Developing a National Volunteer Registry of Persons with Deafblindness in Canada:

Results from the study, 1999-2001

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The Canadian Deafblind and Rubella Association
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About the Author

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Stan is a founding member of the Canadian Deafblind and Rubella Association (CDBRA), has served as president and currently serves as its Special Projects Consultant. He is the author of the CDBRA publication, A Survey of Late Emerging Manifestations of Congenital Rubella In Canada, 1999. Stan serves as Canada's representative on the World Council of Deafblind International (DbI).

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SUMMARY

- Fifty (50) organizations throughout Canada were involved in locating persons with deafblindness to participate in this study.
- Seven hundred and seventy-seven (777) individuals volunteered to be part of the Registry, 370 males and 407 females. The oldest person reporting their age was born in January, 1900, while the youngest was born in June,1998. Twenty persons or 2.6% of the sample were of pre-school age; 121 or 15.7% were school aged (6 to 21 years); 463 or 60.1% were aged 22-64 and 166 or 21.6% were aged 65+.
- The vast majority (86.4%) of these individuals had been previously identified as having the disability and were receiving some level of service from an organization.
- In the sample of 777 individuals, 44.7% of the respondents had congenital deafblindness, while 55.3% reported their deafblindness as acquired.
- Causes of deafblindness for all respondents are summarized in the following categories: inherited rare disorders (35%); intra-uterine infections (19.5%); congenital brain damage (8%); aging (8%); non-inherited chromosomal abnormalities (6.5%); post natal/early childhood infections (5.4%); acquired brain injury (5.2%); prematurity (3.6%); metabolic conditions (2.7%); birth trauma (2.3%); Sexually Transmitted Diseases (1%) and others (2.5%).
- Usher Syndrome and Congenital Rubella Syndrome are the primary reported causes of deafblindness, accounting for 29.9% and 18.7%, respectively.
- Fifty-five percent (55%) of the respondents reported receiving the services of an Intervenor
- Almost 26% reported completing some form of education. Just over 19% of respondents are currently in an education program.
- Eighty-one (81) persons or 10.4% of the total respondents indicated they were working, 51 working in competitive employment and 30 working in sheltered programs. The majority (74.5%) working competitively had acquired deafblindness; the majority (86.7%) working in sheltered programs had congenital deafblindness.
- Persons with acquired deafblindness tend to have a higher rate of use of such modes of communication as writing/printing, sign language (American Sign Language and Signed English), manual alphabet, and speech, than those with congenital deafblindness.
- The living arrangements of the respondents were reported as follows: lives independently (34.3%); lives with birth family (23.3%); lives in Provincial facility (15.5%); lives in supported living facility (8.6%); lives in retirement home (8.5%); lives in supervised living facility (5.5%); lives with adoptive parents or in foster care (3.7%) and lives in nursing home (0.6%).
- Of those respondents born before 1980, 72% reported their marital status as single; 18% married; 9% widowed and 1% divorced.
- A safe estimate of the deafblindness ratio in Canada is 10 to 15/100,000, or a population of 3,100 to 4,650 persons.
- The findings of this report show significant distinctions between congenital and acquired deafblindness in almost every category where comparisons were available. In addition, one major distinction exists; the wide variance between the two groups in terms of the type and extent of services available to each.

1.0 INTRODUCTION

Professionals, advocacy organizations and people who are deafblind have long recognized the necessity to develop a better understanding of the number of persons in Canada with the disability of deafblindness.

Almost two decades ago, a special task force (The Task Force on Services to Deaf-Blind Persons in Canada) was appointed by the Federal Government to report on service needs for persons who are deafblind in Canada. The task force tabled a report 'Task Force On Services to Deaf-Blind Persons in Canada' in 1984 to the Federal Advisory Committee on Services to Deaf-Blind Persons in Canada. Recommendation Number 21 of that report stated "only through accurate statistical data on deafblindness and deafblind persons in Canada can appropriate services be planned and delivered." Unfortunately, nothing concrete developed from these recommendations, until recently.

The Canadian National Institute for the Blind (Deaf-Blind Services) attempted to develop a Registry to meet the task force recommendations, but found it necessary to abandon its efforts for financial reasons. It was with this background that the Canadian Deafblind and Rubella Association (CDBRA) proposed in 1998, to initiate the establishment of a National Registry of Persons who are Deafblind in Canada.

1.1.1 Project Description

Proposal to Human Resources Development Canada (HRDC)

In the summer of 1998, CDBRA prepared an 'Application for Special Project Funding' to the Office of Disability Issues, (HRDC), titled ***"****Project to Develop a Registry of Persons who are Deafblind in Canada".*

The purpose for the project submission (as taken from the application) was:
To establish a voluntary registry of all persons who are deafblind in Canada for the purposes of:

(a) Determining the number of persons, by geographic region, who have been previously identified as deafblind and are in receipt of services or who are identified as deafblind but are not receiving adequate services;

(b) Providing a means of locating individuals who have not previously been identified as deafblind for the purpose of ensuring they have access to a training or education program and receive Intervention services;

(c) Establishing a means of communicating with all people who are deafblind, their families and/or caregivers and agencies who serve them to disseminate current information about deafblindness, the availability of services and programs for individuals who are deafblind in various regions of the country;

(d) Collect vital demographic information on a select population to assist government agencies and volunteer organizations to plan for and implement appropriate services.

The project submission was subsequently approved by HRDC and implementation began in October, 1998.

1.1.2 Project Management

The CDBRA Board of Directors determined that, to be successful with this project it must partner with all organizations providing services to, or advocating for, those with either acquired or congenital deafblindness.

An Advisory Committee reporting to the CDBRA Board of Directors was established, consisting of representatives from the CDBRA (a parent-professional advocacy group); Canadian National Society of the Deaf-Blind (CNSDB - an adult consumer advocacy organization); Rotary Cheshire Apartments (RCA - an independent living facility for adults with acquired deafblindness, located in Toronto); Canadian National Institute for the Blind (CNIB - an organization providing, through its district offices located throughout Canada, case management and specialized services to adults with deafblindness) and the W. Ross Macdonald School for Students who are Blind or Deafblind (WRMS - an educational facility providing residential and outreach educational services primarily for children and youth who are congenitally deafblind).

The intent of this committee was to guide the project from start to finish, including suggestions about: how to survey the various populations, develop contacts, establish project phasing and timetables, maintenance of the Registry database, confidentiality, and data reporting.

A Project Coordinator was appointed, who would report to the CDBRA Board of Directors, and work closely with the Advisory Committee Members. The Project Coordinator's responsibilities included: gathering representatives to form the Advisory Committee; consulting with all members to seek their assistance and advice; bringing the project to completion; maintaining financial records for CDBRA and reporting them to HRDC, and, producing the final report.

The make-up of the Advisory Committee was as follows:

Linda Mamer	CDBRA and Chair
Jim Thompson	CNSDB
Barb Davis	CNSDB
Bill Thompson	WRMS
Bev Ginou	CNIB
Joyce Thompson	RCA
Stan Munroe	Project Coordinator

1.1.3 Project Phases

The Advisory Committee suggested the project be implemented in two phases. **The first phase (Phase 1)** would focus on finding out who is 'known to be deafblind' in Canada. This consisted of developing a questionnaire intended to gather basic demographic information about the individual; learn the status of their vision and hearing loss and its etiology; determine their living arrangement, personal status (in school, retired, working, etc.), and their access to various services (Intervenor, interpreter, friend, etc). In fact, two questionnaires, referred to as the CDBRA and CNSDB forms, respectively, were developed. Both questionnaires contained basically the same questions but presented in different formats. The CDBRA form (see Appendix 5.1.1.1) was intended for those who were congenitally deafblind; and it was expected that they would be completed by family members or advocates. The CNSDB form (see Appendix 5.1.1.2), was designed to be completed by persons who are deafblind. Braille and large print formats were provided to persons known to the organizations partnering in the project.
Each questionnaire (produced in English and French) was accompanied by a letter (see Appendix 5.1.1.1 and 5.1.1.2) outlining the purpose of the survey, the definition of deafblindness, and emphasizing that participation in the survey was strictly voluntary and that any identifying data submitted would be held in strict confidence (See Appendix 5.2.2 for copy of Registry Project Confidentiality Guidelines).

It was well understood by the Advisory Committee that gaining support for this study would be much easier from the population of persons with congenital deafblindness than from the larger and more complex community of those with acquired deafblindness. Persons in the congenital group usually have strong advocates, or family involvement; they are usually well connected with an educational facility, a group home or an independent facility. It is much more challenging to reach individuals from the acquired group. Probably less than half of this population are known to advocacy and service organizations. Many are reluctant to provide confidential information; many have concerns about participating in yet another survey since they have seen little or no action resulting from previous ones. Some from the acquired group are residents of Provincial Institutions or group homes operated by Community Living Organizations and do not receive other services. Many, too, are seniors, either living alone or in Seniors' residences; without appropriate services.

The second phase (Phase 2) was intended to gather further information from those who participated in Phase 1. A questionnaire (see Appendix 5.1.2) was developed which included questions regarding employment, Intervenor Services, living arrangements, training, technical aids, education and leisure activities.

1.1.4 Data management

The services of a statistician was contracted to set up the data entry package for Phase 1 and Phase 2 of the project and to produce statistical compilations. MS Access software was used for the Registry data entry and all data analyses.

1.1.5 Establishing the Cooperative Network

Locating individuals to make them aware of the volunteer Registry Project required networking with a wide array of non-profit organizations, educational and residential facilities, professionals, administrators, educators, care givers and family members, throughout Canada.

Through various sources, in particular the publication, *'Directory of Disability Organizations in Canada, 1998'*, the steering committee located the names and addresses of organizations and facilities believed to advocate for, or provide services to, persons with deafblindness. Information letters were mailed to 148 contact organizations and facilities in Canada in early January, 1999, requesting their participation in this project. The regional breakdown for the contacts is as follows: Maritime Provinces (52); Quebec (5); Ontario (48); Manitoba (6); Saskatchewan (18); Alberta (9) and BC (10).

A total of 50 organizations agreed to participate in this project. Some were national organizations having provincial Chapters or Divisions**.** Appendix Table 1 details the names of the cooperating organizations, organized by province, and the name of key contact persons.

Each cooperating organization agreed to distribute the survey material (including questionnaires, introductory letters and stamped, pre-addressed envelopes), confidentially, to all persons they knew to be deafblind, according to the definition of deafblindness. Questionnaires were produced in braille and large print, in both English and French, and were distributed as requested.

1.1.6 Project Timetable

Phase 1 of the project was fully implemented by March 31, 1999. Two thousand, four hundred and twenty (2,420) questionnaires were distributed among the 50 cooperating organizations throughout Canada. Potential registrants were asked to return the completed questionnaires by June 30, 1999. Twenty six (26) of the questionnaires were returned by Canada Post 'undelivered'.

An interim summary of the statistics was produced at the end of 1999 for distribution to the Phase 1 participants. A total of 654 persons volunteered their personal information for the Registry by December 31, 1999.

Phase 2 was launched by February 2000. Most persons registered during Phase 1, except those who indicated they were not prepared to participate further, were sent a follow-up questionnaire (see Appendix 5.1.2), along with the preliminary statistics.

Up until December 2000, the Project Leader made further contacts of organizations and facilities to further enhance the registry data base. An additional 123 individuals joined the Registry by the end of 2000, for a total of 777. Of the original Phase 1 participants, 224 completed Phase 2 questionnaires.

1.2 Definition of Deafblindness

Since this project was based on a mail-in survey and not a hands-on assessment of the disability, cooperating organizations and participants were provided criteria to assist them in establishing who fit the criteria of being deafblind. The following functional definition of deafblindness, which was developed in Canada as a consensus of educators and other professionals (Bulmer and Newbery, 1995), was the cornerstone for this study: *"Individuals are considered to be deafblind if they have a substantial loss of both sight and hearing such that neither sense provides reliable access to information. The combination of these sensory losses results in significant difficulties in acquiring educational, vocational, avocational and social skills."*

To aid those individuals who would be completing the questionnaire themselves, (the CNSDB questionnaire) this definition was clarified in the following manner.
A person is deaf-blind (CNSDB spelling) if they:

- are deaf and blind
- cannot clearly hear speech or alarms without hearing aids, and are blind
- cannot clearly hear speech or alarms without hearing aids, and cannot read the newspaper even with the best glasses on
- are deaf and have severe tunnel vision
- cannot clearly hear speech or alarms without hearing aids, and have severe tunnel vision
- cannot clearly hear speech or alarms without hearing aids, cannot see road signs or bus numbers even with the best glasses on, and need help to travel in unfamiliar areas.

1.3 Classifications of Deafblindness

The data supplied by all of the respondents allowed the steering committee to assign most persons to one of four broad classifications of deafblindness: 1) *congenital deafblind*; 2) *acquired deafblind: pre-lingually deaf with acquired vision loss*; 3) *acquired deafblind: post-lingually deaf with acquired vision loss;* and 4) *acquired deafblind: congenital blind with acquired hearing loss*. There were some persons who had acquired deafblindness without sufficient information to assign them to a specific classification within the acquired group. They were subsequently assigned the classification: *acquired deafblind: classification unknown*.

- **Congenital Deafblindness**

Persons with congenital deafblindness have minimal or no vision or hearing at birth, or they lose their hearing or vision before the age of two. Causes are usually due to prenatal insults (virus); prematurity; chromosomal abnormalities; rare disorders; post natal influences, etc., up to the age of two. Education must be quite specialized for this population; individuals usually attend special educational programs, with the support of Intervenors, in public schools or in residential settings, where the opportunity exists. Generally, communication abilities are quite limited among this group. Persons with congenital deafblindness do not usually live independently. Many of those born since the early to mid-1960's live in supported living residences with Intervention Services. Older individuals, largely continue to live in institutions. In the past two decades there has been an increase in the number of children with multiple disabilities, due to a higher survival rate of children born prematurely, and/or born with rare disorders.

- **Pre-Lingual Deafness with Acquired Vision Loss**

These individuals are, or become, deaf or hard of hearing before the age of three and lose their vision at a later time. Causes of this type of deafblindness include Usher Syndrome (Type 1), a genetically inherited condition; accidents and post-natal infections such as meningitis, contracted before the age of three. These individuals usually do not acquire normal speech. Communication skills are often delayed in this group because of lack of exposure to a visual language system, such as signing, or concomitant tactual developmental disabilities. If their primary language is through the use of visual sign language, they will adapt to non-visual methods of communication, such as tactile signing, as their visual loss progresses. Persons with this disability are usually educated in mainstream classrooms or, where facilities exist, in residential schools for individuals who are deaf. Individuals in this category may live independently due to their ability to cope visually during their early years. As their vision declines they must learn to adapt their forms of communication.

- **Post-Lingual Deafness with Acquired Vision Loss**

Individuals with this type of deafblindness acquire both vision and hearing loss, often separately, after the age of three. Causes are varied and include several genetically inherited conditions (e.g. Usher Syndrome Types 2 and 3), head trauma, metabolic conditions (e.g. diabetes), various medical conditions, stroke, ageing, etc. Since they acquired regular speech early in their lives, they have usually attended regular schools and had work histories like those of the regular population. Acquiring this disability is very traumatic to the individual and requires psychological adjustment as well as the need to learn new communication methods.

- **Congenital Blindness with Acquired Hearing Loss**

A less common form of deafblindness, individuals lose their vision before the age of two years and acquire their hearing loss at a later age. Causes include genetically inherited disorders (e.g. Alstrom and Norries Syndromes), birth trauma and early post-natal infections, etc. These individuals rely on normal speech expressively and receptively, but will require tactile communication methods as their hearing loss progresses. Persons with this disability are usually educated in regular classrooms or, where facilities exist, in special schools for individuals who are blind.

1.4 Other Registries or Census of Persons with Deafblindness

Several countries have undertaken efforts to count the population of persons with deafblindness and/or establish registries in their respective countries. A few are outlined here.

- **Norway**

The Directorate of Health in Norway made a survey of deafblind individuals first starting in 1976-1977. Their approach was to establish the total number of deafblind people, obtain basic demographic and social data about them, determine their social, medical and educational needs, and recommend measures to reduce the impact of this very severe disability (Svingen, Friele and Jacobsen, 1987). Since that time, Norway has maintained a registry through an ongoing process of identification, based on accepted criteria. Through this process, persons with deafblindness in Norway become eligible to receive the country's full network of educational and social support. The most recent numbers from Norway (December, 2000) indicate there are 302 persons on its Registry, 231 with acquired deafblindness and 71 with congenital deafblindness. Based on their population of 4.4 million people, their deafblindness index is 6.9/100,000 persons. Managers of the Norwegian registry recognize that not all persons with deafblindness in Norway are on their Registry; most of those not registered are older people (Oystein Stette, personal communication, 2001).

- **United Kingdom**

SENSE has estimated that there are 23,000 deafblind or dual sensory impaired people in the UK (Ellis, 1998). The incidence rate is calculated as 40/100,000. This was based on local studies in Devon, Sunderland and Lincolnshire extrapolated for the entire UK. Subsequent recent estimates of 30/100,000 and 58/100,000 were obtained, respectively, in Devon and Northern Ireland, corroborating the 40/100,000 index (Lewin-Leigh, personal communication, 2001).

- **United States**

Interestingly, there is no complete national census or registry of persons with deafblindness in the USA. There are several separate programs designed to obtain information about deafblindness in children and youth (up to age 21).

There is the National Census for Deaf-Blind Children and Youth, ages 0-21. This census is maintained by the Teaching Research Division, Western Oregon University (Monmouth), for the Federal Office of Special Education Programs. This census is produced annually and it is the duty of each state to report this information for the purpose of the national census. The most recent information (December 1, 1999) indicated 10,198 persons aged 0-21 were on this Registry (Hembree, 2000).

There is no comparable estimate of the adult deafblind population (over 21 years of age) in the USA. In 1997, the United States Congress authorized the Helen Keller National Center (HKNC, Sands Point, New York) to develop and maintain a national registry of persons who are deafblind. At the present time, the HKNC registry contains information on over 6,700 individuals in areas such as etiology, communication, employment, living situations and services. Since participation in the registry is voluntary, efforts are underway to work collaboratively with adult service agencies and children's programs to gather additional information (Nancy O'Donnell, personal communication, 2001).

2.0 RESULTS

2.1 Registry Participation

A total of 931 Registry forms were submitted for inclusion in the Registry. For the purpose of reporting on the study project, the final Registry numbers total 777. The remaining one hundred and fifty four individuals (154) were not included as pertinent information about the nature of their vision and hearing impairment was incomplete. It is the intention to include these 154 individuals (one from British Columbia and 153 from Quebec) on the permanent Registry data base once these additional details are obtained.

Table 1 summarizes, by Province, the number of persons who participated in Phases 1 and 2 of the project and the number of persons known as having deafblindness, by various organizations and facilities in Canada.

