Trends in the Epidemiology of Cerebral Palsy

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Centers for Disease Control and Prevention
Presentation Overview

- History of definition and classification of cerebral palsy
- Epidemiology of Cerebral Palsy
- CP and co-occurring developmental disabilities
  - MADDSP 2004
- ADDM Network period prevalence 2002-2004
- Use of surveillance data for etiologic research
- Summary
History of Definition and Classification of CP
William John Little described 47 children with spastic rigidity: 1) hemiplegic rigidity (one side only); 2) paraplegia (legs more than arms); 3) generalized rigidity.

Sir William Osler coined the term CP; described cases as 1) infantile hemiplegia; 2) bilateral spastic hemiplegia or 3) spastic paraplegia.

Sigmund Freud devised classification scheme with ‘diplegia’ used to refer to all bilateral disorders of central origin. Disagreed with Little on the etiology of CP. Three groups based on possible etiologies: 1) maternal and idiopathic congenital; 2) perinatal; 3) postnatal.

Little Club convened by Mac Keith and Polani.

Bax: Definition redefined but continued recognition of inconsistencies led to conclusion that “it is impossible to proceed definitely with classifying cerebral palsy.” (Bax, 1964)
Definition by Mutch et. al., “an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of development.”

Palisano et al. developed Gross Motor Function Classification System in response to a need to classify the severity of motor disability in children with CP.

Surveillance of Cerebral Palsy in Europe published standardized procedures for ascertaining and describing children with CP for registers. Definition: reiteration of Mutch et. al.

An International Workshop on Definition and Classification of CP was held in Bethesda, Maryland, July 11-13 2004

Manual Ability Classification System described for upper extremity functioning.
Cerebral Palsy Definition

- A group of permanent disorders of the development of movement and posture that are attributed to non-progressive disturbances that occurred in the developing brain.
- Often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour; by epilepsy; and by secondary musculoskeletal problems.*


<table>
<thead>
<tr>
<th>Developmental Disability</th>
<th>Total Lifetime Costs for Incident Cohort (in millions)</th>
<th>Per-person Lifetime Costs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mental Retardation</td>
<td>$51,237</td>
<td>$1,014,000</td>
</tr>
<tr>
<td>Cerebral Palsy</td>
<td>$11,470</td>
<td>$921,000</td>
</tr>
<tr>
<td>Vision Impairment</td>
<td>$2,102</td>
<td>$417,000</td>
</tr>
<tr>
<td>Hearing Loss</td>
<td>$2,484</td>
<td>$566,000</td>
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</tbody>
</table>

## Prevalence of CP in Population Studies

<table>
<thead>
<tr>
<th>Study</th>
<th>Denominator Population</th>
<th>Rate / 1000</th>
</tr>
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<tbody>
<tr>
<td>Hagberg; Sweden, 2001</td>
<td>Cross-sectional; live births (LB)</td>
<td>2.1</td>
</tr>
<tr>
<td>Johnson, Europe, 2002</td>
<td>SCPE birth cohorts, live births</td>
<td>2.1</td>
</tr>
<tr>
<td>Winter; US, 2002</td>
<td>MADDSP birth cohorts; live births</td>
<td>2.0</td>
</tr>
<tr>
<td>Sundrum; UK, 2005</td>
<td>Retrospective cohort; live births</td>
<td>2.8</td>
</tr>
<tr>
<td>Serdarogulu; Turkey, 2006</td>
<td>Cross-sectional; children 2-16</td>
<td>4.4</td>
</tr>
<tr>
<td>Ozturk; Turkey, 2006</td>
<td>Cross-sectional; live births</td>
<td>1.1</td>
</tr>
<tr>
<td>Bhasin; US, 2006</td>
<td>MADDSP; cross-sectional, children age 8</td>
<td>3.1</td>
</tr>
<tr>
<td>Yeargin-Allsopp; US, 2008</td>
<td>ADDM; cross-sectional, children age 8</td>
<td>3.6</td>
</tr>
<tr>
<td>Boulet; US, 2009</td>
<td>NHIS; cross-sectional, children 3-17</td>
<td>3.9</td>
</tr>
<tr>
<td>Arneson; US, 2009</td>
<td>ADDM; cross-sectional, children age 8</td>
<td>3.3</td>
</tr>
</tbody>
</table>
Epidemiology of Cerebral Palsy
Public Health Model for Prevention of CP

