Understanding the Epidemiology of Neurological Conditions and Building the Methodological Foundation for Surveillance

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Webinar September 12, 2013

15 Priority Neurological Conditions

- Epilepsy
- Multiple Sclerosis
- Dementia
- *Injuries*
  - Spinal cord injuries
  - Traumatic brain injuries
- *Neuromuscular diseases*
  - Amyotrophic lateral sclerosis
  - Muscular dystrophy
- *Congenital disorders*
  - Spina bifida
  - Hydrocephalus (adult onset also)
  - Cerebral palsy
- *Movement disorders*
  - Tourette syndrome
  - Dystonia
  - Huntington disease
  - Parkinson's disease
- Brain Tumour

Objectives

1. To perform systematic reviews on the international incidence and prevalence of all of the priority neurological conditions identified by PHAC and the NHCC.

2. To summarize and make recommendations as to the best ascertainment sources for surveillance purposes for each of the priority conditions.

3. To develop an inventory of existing neurological registries in Canada and other developed countries.

STUDY RESULTS

Objective 1

- To perform international systematic reviews on the incidence and prevalence of all of the priority neurological conditions identified by PHAC and the NHCC

Caveats and Challenges of Providing Single Estimates of Incidence and Prevalence for each Conditions

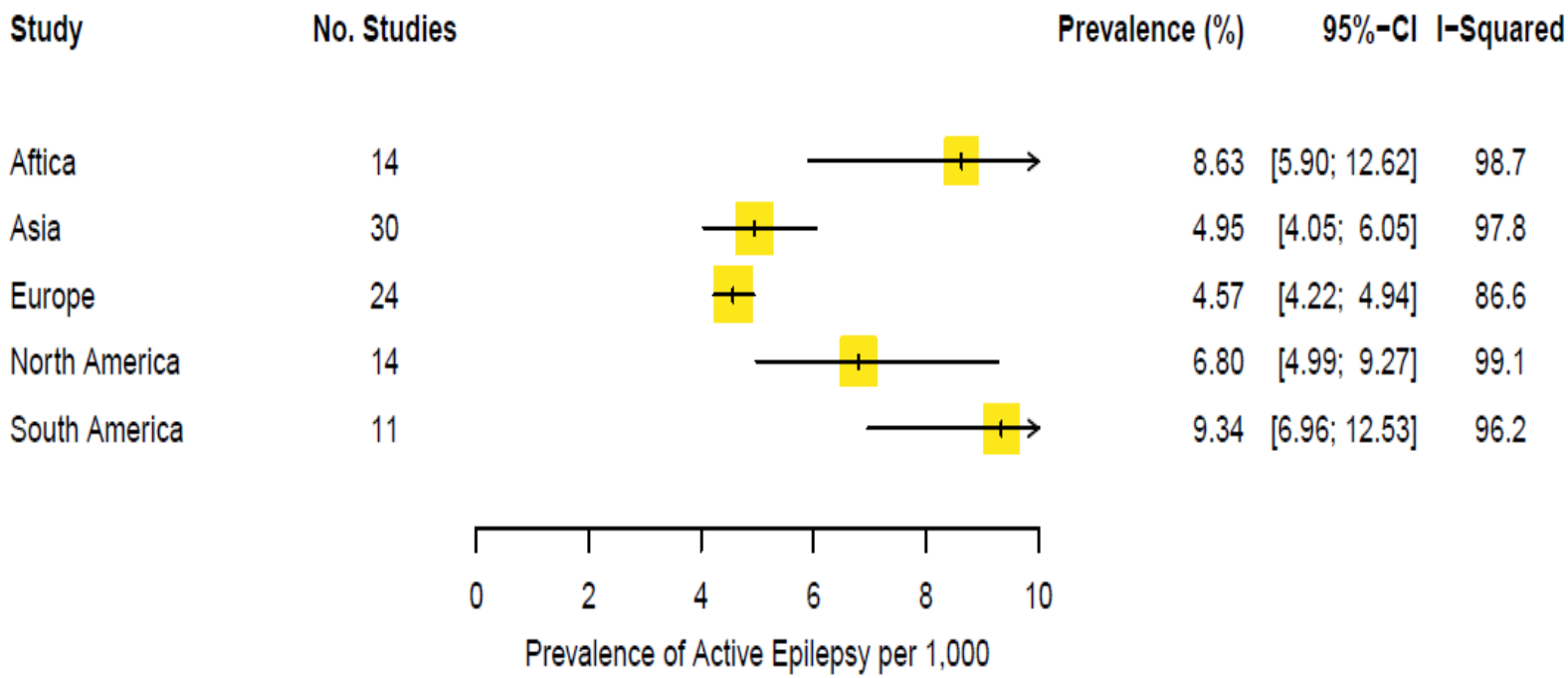
- Different years of data
- Different world regions
- Different settings (e.g. long term care institutions vs. community)
- Different sources of ascertainment
- Different diagnostic criteria
- Different age groups included
- Variables stratified differently from one study to the next
  - age categories often very different from one study to the next
- Adjusted estimates vs. unadjusted estimates
- Need for double checking (even after already reviewed by 2 investigators) – still ongoing for a few conditions
- Overwhelming amount of data for some of the conditions

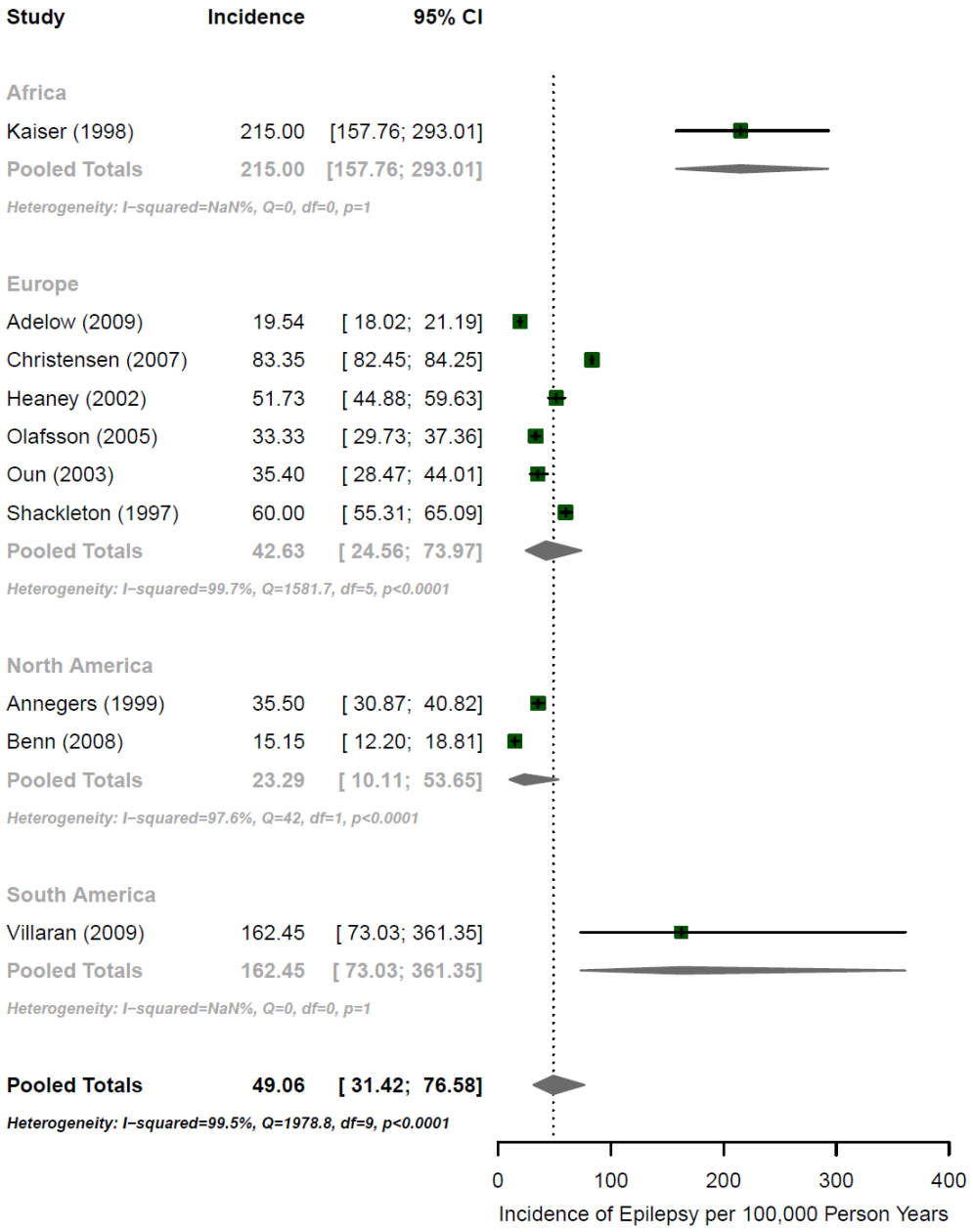
Objective 1: Systematic Reviews of Incidence and Prevalence

Condition	Total # Abstracts	# Abstracts Selected Full Text Review	# Articles Included
ALS (Motor Neuron Disease)	1,909	108	28
Brain Tumors	10,263	241	54
Cerebral Palsy	1,521	91	56
Dementia	16,066	707	177
Dystonia	724	49	16
Epilepsy	13,252	442	202
Huntington's Disease	485	82	20
Hydrocephalus	1289	137	48
Multiple Sclerosis	3,933	491	182
Muscular Dystrophy	1,104	167	31
Parkinson's Disease	4,219	219	134
Spina Bifida	3,336	742	170
Spinal Cord Injury	2,717	202	42
Tourette Syndrome	746	53	31
Traumatic Brain Injury	3,602	167	69
TOTAL	**65,655**	**3,898**	**1,260**

Epilepsy

Epilepsy Prevalence → 0.5 to 0.9%





Epilepsy Incidence 49 per 100,000 person years

Higher in developing countries

Results – Canadian Epilepsy Studies

- 7 Canadian prevalence studies
- Most based on self report (health surveys) – 2 using administrative data
- Prevalence ranges from:
  - 5.2-6.0 per 1,000 in Canada

No Canadian validation of epilepsy self-report

Multiple Sclerosis

Original Paper

Neuro

Neuroepidemiology 2013;40:195–210
DOI: 10.1159/000342779

Received: July 17, 2012
Accepted: August 17, 2012
Published online: January 24, 2013

Incidence and Prevalence of Multiple Sclerosis in the Americas: A Systematic Review

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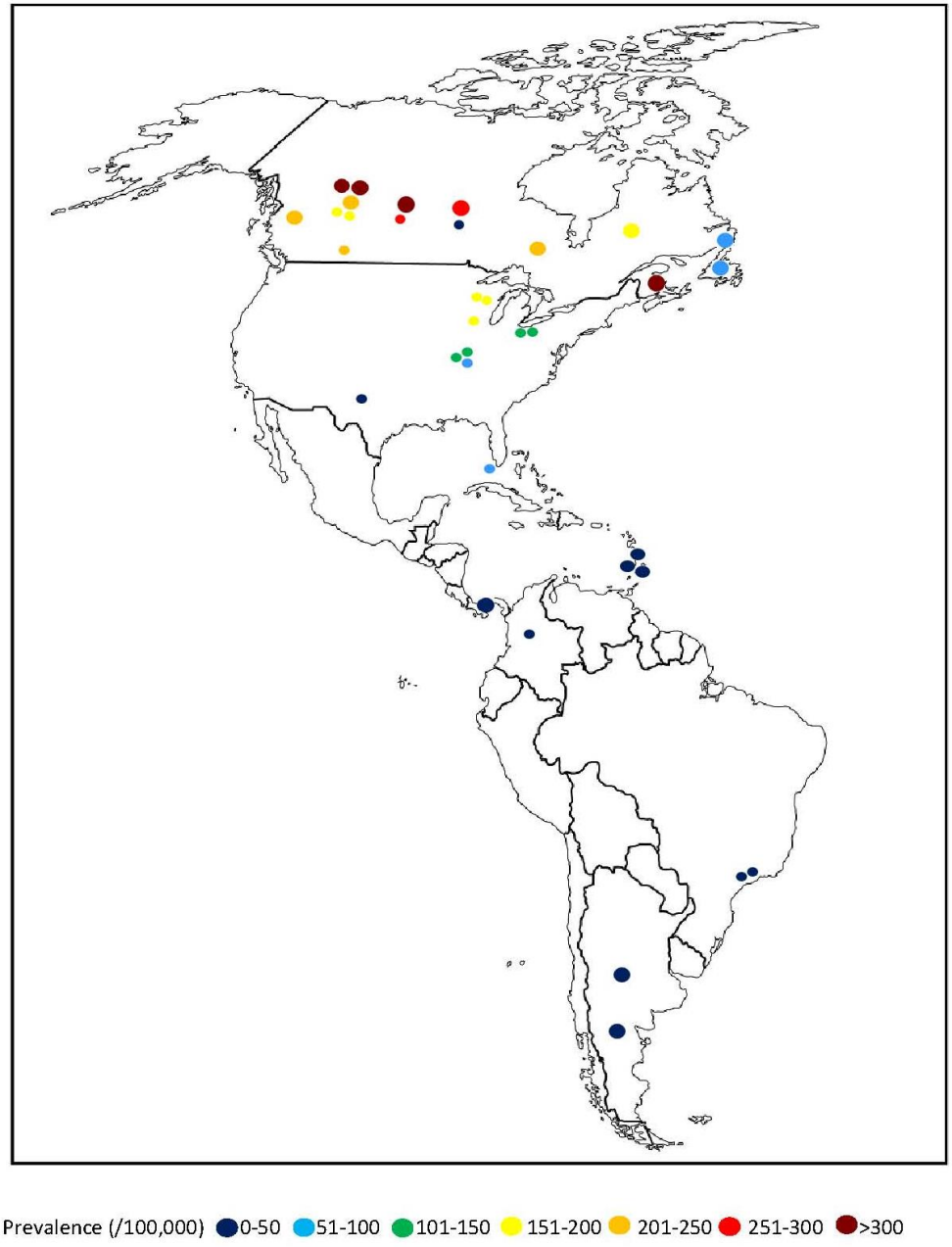
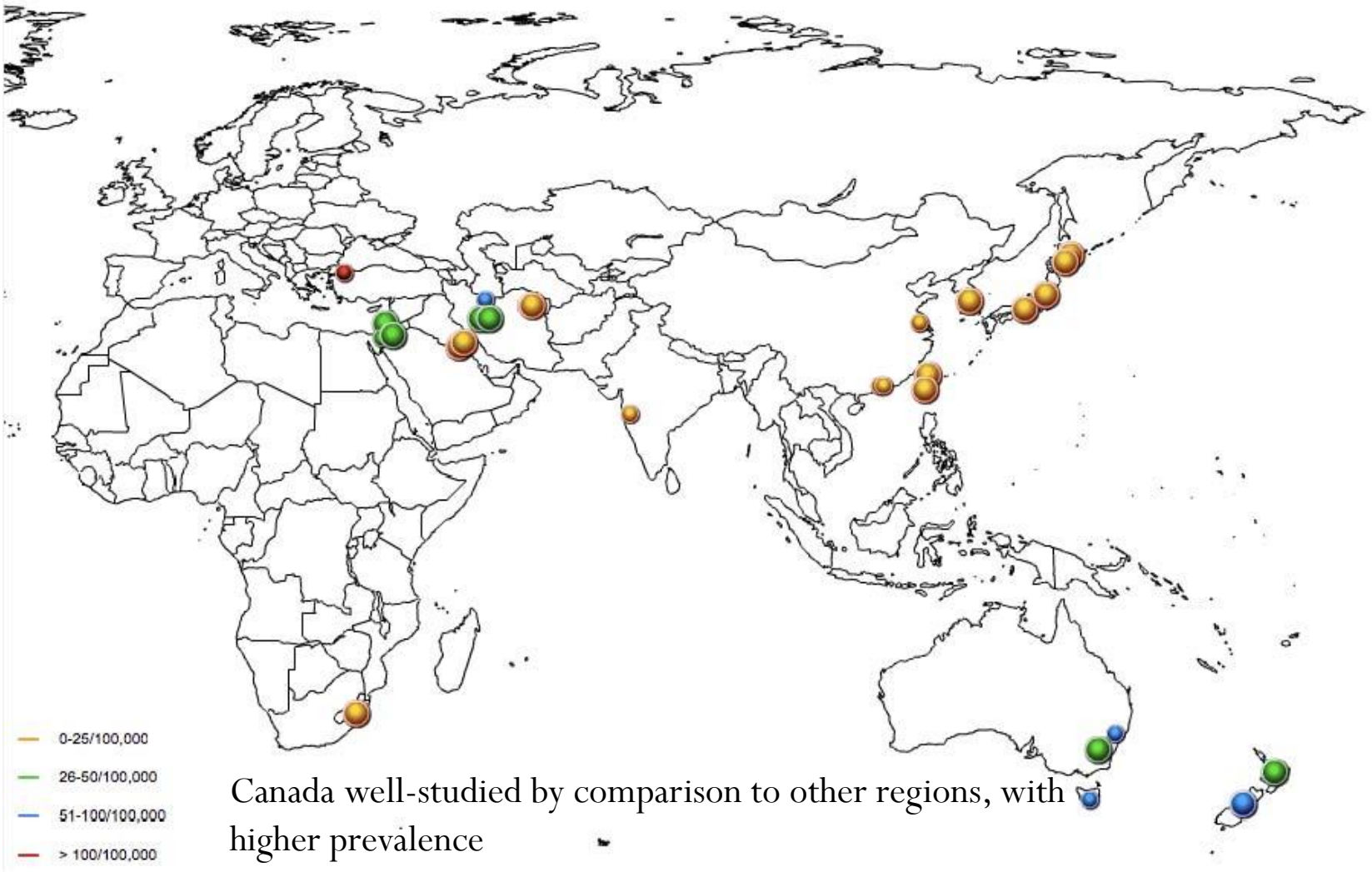
Prevalence of MS in the Americas: studies published 1985-2011

- 31 studies with 12 from Canada → only 1 national (CCHS)
- Larger circles: national or provincial/ state studies
- Smaller circles: county or city studies.
- ***Most recent studies have prevalence exceeding 1/500***



