participants reported having the following special gastrointestinal issues:

- One individual had diverticulosis diagnosed at age 36
- One individual was diagnosed with a bowel blockage at age 30 resulting in surgery followed by a feeding tube for 3 months; in the same individual was discovered at age 38 a congenital diaphragmatic hernia[65]

[61] http://www.nlm.nih.gov. (Gastroesophageal reflux disease (GERD) is a condition in which the stomach contents leak backwards from the stomach into the esophagus (the tube from the mouth to the stomach). This can irritate the esophagus and cause heartburn and other symptoms.)

[62] www.niddk.nih.gov/health-information/.../cyclic-vomiting-syndrome

[63] https://www.google.ca/webhp?sourceid=chrome-instant&ion=1&espv=2&ie=UTF-8#

[64] www.webmd.com. (Diverticulosis is a condition that develops when pouches (diverticula) form in the wall of the colon, part of the large intestine).

[65] http://www.nlm.nih.gov. (A diaphragmatic hernia is a rare birth defect in which there is an abnormal opening in the diaphragm.

- Two individuals were diagnosed with a narrow palate
- One individual developed a hiatal hernia[66] at age 40

2.2.14 Medications

Participants were asked to document the types of medication they were prescribed, their purpose and frequency. For the project leader, this was usually a confirmation of any reported health issues or a reflection of a possible problem that was not reported. The responses from those who reported about this topic (50 of 53) are summarized in medication-type categories and presented in Table 25.

Table 25(a): Summary of responses regarding the use of various types of medications

Medication Type N=50	No. using	Medication Type	No. using	Medication Type	No. using
Anti-anxiety; mood stabilizers; anti-depressants; PRN's[67]	28 (56.0%)	Pain medication	8 (16.0%)	Osteoporosis	3 (6.0%)
Intestinal issues: laxatives; stool softeners etc.	13 (26.0%)	Diabetes	7 (14.0%)	Ulcers	2 (4.0%)
Seizures	12 (24.0%)	Acid reflux	6 (12.0%)	Birth Control	2 (4.0%)
Allergies; Asthma; Breathing issues	12 (24.0%)	Blood pressure	7 (14.0%)	Menstrual Cycle	2 (4.0%)
Ophthalmic	11 (22.0%)	Skin problems	6 (12.0%)	Arthritis	2 (4.0%)
Glaucoma	10 (20.0%)	Cholesterol	6 (12.0%)	COPD	1 (2.0%
Thyroid	10 (20.0%)	Sleeping aids	5 (10.0%)	Incontinence	1 (2.0%
Health and nutritional support – including vitamins and minerals	10 (20.0%)	Heart related	3 (6.0%)		

It is quite obvious from this table that most of these individuals with CRS face a large number of medical conditions that require medications. Twenty-three medication general types were identified; all but perhaps one (Health and Nutritional support) for which medical prescriptions were necessary. The major type of medication reported was in the category 'Anti-anxiety, mood stabilizers, PRN's[68]', for which 56.0% were prescribed various medications. Together with the 24.0% prescribed medication to

The diaphragm is the muscle between the chest and abdomen that helps you breathe. The opening allows part of the organs from the belly (stomach, spleen, liver, and intestines) to go up into the chest cavity near the lungs.)

[66] http://www.mayoclinic.org. (A hiatal hernia occurs when part of your stomach pushes upward through your diaphragm. Your diaphragm normally has a small opening (hiatus) through which your food tube (esophagus) passes on its way to connect to your stomach. The stomach can push up through this opening and cause a hiatal hernia.)

[67] medical-dictionary.thefreedictionary.com/PRN

[68] www.medicinenet.com (p.r.n.: Abbreviation meaning "when necessary" (from the Latin "pro re nata", for an occasion that has arisen, as circumstances require, as needed).

control seizures, this represented 80% of all the participants taking medication for neurological and psychosocial/behavioral purposes.

