Early Diagnosis of Cerebral Palsy in Clinical Practice

A resource for health professionals to detect and diagnose cerebral palsy early, communicate the news of diagnosis and support infants and families using best-practice recommendations.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
Cerebral Palsy CENTRE OF RESEARCH EXCELLENCE
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Acknowledgements

CONTRIBUTING AUTHORS

Professor Iona Novak – Head of Research Cerebral Palsy Alliance, University of Sydney

Professor Dinah Reddihough – Chief Investigator NHMRC-funded Centre of Research Excellence in Cerebral Palsy (CRE-CP), Murdoch Children's Research Institute

Professor Roslyn Boyd – Scientific Director Queensland Cerebral Palsy and Rehabilitation Research, Australasian Cerebral Palsy Clinical Trials Network.

Dr Margot Bosanquet – Queensland Health, James Cook University

Associate Professor Karen Scott – The University of Sydney

Dr Joanne George - Queensland Early Detection and Intervention Network

Dr Cathy Morgan – Cerebral Palsy Alliance Research Institute

Ms Anna te Velde – Cerebral Palsy Alliance Research Institute

Associate Professor Andrea Guzzetta – University of Pisa

Ms Olena Chorna –Fondazione Stella Maris, Tierrenia, Italy

Professor Nadia Badawi – Chair of Cerebral Palsy, Cerebral Palsy Alliance, Westmead Children's Hospital, University of Sydney.

Associate Professor Michael Fahey – Monash University, Monash Children's Hospital

Associate Professor Luke Jardine – Mater Health Services, University of Queensland

Associate Professor Alicia Spittle – University of Melbourne

Parent Consultation Groups – NHMRC-funded Centre of Research Excellence in Cerebral Palsy (CRE-CP)

Parent Consultation Groups – Cerebral Palsy Alliance

Ms Lynda McNamara – Knowledge Translation Fellow Centre of Research Excellence in Cerebral Palsy (CRE-CP) and PhD student The University of Sydney

ACKNOWLEDGEMENTS

We thank Professor Dinah Reddihough, Chief Investigator of the NHMRC-funded Centre of Research Excellence in Cerebral Palsy (CRE-CP), parent consultation groups and advisory committee members of the CRE-CP who contributed to the development of the fact sheet resources.

We thank The Cerebral Palsy Alliance for funding associated with the development of this resource.

Lynda McNamara is in receipt of a NHMRC funded Centre of Research Excellence in Cerebral Palsy (CRE-CP) Knowledge translation fellowship (2016-2019); NHMRC Postgraduate Scholarship (APP1169076) and is supported by The Australasian Cerebral Palsy Clinical Trials Network (AusCP-CTN, NHMRC Number 1116442).

Cerebral Palsy ALLIANCE

THE UNIVERSITY OF SYDNEY

AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Cerebral Palsy Background Information

What is Cerebral Palsy?

Cerebral palsy is:

(1) an umbrella term for a group of disorders
(2) a condition that is permanent but not unchanging
(3) a disorder of movement and/or posture and of motor function
(4) due to a non-progressive interference, lesion, or abnormality, and
(5) the interference, lesion, or abnormality originates in the immature brain.[1-5]

Cerebral palsy is the most common cause of physical disability in childhood.[1]

Classification of Cerebral Palsy

Three major classifications are used to describe cerebral palsy - motor type, topography and function.

Classification by Motor Sub-Types

There are four traditional major motor subtypes (from registries in western industrialised countries):

- **Spasticity:** increased muscle tone, increased deep tendon reflexes, weakness and abnormal gait and posture.[1-8]
- **Dyskinetic:** may have dystonic, athetoid or choreoathetoid movement patterns including involuntary, uncontrolled, recurring, occasionally stereotyped movements and fluctuating muscle tone.[1-9]
- **Ataxia:** characterised by problems with balance and depth perception, loss of co-ordination, so that movements are poorly organised in terms of force, rhythm and accuracy.[1-9]
- **Hypotonia:** Only a very small group of children with cerebral palsy exhibit pure hypotonia with generalised decreased tone.[1] Hypotonic cerebral palsy is characterised by generalised hypotonia that persists beyond 3 years of age and does not result from a primary disorder of peripheral nerves.[9]

There may be more than one motor disorder. A combination of spasticity and dystonia is common. The motor sub-types may emerge and change over the first few years of life.[1,6]

