WESTERN AUSTRALIAN REGISTER OF DEVELOPMENTAL ANOMALIES – CEREBRAL PALSY

WARDA-CP

FIELD NAMES, DESCRIPTIONS AND VALUES

For clinical description of cerebral palsy and associated impairments

INCLUDES
BIRTH YEAR 1980 ONWARDS
WA-BORN CASES ONLY

Data held on CARES

INDEX

Field name	Description	Type	Page
cpsev	CP severity at age 5 years	smallint	1
pnncause	Postneonatal cause of CP	smallint	2
kncause	Known cause of CP	smallint	2
intellect	Intellectual status	smallint	3
epilepsy	Epilepsy	smallint	3
vision	Severity of visual impairment	smallint	3
strabismus	Presence of strabismus	smallint	3
hearing	Severity of hearing impairment	smallint	3
speech	Severity of speech delay/impairment	smallint	4

Calculated fields

Field name	Description	Type	Page
cptypepredom1	Predominant CP type using classical terms	derived	5
cptypepredom2	Predominant CP type using finer categories	derived	5
gmf	cpsev regrouped into approximate GMFCS levels	derived	5
iqgrp1	Intellectual ability in two groups: ID / No ID	derived	5
iqgrp2	Intellectual ability in five groups by severity	derived	5
dscore	Overall disability score	derived	6

Appendix A **Classification of CP for the ACPR**

Appendix B **Disability Score**

Field Name:	**cpsev**
Description:	CP severity at 5 years (used for cases back to birth year 1956. As GMFCS / MACS have now been widely adopted as CP severity indicators, **cpsev** has been converted to **gmf** to enable comparison with GMFCS.
Field Size:	1
Data Type:	smallint
Possible values:	1 = Minimal 2 = Mild 3 = Moderate 4 = Severe
Notes:	Definitions: Minimal – motor signs present without functional impairment. Mild – symptoms result in some functional impairment. Moderate – between mild and severe; eg, ambulant with walking frame. Severe – little purposeful voluntary action, though function may be acquired, IQ permitting. **cpsev** is consistent throughout all years of WARDA-CP data. It refers to the severity of motor impairment in the most affected limb whereas GMFCS relates to functional severity in lower limbs. **cpsev** is therefore not comparable with GMFCS. A new variable, **gmf**, derived from **cpsev** and **gmfcs** using an algorithm that takes these differences into consideration, converts **cpsev** to a GMFCS-equivalent, three-category scale of I-II, III and IV-V.

Field Name: **pncause**
Description: Definite postneonatal cause of CP (after 28 days and before age 5 years)

Field Size: 1
Data Type: smallint

Possible values:
0 = Pre/perinatal cause
1 = Definite postneonatal cause occurring before the age of 2yrs (includes neonatal injury in an undisputedly normal infant *)
2 = Definite postneonatal cause occurring at >=2yrs but < 5yrs

Notes: Coding changed in 2010:

(1) Postneonatal cause divided into two codes (1,2) in order to separate those occurring up to the age of 2 years which are included in the ACPR minimum data set (pncause = 1) and those occurring after age 2 years and before age 5 years which are included only in the WA data set (pncause = 2).

Field Name: **kncause**
Description: Single cause of CP where known with certainty (ie, no doubt is expressed by any member of the medical team)

Field Size: 2
Data Type: smallint

Values:

Pre-/perinatal causes
02 = Intrauterine CMV
03 = Other TORCH infection
08 = Other definite pre-/perinatal cause

Postneonatal causes
Infection:
21 = Dehydration due to gastroenteritis
22 = Other bacterial infection
23 = Other viral infection
28 = Infection nos

Postneonatal causes
Infection:
21 = Dehydration due to gastroenteritis
22 = Other bacterial infection
23 = Other viral infection
28 = Infection nos

Cerebrovascular accident:
31 = Associated with surgery
32 = Associated with cardiac complications (not during/post surgery)
38 = Spontaneous / other CVA

Head injury
41 = MVA – Passenger in vehicle
42 = MVA – Pedestrian
43 = MVA – Unknown if passenger or pedestrian
44 = Non-accidental
45 = Fall
48 = Other head injury / nos

Other causal events:
51 = Near drowning
52 = Apparent life-threatening event (includes near-SIDS)
53 = Post-immunisation
54 = Post-seizure
55 = Peri-operative hypoxia
58 = Other postneonatal event

Neonatal causes
Events occurring in the neonatal period are likely to be associated with the intrauterine environment and therefore are coded as Pre-/Perinatal cause (**pncause** = 0), with the exception of neonatal injury in an undisputedly normal infant (code **pncause** = 1 and make note in **kncause** field).

Notes: Code only if known conclusively to be the immediate cause of the CP. Do not code possible or contributing causes.

Field Name: **intellect**
Description: Intellectual ability (IQ or DQ)

Field Size: 1
Data Type: smallint

Possible values:
0 = Normal / borderline (IQ/DQ >= 70 or so described)
2 = Mild impairment (IQ/DQ 50-69 or so described)
3 = Moderate impairment (IQ/DQ 35-49 or so described)
4 = Severe impairment (IQ/DQ < 35 or so described)
5 = Probably intellectually disabled (IQ/DQ < 70), severity uncertain
6 = Probably no impairment, or only borderline
Null = Unknown / missing data

Field Name: **epilepsy**
Description: Epilepsy

Field Size: 1
Data Type: smallint

Possible values:
0 = None
1 = Resolved by age 5 years (seizure-free for two or more years without medication)
2 = Epilepsy
Null = Unknown / missing data

Notes: Epilepsy defined as two or more afebrile seizures before age 5 years; does not include neonatal seizures.

Field Name: **vision**
Description: Severity of visual impairment

Field Size: 1
Data Type: smallint

Possible values:
0 = No impairment
2 = Some visual impairment (wears glasses)
3 = Functionally blind
Null = Unknown / missing data

Field Name: **strabismus**
Description: Presence of strabismus at age 5 years

Field Size: 1
Field Type: smallint

Values:
0 = No strabismus
1 = Strabismus (includes surgically corrected)
Null = Unknown / missing data

Field Name: **hearing**
Description: Severity of hearing impairment

Field Size: 1
Data Type: smallint

Possible values:
0 = No impairment
2 = Some impairment (includes conductive loss)
3 = Bilateral deafness
Null = Unknown / missing data

Field Name: **speech**
Description: Severity of speech delay / impairment

Field Size: 1
Data Type: smallint

Possible values: 0 = No impairment
2 = Some impairment
3 = Non-verbal
Null = Unknown / missing data

CALCULATED FIELDS

Field Name: **cptypepredom1**
Description: Classical terminology for CP types:
- Hemiplegia
- Diplegia
- Quadriplegia
- Ataxia
- Dyskinesia
- Hypotonic CP

Data Type: Derived

Obtained from: **cptype1**
See H:SQL 2009/altertable cptypepredom.sql
See H:SQL 2009/altertable cptypesecond.sql
See also Appendix A

Field Name: **cptypepredom2**
Description: Classical terminology for CP types (finer categories)
- Right hemiplegia
- Left hemiplegia
- Diplegia
- Triplegia
- Quadriplegia
- Ataxia
- Athetosis
- Dystonia
- Hypotonic CP

Data Type: Derived

Obtained from: **cptype1, cplimb1**
See H:SQL 2009/altertable cptypepredom.sql
See H:SQL 2009/altertable cptypesecond.sql
See also Appendix A

Field Name: **gmf**
Description: **cpsev** regrouped into equivalent GMFCS levels:

Minimal/Mild = I-II
Moderate = III
Severe = IV-V

Data Type: Derived

Obtained from: **cpsev**
See H:SQL2009/altertable gmf.sql

Notes: GMFCS relates to LL function while **cpsev** relates to severity in worst affected limb (in hemiplegia usually UL). Therefore the algorithm to convert **cpsev** to **gmf** takes this into consideration.

Field Name: **iqgrp1**
Description: Intellectual ability grouped as
ID (IQ < 70); No ID (IQ >= 70)

Data Type: Derived

Obtained from: **intellect**
See H:SQL 2009/altertable iqgrp1.sql

Field Name: **iqgrp2**
Description: Intellectual ability grouped by severity ranges:
Normal/borderline, Mild, Moderate, Severe, Unknown

Data Type: Derived

Obtained from: **intellect**
See H:SQL 2009/altertable iqgrp2.sql

Field Name: **dscore**

Description: Disability score as an estimate of overall disability.

Scores from 1 to 12:

Mild	= 1-4
Moderate	= 5-8
Severe	= 9-12

Data Type: Derived

Obtained from: **cptype1, cpsev, epilepsy, intellect, vision, hearing**

For program see SOLO.Wacpreg/Programmability/Functions/Scalar-valued functions/dbo.udf_setdscore

See also Appendix C

Appendix A

Classification of Cerebral Palsy for the Australian Cerebral Palsy Register

Love S, Gibson N, Gubbay A, Blair E, Watson L

Cerebral palsy (CP) is a term that refers to a number of different movement disorders which, for the purposes of the Australian Cerebral Palsy Register (ACPR), are grouped into the following categories:

- Spastic CP is the most common type, occurring as the predominant CP type in about 80% of cases in Western Australia. It is characterised by increased muscle tone and is further classified according to the limb distribution of the hypertonia and ASAS score (see App B):

    - Spastic monoplegia, though rare, is the involvement of one limb only.
    - Spastic hemiplegia is the involvement of only one side of the body, usually more pronounced in the upper limb. Very minimal signs may also be present on the contralateral side.
    - Spastic diplegia means the lower limbs are more affected than the upper limbs. A significant difference in the amount of spasticity (at least 1 point difference as measured by the Modified Ashworth Scale) between the right and left lower limbs is referred to as asymmetric diplegia.
    - Spastic triplegia has been accepted as a separate category by the ACPR and is used to describe involvement of all four limbs but with the relative sparing of one upper limb, and spasticity in the other upper limb being greater than or equal to that in the lower limbs.
    - Spastic quadriplegia means that the upper limbs are equally or more affected than the lower limbs, regardless of any difference in the amount of spasticity between the right and left sides.

    In all types of spastic CP truncal tone will vary, and bulbar signs may or may not be present.

- Dyskinetic cerebral palsy has two forms:

    - Athetoid cerebral palsy is characterised by increased activity with involuntary, unpredictable movements that may be present even at rest. Muscle tone tends to be decreased.

    - Dystonic cerebral palsy is characterised by reduced activity with fluctuating muscle tone, increased at times, depending on posture, mood and effort.

- Ataxic cerebral palsy is characterised by unsteady, wobbling movements or tremor, and problems with balance.

These motor disorders may occur singly or in combination. The presence of other conditions, such as impaired hearing or vision, epilepsy, intellectual disability or speech delay/impairment should be recorded separately and do not have a bearing on the classification of CP type.

Appendix B

Disability score

In order to assess the combined impact on survival of several co-existing disabilities, an overall disability score was derived by summing the score assigned to each disability as follows:

Category of movement disorder:

Hemiplegia = 1, Diplegia = 2, Other = 3

Severity of movement disorder:

Minimal = 0, Mild = 1, Moderate = 2, Severe=3

Severity of cognitive deficit:

IQ 50-69 = 1, IQ 35-49 = 2, IQ<35 = 3

Other impairments:

Blind = 1, Bilateral deafness = 1; Current epilepsy = 1

Thus the maximum possible disability score is 12 and the minimum is 1 (minimal hemiplegia without other impairment). The most frequently occurring score was 4 (17.2%).

This scoring system entails assumptions, for example, that the disability conferred by being blind is equal to the disability conferred by increasing one category in IQ deficit or severity of movement disorder. However it has the advantage of simplicity and reflects therapists' perceptions of overall disability.

Please note (2017): Bilateral deafness (2.2% of cases) to be replaced by non-verbal (24.1% of cases), but as speech impairment data is unavailable for cases born 1956-1974, deafness continues to be used for "Survival with CP" papers.

References

1. Blair E, Wallman A. Changing rates of severity of cerebral palsy and implications for practice. *Action Packed* 2000, 5(3): 18-20
2. Blair E. Life expectancy among people with cerebral palsy in Western Australia [letter]. *Developmental Medicine and Child Neurology* 2001; 43: 792