

**A SURVEY OF
LATE EMERGING MANIFESTATIONS
OF CONGENITAL RUBELLA
IN CANADA**

**STAN MUNROE
PROJECT LEADER
CANADIAN DEAFBLIND AND RUBELLA ASSOCIATION**

About the Author

Stan Munroe graduated from the University of New Brunswick with a B.A. (Biology and Psychology) and M.Sc. (Wildlife Biology) in 1967 and 1968, respectively. He worked for the Ontario Government (1969-1997) in Fish and Wildlife Management. Stan is a past president of the Canadian Deafblind and Rubella Association (CDBRA) and currently is Special Projects Coordinator for the CDBRA. His interest in this topic developed as a result of his parenting a young deafblind adult with congenital rubella syndrome.

Copyright © February, 1999

ISBN 0-921434-02-2

All rights reserved. The contents of this publication may not be reproduced in part or in full without the consent of the publisher.

The Canadian Deafblind and Rubella Association

350 Brant Avenue

Brantford, ON

Canada N3T 3J9

Web Site: www.cdbra.ca

Table of Contents

	Page Number
List of Tables	iii
Acknowledgments	v
Summary	viii
1.0 Introduction	1
1.1 Purpose of Study	1
1.2 Study Methodology	1
1.3 Developing the Network	2
1.4 Cooperator Network	3
1.5 Nature of Rubella	3
1.6 History of Rubella	4
1.7 Current Status of Rubella	5
1.8 Manifestations of Rubella	5
1.9 Early Manifestations of Congenital Rubella	6
1.10 Late Manifestations of Congenital Rubella	7
2.0 Results and Discussion	7
2.1 Section A	7
2.1.1 Sex and Age Distribution	7
2.1.2 Racial Description of Respondents	8
2.1.3 Regional Distribution of Sample	8
2.1.4 Incidence of Rubella	8
2.1.5 Regional Incidence of Rubella	9
2.1.6 Nature of Disability	10
2.1.7 Nature of Rubella Infection	10
2.1.8 Birth Situation	12
2.1.9 Estimated Gestational Age at time of Maternal Infection	13
2.1.10 Birth Mother's Health Issues	14
2.2 Section B	16
2.2.1 Congenital Birth Defects	16
2.2.2 Visual History/Loss	18
2.2.3 Causes of Visual Loss	20
2.2.4 Hearing Loss	22
2.2.5 Neurological Conditions and Symptoms	23
2.2.6 Neurological Development	25
2.2.7 Mental Health	28
2.2.8 Behaviour	29
2.2.9 Seizures	32
2.2.10 Sexual Development	34

Table of Contents, continued

Page Number

2.2.11	Sleep/Wake Patterns	36
2.2.12	Size (Height and Weight) of individuals	37
2.2.13	Skeletal Conditions	38
2.2.14	Motor Skills	39
2.2.15	Urogenital Tract	41
2.2.16	Bowel Function	42
2.2.17	Gastrointestinal	44
2.2.18	Endocrine Function	46
2.2.19	Heart and Circulatory System	47
2.2.20	Lung Condition	49
2.2.21	Allergy/Asthma	50
2.2.22	Osteoporosis	51
2.2.23	Cancer	52
2.2.24	History of Hospitalizations	52
2.2.25	Record of Medications	53
2.2.26	Family Dynamics	54
2.2.27	Educational Activity and Placement	55
2.2.28	Living Situation	56
2.2.29	Working Situation	58
2.2.30	Communication	59
2.2.31	Support Resources for Individuals who are Deafblind	60
2.2.32	Support Resources for Individuals who are Deaf	62
3.0	Conclusions and Recommendations	63
3.1	About John	63
3.2	Mechanisms involved with Manifestations of Congenital Rubella	65
3.3	Implications of the Late Manifestations Study	66
3.4	Study Evaluation	66
3.5	Future Studies	67
3.6	Significance of Project Results for Policy Formulation and Program Development	68
3.7	Final Words	70
	Bibliography	71

List of Tables	Page Number	
Table 1	Sex distribution by age groups	7
Table 2	Numbers reporting region of residency	8
Table 3	Distribution of period rubella virus contracted by mother	9
Table 4	Summary of reported disability by age groupings	10
Table 5	Nature of rubella infection	11
Table 6	Summary of birth description according to sensory disability	12
Table 7	Summary of birth weight according to sensory disability	13
Table 8	Summary of reported gestational age of maternal infection with rubella according to disability	13
Table 9	Summary of month during first trimester when rubella contracted	14
Table 10	Summary of birth mother's responses to health related issues	15
Table 11	Summary of number of years between age birth mother contracted rubella and age reported initial symptoms	15
Table 12	Summary of various congenital birth defects according to disability	17
Table 13	Summary of responses regarding visual history/visual loss	19
Table 14	Summary of responses regarding causes of visual loss	21
Table 15	Summary of responses regarding hearing loss	22
Table 16	Summary of responses regarding neurological conditions and symptoms	24
Table 17	Summary of responses regarding neurological development	26
Table 18	Summary of responses regarding mental health	28
Table 19	Summary of responses regarding behaviour	30
Table 20	Summary of responses regarding seizures	33
Table 21	Summary of responses regarding sexual development	35
Table 22	Summary of responses regarding sleep/wake patterns	36
Table 23	Summary of responses regarding size (height and weight) of individuals	37
Table 24	Summary of responses regarding skeletal conditions	38
Table 25	Summary of responses regarding motor skills	40
Table 26	Summary of responses regarding the urogenital tract	41
Table 27	Summary of responses regarding bowel function	43
Table 28	Summary of bowel function responses according to disability level	44
Table 29	Summary of responses regarding the gastrointestinal system	45
Table 30	Summary of responses regarding endocrine function	46
Table 31	Summary of responses regarding the heart and circulatory system	48
Table 32	Summary of responses regarding lung condition	50
Table 33	Summary of responses regarding allergy/asthma	50
Table 34	Summary of responses regarding osteoporosis	51
Table 35	Summary of responses regarding cancer	52
Table 36	Summary of responses regarding history of hospitalizations	52
Table 37	Summary of responses regarding use of medications	54
Table 38	Summary of responses regarding family dynamics	55
Table 39	Summary of responses regarding educational activity and placement	56

List of Tables, continued	Page Number	
Table 40	Summary of responses regarding living situation	57
Table 41	Summary of responses regarding working situation	58

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

Table 42a	Summary of responses regarding communication	59
Table 42b	Summary of responses regarding communication (without age of first reporting)	59
Table 43	Summary of responses regarding support resources for deafblind	61
Table 44	Summary of responses regarding support resources for deaf	62

Acknowledgments

This project was a venture that required the support and cooperation of many individuals who I will acknowledge. My apologies if I have missed anyone.

First, I wish to acknowledge Linda Mamer, President of CDBRA, who suggested that we apply for funding for this project, and who, throughout this study, has provided constant inspiration and excellent editorial advice.

Others who inspired this study include Joan Brintnell, Executive Director of McInnes House, who suggested that further study was required into the causes for the changes taking place among the congenital rubella young adults like my son, Andrew; and Evabritt Andreassen, Leader of Western Norway Regional Centre for Deafblind, who planted the seed that a study into the late manifestations of congenital rubella would be an excellent retirement project for me.

I also wish to thank Nancy O'Donnell, Helen Keller National Centre for Deafblind for the support she provided to the project, including supplying copies of the many research papers on Rubella.

To all those on the CDBRA National Board of Directors, I offer my sincere appreciation for the opportunity to undertake this project which was so personal to me.

To members of the steering committee, Cathy Proll, Connie Taylor Southall, Joan Brintnell and Linda Mamer, I acknowledge their assistance in putting together the detailed questionnaire and for their suggestions for analysis.

I also wish to acknowledge Harry Taylor, statistician, who, despite his concerns about the number and types of questions, created the final format of the questionnaire and the data entry packages and did the data crunching. (I should add that Harry's early concerns became reality during the complicated data analysis phase).

Jacques Pinault, I thank for his skills to translate into french, several pieces of correspondence, and the lengthy questionnaire. As well, I thank him for his assistance with an english translation of the written comments made on the french questionnaires.

Karen Munro (no relation) should be acknowledged for the many hours spent photocopying and distributing the survey packages (which included a questionnaire and a copy of several research papers) and for the final document editing. I also acknowledge Cindy Rock for her promptness with the financial aspects of the study.

This project would not have been the success it is without the cooperation and participation of the following family members who were involved in completing the questionnaires and/or provided follow up information: Ann Chan, Juanita Crawford, Margaret Hatley, Sharon Haughn, Judy Leonard, Joan Leitch, Ann DeCastro, Diane Lanthier, J. Ray Lanthier, Mildred Hemstock, Sophie Golinsky, Grace Ronald, Mary Chenail, Roseanne Hunt, Reta Bernard, Kateri Chiasson, Betty Chung, Paul Chung, Margaret Milotte, Elaine Parsons, Roberta Byrne, Bea LeMon, Hank LeMon, Marjorie Gurski, Erika Kaukel, Heather Roy, Gillian Roy, Ann McManus, Loretta Neustaeter, Cynthia Goulden, Cherry Bulmer, Dorothy Parker, Dinah Pilgrim, Sharon Rumley, Elaine Benmore, Eleanor Young, Dorothy Boyer, Cynthia Bolwig, Dorothy Grainger, Janet McEachern, Dorothy Chapman, Paulette Scott, Jill Hardy, Pat Andrews, Carrie Paris, Marilyn Dirks, Becky Heck, Ann Floyd, Patricia Curtis, Lillian Mack, Rose Smith, Joanne Jecklin, Cathy Alpaugh, Beryl Champion, Maudline Pink, Phyllis Bruce, Edith McGrath, Mary Cosentino, Millie Dowe, Ruth Evert, Sheila Collett, Betty Hughes, Velma Mallett, Barbara Nolt, Diane McKenzie, Madeleine Doyon, Henri Doyon, Zita Kato, Denise Vigneau Martinet, Hilda LeBlanc, Francis Quarrington, Karyn Krell, Mrs. Lang, Margaret Small, Chloe Maes, Rob Maes, and a mother who wished to be unnamed.

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

Many other individuals need to be acknowledged who, in addition to the above mentioned family members, assisted in completing the questionnaires and/or supplied follow up information. They included: Patti McDougall, Janice McGlincey, Paul Lingen, Samantha Smiley, Pamela Strong-Smith, Audrey Ramsden, Alice Banford, Susan Szczechura, Jackie McKenzie, Barb Usher, Pauline Russell, Carol Wilyman, Dana Janzen, Ghita Gaudet, Belle Maidment, Pat Misener, Don Hendry, David Sutton, Amber Long, Keryi Maynard, Linda Mamer, Joan Brintnell, Connie Taylor Southall, Carla McDonald, Diana Kanz, Melissa Zollner, Marie Claire LaPierre, Anne Wrightson, Louise Carriveau, Marie Paquin, Marie Flouriot, Carole Bousquet, Emiline Francois and Jean Chevarie.

To reach the many individuals and families of individuals who had congenital rubella, required the support and assistance of many people working with non-profit organizations, facilities and schools. The following are acknowledged for their assistance: Jennifer Grondin and Linda Mamer (BC Chapter, CDBRA); Lorraine Williams (Saskatchewan Chapter, CDBRA); Cheryl Ramey (Manitoba Chapter, CDBRA); Cathy Proll and Gail Malcolm (Ontario Chapter, CDBRA); Patricia Curtis (NB-PEI Chapter, CDBRA); Cynthia Goulden (NS Chapter, CDBRA); Hilda Nanning (BC-CNIB); Myrna Matheson (APSEA); Amber Long (Alberta-CNIB); Katherine Hesson and Kerry Wadman (Ontario-CNIB); Len Baker (NFLD-CNIB); Gilles Lefebvre and Christine Cabedoce (Institut Raymond - Dewar); Margaret Phillips (Western Institute for the Deaf); Joanne Deluzio (Canadian Hearing Society); Joan Brintnell (McInnes House); Connie Southall (Independent Living Residences for Deafblind Persons); Deborah Duncan and Bob Follett (Huron Regional Centre); Donna Morrow (Rideau Regional Centre); Ray Brown and Ming Yeh (Southwest Regional Centre); Diane Rimmer (Tayside Community Options); Mary Zadow (James Whitney School); Bill Thompson, Janice McGlincey and Lynn Watkins (W. Ross Macdonald School); Sheena Kay (E.C. Drury School); Mary Ellen Bolt (Robarts School); Jim Thompson (Canadian National Society of Deafblind Persons); Joyce Thompson (Rotary Cheshire Homes); Linda Clark (Newfoundland School for the Deaf); Agnes O'Neill (Regional Residential Services Society); Dr. Aubrey Tingle (BC Research Institute for Children's and Women's Health); Dr. Digby Horne, (Manitoba Health); Paul Bartu (Ontario Ministry of Education) and Viola Sangster (Amherst).

This project would not have been possible without the financial support of the Federal Government of Canada. I wish to acknowledge Human Resources Development Canada for its generous financial support under the Disabled Persons Participation Program. Furthermore, I express much appreciation to Lise Labonte, project advisor, Employability and Social Partnerships Division of HRDC, for her strong support of this project which ensured that the CDBRA received initial and supplementary funding to carry the project through to completion.

Finally, I dedicate this work to my son Andrew, who contracted congenital rubella in New Brunswick in 1966, and I acknowledge being a statistic in this report. Andrew has given me the wonderful privilege to meet and work with so many dedicated people in the field of deafblindness within Canada and all over the world. He has also given me the opportunity to add a real purpose to my life. My wish is that this work can somehow contribute to an improved quality of life for individuals with congenital rubella.

Summary

One hundred individuals with (or with suspected) congenital rubella syndrome participated in a study to document the incidence, within the Canadian population, of late emerging behavioural and medical conditions (manifestations) believed related to the congenital rubella syndrome. This project was carried out by the Canadian Deafblind and Rubella Association in 1997-1998 through funds provided by Employability and Social Partnerships Division of Human Resources Development Canada.

Participants in the study ranged in age from five to sixty-two years, with two thirds of respondents aged 20-39. The remaining were equally divided between nineteen years and under and forty years and over. Respondents represented all regions of the country, with almost 60% coming from Canada's two largest provinces, Ontario and Quebec. Three quarters of the respondents reported contracting congenital rubella during the major epidemic period of the 1960's and 1970's. Eight reported contracting rubella since the 1980's while twenty contracted rubella prior to 1960.

Seventy individuals reported their functional disability as deafblindness, twenty-one as deafness, seven as blindness, while two did not consider themselves functionally blind, deaf nor deafblind. Despite this categorization, 92% reported a visual loss, 96% reported a hearing loss and 91% reported a combination of a visual loss and a hearing loss.

Current 1998 living status of the respondents: 30 live in supported independent living situations (with Intervention); 15 live in an institution for the mentally disabled; 13 live independently in their home communities; 10 live in another type of group home; 28 live at home (eleven attending school, 17 finished school); 3 in a nursing home. Since the survey was undertaken, one of the respondents, a 36 year old woman from Ontario, has died.

To recognize the early manifestations of congenital rubella, individuals reported the following incidence rate for: visual loss (86%); hearing loss (93.9%); heart defect (65.7%); microcephaly (33.3%); combined visual loss and hearing loss (80.2%); combined visual loss, hearing loss, and heart defect (58.9%); and combined visual loss, hearing loss, heart defect and microcephaly (23.9%). Congenital cataracts, reported by 79.6% of individuals, was responsible for most early visual loss, followed by glaucoma, which was reported overall by thirty-one individuals (32.3%), eleven of which reported having this condition since they were two years old.

The incidence of later manifestations of congenital rubella, those which first occur during later childhood, i.e. after age six are reported as follows (where there is an age of first onset of the condition reported):

- X glaucoma (20 of total 31 reporting condition or 20.8% overall)
- X detached retina (11%)
- X change in visual acuity, all causes (27%)
- X change in hearing ability (24.1%)
- X treatment for mental health (26.5%)
- X reporting at least one of these behaviours...aggression, self injury, tantrums, and property destruction (23.2%)
- X seizures (30%) overall, 23% incidence reported after age six)
- X incidence of one of scoliosis, kyphosis or lordosis (overall incidence of 26%)
- X deterioration in energy level, stamina and endurance (21%)
- X clinically diagnosed with thyroid dysfunction (10%)
- X clinically diagnosed with diabetes (12.1%)
- X diagnosed with osteoporosis (7.1%)

Birth mothers were asked questions about several of their health issues. For those reporting, 41.9% reported early symptoms of non-specified arthritis; 25.4% reported early symptoms of menopause (eight

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

of seventeen reported non-specified symptoms before the age of 40); 9.4% reported asthma. Over one quarter of birth mothers reported a host of other health issues including glaucoma, cataracts, multiple sclerosis, cardiac problems, thyroid dysfunction, psychiatric problems, endometriosis etc. The suspicion is that an enhanced incidence of auto-immune health issues might have some relationship to carrying a child with congenital rubella.

1.0 INTRODUCTION

1.1 Purpose of Study

To document the incidence, within the Canadian population, of late emerging behavioural and medical conditions related to the congenital rubella syndrome.

The study was conceived for several reasons. Firstly, concerns were expressed within independent living residences for individuals with deafblindness, about an increasing number of individuals experiencing early ageing health issues such as osteoporosis, diabetes, behavioural changes, physical deterioration etc. Secondly, a study had been recently published by the Helen Keller Centre in the USA (O'Donnell, 1996) that revealed startling statistics about emerging health issues related to individuals with congenital rubella.

Consequently a proposal was prepared and submitted during late summer 1997 by the Canadian Deafblind and Rubella Association (CDBRA) to Human Resources Development Canada, the branch of the Federal Government which administers projects to non-profit organizations serving disabled persons.

In September 1997, the Employability and Social Partnerships Division of Human Resources Development Canada approved the project under the Terms and Conditions governing the Disabled Persons Participation Program funding. A total allocation of \$40,000 was provided for this project, which commenced the end of October 1997 and completed September 30, 1998.

1.2 Study Methodology

A Steering Committee was appointed to guide the study. Members of the committee included Cathy Proll, Joan Brintnell, Connie Southall, Linda Mamer and Stan Munroe, project leader. All of these individuals had extensive experience as professionals working with individuals with deafblindness from congenital rubella.

The Committee developed terms of reference for the study, designed a detailed survey questionnaire, and determined how the survey would be carried out.

The Questionnaire was designed in two sections: Section A was set up to gather information about the general demographics of congenital rubella individuals; to obtain information from the birth mothers about the nature of their infection with rubella, the period in which they contracted rubella, and certain health parameters. Section B was set up to capture a description (life history) of the developmental, health, educational and social profile of individuals with congenital rubella. Section B contained 31 sections, each containing a number of questions. Individuals were asked to answer each question with a "yes" or "no," then, if the answer was "yes," indicate how the "yes" answer applied according to the age groupings: 0-2 years; 3-6 years; 7-12 years; 13-21 years; 22-30 years; 31-40 years; 40+ years.

A statistical consultant assisted in the design of the questionnaire, developed the data entry format (using MS Access and MS Excel software programs) and produced the statistical results.

The focus of the study was to locate as many individuals throughout Canada known or suspected to have congenital rubella and who were deaf, deafblind or blind.

1.3 Developing the Network

Implementing the study to locate individuals for this survey required networking with a wide array of non-profit organizations, educational and residential facilities, professionals, administrators, educators and care givers and families.