Table 1. **Summary of Registry participation rate compared to number known by various organizations as having deafblindness**

Province	Number of participants in Registry Project Phase 1	Number of participants in Registry Project Phase 2	Number of persons with deafblindness known by various organization and facilities
Alberta	**39**	11	112
British Columbia	**93**	25	285
Quebec	**168**	64	515
Saskatchewan	**14**	4	30
Manitoba	**38**	18	80
New Brunswick	**21**	7	70
Newfoundland	**9**	3	11
Nova Scotia	**30**	9	39
Ontario	**361**	83	549
Prince Edward Island	**4**	0	4
Total	**777**	**224**	**1725**

The last column, for the purpose of this report, represents the current known numbers of persons with deafblindness in the various provinces of Canada. It is quite likely an underestimate of the true population in Canada. It was these individuals that the various cooperating organizations initially invited to participate in this project. For comparison purposes, the Task Force Report in 1984 (Task Force, 1984) indicated that the various organizations were aware of 830 persons with deafblindness. The current estimated total of 1725 would indicate there has been some, but slow, progress, in identification of this population.

2.2 Birth Period and Gender of Registry Participants

The reported birth dates of all participants (who provided a date of birth) was organized into ten year intervals from 1900 -1998 and presented in Table 2. Six participants were not willing to provide their date of birth for personal privacy issues.

Table 2. Distribution of birth period and gender of registry participants

Birth Period	Number of Respondents	Male	Female
1900-1909	16	4	12
1910-1919	64	11	53
1920-1929	54	23	31
1930-1939	53	22	31
1940-1949	111	59	52
1950-1959	118	55	63
1960-1969	107	60	47
1970-1979	99	57	42
1980-1989	89	51	38

1990-1998	60	26	34
Total age specified	**771**	368	403
Age not specified	6	2	4
Total responded	**777**	**370**	**407**

The oldest person reporting their age in this study was born in January, 1900, while the youngest was born in June,1998. Twenty persons or 2.6% of the sample were of pre-school age; 121 or 15.7% were school aged (6 to 21 years); 463 or 60.1% were aged 22-64 and 166 or 21.6% were aged 65 and older.

It is not believed that the age distribution in this sample reflects, in any way, the true picture of the population with deafblindness in Canada. In particular, the component aged 65+ is likely two to three times the proportion (21.6%) indicated by this sample. Lewin-Leigh (personal communication, 2001) indicated that in the UK study, children and young adults (0-21 years) represented 17% of the deafblind population, other adults up to age 64 represented 17% of the population, while those 65 and older comprised 66% of the population with deafblindness.

Eleven registrants, who volunteered to be part of the Registry, have since died. Their birth years are as follows: 1908, 1909, 1918, 1919, 1922, 1926, 1929, 1939, 1947, 1962 and 1992. Despite their deaths, their information has not been removed from the data base for the purpose of this report.

The ratio of total males to females in the sample of 777 is 48:52. For the birth period 1900-1919, the male to female ratio of 80 persons in the sample is 19:81. In the remainder of the sample (691) of persons born 1920 +, the male to female ratio is 51:49. It would appear that this gender information reflects the typical Canadian population.

2.3 Occurrence Levels of Types or Classifications of Deafblindness

The reported occurrence of the types or classifications of deafblindness as reported by the respondents is found in Table 3.

Table 3. Reported occurrence of the types or classifications of deafblindness among the registry participants

Classifications of Deafblindness	**Number**	**Total**
Congenital Deafblind	347	**347 (44.7%)**
Acquired Deafblind: Pre-lingual Deaf with Acquired Vision Loss	195	
Acquired Deafblind: Post-lingual Deaf with Acquired Vision Loss	180	
Acquired Deafblind: Congenital Blind with Acquired Hearing Loss	33	
Acquired Deafblind: Classification Unknown	22	
Acquired Deafblind (Total of above)		**430 (55.3%)**
Total		**777**

In the sample of 777 individuals, those with congenital deafblindness accounted for 44.7% of the respondents, while 55.3% reported their deafblindness was of the acquired type. Within the acquired sample of 430, it was possible to assign all but 22 to the three accepted categories as shown in Table 3. These 22 respondents did not supply enough information to allow a proper assignment and were placed in the category of Acquired: Classification Unknown.

Within the assigned Acquired Group (408), persons with Pre-lingual Deafness and Acquired Vision Loss accounted for 47.8%; persons with Post-lingual Deafness and Acquired Vision Loss accounted for 44.1%, while persons with Congenital Blindness and Acquired Hearing Loss accounted for 8.1%.

It is believed that the occurrence rate of congenital (44.7%) and acquired (55.3%) deafblindness presented here

does not truly represent the occurrence of these groups within the population of deafblind persons in Canada, and this must be recognized when evaluating any of the statistics.

There are a number of reasons for the apparent over-representation of the congenital component and under-representation of the acquired component:

- The Registry was voluntary and participation was dependent upon personal interest and concern.
- The size of sample of those with congenital deafblindness is likely influenced by a greater awareness by organizations and various facilities of these individuals and consequently the great cooperation by these organizations, family members and advocates, to complete the surveys on behalf of these persons.
- There may have been a tendency of some organizations or facilities to identify persons as congenitally deafblind when they may have not fit the functional definition of deafblindness as set out in the questionnaire.
- In comparison, many with acquired deafblindness are unknown to organizations like the CNIB and not being on their mailing list, were not aware of the project; if they were aware, they may not have had support services and/or supporting individuals (Intervenors, advocates, family members, etc.) in place to assist them with completing forms. Many in this group are suspicious of more studies and like others within the mainstream population, are reluctant to voluntarily complete yet another survey.

Despite reporting that the above mentioned statistics present a disproportionate picture, full confidence in estimating the actual occurrence of the various components of the deafblind population in Canada is not possible without knowing the total population.

Only the Norwegian Registry offers hard numbers to which we can compare. From the total of 302 reported on its Registry at the end of 2000, those with congenital deafblindness accounted for 23.5% and those with acquired deafblindness accounted for 76.5% (Oystein Stette, personal communication, 2001). The UK study does not separate the components with any accuracy and, in the US study, there is only information about children 0-21 years.

2.3.1 Birth Period and Classifications of Deafblindness

Classifications of deafblindness (congenital and acquired) were cross-tabulated with birth period and presented in Table 4.

Table 4. Distribution of deafblind classifications by birth period

Birth Period	Number	Total no. with Congenital Deafblindness	Total no. with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness	Acquired: Unknown Classification
1900-1909	16	0	16 (100%)	2	14		
1910-1919	64	3 (4.7%)	61 (95.3%)	11	48	2	
1920-1929	54	1 (1.9%)	53 (98.1%)	17	34	2	
1930-1939	53	5 (9.5%)	48 (90.5%)	25	19	3	1
1940-1949	111	26 (23.4%)	85 (76.6%)	45	27	10	3
1950-1959	118	42 (35.6%)	76 (64.4%)	44	17	5	10
1960-1969	107	65 (60.7%)	42 (39.3%)	24	6	4	8
1970-1979	99	81 (81.8%)	18 (18.2%)	13	4	1	
1980-1989	89	70 (78.7%)	19 (21.3%)	12	4	3	
1990-1998	60	54 (90%)	6 (10%)	1	2	3	
Total age specified	771	347	424	194	175	33	22
Age not specified	6	0	6	1	5		
Total	777	347	430	195	180	33	22

It is interesting to observe the changes in relative proportion of each classification of deafblindness throughout the entire period from 1900-1998, which covers the period when all persons in the study were born. Broadly speaking, for those born before 1940 (187), the vast majority (95.2%) have acquired deafblindness. The proportion of those with congenital deafblindness tends to increase through the birth periods until the period 1960-69 when the number with congenital deafblindness (60.7%) first exceeds the number with acquired deafblindness. This trend continues through the remainder of the more recent birth periods, with congenital deafblindness accounting for 90% of those born since 1990. For those born since 1980, those with congenital deafblindness account for 83.2% of the total of 149 persons. For a comparable age group in the US study (Hembree, 2000), it was observed that about 73.5% of them were congenital deafblind.

According to Joyce Thompson (personal communication, 2001), those with acquired deafblindness should be more represented among this age group of persons with deafblindness. This is likely because many younger people with Usher Syndrome, the major cause of acquired deafblindness, did not participate in this study for a variety of reasons. Some of these persons are still in denial about being identified as deafblind. Others prefer to strike out on their own, securing their own funding and developing services that meet their individual needs; thus may not have known about this study. Furthermore, there is the increase of new Canadians with language barriers, and different cultural and religious beliefs that often restrict them seeking services, and hence being unaware of this study (Bev Ginou and Nancy Lord, personal communication, 2001). Another reason, but more seriously, is inadequate screening of Usher Syndrome in schools where students who are deaf or hard of hearing are educated.

Several explanations will be attempted, to describe what influences the change in percentage of the two deafblindness classifications through the range of birth periods as presented in Table 6.

First, acquired deafblindness is recognized to be highly influenced by ageing. About one half of those born between 1900 and 1919 indicated the cause of their deafblindness was ageing, while about a quarter of those born in the 1920's attributed their deafblindness to ageing. Thus, the higher proportion of acquired deafblindness in this sample of the older population born up to 1929 (134 persons or 17.3% of the total sample) can be safely attributed to ageing related issues. This includes the Usher Syndrome group of individuals who may not acknowledge the full extent of their vision loss.

Second, the period of the mid 1960's and 1970's witnessed a huge epidemic of maternal rubella. The highest proportions of congenital rubella syndrome occurred during the 1960's and 1970's (see Table 9) which likely accounts, in part, for the disproportionate number of persons with congenital deafblindness beginning in that period.

Third, the continuing disproportionately high ratio of congenital to acquired deafblindness in the period 1980-1998, is largely due to a high reporting rate by family members and advocates of individuals with this condition. For many of those with acquired deafblindness, most would be in the early stages of this disability and be either not fully aware of, or still in denial of, their disability. The interest in volunteering for such a survey would be lower among this group than for the congenital group.

2.4 Causes of Deafblindness

All the causes contributing to deafblindness (or deafblindness etiologies) as reported by the respondents are briefly described and summarized according to primary contributing causes or etiologies. The frequency distribution of these primary etiology categories are further organized by the classifications of deafblindness. This material is presented in Tables 5 and 6.

The following describes the major categories of contributing causes or etiologies that have been reported as leading to the condition of deafblindness. No individual follow-ups were made to inquire further about any of the questionable responses (see Table 6).

- **Acquired Brain Injury**

An injury to the brain which can result in various degrees of vision, hearing, cognitive and motor impairments. Visual and hearing loss from these injuries may also be the result of the loss of visual and auditory processing without any effects on the eyes or ears themselves. Seventeen different types of occurrences were reported for this causative factor, including: aphasia, various accidents, near drownings, liver septicaemia, several syndromes, brain tumours, haemorrhages, and abuse.

- **Ageing**

The ageing process increases the incidence of both vision and hearing deterioration. A reduction in hearing acuity is a normal expectation of ageing. With ageing also comes an increasing incidence of such eye diseases as cataracts, glaucoma and macular degeneration; although these diseases are not strictly age related, since glaucoma and macular degeneration can occur in persons as young as 35 years old (Joyce Thompson, personal communication, 2001). The complication is when an older person with one of the senses already affected, experiences the onset of deterioration of the second sense. Often deterioration of both senses can occur simultaneously. Loss of both of these senses are frightening and often leads to loss of self-esteem. Learning new modes of communication (typically, tactually) for older people is often impeded by the reduction in manual dexterity due to the common affliction of arthritis. Six different types of ageing related conditions were reported by the respondents.

- **Birth Trauma (causing Brain Damage)**

Birthing irregularities which lead to reduced oxygen to the brain (hypoxia/anoxia), causing irreversible neuro-sensory damage. These trauma may lead to various degrees of hearing and visual loss. Conditions reported in this study included perinatal hypoxia, Meconium aspiration, and brain haemorrhage.

- **Congenital Brain Damage**

Damage to the brain which is present at birth. Damage may be caused by an intra-uterine infection, genetic defect, fetal injury or any insult to the developing fetus that affects normal brain development. Fifteen conditions or symptoms of this category were reported.

- **Intra-Uterine Infections**

Various diseases that invade the developing fetus in-utero cause sensorineural damage. Examples reported in this study, including rubella, cytomegalovirus and toxoplasmosis, can all cause hearing and vision loss.

- **Fetal Alcohol Syndrome (FAS)**

A cluster of related birth defects including: characteristic facial shape or appearance, small head size, poor growth before and after birth, developmental delay, short attention span and behavioural problems. Often there is a malformation of the eustachian tube leading to hearing impairment. Symptoms of FAS can appear in children born to mothers who drink during pregnancy.

- **Inherited Rare Disorders**

This includes rare chromosomal disorders which are transmitted from parent to offspring. Inheritance can involve numerous methods, including the blending of parental genetic material, transmission of recessive traits, transmission of dominant traits, sex linked transmission and maternal transmission. Eighteen different disorders fitting this category were identified in this study.

- **Medical Procedures**

Two incidences of medical procedures were reported as responsible for their deafblind condition: tonsillectomy and kidney dialysis.

- **Metabolic Disorders**

The result of a critical enzyme in the metabolic process being disabled or if a control mechanism for a metabolic pathway is affected. These diseases may be the result of inherited mutations or accidental protein mutations. Four different diseases were reported in this category, including diabetes and several rare syndromes.

Mitochondrial Condition

A rare neuromuscular disorder in which a defect in genetic material of the mitochondria (the intracellular structure which releases energy) causes the brain and muscles to function improperly (i.e. encephalomyopathies). In these disorders a high number of defective mitochondria are present. Two specific diseases were reported in this category.

- **Non-inherited Genetic/Chromosomal Abnormalities or Syndromes**

A disease or disorder that has more than one identifying feature or symptom, is a syndrome. Chromosomal abnormalities (including deletions, duplications, rings, inversions and trans-locations) can lead to birth defects, cognitive abnormalities and increased risks of miscarriage. Most chromosomal abnormalities occur spontaneously during meiosis in the egg or sperm; some happen after conception. Fourteen different syndromes fitting this category were reported for this study.

- **Prematurity** (**Complications)**

A child is premature when born less than 37 weeks gestation. Complications of prematurity include blindness, hearing loss and physical impairment. Conditions reported include Retinopathy of Prematurity (ROP), oxygen exposure (which is believed to cause ROP), cerebral palsy and spastic quadriplegia.

- **Prescribed Drugs (Complications)**

Large doses of prescribed drugs, in particular those of the *mysin* family of drugs, and massive life saving doses have been cited as causes of visual and hearing loss in some persons. In this study two specific incidents were reported, one attributed to quinine and another to a prenatal prescribed medication.

- **Post Natal / Early Childhood Infections**

Various bacterial and viral infections occurring in the immediate post-natal period or in early childhood. The implication in this context is the development of meningitis (infection of the neural lining) causing encephalitis and sensori-neural damage. Most who reported this causative factor did not know the particular disease, others reported such diseases as scarlet fever, red measles, mumps, whooping cough and tuberculosis.

- **Self abuse**

Self inflicted abuse (e.g., repeated head banging, poking foreign objects in ears and eyes) resulting in visual and hearing impairment and brain damage.

- **Sexually Transmitted Diseases (STD's)**

Viral and bacterial diseases transmitted by sexual contact. Examples include syphilis, gonorrhea and various herpes diseases. Can cause damage to sensori-neural centres of the brain, if transmitted to a fetus.

Table 5. Summary of contributing causes of deafblindness according to the various categories of the disability

Contributing Causes to Deafblindness	Acquired: Pre-Lingual Deaf / Acquired Blindness	Acquired: Post-Lingual Deaf / Acquired Blindness	Acquired: Congenital Blind / Acquired Deafness	Acquired: Unknown Classification	Total with Acquired Deafblindness	Total with Congenital Deafblindness	Total
Inherited Rare Disorders	145	42	16		203 (61.4%)	12 (4.2%)	215 (35%)
Intra-Uterine Infections	3		1		4 (1.2%)	116 (40.8%)	120 (19.5%)
Congenital Brain Damage		2			2 (0.6%)	47 (16.6%)	49 (8%)
Ageing		49			49 (14.9%)	0	49 (8%)
Non-inherited Genetic/ Chromosomal Abnormalities					0	40 (14.1%)	40 (6.5%)
Post-Natal/ Early Childhood Infections	8	9	3		20 (6%)	13 (4.6%)	33 (5.4%)
Acquired Brain Injuries		21		1	22 (6.6%)	10 (3.5%)	32 (5.2%)
Complications from Prematurity			1		1 (0.3%)	22 (7.7%)	23 (3.6%)
Metabolic Conditions	2	13			15 (4.5%)	2 (0.7%)	17 (2.7%)
Birth Trauma Causing Brain Damage			2		2 (0.6%)	13 (4.6%)	15 (2.3%)
Sexual Transmitted Diseases		2			2 (0.6%)	4 (1.4%)	6 (1%)
Fetal Alcohol Syndrome					0	3 (1.1%)	3 (0.5%)
Mitochondrial Condition		2			2 (0.6%)	0	2 (0.3%)
Complications from Prescribed Drugs		2			2 (0.6%)	0	2 (0.3%)
Complications from Medical Procedures		2			2 (0.6%)	0	2 (0.3%)
Other Causes		2	1	2	5 (1.5%)	2 (0.7%)	7 (1.1%)
Sub Total Reporting Known Cause	**158**	**146**	**24**	**3**	**331**	**284**	**615**

Sub Total Unspecified or Unknown Causes	**37**	**34**	**9**	**19**	**99**	**63**	**162**
Grand Total	**195**	**180**	**33**	**22**	**430**	**347**	**777**

Causes to Deafblindness	Deafblindness		Congenital Deafblindness		
Inherited Rare Disorders	**203**		**12**		**215**
	184	Usher Syndrome (1-reported as mentally disabled; 7-Type 2)	1	Atypical Leukodystrophy	
			1	Infantile Refsum Syndrome	
			1	Treacher Collins Syndrome	
	1	Hallgren Syndrome (Usher Syndrome with learning difficulties)	1	Smith Lemli-Opitz Syndrome (RHS Syndrome)	
			1	Cockayne Syndrome (early onset epilepsy)	
	1	Stargardt's Syndrome (high fever @ 3 mos.)	1	Acrocolossal Syndrome	
	1	Cockayne Syndrome (early onset epilepsy)	2	Alstrom Syndrome	
			1	Myers Syndrome (Smith-Fineman-Myers Syndrome)	
	4	Alstrom Syndrome			
	6	Norries Syndrome	1	Rud's Syndrome	
	2	Charcot Marie Tooth Syndrome	1	Tuberous Sclerosis	
			1	Phenylketurenia (PKU)	
	1	Friedrich Ataxia Syndrome			
	1	Lebers Disease			
	2	Neurofibromotosis			
Intra-uterine Infections	**4**	Congenital Rubella	**116**		**120**
	4	Syndrome	111	Congenital Rubella Syndrome	
				Cytomegalovirus	
			4	Toxoplasmosis	
			1		
Congenital Brain Damage	**2**		**47**		**49**
	1	not specified	11	not specified	
	1	Cerebral Palsy	4	developmental encephalopathy	
			1	holoprosencephaly	
			7	Cerebral Palsy	
			1	Hypoxic Ischemic	
			1	Encephalopathy (HIE)	
			1	prenatal hypoxia	
			1	cerebral haemorrhage	
				congenital brain malformation	
			6	hydrocephalus	
			1	enlarged left ventricle	
			1	Lennox-Gastaut Syndrome	
			1	neonatal bleeding	
			1	cerebral atrophy	
			4	microcephaly	
			1	Lissencephaly (with	
			1	microcephaly)	
			1	epilepsy	
			2	encelocephale	
				multi-disabled	
				Dandy-Walker Syndrome	

	18	cataracts			
	1	degenerating myopia			
	2	detached retina			
	2	eye thrombosis			
	2	unspecified			
Non-inherited Genetic/ Chromosomal Abnormalities			**40**		**40**
			1	18q minus syndrome	
			2	Ring Chromosome18 Syndrome	
			1	Wolf Hirschhorn Syndrome (4P-deletion syndrome)	
			1	Distal Trisomy 10Q	
			12	Downs Syndrome (Trisomy 21)	
			1	Trisomy 22 (partial Trisomy 11/22)	
			1	13Q minus ring	
			1	Pallister-Killian Mosaic	
			1	Syndrome (chromosome 12p tetrasomy)	
			1	Velo Cardio Facial	
			1	(chromosome 22q deletion)	
			14	Charge Association (22q11 deletion syndrome)	
			2	Cornelia de Lange Syndrome	
			1	Klippel Fiel /Arnold Chiarr Syndrome	
			1	Septo Optic Dysplasia	
			1	Apert Syndrome	
Post-Natal / Early Childhood Infections	**20**		**13**		**33**
	3	meningitis (1@age 3yr; 2@age 6yr)	11	meningitis (1-@ age 3days; 1@ age 5mo; 1@ age 6mo; 1@age 1yr; 7-age of contact not specified)	
	1	unknown (causing encephalitis)	1	viral infection (causing encephalitis @1.5yr)	
	1	unknown	1	unspecified infection (causing encephalitis @1.5yr)	
	1	congenital tuberculosis/epilepsy			
	6	scarlet fever			
	1	unspecified early childhood disease			
	1	measles and mumps			
	1	measles			
	1	whooping cough			
	2	childhood viral infections			
	1	fever (causing brain damage)			
	1	unspecified infection			

	2	work accidents	1	Shaken Baby Syndrome	
	1	car accident @age 26	4	accidents (1-near drowning @ age 1; 1-near drowning @ age 1.5; 1-near drowning @ age 2;	
	1	car accident @age 10+CP			
	1	accident @ age 10			
	1	accident @ age 6	1	1-head injury @ 2 mo)	
	1	blows to head @age 10	1	Sudden Infant Death	
	1	abuse @ age 30	1	Syndrome (SIDS)	
	1	brain damage @age 18	1	progressive neurological deterioration	
	1	accident @age 55			
	1	traumatic brain injury		liver septicaemia @ age 2	
	1	blood clot			
	1	operation for brain tumour			
	1	nothing specified			
	2	aphasia			
	1	brain haemorrhage			
Complications of Prematurity	**1**		**22**		**23**
	1	oxygen over-exposure (likely ROP)	1	CP	
			4	oxygen exposure (likely ROP)	
			4	retinopathy of prematurity (ROP)	
			1	spastic quadriplegic	
			12	nothing specified	
Metabolic Conditions	**15**		**2**		**17**
	13	diabetes (1@age 20; 12-age not specified.)	1	Progressive Metabolic Disease	
				diabetes @age 10mo	
	1	Morquio Syndrome	1		
	1	Pagets Disease			
Birth Trauma Causing Brain Damage	**2**		**13**		**15**
	1	anoxia at birth	3	birthing	
	1	nothing specified	3	hypoxia at birth	
			1	subarachnoid haemorrhage	
			1	epilepsy and multidisabled	
			3	nothing specified	
			1	Meconium aspiration	
			1	Syndrome	
			1	perinatal hypoxia	
Sexual Transmitted Diseases	**2**		**4**		**6**
	2	syphilis	1	herpes simplex (causing encephalitis)	
			3	syphilis	
Fetal Alcohol Syndrome			**3**		**3**
			1	Causing Brain Damage	
			2	Fetal Alcohol Syndrome (1 including cocaine)	
Mitochondrial Condition	**2**				**2**
	1	Kearns Sayre-Sky Syndrome			
	1	mitochondrial disease (non-specified)			

	1	Reported Prenatal Medication			
Complications from Medical Procedures	**2** 1 1	 tonsillectomy kidney dialysis			**2**
Other Causes	**5** 1 1 1 2	 bone marrow cancer influenza epilepsy self abuse	**2** 2	 epilepsy	**7**
Sub Total Known Causes	**331**		**284**		**615**
Unknown/ Unspecified Causes	**99**		**63**		**162**
Total	**430**		**347**		**777**

As is indicated in Table 5, not all respondents knew the cause of their deafblindness. Several respondents did not want to disclose this information for personal reasons. In total, 613 of 777 (78.9%) persons reported they knew the contributing cause to their deafblindness, which is the sample size used in the subsequent statistics.

According to Table 5, two categories of contributing factors, inherited rare disorders (35%) and intra-uterine infections (19.5%), together account for over half (54.5%) of the known causes contributing to deafblindness. In descending order, the following categories account for the remainder of the causes: congenital brain damage (8%), aging (8%), non-inherited chromosomal abnormalities (6.5%), post natal/early childhood infections (5.4%), acquired brain injury (5.2%), prematurity (3.6%), metabolic conditions (2.7%), birth trauma (2.3%), STD's (1%) and others grouped together (2.5%).

almost half (48.6%) of the reported causes of deafblindness (see Table 6). An explanation for each of the specific causes reported in Table 6 is found in the Appendix 5.4.

The relative proportion of the total reported causes of deafblindness in Table 5 takes on a different picture when the sample is examined according to whether the individuals reported having congenital or acquired deafblindness. It should be mentioned at this point that, with respect to individuals with Congenital Rubella, Alstrom and Cockayne Syndromes, some individuals have reported congenital deafblindness while others have reported acquired deafblindness (see Table 6). While these are congenital related causes, the manifestations may be acquired beyond the age criteria for congenital (i.e. by age 2), therefore they were assigned as acquired deafblindness.

2.4.1 Causes of Congenital Deafblindness

For the congenital group who reported a known cause of their deafblindness, intra-uterine infections (40.8%), congenital brain damage (16.6%) and non-inherited chromosomal abnormalities (14.1%) combined, accounted for over two-thirds or 71.5% of the reported known causes. In descending order, the following account for the remainder of the causes of deafblindness among the congenital group who reported a known cause: prematurity (7.7%), inherited rare disorders (4.2%), post-natal/early childhood infections (4.6%), birth trauma (4.6%), acquired brain injury (3.5%), sexually transmitted diseases (1.4%), Fetal Alcohol Syndrome (1.1%), and other causes (0.7%).

More specifically, congenital rubella syndrome, a congenital viral infection affecting the unborn fetus during the first trimester of pregnancy, represented the primary cause of deafblindness (40.4%) reported by those with congenital deafblindness (who knew their cause of deafblindness).

In Table 7, the major causes of congenital deafblindness have demonstrated considerable differences through the major birth periods. The most noticeable is with maternal rubella. The highest incidence of this condition occurred during the 1960's and 1970's, corroborating the period during which the major epidemic of rubella occurred in Canada and throughout the world. The incidence of rubella as a causative factor in deafblindness has shown a significant decline through the 1980's and 1990's. This major reduction in incidence of rubella is believed largely, to be the result of widespread immunization programs.

As the incidence of congenital rubella syndrome as being responsible for the cause of deafblindness decreased during the past two decades, the incidence of other conditions has increased.

The group of Chromosomal Abnormalities (see Table 7) stand out as showing an increase in incidence during the 1980's (11.2%) and 1990's (27.8%). Persons with Down Syndrome represents one third of the 35 persons reporting chromosome abnormalities. None of the persons with chromosomal abnormalities born during the 1990's had Down Syndrome.

Prematurity as the contributor to deafblindness showed the highest percentage (11.1%) during the 1990's. Congenital brain damage shows a higher incidence rate during the 1980's and 1990's as compared to the 1960's and 1970's. However, during the 1940's and 1950's the incidence of congenital brain damage have similar high rates of incidence as reported during the more recent decades.

The ‘Other category’ includes such causes as cytomegalovirus, meningitis, fetal alcohol syndrome, acquired brain injuries and various unknown causes. For the period 1980-1998, the incidence rate of the various known and unknown causes is shown as a footnote.

The findings in this study, that congenital rubella syndrome is no longer the most common cause of congenital deafblindness, has been reported similarly in the UK (Brown,1997) and the United States (Collins, Majors and Riggio,1991). Collins et. al reported that, by 1991 in four New England states, maternal rubella accounted for 15% of those students having deafblindness. In Brown’s study of 100 children with deafblindness in the UK, only two had reported rubella as the cause of their deafblindness.

causes of congenital brain damage. There is no substantive evidence relating environmental factors to the higher incidence of persons born with rare disorders or congenital defects. The higher incidence rate of children born with these rare disorders could relate to better post-natal assessment of the condition by medical personnel, better genetic testing and improved efforts to locate services for these children. As a result, many of these children are receiving services from organizations that have developed specialized programs for deafblindness.

Concern has been expressed that perhaps some of these individuals, despite their disability, may not truly fit the functional definition of deafblindness. This is an issue for professionals to examine in further studies.

	deafblindness	congenital rubella syndrome	prematurity	inherited rare disorders	non-inherited chromosomal abnormalities	congenital brain damage	
1900-1909	0						
1910-1919	3	1 (33.3%)				2 (66.7%)	
1920-1929	1						1 (100%)
1930-1939	5	2 (40%)					3 (60%)
1940-1949	26	3 (11.4%)	1 (3.9%)	1 (3.9%)	4 (15.4%)	4 (15.4%)	13 (50%)
1950-1959	42	14 (33.3%)	2 (4.8%)	1 (2.4%)	3 (7.1%)	8 (19.1%)	14 (33.3%)
1960-1969	65	37 (56.9%)	3 (4.6%)		2 (3.1%)	8 (12.3%)	15 (23%)
1970-1979	81	41 (50.6%)	7 (8.6%)	5 (6.2%)	3 (3.7%)	1 (1.2%)	24 (29.6%)
1980-1989	71	13 (18.3%)	3 (4.2%)	3 (4.2%)	8 (11.2%)	16 (22.4%)	28* (39.4%)
1990-1998	54	4 (7.4%)	6 (11.1%)	3 (5.5%)	15 (27.8%)	10 (18.5%)	16** (29.6%)
Total	**348**	**115**	**22**	**13**	**35**	**49**	**114**

*Other causes during the birth period 1980-89 include: birth trauma (1); metabolic disease (1); meningitis (3); accidents (3); brain damage unspecified (2); Alstrom Syndrome (1); liver septicaemia (1); Fetal Alcohol Syndrome (FAS-1); Shaken Baby Syndrome (1); Cytomegalovirus (CMV-2); unknown (12);

**Other Causes during the birth period 1990-98 include: CMV (2); recovered Sudden Infant Death Syndrome (2); Alstrom Syndrome(2); birth trauma (1); meningitis (1); FAS/Drugs (3); unknown (6).

2.4.2 Causes of Acquired Deafblindness

For the acquired group who reported a known cause of their deafblindness (Table 5), genetically inherited disorders (61.4%) and ageing (14.9%) together accounted for over three quarters or 76.3% of the reported known causes. In descending order, the following account for the remainder of the causes of deafblindness among the acquired group (who reported a cause): post-natal infections/early childhood infections (6%), acquired brain injury (6.6%), metabolic conditions (4.5%), inter-uterine infections (1.2%), and other causes (3.3%). Additional details about the specific causes or conditions for the acquired deafblindness can be found in Table 6.

The genetically inherited disorder Usher Syndrome was reported by 55.6% of persons with acquired deafblindness who reported the cause of their disability. It is suspected that some of those who reported an unknown cause of their deafblindness would have been people with Usher Syndrome. Many of these persons are elderly and have not been diagnosed with this condition. Usher Syndrome is said to account for between 4-6% of congenitally deaf children (William Green, personal communication, 2001). Some studies suggest that persons with Usher represent about two-thirds of the population of persons with acquired deafblindness.

Usher Syndrome has been identified as occurring in three types: Type 1: born deaf and acquiring vision loss within the first 10 years or early adolescence; Type 2: born with mild to severe hearing loss and acquiring vision loss from late adolescence to late 20's, and Type 3: acquiring both hearing and vision loss with advancing age. In this study very few indicated the Type of Usher Syndrome, but research has indicated (Joyce Thompson, personal communication, 2001) that 90% of the incidences are Type 1. This would suggest that of the 184 cases reported,

As the elderly component of the regular population rises, combined vision and hearing loss or deafblindness is expected to increase in Canada. Gradual loss of hearing is recognized as a condition of ageing. This, combined with such common eye problems as macular degeneration (a condition altering the vascular supply to the retina which often occurs after the age of 60), glaucoma (an eye disease affecting the optic nerve that often begins after age 55 and can progress slowly) and cataracts (opacity of the crystalline lens, a condition which occurs in the later years) can lead to the gradual loss of vision.

2.5 Information about Reported Hearing Loss

Respondents were asked to report the age of onset of their hearing loss, the degree (mild, moderate, severe or profound) of their hearing loss and also about the prognosis (fluctuating, progressive or stable) of their hearing loss. This information is reported, according to the classifications of deafblindness, respectively, in Tables 8, 9 and 10. It should be pointed out that sample sizes will vary from table to table since not all 777 individuals from the Registry responded to each question about hearing loss.

hearing loss	age of onset	Congenital Deafblindness	Acquired Deafblindness	Deaf/ Acquired Blindness	Acquired Blindness	Blind/ Acquired Deafness	Classific'n
0-2	542	347	195	195			
3-5	14		14		7	7	
6-9	25		25		18	7	
10-20	31		31		28	3	
21-29	15		15		8	2	5
30-39	12		12		8	3	1
40-49	12		12		8	3	1
50-59	10		10		8	2	
60-65	14		14		14		
66+	35		35		34	1	
Total sample size	**710**	**347**	**363**	**195**	**133**	**28**	**7**

All those who were congenitally deafblind (347) or were acquired deafblind: pre-lingually deaf with acquired vision loss (195) are shown in Table 8 as reporting the onset of their hearing loss in the 0-2 years period, as per the criteria of their condition.

For the rest of the sample (of individuals reporting age of onset of hearing loss), the major peak periods of onset of hearing loss occur during: childhood (3-9 years), adolescence to early adulthood (10-20) and during old age (66+years).

Table 9. Summary of responses: Degree of hearing loss

Degree of Hearing Loss	Total reporting	Total number with Congenital Deafblindness	Total number with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness	Acquired: Unknown Classific'n
Mild	63 (8.7%)	38 (12.5%)	25 (6%)	4	12	2	7
Moderate	120 (16.6%)	50 (16.4%)	70 (16.7%)	9	42	8	11
Severe	185 (25.6%)	84 (27.5%)	101 (24.2%)	21	64	13	3
Profound	355 (49.1%)	133 (43.6%)	222 (53.1%)	158	56	7	1
Total sample size Total	**723**	**305**	**418**	**192**	**174**	**30**	**22**

The criteria for the four categories of hearing loss are as follows: **mild:** 26-49dB loss; **moderate:** 41-70dB loss; **severe:** 71-90dB loss and **profound:** 91-110dB loss.

For those with congenital and acquired deafblindness, most persons reported their hearing loss as profound. For

congenital (pre-lingual) deafness. The acquired group also has a high number of elderly persons where deafness is a progressive issue.

The higher percentage of mild hearing loss among the congenital group might reflect the higher number of younger persons in this group whose hearing loss is not yet complete, as well as the prevalence of persons with rare disorders where hearing loss may not be as severe.

Table 10. Summary of responses: Prognosis of hearing loss

Prognosis of Hearing Loss	Total reporting	Total number with Congenital Deafblindness	Total number with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness	Acquired: Unknown Classific'n
Fluctuating	22 (3.6%)	19 (8.4%)	3 (0.8%)	2	1		
Progressive	159 (26.2%)	28 (12.3%)	131 (34.4%)	25	84	22	
Stable	427 (70.2%)	180 (79.3%)	247 (64.8%)	150	82	9	6
Total	**608**	**227**	**381**	**177**	**167**	**31**	**6**

An evaluation of the prognosis of hearing loss is reported in Table 10. Of the 608 persons who reported this information, the majority, (70.2%) reported their hearing loss was stable or unchanged in recent years; 26.2% reported their hearing loss was progressive or deteriorating, and only 3.6% reporting a fluctuating loss of hearing (deteriorating/improving).

Comparing congenital and acquired deafblindness in Table 10, those with congenital deafblindness reported a higher level of both fluctuating and stable hearing loss than the acquired group. Persons with acquired deafblindness showed a much higher level of progressive hearing loss than their congenital counterparts.

The reasons for any differences between congenital and acquired would be similar to the explanations posed for differences in degree of hearing loss in Table 9.

2.6 Information about Reported Vision Loss

Respondents were asked to report the age of onset of their vision loss, the degree (low vision, light perception only, legally blind or totally blind) of their vision loss and also the prognosis (fluctuating, progressive or stable) of their vision loss. This information is reported, according to the classifications of deafblindness, respectively, in Tables 11, 12 and 13. The criteria for congenital deafblindness and congenital blindness is that the condition occurs up to and including the age of two years. It should be pointed out that sample sizes will vary from table to table since not all 777 individuals from the Registry responded to each question about vision loss.

Table 11. Summary of responses: Age of onset of vision loss

Age of onset of vision loss	Total reporting	Total number with Congenital Deafblindness	Total number with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness	Acquired: Unknown Classific'n
0-2	380	347	33			33	
3-5	15		15	8	7		
6-9	20		20	9	11		
10-20	78		78	53	22		3

50-59	24		24	12	12		
60-65	10		10		10		
66+	60		60	6	54		
Total	**668**	**347**	**321**	**137**	**146**	**33**	**5**

All those who were congenitally deafblind (347) and those 33 persons with congenital blindness (and acquired hearing loss) are shown as having the onset of their vision loss during the 0-2 years period, as per the criteria of their condition.

For the remainder of the sample (of individuals reporting age of onset of vision loss), the major peaks occur during: the period of adolescence to early adulthood (10-20 years), the typical period when tunnel vision and night blindness impacts the ability of those with Usher Syndrome to function, and during old age (66+ years).

Table 12. Summary of responses: Degree of vision loss

Degree of Vision Loss	Total reporting	Total number with Congenital Deafblindness	Total number with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness	Acquired: Unknown Classific'n
Low Vision	175 (23.6%)	80 (24.8%)	95 (22.8%)	27	49	2	17
Legally Blind	448 (60.6%)	178 (53.3%)	270 (64.5%)	143	110	15	2
Totally Blind	117 (15.8%)	64 (19.9%)	53 (12.7%)	19	17	15	2
Total	**740**	**322**	**418**	**189**	**176**	**32**	**21**

The criteria for the categories of vision loss are as follows: **low vision:** central visual acuity 20/70 to 20/200; **legally blind:** central visual acuity is greater than 20/200 or visual field is less than 20 degrees; **totally blind:** absence of vision. Ninety-five percent or 740 persons responded to the question about degree of vision loss in Table12.