For those who reported (50) using medications, every individual reported taking a least one type of prescription medications. The frequency distribution of individuals taking different types of mediations is shown in Table 25(b). Taking medications for three types of conditions showed the highest frequency (38.0%), followed by taking four types (28.0%), taking two types (26.0%). The least frequent was four individuals (8.0%) reporting taking just medication for one condition. It should be mentioned that, except for those reporting one medication, that most of the individuals reported taking several different types of medications for pain, skin conditions, health and nutritional, eye problems etc. There were too many to mention.

Table 25(b): Rate of taking medications

Number of medical conditions for which medications are prescribed:	Number (n=50)
One	4 (8.0%)
Two	13 (26.0%)
Three	19 (38.0%)
Four	14 (28.0%)

It is interesting to compare some of the rates of use of medications mentioned in Table 25(a) with the results in the 1999 survey. See Table 25(c).

Interestingly, the rate of medication use for the 'Anti-anxiety; mood stabilizers; anti-depressants; PRN's' category was much higher in the 1999 survey (79.5%) than the 55.1% reported in this study. For the other conditions where medication was prescribed, the reported rate was much higher in 2014 for the treatment of diabetes, seizures, thyroid condition and high blood pressure. In the 1999 survey, more individuals were treated for osteoporosis than in the current survey.

Table 25(c): Comparison of the rate of taking certain medications, 1999 vs 2014 survey

Medication Type for:	Rate of use of medications 2014	Rate of use of medications 1999
Anti-anxiety; mood stabilizers; anti-depressants; PRN's	56.0%	79.5%
Seizures	24.0%	16.0%
Thyroid condition	20.0%	9.2%
Diabetes	14.0%	8.2%
High blood pressure	14.0%	6.2%
Osteoporosis	6.0%	8.3%

2.3 Section C: Lifestyle, educational history and communication

While this study had a major focus to report on health issues experienced by the individuals with CRS, a secondary focus was to report on the lifestyle (living arrangement) of these individuals, their educational history, their communication capabilities and their employment history.

2.3.1 Living Situation of Participants

Table 26 outlines the responses from the fifty-three participants about their current living arrangements.

Table 26: Living arrangement reported by participants

	Number reported
Living in supported independent living residence for persons who are deafblind	20 (37.7%)
Living in other type of supported group home	18 (34.0)
Living with their natural family	13 (24.5%)
Living with adopted family	1 (1.9%)
Living Independently	1 (1.9%)
Total	53

The vast majority (71.7%) of these individuals live in supported living facilities. Of the remainder, 26.4% live in a family setting while only one individual reported living totally independently.

2.3.2 Educational History

The educational experiences reported by the participants are presented in Table 27. The vast majority (80%) report having attended a specialized educational program. Eighteen percent (9) reported combining either home or public schooling along with attending a specialized program. Of the 40 individuals who reported attending a specialized school, 35 of those were known to have attended the W. Ross Macdonald School for the Visually Impaired and Deafblind[69] in Brantford, Ontario. Four were known to have attended the Interprovincial School for the Deaf[70] in Amherst Nova Scotia which had a specialized program for deafblindness before it closed in 1995. Details were not provided about the other individual who reported attending a specialized school.

For those 40 individuals reported having attended a specialized school or combination with home or public school, with two exceptions they reported attending school for 12-15 years. The two exceptions reported attending for four and eight years.

[69] www.psbnet.ca
[70] http://www.archivescanada.ca

Only two individuals of the total reporting indicated they had post-secondary education.

Table 27: Educational history reported by participants

	Number reporting
Specialized school	31 (62.0%)
Home school plus specialized school	5 (10.0%)
Public school plus specialized school	4 (8.0%)
Public school	4 (8.0%)
Home school	4 (8.0%)
No schooling	2 (4.0%)
Total reporting	50

2.3.3 Employment History

To gain a further understanding of the lifestyle of those individuals with congenital rubella syndrome, questions were asked about their employment history. Their responses about are summarized in Table 28.