Surveillance Systems
- prevalence
- registry of cases
- monitor prevention

Epidemiological Studies
- risk factors
- protective factors
- public concerns

Prevention Programs
- prevention strategies
- public policy
- education
Public health surveillance is the systematic, ongoing assessment of the health of a community, based on the collection, interpretation, and use of health data and information. Surveillance provides information necessary for public health decision making. *

Methods for Conducting CP Surveillance

- Notification (reportable disease surveillance)
- Periodic population-based surveys
- Aggregate data
- Disease registries
- Ongoing population based record review
**Ongoing, Population-Based Record Review**

Surveillance systems where information is systematically collected via standardized data collection instruments by review of existing records at data sources.

**Strengths**
- No contact with participants
- Representative
- Objective, reliable measures
- Individual-level data; Ability to link to other data
- Multiple source methods
- Active
- Minimal burden

**Ongoing program to monitor trends**
- Extensive QC measures
- Does not depend solely on previously documented CP diagnoses
- Can examine CP by subtype by race/ethnicity & co-occurring DDs
- Can be used to identify subjects for research
**Ongoing, Population-Based Record Review**

Surveillance systems where information is systematically collected via standardized data collection instruments by review of existing records at data sources.

Limitations
- More labor intensive and costly to operate than passive systems
- Timeliness
- May underestimate children with mild CP who have not come to the attention of service providers early in childhood
- Lack of completeness in some existing records;
- Dependent on the availability of records

Considered the Gold Standard approach (and CDC believes worth the investment) to produce the most accurate estimates of the prevalence of CP and track changes in prevalence over time.
Metropolitan Atlanta Developmental Disabilities Surveillance Program (MADDSP)

- Ongoing, active monitoring program since 1991
- 5 counties of metro Atlanta
- Multiple sources (educational, clinical, service sources)
- 5 Disabilities:
  - Mental Retardation/Intellectual Disability
  - Cerebral Palsy
  - Hearing Loss
  - Vision Impairment
  - Autism Spectrum Disorders (since 1996)
MADDSP Surveillance Case Definitions

Intellectual Disability (ID)*
I.Q. ≤ 70 on most recently administered psychometric test

Cerebral Palsy (CP)
A group of permanent disorders of the development of movement and posture that are attributed to non-progressive disturbances that occurred in the developing brain.

Hearing Loss (HL)
Measured bilateral pure tone hearing loss averaging 40 decibels or higher (unaided) in the better ear

Vision Impairment (VI)
Measured visual acuity of 20/70 or worse in the better eye with correction

*Formerly referred to as Mental Retardation (MR)
MADDSP: Types of Data Collected

- Demographics: child and parent names, race, ethnicity, gender, residence, DOB
- School service data: school, spec ed eligibility category
- Psychometric test results: intelligence, adaptive, autism
- Hearing and vision test results
- Physical findings (CP)
- Verbatim descriptions of behaviors (Autism)
- Associated medical conditions
- Other developmental disabilities monitored by MADDSP
Co Occurring Developmental Disabilities and Cerebral Palsy
Presence and Type of Co-occurring DDs among children with CP (MADDSP, 2004)

Proportion of 8-year-olds

Isolated CP | 55.9
CP & Co-occurring DDs | 44.1
CP & ID | 9.8
CP & AU | 3.5
CP & HL | 14.7
CP & VI | 37.1
Isolated CP | 16.8
CP+1 DD | 2.1
Severity of Intellectual Disability among children with CP (MADDSP, 2004)

Mean IQ score for CP cases with ID was 38.3 (std dev = 17.5)