Less than one quarter of the sample of respondents indicated they had low vision. Just over sixty percent (60.6%) reported they were legally blind, while 15.8% indicated they were totally blind. Many respondents may not have discriminated between legally blind and totally blind. For this reason, any comments about the differences between the two groups, congenital and acquired, is not warranted.

Table 13. Summary of responses: Prognosis of vision loss

Prognosis of Vision Loss	Total reporting	Total number with Congenital Deafblindness	Total number with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness	Acquired: Unknown Classific'n
Fluctuating	18 (2.8%)	14 (5.5%)	4 (1%)	1	2		1
Progressive	326 (50.7%)	56 (22%)	270 (69.6%)	146	109	13	2
Stable	299 (46.5%)	185 (72.5%)	114 (29.4%)	32	56	18	8
Total	**643**	**255**	**388**	**179**	**167**	**31**	**11**

tests are not measured each year.

Comparing congenital and acquired deafblindness in Table 13, those with congenital deafblindness had higher incidences of both Fluctuating and Stable vision loss. With respect to the Progressive Vision Loss category, those with acquired deafblindness showed a much higher rate, which largely reflects vision loss progression of those with Usher Syndrome. (Joyce Thompson, personal communication)

The higher rate of progressive vision loss among the acquired group reflects ageing and the advancing loss of vision among those with Usher Syndrome. The higher rate of stable vision loss among those with congenital deafblindness reflects the etiology of this condition. Most of the causes of congenital deafblindness affect the visual system early in the individual's life, and are less likely to be of a progressive nature.

2.7 Miscellaneous Information Reported by the Respondents

The questionnaires distributed in both phases of this Project asked many different questions of the participants. The area of questioning included assistance (types and extent), personal status (working, education, etc.), communication, special aids and lifestyle. The remainder of this report is devoted to this portion of the project.

2.7.1 Types of Assistance

Respondents were asked to indicate whether they received assistance from a wide range of possibilities, including Intervenor, interpreter, volunteer, friend, driver/guide, family member, institutional care giver. Respondents were not limited to indicate one type of assistance, thus more than one type of assistance was indicated by many of the respondents. Table 14 summarizes the responses to the question about the type of assistance. The percentages listed in column 'number reporting type of assistance' is calculated using sample size 697, since 80 persons did not reply to this question. Percentages are calculated for congenital and acquired deafblindness, but not for each classification of the acquired group.

Table 14. Summary of responses: Types of assistance received

Type of Assistance	Total reporting type of assistance	Total number with Congenital Deafblindness	Total number with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness	Acquired: Unknown Classific'n
Intervenor	384 (55.1%; n=697)	201 (52.3%; n=384)	183 (47.7%; n=384)	103	63	17	0
Interpreter	124 (17.8%; n=697)	16 (12.9%; n=124)	108 (81.1%; n=124)	81	23	4	0
Volunteer	110 (15.8%; n=697)	8 (7.3%; n=110)	102 (92.7%; n=110)	62	32	8	0
Friend	106 (15.2%; n=697)	11 (10.4%; n=106)	95 (89.6%; n=106)	48	39	8	0
Driver / Guide	68 (9.8%; n=697)	2 (1.9%; n=68)	66 (98.1%; n=68)	44	16	6	0
Family	66 (9.5%; n=697)	9 (13.6%; n=66)	57 (86.4%; n=66)	22	32	3	0
Institutional Care	177 (25.4%; n=697)	107 (60.5%; n=177)	70 (39.5%; n=177)	22	24	2	22
Other care givers	10 (1.4%;	5 (50%; n=10)	5 (50%; n=10)	2	2	1	0

Did not Respond	80 (10.3%; n=777)	30 (37.5%; n=80)	50 (62.5%; n=61)	18	28	4	0

The most interesting and troubling statistic in Table 14 is that less than ten percent (8.8%) indicated receiving no assistance. However, it was not clear from the reports whether these 61 individuals could not get assistance, did not know where to seek assistance, or believed that they did not require assistance. Most, (72.1%) of those indicating they received no assistance, had acquired deafblindness. Further, it is unknown whether those who did not respond (80), simply missed the question, or possibly, fit the category of receiving no assistance at all.

Upon examining the congenital versus acquired statistics in Table 14, it can be observed that those with acquired deafblindness receive more assistance from interpreters (81.1%), volunteers (92.7%), friends (89.6%), driver/guide (98.1%) and family members (86.4%), than did those with congenital deafblindness. Those, with the exception of interpreters, are considered volunteer services. The subject of Intervenor assistance will be discussed in more detail in the following section.

What stands out in Table 14 is that 25.4% of respondents who reported receiving assistance, received it in the form of institutional care. For the purpose of this particular discussion, institutional care refers to group homes, institutions for individuals who are mentally disabled, and nursing homes. The majority of those receiving this form of assistance (60.5%), were those with congenital deafblindness. It should also be pointed out that Nursing Homes provide nursing care, and in the vast majority of cases, do not provide auditory or visual information assistance to the person with deafblindness.

2.7.1.1 Intervenor Services

The Intervenor forms a vital link with the person with deafblindness for the purpose of communication and interpreting the environment.

For the purpose of this discussion, Intervenor Services include services offered by accompagnateurs and Intervenors. An accompagnateur is the name, in Quebec, of the individual who assists persons who are deafblind with shopping, social and leisure activities, sports and medical appointments (Gilles Lefebvre, personal communication, 2000). An Intervenor is the name, in the rest of Canada, of the individual who serves as “the eyes and ears” of persons who are deafblind and provides Intervention or Intervention Services. The major difference between the two is that communication is not the focus of the accompagnateur, whereas it is the main focus of an Intervenor.

According to Table 14, Intervenors provide assistance to only 55.1% of those who responded to the question about types of assistance. Intervenor services were reported as being used by 52.3% and 47.7% of those who were congenitally deafblind and acquired deafblind, respectively.

The utilization of Intervenor Services by persons on the Registry is summarized in Table 15. This information serves as providing a very cursory review of the utilization of Intervenor Services by persons with deafblindness across Canada. The rates of utilization of Intervenor Services reflects availability of this service, and Table 15 indicates the wide range of availability across Canada and the differences in availability within the congenital population and the acquired population.

	on Registry	Intervenor Services	Deafblindness	Deafblindness receiving Intervention	Deafblindness	Deafblindness receiving Intervention
British Columbia	93	45 (48.4%)	48	32 (66.6%)	45	13 (28.9%)
Alberta	39	11 (28.2%)	16	2 (12.5%)	23	9 (39.1%)
Saskatchewan	14	13 (92.9%)	12	12 (100%)	2	1 (50%)
Manitoba	38	29 (76.3%)	24	22 (91.7%)	14	7 (50%)
Ontario	361	186 (51.5%)	205	112 (54.6%)	156	74 (47.4%)
Quebec	168	78 (56.4%)	11	7 (63.6%)	157	71 (45.2%)
Prince Edward Island	4	2 (50%)	4	2	0	0
New Brunswick	21	10 (47.6%)	10	6 (60%)	11	4 (36.4%)
Nova Scotia	30	8 (26.7%)	12	4 (25%)	18	4 (22.2%)
Newfoundland	9	2 (22.2%)	5	2 (40%)	4	0 (0%)
Total	**777**	**384 (55.1%)**	**347**	**201 (52.3%)**	**430**	**183 (47.7%)**

Additional details on Intervenor Services were obtained from Project Phase 2 participants. These further details about Intervenor Services are summarized in Table 16.

Table 16. Summary of additional details on Intervenor services

Number of hours of Intervention Services accessed per week	Total reporting	Total number with Congenital Deafblindness	Total number with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness
1 to 5	38 (28.1%; n=135)	7 (18.4%; n=38)	31 (81.6%; n=38)	15	13	3
6 to 10	26 (19.3%; n=135)	7 (26.9%; n=26)	19 (73.1%; n=26)	3	11	5
11 to 20	13 (9.6%; n=135)	5 (30.8%; n=13)	8 (69.2%; n=13)	7	0	1
21 to 40	32 (23.7%; n=135)	22 (68.8%; n=32)	10 (31.2%; n=32)	5	3	2
40+	26 (19.3%; n=135)	26 (100%; n=26)	0	0	0	0
Number not reporting number of hours	15	2	13	7	6	0
Total Receiving Intervention Services	150 (67%; n=224)	69 (46%; n=150)	81 (54%; n=150)	37	33	11

with Intervention hours						

Of the 224 persons participating in Phase 2, 150 or 67% indicated they received Intervenor Services. Slightly more, (54%) who reported receiving those services, were persons with congenital deafblindness.

Table 16 demonstrates wide differences between the acquired and congenital deafblind groups with respect to the number of hours Intervenor services were utilized. For those 77 persons reported receiving 1-20 hours per week, fifty-eight or 75.3% were individuals with acquired deafblindness. Nineteen or 24.7% of those receiving this level (1-20 hours) of Intervenor Services were congenitally deafblind. Persons with congenital deafblindness were the prime recipients of Intervenor services at 21+ hours per week. Those persons who received 40+ hours per week are extremely high need, congenitally deafblind persons, who live in various Independent Living Residences for Deafblind in Ontario, Manitoba, Saskatchewan and British Columbia.

2.7.1.2 Alternative Forms of Assistance

Respondents to Phase 2 of the project were asked to supply information about alternative forms of assistance (referred to in the Questionnaire as Methods of Travel). Their responses are summarized in Table 17. Of the 224 persons responding to Phase 2, 194 or 86.6% indicated receiving alternative forms of assistance other than from Intervenors and Interpreters. Of those indicating this other assistance, 64.9% had acquired deafblindness and 35.1% had congenital deafblindness.

Table 17. Summary of responses to "Alternate forms of assistance"

Alternate Forms of Assistance	Total reporting	Total number with Congenital Deafblindness	Total number with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness
Using Sighted Guide (not including Intervenor)	63 (32.5%; n=194)	33 (42.3%; n=63)	30 (57.7%; n=63)	15	10	5
Cane Travel	72 (37.1% n=194)	7 (9.9%; n=72)	65 (90.1%; n=72)	28	28	9
Guide Dog	12 (6.2%; n=194)	1 (8.3%; n=12)	11 (91.7%; n=12)	3	5	3
Uses Remaining Vision	94 (48.5%; n=194)	36 (38.3%; n=94)	58 (61.7%; n=94)	24	31	3
Other*	10 (5.2%; n=194)	4 (40%; n=10)	6 (60%; n=94)	1	3	2
Total responding to using alternative forms of assistance	194 (86.6%; n=224)	68 (35.1%;n=194)	126 (64.9%;n=194)			
Did not respond	30 (13.4%; n=224)	17 (56.7%; n=30)	13 (43.3%; n=30)	6	7	0

*The ten other were: wheel chair (7); wheel-trans (1); handi-cart (1) and taxi (1).

2.8 Personal Status

Respondents were asked to provide information about their personal status regarding education, working situation, whether they were retired, receiving medical care, etc. Respondents were not limited to one answer. Their responses are summarized in Table 18. Further discussion about education and working is discussed in separate sections.

Table 18. Summary of responses regarding personal status

Personal Status	Total reporting	Total number with Congenital Deafblindness	Total number with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness	Acquired: Unknown Classifc'n
Currently in School	149 (19.2%; n=777)	120 (80.5%; n=149)	29 (19.5%; n=149)	19	3	7	0
Reported Finishing School (various levels)	201 (25.9%; n=777)	82 (40.8%; n=201)	119 (59.2%; n=201)	57	53	9	0
Working (Competitive and self-	51 (6.6%; n=777)	13 (25.5%; n=51)	38 (74.5%; n=51)	27	8	3	0

	(21.6%;n=777)	(69%; n=168)	(31%; n=168)				
Homemaker	8 (1%; n=777)	1 (12.5%; n=8)	7 (87.5%; n=8)	3	4	0	0
Homebound	4 (0.5%; n=777)	1 (25%; n=4)	3 (75% n=4)	0	2	1	0
Retired	30 (3.9%; n=777)	0	30 (100%; n=30)	9	20	1	0
Receiving Medical Care	5 (0.6%; n=777)	2 (40%; n=5)	3 (60%: n=5)	0	2	1	0

2.8.1 Education

According to Table 18, 201 or 25.9% reported completing some form of education. Additional statistics about educational history are found in Table 19 and statistics about current educational activities are found in Table 20. Referring first to Table 19, one hundred and forty-nine (149) or 19.2% of respondents were currently in an education program. Seven persons indicated they had received no formal schooling and 411 did not respond to this question.

Table 19. Summary of statistics about education

Specified School Program	Number
Currently attending public or post-secondary school (see Table 20)	149
Completed some public school but not High School	20
Completed Deafblind Programs: •WRMS, Brantford, ON (36) •APSEA, Amherst, NS (12) •Saskatchewan (3)	51
Attended Schools for Deaf: •E.C. Drury , Milton, ON (3) •Sir James Whitney, Belleville, ON (2)	5
Completed High School Programs: •HS - not specified (17) •Grade 11 (1) •Grade 12 (32) •Secondary II (1) •Secondary III (2) •Secondary IV (1) •Secondary V(5)	59
•Completed home schooling	1
Completed Specialized School Programs: •High School Life Skills (1) •Special Education (2) •Community Living Skills (1)	4
Completed Post Secondary Education: •University-not specified (14) •University-Post Graduate (3) •Community College (11) •Technical School (5)	33
•Completed School - not specified	28
•Received no formal education	7

It is difficult to make any conclusive statements about these statistics on education because of the difficulty with evaluating the responses to the different questionnaires. Questions about education were phrased in different ways on the questionnaires. The CDBRA questionnaire asked if the person completed school; the CNSDB questionnaire asked if the person was in school or not. The statistics on school completion was largely gleaned from the respondents who completed the Phase 1 CDBRA questionnaire supplemented by information supplied by those individuals who completed the Phase 2 questionnaire. Thus it is suspected that many more of the respondents completed some aspect of an education program than the above statistics suggest. According to Joyce Thompson (personal communication, 2001) many individuals with acquired deafblindness have very few years of formal education; many are self taught.

Some specifics on the type of education program currently attended by the 149 persons who reported they were still in school is found in Table 20. As indicated in Table 18, over eighty percent (80.5%) of those currently attending school have congenital deafblindness. This is attributed to the fact that most persons of school age with deafblindness are congenitally deafblind (Table 4).

Table 20. Specifics on current educational programs attended by persons with deafblindness

Type of School Program	**Number**
Public School - nothing specified	57
Public School - special classes	10
Special Deafblind Programs •Evan Hardy Public School, SK (3) •Other Public School (5) •WRMS (13)	21
Schools for Deaf •E.C. Drury, Milton, ON (4) •Newfoundland School for the Deaf (1)	5
•Pre-school programs	9
•Home schooling	3
•University	2
•Community College	2
•Did not specify	40
•**Total currently in school programs**	•**149**

2.8.2 Working

According to Table 18, some 81 persons or 10.4% of the total respondents indicated they were working. Of those, 51 were working in competitive employment, while 30 were in sheltered programs. These statistics will be a little more meaningful if the number of persons who are of school age and younger, born after 1980 (149), and those seniors older than 65 years, born before 1936 (166), are removed from the sample size used to calculate percentage of persons working. Therefore, based on a revised sample size of 462, the percentage of persons with deafblindness who are working now becomes 17.5%.

In Phase 2 of the project, persons were asked a number of questions about working, such as: were they working in competitive or sheltered work environments? Were they actively looking for work? Did they work or was their work of a volunteer nature? This subset of information on working is summarized in Table 21. Out of the original sample size of 224, fifty-seven were removed from this calculation, twenty five who were in school and thirty-two who are retired.

Table 21. Additional details about working from a sample of persons with deafblindness

Status of Working	Total reporting	Number with Congenital Deafblindness	Number with Acquired Deafblindness	Pre-Lingual Deaf/ Acquired Blindness	Post-Lingual Deaf/ Acquired Blindness	Congenital Blind/ Acquired Deafness
Working (competitively or self-employed)	28 (16.8%; n=167)	5 (17.9%; n=28)	23 (82.1%; n=28)	11	9	3
Sheltered Workshop	7 (4.2%; n=167)	2 (28.6%; n=7)	5 (71.4%; n=7)	3	2	0
Looking for Work	18 (10.8%; n=167)	9 (50%; n=18)	9 (50%; n=18)	6	3	0
Volunteering	33 (19.8%; n=167)	12 (36.4%; n=33)	21 (63.6%; n=33)	13	6	2

2.9 Modes of Communication

One of the significant attributes that the survey was trying to determine was the various modes of communication that persons with deafblindness were using. Individuals were asked to indicate all the modes of communication that they used. The responses to questions regarding communication modes used, is summarized in Table 22. The rate or percentage of response, for use of each mode was calculated using the sample size of 710, the number who responded to this question.

Some caution must be used when interpreting these results. The question about modes of communication was not phrased to distinguish between expressive and receptive use of a particular mode of communication. The rate or percentage of response for use of each mode may lead one to believe that the person uses a particular mode both ways, but this may not always be the case.

Table 22. Summary of responses: Modes of communication

Modes of * Communication	Total reporting	Total number with Congenital Deafblindness	Total number with Acquired Deafblindness	Acquired: Pre-Lingual Deaf/ Acquired Blindness	Acquired: Post-Lingual Deaf/ Acquired Blindness	Acquired: Congenital Blind/ Acquired Deafness	Acquired: Unknown Classific'n
Writing/Printing	330 (46.5%; n=710)	68 (20.6%; n=330)	262 (79.4%; n=330))	108	143	11	0
Sign Language: ASL and/or Signed English	418 (58.9%; n=710)	141 (33.7%; n=418)	277 (66.3%; n=418)	146	24	7	0
Adapted Sign Language	25 (3.5%; n=710)	20 (80%; n=25)	5 (20%; n=25)	4	0	1	0

Gestures	179 (25.2%; n=710)	150 (83.8%; n=179)	29 (16.2%; n=179)	14	7	2	6
Speech	247 (34.8%; n=710)	59 (23.9%; n=247)	188 (76.1%; n=247)	35	133	20	0
Tellatouch	13 (1.8%; n=710)	1 (7.7%; n=13)	12 (92.3%; n=13)	5	4	3	0
Audio tapes	28 (3.9%; n=710)	2 (7.1%; n=28)	26 (92.9%; n=28)	5	13	8	0
Other skills - Unspecified	194 (27.3%; n=710)	90 (46.4%; n=194)	104 (53.6%; n=194)	67	30	6	1
TTY	129 18.2%; n=710)	29 (22.5%; n=129)	100 (77.5%; n=129)	76	19	5	0
Did not respond	67	47	20	3	16	1	0

* Definitions of each mode of communication is found in Appendix 5.5

The statistics in Table 22 suggest that persons with acquired deafblindness tend to have a higher rate of use of using writing/printing, sign language (ASL and Signed English), manual alphabet, and speech, than those with congenital deafblindness. This is related to the ability to use either the auditory or visual sense to develop a good language base, which was more prevalent in the individual before acquiring deafblindness.