Table 28: Reported employment history

	Number reporting	Details
Reporting not Currently Employed and/ unable to work	41 (77.3%)	
Currently Working Part Time	10 (18.9%)	5 work part time but work not specified; 2 work in delivery services; 1 reported working in a cafeteria; 1 stocks shelves in a pharmacy; 1 is an Avon sales representative
Currently Working Full Time	2 (3.8%)	1 works as a cashier; 1 works in delivery services
Total	53	

The vast majority of the respondents (77.3%) reported currently not employed or not able to work. Ten or 18.9% report various part time work activities while only 2 (3.8%) report having a full-time job.

2.3.4 Communication

The questionnaire was set up to ask individuals to respond to their usage of the many modes of communication that are available to individuals with deafblindness. Table 29 summarizes the reported use of the various modes of communication by the 53 individuals who responded. Their responses are organized into those individuals fitting

in three levels of communication capabilities: Level 1, characterized by Speech, American Sign Language (ASL) and Manual Alphabet, and used by individuals having some limited vision and hearing ability or those deaf individuals with some vision; Level 2, characterized by Signed English (see footnote 72) and individually adapted sign language used by individuals primarily deaf with very limited vision; and Level 3, characterized by individuals with very limited communication abilities due to vision and hearing loss, primarily using touch.

Table 29: Reported use of the various modes of communication

Level 1	
Speech (only)	2
American Sign Language[71] (only)	3
American Sign Language + Speech	2
American Sign Language + Manual alphabet[72]	3
Manual alphabet (only)	1
American Sign Language + Signed English[73]	2
Level 2	
Signed English (only)	4
Signed English + Individually adapted sign language[74] + Gestures/Objects/Body Language	12
Signed English + Gestures/Objects/Body Language	14
Level 3	
Individually adapted sign language (only)	2
Individually adapted sign language + Gestures/Objects/Body Language	5
Gestures/Objects/Body Language (only)	3
Total	53

According to these criteria, the communication ability of the 53 individuals is as follows: Level 1 (24.5%), Level 2 (56.6%) and Level 3 (18.9%).

Participants were asked about their communication opportunities. Their responses to the questions are organized in Table 30: were there people to communicate with at home and/or in a work or volunteer setting. They were also asked if they had the availability of Intervention Services.[75]

[71] http://en.wikipedia.org/wiki/American_Sign_Language. (American Sign Language (ASL) is the predominant sign language of deaf communities in the United States and most of Anglophone Canada)
[72] http://en.wikipedia.org/wiki/American_manual_alphabet. (The American Manual Alphabet is a manual alphabet that augments the vocabulary of American Sign Language when spelling individual letters of a word is the preferred or only option, such as with proper names or the titles of works. Letters are signed with the dominant hand, and in most cases with the palm facing the viewer.)
[73] http://en.wikipedia.org/wiki/Manually_coded_English#Signed_English_.28SE.29_.E2.80.93_American. (Different systems called 'Signed English' have been developed in Australia, New Zealand, Ireland, the UK, the US, Kenya, and South Africa. However each 'Signed English' has borrowed signs from the local deaf sign language and invented new signs to represent the words and grammar of English. They tend to follow a loose logic of sound rather than the strict phonetic structure of Cued Speech. Signed English tends to be slower than spoken English, and teachers using it have usually found themselves 'cutting corners' and reverting to Contact sign.
[74] Custom made sign language to fit the individual's ability
[75] http://www.cdbaontario.com/about/intervenor_e.php

Table 30: Communication opportunities

	Reporting Yes
a) Are there people with whom the participant can communicate at home? (n=53)	51 (96.2%)
b) Are there people with whom the participant can communicate in a work or volunteer setting? (n=16)	14 (87.5%)
c) Does the participant have the availability of Intervention Services? (n=51)	44 (86.3%)

According to Table 30(a), most participants (96.2%) reported that there were people in their home that they could communicate with. For the much smaller number of individuals reporting who worked or volunteered (see Table 28) in various settings, Table 30(b) indicates that most of them (87.5%) had people available in those settings to communicate with them. Primarily, this communication occurs through Intervention Services (Table 30(c)), which indicates that 86.3% of the participants receive these services. As indicated in Table 26, many of these participants live in independent living residences for individuals who are deafblind which provide up to 24-hour Intervention services or on a need basis. These residences are operated in the Province of Ontario by the Canadian Deafblind Association (Ontario Chapter), Deafblind Ontario Services, Lions McInnes House and Rotary Cheshire Homes. In the Province of Saskatchewan, they are operated by Canadian Deafblind Association (Saskatchewan Chapter). Some individuals live in other types of supported independent living facilities, including family homes, and also receive varying levels of Intervention services support.