MS Prevalence – Africa, Asia, Australia and New Zealand

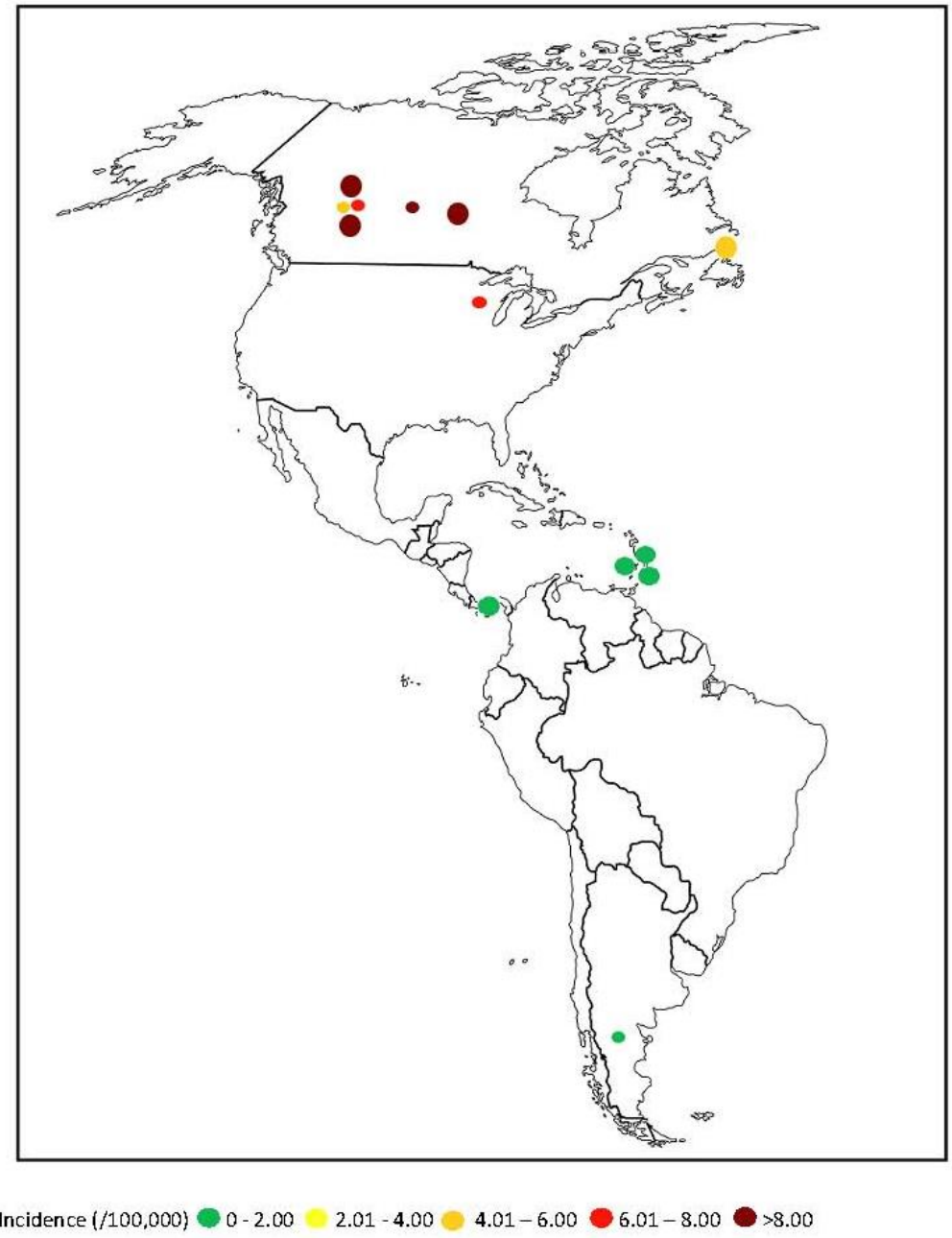


Canada well-studied by comparison to other regions, with higher prevalence

Figure 1. MS Prevalence in Africa, Asia, Australia, and New Zealand
Smaller circles represent studies conducted in a single city; larger circles represent studies conducted in a province, territory, or country.

Incidence of MS - the Americas

- Far fewer incidence studies than prevalence studies
- ***Most conducted in Canada***
- Most regions understudied with respect to incidence & prevalence

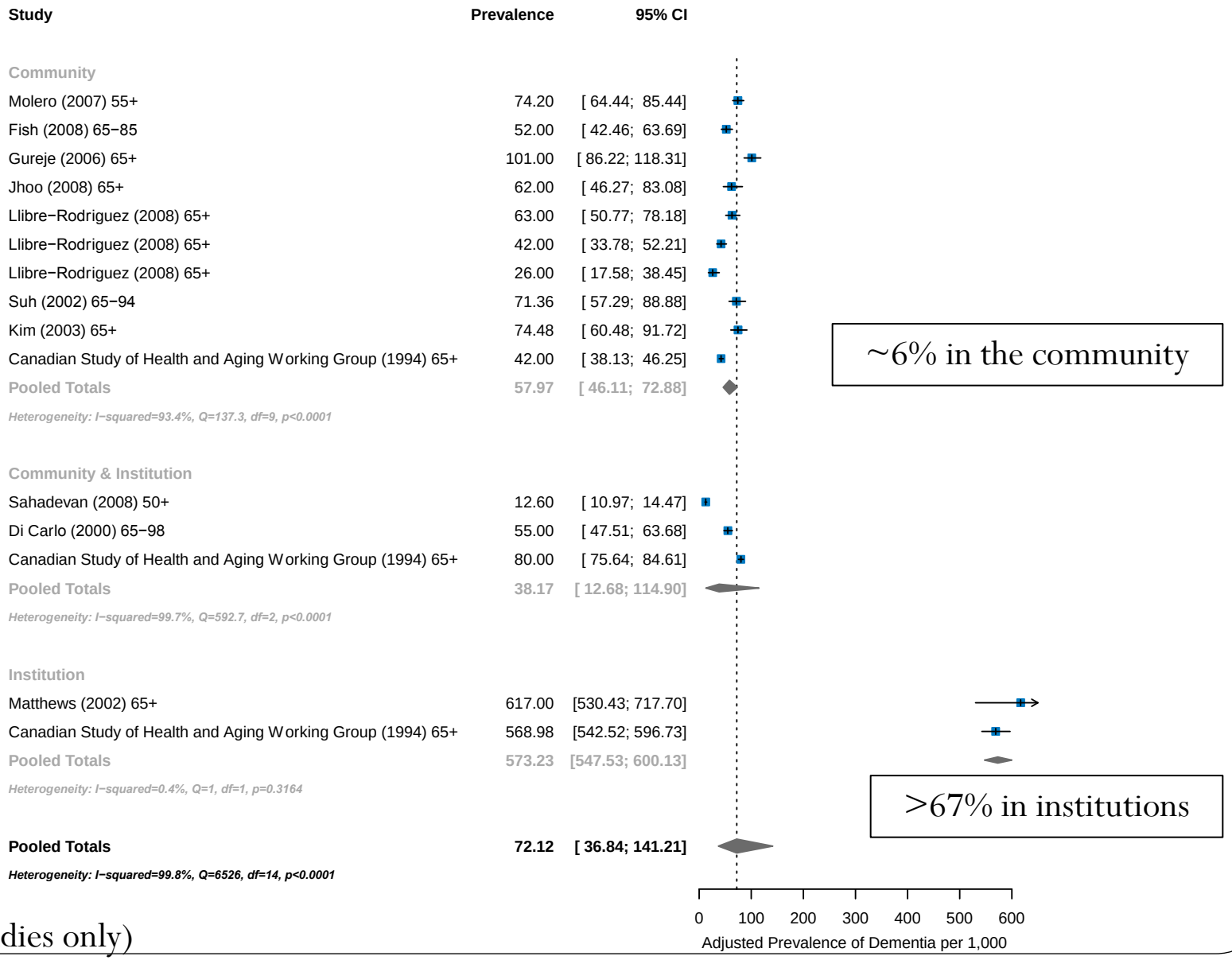


Dementia

Dementias (n=178 studies)

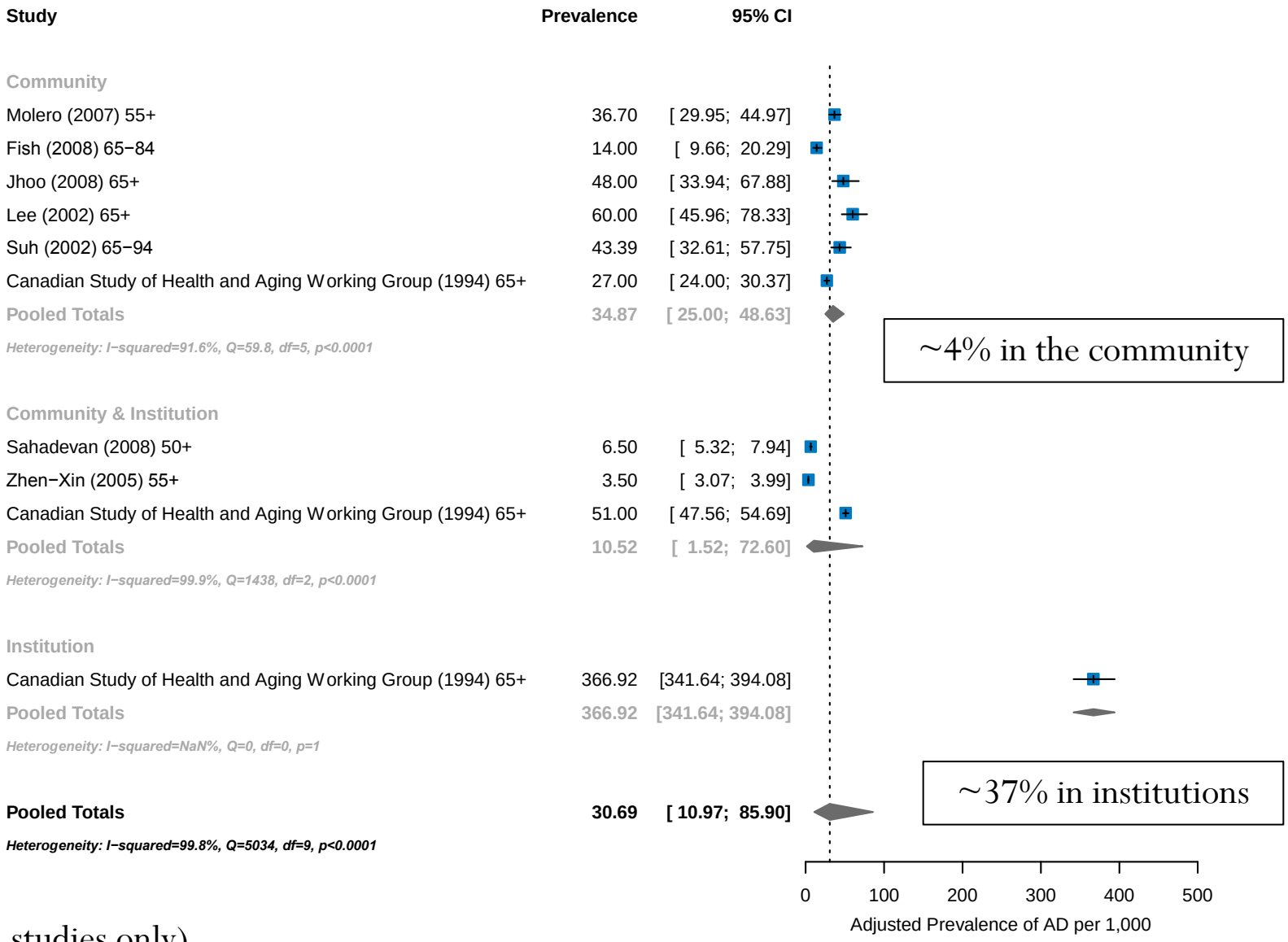
Dementia Type (Unadjusted)	Number of Studies Overall (Incl. in M-A) [Total]	Studies Included in Analyses	Pooled Prevalence per 1000 (95% CI) [Range of Estimates]	Studies Included in Analyses	Pooled Incidence per 1000 (95% CI) [Range of Estimates]
Dementia	109 **[123]**	78	69.4 (53.9-89.3) [0.9 – 676.8]	7	86.5 (56.4-132.6) [8.7 – 232.0]
Alzheimer's Disease	75 **[86]**	35	46.0 (32.9-64.3) [3.8 – 352.0]	6	54.4 (23.1-128.3) [12.3 – 126.1]
Vascular Dementia	53 **[62]**	27	18.4 (11.6-29.0) [2.5 – 294.34]	4	33.6 (14.6-77.5) [14.4 – 95.0]
Frontotem-poral Dementia	**[14]**	---	[0.3 – 31.04]	---	---
Dementia with Lewy Bodies	**[10]**	---	[0.6 – 49.92]	---	---

Dementia Prevalence



(Adjusted studies only)

Alzheimer's Disease Prevalence



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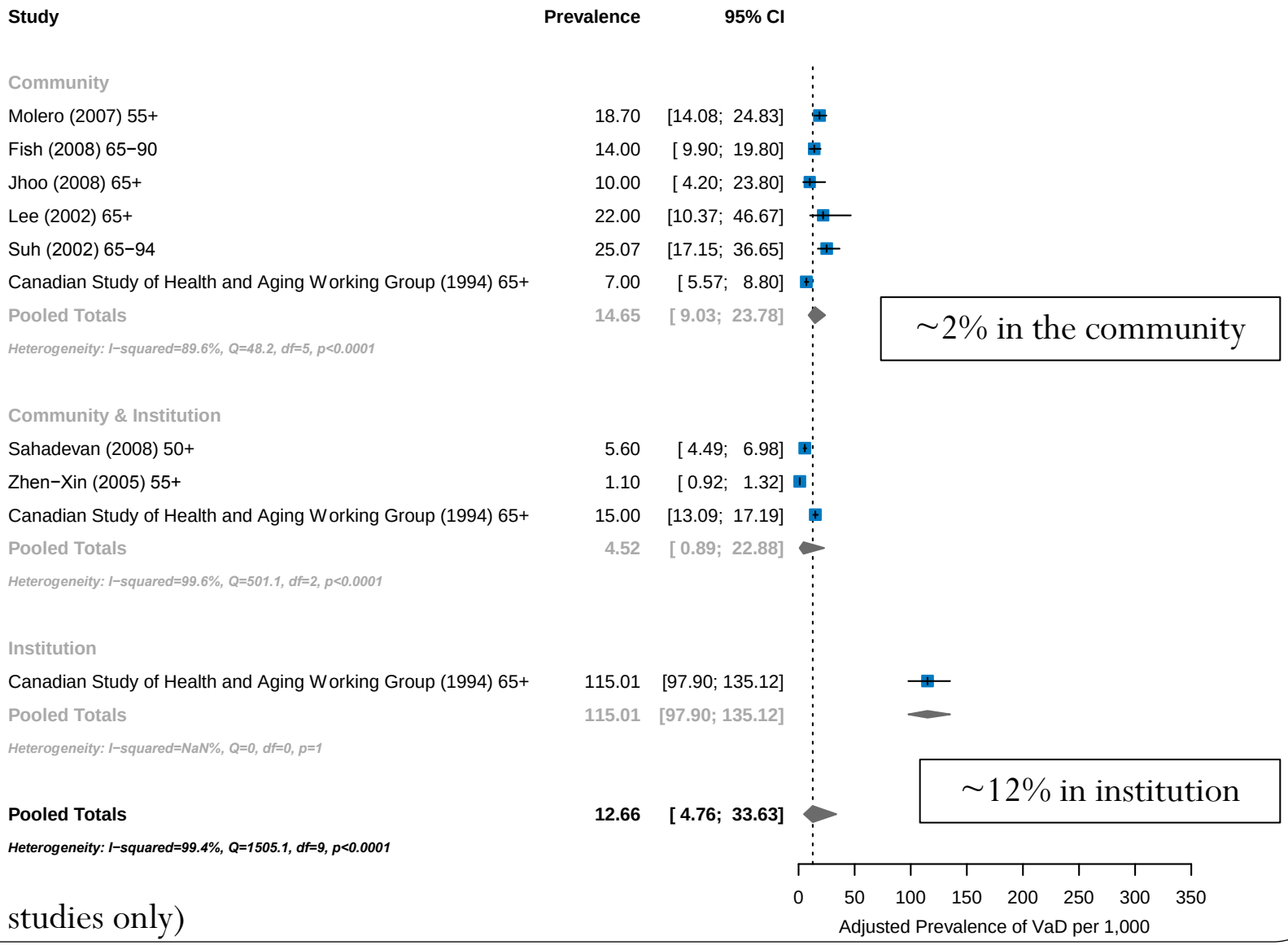
Vascular Dementia Prevalence



(Adjusted studies only)

Lewy Body Dementia

- Relatively few studies available
  - No Canadian data
- Incidence
  - 1.1 - 1.4 per 1000 person-years among those 65+ in the community
- Prevalence
  - More variable but among studies focusing on those 60-65+ in the community, the reported rate is between 0.27 and 5.3 per 1000

Frontotemporal Dementia

- Relatively few studies available
  - No Canadian data
- Affects relatively younger individuals and more variability in diagnostic criteria (i.e., determining whether it is present)
- Incidence: 0.04 - 1.1 per 1000 person-years (higher rates in institutions) among those 30 and up
- Prevalence: 0.7 - 2.6 per 1000 among those 65+ in the community

Canadian Studies of Dementia

- 10 Canadian Studies
  - 7 included in meta-analysis
  - 7 studies using Canadian Study of Health & Aging data (5 included in meta-analysis)
- 8 for Canada overall (CSHA and CCHS), 2 for Manitoba
- Prevalence ranging from 38 to 215 per 1000 persons
- Incidence ranging from 3.4 to 25.3 per 1000 persons