Those with congenital deafblindness, instead, use gestures at a much higher level instead of other communication modes. This confirms that the congenital population experience serious difficulties with most communication modes because of the level of their disability. One should put in context, also, that many of those with congenital deafblindness are younger children and, many have not yet mastered some of these modes of communication used by those with acquired deafblindness.

Additional details about communication were obtained from Phase 2 of the project. These further details are summarized in Table 23.

Of those 224 persons who responded to Phase 2 of the project, 183 or 81.7% reported using at least one specialized aid to communicate. The vast majority (73.2%) of those reporting use of various communication aids or devices, are those with acquired deafblindness. Most of the 41 individuals who did not respond (to this part of Phase 2) were, in fact, those with congenital deafblindness, whose communication abilities are limited in not being able to utilize the specialized communication aids. Those 41 persons were removed from the calculations of the rate of use of the various communication modes in Table 23.

		Deafblindness	Deafblindness	Deaf/ Acquired Blindness	Lingual Deaf/ Acquired Blindness	Blind/ Acquired Deafness
Hearing aid	115 (62.8%; n=183)	35 (30.4%; n=135)	80 (69.6%; n=135)	17	52	11
FM/Infrared System	31 (16.9%; n=183)	13 (41.9%; n=31)	18 (58.1%; n=31)	4	10	4
Cochlear Implants	2 (1.1%; n=183)	0	2 (100%; n=2)	0	1	1
Magnifiers	70 (38.3%; n=183)	12 (17.1%; n=70)	58 (82.9%; n=70)	26	31	1
Telebraille	13 (7.1%; n=183)	1 (7.7%; n=13)	12 (92.3%; n=13)	7	1	4
Teletouch	12 (6.6%; n=183)	0	12 (100%; n=12)	7	4	1
Computer	59 (32.2%; n=183)	15 (25.4%; n=59)	44 (74.6%; n=59)	20	18	6
Internet	33 (18%; n=183)	4 (12.1%; n=33)	29 (87.9%; n=33)	15	9	5
CCTV reader	50 (27.3%; n=183)	4 (8%; n=50)	46 (92%; n=50)	19	25	2
Special Glasses	62 (33.9%; n=183)	17 (27.4%; n=62)	45 (72.6%; n=62)	22	21	2
Monoculars	10 (5.5%; n=183)	3 (33.3%; n=10)	7 (66.7%; n=10)	1	5	1
Brailler	24 (13.1%; n=183)	3 (12.5%; n=24)	21 (87.5%; n=24)	10	3	8
Slate & Stylus	21 (11.5%; n=183)	0	21 (100%; n=21)	9	3	9
Closed Caption Decoder	36 (19.7%; n=183)	5 (13.9%; n=36)	31 (86.1%; n=36)	23	7	1
TTY	47 (25.7%; n=183)	10 (21.3%; n=47)	37 (78.7%; n=47)	29	6	2
Did not respond	41	36	5	3	1	1

*Definitions of each specialized aid to communication is found in Appendix 5.6

2.10 Living Arrangements

Respondents were asked to indicate their living arrangement. The choices on the questionnaires were: if they were children, did they live with their birth, foster or adoptive family; if they were adults, did they live independently, live in provincial facilities, in supported independent living homes, or in independent living facilities. The responses in Table 24 are grouped by category of living arrangement and by type of deafblindness.

Table 24. Details on reported living arrangements organized by type of deafblindness

Type of Living Arrangement	Number	Number with Congenital Deafblindness	Number with Acquired Deafblindness	Locations Reported	
Lives Independently	266 (34.3%; n=777)	19 (7.1%; n=266)	247 (92.9%; n=266)	Lives with family members	99
				Lives in own apartment	6
				Lives in boarding house	3
				Lives in private apartments (Rotary Cheshire Apt)	16
				Lives in facility for deaf	3
				Lives in facility for deafblind	
				Lives in school or University dorm	17
				Lives in religious	2

Facility	(15.5%; n=777)	(66.7%; n=120)	(33.3%; n=120)	(Ontario) Rideau Regional Centre (Ontario) Manitoba Developmental Centre	 6 17
Supported Living Facilities**	66 (8.6%; n=777)	65 (98.5%; n=66)	1 (1.5%; n=66)	Independent Living Residences for Deafblind Persons CDBRA-ON CDBRA- SK Intervention Manitoba Lions McInnes House Regional Residential Support Services (NS) Small Options Group Home (NS) LaHave Manor (NS) WINGS Deafblind Residential Housing (BC) Apartment in parents home Non-specified Group Home	 15 8 11 5 13 3 1 1 1 4 3 1
Supervised* Living Facilities**	43 (5.5%; n=777)	25 (58.1%; n=43)	18 (41.9%; n=43)	Community Living Home Other Group Home Group Home for Deaf (PQ) Apartment in parent's home Shared apartment Special Care Centres Bloorview MacMillan Centre Not specified	8 16 1 1 1 10 1 1
Retirement Home	66 (8.5%; n=777)	1 (1.5%; n=66)	65 (98.5%; n=66)	Manoir Cartierville Residence Louis Hebert Pavillon St. Dominique Pavillon Mille Fleurs Richmond Manoir Little Mountain Place Residence Beausejour Residence aux 3 Pignols Manoir Sully Willowdale Personal Care Home Seniors Home for Deaf CHSLD Bob Rumball Centre for Deaf Not specified	25 3 1 1 1 1 1 1 1 1 1 1 12 16
Living with Adoptive Parents or in Foster Care	29 (3.7%; n=777)	26 (89.7%; n=29)	3 (11.3%; n=29)	n/a	

				Portneuf Not specified	1 2
Total	777	347	430		

*44 indicated they were born before 1975
** Supported means with Intervention services
***Supervised means without Intervention services

The distribution of where people live is self explanatory and needs little discussion. The interesting part of this table is with respect to the difference in living arrangements between the congenital and acquired groups. A few observations are noted:

- Most of the individuals living with their birth families (71.4%) are congenitally deafblind, but this should be no surprise considering most young persons who are deafblind have this condition. An interesting statistic is that 44 of the persons living with their birth parents were over 25 years of age. Twenty one of these 44 individuals were congenitally deafblind.
- While only 29 were living with adoptive or foster families, most of these persons (26 or 89.7%) were congenitally deafblind.
- For those living in retirement homes, all but one had acquired deafblindness.

For those living in supported living facilities, all but one were high needs individuals with congenital deafblindness.

- The vast majority of those living independently (92.9%) had acquired deafblindness.

2.11 Marital Status

All Registry participants were asked to indicate their marital status as single, married, divorced (or separated) or widowed. Information reported on marital status of the respondents is summarized in Table 25.

Table 25. Summary of personal marital status of respondents to registry

Marital Status	Total number of respondents born after 1980	Total number of respondents born before 1980	No. with congenital deafblindness	No. with acquired deafblindness
Single	150	450 (72%; n=627)	214 (97.3%; n=220)	236 (58% n=407)
Married		112 (18%; n=627)	3 (1.4%; n=220)	109 (26.8%; n=407)
Widowed	0	55 (9%; n=627)	2 (0.9%; n=220)	53 (13%;n=407)
Divorced	0	10 (1%; n=627)	1 (0.4%; n=220)	9 (2.2%; n=407)
Total	**150**	**627**	**220**	**407**

To make it more appropriate for comparison, the respondents were separated into those born before1980 (627) and those born after 1980 (150), since it was assumed that the former had a greater likelihood, because of their age, of establishing a partnership or marriage. Using the sample size of 627 (i.e., those born before 1980), 72% of the respondents reported being single; 18% married; 9% widowed and 1% divorced. The implications of these statistics point to the difficulty that persons with deafblindness have in achieving a relatively 'normal' lifestyle. The low percentage of those having established a partnership (28%) reflects many things, including the extent of their disability limiting relationships, their relative isolation, etc. However, it was not clear from the respondents whether some of those who indicated being single had been previously married.

The marital status was distinguished between whether the person had congenital or acquired deafblindness. There are stark differences between these groups regarding the percentages who are single versus those who had

3.0 DISCUSSION

3.1 How well did the project achieve its goal and objectives?

The overall purpose or goal of this project was to establish a voluntary Registry of Persons with Deafblindness in Canada. Additional objectives of establishing the National Registry were to facilitate contact with previously identified persons with deafblindness; locate persons previously unidentified as having deafblindness; establish a means of communication with persons who are deafblind and also have access to vital demographic information from the Registry data.

- This project achieved its overall goal which was to establish a voluntary Registry of Persons with Deafblindness in Canada. The project was successful in meeting the four additional objectives of the National Registry project; some satisfactorily, others not so satisfactory.

- The project was able to attract 777 participants from across Canada. The vast majority (86.4%) of these individuals had been previously identified as having the disability and were receiving some level of service from an organization. Unfortunately not all those who were informed of the study, wished to participate.

- The objective of locating individuals not previously identified with deafblindness, was not well achieved. In the Province of Ontario we were able to locate a large number of persons from Provincial Institutions who were not previously identified with the disability. They made up 106 or 13.6% of the total voluntary participation. An additional 65 were reported living at another Ontario Provincial facility, but Registry information was not received. We expect there are many others not identified, including those living in various group homes and elderly persons living in nursing facilities.

- As a means of establishing communication with 777 in the deafblind community, their families and/or caregivers who participated in the study, the National Registry project was successful. The key is to maintain an up-to-date Registry data base to enable periodic communication.

- It is with respect to the last objective of the study, that of collecting vital demographic information, that the project achieved its best success. The study did an excellent job at collecting an extensive array of vital information from the participants, including basic demographics such as age and gender; an exhaustive review of the varied causative factors of the disability together with obtaining a basic understanding of individual lifestyle, marital status, education experiences, and work history. While it has satisfactorily determined some basic information about communication and assistance, it has only begun to scratch the surface regarding the populations' need for services. In all fairness, the study's purpose and level of funding did not permit the study of services and service needs that is required for this disability group.

3.2.1 Population Complexity

It would not be possible to stereo-typically define a person with deafblindness. As the data reveals in this study, deafblindness is an incredibly complex disability. The causes of the condition are diverse as are the consequences of the disability.

The relative progress that persons (with deafblindness) achieve in their life, including their quality of life, is highly dependent upon the time of onset of the disability. For some persons, the various attributes of the disability begin at birth or within their first two years of life (congenital deafblindness); while for most others the features of their disability are acquired and worsen throughout life (acquired deafblindness). Within this latter group, many persons acquire one form of the disability early (e.g. deafness) in life, then progressively lose the other. Others are born blind and acquire hearing loss later in life. The majority, which is within the growing Seniors population, lose their vision and hearing (sometimes together, often at different times) much later in life.

Therefore, whether the person with deafblindness was born that way (congenital) or whether they acquired their disability early or later in life, has much to do with determining the language and communication skills each person develops, the lifestyle they can achieve, the extent of medical and rehabilitation care they require, the kind of education programs they can attend, the level of independence they can achieve and/or the type of care and Intervenor services they require.

Whatever the cause of the disability, whatever the type of deafblindness and its manifestations, a common aspect of this disability, and not often discussed, are the psycho-social consequences experienced by the individual. Common challenges faced by this group are extreme isolation, loneliness, lack of self esteem and absence of purpose to their lives. As an example of isolation, see copy of letter from a senior in Appendix 5.2.2.

3.2.2 Classifications of Deafblind People in Canada

It was not possible from this study of 777 registrants to construct an accurate representative breakdown of the population of deafblind persons in Canada. In this study, 347 or 44.7% of the participants were congenitally deafblind, while 430 or 55.3% were acquired deafblind. This high proportion of the congenital component is due, in part, to the increased awareness (especially of the younger ages) and level of services for persons with congenital deafblindness in Canada. The lower than anticipated representation of the acquired group is indicative of the opposite effects, that is, lack of awareness and available services.

Current statistics from Norway indicate that 23.5% of its known deafblind population were congenitally deafblind (Oystein Stette, personal communication, 2001). There were no easily comparable statistics from the UK study (Lewin-Leigh, personal communication, 2001).

In this Canadian study, those born pre-lingually deaf with acquired vision loss accounted for just over 25% (195 of 777) of the total sample (see Table 5). Other studies have shown that this component represents one-third to one-half of persons with deafblindness (William Green, personal communication, 2001). Those who were born blind with acquired deafness represent 4% of the total Canadian sample, which is similar to other studies.

It is the elderly component that our study vastly under-represents. In this study, people with deafblindness over the age of 65 represented 21.6% of the total. Other studies suggest that ageing related deafblindness represents almost half of the population with the disability. The UK study indicates that two-thirds of its population of persons with deafblindness were over 65 years.

3.2.3 Population Size Estimate

Earlier in the report it was stated that, collectively, organizations working with persons who are deafblind knew of about 1725 persons with this disability**.** Based on Canada's population of 31 million, this would represent a deafblindness index of 4.8/100,000. The Task Force Report (1984) indicated that service providers knew of about 830 persons with deafblindness in Canada. That report estimated that at that time, the Canadian population of persons with deafblindness was between 1500 and 2000, for an index of between 6 and 8/100,000.

What is a proper Canadian population of persons with deafblindness? It is definitely above the level of 1725 (current numbers of people known; Table 2) and believed to be well below the UK estimates. A safe estimate would be between 10 to 15/100,000, for a population of 3,100 to 4,650 persons.

3.2.4 The Need for Identification and Assessment

Some believe that this estimate of 3,100 to 4,650 persons with deafblindness underestimates the extent of the disability in Canada. The challenge ahead is to obtain a better estimate, and, if that happens, those with this disability will be well served. In the meantime, what was stated almost two decades ago (Task Force, 1984) still applies in the new century, that only through accurate assessment of the population of persons with deafblindness can appropriate services be planned and delivered. Governments and service delivery organizations must know who their potential clients are, where they live, and what their needs are.

Better identification and assessments are the key. The people with the disability must be located and consistent assessment tools must be implemented. Professionals, with advice from deafblind persons, must establish rigorous definitions that account for any differences that may occur between functional and medical definitions of deafblindness. It should also be recognized that differences exist between provinces regarding the thresholds of hearing and visual loss (medical definition) which constitute services for deafblindness. In Quebec, for example, they use the medical rather than the functional definition and the criteria is that deafblind services will be provided if the individual has a vision loss of 20/70 and hearing loss 25dB or greater (Alain Levesque, personal communication, 2000).

Another instance needing deliberation is whether the functional definition continues to be applicable to all individuals identified with congenital deafblindness. The changing face of the congenital population resulting from the emergence or identification of children with visual and hearing impairments due to chromosomal disorders, brain damage and prematurity is causing some to question the current applicability of the functional definition. The definition in place was developed almost three decades ago when congenital rubella syndrome was the leading cause of congenital deafblindness. The definition was subsequently accepted by those professionals working with the acquired population and adopted as the Canadian definition. Some questions have come to the forefront about the continued appropriateness of applying the current functional definition of deafblindness to many of the younger individuals identified as congenitally deafblind; according to some professionals (including David Brown, 1997), visual and hearing impairment of some of these severely disabled children may be “the least of their problems”, and presumably not the primary factor limiting their acquiring communication skills.

Another critical issue is the need to recognize the extent of the growing problem of deafblindness within the senior population. In this study, seniors are believed to be vastly under-represented. The UK study recognized that two-thirds of their deafblind population are seniors, who acquired deafblindness due to ageing. It is very likely that similar statistics exist in Canada, meaning a population of 2,000-2,500 seniors with deafblindness, based on the suggested population ratio as 10-15/100,000 persons. Furthermore, this number is expected to rise significantly in the years ahead, if the trend in population demographic changes continue as expected.

Increased sensory loss within the senior population is a given fact. According to 1997 statistics from the UK Institute of Hearing Research (William Green, personal communication, 2001), 70% of persons over the age of 60 have some degree of hearing loss; and 10% of persons over the age of 60 are blind. Furthermore, other European research indicates that the deafblindness index within the population of seniors (over 60 years of age) is 150 per 100,000. The population of seniors in Canada (aged 65 and older) is predicted to represent twenty-three percent (or 8.3 million) of an anticipated population of 36.2 million by 2026 (Source: Statistics Canada). If we apply the European deafblindness ratio (150/100,000 for those over 60 years of age) to the Canadian population, there could be upwards of 12,450 older deafblind people in Canada in the next twenty five years. Governments and service agencies must be prepared for this and be able to supply services which, currently, are totally inadequate!

3.3 Overall Comments about the Registry Project

In addition, it can safely be concluded that the results of this study have contributed immensely toward a better

many factors, including the description of the project and the goals and objectives, the fact that participation in the Registry was voluntary, the design of the questionnaires, and finally the absence of any verification participants fit the identification criteria as being functionally deafblind according to the definition.
The report very satisfactorily describes the demographics, characteristics and provincial distribution of the participants. While the current registrants of 777 persons with deafblindness represent a good sample for analyses, there is no confidence that this sample is representative of the actual Canadian population of persons with deafblindness. There still remains much uncertainty about the actual size of the total population, its true characteristics (congenital vs. acquired), demographics (gender and age) and distribution across Canada. For example, it was reported that the congenital component of the sample was just under 45%. It is believed that the congenital component of the actual population in Canada would represent at the most, one quarter of the population. The study also reported that seniors over age 65 represented just over one-fifth of the sample. In reality, seniors likely should represent three times that number in the Canadian population.

These points are made to further call attention to the fact that if appropriate services are to be developed and offered to this disability group, that a much better determination of the actual deafblind population in Canada, with all its demographic patterns, is essential.

This study made some very significant advances. It has begun the process of revealing the real nature of deafblindness in Canada. It has alluded to the fact that there are problems with identifying deafblindness, and there is great disparity in services available to the vast majority of persons in Canada, with this disability. For example, the fact that the congenital population was so highly represented reflects a greater awareness by family members, the public and non-governmental agencies, of services available to this group, who tend to be a very high need segment of the deafblind population. The under-representation of the acquired group in the sample reflects many things; there are fewer services available to the acquired population, and people with this disability do not go looking for services, even if they do exist. All this contributes to a reduced opportunity for more of them to be identified as deafblind and to give a voice to their special needs.

There are definitely two distinct ‘worlds’ within the population of persons with deafblindness; the congenital world and the acquired world. The findings of this report show significant distinctions between the two groups in almost every category where comparisons were available. In addition, one major distinction exists; the wide variance between the two groups in terms of the type and extent of services available to each. However, since this was not one of the terms of reference of the study, further dialogue on this issue must be left for another discussion.

disability group.

- That all governments in Canada recognize deafblindness as a unique disability.
- All Canadians with the disability deafblindness be given every opportunity to enjoy a decent quality of life, contribute to their community, and be recognized as participating members of Canadian society.
- A special study be launched focussing on the current state of services available to all persons with deafblindness in Canada.
- Appropriate measures be developed to ensure that equitable services are available to all Canadians with deafblindness.
- Deafblindness identification criteria be the subject of continual debate, re-examination and re-evaluation to ensure it fits the changing face of the congenital population with special needs.
- Consistent identification criteria be developed and used across Canada in all medical, educational and institutional settings that serve people with combined visual and hearing impairments.
- The current National Volunteer Registry data base be maintained and continually updated.
- All persons with deafblindness be identified and encouraged to register with this National data base. This will require the support of all agencies serving or advocating for persons who are deafblind, provincial governments who maintain psychiatric facilities and community living organizations that support group homes for disabled persons, nursing homes, etc. Forms should be developed and distributed to intake departments that will facilitate keeping the Registry current. Notices should be placed in Deafblindness Newsletters and News Magazines reminding people to update their addresses and information, if moving.
- A network of Centres of Expertise be established across Canada to advise provincial governments and various non-governmental agencies and organizations about all aspects of deafblindness.

National Registry of Persons who are Deafblind in Canada

Canadian Deafblind and Rubella Association

1658 4th Av. West, Owen Sound, Ontario N4K 4X4
Ph:519-372-0887 Fax:519-372-0312 email:stan.munroe@sympatico.ca

Dear Participant:

The Canadian Deafblind and Rubella Association (CDBRA) received funding from the Federal Government to establish a Registry of Persons who are Deafblind in Canada. This Registry project is being implemented in partnership with the Canadian National Society of the Deaf-Blind (CDSDB), the Canadian National Institute for the Blind (CNIB), the W. Ross Macdonald School (WRMS) and Rotary Cheshire Homes Inc. These organizations together believe that the information gathered from this Registry will greatly assist to improve services for all Canadians who are deafblind.

The Registry will determine how many people are deafblind in Canada, information about their condition, where they live in Canada and the kind of services they receive. This information will be supplied to governments and various non-profit organizations that work with individuals who are deafblind.

Individuals are considered to be deafblind if they have a substantial loss of both sight and hearing such that neither sense is a reliable access to information. The combination of these sensory losses results in significant difficulties in acquiring educational, vocational, avocational and social skills.

If you believe that you fit this definition of deafblindness, please complete the attached questionnaire. The information collected is private and confidential. Your name and personal information will not be provided to anyone else without your permission. Participation in this survey is strictly voluntary. Once you have completed the questionnaire please return in the envelope provided.

Thank you for participating in this registry project.

1658 4th Av. West, Owen Sound, Ontario N4K 4X4
Ph:519-372-0887 Fax:519-372-0312 email:munroes@bmts.com

First name:_____ Last name_____ Address:_____Ci ty/town_____Provi nce_____Postal code_____ Date of Birth: _____ Sex: M____ F____ Marital Status: single____married_____	Phone: _____ TTY_____ Fax: _____ Email address: _____ Preferred method for future communication: Braille 1___Braille 2___ Large Print___Diskette___ Email____Fax___ Which language? English___ French___Other Language____

Communication Ability (circle predominant methods) Primary Living Accommodation (circle appropriate item)

1. Writing and Printing (Typing) 2. Sign language: ASL, Signed English 3. Speech 4. Manual alphabet/finger spelling 5. Braille 1 6. Braille 2 7. Gestures 8. Other (specify)_____ 9. Unknown	1. Lives with birth family 2. In foster care 3. Independent 4. Retirement Home 5. Provincial or Regional facility (Name) _____ 6. Supervised living (e.g. group home - Name)_____ 7. Supported living (e.g. with Intervention-Name) _____ 8. Other (specify)_____ 9. Unknown

Degree of Vision Loss (Circle only one in category)

1. Low Vision (20/70 - 20/200) 2. Legally blind (less than 20/200 or visual field less than 20 degrees) 3. Light perception only	4. Totally blind 5. Unable to test - uses vision 6. Unable to test - does not use vision 7. Unknown

Degree of Hearing Loss (Circle only one in category)

1. Mild (26 - 40 dB) 2. Moderate (41-70 dB) 3. Severe (71-90 dB) 4. Profound (91- 110+ dB)	5. Unable to test - uses hearing 6. Unable to test - does not use hearing 7. Unknown

Onset of Visual Loss (circle appropriate item) Onset of Hearing Loss (circle appropriate item)

1. Congenital 2. Acquired (later in life)....indicate age if known_____ 3. Unknown	1. Congenital 2. Acquired (later in life)....indicate age if known_____ 3. Pre-lingual (before language 4. Unknown

1. Stable 2. Progressive 3. Fluctuating 4. Unknown	1. Stable 2. Progressive 3. Fluctuating 4. Unknown

Etiology of Visual Loss (circle appropriate items)	Etiology of Hearing Loss (circle appropriate items)
1. Accident or Trauma (specify type and age)_____ 2. Brain Damage (specify)_____ 3. Syndrome a) Rubella b) Usher (specify type)_____ c) CHARGE d) Other_____ e) Unknown 4. Cataracts a) congenital b) late onset (specify age)_____ 5. Glaucoma (specify age of onset)_____ 6. Medical condition a) diabetes (age of onset)_____ b) Multiple sclerosis (age of onset)_____ c) other (specify)_____ 7. Other causes (specify)_____ 8. Unknown	1. Accident or Trauma (specify type and age)_____ 2. Brain Damage (specify)_____ 3. Syndrome a) Rubella b) Usher (specify type)_____ c) CHARGE d) Other_____ e) Unknown 4. Auditory neuritis 5. Otitis media 6. Medical condition a) diabetes (age of onset)_____ b) Multiple sclerosis (age of onset)_____ c) other (specify)_____ 7. Drug related (specify)_____ 8. Other causes (specify) _____ 9. Unknown

Personal Status: (circle appropriate items)

1. In school (specify school and level)_____ 2. Finished school (specify level completed)_____ 3. Competitive employment	4. Self employed 5. Sheltered employment 6. Workshop setting 7. Cannot work (specify reasons)_____	8. Homemaker 9. Homebound 10. Retired 11.Receiving medical care (specify)_____	12.Unknown 13.Other (specify): _____ _____ _____

This form completed by:

Individual yes___no_____If no complete: Name_____ Address_____ _____ _____	Relationship to individual (specify): _____ If facility, specify:_____ Date Completed:_____

How did you learn of this Registry?
Organization of facility (specify)_____ Family member____Other(specify_____

National Registry of Persons who are Deafblind in Canada

Canadian Deafblind and Rubella Association
1658 4th Av. West, Owen Sound, Ontario N4K 4X4
Ph: 519-372-0887 Fax: 519-372-0312 email:stan.munroe@sympatico.ca

Dear Participant:

This is the question paper for the Registry of Persons who are Deaf-Blind. It is important for everyone to get their name on this list. A registry is a list of people. The information will help to explain how many deaf-blind people live in Canada. It is important to know this information so that the governments can understand about your needs. Please answer the questions as soon as possible. Send the papers back in the envelope given to you.

A person is Deaf-Blind if they:

- are Deaf and Blind
- cannot clearly hear speech or alarms without hearing aids and are blind
- cannot clearly hear speech or alarms without hearing aids and cannot read the newspaper even with the best glasses on
- are deaf and have severe tunnel vision
- cannot clearly hear speech or alarms without hearing aids and have severe tunnel vision
- cannot clearly hear speech or alarms without hearing aids, cannot see the road signs or bus numbers even with the best glasses on and need help to travel in unfamiliar areas.

1.0 According to these points are you Deaf-Blind?
Yes No
2.0 PERSONAL and CONFIDENTIAL INFORMATION

For office use only: Individual Code # _____

Last Name

Address:
Street & # Apt #

City/Town

Prov Postal Code

Telephone - area code and # ( )

TTY Voice Other

Fax #

E-mail address

Your birth date? Month Day Year

Sex: Male___ Female____

Are you married? Yes No

Do you live with your parents? Yes No

Do you live alone? Yes No Live with others? Yes No__

If Yes: Group home Institution Shared Apt.

3.0 ACCESS TO INFORMATION

How do you want information sent to you? Check one.

Braille Grade 1 or Braille Grade 2

Large print Regular print

Computer Disk Fax E-mail

What language do you wish the information? English French Other

4.0 COMMUNICATION

Which languages do you use for communication? Check all those that apply.

ASL Other 2-Hand Manual

Finger Spelling Print Notes

Tellatouch or other machine

Audio Tapes Speech Other

5.0 VISION DIAGNOSIS

Do you know the cause of your vision problem? Yes No____

What is it called?

How old were you when your vision problem started?_____

Is your vision loss getting worse? Yes____no____

6.0 HEARING LOSS

How old were you when your hearing loss began?_____

Is your hearing loss getting worse? Yes____ No_____

7.0 ACTIVITIES

Are you employed? Yes No

Are you in school? Yes No

8.0 ASSISTANCE

Check the services you get:

Interpreter Intervenor Volunteer

Friend Driver/Guide____ Other_____ None_____

How did you find out about this survey? Friend____Family

member_____Organization(specify)_____

Please tell other people about the Register.

If anyone needs a form or has questions please call Joyce Thompson at (416) 730-9501 Voice or (416) 730-9187 TTY.
or email at rcheshire@onramp.ca

Thank you for taking time to answer these questions. There may be another set of different questions sent to you in about six (6) months.

Canadian Deafblind and Rubella Association
1658 4th Av. West, Owen Sound, Ontario N4K 4X4
Ph: 519-372-0887 Fax: 519-372-0312 email:stan.munroe@sympatico.ca

January, 2000

Dear Participant:

Thank you for your participation in the Registry of Persons who are Deafblind in Canada.

We are asking for your cooperation in a follow-up survey. The purpose of this segment of the study is to obtain further information about the population of individuals who are Deafblind, including areas such as education, employment, living arrangements, technical aids, Intervenor Services, leisure and advocacy.

We would appreciate it if you would complete the attached questionnaire and return it by March 31, 2000. As with the first questionnaire, we have attached a stamped, self addressed envelope for your return.

Some people have asked, "Why another survey? Wouldn't it be better to spend the money on Intervenor Services, improving employment opportunities, etc?" The fact is that statistics gathered in this survey are very important to yourself and to the advocacy organizations participating in this study. These organizations, which include CNSDB, CDBRA, RCH and CNIB, will use the findings to advocate for improved services for all persons who are Deafblind.

For this reason, we believe that the Registry Study is extremely important. It is the only way we can advise the government of the number of individuals in Canada with both auditory and visual loss and that these individuals have special and unique needs.

There is a similar Registry Project going on in the United States. The agencies in the USA are updating a previous Registry as a means of proving to various USA governments that more services are required for individuals with both hearing and visual problems.

But there, persons who are Deafblind are not given a choice - they must answer the questions. Here in Canada we believe that you should not be forced to answer this survey but that you will recognize the need and do it voluntarily.

Attached is a brief summary of the statistics gathered so far in the Survey. As of December 31, 1999, 657 individuals have volunteered to be part of the Canadian Registry. This represents about 55% of the people known to be Deafblind in Canada and who have already received a questionnaire. We believe this only represents 25 to 30 percent of the total Deafblind population in Canada. For this reason we are asking you to help further by asking others you know, who are Deafblind, to get registered so that we can have the most accurate information possible.

For your information, we are enclosing additional resource material. We suggest that you join as many organizations as you can that are connected to Deafblindness. You will become more familiar with current research, new technology and networking with other individuals in Canada or across the world through newsletters and meetings.
This survey is important for you and for all other persons who are Deafblind in Canada! Please complete and return it now.

Yours Sincerely

Stan Munroe
Registry Project Leader

Ph: 519-372-0887 Fax: 519-372-0312 email: stan.munroe@sympatico.ca

1.0 PERSONAL INFORMATION
First Name_____
Last Name_____
Address_____
_____

2.0 EMPLOYMENT
Please check off which applies to you:
Working _____What job do you do?_____
Self Employed?_____ Working in sheltered workshop?_____
Looking for work?_____ Volunteering?_____
Want to work?_____

3.0 INTERVENOR SERVICES
Do you receive Intervenor Services? Yes___ No____
If Yes: Number of hours per week_____
Are you happy with these hours? Yes____ No_____
Are Intervenors available when you need them? Yes___ No__
How much notice must you give to get Intervenor Services? ___days.
Are your Intervenors fluent in the communication methods you use? Yes___ No___
Additional comments about Intervenors: _____
Do you need an Intervenor to communicate with your own family?
Yes_____ No_____
Are you happy with your communication skills? Yes_____ No_____

4.0 INDEPENDENT LIVING
Are you happy with your current accommodation?
Yes_____ No_____
If No, what do you need? _____
_____
Have you received training in the following:
Orientation and Mobility? Yes_____ No_____
Which method of travel do you use most often: sighted guide_____;
cane travel_____; guide dog_____; remaining vision_____; other (specify)_____
Daily Living skills (meal preparation, cleaning, budget management etc)
Yes_____ No_____
Are you happy with the daily living and orientation and mobility training that you received?
Yes____ No____
If No, what do you need?_____
_____

5.0 TECHNICAL AIDS
Have you received training to use communication devices?
Yes_____ No____
If No, what training do you need or want?_____
Please check the devices you use for communication:
Hearing Aids_____ FM/Infrared systems_____
Cochlear Implant _____
Magnifiers_____ Telebraille_____ Tellatouch_____
TTY_____ Computer_____ Internet _____

Closed Caption Decoder_____

Please check the signal systems you use?
Flashing Signals____Vibrating Signals_____
Fire Alarms/Smoke Detectors_____
Please list other signal systems you use _____

6.0 LEVEL OF EDUCATION & TRAINING
High School: Completed?_____ Did not complete_____
College: Completed?_____ Did not complete_____
Technical Training: Completed?___ Specify course_____
University: Completed?_____Specify degree_____ Did not complete_____
If you had support services would you want to continue your education? Yes_____ No_____

7.0 LEISURE ACTIVITIES
Do you have any hobbies? Yes_____ No_____

Please list your hobbies:_____

Do you enjoy crafts? Yes_____ No_____
Please list the crafts you do_____

8.0 ADVOCACY
Do you have an opportunity to evaluate the services you receive?
Yes_____No_____
Do you have an opportunity to advocate for your rights as a person who is Deafblind? Yes_____No_____
Are you a member of:
CNSDB (Canadian National Society of the Deaf Blind)____
CDBRA (Canadian Deafblind and Rubella Association)____
DBAT(Deaf-Blind Association of Toronto___
BCADB (British Columbia Association of the Deaf-Blind)____
MDBA (Manitoba Deaf-Blind Association) ____
Other Organization (name:_____)
Are you an advocate for services by being a Board Member or a Spokesperson for an organization? Yes___
No____
If yes, please name the organization_____

For Office use only:
Date Received_____ ID #_____

Implementation and Management Guidelines

- The registry project will be implemented and managed according to the rules of confidentiality and as per the confidentiality requirements indicated by the funding agent.

- Participation in the Registry is strictly voluntary. Participants in the Registry are not obligated to provide their name and full address, but the town and province they are living is important. Those who prefer not to provide a complete address will not receive the survey results and thus indicate their unwillingness to participate in any follow up surveys or requests for information.

- It is presumed that individuals who supply their name and full address are volunteering for follow-up information, unless they indicate otherwise.

- The results from the Registry study will be a summary of the information provided by all volunteer participants and will not identify individual participants.

- A copy of the Registry report will be distributed to all volunteer participants, pertinent Federal and Provincial agencies, not-for-profit service, consumer and advocacy organizations involved with deafblind issues etc.

- Strict rules of confidentiality will be adhered to with respect to information provided by volunteer participants. Names of individuals, their addresses, phone, fax or email addresses will be protected. Specific individual related information will not be supplied to any organization or individual without the consent of that individual, their family or advocate.

- The confidential material contained in the data bank will be managed by the Registrar of the Data Bank and will not be available to any organization, including the organization (CDBRA) coordinating this Registry project or its partner organizations. Organizations wishing to contact specific individuals or groups of individuals will contact the Registrar who will coordinate the distribution of material to individuals. Organizations will be expected to cover the costs of distribution of any material or follow up surveys.

May 4, 1987

Dear Sirs:

I very strongly hope that any plan for the building of homes of the Deaf/Blind people of Canada will go ahead as quickly as possible. These Deaf/Blind people certainly should receive specially trained help that will enable them to lead happy and constructive lives. I believe there are enough cases of Deaf/Blindness in Canada to deserve this assistance.

I have a special interest in this matter as I, myself am totally deaf and 3/4 blind. I understand the special needs of these handicapped people.

Although I live in an excellent Nursing Home, the problem of communication with nurses and officials of the Nursing Home frustrates me every day. I communicate with others by having them spell words on my hand. The moment I mention this, people clam up and back away. I am a different person than most and people are suspicious of me. They are not educated in this field and are afraid of the unknown.

The doctors and nurses do try to talk to me, although they are amateur at it. Out of the 105 people who live with me, only one even attempts to talk to me. That is how alone I am. I try to be friendly with everyone, but the moment I ask them to spell on my hand, the conversation ends. Although I am always with 100 people, I am always alone.

A Deaf/Blind person has to entertain himself only with the contents of his own mind, because most people don't want to be bothered learning to communicate. I am luckier than most because I am well educated and have travelled extensively, leaving me with many memories. The only activity I can think of is to memorize poetry, though I am not a poetry fan. This can only be done at the convenience of someone who can help me. I cannot read, write or watch TV.

The idea of having people live together who can talk to each other will mean they won't be alone, as I am.

signed Canon R.T. Appleyard/BL

Reverend Canon Reginald T. Appleyard
Retired Anglican Priest

Province	Sponsoring Organization	Contact Persons
Alberta	• CNIB-Alberta Deafblind Services (Calgary) • CNIB-Alberta/ NWT Division (Edmonton) • Calgary Board of Education • CDBRA-Alberta	• Amber Long • Erin Martin • Janice Northcott • Wendy Johnson
British Columbia	• CNIB-BC/Yukon Division and Deafblind Services • Western Institute for the Deaf • BC-Provincial Outreach Program for Deafblind • CDBRA-BC • DBR Housing • Canadian Council for the Blind BC/Yukon Division • BC Ministry for Children and Families • Mainstream Association for Proactive Living • BC Deaf-Blind Association	• Hilda Nanning/Janet Hanevelt • Margaret Phillips • Linda Mamer/Joyce Olson • Linda Mamer • Allison Seez • Shar on Wagner • Gord Tullock • Craig McLean and Monique McDonald
Quebec	• Institut Raymond Dewar (Montreal) • Institut readaptation Physique de Quebec (Quebec City) • L'Interaction Mont Joli	• Gilles Lefebvre • Lucille St. Denis • Gilles Lavoie
Manitoba	• Intervention-Manitoba (CDBRA-MB) • Manitoba Deaf-Blind Resource	• Cheryl Ramey • Jane

	• CNIB-NB Division		ia Curtis • Jean Ann Ledwell
Newfoundland	• Deaf • Division •	Newfoundland School for the CNIB-Newfoundland/ Labrador Individual families	• Linda Clark • Len Baker • confi dential
Nova Scotia	• • • Society • • (Canada)	APSEA CNIB-NS/PEI Division Regional Residential Services Small Options Group Homes Society of Alstrom Syndrome	• Dr. Ann MacCuspie • Eliza beth Hamilton • Agne s O'Neill • nume rous • Sand ra Surette

	Falls) • Southwest Regional Centre (Blenheim) • E.C. Drury Provincial School for the Hearing Impaired (Milton) • Sir James Whitney Provincial School for the Hearing Impaired (Belleville) • Robarts Provincial School for the Hearing Impaired (London) • CNIB-Ontario Division • Rotary Cheshire Homes, Inc. (Toronto) • CNSDB (Canada) • Bob Rumball Mission for the Deaf (Toronto) • BALANCE • McInnes House for Deafblind Persons (Brantford) • Independent Living Residences for Deafblind Persons (Richmond Hill) • Ministry of Education - Ottawa • Ministry of Education - Sudbury • CDBRA-ON • W Ross Macdonald School, Ministry of Education (Brantford) • Mijiwam Support Home (Eastern Ontario) • Individuals • Parents of Deaf-Plus Ontarians • ReCharge Ontario	Marie Carruthers • Donna Morrow • Ray Brown • Margaret Bulligan • • Mary Zadow • • Mary Ellen Bolt • • Bev Ginou/Nancy Lord • Joyce Thompson • Jim Thompson /Anna Bloom • Bob Rumball • Sue Archibald • Joan Brintnell • Jo Ann Newbery • • Jean-Marc Cholette • Carole Landry • Cathy Proll • Bill Thompson • • Keryl Maynard Banks • confidential • Susan Popper • Deb Caccia
Prince Edward Island	See CNIB-NS/PEI and CDBRA-NB/PEI	Patricia Curtis

	• of Hearing • •	Prince Albert Association of Hard Sask. Deaf and Hard of Hearing individual
		ne Williams • Lynn Lada • Fern Lamb • George Thompson/ • Leona Ames • confidential

Syndrome); loss of oxygen (from near drowning); brain tumour, abscess or haemorrhage; stroke, Sudden Infant Death Syndrome (SIDS). May result in various degrees of vision, hearing, cognitive and motor impairments. Eye and ear function may actually be intact but the brain cannot process the visual or auditory information.

Aphasia

An impairment of the ability to use or comprehend words, usually acquired as the result of a stroke or other brain injury. Source: http://www.aphasia.org

Septicaemia (Liver)

Systemic disease caused by presence of microorganisms or their toxins in the blood stream, resulting in a serious fever. In this instance the child had a ruptured liver abscess which led to the septicaemia, high fever and brain damage.

Shaken Baby Syndrome

Serious head injury the result of physical abuse; observed most often in infants less than 6 months of age. Symptoms can include retinal haemorrhage which can lead to blindness; subdural and/or sub-arachnoid haemorrhage which can lead to cerebral atrophy; mental retardation; spastic quadriplegia and severe motor dysfunction. Source: American Academy of Pediatrics (December, 1993) Shaken Baby Syndrome: Inflicted Cerebral Trauma (RE9337). Pediatrics Vol. 92(6) 872-875 http://www.aap.org/policy/05126.html

Sudden Infant Death Syndrome (SIDS) - Survival

Normally SIDS is fatal and its cause unknown. In this particular instance the child was revived after been pronounced dead from SIDS. The child suffered a stroke and accompanying seizures and is experiencing some neurological dysfunction and sensory loss. Source: A family member

5.4.2 Ageing

The ageing process increases the incidence of both vision and hearing deterioration. A reduction in hearing acuity is a normal expectation of ageing. With ageing also comes an increasing incidence of such eye diseases as cataracts, glaucoma and macular degeneration; although these diseases are not strictly age related, since glaucoma and macular degeneration can occur in persons as young as 35. Source: Joyce Thompson (2001)

Cataracts

The progressive clouding or opaqueness of the crystalline lens which blocks the passage of light rays thus limiting vision. Cataracts are usually associated with ageing, although they can occur during the early years Source: A Guide to the Human Eye. Merck Frosst Canada Inc (1994)

National Society for the Prevention of Blindness (1977)

Detached Retina
Condition in which the rod and cone cells (bacillary layer) of the retina is partially or completely separated from the pigment epithelial layer, resulting in a loss of vision in the area that is detached. Source: Hoffman, 1998. Quick Reference Glossary of Eye Care Terminology

Eye Thrombosis
Blood clot within the eye. Source: Stedman's Medical Dictionary (1972)

Glaucoma
An eye disease that can permanently damage the optic nerve. Usually associated with excessive pressure in the eye from the accumulation of aqueous humour. Glaucoma gradually leads to a loss of visual field, beginning with side vision. Glaucoma may affect one eye only, but usually both eyes are involved. Source: Understanding Language. National Society for the Prevention of Blindness (1977)

Macular Degeneration
Central vision impairing condition caused by alteration in vascular supply to the retina. This is a painless disease that usually affects the elderly, but sometimes affects younger persons. The degeneration begins when the blood supply behind the retina leaks. Subsequently abnormal blood vessels grow, replacing retinal nerves. The macula then deteriorates, impairing the central vision. Source: A Guide to the Human Eye. Merck Frosst Canada Inc. (1994)

5.4.3 Birth Trauma causing Brain Damage

Birthing irregularities leading to hypoxia/anoxia (reduced oxygen to the brain) causing irreversible neuro-sensory damage. May lead to various degrees of hearing and visual loss.

Hypoxia/Anoxia
Oxygen deficiency; more precisely called hypoxia.

Meconium Aspiration Syndrome
Aspiration occurs when the newborn inhales meconium (the first stool of the newborn) when it is mixed with amniotic fluid during labour and delivery. Meconium is released if there is a lack of oxygen suffered by a fetus during a stressful labour and delivery. When the child inhales the meconium mixture while still in the uterus, a partial to complete blockage of the airwaves can occur. This can lead to intra-uterine hypoxia (decreased oxygen to the infant while still in the uterus) leading to brain damage. Source: http://health.yahoo.com/health/Diseases

Source: http://www.ion.ucl.ac.uk/library/patient/sah.htm

5.4.4 Congenital Brain Damage

Damage to the brain which is present at birth. May be caused by an inter-uterine infection, genetic defect, fetal injury or any insult to the developing fetus that affects normal brain development.

Cerebral Atrophy

Pertains to the wasting away or deterioration of the neural tissue in that part of the brain called the cerebrum.

Cerebral Haemorrhage

A profuse flow of blood into the cerebrum section of the brain due to a rupture of the lenticulostriate artery. Source: Stedman's Medical Dictionary (1972)

Cerebral Palsy

A term used to describe a group of disorders affecting body movement and muscle coordination, the result of an insult to, or anomaly of, the developing brain. Effects of this condition may include involuntary movement, muscle tightness or spasms, difficulty with gross and fine motor skills, abnormal perception and sensation. The associated brain damage may lead to seizures, learning difficulties and developmental delay. Causes are numerous, including: genetic or developmental disorders during pregnancy; labour difficulties and prematurity, head injuries, drowning injuries, brain haemorrhages and infections like meningitis during early childhood. Source: http://www.cerebralpalsycanada.com

Congenital Brain Malformation

Pertaining to brain deformity or malfunction which may be hereditary, or due to some influence arising during gestation. Source: Stedman's Medical Dictionary (1972)

Dandy-Walker Syndrome

A rare congenital malformation of the brain characterized by an abnormally enlarged space at the back of the brain that interferes with normal flow of cerebrospinal fluid within the brain. Accumulation of fluid around the brain causes abnormally high pressure within the skull and swelling of the head (congenital hydrocephalus) leading to neurological impairment. Symptoms may include learning disabilities, motor and cognitive delays, impaired hearing and impaired vision through swelling of optic nerve and nystagmus. Causes considered multi-factorial; in rare cases inherited as an autosomal trait. Boys appear to be more affected than girls. The malformation occurs during early embryological development. Source: National Organization on Rare Disorders: http://www.rarediseases.org; email: orphan@rarediseases.org); Canadian Organization on Rare Disorders; www.cord.ca; Dandy Walker Syndrome Network www.geocities.com/Heartland/Hills/3919/dws.html

Such exposure of brain material can lead to infection or swelling of the brain (meningitis). May also lead to hydrocephalus with the corresponding neural affects previously described. Source: National Organization on Rare Disorders: http://www.rarediseases.org

Encephalopathy (developmental)
Pertaining to any disease of the brain and diagnosed in relation to the pre-natal developing brain. Source: Stedman's Medical Dictionary (1972)

Enlarged Left Ventricle
An enlargement of the left ventricle or cavity (*ventriculus sinister*) of the brain, due to accumulation of cerebrospinal fluid. Source: Stedman's Medical Dictionary (1972)

Epilepsy
A chronic disorder characterized by spasm attacks (electrical impulses) of brain dysfunction, usually associated with some alteration of consciousness. Epileptic attacks may remain confined to elementary or complex impairment of behaviour or may progress to a generalized convulsion. Source: Stedman's Medical Dictionary (1972)

Holoprosencephaly
Failure of the forebrain to divide into lobes during fetal brain development. Can result in different degrees of severity. The most severe form can lead to severe mental retardation, seizures, microcephaly, seizures and cleft lip/palate. This disorder is associated with chromosome abnormalities. Source: National Institute for Neurological Disorders and Stroke; http://www.ion.ucl.ac.uk/library/holopro.htm; Stedman's Medical Dictionary (1972)

Hydrocephalus
A condition in which abnormally widened spaces in the brain (ventricles) inhibit the normal flow of cerebrospinal fluid that surrounds the brain and spinal cord. The fluid accumulates in the skull and puts pressure on brain tissue. An enlarged head (cephalomegaly), increase in pressure within the brain, convulsions and abnormal reflexes are symptoms of hydrocephalus. This condition may lead to Dandy-Walker Syndrome. Congenital hydrocephalus is thought to be caused by complex interaction of environmental and genetic factors. Occurs in 1 to 2 per 1000 births. Source: National Organization on Rare Disorders; http://www.rarediseases.org; Hydrocephalus Association; http://www.HydroAssoc.org/

Hypoxic-Ischemic Excelopathy (H.I.E.)
A clinical diagnosis of impaired neurological function in a newborn. A frequent cause of this brain injury is associated with birth or perinatal asphyxia due to decreased or inadequate blood supply to the child. Seizures often develop in infants who sustained severe birth asphyxia. Source: Birth Trauma Website: Newborn Conditions Associated with Asphyxia: http://www.alfelgreene.com/newborn.html

delays in acquisition of skills that require coordination of mental and physical activity. Mental retardation is present in most individuals after 5 years. Incidence is 0.1/100,000 children and accounts for 3.2% of children with epilepsy. This syndrome can occur with individuals suffering previous brain damage and other neurological conditions resulting from a brain tumour, severe head injury or a congenital condition called tuberous sclerosis. Genetic factors believed to account for a small number of cases - less than 3%. Source: Gilbert (1996); National Organization on Rare Disorders (www.stepstn.com/cgi-win/nord.exe) and www.icondata.com/pedbase/files/LENNOX-G.HTM

Lissencephaly (with microcephaly)

A malformation of the brain characterized by the absence of the convoluted pattern of the cerebral cortex. Conditions observed in the child include poor visual tracking, poor feeding, and weight gain and seizures. Some children have microcephaly. The conditions are observed usually several months after birth. Causes may be either genetic or non-genetic. The causes may include viral infections in first trimester of pregnancy, insufficient blood supply to the brain during early development, a genetic disorder (recessive inheritance) and damage or mutation to a specific region of chromosome 17. There are a number of syndromes associated with this condition. Source: Lissencephaly Information for Parents: http://www.lissencephaly.org/about/lissen.htm

Microcephaly

Abnormal smallness of the head; also denoting a skull with a capacity below 1350 cc. Source: Stedman's Medical Dictionary (1972)

Neonatal Bleeding

Pertains to bleeding during the period preceding birth; the result of a trauma. Source: Stedman's Medical Dictionary (1972)

Perinatal

Trauma occurring or pertaining to, before, during or after the time of birth. Time designations before or after birth are usually arbitrary. Source: Stedman's Medical Dictionary (1972)

Perinatal Hypoxia

Lack of oxygen occurring before, during or after the time of birth. Source: Stedman's Medical Dictionary (1972)

Prenatal Hypoxia or Anoxia

See Birth Trauma leading to Brain Damage

5.4.5 Fetal Alcohol Syndrome

A cluster of related birth defects including characteristic facial shape or appearance, small head size, poor growth before and after birth, developmental delay, short attention span together with behavioural problems. Often there is a malformation of the eustachian tube leading to hearing impairment. Symptoms are shown in children born to mothers who drink alcohol heavily during pregnancy. Incidence to some degree in 1 of every 100 live births; rising to 1 of 3 if drinking habits are heavy. Source: Gilbert (1996)

5.4.6 Intra-Uterine Infections

Various diseases that invade the developing fetus in-utero causes sensorineural damage. Examples are rubella, cytomegalovirus and toxoplasmosis which can cause hearing and vision loss.

Congenital Rubella Syndrome (CRS)

Refers to the manifestation of various defects to the new born child that frequently result when the pregnant mother contacts the rubella virus during her first trimester of pregnancy. Rubella generally causes hearing and vision loss and various cardiac defects. Other manifestations may occur later, (e.g. diabetes and hypothyroidism), often around the age of puberty. Rubella is known to be a cause of hearing loss without the

spasticity, intercranial calcifications, mental retardation and seizures. In some cases hearing and vision impairments can occur. Source: Cytomagalovirus. Dbl Review: July-December, 1995

Toxoplasmosis

An infection caused by the reaction to the protozoan parasite, *Toxoplasma gondii.* The infection is usually mild producing symptoms like a mild flu. When the infection occurs in a pregnant woman, in many cases (40%) can cause a congenital abnormality to the unborn child. When the case is acute, in association with the fever it can cause encephalomyelitis, ophthalmic lesions, jaundice, etc. Source: Stedman's Medical Dictionary (1972)

5.4.7 Inherited Rare Disorders

Rare disorders, the characteristics or qualities which are transmitted from parent to offspring. Inheritance can involve numerous methods: blending of parental genetic material, transmission of recessive traits (which may occur irregularly), transmission of dominant traits (from one generation to the next), sex linked transmission (via X-chromosome), maternal transmission (via peculiarities of the egg cytoplasm), hologynic (mothers to daughters) etc. Source: Stedman's Medical Dictionary (1972)

typically characterized by an underdevelopment or absence of the band of nerve fibres joining the brain hemispheres or corpus colossum and moderate to severe mental retardation. Brain malformations may also be associated with additional complications, including brain seizures and hydrocephalus. Individuals may have physical abnormalities affecting the eyes (e.g. spacing, slanting eyelids, drooping of upper eyelids and vertical skin folds covering part of eye), decreased retinal pigmentation and optic atrophy. Recessive inheritance has been suggested as the cause; but the syndrome may often appear randomly. Source: National Organization of Rare Disorders: http://www.rarediseases.org

Alstrom Syndrome

A hereditary recessive syndrome which can be inherited by males and females with equal probability. Manifestations of the syndrome include: vision and hearing impairment due to nystagmus and photodysphoria in early infancy; progressive pigmentary retinopathy; mild to moderate bilateral sensoririneural hearing loss. Individuals have normal intelligence with some reports of delayed early developmental milestones.
Source: http://www.lena.jax.org/alstrom/documents

Charcot Marie Tooth (CMT) Syndrome

A common inherited neurological disorder, recognized largely as autosomal (non sex-linked) dominant. It has been known to be inherited in a recessive or X-linked pattern. Recent evidence is that it is associated with duplication of chromosome 17. It is known to affect 130,000 Europeans and 150,000 Americans. Symptoms, which usually occur by age 10, include atrophy of the limbs and extremities, renal nephritis and progressive sensori-neural hearing loss, beginning during teen years. Source: Gilbert (1996); www.charcot-marie-tooth.org

Cockayne Syndrome (early onset epilepsy)

A genetic and recessively inherited rare form of dwarfism. Both parents must be carriers of the recessive gene to cause a child to have this condition. Common symptoms of this syndrome include early onset epilepsy, dwarfism, microcephaly, progressive neuro-developmental delay, progressive mental deficiency, progressive retinopathy and/or cataracts and progressive hearing loss. Onset of symptoms usually appear in the child's second year of life. Source: http://www.kimbanet.com; Share and Care Cockayne Syndrome Network, email: cockayne@kimbanet.com

Friedrich Ataxia Syndrome

An inherited autosomal recessive disorder, affecting gene on chromosome 9, causing atrophy of the spinal cord. A progressive condition, the age of first manifestation is variable, but often occurs around puberty. Eye effects include nystagmus and optic atrophy. Deafness is a less common occurrence. Source: Gilbert (1996); **DisplayText cannot span more than one line!**

Infantile Refsum Syndrome

A rare metabolic disorder characterized by the reduction or absence of peroxisomes (cell structures that rid the body of toxic substances) in the body, and the accumulation of phytanic acid in the blood plasma and tissue. This syndrome is one of a group of genetic disorders called leukodystrophies that affect the myelin sheath which covers the nerve fibres. It has an autosomal recessive pattern of inheritance. Symptoms include such vision impairments as retinitis pigmentosa and nystagmus, developmental delay, hearing impairments and mild facial abnormality. Onset of the disorder occurs early in infancy. Source: National Organization on Rare Disorders: http://www.rarediseases.org; http://www.medemall.com/general/ird.htm; and http://www.oil4kids.com/

Kearns-Sayre Syndrome (KSS)

See description under Mitochondrial diseases.

Leber's Optic Neuropathy (Leber's Disease)

A rare genetic condition which can cause the loss of central vision through damage to the retina and optic nerve. It usually affects men, most commonly in their late 20's or early 30's; but it can affect men and women at any age. Usually Leber's affects one eye first then the other eye several months later. The condition is transmitted through the female parent. Leber's is linked to a number of genes, all in the DNA of the mitochondria. Source: Leber's Optic Nerve Home Page: http://www.leeder.demon.co.uk/blacktxt/lhonhome.htm

Leukodystrophy (Atypical)

Genetically determined disorder that affects the myelin sheath of the brain, spinal cord and peripheral nerves. Pattern of inheritance is either autosomal recessive or X-linked. Mental function is affected. Source: http://www.oil4kids.com/aldinfo.htm

Myers Syndrome (Smith-Fineman-Myers Syndrome)

A rare syndrome featuring microcephaly (small head), unusual facial features and mental deficiency etc. Other symptoms may include strabismus and optic nerve effects, occasional seizures, growth, motor and mental retardation. Genetic inheritance transmitted as an X-linked trait. Source: http://www.nim.nih.gov/mesh/jablonski/syndromes

Neurofibromotosis (NF; also known as von Recklingshausen disease)

A genetic disorder of the nervous system that primarily affects the development and growth of neural cell tissues through the growth of tumours. The disorder is classified as either NF1 or NF2; the former is more common and is apparent as changes in skin appearance, presence of tumours and bone abnormalities; NF2 is less common and is characterized by bilateral tumours on the 8th cranial nerve. Individuals affected with NF2 may notice hearing loss as early as the teen years. The tumours associated with NF2 can, in some cases, cause damage to nearby vital structures such as other cranial nerves and the brainstem. While NF is transmitted genetically, the disorder can occur spontaneously as a mutation in 30-50% of cases. Once these occur, the disorder can be transmitted in succeeding generations. NF1 is associated with chromosome 17 while NF2 is associated with chromosome 22. Source: National Institute of Neurological Disorders and Stroke: http://www.ninds.nih.gov/health and www.medical/disorders/neurofibr.htm

Norries Syndrome

A rare syndrome of retinal malformation characterized by dysphasia of the retina and associated with mental retardation and severe sensorineural deafness. Genetic inheritance transmitted as X-linked recessive (chromosome 11).
Source: http://www.icondata.com/health/pedbase/files/NORRIEDI.HTM

Phenylketonuria (PKU)

A metabolic disease caused by a deficiency of a vital enzyme (PAH) necessary for the conversion of the amino acid phenylalanine to the amino acid tyrosine, resulting in an excess of phenylalanine in the blood. This condition may be controlled by diet. If not diagnosed at birth this can lead to brain damage. PKU is inherited in an

A common form of inherited juvenile macular degeneration. Macular degeneration is characterized by a reduction of central vision with preservation of peripheral vision. Progression of visual loss with Stargardt's disease is variable. The disease is inherited as an autosomal recessive disorder. Source: The Foundation Fighting Blindness;
http://blindness.org/html/vision_disorder/wstargrdt.html

Smith Lemli-Opitz Syndrome (RHS Syndrome)

A rare syndrome that is inherited in an autosomal recessive manner with an incidence rate of about 1 birth per 40,000. Symptoms include learning disabilities, microcephaly, facial deformities, and to a lesser extent heart and renal problems. Source: Gilbert (1996); Opitz Family Network: email: opitznet@rymtnhi.com

Treacher Collins Syndrome

This syndrome, which solely affects the face and associated anatomical features, is inherited as an autosomal dominant syndrome. There is also a fifty percent chance of the syndrome to be the result of mutations. Symptoms include: abnormal development of the middle ear and external ear canal, causing conductive deafness in some instances; external eye effects but not recognized as being associated with visual loss; defects to jaw and face, etc. Source: Gilbert (1996); Treacher Collins Network: http://www.geocities.com/Heartland/Plains/6153/

describes a group of hereditary diseases of the retina. With RP, the retina slowly degenerates and loses its ability to transmit pictures to the brain. Three different types of Usher Syndrome have been identified: Type 1 is characterized by profound congenital deafness and RP evident within the first 10 years of age (identified in up to 90% of cases).Type 2, is characterized with mild to severe congenital hearing impairment, with RP becoming evident from late adolescence to late 20's. Type 3 is characterized by both progressive hearing loss and RP with age. Source: The Collaborative Usher Syndrome Project. Boys Town National Research Hospital: http://www.boystown.org/btnrh/usher.html

5.4.8 Metabolic Diseases

The result of a critical enzyme in the metabolic process being disabled or if a control mechanism for a metabolic pathway is affected. These diseases may be the result of inherited mutations or accidental protein mutations. Includes Infantile Refsum Syndrome and Phenylketonuria, both inherited disorders. Source: http://www.nccbi.nlm.gov/disease/Metabolism.html

Diabetes Mellitus

A metabolic disease in which carbohydrate (sugars) utilization is reduced and that of lipids (fats) and proteins are enhanced. It is caused by a deficiency of insulin. Complications may include neuropathy (a disease of the peripheral nerves which involves primarily sensory fibres), retinopathy (non-inflammatory degenerative disease of the retina characterized by haemorrhages), nephropathy (disease of the kidneys) and degenerative changes in the large and small blood vessels. Source: Stedman's Medical Dictionary (1972)

Morquio's Syndrome

One of the mucopolysaccharide (complex sugar) abnormalities that has a particular enzyme defect being the accumulation of complex sugars in the various organs. By about 18 months of age the individual may demonstrate the following symptoms: growth retardation, skeletal deformation, deafness, cardiac defects and/or visual loss (corneal clouding). Incidence 1 per 100,000 live births; both sexes equally affected. Inherited as an autosomal recessive. Source: Gilbert (1996)

Paget's disease

A metabolic bone disorder of unknown origin that normally affects older people. Complications include arthritis, deafness, pain and fractures. The progressive nature of the disease causes the skull to increase in size causing pressure on nearby nerves. Severe skull enlargement, resulting from such pressure can result in deafness, disturbed vision, dizziness and tinnitis. This disease occurs more commonly in Britain than anywhere else in the world, where there are 0.75 to 1.0 million sufferers. Source: The National Association for the relief of Pagets Disease: http://paget.org.uk/paget.html

disorders a high number of defective mitochondria are present. Source: http://www.kumc.edu/gec/support/mitochon.html

Kearns-Sayre Syndrome (KSS)

A rare disorder that belongs to the above mentioned neuromuscular disorders. In 80% of the cases, missing chromosomal material (deletion) involves the unique DNA in mitochondria. Observed symptoms may include atypical retinitis pigmentosa (accumulation of pigmented material), progressive deterioration of certain eye structures, optic nerve atrophy, hearing loss leading to deafness, heart disease, mental retardation, retardation of growth, ataxia, deterioration of intellectual abilities, etc. Males and females are equally affected. Symptoms of the disorder are usually apparent by age 20, with eye abnormalities and developmental delay observed before the age of 5 years. Source: National Organization on Rare Disorders: www.rarediseases.org

Leber's Optic Neuropathy

See Leber's described under Genetic Disorders.

5.4.10 Non-inherited Genetic/Chromosomal Abnormalities

Chromosomal abnormalities may include chromosomal deletions (when a part of a chromosome has been deleted); chromosome duplication (duplication of a section of a chromosome also referred to as partial trisomy which results in three copies of the genetic material); chromosomal ring (fusion of arms of chromosomes which can, may or may not, result in a loss of chromosomal material); chromosomal inversions (two breaks in one chromosome) and chromosomal trans-locations (unbalanced rearrangement of parental chromosomal material). These abnormalities can lead to birth defects, cognitive abnormalities and increased risks of miscarriage. Most chromosomal abnormalities occur spontaneously as an accident during meiosis in the egg or sperm. Some abnormalities can happen after conception, while some abnormalities can be inherited from a parent. A disease or disorder that has more than one identifying feature or symptom is a syndrome. Source: Chromosome Deletion Outreach, Inc, July 2000: Introduction to Chromosome Abnormalities: http:/www./members.aol.com

Apert's Syndrome (also known as Acrocephalo-syndactyls Type 1)

One of a group of syndromes which are characterized by the premature fusion of the bones of the skull, together with malformations of the feet. Occurs in between 1/100,000 and 1/150,000 births. Most incidences are recognized as new mutations. Congenital hearing loss is prevalent and mild learning disability occurs in half the cases. Hydrocephalus is a frequent complication of this syndrome which likely contributes further to multi-sensory deprivation. Source: Gilbert (1996)

Arnold Chiarr Syndrome

A rare malformation of the brain that is present at birth. The abnormality at the base of the brain may include the displacement of the lower portion of the brain and/or brain stem through the opening at the back of the skull. Hydrocephalus is often associated with this rare disorder. Symptoms usually include varying degrees of mental impairment, muscle weakness in head and face; other symptoms may include double vision, deafness and ataxia. Source: National Organization on Rare Disorders: http://www.rarediseases.org

CHARGE Syndrome/Association

A disorder which is an acronym for the conditions: C (coloboma of the eye); H (heart defects); A (choanal atresia); R (retarded growth); G (genital hypoplasia) and E (ear abnormalities). CHARGE occurs in males twice as frequently as females. Most cases occur spontaneously at conception, but both autosomal dominant and recessive modes of inheritance have been described. The disorder has been associated with deletion of chromosome 22q11. (See Velocardiofacial Syndrome) Source: Gilbert (1996); http://www.ibis-birthdefects.org

Cornelia de Lange Syndrome

A congenital syndrome of unknown cause. Not believed to be caused by an inherited gene, but believed to be the result of spontaneous chromosomal abnormality at conception. Common characteristics include low birth weight,

translocation or unbalanced rearrangement of parental chromosomal material. The disorder includes defects to head and brain (including microcephaly), ears, eyes (including microphthlmia and coloboma), urogenital and cardiovascular system. Growth and mental development are severely affected. Source: www.proaxis.com and United States National Library of Medicine at www.nlm.gov/mesh/jablonski/syndromes/

Down Syndrome (Trisomy 21)

The most well known chromosomal abnormality with an incidence of between 1 in 660 and 1 in 800 live births. Can arise in two ways: an extra chromosome can spontaneously occur in the Chromosome 21 position (hence trisomy 21) during the production of eggs or sperm; or an extra chromosome can be added by translocation to another chromosome. The former occurs in 95% of the cases, while the latter in 5% of the cases. The risk of occurrence in the first case rises with maternal age. Symptoms include developmental delay, eye abnormalities (including cataracts and nystagmus), congenital heart disease, thyroid disease and middle ear infections that can lead to deafness. Source: Gilbert (1996)

Klippel Fiel Syndrome

A rare disorder characterized by complications to the spinal column, and in some circumstances associated with eye defects, deafness (either conductive or sensorineural), and defects to the urinary and cardiovascular system. Most cases arise sporadically, but there have been observations of genetic transmission. Inheritance can be either autosomal recessive or autosomal dominant. It is believed that more than two separate genes may be involved. Incidence reported to be 1 to 40,000 live births, girls being affected more than boys. Source: Gilbert (1996)

Pallister-Killian Mosaic Syndrome (chromosome 12p tetrasomy)

A rare congenital disorder caused by chromosome 12p tetrasomy (four copies of chromosome) in the affected cells, resulting in distinct cranial defects and neurological manifestations. Symptoms include seizures, profound mental retardation, hydrocephalus, sensorineural hearing loss, craniofacial defects involving the eye (strabismus, ptosis and occasionally cataracts). Males and females are affected equally.
Source: http://icondata.com/health/pedbase/files/PALLISTE.HTM

Ring Chromosome 18 Syndrome

This is one of the three deletion abnormalities (18q-, 18p- and ring 18) of chromosome 18, and some of the most common autosomal deletion syndromes. This particular syndrome occurs when the usually linear chromosome forms a ring. Ring 18 can appear to be like 18q- or 18p- or a combination of both. Most 18q deletions occur spontaneously upon conception. But in some cases this syndrome can occur if the parent has a balanced translocation (showing no personal symptoms) but may pass it on to their child. Symptoms include mental retardation, seizures, microcephaly, congenital heart disease, nystagmus, strabismus, impaired hearing, coloboma of the eye, etc. Source: The Chromosome 18 Registry & Research Society. The Deletion Syndromes of Chromosome 18. 9 pages, 2001; http://www.chr18.uthscsa.edu/syndrome.html

Septo-Optic Dysplasia (de Morsier's Syndrome)

A rare disorder characterized by abnormal development of the optic disk, pituitary difficulties and often the absence of the septum pellicidum or that part of the brain that separates the lateral ventricles of the brain. Symptoms may include blindness in one or two eyes, seizures and intellectual difficulties in some instances. Most individuals with this disorder are developmentally delayed and visually impaired.
Source: National Institute of Neurological Disorders and Stroke: http://www.ninds.nih.gov.health and www.medical/disorders/septo-optic.htm

Trisomy 22 (partial Trisomy 11;22; unbalanced 11;22)

Disorder caused by translocation of 11^{th} and 22^{nd} chromosomes. Children born with this disorder have a marker chromosome made up of the upper (p section), a part of the lower (q section) arms of chromosome 22 and a small portion of the lower (q section) arm of chromosome 11. This disorder is usually inherited from a carrier parent, although the disorder can occur spontaneously at conception. Common symptoms of the disorder may include cleft palate, heart defects, ear abnormalities, male genital abnormalities and moderate to severe mental

behavioural problems (in 50% of the cases). Incidence reported as between 1 to 30/40000 live births. In most cases, the disorder is caused by sporadic mutations. It also has been observed as an inherited autosomal dominant trait. Source: Gilbert (1996); National Organization on Rare Disorders: http://www.rarediseases.org

Wolf Hirschhorn Syndrome (4-p deletion syndrome)

A rare disorder resulting from the deletion of genetic material on the short arm (p) of chromosome 4. In most cases the condition arises spontaneously, but in about 10% of the cases it can result from "balanced translocation". Symptoms include severe growth retardation and mental defect, microcephaly, cleft palate or lip, coloboma of the eye, seizures and cardiac defects. Source: Gilbert (1996); http://www.ibis-birthdefects.org/start/wolfhirs.htm

Velo Cardio Facial Syndrome (also called chromosome 22q11 deletion, Shprintzen, DiGeorge and Catch 22 Syndrome)

A specific genetic disorder of chromosome 22 characterized by a small piece missing. A disorder that has been associated with over thirty different features. May occur spontaneously at conception, in most instances, and through parental inheritance to a lesser degree. Syndrome includes a range of conditions, e.g. congenital heart or cardiac disease, palatal or velum abnormalities, characteristic facial features. Disorder is linked to learning disabilities, behavioural disorders, speech and language deficits and a broad range of cognitive disabilities. Many individuals have physical defects with the eyes. Source: http://www.ibis-birthdefects.org

13q Minus Ring Syndrome

An abnormality of chromosome 13 whereby chromosomal material from the q arm breaks off and fuses with the p arm, forming a ring chromosome with the consequential loss of chromosomal material. Source: See details on Ring Chromosome 18 Syndrome

18q Minus Ring Syndrome

Source: See details on Ring Chromosome 18 Syndrome

5.4.11 Prematurity (Complications)

A child is premature if born less than 37 weeks gestation. Complications of prematurity can include blindness, hearing loss and physical impairment. Blindness is common among premature children and is referred to as Retinopathy of Prematurity (ROP). ROP is a disease of the retina that prevents the eye from developing properly and can lead to permanent damage and blindness. Source: http://www.rdbraille.com/pbpb.html

5.4.12 Prescribed Drugs (Complications)

Large doses of prescribed drugs, in particular those of the *mysin* family of drugs, and massive life saving doses have been cited as causes of visual and hearing loss in some persons. In this study two specific incidents were reported, one attributed to quinine and another to an unspecified prenatal prescribed medication.

5.4.13 Post Natal / Early Childhood Infections

Various bacterial and viral infections occurring in the immediate post-natal period or in early childhood.

Meningitis

An infection of the meninges or membranes that surround the brain and spinal column, as the result of a bacterial or viral infection. An especially serious infection is caused by the meningococcus *Neisseria meningitidis*, which can result in convulsions, paralysis, blindness, deafness, neck stiffness, headache, etc, Source: Stedman's Medical Dictionary (1972)

Scarlet Fever

An acute disease characterized by a skin eruption of bright red colour accompanied by a fever. Source: Stedman's Medical Dictionary (1972)

Whooping Cough or Pertussis

Mumps or Parotiditis

An acute contagious viral infectious disease characterized by inflammation and swelling of the parotid gland. Occasionally causes inflammation of testes, ovaries, pancreas and meninges (membrane surrounding the brain and spinal cord).

5.4.14 Self abuse

Self inflicted abuse that can result in visual and hearing impairment.

5.4.15 Sexually Transmitted Diseases

Viral and bacterial diseases transmitted by sexual contact. Examples include syphilis, gonorrhea and various herpes diseases.

Congenital Syphilis

An acute and chronic infectious bacterial disease caused by *Tripenema pallidum* and transmitted by direct contact, usually by sexual intercourse. Referred to as congenital if acquired by the fetus, in utero and present at birth. Symptoms include cardiovascular and central nervous system lesions during the third or final stage of the disease. Source: Stedman's Medical Dictionary (1972)

Neonatal Herpes Simplex (HSV)

A pathogen of herpes simplex in man, causing acute stomatitis (inflammation of the mucous membrane of the mouth), especially in children, and so called fever blisters usually on the lips and external nares. Source: Stedman's Medical Dictionary (1972)

5.5 Definitions of various communication modes (Source: Joyce Thompson, personal communication, 2001)

Adapted Sign Language - used by a person with deafblindness who places their hands over the hands of the signer to feel the hand movements that indicate words, actions or letters. Also used by persons who are deaf and have little to no vision.

Audio Tapes - receptive communication system used by persons who have hearing. Expressive communication system used by persons with speech to send taped messages to hearing family members and friends.

Braille - Grade 1 - a system of raised dots that indicate letters of the alphabet.

Braille - Grade 2 - a sophisticated system that takes considerable effort to learn where words are contracted using the embossed/raised dots of braille. Used by persons whose severe vision loss prevents them from reading print, even with magnifiers.

Closed Caption Decoders - are used to pick up captions transmitted over the TV cable which are hidden from view without the decoder. Text of some programs are available for access to television if the person has enough vision to catch the main idea of the program.

Gestures - are usually a personally created system of body and hand movements developed by family members or institutional care workers who have never taken the extensive sign language courses required to communicate with persons who are deaf or deafblind.

Manual Alphabet - a tactile system of spelling each word by tapping different points on the palm of the person with deafblindness. Used by many persons who have total deafblindness.

vision.

Speech - Can be expressive and/or receptive and means that the person with deafblindness once heard or still can hear, through amplification, and is usually augmented by lip reading.

deafblind and a braille reader, when travelling alone or in hospital. This machine is no longer produced.

TeleBraille - is a telephone device similar to the Tellatouch but has a braille display for persons who are deafblind and able to read braille.

TTY-TDD (Teletype or Telephone Device for the Deaf) are typical keyboard devices with printer displays which connect to the telephone line by an acoustic coupler for the telephone receiver or directly connected to the telephone line by a jack. Messages are typed back and forth to another similar machine; or to the telephone relay operator, who voices the transmitted message to the hearing person being called and keys back the hearing person's message to the deafblind person's device.

Writing/Printing - used by a person who is deafblind (deaf with some remaining vision) to communicate with persons who do not know the deafblind person's communication system.

5.6 Definitions of various aids to communication (Source: Joyce Thompson, personal communication, 2001)

Brailler is a mechanical device with 6 keys and space bar that embosses on braille paper the dots that symbolize letters of the alphabet. The most common mechanical device is the sturdy and heavy Perkins Braillewriter that has not changed since it was first produced 50 years ago. Price: in excess of $1,000 CAD.

CCTV Reader (Closed Circuit TV readers) is a camera that magnifies print (up to 60 times) onto a TV-like screen, with the option of converting the text to the negative mode (white print on a black background), which is preferred by many people with low vision. This device costs approximately $3,000.

Cochlear implants are made up of a microphone placed outside the ear and a processor implanted inside the ear. The processor selects and codes sounds so the brain will understand them. The "messages" are sent directly to the brain, bypassing the parts of the inner ear that don't work. Cochlear implants are now being sought by more persons with deafblindness.

Computers are often used in meetings. A note taker will type the proceedings of a meeting on the computer keyboard and the deaf/low vision person will read the scrolling text set at the font size and contrast needed by the individual. Several screens can be linked so that many people are served by the note takers; or the text can be projected onto a screen if the audience is deaf. This is not satisfactory for persons with deafblindness.

audience.

Hearing aids are used by many people who cannot hear speech but have residual hearing to hear sirens, and environmental sounds. This ability to hear sounds provides persons with deafblindness some awareness of the world around them and greatly assists them with their safety.

Internet is a system for individuals to electronically connect with people, via a computer, and obtain information via the World Wide Web. Unfortunately many people with deafblindness do not have access to training or the resources to purchase a computer to access the Internet.

Magnifiers are devices used to magnify print, and may include the hand and pocket variety, large and small, and can include a lighted part.

Monoculars are ocular devices used outside to read street signs and numbers or used inside to read chalk boards.

Slate and Stylus is a metal or plastic plate with a frame to hold braille paper. The user uses a stylus (a blunt metal rod with a knob) to impress the combination of dots used to indicate letters.

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