Further to Table 30(c), the 44 individuals receiving Intervention services reported on the amount of services they received on a daily basis. The results are summarized in Table 31.

Table 31: Amount of Intervention Services reported

Reporting Yes to Receiving Intervention Services (44)	Number reported
No. reporting IV 24 hours per day	28 (63.6%)
No. reporting IV 16-20 hours per day	0
No. reporting IV 11-15 hours per day	2
No. reporting IV 6-10 hours per day	6
No. reporting IV 2-5 hours per day	6
No. reporting only as required (not specified)	2
Total	44

Of those reporting intervention hours, 63.6% received these services 24 hours per day in the supported living environments. The reasons the remaining one third reporting less than 24-hour Intervention services were varied: some lived at home and needed less support, others were receiving services from agencies other than those mentioned above

that did not provide the degree of financial support.

2.3.5 Mortality

Three individuals whose report is included in this study had deceased: one male deceased at age 48, one male deceased aged 39 and one female deceased aged 36. Two of these individuals died of congestive heart failure (ages 48 and 36), while the third (aged 39) died of unspecified heart failure. These three individuals had participated in the 1999 study.

While not specifically related to this study, the writer received information that six other individuals (four females and 2 males) who participated in the 1999 study had deceased. Their birth years were 1952, 1957, 1962, 1963, 1974 and 1993. The age of death was known for only one of those individuals (born 1963 and died age 48).

Further to this, the writer also received unconfirmed details that two individuals from the Registry project reporting CRS had deceased. Their birth years were 1965 and 1983.

2.3.6 Who completed the survey?

At the end of questionnaire, participants were asked to indicate who completed the questionnaire.

A summary of who completed the questionnaire is presented in Table 32. The majority (58.5%) of the questionnaires were completed by professionals, while parent members completed 20.2%. Only one individual with CRS completed the survey and that person had the assistance of a parent member.

Table 32: Who completed the survey?

Professionals	31 (58.5%)
Parents	16 (30.2%) (13 Mothers; 3 Fathers)
Parent + Professional	3 (5.7%)
Sibling	1(1.9%)
Foster family member	1(1.9%)
Person with rubella + Parent	1 (1.9%)
Total	53

3.0 DISCUSSION

3.1 Study Rationale

The rationale for conducting this project was to describe the manifestations of congenital rubella syndrome through several recognized stages: a) **early onset** - those largely congenital manifestations such as deafness, congenital cataracts, congenital heart defects, etc. observed initially in newborns or during early childhood, and b) **delayed or later onset** – those conditions developing during later childhood into adulthood, which include glaucoma and corneal conditions affecting vision, epilepsy, endocrine dysfunction issues (diabetes and thyroid issues), and advanced vascular issues, etc.

Based on this understanding, this project was developed to obtain observations about the general age of onset of the various stages of manifestations affecting each of the major body systems: sensory, neurological, vascular, musculoskeletal, endocrine, respiratory and gastrointestinal. Further to the medical aspect of the study, the project also attempted to describe a sociological snapshot of these individuals through an examination of their educational, employment, communication and lifestyle history.

Early onset manifestations, according to Remington and Klein (1990)[76], can be explained 'simplistically' as the rubella virus tampering with embryonic cell division, possibly by decreasing cell multiplication, promoting chromosomal breaks and arresting the development of certain cell types. The exact mechanisms involved, however, are not clearly known.

With respect to the delayed manifestations, the mechanisms continue to be speculative. Sever, South and Shaver (1985)[77] suggest that several mechanisms have to be considered: growth of the virus in the tissues leading to reduced growth and shortened life span of the cells; auto immune responses initially stimulated by the rubella infection; the individual's genetic susceptibility; the early vascular damage by the infection may lead to further cardiac damage, and possible chronic persistence of the virus in tissues leading to extension of the virus to other areas of the body.