Mean Adaptive functioning score for CP cases among those with adaptive data (90%) was 45.7 (std dev = 15.7)

41% of CP cases with ID scored higher on adaptive test
<table>
<thead>
<tr>
<th></th>
<th>Isolated CP</th>
<th>CP &amp; Co-occurring DDs</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>N</strong></td>
<td>63</td>
<td>80</td>
</tr>
<tr>
<td><strong>Spastic</strong></td>
<td>88.9%</td>
<td>82.5%</td>
</tr>
<tr>
<td><strong>Unilateral</strong></td>
<td>41.1%</td>
<td>27.7%</td>
</tr>
<tr>
<td><strong>Bilateral</strong></td>
<td>58.9%</td>
<td>72.3%</td>
</tr>
<tr>
<td><strong>Dyskinetic</strong></td>
<td>1.6%</td>
<td>1.3%</td>
</tr>
<tr>
<td><strong>Hypotonic</strong></td>
<td>1.6%</td>
<td>3.8%</td>
</tr>
<tr>
<td>*<em>Other</em></td>
<td>7.9%</td>
<td>12.5%</td>
</tr>
</tbody>
</table>

* Includes spastic-ataxic, spastic-dyskinetic, dyskinetic-ataxic, and CP not otherwise specified (CP NOS).
Use of Special Education Services among Children with CP by the Presence of Co-occurring DDs (MADDSP, 2004)

- Intellectual Disability: 37%
- Orthopedic Impairment: 10.9%
- Other Health Impairment: 6.5%
- Speech and Language: 4.3%
- Significant Developmental Delay: 2.4%
- Learning Delay: 2.4%
- Traumatic Brain Disorder: 4.3%

CP & Co-occurring DDs

- Intellectual Disability: 73.1%
- Orthopedic Impairment: 6.4%
- Other Health Impairment: 5.1%
- Autism: 5.1%
- Visual Impairment: 3.8%
- Significant Developmental Delay: 3.8%
CP and Co-Occurring DDs, MADDSP 2004

Conclusions

CP & ID
- Adaptive functioning data indicates functioning at a slightly higher level than IQ data.

Subtype
- Spastic bilateral CP is most common subtype, approximately 10% more children with CP & co-occurring DDs have this subtype than children with isolated CP.

Special Education
- Majority of children with CP and co-occurring DDs are served through ID classes.
- Children with isolated CP commonly served through other health impairment and orthopedic impairment classes.
The goals of the ADDM CP Network

- Obtain a complete count of the number of children with CP in each project area.
- Provide comparable, population-based CP prevalence estimates in different sites.
- Study if CP is more common in some groups of children than in others, and if rates are changing over time.
- Improve the consistency of identification of children with CP.
3 sites (AL, GA, WI) conducted CP surveillance in 2002.

Site-specific rates were similar (3.3-3.8 per 1,000) with average of 3.6 per 1,000 8-year-old children.

Male: female ratio was 1.1 (GA) to 1.6 (AL, WI).

Spastic CP was the most common subtype (77% of all cases), primarily represented by bilateral spastic CP.
ADDM: Expanded Cerebral Palsy Monitoring

- 3 sites (AL, GA, WI) conducted CP surveillance in 2004.
- Site-specific rates (4.1-3.1 per 1,000) with average of 3.3 per 1,000 8-year-old children.
- Male: female ratio was 1.3 (GA, AL) to 2.5 (WI).
- Spastic CP was the most common subtype (86.8% of all cases), primarily represented by bilateral spastic CP.