The first step was to locate and contact organizations and facilities serving or advocating for individuals with sensory losses, explain the nature of the project and request cooperation and/or participation in the study. Once the level of participation was determined, a key contact person was assigned to work with the project leader.

The focus, as indicated, was on locating individuals with congenital rubella. Cooperating organizations and facilities assisted by locating individuals for the study in several ways: (a) names and addresses were provided directly to the project leader who then directly contacted individuals or their families; (b) cooperating organizations contacted individuals themselves (directly or through family members or care givers) seeking their participation in the study. Cooperating organizations and facilities also sought consent from individuals, their families or their advocates to participate in this study.

A number of individual participants were also located through "word of mouth" contacts with family members who were not connected to an organization or facility.

Once the individuals were located, a survey package was then distributed directly or through cooperating organizations to individuals consenting to participate in the study.

The questionnaires were completed by individuals themselves (who could do so), by their family members or their advocates, and/or (if the individuals lived in an independent living facility or institution) by care givers. Completing the questionnaire was often a joint effort between the family and facility staff. On numerous occasions the project leader contacted the individuals who completed the survey to obtain clarification or to complete sections that had not been completed.

1.4 Cooperator Network

The following list represents those organizations and facilities who participated in this study in one form or another:

- Atlantic Provinces Special Education Authority, Halifax, NS
- British Columbia Research Institute for Children's & Women's Health, Vancouver, BC
- Canadian Hearing Society, Toronto, ON
- Canadian National Society of the Deaf-Blind, Toronto, ON
- CDBRA - New Brunswick/Prince Edward Island Chapter
- CDBRA - Ontario Chapter
- CDBRA - Manitoba Chapter
- CDBRA - Saskatchewan Chapter
- CDBRA - British Columbia Chapter
- CDBRA - National Office
- Clair-Foyer Inc., Val D'or, PQ
- CLSC - Iles de la Madeleine, PQ
- CNIB - National Office, Toronto, ON
- CNIB - Newfoundland, St. John's, NF
- CNIB - Ontario, Toronto, ON
- CNIB - Alberta, Calgary, AB
- CNIB - British Columbia, Vancouver, BC
- DeafBlind Housing Society, Richmond, BC
- Ernest C. Drury School, Milton, ON
- Huronia Regional Centre, Orillia, ON
- James Whitney School, Belleville, ON
- Independent Living Residences (for Persons who are) Deafblind, Richmond Hill, ON
- Institut Raymond-Dewar, Montreal, PQ
- McInnes House, Brantford, ON
- Mijiwam Special Support Home, Almonte, ON
- Newfoundland School for the Deaf, St. John's, NF
- Regional Residential Services Society, Dartmouth, NS
- Rideau Regional Centre, Smiths Falls, ON
- Robarts School, London, ON
- Tayside Community Options, Perth, ON
- Southwestern Regional Centre, Blenheim, ON
- West Montreal Re-Adaption Centre, Verdun, PQ
- Western Institute for the Deaf, Vancouver, BC
- W. Ross Macdonald School, Brantford, ON

1.5 Nature of Rubella

Rubella, known more commonly as German measles, was once recognized as the least troublesome of the familiar childhood communicable diseases. Its infectious nature is characterized by a mild fever, slightly swollen glands and a quickly disappearing rash. Often, when there was a rash, it lasted only several hours. These rather benign symptoms once rendered rubella to a much less feared status than the red measles. In comparison, the red measles, a more dangerous communicable childhood disease, was characterized

by a high fever, sore throat, stuffy nose, usually an eye infection along with a very distinctive and longer lasting rash.

Individuals of all ages contracting rubella were not totally without complications, although their implications were less severe than that of the more feared red measles. The most frequently encountered complication from natural rubella is arthralgia, an inflammation in the joints, with the hands most frequently affected. Arthralgia is more prevalent in adult women than men or children. Another complication, although less frequent, affecting about one in 3,000 individuals, and mostly children (Waxham and Wolinsky, 1984), is purpura, a hemorrhaging of the skin and mucous membranes. The most serious complication of acquired or post natal rubella, affecting about one in 5,000 rubella cases, and those mostly young adults, is encephalitis, an inflammation of the brain. Examples reported of other, although rare, complications of rubella to name a few, include progressive parencephalitis, carotid artery thrombosis, myocarditis and pericarditis (Cherry, 1987).

1.6 History of Rubella

Rubella was first described by an English physician, Dr. George Maton, who, in 1815 noted, a mild disease resembling scarlet fever. Rubella epidemics occurred in Germany in the first half of the 19th Century during which time the disease was called by the German name rotheln. An English physician proposed the name rubella (meaning red and pretty) as opposed to its harsh German name, rotheln (Cooper, 1966). Due to the great interest of German physicians in this disease during this period, the name "German measles" was attached to the disease (Cherry, 1987).

Little attention was given to the disease until 1941, when there was a widespread epidemic of rubella in Australia. During this time an Australian ophthalmologist, Sir Norman Gregg, noted a high incidence of congenital cataracts among many of the infants he examined. He startled the medical community with what he believed to be a relationship between rubella and congenital cataracts. In 1941, of 78 patients he examined with cataracts, 68 of their mothers remembered having rubella during the early part of their pregnancy. Gregg also reported within this small population of infants, a high incidence of low birth weight, feeding difficulties, congenital heart defects, deafness and a high mortality rate (Givens et al 1993).

Gregg's observations were eventually confirmed to reveal the most sinister side of the previously believed benign rubella, that related to its assault or impact on the developing fetus if contracted by the mother during her first trimester of pregnancy. This impact became widely known during the 1960's with the worldwide epidemic of rubella between 1962 and 1965. During this period, rubella was believed to have affected approximately 10% of pregnant women, with about 30% of those infected bearing infants with congenital manifestations from rubella (Givens et al 1993).

The realization that the mother may have contracted rubella was obscured within one-third to two-thirds of the cases, since in many individuals the illness from rubella was sub-clinical or showed no observable consequences from the disease (Cooper, 1966).

The rubella virus was eventually isolated and tissue was cultured in 1962 which led to the production of vaccines and the introduction of immunization programs throughout the world beginning in 1969. This did not occur in time to limit the damage that the 1960's rubella pandemic created, however. The United States epidemic in 1964-65 was estimated to have infected over 12.5 million individuals, caused about 11,000 instances of fetal death and is implicated in congenital birth defects in about 20,000 infants (Cochi, 1989). There are no Canadian statistics for that period, but since the epidemic moved through Canada, it would not be surprising if 2-3,000 children were born during the 1960's with congenital birth defects related to congenital rubella.

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

1.7 Current Status of Rubella

With widespread immunization a major abatement of rubella has occurred since the 1960's, but not its complete eradication. Countries without systematic vaccination programs are still witnessing the birth of relatively large number of infants with congenital rubella (van Dijk, 1991). Startling statistics are coming out of Mexico, based on a recent report from the Texas Department of Health. Health officials reported over 7,000 cases of rubella in Mexico in 1998. Almost 30% of these cases occurred in Mexican states that border Texas. The first 19 of 35 cases reported in Texas this year were in a federal immigration and naturalization service detention centre in south Texas.

Even countries with mandatory immunization programs are witnessing rubella outbreaks. An outbreak of rubella was observed in the Canadian province of Manitoba over the period October 1996-May 1997. According to Dr. Horne (personal communication, 1998), this epidemic affected approximately 4,000 individuals. There is one confirmation to date of a child born with congenital rubella syndrome with both hearing and visual loss. These incidences of rubella in Manitoba accounted for all of the reported Canadian cases in 1996 and 1997. They significantly eclipsed the 236 cases of rubella reported in Canada in 1995 (Source: The 1997 Canadian Federal Health Department Survey as reported by The Toronto Star, October 27, 1998).

It is safe to say that the rubella virus has been largely contained, especially in the western world, and this should continue, providing countries maintain an aggressive immunization campaign. Further outbreaks of the disease will continue unless mandatory immunization regimes are maintained and mechanisms are established to screen individuals moving from other countries which do not have mandatory rubella immunization policies.

1.8 Manifestations of Rubella

The rubella virus is remarkable, firstly, with its association with a diverse group of clinical diseases and, secondly, in the various processes by which it attacks its host congenitally and continues post-natally.

As previously indicated, the rubella virus causes minor external symptoms associated with the disease. In addition, rubella may cause such complications as secondary rubella arthritis or arthralgia, post-infectious encephalitis and a slow progressive neurologic disease called progressive rubella parencephalitis (PRP). Usually fatal, PRP is believed to be a late complication of natural childhood rubella (Slagle and Wolinsky, 1989).

It is the various processes of infection connected with congenital rubella (and which can be related to the later manifestations of the disease) that has been the focus of recent research and is the subject of this paper. The rubella infection process is known to work in various ways: Firstly, the rubella virus reduces cell division in the embryo or developing foetus, resulting in incomplete, delayed or defective growth of organs and body parts. An attack during the critical first trimester of development can infect and impact virtually every developing organ. Secondly, the virus results in damage to cells and inflammation throughout the unborn fetus' body (Menser and Reye, 1974; Vernon and Hicks, 1980 as reported in O'Donnell, 1996). This cellular damage generally affects early inner ear development and generalized vascular damage (Cherry, 1987). Thirdly, the rubella virus continues to infect the post-natal individual, as evidenced by the isolation of the rubella virus in many congenital rubella children for up to eighteen months and even older. Finally, circulating rubella-specific immune complexes are believed to be implicated in later manifested immune-mediated conditions as seizures, thyroiditis, glaucoma and abnormal glucose tolerance etc (Gilbert, 1991).

Merth et al (1987) provided the following framework for examining the late onset of defects: (a) in the first

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

two months or neonatal period, the major observable defects are usually hearing impairment, cataracts, cardiac defects and purpura ; (b) in months two to six, late onset observable defects include failure to thrive, intestinal pneumonia, chronic diarrhea, immunological system defects, mental retardation and hearing impairment; (c) from three to 15 years, delayed type defects observed include diabetes mellitus or other endocrinological defects, progressive involvement of the central nervous system, deafness and glaucoma, hyperkinesia and sub-retinal non-vascularization. These three categories were not considered mutually exclusive, since symptoms in the third category may occur in the first or second category.

1.9 Early Manifestations of Congenital Rubella

A host of publications have presented statistics collected from the 1960's epidemic in the United States about the disabling conditions rendered by congenital rubella. These are summarized in O'Donnell (1996) as follows:

- One-third of infants born of mothers who had rubella during pregnancy had some sign of rubella infection. Of those infants, 85% had various handicapping conditions, including hearing loss (73%); cardiac problems (35-76%); some form of visual loss, including glaucoma, cataracts and microphthalmia (33%) and cognitive difficulties (42%).
- Forty percent of deaf rubella infants weighed less than 5.5 pounds at birth, the result of impaired cell division, which placed them in the "premature category" according to the World Health Organization.

1.10 Late Manifestations of Congenital Rubella

The most recent study on late manifestations of congenital rubella was reported by O'Donnell (1996) from a survey of 88 individuals with congenital rubella in the United States. Twenty-eight of the survey respondents (32%) were individuals 25-26 years of age, and progeny of the 1960's rubella epidemic. O'Donnell reported nearly 73% of the respondents to be deaf with significant vision problems or total blindness. About 90% had no functional hearing. One case of late onset hearing loss was reported in the study. Two cases of cataracts were identified at age 7, one case each at ages 16 and 18. Thirty percent of the respondents had glaucoma, most acquired since infancy. Over 13% reported detached retinas. The study reported a 5.7% incidence of diabetes, 6.8% incidence of thyroid problems, 6.8% showing degenerative processes, several of which were progressive rubella parencephalitis or PRP. In addition, 6.8% of respondents reported incidences of esophageal stricture, gagging, swallowing and cyclic vomiting. These latter conditions have not been previously reported in the literature connected with congenital rubella.

2.0 RESULTS AND DISCUSSION

The report is prepared to correspond to the design of the survey questionnaire which was in two parts: Section A and Section B. Section A was demographic in nature, and designed to collect information on history of the rubella infection and information on the birth mother's health. Section B focused on the developmental, social, educational and health profile of the individual.

2.1 Section A

2.1.1 Sex and Age Distribution

Fifty-eight of the 100 respondents were female, forty-two were male. The ages of the respondents ranged from 5 years to 62 years of age. These are summarized by age groups in Table 1.

Table 1: Sex distribution by age groups of respondents

Age Group	Males	Females	Total
0-12	3	2	5
13-19	7	4	11
20-29	15	19	34
30-39	12	21	33
40+	5	12	17
Totals	42	58	100

Since completing a questionnaire, it was learned that one of the respondents with congenital rubella, a 36 year old woman from Ontario, had died during the summer of 1998. The cause of death was described as aspiration due to fluid on the lung causing congestive heart failure. She had a congenital heart defect. During the month prior to her death, she had a urinary tract infection, septicemia (blood poisoning), ulceritis in the colon and demonstrated liver and kidney failure.

2.1.2 Racial Description of Respondents

Ninety-three reported being white Anglophone or Francophone Canadians. The remaining indicated their racial origin to be Black (3), Black-Caucasian (1), Aboriginal-Caucasian (1), Taiwanese (1) and Chinese-Filipino (1).

2.1.3 Regional Distribution of Sample

The current residence of respondents is summarized in Table 2 . The bulk of respondents (51) report currently residing in the Province of Ontario. The remaining 49 live in the Atlantic Provinces of Newfoundland, Prince Edward Island, Nova Scotia and New Brunswick (15); the Prairie Provinces of Manitoba, Saskatchewan and Alberta (10); British Columbia (16) and Quebec (8).

Table 2: Comparison of number of respondents reporting region of residency versus region where rubella contracted

Region	Region Currently Residing	Region Where Rubella Contracted
Atlantic Provinces	15 (Nfld 3; NS 5; NB 6; PEI 1)	20
Quebec	8	10
Ontario	51	36
Prairie Provinces	10 (MB 2; SK 6; AB 2)	10
Yukon-BC	16	17
Unknown Canada	0	1

Outside Canada	0	6 (UK 2; Taiwan 1; France 1; Honduras 1; USA 1)
Total	100% (100)	100% (100)

2.1.4 Incidence of Rubella

Individuals were asked to report the year their birth mother came into contact with the rubella virus. The purpose was to observe the incidence of past rubella infections throughout Canada. The results are summarized in Table 3.

Table 3: Distribution of respondents according to period rubella virus contracted by mother

Contract Period	Before 1950	1950's	1960 - 1965	1966 - 1969	1970 - 1975	1976 - 1979	1980's	1990's
Number n=100	4	16	23	12	25	12	5	3

Respondents reported being in contact with rubella from 1935 to 1993. Table 3 verifies that the major wave of rubella first swept across North America during the first half of the 1960's then persisted to affect individuals through the end of the 1970's. The continued incidence of congenital rubella cases since 1975, although progressively decreasing through time, suggests a considerable delay in the absolute effectiveness of the rubella vaccine which has been in widespread use since the beginning of the 1970's.

What is interesting is the 16 individuals who contracted rubella during the 1950's and who represent 16% of the total. This raises the question whether this is indicative of previously unreported epidemics in Canada. Congenital rubella was not a reportable disease until the mid 1960's in the United States (Cherry, 1987). If Canada is similar to the United States in that regard, little, if anything, is available on the incidence of this disease during this period in Canada.

Cherry (1987) reported information on the past incidence of rubella from the eastern United States, 1928-1983. He observed that the basic pattern of rubella epidemics follow a 3 year cycle of build up and fall, occurring generally over six to nine year intervals. The data they report upon indicated peaks of high rubella incidence in the eastern USA during 1934-1936, 1941-1944, 1952-1953, 1956-1959 and 1963-1965. They did not report any further peaks of incidence following the 1960's pandemic.

In Table 3, the sample of reported congenital rubella for the 1950's in Canada corresponds to the two peaks (1952-1953 and 1956-1959) reported in the eastern United States.

2.1.5 Regional Incidence of Rubella

Individuals were asked to indicate in what province of Canada, or elsewhere, the birth mother contracted the rubella virus. Their responses are summarized in Table 2. Ninety-four individuals report the country of contact as being Canada. One, who verified Canada as the country in which rubella was contracted, was unsure of the specific region. The remaining six individuals reported contracting rubella in the following locations and years, outside of Canada: UK (1 in 1940 and 1 in 1966); Taiwan (1 in 1976); France (1 in 1962); Honduras (1 in 1990) and New York State, USA (1 in 1974).

Thirty-six respondents reported contracting the rubella virus in Ontario. This was followed by 20 reporting

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

the Atlantic Provinces; 17 reporting the Yukon-BC Region; 10 reporting the Prairie Provinces; 10 reporting the Province of Quebec; and 6 reporting outside Canada.

While most respondents (36) reported contracting rubella in Ontario, the number reporting Ontario as the region of residency was substantially higher at 51 individuals.

2.1.6 Nature of Disability

The questionnaire asked whether the individual was blind, deaf or deafblind. The respondents noted their functional disability as follows: 70 identified themselves as deafblind; 21 as deaf and 7 as blind. Two individuals reported not being blind, deaf nor deafblind.

Table 4 summarizes the reported visual or hearing loss among the 100 respondents. These figures differ from what the individuals described as their major disability. Ninety-one reported a visual loss, ninety-six reported a hearing loss and ninety-one reported having a combined visual and hearing loss. In three cases, the verification of a hearing loss was not given. Five of the seven (71.4%) individuals identified as blind also had a hearing loss, while thirteen of the twenty-one (61.9%) individuals who were deaf reported a visual loss. Only 9 of the total respondents reported either a visual or hearing loss but not both.

Table 4: Summary of reported disability according to age groupings

Age Grouping	Sample Size	Visual Loss Only	Hearing Loss Only	Hearing and Visual Loss	Unknown
3-12	5	1	0	4	0
13-19	11	0	1	10	0
20-29	34	1	0	33	0
30-39	33	1	3	27	2
40+	17	1	4	9	3
Total	100	4	8	83	5

2.1.7 Nature of Rubella Infection

Ninety-two respondents indicated they had congenital rubella. Eight respondents indicated their disability was suspected to be caused by congenital rubella. Four of these suspected cases were identified as deafblind while four were deaf.

Individuals were asked if they could relate any history about the birth mother's rubella infection. Table 5 summarizes their responses. The high numbers reporting "unknown" is largely due to individuals other than mothers or family members, completing the questionnaire.

Table 5: Number of respondents answering questions concerning nature of rubella infection

Question asked about nature of infection	Yes	No	Unknown
Presence of rash and fever	47	23	30

Subclinical symptoms	21	48	31
Rubella infection confirmed by doctor	32	33	35
A known rubella re-infection	6	46	48
Mother immunized against rubella	12	38	50

While 47% of total respondents report the presence of a rash or fever, 21% of the total respondents indicated subclinical symptoms of rubella, that is, no presence of a rash or fever. This latter percentage would rise to 30.4% if the “unknown” number was removed from the sample. This compares with Cooper’s (1966) statement that at least one third of individuals infected with rubella fail to demonstrate the symptoms.

Under the question as to whether rubella was confirmed by a doctor, responses to all three categories were quite similar. Unfortunately the question was not specific whether the confirmation was by a clinical antibody test or medical observation. Clinical tests of rubella antibodies have been available since the early 1970’s. Of the 33 who said “no” to the question on medical confirmation, 25 were born in or before 1975. Therefore, it is not likely that a clinical confirmation would have been made with most of that group.