Regarding the possible autoimmune response, Wolinsky (1990)[78] believes that late endocrine dysfunction (leading to diabetes and thyroid problems) is related to viral-induced autoimmune mechanisms, suggesting some persisting rubella viral activities.

3.2 Implications from this updated study

The initial Canadian project to describe the late manifestations of congenital rubella syndrome completed in 1999, over fifteen years ago, answered many questions and concerns for family members and Intervenors (who live and work in different situations across the country) about the changing health conditions of individuals with CRS. It helped to improve the awareness of the many similar medical conditions affecting individuals with CRS and clarify the underlying causes of these delayed manifestations. The 1999 study also provided some clear messages to family members and professionals that they need to prepare for the predictable health challenges that will likely as these individuals age.

Since the initial study, family members and professionals have been requesting more information about this topic. They wanted further confirmation that the health changes they observe over time are in fact delayed manifestations of CRS. Further to these internal requests, international professionals connected with Deafblind International's

[76] Remington, J.S. and Klein, J.O. (Editors*). Infectious Diseases of the Fetus and Newborn Infant*. 3rd Edition (1990). W.B.Saunders, Philadelphia

[77] Sever, J. L., South, M. A. & K. A. Shaver. (1985). *Delayed Manifestations of Congenital Rubella*. Clinical Infectious Diseases,, Volume 7, Issue Supplement 1, Pp S164-S169

[78] Wolinsky, J.S. (1990). *Rubella*. In: Fields, B.N., Knipe, D.M. (Editors). *Virology*, 2nd Edition. Raven Press, New York. 815-836.

Rubella Network have advocated that countries undertake follow up studies to track this developing issue.

These discussions and recommendations encouraged CDBA National to undertake a follow up project to validate the initial findings from the 1999 study and to monitor whether additional medical changes are occurring within this unique population over the passage of time. As an important part of an updated study, family members and professionals recommended to CDBA National that a new survey should document any evidence of progressive neurological deterioration, clinical depression, and report any further changes in behavioral/psychosocial issues which characterizes many individuals with CRS. To assist with this latter request, CDBA sought the advice of neuropsychologist Dr. Jude Nicholas, a recognized expert in this field, to suggest appropriate questions for this part of the questionnaire.

In a brief summary, the findings reported in this follow-up study not only validate the results from the previous study but demonstrate 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); the incidence of spinal conditions (35.3% in 2014 vs 26% 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 degeneration of cognitive skills between 1999 and 2015. Surprisingly the report results did not indicate appreciable deterioration in motor skills.

Interestingly, difficulties with behavioural control were similar between the two surveys, confirming the link of these behaviours to those emotional disorders observed in individuals who are deafblind. This observation would suggest that, like the similar incidence of seizures (27.5% in 2014 survey versus 30% in 1999), these issues while established as congenital issues eventually appear as late onset manifestations.

The indicators of depression were similar in incidence between the two surveys, confirming a close connection of these aggressive-disruptive behaviours to mood control and emotional depression, being congenitally determined but not manifested until later.

An additional implication to report from this follow up study was reported mortalities among the current participants. As noted earlier, 3 of the 53 participants had deceased; all three at relatively young ages: 36, 39 and 48. Six of the original 100 individuals involved in the 1999 study were reported having deceased. None would have been older than 60, according to their birth ages. It would not be surprising to learn that more from that original 1999 population are not with us!

3.3 Writers personal comments about the study

First it is important to take into account several things about the make-up of the participants in the 2014 study versus that of 1999.

- The 2014 sample size (53) is only half of the 1999 project (100) sample size.
- Of the 53 individuals, 28 or 52.8% were part of the 1999 Rubella study.

While the current sample size is lower, it still does provide information from a core group of individuals we have studied before. Many of these individuals were supported early in their lives through specialized educational programs; and many live today in supported independent living facilities with Intervenor services or receive Intervenor services in their family homes.

Admittedly an impersonal mail out questionnaire is not a perfect method to guarantee detailed and precise answers about particular health and personal history. In some cases, professionals (who provided most of the information) did not always have the complete detailed historical information about the participant. Many sections of the questionnaire were not completed and/or information stated as unknown, which accounts for sample sizes being less than 53 for many of the questions.

Another weakness with the study was the lack of emphasis on the functional status of vision, hearing, motor activities, communication, etc.

Despite some of these weaknesses, it appears significant to the writer how the currently reported results validated the extent of the level of disability reported in the 1999 survey, despite the lower sample size and the inclusion of many new individuals. The incidences of various manifestations are similar in some cases and show higher levels for others. This is consistent with the fact that some of the conditions are early onset manifestations (and should be somewhat similar) while others fit the delayed stage of manifestations. Since many of the individuals are fifteen years older now, one should expect a higher incidence of these delayed conditions than in 1999.

3.4 Final Observations and Comments

It has been a wonderful privilege to be the project leader for two projects sponsored by the Canadian Deafblind Association to study the manifestations of congenital rubella syndrome of a small population of Canadians affected by this virus.

It continues to astound the degree of disability that this rather benign virus has thrust upon these individuals. The statistics are startling when one considers that a developing fetus exposed to the rubella virus in the first trimester of their life has such a high chance of being born visually impaired (over 96% in this study were), hearing impaired (over 92% in this study) and with a congenital cardiac condition (over 64% in this study). Almost 89% of the individuals in this study were born visually and hearing impaired and over 64% of them reported having the three disabilities. Then, in addition to starting off life facing these problems, many face a lifetime (and it seems like a shorter than normal one) of gradually deteriorating medical conditions affecting almost every bodily function.

Now the good news! Since the development of the rubella vaccine over fifty years ago, children need not be born with these devastating conditions. Canada like most of the western developed countries has observed incidences of the wild rubella virus plunge over the past 4-5 decades and consequently with minimal chance of children being born with CRS.

The numbers of children born with CRS are unknown in Canada during the era when the country experienced periodic epidemics of the rubella virus. Many children would have died at birth or in utero when post-partum birth conditions were different. Those that did survive were likely placed in institutions, the cause of their condition unknown. Many individuals born in the pre-immunization era with deafness or with hearing impairments were likely victims of congenital rubella as well.
Past reports from the World Health organization (WHO)[79] for Canada show previous high counts of wild rubella as follows: 70,000 (1936), 57,000 (1941), 52,000 (1956), 12,000 (1970&1975), 8,000 (1979), 6,500 (1983), fluctuating up and down to 246 (1994). It is during the 1960's through 1980's that the subjects in this study were born. This was during the period just before and after the rubella vaccine was taking effect to reduce the incidence of rubella. In the last two decades, the largest number of wild rubella cases reported in Canada was in 1997 when 4008 cases were observed. Since that time, cases have steadily declined. According to a WHO report (2013)[80], Canada reported 2 incidences of wild rubella and 1 case of CRS in 2011. Records since 2011 are unavailable.

It was rather disconcerting this past winter period 2015 when a small epidemic of red measles was reported in California then spread throughout the continent, fortunately not in large numbers. This demonstrated the alarming influence of 'anti-vaccination'[81] campaigns. The publicity is primarily about measles and risk of this virus to unvaccinated children. Earlier this year we learned also about small outbreaks of mumps with famous National Hockey League players sidelined due to either not been vaccinated or the vaccine losing its influence. Fortunately, there has not been a reported case of rubella in Canada amid this controversy. It must be kept in mind that the rubella vaccine is combined with the vaccine for red measles and mumps…hence MMR. If the anti-vaccine movement continues and minor outbreaks of measles and mumps continues, eventually cases of the wild rubella will be witnessed and inevitably a child will be born with symptoms of congenital rubella syndrome. Let's not let that happen!

3.5 Acknowledgements

This project was a venture involving the support and cooperation of many individuals from numerous organizations supporting individuals who are deafblind.

[79] www.who.int
[80] www.who.int/gho/publications/world_health_statistics/EN_WHS2013_Full.pdf - 2560k
[81] http://en.wikipedia.org/wiki/Vaccine_controversies

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