## ADDM Prevalence Comparisons 2002 and 2004

<table>
<thead>
<tr>
<th></th>
<th>AL</th>
<th>GA</th>
<th>WI</th>
<th>Average Prevalence</th>
</tr>
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<tbody>
<tr>
<td><strong>2002 Prevalence</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>(95% CI) per 1000</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3.7 (3.1-4.4)</td>
<td>3.8 (3.3-4.4)</td>
<td>3.3 (2.8-4.0)</td>
<td>3.6 (3.3-4.0)</td>
<td></td>
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<tr>
<td><strong>2004 Prevalence</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(95% CI) per 1000</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4.1 (3.0, 5.4)</td>
<td>3.1 (2.7-3.7)</td>
<td>3.3 (2.4-4.6)</td>
<td>3.3 (2.9-3.8)</td>
<td></td>
</tr>
</tbody>
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*2004 was an optional SY. WI and AL had reduced study areas*
Use of Surveillance Data: Etiologies and Risk Factors for CP

- Linkage with birth certificate data
- Approximately 60% of period prevalence cases born in surveillance area
- Population-based case series for special studies
Birth Weight Distribution of 3-10 Yr Old Children with Developmental Disabilities

(Referent population: 3-yr survivors, 1981-1993 Atlanta births)
## Prevalence of CP by Birth Cohort


<table>
<thead>
<tr>
<th></th>
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</thead>
<tbody>
<tr>
<td># Survivors</td>
<td># Cases</td>
<td>Rate 1000</td>
<td># Survivors</td>
</tr>
<tr>
<td>&lt;1500</td>
<td>414</td>
<td>27</td>
<td>65.2</td>
</tr>
<tr>
<td>1500-2499</td>
<td>4361</td>
<td>30</td>
<td>6.9</td>
</tr>
<tr>
<td>&gt;2500</td>
<td>60011</td>
<td>53</td>
<td>0.9</td>
</tr>
<tr>
<td>Total</td>
<td>64,786</td>
<td>110</td>
<td>1.7</td>
</tr>
</tbody>
</table>

*CP cases identified at 10 years of age
†CP cases identified at 3 to 10 years of age
The Association between Cerebral Palsy or Mental Retardation and Prenatal Magnesium Sulfate Exposure in Atlanta Infant Survivors*

<table>
<thead>
<tr>
<th></th>
<th>Prenatal Magnesium Sulfate Exposure</th>
<th></th>
<th>OR</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Yes</td>
<td>No</td>
<td></td>
<td>95% CL</td>
</tr>
<tr>
<td>CP</td>
<td>1 (0.9%)</td>
<td>30 (7.7%)</td>
<td>0.11</td>
<td>0.02, 0.81</td>
</tr>
<tr>
<td>MR</td>
<td>2 (1.8%)</td>
<td>22 (5.8%)</td>
<td>0.30</td>
<td>0.07, 1.29</td>
</tr>
</tbody>
</table>

Schendel et al.  JAMA 1996:276(22);1805-1810
Magnesium Sulfate Reduces Cerebral Palsy Risk after Preterm Birth

- 2,241 women at "imminent risk" of preterm birth
  - 24 through 31 weeks gestation
- Cases randomized to intravenous magnesium sulfate or placebo
- Results:
  - Overall prevalence of CP was lower in the mag sulfate treated group (1.9% versus 3.5%).
  - The risk of death did not differ significantly between groups and no woman had a life-threatening event.

Summary

- The epidemiology of cerebral palsy is a rapidly evolving field
- Trends in CP prevalence indicate:
  - Overall increase from 1991-2006
  - Consistently higher among children who are/have
    - Boys
    - Black non-Hispanic
    - Spastic bilateral CP with and without co-occurring DDs
- Surveillance data can be used for etiologic research on cerebral palsy
- CDC is committed to understanding more about CP using the public health model in order to ultimately guide prevention programs and inform public policy.
Acknowledgements
“It Takes A Village”

- It takes many individuals for MADDSP and each ADDM site to run our monitoring programs.
- Primary investigators, project coordinators, abstractors, data managers, programmers, clinician reviewers, epidemiologists and other project staff.
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Cathy Yungbluth

SAFER • HEALTHIER • PEOPLE™
Thank you!!

For learn more about CDC’s work on developmental disabilities, go to:

http://www.cdc.gov/ncbddd

The findings and conclusions in this report are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention.