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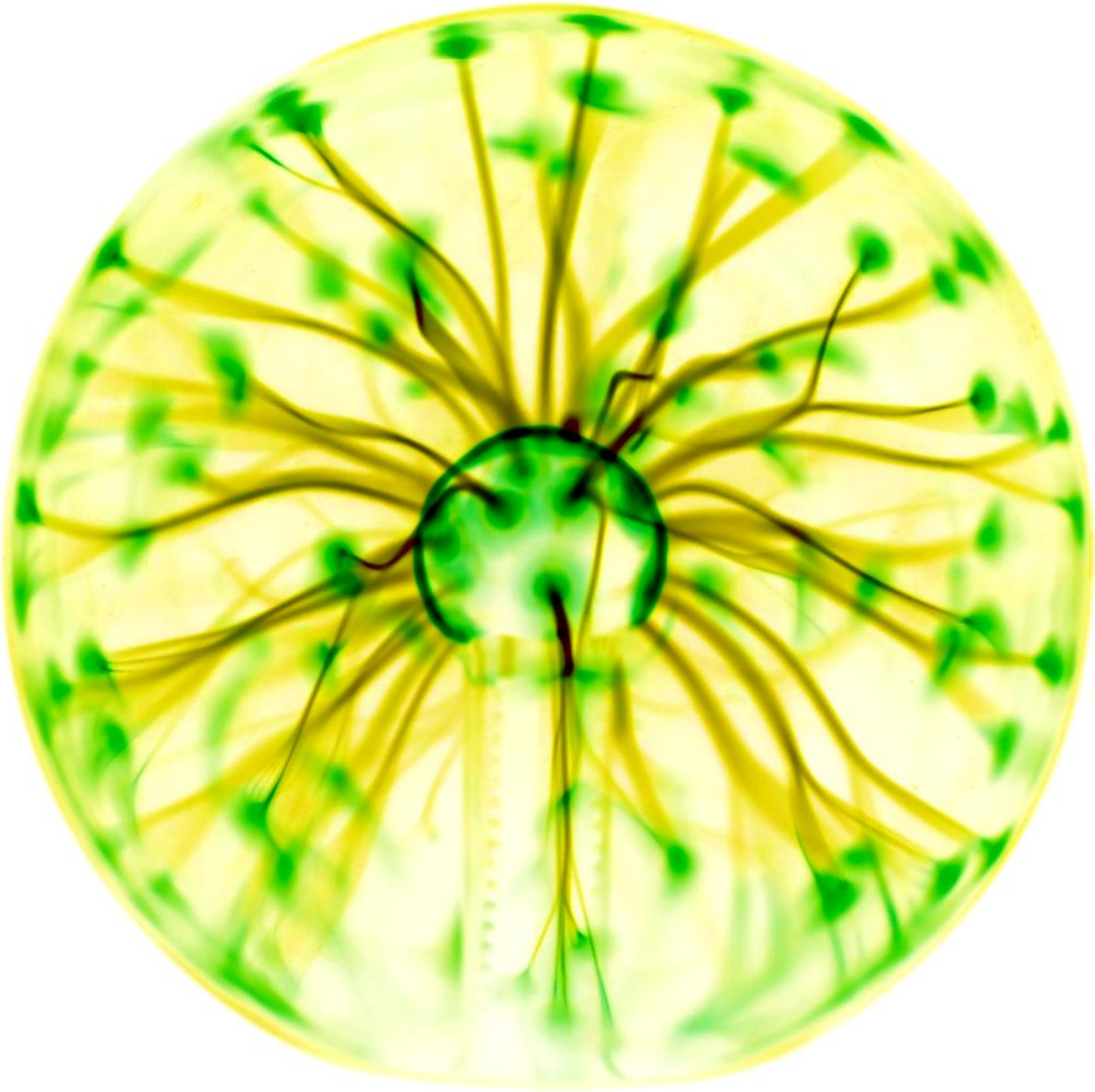


The socioeconomic impact of inherited retinal dystrophies (IRDs) in Canada

Retina International

October 2020

Deloitte
Access Economics

Contents

Glossary

Acronym	Full name
AMD	age-related macular degermation
AWE	average weekly earnings
BBS	Bardet-Biedl Syndrome
BCM	Blue cone monochromacy
CADTH	Canadian Agency for Drugs and Technologies in Health
CBA	cost benefit analyses
CCC	Canada caregiver credit
CDR	Common Drug Review
CEA	cost effectiveness analyses
CIHI	Canadian Institute for Health Information
CIHR	Canadian Institutes of Health Research
CMA	cost minimisation analyses
CNIB	Canadian National Institute for the Blind
CRX	cone-rod homeobox
CUA	cost utility analyses
DALY	disability adjusted life year
DLTV	Daily Living Tasks Dependent on Vision Questionnaire
DTC	disability tax credit
DWL	deadweight loss
EMA	European Medicine Agency
GP	general practitioner
GPS	global positioning system
GUCY2D	guanylyl cyclase 2D
IRD	inherited retinal dystrophy
LCA	Leber congenital amaurosis

Acronym	Full name
LHON	Leber hereditary optic neuropathy
NA	not applicable
NCT	National Clinical Trial
NHS	National Health Service
ODSP	Ontario Disability Support Program
OECD	Organisation of Economic Cooperation and Development
OHIP	Ontario Health Insurance Plan
RAMQ	*Régie de l'Assurance Maladie du Québec*
RP	Retinitis pigmentosa
RPE65	retinal pigment epithelium-specific 65
SEN	special education needs
UK	United Kingdom
US	United States
USA	United States of America
USH1	Usher syndrome Type I
USH2	Usher syndrome Type II
USH3	Usher syndrome Type III
UV	ultraviolet
VI	visual impairment
WHO	World Health Organisation
XLRS	X-linked retinoschisis
YLD	years lived with disability
YLL	years of life lost due to premature death

Foreword

This work was funded and supported by a consortium of patient organisations and industry partners led by Retina International, who recognized the need to better understand the real-world impact of inherited retinal dystrophies (IRDs) in Canada and the United States of America (USA). The consortium partners involved in this study were AGTC, Fighting Blindness Canada, Foundation Fighting Blindness USA, Janssen Global, Novartis, and Retina International. Each partner funded and assisted in the design of the project and recruitment of survey participants.

The socioeconomic impact of inherited retinal dystrophies (IRDs) in Canada in 2019

Methodology

The socioeconomic burden of inherited retinal dystrophies (IRDs) in Canada was estimated using cost-of-illness methodology applying a prevalence approach (Larg & Moss, 2011). This approach involves estimating the number of people with IRDs in a base period (2019) and the costs attributable to IRDs in that period. The analysis was based on a targeted literature review and primary data (survey) collection.

Prevalence

Overall, the prevalence of IRDs in Canada was estimated to range between 0.056% to 0.228% (lower to upper bound) or between 20,947 to 85,672 prevalent cases in 2019. The highest proportion of the overall prevalence was attributed to rod-cone dystrophy (between 27.8% to 27.9% (lower to upper bound) or between 803 to 23,891 people).

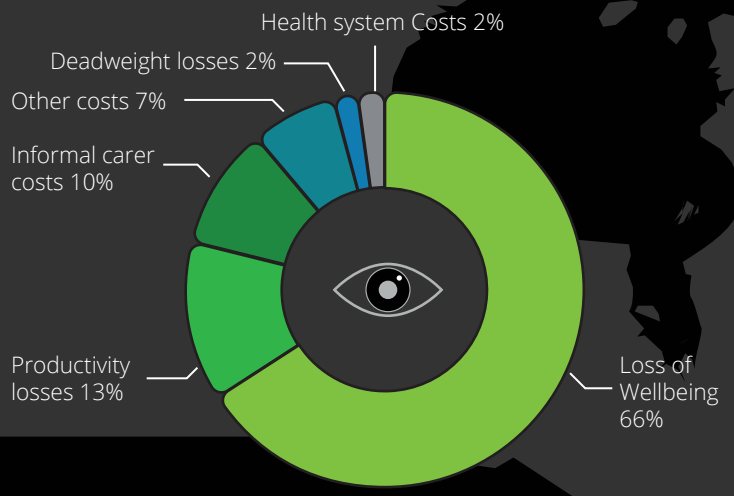
Prevalence of IRDs in Canada (n, 2019)

IRD	Cases (Lower)	Cases (Upper)
Rod-cone dystrophy	5,841	23,891
RP	5,803	23,736
Autosomal dominant RP	*1,818*	*7,347*
Autosomal recessive RP	*2,679*	*9,607*
X-linked RP	*1,307*	*6,782*
Stargardt disease	4,163	17,027
Cone-rod dystrophy	1,302	5,323
Choroideremia	748	3,060
Best disease	694	2,840
Usher syndrome	584	2,387
XLRS	577	2,360
LCA	329	1,347
Cone dystrophy	310	1,270
BCM	217	889
BBS	146	596
LHON	131	535
Achromatopsia	101	412
Total	**20,947**	**85,672**

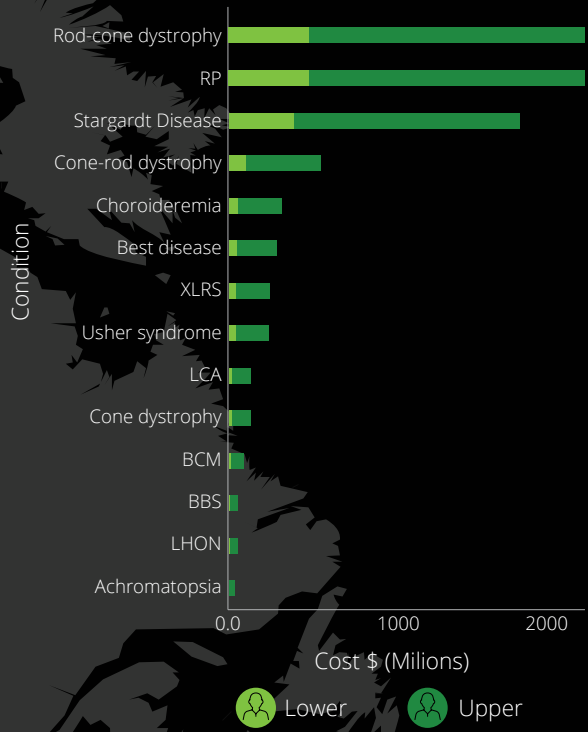
Total costs

Total costs attributable to IRDs in Canada were estimated to range between **$1,638 to $6,688** million in 2019.

Proportion (%) of total costs of IRDs in Canada by cost type (2019) (lower and upper bound scenario)



Total costs of IRDs ($ million, 2019) in Canada by condition (lower and upper bound scenario)



Conclusion

IRDs imposed significant economic and wellbeing costs in the Canadian population in 2019. Persons living with an IRD incur significant economic costs and reductions in their quality of life. Significant economic costs are also borne by families, friends, government, employers and society.

1 Inherited retinal dystrophies

Deloitte Access Economics (Australia) was engaged by Retina International to estimate the socioeconomic burden of 14 inherited retinal dystrophies (IRDs) in Canada.

IRDs represent a diverse group of progressive, visually debilitating diseases in which genetic mutations that are critical to retinal function lead to progressive photoreceptor cell death and associated vision loss (Duncan et al, 2018). IRDs are a significant cause of vision loss and are characterized by the loss of photoreceptors and the retinal pigment epithelium (RPE) (Hafler et al, 2017).

For patients with IRDs, there have traditionally been no effective treatments to restore vision. The development of treatments and cures to modify the rate of disease progression has been limited to date, with some success of neurotrophic factor therapy and gene therapies reported from clinical trials. The best example of treatment success, which recently received US Food and Drug Administration (FDA) approval, and represented the first FDA-approved gene therapy for any genetically inherited disease, is gene augmentation therapy for IRDs caused by mutations in the *RPE65* gene (RPE65-associated Leber congenital amaurosis) (Duncan et al, 2018; Hafler et al, 2017).

There are three types of inheritance patterns that can lead to an IRD, including autosomal dominant, autosomal recessive, and X-linked. However, some IRDs can also arise through new genetic variants meaning that individuals with no family history can be affected.

The 14 IRDs included in this analysis are described below.

1.1 Retinitis pigmentosa (RP)

Retinitis pigmentosa (RP) is a group of rare, genetic disorders which result in progressive vision loss (Campochiaro and Mir, 2018; Ferrari et al, 2011). Symptoms of RP include impaired night vision, deteriorated central vision and a loss of side (peripheral) vision (Verbakel et al, 2018).

There are currently over 80 genes associated with RP (Verbakel et al, 2018). Genetic testing can be undertaken to determine the presence of these genetic variants; however, this is challenging due to the significant genetic diversity observed in RP causing variants (Birtel et al, 2019). Diagnostic imaging techniques can be used to diagnose RP and assess disease severity and progression (Verbakel et al, 2018). One form of RP occurs when there are variants in both copies of a gene called 'RPE65'.

RP is typically diagnosed in young adulthood, however the age of onset may range from early childhood to the mid-30s to 50s. Research has shown that the average age at diagnosis for RP was 35.1 years, while the study also found that individuals had been diagnosed with RP from 1 year up to 89 years of age (Tsujikawa et al, 2008).

RP can be inherited through one of three inheritance patterns including autosomal dominant (about 30–40% of cases), autosomal recessive (50-60% of cases) or X-linked recessive (5-15% of cases) (Campochiaro and Mir, 2018; Daiger et al, 2015; Hartong et al, 2006). These inheritance patterns are further described below.

Voretigene neparvovec (Luxturna) is a new gene therapy for the treatment of patients with confirmed biallelic *RPE65* mutations, often clinically diagnosed as RP or Leber congenital amaurosis (LCA, described in Section 1.9). Patients must have sufficient viable retinal cells in order to qualify for treatment with the gene therapy (European Medicines Agency (EMA), 2019). Luxturna is designed to deliver a normal copy of the *RPE65* gene directly to retinal cells to help restore patients' vision (Ziccardi et al, 2019; US Food and Drug Administration, 2017).

The Canadian Agency for Drugs and Technologies in Health (CADTH) is currently evaluating the use of Luxturna through the Common Drug Review (CDR) process. The CDR process involves a thorough and objective review of studies that report on clinical effectiveness, safety, and cost-effectiveness of drugs under review. The draft recommendation for the approval and reimbursement of Luxturna is expected to be issued in September 2020 (CADTH, 2020). If Luxturna is approved for use in Canada, Novartis will hold the rights to the gene therapy, as it does for countries outside the US which have already approved the treatment (CADTH, 2020).

More recently, the safety and effectiveness of a bionic vision system (Intelligent Retinal Implants System II) has been evaluated for blind patients with RP, cone-rod dystrophy (described in Section 1.4) or choroideremia (described in Section 1.5); however, the results from the clinical trial have not yet been published (National Clinical Trial (NCT) number: NCT02670980 ClinicalTrials.gov, 2020).

1.1.1 Autosomal dominant RP

In autosomal dominant RP, only one copy of the mutated gene needs to be inherited to bring about the condition. An individual with autosomal dominant RP has a 50% chance of passing the mutated gene onto each of their offspring. Like other forms of inherited retinal disease, autosomal dominant RP is exceptionally diverse with more than 1000 variants having been reported in the 25 genes which are known to cause the condition (Daiger et al, 2015).

1.1.2 Autosomal recessive RP

In autosomal recessive RP, two copies of the mutated gene must be inherited to give rise to the condition. An individual with only one recessive gene variant is known as a carrier and will not be affected by the condition. When two carriers of an autosomal recessive variant have a child there is a one in four (25%) chance that that child (and each of their subsequent children) will have the disorder. At least 35 genes have been associated with autosomal recessive RP (Daiger et al, 2013; Daiger et al, 2007).

1.1.3 X-linked recessive RP (XLRP)

In X-linked RP (XLRP), the mutated gene is stored on the X chromosome which is a sex chromosome as opposed to an autosomal (non-sex) chromosome. This means women can either be affected by XLRP, carrier of XLRP or not affected by XLRP whereas males can only be affected or not affected by XLRP. This is because women have two X-chromosomes while men have one X-chromosome and one Y-chromosome.

XLRP is particularly severe in males, characterised by early onset and rapid progression of vision loss, resulting in legal blindness by the third decade. Most female carriers do not report symptoms or present with clinically relevant changes (Verbakel et al, 2018; Martinez-Fernandez et al, 2018).

1.2 Rod-cone dystrophy

Rod-cone dystrophy is characterised by progressive night blindness and visual field loss resulting from degeneration to the 'rod' components of the retina and, to a lesser extent, the 'cone' components (Ahn et al 2020). There is wide clinical diversity in rod-cone dystrophy and more than 80 genes have been associated with the condition so far (Ahn et al 2020). In addition, new genetic causes of rod-cone dystrophy are rapidly being discovered due to recent advances in genetic analysis techniques. These discoveries are anticipated to assist researchers in uncovering future therapeutics for the condition (Sorrentino et al, 2016; Salmaninejad et al, 2019).

Rod-cone dystrophy is typically diagnosed in young adulthood (ages 18 to 24 years); however, the age of onset may vary from early childhood (ages 0 to 8 years) through to a person's 30s or 50s. Rod-cone dystrophy is suspected in patients with poor night vision, and confirmed following genetic testing, electroretinography or visual field testing (Pennesi et al, 2010).

Low vision aids including high-intensity lamps, contrast enhancing filters, infrared blocking sun lenses and magnifiers can be used by people with rod-cone dystrophy to maintain or improve their quality of life. Regular eye examinations are also necessary to track the rate and degree of visual deterioration (Hamel et al, 2006).

1.3 Stargardt disease

Stargardt disease is an autosomal recessive disease characterised by declining central vision including an inability to perceive colors (dyschromatopsia) and the emergence of blind spots (scotoma) (Han et al, 2014; Tanna et al, 2017). Stargardt disease is one of the most common causes of macular disease in childhood, with symptoms typically first presenting in early childhood or adolescence (Soriano and López, 2017).

ABCA4 is the primary gene associated with Stargardt disease, though there are over 900 disease-causing variants identified in this gene (Tanna et al, 2017). Genetic testing for Stargardt disease is particularly difficult due to the genetic diversity of the disease (Tanna et al, 2017). As such, retinal imaging technologies can be used as a diagnostic tool (Ergun et al, 2015).

There is currently no treatment for Stargardt disease; however, limiting exposure to ultraviolet (UV) light by wearing UV screening glasses and limiting intake of vitamin A is advised as part of disease management strategies. Research into treatment strategies is highly intensive with gene therapy emerging as a potential option (Han et al, 2014; Kumaran et al, 2018a). Stem cell therapies are also being investigated (Tanna et al, 2017; Schwartz et al, 2015). In addition, two late stage clinical trials investigating an oral therapy are underway (Fighting Blindness Foundation, 2019).

1.4 Cone-rod dystrophy

Cone-rod dystrophy is characterised by poor visual acuity, light sensitivity (photophobia), nystagmus, colour blindness (dyschromatopsia) and impaired night vision (nyctalopia) (Gill et al, 2019). Cone-rod dystrophy differs from cone dystrophy in that it is associated with earlier degeneration of cone components of the retina (Thiadens, 2012).

Diagnosis of cone-rod dystrophy can be facilitated by genetic testing and imaging techniques (Gill et al, 2019). The prognosis of cone-rod dystrophy is typically more severe and rapid relative to other IRDs, leading to earlier total blindness. However, the age of onset varies ranging from the second to the fifth decade of life (Thiadens et al, 2012).

There are currently no treatments to prevent the progression of cone-rod dystrophy (Gill et al, 2019). Care strategies focus on alleviating symptoms, including refractive correction, tinted lenses and low vision aids (Gill et al, 2019).

1.5 Choroideremia

Choroideremia is an X-linked retinal dystrophy characterised by progressive night-blindness, tunnel vision and vision loss with onset typically in the first two decades of life (Tsang and Sharma, 2018; Sahel et al, 2015). Other symptoms of choroideremia include tunnel vision and, in some cases, central blindness (MacDonald et al, 2015). Choroideremia typically affects males, although some symptoms have been reported in female carriers.

Choroideremia is caused by variants in the *CHM* gene and genetic testing can be used to screen for the presence of this genetic variant (MacDonald et al, 2015). Diagnosis of choroideremia can be

confirmed through diagnostic imaging technologies (Coussa and Traboulsi, 2012).

Current approaches to choroideremia management focus on symptom alleviation through surgery as well as through non-surgical methods such as dietary modification and visual aids (MacDonald et al, 2015). Clinical trials of gene therapies are ongoing (Kumaran et al, 2018a; Mitsios et al, 2018; NCT02670980 ClinicalTrials.gov, 2020).

1.6 Best disease

Best disease is an autosomal dominant IRD that is characterised by the build-up of fatty yellow pigments in cells underneath the macula which, over time, can damage cells that are essential for clear central vision (Parodi et al, 2014). As such, Best disease is often referred to as the genetic form of macular degeneration. The symptoms associated with Best disease include slowly worsening visual clarity, central vision and occasionally blurry or distorted perception of straight lines (Zerbib et al, 2016).

The usual age at onset is within the first two decades and detection is usually incidental (Tripathy et al, 2018). It has also been noted that the macular lesion can be detected for the first time as late as 75 years of age (Mullins et al, 2005). Diagnosis of Best disease can be facilitated by genetic testing and imaging techniques (Parodi et al, 2014; Budiene et al, 2014). There are no treatments available to prevent the onset or progression of Best disease, though symptoms can be managed with visual aids (Budiene et al, 2014). Clinical trials, using gene and cell-based therapies, are ongoing (Yang et al, 2015). A current clinical trial is exploring the use of stem cells to identify and test therapeutic approaches. The study is expected to reach completion in 2021 (NCT02162953 ClinicalTrials.gov, 2020).

1.7 Usher syndrome

Usher syndrome is an autosomal recessive condition characterised by hearing loss and progressive retinal degeneration. Usher syndrome is the most common deaf-blindness caused by genetic factors with 11 genes currently linked to the condition (Mathur and Yang, 2019; Yang et al, 2012). The visual symptoms of Usher syndrome are identical to those associated with RP while individuals with Usher syndrome will also experience a loss of auditory function

and may encounter difficulties with balance (Mathur and Yang, 2015; Mathur and Yang, 2019).

There are three major clinical subtypes of Usher syndrome; Type I (USH1) Type II (USH2) and Type III (USH 3). Usher syndrome Type I is further subdivided into 5 types; mutations in the *MYO7A*, *USH1C, CDH23, PCDH15,* and *USH1G* genes cause Usher syndrome Type 1B, Type 1C, Type 1D, Type 1F and Type 1G respectively. Mutations in these genes account for most cases of USH1. Mutations in *MYO7A* are the most common accounting for 39-55% of cases (Keats and Lentz, 2006).

Persons with Type I (USH1) have congenital severe-to-profound deafness, disrupted sensory function and onset of RP within the first decade of life. While persons with Type II (USH2) have congenital moderate-to-severe hearing loss, normal vestibular function and onset of RP within the second decade of life. By comparison, persons with Type III (USH3) experience progressive hearing loss, sporadic vestibular dysfunction and variable onset of RP are progressive (Mathur and Yang, 2015).

Diagnosis of Usher syndrome can be facilitated by genetic testing and imaging techniques (Mathur and Yang, 2015). While the auditory symptoms of Usher syndrome can be managed using aid devices, there is no treatment currently available for the visual component of the condition (Mathur and Yang, 2019).

Gene replacement therapy is emerging as a potential approach to prevent progression to total blindness. For example, a current clinical trial is evaluating the long-term safety, tolerability and biological activity of a sub-retinally injected gene therapy for persons with Usher Syndrome Type 1B (NCT02065011 ClinicalTrials.gov, 2020; Mathur and Yang, 2015; Takahashi et al, 2018).

1.8 X-linked retinoschisis (XLRS)

X-linked retinoschisis (XLRS) is a rare congenital disorder characterised by central vision loss, involuntary eye movements and/or crossed eyes. More severe cases will exhibit signs of retinal detachment and/or glaucoma which can result in extensive vision loss (Vincent et al, 2013; Rao et al, 2018; Sahel et al 2015). Significant progress has been made in understanding XLRS in recent

years with over 190 different variants having been identified in the gene which is known to cause XLRS (*RS1)* (Molday et al, 2012).

Management of XLRS generally relies on pharmacological or vision-aid interventions (Molday et al, 2012). Surgical options may also be used to address more severe complications associated with the condition, such as retinal detachment (Rao et al, 2018).

XLRS can be diagnosed using retinal imaging techniques (Vincent et al, 2013; Sieving et al, 2014). There is currently no cure for XLRS; however, gene replacement therapy is being explored (Kumaran et al, 2018a; Dalkara et al, 2013; Delgado et al, 2012).

1.9 Leber congenital amaurosis (LCA)

Leber congenital amaurosis (LCA) is a congenital disorder that results in severe vision loss at an early age, typically presenting between birth and the first few months of life (Kumaran et al, 2017; Kumaran et al, 2018b). LCA is usually inherited through an autosomal recessive pattern, although an autosomal dominant inheritance pattern can occur in rare cases (Kumaran et al, 2018b; Sahel et al, 2015). Presently, 25 genes responsible for LCA have been identified; accounting for 70-80% of cases (Kumaran et al, 2017).

Symptoms commonly associated with LCA include early-onset visual impairment, eye oscillations, slow/absent pupil reactions, heightened light sensitivity, far-sightedness and cornea bulging (Kumaran et al, 2017). In some cases, children with LCA may present with neurodevelopmental delays, intellectual disability, autism and reduced sense of smell (Chacon, Camacho and Zenteno, 2015; Sahel et al, 2015).

LCA can be screened for through genetic testing which aims to detect variations in the genes most commonly associated with the conditions (Kumaran et al, 2018b). Other diagnostic tools can be used to assess symptoms of LCA (Kumaran et al, 2018b).

Treatment of LCA currently focuses on symptom management such as the correction of refractive error and provision of low vision supports (Kumaran et al, 2018b). In addition, Luxturna, a new gene therapy, is being evaluated by The Canadian Agency for Drugs and Technologies in Health (CADTH), through the Common Drug Review (CDR) process, for the treatment of patients with inherited retinal

disease (including cases of LCA) caused by biallelic mutation of *RPE65* gene (Marlhens et al, 1997; den Hollander et al, 2008; Cideciyan, 2010). The draft recommendation for the approval and reimbursement of Luxturna is expected to be issued in September 2020 (CADTH, 2020).

Ongoing clinical trials are continuing to investigate other gene therapies as well as stem cell therapies (Kumaran et al, 2018c).

1.10 Cone dystrophy

Cone dystrophy is characterised by degeneration of the cone components of the retina (Sisk et al, 2018). Symptoms that most commonly present include difficulties in adjusting to bright light, colour vision disturbances, reduced visual clarity and involuntary eye movements (nystagmus) (Sisk et al, 2018). Persons with cone dystrophy present as either stable or progressively declining. The former typically present with cone dystrophy symptoms during early childhood and remain stable. Progressive cone dystrophy usually develops in late childhood or during early adulthood and worsens over time (Gill et al, 2019).

Six genes are responsible for most cone dystrophy cases; inherited in an autosomal dominant or autosomal recessive pattern (Gill et al, 2019). Individuals can be screened for cone dystrophy disease-causing genes using molecular genetic testing (Gill et al, 2019). There are also several diagnostic imaging techniques that can be used to diagnose cone dystrophy (Sisk et al, 2018).

There are currently no treatments available for slowing progression or restoring visual impairment. Management of cone dystrophy relies on the alleviation of visual symptoms (Gill et al, 2019). However, there are several clinical trials underway evaluating the use of gene replacement therapy (Gill et al, 2019).

1.11 Blue cone monochromacy (BCM)

Blue cone monochromacy (BCM) is a rare congenital disorder with an X-linked recessive inheritance pattern. BCM is characterised by severe colour vision loss (Nathans et al, 1989). BCM is caused by variants in genes located on the X chromosome which are responsible for controlling the perception of red and green colours. The perception of blue colours is controlled by genes located on

autosomal chromosomes, meaning they are not affected by BCM (Luo et al, 2015; Katagiri et al 2018).

Persons living with BCM typically experience considerably reduced central vision, severely impaired color discrimination, reduced visual clarity, bright light sensitivity, and involuntary eye movements (Katagiri et al, 2018; Michaelides et al, 2005).

A family history analysis in addition to several diagnostic tools including a colour vision test and diagnostic imaging scan are often required to confirm a diagnosis of BCM (BCM Families Foundation, 2017).

BCM is considered a non-progressive disease, although there is evidence of disease progression with macular degeneration in many patients (Michaelides et al, 2005). There is currently no cure for BCM; however, there is a major research focus on gene therapy which would involve the delivery of a 'normal' copy of the mutated gene(s) into the retina. Research has also focused on determining what outcome measures could be used in future clinical trials (Luo et al, 2015).

1.12 Bardet-Biedl syndrome (BBS)

Bardet–Biedl syndrome (BBS) is a rare autosomal recessive condition characterised by retinal degeneration, obesity, duplication of the fifth digit on a hand or foot (post-axial polydactyly), renal dysfunction, learning difficulties and hypogonadism (Forsythe and Beales, 2013).

The BBS phenotype evolves slowly throughout the first decade of life. By age 20 around 75% of patients with BBS will suffer from major vision loss. Most patients with BBS will be legally blind by age 30 (Puech et al, 2014). Obesity is present in approximately 70% of cases and often results in increased complications (Puech et al, 2014).

Diagnosis is possible based on clinical signs and can be confirmed by genetic sequencing with most patients being diagnosed by late childhood or early adulthood (Forsythe and Beales 2013). While there is research in progress, there is no targeted treatment available for BBS. Thus, a multidisciplinary approach is required to effectively manage the condition (Forsythe and Beales 2013).

1.13 Leber hereditary optic neuropathy (LHON)

Leber hereditary optic neuropathy (LHON) is a rare neurodegenerative condition caused by a variant in the DNA found in mitochondrial cells that results in blindness due to degeneration of the optic nerve (Brown et al, 1992; Man et al, 2002; Orssaud 2003).

Progressive central vision loss between the ages of 15 and 35 years is usually the only clinical indicator of LHON. Other common features include partially or entirely diminished visual clarity, impaired colour vision and optic nerve degeneration (Carelli et al, 2017). Cardiac defects and minor neurologic abnormalities have also been noted in some LHON pedigrees (Brown et al, 1992).

A definitive diagnosis of LHON can be obtained through molecular identification of one of the three common mitochondrial gene variants, supported by a family history analysis (Carelli et al, 2017).

There are no proven therapies for LHON; however, there are currently several treatments being tested (Myerson et al, 2015). In 2015, the European Medicine Agency (EMA) approved idebenone (Raxone) under exceptional circumstances for the treatment of visual impairment in adolescent and adult patients with LHON due to the rarity and severity of the condition (EMA, 2015; Carelli et al, 2017). Raxone is not approved for use, nor available for purchase, in the US or in Canada.

The management of LHON includes genetic counselling and avoiding environmental risk factors for vision loss such as tobacco use, heavy alcohol consumption, medications with mitochondrial toxicity and environmental toxins (Myerson et al, 2015; Carelli et al, 2017).

1.14 Achromatopsia

Achromatopsia is an inherited macular dystrophy characterised by dysfunction or absence of the cone components of the retina (Yu et al, 2014). Achromatopsia is a diverse condition involving a range of symptoms; however, the primary symptom is a partial or total absence of colour vision (Hirji et al, 2018). The lack of function of or absence of cone photoreceptors occurs from birth or early infancy and the early onset of sight impairment can be severely disabling (Hirji et al, 2018). Other symptoms may include light sensitivity (photophobia), involuntary eye movements (nystagmus) and poor visual acuity (Hirji et al, 2018).

While most people experience complete achromatopsia with no function present in all cones, some people have incomplete achromatopsia in which one or more cone types remains functional (Kohl et al, 2018).

The genes associated with achromatopsia are inherited in an autosomal recessive pattern indicating that hereditary factors are most significant risk factor for the condition (Kohl and Hamel, 2013). Achromatopsia can be diagnosed via genetic testing and diagnostic imaging (Kohl et al, 2018).

While there is currently no cure for achromatopsia, early phase clinical trials are currently being undertaken to determine the extent to which gene therapy might benefit visual acuity, colour discrimination, photophobia and nystagmus (Kumaran et al, 2018a). Current strategies available to manage the symptoms associated with the condition include the use of special-filter glasses and low vision aids (Kohl et al, 2018).

1.15 This report

This report describes the socioeconomic burden of 14 IRDs in Canada. To facilitate accessibility, non-standard formatting has been used throughout this report. This includes size 14 font and darker headings. In addition, spacing within the tables has been increased and pages containing detailed charts and tables have been converted to landscape.

The remaining sections of this report are set out as follows:

- Section 2: Methodology
- Section 3: Survey results
- Section 4: Prevalence
- Section 5: Health system costs
- Section 6: Productivity losses
- Section 7: Other costs
- Section 8: Loss of wellbeing
- Section 9: Sensitivity analysis
- Section 10: Conclusion
- References
- Appendices.

2 Methodology

This section describes the cost-of-illness methodology used to estimate the socioeconomic burden of 14 IRDs in Canada in 2019.

2.1 Overview

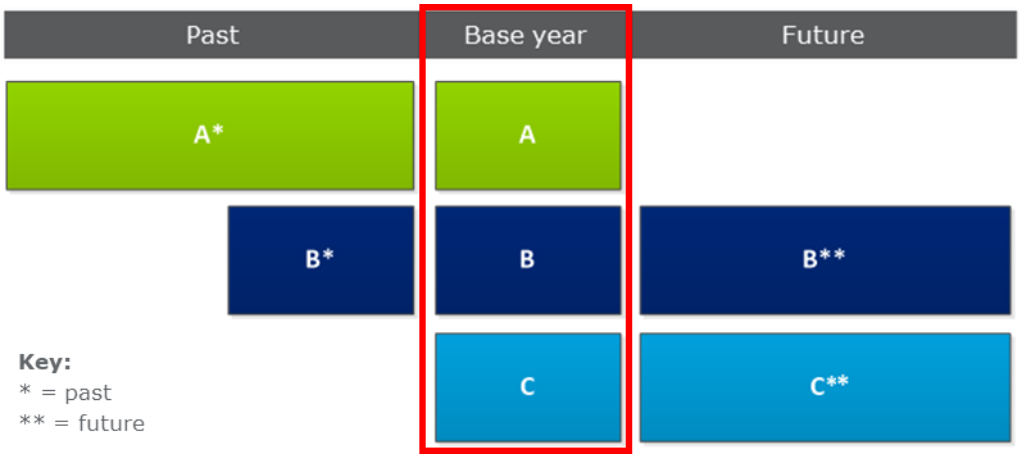
The socioeconomic burden of IRDs in Canada was estimated using cost-of-illness methodology applying a prevalence approach (Larg & Moss, 2011).

This approach involves estimating the number of persons living with an IRD in a base period (2019) and the costs attributable to IRDs in that period. Figure 2.2 provides an overview of the cost-of-illness model and indicates epidemiology is considered by the type of IRD. The analysis was based on a targeted literature review and primary data (survey) collection, described in the following sub-sections.

2.2 Epidemiological approach

This report uses a prevalence approach to estimate the costs of IRDs in Canada in 2019. The alternative approach is the incidence (lifetime cost) approach. In the prevalence approach, only the costs incurred in the base year (2019) are considered (i.e. A + B + C in Figure 2.1).

Figure 2.1 Prevalence approach



Source: Deloitte Access Economics.

No nationwide Canadian-specific prevalence estimates of IRDs (overall, or condition-specific) were identified. As such, this report

sets out two approaches for deriving the prevalence estimates required for the cost-of-illness modelling – approach 1 (lower bound) and approach 2 (upper bound). Further detail regarding the estimation of the lower and upper bound scenarios is provided in Section 4.

2.3 Costing approach

Costs were categorised by type, as:

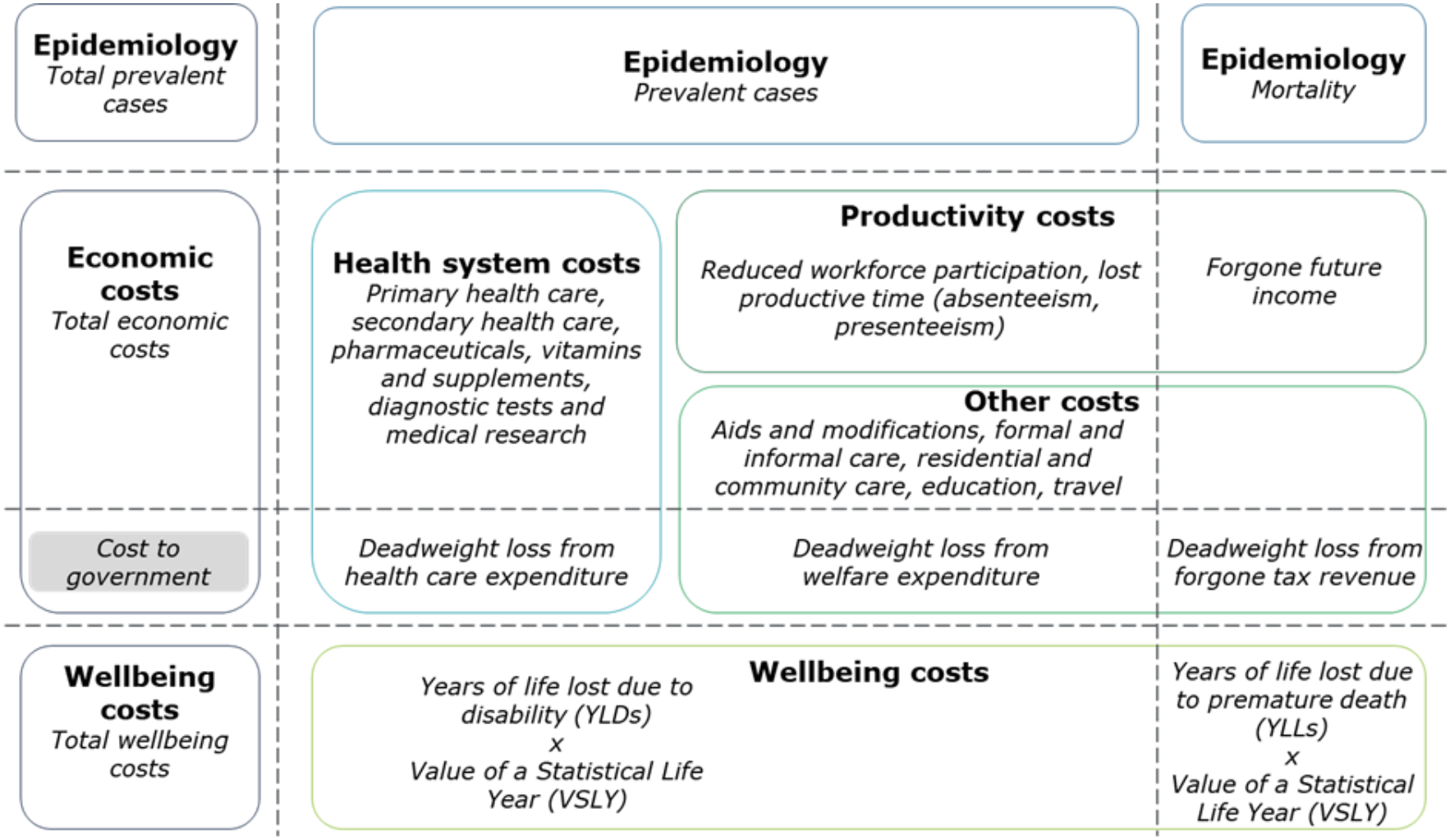
- Costs to the health system including primary and secondary health care, pharmaceuticals, vitamins and supplements, diagnostic tests and medical research
- Individual productivity losses due to reduced workforce participation, absenteeism and presenteeism (reduced productivity while at work)
- Other costs such as aids and modifications, formal and informal care, residential and community care, education, travel and deadweight losses (society-wide efficiency losses arising from loss of taxation revenue due to reduced workforce participation and higher government expenditures)
- Wellbeing cost measured by years of healthy life lost to IRDs.

Different costs of disease are borne by different individuals or sectors of society. Understanding how costs are shared helps to make informed decisions regarding interventions. While persons living with an IRD are most severely affected by the condition, family members and other parts of society also face costs attributable to IRDs.

This analysis estimated first round societal impacts. No second round, longer term dynamic impacts or mortality estimates were modelled (i.e. changes in wages or labour market outcomes associated with the economic burden of IRDs).

Component figures or calculations may not sum to totals due to rounding.

Figure 2.2 Cost-of-illness model overview



Source: Deloitte Access Economics.
Note: There is a lack of available data on the risk of premature mortality attributable to IRDs. As such, no costs associated with premature mortality were estimated

2.4 Data collection methods

2.4.1 Targeted literature review

A targeted review of the scientific literature and publicly available databases was conducted to identify the most relevant inputs for this report. The review involved a targeted (non-systematic) search of the PubMed and Cochrane Library databases to:

- Identify the target indication (IRDs)
- Identify epidemiological parameters of the target indication
- Identify economic parameters of the target indication.

Additional ad-hoc searches were performed, as required. Where possible, inputs specific to IRDs and Canada were used. Where this was not possible, inputs from similar populations in other countries, and diseases of comparable severity and symptoms, but different aetiology, were used.

2.4.2 Primary data (survey) collection and analysis

A survey was distributed to persons living with an IRD and/or the parent of children (under 18) living with an IRD in Canada or the US. The survey was designed to:

- Collect more specific and detailed primary data regarding the health care resource utilisation and productivity impacts of persons living with an IRD and the parents of children (under 18) living with an IRD, attributable to IRDs
- Address any data gaps identified in the targeted literature review.

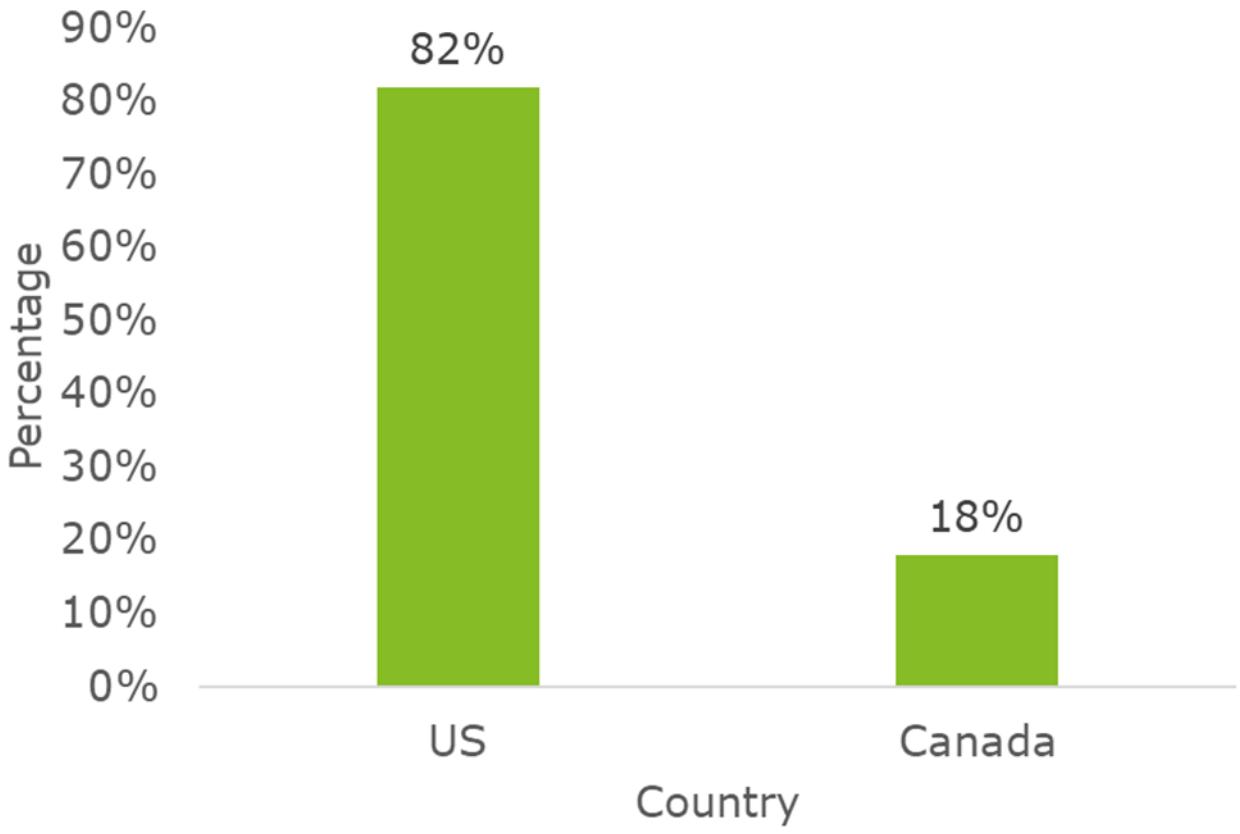
The survey questionnaire was co-designed with Retina International and consortium partners, and with reference to the metrics identified in the targeted literature review. Skip logic was incorporated into the programming of the survey to ensure only relevant questions would be asked of participants, depending on the answers provided to previous questions.

Retina International and consortium partners were responsible for inviting participants to self-register their interest in completing the survey via an online registration form. This form of non-probabilistic convenience sampling was employed to ensure the greatest number of respondents; however, may limit the generalisability of the survey to the broader Canadian population living with an IRD. Participation was restricted to persons living with an IRD and the parents of

children (under 18) living with an IRD. Ethical and patient consent procedures complied with relevant data protection, ethical and compliance obligations. Please refer to Section 3 for a breakdown of persons living with an IRD and/or the parents of children (under 18) living with an IRD invited to complete the survey, and responses received.

A total of 843 responses were received from persons with an IRD or the parents of children (under 18) living with an IRD, with 151 responses (18%) received from Canada and 687 responses (81%) received from the US (Chart 2.1).

Chart 2.1 Distribution of survey participants by country (n = 843)



Source: Deloitte Access Economics analysis.

2.5 Limitations

In many cases, the inputs underlying the cost-of-illness analysis are uncertain and changes in these inputs may have a significant impact upon the total estimate of the costs of IRDs in Canada in 2019. As such, when reviewing this report, it is important to consider the following limitations:

- Published population prevalence estimates of IRDs (overall or condition specific) are limited. Recent and nationwide prevalence estimates for IRDs in Canada were not identified. Please refer to Section 2.5.1 for further detail regarding how this limitation has been accounted for in the modelling.
- There are discrepancies in the definition and grouping of IRDs within the literature. For example, in some cases RP is considered to be a distinct condition while in other cases RP is considered to be an umbrella condition encompassing a range of other IRDs.
- There is a large range between the lower and upper bound prevalence rates estimated. This is reflective of the inherent uncertainty in the prevalence inputs available within the published peer-review literature.
- Aspects of the cost of illness model rely on survey data. Although participants were selected through a non-probabilistic convenience sampling method, it should be noted that bias may exist in the survey results as responses skew towards participants who have access to online resources such as smart phones and computers. As such the survey responses may limit the generalisability of the survey to the broader Canadian population living with an IRD.

2.5.1 Sensitivity analysis

Given the limited published data on the prevalence of IRDs in Canada, the socioeconomic burden of IRDs in Canada has been estimated for a lower and upper bound prevalence rate. The results of additional sensitivity analyses are set out in Section 10.

3 Survey results

This section describes the results from the survey of persons living with an IRD and/or the parents of children (under 18) living with an IRD in Canada.

Key findings

- A total of 843 survey respondents were received, with 151 (18%) survey respondents from Canada
- Survey respondents were skewed towards an older population, with the majority (59%) of respondents over 45 years of age
- There was a relatively even distribution of survey respondents by sex (53% female, 47% male)

The following key findings are specific to the survey respondents from Canada:

- Over one third of respondents (36%) were currently employed full-time
- The majority of respondents had been diagnosed with RP (69%)
- Over 70% of respondents were registered as legally blind
- Almost two thirds of respondents (62%) had received a genetic test for their IRD, with most having undergone testing the last 10 years.

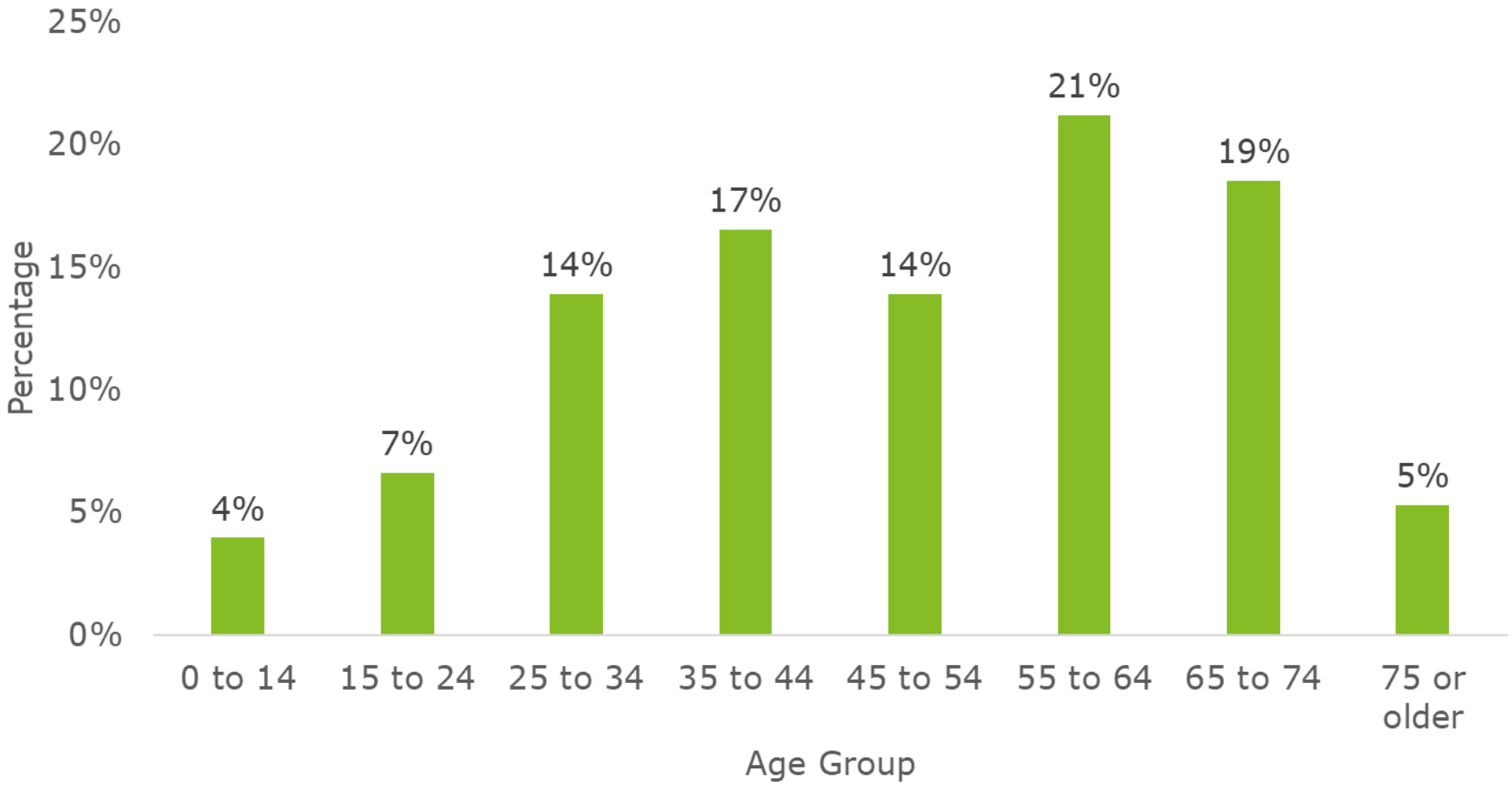
3.1 Participant characteristics

3.1.1 Participant type and age

Of the 151 responses from Canada, 138 (91%) were persons living with an IRD and 13 (9%) were the parents of children (under 18) living with an IRD.

Survey respondents were generally older with the majority of respondents (59%) over 45 years of age, while 25% of respondents were under 35 years of age (Chart 3.1).

Chart 3.1 Distribution of Canadian survey respondents by age (n=151)

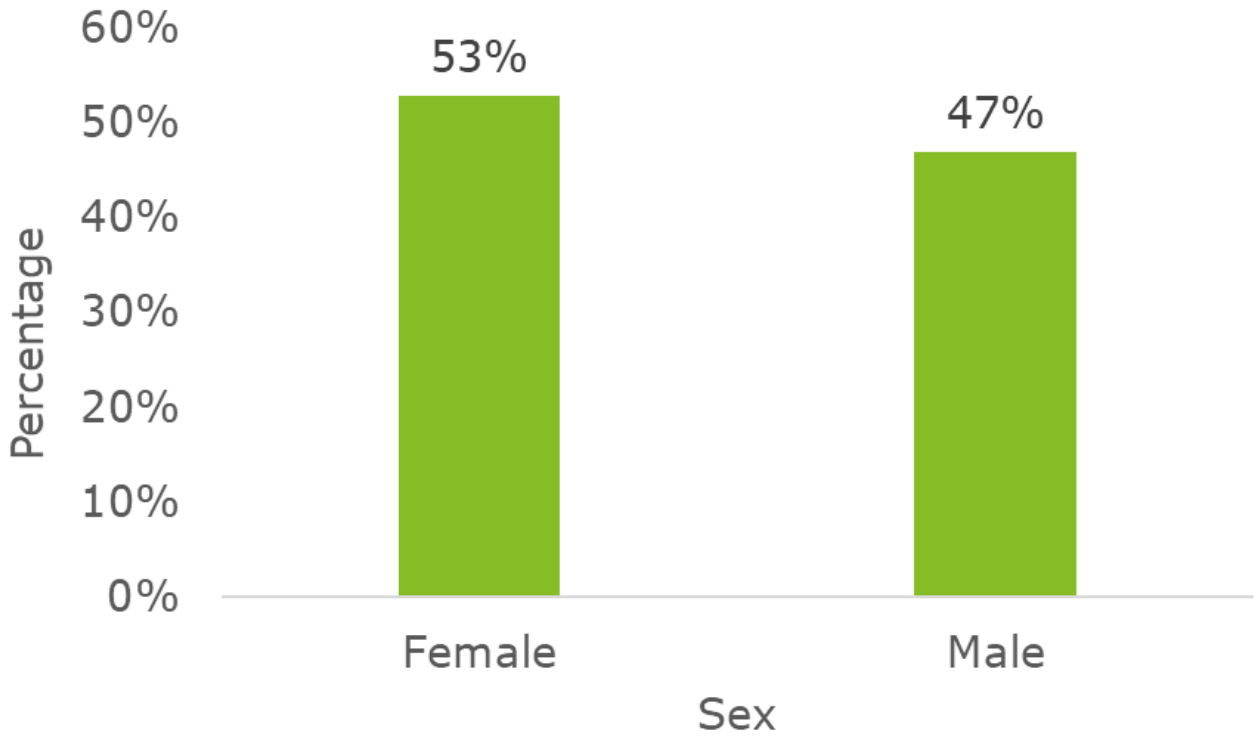


Source: Deloitte Access Economics analysis.

3.1.2 Sex

A slightly greater number of survey respondents were female, comprising 53% of all respondents, with Males comprising the remaining 47% (Chart 3.2).

Chart 3.2 Distribution of Canadian survey participants by sex (n=151)

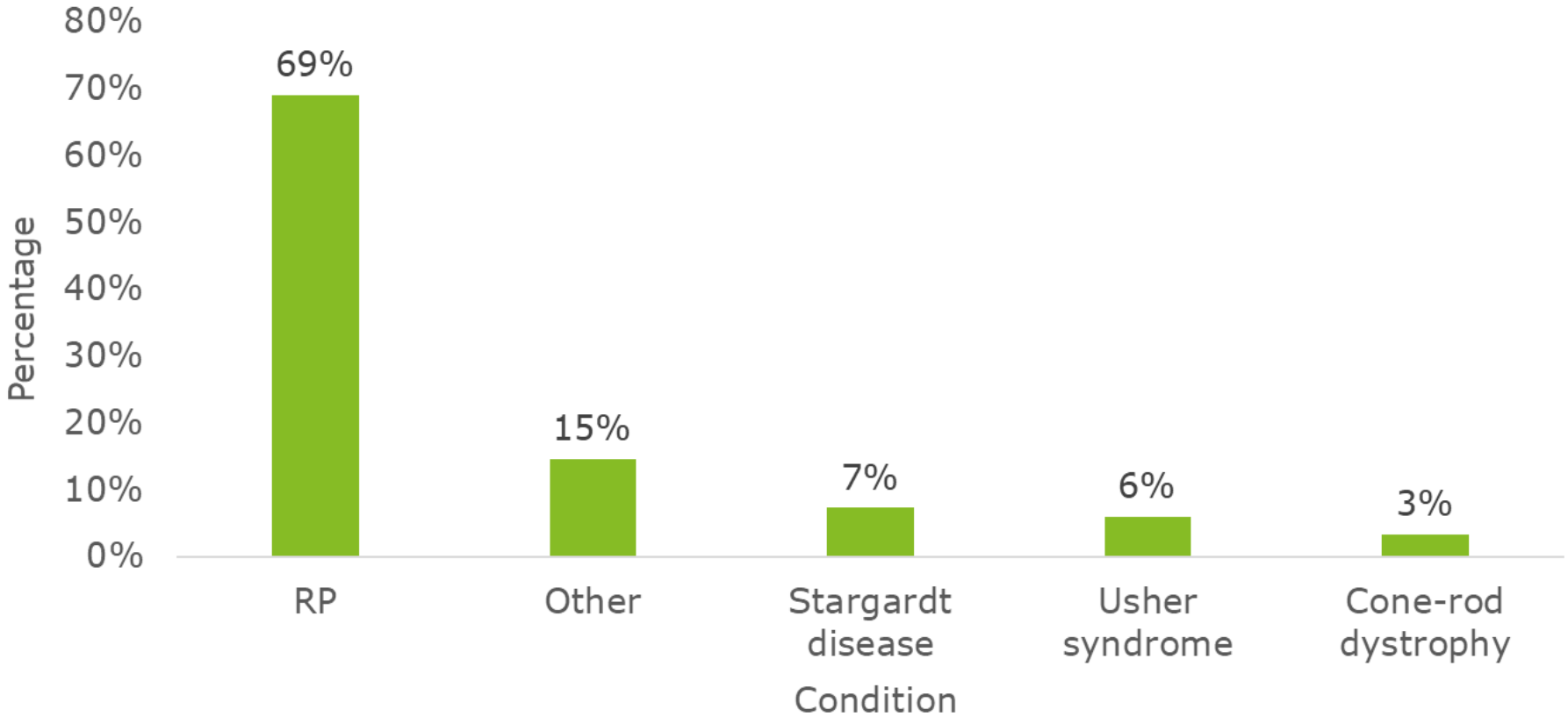


Source: Deloitte Access Economics analysis.

3.1.3 Condition

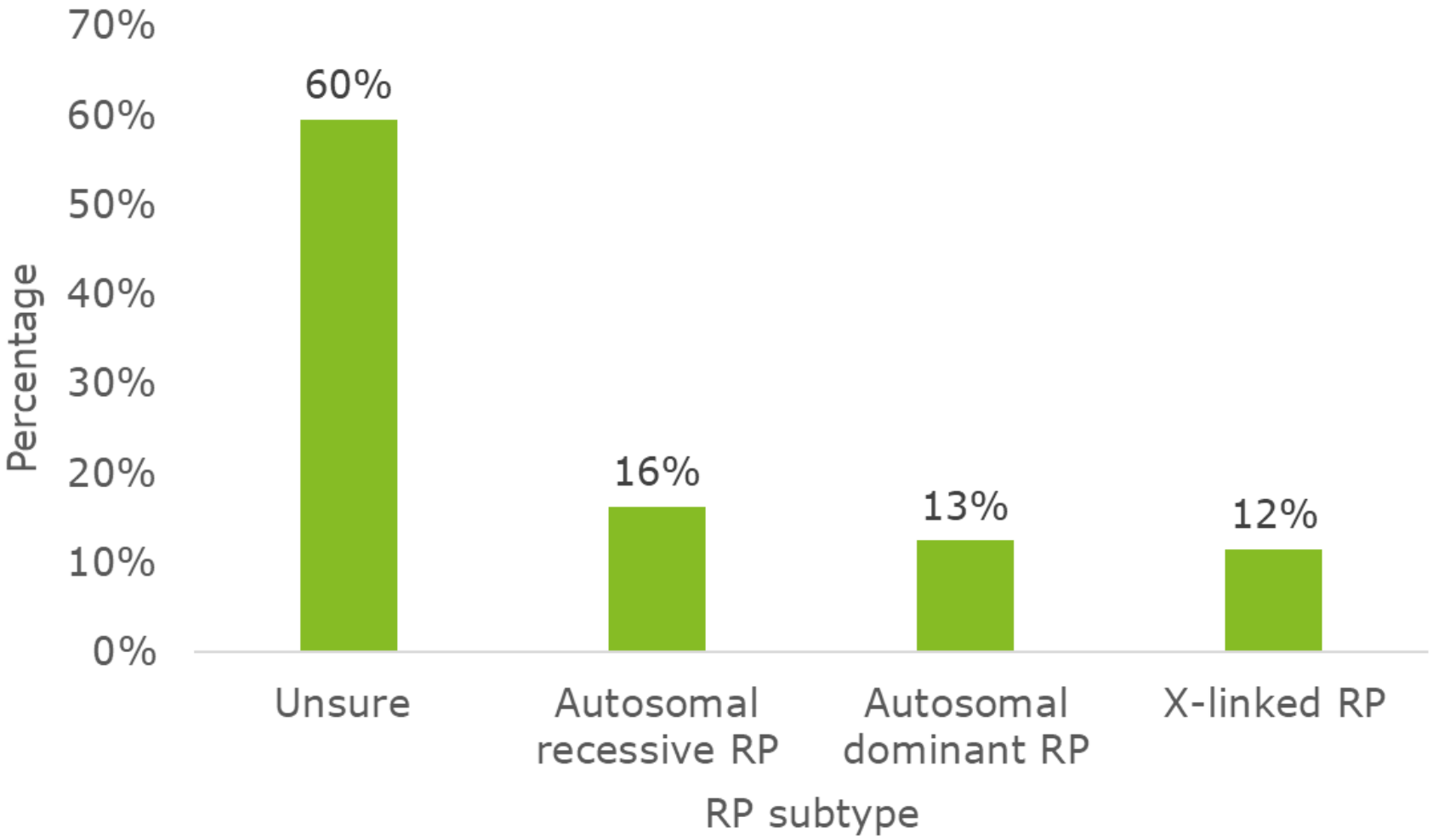
The majority of respondents (69%) identified RP as the IRD they had been diagnosed with (Chart 3.3). Stargardt disease and Usher syndrome made up 7% and 6% of respondent IRDs, respectively. A small number of responses were received for cone-rod dystrophy, LCA, LHON, BBS, rod-cone dystrophy, XLRS, cone dystrophy, choroideremia and Best disease. Together these conditions accounted for 15% of all responses. No responses were received for BCM or achromatopsia. Respondents who identified as having RP were asked about the subtype they had been diagnosed with. The majority (60%) of respondents were unsure of their subtype (Chart 3.4). Of the remaining 40% of respondents who knew their subtype, the majority identified as having autosomal recessive RP (40%), followed by autosomal dominant RP (31%) and X-linked RP (29%).

Chart 3.3 Distribution of Canadian survey respondents by condition (n=151)



Source: Deloitte Access Economics analysis.
Note: For confidentiality a number of other IRDs have been reported as 'other'.

Chart 3.4 Distribution of Canadian survey participants with RP, by RP subtype (n=104)

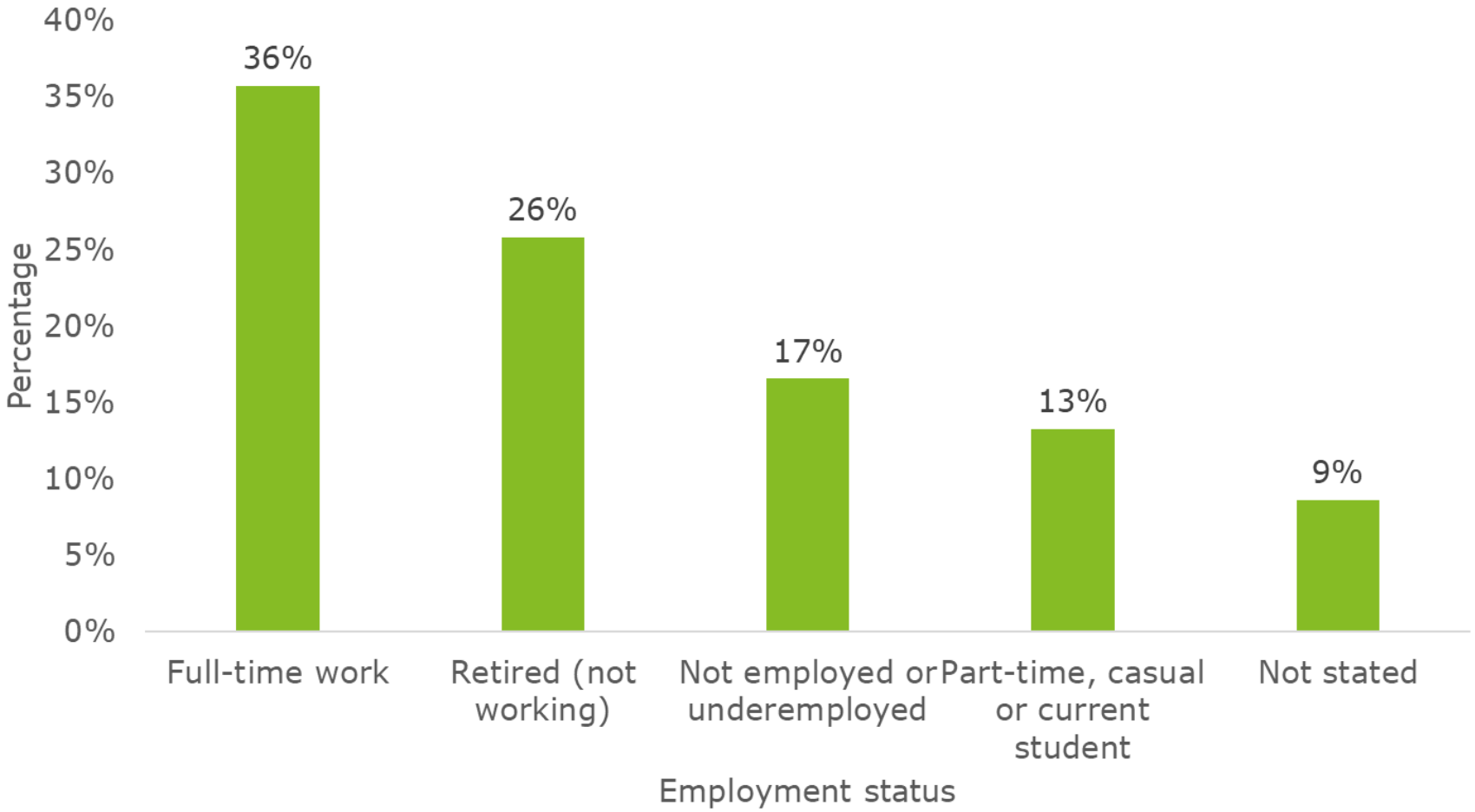


Source: Deloitte Access Economics analysis.
Note: Percentages may not add due to rounding. Due to the relatively small number of responses RP subtypes could not be disaggregated by sex.

3.1.4 Employment status

More than one third (36%) of survey respondents were employed full-time while 13% were working part-time, casually or were studying (Chart 3.5). The remaining respondents were retired (26%) or were not employed or underemployed (17%).

Chart 3.5 Distribution of Canadian survey respondents by employment status (n=151)

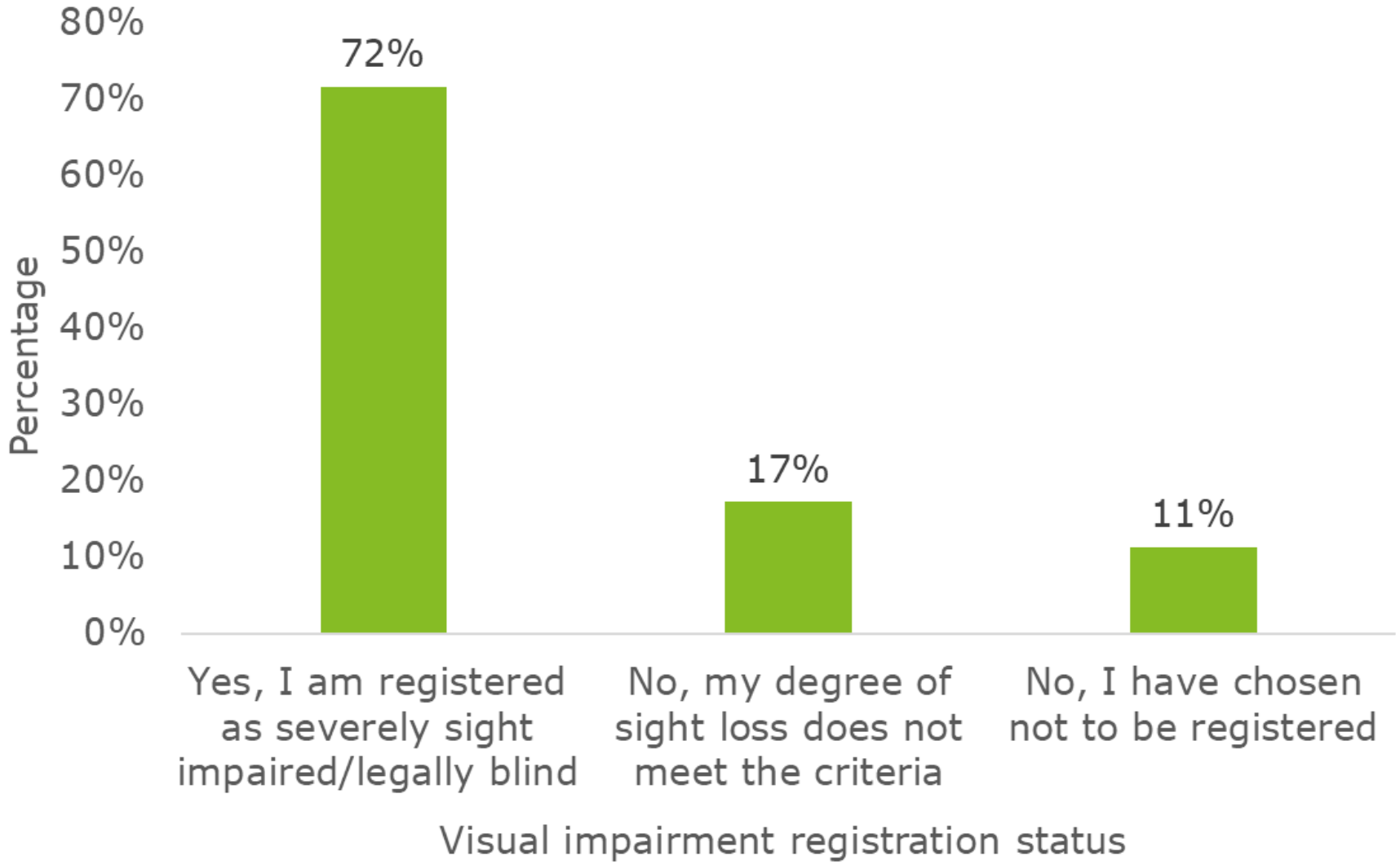


Source: Deloitte Access Economics analysis.
Note: Part-time and casual work included participants who do not work on a regular basis. Not employed or underemployed participants include individuals who desire to work additional hours or who are working in a role that does not use their skills or qualifications.

3.1.5 Visual impairment registration

The majority of respondents (72%) reported that they registered as legally blind with the Canadian National Institute for the Blind (CNIB) Foundation (Chart 3.6). Less than one third of respondents (28%) were not registered.

Chart 3.6 Distribution of Canadian survey respondents by visual impairment registration (n=151)

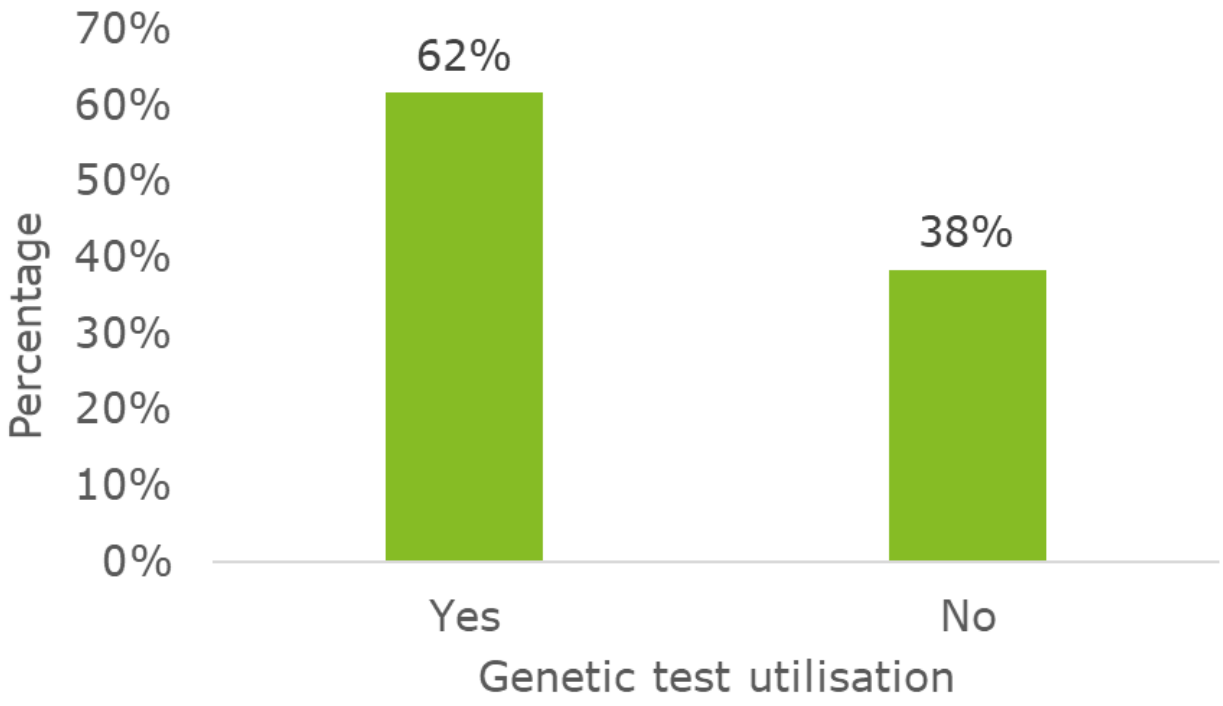


Source: Deloitte Access Economics analysis.

3.1.6 Genetic testing

In some instances, persons living with an IRD can receive genetic testing to investigate the underlying genetic cause(s) of their specific IRD. A total of 93 survey respondents (62%) reported that they had received a genetic test for the purpose of diagnosing their IRD (Chart 3.7).

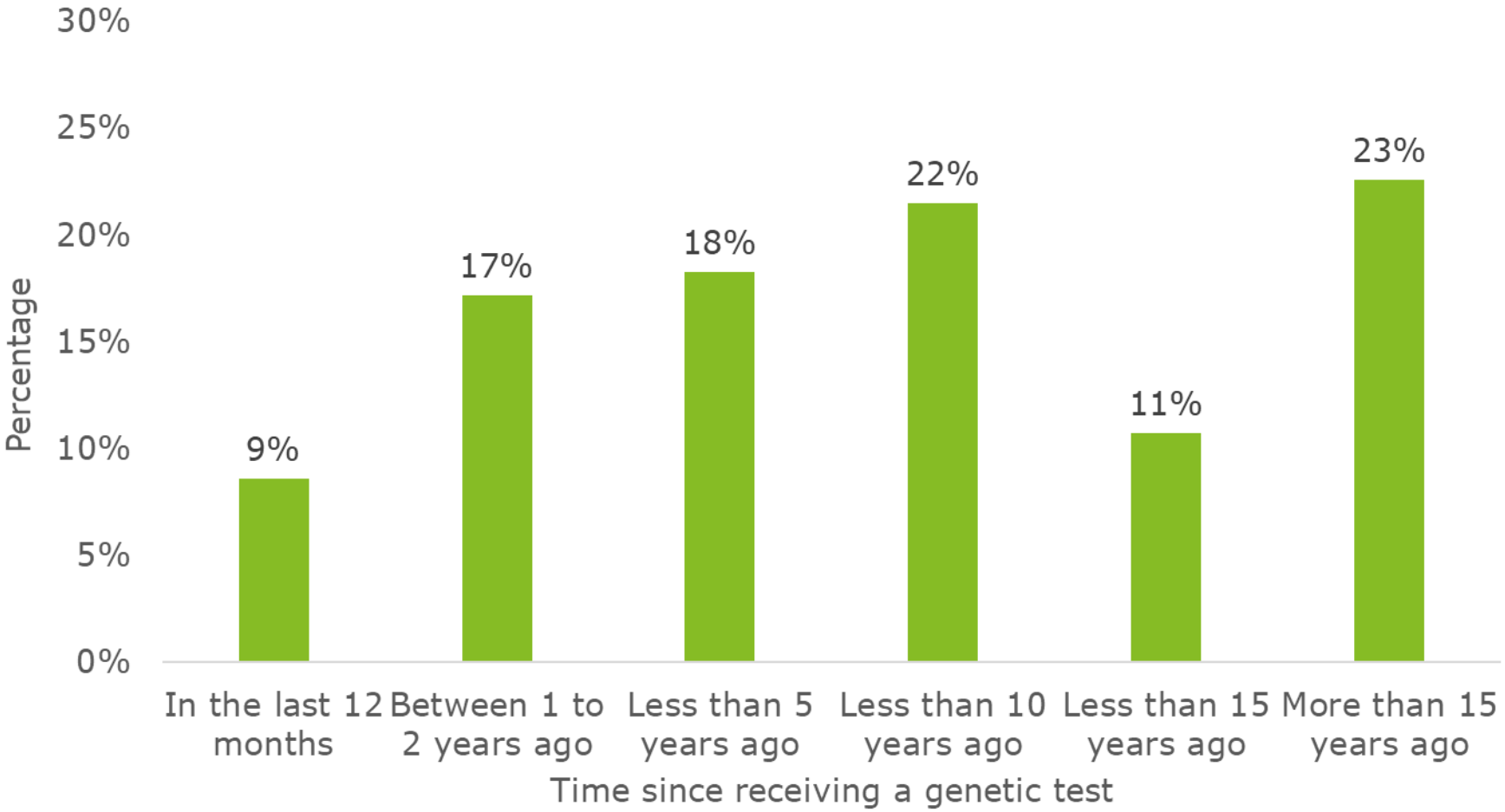
Chart 3.7 Genetic test utilisation of Canadian survey participants (n=151)



Source: Deloitte Access Economics analysis.

Those who had received a genetic test were asked about when they undertook this test (Chart 3.8). Two thirds of respondents (66%) had received their genetic test within the last 10 years, while 23% had received their test over 15 years ago.

Chart 3.8 Distribution of Canadian survey respondents by time since they received a genetic test (n=93)



Source: Deloitte Access Economics analysis.
Note: A small number or respondents are not included in the chart where they could not remember when they received their genetic test.

4 Prevalence

This section describes the approach used to estimate the prevalence of IRDs in Canada in 2019.

Key findings

This report includes a lower and upper bound estimate of the prevalence of IRDs in Canada in 2019:

- The overall prevalence of IRDs was estimated to range between 0.056% to 0.228% (lower to upper bound), which represented between 20,947 to 85,672 cases in Canada in 2019
- Across the 14 IRDs included in this analysis, rod-cone dystrophy contributed the highest proportion of cases between 27.8% to 27.9% (lower to upper bound) or between 5,841 to 23,891 cases in Canada
- The lowest proportion of cases was for achromatopsia representing 0.48% (for both lower and upper bound estimates) or between 101 to 412 cases in Canada
- Prevalence of IRDs was estimated to be slightly higher in males (54.5%) and tended to increase with age, up to the 55-59 year age group, after which prevalence gradually declines.

4.1 Summary of approach

A targeted literature review was conducted to identify available prevalence estimates to inform the cost-of-illness analysis. However, no nationwide Canadian-specific prevalence estimates of IRDs (overall, or condition-specific) were identified.

As such, this section sets out two approaches for deriving the prevalence estimates required for the cost-of-illness modelling – approach 1 (lower bound) and approach 2 (upper bound). Both of these options rely upon an available point prevalence estimate (either for a specific condition, or for total IRDs) which was used to derive the relative prevalence of the remaining IRDs (see Section 4.1.2 for a description of options 1 and 2). The prevalence of IRDs in Canada was then distributed by age (see Section 4.1.3) and sex (see Section 4.1.4).

A summary of each of the two approaches is provided in Table 4.1, below, with a more detailed description provided in the following sub-sections. In addition, two other approaches for estimating the prevalence of IRDs are provided in Appendix A. Further detail regarding the studies containing estimates of the prevalence of IRDs in Canada that were identified and considered but not included is provided in Appendix B.

Table 4.1 Summary of the two approaches detailed in this report

Approach	Sources used to estimate the prevalence of IRDs in Canada in 2019 (Section 4.1.2)	Sources used to distribute the prevalence of IRDs in Canada by age (Section 4.1.3)	Sources used to estimate distribute the prevalence of IRDs in Canada by sex (Section 4.1.4)
Lower bound	Bertelsen et al (2014); National Eye Institute (2019); Bocquet et al (2013); Orphanet (2013)	National Eye Institute (2019)	National Eye Institute (2019); Holtan et al (2020); Poincenot (2020)
Upper bound	Statistics Canada (2019); Bunce and Wormald (2006); Bunce et al (2010); Quartilho et al (2016); Motta et al (2018); National Eye Institute (2019); Bocquet et al (2013); Orphanet (2013)	National Eye Institute (2019)	National Eye Institute (2019); Holtan et al (2020); Poincenot (2020)

4.1.2 Estimating the prevalence of IRDs in Canada

4.1.2.1 Lower bound approach

Bertelsen et al (2014) estimates the underlying population-based prevalence of RP based on a national registry from Denmark. The prevalence of RP in the Danish population was estimated on 1 January 2013 using data from the national registry, death records and emigration data, and demographics data from Statistics Denmark.

The RP point prevalence estimated from Bertelsen et al (2014) was then used to derive the prevalence of each of the remaining IRDs relative to RP. Where available, preference was given to eyeGENE since it captures 10 of the 14 included IRDs within a US and Canadian cohort of 4,635 participants. As such, eyeGENE was used to derive the relative prevalence rates of achromatopsia, Best disease, choroideremia, cone rod dystrophy, LHON, RP, Stargardt disease, Usher syndrome, and XLRS.

The relative prevalence of BBS, cone dystrophy, LCA, and rod-cone dystrophy was informed by a genetic sequencing study of individuals with IRDs in France (Bocquet et al, 2013). In this study, 1,957 patients were recruited over a 21-year period in a specialised outpatient clinic for rare genetic sensory diseases.

In the absence of other available peer-reviewed literature and published data, the relative prevalence of BCM was derived using data from Orphanet (2013). Orphanet is an online inventory and classification of rare diseases with data contributed from 41 countries.

The lower bound approach which relies upon Bertelsen et al (2014) is considered the most robust and recent population-based prevalence estimate of IRDs and includes six of the 14 IRDs in-scope of this analysis.

4.1.2.2 Upper bound approach

Statistics Canada (2019) estimates the number of individuals in the Canada who experience a seeing disability. Individuals are identified as persons whose daily activities are limited because of difficulties with their ability to see. To determine the prevalence of IRDs within this population, the proportion of individuals experiencing blindness or visual impairment with IRDs as a primary or contributory cause

was applied, using data based on UK sight loss certifications (Bunce and Wormald, 2006; Bunce et al, 2010; Quartilho et al, 2016). Motta et al (2018), a retrospective analysis of medical reports which includes 10 of the 14 IRDs in-scope was used to adjust for the relative frequency of IRDs within the analysis. The overall estimate of IRDs in Canada was disaggregated using the sources described in Table 4.1.

Where available, preference was given to eyeGENE since it captures 10 of the 14 included IRDs within a US and Canadian cohort of 4,635 participants. As such, eyeGENE was used to derive the relative prevalence rates of achromatopsia, Best disease, choroideremia, cone rod dystrophy, LHON, RP, Stargardt disease, Usher syndrome, and XLRS.

The relative prevalence of BBS, cone dystrophy, LCA, rod-cone dystrophy and BCM were derived by the same method as described in the lower bound approach (Section 4.1.2.1).

For the two approaches, the prevalence of RP subtypes was derived using country-specific survey results. In Canada, it was found that the largest proportion of RP cases was accounted for by an autosomal recessive inheritance pattern (40.5%), followed by autosomal dominant (31.0%) and X-linked (28.6%) inheritance patterns.

The upper bound approach relies on Statistics Canada to derive an overall estimate of the prevalence of blindness and visual impairment and uses UK sources to estimate the proportion of blindness and visual impairment that is attributable to IRDs. There is insufficient evidence to determine whether the proportion of blindness and visual impairment that is attributable to IRDs in the UK is representative of that in Canada. Furthermore, it is possible that not all persons living with an IRD in the UK have a sight loss certification which may also limit its application to Canadian context.

4.1.3 Distributing the prevalence of IRDs in Canada by age

Age specific prevalence rates for the IRDs were utilised where they were available, from the literature. Data from the eyeGENE registry, which captures 10 of the 14 included IRDs within a US and Canadian cohort of 4,635 participants, was used to inform the age distribution

in the absence of condition-specific literature (National Eye Institute, 2019).

4.1.4 Distributing the prevalence of IRDs in Canada by sex

Data from the eyeGENE registry, was used to inform the condition-specific sex distribution for achromatopsia, Best disease, choroideremia, cone-rod dystrophy, RP, Stargardt disease, Usher syndrome, XLRS and rod-cone dystrophy.

Due to the lack of available peer-reviewed literature and published data, it was assumed that the sex distribution of cone dystrophy matched the profile of cone-rod dystrophy reported based on the eyeGENE registry (National Eye Institute, 2019). It was also assumed that the sex distribution of BCM matched the profile of XLRS, as both are X-linked conditions. The sex distribution of BBS and LCA were informed by Holtan et al (2020), while the sex distribution of LHON was informed by Poincenot et al (2020).

4.2 Results

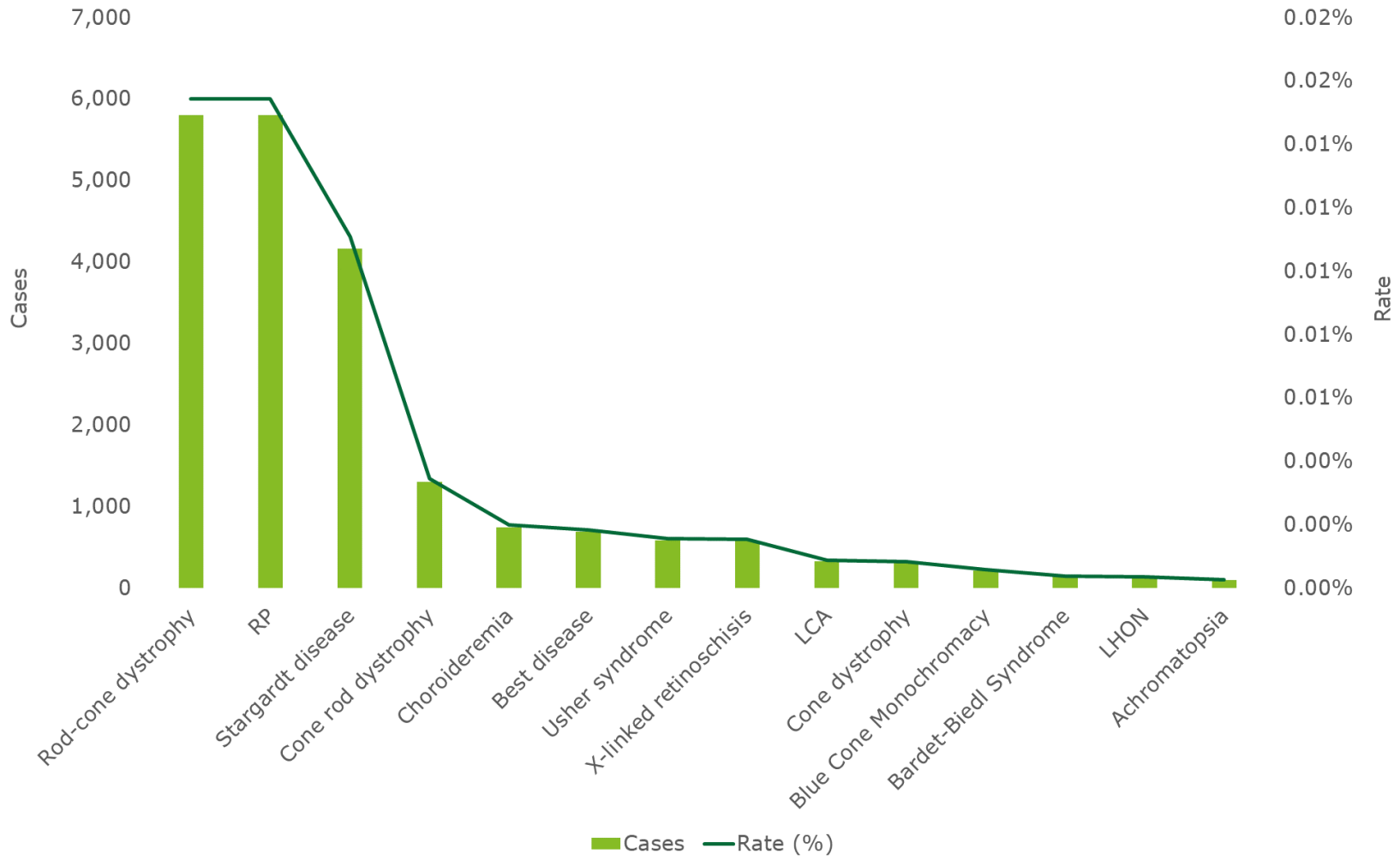
Overall, the prevalence of IRDs in Canada was estimated to range between 0.0556% to 0.2279% (lower to upper bound), or between 20,947 to 85,672 prevalence cases in 2019. The highest proportion of the overall prevalence was attributed to rod-cone dystrophy (between 27.8% to 27.9%, or between 5,841 to 23,891 people), followed by RP (between 27.7% to 27.8%, or between 5,803 to 23,736 people).[1] Chart 4.1 (lower bound), Chart 4.2 (upper bound) and Table 4.2 provide detailed prevalence estimates by condition, including the prevalence rate, number of cases, and proportion represented by each condition.

Overall prevalence of IRDs in Canada by age and sex is shown in Chart 4.3 (lower bound) and Chart 4.4 (upper bound). The prevalence rate of IRDs increases progressively with age until the

[1] This study utilises mutually exclusive prevalence rates for each condition including RP and rod-cone dystrophy. However, in some studies, RP and rod-cone dystrophy are used interchangeably. This may result in some misclassification in the diagnosis of RP and rod-cone dystrophy. However, the prevalence estimates are mutually exclusive. See: Hamel, C.P. Cone rod dystrophies. Orphanet J Rare Dis 2, 7 (2007). https://doi.org/10.1186/1750-1172-2-7.

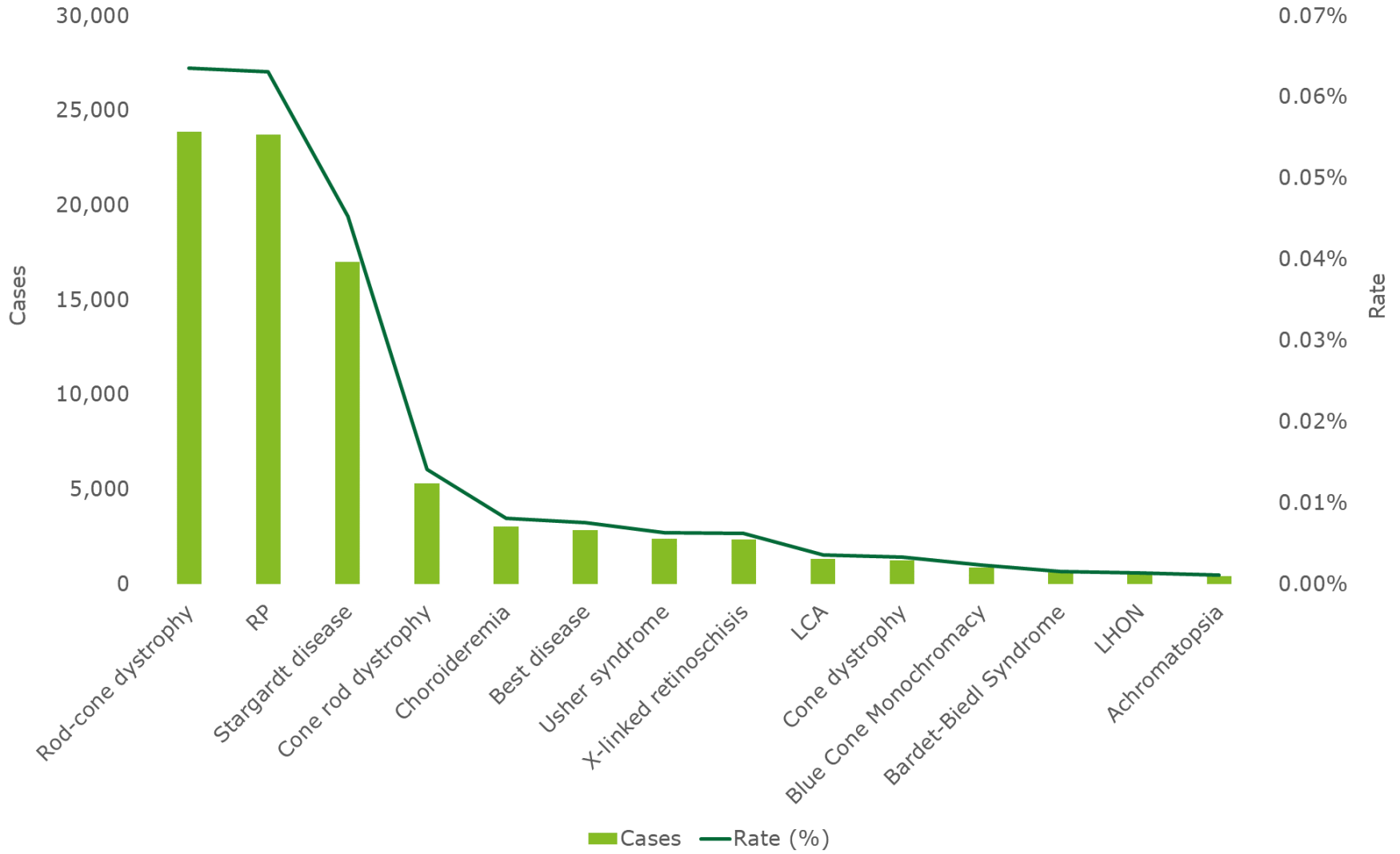
55-59 year age group, after which prevalence gradually declines. Overall prevalence is slightly higher in males (57.5%) compared to females (42.5%).

Chart 4.1 Prevalence of IRDs in Canada (2019) by condition – lower bound scenario



Source: Deloitte Access Economics analysis.

Chart 4.2 Prevalence of IRDs in Canada (2019) by condition – upper bound scenario



Source: Deloitte Access Economics analysis.

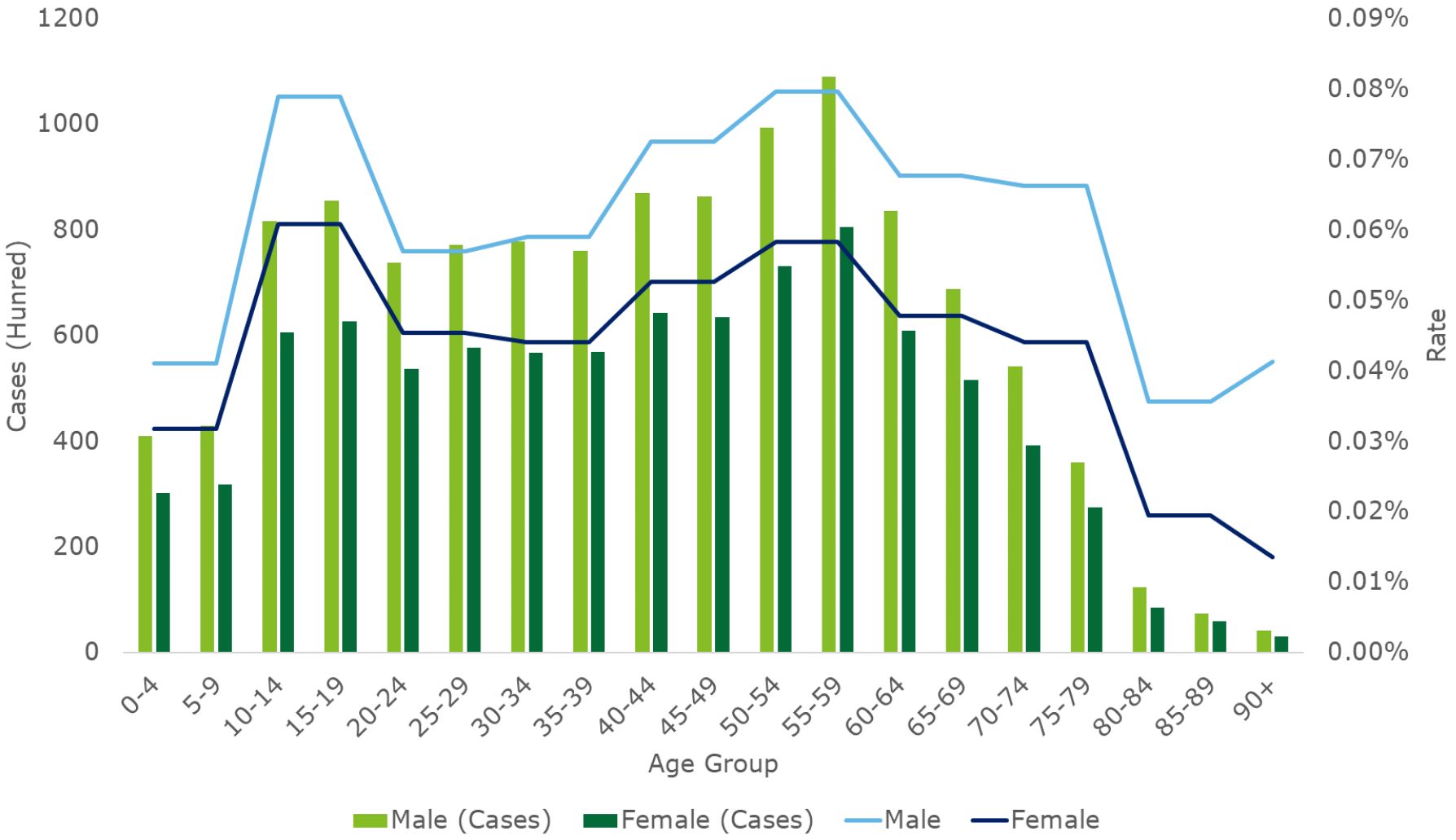
Table 4.2 Prevalence of IRDs in Canada (2019) by condition – lower and upper bound estimates

IRD	Approach 1 (lower bound) Rate (%)	Approach 1 (lower bound) Cases (n)	Approach 2 (upper bound) Rate (%)	Approach 2 (upper bound) Cases (n)
Rod-cone dystrophy	0.0155	5,841	0.0636	23,891
Retinitis pigmentosa (RP)	0.0154	5,803	0.0631	23,736
Autosomal dominant RP	0.0048	1,818	0.0198	7,434
Autosomal recessive RP	0.0071	2,679	0.0291	10,955
X-linked RP	0.0035	1,307	0.0142	5,347
Stargardt disease	0.0111	4,163	0.0453	17,027
Cone-rod dystrophy	0.0035	1,302	0.0142	5,323
Choroideremia	0.0020	748	0.0081	3,060
Best disease	0.0018	694	0.0076	2,840
Usher syndrome	0.0016	584	0.0064	2,387

IRD	Approach 1 (lower bound) Rate (%)	Approach 1 (lower bound) Cases (n)	Approach 2 (upper bound) Rate (%)	Approach 2 (upper bound) Cases (n)
X linked retinoschisis (XLRS)	0.0015	577	0.0063	2,360
Leber congenital amaurosis (LCA)	0.0009	329	0.0036	1,347
Cone dystrophy	0.0008	310	0.0034	1,270
Blue Cone Monochromacy (BCM)	0.0006	217	0.0024	889
Bardet-Biedl Syndrome (BBS)	0.0004	146	0.0016	596
Leber's hereditary optic neuropathy (LHON)	0.0003	131	0.0014	535
Achromatopsia	0.0003	101	0.0011	412
Total	**0.0557**	**20,947**	**0.2279**	**85,672**

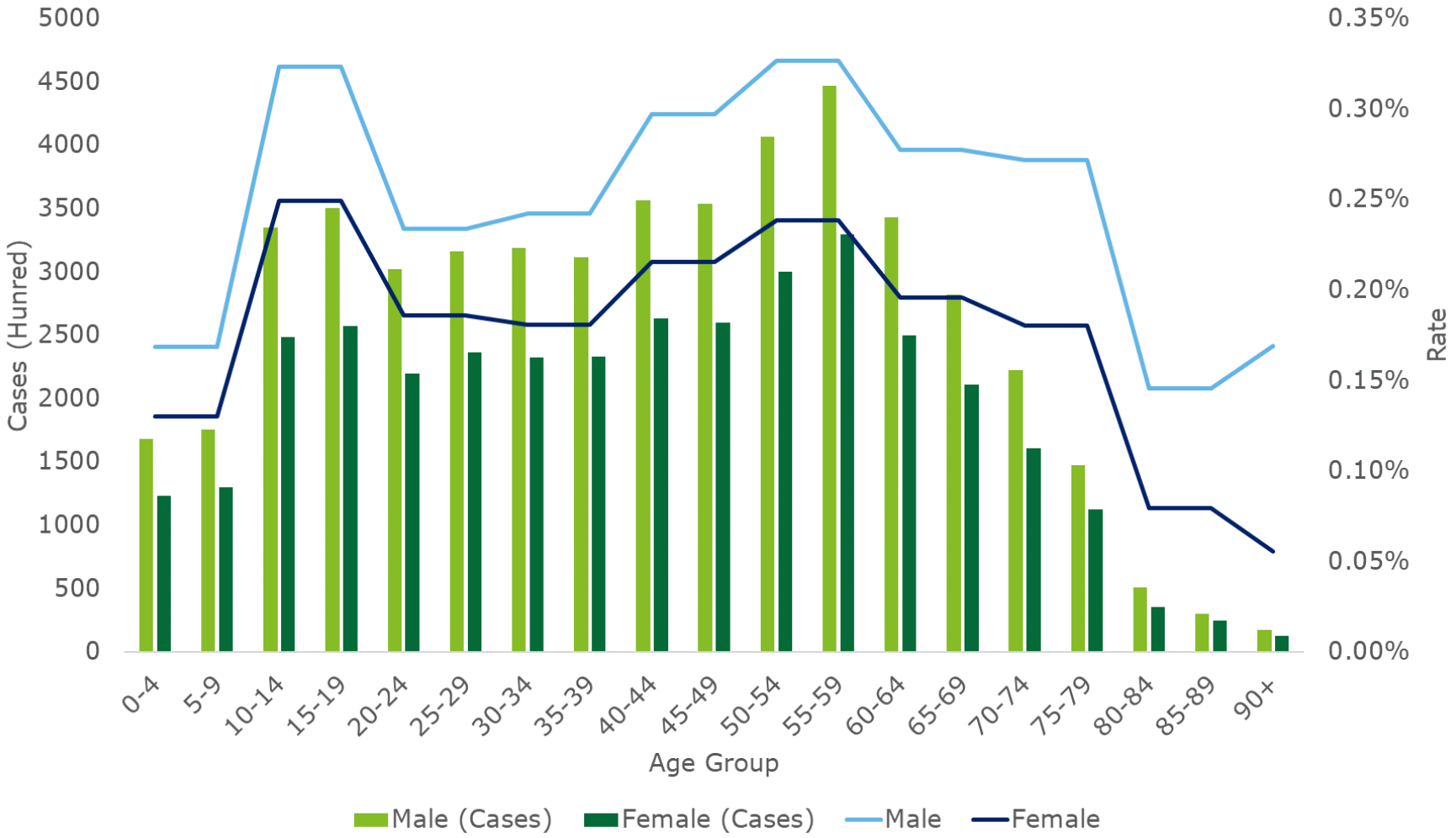
Source: Deloitte Access Economics analysis. using Bertelsen et al (2014) and Statistics Canada (2019).

Chart 4.3 Prevalence of IRDs in Canada (2019) by age and sex – lower bound scenario



Source: Deloitte Access Economics analysis.

Chart 4.4 Prevalence of IRDs in Canada (2019) by age and sex – upper bound scenario



Source: Deloitte Access Economics analysis.

5 Health system costs

This section describes the approach used to estimate the health system costs – including primary and secondary health care, diagnostic tests, pharmaceuticals, vitamins and supplements, and medical research – associated with IRDs in Canada in 2019.

Key findings

- Total health system costs associated with IRDs in Canada in 2019 were estimated to range between $37.8 to $144.3 million (lower to upper bound).
- The largest component of health system costs was primary health care (between $22.5 to $92.1 million), followed by diagnostic tests (between $7.3 to $29.9 million). The least proportion of cost was accounted for by pharmaceuticals and medical research (lower and upper bound respectively), totalling $1.0 and $2.0 million respectively.

5.1 Summary of approach

Health system costs include primary and secondary care, diagnostic tests, pharmaceuticals, vitamins and supplements, and medical research. Health system costs in Canada are primarily paid for by government; however, other sources include out-of-pocket patient payments and funding from other parties such as private health insurers. Canada's publicly funded health care system is financed through federal, provincial and territorial taxation, and includes some provinces charging a health premium to fund for publicly funded health care services (Government of Canada, 2020).

The targeted literature review identified no sources estimating the health system cost of IRDs in Canada. International literature was also scarce. The approaches and evidence for estimating categories of health system costs are described in each subsection below. Where available, utilisation of health services was derived from country-specific survey results and unit costs were sourced from national reference prices.

While survey data collected provides an estimate of out-of-pocket expenditures, it does not provide a method for attributing costs between society, government, family members and the individual. For this reason, the breakdown of total health system costs estimated by payer was based on the Canadian Institute for Health Information (CIHI, 2020) which publishes an estimated share of health system costs borne by government, private health insurers, patients (i.e. out-of-pocket costs) and other.

5.2 Primary health care

Primary health care includes general practitioner (GP) visits, allied health services, and other community-based services.

The cost of primary health care for persons living with an IRD was estimated by using the:

- Proportion of persons living with an IRD that used each health care service
- Average utilisation of each health service per person living with an IRD
- Incremental unit cost of each health care service.

Average utilisation was estimated using country-specific survey results of the proportion of the IRD population accessing each service and the annual per-person frequency of service use. Out-of-pocket travel costs for each service were also obtained from country-specific survey results. Where available, incremental unit costs were sourced from three Canadian provinces agreed with Retina International (Ontario, Alberta and Quebec), and were used to calculate a weighted-average Canadian unit cost. The incremental unit costs for Ontario, Alberta and Quebec were sourced from:

- Ontario Health Schedule of Benefits Physician Services (Ontario Health Insurance Plan (OHIP), 2019a) and Optometry Services (OHIP, 2019b)
- Alberta Health Schedule of Medical procedure list (Alberta Health Care Insurance Plan, 2019)
- Quebec Optometrists Manual Agreement and Rates (*Régie de l'Assurance Maladie du Québec* (RAMQ), 2019a)
- Quebec Manual of General Practitioners (RAMQ, 2019b)
- Quebec Manual of Specialist Physicians (RAMQ, 2019c).

5.2.1 Utilisation of primary health care services

Country-specific survey results were used to inform the proportion of persons living with an IRD accessing each health service and the relative frequency that each service was accessed.

Health services accessed by respondents at least once in the past 12 months included an ophthalmologist (21%), psychologist; psychiatrist or counsellor (11%), GP (9%), habilitation specialist (9%), specialist outpatient clinic (5%), genetic counsellor (4%) sight support volunteer (3%), occupational therapist (3%) and hospital emergency department (1%) (Table 6.1).

Of those health services accessed by persons living with an IRD, those accessed most frequency included sight support volunteer (12.9 times per annum), habiltation specialist (7.4 times per annum), psychologists, psychiatrist or counsellor (6.7 times per annum), admitted to hospital as an inpatient (5.5 times per annum) and specialist outpatient clinic (4.6 times per annum) (Table 6.1).

Table 5.1 Primary health care service utilisation of survey participants in Canada (n=151)

Health service	Proportion (%)	Frequency (count)
Ophthalmologist	21.2	1.5
Psychologist, psychiatrist or counsellor	11.3	6.7
General practitioner	9.3	2.3
Habilitation rehabilitation specialist or service, or low vision specialist	8.6	7.4
Specialist outpatient clinic or unit	4.6	4.6
Genetic counsellor	4.0	1.3
Sight support volunteer	2.7	12.9
Occupational therapist	2.7	2.8
Hospital emergency department	0.7	1.4
Admitted to hospital as an inpatient	0.0	5.5
My Retina Tracker app counselling service	0.0	0.0
Optometrist/optician	NA	1.7

Source: Deloitte Access Economics analysis. and utilisation from Deloitte Access Economics IRD survey.
Note: This question allowed for the selection of multiple answers; hence, the percentages do not sum to 100.

5.2.2 Incremental unit costs of primary health care services

The incremental unit cost for each service was assumed to comprise a reimbursed amount, out-of-pocket expenditure and travel costs. Where possible, the reimbursed amount for each service was estimated based on the national reference price (OHIP, 2019a; OHIP, 2019b; Alberta Health Care Insurance Plan, 2019; RAMQ, 2019a; RAMQ, 2019b; RAMQ, 2019c).

For services where a national reference price could not be located, a cost estimate was obtained from the literature or based on an average across a sample of provider-reported prices. For sight support volunteers, no official estimate could be obtained. Therefore, the average hourly cost of volunteer time was estimated based on the country-specific Organisation of Economic Cooperation and Development (OECD) calculations obtained from the OECD Survey of Adult Adults and inflated to 2019 dollars (OECD, 2015).

Out-of-pocket expenditures and out-of-pocket travel costs for each service were obtained from country-specific survey results. Where applicable, a measure of health inflation was utilised to convert unit values to 2019 terms (OECD, 2015).

Incremental health resource unit costs by service type are provided in Table 5.2.

Table 5.2 Primary health care service incremental unit costs ($ 2019)

Health service	Incremental unit cost
Psychologist, psychiatrist or counsellor	192
Hospital emergency department	93
Specialist outpatient clinic or unit	93
Admitted to hospital as an inpatient	81
Ophthalmologist	79
General practitioner	72
Genetic counsellor	57
Optometrist/optician	46
Sight support volunteer	24
Habilitation rehabilitation specialist or service, or low vision specialist	22
Occupational therapist	13
My Retina Tracker app counselling service	0

Source: Unit costs from OHIP (2019a), OHIP (2019b), Alberta Health Care Insurance Plan (2019), RAMQ (2019a), RAMQ (2019b), RAMQ (2019c), OECD (2015), Deloitte Access Economics analysis.

The average primary health care cost for persons living with an IRD is summarised in Table 5.3.

Table 5.3 Average cost of primary health care per person living with an IRD ($ 2019)

Service	Cost
Average primary health care costs per person living with an IRD	1,075

Source: Utilisation from Deloitte Access Economics IRD survey. Unit costs from OHIP (2019a), OHIP (2019b), Alberta Health Care Insurance Plan (2019), RAMQ (2019a), RAMQ (2019b), RAMQ (2019c), OECD (2015), Deloitte Access Economics analysis.

5.3 Secondary health care

Secondary health care includes hospital inpatient and outpatient services. Inpatient services include hospitalisations related to a primary or secondary diagnosis of an IRD and falls attributable to IRDs. Outpatient services include ophthalmologist attendances and associated procedures performed in an outpatient clinic.

The total cost associated with secondary health care services was estimated by applying the average cost of services to the number of services delivered.

5.3.1 Inpatient services

5.3.1.1 Inpatient admissions

There is limited evidence regarding the number of inpatient admissions related to a primary or secondary diagnosis of an IRD in Canada. To estimate the number of inpatient services related to a diagnosis of an IRD, hospitalisation data was used from the United Kingdom (UK) National Health Service (NHS, 2019) where the ratio of the number of hospitalisations due to an IRD in the UK to the number of hospitalisations due to visual impairment in the UK was applied to the number of hospitalisations due to visual impairment in Canada (CIHI, 2016).While the hospitalisation data is not specific to the Canadian health system, the two countries report similar health spending per capita (CIHI, 2020).

The average cost of admission was estimated by taking an average of major ophthalmology disorder and other ophthalmology disorder costs weighted by the relative frequency of each service (CIHI, 2019a; Gonder, 2014).

To estimate the total cost of inpatient services, the estimated number of admissions was applied to the average cost of an admission. The estimated cost totalled $1.3 million.

Table 5.4 Average cost of inpatient health care per person living with an IRD (2019)

Parameter	Estimate
Frequency (count) of inpatient admission for person living with an IRD	358
Unit cost ($) of inpatient admission per person living with an IRD	3,635
Average cost ($) of inpatient admission per person living with an IRD	**62**

Source: Deloitte Access Economics analysis. and utilisation from Deloitte Access Economics IRD survey.

5.3.1.2 Emergency department admissions

There is limited evidence regarding the number of emergency department admissions related to a diagnosis of an IRD in Canada. To estimate the number of emergency department related to a diagnosis of an IRD, hospitalisation data was used from the NHS (2019) where the ratio of the number of emergency department admissions due to an IRD in the UK to the number of emergency department due to an IRD in the UK was applied to the number of hospitalisations due to an IRD in Canada (CIHI, 2019b).

The average cost of an emergency department admission was derived from Gonder et al (2014), which estimated the cost of emergency department visit associated with visual impairment, adjusted to 2019 dollars (CIHI, 2019c).

Table 5.5 Average cost of emergency department admission per person living with an IRD (2019)

Parameter	Estimate
Frequency (count) of emergency department admissions for person living with an IRD	17
Unit cost ($) of emergency department admission per person living with an IRD	1,047
Average cost ($) of emergency department services per person living with an IRD	**1**

Source: Deloitte Access Economics analysis. and utilisation from Deloitte Access Economics IRD survey.

5.3.2 Outpatient services

Country-specific survey results were used to estimate the utilization of outpatient services by persons living with an IRD and applied to the number of prevalent cases. A weighted average cost for specialist outpatient clinic was obtained from the three provinces.

Out-of-pocket expenditures and travel costs were taken from the survey responses and added to the average service cost to estimate the total incremental unit cost.

Table 5.6 Average cost of outpatient health care per person living with an IRD (2019)

Parameter	Estimate
Proportion (%)	5
Unit cost ($)	1,108
Average cost ($) of outpatient services per person living with an IRD	**51**

Source: Deloitte Access Economics, CIHI (2019c).

5.3.3 Cost of falls

Falls may result from the visual impairment associated with IRDs. The CIHI (2018) estimates the number of individuals in Canada who experienced a fall injury disaggregated by presentation type (inpatient or emergency department setting) in 2017-18. To determine the number of persons with an IRD who experienced a fall within this population, a population attributable fraction approach, which was derived from converting an adds ratio to relative risk ratio, was applied to estimate the proportion of attributable falls (Deandrea, 2010) and inflated to 2019 terms using population growth (Statistics Canada, 2019). An average cost was then estimated (CIHI, 2019; Woolcott et al, 2012) and inflated to 2019 terms (CIHI, 2020). Overall, expenditure on cost of falls attributed to IRDs estimated to range between $0.1 to $0.5 million.

Table 5.7 Average cost of falls per person living with an IRD (2019)

Parameter	Estimate
Frequency (count) of inpatient admissions due to falls for persons living with an IRD	79
Unit cost ($) of inpatient admission due to falls per person living with an IRD	6,169
Frequency (count) of emergency department admissions due to falls for persons living with an IRD	24
Unit cost ($) of emergency department admission due to falls per person living with an IRD	1,102
Average cost ($) of falls per person living with an IRD	**6**

Source: Deloitte Access Economics.

5.3.4 Average cost of secondary health care

The average secondary health care cost for persons living with an IRD is summarised in Table 5.8.

Table 5.8 Average cost of secondary health care per person living with an IRD ($ 2019) (lower and upper bound)

Service	Average secondary health care costs per person living with an IRD
Lower bound estimate	120
Upper bound estimate	73

Source: Utilisation from Deloitte Access Economics IRD survey. Unit costs from CIHI (2019), Woolcott et al (2012), CIHI (2020), Deloitte Access Economics modelling.

5.3.4.2 Average cost of secondary care for family members

The cost of secondary care for family members of persons living with an IRD was assumed to include the cost of specialist outpatient consultation and genetic counselling. Survey data was used to inform the proportion of persons with an IRD whose family members had accessed these services, as shown in Table 5.9.

The number of family members accessing specialist consultation was estimated by calculating the number of persons with an IRD who reported having one or more family members who had accessed specialist consultation. It was then assumed that one of these family members had accessed this service within the past year.

To derive the number of family members accessing genetic counselling, it was assumed that family members accessed this service in-line with the ratio of genetic counselling utilisation to genetic testing utilisation among persons with an IRD.

Family members were assumed to access one service per annum. Incremental unit costs for genetic counselling and specialist outpatient consultation were assumed to equal the respective unit costs for these services among people with an IRD.

Table 5.9 Average cost of secondary health care for family members per person living with an IRD ($ 2019)

Service	Estimate
Proportion (%) of people with an IRD whose family member received a specialist consultation	22
Average secondary health care costs ($) for family members per person living with an IRD	**57**

Source: Utilisation from Deloitte Access Economics IRD survey. Unit costs from OHIP (2019a), OHIP (2019b), Alberta Health Care Insurance Plan (2019), RAMQ (2019a), RAMQ (2019b), RAMQ (2019c), OECD (2015), Deloitte Access Economics modelling.

5.4 Diagnostic tests

There is limited evidence regarding the cost of diagnostic genetic testing in Canada. To estimate the cost of genetic testing for persons with an IRD, cost data was used from the NHS (2019). While the unit cost estimate is not specific to the Canadian health system, the two countries report similar health spending per capita (CIHI, 2020). The cost of genetic testing for persons with an IRD was estimated at £850.00 based on the average cost of retinal dystrophy 235 gene exome panel sequencing of the entire coding region of genes (NHS, 2019d). This value was converted to Canadian terms using health purchasing power parity (OECD, 2020).

The cost of genetic testing for persons with an IRD ($1,493) was applied to the utilisation and frequency of service use, based on country-specific survey results, to derive an average cost per person. The cost of genetic testing for persons with an IRD includes the cost of obtaining a genetic test (and associated costs) and does not necessarily include the cost of interpretation of the genetic test results (which may be included as a separate cost incurred through a genetic counsellor consultation).

The average cost of diagnostic testing for persons with an IRD is summarised in Table 5.10.

Table 5.10 Average cost of diagnostic test per person living with an IRD

Service	Cost
Average cost of diagnostic tests per person living with an IRD	172

Source: NHS (2019), OECD (2020), Deloitte Access Economics analysis.

5.4.1 Genetic testing for family members of persons with an IRD

The number of family members accessing genetic testing was estimated by calculating the number of persons with an IRD who reported having one or more family members who had accessed genetic testing. It was then assumed that one of these family members had accessed this test within the past year.

The unit cost of genetic testing for family members was also derived from the NHS (2019) in the absence of publicly available Canada specific unit costs. The unit cost of genetic testing for family members was estimated at £157.50 based on retinal dystrophy 235 gene exome panel testing for known mutations in family members (NHS, 2019d). This value was converted to Canadian terms using health purchasing power parity (OECD, 2020). Utilisation and frequency of service use for family members was based on survey results.

The cost of genetic testing for family members of persons with an IRD ($277) was applied to the utilisation and frequency of service use, based on country-specific survey results, to derive an average cost per person. The cost of genetic testing for family members of persons with an IRD includes the cost of obtaining a genetic test (and associated costs) and does not necessarily include the cost of the interpretation of the genetic test results (which may be included as a separate cost incurred through a genetic counsellor consultation).

The average cost of diagnostic tests for family members of persons with an IRD is summarised in Table 5.11.

Table 5.11 Average cost of diagnostic tests per family member of persons with an IRD ($ 2019)

Service	Cost
Average cost of diagnostic tests for family members per person living with an IRD	176

Source: NHS (2019), OECD (2020), Deloitte Access Economics analysis.

5.5 Pharmaceuticals

The cost of pharmaceuticals incurred by persons with an IRD was estimated by applying the utilisation of medications to their incremental unit cost. Utilisation of medication was informed by survey results and the cost of each medication was obtained from online retail and pharmacy shops.

The average cost of pharmaceuticals for persons living with an IRD is summarised in Table 5.12.

Table 5.12 Average cost of pharmaceuticals per person living with an IRD ($ 2019)

Service	Cost
Average pharmaceutical costs per person living with an IRD	47

Source: Deloitte Access Economics analysis.

5.5.1 Gene therapy

One gene therapy – Voretigene neparvovec (Luxturna) – is currently under review by Health Canada for the treatment of adult and paediatric patients with confirmed biallelic *RPE65* mutations, often clinically diagnosed as RP or LCA (CADTH, 2020). Patients must have sufficient viable retinal cells in order to qualify for treatment with the gene therapy (EMA, 2018). At present this product is not available in Canada and as such is not included in this analysis.

5.6 Vitamins and supplements

Country-specific survey results were used to estimate the utilisation and online retail shops were used to obtain the unit cost incurred by persons living with an IRD on vitamins and supplements.

The average cost of vitamins and supplements for persons living with an IRD is summarised in Table 5.13.

Table 5.13 Average cost of vitamins and supplements per person living with an IRD ($ 2019)

Service	Cost
Average cost of vitamins and supplements per person living with an IRD	61

Source: Deloitte Access Economics.

5.7 Medical research

Medical research costs represent the average annual value of all grants relating to IRDs that are active in 2019 (Canadian Institutes of Health Research (CIHR), 2019). This cost does not vary with prevalence.

The value of medical research costs associated with IRDs was estimated by filtering the grants database published by the CIHR Canadian Research Information System (2019) for key terms. An average of the past five years of grants related to these conditions was calculated to estimate average annual medical research costs in 2019 (Table 5.14).

This estimate is likely to be conservative since it does not include broader research related to blindness or visual acuity which may partly be attributable to IRDs. For example, UK blindness certifications suggest that IRDs account for 5.7% of blindness and visual impairment in the UK (Quartilho et al, 2016; Bunce et al, 2010; Bunce & Wormald, 2006). There is limited evidence on the proportion of research related to blindness or visual acuity which can be attributable to IRDs in Canada. To estimate the medical research costs from broader research related to blindness or visual acuity due to an IRD, the attributable proportion was derived from the UK blindness certification sources. The value of grants related to

blindness and visual impairment allocated on this basis and attributed to research related to IRDs was valued at $237,103.

Table 5.14 Total medical research costs associated with IRDs ($ million 2019)

Parameter	Cost
Total medical research costs	2.0

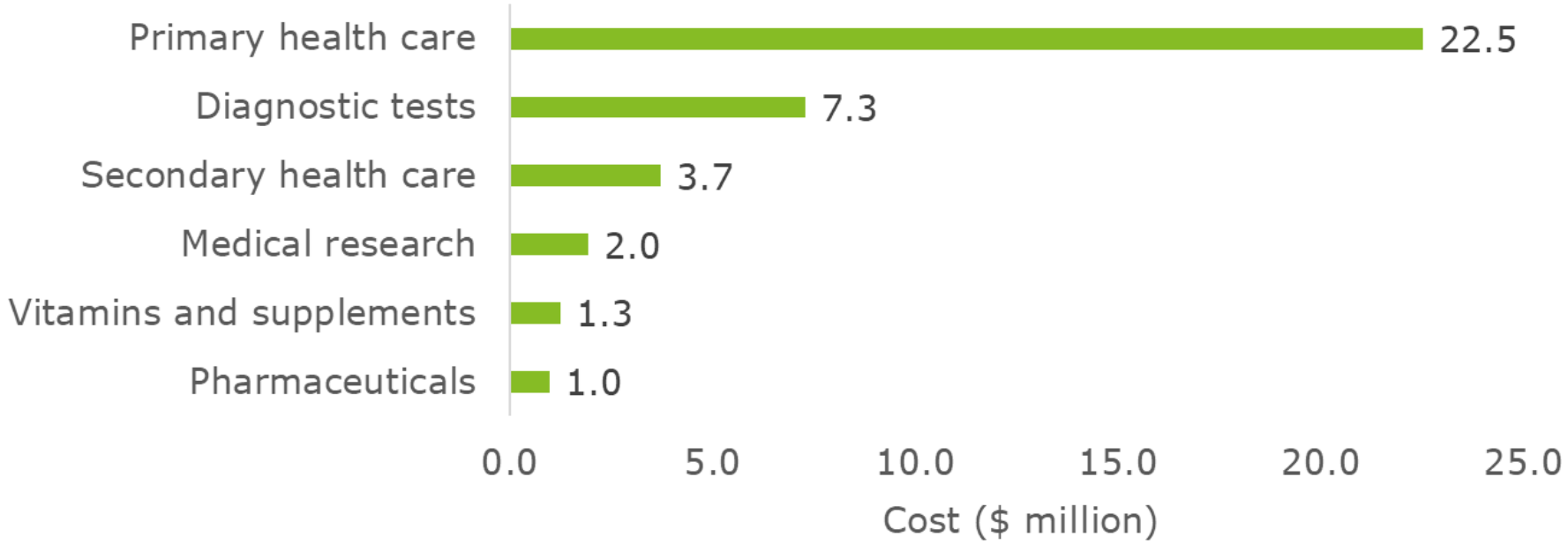
Source: CIHR (2019), Deloitte Access Economics analysis.

5.8 Results

Total health system costs associated with IRDs in Canada in 2019 were estimated to range between $37.8 to $144.3 million (lower to upper bound).[2] The largest component of health system costs was primary health care (between $22.5 to $92.1 million), followed by diagnostic tests (between $7.3 to $29.9 million). The least proportion of cost was accounted for by pharmaceuticals and medical research (lower and upper bound respectively), totalling $1.0 and $2.0 million respectively.

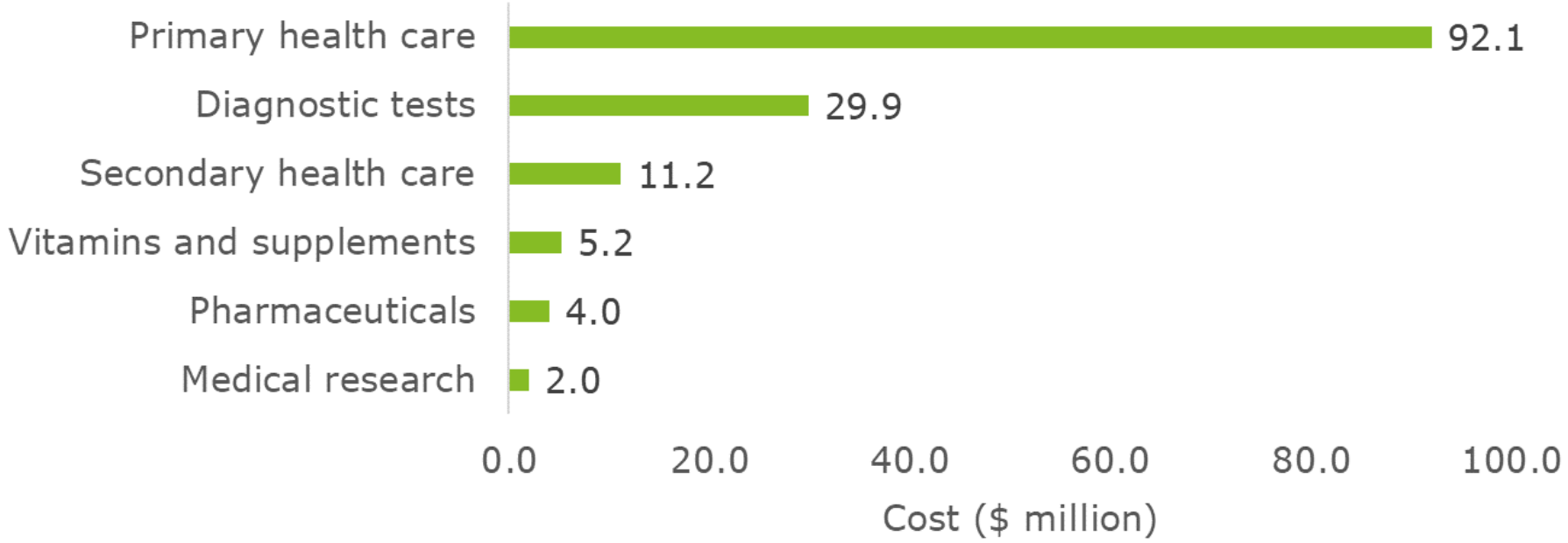
[2] Note: Psychologist costs are contributing substantially to elements of the overall health system cost.

Chart 5.1 Total health system costs of IRDs ($ million, 2019) in Canada by cost type (lower bound scenario)



Source: Deloitte Access Economics.

Chart 5.2 Total health system costs of IRDs ($ million, 2019) in Canada by cost type (upper bound scenario)



Source: Deloitte Access Economics.

Table 5.15 Total health system costs of IRDs ($ million, 2019) in Canada by cost type (lower to upper bound scenario)

Cost type	Lower bound cost	Upper bound cost
Primary health care	22.5	92.1
Diagnostic tests	7.3	29.9
Secondary health care	3.7	11.2
Medical research	2.0	2.0
Vitamins and supplements	1.3	5.2
Pharmaceuticals	1.0	4.0
Total	**37.8**	**144.3**

Source: Deloitte Access Economics analysis.
Note: Components may not sum to totals due to rounding.

6 Productivity losses

This section describes the approach used to estimate individual productivity losses – including reduced workforce participation and lost productive time – associated with IRDs in Canada in 2019.

Key findings

- In 2019, the individual productivity loss of IRDs in the Canada were estimated to range between $205.2 to $839.2 million.
- 72.5% of the individual productivity loss were due to forgone income as a result of reduced workforce participation.
- In Canada, persons with an IRD were 24.4% less likely to be in paid employment than the general population.
- IRDs resulted in a 1.4% reduction in productivity while at work.

6.1 Summary of approach

Persons living with an IRD can experience reduced capacity to effectively participate in the workforce. Consequently, there are significant productivity losses in terms of reduced workforce participation and lost productive time associated with IRDs.

A human capital approach was adopted to estimate the productivity losses attributable to IRDs. This involved calculating the difference in employment rate between persons living with an IRD and the general population, multiplied by average weekly earnings (AWE). Similarly, costs incurred through lost productive time were estimated by multiplying the average number of weeks of productive time lost by AWE.

The country-specific survey results informing this section were validated through triangulation with peer-reviewed literature estimating the productivity losses of IRDs, or conditions with comparable aetiology, in Canada and internationally.

6.2 Reduced workforce participation

Workforce participation refers to the section of the working population (aged 15-64) who are currently employed or seeking employment. Persons living with an IRD may experience reduced

workforce participation relative to the general population due to disadvantages in job-seeking or self-selection out of the labour force. This can lead to significant productivity losses through lost wages.

Costs incurred through reduced workforce participation were estimated by applying the time lost to reduced workforce participation to the Canadian general population employment rates and AWE by age and sex.

Based on the latest data from Statistics Canada (2020), the employment rate for individuals aged 15 to 64 years with a seeing disability in 2019 was 37.6%. Compared to the employment rate of 62.0% in the working population without a disability, this represents a 24.4% relative reduction in employment for people with a seeing disability in Canada.

The most common survey response indicated that most participants were employed full-time (36%). This result is comparable to the employment rate obtained from Statistics Canada (2020), outlined above. However, this was closely followed by survey respondents who indicated they were retired (not working) (26%); and not employed or underemployed (17%). Individuals who were employed part-time, casually or were a current student comprised 13% of survey respondents. 9% of survey respondents did not provide a response regarding their employment status (Table 6.1).

Table 6.1 Distribution of survey participants by employment status in Canada (n=151)

Employment status	Count (n)	Percentage (%)
Employed full-time	54	36
Retired (not working)	39	26
Not employed or underemployed	25	17
Employed part-time, casually, or current student	20	13
Not stated	13	9
Total	**151**	**100.0**

Source: Deloitte Access Economics analysis.
Note: Components may not sum to totals due to rounding.

6.2.2 Reduced workforce participation - summary of available literature

A targeted literature review was undertaken to identify studies that could be used to triangulate the findings for reduced workforce participation based on the survey results. There was only one published study found that estimated productivity losses due to vision loss in Canada.

In 2008, the CNIB Foundation and the Canadian Opthamological Society Commissioned Access Economics Pty Limited to undertake a comprehensive study on the cost of vision loss in Canada, which was the first study of this kind to be conducted. Access Economics' report *'The Cost of Vision Loss in Canada'* was released in 2009, and results were subsequently published by Creuss et al in the academic literature in 2011.

Creuss et al (2011) based costs associated with productivity losses on employment information compiled by Statistics Canada. The study reported that 28.0% ($4.4 billion) of the costs associated with

vision loss in Canada were attributable to productivity lost due to lower employment, higher absenteeism and premature death of Canadians with vision loss. This is comparable to more recent literature estimating the economic impact of blindness in Europe which assumed that productivity losses were 34.5% for people with moderate to severe visual impairment (Chakravarthy et al, 2017). The findings from Chakravarthy et al (2017) align very closely with the survey results from Canada, which indicated that most participants were employed full-time (36%) and this figure was also comparable to the employment rate (37.5%) for individuals with a seeing disability based on data from Statistics Canada (2020).

In addition, a number of studies estimating the employment rates of persons living with an IRD or condition of a comparable aetiology in other countries are summarised in Table 6.2.

Table 6.2 Summary of evidence on reduced workforce participation due to IRDs or visual impairment (VI)/blindness

Study	Condition	Country	Employment No VI (%)	Employment VI (%)	Reduction (% points)
Sherrod et al (2014)	VI – General	US	69.5	42.0	27.5
McDonnall and Sui (2019)	VI – General	US	77.0	44.0	33.0
Chaumet-Riffaud et al (2017)	RP	France	80.2	70.1	10.1
Sander et al (2005)	VI – General	France	65.0	29.0	36.0
Hewett and Keil (2016)	VI – General	UK	73.8	45.1	28.7
Slade and Edwards (2015)	VI – General	UK	73.0	27.0	46.0

Source: Sherrod et al (2014), McDonnall and Sui (2019), Chaumet-Riffaud et al (2017), Sander et al (2005), Hewett and Keil (2016), and Slade and Edwards (2015).

6.3 Lost productive time

Lost productive time is comprised of absenteeism and presenteeism.

6.3.1 Absenteeism

Absenteeism is the average number of days per year that an employee is away from work due to their condition.

Absenteeism incurs a productivity loss to:

- **Employers:** Lost output from work absences whilst incurring leave entitlements, management time in processing employee absences, line manager time in rearranging work, time of back office personnel, and overhead costs of employees such as office space and insurance costs
- **Government:** Sickness benefit entitlement paid through social insurance
- **Individuals:** Lost wages due to the gap between paid wages and the sick leave entitlement received from employers and/or government.

The survey results indicate that persons living with an IRD took an additional 21.04 days of personal leave due to their condition. Additional information on survey results that informed this estimation can be found in Appendix C (Table C.18).

By comparison, a study estimated that people with sight loss in the US were likely to take an average of 4.1 additional days off work annually by Pezzullo et al (2018). This study was excluded from this analysis in favour of the survey results that are specific to the population of persons living with an IRD in Canada.

6.3.2 Presenteeism

Presenteeism is the average number of hours per day that an employee loses to reduced performance or impaired function as a result of their condition. This is measured as a reduction in the quality and efficiency of work produced. Relative to absenteeism, presenteeism may occur more frequently and have a larger effect (Van den Heuvel et al, 2010). Persons living with an IRD may be more likely to experience reduced productivity at work as a result of their condition.

Costs incurred through presenteeism were estimated by applying the time lost to presenteeism to the Canadian general population employment rates and AWE by age and sex.

Presenteeism was estimated to be 1.43% for persons living with an IRD in the Canada in 2019 based on survey results. Additional information on survey results that informed this estimation can be found in Appendix C (Table C.19).

Following a targeted literature review, there was only one study identified that could be used to estimate presenteeism in a visually impaired cohort. Schakel et al (2018) collected cross-sectional data through interviews and surveys of visually impaired and normally sighted adults in the Netherlands between 2015 and 2016. Using a scale between one and ten, visually impaired and normally sighted respondents placed a value on their level of efficiency while at work. Deloitte Access Economics used the relative decline in self-reported work efficiency scores – measured on a scale of 1-10 – between the visually impaired (6.6) and normally sighted (7.3) respondents to estimate presenteeism of 9.6%.

However, the Schakel study was excluded from this analysis given that it was conducted in another jurisdiction and the impact of visual impairment and employment was also associated with other co-morbidities described in the study. As this analysis estimates the socioeconomic burden attributable to IRDs, costs associated with comorbidities were excluded from the analysis.

6.4 Other productivity losses not estimated

Premature mortality results in productivity losses due to forgone potential earnings. This can also result in increased costs associated with employee turnover such as search, hiring and training costs. There is a lack of available evidence regarding on the risk of premature mortality attributable to IRDs. As such, no cost has been allocated to forgone income or search, hiring and training costs due to IRDs in Canada.

6.5 Results

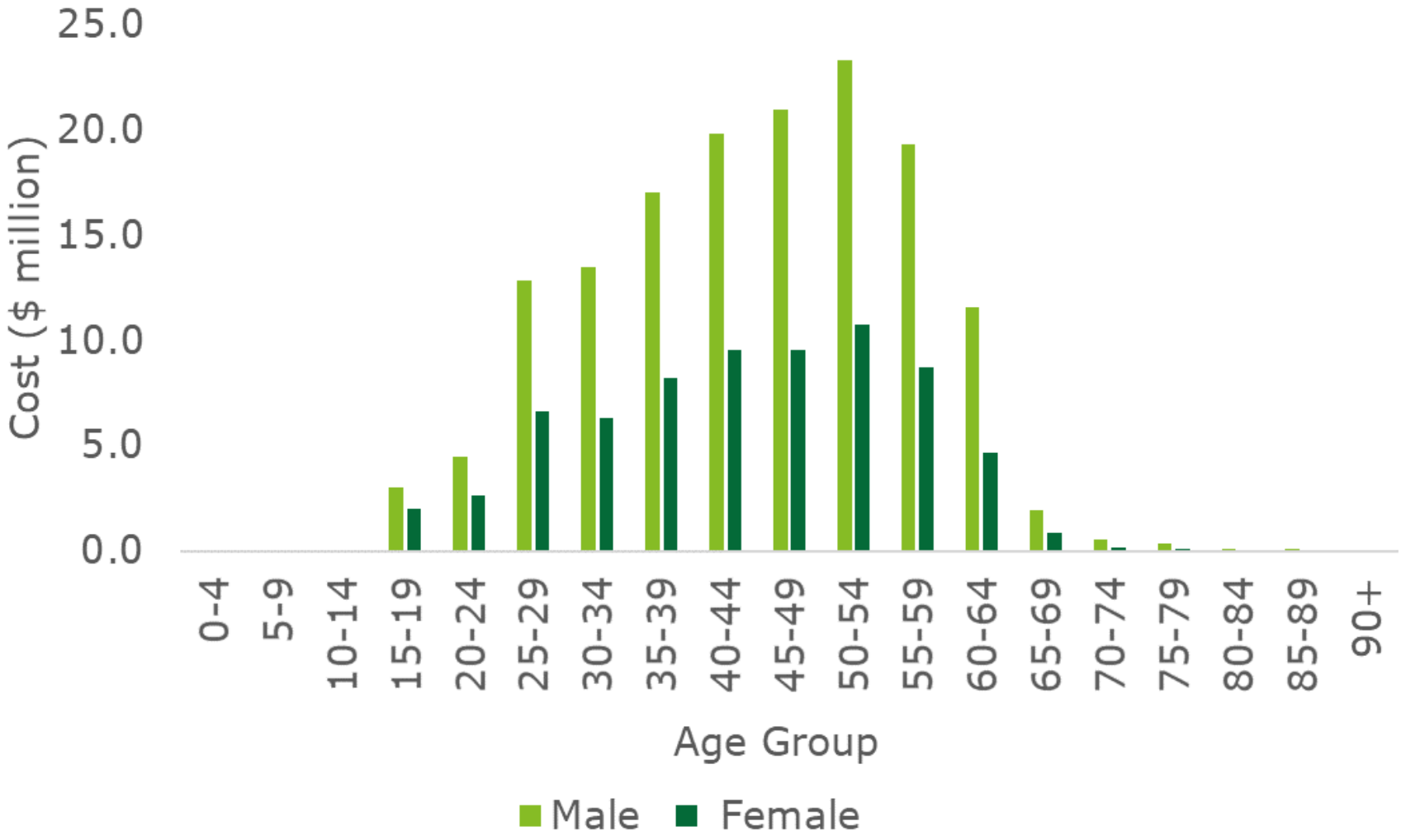
Total productivity losses due IRDs in Canada in 2019 were estimated to range between $205.2 to $839.2 million (lower to upper bound). The largest component of productivity losses were for reduced workforce participation (between $148.7 to $608.1 million), followed by absenteeism (between $49.9 to $204.2 million) and presenteeism (between $6.6 to $27.0 million) (Table 6.3 (lower to upper bound), Chart 6.1 (lower bound), Chart 6.2 (upper bound).

Table 6.3 Total productivity losses of IRDs ($ million, 2019) in Canada by cost type (lower and upper bound scenario)

Cost type	Lower bound cost	Upper bound cost
Reduced workforce participation	148.7	608.1
Absenteeism	49.9	204.2
Presenteeism	6.6	27.0
Total	**205.2**	**839.2**

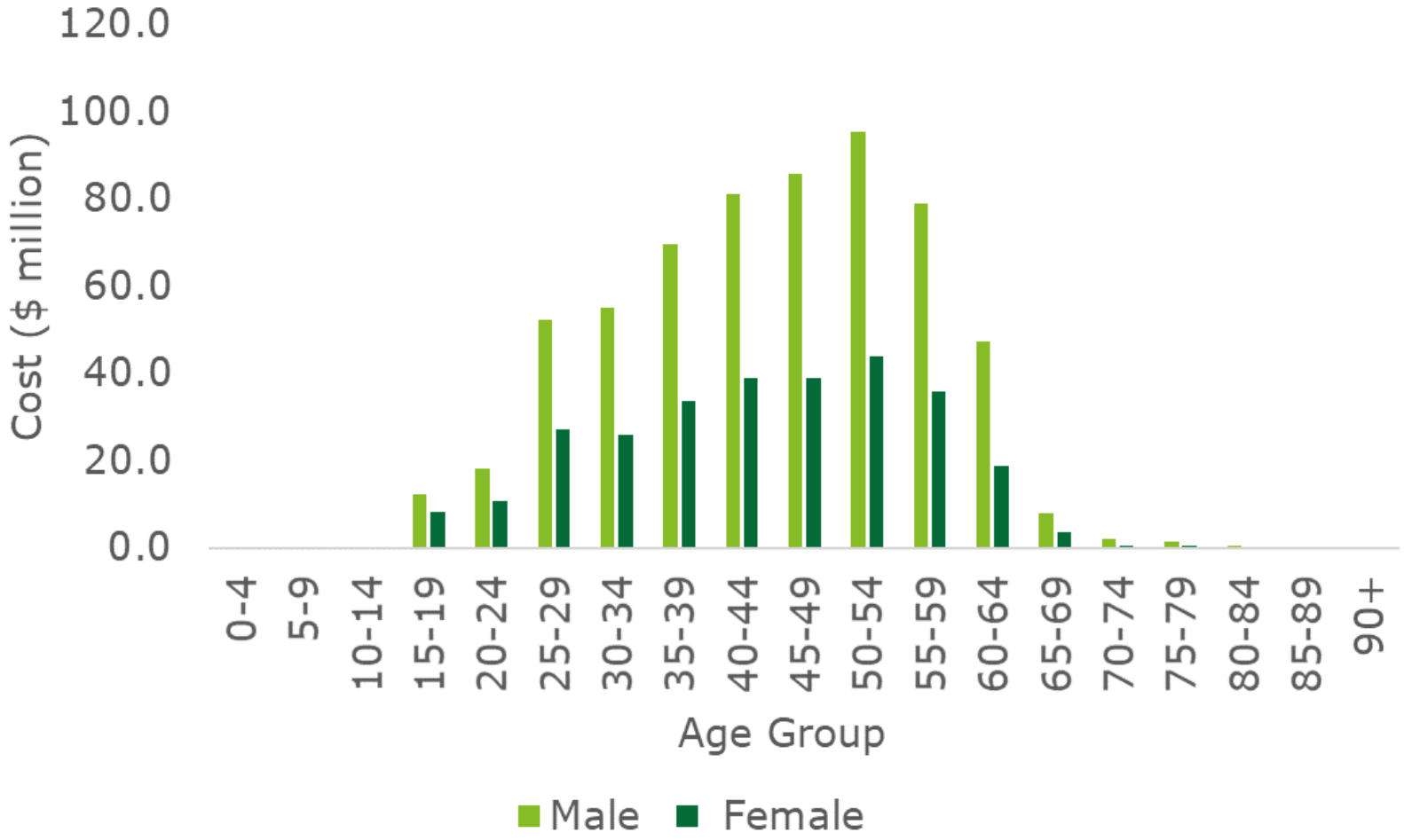
Source: Deloitte Access Economics analysis.

Chart 6.1 Total productivity losses of IRDs ($ million, 2019) in Canada by age and sex (lower bound scenario)



Source: Deloitte Access Economics analysis.

Chart 6.2 Total productivity losses of IRDs ($ million, 2019) in the Canada by age and sex (upper bound scenario)



Source: Deloitte Access Economics analysis.

7 Other costs

This section describes the results from the survey of persons living with an IRD and/or the parents of children (under 18) living with an IRD in Canada.

Key findings

- In 2019, other costs attributable to IRDs in Canada were estimated to range between $309.8 to $1,265.9 million (lower to upper bound), including formal and informal carer costs between $145.9 to $596.7 million, aid and modifications costs between $109.4 to $447.4 million, deadweight losses between $38.3 to $155.7 million and residential care costs between $16.1 to $66.0 million.
- Carer costs associated with carers of persons with an IRD include between $16.4 to $67.3 to million of formal care and between $129.5 to $529.5 million of informal care.

7.1 Summary of approach

Other costs attributable to IRDs include aids and modifications, education, travel, formal and informal care, and deadweight losses of taxation payments.

Survey results informing this section were supplemented with data obtained from grey literature (i.e. government publications and research reports). Where possible, these estimates were validated through triangulation with published literature estimating the other costs of IRDs, or conditions with comparable aetiology, in Canada and internationally.

7.2 Aid and modifications

Aids and modifications include items such as guide dogs, installing handrails in the bathroom, magnifying glasses, Global Positioning System (GPS), electronic mobility devices and other similar products used to assist persons living with an IRD with their daily living.

The cost of aids and modifications for persons living with an IRD was estimated by using the:

- Proportion of persons living with an IRD that used each aid or modification
- Average utilisation of each aid or modification per person living with an IRD
- Incremental unit cost of each aid or modification.

7.2.1 Utilisation of aids and modifications

Country-specific survey results were used to inform the proportion of persons living with an IRD using each aid and/or modification.

7.2.1.1 Aids

Average utilisation was estimated using survey data of the proportion of the IRD population using each aid and the average number of aids used per annum.

The greatest number of survey participants reported using handheld magnifiers (51.7%), followed by a white/long cane (50.3%), mobile phone modifications (48.3%), book alternatives (46.4%), screen magnification technology (31.8%), specialised laptops (29.8%) and screen reading software (28.5%). Other aids are outlined in Table 7.1.

The number of aids used per annum was also informed based on country-specific survey response data. Of people using a specific aid, the largest quantity of aids used per annum was recorded for tactile or large print labels (7.8), large keyboards (6.0), handheld magnifiers (5.2), mobile phone modifications (4.3), book alternatives (4.3) and verbal calculators (4.2). Other aids are outlined in Table 7.1.

7.2.1.2 Modifications

Average utilisation was estimated using survey data of the proportion of the IRD population using each modification and the average number of modifications used per annum. Of survey participants, only a small proportion reported they had a home modification (7.3%) or bathroom modification (4.0%).

Table 7.1 Aids and modification used by survey participants in Canada (n=151)

Aids and modifications	Percentage (%)	Number used per annum
Handheld magnifiers	51.7	5.2
White cane	50.3	NA
Mobile phone modifications	48.3	4.3
Book alternatives	46.4	4.3
Screen magnification technology	31.8	1.6
Specialised laptops	29.8	1.9
Screen reading software	28.5	2.9
Green or blue blocking sunglasses	26.5	3.2
Books with enlarged font	24.5	2.6
High vision lamps	21.2	3.5
Mini guide or other electronic mobility device	20.5	NA
Customised clocks or timers	18.5	2.9
Tactile or large print labels	16.6	7.8

Large keyboards	15.9	6.0
Ergonomic adaptations at home	15.2	1.7
Contrast enhancing filters	14.6	1.8
Spoken word processors	14.6	1.4
Ergonomic adaptations at work	14.6	2.4
Verbal calculators	12.6	4.2
Magnifying mirrors	10.6	1.4
Guide dog	8.0	NA
Portable note takers	3.3	1.6
Braille displays	2.7	3.0
Home modification	7.3	NA
Bathroom modification	4.0	NA

Source: Deloitte Access Economics analysis.
Note: This question allowed for the selection of multiple answers; hence, the percentages do not sum to 100.

7.2.2 Incremental unit cost of aids and modifications

The cost of each aid was obtained from the out-of-pocket expenditures recorded by the survey, or online shops.

For higher cost items, an annual depreciable amount was used as the basis for estimating an annual cost. This method is used to allocate the cost of assets over their useful life, for example, guide dogs were assumed to have a working life of nine years – meaning that in a given year, it was assumed that, on average, 11.1% of lifetime guide dog costs would be incurred.

7.2.2.1 Aids

The incremental unit cost for each aid was collected based on prices reported by online shops except for costs related to 'laptop related items (including cost of laptop)' and 'modifications to mobile phone such as applications' as these aids have specific costs to persons with an IRD. For higher cost items, the total purchase cost was attributed to an annual value using a measure of straight-line depreciation. A useful life of five years was assumed for electronic products including large keyboards, screen magnification technology, screen reading software, portable note takers and electronic mobility devices. This assumption was informed by the classes of depreciable property (2019) commissioned by the Government of Canada. An annual depreciable value of guide dogs was also assumed, based on the working life of nine years reported by Lions Foundation of Canada Dog Guides (2019). The annual unit cost of each item is summarised in Table 7.2.

7.2.2.2 Modifications

The cost of modifications were estimated from country-specific survey results. No studies estimating the cost of modifications of persons living with an IRD in Canada were identified. Unit costs were applied to the utilisation obtained from the survey. This yielded an estimated cost of modifications of $766 per person in 2019 terms.

Table 7.2 Aids and modification unit costs for persons living with an IRD in Canada ($ 2019)

Aids and modifications	Annual unit cost
Guide dog	2,778
Electronic mobility device	767
Portable note takers	739
Braille display	278
Screen magnification technology	160
Spoken word processor	110
Screen reading software	110
High intensity lamps	80
Ergonomic adaptations (e.g. raised screens) at work	59
Ergonomic adaptations (e.g. raised screens) at home	59
Verbal calculator	49
Customised clocks or timers	48
Contrast enhancing filters	48
Large keyboards	40
Tactile or large print labels	36
White cane	34
Magnifiers	25

Aids and modifications	Annual unit cost
Books with enlarged font	20
Green or blue blocking sunglasses	18
Book alternatives (such as audio books)	16
Magnifying mirrors	15
Laptop related items (including cost of laptop)	NA
Modifications to mobile phone such as applications (apps)	NA

Source: Deloitte Access Economics analysis.

These unit costs were multiplied by the survey utilisation data to estimate the cost of aids and modifications in Canada in 2019.

The average cost of aids and modifications for persons living with an IRD is shown in Table 7.3. Overall, expenditure on aids and modifications was estimated to range between $109.4 to $447.4 million.

Table 7.3 Average cost of aids and modifications per person living with an IRD ($ 2019)

Product	Cost
Average cost of aids per person living with an IRD	4,457
Average cost of modifications per person living with an IRD	766

Source: Deloitte Access Economics analysis.

7.2.3 Education

Persons living with an IRD can experience significant barriers to effective learning, thus necessitating specialist support and additional resourcing.

In Canada, there is no special education needs (SEN) funding provided by the national government, instead SEN programs for students up to grade 12 are designed by each province (National Centre on Education and the Economy 2020). Given that there is no SEN funding provided at a national level, the estimated average cost of SEN was excluded from the cost analysis. However, in Canada, persons with an IRD (or persons living with a disability more generally) may be eligible for SEN funding and financial support depending on the province/territory they reside in.

For example, in Ontario funding for students from kindergarten to grade 12 with SEN is provided by the Ontario's Ministry of Education who distributes funds to individual disctrict school boards based on a needs assessment referred to as an Individual Education Plan. As at June 2019, approximately 360,450 million students in Ontario were classified as having SEN with $3.01 billion in funding allocated to

schools to support students with high needs in 2018 -2019 (Ministry of Education 2019).

In Alberta, funding for students from Junior Kindergarten to Grade 12 with SEN is provided to charter schools, private schools and designated special education private schools by Alberta Education. To receive funding from Alberta Education, students with severe or mild/moderate disbilities must be identified and documented to be assigned with special education codes for approval. Funding is then allocated to schools based on the approval of students (Alberta Goverment 2019).

Students in Alberta with a visual impairment must satisfy the following criteria to be eligible for SEN funding:

- The child/student has limited capabilities to understand instrucational situations after vison correction and information must be presented in other forms
- The child/student only has up to to 6/60 (20/200) visual acuity in the better eye after vision correction.

For students that are difficult to assess, visual assessment conducted by a qualified medical professional or specialist in the field of vision may be sufficient to support eligibility for special education funding of the child (Alberta Goverment 2019).

7.2.4 Travel

Persons living with an IRD can experience significant barriers to driving a motor vehicle, meaning they may need to rely on public transport as their primary mode of travel. As a national travel concession program for persons with disabilities does not exist in Canada, the estimated average cost of transportation for reasons other than health care visits were excluded from the cost analysis. However, in Canada, persons with an IRD (or persons living with a disability more generally) are eligible for concessions and financial support depending on the province/territory they reside in.

For example, eligible Toronto residents with a disability who receive financial support from the Ontario Disability Support Program (OSDP) are eligible for the 12-month Fair Pass Transit Discount Program (City of Toronto, 2020). The fair pass transit discount program offers residents reduced fares when using the public transit system in Ontario. Whereas, in British Columbia, persons with

disabilities receive an extra $52 each month as a transportation supplement, which can be used for an annual bus pass or for other transportation needs (British Columbia, 2020). By contrast, a monthly taxi allowance of $56 is available for eligible people with disabilities that reside in the city of Calgary in Alberta, who are unable to use the public transit services (Calgary Transit, 2020).

7.3 Informal, formal and residential care

IRDs can result in impairment and disability that not only affects those persons living with the condition but can also affect their family and friends. Individuals with severe impairment resulting from an IRD can have limited ability to engage in day-to-day and self-care activities, requiring others to do these activities in their place.

7.3.1 Informal care

A range of informal care activities may be provided to persons living with an IRD. Impairment and disability not only affect the individual but can also affect their family and friends. This can limit the ability of the individual to engage in day-to-day and self-care activities, requiring others to do these activities in their place. Informal care activities depend on the level of impairment experienced by persons living with an IRD, and can include:

- Collecting relevant prescriptions and organising and timing the administration of medication
- Assistance in daily domestic activities such as cooking and laundry
- Ad-hoc tasks, such as shopping, transport and cleaning activities
- Monitoring of the patient's physical and mental wellbeing.

The opportunity cost method was used to estimate the cost of informal care. This method assumes that time spent providing informal care could be used to engage in paid employment or in leisure activities.

The cost of informal care for persons living with an IRD was estimated by using the:

- Proportion of persons living with an IRD that receive informal care
- Number of hours of informal care each person with an IRD receives
- Hourly cost of providing informal care to a person with an IRD, which was calculated based on the AWE in Canada (see Table 7.6).

The utilisation and average weekly hours of informal care were estimated using country specific survey results (Table 7.4 and Table 7.5). 108 survey participants (71.5%) reported receiving informal care, while 43 participants (28.5%) reported that they did not receive any informal care. Survey participants who reported receiving informal care received 11.2 hours of informal care per week on average.

Table 7.4 Percentage of survey participants who receive informal care in the Canada (n=151)

Informal care received	Count (n)	Percentage (%)
Does not receive informal care	43	28.5
Receives informal care	108	71.5
Total	**151**	**100.0**

Source: Deloitte Access Economics analysis.
Note: Components may not sum to totals due to rounding.

Table 7.5 Average weekly hours of informal care received by survey participants receiving informal care in Canada (n=108)

Service	Hours
Average weekly hours of informal care per person	11.2

Source: Deloitte Access Economics analysis.

Table 7.6 Hourly cost of informal care ($ 2019) in Canada

Service	Cost
Hourly cost of informal care	23.0

Source: Deloitte Access Economics analysis.

In order to estimate the costs of informal care, country specific survey results were chosen in favour of published literature given their relevance to the specific IRD cohort. The proportion of persons living with an IRD that receive informal care, the number of hours of informal care each person with an IRD receives and the hourly cost of providing informal care to a person with an IRD were multiplied together to estimate the cost of informal care in the Canada in 2019. Overall, informal care costs were estimated to range between $129.5 to $529.5 million.

For triangulation purposes, the following studies estimating the cost of informal care for persons living with an IRD or condition with a comparable aetiology in Canada were found:

- **Aljied et al (2019)** used data from the Canadian Longitudinal Study on Aging Comprehensive Cohort (for people aged 45-85) and found that 15.1% of people with a visual impairment used informal care at home for an average of 8.3 hours per week, over 28.3 weeks of the year
- **Schmier et al (2006)** conducted a survey of 803 patients with age-related macular degeneration (AMD) in Canada using the Daily Living Tasks Dependent on Vision Questionnaire (DLTV). The average level of informal care usage by all patients was 41.2%
- **Khan et al (2016)** studied the burden among caregivers of people with a visual impairment in Canada. Of the 236 participants (32% legally blind; 68% low vision), carers reported providing a daily average of 2.2 hours of close supervision, or 15.4 hours per week.

7.3.2 Formal care

Formal care can include help from a private nurse or assistance with activities such as childcare, housekeeping and shopping – these costs are not subsidised by private health insurance or the government but instead are out-of-pocket expenses borne by the individual.

The cost of formal care for persons living with an IRD can be estimated by establishing the:

- Proportion of persons living with an IRD that receive formal care
- Number of hours of formal care each person with an IRD receives
- Cost per hour of formal care for a person with an IRD.

The utilisation and average weekly hours of formal care were estimated using the survey data (see Table 7.7, and Table 7.8). 7 survey participants (4.6%) reported receiving formal care, while 144 participants (95.4%) reported that they did not receive any formal care. Survey participants who reported receiving formal care received 15.4 hours of formal care per week on average.

The hourly cost of formal care was estimated using the average hourly wage rates by occupation in the Annual Labour Force Survey published by Statistics Canada (see Table 7.9) (Statistics Canada, 2020).

Table 7.7 Percentage of survey participants who receive formal care in Canada (n=151)

Formal care received	Count (n)	Percentage (%)
Does not receive formal care	144	95.4
Receives formal care	7	4.6
Total	**151**	**100.0**

Source: Deloitte Access Economics analysis.

Table 7.8 Average weekly hours of formal care received by survey participants receiving formal care Canada (n=7)

Service	Hours
Average weekly hours of formal care	15.4

Source: Deloitte Access Economics analysis.

Table 7.9 Hourly cost of formal care ($ 2019) in Canada

Service	Source	Cost
Hourly cost of formal care	Statistics Canada (2020)	21.0

Source: Deloitte Access Economics analysis.

The proportion of persons living with an IRD that receive formal care, the number of hours of formal care each person with an IRD receives, and the cost per hour of formal care for a person with an IRD were multiplied together to estimate the cost of community based formal care in Canada in 2019. Overall, formal care costs were estimated to range between $16.4 to $67.3 million.

For triangulation purposes, the following studies estimating the cost of formal care for persons living with an IRD or condition with a comparable aetiology in Canada were found:

- **Aljied et al (2019)** used data from the Canadian Longitudinal Study on Aging Comprehensive Cohort on the population aged 45-85 and found that 13.3% of people with a visual impairment (binocular acuity worse than 20/60) utilised formal care at home. Aljied et al (2019) also estimated that those who used formal care for a visual impairment did so for an average of 5 hours per week, over 27.6 weeks of the year
- **Jin et al (2019)** analysed the Canadian Community Health Survey – Healthy Aging 2008/2009 and found that Canadians with visual impairment were more likely to utilise formal home care services compared to those without a visual impairment. 23.9% of Canadians aged 45+ years with visual impairment utilised home care services compared to 4.8% of individuals without a visual impairment
- **Schmier et al (2006)** conducted a survey of 803 patients with age-related macular degeneration (AMD) in Canada using the Daily Living Tasks Dependent on Vision Questionnaire (DLTV). The average level of formal care usage by all patients was 16.6%.

7.3.3 Residential care

Persons living with an IRD can experience severe (visual) impairment or disability requiring continuous care and supervision. Such persons may require long term care in the form of a residential care service (Wang et al, 2003). Residential care includes the independent sector residential care, local authority residential care and nursing care.

A targeted literature review was conducted to estimate the cost of residential care due to IRDs in Canada. Where IRD-specific literature was unavailable, the scan was expanded to include conditions with comparable aetiology and international sources. Despite this, no

literature was identified that estimated the cost of residential care for individuals with an IRD or conditions of comparable aetiology (for example visual impairment or blindness). There is also limited evidence on the proportion of residential costs related to blindness or visual acuity that could be attributed to IRDs in Canada.

To overcome the lack of available literature, the cost of residential care was estimated by multiplying the annual average cost per recipient of residential care in Canada and an attributable proportion of costs for individuals with an IRD. The average annual cost per recipient of residential care was based on an estimate of $60,200 per year from Blomqvist et al (2014) and subsequently inflated to 2019 dollars (Table 7.10). UK blindness certifications which suggest that IRDs account for 5.7% of blindness and visual impairment in the UK were used as a proxy for the proportion of individuals with an IRD who utilised residential care in Canada in 2019. The total expenditure on long term residential care for individuals was estimated to range from $16.1 to $66.0 million in 2019 (Table 7.11).

The proportion of total expenditure on long term residential care for IRDs by payer was based on Jain et al (2019) who reported that of the $53.4 billion spent on long-term residential care in 2014, $18.5 billion was paid for by the government (Table 7.12).

The total expenditure on long term residential care for IRDs by payer is shown in Table 7.12.

Table 7.10 Annual average cost per recipient of residential care in Canada in 2019 ($)

Source	Annual average cost per recipient of residential care
Blomqvist et al (2014)	64,909

Source: Blomqvist et al (2014) and Deloitte Access Economics analysis.

Table 7.11 Total expenditure on long term residential care for individuals with an IRD ($ million 2019)

Parameter	Total expenditure on long term residential care
Lower bound estimate	16.1
Upper bound estimate	66.0

Source: Deloitte Access Economics analysis.

Table 7.12 Total expenditure on long term residential care for IRDs ($ million 2019)

Payer	Percentage (%)	Total cost lower bound ($ million)	Total cost upper bound ($ million)
Individuals	65.4	10.6	43.2
Government	34.6	5.6	22.9
Total	**100.0**	**16.1**	**66.0**

Source: Deloitte Access Economics analysis.
Note: Components may not sum to totals due to rounding.

7.4 Deadweight losses (DWL) of taxation payments

The act of taxation and redistribution creates distortions and inefficiencies in the economy, so transfers also involve real net costs to the economy, known as deadweight losses. This section calculates the deadweight losses resulting from lost taxation (which must be raised elsewhere) and government expenditure on welfare payments and the health system.

This section estimates the deadweight losses arising from raising and administering taxation (i.e. transfer) payments, including government and social insurance expenditure on health and welfare.

Taxes alter the price and quantity of goods sold compared to what they would be if the market were not distorted. In a practical sense, this distortion reveals itself as a loss of efficiency in the economy, which means that raising $100 of taxation revenue requires consumers and producers to give up more than $100 of value. This, in turn, creates a reduction in consumer and producer surplus by diminishing the value of trade between these parties that would otherwise be achieved – a deadweight loss. As such, deadweight losses represent a real net cost to the economy.

7.4.1 Welfare payments

The cost of welfare payments themselves are not included in the cost of IRDs. However, the deadweight losses arising from raising and administering welfare payments are included in the total cost of IRDs.

Persons living with an IRD may be eligible for several forms of government support, the:

- Canada Pension Plan Disability Benefit/ Quebec Pension Plan Disability Benefit
- Disability Tax Credit (DTC).

Additionally, informal carers of a person or persons living with an IRD may be eligible to receive the Canada Caregiver Credit (CCC).

Welfare payments were estimated by applying the average utilisation of welfare payments to the weekly entitlement associated with each payment. Average utilisation was estimated using country-specific survey data of the proportion of respondents who received each payment. The weekly entitlement amount for each welfare payment was calculated based on the weighted average amount received by all recipients of that welfare type.[3]

Most Canadian survey respondents (69.5%) reported receiving support from their government (Table 7.13).

[3] Individuals receive different amounts for the same welfare type depending on their level of need.

Table 7.13 Welfare payment utilisation of survey participants in Canada (n=151)

Government support utilisation	Count	Percentage (%)
Supported	105	69.5
Unsupported	46	30.5
Total	**151**	**100.0**

Source: Deloitte Access Economics survey.

Table 7.14 Welfare payments received by persons living with an IRD ($ 2019) by welfare type

Type	Welfare	Entitlement ($)
Individual	Canada Pension Plan/Quebec Pension Plan Disability Benefit	12,126.8
Individual	Disability Tax Credit	6,662.5
Carer	Canada Caregiver Credit	2,230 plus up to a maximum of 7,140 per year

Source: Deloitte Access Economics analysis.

These average welfare payments were applied to data on individual payment types to estimate the total cost of welfare payments to persons living with an IRD, which ranged between $100.2 to $409.7 million in 2019.

In addition, the total cost of welfare payments to carers of persons living with an IRD was estimated to range between $13.0 to $53.0 million in 2019. Therefore, the total welfare payments due to IRDs were estimated to range between $113.1 to $462.6 million in 2019.

7.4.2 Reduced taxation revenue

Lower employment participation and lower output (e.g. due to absenteeism or presenteeism) reduce the possible taxation revenue government can collect. The reduction in taxation revenue was estimated by applying an average personal income or company tax rate – which were assumed to be 23.2% and 15.0%, respectively – to lost individual or company earnings.

The total reduction in taxation revenue was estimated to range between $76.3 to $312.2 million in 2019.

7.4.3 Deadweight loss of taxation payments and administration

The deadweight loss due to lost taxation revenue (given an assumption of no change in spending) or additional expenditure on government programs (e.g. health or welfare) can be estimated by applying the marginal burden of taxation to the total of lost taxation and government expenditures. This marginal burden was estimated to be 14% (Dahlby and Ferede, 2011).

The total deadweight loss due to IRDs were estimated to range between $38.3 to $155.7 million in 2019, as shown in Table 7.15.

Table 7.15 Deadweight losses due to IRDs ($ million 2019) (lower and upper bound scenario)

Cost type	Total cost Lower bound	Total cost Upper bound	Resulting deadweight loss Lower bound	Resulting deadweight loss Upper bound
Welfare payments	113.1	462.6	16.2	66.2
Government programs (e.g. health)	78.5	314.0	11.2	44.9
Lost individual taxes	38.1	155.7	5.4	22.3
Lost carer taxes	30.0	122.8	4.3	17.6
Lost company taxes	8.2	33.7	1.2	4.8
Total	**267.9**	**1,088.8**	**38.3**	**155.7**

Source: Deloitte Access Economics analysis.
Note: Components may not sum to totals due to rounding.

7.5 Results

Total other costs due IRDs in Canada in 2019 were estimated to be range between $309.8 to $1,265.9 million (lower to upper bound). Informal care resulted in the greatest proportion of other costs between $129.5 to $529.5 million (41.8%), followed by aids and modifications (between $109.4 to $447.4 million, 35.5%), deadweight losses (between $38.3 to $155.7 million, 12.4%), formal care ($16.4 to $67.3 million, 5.3%) and residential care costs (between $16.1 to $66.0 million, 5.2%) (Table 7.16).

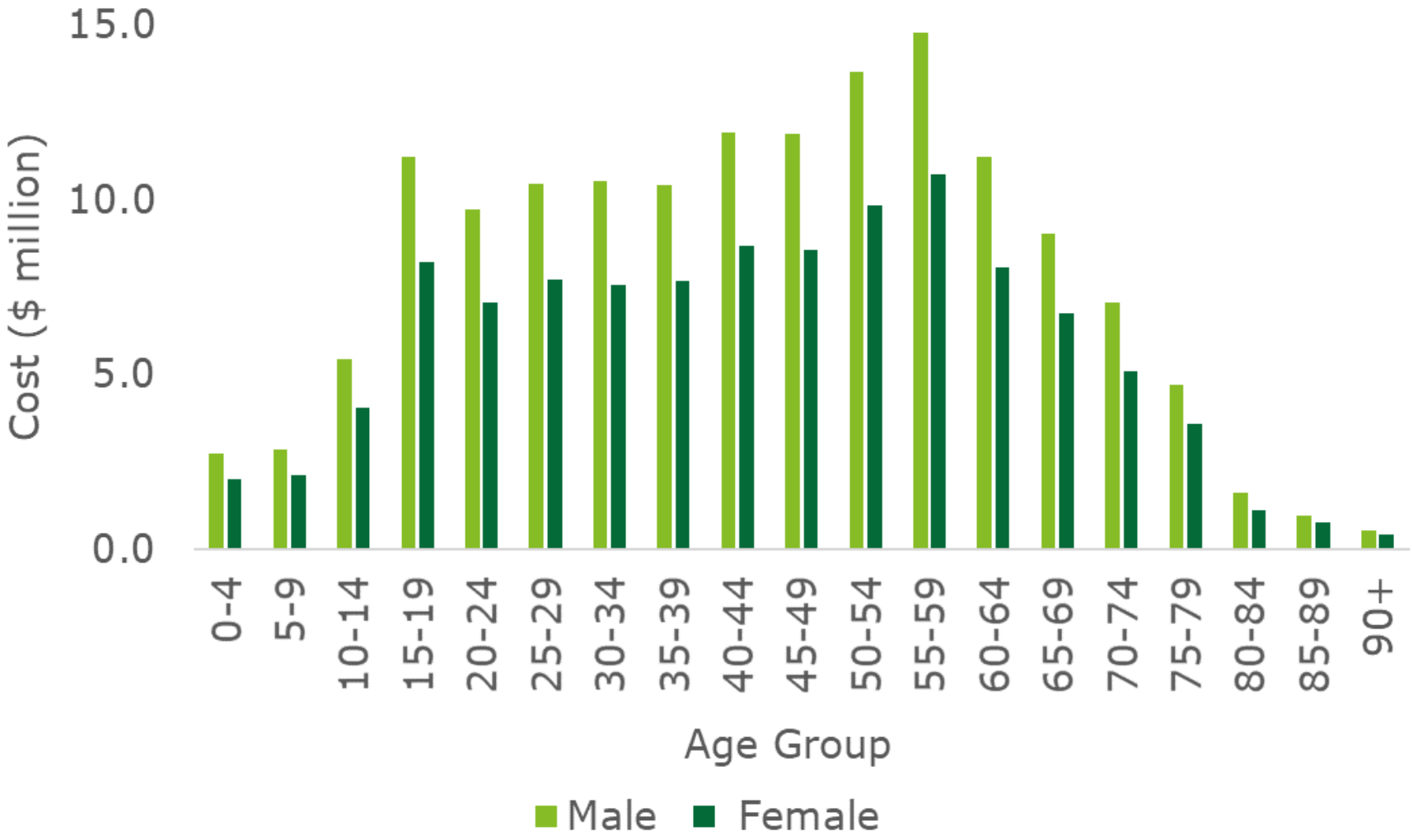
The largest component of this cost is borne by families (37%), with government, individuals and society incurring 17%, 34% and 12% of the costs, respectively. Other costs by payer are summarised in Chart 7.3.

Table 7.16 Total other costs of IRDs ($ 2019) in Canada by cost type (lower and upper bound scenario)

Cost type	Cost per person ($) Lower bound	Cost per person ($) Upper bound	Total cost ($ million) Lower bound	Total cost ($ million) Upper bound
Informal care	6,180	6,180	129.5	529.5
Aids and modifications	5,223	5,223	109.4	447.4
Deadweight losses	1,829	1,817	38.3	155.7
Formal care	785	785	16.4	67.3
Residential care	771	771	16.1	66.0
Total	**14,788**	**14,776**	**309.8**	**1,265.9**

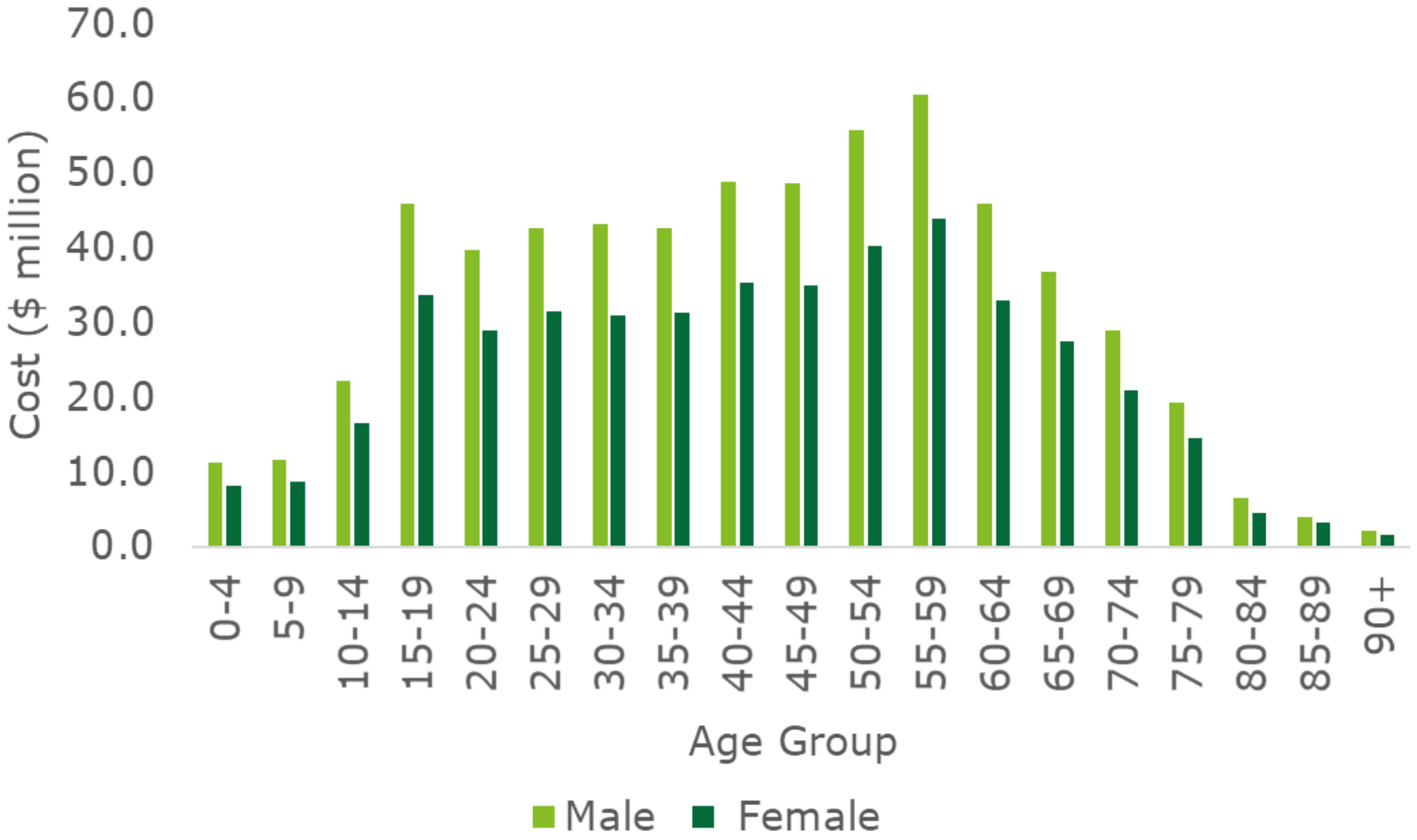
Source: Deloitte Access Economics analysis.
Note: Components may not sum to totals due to rounding.

Chart 7.1 Total other costs of IRDs ($ million, 2019) in the Canada by age and sex (lower bound scenario)



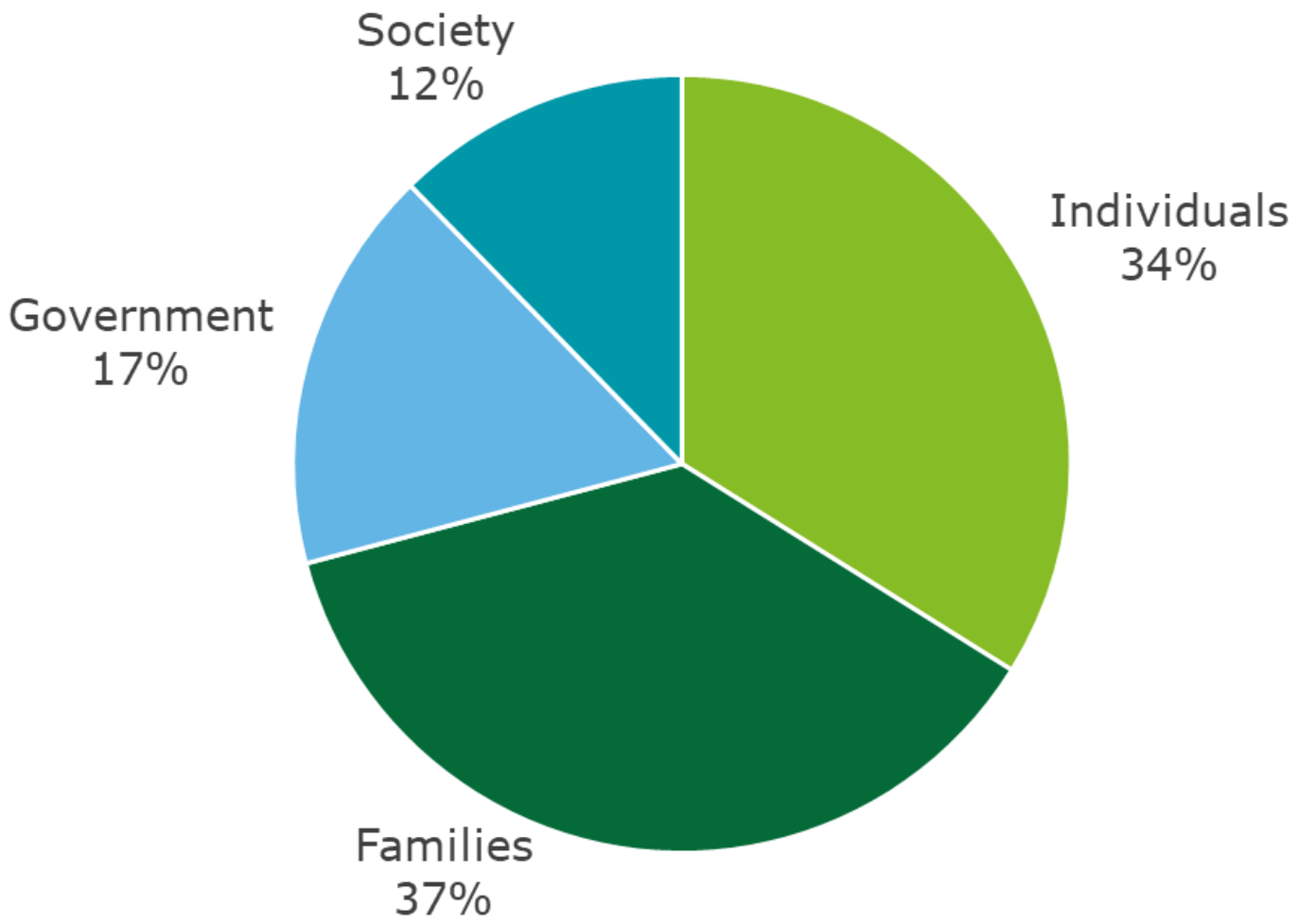
Source: Deloitte Access Economics analysis.

Chart 7.2 Total other costs of IRDs ($ million, 2019) in Canada by age and sex (upper bound scenario)



Source: Deloitte Access Economics analysis.

Chart 7.3 Proportion (%) of other costs of IRDs in Canada by payer (low and upper bound scenario)



Source: Deloitte Access Economics analysis.

8 Loss of wellbeing

This section describes the results from the survey of persons living with an IRD and/or the parents of children (under 18) living with an IRD in Canada.

Key findings

- In 2019, persons living with an IRD in Canada were estimated to experience between 3,205 to 13,108 (lower to upper bound) disability adjusted life years (DALYs).
- Total wellbeing costs associated with IRD in 2019 were estimated to be $51,147 per person and between $1,071.4 to $4,381.9 million in total.

8.1 Summary of approach

Loss of wellbeing were estimated using the World Health Organisation (WHO) burden of disease methodology. This is a non-financial approach, where pain, suffering and premature mortality are measured in terms of DALYs.

DALYs are composed of premature mortality (years of life lost due to premature death – YLL) and morbidity (years of healthy life lost due to disability – YLD) components. DALYs are calculated by assigning disability weights to various health states, where zero represents a year of perfect health and one represents death. Other health states are given a weight between zero and one to reflect the quality of life that is lost due to a particular condition. For example, a disability weight of 0.2 is interpreted as a 20% loss in the quality of life relative to perfect health for the duration of the condition.

Disability weights were obtained from the IHME's *Global Burden of Disease Study 2017* and are classed into three levels of severity (Table 8.1). A disability weight were applied to the prevalence of IRDs in Canada and discounted at a rate of 3% consistent with WHO methodology (WHO, n.d.).

Table 8.1 Disability weights per person due to 'other vision loss' (2017)

Sequela	Health state (severity of VI)	Health state description	Disability weight
Moderate vision impairment due to other vision loss	Distance vision, moderate impairment	Vision problems that make it difficult to recognise faces or objects across a room	0.031
Severe vision impairment due to other vision loss	Distance vision, severe impairment	Severe vision loss, which causes difficulty in daily activities, some emotional impact, and some difficulty going outside the home without assistance	0.184
Blindness due to other vision loss	Distance vision blindness	Completely blind, which causes great difficulty in some daily activities, worry and anxiety, and great difficulty going outside the home without assistance	0.187

Source: IHME (2018).

For the purposes of this report, a single disability weight of 0.153 was applied to DALYs. This reflects an average of the three disability weights by severity, weighted by the estimated distribution of prevalent IRD cases in Canada across the three levels of severity.[4]

DALYs were converted into pounds using an estimate of the value of a statistical life year (VSLY). The VSLY is an estimate of the value society places on an anonymous life. No national guidance on the empirical VSLY in Canada was found. As such, the VSLY is drawn from a systematic review of economic studies providing data on the empirical data on the value of a statistical life (VSL) published between 1995 and 2015 by Schlander et al (2018). This study reports an estimated VSLY for North America, Europe, Asia and Other. The VSLY for North America is $417,050 per person which was inflated to 2019 terms for use in the cost-of-illness modelling.

The disability weight estimates are supported by the survey results wherein the vast majority of participants reported experiencing negative impacts on their wellbeing as a result of their IRD, with 84.8% reporting experiencing anxiety, 74.2% depression, 48.4% social isolation, 55.1% financial stress and 13.2% 'other' wellbeing effects (Table 8.2). The 'other' category included negative impacts on persons living with an IRD wellbeing due to fear, stress, fatigue, hopelessness, loneliness, panic attacks, suicidal thoughts and the development of tics.

[4] The distribution of cases of VI/blindness across the severities was estimated based on the average of studies by Bunce (2006), Bunce (2010) and Quartilho (2016), using vision loss certifications in the UK (NHS, 2017) and the estimated prevalence of IRDs in the UK relative to the estimated prevalence of IRDs in Canada.

Table 8.2 Wellbeing status of survey participants in Canada (n=151)

Wellbeing (persons living with an IRD)	Percentage (%)
Anxiety	84.8
Depression	74.2
Social isolation	48.4
Financial stress	55.1
Other	13.2

Source: Deloitte Access Economics analysis.
Note: This question allowed for the selection of multiple answers; hence, the percentages do not sum to 100.

Furthermore, the wellbeing effects of IRDs are not limited to persons living with an IRD. 69.2% of survey participants reported that close family members had experienced feelings of depression, anxiety, or another mental health condition as a result of their IRD. The parents of children (under 18) living with an IRD reported feelings of anxiety (84.6%), depression (61.5%), and 'other' mental health impacts (92.3%) (Table 8.3). The 'other' category included negative impacts on the parents of children (under 18) living with an IRD wellbeing due to stress, loneliness and concerns for the safety of their child.

In addition, the majority of persons living with an IRD (66.6%) and the parents of children (under 18) living with an IRD (84.7%) were frustrated by the lack of awareness and support for IRDs (Table 8.4).

Table 8.3 Wellbeing status of survey participants (the parents of children (under 18)) in Canada (n=13)

Wellbeing (the parents of children (under 18) living with an IRD)	Percentage (%)
Anxiety	84.6
Depression	61.5
Other mental health impacts	92.3

Source: Deloitte Access Economics analysis.
Note: This question allowed for the selection of multiple answers; hence, the percentages do not sum to 100.

Table 8.4 Degree of survey participant frustration with the lack of awareness and support for IRDs in Canada (n=13)

Participant type	Strongly disagree (%)	Disagree (%)	Neutral (%)	Agree (%)	Strongly agree (%)
Persons living with an IRD	5.8	5.1	22.5	30.4	36.2
The parents of children (under 18) living with an IRD	0.0	0.0	15.4	46.2	38.5

Source: Deloitte Access Economics analysis.

8.2 Results

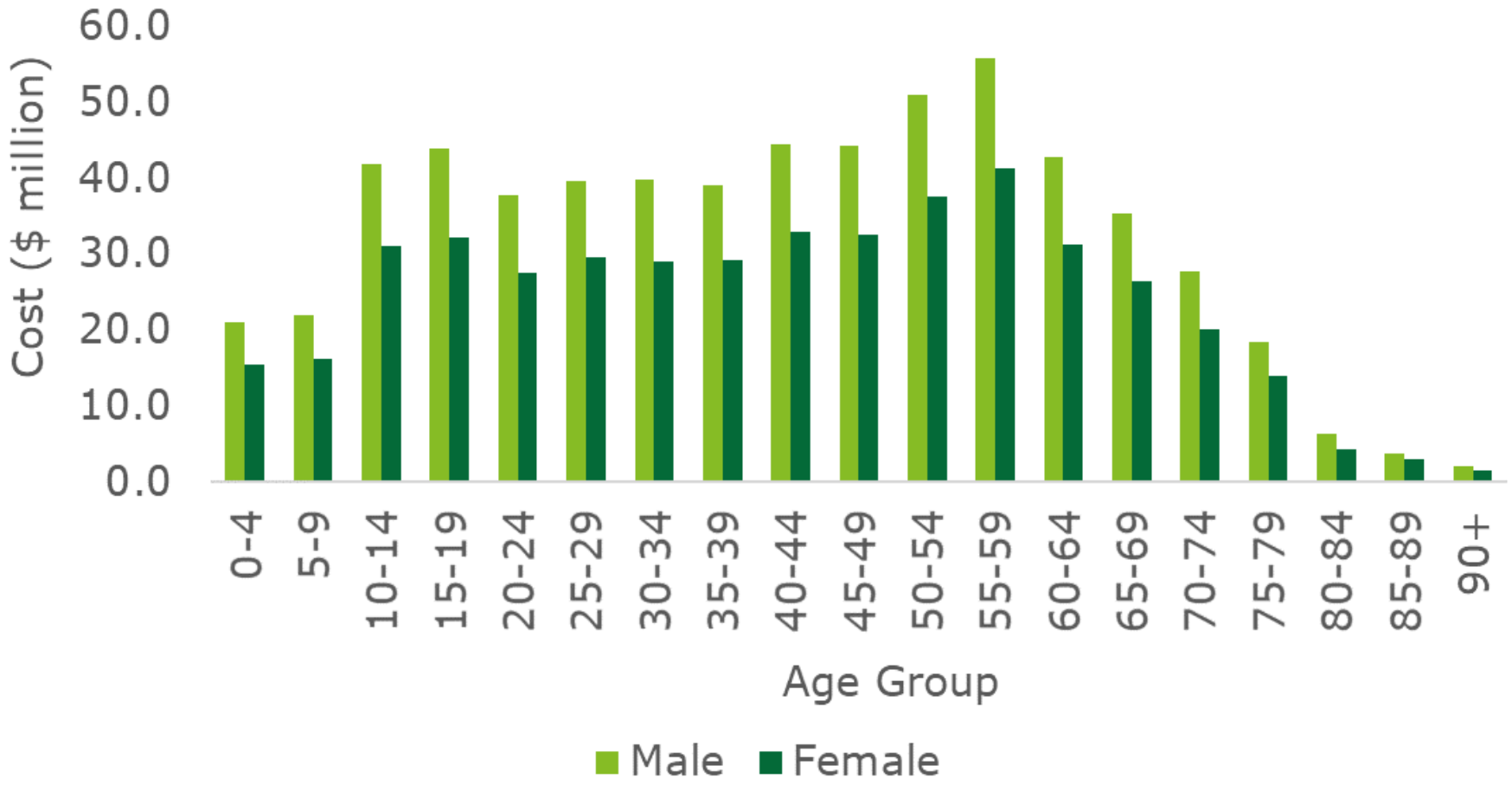
In 2019, persons living with an IRD in Canada were estimated to experience between 3,205 to 13,108 (lower to upper bound) DALYs overall. This equated to total wellbeing costs of $1,071.4 to $4,381.9 million or $51,147 per person (see Table 8.5). Loss of wellbeing by age and sex are displayed in Chart 8.1.

Table 8.5 Total DALYs and wellbeing costs of IRDs (2019) in Canada (lower and upper bound scenario)

Parameter	Per person ($) Lower bound	Per person ($) Upper bound	Total ($ million) Lower bound	Total ($ million) Upper bound
DALYs	0.15	0.15	3,205	13,108
Loss of wellbeing	**51,147**	**51,147**	**1,071.4**	**4,381.9**

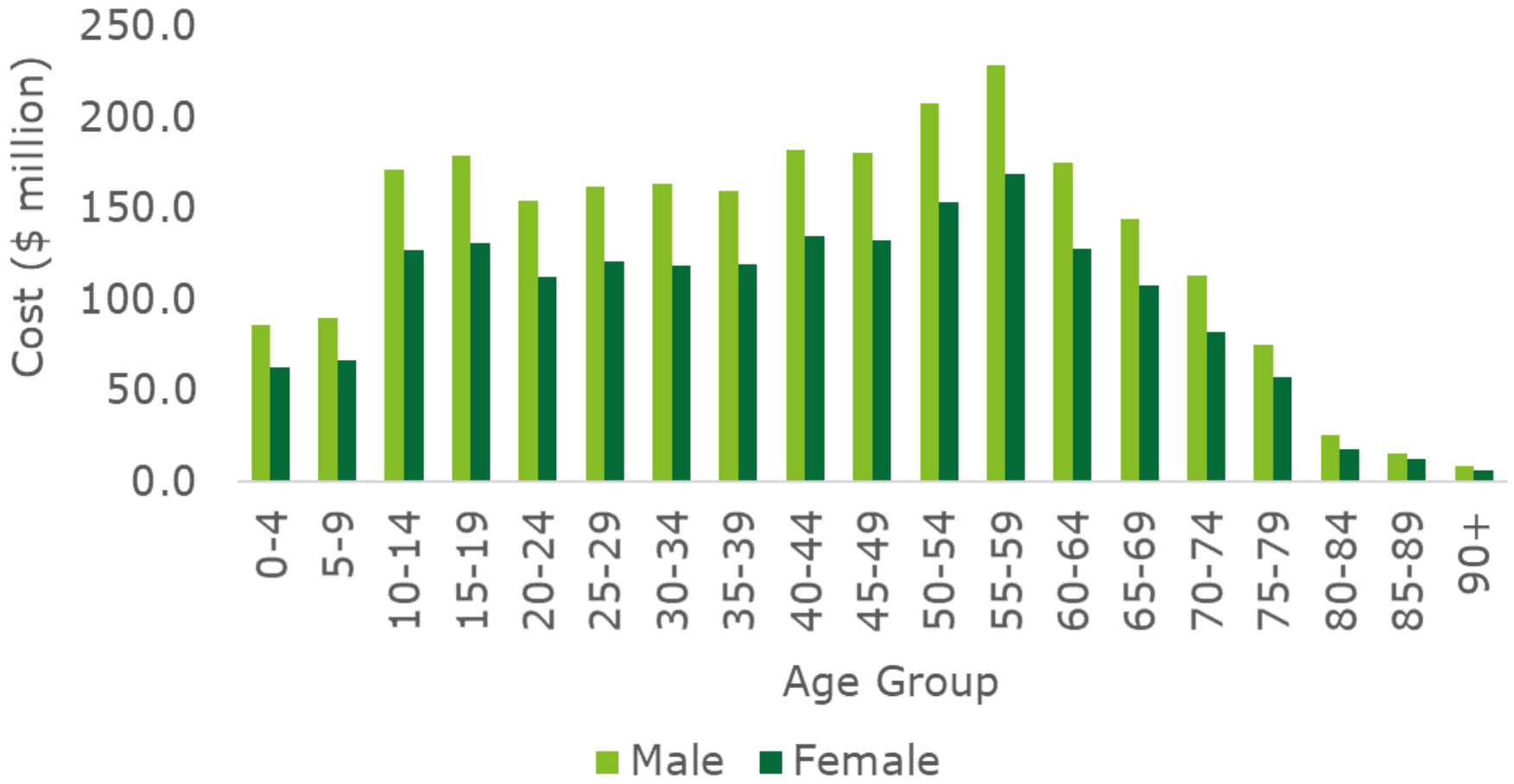
Source: Deloitte Access Economics analysis.

Chart 8.1 Total wellbeing costs of IRDs ($ million, 2019) in Canada by age and sex (lower bound scenario)



Source: Deloitte Access Economics analysis.

Chart 8.2 Total wellbeing costs of IRDs ($ million, 2019) in Canada by age and sex (upper bound scenario)



Source: Deloitte Access Economics analysis.

9 Sensitivity analysis

In many cases, the inputs underlying the cost-of-illness analysis are uncertain and changes in these inputs may have a significant impact upon the total estimate of the costs of IRDs in Canada in 2019. Sensitivity analysis was undertaken by adjusting the unit costs of each cost component by +10% (High scenario) and -10% (low scenario).

9.1 Results

The high sensitivity scenario yielded an overall estimate of the cost of IRDs in Canada between $1,813.4 to $7,404.2 million (lower to upper bound). The variance between components under each prevalence estimate are shown in Table 9.1. The most sensitive components were wellbeing costs, followed by productivity losses as shown in Chart 9.1 and Chart 9.2.

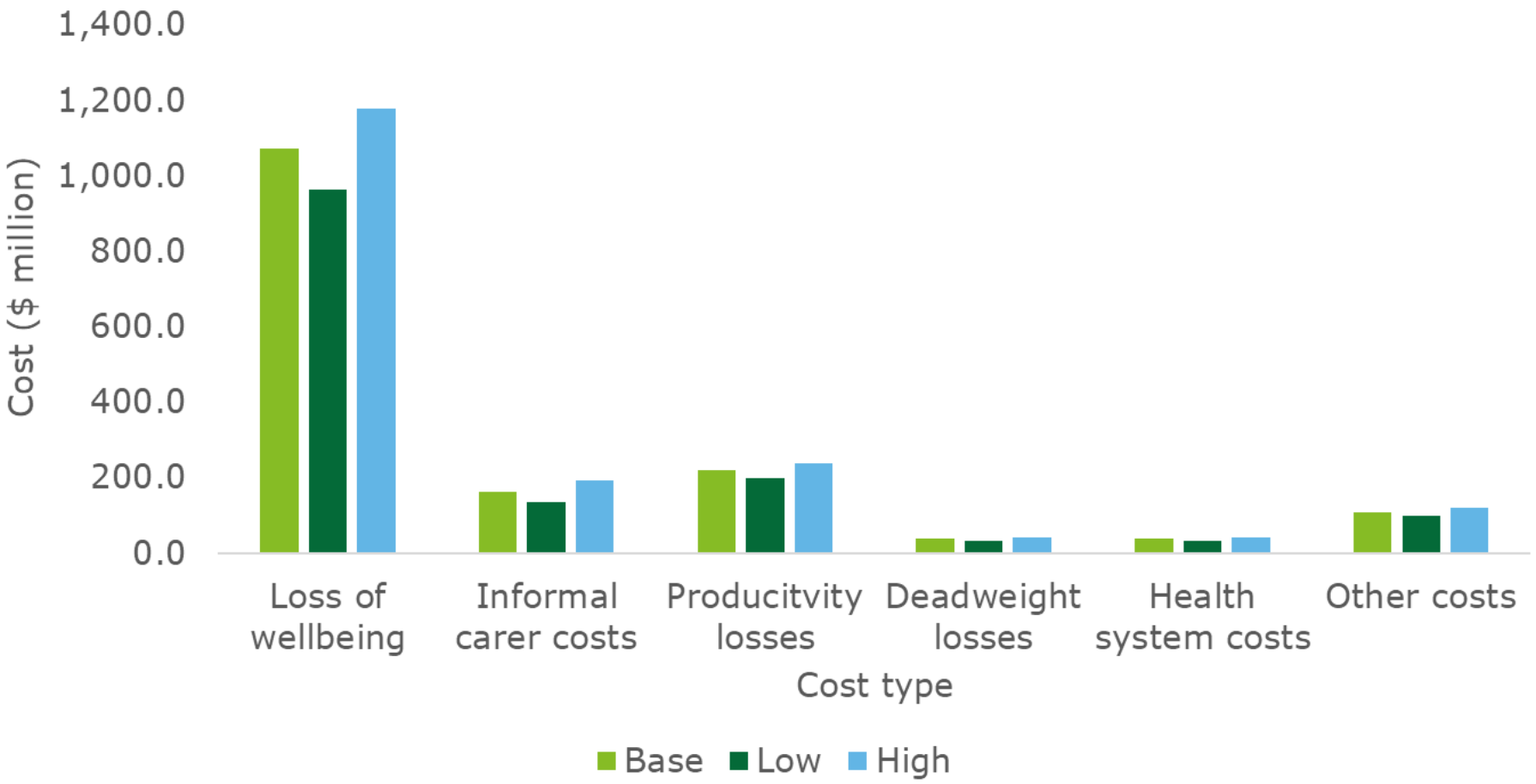
Table 9.2, Chart 9.3 and Chart 9.4 show the total costs under each scenario disaggregated by condition.

Table 9.1 Sensitivity analysis results ($ million, 2019) in the Canada by cost type (lower and upper bound scenario)

Cost type	Base case Lower bound	Low	High	Base case Upper bound	Low	High
Loss of wellbeing	1,071.4	964.2	1,178.5	4,381.9	3,943.7	4,820.0
Informal carer costs	162.0	134.2	192.5	662.7	548.8	787.3
Productivity losses	219.0	200.1	237.5	895.5	818.3	971.2
Deadweight losses	38.3	33.8	43.0	155.7	137.3	174.8
Health system costs	37.8	34.0	41.5	144.3	129.9	158.7
Other costs	109.4	98.5	120.3	447.4	402.7	492.2
Total costs	**1,637.8**	**1,464.7**	**1,813.4**	**6,687.5**	**5,980.6**	**7,404.2**

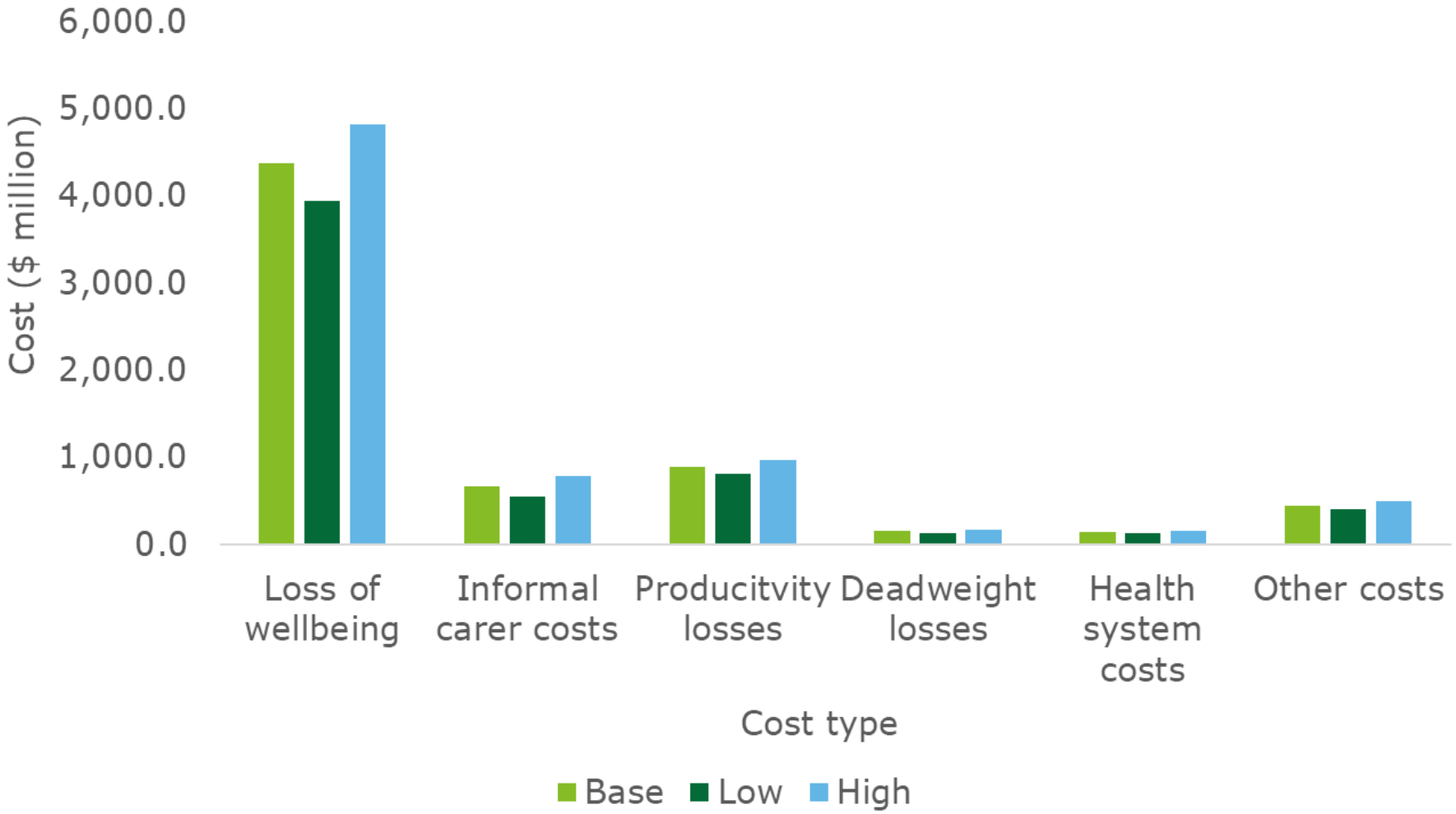
Source: Deloitte Access Economics analysis.

Chart 9.1 Sensitivity analysis results ($ million, 2019) in Canada by cost type (lower bound scenario)



Source: Deloitte Access Economics analysis.

Chart 9.2 Sensitivity analysis results ($ million, 2019) in Canada by cost type (upper bound scenario)



Source: Deloitte Access Economics analysis.

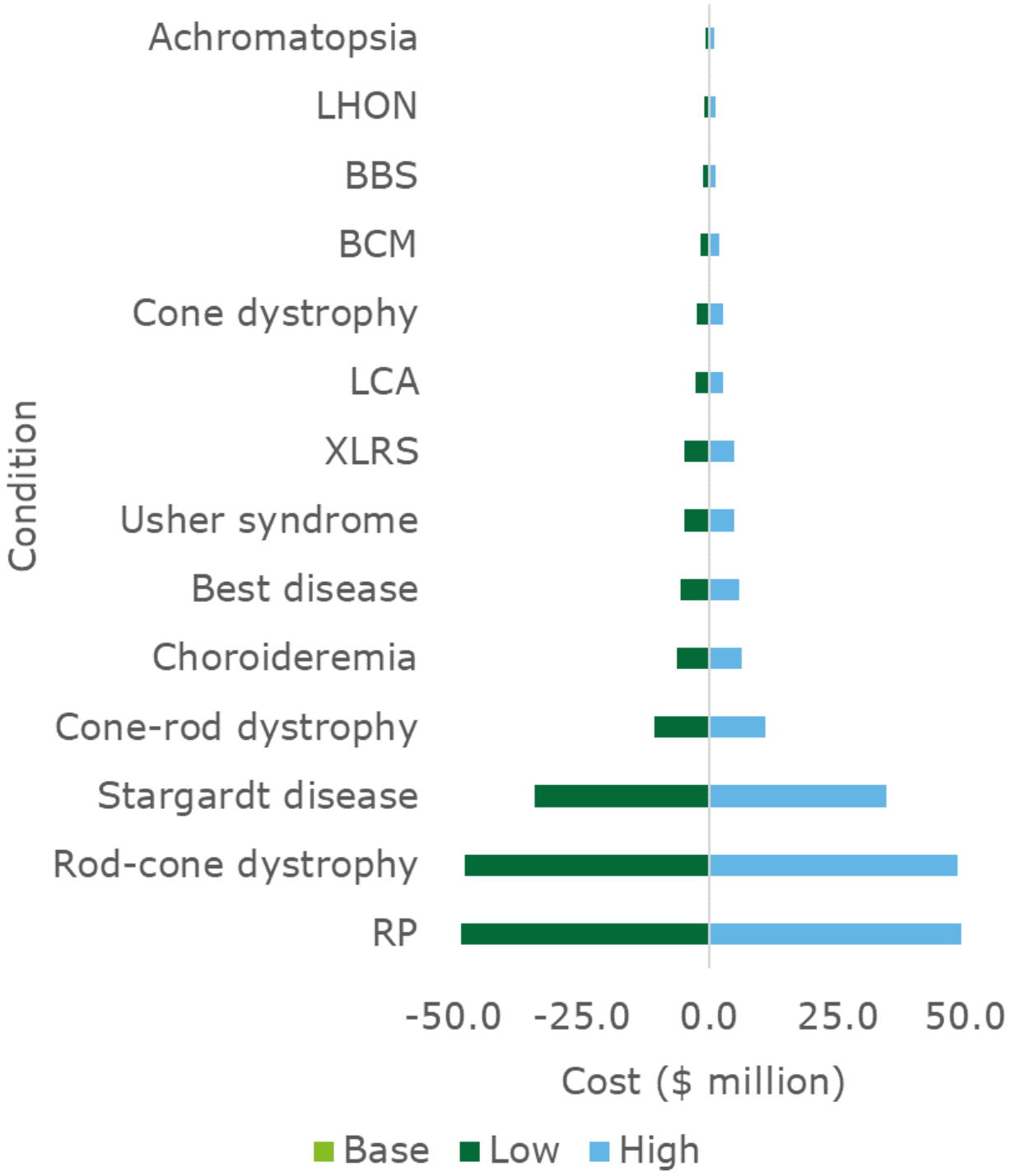
Table 9.2 Sensitivity analysis results ($ million, 2019) in the Canada by condition (lower and upper bound scenario)

IRD	Base Lower bound	Low	High	Base Upper bound	Low	High
Rod-cone dystrophy	459.1	410.6	508.2	1,874.4	1,676.4	2,075.1
Retinitis pigmentosa (RP)	452.5	404.6	501.0	1,847.5	1,652.1	2,045.6
Stargardt disease	322.6	288.5	357.3	1,317.3	1,177.8	1,458.7
Cone-rod dystrophy	102.3	91.5	113.2	417.6	373.5	462.4
Choroideremia	59.4	53.1	65.7	242.5	216.9	268.4
Best disease	53.9	48.2	59.7	220.3	197.0	243.9
X linked retinoschisis (XLRS)	46.1	41.3	51.0	188.3	168.5	208.4
Usher syndrome	45.2	40.4	50.0	184.5	165.0	204.3
Leber congenital amaurosis (LCA)	25.6	22.9	28.4	104.7	93.6	115.9
Cone dystrophy	24.4	21.8	27.0	99.6	89.1	110.3
Blue Cone Monochromacy (BCM)	17.4	15.5	19.2	70.9	63.5	78.5
Bardet-Biedl Syndrome (BBS)	11.2	10.0	12.4	45.9	41.0	50.8

Leber's hereditary optic neuropathy (LHON)	10.3	9.2	11.4	42.2	37.8	46.7
Achromatopsia	7.8	7.0	8.7	31.9	28.5	35.3
Total IRDs	**1,637.8**	**1,464.7**	**1,813.4**	**6,687.5**	**5,980.6**	**7,404.2**

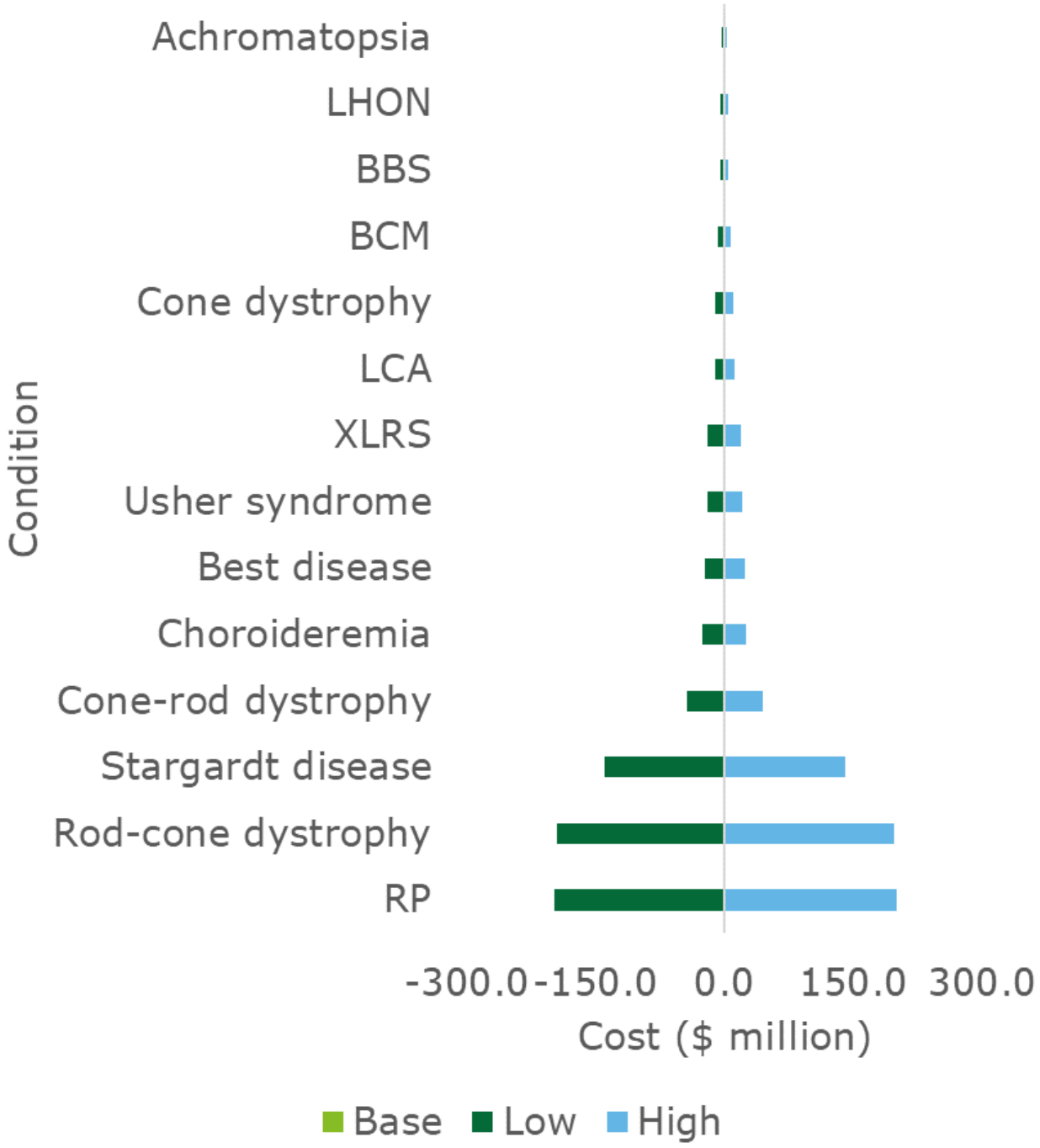
Source: Deloitte Access Economics analysis.

Chart 9.3 Sensitivity analysis results ($ million, 2019) in Canada by condition (lower bound scenario)



Source: Deloitte Access Economics analysis.

Chart 9.4 Sensitivity analysis results ($ million, 2019) in Canada by condition (upper bound scenario)



Source: Deloitte Access Economics analysis.

10 Conclusion

Cost-of-illness studies are conducted with the intent of describing the economic burden imposed by a disease, on a specific population (Larg et al, 2011). The results from cost-of-illness studies can be used to justify investment in preventive or treatment interventions, inform funding allocation and prioritisation, provide a basis for policy and planning, and provide inputs for economic analyses. This could include cost benefit analyses (CBA), cost effectiveness analyses (CEA), cost utility analyses (CUA), and cost minimisation analyses (CMA) (Rice et al, 2000; The Joanna Briggs Institute, 2015).

This study fills an important gap by bringing together the best available evidence to provide an estimate of the economic and wellbeing cost of IRDs in Canada in 2019. The results from this cost-of-illness study could be used to support increasing the awareness of IRDs and/or provide inputs for screening and treatment reimbursement decisions.

Many of the epidemiological, economic and wellbeing parameters required to model the socioeconomic burden of IRDs in Canada in 2019 were supported by limited available evidence. This means that in many cases the inputs underlying this study are uncertain, and changes in these inputs and parameters may have a significant impact upon the total estimate of the costs of IRDs in Canada in 2019. To assist in quantifying the impact of this uncertainty, a scenario analysis approach was used whereby the most uncertain estimate (the prevalence of IRDs) was estimated via both a lower and upper bound approach. These lower and upper bound scenarios are presented within each results section throughout the report.

In summary, this analysis found that IRDs imposed significant economic and wellbeing costs on the Canadian population in 2019. As it currently stands, persons living with an IRD incur significant economic costs and reductions in their quality of life. In addition to the costs incurred by these persons, their families, friends, government, employers and society all incur significant economic costs due to IRDs.

10.1 Summary of total costs

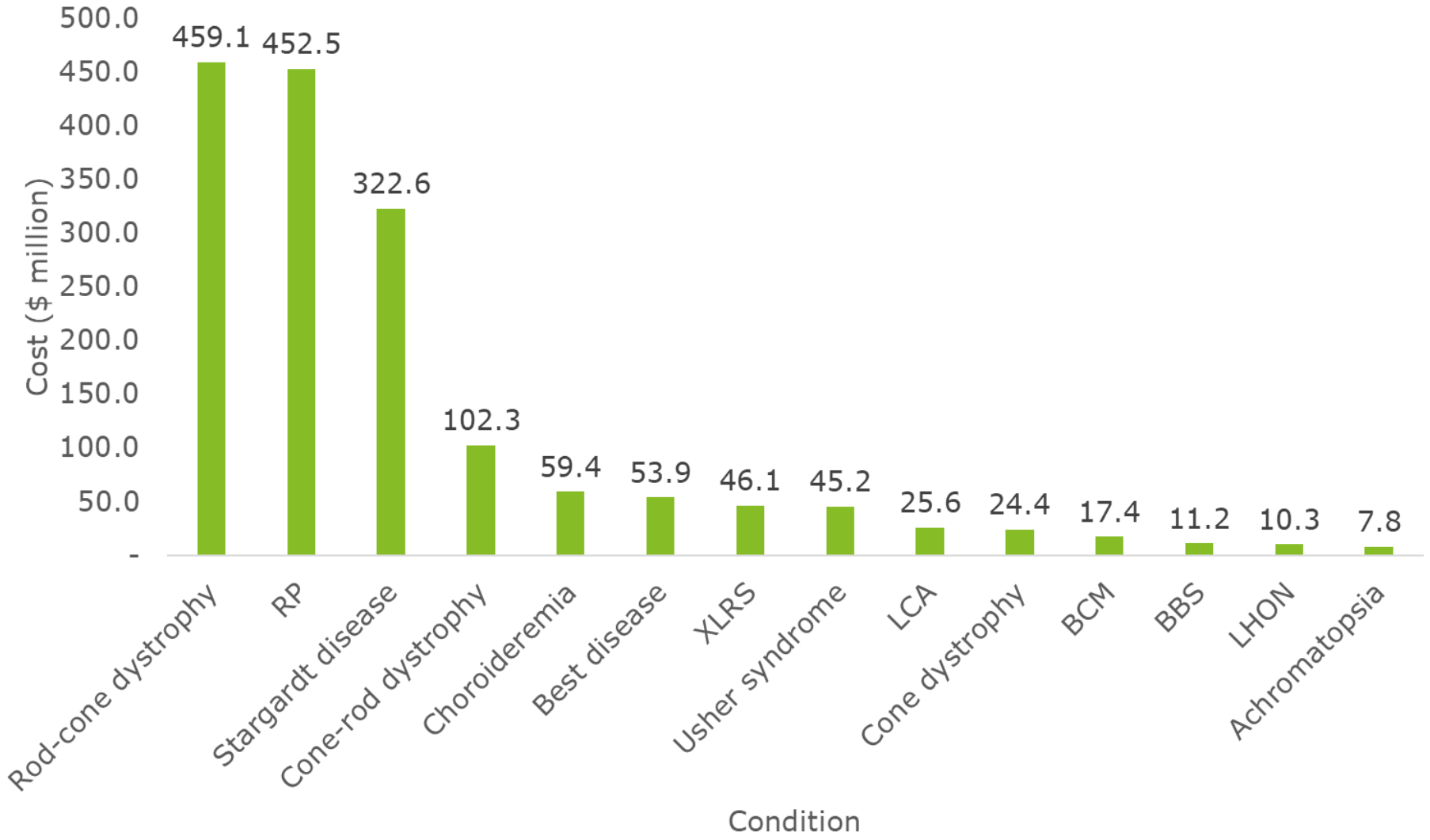
Total costs attributable to IRDs in Canada were estimated to range between $1,637.8 to $6,687.5 million in 2019, comprising both economic costs (between $566.6 to $2,305.7 million) and wellbeing costs (between $1,071.4 to $4,381.9 million).

Of the 14 IRDs within scope, rod-cone dystrophy incurred the greatest proportion of total costs between $459.1 to $1,874.4 million (28.0%), followed by RP (between $452.5 to $1,847.5 million, 27.6%), Stargardt disease (between $322.6 to $1,317.3 million, 19.7%) and cone-rod dystrophy (between $102.3 to $417.6 million, 6.2%) (Chart 10.1, Chart 10.2 and Table 10.1).

Loss of wellbeing comprised the largest share of total costs between $1,107.4 to $4,381.9 million (65.4%), followed by followed by productivity losses (between $219.0 to $895.5 million, 13.4%), informal carer costs (between $162259.5 to $1,063.3 million, 9.9%, other costs (between $109.4 to $447.4 million, 6.7%), deadweight losses (between $38.3 to $155.7 million, 2.3%) and health system costs (between $37.8 to $144.3 million, 2.3%) (Chart 10.3, Chart 10.4, Chart 10.5, Chart 10.6, Chart 10.7, Table 10.2, Table 10.3 and Table 10.4). This means that in addition to imposing significant economic costs, IRDs result in pain and suffering that leads to a significant wellbeing cost for persons living with an IRD.

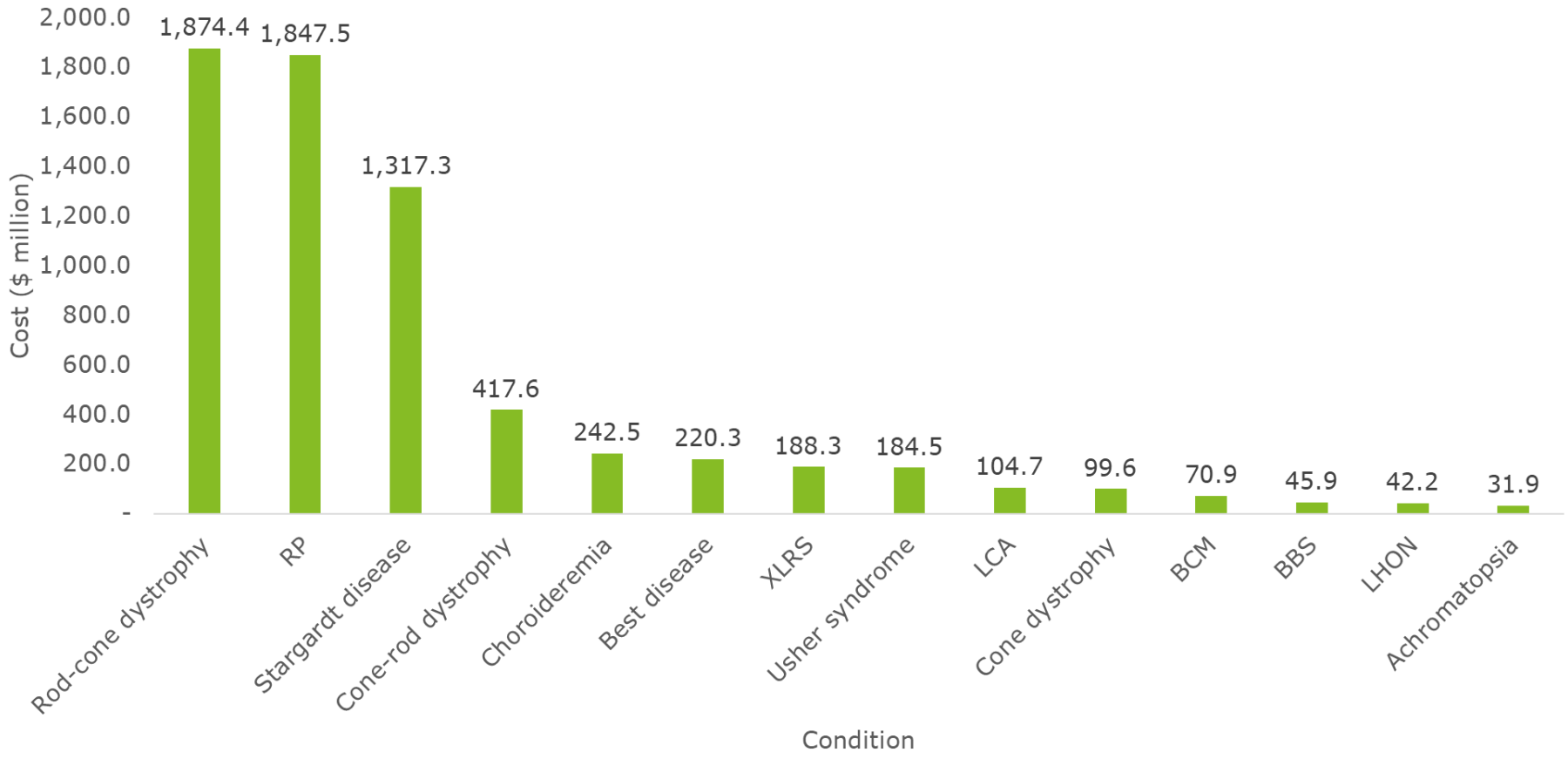
Individuals bear the largest share of total costs at 80.0%, followed by government (7.5%), family/friends (7.0%), employers (2.9%), and society/other (2.7%) (Chart 10.8).

Chart 10.1 Total cost of IRDs ($ million, 2019) in Canada by condition (lower bound scenario)



Source: Deloitte Access Economics analysis.

Chart 10.2 Total cost of IRDs ($ million, 2019) in Canada by condition (upper bound scenario)



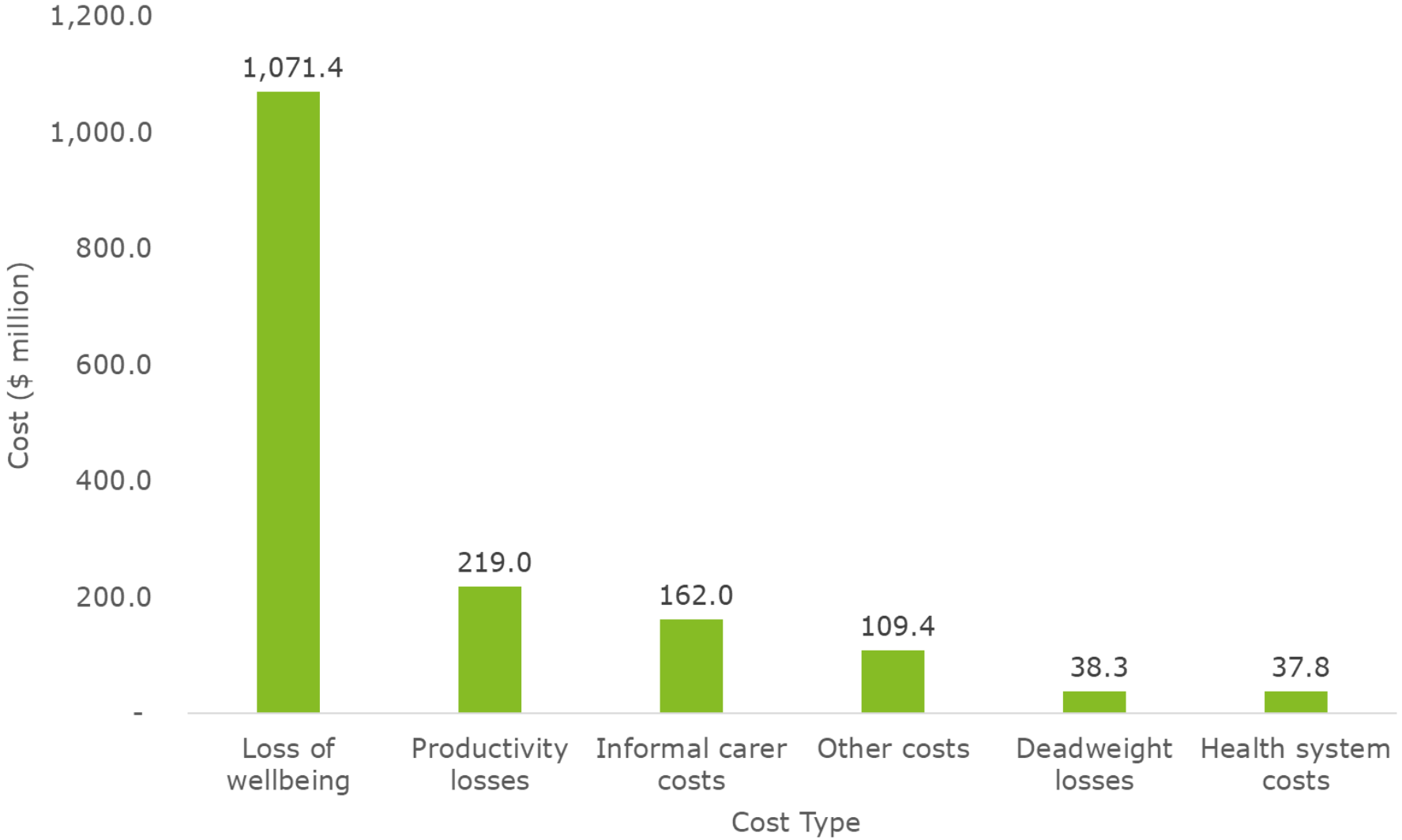
Source: Deloitte Access Economics analysis.

Table 10.1 Total costs of IRDs ($ million, 2019) in Canada by condition (lower and upper bound scenario)

IRD	Cost Lower bound	Cost Upper bound
Rod-cone dystrophy	459.1	1,874.4
RP	452.5	1,847.5
Stargardt disease	322.6	1,317.3
Cone-rod dystrophy	102.3	417.6
Choroideremia	59.4	242.5
Best disease	53.9	220.3
XLRS	46.1	188.3
Usher syndrome	45.2	184.5
LCA	25.6	104.7
Cone dystrophy	24.4	99.6
BCM	17.4	70.9
BBS	11.2	45.9
LHON	10.3	42.2
Achromatopsia	7.8	31.9
Total	**1,637.8**	**6,687.5**

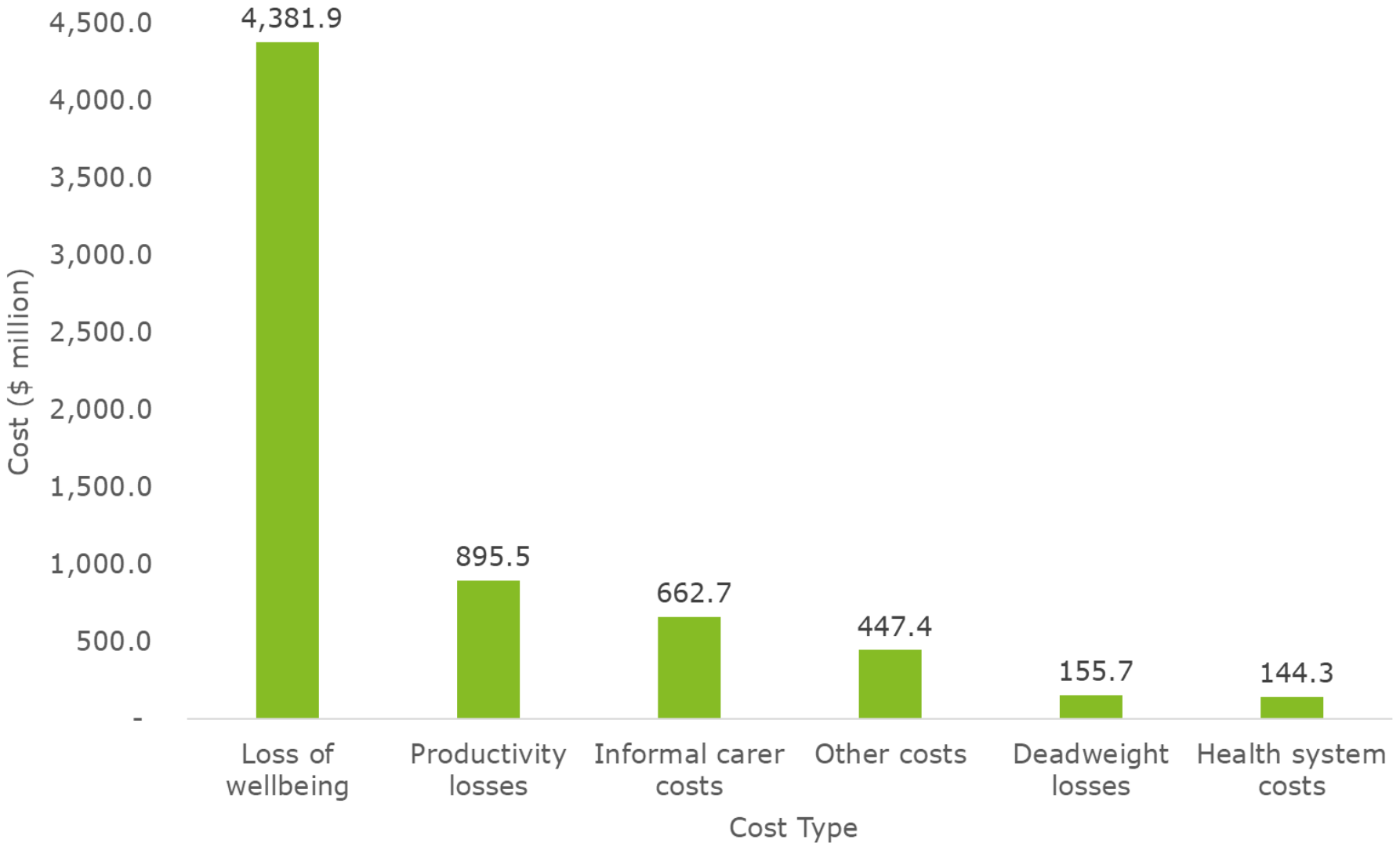
Source: Deloitte Access Economics analysis.
Note: Components may not sum to totals due to rounding.

Chart 10.3 Total costs of IRDs ($ million, 2019) in Canada by cost type (lower bound scenario)



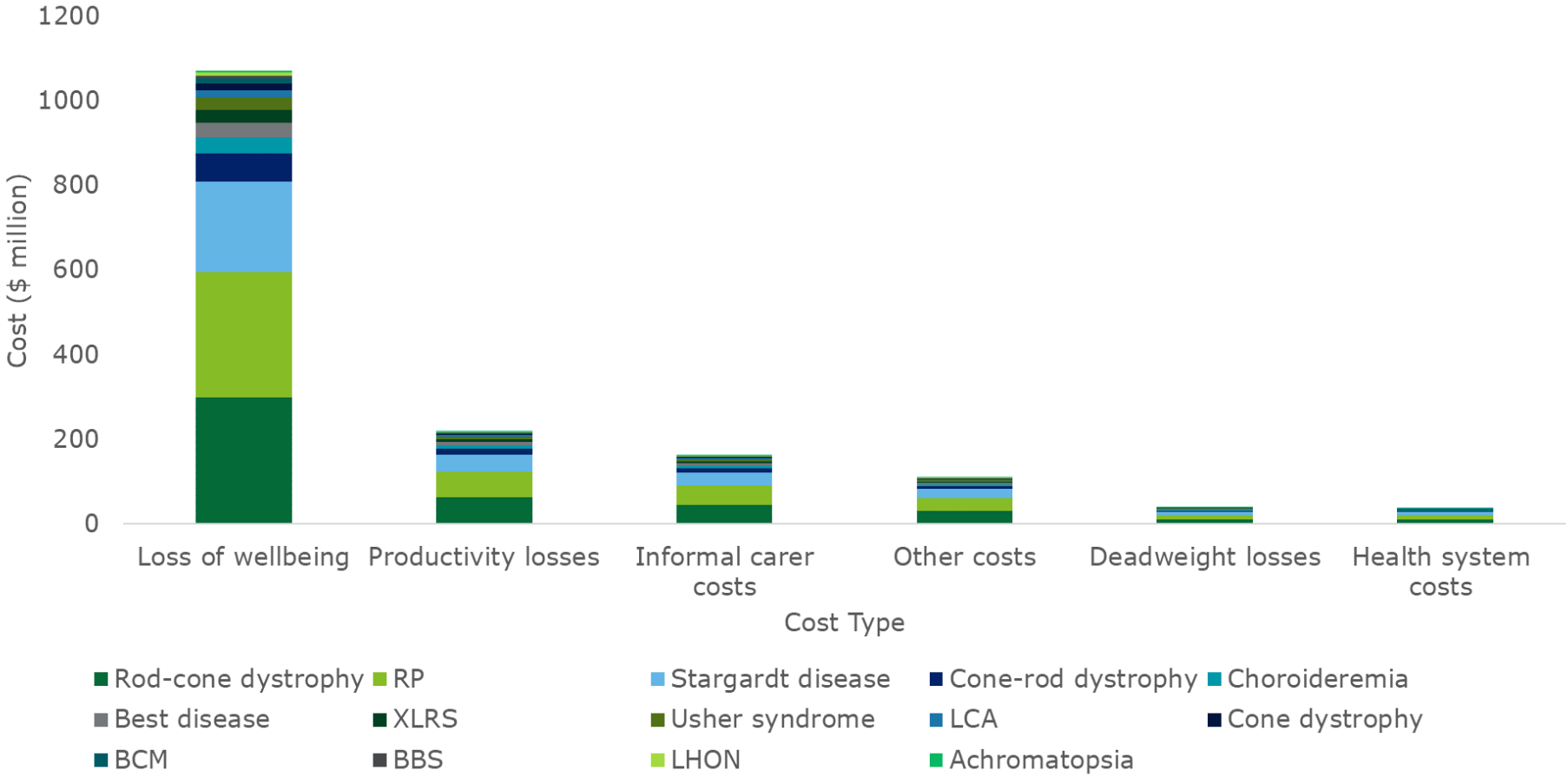
Source: Deloitte Access Economics analysis.

Chart 10.4 Total costs of IRDs ($ million, 2019) in Canada by cost type (upper bound scenario)



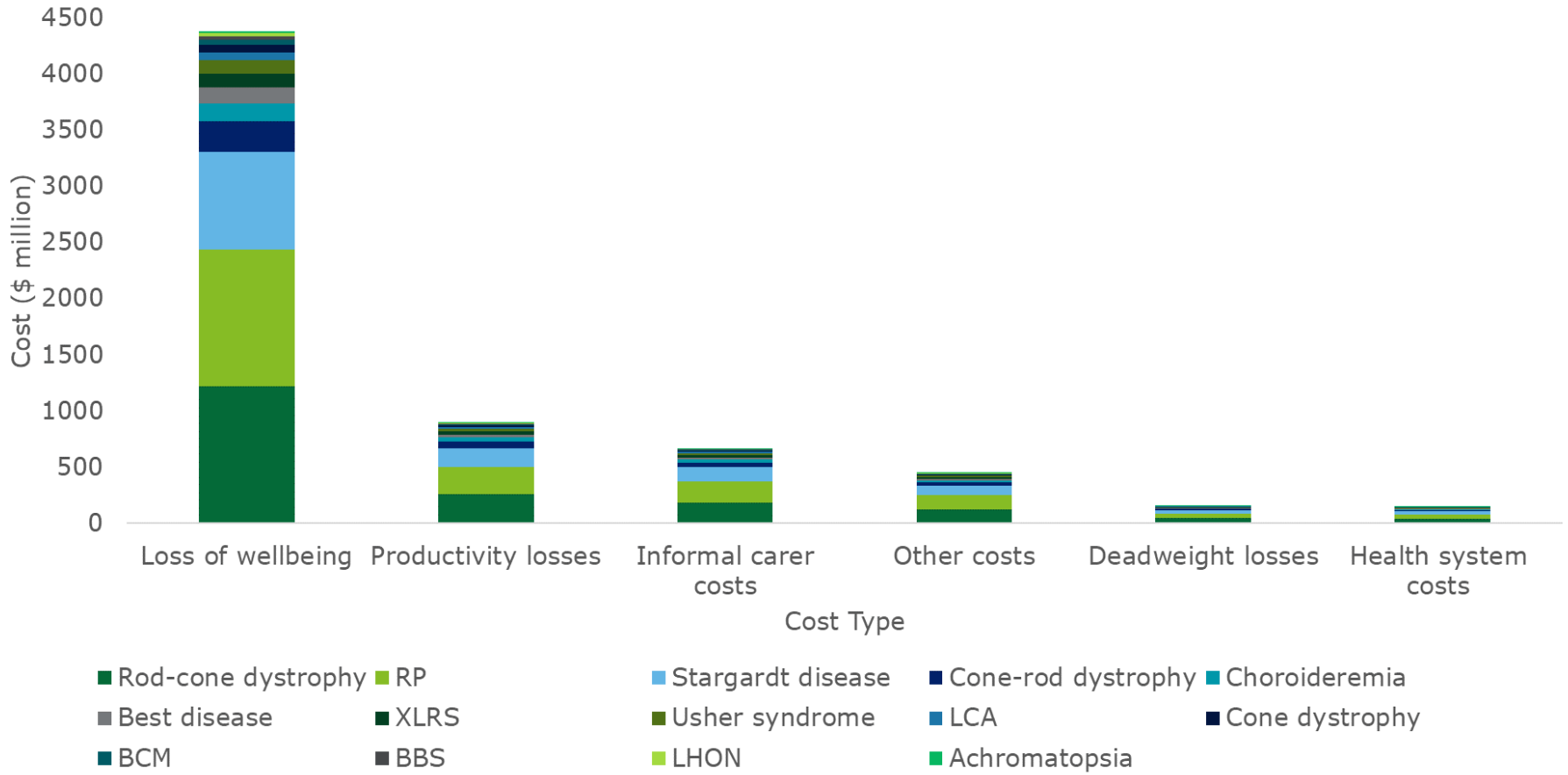
Source: Deloitte Access Economics analysis.

Chart 10.5 Cost type ($ million, 2019) in Canada by condition (lower bound scenario)



Source: Deloitte Access Economics analysis.

Chart 10.6 Cost type ($ million, 2019) in Canada by condition (upper bound scenario)



Source: Deloitte Access Economics analysis.

Table 10.2 Cost type ($ million, 2019) in Canada by condition (lower bound scenario)

IRDs	Loss of wellbeing	Productivity losses	Informal carer costs	Other costs	Deadweight losses	Health system costs
Rod-cone dystrophy	298.8	63.3	45.2	30.5	10.8	10.5
RP	296.8	59.4	44.9	30.3	10.6	10.5
Stargardt disease	212.9	40.7	32.2	21.7	7.5	7.5
Cone-rod dystrophy	66.6	14.1	10.1	6.8	2.4	2.3
Choroideremia	38.3	8.7	5.8	3.9	1.4	1.3
Best disease	35.5	6.9	5.4	3.6	1.3	1.3
XLRS	29.5	7.0	4.5	3.0	1.1	1.0
Usher syndrome	29.9	5.7	4.5	3.0	1.1	1.1
LCA	16.9	3.3	2.5	1.7	0.6	0.6
Cone dystrophy	15.9	3.4	2.4	1.6	0.6	0.6

BCM	11.1	2.6	1.7	1.1	0.4	0.4
BBS	7.5	1.4	1.1	0.8	0.3	0.3
LHON	6.7	1.5	1.0	0.7	0.2	0.2
Achromatopsia	5.1	1.0	0.8	0.5	0.2	0.2
Total	**1,071.4**	**219.0**	**162.0**	**109.4**	**38.3**	**37.8**

Source: Deloitte Access Economics analysis.
Note: Components may not sum to totals due to rounding.

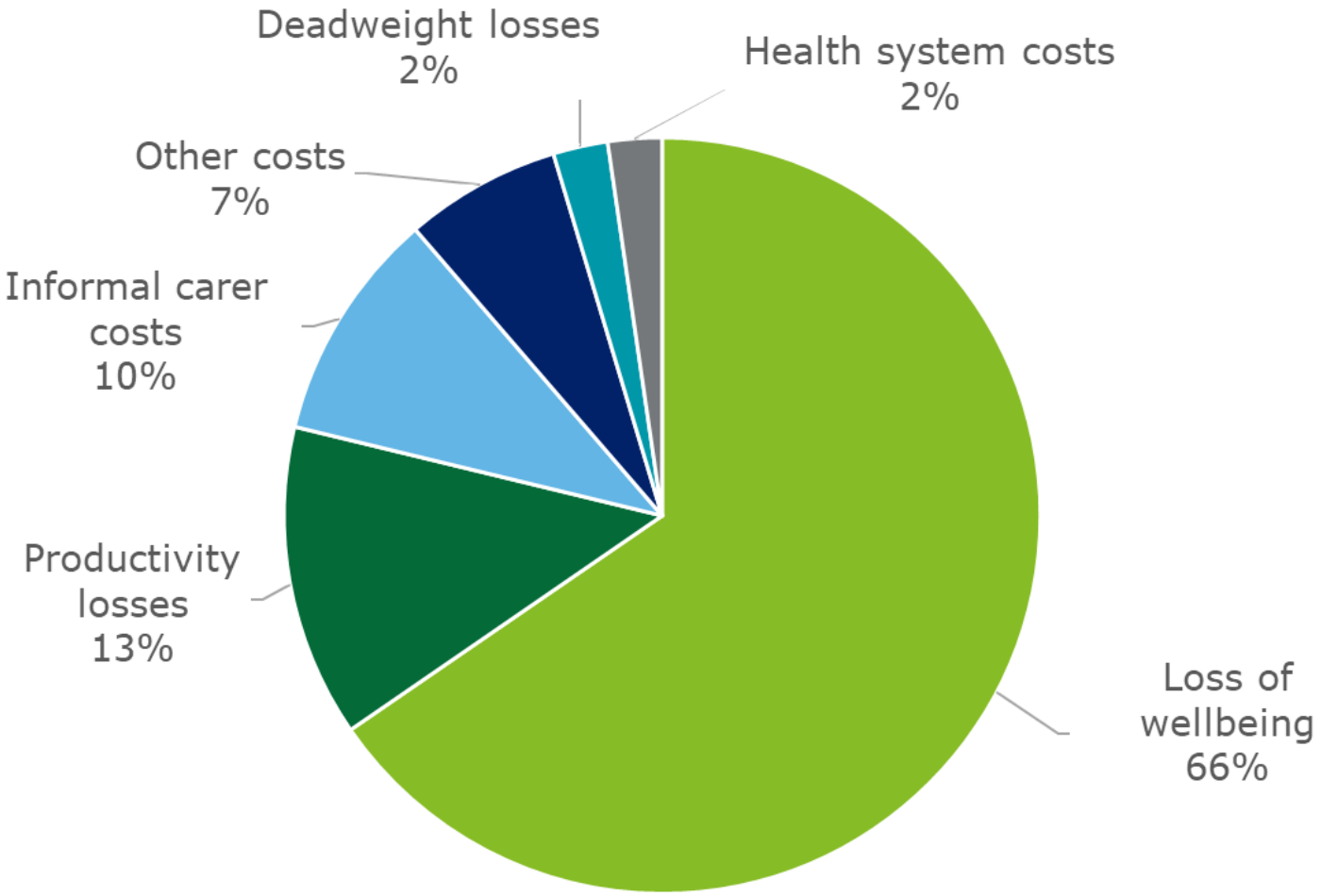
Table 10.3 Cost type ($ million, 2019) in Canada by condition (upper bound scenario)

IRDs	Loss of wellbeing	Productivity losses	Informal carer costs	Other costs	Deadweight losses	Health system costs
Rod-cone dystrophy	1,222.0	258.9	184.8	124.8	43.7	40.2
RP	1,214.0	242.9	183.6	124.0	43.0	40.0
Stargardt disease	870.9	166.5	131.7	88.9	30.6	28.7
Cone-rod dystrophy	272.3	57.7	41.2	27.8	9.7	9.0
Choroideremia	156.5	35.5	23.7	16.0	5.7	5.2
Best disease	145.3	28.3	22.0	14.8	5.1	4.8
XLRS	120.7	28.6	18.3	12.3	4.4	4.0
Usher syndrome	122.1	23.2	18.5	12.5	4.3	4.0
LCA	68.9	13.6	10.4	7.0	2.4	2.3
Cone dystrophy	64.9	13.8	9.8	6.6	2.3	2.1

BCM	45.5	10.8	6.9	4.6	1.7	1.5
BBS	30.5	5.6	4.6	3.1	1.1	1.0
LHON	27.4	6.0	4.1	2.8	1.0	0.9
Achromatopsia	21.1	4.1	3.2	2.1	0.7	0.7
Total	**4,381.9**	**895.5**	**662.7**	**447.4**	**155.7**	**144.3**

Source: Deloitte Access Economics analysis.
Note: Components may not sum to totals due to rounding.

Chart 10.7 Proportion (%) of total costs of IRDs in Canada by cost type (2019) (lower and upper bound scenario)



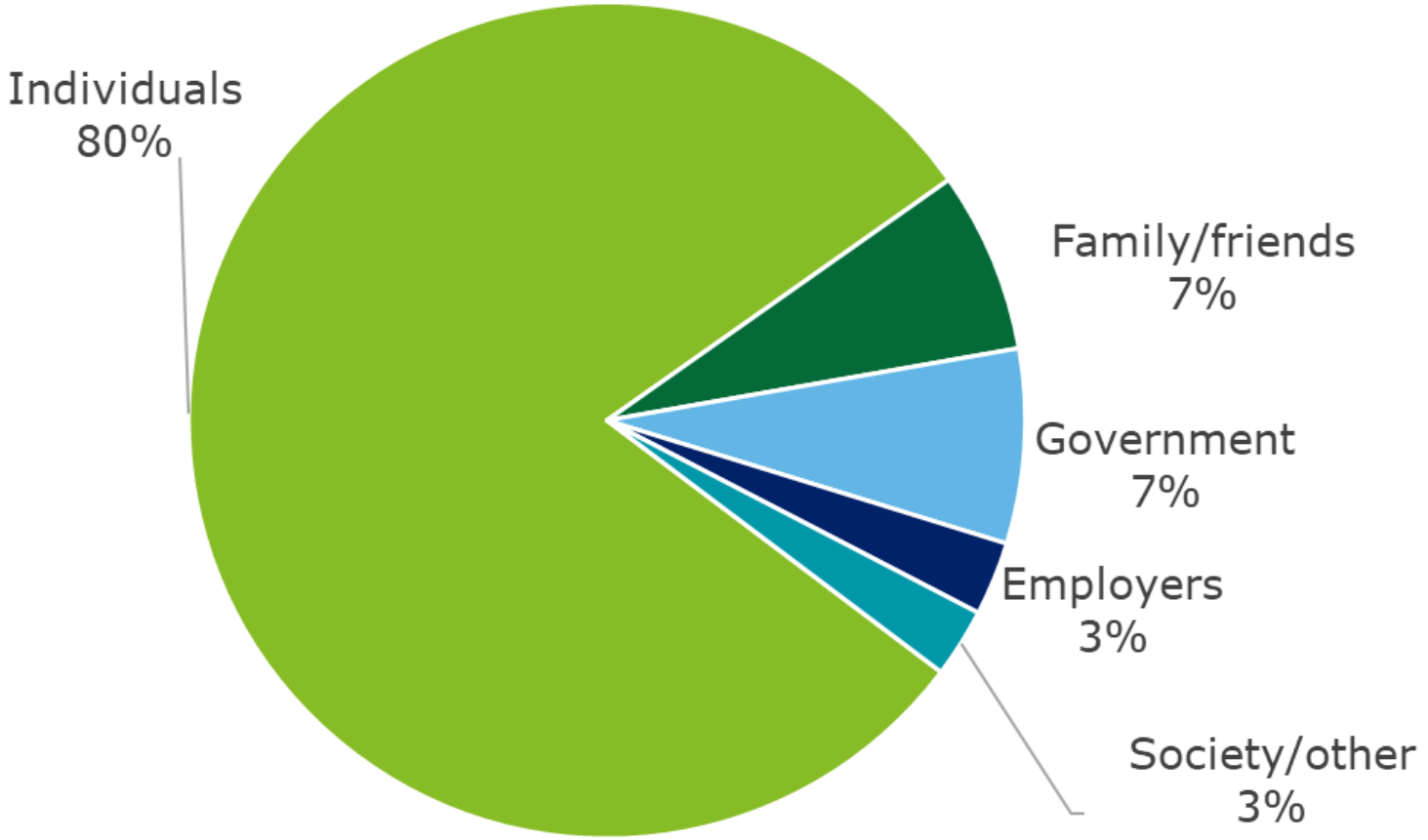
Source: Deloitte Access Economics analysis.

Table 10.4 Total costs of IRDs ($ million, 2019) in Canada by cost type (lower and upper bound scenario)

Cost type	Total cost ($) Lower bound	Total cost ($) Upper bound	Percentage of total cost (%)
Loss of wellbeing	1,071.4	4,381.9	65.4
Productivity losses	219.0	895.5	13.4
Informal carer costs	162.0	662.7	9.9
Other costs	109.4	447.4	6.7
Deadweight losses	38.3	155.7	2.3
Health system costs	37.8	144.3	2.3
Total costs	**1,877.53**	**6,687.5**	**100**

Source: Deloitte Access Economics analysis.
Note: Components may not sum to totals due to rounding.

Chart 10.8 Proportion (%) of total costs of IRDs in Canada by payer (2019) (lower and upper bound scenario)



Source: Deloitte Access Economics analysis.

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Appendix A Alternative approaches for estimating the prevalence of IRDs in Canada

A.1. Approach 3

Green et al (2020) estimates the prevalence of Stargardt disease based on a Canadian cohort study in Newfoundland, Canada. Provincial records of the Canadian National Institute for the Blind (between 1978 and 2018) were used to estimate the prevalence of Stargardt disease. To estimate a national Canadian population of Stargardt disease, the estimates published by Green et al (2020) were extrapolated by adjusting for geographic-related differences in prevalence rates using a ratio of seeing disability in the province relative to the prevalence of seeing disability in the total Canadian population using data from Statistics Canada (2019).

The Stargardt disease point prevalence estimated from Green et al (2020) was then used to derive the prevalence of each of the remaining IRDs relative to Stargardt disease. Where available, preference was given to eyeGENE since it captures 10 of the 14 included IRDs within a US and Canadian cohort of 4,635 participants. As such, eyeGENE was used to derive the relative prevalence rates of achromatopsia, Best disease, choroideremia, cone-rod dystrophy, LHON, RP, Usher syndrome, and XLRS.

The relative prevalence of BBS, cone dystrophy, LCA, and rod-cone dystrophy was informed by a genetic sequencing study of individuals with IRDs in France (Bocquet et al, 2013). In this study, 1,957 patients were recruited over a 21-year period in a specialised outpatient clinic for rare genetic sensory diseases.

In the absence of other available peer-reviewed literature and published data, the relative prevalence of BCM was derived using data from Orphanet (2013). Orphanet is an online inventory and classification of rare diseases with data contributed from 41 countries.

A.2. Approach 4

The Fighting Blindness Canada site refers that the prevalence of RP in the Canadian population is between 1/3,500 and 1/4,000. The original reference for the RP prevalence is a study conducted in Denmark by Haim et al (2002). Haim et al (2002) has been superseded by Bertelsen et al (2014) (option 1) as both studies use the same database to determine prevalence of IRDs in the Danish population. The midpoint of the prevalence estimate reported on the Fighting Blindness Canada site, 1/3,750, was used as the estimated prevalence point of RP in the Canadian population.

The RP point prevalence estimated from Fighting Blindness Canada site was then used to derive the prevalence of each of the remaining IRDs relative to RP. Where available, preference was given to eyeGENE since it captures 10 of the 14 included IRDs within a US and Canadian cohort of 4,635 participants. As such, eyeGENE was used to derive the relative prevalence rates of achromatopsia, Best disease, choroideremia, cone-rod dystrophy, LHON, Stargardt disease, Usher syndrome, and XLRS.

The relative prevalence of BBS, cone dystrophy, LCA, and rod-cone dystrophy was informed by a genetic sequencing study of individuals with IRDs in France (Bocquet et al, 2013). In this study, 1,957 patients were recruited over a 21-year period in a specialised outpatient clinic for rare genetic sensory diseases.

In the absence of other available peer-reviewed literature and published data, the relative prevalence of BCM was derived using data from Orphanet (2013). Orphanet is an online inventory and classification of rare diseases with data contributed from 41 countries.

Table A.1 Prevalence of IRDs in Canada (2019) by condition – approach 3 and 4

IRD	Approach 3 Rate (%)	Approach 3 Cases (n)	Approach 4 Rate (%)	Approach 4 Cases (n)
Rod-cone dystrophy	0.0630	23,678	0.0268	10,089
Retinitis pigmentosa (RP)	0.0112	4,224	0.0084	3,139
Autosomal dominant RP	0.0035	1,323	0.0123	4,626
Autosomal recessive RP	0.0052	1,950	0.0060	2,258
X-linked RP	0.0025	952	0.0076	2,864
Stargardt disease	0.0081	3,030	0.0191	7,190
Cone-rod dystrophy	0.0036	1,335	0.0060	2,248
Choroideremia	0.0033	1,258	0.0034	1,292
Best disease	0.0025	947	0.0032	1,199
Usher syndrome	0.0016	591	0.0027	1,008
X-linked retinoschisis (XLRS)	0.0014	545	0.0027	997

Leber congenital amaurosis (LCA)	0.0013	505	0.0015	569
Cone dystrophy	0.0011	425	0.0014	536
Blue cone monochromacy (BCM)	0.0011	420	0.0010	375
Bardet-Biedl Syndrome (BBS)	0.0008	303	0.0007	252
Leber’s hereditary optic neuropathy (LHON)	0.0003	95	0.0006	226
Achromatopsia	0.0002	73	0.0005	174
Total	**0.0996**	**37,431**	**0.0963**	**36,180**

Source: Deloitte Access Economics analysis. using Green et al (2020), Fighting Blindness Canada.

Appendix B Canadian prevalence estimates identified and considered but not included

Table B.1 Canadian prevalence estimates identified and considered but not included

Source	Description	Reason(s) for exclusion
Dharmaraj et al (2009)	This study examined patients with LCA in order to assess the relative burden of the three known genes involved in LCA, guanylyl cyclase 2D (GUCY2D), Retinoid Isomerohydrolase RPE65 (RPE65) and Cone-Rod Homeobox (CRX). Variant analysis and detailed clinical examinations were performed in patients diagnosed with LCA at one hospital in the US and one hospital in Canada. 100 patients were included in the final analysis.	Does not provide a Canadian-specific prevalence estimate; findings provide proportion of patients with a specific gene variant.

Source	Description	Reason(s) for exclusion
Hammed et al (2001)	A systematic search of an ocular database in the Polaris literature search engine and an MSN Web-based search engine was used to find information across three areas: health administrative databases, health surveys and registries. Information on blindness and visual impairment was obtained through online platforms (e.g. website or published literature), by directly contacting the institution (e.g. telephone or email).	Age of study; superseded by the Statistics Canada survey data included in option 2 that includes province and territory-specific prevalence estimates for visual impairment.
Koenekoop et al (2004)	A review article summarising the recent advances in the understanding of Leber congenital amaurosis.	Does not report a Canadian-specific prevalence estimates includes only a worldwide prevalence of Leber congenital amaurosis referencing a 1957 publication.

Source	Description	Reason(s) for exclusion
Lotery et al (2000)	This study was undertaken to determine the frequency of variants in the GUCYD2D, RPE6 and CRX genes in patients affected by LCA. Single-strand conformation polymorphism analysis and DNA sequencing was used screen patients' genes. 100 patients from US, 39 from India, 28 from Canada, 3 from Israel, 2 from Switzerland, 1 from Italy, 1 from Jordan, 1 from Pakistan, and 1 from Honduras were included in the analysis.	Does not report a Canadian-specific prevalence estimate; sample size includes patients from a range of countries which are likely not representative of the Canadian-population.
Maberley et al (2006)	A retrospective cohort study to ascertain the prevalence and primary causes of visual impairment in a representative Canadian population Patients who attended ophthalmologists' offices between 1996 and 2001 at Prince George, British Columba, Canada were eligible for inclusion. Review of ophthalmic charts in the community was performed in August 2003/ 2466 charts were evaluated, of which 962 participants were included in the analysis.	Age of study; superseded by the Statistics Canada survey data included in option 2 that includes province and territory-specific prevalence estimates for visual impairment.

Source	Description	Reason(s) for exclusion
Morimura et al (1998)	This study explored the role that defects in gene RPE65 might play in the aetiology of retinal degenerations. A large cohort of unrelated patients with non-syndromic RP and non-syndromic LCA were screened for variants. Patients in this study, who resided in the US or Canada, were examined and diagnosed by one or more of the authors. 15 patients with isolated RP and 45 patients with LCA were included in the analysis.	Age of the study; does not report a Canadian-specific prevalence estimate.
Zernant et al (2005)	An LCA genotyping microarray was used to screen patients with genetically heterogeneous condition of RP. The LCA array allowed for the simultaneous detection of all known disease-associated alleles in patients with early onset RP. LCA patients' cohorts were derived from two locations in North America and two centres in Europe. 298 unrelated participants were included in the analysis	Does not report a Canadian-specific prevalence estimate; prevalence reported are for detecting an occurrence of alleles.

Appendix C Additional information on survey results

This appendix details the survey questionnaire distributed to persons living with an IRD and/or the parents of children (under 18) living with an IRD.

The questionnaire comprised 12 modules:

1. About you
2. Your current condition
3. Work
4. Education
5. Management of your condition – health care providers
6. Management of your condition – medications and supplements
7. Management of your condition – vision aids and modifications and travel
8. Formal care
9. Informal care
10. Government support
11. Other impacts of your condition
12. Impact of your child's IRD on your mental wellbeing – from your own perspective as a parent/guardian.

Skip logic was incorporated into the programming of the survey to ensure only relevant questions were asked of participants, depending on the answers provided to previous questions.

The questions asked, answers available and results received for each question are outlined in this appendix. In instances where a low number of results were received, results are aggregated to protect participant privacy and confidentiality.

C.1. Introduction

Deloitte Access Economics (Australia) is assisting Retina International and its partner organisations to better understand the real impact of inherited retinal dystrophies (IRDs). This survey aims to evaluate the financial and social impact of IRDs.

The findings will lead to two reports, one each for Canada and the USA. The reports will be published on the websites of Retina International, Fighting Blindness Canada, and the Foundation Fighting Blindness.

Information collected through this survey will inform knowledge and understanding of:

- The impact on the wellbeing of individuals affected and their families; and
- The economic cost of irds to health and social care, education and employment and carers.

C.1.1. Purpose of the study

You are invited to participate in an online survey, which is being conducted in order to better understand IRD impacts. The survey is open to people who have or are the parent/guardian of someone with any of the following conditions:

- Retinitis pigmentosa (RP)
- Choroideremia
- Stargardt disease
- Usher syndrome
- Rod-cone dystrophy
- Bardet-Biedl Syndrome (BBS)
- Leber congenital amaurosis (LCA)
- X-linked retinoschisis (XLRS)
- Cone-rod dystrophy
- Best disease
- Cone dystrophy
- Achromatopsia
- Leber's hereditary optic neuropathy (LHON)
- Blue Cone Monochromacy (BCM)

The survey will collect personal information about you including demographic information (such as your age and gender at birth) and details about your work and education. It will also collect personal information about you that may be considered “sensitive” personal information, including your ethnicity, details about your IRD, how you manage your IRD, your use of healthcare services, and your wellbeing.

Your personal information will be used to estimate the economic costs of IRDs and to describe the characteristics of groups of people (not individuals) who have or are the parent/guardian of someone with a diagnosed IRD. Your data will be combined with all survey responses

received and any personal information will be removed prior to publication.

C.1.2. Confidentiality

All personal information collected as part of this survey will be kept strictly confidential. Your identity will not be revealed and your confidentiality will be protected in any reviews and reports of this study that may be published.

While we use a third party survey platform provider to assist us in administering this survey, this provider is bound by strict confidentiality and privacy obligations. This means that your information will only be accessed by authorised personnel where required for the purpose of administering the survey and conducting the analysis described above.

C.1.3. The Legal basis for processing personal information

Before you complete this survey you will be asked to provide your consent to the collection and processing of your personal information and special category information for the purpose described above. Your consent will then form the legal basis upon which we are able to process your information.

C.1.4. Voluntary participation/right to refuse or withdraw

Your participation in this survey is voluntary; and you can withdraw at any time if you change your mind. If you decide to withdraw your consent by exiting the survey mid-way through, your responses will not be recorded.

You have various rights in relation to your personal information, including the right to seek access to, or to correct, your information (for more information, please see our Privacy Statement).

If you are located in Europe, you also have the right to:

- Ask that we delete personal information that we hold about you, or restrict the way in which we use your personal information;
- Withdraw consent to our processing of your personal information; and
- Obtain and/or move your personal information to another service provider.

However, we may still be required to retain a copy of certain information where it is necessary to comply with applicable laws or professional standards.

C.1.5. Contact details should you experience any unsettled feelings during this survey

Your participation in this survey is voluntary and you can withdraw at any time if you change your mind. If you decide to withdraw your consent by exiting the survey mid-way through, your responses will not be recorded.

You have various rights in relation to your personal information, including the right to seek access to, or to correct, your information (for more detail, please see our Privacy Statement).

Contact details should you experience any unsettled feelings during this survey.

If you have any concerns about your IRD care and management, please contact your general practitioner/preferred health provider. If you are in Canada, you can access health information services through Fighting Blindness Canada by emailing healthinfo@fightingblindness.ca or calling 1-888-626-2995. If you are in the USA, you can access health information by visiting https://www.usa.gov/health-resources#item-36417 or calling 1-844-872-4681.

For any questions about the objectives of the survey, how your data will be used or to exercise any of your rights, please contact the Deloitte Privacy Officer at privacy@deloitte.com.au.

For any other questions please contact: irds@deloitte.com.au

C.2. Eligibility and Consent

Who are you completing this survey for?

- Myself as a person with an IRD
- As the parent/guardian of a child (aged up to 18 years old) with IRD
- On behalf of an adult with IRD as their carer

If you would like to proceed with participating in this survey, please provide consent.

By providing consent you are telling us that you:

- Understand what you have read.
- You are 18 years or over.
- Consent to take part in the research project.
- Consent to the processing of your personal information for the purpose described in the introduction to this survey

- Consent to the processing of your health and other types of special category information for the purpose described in the introduction to this survey.

Please confirm you have read and understand the information provided. By selecting 'yes' you agree to your involvement in the survey to understand the real impact of IRDs.

- Yes, I voluntarily consent
- No, I do not consent

C.3. Preliminary questions

Which of the following best describes how you will respond to this survey?

- As myself a person affected with an IRD
- As the parent/guardian of a child (aged up to 18 years old) with an IRD

Table C.1 Which of the following best describes how you will respond to this survey? (n=151) (Canada specific)

Participant type	Count (n)	Percentage (%)
As myself a person affected with an IRD	138	91.4
As the parent/guardian of a child (aged up to 18 years old) with an IRD	13	8.6
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

C.4. Module 1: About you

Which IRD are you diagnosed with?

- Retinitis pigmentosa (RP)
- Choroideremia
- Stargardt disease
- Usher syndrome
- Rod-cone dystrophy
- Bardet-Biedl Syndrome
- Leber congenital amaurosis (LCA)
- X-linked retinoschisis
- Cone-rod dystrophy
- Best disease
- Cone dystrophy
- Achromatopsia
- Leber's hereditary optic neuropathy (LHON)
- Blue Cone Monochromacy

Table C.2 Which IRD are you diagnosed with? (n=151) (Canada specific)

Participant condition	Count (n)	Percentage (%)
Retinitis pigmentosa (RP)	104	68.9
Other	22	14.5
Stargardt disease	11	7.3
Usher syndrome	9	6.0
Cone-rod dystrophy	5	3.3
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Did you receive a genetic diagnosis for your IRD (for example, identification of a disease causing gene)?

- Yes
- No

Table C.3 Did you receive a genetic diagnosis for your IRD (for example, identification of a disease causing gene)? (n=151) (Canada specific)

Genetic diagnosis	Count (n)	Percentage (%)
Yes	93	61.6
No	58	38.4
Total	**151**	**100.0**

Source: Deloitte Access Economics analysis.

How long ago did you receive a genetic diagnosis for your IRD (for example, identification of a disease causing gene)?

- In the last 12 months
- Between 1 to 2 years ago
- Less than 5 years ago
- Less than 10 years ago
- Less than 15 years ago
- Less than 20 years ago
- More than 20 years ago
- I do not remember when I received my genetic diagnosis

Table C.4 How long ago did you receive a genetic diagnosis for your IRD (for example, identification of a disease causing gene)? (n=93) (Canada specific)

Genetic diagnosis	Count (n)	Percentage (%)
In the last 12 months	8	8.6
Between 1 to 2 years ago	16	17.2
Less than 5 years ago	17	18.3
Less than 10 years ago	20	21.5
Less than 20 years ago	14	15.1
More than 20 years ago/I do not remember when I received my generic diagnosis	18	19.4
Total	**93**	**100**

Source: Deloitte Access Economics analysis.

What form of retinitis pigmentosa (RP) are you diagnosed with?

- Autosomal dominant RP
- Autosomal recessive RP
- X-linked recessive RP
- Unsure

Table C.5 What form of retinitis pigmentosa (RP) are you diagnosed with? (n=104) (Canada specific)

Participant condition	Count (n)	Percentage (%)
Unsure	62	59.6
Autosomal recessive RP	17	16.3
Autosomal dominant RP	13	12.5
X-linked recessive RP	12	11.5
Total	**104**	**100**

Source: Deloitte Access Economics analysis.

What was your biological sex at birth?

- Female
- Male
- Prefer not to say

Table C.6 What was your biological sex at birth (n=151) (Canada specific)

Biological sex	Count (n)	Percentage (%)
Female	80	53.0
Male	71	47.0
Not stated	0	0.0
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Which Province do you currently reside in?

- British Columbia
- Alberta
- Quebec
- Ontario
- Nova Scotia
- Manitoba
- Newfoundland and Labrador
- Saskatchewan

Table C.7 Which Province do you currently reside in (n=151) (Canada specific)

Age	Count (n)	Percentage (%)
Ontario	75	49.7
British Columbia	26	17.2
Alberta	22	14.6
Quebec	11	7.3
Manitoba / Newfoundland and Labrador	7	4.6
Nova Scotia	5	3.3
Saskatchewan	5	3.3
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Which of the following options best describes the geographic area you live in?

- Large urban population centre
- Medium population centre
- Small population centre
- Rural area
- Urbanized Area (UA) of 50,000 or more people
- Urban Cluster (UC) of at least 2,500 and less than 50,000 people
- Rural Area encompassing all population, housing, and territory not included within an Urban Area

Table C.8 Which of the following options best describes the geographic area you live in? (n=151) (Canada specific)

Age	Count (n)	Percentage (%)
Large urban population centre	76	50.3
Medium population centre	47	31.1
Small population centre	19	12.6
Rural area	9	6.0
Urbanized Area (UA) of 50,000 or more people	0	0.0
Urban Cluster (UC) of at least 2,500 and less than 50,000 people	0	0.0
Rural Area encompassing all population, housing, and territory not included within an Urban Area	0	0.0
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

What is your age (all ages)?

- 5 to 9
- 10 to 14
- 15 to 17
- 18 to 24
- 25 to 34
- 35 to 44
- 45 to 54
- 55 to 64
- 65 to 74
- 75 or older

Table C.9 What is your age (all ages)? (n=151) (Canada specific)

Age	Count (n)	Percentage (%)
Under 25	16	10.6
25-34	21	13.9
35-44	25	16.6
45-54	21	13.9
55-64	32	21.2
65-74	28	18.5
75+	8	5.3
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

What is your ethnicity?

- White
- Asian including Indian/Chinese/Vietnamese/Filipino
- Hispanic or Latino or Spanish Origin including but not limited to Cuban, Mexican, Puerto Rican, South or Central American, or other Spanish culture or origin
- French Canadian
- First Nations Canadian/Native Alaskan/American Indian
- Mixed/Multiple ethnic groups
- Middle Eastern including but not limited to Lebanese/Iranian
- Black including but not limited to African or Caribbean
- Other ethnic groups

Table C.10 What is your ethnicity? (n=151) (Canada specific)

Participant ethnicity	Count (n)	Percentage (%)
White	111	73.5
Other ethnic groups	21	13.9
French Canadian	11	7.3
Asian including Indian/Chinese/Vietnamese/Filipino	8	5.3
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

C.5. Module 2: Your current condition

Which of these best describes the vision (if any) you have remaining?

- No vision
- Light perception only (or shadows only)
- Some useful central vision
- Good central vision
- Some useful peripheral vision
- Good peripheral vision
- I still have good overall vision
- Other – please specify

Table C.11 Which of these best describes the vision (if any) you have remaining? (n=151) (Canada specific)

Participant condition	Count (n)	Percentage (%)
I have problems with light and glare sensitivity	87	57.6
I have night blindness	87	57.6
I have tunnel vision	57	37.7
Some useful central vision	48	31.8
Moderate peripheral vision loss	45	29.8
Good central vision	39	25.8
Other / No vision	26	17.2
Light perception only (or shadows only)	18	11.9
Good peripheral vision / I still have good overall vision	14	9.9
Total	**151**	**100.0**

Source: Deloitte Access Economics analysis. Note "Other" responses were reclassified into the relevant categories. This question allowed for the selection of multiple answers; hence, the percentages do not sum to 100.

Are you registered as legally blind with the Canada National Institute for the Blind (CNIB) Foundation?

- Yes, I am registered as severely sight impaired/legally blind (defined as best corrected visual acuity of less than 20/200, or visual field of 20 degrees or narrower)
- No, my degree of sight loss does not meet the criteria
- No, I have chosen not to be registered

Table C.12 Are you registered as legally blind with the National Council for the Blind (NCBI)? (n=151) (Canada specific)

Participant condition	Count (n)	Percentage (%)
Yes, I am registered as severely sight impaired/legally blind (defined as best corrected visual acuity of less than 20/200, or visual field of 20 degrees or narrower)	108	71.5
No, my degree of sight loss does not meet the criteria	26	17.2
No, I have chosen not to be registered	17	11.3
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Has a doctor or specialist said that you have hearing loss?

- Yes
- No

Table C.13 Has a doctor or specialist said that you have hearing loss? (n=9) (Canada specific)

Diagnosed	Hearing loss Count (n)	Percentage (%)
Yes	9	100.0
No	0	0.0
Total	**9**	**100.0**

Source: Deloitte Access Economics analysis.

Do you currently use a hearing aid or other device because of your hearing loss I do not have any hearing loss?

- Yes, I use a hearing aid
- No, I do not currently use a hearing device
- Yes, I use a cochlear implant
- Other (Please describe)

Table C.14 Do you currently use a hearing aid or other device because of your hearing loss I do not have any hearing loss? (n=9) (Canada specific)

Self-reported hearing	Count (n)	Percentage (%)
Yes, I use a hearing aid / Other (please describe)	9	100
No, I do not currently use a hearing device	0	0.0
Yes, I use a cochlear implant	0	0.0
Total	**9**	**100**

Source: Deloitte Access Economics analysis.

C.6. Module 3: Work

Prior to any COVID-19 interruptions, which of the following best described your state of work and study?

- Full time work
- Part time work
- Casual work (Not working on a regular basis)
- Underemployed (e.g. working part time but desiring full time work, or working in a role that does not use your skills or qualifications)
- Student only
- Retired (not working)
- Not employed and not studying, and looking for work
- Not employed, and not looking for work
- Which of the following best describes your current state of work and study?

Table C.15 Prior to any COVID-19 interruptions, which of the following best described your state of work and study? (n=138) (Canada specific)

Employment/study status	Count (n)	Percentage (%)
Full time work	54	39.1
Retired (not working)	39	28.3
Not employed, and not looking for work / Student only	14	10.1
Part time work	13	9.4
Not employed and not studying, and looking for work	12	8.7
Underemployed (e.g. working part time but desiring full time work, or working in a role that does not use your skills or qualifications) / Casual work (Not working on a regular basis)	6	4.4
Total	**138**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, what were your average weekly earnings after tax?

- $0 to $250
- $250 to $500
- $500 to $1,000
- $1,000 to $1,500
- $1,500 or more
- Prefer not to say

Table C.16 Prior to any COVID-19 interruptions, what were your average weekly earnings after tax? (n=73) (Canada specific)

Average weekly earnings (post tax, $)	Count (n)	Percentage (%)
$0 to $250	8	11.0
$250 to $500	9	12.3
$500 to $1,000	24	32.9
$1,000 to $1,500	13	17.8
$1,500 or more	9	12.3
Prefer not to say	10	13.7
Total	**73**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, how many hours did your employer expect you to work in a typical 7-day week?

If your hours varied, estimate the average. If you were self-employed, estimate the number of hours you considered a full work week. If you had more than one job, combine total number of hours for all jobs.

- 0 hours to 10 hours
- 10 hours to 20 hours
- 20 hours to 30 hours
- 30 hours to 40 hours
- 40 hours or more

Table C.17 Prior to any COVID-19 interruptions, how many hours did your employer expect you to work in a typical 7-day week? (n=71) (Canada specific)

Expected work hours	Average	Standard Deviation
0 hours to 10 hours	5	7.0
10 hours to 20 hours	6	8.5
20 hours to 30 hours	8	11.3
30 hours to 40 hours	36	50.7
40 hours or more	16	22.5
Total	**71**	**100**

Source: Deloitte Access Economics analysis.

Now please think of your work experiences prior to any COVID-19 interruptions. In a typical 4-week period prior to any COVID-19 interruptions, how many full and partial days of work (rounded to the nearest whole number) did you miss because of your IRD? Please include only days missed for your own health, not someone else's health.

- 0 days
- 1 day
- 2 days
- 3 days
- 4 days
- 5 days
- 6 to 10 days
- 11 to 15 days
- 16 to 20 days
- Other

Table C.18 Prior to any COVID-19 interruptions, how many full and partial days of work did you miss because of your IRD? (n=69) (Canada specific)

Work days missed due to IRDs	Average	Standard Deviation
0 days	49	71.0
2 or more days	14	20.2
1 day	6	8.7
Total	**69**	**100**

Source: Deloitte Access Economics analysis.

About how many hours altogether did you work in a typical 4-week period prior to any COVID-19 interruptions? If you did not work at all, select "0 hours".

- 0 hours
- Between 1 and 40 hours
- Between 40 and 80 hours
- Between 80 and 120 hours
- Between 120 and 160 hours
- 160 hours or more

Table C.190 Prior to any COVID-19 interruptions, how many full and partial days of work did you miss because of your IRD? (n=73) (Canada specific)

Work days missed due to IRDs	Average	Standard Deviation
Between 0 and 40 hours	16	21.9
Between 40 and 80 hours	14	19.2
Between 80 and 120 hours	5	6.8
Between 120 and 160 hours	25	34.2
160 hours or more	13	17.8
Total	**73**	**100**

Source: Deloitte Access Economics analysis.

In a typical 7 day period prior to any COVID-19 interruptions, how much did your health problems affect your productivity while you were working?

(Help text: Think about days you were limited in the amount or kind of work you could do, days you accomplished less than you would like, or days you could not do your work as carefully as usual. If health problems affected your work only a little, choose a low number. Choose a high number if health problems affected your work a lot.)

- 0 (health problems had no effect on my work)
- 1
- 2
- 3
- 4
- 5
- 6
- 7
- 8
- 9
- 10 (health problems completely prevented me from working)

Table C.20 How would you rate the usual performance of most workers in a job similar to yours? (n=70) (Canada specific)

Perceived workplace performance rating	Count (n)	Percentage (%)
0	25	35.7
1	5	7.1
2	16	22.9
3 to 10	24	34.3
Total	**70**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, which of the following modes of transport did you most commonly use to get to work?

- Public transport
- Private transport
- Taxi
- Other (free text)

Table C.21 Prior to any COVID-19 interruptions, which of the following modes of transport did you most commonly use to get to work? (n=70) (Canada specific)

Public transport	Count (n)	Percentage (%)
Public transport	36	51.4
Private transport	21	30.0
Taxi or other (free text)	13	18.6
Total	**70**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, how much did you spend on an average week travelling to work?

- $0 to $10
- $10 to $20
- $20 to $30
- $30 to $40
- $40 or greater

Table C.22 How would you compare your overall job performance on the days you worked during the past 4 weeks with the performance of most other workers who have a similar type of job? (n=70) (Canada specific)

Travel cost (average amount spent per week)	Count (n)	Percentage (%)
$0 to $10	32	45.7
$10 to $20	6	8.6
$20 to $30	10	14.3
$30 to $40	8	11.4
$40 or greater	14	20.0
Total	**70**	**100**

Source: Deloitte Access Economics analysis.

C.7. Module 4: Education

Immediately prior to any COVID-19 interruptions, were you enrolled in an educational program or at an educational institution?

- Yes
- No

Table C.23 Are you currently studying or learning? (n=90) (Canada specific)

Study Status	Count (n)	Percentage (%)
No	69	76.7
Yes	21	23.3
Total	**90**	**100**

Source: Deloitte Access Economics analysis.

Immediately prior to any COVID-19 interruptions, where were you receiving your education?

- Higher education (university)
- Further education
- Secondary education/post primary education
- Primary education
- Pre-school/early childhood education
- Other education provider

Table C.24 Immediately prior to any COVID-19 interruptions, where were you receiving your education? (n=21) (Canada specific)

Study Status	Count (n)	Percentage (%)
Higher/tertiary education (university)	10	47.6
Secondary education/high-school education/other education provider	6	28.6
Pre-school/kindergarten/early childhood education/Primary/elementary education	5	23.8
Vocational/technical education	0	0.0
Total	**21**	**100**

Source: Deloitte Access Economics analysis.

About how many hours altogether did you spend attending class in your place of learning in a typical 4-week period prior to any COVID-19 interruptions?

- 0 hours
- 1 to 20 hours
- 20 to 30 hours
- 40 to 50 hours
- Greater than 60 hours

Table C.25 About how many hours altogether did you spend attending class in your place of learning in a typical 4-week period prior to any COVID-19 interruptions? (n=21) (Canada specific)

Study Status	Count (n)	Percentage (%)
0 to 20 hours	10	14.3
20 to greater than 60 hours	11	85.7
Total	**21**	**100**

Source: Deloitte Access Economics analysis.

How many hours did you spend on education-related activities (e.g. studying, completing homework and/or assignments, research etc.) outside of the classroom in a typical 7-day week prior to any COVID-19 interruptions? If it varies, estimate the average.

- 0 hours
- 1 to 20 hours
- 20 to 30 hours
- 40 to 50 hours
- Greater than 60 hours

Table C.26 How many hours did you spend on education-related activities (e.g. studying, completing homework and/or assignments, research etc.) outside of the classroom in a typical 7-day week prior to any COVID-19 interruptions? (n=21) (Canada specific)

Study Status	Count (n)	Percentage (%)
0 to 10 hours per week	13	61.9
10 to 40 hours per week	8	38.1
Total	**21**	**100**

Source: Deloitte Access Economics analysis.

In a typical 4-week period prior to any COVID-19 interruptions, how many full and partial days of class (round to the nearest whole day) did you miss because of your IRD? Please include only days missed for your own health, not someone else's health.

- 0 days
- 1 day
- 2 days
- 4 days
- 6 to 10 days
- 16 to 20 days
- Other

Table C.27 In a typical 4-week period prior to any COVID-19 interruptions, how many full and partial days of class (round to the nearest whole day) did you miss because of your IRD? Please include only days missed for your own health, not someone else's health. (n=21) (Canada specific)

Study Status	Count (n)	Percentage (%)
0 days	7	33.3
1 day / Other	14	66.7
Total	**21**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, did you receive additional support from a special needs teacher/assistant in your learning environment?

- Yes
- No

Table C.28 Prior to any COVID-19 interruptions, did you receive additional support from a special needs teacher/assistant in your learning environment? (n=21) (Canada specific)

Special needs teacher/assistant	Count (n)	Percentage (%)
No	13	61.9
Yes	8	38.1
Total	**21**	**100**

Source: Deloitte Access Economics analysis.

On average, prior to any COVID-19 interruptions, approximately how many hours per week did you receive additional support from a special needs assistant?

- 1 to 5 hours
- 5 to 10 hours
- 10 to 20 hours
- Greater than 20 hours

Table C.29 On average, prior to any COVID-19 interruptions, approximately how many hours per week did you receive additional support from a special needs assistant? (n=7) (Canada specific)

Study Status	Count (n)	Percentage (%)
1 to greater than 20 hours	7	100
Total	**7**	**100**

Source: Deloitte Access Economics analysis.

C.8. Module 5: Management of your condition – health care providers

In a typical year prior to any COVID-19 interruptions, did you see any health providers specifically for or relating to the management of your IRD?

- Yes
- No
- Unsure

Table C.30 Have you seen any health providers specifically for or relating to the management of your IRD in the last year? (n=151) (Canada specific)

Health system utilisation (total this year)	Count (n)	Percentage (%)
Yes	108	71.5
No / Unsure	43	28.5
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

When was the last time you saw a health provider specifically for or relating to the management of your IRD?

- 1 to 2 years
- 2 to 3 years
- 4 or more years
- Unsure

Table C.31 When was the last time you saw a health provider specifically for or relating to the management of your IRD? (n=41) (Canada specific)

Health system utilisation (total)	Count (n)	Percentage (%)
1 to 2 years	11	26.8
2 to 3 years	12	29.3
4 or more years	13	31.7
Unsure	5	12.2
Total	**41**	**100**

Source: Deloitte Access Economics analysis.

The following questions ask about which providers you have seen for management specifically or including for your IRD in a typical year prior to any COVID-19 interruptions. Please indicate how many times you accessed each service, and the total amount you spent for management of your IRD over a typical year prior to any COVID-19 interruptions.

Please enter a numerical answer rounded to the nearest whole number (i.e. no '$' symbol). For example, if you spent $20, please enter 20.

How many times did you access the service in the last year for your IRD? (If you did not access the service, please enter '0').

- General practitioner / Family physician
- Ophthalmologist
- Optometrist/optician
- Sight support volunteer
- Psychologist, psychiatrist or counsellor
- Genetic counsellor
- Occupational therapist
- Hospital emergency department
- Admitted to hospital as an inpatient
- Specialist outpatient clinic or unit
- Habilitation, Rehabilitation specialist or service, or low vision specialist
- My Retina Tracker Registry app counselling service

Table C.32 Which health providers have you seen for management specifically or including for your IRD in the last year? (n=151) (Canada specific)

Health System Utilisation (specific)	Count (n)
Admitted to hospital (inpatient)	5.5
Emergency department	1.4
Genetic counsellor	1.3
General practitioner	2.3
Occupational therapist	2.8
Ophthalmologist	1.5
Optometrist	1.7
Psychologist	6.7
Habilitation specialist	7.4
My Retina Tracker Registry app counselling service	NA
Sight support volunteer	12.9
Specialist outpatient clinic	4.6

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, how much money did your last visit to the health professional cost you out-of-pocket, after refunds, reimbursements and financial support?

- General practitioner / Family physician
- Ophthalmologist
- Optometrist/optician
- Sight support volunteer
- Psychologist, psychiatrist or counsellor
- Genetic counsellor
- Occupational therapist
- Hospital emergency department
- Admitted to hospital as an inpatient
- Specialist outpatient clinic or unit
- Habilitation, Rehabilitation specialist or service, or low vision specialist
- My Retina Tracker Registry app counselling service

Table C.33 Prior to any COVID-19 interruptions, how much money did your last visit to the health professional cost you out-of-pocket, after refunds, reimbursements and financial support? (n=19) (Canada specific)

Health System Utilisation (cost)	Average ($)
Sight support volunteer	1119
Specialist outpatient clinic	966
General practitioner	823
Optometrist	490
Ophthalmologist	438
Psychologist	409
Habilitation specialist	271
Occupational therapist	253
Genetic counsellor	74
Admitted to hospital (inpatient)	60
Emergency department	50
My Retina Tracker Registry app counselling service	0

Source: Deloitte Access Economics analysis.

In a typical year prior to any COVID-19 interruptions, did you receive genetic testing and counselling specifically for or including for your IRD?

- Diagnostic Genetic Test
- Genetic Counselling
- Both
- No
- Unsure

Table C.34 In a typical year prior to any COVID-19 interruptions, did you receive genetic testing and counselling specifically for or including for your IRD? (n=110) (Canada specific)

Genetic testing	Count (n)	Percentage (%)
Genetic Counselling	76	69.1
Diagnostic Genetic Test	12	10.9
Both / No / Unsure	22	20.0
Total	**110**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, approximately much money did you spend on travel (e.g. airfares, gas, bus fare, train fare, or taxi fare) to receive the results of your genetic test?

If you are unsure please answer not applicable.

- $0 to $20
- $20 to $50
- $50 to $100
- $100 to $200
- $200 to $500
- Greater than $500
- Not applicable

Table C.35 In a typical year prior to any COVID-19 interruptions, did you receive genetic testing and counselling specifically for or including for your IRD? (n=110) (Canada specific)

Genetic testing	Count (n)	Percentage (%)
$0 to $20	20	18.2
$20 to $50	8	7.3
$50 to great than $500	13	11.6
Not applicable	69	62.7
Total	**110**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, approximately how long did it take for you to travel one way to your doctor/clinician for management of your IRD?

Estimate this time for the doctor/clinician you visit most often for your condition.

- Less than 15 minutes
- Between 15 minutes and 30 minutes
- Between 30 minutes and an hour
- Between 1 hour and 2 hours
- Greater than 2 hours

Table C.36 Approximately how long does it take for you to travel one way to your doctor/clinician for management of your IRD? (n=75) (Canada specific)

Average hours spent travelling to medical appointments	Count (n)	Percentage (%)
Less than 15 minutes	12	16.0
Between 15 minutes and 30 minutes	22	29.3
Between 30 minutes and an hour	29	38.7
Between 1 hour and 2 hours	12	16.0
Greater than 2 hours	0	0.0
Total	**75**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, approximately how much did you spend to travel one way to your doctor/clinician for management of your IRD?

- $0 to $10
- $10 to $20
- $20 to $30
- $30 to $40
- $40 to $60
- $60 to $80
- $80 to $100
- Greater than $100

Table C.37 Prior to any COVID-19 interruptions, approximately how much did you spend to travel one way to your doctor/clinician for management of your IRD? (n=110) (Canada specific)

Average travel cost of the medical appointment	Count (n)	Percentage (%)
$0 to $10	55	50.0
$10 to $20	20	18.2
$20 to $30	17	15.5
$30 to $100	11	9.9
Greater than $100	7	6.4
Total	**110**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, did you take time off work or education to attend these appointments?

- Yes, for all of my appointments
- Yes, for about three quarters of my appointments
- Yes, for about half of my appointments
- Yes, for about one quarter of my appointments
- No, I do not take time off work or education to attend my appointments

Table C.38 Prior to any COVID-19 interruptions, did you take time off work or education to attend these appointments? (n=110) (Canada specific)

Time off for appointments (total)	Count (n)	Percentage (%)
Yes, for all of my appointments	52	47.3
Yes, for about one quarter of my appointments	5	4.5
Yes, for about half to three quarters of my appointments	6	5.4
No, I do not take time off work or education to attend my appointments	47	42.7
Total	**110**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, did you attend clinic visits alone or with a friend, family member or carer?

- I attend consultations alone
- I attend consultations with a friend or family member
- I attend consultations with a formal carer
- I attend consultations with a friend or family member and a formal carer

Table C.39 Do you attend consultations alone or with a friend, family member or carer? (n=47) (Canada specific)

Appointment companion type	Count (n)	Percentage (%)
I attend clinic visits with a friend or family member / I attend clinic visits with a friend or family member and a formal carer / I attend clinic visits with a formal carer	70	63.6
I attend clinic visits alone	40	36.4
Total	**110**	**100**

Source: Deloitte Access Economics analysis.

Does your friend, carer or family member take time off work to attend these appointments?

- Yes, for all of my appointments
- Yes, for about three quarters of my appointments
- Yes, for about half of my appointments
- Yes, for about one quarter of my appointments
- No, I attend these appointments alone

Table C.40 Does your friend, carer or family member take time off work to attend these appointments? (n=70) (Canada specific)

Appointment companion time off	Count (n)	Percentage (%)
Yes, for all of my appointments	40	57.1
Yes, for about half to three quarters of my appointments	13	18.6
Yes, for about one quarter of my appointments	10	14.3
No, I attend these appointments alone	7	10.0
Total	**70**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, had any other members of your family seen a specialist to check for IRD symptoms?

- Yes
- No
- Unsure

Table C.41 Have you seen any health providers specifically for or relating to the management of your IRD in the last year? (n=70) (Canada specific)

Family specialist check	Count (n)	Percentage (%)
Yes	33	47.1
No	31	44.3
Unsure	6	8.6
Total	**70**	**100**

Source: Deloitte Access Economics analysis.

How many of your family members have seen a specialist to check for IRD symptoms?

- One
- Two
- Three
- Four
- Five or more
- Unsure

Table C.42 How many of your family members have seen a specialist to check for IRD symptoms? (n=33) (Canada specific)

Family specialist check (number)	Count (n)	Percentage (%)
One	10	30.3
Two	11	33.3
Three or more	12	36.4
Total	**33**	**100**

Source: Deloitte Access Economics analysis.

Have your family members ever received a genetic test specifically for an IRD?

- Yes
- No
- Unsure

Table C.43 Have your family members ever received a genetic test specifically for an IRD? (n=33) (Canada specific)

Family genetic testing	Count (n)	Percentage (%)
No / Unsure	17	51.3
Yes	16	48.5
Total	**33**	**100**

Source: Deloitte Access Economics analysis.

C.9. Module 6: Management of your condition – medications and supplements

Prior to any COVID-19 interruptions, did you use any medications to help manage your IRD?

- Yes
- No

Table C.44 Prior to any COVID-19 interruptions, did you use any medications to help manage your IRD?? (n=151) (Canada specific)

Medication usage	Count (n)	Percentage (%)
No	122	80.8
Yes	29	19.2
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

What medications do you use to manage your IRD? (Tick all which apply).

(Please check with your healthcare provider before taking any supplements).

- Diamox
- Dexamethasone
- Eye drops
- Other (please specify)

Table C.45 What medications do you use to manage your IRD? (n=35) (Canada specific)

Medication usage	Count (n)	Percentage (%)
Dexamethasone	0	0
Eye drops	21	13.9
Diamox / Other	14	9.1
Total	**35**	**100**

Source: Deloitte Access Economics analysis. Note: This question allowed for the selection of multiple answers, hence, the percentages do not sum to 100.

Prior to any COVID-19 interruptions, did you use any vitamins or nutritional supplements to help manage your IRD?

- Yes
- No

Table C.46 Prior to any COVID-19 interruptions, did you use any vitamins or nutritional supplements to help manage your IRD? (n=151) (Canada specific)

Vitamin usage	Count (n)	Percentage (%)
Yes	88	58.3
No	63	41.7
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

What vitamins or nutritional supplements do you use to manage your IRD? (Tick all which apply)

- Multivitamins and iron
- Omega 3
- Omega 7
- Omega 7 mix brand
- Vitamin A palmitate
- Other (please specify)

Table C.47 What vitamins or nutritional supplements do you use to manage your IRD? (n=15) (Canada specific)

Vitamin usage	Count (n)	Percentage (%)
Omega 3	51	33.8
Other	47	31.1
Multivitamins and iron	39	25.8
Vitamin A palmitate	29	19.2
Omega 7 or Omega 7 Mix Brand	5	3.3

Source: Deloitte Access Economics analysis. Note: This question allowed for the selection of multiple answers, hence, the percentages do not sum to 100

Prior to any COVID-19 interruptions, approximately how much money, on average, did you spend on medications and supplements specifically for your IRD, after refunds, reimbursements and financial supports in a typical 4-week period? Please only include out of pocket costs.

- $0 to $10
- $10 to $20
- $20 to $30
- $30 to $50
- $50 to $100
- Greater than $100

Table C.48 Prior to any COVID-19 interruptions, approximately how much money, on average, did you spend on medications and supplements specifically for your IRD, after refunds, reimbursements and financial supports in a typical 4-week period? (n=91) (Canada specific)

Average cost of medications & supplements	Count (n)	Percentage (%)
$0 to $10	13	14.3
$10 to $20	18	19.8
$20 to $30	15	16.5
$30 to $50	12	13.2
$50 to $100	14	15.4
Greater than $100	19	20.9
Total	**91**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, which of the following options best described your health coverage?

- Covered by a private health insurance policy
- Covered by an organizational or industry health care plan
- Currently uninsured and not covered by a health care plan
- Other (please specify)

Table C.49 Prior to any COVID-19 interruptions, which of the following options best described your health coverage? (n=138) (Canada specific)

Average cost of medications & supplements	Count (n)	Percentage (%)
Covered by an organizational or industry health care plan	54	59.3
Covered by a private health insurance policy	35	38.5
Other	33	36.3
Currently uninsured and not covered by a health care plan	16	17.6
Total	**138**	**100**

Source: Deloitte Access Economics analysis.

Table C.50 Prior to any COVID-19 interruptions, had you received Luxturna (voretigene neparvovec-rzyl) gene therapy to treat your IRD? (n=151) (Canada specific)

Luxturna treatment	Count (n)	Percentage (%)
No, I have not received Luxturna gene therapy / Yes, I have received Luxturna gene therapy / No response	151	100.0
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, approximately how much money did you spend on Luxturna (voretigene neparvovec-rzyl) gene therapy after refunds, reimbursements and financial supports? Please only include out of pocket costs. This refers to the treatment only and not to transport or accommodation etc. Please enter a numerical answer rounded to the nearest whole number (i.e. no '$' symbol). For example, if you spent $20, please enter 20.

Table C.51 Prior to any COVID-19 interruptions, approximately how much money did you spend on Luxturna (voretigene neparvovec-rzyl) gene therapy after refunds, reimbursements and financial supports? Please only include out of pocket costs. This refers to the treatment only and not to transport or accommodation etc. (n=2) (Canada specific)

Luxturna treatment cost	Count (n)	Percentage (%)
No responses	151	100.0
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Have you travelled within the US or to some other country to be treated with the gene therapy Luxturna (voretigene neparvovec-rzyl)?

- No, I have not travelled within the US or some other country
- Yes, I have travelled within the United States

Table C.52 Have you travelled within the US or to some other country to be treated with the gene therapy Luxturna (voretigene neparvovec-rzyl)? (n=151) (Canada specific)

Luxturna treatment travel	Count (n)	Percentage (%)
Yes, I have travelled within the United States / No, I have not travelled within the US or some other country / No response	151	100.0
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, approximately how much money did you spend on travel, including transportation, accommodations, and all other related expenses, to receive Luxturna? This does not include the cost of the treatment itself. Please enter a numerical answer rounded to the nearest whole number (i.e. no '$' symbol). For example, if you spent $20, please enter 20.

Table C.53 Prior to any COVID-19 interruptions, approximately how much money did you spend on travel, including transportation, accommodations, and all other related expenses, to receive Luxturna? This does not include the cost of the treatment itself. (n=151) (Canada specific)

Luxturna treatment travel cost	Count (n)	Percentage (%)
No response	151	100.0
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

C.10. Module 7: Management of your condition – vision aids and modifications

For each of the following aids and modifications, please specify whether or not you used the aid or modification for your IRD in a typical year prior to any COVID-19 interruptions.

- Book alternatives (such as audio books)
- Books with enlarged font
- Braille display
- Customised clocks or timers
- Ergonomic adaptations (e.g. raised screens) at home
- Ergonomic adaptations (e.g. raised screens) at work
- Contrast enhancing filters
- High vision lamps
- Large keyboards
- Tactile or large print labels
- Laptops (only select if specialised to you)
- Magnifiers
- Magnifying mirrors
- Portable note takers
- Modifications to mobile SMART phones (IOS or Android) such as applications (apps)
- Screen magnification technology
- Screen reading software
- Spoken word processor
- Green or blue blocking sunglasses
- Verbal calculator

For each aid or modification that you used, please specify approximately how many you used and how much money you spent in total in a typical year prior to any COVID-19 interruptions.

Please enter a numerical answer rounded to the nearest whole number (i.e. no '$' symbol). For example, if you spent $20, please enter 20.

Table C.54 For each of the following aids and modifications, please specify whether or not you used the aid or modification for your IRD in a typical year prior to any COVID-19 interruptions.? (n=151) (Canada specific)

Aid or Modification	Count (n)	Percentage (%)	Average ($)
Magnifiers	78	51.7	485
Modifications to mobile SMART phones (IOS or Android) such as applications (apps)	73	48.3	713
Books with enlarged font	70	46.4	364
Screen magnification technology	48	31.8	1383
Laptops (only select if specialised to you)	45	29.8	1543
Screen reading software	43	28.5	658
Green or blue blocking sunglasses	40	26.5	391
Book alternatives (such as audio books)	37	24.5	254
High vision lamps	32	21.2	146
Customised clocks or timers	28	18.5	276
Tactile or large print labels	25	16.6	108
Large keyboards	24	15.9	326
Ergonomic adaptations (e.g. raised screens) at home	23	15.2	314

Aid or Modification	Count (n)	Percentage (%)	Average ($)
Ergonomic adaptations (e.g. raised screens) at work	22	14.6	283
Contrast enhancing filters	22	14.6	639
Spoken word processor	22	14.6	942
Verbal calculator	19	12.6	122
Magnifying mirrors	16	10.6	66
Portable note takers	5	3.3	2178
Braille display	4	2.7	2667

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, were you a guide dog owner?

- Yes
- No

Table C.55 Prior to any COVID-19 interruptions, were you a guide dog owner? (n=151) (Canada specific)

Guide dog ownership	Count (n)	Percentage (%)
No	139	92.1
Yes	12	7.9
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

In a typical year prior to any COVID-19 interruptions, approximately how many days in total did you spend learning with a trained professional to work with your guide dog or dogs if you have had more than one? If you did not receive any training, please answer not applicable.

- Not applicable
- 1 day
- 2 days
- 3 to 5 days
- 5 to 7 days
- 7 to 10 days
- 10 to 14 days
- 14 to 30 days
- More than 30 days

Table C.56 In a typical year prior to any COVID-19 interruptions, approximately how many days in total did you spend learning with a trained professional to work with your guide dog or dogs if you have had more than one? (n=12) (Canada specific)

Guide dog – average training time	Count (n)	Percentage (%)
1 to 14 days	7	58.3
14 to 30 days / Not applicable	5	41.7
Total	**12**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, approximately how much money did you spend on caring for your guide dog in a typical month?

- $0 to $20
- $20 to $50
- $50 to $100
- Greater than $100

Table C.57 Prior to any COVID-19 interruptions, approximately how much money did you spend on caring for your guide dog in a typical month? (n=12) (Canada specific)

Money spent on guide dog in a month	Count (n)	Percentage (%)
$0 to $20	0	0.0
$20 to $50	0	0.0
$50 to $100	5	41.7
Greater than $100	7	58.3
Total	**12**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, did you use a 'white' cane to aid your mobility?

- Yes
- No

Table C.58 Prior to any COVID-19 interruptions, did you use a 'white' cane to aid your mobility? (n=151) (Canada specific)

White cane owners	Count (n)	Percentage (%)
No	75	49.7
Yes	76	50.3
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

In a typical year prior to any COVID-19 interruptions, approximately how many days in total did you spend learning with a trained professional to use your cane? If you did not receive any training, please answer not applicable

- Not applicable
- 1 day
- 2 days
- 3 to 5 days
- 5 to 7 days
- 7 to 10 days
- 10 to 14 days
- 14 to 30 days
- More than 30 days

Table C.59 In a typical year prior to any COVID-19 interruptions, approximately how many days in total did you spend learning with a trained professional to use your cane (n=74) (Canada specific)

White cane – average training time	Count (n)	Percentage (%)
1 day	13	17.6
2 days	8	10.8
3 to 5 days	9	12.2
5 to more than 30 days	11	14.8
Not applicable	33	44.6
Total	**74**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, did you use the aid of a mini guide or other electronic mobility devices (GPS or sensor devices)?

- Yes
- No

Table C.60 Prior to any COVID-19 interruptions, did you use the aid of a mini guide or other electronic mobility devices (GPS or sensor devices)? (n=151) (Canada specific)

Electronic mobility devices	Count (n)	Percentage (%)
Yes	76	50.3
No	75	49.7
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

What was the approximate cost of this device?

- $0 to $50
- $50 to $50
- $100 to $200
- Greater than $200

Table C.61 What was the approximate cost of this device? (n=31) (Canada specific)

Average cost of electronic mobility device	Count (n)	Percentage (%)
$0 to $100	14	45.2
Greater than $200	17	54.8
Total	**31**	**100**

Source: Deloitte Access Economics analysis.

Approximately how many hours did you spend training with the instructor for your mobility device?

- Not applicable
- 1 day
- 2 days
- 3 to 5 days
- 5 to 7 days
- 7 to 10 days
- 10 to 14 days
- 14 to 30 days
- More than 30 days

Table C.62 Approximately how many hours did you spend training with the instructor for your mobility device? (n=31) (Canada specific)

Electronic mobility device – average training time	Count (n)	Percentage (%)
1 to more than 30 days	5	16.1
Not applicable	26	83.9
Total	**31**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, did you have customised door frames or light switches in your home specifically for your IRD?

- Yes
- No

Table C.63 Prior to any COVID-19 interruptions, did you have customised door frames or light switches in your home specifically for your IRD? (n=151) (Canada specific)

Customised door frames or light switches	Count (n)	Percentage (%)
No	137	90.7
Yes	14	9.3
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

If you have customised door frames or light switches in your home specifically or including for your IRD, please estimate the cost of this modification. If you are unsure please answer not applicable.

- Not applicable
- $0 to $100
- $100 to $500
- $500 to $1000
- $1000 to $5000
- Greater than $5000

Table C.64 If you have customised door frames or light switches in your home specifically or including for your IRD? (n=14) (Canada specific)

Customised door frames or light switches – average cost	Count (n)	Percentage (%)
$0 to $500	8	57.8
$500 to greater than $5000 / Not applicable	6	42.8
Total	**14**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, did you have customised bathtubs, showers or other bathroom modifications in your home specifically for your IRD?

- Yes
- No

Table C.65 Prior to any COVID-19 interruptions, did you have customised bathtubs, showers or other bathroom modifications in your home specifically for your IRD? (n=151) (Canada specific)

Bathroom modifications	Count (n)	Percentage (%)
No	143	94.7
Yes	8	5.3
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

If you have customised bathtubs, showers or other bathroom modifications in your home specifically or including for your IRD, please estimate the cost of these modifications. If you are unsure, please answer not applicable.

- $0 to $100
- $100 to $500
- $500 to $1000
- $1000 to $5000
- Greater than $5000
- Not applicable

Table C.66 If you have customised bathtubs, showers or other bathroom modifications in your home specifically or including for your IRD, please estimate the cost of these modifications. If you are unsure, please answer not applicable. (n=7) (Canada specific)

Bathroom modifications – average cost	Count (n)	Percentage (%)
$0 to greater than $5000 / Not applicable	7	100
Total	**7**	**100**

Source: Deloitte Access Economics analysis.

C.11. Module 8: Formal care

Prior to any COVID-19 interruptions, did you have a formal carer (a person who is paid to give care) to assist you with your activities of daily living specifically or including for your IRD?

- Yes
- No

Table C.67 Prior to any COVID-19 interruptions, did you have a formal carer (a person who is paid to give care) to assist you with your activities of daily living specifically or including for your IRD? (n=151) (Canada specific)

Formal care	Count (n)	Percentage (%)
No	144	95.4
Yes	7	4.6
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, approximately how many hours per week did you receive assistance from a formal carer specifically or including for your IRD?

- Less than 5 hours
- 5 to 9 hours
- 10 to 19 hours
- 20 to 29 hours
- 30 to 39 hours
- 40 hours or more

Table C.68 Prior to any COVID-19 interruptions, approximately how many hours per week did you receive assistance from a formal carer specifically or including for your IRD? (n=7) (Canada specific)

Formal care hours	Count (n)	Percentage (%)
Less than 5 hours to greater than 40 hours	7	100
Total	**7**	**100**

Source: Deloitte Access Economics analysis.

C.12. Module 9: Informal care

Prior to any COVID-19 interruptions, did you receive any assistance from an informal carer? An informal carer includes any person, such as a family member, friend or neighbor, who gave regular, ongoing assistance to another person without payment for the care given. If so, from whom?

(Please identify your primary informal carer if you have more than one informal carer).

- Spouse
- Family member or relative
- Friend or other informal carer
- I do not receive any assistance from an informal carer

Table C.69 Prior to any COVID-19 interruptions, did you receive any assistance from an informal carer? An informal carer includes any person, such as a family member, friend or neighbour, who gave regular, ongoing assistance to another person without payment for the care given. If so, from whom? (n=117) (Canada specific)

Informal carer type	Count (n)	Percentage (%)
Spouse	58	49.6
I do not receive any assistance from an informal carer	43	36.8
Friend or other informal carer	16	13.7
Family member or relative	0	0.0
Total	**117**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, approximately how many hours per week did you receive assistance from an informal carer?

- Less than 5 hours
- 5 to 9 hours
- 10 to 19 hours
- 20 to 29 hours
- 30 to 39 hours
- 40 hours or more

Table C.70 Prior to any COVID-19 interruptions, approximately how many hours per week did you receive assistance from an informal carer? (n=108) (Canada specific)

Informal care hours	Count (n)	Percentage (%)
Less than 5 hours	55	50.9
5 to 9 hours	17	15.7
10 to 19 hours	16	14.8
20 to 39 hours	7	6.5
40 hours or more	13	12.0
Total	**108**	**100**

Source: Deloitte Access Economics analysis.

What was the sex/gender of your informal carer? (Please choose the option as it applies to your primary informal carer if you have more than one informal carer)

- Male
- Female
- Other/unsure

Table C.71 What was the sex/gender of your informal carer? (n=108) (Canada specific)

Informal carer gender	Count (n)	Percentage (%)
Female	57	52.8
Male / Other / Unsure	51	47.2
Total	**108**	**100**

Source: Deloitte Access Economics analysis.

What was the approximate age of your informal carer? (Please choose the option as it applies to your primary informal carer if you have more than one informal carer and guess if you are unsure exactly)

- 15 to 24
- 25 to 34
- 35 to 44
- 45 to 54
- 55 to 64
- 65 to 74
- 75 or older

Table C.72 What was the approximate age of your informal carer? (n=108) (Canada specific)

Informal carer age	Count (n)	Percentage (%)
15 to 34	7	6.5
35 to 44	14	13.0
45 to 54	23	21.3
55 to 64	34	31.5
65 to 74	22	20.4
75 or older	8	7.4
Total	**108**	**100**

Source: Deloitte Access Economics analysis.

Prior to any COVID-19 interruptions, was your informal carer employed and if so, in what capacity? (Please choose the option as it applies to your primary informal carer if you have more than one informal carer).

- Retired
- Casual work (Not working on a regular basis)
- Part time work
- Full time work
- My informal carer is not employed, and not looking for work
- My informal carer is not employed, and is looking for work
- My informal carer is a student
- Unsure
- Underemployed (e.g. working part time but desiring full time work; working in a role that does not use their skills or qualifications)

Table C.73 Prior to any COVID-19 interruptions, was your informal carer employed and if so, in what capacity? (n=105) (Canada specific)

Informal carer employment	Count (n)	Percentage (%)
Full time work	42	40.0
Retired	37	35.2
Part time work / Casual work (Not working on a regular basis)	14	13.3
My informal carer is not employed, and not looking for work	7	6.7
Underemployed (e.g. working part time but desiring full time work; working in a role that does not use their skills or qualifications) / My informal carer is not employed, and is looking for work / Unsure	5	4.8
Total	**105**	**100**

Source: Deloitte Access Economics analysis.

C.13. Module 10: Government support

Prior to any COVID-19 interruptions, did you receive any government support specifically or including for your IRD? If so, which of the following forms of assistance did you receive?

- Social Security Disability Insurance program
- Supplemental Security Income (SSI) program
- Other (please specify)
- Unsure
- I do not receive any government support for my condition

Table C.74 Do you currently receive any government support specifically or including for your IRD (n=151) (Canada specific)

Government Payment	Count (n)	Percentage (%)
Canadian Disability Tax Credit for Blindness	82	54.3
Canada National Institute for the Blind (CNIB) Card	74	49.0
I do not receive any government support for my condition	46	30.5
Canada Pension Plan (CPP) / Quebec Pension Plan (QPP) Disability Benefits	24	15.9
Other (please specify)	16	10.6
Total	**151**	**100**

Source: Deloitte Access Economics analysis. Note: This question allowed for the selection of multiple answers, hence, the percentages do not sum to 100.

Prior to any COVID-19 interruptions, did your informal carer receive any government support specifically or including for your IRD? If so, please provide details.

- My informal carer does not receive any government support for my condition
- Yes, my informal carer receives government support for my IRD (please specify)
- Unsure

Table C.75 Does your informal carer receive any government support specifically or including for your IRD? (n=151) (Canada specific)

Government Payment	Count (n)	Percentage (%)
My informal carer does not receive any government support for my condition	131	86.8
Yes, my informal carer receives government support for my IRD (please specify)	10	6.6
Unsure	10	6.6
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

C.14. Module 11: Other impacts of your condition

Please reflect on your life prior to the COVID-19 pandemic, did you ever experience feelings of depression because of your IRD?

- Yes
- No

Table C.76 Please reflect on your life prior to the COVID-19 pandemic, did you ever experience feelings of depression because of your IRD? (n=151) (Canada specific)

Experience of depression	Count (n)	Percentage (%)
Yes	112	74.2
No	39	25.8
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Please reflect on your life prior to the COVID-19 pandemic, did you ever experience feelings of anxiety because of your IRD?

- Yes
- No

Table C.77 Please reflect on your life prior to the COVID-19 pandemic, did you ever experience feelings of anxiety because of your IRD? (n=151) (Canada specific)

Experience of anxiety	Count (n)	Percentage (%)
Yes	128	84.8
No	23	15.2
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Please reflect on your life prior to the COVID-19 pandemic, did you ever experience another mental health condition because of your IRD?

- Yes, please describe
- No

Table C.78 Have you ever experienced feelings of another mental health condition because of your IRD? (n=151) (Canada specific)

Experience of other mental health conditions	Count (n)	Percentage (%)
No	131	86.8
Yes	20	13.2
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

Please reflect on your life prior to the COVID-19 pandemic, did any of your close family members or your informal carer(s) ever experience depression, anxiety, or another mental health condition as a result of your IRD?

- Yes
- No

Table C.79 Have any of your close family members ever experienced feelings of depression, anxiety, or another mental health condition as a result of your IRD? (n=138) (Canada specific)

Family experience of other mental health conditions	Count (n)	Percentage (%)
No	85	61.6
Yes	53	38.4
Total	**138**	**100**

Source: Deloitte Access Economics analysis.

Please reflect on your life prior to the COVID-19 pandemic. On a scale of 1 to 5 with 5 being strongly agree and 1 being strongly disagree, please answer the following statement. I feel frustrated from the lack of awareness and support for IRDs.

- 1 - I strongly disagree that I feel frustrated from the lack of awareness and support for IRDs.
- 2 - I disagree that I feel frustrated from the lack of awareness and support for IRDs.
- 3 - I neither agree nor disagree that I feel frustrated from the lack of awareness and support for IRDs.
- 4 - I agree that I feel frustrated from the lack of awareness and support for IRDs.
- 5 - I strongly agree that I feel frustrated from the lack of awareness and support for IRDs.

Table C.80 Feeling of frustration from the lack of awareness and support for IRDs (n=138) (Canada specific)

Frustration	Count (n)	Percentage (%)
1 – I strongly disagree that I feel frustrated	8	5.8
2 - I disagree that I feel frustrated	7	5.1
3 - I neither agree nor disagree	31	22.5
4 - I agree that I feel frustrated	42	30.4
5 - I strongly agree that I feel frustrated	50	36.2
Total	**138**	**100**

Source: Deloitte Access Economics analysis.

Please reflect on your life prior to the COVID-19 pandemic, did you experience any financial stress because of your IRD?

- Yes
- No
- Unsure

Table C.81 Please reflect on your life prior to the COVID-19 pandemic, did you experience any financial stress because of your IRD? (n=138) (Canada specific)

Experience of other mental health conditions	Count (n)	Percentage (%)
Yes	76	55.1
No / Unsure	62	54.9
Total	**138**	**100**

Source: Deloitte Access Economics analysis.

Please reflect on your life prior to the COVID-19 pandemic, did you experience social isolation because of your IRD? Social isolation is a state of complete or near complete lack of contact between an individual and society.

- Yes
- No
- Unsure

Table C.82 Please reflect on your life prior to the COVID-19 pandemic, did you experience social isolation because of your IRD? (n=151) (Canada specific)

Experience of social isolation	Count (n)	Percentage (%)
No / Unsure	78	51.6
Yes	73	48.4
Total	**151**	**100**

Source: Deloitte Access Economics analysis.

C.15. Module 12: Impact of your child's IRD on your mental wellbeing – from your own perspective as a parent/ guardian:

Please reflect on your life prior to the COVID-19 pandemic, did you ever experience feelings of depression due to your caring requirements for your child with an IRD?

- Yes
- No

Table C.83 Please reflect on your life prior to the COVID-19 pandemic, did you ever experience feelings of depression due to your caring requirements for your child with an IRD? (n=13) (Canada specific)

Experience of depression	Count (n)	Percentage (%)
Yes	8	61.5
No	5	38.5
Total	**13**	**100**

Source: Deloitte Access Economics analysis.

Please reflect on your life prior to the COVID-19 pandemic, did you ever experience feelings of anxiety due to your caring requirements for your child with an IRD?

- Yes
- No

Table C.84 Please reflect on your life prior to the COVID-19 pandemic, did you ever experience feelings of anxiety due to your caring requirements for your child with an IRD? (n=13) (Canada specific)

Experience of anxiety	Count (n)	Percentage (%)
Yes / No	13	100.0
Total	**13**	**100**

Source: Deloitte Access Economics analysis.

Please reflect on your life prior to the COVID-19 pandemic, did you ever experience feelings of another mental health condition due to your caring requirements for your child with an IRD?

- Yes, please specify
- No

Table C.85 Please reflect on your life prior to the COVID-19 pandemic, did you ever experience feelings of another mental health condition due to your caring requirements for your child with an IRD? (n=13) (Canada specific)

Experience of other mental health conditions	Count (n)	Percentage (%)
Yes / No	13	100
Total	**13**	**100**

Source: Deloitte Access Economics analysis.

Please reflect on your life prior to the COVID-19 pandemic, on a scale of 1 to 5 with 5 being strongly agree and 1 being strongly disagree, please answer the following statement. I feel frustrated from the lack of awareness and support for IRDs.

- 1 - I strongly disagree that I feel frustrated from the lack of awareness and support for IRDs.
- 2 - I disagree that I feel frustrated from the lack of awareness and support for IRDs.
- 3 - I neither agree nor disagree that I feel frustrated from the lack of awareness and support for IRDs.
- 4 - I agree that I feel frustrated from the lack of awareness and support for IRDs.
- 5 - I strongly agree that I feel frustrated from the lack of awareness and support for IRDs.

Table C.86 Parent feeling of frustration from the lack of awareness and support for IRDs (n=13) (Canada specific)

Frustration	Count (n)	Percentage (%)
1 – I strongly disagree that I feel frustrated	0	0.0
2 - I disagree that I feel frustrated	0	0.0
3 - I neither agree nor disagree / 4 - I agree that I feel frustrated	8	61.6
5 - I strongly agree that I feel frustrated	5	38.5
Total	**13**	**100**

Source: Deloitte Access Economics analysis.

[Parent] Please reflect on your life prior to the COVID-19 pandemic, did you experience any financial stress because of your IRD?

- Yes
- No
- Unsure

Table C.87 [Parent] Please reflect on your life prior to the COVID-19 pandemic, did you experience any financial stress because of your IRD? (n=13) (Canada specific)

Financial stress	Count (n)	Percentage (%)
Yes / No	13	100.0
Unsure	0	0.0
Total	**13**	**100**

Source: Deloitte Access Economics analysis.

Please reflect on your life prior to the COVID-19 pandemic, did any of your close family members or your informal carer(s) ever experience depression, anxiety, or another mental health condition as a result of your IRD?

- Yes
- No
- Unsure

Table C.88 Please reflect on your life prior to the COVID-19 pandemic, did any of your close family members or your informal carer(s) ever experience depression, anxiety, or another mental health condition as a result of your IRD? (n=13) (Canada specific)

Experience of other mental health conditions	Count (n)	Percentage (%)
Yes / No	13	100.0
Unsure	0	0.0
Total	**13**	**100**

Source: Deloitte Access Economics analysis.

Limitation of our work

General use restriction

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