1. ACPR Group. Australian cerebral palsy register report 2018, birth years 1995-2012. Sydney, Australia: Cerebral Palsy Alliance; (2018)
2. Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys and registers. Surveillance of Cerebral Palsy in Europe (SCPE). (2000). Dev Med Child Neurol, 42(12), 816-824. doi:10.1017/s0012162200001511
3. Bax, M. C. O. (1964). TERMINOLOGY AND CLASSIFICATION OF CEREBRAL PALSY. Developmental Medicine & Child Neurology, 6(3), 295-297. doi:10.1111/j.1469-8749.1964.tb10791.x
4. Rosenbaum, P., Paneth, N., Leviton, A., Goldstein, M., Bax, M., Damiano, D., . . . Jacobsson, B. (2007). A report: the definition and classification of cerebral palsy April 2006. Dev Med Child Neurol Suppl, 109, 8-14.
5. Mutch, L., Alberman, E., Hagberg, B., Kodama, K., & Perat, M. V. (1992). Cerebral palsy epidemiology: where are we now and where are we going? Dev Med Child Neurol, 34(6), 547-551. doi:10.1111/j.1469-8749.1992.tb11479.x
6. Rice, J., Skuza, P., Baker, F., Russo, R., & Fehlings, D. (2017). Identification and measurement of dystonia in cerebral palsy. Dev Med Child Neurol, 59(12), 1249-1255. doi:10.1111/dmcn.13502
7. Sanger, T. D., Delgado, M. R., Gaebler-Spira, D., Hallett, M., & Mink, J. W. (2003). Classification and definition of disorders causing hypertonia in childhood. Pediatrics, 111(1), e89-97. doi:10.1542/peds.111.1.e89
8. Oskoui, M., Coutinho, F., Dykeman, J., Jette, N., & Pringsheim, T. (2013). An update on the prevalence of cerebral palsy: A systematic review and meta-analysis. Dev Med Child Neurol, 55. doi:10.1111/dmcn.12080
9. Koman, L. A., Smith, B. P., & Shilt, J. S. (2004). Cerebral palsy. Lancet, 363(9421), 1619-1631. doi:10.1016/s0140-6736(04)16207-7

Cerebral Palsy Background Information

Classification by Topography

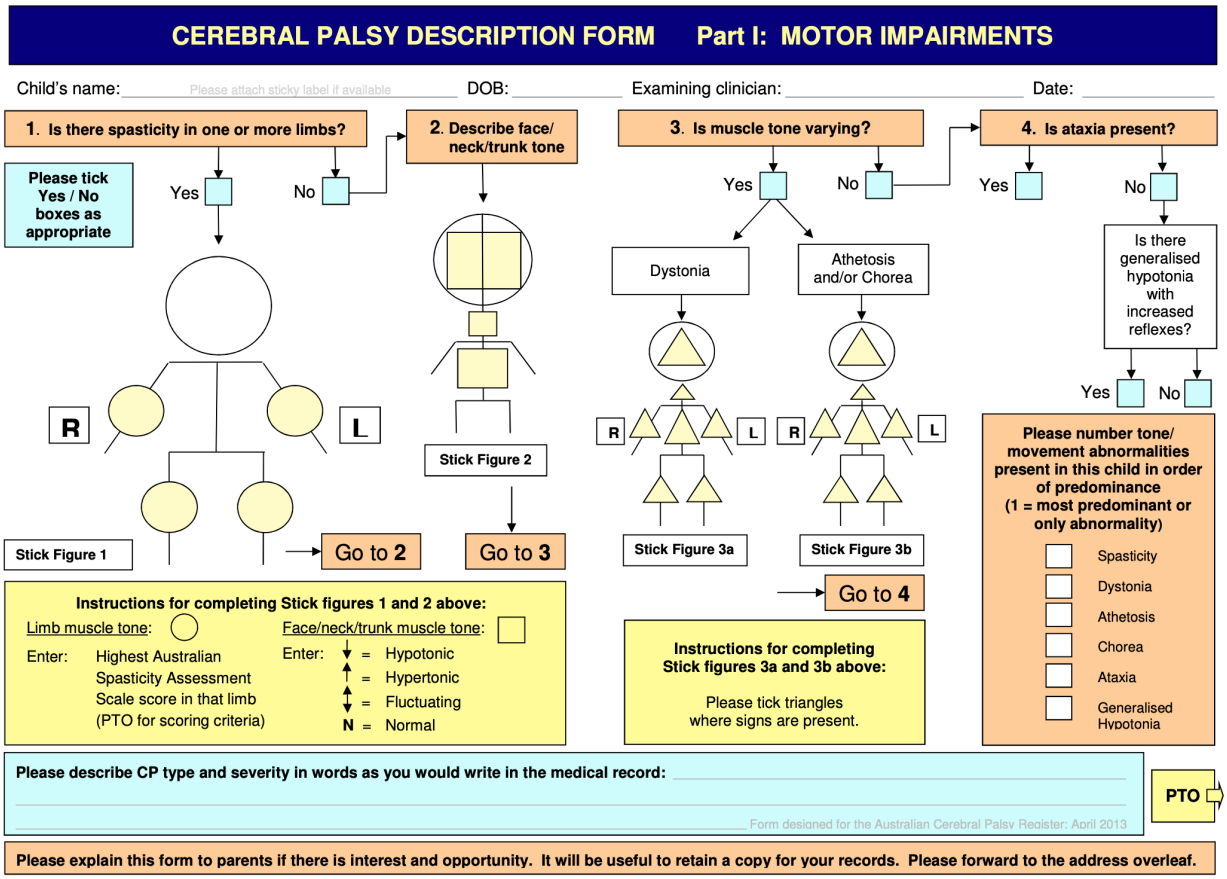
Categorisation by topography also guides intervention. The early identification of unilateral versus bilateral cerebral palsy is important, as the interventions and long-term musculo-skeletal outcomes differ.[1]