What may be of concern is the reported incidence, although small (6), of respondents who reported that the birth mother’s rubella was a re-infection. By removing the unknown group from this sample, this reported rate of re-infection, although unconfirmed, is 11.5%. Others have reported incidences of re-infection, but they were believed to be rare. Sidle’s (1985) review of the literature described an infrequent number of incidences of rubella re-infections. Cherry (1987) suggested that the past history of rubella infection may not be particularly reliable. They also suggest, that because of the prevalence of other viral diseases which resemble rubella, descriptions of clinical rubella made prior to the modern virological diagnostic techniques are not always accurate.

Of further concern is the number of mothers (12) who reported that they had already been immunized against rubella. Again removing the unknown group from this sample, this rate of immunization rises to an astonishing 24%. Three of the twelve did not indicate when they were immunized. Of the nine who did, two reported being immunized during the year their child was born, two reported being immunized two years prior to their child’s birth, and one each reported being immunized at age 5, 8, 9, 12 and 14. The concern here is that for these respondents, prior immunization may not have been a guarantee to prevent infection from wild rubella. Sidle’s (1985) review of the literature also described incidences of rubella infection following vaccinations. Tingle (Personal Communication, 1998) indicated that the early vaccines were not as effective as those developed later.

2.1.8 Birth Situation

Individuals were asked to describe the nature of the child’s birth situation, such as the length of pregnancy, the known birth weight, type of delivery and whether or not the child was born with a rubella rash.

Of the eighty-six who reported about the nature of their birth, seventy-three (84.9%) reported natural childbirth, eight (9.3%) were born by caesarian section, while five (5.8%) reported a forceps birth.

Table 6 indicates the distribution among the three disability groups according to whether the birth was term or premature. Twenty reported a premature birth. Of the twenty individuals reporting premature births, one (5%) reported their disability as blindness, three (15%) reported their disability as deafness while 16 (80%) reported their disability as deafblindness.

Table 6: Summary of birth description according to sensory disability

Disability	Term Pregnancy	Premature Delivery	Not Specified	Total
Blindness	6	1	-	7
Deafblindness	49	16	5	70
Deafness	18	3	-	21
Total	73	20	5	98

Where there was a birth weight provided (78 of 98 respondents),**** forty-three (55.1%) reported the birth weight as 2500 grams or less (5.5 pounds), which is the weight categorized by the World Health Organization as premature. As indicated already, only 20% of the respondents reported their births as premature. Vernon (1969) reported that 40% of rubella deaf infants born during the 1960's in the USA, were below the 5.5 pound category.

According to sensory disability (Table 7), those who are deafblind had the most individuals (35) reporting low birth weight, and had individuals with the lowest mean weight (5.32 pounds).

Table 7: Summary of birth weight particulars according to sensory disability. Sample sizes shown in brackets.

Disability	Percent reporting birth weight 5.5 pounds and less	Mean reported birth weight (in pounds)
Blindness	2.6 (2)	5.98 (5)
Deafblindness	44.8 (35)	5.32 (58)
Deafness	7.7 (6)	5.92 (15)
Total	55.1 (43)	5.74 (78)

As a possible indication of the severity of the viral infection, individuals were asked whether the congenital rubella child had a rash at birth. Only sixteen of the 77 respondents to the question (20.8%) acknowledged the presence of a rash at birth.

2.1.9 Estimated Gestational Age at time of Maternal Infection

Individuals were asked to report an estimated gestational age in terms of which trimester of pregnancy they contracted maternal rubella. If they were infected during the first trimester, they were asked to verify which month the infection occurred. The responses are summarized in Tables 8 and 9 and are organized according to whether the individual reported their disability as blindness, deafblindness or deafness.

Table 8: Summary of reported gestational age of maternal infection with rubella according to disability

Disability	First Trimester	Second Trimester	Third Trimester	Unknown	Total
Blindness	4	1	0	2	7
Deafblindness	54	0	0	16	70
Deafness	15	3	0	3	21
Totals	73	4	0	21	98

Based on those individuals who reported the trimester of pregnancy (n=77 respondents), the vast majority (94.8%) of sensory deprived respondents contracted rubella during the first trimester, while 5.2% contracted rubella during their second trimester. No individual reported contracting rubella during their third trimester. Twenty-one of the 98 total respondents (who indicated their disability) did not know the period of pregnancy when rubella was contracted.

As Table 8 indicates, none of the individuals who are deafblind reported a trimester other than the first. Eighty percent (4 of 5) of individuals who are blind and 83.3% (15 of 18) of individuals who are deaf reported contracting rubella during the first trimester.

Table 9: Summary of month during first trimester when rubella contracted

Disability	First Month	Second Month	Third Month	Unknown	Total
Blindness	2	2	0	0	4
Deafblindness	21	19	9	5	54
Deafness	3	8	2	2	15
Totals	26	29	11	7	73

Sixty-six individuals reported an actual month during the first trimester when they contracted rubella. Fifty-five (83.3%) reported contracting rubella during the first two months, while eleven (16.7%) reported contracting rubella during the third month. Inadequate sample sizes prevent any clear correlation between month and sensory disability.

van Dijk (1991) reported that 17 children, whose mean time of infection was 4-7 weeks gestation, exhibited both visual and hearing impairments, while 54 children whose mean infection time was 10.6 weeks suffered hearing impairment only. Sallomi (1966) reported the following rates (based on analysis of other published studies) of congenital anomalies by gestational ages: weeks 1-4, 61%; weeks 5-8, 26% and weeks 9-12, 8%. Anomalies or congenital malformations are reported to be minimal beyond 12 weeks gestational age, with no defects reported after 20 weeks or five months.

2.1.10 Birth Mother's Health Issues

According to an article published in Newsday (07/22/97), researchers may have found a link between fetal cells left behind in a mother's blood and several serious auto-immune diseases (reported by N. O'Donnell in News From Advocates for Deaf-Blind, 1997). These auto-immune diseases are rheumatoid arthritis and Sjorgen's syndrome. The latter is an uncomfortable disorder that includes mucous membranes in the mouth and around the eyes, and a dry skin disease called scleroderma. Individuals with Sjorgen's syndrome, are observed to have symptoms of arthritis in one-third of the cases. Dr. Diana Bianchi, head of pediatric genetics at Tufts New England Medical Centre in Boston, reported in a news conference in 1997, that it was discovered several years ago that left-over white blood cells from the fetus may last as long as 27 years in the mother's bloodstream. This same unpublished research also reported that the fetal cells have been observed as early as seven weeks into the pregnancy. If this is indeed true, then there is a possibility that foreign rubella virus antigens could impair the mother's health for some years following their child's birth, or in the event of a miscarriage or abortion.

The survey asked individuals whether or not their birth mother had experienced, and, if so, at what age, symptoms of non-specified arthritis, asthma, early menopause and any other health issues that they wished to specify. Their responses are summarized in Table 10. The high number of unknowns reflects the questionnaires being completed by individuals other than the birth mother.

Table 10: Summary of birth mother's responses to health related issues

Health Symptoms	Yes	No	Unknown	Total
Arthritis (non-specified)	26	36	38	100
Early Symptoms of Menopause	15	44	41	100
Asthma	6	58	36	100
Other Symptoms	18	51	31	100

After removing the unknowns from each health issue category, the following percentage of respondents acknowledged "yes" to these health categories: non-specified arthritis - 41.9%; early symptoms (not specified) of menopause - 25.4%; asthma - 9.4% and other - 26.1%. The list of "other" symptoms which respondents reported include thyroid conditions, back problems, various allergies, cataracts, glaucoma, heart problems, psychiatric problems, multiple sclerosis, removal of spleen, fibromyalgia, endometriosis, elevated blood pressure, myopia and one individual stated "too many to mention."

To put a perspective on these results on birth mother's health issues, the number of years between the age the individual reported contracting rubella and the age at which she first reported the onset of the various conditions, is summarized in Table 11. No ages were provided for the category "other" health issues. For the 26 respondents acknowledging non-specified arthritis, one did not provide an age and another reported having arthritis symptoms prior to the child's birth.

Table 11: Summary of number of years between age birth mother contracted rubella and age reported initial symptoms (n=sample size)

Number of years between the age birth mothers contracted rubella and the age reporting initial	Symptoms of Arthritis	Symptoms of Asthma	Symptoms of menopause
0-10 years	1, 8, 9, 9, 9,10,10 (n=7)	1, 2 (n=2)	4, 5, 5, 10 (n=4)
11-20 years	11, 12, 13, 14, 16, 18, 18, 19 (n=8)	16, 18 (n=2)	11, 11, 12, 13, 14, 18, 18, 19, 19, 20, 20, 20 (n=12)
21-29 years	21, 22, 23, 25, 25, 26, 27, 27, 28, 29 (n=10)		23 (n=1)

For the six reporting asthma, one mother indicated she had asthma all her life while another did not report an age.

The birth mothers reported observing their initial symptoms of menopause at ages as follows: 27, 28, 29, 29, 30, 36, 38, 39, 40, 41, 41, 41, 43, 43, 43, 47 and 48. According to a physician (K. Van Alphen, personal communication, 1998), menopause symptoms occurring earlier than 45 years of age is not the norm.

At this point we cannot state definitely that any of these symptoms reported are directly related to rubella. The possibility that rubella viral infection is related to rheumatoid arthritis has been studied on several occasions according to Cherry (1987). Tingle (personal communication, 1998) reported his research in connection with 35 women from British Columbia who contracted rubella following a rubella outbreak in that province in 1985. Dr. Tingle observed that 10 of the women reported acute arthritis and 6 reported arthralgia (aching joints) for a total of 45.7% of his sample experiencing these auto-immune symptoms. Dr. Tingle also reported that two of the ten women had persistent symptoms of arthritis lasting longer than six months. He also suggested a link between rubella and fibromyalgia.

2.2 Section B

The remainder of the report presents the results of Section B of the survey questionnaire. Table 12 is a compendium table representing various sections of Section B. Tables (Tables 13 through 44 represent each of the 31 parts of Section B. Each table is set up similarly. The first column presents the question along with the size of the sample answering each particular question. For example, only 95 of the 100 individuals may have answered the question at hand. The second column indicates the percentage of the sample size answering "yes" to the question in the first column. The number shown in brackets in the second column (called response size) reflects the number answering "yes" to each question. The subsequent eight columns were set up to record the rate or percentages of first occurrence according to one of the seven age groupings in the survey questionnaire: 0-2, 3-6, 7-12, 13-21, 22-29, 30-39, 40 + . A column is also provided and labeled "?" to itemize those for which the age of first occurrence was unknown or not reported. The number in brackets in each age column indicates the number reporting the age of first occurrence, including "?".

2.2.1 Congenital Birth Defects

Table 12 presents the results about congenital birth defects as the respondents reported in various

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

sections in the questionnaire on visual loss, hearing loss, neurological conditions and heart and circulatory system. This table is not set up according to the format described for the subsequent tables. In Table 12, the specific birth defect is mentioned in Column 1, with the sample size shown in brackets. The reported incidence for all respondents is shown in Column 2, while the rate of incidence for individuals indicated their disability as deafblindness, deafness or blindness is indicated in Columns 3, 4 and 5, respectively. (Note that two of the individuals did not fit either of the three categories which accounts for in most incidences why the sum of the sample sizes in Columns 3, 4 and 5 does not equal the sample size in Column 2.)

Table 12: Summary of various congenital birth defects according to disability (n=sample sizes of respondents according to condition)

Birth defect	Incidence Among All Respondents	Incidence Among Respondents who were individuals with Deafblindness	Incidence Among Respondents who were individuals with Deafness	Incidence Among Respondents who were individuals with Blindness
Column #1	Column #2	Column #3	Column #4	Column #5
Microcephaly n = 96	33.3% (32)	36.4% (24)	4.8% (1)	71.4% (5)
Bulging anterior fontanelle n = 96	6.3% (6)	8.6% (6)	0.0%	0.0%
Congenital Heart Defect n = 99	65.7% (65)	73.9% (51)	47.6% (10)	42.9% (3)
Microphthalmia n = 95	31.6% (30)	35.7% (25)	14.3% (3)	14.3% (1)
Congenital Visual Loss n = 100	86% (86)	98.6% (69)	42.9% (9)	100% (7)
Congenital Hearing Loss n = 96	93.9% (92)	98.6% (69)	90.5% (19)	83.3% (5)
Congenital Hearing Loss + Visual Loss n = 96	80.2 (77)	98.6 (69)	38.1 (8)	60.0 (3)
Congenital Hearing Loss + Visual Loss + Heart Defect n = 95	58.9 (56)	72.1 (49)	28.6 (6)	20.0 (1)

Congenital Hearing Loss + Visual Loss + Heart Defect + Microcephaly n = 92	23.9 (22)	32.3 (21)	0	14.3 (1)
---	--------------	--------------	---	-------------

As Table 12 shows, congenital hearing loss represents the most frequently observed individual congenital defect, occurring in 92 (93.9%) individuals where this was reported. This is followed by visual loss at 86% and cardiac defects representing 65.7%. Individuals who are deafblind had the highest percentages of these congenital defects.

Cataracts were reported as the most common cause of congenital visual loss, accounting for all but nine (89.5%) of the eighty-six individuals reporting a congenital visual loss (see Table 14).

Another congenital birth defect, microcephaly, a medical term for a very small head, was reported in 33.3% of all individuals. This defect was observed in five of seven, (71.4%) of those who are blind. Just over one third (36.4%) of deafblind individuals reported microcephaly, while only 4.8% of those who were deaf reported this congenital condition. Microcephaly is caused by the premature fusion of the cranial sutures early in childhood. In some instances, microcephaly can lead to mental retardation if rapid brain growth occurs during the first six months following premature brain skull fusion. Givens et al (1993) reported microcephaly in 42% of congenital rubella cases examined in the Mayo Clinic study while Cherry (1987) reported that the frequency of occurrence is rare from the studies he has examined.

Only six individuals of ninety-five responding (6.3%) reported bulging anterior fontanelle, a clinical manifestation in infancy indicative of central nervous system involvement of congenital rubella (Slague and Wolinsky, 1989). The six reporting this condition are individuals who are deafblind. Cherry (1987) reported from two studies that the incidence of this condition was about 10% among individuals with congenital rubella. They suggest this condition relates to severe initial meningoencephalitis which can have later neurological implications, either motor, or mental, retardation.

Another congenital defect was that of small eyes or microphthalmia. Almost thirty-two percent reported this condition. The highest percentage was reported by those who were deafblind, 35.7%, while those who are blind and those who are deaf reported the same incidence rate at 14.3%. This condition was reported in 23% of the 125 cases examined by Slague and Wolinsky (1989). These authors suggest microphthalmia is a variable that may influence rubella related glaucoma. Thirteen (43.3%) of individuals reporting microphthalmia also had reported glaucoma, two of which were congenital glaucoma. Givens et al (1993) reported significant correlation between microphthalmia and glaucoma in the Mayo Clinic ophthalmic study.

The discussion above refers to the impact of rubella on one body system. A further complication of rubella is its impact on multiple systems. As Table 12 indicates, seventy-seven (80.2%) of the sample had two congenital defects (hearing and visual loss), fifty-six (58.9%) had demonstrated three congenital defects (hearing loss, visual loss, heart defect), while twenty-two (23.9%) had four congenital defects (hearing loss, visual loss, heart defect, microcephaly). The individuals who are deafblind (70) reported the highest rates of multiple disabilities.

2.2.2 Visual History/Visual Loss

Individuals were asked a series of questions about their visual history, including questions about visual loss, if any, and other questions in the category of vision. Their responses are summarized in Table 13.

Table 13: Summary of responses regarding visual history/visual loss

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Visual Loss n=100	91.0 (91)	-	94.5 (86)	4.4 (4)	-	1.1 (1)	-	-	-
Visual Loss Both Eyes n=100	84.0 (84)	-	90.5 (76)	8.3 (7)	-	1.2 (1)	-	-	-
Vision Severely Impaired n=100	71.0 (71)	4.2 (3)	83.1 (59)	8.5 (6)	1.4 (1)	1.4 (1)	-	-	1.4 (1)
Legally Blind n=98	74.5 (73)	-	-	-	-	-	-	-	-
Light Perception Only n=91	19.8 (18)	38.8 (7)	55.6 (10)	-	-	5.6 (1)	-	-	-
Other Symptoms Reported n=92	41.3 (38) See below	-	-	-	--	-	-	-	-
Reported Unable to Test n=94	22.2 (28)	-	-	-	-	-	-	-	-
Surgical Correction n=99	72.7 (72)	-	-	-	-	-	-	-	-
Used Corrective Lenses n=100	58.0 (58)	5.2 (3)	60.3 (35)	20.7 (12)	8.6 (5)	5.2 (3)	-	-	-
Reported Change in Visual Acuity n=89	48.3 (43)	4.7 (2)	18.6 (8)	20.9 (9)	16.3 (7)	27.9 (12)	9.3 (4)	-	2.3 (1)

All but 9 of respondents reported a visual loss, with the vast majority (94.5%) reporting the loss being congenital (0-2 years). Four reported the loss occurring during the period 3-6 years, while one reported the loss developing during the teen age period.

Respondents reported severe visual impairment at birth or during the first two years in 83.1% of cases. Six (8.5%) reported the impairment as being severe during the 3-6 year stage, while one each reported severe impairment occurring in the pre-puberty years, teens and 40+.

Eighteen respondents reported having light perception only, with seven (38.8%) not reporting an age of first occurrence. Of the eleven that did, ten reported light perception only during the period 0-2 years, and one showing first onset of having only light perception only during the teen period.

Almost seventy-three percent of the respondents (72.7%) had some form of surgery to correct their visual loss, and fifty-eight reported having worn corrective lenses sometime during their lives. Just over sixty percent of respondents (60.3%) reported use of corrective lenses sometime during their first two years.

Almost half of the respondents (48.3%) reported a change in visual acuity occurring sometime in their lives. Most of those reported a change by the time they were 21 years old, while four reported first occurrence during their twenty's and one during middle age.

Thirty-eight individuals (41.3%) reported other visual or ocular symptoms. They included the following

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

symptoms:

Astigmatism	9	Bilateral keratoconus	1
Nystagmus	6	Chorioretinopathy	1
Myopia	4	Cross-eyes	2
Hyperopia	1	Crystalline eye	1
Retinal Pigmentation	5	Extropiasia	1
Bilateral Aphakia	2	Hyperopia	1
Strabismus	3	Horizontal pendula	1
Micro-Cornea	2	Keratopathy	2
Amblyopia	1	Not specific	2
Aphakia	2	Dychromotopsie	1
Phthisis	2	Coloboma	1

2.2.3 Causes of Visual Loss

Individuals were asked a number of questions about their vision according to a number of parameters, including possible causes of their visual loss, if any. Their responses are summarized in Table 14.

Table 14: Summary of responses regarding causes of visual loss

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
n = sample size	(response size)	?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Cataracts One Eye n = 98	16.3 (16)	-	-	-	-	-	-	-	-
Cataracts Both Eyes n = 98	64.3 (63)	-	-	-	-	-	-	-	-
Cataracts One or Both Eyes n = 98	79.6 (78)	-	94.9 (74)	3.8 (3)	-	-	-	-	1.3 (1)
Glaucoma One Eye n = 96	13.5 (13)	-	-	-	-	-	-	-	-
Glaucoma Both Eyes n = 96	18.8 (18)	-	-	-	-	-	-	-	-
Glaucoma One or Both Eyes n = 96	32.3 (31)	-	35.5 (11)	9.7 (3)	22.6 (7)	25.7 (8)	6.5 (2)	-	-
Detached Retina n = 91	11.0 (10)	-	10.0 (1)	-	50.0 (5)	20.0 (2)	10.0 (1)	-	10.0 (1)
Eye Infections n = 90	27.8 (25)	28.0 (7)	36.0 (9)	24.0 (6)	-	4.0 (1)	4.0 (1)	4.0 (1)	-

Causes of visual loss are categorized as primary or secondary causes. Cataracts was the major cause of primary visual loss, occurring in one eye or both eyes in 79.6% of respondents. Most of the respondents reported congenital (reported during years 0-2) cataracts (94.9%), while four reported post congenital first

occurrences, three during ages 3-6 and one during middle age. For this discussion, congenital refers to that reported 0-2 years. O'Donnell (1996) similarly reported several incidences of cataracts occurring post congenitally.

Glaucoma, a secondary cause of visual loss, was reported in one eye or both eyes in almost one third (32.3%) of respondents. Eleven individuals (35.5%) reported congenital glaucoma. Three reported first occurrence during age period 3-6, while fifteen reported first occurrence during pre-puberty (7-12 years) and teen periods. Two individuals reported first occurrence of glaucoma during their 20's. This high rate of congenital glaucoma is at odds with Cherry (1987) who reported congenital glaucoma frequency rate at 5%.

Another secondary cause of visual loss, detached retina, was reported by 10 (11%) of respondents. One reported first onset during ages 0-2, five during their preteens and two during the teen period. One each reported first occurrence during their 20's and 40+ years. Twenty-five (27.8%) reported eye infections during their lives. Of the eighteen who could identify the age of first onset, fifteen reported the onset of eye infections during their first six years of life.

2.2.4 Hearing Loss

Individuals were asked a series of questions to determine their auditory history. Their responses are summarized in Table 15.

Table 15: Summary of responses regarding hearing loss

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Reported Hearing Loss n=98	96.9 (95)	2.1 (2)	94.7 (90)	3.2 (3)	-	-	-	-	-
Reported Hearing Loss One Ear n=96	2.1 (2)	-	-	-	-	-	-	-	-
Reported Hearing Loss Both Ears n=96	90.6 (87)	-	-	-	-	-	-	-	-
Hearing Loss Reported Mild to Moderate n=98	15.3 (15)	-	-	-	-	-	-	-	-
Hearing Loss Reported Severe to Profound n=98	83.7 (82)	-	-	-	-	-	-	-	-
Uses Hearing Device n=100	65.0 (65)							-	
Unable to Test for Hearing Loss n=99	27.3 (27)	-	-	-	-	-	-		-

Chronic Ear Infections n=93	26.9 (25)	16.0 (4)	64.0 (16)	12.0 (3)	4.0 (1)	-	4.0 (1)	-	-
Reported Change in Hearing Ability n=83	30.1 (25)	4.0 (1)	4.0 (1)	12.0 (3)	20.0 (5)	20.0 (5)	28.0 (7)	12.0 (3)	-

Hearing loss was reported by ninety-five (96.9%) of the respondents. Like visual loss, hearing loss was largely a congenital condition, with 94.7% reporting 0-2 years as the age of first occurrence or first observation. Although considered congenital, because of the nature of deafness, diagnosis of this condition in most, if not all individuals was not confirmed for sometime following birth. Consistent with this comment, three individuals reported first onset during the age period 3-6 years.

According to Cherry (1987), sensorineural deafness is the most common manifestation of congenital rubella, occurring in 80-90% of all patients. Together with central deafness and middle ear damage, he reported that almost all individuals born with congenital rubella have some form of hearing loss. The findings in this study correspond well to Cherry's findings.

The hearing loss reported was primarily bilateral, with only two (2.1%) reporting a hearing loss in only one ear. Most respondents (83.7%) reported a hearing loss in the severe to profound range (greater than 70 dB loss), while 15.3% of individuals reported their hearing was in the mild to moderate range (26-70 dB loss). Twenty-seven (27.3%) reported not able to be tested for hearing, with reasons stated for this as behavioural and communication difficulties. Over two thirds (65.3%) of respondents reported using a hearing device sometime during their life. Just over one-quarter (26.9%) reported having ear infections, with 64% of those reporting ear infections during their first two years of life.

Twenty-five individuals (30.1%) reported a change in hearing or auditory acuity. Similar numbers reported a change in condition during their preteens (5), teens (5) and twenties (7). Three reported a change during their 30's. In most cases, respondents did not specify whether the change was an improvement or deterioration. Six reported that the change was a deterioration, two reported this change as occurring in their preteen period (one reported a clinically diagnosed 10 dB loss at 8.75 years), one indicated their hearing became worse during their teens, two reported the deterioration occurring during their 20's (one specified at age 22), while one indicated that the hearing loss was gradual during their life. Where an improvement was indicated, the reasons reported included benefits from using a hearing device to being able to use their residual hearing better.

According to Gilbert (1991), most studies have reported a high prevalence of both severe and mild-moderate hearing loss that can progress after birth. While this study may suggest that just over thirty percent showed a change in hearing, it is unclear from the nature of the responses how much of that incidence represents a progressive loss.

2.2.5 Neurological Conditions and Symptoms

This section of the survey was to report specified symptoms which may be related to a change in neurological condition. Respondents were asked whether there was any demonstrated memory loss, irritability, lethargy or an inappropriate response to pain. Furthermore, they were asked whether they were ever diagnosed with any Alzheimer-like conditions, including dementia, disorientation, wandering, forgetfulness, aggression, sleeping or eating disorders, loss of sense of hot or cold. A summary of their responses is found in Table 16.

Table 16: Summary of responses regarding neurological conditions and symptoms

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Diagnosed with Alzheimer-like symptoms (showing 3 of those indicated) n=98	14.3 (15)	26.7 (4)	20.0 (3)	6.7 (1)	6.7 (1)	13.2 (2)	20.0 (3)	6.7 (1)	-
Lethargy n=98	32.7 (32)	28.1 (9)	28.1 (9)	6.3 (2)	3.1 (1)	9.4 (3)	9.4 (3)	12.5 (4)	3.1 (1)
Irritability n=98	59.2 (58)	20.7 (12)	32.7 (19)	6.9 (4)	22.5 (13)	8.6 (5)	8.6 (5)	-	-
Other Neurological Conditions n=99	17.2 (17)	-	-	-	-	-	-	-	-
Memory Loss n=87	12.6 (11)	18.2 (2)	-	-	-	36.3 (4)	27.3 (3)	9.1 (1)	9.1 (1)
Inappropriate Response to Pain n=93	24.7 (23)	34.8 (8)	43.6 (10)	4.3 (1)	13.0 (3)	-	4.3 (1)	-	-

For the Alzheimer questions, if the respondent noted the presence of three or more of the conditions, the answer was placed in the affirmative. Fifteen (14.3%) responded “yes” to this question, with the conditions noted primarily aggression, sleeping disorders, memory loss, eating disorders and loss of sense of hot and cold. The conditions of dementia, wandering or disorientation were not mentioned by any of the respondents. The responses are too scattered across the age groups, with almost a third in the “unknown” category, to make any further comments about this question.

The conditions in this section seem to be most prominent during the early infant years. Respondents to the questions on lethargy, irritability and inappropriate response to pain, and Alzheimer-like symptoms, noted first occurrence in the 0-2 age category at rates of 28.1%, 32.7%, 43.6% and 29.0%, respectively. The prominence of lethargy and irritability during the first two years likely relate to the transitory neurological conditions caused by rubella. Irritability appeared to increase in the preteen period (22.5%), and memory loss was observed to a larger degree in respondents in their teens and older. The large number of respondents in the unknown age at initial onset make it difficult to make any further observations about this section of the survey.

Gilbert (1991) indicated that lethargy and irritability are symptoms of neurological manifestations of congenital rubella, which confirms the high incidence (especially for irritability) observed during the age period 0-2 years.

Seventeen (17.2%) of respondents noted “Other Neurological Conditions.” These included the following:

Autism	3	Tourettes’s Syndrome	1
Cerebral Palsy	4	Suspected vestibular syndrome	1
Attention Deficit Syndrome (ADHD)	2	Haematoma due to skull fracture	1

Bipolar Psychosis	1	Asymmetry of face	1
Diagnosed Schizophrenia	1	Quadripareisis	1
Cold aglutein (intolerance to cold)	1	Weakness on left side	1

A number of these conditions are not typical neurological conditions but were reported by respondents for this question.

2.2.6 Neurological Development

Individuals were asked to comment on their neurological or intellectual development according to a series of questions. Their responses are summarized in Table 17.

Table 17: Summary of responses regarding neurological development

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
n=sample size	(response size)	?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Normal Intelligence n=95	38.9 (37)	-	97.3 (36)		2.7 (1)	-	-	-	-
Above Average Intelligence n=95	9.5 (9)	11.1 (1)	55.6 (5)	22.2 (2)	11.1 (1)	-	-	-	-
Intelligence Evaluation Downgraded n=96	7.3 (7)	42.9 (3)	-	-	-	42.9 (3)	-	14.2 (1)	-
Diagnosed with Mild Mental Retardation n=96	13.5 (13)	7.7 (1)	61.5 (8)	7.7 (1)	15.4 (2)	7.7 (1)	-	-	-
Diagnosed with Moderate Mental Retardation n=93	10.8 (10)	50.0 (5)	10.0 (1)	40.0 (4)					
Diagnosed with Profound Mental Retardation n=93	35.5 (33)	18.2 (6)	48.5 (16)	12.1 (4)	12.1 (4)	6.1 (2)	3.0 (1)	-	-
Initial MR Diagnosis Upgraded n=88	11.4 (10)	20.0 (3)	-	20.0 (2)	20.0 (2)	30.0 (3)	-	-	-
Diagnosed with Autism n=93	16.7 (20)	20.0 (4)	20.0 (4)	45.0 (9)	15.0 (3)	-	-	-	-

Over one-third (38.9%) of those who responded about intelligence indicated their intelligence was normal,

while nine (9.5%) suggested having above average intelligence. Of those who categorized themselves as having above average intelligence, four were individuals who were deaf and four were individuals who were deafblind, while one was an individual who was blind. This evaluation was a subjective appraisal by the parent or advocate and not verified medically or by intelligence testing. This subjective evaluation was subsequently downgraded or re-evaluated to be something less than normal for seven (7.3%) of those respondents whose intelligence was first judged as "normal." This downgrading occurred for three during their teen period and one during their 30's. Three of those individuals did not have a known age assigned for this downgrading.

Fifty-six respondents also received an objective medical diagnosis about their neurological development sometime during their lives. Thirteen (13.5%) were identified as having mild mental retardation, ten (10.8%) as being moderately mentally retarded and thirty-three (35.5%) as profoundly mentally retarded. Fourteen of the nineteen reported as profoundly retarded were placed in a special facility for individuals with developmental disabilities at an early age. Ten (11.4%) of the individuals had their early diagnosis modified or upgraded presumably on the basis of communication development.

This reported incidence compares to the findings from the Rubella Birth Defects Evaluation Study (Cooper et al, 1969) which reported that nearly 50% of infants studied developed psychomotor retardation ranging from mild intellectual impairment to severe mental retardation with spastic diplegia.

Cherry (1987) suggested, however, that many infants (and this can be extended for those older individuals) labeled "retarded" are really individuals with auditory and visual defects who initially did not have proper diagnosis and training for their handicaps. He inferred that only about 10% of congenitally infected rubella individuals have a central nervous system defect that precludes normal development.

Ziring (1978) reported about a study of 74 congenital rubella children considered to be mentally retarded by the age of four or five, that 21 of them were no longer classified as mentally retarded, by the age of eight or nine. Ziring also suggested that once the encephalopathic process "clears up" (between the age of 3 and 5), there occurs a spurt in the child's development, making many early developmental evaluations premature and inaccurate.

Twenty (16.7%) of individuals responding in this study to the question about autism indicated being identified as autistic. Autism is an organic condition or disorder of the central nervous system caused by neurological abnormalities (Wagman et al, 1988). Those who reported an age of diagnosis indicated being identified with this condition during their early youth. It is not clear from this survey which of those labeled autistic had this diagnosis based on a medical diagnosis or a subjective evaluation.

Like mental retardation, the relationship of autism to multi-sensory impairment and hence in many instances, to congenital rubella, is one that has received considerable debate and discussion. Chess et al (1971) among others have argued that congenital rubella is a causative factor in early infantile autism. Cooper (1985) reported a study led by Dr. Stella Chess and associates that suggested an autism incidence rate of 6%, with or without associated hearing and visual defects. The symptoms of withdrawal, the delay in developing social relationships and the presence of quasi-obsessive and ritualistic types of behaviours make children with congenital rubella to be easily labeled or mis-labeled, whatever the case may be. According to Dr. Linda Mamer (personal communication, 1998), current research has determined that genetic factors alone do not cause autism but they may increase susceptibility to developing this condition. Autism may then develop as a result of some environmental exposure or trigger.

Indeed it may be plausible that congenital rubella may be that trigger or causative factor, suggested by Chess (1971), that influenced the rather high percentage of individuals (16.7%) diagnosed as autistic.

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

van Dijk (1991) reported that researchers in the field suggest that as multi-sensory deprived individuals (who are deafblind) develop their communicative skills further, their aberrant behaviour diminishes and consequently, in many cases their autism tendencies largely disappear. van Dijk (1991) also reported that there is a diagnostic instrument currently available to differentiate between individuals diagnosed as autistic, and those who are diagnosed as deafblind.

2.2.7 Mental Health

Individuals were asked several questions concerning mental health. Table 18 summarizes their responses.

Table 18: Summary of responses regarding mental health

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Change in Level of Anxiety n=96	32.3 (31)	19.4 (6)	9.7 (3)	16.0 (5)	9.7 (3)	25.8 (8)	12.9 (4)	6.5 (2)	-
Indications of Depression n=99	44.4 (40)	10.0 (4)	5.0 (2)	15.0 (6)	22.5 (9)	12.5 (5)	27.5 (11)	5.0 (2)	2.5 (1)
History of Traumatic Event n=97	40.2 (39)	10.3 (4)	23.1 (9)	7.7 (3)	12.8 (5)	25.6 (10)	10.3 (4)	7.7 (3)	2.5 (1)
Treatment for Mental Health n=98	40.8 (40)	12.5 (5)	5.0 (2)	15.0 (6)	10.0 (4)	25.0 (10)	27.5 (11)	5.0 (2)	-

Almost one-third of respondents (32.3%) indicated a change in their level of anxiety. Across the age groups, the teen period represented the age showing the highest percentage of first occurrence for this condition, with eight (25.8%) indicated a change during this period.

Forty respondents (44.4%) responded “yes” to indications of depression. For this indicator of mental health, the preteen and 20’s periods had the highest percentage of respondents indicating first onset. Surprisingly, the older age periods had the lowest percentages of first onset. Four individuals did not indicate an age of onset of depression.

Thirty-nine (40.2%) of all respondents indicated a traumatic event occurring sometime during their lives. Respondents did not specifically indicate what these events were in this question, but throughout the questionnaire respondents noted such events as family separation, multiple hospitalizations with medical treatments, physical abuse, death of a family member, living apart from the family a large part of the year in a residential school, and the disability itself of hearing and/or visual loss, as representing examples of traumatic events in their lives. Many of these individuals reported more than one of these events, with their frequency tending to be higher during their early years.

A surprising forty (40.8%) of respondents indicated being treated sometime during their lives for mental health. The treatment included counseling and medication. Thirty of these had received medication while fourteen had received counseling services for their mental health problems.

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

2.2.8 Behaviour

Individuals were asked to answer numerous questions on the broad topic of behaviour. Their responses are summarized in Table 19.

Table 19: Summary of responses regarding behaviour

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Change in Attention Span n=94	28.7 (27)	11.1 (3)	7.4 (2)	11.1 (3)	22.3 (6)	25.8 (7)	22.3 (6)	-	-
Demonstrated Continuous Random Purposeless Movements n=99	55.6 (55)	-	-	-	-	-	-	-	-
Demonstrated Persistent Unusual Behaviour n=100	51.0 (51)	-	-	-	-	-	-	-	-
Demonstrated Ritualistic Compulsive Behaviour n=98	30.6 (25)	-	-	-	-	-	-	-	-
Demonstrated at least One of these Behaviours n=100	70.0 (70)	18.6 (13)	45.7 (32)	12.9 (9)	11.4 (8)	8.6 (6)	2.8 (2)	-	-
Performed Self Stimulating Behaviour n=97	59.8 (58)	20.7 (12)	43.1 (25)	19.0 (11)	6.9 (4)	6.9 (4)	1.7 (1)	1.7 (1)	-
Demonstrated Aggressive Behaviour n=100	65.0 (65)	-	-	-	-	-	-	-	-
Demonstrated Self Injurious Behaviour n=100	61.0 (61)	-	-	-	-	-	-	-	-
Demonstrated Tantrums or Outbursts n=99	73.7 (73)	-	-	-	-	-	-	-	-
Destroyed or Attempted to Destroy Property n=98	42.9 (42)	-	-	-	-	-	-	-	-
Demonstrated One of: Aggressive, Self Injurious, Tantrums or Destruction of Property n=100	80.0 (80)	8.8 (7)	26.1 (21)	23.7 (19)	25.0 (20)	11.3 (9)	3.8 (3)	1.3 (1)	-
Increase in the Incidence of these Behaviours n=99	30.3 (30)	10.0 (3)	3.3 (1)	10.0 (3)	13.3 (4)	30.0 (9)	23.4 (7)	10.0 (3)	-
Decrease in the Incidence of these Behaviours n=99	53.5 (53)	15.1 (8)	1.9 (1)	5.7 (3)	9.4 (5)	26.3 (14)	20.8 (11)	17.0 (9)	3.8 (2)
Demonstrated any Sexual Frustration n=89	27.0 (24)	8.3 (2)	-	8.3 (2)	16.8 (4)	20.8 (5)	41.6 (10)	4.2 (1)	-

As Table 19 indicates, many individuals with congenital rubella demonstrate, during their lives, numerous behavioural characteristics which may or may not be indicators of undisclosed problems.

Twenty-seven (28.7%) of respondents indicated a change in attention span. Sixteen of those indicated

an improved attention span, nine indicated a deterioration, and two did not indicate the nature of the change. It would appear that real change in attention span improves with age in this sample as indicated by higher percentages of age of first onset from the preteens through the 20's. There were no respondents that showed a first onset of change in attention span, older than 30 years of age. Of the nine who indicated a decrease in attention span, one indicated age of first occurrence during age period 3-6, one during the preteens, two during their teens, four during their 20's and one during their 30's.

Over half (55.6%) demonstrated continuous random purposeless movements, fifty-one percent demonstrated persistent unusual behaviour, while 30.6% demonstrated ritualistic compulsive behaviour. Seventy percent demonstrated at least one of these behaviours. The prime period during which these behaviours showed first onset was during the first two years of life. These behaviours appear to be an early manifestation of visual and hearing impairments, at the least, since their age of first onset decreases across the age continuum. This was not to suggest that the behaviours disappeared as the individuals became older. In fact, several respondents indicated a worsening of these behaviours during their 20's and 30's. One individual had their worsening behavioural condition medically diagnosed as possible Obsessive-Compulsive Mental Disorder. It is unknown about whether congenital rubella is the cause of these complex behaviours.