Spinal Cord Injury

Average Annual Incidence of SCI per 100,000 people

Study	Continent	Incidence	95% CI
Gur (2005)	Asia	12.34	[9.45; 16.12]
Ide (1993)	Asia	28.69	[22.03; 37.37]
Karacan (2000)	Asia	12.69	[11.70; 13.76]
Karamehmetoglu (1997)	Asia	20.80	[17.74; 24.38]
Karamehmetoglu (1995)	Asia	16.98	[13.54; 21.30]
Lan (1993)	Asia	56.09	[36.11; 87.13]
Otom (1997)	Asia	17.98	[12.16; 26.57]
Shingu (1994)	Asia	40.23	[39.12; 41.36]
O'Connor (2002)	Australia	14.50	[12.86; 16.36]
Hagen (2010)	Europe	18.48	[9.86; 34.63]
Silberstein (1995)	Europe	28.00	[20.47; 38.29]
van Asbeck (2000)	Europe	12.15	[10.51; 14.04]
Buechner (2000)	North America	56.00	[43.04; 72.87]
Price (1994)	North America	39.84	[33.45; 47.47]
Warren (1995)	North America	82.80	[62.24; 110.16]
Woodruff (1994)	North America	25.00	[18.82; 33.21]
Burke (2001)	North America	25.20	[17.45; 36.40]
Dryden (2005)	North America	43.50	[36.28; 52.16]
Johnson (1997)	North America	44.89	[38.52; 52.31]
Surkin (2000)	North America	76.75	[66.76; 88.24]
Pooled Totals		**28.25**	**[21.37; 37.36]**

Heterogeneity: I–squared=98.7%, Q=1453.9, df=19, p<0.0001

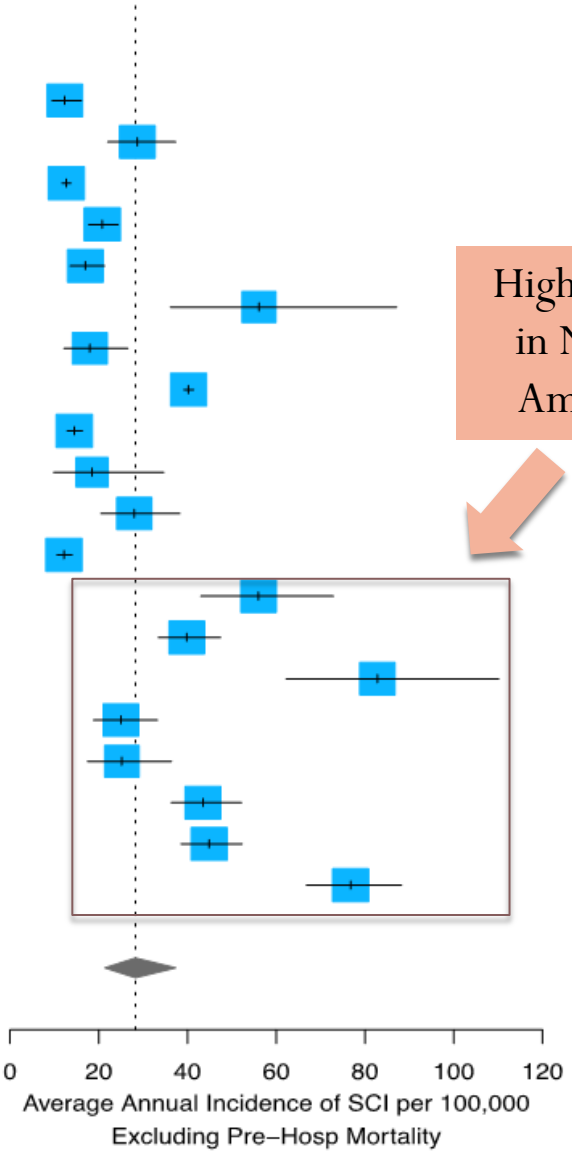


Figure 1: Forest plot of AAI of studies who excluded pre-hospital mortality from their estimates of SCI.

Average Annual Incidence of SCI per 100,000 – Canadian Studies

Study	Incidence	95% CI
Couris (2010)	24.00	[21.11; 27.28]
Dryden (2005)	43.50	[36.28; 52.16]
Pickett (2003)	37.20	[34.01; 40.69]
Pooled Totals	**33.75**	**[24.31; 46.86]**

Heterogeneity: I-squared=94.9%, Q=39.2, df=2, p<0.0001

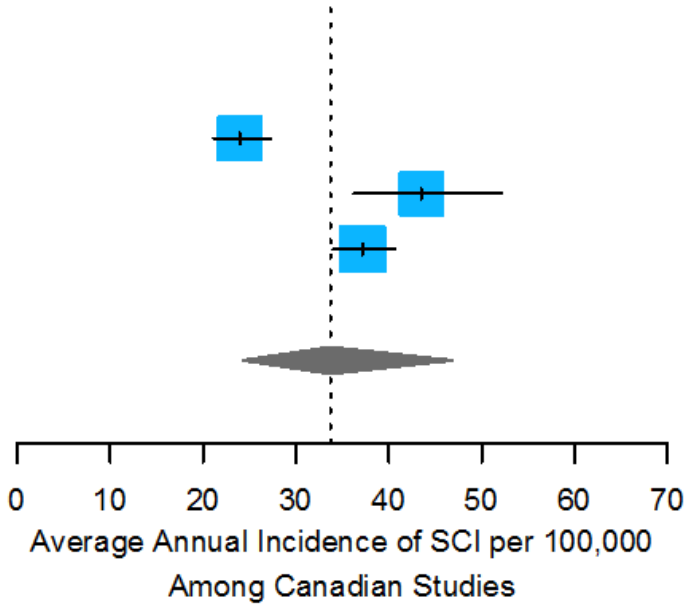
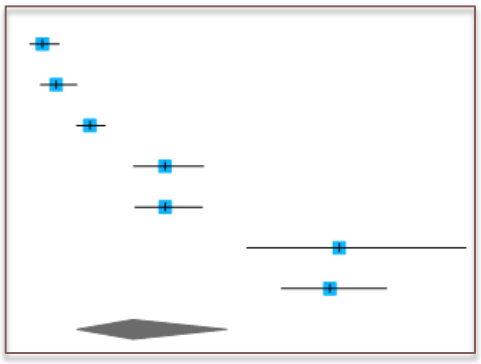


Figure 2: Forest plot of AAI among studies completed in Canada.

AAI in Canada (33.8) slightly higher than overall AAI internationally (28.3)

Average Annual Incidence of SCI per 100,000 by Sex

Study	Continent	Incidence	95% CI
Female			
Gur (2005)	Asia	5.66	[3.24; 9.90]
Ide (1993)	Asia	5.22	[2.81; 9.70]
Couris (2010)	North America	12.10	[9.40; 15.57]
Dryden (2003)	North America	23.80	[17.01; 33.31]
Price (1994)	North America	16.00	[10.84; 23.62]
Warren (1995)	North America	28.95	[14.35; 58.41]
Surkin (2000)	North America	28.70	[20.93; 39.36]
Pooled Totals		14.43	[9.24; 22.52]
Heterogeneity: I-squared=88.4%, Q=51.8, df=6, p<0.0001			
Male			
Gur (2005)	Asia	18.21	[13.44; 24.68]
Ide (1993)	Asia	23.48	[17.53; 31.44]
Couris (2010)	North America	36.30	[31.28; 42.12]
Dryden (2003)	North America	64.90	[52.93; 79.58]
Price (1994)	North America	65.00	[53.43; 79.07]
Warren (1995)	North America	131.19	[95.98; 179.32]
Surkin (2000)	North America	127.60	[109.12; 149.20]
Pooled Totals		52.66	[31.35; 88.47]
Heterogeneity: I-squared=97.7%, Q=256.9, df=6, p<0.0001			



Higher AAI in Males

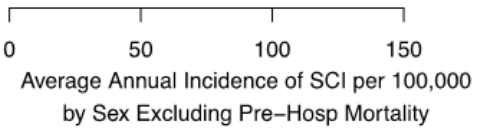
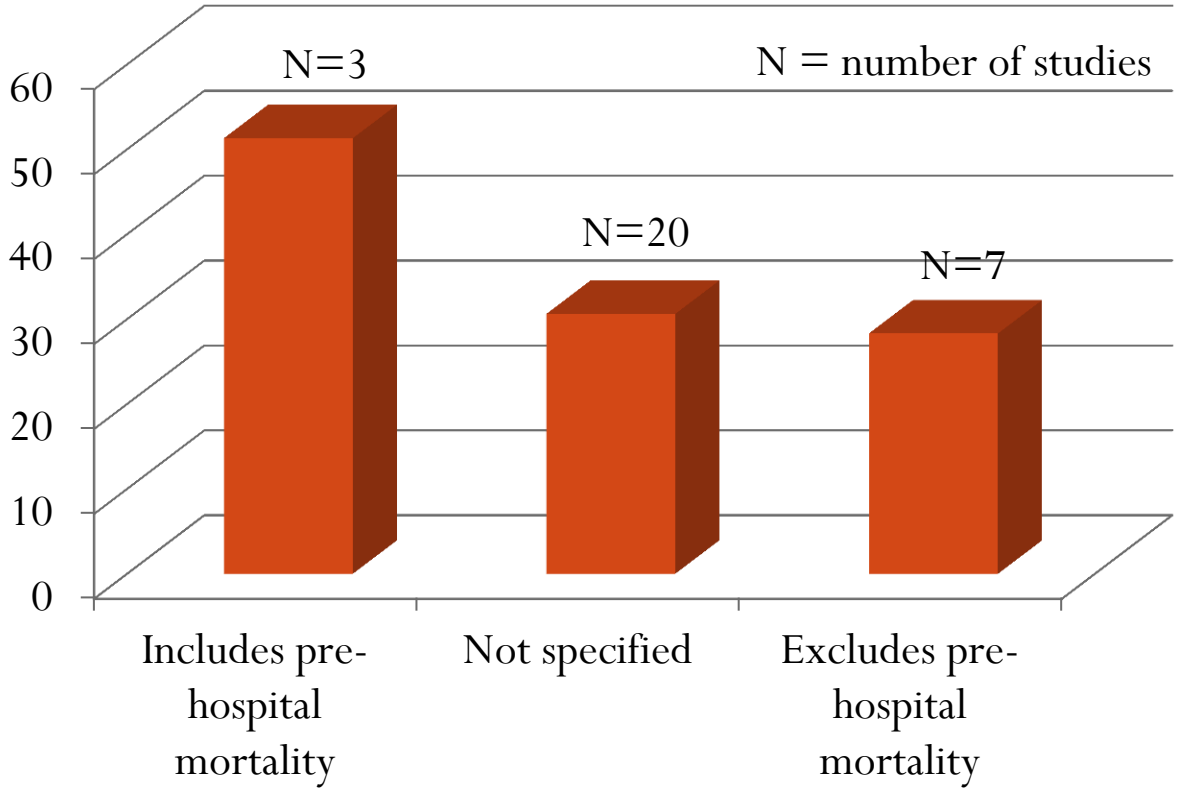


Figure 3: Forest plot of AAI stratified by male and female sex.

Spinal Cord Injury Average Annual Incidence per 100,000 Based on Inclusion of Pre-Hospital Mortality Cases or Not

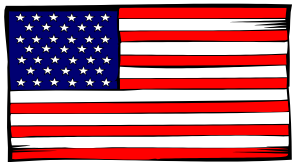


**there is an urgent need for prevalence studies of SCI nationally and internationally

Traumatic Brain Injuries

Results Prevalence TBI

- Only 2 prevalence studies
  - Australia and USA
  - Both report on all severities of TBI
  - 1999-2005
  - Prevalence
    - Australia: 1 per 100
    - USA: 0.02 per 1000

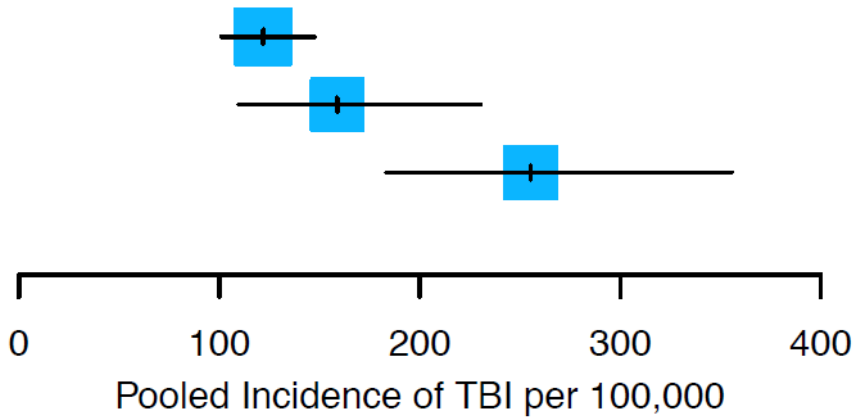


- Lifetime prevalence (Australia): 5.7 per 100

- Prevalence of TBI is higher in males than in females

Results Incidence TBI

Area	No. of Studies	Incidence
Europe	22	121.99
North America	24	158.82
Africa/Asia/South America/Australia	8	255.34

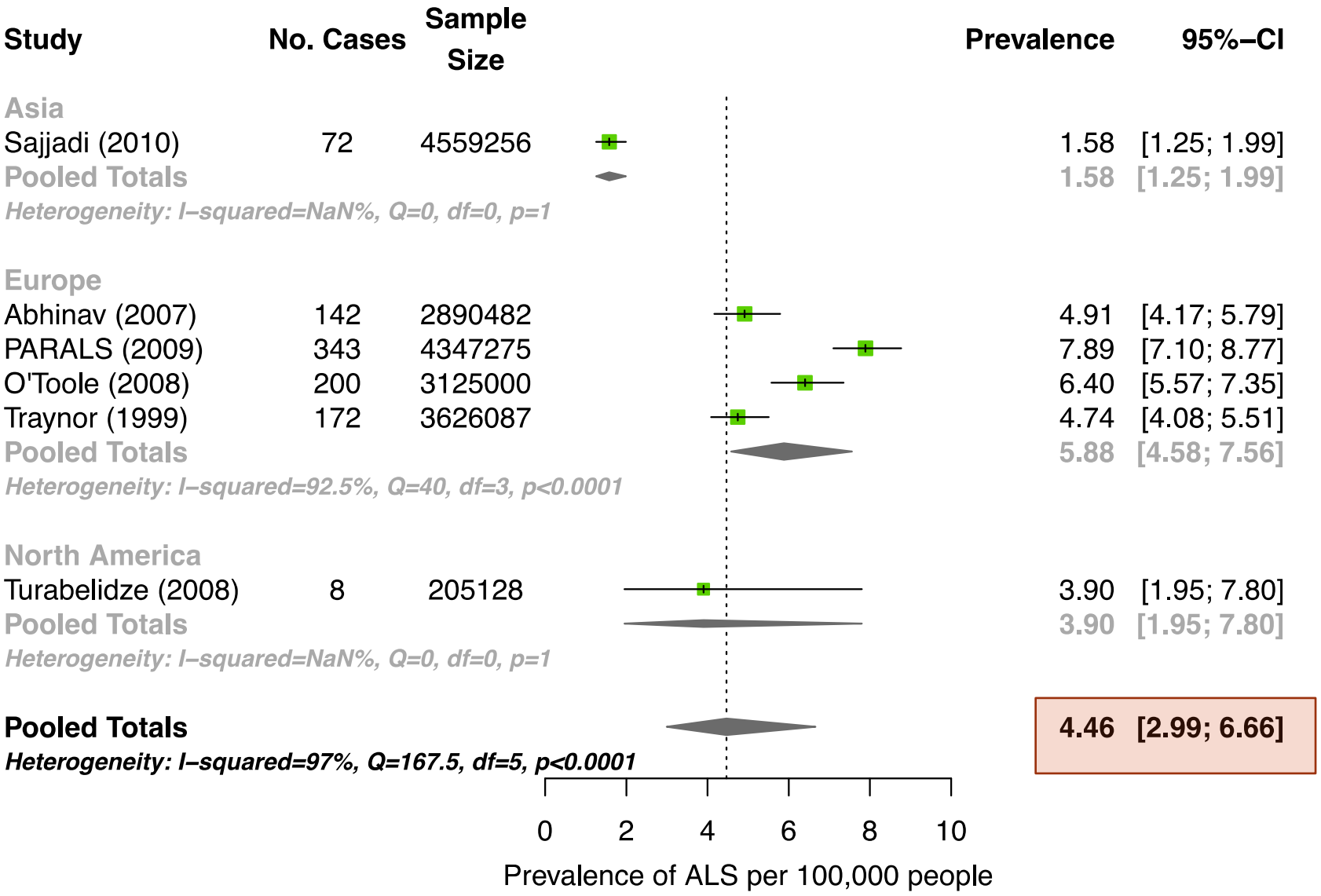


Traumatic Brain Injury Canadian Data

- 4 Canadian TBI studies (all incidence)
  - 2 focused on mild TBI (ON and national)
  - 1 looked at severe TBI (regional)
  - 1 looked at all types of TBI (national)
- Incidence highest in youngest and oldest age groups
- Incidence higher in males than females
- Overall incidence estimates ranged from 10.3 to 810 per 100,000 (e.g. year 2000)