Dr. Collins, a psychiatrist from McMaster University indicated he was not previously aware of Obsessive-Compulsive Disorder with individuals with congenital rubella syndrome but suggested it was a late onset condition observed in some individuals with Down's Syndrome (personal communication, 1998). It is unknown if congenital rubella is the cause of these complex behaviours.

Fifty-eight (59.8%) of respondents acknowledged performing self-stimulation behaviour, with the vast majority confirming their first onset during their first six years of life. Only ten reported age of first onset of self-stimulation behaviour after the age of six. Twelve did not indicate an age of first onset since no information was available for their early lives. It could be suggested that self-stimulating behaviour is primarily an early manifestation of congenital rubella. Since most of those responding to self-stimulation were individuals who are deafblind, it can also be similarly suggested that the combined visual and hearing loss contributes to this behaviour, regardless of whether congenital rubella was the cause of the sensory losses.

Respondents acknowledged a very high response rate to such behaviour as aggression, self-injury, tantrums or outbursts and destruction or attempting to destroy property. Eighty percent demonstrated one of these behaviours. Table 19 suggests the incidence of these behaviours (as indicated by age of first onset) is highest during the first twelve years of life. Only four reported first onset of these behaviours after their teen years. This is not to suggest that these problems disappear with age, in fact, 30.3% indicated an increase in these behaviours.

There is a positive side to this behavioural situation, however, since over half (53.5%) of respondents noted a decrease in the incidence of these behaviors.

Twenty-four (27%) of respondents acknowledged some form of sexual frustration, although not specified. Fourteen of these individuals were males, ten were females. Where an age was indicated, the primary period for sexual frustration was noted to occur between the preteen period through the 20's.

Thirty-two, or one third demonstrated noticeable lethargy or loss of energy

Numerous authors discuss the frequencies of such maladaptive disorders as aggressiveness, destructiveness, stereotyped and ritualistic behaviour and their association with congenital rubella (van Dijk, 1991). They usually attribute many of the behavioural disorders to neurological damage initiated

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

during the early gestational developmental phase. There are also reports of later emergence or escalation of behaviours, which Ziring (1967) believed may be the result of progressive or degenerative neurological damage caused by congenital rubella. Several of these authors referred to by van Dijk (1991) stress that when relating behavioural disorders with neurological damage not to overlook the implications of multiple sensory impairments and the problems created from lack of communication.

John Walters, Helen Keller National Center, Sands Point New York, reported in the Fall 1994 edition of the *National Family Association of Deaf-Blind Newsletter*, that he has observed many common behaviours within the population of congenital rubella individuals. From a functional perspective, Walters characterized these behaviours into three categories: the need for ordered environments, ritualized behaviours and item adoration. Walters described an ordered environment as the desire or practice of an individual to maintain a consistent, non-changing world; a ritualized behaviour as repetitive actions which continue in duration, process or frequency past that which is necessary to complete, and item adoration as the practice where an individual places a great deal of emphasis on a specific object or objects.

Walters believes that, for individuals with Congenital Rubella Syndrome who demonstrate these two behaviours, an ordered environment and ritualized behaviour, perhaps relate to some sense of control and a feeling of safety and comfort. Material items, Walters suggests, may point to predictability and reliability. These attributes, together with control, safety and comfort mentioned by Walters, together point to some need for a structured environment for these individuals. Walters also mentioned that in cases where ritualized behaviours have been changed by someone or something in the environment, the result is often that the person who is deafblind becomes self-injurious, aggressive toward others or destructive of property.

2.2.9 Seizures

Individuals were asked whether they had any history of seizure activity. Their responses are summarized in Table 20.

Table 20: Summary of responses regarding seizures

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Experienced Seizures n=100	30.0 (30)	6.7 (2)	6.7 (2)	10.0 (3)	16.6 (5)	30.0 (9)	20.0 (6)	10.0 (3)	-
Petit Mal Seizures n=95	6.3 (6)	-	-	-	-	-	-	-	-
Psychomotor Seizures n=95	1.1 (1)	-	-	-	-	-	-	-	-
Grand mal Seizures n=95	16.8 (16)	-	-	-	-	-	-	-	-
Focal Seizure n=96	7.3 (7)	-	-	-	-	-	-	-	-
Change in frequency of occurrence n=30	46.7 (14)	7.1 (1)	-	7.1 (1)	-	35.8 (5)	21.4 (3)	14.3 (2)	14.3 (2)
Seizures treated by medication n=30	53.3 (16)	-	-	6.2 (1)	18.8 (3)	43.8 (7)	18.8 (3)	6.2 (1)	6.2 (1)

Thirty of the respondents (30%) had indicated an occurrence (not specified as clinical) of seizures at least once during their lives. One could conclude from Table 20 that seizure activity is a late manifestation of congenital rubella. Two-thirds or twenty reported the first onset of seizures occurred during the period age 7 through age 30. Five of those acknowledging seizures experienced seizures during their first six years of life, while three reported an initial onset of seizures during their 30's. Two individuals reported seizures during their first two years. Cherry (1987) reported that infantile seizures are rare, transitory and probably related to meningoencephalitis, a condition which may, with congenital infants last up to one year.

The onset of puberty is strongly suspected as precipitating this neurological condition. The teen period had the highest first onset percentage, nine (30%), while five (16.6%) noted first occurrence during the preteen period. Of the five from the preteen period, one was reported at age 7.75, one was age 12 while the ages of the other three were unstated.

Sixteen of the thirty reporting seizures (53.3%) experienced grand mal seizures and six (20.04%) experienced petit mal seizures. Focal seizures were experienced by seven (23.3%) while psychomotor seizures were reported by one (3.3%). No individuals reported their seizure as focal or psychomotor alone. Individuals reporting these types of seizures were always together with grand mal or petit mal. In one instance, an individual reported both a focal and psychomotor seizure.

Fourteen of those reporting seizures (46.7%) indicated a change in the frequency of occurrence. Nine of those reported an absence or infrequency of seizures, one reported an increase in the frequency of seizures, the other six did not indicate whether the change was an improvement or a deterioration.

Sixteen of those reporting seizures (53.3%) had taken medication to prevent seizures sometime during their lives. Currently ten, or one-third, report they continue to take medication for the prevention of their seizure activity.

Ziring (1978) reported an increase in seizure disorders appearing during or shortly after puberty in a USA population. He mentioned an incidence rate of 2.1% (eleven of 517 individuals) with clinical and electroencephalographic evidence of a seizure compared to 0.5% in the general population. The results reported in this study suggest a much higher rate of incidence of seizures than that indicated by Ziring, while recognizing that the reported occurrences are often anecdotal and may not always reflect a clinical diagnosis. The Canadian statistics also confirm Ziring's observations about an increase in seizures around the age or period of puberty.

2.2.10 Sexual Development

Individuals were asked about their history of sexual development and any issues around sexual function. The summary of their responses is found in Table 21.

Table 21: Summary of responses regarding sexual development

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Age of Onset of Sexual maturity n=90	93.3 (84)	6.0 (5)	-	-	40.5 (34)	46.3 (39)	6.0 (5)	1.2 (1)	
Inability With Sexual Function n=85	10.6 (9)	11.1 (1)	-	-	11.1 (1)	44.5 (4)	22.2 (2)	11.1 (1)	-
Females Experiencing Menstrual Problems n=58	60.3 (35)	5.8 (2)	-	-	14.3 (5)	42.8 (15)	34.2 (12)	2.9 (1)	-
Females Demonstrating Symptoms of Menopause Problems n=57	12.3 (7)	-	-	-	-	14.3 (1)	42.9 (3)	28.5 (2)	14.3 (1)
Females Demonstrating Noticeable Increases in Breast Size n=56	19.6 (11)	-	-	-	9.1 (1)	45.4 (5)	36.4 (4)	9.1 (1)	-

The period of onset of sexual maturity for all individuals was spread over periods 7-12 (40.5%) and 13-21 (46.3%). For the eight males who reported an exact age of onset, their ages reported ranged from 10 to 14 years. One male respondent age 18, reported no evidence yet of sexual maturation.

There was a much wider range of ages among the thirty-two females who reported an exact age of onset of sexual maturity. Their ages ranged from 7 to 34 years of age. Just over half of the female respondents to age of sexual maturity reported the occurrence between the ages of eleven and fourteen. Below that range of ages, one each reported onset of sexual maturity at ages seven and nine and three reported onset at age ten. Above the age of 14, six reported age fifteen, and one each at age 16, 18, 20, 21, 23 and 34. All but one of the females reporting first onset at age 15 and older live or have lived in an institution. Of interest, is that one individual reported no menses since she was born without a uterus.

Nine (10.6%) report a problem with sexual function. Of these nine individuals, one reported this condition occurring during their preteens, four in their teens, two in their 20's and one during their 30's. Six of these individuals were males and three were females.

Thirty-five (60.3%) of female respondents report experiencing problems with their menstrual cycle, including irregular periods and premenstrual syndrome.

Symptoms of menopause was reported by seven (12.3%) of females responding. One reported symptoms first occurring during her teen period, three during their 20's, two during their 30's and one during her 40's.

Eleven (19.6%) of females reported a noticeable increase in breast size, of which normal teen age breast development accounted for six of these observations. Three respondents, however, did report observations about breast development which would be considered unusual. These three observations included one reporting large pendulous breasts, one reporting fibrocystic disease and one reporting very noticeably large breasts (with one being larger than the other).

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

2.2.11 Sleep/Wake Patterns

Sleeping is a common concern within the population of individuals who are deafblind. Several questions were asked about the sleep/wake issue. The responses are summarized in Table 22.

Table 22: Summary of responses regarding sleep/wake patterns

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Experiencing Persistent Sleep Wake Problem n=99	39.4 (39)	12.8 (5)	53.8 (21)	10.3 (4)	7.7 (3)	7.7 (3)	2.6 (1)	5.1 (2)	-
Experiencing Change in Sleep/Wake Pattern n=97	30.9 (30)	10.0 (3)	-	6.7 (2)	16.7 (5)	23.3 (7)	23.3 (7)	20.0 (6)	-

Thirty-nine (39.4%) indicated they had experienced persistent problems with sleep. As Table 22 indicates, the sleeping issue is particularly a problem during the first two years of age since 53.8% indicated initial onset of persistent sleep/wake problems during this period. The lower percentages after age two does not suggest that sleeping problems disappear, as most respondents report that the sleep issue is an ongoing problem.

Thirty (30.9%) indicated that the individual experienced a change in sleep/wake patterns. Sixteen indicated an improvement, five noted a worsening, while nine did not suggest the nature of the change. For those noting an improvement in sleep/wake patterns, the reasons provided were mixed between general improvements with age, to the use of medication.

It can be observed in Table 22 that for those reporting a change in sleeping patterns the change tends to occur with increasing age.

Much is written about sleep in the general population and about the many negative implications to the individual receiving insufficient or interrupted sleep. Dr. Ann Poindexter was quoted in the Summer 1997 edition of *News From Advocates for Deaf-Blind* that many mentally retarded individuals with whom she is familiar, who are blind and without light perception, and who are living in institutions, have significant sleep disorders, with disturbances of sleep-wake patterns. She reported that many of these individuals demonstrate an increase of maladaptive behaviours, in particular self-injurious behaviour, when their day does not fit society's day. Many of these individuals, according to Dr. Poindexter, are consequently diagnosed as "psychotic disorders", "bipolar disorder" and/or with autism.

2.2.12 Size (Height and Weight) of individuals

Individuals were asked several questions to gather information about their size. Their responses are summarized in Table 23.

Table 23: Summary of responses regarding size (height and weight) of individuals

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	22-30	40+
Born Low Birth Weight n=82	54.9 (45)	-	-	-	-	-	-	-	-
Weight Normal Throughout Life n=99	50.5 (50)	16.0 (8)	66.0 (33)	14.0 (7)	2.0 (1)	-	2.0 (1)	-	-
Significant Changes in Body Weight n=98	35.7 (35)	17.1 (6)	2.9 (1)	2.9 (1)	11.4 (4)	31.4 (11)	28.6 (10)	5.7 (2)	-
Height Normal Throughout Life n=100	53.0 (53)	-	-	-	-	-	-	-	-

Forty-five (54.9%) of respondents reported a birth weight of 2500 grams (5.5 lbs) or less and are referred to subsequently in this study as born with low birth weight (reported in Table 7 with slight differences in sample size and percentage occurrence).

Approximately fifty percent of respondents reported that their weight (50.5%) and height (53%) was considered normal throughout their lives. Conversely the other fifty percent reported their weight (49%) and height (47%) was below normal at least sometime during their lives.

It would appear that a manifestation of congenital rubella is physical development delay. Table 23 confirms that over half of the respondents are affected. This effect began with a high number being born with low birth weight (54.9%) and continued through life with many individuals becoming small adults. Cherry (1987) reports that the most common manifestation of congenital rubella, and readily apparent at birth, is growth retardation, with between 50 and 85% of all babies being born below 2500 grams or 5.5 pounds, even at normal gestational age.

While the questionnaire did not request that the relative size of the individual be noted, many respondents made comments about the individual's smaller than average size. Size comments were made in the questionnaires for 15 adult males and 13 adult females. Of the males reporting, five indicated they were generally slight and slim, six provided the following heights: 5'8", 5'5", 5'5", 5'3", 5'0" and 4'11", two provided weights of 108 and 118 pounds, and one indicated they had always been 2-4 years behind in physical development. Of the females reporting, five indicated they were small, eight provided the following heights: 4'0", 4'3", 4'8", 4'10", 4'11", 5'3", one less than 5' and one 5'6" (weight 95 pounds). An issue that may be contributing to the generalized short stature and relatively small size of individuals with congenital rubella is a growth hormone deficiency, relating to a disorder of the hypothalamus (O'Day and Marshall, 1988).

2.2.13 Skeletal Conditions

Individuals were asked whether they had experienced any spinal related physical conditions. Their responses are summarized in Table 24.

Table 24: Summary of responses regarding skeletal conditions

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Presence of Scoliosis n=100	16.0 (16)	31.3(5)	12.5(2)	-	18.7(3)	12.5(2)	18.7(3)	6.3(1)	-
Presence of Kyphosis n=100	6.0 (6)	16.7(1)	-	-	66.6(4)	16.7(1)	-	-	-
Presence of Lordosis n=100	9.0 (9)	33.3(3)	-	-	44.5(4)	11.1(1)	11.1(1)	-	-
Presence of One of Those 3 Conditions n=100	26.0 (26)	23.1(6)	7.7(2)	-	38.5(10)	15.4(4)	11.5(3)	3.8(1)	-
Changes in Posture n=100	16.0 (17)	5.9(1)	-	5.9(1)	17.7(3)	35.3(6)	23.5(4)	11.7(2)	-
Other Physical Changes n=99	12.1 (12)	25.0(3)	8.3(1)	8.3(1)	25.1(3)	8.3(1)	25.0(3)	-	-

Individuals were asked to report about any developing conditions affecting their skeletal system. Respondents reported the presence of the following spinal curvatures: sixteen (16%) with scoliosis, a lateral curvature of the central part of the spine; nine (9%) with lordosis, an exaggerated forward curvature of the spine caused usually by a hip deformity or defect in posture, and six (6%) with kyphosis, an exaggerated backward spinal curvature characterized by a humpback appearance.

Twenty-six (26%) reported the presence of at least one of these spinal conditions. Two individuals reported the onset of one of these conditions during their first two years, ten reporting initial onset during the preteen period, four during their teens, three occurring during their 20's and one during their 30's. Six did not know the age of first onset. Spinal deterioration appears to be a later manifestation of congenital rubella and is possibly related to the impact of rubella on physical development delay.

Seventeen (16%) reported a change in posture, with the deterioration manifesting itself largely during late childhood through the teens, twenties and thirties.

Twelve (12.1%) of respondents reported the following "other physical changes":

X Distended stomach	1	X Deterioration due to lack of physical exercise	1
X Malformed stomach	1	X Asymmetry of face	1
X Problem with toes and joints	1	X Mild dystonia paraplegia	1
X Pain in knees and joints	1	X Hemi-paresis dysphasia	1
X Problem with neck	1	X Vertigo... Meuniere's syndrome	1
X Left leg shorter than right leg	1	X Didn't specify	1
X Quadriplegic	1		
X Weakness on left side	1		
X Chest larger on one side	1		

Three of these, one with distended stomach, one with malformed stomach (possibly the same condition) and vertigo, do not appropriately fit this section but there was no other section in which to place them.

The two referring to knees, toes and joints are possibly symptoms of arthritis. The respondents who noted quadriplegia, weakness on one side, dystonia paraplegia and hemi-paresis dysphasia are more appropriately symptoms of motor ability deterioration.

2.2.14 Motor Skills

Respondents were asked a series of questions about motor skills to learn more about early motor development and assess whether any late emerging deterioration in motor skills was being observed. Responses are summarized in Table 25.

Table 25: Summary of responses regarding motor skills

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Symptoms of Cerebral Palsy-like Movements n=99	23.2 (23)	8.7 (2)	56.6 (13)	13.0 (3)	8.7 (2)	-	4.3 (1)	8.7 (2)	-
Change in Cerebral Palsy-like Movements n=100	6.0 (6)	16.7 (1)	-	33.2 (2)	16.7 (1)	-	16.7 (1)	16.7 (1)	-
Learning to Walk n=100	96.0 (96)	11.5 (11)	36.4 (35)	45.7 (44)	4.2 (4)	1.1 (1)	1.1 (1)	-	-
Deterioration in Mobility (Walking) n=99	21.2 (21)	14.3 (3)	4.8 (1)	4.8 (1)	9.5 (2)	9.5 (2)	38.1 (8)	9.5 (2)	9.5 (2)
Deterioration in Balance n=99	23.2 (23)	17.4 (4)	13.0 (3)	8.7 (2)	4.3 (1)	-	34.9 (8)	13.0 (3)	8.7 (2)
Deterioration in Energy Level, Stamina and Endurance n=100	21.0 (21)	14.3 (3)	4.8 (1)	-	9.5 (2)	19.0 (4)	38.1 (8)	9.5 (2)	4.8 (1)
Change in Ability to Sit n=100	1.0 (1)	-	-	-	-	-	-	-	-
Deterioration in Fine Motor Skills n=97	4.1 (4)	25.0 (1)	25.0 (1)	-	-	-	50.0 (2)	-	-

Twenty-three of respondents (23.2%) reported cerebral palsy-like movements. The highest incidence of first observation of this condition was during the first two years (56.6%). Cases of first onset was observed during age periods 3-6 (3), 7-12 (2), 20's (1) and 30's (2). Three of these respondents did confirm these conditions were related to diagnosed cerebral palsy. Six (6%) reported a change in these

palsy like movements, two during age period 3-6, one each during their preteen period, 20's and 30's. Gilbert (1991) indicated that signs of cerebral palsy and clinical cerebral palsy are neurological manifestations of congenital rubella.

All but four, of the respondents reported being able to walk. The four who did not walk had lived all their lives in long term care facilities for individuals with severe disabilities. One of those individuals was a quadriplegic.

The onset of walking is considerably delayed in this population as indicated by the unusually large proportion commencing walking during the age period 3-6. Most individuals were walking by age six. Six indicated beginning to walk after the age of six, four during their preteen period, one each during their teens and 20's.

Twenty-one (21.2%) reported a deterioration in mobility or walking, twenty-three (23.2%) reported a deterioration in balance and twenty-one (21%) also reported a deterioration in energy level, stamina and endurance. For these three questions, the majority showed first onset after the beginning of their teens, primarily during their 20's, although there were incidences of first onset during their 30's and 40's.

Only one reported a change in the ability to sit, and that individual reported they had a problem with sitting throughout their life. The same individual responded affirmatively to problems with mobility, balance and energy, stamina and endurance all during their life.

Four (4.1%) indicated a deterioration in fine motor skills, one reported this condition during their infancy period, two with first onset reported during their 20's, while one reported unknown first occurrence.

2.2.15 Urogenital Tract

Individuals were given several questions about medical issues concerning their reproductive tract. Table 26 summarizes their responses to these questions .

Table 26: Summary of responses regarding the urogenital tract

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Born With Reproductive Tract Problems n=96	13.5 (13)	-	-	-	-	-	-	-	-
Treatment or Surgery n=98	9.2 (9)	11.1 (1)	33.3 (3)	11.1 (1)	22.2 (2)	22.2 (2)	-	-	-
Incontinence n=99	30.3 (30)	13.3 (4)	46.6 (14)	3.3 (1)	16.7 (5)	6.7 (2)	6.7 (2)	6.7 (2)	-
Other Urogenital Tract Changes n=98	8.2 (8)	11.1 (1)	11.1 (1)	11.1 (1)	22.2 (2)	33.4 (3)	-	-	-

Thirteen respondents (13.5%) reported being born with reproductive tract problems. This appears to be largely a male issue, since twelve of those reporting this problems were males (30% of males) while one

was a female (1.8% of females). Where the problem was specified for males, it was usually related to an undescended testes. Nine of these individuals reported receiving surgery for this problem. As Table 26 indicates, the treatment or surgery was done primarily during the individual's youth. For the one female, the congenital problem was being born without a uterus.

Thirty (30.3%) reported a problem with incontinence, with the majority, not surprisingly, reporting first onset during their infancy period. For most of these individuals, incontinence has been a problem throughout their lives. Eleven reported first onset from the preteen period on, two of those occurring during their 20's and two occurring during their 30's. Seven individuals reported never being toilet trained, all having lived most of their entire lives in a facility for individuals with severe disabilities.

Eight respondents reported incidences of the following "other urogenital changes":

- | | |
|--|--|
| X Ovarian cysts | X Twisted urethra |
| X Vaginal infection | X Urethra implant |
| X Urine retention difficulty | X Absence of right kidney (congenital) |
| X Frequent urogenital tract and kidney infection | X Acid in bladder |

While several of these other changes were first reported during the very early years, two were reported to occur during the preteens and three during their teens.

Gilbert (1991) reported that undescended testes and renal abnormalities are examples of abnormal development which are believed to occur with increased frequency with congenital rubella syndrome. Remington and Klein (1990) report from several studies a frequency rate for undescended testes to be less than 20% in congenital rubella males. Wagman (1988) suggests that the incidence of undescended testes in the general population is one per 200 births (0.5%).

2.2.16 Bowel Function

Respondents were asked to comment about whether there had been a persistent problem with bowel function or any change in bowel function throughout their lives. Responses are summarized in Table 27.

Table 27: Summary of responses regarding bowel function

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Persistent Bowel Problems n=100	31.0 (31)	19.4 (6)	48.4 (15)	9.6 (3)	6.5 (2)	6.5 (2)	9.6 (3)	-	-
Change in Bowel Function n=99	11.1 (11)	-	-	9.1 (1)	18.2 (2)	27.3 (3)	36.3 (4)	9.1 (1)	-

Thirty-one (31%) reported persistent bowel problems, again with the majority, not surprisingly, reporting first occurrence during their infancy period. This problem with toileting was reported to persist throughout life for almost all of those reporting first onset during their infancy. Ten reported first onset following

infancy, three during years 3-6, two 7-12 , two during their teens and three during their 20's. The age of first onset of this problem was unknown for six individuals. To repeat the statement mentioned above regarding continence, seven individuals reported never being toilet trained.

Eleven reported a change in bowel function, five which were reported as a deterioration, four were reported as an improvement while two did not indicate the nature of change. One of those reporting a deterioration indicated having irritable bowel syndrome and two reported a bowel obstruction. Three indicating an improvement reported the use of medication and improved diet as contributing to their improvement.

Responses to the section on bowel function were arranged according to their disability level, that is, whether they indicated they were individuals who are deafblind, deaf or blind. Their responses are summarized in Table 28.

Table 28: Summary of bowel function responses according to disability level

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
n=sample size									
Individuals Who are Deafblind n=70	37.1 (26)	15.4 (4)	61.4 (16)	3.9 (1)	7.7 (2)	3.9 (1)	7.7 (2)	-	-
Individuals Who are Blind n=7	42.9 (3)	-	66.7 (2)	-	-	33.3 (1)	-	-	-
Individuals Who are Deaf n=21	14.3 (3)	66.7 (2)	-	-			33.3 (1)	-	-
Total n=98	32.7 (32)	18.8 (6)	56.3 (18)	3.1 (1)	6.2 (2)	6.2 (2)	9.4 (3)	-	-

It is readily apparent that individuals who are deafblind experience the greatest problems with bowel function.

2.2.17 Gastrointestinal

Questions were posed about eating and whether there were any difficulties being experienced relating to chewing and subsequent digestion. Their responses are summarized in Table 29.

Table 29: Summary of responses regarding the gastrointestinal system

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
n=sample size									
Deterioration in Ability to Chew n=100	16.0 (16)	12.5 (2)	18.7 (3)	-	12.5 (2)	-	37.5 (6)	12.5 (2)	6.3 (1)

Deterioration in Ability to Swallow n=100	8.0 (8)	-	-	-	-	-	-	-	-
Increased Incidence in Gagging n=100	14.0 (14)	-	-	-	-	-	-	-	-
Increased Incidence in Vomiting n=100	11.0 (11)	-	-	-	-	-	-	-	-
Problems with Swallowing, Gagging or Vomiting n=99	25.3 (24)	20.8 (5)	37.5 (9)	4.2 (1)	-	8.3 (2)	12.5 (3)	12.5 (3)	4.2 (1)

Sixteen (16%) reported a deterioration in the ability to chew. Two reported this difficulty in their preteens while nine reported first onset of this condition from age group 20's on.

Twenty-four (25.3%) reported a problem with either swallowing, gagging or vomiting. While most reported first onset in early childhood, nine reported first onset from the teen period on.

One respondent reported an incidence of a distended stomach and another reported a malformed stomach (see Section 2.2.13).

An article in the fall 1993 edition of *National Parent Network Newsletter* mentions gagging and vomiting problems as another reported symptom of late onset manifestations of congenital rubella. O'Donnell (1996) reported incidences of esophageal stricture, gagging, swallowing and cyclic vomiting disorders in the 1991 Helen Keller National Centre study. Since this study in the United States, O'Donnell reported that documented cases of these disorders have been reported in Puerto Rico, Canada, England and Sweden. There is no reference in the literature about these disorders being related to the congenital rubella syndrome.

2.2.18 Endocrine Function

The endocrine glands have the all important role of regulating the body's internal chemistry. Consequently, it was considered crucial to inquire about any medical history around the endocrine system, including thyroid and pancreas function. The responses to questions regarding the endocrine system are summarized in Table 30.

Table 30: Summary of responses regarding endocrine function

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Diagnosed with Thyroid n=100	10.0 (10)	-	-	-	20.0 (2)	50.0 (5)	10.0 (1)	10.0 (1)	10.0 (1)
Hyperthyroid n=100	0	-	-	-	-	-	-	-	-
Hypothyroid n=100	10.0 (10)	-	-	-	20.0 (2)	50.0 (5)	10.0 (1)	10.0 (1)	10.0 (1)

Not Medically Diagnosed With Thyroid But Shows Symptoms n=100	12.0 (12)	25.0 (3)	8.3 (1)	-	16.7 (2)	16.7 (2)	8.3 (1)	25.0 (3)	-
Diagnosed With Diabetes n=99	11.1 (11)	-	9.1 (1)	-	9.1 (1)	36.5 (4)	18.2 (2)	18.2 (2)	9.1 (1)
Not Medically Diagnosed With Diabetes But Shows Symptoms n=100	5.0 (5)	-	20.0 (1)	-	-	20.0 (1)	40.0 (2)	20.0 (1)	-
Diagnosed With Diabetes and Insulin Dependent n=12	58.3 (7)	-	-	-	-	-	-	-	-

Ten (10.0 %) of respondents reported being clinically diagnosed with a thyroid condition, all reporting the condition to be an underactive thyroid or hypothyroidism. Symptoms of this condition, caused by the underproduction of the thyroid hormone thyroxin, are usually drowsy or sluggishness, weight gain and the feeling of being tired and weary with everyday activities (Wagman et al, 1988). The first onset of hypothyroidism was reported by two during preteens, five during their teens, one during their 20's, one individual during their 30's and one during their 40's+. All of the individuals clinically diagnosed with hypothyroidism were on medication. Remington and Klein (1990) report from several studies that thyroid dysfunction has been reported in about 5% of patients with congenital rubella, and the dysfunction manifests itself as hyperthyroidism, hypothyroidism and thyroiditis. Auto-immune mechanisms appear to be responsible for these abnormalities.

In addition, twelve (12%) showed symptoms of a thyroid condition, but were not clinically diagnosed. These respondents did not indicate whether the symptoms were related to hyperthyroidism or hypothyroidism. While one indicated first onset of the undiagnosed condition during the infancy period, two reported first onset during preteen, two during their teens, one during their 20's and three during their 30's.

Regarding diabetes, twelve (12.1%) reported being clinically diagnosed with diabetes mellitus. Diabetes is a disease caused by the insufficient production of the hormone insulin by the pancreas. The first onset of diabetes was observed at the following ages: 0-2 (1); age 9 (1); age 14 (1); 19 (1); 21 (2); 20's (2); 31 (1); 32 (1); 35(1) and 53 (1). Seven of the twelve (58.3%) of those diagnosed with diabetes were currently on medication. One of these individuals has been on insulin since infancy, one beginning at age nine and another two began medication during their teens. The other individuals with diabetes and not on medication have their condition controlled by diet.

In addition to the medical diagnoses, five (5%) reported they suspected, but were not clinically diagnosed with diabetes because they demonstrated several classic symptoms including unusual thirst, rapid weight loss, irritability, frequent urination, weakness, itching, etc. One respondent reported first onset during infancy, one during their teens, two during their 20's and one during their 30's.

The prevalence of clinical diagnosis of diabetes mellitus (11.1%) in this study compares well to other studies of diabetes and congenital rubella. Seven studies reported by Tingle (personal communication, 1998) indicated the prevalence of clinical diabetes to be 2%, 3.5%, 11%, 11%, 12%, 12.4% and 12.5%. Menser et al (1982) evaluated a group of adult survivors from the 1942 Australia epidemic and reported that 40% had diabetes mellitus. Like thyroid dysfunction, auto-immune mechanisms appear to be

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

responsible for an abnormality of the pancreas.

It is interesting to note that three individuals had a coexistence of clinical thyroid dysfunction and diabetes mellitus. This has also been reported in Remington and Klein (1990).

2.2.19 Heart and Circulatory System

Respondents were asked to document their medical history related to the heart and circulatory system. Their responses are summarized in Table 31.

Table 31: Summary of responses regarding the heart and circulatory system

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Diagnosed With a Heart Defect n=99	65.7 (65)	3.1 (2)	95.4 (62)	-	-	1.5 (1)	-	-	-
Hole in Heart or Patent Ductus Arteriosus n=94	34.0 (32)	-	-	-	-	-	-	-	-
Pulmonary Artery Stenosis n=93	17.2 (16)	-	-	-	-	-	-	-	-
Ventricle Septal Defect n=93	9.7 (9)	-	-	-	-	-	-	-	-
Renal Artery Stenosis n=93	2.2 (2)	-	-	-	-	-	-	-	-
Diagnosed With High Blood Pressure n=97	7.2 (7)	14.3 (1)	14.3 (1)	-	-	14.3 (1)	28.5 (2)	14.3 (1)	14.3 (1)
Other Heart or Circulatory Problems Diagnosed n=97	19.6 (19)	5.3 (1)	73.6 (14)	10.5 (2)	-	5.3 (1)	5.3 (1)	-	-
Surgery For Any Problems Noted n=94	24.5 (23)	4.3 (1)	74.0 (17)	17.4 (4)	-	4.3 (1)	-	-	-

Sixty-five (65.7%) reported being diagnosed with various heart defects. The four conditions specified in the questionnaire were reported as follows:

- Patent Ductus Arteriosus or hole in the heart 32
- Pulmonary Artery Stenosis 16
- Ventricle Septal Defect 9
- Renal Artery Stenosis 2

Ten individuals reported multiple conditions as follows: **three** with a hole in heart and pulmonary artery stenosis; **one** with a hole in heart, pulmonary artery stenosis and ventricle septal defect; **four** with a hole in heart and ventral septal defect; **one** with pulmonary artery stenosis and renal artery stenosis and **one** reported pulmonary artery stenosis and ventricle septal defect.

Twenty-three of those diagnosed with a heart defect (24.5%) had surgery. Most surgery was performed during the child's early life.

Most (95.4%) reported these conditions as congenital or reported during the first two years of life. The one not reporting the conditions as congenital, reported first onset during their teens. The one incidence was an "other condition" and was described as a heart murmur detected at age 17. It is likely that this respondent had this condition from birth. Two respondents did not attach an age of onset, but it is presumed the condition was congenital.

Seven (7.2%) reported high blood pressure. One of these reported the condition since birth, one reported first onset during their teens, two reported first onset during their 20's and one each during their 30' and 40's. One did not report the age of first onset of high blood pressure.

Nineteen (19.6%) reported other "Circulatory Problems." The incidence of these other problems are as follows:

- Heart murmur 11 (8 reported; 1 age 3-6; 1-teens; 1-20's)
- Heart rhythm 1 (unknown age)
- Passage too small for valve 1 (congenital)
- Valve outside of heart 1 (congenital)
- Hypertrophy non-destructive cardiomyopathy 1 (congenital)
- Sinus tachy-cardia 1 (congenital)
- Clockwise rotation of heart 1 (congenital)
- Severe nose bleeds 1 (age 3-6)
- Atrial ventricle valve (DAP) 1 (congenital)
- Narrow valve 1 (congenital)
- Coarctating of aorta 1 (congenital)

As this list indicates, many of these conditions are congenital and reflect heart defects the questionnaire did not address.

Cherry (1987) reports, from various studies, the frequency of congenital cardiovascular irregularities in congenital rubella individuals as follows: patent ductus arteriosus (30%); pulmonary arterial hypoplasia, supravalvular stenosis, vulvular stenosis and peripheral branch stenosis (25%); and, ventricle and atrial septal defect (2-5%).

2.2.20 Lung Condition

Several questions were posed to gather information about lung congestion and history of pneumonia. Summary of responses is presented in Table 32.

Table 32: Summary of responses regarding lung condition

Question	Percent Yes	Percent First Responding by Age Group								
		n=sample size	(response size)	?	0-2	3-6	7-12	13-21	22-30	31-40
Symptoms of Lung Congestion n=99	5.1 (5)		20.0 (1)	40.0 (2)	20.0 (1)	-	-	-	20.0 (1)	-
History of Pneumonia n=98	17.4 (17)		11.8 (2)	52.9 (9)	5.9 (1)	23.5 (4)	-	-	5.9 (1)	-

Five (5.1%) of respondents reported symptoms of lung congestion. Two reported first onset during infancy, one during period 3-6 years and another during their 30's. One did not report the age of first onset.

Seventeen (17.4%) of respondents reported a history of pneumonia. This condition appears to be youth related, with only one first incidence reported after the preteen period. Gilbert (1991) reported that pneumonitis was another early manifestation of congenital rubella due to a continuing viral infection. Cherry (1987) reported from the literature an incidence rate of life threatening interstitial pneumonitis as 5-10%.

2.2.21 Allergy/Asthma

Respondents were questioned about their history of allergies and asthma. A summary of responses is presented in Table 33.

Table 33: Summary of responses regarding allergy/asthma

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Experienced Allergy Problems n=99	37.4% (37)	21.6 (8)	21.6 (8)	16.2 (6)	10.9 (4)	8.1 (3)	16.2 (6)	5.4 (2)	
Experienced Asthma Symptoms n=95	7.4% (7)	14.3 (1)	14.3 (1)	-	14.3 (1)	14.3 (1)	42.8 (3)	-	-

Thirty-seven (37.4%) of respondents reported allergy symptoms. First onset of allergy symptoms generally ranged across the age spectrum.

Respondents were asked to categorize the type of allergies, whether food, drug or environmental. The following lists the responses:

- drug (examples: penicillin and sleeping medication) 11
- environmental (hay fever) 9
- food (example: eggs) 2
- food and drug 2
- food and environmental 5
- drug and environmental 5
- unsure 3
- Total 37**

Seven (7.4%) of respondents reported symptoms of asthma, with first onset of asthma symptoms noted by late childhood through to the 20's.

2.2.22 Osteoporosis

Osteoporosis is a metabolic disorder marked by porousness and fragility of the bones and is a condition usually associated with old age. While its exact cause is unknown, contributing factors are believed to be improper metabolism of calcium and phosphorus beginning at youth, deficiency of sex hormones following menopause, and atrophy due to disuse and lack of stress and strain on the bones (Wagman et al, 1988).

Respondents were asked whether there was any reported incidence of osteoporosis and responses are summarized in table 34.

Table 34: Summary of responses regarding osteoporosis

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Diagnosed With Osteoporosis n=99	7.1% (7)	-	-	-	14.3 (1)	-	28.6 (2)	57.1 (4)	-

Seven (7.1%) of respondents reported clinical osteoporosis, with first onset reported as follows: one during their preteens, two during their 20's and four during their 30's. While osteoporosis is often recognized as a female issue, two of the seven individuals with osteoporosis, were males. Statistics on this condition indicate that usually males represent less than 20% of those diagnosed with osteoporosis (Bonnick, 1994).

It is believed that the incidence of this condition would be much higher if more of the adult individuals were tested. A lack of physical activity combined with a higher than normal rate of symptoms of early menopause, (12.3% of females, Table 21) make this population at high risk to osteoporosis.

2.2.23 Cancer

Respondents were asked whether there was a history of cancer in the population. Respondents were asked whether there was any reported incidence of cancer and responses are summarized in Table 35.

Table 35: Summary of responses regarding cancer

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Diagnosed With Cancer n=99	2% (2)	-	-	-	-	-	50.0 (1)	50.0 (1)	-

Two individuals (2%) reported cancer, one reporting the onset during their 20's and one during their 30's. One reported form of cancer was basal cell carcinoma while the other was testicular cancer. As a matter of interest, the one individual with testicular cancer also reported his testes had never descended, despite surgery.

2.2.24 History of Hospitalizations

Individuals were asked to provide an insight into their history of hospitalization and to summarize the treatments they received. Table 36 summarizes the response to the question.

Table 36: Summary of responses regarding history of hospitalizations

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Individual Hospitalized n=100	94% (94)	2.1 (2)	73.4 (69)	6.4 (6)	5.3 (5)	6.4 (6)	4.3 (4)	2.1 (2)	-

The responses confirm that congenital rubella individuals have a significant history with the medical community, especially during their youth. Ninety-four (94%) of respondents reported being hospitalized, and for most, this involved multiple visits. The incidence of multiple visits is not shown here. Sixty-nine (73.4%) reported undergoing hospital procedures during their infancy, with few showing initial hospitalizations later in life.

Respondents provided details on their hospitalization treatments which are summarized as follows:

• Cataracts	54	• Hernia	6
• Detached retina	2	• Intestinal problems (ulcers, gastroenteritis, reflux, gas)	11
• Glaucoma	7	• Appendicitis	2
• Other eye surgery	17	• Laparotomy (bowel)	1
• Lens implant	1	• Problems with vomiting and diarrhea	3
• Eye examinations	1	• Acute maxillary sinusitis	
• Ear surgery	2	• Bowel obstruction	2
• Ear tubes (myringotomy)	1	• Psychiatric examination/ Treatment	8
• Ear infections	1	• Gall Bladder	2
• Ear examinations	1	• Infections	2
• Heart surgery	24	• Testing for rubella in spine	1
• Anemia	1	• Surgery on legs/hips	3
• Diabetes	2	• Jaw surgery	1
• Dental work (non-specified)	13	• Shoulder surgery	1
• Pneumonia	7	• Nose surgery	1
• Bronchitis	1	• Head surgery (accident)	1
• Influenza	2	• Excision of femoral head	1
• Breathing problem	1	• Bilateral tendon addiction	1
• Allergies	1	• Veins stripped	1
• Tonsils and Adenoids	5	• Back surgery	4
• Pharyngitis	1	• Planters wart	1
• Esophageal dilation	1	• Thyroid	1
• Seizures	3	• Hospitalized as an infant re: rubella	6
• Fractures	6	• General evaluation	1
• Testes	6		
• Lupus	1		
• Removal of cyst	1		
• Urinary problems	4		

2.2.25 Record of Medications

Individuals were asked to provide a record of the use of various medications for specific medical

purposes. The summary of responses is found in Table 37.

Table 37: Summary of responses regarding use of medications

Medication n=sample size	Percent Yes (response size)	Medication n=sample size	Percent Yes (response size)
Anti-depressives n=97	34.0 (33)	High Blood Pressure n=97	6.2 (6)
Anxiety n=94	28.7 (27)	Osteoporosis n=97	8.3 (8)
Hyperactivity n=95	16.8 (10)	Birth Control n=55	47.3 (26)
Seizures n=100	16.0 (16)	Menstrual Suppression n=56	33.9 (15)
Lethargy n=95	4.2 (4)	Allergies n=94	16.0 (15)
Thyroid n=98	9.2 (9)	Asthma n=95	3.2 (3)
Diabetes n=98	8.2 (8)	Surgery n=97	68.0 (66)
Glaucoma n=98	21.4 (21)	Other n=93	36.6 (34)
Migraines n=96	10.4 (10)		

Medications were used for other medical purposes as follows:

Dermatological	Stool softening	Sleeping
Aggression	Urination	Gastrointestinal problems
Lupus	Eye infections	Behaviour
Constipation	Menstrual Cycle Regulation	Dry nasal passage
Self-injury	Psychological	Rheumatism
Infection	Cough suppression	Bed wetting
Pain		

2.2.26 Family Dynamics

Individuals were asked several questions about their family history. Their responses are summarized in Table 38.

Table 38: Summary of responses regarding family dynamics

Question n=sample size	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Family Intact n=94	63.8 (60)								
Change in Family Unit n=95	46.3 (44)	15.9 (7)	34.0 (15)	11.4 (5)	9.1 (4)	13.6 (6)	11.4 (5)	-	4.6 (2)
Placed in an Institution n=100	36.0 (36)	-	11.1 (4)	47.2 (17)	33.3 (12)	2.8 (1)	5.6 (2)	-	-

Siblings n=96	89.6 (86)								
Death in the Family n=90	34.4 (31)	19.4 (6)	6.5 (2)	9.7 (3)	12.9 (4)	29.0 (9)	16.0 (5)	-	6.5 (2)

Sixty (63.8%) of respondents reported their natural family remained intact during their lives.

Forty-four (46.3%) of respondents reported a change in their birth family unit, with a significant amount of the change occurring during the individual's early childhood. For example, fifteen (34%) reported the first change in family unit occurred during their infancy period. Twelve individuals reported being placed in foster care or being adopted.

Thirty-six (36%) of respondents reported being placed in an institutional setting other than the original family home sometime during their life. For the purpose of this discussion, institutional setting does not include residential schools where an individual may have spent part of his year. Twenty-one of them (58.3%) were placed in another facility by the age of six, while thirty-three (91.6%) were placed before their teen years.

Thirty-one (34.4%) of respondents reported a death in the family. This question was asked to gather background data surrounding possible stresses in an individual's life that could manifest itself in some change of behaviour. Thirteen (41.9%) acknowledged a death in the family while they were in their preteen and teen period, a period during which other changes manifest themselves in this population.

2.2.27 Educational Activity and Placement

Individuals were asked to indicate their educational history and Table 39 summarizes their responses.

Table 39: Summary of responses regarding educational activity and placement

n=sample size	(response size)	n=sample size	(response size)
Attended a Pre-School Program n=94	47.9 (45)	Attended School for Deaf n=100	25.0 (25)
Attended Regular Classes in Community Public School n=97	21.7 (21)	Attended School for Blind n=99	3.0 (3)
Attended Special Classes in Community Public School n=97	19.6 (19)	Attended a Residential School n=99	56.6 (56)
Attended Home Schooling n=97	6.2 (6)	Attended Another Type of Educational Program n=100	25.0 (25)
Attended School for Deafblind n=99	43.4 (43)	Attended Post-Secondary School n=98	13.3 (13)

Respondents indicated the following educational history:

- Forty-five (47.9%) had attended a pre-school program.

- Twenty-one (21.7%) had attended regular classes in a community public school.
- Nineteen (19.6%) had attended special classes in a community public school.
- Six (6.2%) had attended home based schooling.
- Forty-three (43.4%) had attended a school for individuals who are deafblind.
- Twenty-five (25%) had attended a school for individuals who are deaf.
- Three (3%) had attended a school for individuals who are blind.
- Fifty-six (56.6%) had attended a residential school for individuals who deaf, deafblind or blind.
- Twenty-five (25%) had attended another type of educational program (unspecified).
- Thirteen (13.3%) had attended a post-secondary school.

2.2.28 Living Situation

Individuals were asked to indicate their past or current living situation and their responses are summarized in Table 40.

Table 40: Summary of responses regarding living situation

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Lives in Family Home n=100	28.0 (28)	-	-	7.1 (2)	7.1 (2)	39.4 (11)	21.4 (6)	21.4 (6)	3.6 (1)
Lives in Apartment in Family Home n=99	3.0 (3)	-	-	-	-	66.7 (2)	33.3 (1)	-	-
Lives Independently in Community n=99	13.1 (13)	7.7 (1)	-	-	-	38.5 (5)	38.5 (5)	15.3 (2)	-
Lives in Supported Independent Living Setting n=100	30.3 (30)	-	-	2.9 (1)	6.0 (2)	44.1 (12)	44.1 (15)	-	-
Lives in Another Facility (other than a family setting) n=100	30.0 (30)	-	13.2 (4)	46.7 (14)	26.7 (8)	6.7 (2)	6.7 (2)	-	-
Lives in Nursing Home n=99	3.0 (3)	-	-	-	33.3 (1)	-	33.3 (1)	-	33.4 (1)
Lives With Spouse n=99	6.1 (6)	33.3 (2)	-	-	-	66.7 (4)			

- Twenty-eight (28%) of respondents currently report living at home sometime during the year, either with their natural or foster family. Of these twenty-eight, eleven attend school while the

- remaining seventeen no longer attend school.
- Three respondents (3%) report currently living in an apartment in their family home.
- Thirteen (13.1%) report currently living independently in their community.
- Thirty (30%) report currently living in a supported independent living setting (with one on one Intervention services) established and designed specifically for individuals who are deafblind. Of these thirty individuals, two live in a supported setting while they continue to attend school for individuals who are deafblind, two live in their own apartments while the remaining twenty-six live in group settings.
- Thirty (30%) report having lived for a period of their lives in another type of facility...generally an institutional setting, including a group home. Most of these individuals (86.6%) entered an institutional setting prior to their teenage years, with eighteen (60%) leaving the family home before the age of seven. Currently fifteen live in an institution and eleven live in a group home (without one on one Intervention).
- Three (3%) report living in a nursing home.
- Six (6.1%) report living with a spouse.

Some movement of individuals has occurred among living settings outside the family or foster home which accounts for the difference in the total of the sample sizes.

2.2.29 Working Situation

Individuals were questioned about their employment and volunteer experience. The summary of their responses is presented in Table 41.

Table 41: Summary of responses regarding working situation

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Works in Competitive Employment n=100	20.0 (20)	-	-	-	-	50.0 (10)	50.0 (10)	-	-
Works in Sheltered Workshop n=99	12.1 (12)	16.7 (2)	-	-	-	33.3 (4)	50.0 (6)	-	-
Works As a Volunteer n=99	29.3 (29)	13.8 (4)	-	-	6.9 (2)	51.8 (15)	24.1 (7)	3.4 (1)	-
Does Not Work n=100	54.0 (54)	-	-	-	-	-	-	-	-
Change in the Ability to Work or Volunteer n=99	15.2 (15)	6.6 (1)	-	-	-	20.0 (3)	60.0 (9)	6.7 (1)	6.7 (1)

- Twenty (20%) of respondents report working at one time in competitive employment; fourteen currently.

- Twelve (12.1%) of respondents report currently or recently working in a sheltered workshop.
- Fifty-four (54%) report they are currently not working or volunteering at all.
- Twenty-nine (29.3%) of respondents report currently or recently working as a volunteer.
- Fifteen (15.2%) of respondents report a change in their ability to work or volunteer.

Of the eleven that did make comments, three indicated an **improvement** for such reasons as having increased skills, increased use of language skills and possessing improved concentration abilities. The other eleven indicated a **deterioration** with the following comments: becoming frustrated and angry, less care paid to detail, becoming slow, illness, deterioration in hearing, hearing loss affected work ability and motivation to work, disabled after head injury due to accident, deteriorating physical ability, behaviour and health.

2.2.30 Communication

A series of questions were asked about communication to determine the status of individual's access to communication, Intervention and their use of various communication modalities. Their responses are summarized in Table 42.

Table 42a: Summary of responses regarding communication

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Lived in Environment with Communication n=99	83.8 (83)	1.2 (1)	32.5 (27)	33.8 (28)	22.9 (19)	4.8 (4)	2.4 (2)	1.2 (1)	1.2 (1)
Received Intervention n=99	60.6 (60)	1.7 (1)	13.3 (8)	35.0 (21)	33.4 (20)	8.3 (5)	5.0 (3)	-	3.3 (2)

Table 42b: Summary of responses regarding communication (without age of first reporting)

Question n=sample size	Percent Yes (response size)	Question n=sample size	Percent Yes (response size)
Uses Concrete/ Object Cues n=87	51.7 (45)	Uses One Handed Finger Spelling n=96	22.9 (22)
Uses Speech n=95	31.6 (30)	Uses Two Handed Finger Spelling n=96	13.5 (13)
Uses Vocalization n=95	61.1 (58)	Uses Adapted Sign Language n=98	58.2 (57)
Uses Sign Language n=98	67.3 (66)	Uses Braille n=98	4.1 (4)
Uses Written Language n=97	42.3 (41)	Uses Other Communication Devices n=98	21.4 (21)

- Eighty-three (83.8%) of respondents report having lived (at one time) in an environment with communication (pertinent to their disability). While most individuals (32.5% ages 0-2 plus 33.8% ages 3-6) reported receiving pertinent communication early in their lives (up to age six), many (19) received their initial communication during their preteen years (22.9%), four not until their teens, two

during their 20's, one during their 30's and one during their 40's.

- Sixty (60.6%) of respondents report currently receiving Intervention. Almost fifty percent (29 individuals) report onset of Intervention early in their lives (by the age of six), while an additional 20 (33.4%) report Intervention first beginning during the preteen period. This latter percentage reflects Intervention not beginning until individuals first attended specialized schools for individuals who are deafblind. For ten (16.6%) of respondents, Intervention did not commence until their teenage period and later. The reasons for this late commencement of Intervention for six of them, was the fact that they had lived in an institutional setting without any access to this critical process. For the others, two were originally in programs for deafness before attending a specialized school for individuals who are deafblind, one began receiving Intervention following a deterioration in their vision, and another did not have Intervention available in the community until age 15.
- Thirty (31.6%) of respondents currently use speech as a mode of communication.
- Fifty-eight (61.1%) of respondents currently use some form of vocalization.
- Sixty-six (67.3%) of respondents currently use sign language.
- Fifty-seven (58.2%) of respondents currently use an adapted form of sign language.
- Twenty-two (22.9%) of respondents currently use one handed finger spelling.
- Thirteen (13.5%) of respondents currently use two handed finger spelling.
- Forty-one (42.3%) of respondents currently use written language.
- Four (4.1%) of respondents currently use braille.
- Twenty-one (21.4%) of respondents currently use other, unspecified communication devices.

2.2.31 Support Resources for Individuals who are Deafblind

Individuals who were deafblind were asked to respond to questions pertinent to Support Resources or Intervention. A summary of their responses is found in Table 43.

Table 43: Summary of responses regarding support resources for deafblind

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Receives Intervention at Work n=69	33.3 (23)	5.0 (1)	-	-	-	47.8 (11)	47.8 (11)	-	-
Receives Intervention at School n=70	64.3 (45)	4.4 (2)	-	57.8 (26)	28.9 (13)	8.9 (4)	-	-	-
Receives Intervention in the Community n=69	65.2 (45)	17.8 (8)	2.2 (1)	17.8 (8)	20.0 (9)	13.4 (6)	22.2 (10)	4.4 (2)	2.2 (1)
Receives Intervention at Home on Weekends n=70	54.3 (38)	13.2 (5)	2.6 (1)	18.4 (7)	13.2 (5)	15.8 (6)	34.2 (13)	2.6 (1)	-

Receives Intervention at Home During Evenings n=70	47.1 (33)	9.1 (3)	3.0 (1)	18.2 (6)	12.1 (4)	12.1 (4)	45.5 (15)	-	-
Receives Intervention at Home During Summer Periods n=70	65.7 (46)	15.2 (7)	2.2 (1)	28.3 (13)	15.2 (7)	17.4 (8)	19.5 (9)	2.2 (1)	-

- Twenty-three (33.3%) of respondents reported receiving Intervention while they were working.
- Forty-five (64.3%) of respondents reported that they were either currently receiving or had received Intervention while at school.
- Forty-five (65.2%) of respondents report currently receiving Intervention in the community.
- Thirty-eight (54.3%) of respondents reported receiving Intervention at home on weekends.
- Thirty-three (47.1%) of respondents reported receiving Intervention at home during evenings.
- Forty-six (65.7%) reported receiving Intervention at home during summer periods.

2.2.32 Support Resources for Individuals who are Deaf

Individuals who were deaf were asked to respond to questions pertinent to Interpretive Services. A summary of their responses is found in Table 44.

Table 44: Summary of responses regarding support resources for the deaf

Question	Percent Yes (response size)	Percent First Responding by Age Group							
		?	0-2	3-6	7-12	13-21	22-30	31-40	40+
Receives Interpretive Services at Work n=20	25.0 (5)	40.0 (2)	-	-	-	-	60.0 (3)	-	-
Receives Interpretive Services at School n=20	90.0 (18)	22.2 (4)	-	33.3 (6)	33.3 (6)	11.2 (2)	-	-	-
Receives Interpretive Services in the Community n=20	70.0 (14)	43.0 (6)	14.3 (2)	7.1 (1)	21.4 (3)	7.1 (1)	7.1 (1)	-	-
Receives Interpretive Services at Home on Weekends n=20	25.0 (5)	60.0 (3)	20.0 (1)	-	20.0 (1)	-	-	-	-
Receives Interpretive Services at Home During Evenings n=20	25.0 (5)	60.0 (3)	20.0 (1)	-	20.0 (1)	-	-	-	-
Receives Interpretive Services at Home during Summer Periods n=20	35.0 (7)	57.1 (4)	14.3 (1)	14.3 (1)	14.3 (1)	-	-	-	-

- Five (25%) report receiving interpretive services at work.
- Eighteen (90%) reported receiving interpretive services at school.
- Fourteen (70%) reported receiving interpretive services in the community.
- Five (25%) reported receiving interpretive services at home on weekends.
- Five (25%) reported receiving interpretive services at home during evenings.
- Seven (35%) reported receiving interpretive services at home during summer periods.

3.0 CONCLUSIONS AND RECOMMENDATIONS

It is easy to be overwhelmed by the magnitude of issues that beset many of the individuals whose mothers contract rubella while pregnant with these babies. To give the issue some perspective, let us

describe the life of a hypothetical male individual born in a small town in northern Alberta.

3.1 About John

John was born prematurely in early March, 1967, weighing about 5 pounds. His single teenaged mother knew she had contracted a mild form of measles known as German measles, but was assured by her doctor that things would be fine with her baby. The subsequent results did not fit the doctor's early prognosis. John was first diagnosed with a minor (and not considered threatening) inflammation of the brain called encephalitis. There was also a problem with his breathing, caused by a mild case of pneumonia, and he was covered with a purple rash. For several days he had problems with diarrhea and vomiting. John was hospitalized for several weeks due to his prematurity and the other symptoms. During this initial period in the hospital he was diagnosed with congenital cataracts and microphthalmia (small eyes), a congenital heart condition and undescended testes. Physicians also observed that his head circumference was below normal, and he was recognized as being borderline microcephalic, and believed at risk to profound mental retardation.

John was released to go home with his mother after several weeks recuperating from his initial problems with pneumonia, rash, diarrhea and vomiting. For several months following his birth, John was quite inactive or lethargic. Fortunately by about six months the initial transitory effects of the encephalitis had subsided and he was becoming a more active child.

During John's first year, he returned to hospital several times for procedures which included a needling procedure to remove his cataracts, an operation to repair his congenital heart defect (hole in the heart or patent ductus arteriosus and pulmonary arterial stenosis), and for a procedure to assist with the descent of his testes.

During a hospital visit the family physician advised John's mother that the child would be severely physically and mentally disabled and that the best course of action for her was to give up her child to a foster family or place John in an institution. It was not long before the young single mother, without any support, chose to give up her child to a foster family. The foster family took John into their home and provided nurturing for the child.

Well into John's second year, the foster family became concerned with his lack of awareness of sound, which eventually resulted in the diagnosis of deafness. This was a setback to the foster family who were struggling with the realization that the child was severely visually impaired and possibly mentally delayed. Despite his difficulties, John appeared healthy and began to function reasonably well. After age 3, the foster parents legally adopted John and raised him as their own!

By age four there was concern about schooling and also about John's lack of good communication. Through their connections, the family discovered that John could receive the appropriate education for his disability, but that meant he would have to attend residential school...in Brantford, Ontario. The family wondered whether they would be forced to move to Ontario? They learned that the child could travel by air to Ontario for the fall school period, return home for the Christmas holidays; return again to Ontario and stay until June when he would come home again for the ten week summer period. They would not have to move!

The long periods away from home were difficult for John and for the parents as well. At school he thrived, developed good communication and received an excellent education designed for his deafblindness disability. While John was profoundly deaf, his residual eyesight (which was improved by thick glasses) allowed him to develop remarkably well. The early profound mental retardation label proved to be inappropriate. However, being deafblind severely restricted his intellectual development due to the lack of wide ranging experiences that typically leads to an enriched language.

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

Then, one day, at age eleven John developed a mild eye infection and swiftly his residual vision disappeared, resulting in a quick regression, with most of John's major gains put on hold. He could no longer see the communication signs upon which he depended. To make matters worse, his sleep pattern, which for several years had become regular, had now severely deteriorated. Some nights he could not sleep until early into the morning, and often, not at all. Different medications were prescribed to regulate his sleep. Together, the total vision loss and sleep difficulties caused much agitation and frustration, which severely interfered with his continued classroom and communication development.

Just before his thirteenth birthday, John experienced the first of numerous epileptic seizures. Fortunately the frequency and severity of his seizures were reduced through medication. This medical condition was followed shortly by the diagnosis of diabetes. John's dependence on a wide range of medication was expanded, and now included medication for sleeping, seizures, and insulin for his diabetes. Because of his increased agitation and frustration, a psychotropic medication was also prescribed to further complicate his chemical regime.

John was a small individual. He started out prematurely, was tiny in his early years and continued to be small throughout his life. By his late teens John weighed about 115 pounds and was 5 foot 3 inches.

John stayed in Brantford and did the semi-annual traveling between Ontario and Alberta until age 19, after which he went home permanently, to his adopted parents. He stayed at home for five years without any services until an independent facility was finally located in a larger town 200 km from his home. Fortunately, John's adult living situation was now resolved and his parents were relieved since they could not satisfy John's adult living needs in their home in the small community in northern Alberta.

John was doing well until about age 28. His ability to masturbate began to deteriorate. Then he began to become quite aggressive with his care givers, he demonstrated extreme frustration, and began breaking furniture. This behaviour fluctuated week to week and required continuous consultation with physicians to determine the reasons for change in behaviour. They experimented with medications for agitation, for depression; they adjusted levels of the other medications...all in vain. The behaviours increased one day; subsided another, without any resolve. There continued to be periodic testing for any indications of a neurological change, a physiological change, or possibly a physical change or related to pain.

Today, at age 31, John is living in a comfortable independent facility for individuals who are deafblind, receiving excellent care, including Intervention services. But John's health is at the crossroads!

3.2 Mechanisms Involved with Manifestations of Congenital Rubella

While John is described as a hypothetical individual with deafblindness living in Canada, he is an amalgam of many individuals who suffer the continuous ravages of congenital rubella, 100 of whom were reported in this study.

Many of the individuals in this study, like the hypothetical John, demonstrate a number of symptoms of congenital rubella and in a similar chronology of occurrence. The first symptoms, like encephalitis, pneumonia, the rash, digestive problems, to name a few, are short term transitory symptoms, the effects of congenital rubella. They generally leave no lasting conditions. The other manifestations, mentioned in connection with John, some identified early and some developed later are the subject of this study.

The early manifestations such as the sensorineural problems (vision and hearing), possible neurological difficulties, small birth size and cardiac defects 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,

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

are not clearly known (Remington and Klein, 1990).

Until recently, it was thought that the early manifestations (visual loss, hearing loss, cardiac defects) were the final results of the embryo being victimized in its early stages by rubella. Believing this, it was then up to families and professionals to ensure that developing good life skills, communication and educational programming was enough to ensure a reasonable life. It came as an unwelcome surprise to families, professionals and medical experts that there would be later developing manifestations of the congenital rubella. Such later manifestations, further vision loss, additional hearing loss, epileptic seizures, an onset of diabetes, and thyroid failure, were indicators that the ravages of the rubella virus had not subsided.

It is still unclear how this is happening but there are several opinions that include auto-immune effects, persistence of the virus or re-activation of the virus. The impact of rubella on cell growth and organization during early organ and neural development, as mentioned previously, could possibly account for deteriorating problems as the individual grows older. Wolinsky (1990) believes that late endocrine dysfunction (leading to diabetes and thyroid problems) is related to viral-induced autoimmune mechanisms, suggesting some persisting rubella viral activities. On another front, Wolinsky (1990) also suggests that if not the complete rubella virus, then continued or renewed generation of rubella-virus-specific gene products, has an influence in progressive rubella parencephalitis. This condition was not observed in the Canadian study, but is mentioned as another opinion about how late manifestations of congenital rubella may be occurring.

3.3 Implications of the Late Manifestations Study

This study was undertaken to learn the extent of the harm that the rubella virus has caused to Canadians throughout their lives, and to provide this information as an important resource to others involved with individuals who have the congenital rubella syndrome. This includes the individuals themselves, their families, care givers, professionals and medical personnel.

While not meant to be a scare tactic, everyone involved with individuals with congenital rubella should appreciate the potential health risks connected with this syndrome. Perhaps some individuals have been spared many of the serious health issues in their early lives that others have undergone. This study is also intended to make those congenital rubella individuals aware of the potential health issues that could occur, and urge them to remain vigilant of potential health problems. For example, if diabetes is 100 to 200 times more prevalent (Remington and Klein, 1990) 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.

Another point is that care givers may be noting previously unobserved changes in the individual in their care. Since many individuals with deafblindness have difficulties with communication, often they will communicate a change in their health through a behavioural change. The new behaviour may not suggest precisely what the health issue is, but should alert the family or staff that something is wrong. This should lead care givers to undertake a detailed health surveillance plan. The individual may be undergoing severe pain, there may be a mental health problem, the chemistry of their medication may be confounded, or their natural endocrine hormone levels may be out of balance. Any of these could lead to a behavioural change that must be dealt with. For example, van Dijk (1991) reported a high relationship between head banging and unbalanced glucose levels. He also reported an increase of aggressive behaviour with a "high functioning" rubella child several days before her menstrual period.

3.4 Study Evaluation

This study was very ambitious in its attempt to obtain a life history profile of those who volunteered to

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

participate in this study. What was surprising was the high level of interest and support by family members, professionals, care givers and individuals who are deafblind, to participate in this study. The sample size of one hundred was an astonishing response and represents a 50% return rate based on the number of individuals or their families who were contacted and provided with a survey package. This sample size is comparable to the Helen Keller National Centre Research study (O'Donnell, 1996) 1989-1991, during which time eighty-eight mail-in questionnaires were analyzed in addition to thirty-nine telephone interviews.

The complexity of the questionnaire perhaps was not as much a challenge to the participants as it was to the design team, and those left to analyze and interpret the results. Those who participated are to be congratulated for the effort in completing the questionnaire. The task for family members was often not as much a chore as it was for care givers or professionals, who painstakingly pored through thick files to provide past details that were requested.

One could write many comments about the various inadequacies of the survey. On one hand it can be said that it was too complex and had too many questions, while on the other hand during the analysis phase it was realized that perhaps some the questions were unclear or in some cases, did not ask for enough detail. In many instances there was not enough supporting information accompanying the answers to particular questions. This was partly the fault of the survey design, where in some cases, the question was not specific enough, or where more information would have been useful. In numerous instances, however, answers could not be provided to questions about an individual's early life, because the particular information was not on file, the relevant family member may not have been alive or they could not be contacted for pertinent information.

Several specific weaknesses of the survey will be highlighted, although others may come to light as the study is reviewed. One weakness recognized is that there is a lack of emphasis or appropriate questions about the individual's actual current functional status of their vision, hearing, communication, motor activity, etc. Another weakness recognized was in not obtaining more precise indications of a change in hearing, change in vision, change in motor activities, neurological changes, etc. These are important indicators of later manifestations of congenital rubella, and in some instances the answers to the question are not clear enough. While one could be excused by the fact this was not pure scientific research, it is important to acknowledge weaknesses and point out where improvements could be made by others contemplating surveys of a similar nature.

Finally, it is believed that some (but not all) of the inadequacies of the survey could have been overcome through a better means of obtaining the information from family members, individuals or care givers. While the mail survey is an inexpensive means of collecting the information it is also an impersonal method. It is believed that more detailed and precise results would have been obtained through person to person contact. In many instances the respondents were telephoned to clarify answers and complete unanswered questions on the questionnaires. However, even with follow-up telephone interviews, complete answers were not always possible due to inadequacies with some individuals' personal files.

3.5 Future Studies

Surveys such as this, often create as many questions as they provide answers. One area that is very intriguing is the one on birth mothers' health issues. Even by just scratching the surface, since admittedly the questions were not precise enough (arthritis and early menopause), there seems to be some connection between congenital rubella and birth mothers' health issues; this definitely needs further research.

Another area, and a serious one, is the prevalence of arthritis among the population of young adult individuals with congenital rubella. Although we did not ask the question about arthritis, no respondent

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

mentioned there was a diagnosed arthritic condition. Several did mention pain in legs and knees. If the prevalence of arthritis is as high in birth mothers as this survey has indicated (over 40%) and if the cause-effect relationship is as Dr. Tingle (personal communication, 1998) suggested, then it would not be surprising if there was a high rate of arthritic condition within this population of individuals sampled. This is worth investigating, since some of the late onset behavioural difficulties many of these individuals are experiencing could be connected with elevated pain!

A final area is with respect to learning the actual biological mechanisms responsible for late onset manifestations of congenital rubella. Researchers postulate possible reasons for what is happening, but the intricate biochemical mechanisms needs to be better understood. If so, this could help improve the quality of lives of many of these individuals with congenital rubella. Unraveling this mystery could also help improve the lives of individuals who are disabled by cytomegalovirus (CMV), another rather benign virus which is disabling many young individuals today.

3.6 Significance of Project Results for Policy Formulation and Program Development

The results of this project confirm the need for communications; the need to inform and educate families and care givers of individuals with congenital rubella, as well as the medical profession, about the long term effects of rubella. While, fortunately the population of individuals with congenital rubella in Canada is small (probably less than 2,500), the challenge to contact and inform them is large.

Several approaches will be immediately implemented. This first will involve distributing the results of this study to those many individuals, facilities and organizations who participated in this project. Hopefully this network of people will do their part to communicate the results to others within their particular networks, including family physicians and medical specialists. Also, CDBRA will inform organizations of medical and health professionals within Canada, as well as Federal and Provincial Departments of Health, of the results of this study. They, then hopefully, will advise and inform their networks. It will be CDBRA's intention to contact as many organizations, health and disability networks, etc. to get the message out about the late manifestations of congenital rubella. Other agencies which serve or advocate for individuals who are blind or deaf or hard of hearing will have the responsibility then to inform their clients about the later implications of this disease. The CDBRA will also place the results on its Web Site (www.cdbra.ca) allowing for further dissemination through that medium.

The ability to properly disseminate this vital information to a diverse special population calls for the need for a special registry or data bank for individuals with congenital rubella.

If such a data bank was available it would be possible to communicate with this population of individuals, their advocates, family members, care givers, etc. about the latest findings regarding this health issue. The CDBRA will be establishing a Data Registry for individuals who are deafblind in Canada. This will be a great source for information sharing for individuals who are deafblind resulting from congenital rubella.

The results of the study should convince people involved in the lives of individuals who have congenital rubella, that constant surveillance of their health is paramount. The understanding of their health risks, which are higher than the regular population, should be reason enough to ensure regular health monitoring. Family physicians need to be aware of the issue, be sensitive of the need to undertake additional and on-going testing of the individual with congenital rubella, and medically assess the individual holistically. Families, physicians and specialists in the fields of vision and hearing, need to be vigilant of further loss of vision and/or hearing, that can occur as the result of congenital rubella in an individuals youth, and later in life.

On the point of the need for sensitivity by the medical community to late manifestations of rubella, there needs to be one or several medical specialists fully versed in the rubella issues, available for consultation to family physicians, family members and care givers of individuals with congenital rubella. These specialists would thereby become aware, in consultation with family members and others in charge of their care, of these individuals' special needs, and be empathetic with respect to medical diagnoses and for any resultant medical care, such as prescribing medications or hospitalization care. If not directly available to the individual, at least these specialists could consult with family physicians, who may not always be comfortable dealing with some individuals who have very special and challenging needs.

This suggestion also speaks to the support for the Canada wide need for Intervention services for all congenital rubella individuals with deafblindness. During medical testing, as well as with the prescribing of medication or hospital procedures, these individuals require an Intervenor to be with them to assist with communication. Health changes, such as those caused by neurological, hormonal or blood chemistry changes, often require the use of various medications or even hospital care. This can be very frightening to individuals who have a serious difficulty with communication. The opportunity to have a trained Intervenor to be the individual's "go between" with the medical community is a definite must!

Finally, this study further reinforces the need for continuous publicity about rubella and the mandatory requirement for vaccinations against this disease. The outbreak of rubella in Manitoba in 1997 demonstrates a minor breakdown in the immunization program. Admittedly there is a growing awareness about the side effects of vaccinations, including rubella, and this should not be hidden from the public. While short term arthralgia and, rarer, long term arthritis are recognized symptoms of rubella vaccinations, (Tingle, personal communications, 1998) they underscore the short and long term implications of congenital rubella. Thanks to a wide spread policy of immunizations over the past twenty years in Canada, few incidences of congenital rubella have been observed, although five (5%) of the total sample in this study are twelve years of age or less. This sample is still too high, and is indicative that congenital rubella babies will continue to be born in Canada until everyone is immunized against rubella. There should be no exceptions! Furthermore, the Government of Canada should ensure, if there is no evidence of immunity, that all refugees and new immigrants are tested for rubella and vaccinated.

3.7 Final Words

Society has paid a high price because of past epidemics of rubella, in particular because of its effect upon an early developing fetus. Just looking at the lives of one hundred Canadians who have been unfortunate victims, is enough to confirm the need to be vigilant against this virus. The only known way to combat rubella and keep the statistics described in this study to be just an historical artifact, is by maintaining a vigorous campaign of rubella immunizations. We must recognize that there are known side effects from the vaccinations to some individuals, including short term arthralgia. One cannot rule out long-term arthritis and the connection to diabetes.

The challenge is to review the facts from this report. Look at the effect on the one hundred human lives; look at the costs in medical care; examine the special education requirements and the need for specialized living facilities. Society cannot afford to have this happen again!

Bibliography

Bonnick, S.L. (1994). *The Osteoporosis Handbook*. Taylor Publishing Company, Dallas.

Cherry, J.D. (1987). *Rubella*. In: Feigin, R.D. and Cherry, J.D. (Eds). *Textbook of Pediatric Infectious*

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

Diseases. 2nd Ed. W.B. Saunders, Philadelphia, pp. 1810-1841.

Chess, S., Korn, S.J., and Fernandez, P. (1971). *Psychiatric Disorders of Children with Congenital Rubella*. Brunner/Mazel, New York.

Cochi, S.L., Edmonds, L.E., and Dyer, K. (1989). *Congenital rubella syndrome in the United States, 1970-1985*. American Journal of Epidemiology, 129, 349-361.

Collins, S. (1998). *Personal communication*.

Cooper, L.Z. (1966). *German Measles*. Scientific American 215 (1): 30-37.

Cooper, L.Z. (1985). *The history and medical consequences of Rubella*. Review of Infectious Diseases (Supplemental)7, 2-10.

Cooper, L.Z., Ziring, P.R., and Ockerse, A.B. (1969). *Rubella: clinical manifestations and management*. American Journal of Diseases in Children, 118,1829.

Gilbert, G.L. (1991). *Infectious diseases in pregnancy and the newborn infant*. Monographs in Clin. Pediatrics. 23-62.

Givens, K.T., Lee, D.A., Jones, T. and Ilstrup, D.M. (1993). *Congenital rubella syndrome: ophthalmic manifestations and associated systemic disorders*. British Journal of Ophthalmology, 77, 358-363.

Horne, D. (1998). *Personal communication*.

Mamer, L. (1998). *Personal communication*.

Menser, M.A. and Reye, R.D.K.(1974). *The pathology of congenital rubella: a review written by request*. Pathology, 6, 215-222.

Menser, M., Forrest, J., Bransby, R. and Hudson, J.(1982). *Long-term observation of diabetes and the congenital rubella syndrome in Australia*. In Mimura, G., Baba, S., Goto, Y. and Kebberling, J. (Eds). Clinicogenetic Genesis of Diabetes Mellitus. Excerpta Medica, Amsterdam.

Merth,T., Gerritsen, E.J.A, Van Tol, M.J.D, Stokvis, W.H, Weiland, H.T., and Vossen, J.M. (1987). *The late onset type congenitale rubella syndroom*. Nederlands, Tijdschrift voor Geneeskunde, 1987, 31, 2203-2206.

O'Day, A.F. and Mayhall, C.A. (1988). *Delayed manifestations of congenital rubella*. Journal of Visual Impairment and Blindness, November, 379-381.

O'Donnell, N. (1996). *History of congenital rubella syndrome*. Journal of Vocational Rehabilitation, 6, 149-157.

Preblud, S.R. and Alford, C.A. (1990). *Rubella*. In: Remington, J.S and Klein, J.O. (Editors) *Infectious Diseases of the Fetus and Newborn Infant* 3rd Ed. W.B Saunders, Philadelphia, pp.196-240.

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

Sallomi, S.J. (1966). *Rubella in pregnancy. A review of prospective studies from the literature*. Journal of

Survey of Late Emerging Manifestations of Congenital Rubella in Canada

Obstetrics and Gynecology, 27, 252-256.

Sidle, N. (1985). *Rubella in Pregnancy. A Review of Rubella as an Infection in Pregnancy, its Consequences and Prevention*. Sense. 108 p

Slagle, B.L. and Wolinsky, J.S. (1989). *Rubella virus and central nervous system disease*. Clinical and Molecular Aspects of Neurotropic Virus Information, 303-318.

Tingle, A. (1998). *Personal communication*.

Van Alphen, K. (1998). *Personal communication*.

van Dijk, J. (1991). *Persons Handicapped by Rubella; Victors and Victims: A Follow-up Study*. Swets & Zeitlinger, Amsterdam.

Vernon, M. (1969). *Multiple handicapped deaf children. Medical, educational and psychological considerations*. CEC Research Monographs 18, (1).

Vernon, M. and Hicks, D. (1980). *Overview of rubella, herpes simplex, cytomegalovirus, and other viral diseases: Their relationship to deafness*. American Annals of the Deaf, 125, 529-534.

Wagman, R.J. et al. (Eds)(1988). *Medical and Health Encyclopedia*. 'Round the World Books Inc. Division of MacLean Hunter Ltd., Toronto.

Walters, J. (1994). *Behaviours in persons with CRS - a response*. NFADB Newsletter. Fall edition, 10-12.

Waxham, M.N. and Wolinsky, J.S. (1984). *Rubella virus and its effect on the central nervous system*. Clinical Neurology, 2, 367-385.

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

Ziring, P.R., Fedum, B.A. and Cooper, L.Z. (1967). *Thyrotoxicosis in congenital rubella*. Journal of Pediatrics, 87, 1002.

Ziring, P.R. (1978). *Psychiatric sequelae of the 1964-65 rubella epidemic*. Annals of Psychology, August. 1-6.

Survey of Late Emerging Manifestations of Congenital Rubella in Canada