Amyotrophic Lateral Sclerosis

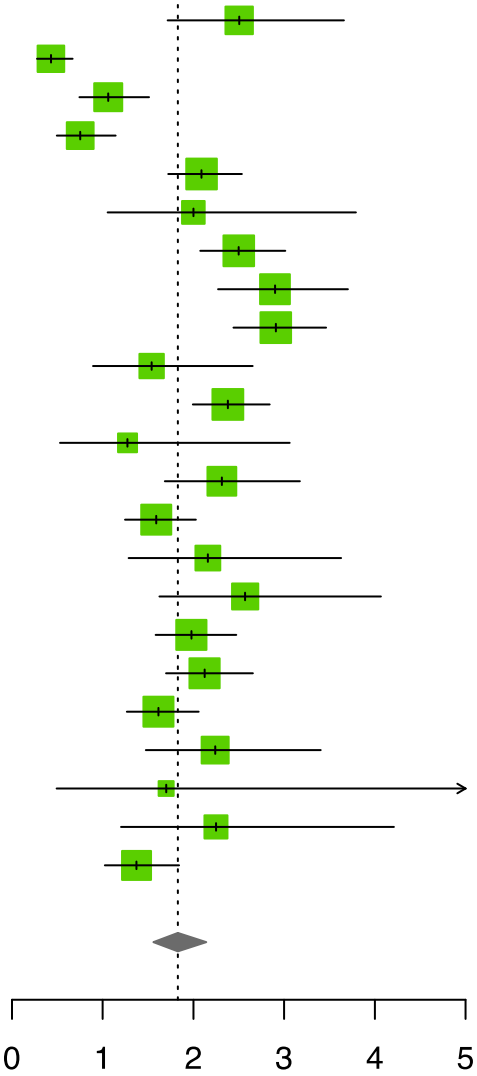
Prevalence ALS per 100,000 people



Average Annual Incidence ALS per 100,000 people

Study	Continent	No. Cases	Sample Size	Incidence	95%–CI
Kihira (2005)	Asia	27	1069912	2.50	[1.72; 3.66]
Sajjadi (2010)	Asia	20	4559256	0.43	[0.28; 0.67]
Abhinav (2007)	Europe	31	2890482	1.06	[0.74; 1.51]
Beghi (2001)	Europe	22	2922920	0.75	[0.50; 1.14]
Beghi (2007)	Europe	103	4947368	2.09	[1.72; 2.53]
Bonvicini (2008)	Europe	9	470000	2.00	[1.06; 3.79]
PARALS (2001)	Europe	110	4420000	2.50	[2.07; 3.01]
Chio (2008)	Europe	64	2218391	2.90	[2.27; 3.70]
PARALS (2009)	Europe	126	4332842	2.91	[2.44; 3.46]
Cima (2009)	Europe	13	844156	1.54	[0.89; 2.65]
Forbes (2007)	Europe	122	5142857	2.38	[1.99; 2.84]
Gross–Paju (1998	Europe	5	392427	1.27	[0.53; 3.06]
Imam (2010)	Europe	38	1659600	2.31	[1.69; 3.17]
Logroscino (2005)	Europe	65	4086613	1.59	[1.25; 2.03]
Mandrioli (2003)	Europe	14	662037	2.16	[1.29; 3.63]
Marin (2009)	Europe	18	710792	2.57	[1.63; 4.07]
O'Toole (2008)	Europe	78	3917203	1.98	[1.58; 2.47]
Traynor (1999)	Europe	77	3626087	2.12	[1.70; 2.65]
Zoccolella (2006)	Europe	65	4025329	1.61	[1.27; 2.06]
Bonaparte (2007)	North America	22	982143	2.24	[1.47; 3.40]
Sorenson (2007)	North America	2	147059	1.70	[0.49; 5.87]
Murphy (2008)	Oceania	10	434343	2.25	[1.20; 4.21]
Vasquez (2008)	South America	44	3241003	1.37	[1.02; 1.84]
Pooled Totals				**1.83**	**[1.56; 2.14]**

Heterogeneity: I–squared=83.6%, Q=133.9, df=22, p<0.0001



Canadian Data Motor Neuron Disease

- 2 Canadian studies
  - One on MND in Alberta (excluded as not ALS specific)
  - One of ALS in Nova Scotia

- Annual age-adjusted incidence of ALS in Nova Scotia:
  - 2.13 per 100,000 people between 2003-2004

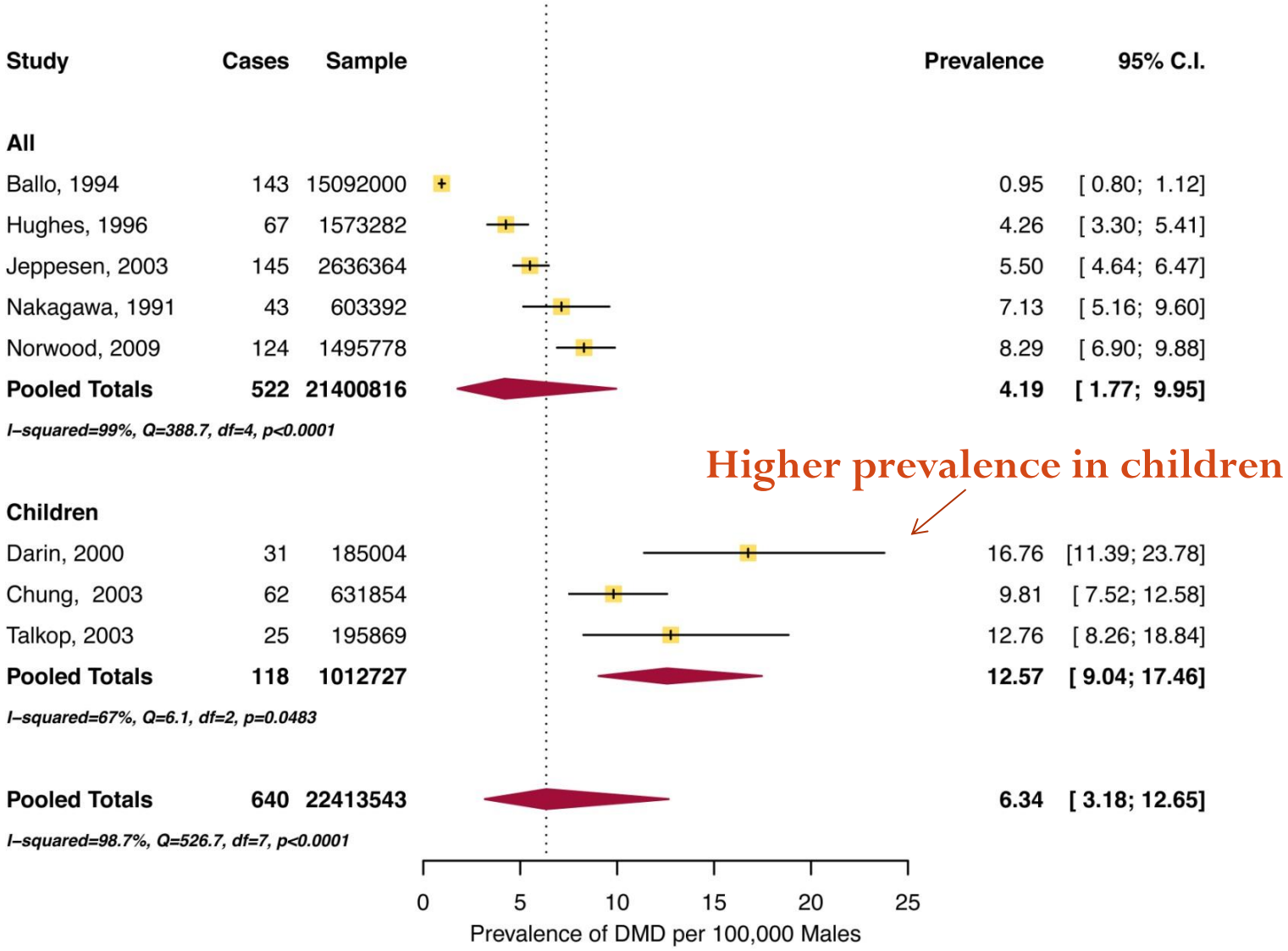
- ***Future research needed to estimate burden of ALS in Canada***

Muscular Dystrophy

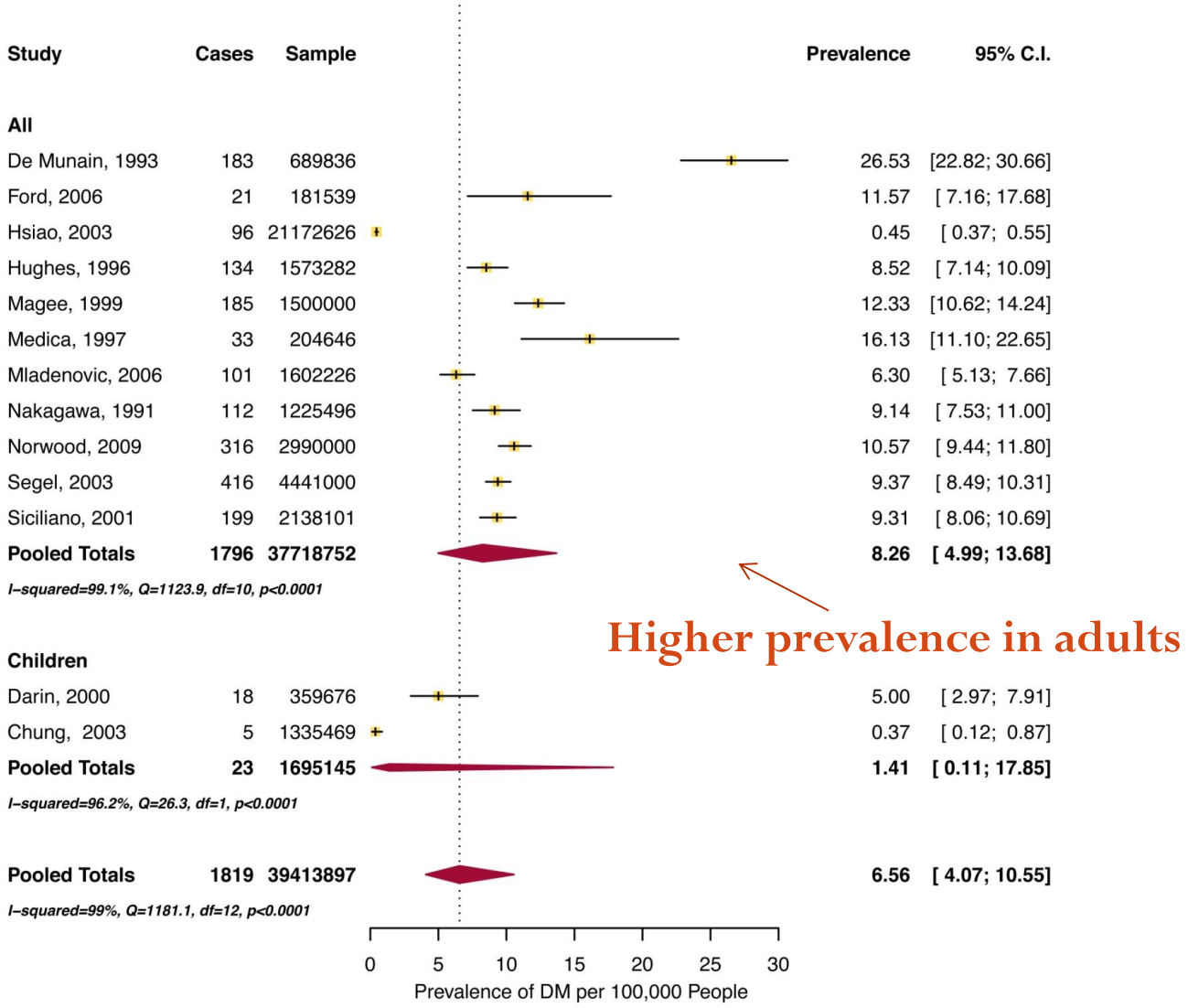
Overall Prevalence Estimates for Muscular Dystrophy per 100,000 people

Muscular dystrophy type	Overall Prevalence Estimate (95% CI)	Childhood Prevalence Estimate (95% CI)
Duchenne	4.2 (1.8-10.0)	-
Facioscapulohumeral	4.0 (2.9-5.4)	0.3 (0.0-3.0)
Type 1 myotonic	8.3 (5.0-13.7)	1.4 (0.1-17.9)
Limb girdle	1.6 (0.9-2.8)	0.5 (0.2-1.3)
Emery-Dreifuss	0.4 (0.1-1.3)	0.2 (0.1-0.7)
Congenital	1.0 (0.6-1.6)	0.8 (0.6-1.5)
Overall	20.9 (16.7-26.2)	-
		Childhood Prevalence Estimate in Boys (95% CI)
Duchenne	-	12.6 (9.0-17.5)
Becker	-	1.4 (0.8-2.5)

Prevalence of Duchenne Muscular Dystrophy per 100,000 males



Prevalence of Myotonic Dystrophy per 100,000 people

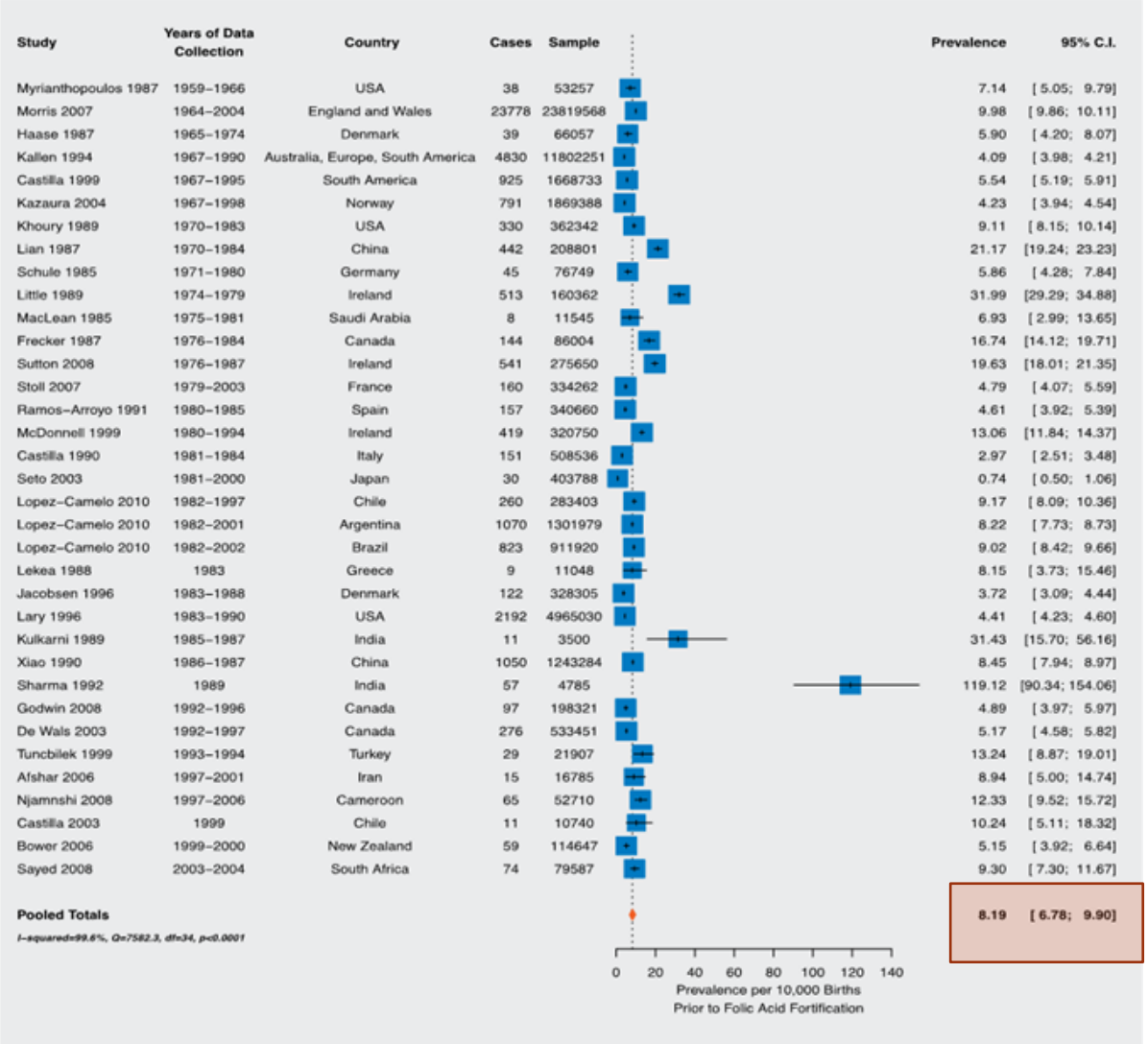


Higher prevalence in adults

Spina Bifida

Birth Prevalence of Spina Bifida per 10,000 Pregnancies *Prior to* Folic Acid Fortification

8.19 per 10,000



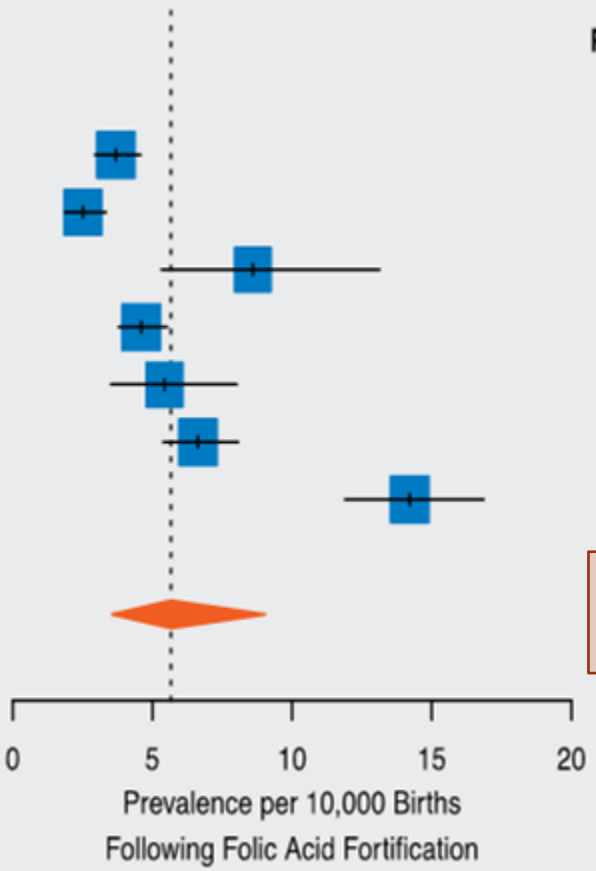
Study	Years of Data Collection	Country	Cases	Sample	Prevalence	95% C.I.
Myrianthopoulos 1987	1959–1966	USA	38	53257	7.14	[5.05; 9.79]
Morris 2007	1964–2004	England and Wales	23778	23819568	9.98	[9.86; 10.11]
Haase 1987	1965–1974	Denmark	39	66057	5.90	[4.20; 8.07]
Kallen 1994	1967–1990	Australia, Europe, South America	4830	11802251	4.09	[3.98; 4.21]
Castilla 1999	1967–1995	South America	925	1668733	5.54	[5.19; 5.91]
Kazaura 2004	1967–1998	Norway	791	1869388	4.23	[3.94; 4.54]
Khoury 1989	1970–1983	USA	330	362342	9.11	[8.15; 10.14]
Lian 1987	1970–1984	China	442	208801	21.17	[19.24; 23.23]
Schule 1985	1971–1980	Germany	45	76749	5.86	[4.28; 7.84]
Little 1989	1974–1979	Ireland	513	160362	31.99	[29.29; 34.88]
MacLean 1985	1975–1981	Saudi Arabia	8	11545	6.93	[2.99; 13.65]
Frecker 1987	1976–1984	Canada	144	86004	16.74	[14.12; 19.71]
Sutton 2008	1976–1987	Ireland	541	275650	19.63	[18.01; 21.35]
Stoll 2007	1979–2003	France	160	334262	4.79	[4.07; 5.59]
Ramos–Arroyo 1991	1980–1985	Spain	157	340660	4.61	[3.92; 5.39]
McDonnell 1999	1980–1994	Ireland	419	320750	13.06	[11.84; 14.37]
Castilla 1990	1981–1984	Italy	151	508536	2.97	[2.51; 3.48]
Seto 2003	1981–2000	Japan	30	403788	0.74	[0.50; 1.06]
Lopez–Camelo 2010	1982–1997	Chile	260	283403	9.17	[8.09; 10.36]
Lopez–Camelo 2010	1982–2001	Argentina	1070	1301979	8.22	[7.73; 8.73]
Lopez–Camelo 2010	1982–2002	Brazil	823	911920	9.02	[8.42; 9.66]
Lekea 1988	1983	Greece	9	11048	8.15	[3.73; 15.46]
Jacobsen 1996	1983–1988	Denmark	122	328305	3.72	[3.09; 4.44]
Lary 1996	1983–1990	USA	2192	4965030	4.41	[4.23; 4.60]
Kulkarni 1989	1985–1987	India	11	3500	31.43	[15.70; 56.16]
Xiao 1990	1986–1987	China	1050	1243284	8.45	[7.94; 8.97]
Sharma 1992	1989	India	57	4785	119.12	[90.34; 154.06]
Godwin 2008	1992–1996	Canada	97	198321	4.89	[3.97; 5.97]
De Wals 2003	1992–1997	Canada	276	533451	5.17	[4.58; 5.82]
Tuncbilek 1999	1993–1994	Turkey	29	21907	13.24	[8.87; 19.01]
Afshar 2006	1997–2001	Iran	15	16785	8.94	[5.00; 14.74]
Njamnshi 2008	1997–2006	Cameroon	65	52710	12.33	[9.52; 15.72]
Castilla 2003	1999	Chile	11	10740	10.24	[5.11; 18.32]
Bower 2006	1999–2000	New Zealand	59	114647	5.15	[3.92; 6.64]
Sayed 2008	2003–2004	South Africa	74	79587	9.30	[7.30; 11.67]
Pooled Totals					**8.19**	**[6.78; 9.90]**

I-squared=99.6%, Q=7582.3, df=34, p<0.0001

Birth Prevalence of Spina Bifida per 10,000 Births After Folic Acid Fortification

Study	Years of Data Collection	Country	Cases	Sample	Prevalence	95% C.I.
De Wals 2003	1998–2000	Canada	82	222190	3.69	[2.94; 4.58]
Godwin 2008	1999–2003	Canada	48	191028	2.51	[1.85; 3.33]
Castilla 2003	2000–2001	Chile	21	24439	8.59	[5.32; 13.13]
Lopez–Camelo 2010	2002–2007	Chile	112	243624	4.60	[3.79; 5.53]
Sayed 2008	2004–2005	South Africa	25	46021	5.43	[3.52; 8.02]
Lopez–Camelo 2010	2005–2007	Argentina	98	147853	6.63	[5.38; 8.08]
Lopez–Camelo 2010	2005–2007	Brazil	132	92843	14.22	[11.90; 16.86]
Pooled Totals					5.66	[3.55; 9.04]

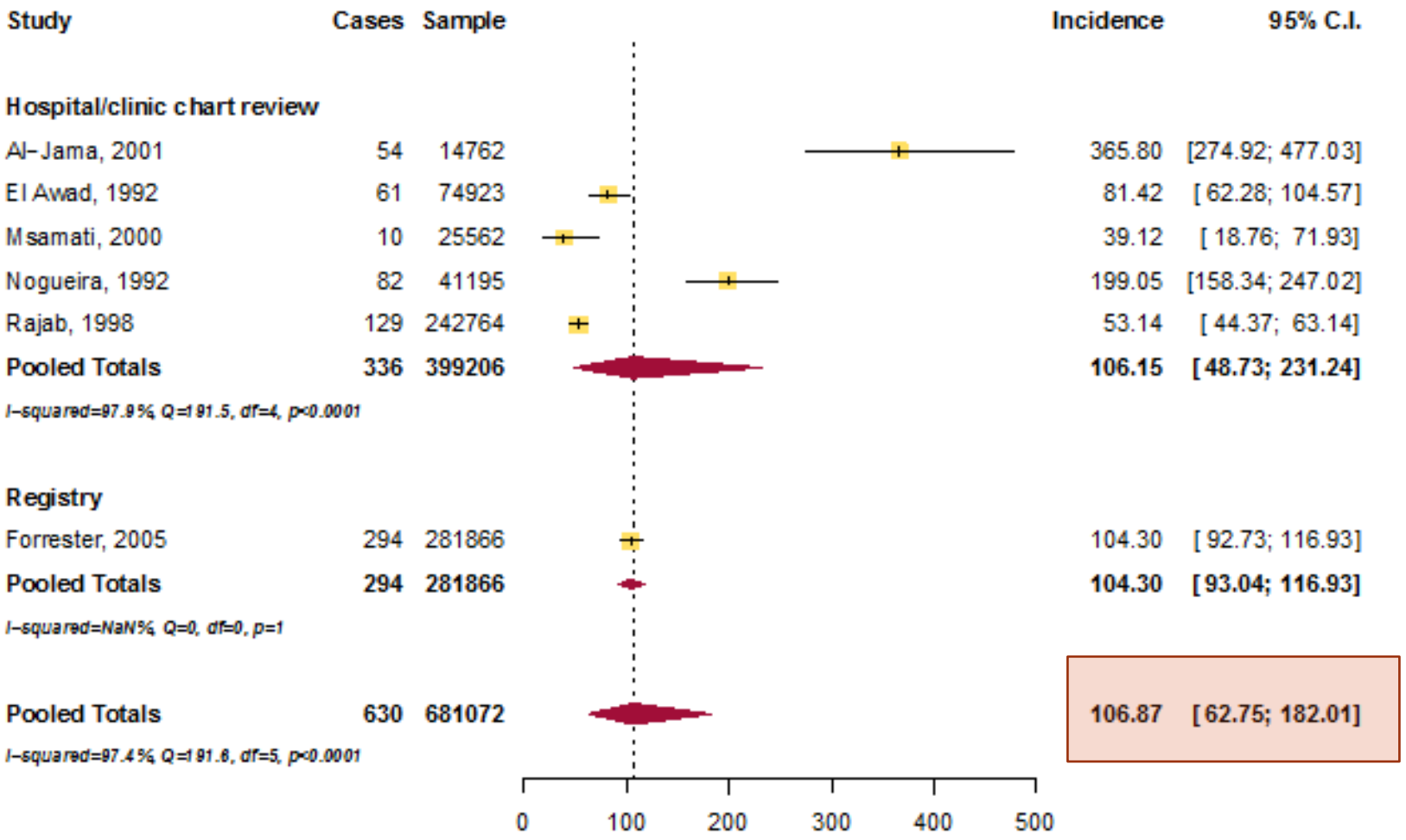
I–squared=96.4%, Q=165.4, df=6, p<0.0001



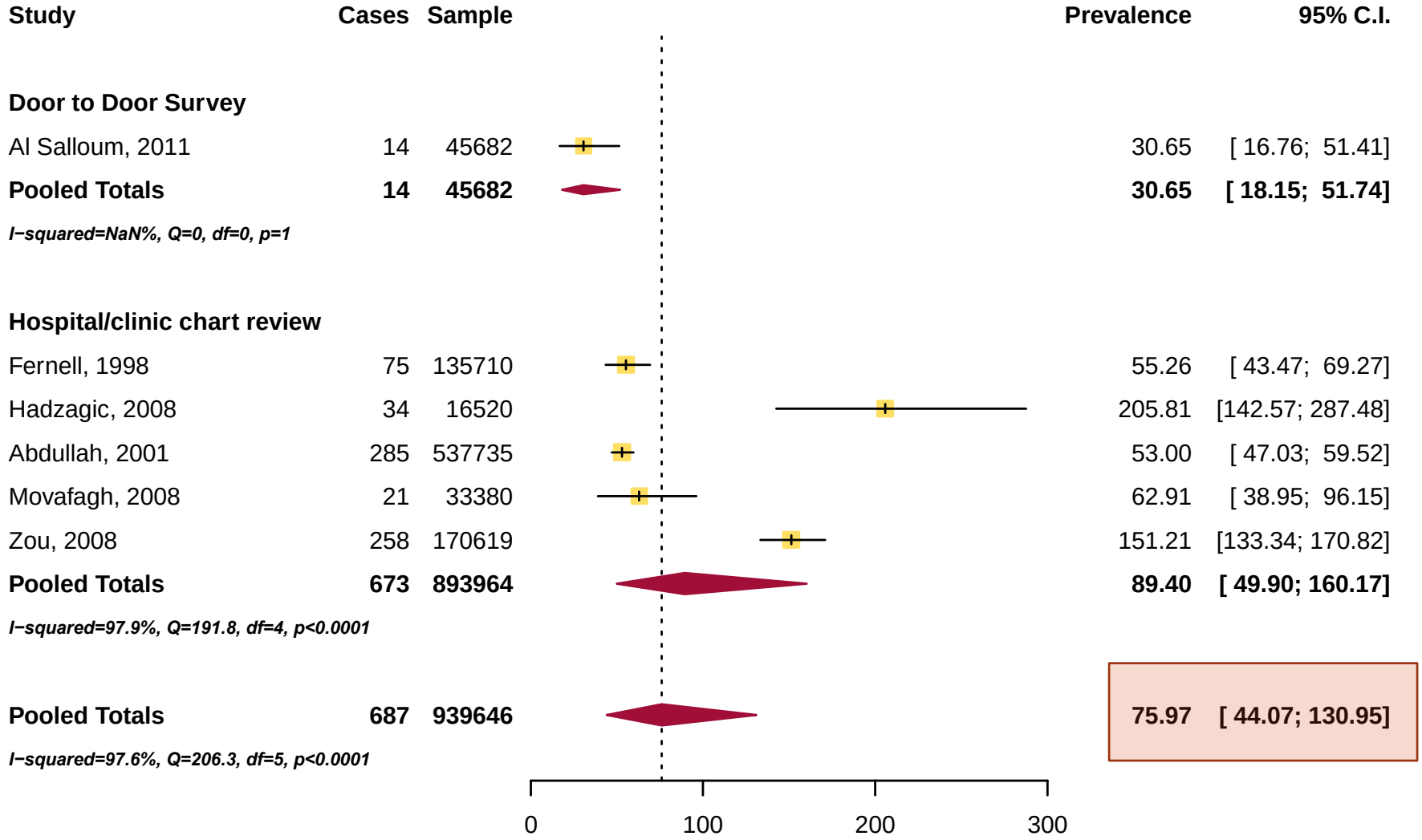
5.66 per 10,000

Hydrocephalus

Incidence of Hydrocephalus in Infants per 100,000 per year



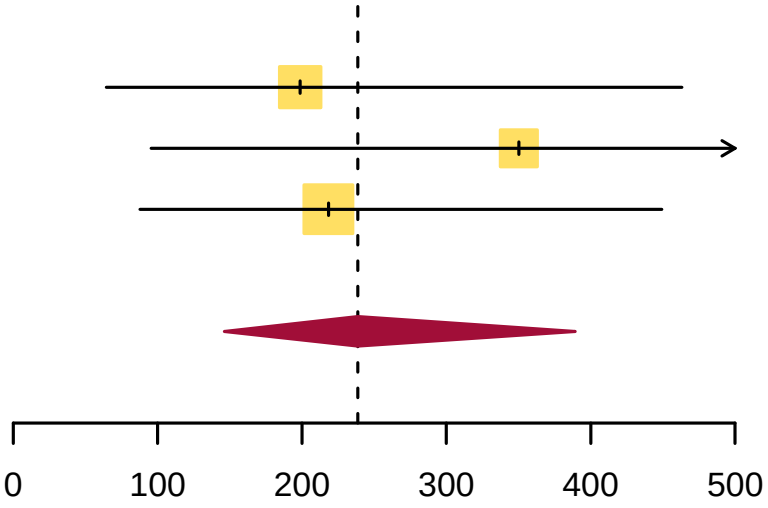
Prevalence of Hydrocephalus, Infants and Children per 100,000



Prevalence of Hydrocephalus, Elderly per 100,000

Study	Cases	Sample	Prevalence	95% C.I.
Hiraoka, 2008	5	2516	198.73	[64.56; 463.15]
Iseki, 2009	4	1142	350.26	[95.51; 894.36]
Tanaka, 2009	7	3207	218.27	[87.80; 449.20]
Pooled Totals	**16**	**6865**	**238.60**	**[146.26; 389.24]**

I−squared=0%, Q=0.8, df=2, p=0.6654



Tic Disorders

Pediatric Neurology 47 (2012) 77–90

Contents lists available at ScienceDirect

Pediatric Neurology

journal homepage: www.elsevier.com/locate/pnu

Review Article

Prevalence of Tic Disorders: A Systematic Review and Meta-Analysis

Tristan Knight [a], Thomas Steeves MD [b], Lundy Day BSc [c], Mark Lowerison MSc [c], Nathalie Jette MD [c,d], Tamara Pringsheim MD [c,e,*]

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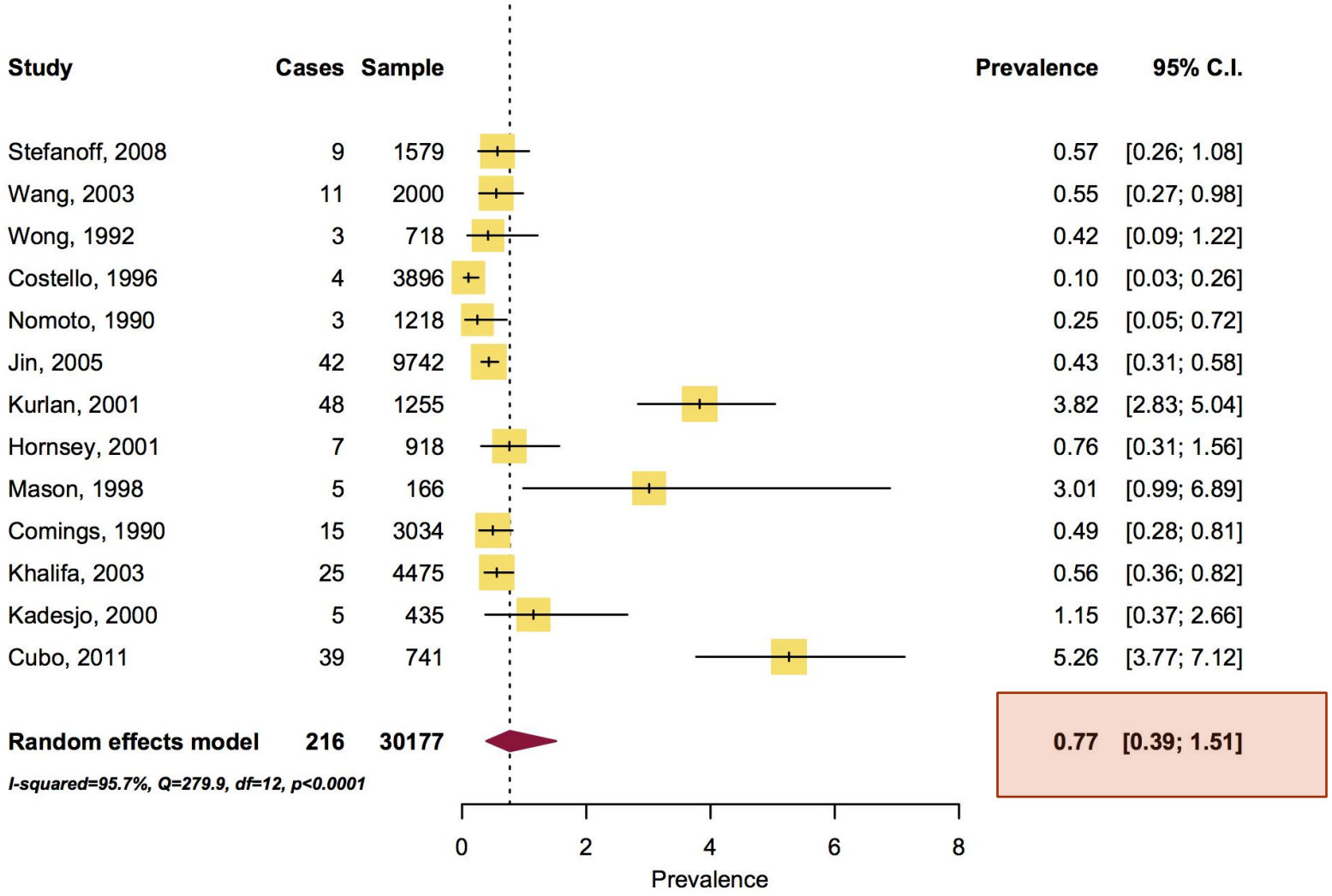
ARTICLE INFORMATION

Article history:
Received 23 March 2012
Accepted 9 May 2012

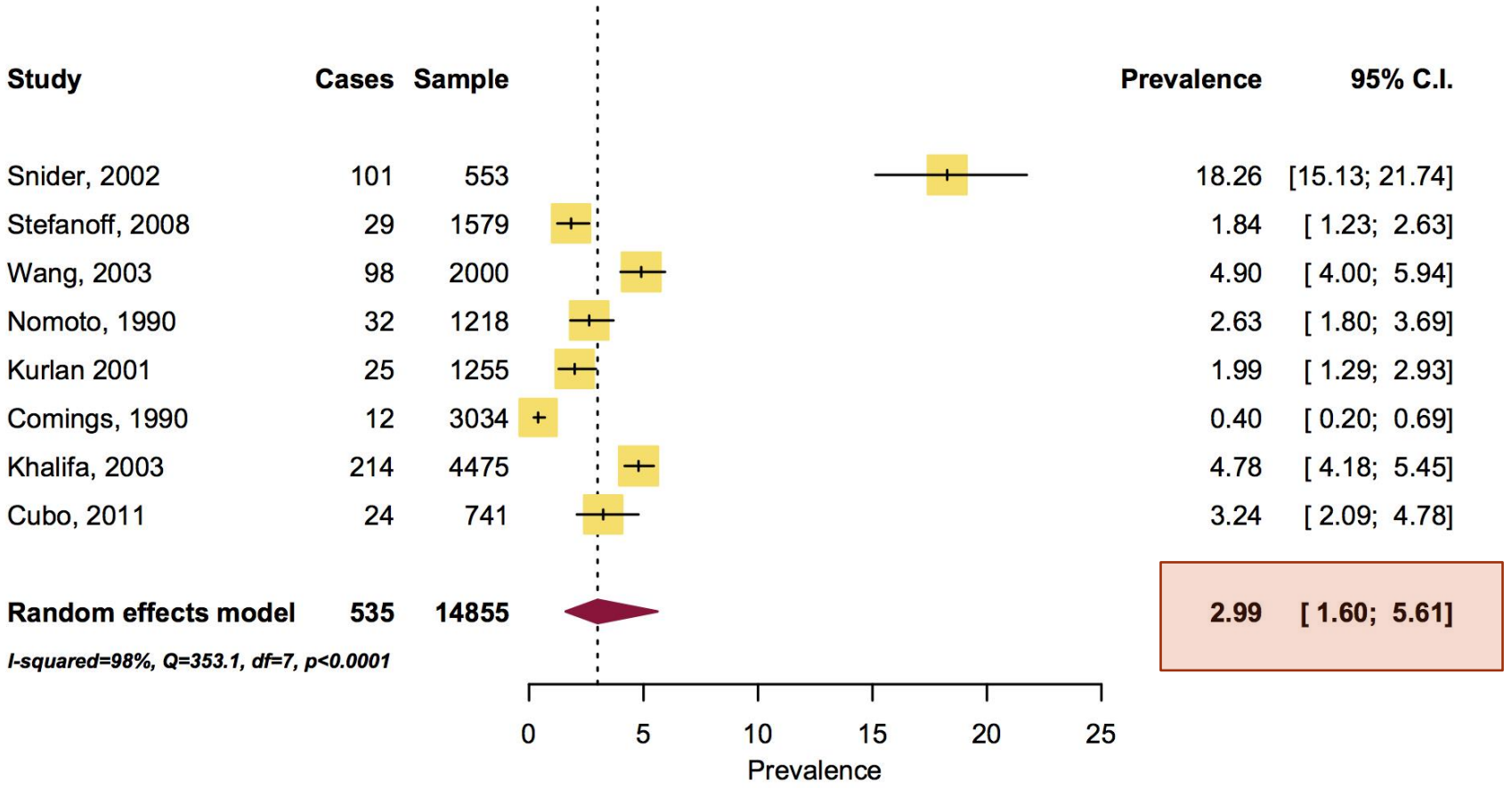
ABSTRACT

This study evaluated the prevalence of tic disorders. MEDLINE and EMBASE databases were searched, using terms specific to Tourette syndrome and tic disorders, for studies of incidence, prevalence, and epidemiology. Thirty-five studies reporting data from 1985-2011 on the incidence or prevalence of tic disorders in a defined population were included. One reported incidence, and 34 reported prevalence. Meta-analysis of 13 studies of children yielded a prevalence of Tourette syndrome at 0.77% (95% confidence interval, 0.39-1.51%). Prevalence is higher in boys: 1.06% of boys were affected (95% confidence interval, 0.54-2.09%) vs 0.25% of girls (95% confidence interval, 0.05-1.20%). Transient tic disorder comprised the most common tic disorder in children, affecting 2.99% (95% confidence interval, 1.60-5.61%). Meta-analysis of two studies assessing adults for Tourette syndrome revealed a prevalence of 0.05% (95% confidence interval, 0.03-0.08%). The prevalence of tic disorders was higher in all studies performed in special education populations. Tic disorders are more common in children than adults, in boys than girls, and in special education populations. Parents, educators, healthcare professionals, and administrators should be aware of the frequency with which tic disorders occur, and ensure proper access to appropriate care.

Prevalence of Tourette Syndrome in Children: School Based Studies →~1%



Prevalence of Transient Tic Disorder in Children: School Based Studies → ~3%



Prevalence (and Incidence) of Tic Disorders

- All studies in ***children show males more affected than females***
- Higher prevalence of tic disorders in special education populations
- Prevalence in adult populations
  - Tourette Syndrome: 5 per 10,000 in military recruits (compulsory exam on admittance)
  - All tic disorders: 42 per 10,000 in adults 50-89 (population based random sample)
- Only 1 study of incidence → ***Incidence studies needed***

Dystonia

RESEARCH ARTICLE

The Prevalence of Primary Dystonia: A Systematic Review and Meta-analysis

Thomas D. Steeves, MD, MSc,[1] Lundy Day, BSc,[2] Jonathan Dykeman, BSc,[2]
Nathalie Jette, MD, MSc,[2] and Tamara Pringsheim, MD, MSc[2]*

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[2]*Department of Clinical Neurosciences, University of Calgary, Calgary, Alberta, Canada*

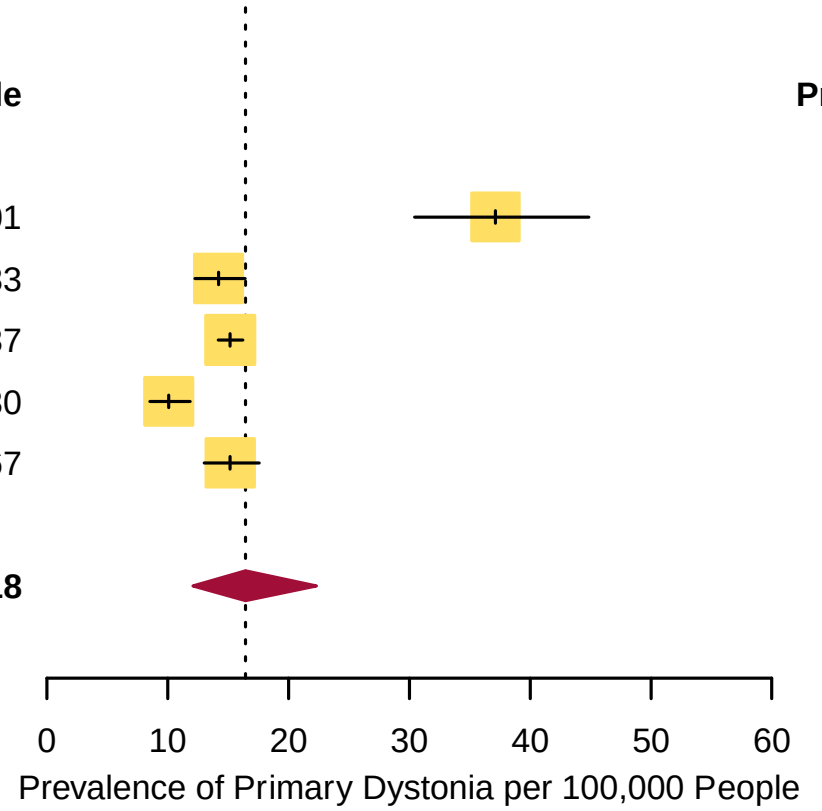
ABSTRACT: Dystonia is a hyperkinetic movement disorder characterized by sustained muscle contractions that produce repetitive movements and abnormal postures. Specific information on the prevalence of dystonia has been difficult to establish because the existing epidemiological studies of the condition have adopted different methodologies for case ascertainment, resulting in widely differing reported prevalence. Medline and Embase databases were searched using terms specific to dystonia for studies of incidence, prevalence, and epidemiology. All population-based studies reporting an incidence and/or prevalence of primary dystonia were included. Sixteen original studies were included in our systematic review. Fifteen studies reported the prevalence of dystonia, including 12 service-based and three population-based studies. We performed a meta-analysis on the results of the service-based studies, and were able to combine data on the prevalence of several dystonia subtypes. From these studies, we calculated an overall prevalence of primary dystonia of 16.43 per 100,000 (95% confidence interval [CI]: 12.09–22.32). The prevalence of dystonia reported in the three population-based studies appears higher than that reported in the service-based studies. Only 1 of the 16 studies reported an incidence of cervical dystonia. This corresponded to a corrected incidence estimate of 1.07 per 100,000 person-years (95% CI: 0.86–1.32). Despite numerous studies on the epidemiology of dystonia, attempting to determine an accurate prevalence of the condition for health services planning remains a significant challenge. Given the methodological limitations of the existing studies, our own prevalence estimate of primary dystonia likely underestimates the true prevalence of the condition.

Key Words: prevalence studies; incidence studies; dystonia; blepharospasm

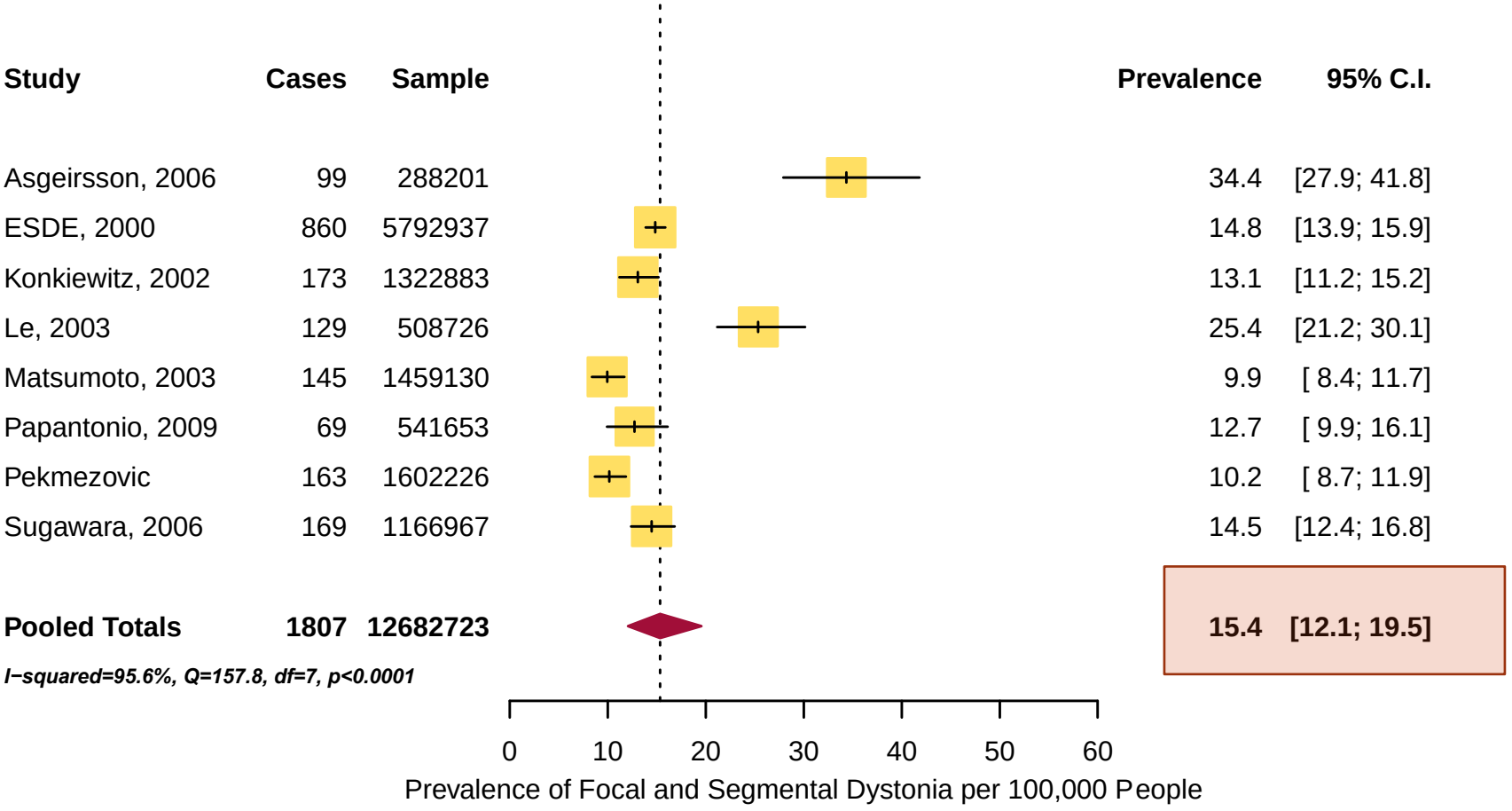
Prevalence of Primary Dystonia per 100,000 people

Study	Cases	Sample	Prevalence	95% C.I.
Asgeirsson, 2006	107	288201	37.1	[30.4; 44.9]
Konkiewitz, 2002	188	1322883	14.2	[12.3; 16.4]
ESDE, 2000	879	5792937	15.2	[14.2; 16.2]
Matsumoto, 2003	147	1459130	10.1	[8.5; 11.8]
Sugawara, 2006	177	1166967	15.2	[13.0; 17.6]
Pooled Totals	**1498**	**10030118**	**16.4**	**[12.1; 22.3]**

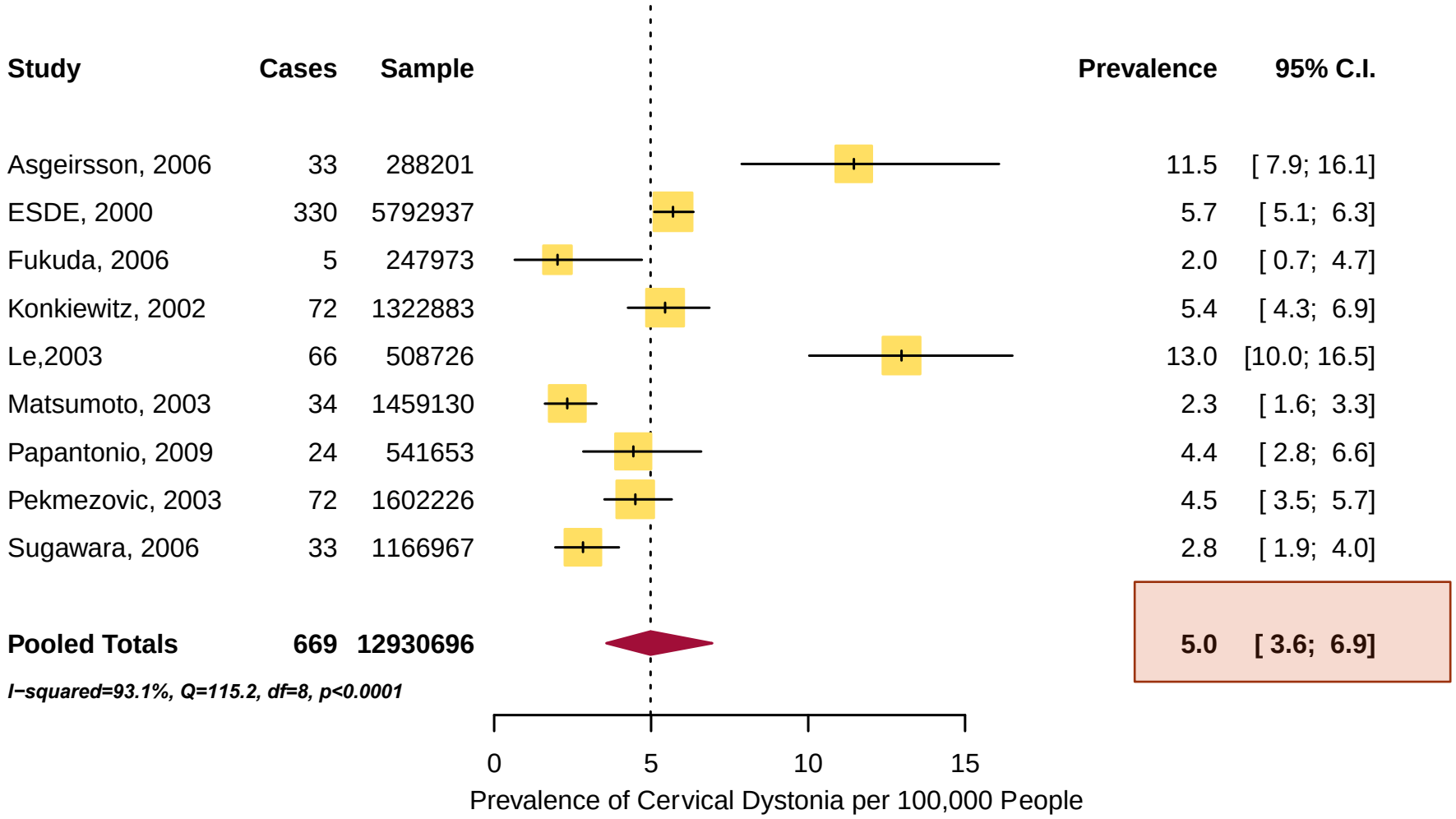
I−squared=96.4%, Q=110.8, df=4, p<0.0001



Prevalence of Focal and Segmental Dystonia per 100,000 people



Prevalence of Cervical Dystonia per 100,000 people



Results of Dystonia Incidence/Prevalence Systematic Review

- Higher estimates with population based studies compared to service-based studies
- No difference in prevalence between men and women
- ***Prevalence increases with age***
- Only 1 incidence study
- ***No Canadian dystonia studies***

Huntington Disease

REVIEW

CME

The Incidence and Prevalence of Huntington's Disease: A Systematic Review and Meta-analysis

Tamara Pringsheim, MD, MSc,[1]* Katie Wiltshire, MD,[1] Lundy Day, BSc,[1] Jonathan Dykeman, BSc,[1] Thomas Steeves, MD, MSc,[2] and Nathalie Jette, MD, MSc[1]

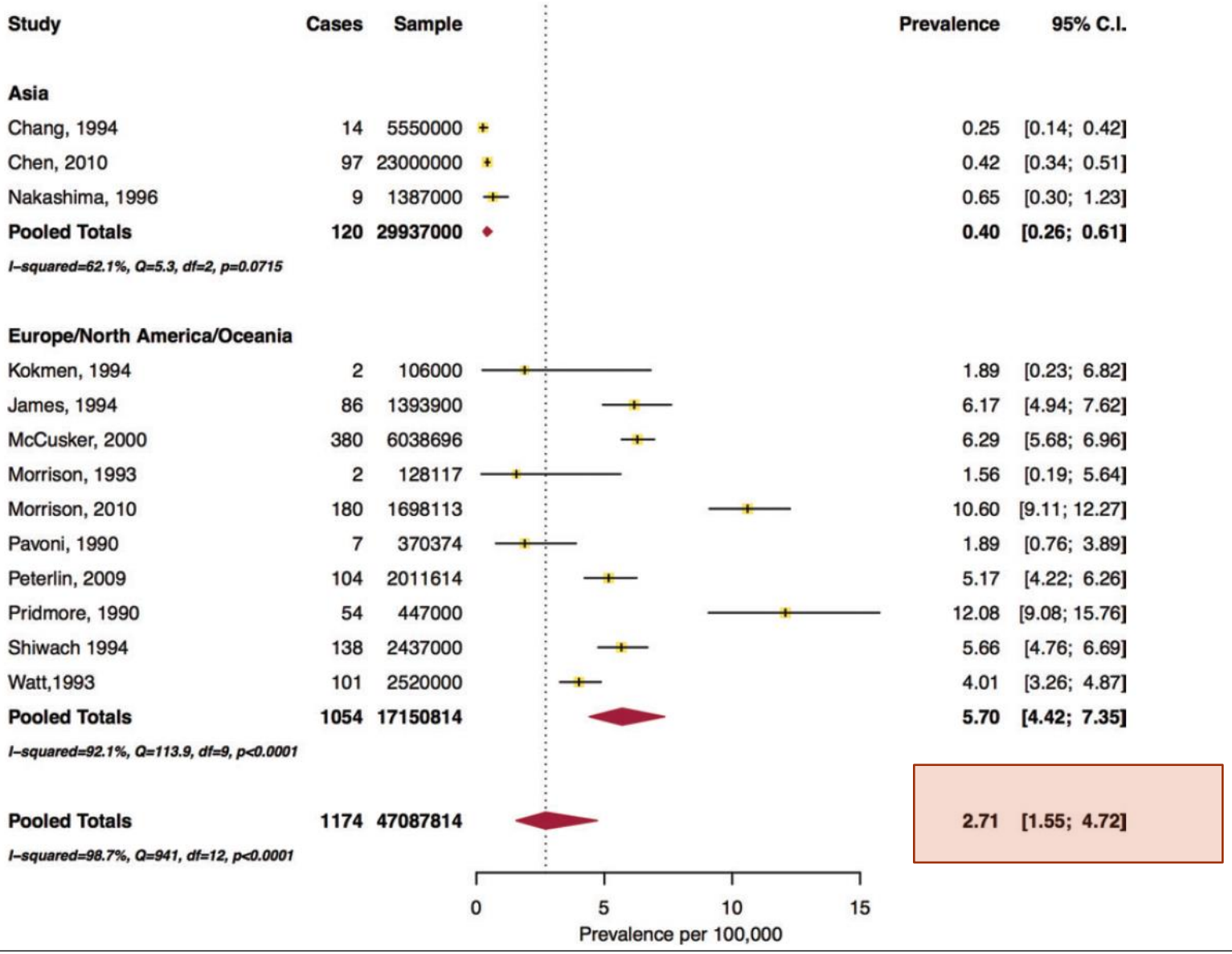
[1]*Department of Clinical Neurosciences, University of Calgary, Calgary, Alberta, Canada*
[2]*Department of Medicine, University of Toronto, Toronto, Ontario, Canada*

ABSTRACT: Huntington's disease (HD) is a rare, neurodegenerative disorder characterized by chorea, behavioral manifestations, and dementia. The aim of this study was to estimate the incidence and prevalence of HD through a systematic review of the literature. Medline and Embase databases were searched using terms specific to HD as well as studies of incidence, prevalence, and epidemiology. All studies reporting the incidence and/or prevalence of HD were included. Twenty original research articles were included. Eight studies examined incidence, and 17 studies examined prevalence. Meta-analysis of data from four incidence studies revealed an incidence of 0.38 per 100,000 per year (95% confidence interval [CI]: 0.16, 0.94). Lower incidence was reported in the Asian studies (n = 2), compared to the studies performed in Europe, North America, and Australia (n = 6). The worldwide service-based prevalence of HD, based on a meta-analysis (n = 13 studies), was 2.71 per 100,000 (95% CI: 1.55–4.72). Eleven studies were conducted in Europe, North American, and Australia, with an overall prevalence of 5.70 per 100,000 (95% CI: 4.42–7.35). Three studies were conducted in Asia, with an overall prevalence of 0.40 per 100,000 (95% CI: 0.26–0.61). Metaregression revealed a significantly lower prevalence of HD in Asia, compared to European, North American, and Australian populations. HD is a devastating neurodegenerative disorder with a higher prevalence in Europe, North America, and Australia than in Asia. The difference in prevalence of this genetic disorder can be largely explained by huntingtin gene haplotypes.

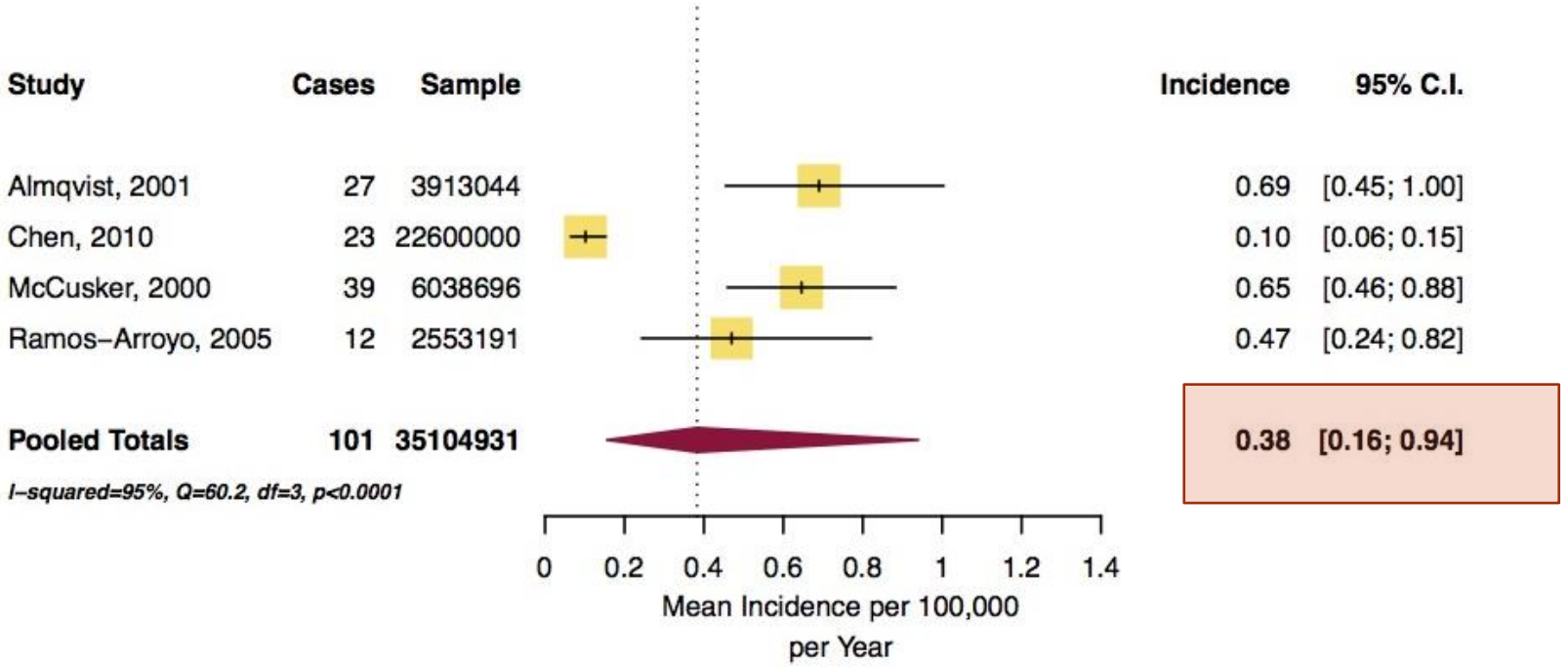
Key Words: Huntington's disease; epidemiology; incidence; prevalence

Movement Disorders, 2012

Prevalence of Huntington Disease per 100,000



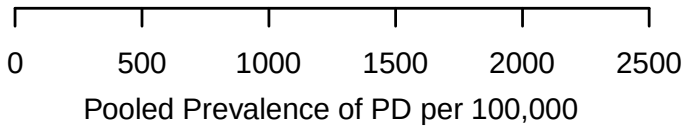
Incidence of Huntington Disease per 100,000 per year



Parkinson's Disease

Prevalence of Parkinson's Disease per 100,000 people

Age Group	No. Studies Combined	Total Sample	Prevalence	95%−CI	I2
40−49 years	5	21376	40.51	[20.26; 81.00]	0
50−59 years	13	399940	106.67	[53.88; 211.18]	85.4
55−64 years	4	26395	172.68	[87.82; 339.54]	74
60−69 years	15	276995	428.48	[235.45; 779.75]	95
65−74 years	5	22300	425.36	[192.67; 939.06]	89.3
70−79 years	20	153969	1086.54	[626.96; 1883.02]	97.4
80+ years	24	51584	1902.98	[1132.23; 3198.41]	95.9



Significant increase in prevalence with age

Brain Tumour

Canadian Brain Tumour Studies

- 7 Canadian incidence studies
  - AB, SK, MB, ON
  - Mostly of tumour subtypes

- Overall incidence in children (all brain tumours)
  - 3-4 per 100,000 person per year

- Overall incidence in all age groups (all brain tumours)
  - 11.1 per 100,000 person per year

Cerebral Palsy

DEVELOPMENTAL MEDICINE & CHILD NEUROLOGY REVIEW

An update on the prevalence of cerebral palsy: a systematic review and meta-analysis

MARYAM OSKOUI[1] | FRANZINA COUTINHO[2] | JONATHAN DYKEMAN[3] | NATHALIE JETTÉ[3] | TAMARA PRINGSHEIM[4]

1 Departments of Pediatrics and Neurology, McGill University, Montreal, Quebec; **2** School of Physical and Occupational Therapy, McGill University, Montreal, Quebec; **3** Department of Clinical Neurosciences and Hotchkiss Brain Institute, Department of Community Health Sciences and Institute of Public Health, University of Calgary, Calgary, Alberta; **4** Departments of Clinical Neurosciences and Pediatrics, University of Calgary, Calgary, Alberta, Canada.

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PUBLICATION DATA
Accepted for publication 22nd October 2012.
Published online 24th January 2013.

AIMS The aim of this study was to provide a comprehensive update on (1) the overall prevalence of cerebral palsy (CP); (2) the prevalence of CP in relation to birthweight; and (3) the prevalence of CP in relation to gestational age.

METHOD A systematic review and meta-analysis was conducted and reported, based on the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-analyses) statement. Population-based studies on the prevalence of CP in children born in 1985 or after were selected. Statistical analysis was carried out using computer package R, version 2.14.

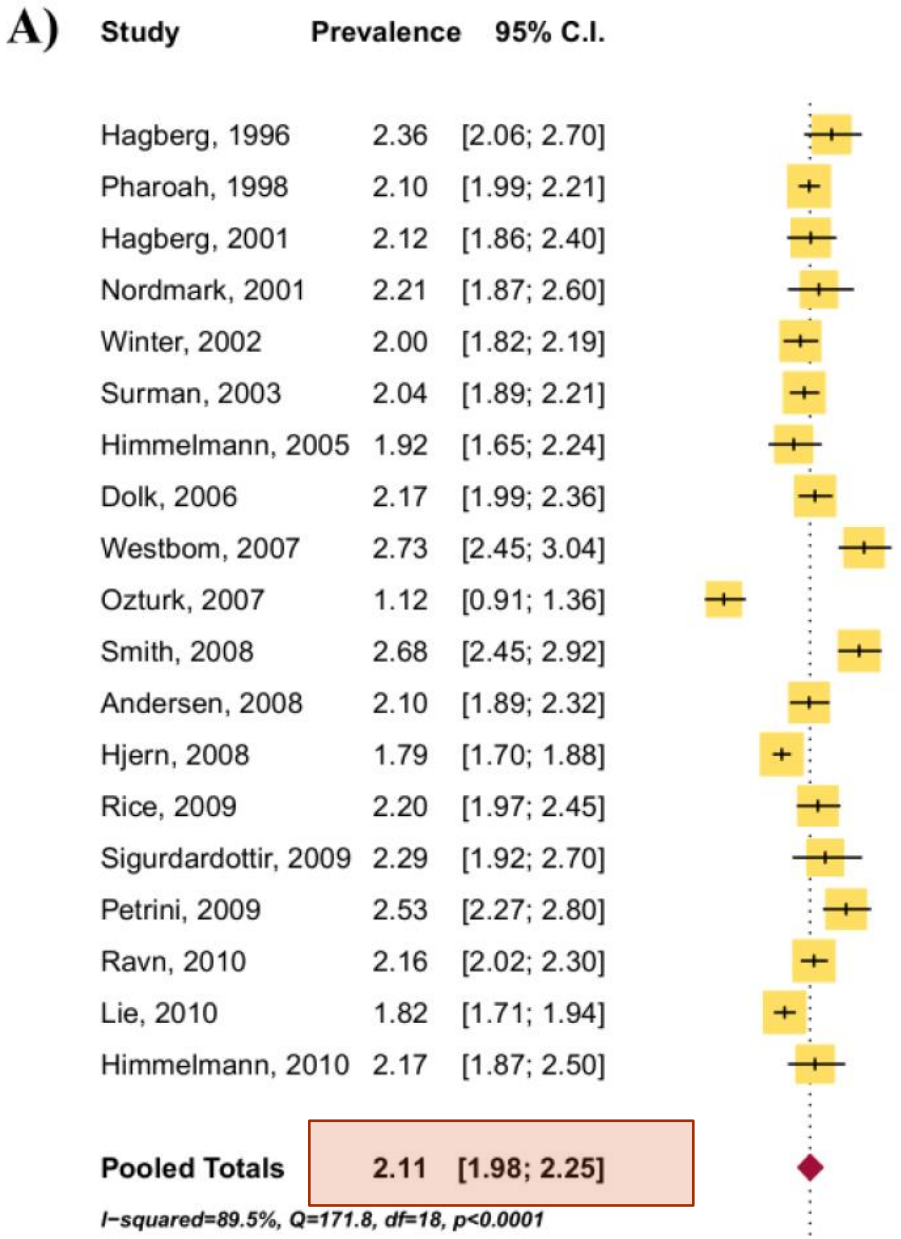
RESULTS A total of 49 studies were selected for this review. The pooled overall prevalence of CP was 2.11 per 1000 live births (95% confidence interval [CI] 1.98–2.25). The prevalence of CP stratified by gestational age group showed the highest pooled prevalence to be in children weighing 1000 to 1499g at birth (59.18 per 1000 live births; 95% CI 53.06–66.01), although there was no significant difference on pairwise meta-regression with children weighing less than 1000g. The prevalence of CP expressed by gestational age was highest in children born before 28 weeks' gestation (111.80 per 1000 live births; 95% CI 69.53–179.78; $p<0.0327$).

INTERPRETATION The overall prevalence of CP has remained constant in recent years despite increased survival of at-risk preterm infants.

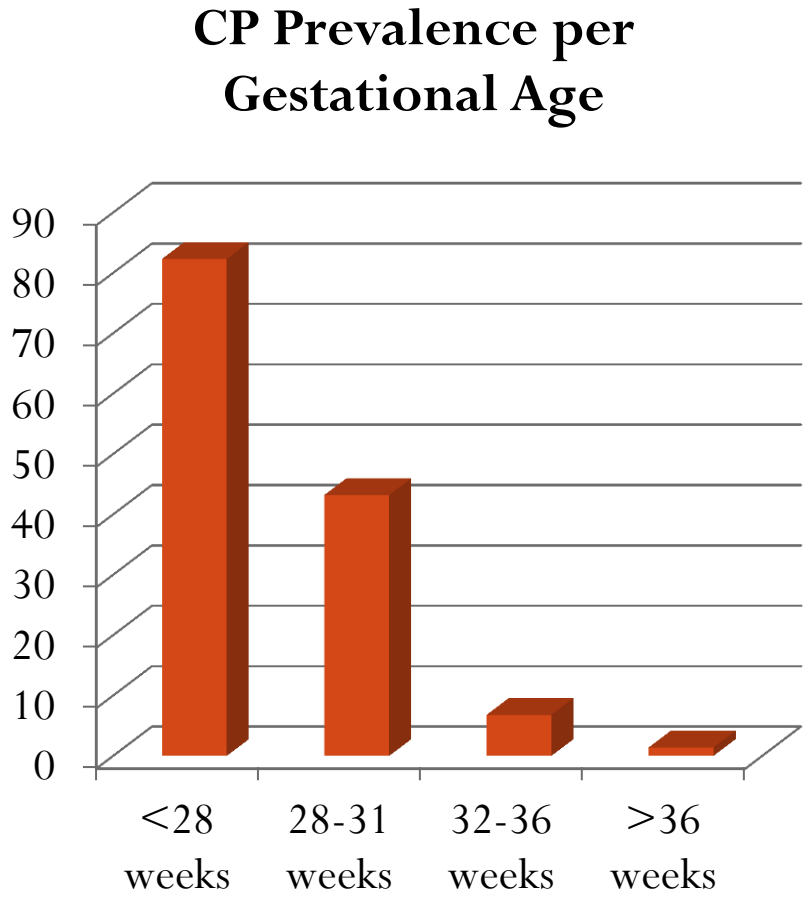
Overall Prevalence of CP
2.11 per 1,000 live births

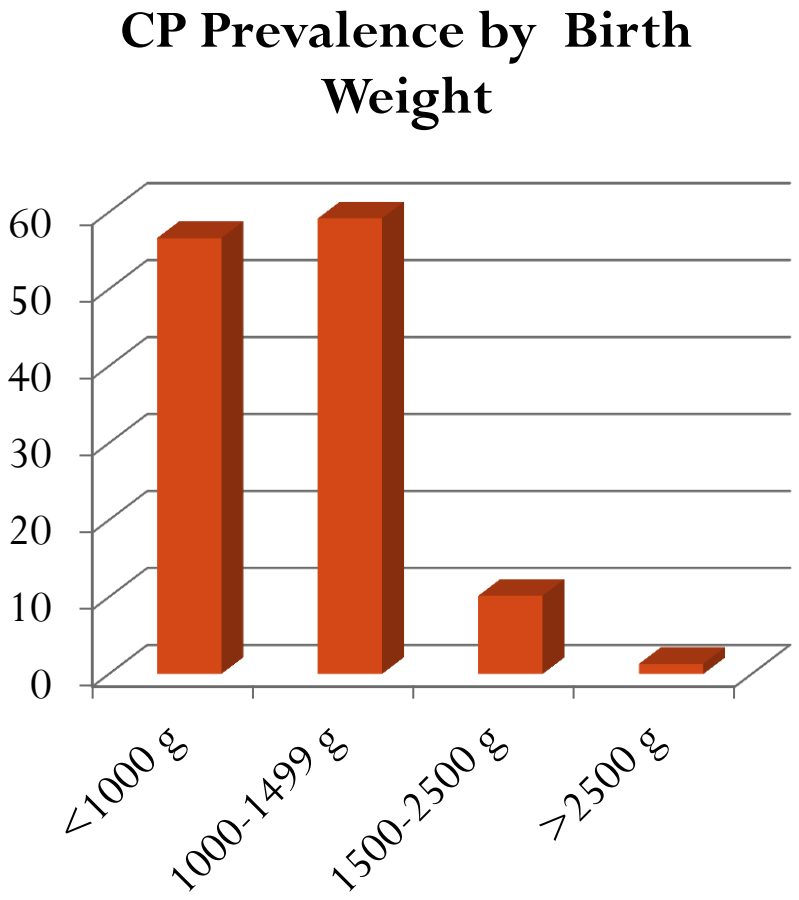
Overall prevalence has remained relatively stable over the past 15 years

No differences in prevalence for studies using administrative data vs. birth cohorts



Prevalence of Cerebral Palsy per 1,000 Live Births





Objective 2

- To summarize ascertainment sources for surveillance purposes for each of the priority conditions

1. Systematic Review of Validated Administrative Data Based Case Definition for Neurological Conditions

2. Summary of Ascertainment Sources from all of the Systematic Reviews of Incidence and Prevalence of each Conditions

Recommendations for optimal ICD codes to study neurologic conditions

A systematic review

Christine St. Germaine-Smith, MSc
Amy Metcalfe, MSc
Tamara Pringsheim, MD
Jodie Irene Roberts, BSc
Cynthia A. Beck, MD
Brenda R. Hemmelgarn, MD, PhD
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ABSTRACT

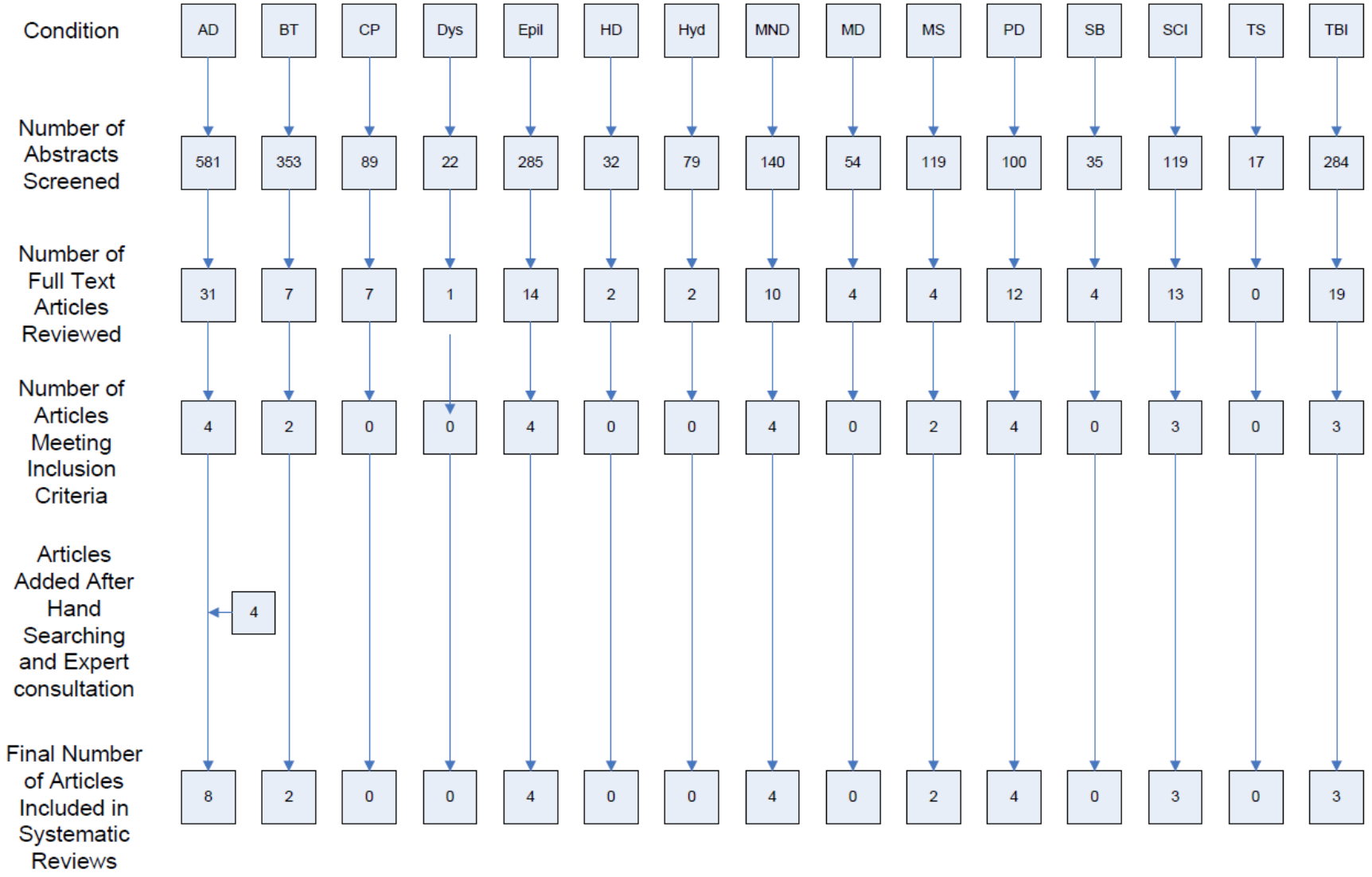
Objective: Administrative health data are frequently used for large population-based studies. However, the validity of these data for identifying neurologic conditions is uncertain.

Methods: This article systematically reviews the literature to assess the validity of administrative data for identifying patients with neurologic conditions. Two reviewers independently assessed for eligibility all abstracts and full-text articles identified through a systematic search of Medline and Embase. Study data were abstracted on a standardized abstraction form to identify ICD code–based case definitions and corresponding sensitivity, specificity, positive predictive values (PPVs), and negative predictive values (NPVs).

Results: Thirty full-text articles met the eligibility criteria. These included 8 studies for Alzheimer disease/dementia (sensitivity: 8–86.5, specificity: 56.3–100, PPV: 60–97.9, NPV: 68.0–98.9), 2 for brain tumor (sensitivity: 54.0–100, specificity: 97.0–99.0, PPV: 91.0–98.0), 4 for epilepsy (sensitivity: 98.8, specificity: 69.6, PPV: 62.0–100, NPV: 89.5–99.1), 4 for motor neuron disease (sensitivity: 78.9–93.0, specificity: 99.0–99.9, PPV: 38.0–90.0, NPV: 99), 2 for multiple sclerosis (sensitivity: 85–92.4, specificity: 55.9–92.6, PPV: 74.5–92.7, NPV: 70.8–91.9), 4 for Parkinson disease/parkinsonism (sensitivity: 18.7–100, specificity: 0–99.9, PPV: 38.6–81.0, NPV: 46.0), 3 for spinal cord injury (sensitivity: 0.9–90.6, specificity: 31.9–100, PPV: 27.3–100), and 3 for traumatic brain injury (sensitivity: 45.9–78.0 specificity: 97.8, PPV: 23.7–98.0, NPV: 99.2). No studies met eligibility criteria for cerebral palsy, dystonia, Huntington disease, hydrocephalus, muscular dystrophy, spina bifida, or Tourette syndrome.

Conclusions: To ensure the accurate interpretation of population-based studies with use of administrative health data, the accuracy of case definitions for neurologic conditions needs to be taken into consideration. *Neurology*® 2012;79:1-1

Figure 1: Flow diagram of article inclusion



AD=Alzheimer's Disease and Dementia; BT= Brain Tumors; CP= Cerebral Palsy; Dys=Dystonia; Epil=Epilepsy; HD=Huntington's Disease; Hyd=Hydrocephalus; MND=Motor Neuron Disease; MD=Muscular Dystrophy; MS=Multiple Sclerosis; PD=Parkinson's Disease; Sb=Spina Bifida; SCI=Spinal Cord Injury; TS=Tourette Syndrome; TBI=Traumatic Brain Injury

A listing of excluded articles and reasons for exclusion can be found in eTable3.

Systematic Review of Validated Case Definitions

Condition	# Studies	# Validations	Sensitivity	Specificity	Positive Predictive Value	Negative Predictive Value
Alzheimer's disease and dementia	8	21	8-86.5	56.3-100	60-97.9	68.0-98.9
Brain Tumors	2	4	54.0-100	97.0-99.0	91.0-98.0	None Reported
Epilepsy	4	13	98.8	69.6	62.0-100	89.5-99.1
Motor Neuron Disease (ALS)	4	6	78.9-93.0	99.0-99.9	38.0-90.0	99
Multiple Sclerosis	2	7	85-92.4	55.9-92.6	74.5-92.7	70.8-91.9
Parkinson's Disease	4	16	18.7-100	0-99.9	38.6-81.0	46.0
Spinal Cord Injury	3	19	0.9-90.6	31.9-100	27.3-100	None Reported
Traumatic Brain Injury	3	8	45.9-78.0	97.8	23.7-98.0	99.2

- Data available for only 8 of 15 conditions
- Significant gaps identified (especially for ICD-10 coded databases)
- ***Excellent coding accuracy in general for epilepsy, motor neuron disease (incl. ALS) and multiple sclerosis***
- Accuracy for Alzheimer's disease and other dementias, Parkinson's disease, spinal cord injury and traumatic brain injury were more varied and at times less accurate

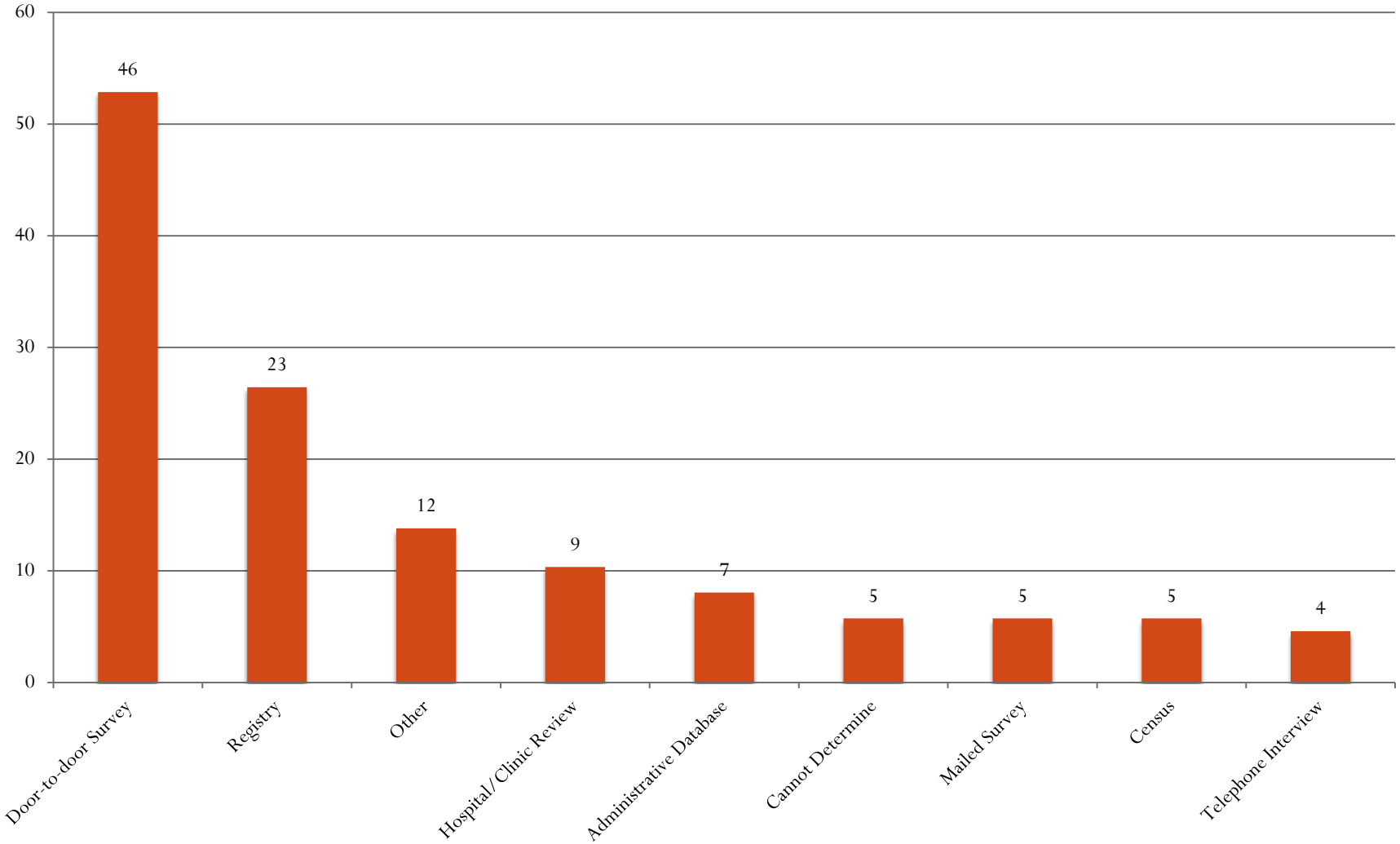
Objective 2

- To summarize ascertainment sources for surveillance purposes for each of the priority conditions

1. Systematic Review of Validated Administrative Data Based Case Definition for Neurological Conditions

2. Summary of Ascertainment Sources from all of the Systematic Reviews of Incidence and Prevalence of each Conditions

Sources of ascertainment used in Alzheimer's disease incidence/prevalence studies



CONDITION	MOST COMMON SOURCES OF ASCERTAINEMENT
Alzheimer's disease and other dementia	Door to door surveys, registry, "other", administrative databases
Amyotrophic lateral sclerosis	Administrative data, hospital/clinic chart review, other, registry
Brain tumour	Registry, hospital/clinic chart review, administrative databases
Cerebral palsy	Prospective birth cohorts, administrative databases
Dystonia	Multiple sources, surveys, hospital/clinic chart review
Epilepsy	Door to door survey, "other", hospital/clinic chart review
Huntington disease	Multiple sources, administrative databases, hospital/clinic chart review
Hydrocephalus	Hospital/clinic chart review, registry, other
Multiple sclerosis	"Other", Hospital/clinic chart review, and administrative data
Muscular dystrophy	Hospital/clinic chart review, "other", mailed survey, registry
Parkinson's disease	Door to door survey, multiple sources, hospital/clinic chart review
Spina bifida	Registry, hospital/clinic chart review, administrative databases
Spinal cord injury	Administrative databases, registry, "other"
Tourette syndrome	Schools, hospital/clinic chart review, mailed survey
Traumatic brain injury	"other", administrative databases, registry, hospital/clinic chart review

Objective 3: To develop an inventory of existing neurological registries in Canada and other developed countries

Summary of International and National Neurological Registries that Document Incidence and/or Prevalence Data

Condition	AD	BT	CP	Dys	Epil	HD	Hyd	MND	MD	MS	PD	SB	SCI	TS	TBI
Number of registries identified	12	307	36	0	5	3	34	15	9	15	3	37	22	1	71
Number of CDN registries (national, provincial or regional)	1	16	2	0	1	0	3	1	0	2	1	3	1	0	0
Number of Canadian registries which are national	0	2	1	0	0	0	1	0	0	0	0	1	1	0	0

AD=Alzheimer's Disease and Dementia; BT=Brain Tumors; CP=Cerebral Palsy; Dys=Dystonia; Epil=Epilepsy; HD=Huntington's Disease; Hyd=Hydrocephalus; MND=Motor Neuron Disease; MD=Muscular Dystrophy; MS=Multiple Sclerosis; PD=Parkinson's Disease; SB=Spina Bifida; SCI=Spinal Cord Injury; TS=Tourette Syndrome; TBI=Traumatic Brain Injury

CONDITION	CDN	NAME OF REGISTRY
Alzheimer + other dementia	No	CJD only (PHAC); Coming soon: CPCSSN
Amyotrophic lateral sclerosis	No	Canadian Neuromuscular Disease Registry (coming soon)
Brain tumour	Yes	Cancer surveillance programs (nationally and provincially)
Cerebral palsy	Yes	Canadian Multi-Regional Cerebral Palsy Registry (+ PQ registry)
Dystonia	No	-
Epilepsy	No	Coming soon: CPCSSN
Huntington disease	No	-
Hydrocephalus	Yes	As part of congenital anomaly registries (nationally and provincially)
Multiple sclerosis	Yes	Dalhousie U.
Muscular dystrophy	No	-
Parkinson's disease	No	Coming soon: CPCSSN
Spina bifida	Yes	Congenital disease surveillance programs (nationally and provincially)
Spinal cord injury	Yes	Rick Hansen Spinal Cord Injury Registry (national) – coming soon! Vertebase – Vancouver General Hospital Spinal Database
Tourette syndrome	Yes	TIC Database- global registry based in Canada but no incidence/prevalence data provided
Traumatic brain injury	No	-

Objective 4

- To make recommendations regarding the best sources of ascertainment for neurological disease surveillance

TECHNICAL REPORT
FOR THE PUBLIC HEALTH AGENCY OF CANADA AND THE
NEUROLOGICAL HEALTH CHARITIES OF CANADA

Recommendations: Best sources of ascertainment for neurological conditions for surveillance purposes

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For recommendations, see technical report for PHAC and the NHCC – April 2013

Thank you for your attention

- This study was part of the National Population Study of Neurological Conditions. We wish to acknowledge the membership of Neurological Health Charities of Canada and the Public Health Agency of Canada for their contribution to the success of this initiative.

- Funding for this study was largely provided by the Public Health Agency of Canada. The opinions expressed in this publication are those of the authors/researchers, and do not necessarily reflect the official views of the Public Health Agency of Canada.

- N. Jette holds a Canada Research Chair in Neurological Health Services Research and an Alberta Innovates Health Solutions Population Health Investigator Award