Recognised typographies for spasticity (Australian Cerebral Palsy Register and Surveillance of Cerebral Palsy Europe):

- Unilateral: one side of the body is predominantly involved. In rare cases, if only one limb is involved, the term monoplegia is applied.
- Bilateral: both sides of the body are involved. It can be further categorised into:

  A. Diplegia: both legs are impaired. Legs impaired more than the arms.
  B. Quadriplegia: all four limbs and trunk are impaired.[2]

The Australian Cerebral Palsy Register utilises the Cerebral Palsy Description Form: Motor Impairments. This form applies a limb by limb approach to provide an objective clinical picture of the child with cerebral palsy.[2]



[View form]

1. Palisano, R. J., Rosenbaum, P., Bartlett, D., & Livingston, M. H. (2008). Content validity of the expanded and revised Gross Motor Function Classification System. Dev Med Child Neurol, 50(10), 744-750. doi:10.1111/j.1469-8749.2008.03089.x
2. ACPR Group. Australian cerebral palsy register report 2018, birth years 1995-2012. Sydney, Australia: Cerebral Palsy Alliance; (2018). Available from: https://cpregister.com/wp-content/uploads/2019/02/Report-of-the-Australian-Cerebral-Palsy-Register-Birth-Years-1995-2012.pdf. Accessed June

Cerebral Palsy Background Information

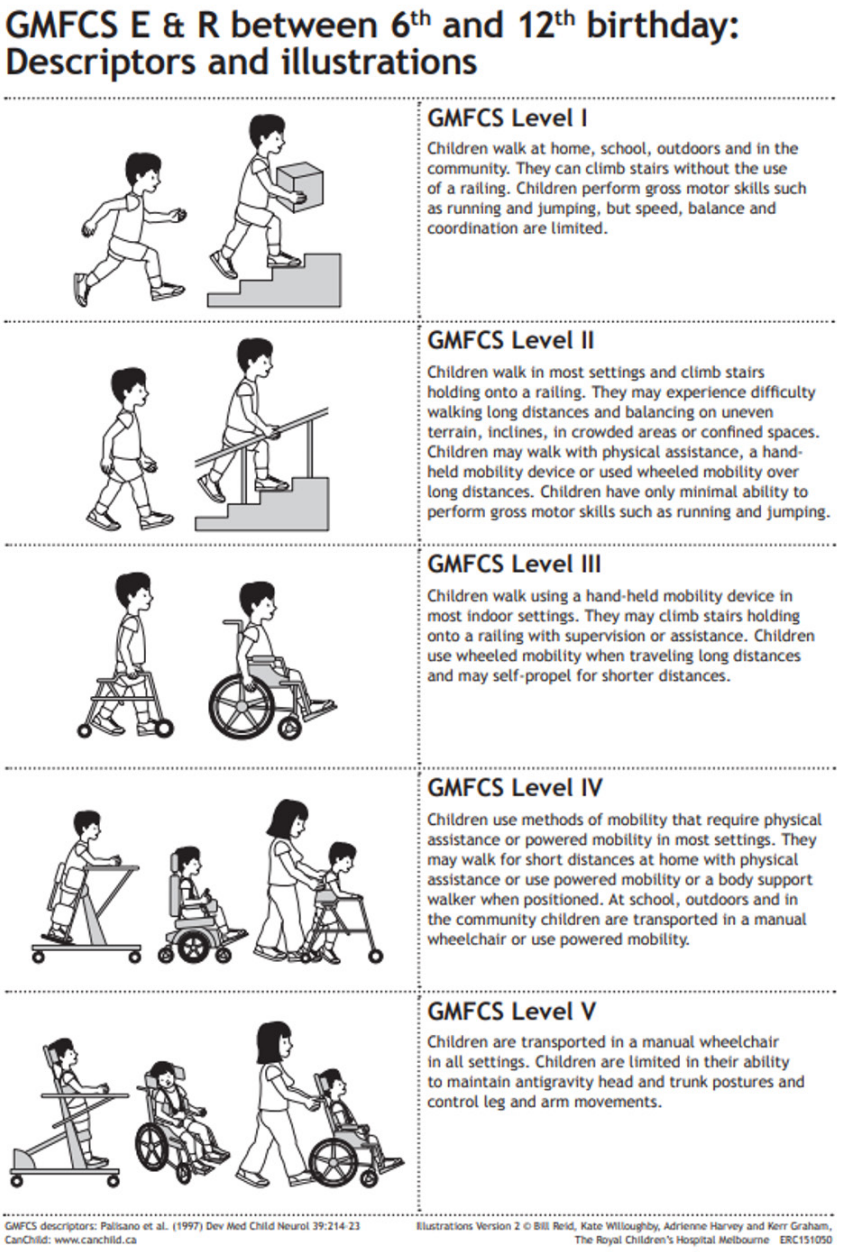
Classification by Motor Function and Severity

The Gross Motor Function Classification System (GMFCS – E& R)[1,2] www.canchild.ca is the "gold standard" five level classification system that provides a common language and functional "picture" of a child with cerebral palsy. Based on a child's ability to self-initiate movement, with a focus on sitting, transferring, and mobilising, different functional classification descriptions exist at different age groups. It provides a prognostic guide for longer term mobility and is most accurate in children over 2 years of age[3]. There are five age bands: Under 2 years, 2 – 4 years, 4 – 6 years, 6 – 12 years and 12 – 18 years.

Classification by Motor Function and Severity

At 2 – 4 years Classification by Gross Motor Function:

- Level I: Floor sits independently, hands-free. Walks without assistive device
- Level II: Floor sits independently, hands-free with balance affected. Walks using assistive mobility device
- Level III: Floor sits using w-sitting. Walks short distances indoors using a hand-held mobility device with assistance
- Level IV: Floor sits when placed, used hands for balance. Rolls, creeps or crawls for short distances
- Level V: Unable to sit independently. No form of independent mobility



Assessment of GMFCS classification at the age of 2 is recommended. Functional severity classification made after 2 years of age it is stable and life-long.[3]

1. Palisano, R. J., Rosenbaum, P., Bartlett, D., & Livingston, M. H. (2008). Content validity of the expanded and revised Gross Motor Function Classification System. Dev Med Child Neurol, 50(10), 744-750. doi:10.1111/j.1469-8749.2008.03089.x
2. Palisano, R. J., Rosenbaum, P., Walter, S., Russell, D, et al. (1997). Development and reliability of a system to classify gross motor function in children with cerebral palsy. Dev Med Child Neurol, 39(4): 214-23.
3. Gorter, J. W., Ketelaar, M., Rosenbaum, P., Helders, P. J., & Palisano, R. (2009). Use of the GMFCS in infants with CP: the need for reclassification at age 2 years or older. Developmental Medicine & Child Neurology, 51(1), 46-52. doi:10.1111/j.1469-8749.2008.03117.x

Cerebral Palsy Background Information

Other Functional Classification Systems

Other functional cerebral palsy classification systems include:

- **Manual Ability Classification Systems (MACS)** classifies typical manual performance and how a child handles an object in daily life.[1] www.macs.nu
- **Bimanual Fine Motor Classification System (BFMF)**[2] describes fine motor function by classifying the ability to grasp, hold and manipulate objects in each hand separately.
- **Communication Function Classification System (CFCS)** classifies everyday communication performance.[3] http://cfcs.us
- **Eating and Drinking Ability Classification System (EDACS)** classifies a child's usual ability to eat and drink considering safety, efficiency and level of assistance required.[4] www.sussexcommunity.nhs.uk
- **Visual Function Classification System (VFCS)** classifies how toddler and youth with Cerebral palsy (CP) use visual abilities in daily life.

1. Eliasson, A.C., Krumlinde-Sundholm, L., Rosbald, B., Beckung, E., et al. (2006). The Manual Ability Classification System (MACS) for children with cerebral palsy: scale development and evidence of validity and reliability. Dev Med Child Neurol. 48 (7): 549-54.
2. Beckung, E., & Hagberg, G. (2002). Neuroimpairments, activity limitations, and participation restrictions in children with cerebral palsy. Dev Med Child Neurol, 44(5), 309-316. doi:10.1017/s0012162201002134
3. Hidecker M.J, Paneth N.,Rosenbaum, P.L., Kent, et al. (2011). Dev Med Child Neurol. 53(8): 704-10.
4. Sellers, D., Mandy,A., Pennington,L.,Hankins, M., Morris,C. (2014). Dev Med Child Neurol. 56:245-51.

Cerebral Palsy Background Information

Associated impairments

All children with cerebral palsy have a motor impairment but this is frequently also accompanied by associated impairments, health issues and functional limitations.[1] Likelihood and severity of associated impairments increases with the severity of motor impairment and may impact more on function and quality of life than the motor impairment.[2-6]

Physicians should routinely screen for associated impairments, diseases and functional limitations co-occurring with cerebral palsy.[7] Co-occurring impairments and diseases are strongly linked to the severity of the motor impairment with the exception of pain and behaviour disorders. Pain is likely to be present with all levels of disability and behaviour disorders common with milder levels of motor impairment.

Keeping up to date with the best available evidence interventions for the prevention and management of cerebral palsy is challenging for clinicians and families. Systematic reviews of cerebral palsy interventions [8-9] assist in guiding families and clinical decision making.

Pain (75%) is likely to be present at all levels of physical disability.

Behaviour disorders (25%) are more common in the presence of mild physical disability.

The co-occurrence of epilepsy and intellectual disability in combination with severe physical disability has the greatest impact on prognosis and life expectancy.[1]

From a meta-analysis of CP registers,[1] the rates of associated impairments and functional limitations are as follows:

3 in 4
have chronic pain

1 in 2
have an intellectual disability

1 in 3
cannot walk

1 in 4
cannot talk

1 in 4
have epilepsy

1 in 3
have hip displacement

1 in 4
have bladder control problems

1 in 5
have a sleep disorder

1 in5
have sialorrhea

1 in 10
are blind

1 in 15
require tube feeding

1 in 25
are deaf

Cerebral Palsy Background Information

1. Novak, I., Hines, M., Goldsmith, S., & Barclay, R. (2012). Clinical prognostic messages from a systematic review on cerebral palsy. Pediatrics, 130(5), e1285-1312. doi:10.1542/peds.2012-0924
2. McIntyre, S., Morgan, C., Walker, K., & Novak, I. (2011). Cerebral palsy--don't delay. Dev Disabil Res Rev, 17(2), 114-129. doi:10.1002/ddrr.1106
3. Stanley, F., Blair, E., & Alberman, E. (2000). Cerebral Palsies: Epidemiology and Causal Pathways. London: MacKeith Press. 8-13.
4. Himmelmann, K., Beckung, E., Hagberg, G., & Uvebrant, P. (2006). Gross and fine motor function and accompanying impairments in cerebral palsy. Dev Med Child Neurol, 48(6), 417-423. doi:10.1017/s0012162206000922
5. Odding, E., Roebroeck, M. E., & Stam, H. J. (2006). The epidemiology of cerebral palsy: incidence, impairments and risk factors. Disabil Rehabil, 28(4), 183-191. doi:10.1080/09638280500158422
6. Delacy, M. J., & Reid, S. M. (2016). Profile of associated impairments at age 5 years in Australia by cerebral palsy subtype and Gross Motor Function Classification System level for birth years 1996 to 2005. Dev Med Child Neurol, 58 Suppl 2, 50-56. doi:10.1111/dmcn.13012
7. Novak, I., Morgan, C., Adde, L., Blackman, J., Boyd, R. N., et al. (2017). Early, Accurate Diagnosis and Early Intervention in Cerebral Palsy: Advances in Diagnosis and Treatment. JAMA Pediatr, 171(9), 897-907. doi:10.1001/jamapediatrics.2017.1689
8. Novak, I., McIntyre, S., Morgan, C., Campbell, L., Dark, L., Morton, N., . . . Goldsmith, S. (2013). A systematic review of interventions for children with cerebral palsy: state of the evidence. Dev Med Child Neurol, 55(10), 885-910.
9. Novak, I., Morgan, C., Fahey, M., Finch-Edmondson, M., Galea, C., Hines, A., . . . Badawi, N. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Cerebral Palsy Background Information

Incidence and Prevalence

The prevalence of cerebral palsy is showing some decline in recent years in both rates and severity in both Australia and Europe.

The prevalence of cerebral palsy in the most recent reporting period (2010 – 2012) of the Australian Cerebral Palsy Register is 1 in 700 children (1.4 per 1000 live births).[1]

Advances in neuroprotective strategies and improvements in public health, obstetric and perinatal care are contributing, all underpinned by Australian and international research.

- The rate of CP per 1000 neonatal survivors for children born 20-27 weeks declined, 1995-2012.[1]
- The rate of CP per 1000 live births for those born 37+ weeks declined, 2004-2012.[1]
- The rate of CP per 1000 neonatal survivors with moderate-severe gross motor function (Gross Motor Function Classifications System, levels III-V) declined, 1995-2012.[1]

Prevalence Rates by Gross Motor Functional Classification Scale

Mild cerebral palsy (classified according to GMFCS levels) is more common than severe cerebral palsy;

GMFCS 1 – 11 62%
GFMCS 111 12%
GMFCS IV – V 26%

Percentage of children with CP by Gross Motor Function Classification System groups (levels I-II, III, IV-V), predominant motor type at 5 years and birth period, all states/territories combined (1995-2012).[1]

Sub group populations at risk of cerebral palsy[1]

Sub group populations of infants considered at risk of cerebral palsy can be identified as:

- **Premature Infants** (30 – 40% of all cerebral palsy whose risk increases as gestational age decreases);
- **Term Born Infants with Perinatal event or birth defect eg Neonatal Encephalopathy** (15 – 20% of all cerebral palsy whose risk of cerebral palsy increases with severity of NE);
- **Perinatal stroke or cerebral birth defect**;
- **Term born infants receiving routine care at birth** (40 – 50% of all cerebral palsy and may not have any perinatal risk factors). [1-2]

1. ACPR Group. Australian cerebral palsy register report 2018, birth years 1995-2012. Sydney, Australia: Cerebral Palsy Alliance; (2018). Available from: https://cpregister.com/wp-content/uploads/2019/02/Report-of-the-Australian-Cerebral-Palsy-Register-Birth-Years-1995-2012.pdf. Accessed June 2020.
2. McIntyre, S., Morgan, C., Walker, K., & Novak, I. (2011). Cerebral palsy--don't delay. Dev Disabil Res Rev, 17(2), 114-129. doi:10.1002/ddrr.1106

Risk factors for cerebral palsy

Clinical history indicating risk for cerebral palsy

The full causal pathway is a complex interplay between several risk factors across multiple epochs. Note that as many as one third of children who are diagnosed with cerebral palsy lack traditional risk factors for cerebral palsy.

Preconception

Previous stillbirths, miscarriage(s), use of reproductive technology, low socio economic status

and/or during pregnancy

Intra-uterine growth restriction, prematurity, maternal thyroid disease, pre-eclampsia, placental abnormalities, bleeds, infection, substance abuse, multiple births, birth defects

and/or perinatal

Acute intrapartum hypoxic event, stroke, seizures, hypoglycaemia, jaundice, infection

and/or postnatal

Stroke, infections, accidental and non-accidental brain injury

Risk factors for cerebral palsy

Newborn detectable risks

Born premature cerebral palsy risk factor

< 28 weeks = 3-9%

28–31 weeks = 3-5%

32–36 weeks = <1%

> 37 weeks = 0.1

Abnormal neuroimaging

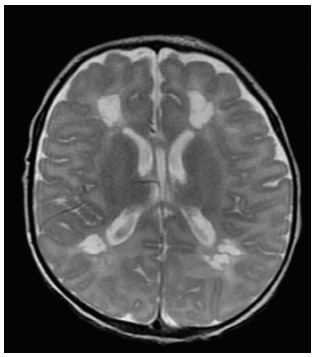


IMAGE 1

White matter Injury PVL, IVH Grade III IV, PVL corona radiata above PLIC, ventriculomegaly, arterial infarction, maldevelopment.

Complex course

Neonatal intensive care unit (NICU) admission, low birth weight, exposure to infection/ inflammation early/postnatal/ late pre-natal, seizures, necrotising enterocolitis, bronchopulmonary dysplasia, chronic neonatal lung disease, severe retinopathy of prematurity (ROP stage 4 or 5), surgery for Patent Ductus Arteriosus. Developmental and neurobehavioural risk factors - difficulty with oral feeding.

Term-born cerebral palsy risk factor

Neonatal encephalopathy

Neonatal stroke

Abnormal neuroimaging

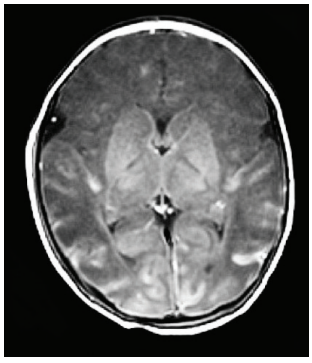


IMAGE 2

Grey matter injury basal ganglia/ thalmus, arterial infarction, haemorrhage, Combined injury white and grey matter, involvement PLIC, myelination asymmetry, congenital malformations

Complex course

NICU admission, low birth weight, intrauterine growth restriction, meconium aspiration syndrome, neonatal seizures, infections, hypoglycaemia, multiple births, surviving twin after death of a co-twin, maternal age over 35 years, low apgars, birth defects, family history of neurodevelopmental condition.

IMAGES 1 AND 2 PROVIDED BY ASSOCIATE PROFESSOR ANDREA GUZZETTA AND DR SIMONA FIORI FROM THE UNIVERSITY OF PISA

Risk Factors for Cerebral Palsy

Preterm infants

The incidence of cerebral palsy in preterm infants is 40% of cases. The risk of cerebral palsy increases as gestational age decreases[1]. The subgroup of extreme prematurity is generally considered less than 28 weeks. Up to 10% of extremely preterm infants have cerebral palsy and up to 5 % of infants born 28 to 32 weeks gestational age have cerebral palsy.[1]

Premature infants are more at risk if they have had cerebral lesions. Neuroimaging indicating abnormal white matter injury patterns of Periventricular leukomalacia and Intraventricular haemorrhage (IVH) (grades 3 and 4) and ventriculomegaly are important predictors of cerebral palsy in very preterm infants.[1-4]

Periventricular leukomalacia lesions in the corona radiate above the posterior limb of the internal capsule (PLIC) have been accurately used to predict motor prognosis.[4] Grey matter lesions are a significant predictor for severe cerebral palsy.[3,4]

Intrauterine infection and inflammation in early postnatal course and late prenatal/ early neonatal exposure to inflammation may predispose higher risk of cerebral palsy.[1]

Neonatal encephalopathy including seizures regardless of cause increases risk. Transient hypothyroxinaemia, bronchopulmonary dysplasia and necrotizing enterocolitis have also been associated with premature birth and later cerebral palsy.[1]

Preterm infants who have had surgery to repair patent ductus arteriosus or who have required home oxygen have an increased risk of cerebral palsy.

Chronic neonatal lung disease and requiring mechanically ventilated until 36 weeks postmenstrual age had least a fourfold increased risk of cerebral palsy.

The most common early developmental risk factor is difficulty with oral feeding skills.[1]

1. McIntyre, S., Morgan, C., Walker, K., & Novak, I. (2011). Cerebral palsy--don't delay. Dev Disabil Res Rev, 17(2), 114-129. doi:10.1002/ddrr.1106
2. Tran, U., Gray, P. H., & O'Callaghan, M. J. (2005). Neonatal antecedents for cerebral palsy in extremely preterm babies and interaction with maternal factors. Early Hum Dev, 81(6), 555-561. doi:10.1016/j.earlhumdev.2004.12.009
3. Beaino, G., Khoshnood, B., Kaminski, M., Pierrat, V., Marret, S., Matis, J., . . . Ancel, P. Y. (2010). Predictors of cerebral palsy in very preterm infants: the EPIPAGE prospective population-based cohort study. Dev Med Child Neurol, 52(6), e119-125. doi:10.1111/j.1469-8749.2010.03612.x
4. Himpens, E., Oostra, A., Franki, I., Van Maele, G., Vanhaesebrouck, P., & Van den Broeck, C. (2010). Predictability of cerebral palsy and its characteristics through neonatal cranial ultrasound in a high-risk NICU population. Eur J Pediatr, 169(10), 1213-1219. doi:10.1007/s00431-010-1207-6
5. Van Marter, L. J., Kuban, K. C., Allred, E., Bose, C., Dammann, O., O'Shea, M., . . . Leviton, A. (2011). Does bronchopulmonary dysplasia contribute to the occurrence of cerebral palsy among infants born before 28 weeks of gestation? Arch Dis Child Fetal Neonatal Ed, 96(1), F20-29. doi:10.1136/adc.2010.183012

Risk Factors for Cerebral Palsy

Term-Born Infants

Identified high-risk factors for cerebral palsy in the term-born subgroup indicative of commencing active routine screening assessments and surveillance:

Neonatal encephalopathy – moderate to severe Sarnat Stage 2 or 3 should be automatically described as high risk infants.[1]

Cerebral birth defect or stroke with involvement of the cerebral peduncle.[1]

Neonatal Encephalopathy

Term infants with moderate to severe neonatal encephalopathy Sarnat stage 2 or 3 and a basal ganglia/ thalamus injury have a positive predictive value for cerebral palsy of 88%[2] and should be automatically described as high-risk infants.[1]

This accounts for 1 in 4 infants with cerebral palsy who are born at term, and is more likely to be severe and involve cognitive impairments, epilepsy, speech and also more likely to affect males.[1]

Of note, less than 30% of new born encephalopathy is associated with intrapartum hypoxia (sentinel birth events , placental abruption, cord prolapse, sever intrapartum haemorrhage, severe shoulder dystocia, and tight nuchal cord).[3]

Other antenatal risk factors such as inter-uterine growth restriction, intrauterine infection, metabolic abnormalities, syndromes and birth defects have been identified in neonatal encephalopathy infants that progress to having cerebral palsy.[1]

Three quarters of children born at term who developed cerebral palsy do not have a history of newborn encephalopathy.[1]

Perinatal stroke or cerebral birth defect

Infants with a cerebral birth defect or stroke with involvement of the cerebral peduncle should be identified as high-risk.[1]

Stroke with abnormalities involving the cerebral peduncle are highly predictive of cerebral palsy positive predictive value of 78%.[4]

Perinatal arterial stroke occurs in 1.7/100 000 live births. It can result in neonatal encephalopathy but majority present after the immediate neonatal period with seizures or hemiparesis.[1]

Preeclampsia and infants who have inter-uterine growth restriction are at risk of perinatal stroke.[1]

1. McIntyre, S., Morgan, C., Walker, K., & Novak, I. (2011). Cerebral palsy--don't delay. Dev Disabil Res Rev, 17(2), 114-129. doi:10.1002/ddrr.1106
2. Shankaran, S. (2008). Prevention, diagnosis, and treatment of cerebral palsy in near-term and term infants. Clin Obstet Gynecol, 51(4), 829-839. doi:10.1097/GRF.0b013e3181870c35
3. Badawi, N., Felix, J. F., Kurinczuk, J. J., Dixon, G., Watson, L., Keogh, J. M., . . . Stanley, F. J. (2005). Cerebral palsy following term newborn encephalopathy: a population-based study. Dev Med Child Neurol, 47(5), 293-298. doi:10.1017/s0012162205000575
4. de Vries, L. S., van Haastert, I. C., Benders, M. J., & Groenendaal, F. (2011). Myth: cerebral palsy cannot be predicted by neonatal brain imaging. Semin Fetal Neonatal Med, 16(5), 279-287. doi:10.1016/j.siny.2011.04.004

Risk Factors for Cerebral Palsy

Conditions not included in cerebral palsy classification[1]

- ◯ Hypotonia as the sole neurological finding
- ◯ Transient disorders
- ◯ Spinal disease
- ◯ Motor dysfunction which results from recognised progressive brain disorder e.g. Ataxia Telangiectasia
- ◯ People with neurodevelopmental disabilities that do not primarily effect movement and posture
- ◯ Child with severely impaired cognition and no motor signs (except for some degree of hypotonicity) is not included in the cerebral palsy diagnosis
- ◯ Metabolic syndromes

1. Krageloh-Mann I, Petruch U, Weber P-M. (2005) SCPE Reference and Training Manual (R&TM). Grenoble: Surveillance of Cerebral Palsy in Europe.

Risk factors for cerebral palsy

Clinical history indicating risk for cerebral palsy

The full causal pathway is a complex interplay between several risk factors across multiple epochs. Note that as many as one third of children who are diagnosed with cerebral palsy lack traditional risk factors for cerebral palsy.

Preconception

- ◯ Previous stillbirths
- ◯ Miscarriage(s)
- ◯ Use of reproductive technology
- ◯ Low socio economic status

and/or during pregnancy

- ◯ Intra-uterine growth restriction
- ◯ Prematurity
- ◯ Maternal thyroid disease
- ◯ Pre-eclampsia
- ◯ Placental abnormalities
- ◯ Bleeds
- ◯ Infection
- ◯ Substance abuse
- ◯ Multiple births
- ◯ Birth defects

and/or perinatal

- ◯ Acute intrapartum hypoxic event
- ◯ Stroke
- ◯ Seizures
- ◯ Hypoglycaemia
- ◯ Jaundice
- ◯ Infection

and/or postnatal

- ◯ Stroke
- ◯ Infections
- ◯ Accidental and non-accidental brain injury

What are General Movements?

The young nervous system endogenously generates a variety of motor patterns such as startles, general movements, isolated limb movements, twitches, yawns, and breathing movements. General Movements (GMs) are the most effective early movement pattern to use for the functional assessment of the young nervous system.

"General Movements are part of the spontaneous movement repertoire and are present from early fetal life (nine weeks postmenstrual age) onwards until the end of the first half a year of life".[1,2,3]

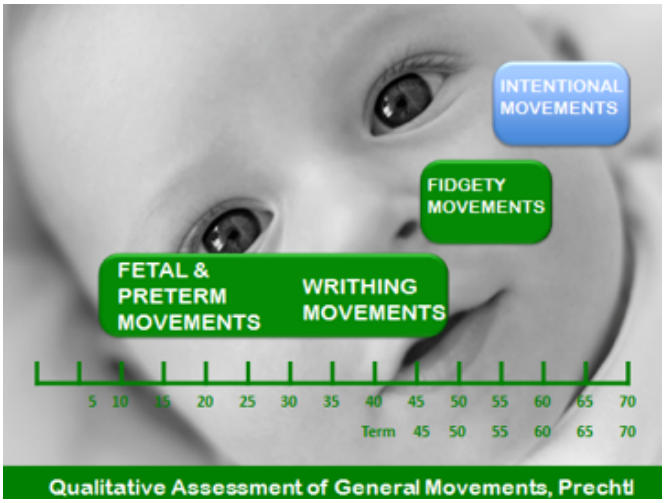
"They are complex and involve the whole body in a variable sequence of arm, leg, neck, and trunk movements. They wax and wane in intensity, force and speed and they have a gradual beginning and end. Rotations along the axis of limbs and slight changes in direction of movement make them appear fluent and elegant and create an impression of complexity and variability".[2]

A sustained and specific maturational progression of GMs is suggested as they are observable in a specific order related to the typical developmental trajectory.

The refinement of GMs occurs in two stages: normal 'writhing' movements (present until 6–9 post-term age as they gradually disappear) followed by 'fidgety' movements (emerging at 6–9 weeks post-term age although most prevalent at 12 weeks adjusted age until 16–20 weeks post-term age). Between 16-20 weeks fidgety movements start decreasing and eventually disappear, become replaced by intentional movements.

'Fidgety' movements are present up to the end of the first half a year of life when intentional and anti-gravity movements start to dominate.

Because GMs include activity of all segments from cervical to limbs to spinal cord, it is likely that the generating neuronal structure is located supraspinally. It is currently thought that the complexity and variation in GMs are generated by the cortical subplate and mediated by its motor efferent connections and can be affected by impairments to these structures.[2] There is an overlap when 'writhing' GMs fade and 'fidgety' movements emerge. It is thought that 'writhing' and 'fidgety' GMs may be generated by different central pattern generators.



Qualitative Assessment of General Movements, Prechtl

Abnormalities of GMs in young infants may be markers of more widespread injury to the brain, the overt signs and symptoms of which may evolve over time. If the nervous system is impaired, there is reduced modulation of the central pattern generators and GMs lose their complex and variable character. 'Writhing' movements become monotonous and 'poor repertoire', 'cramped' and 'synchronised' or 'chaotic'. 'Fidgety' movements can either be absent or abnormal.[2] Abnormal GMs, in particular 'absent fidgety' GMs have been shown to be highly predictive of cerebral palsy.[4]

1. General Movements Trust Training course notes.
2. Einspieler C, Prechtl H. Prechtl's Assessment of General Movements: A diagnostic tool for the functional assessment of the young nervous system. *Mental Retardation and Developmental Disabilities Research Reviews* 2005; 11: 61 – 67.
3. Prechtl H. F. R. (1990). Qualitative changes of spontaneous movements in fetus and preterm infants are a marker of neurological dysfunction. *Early Human. Dev.* 1990; 23: 151–158.
4. Bosanquet M, Copeland L, Ware R, Boyd R. A systematic review of tests to predict cerebral palsy in young children. *Dev Med Child Neurol* 2013; 55: 418-26.

Standardised motor assessment tools

What is the role of standardised motor assessments in the clinical diagnosis of high risk of cerebral palsy?

"Standardised motor assessment tools now exist in early infancy to enable accurate and early detection of high risk of cerebral palsy before the clinical observation of motor delay may be evident." Novak et al (2017)

Cerebral palsy is a clinical diagnosis based on a combination of clinical signs, neurological symptoms and motor activity limitations rather than a laboratory biomarker. Historically, early infancy was regarded as the latent or silent period where cerebral palsy could not be identified accurately.

Motor dysfunction remains an essential criteria in the clinical diagnosis of high-risk of cerebral palsy. This may be assessed by an infant having reduced quality of movements as measured on a standardised assessment (e.g. General Movements Assessment[2]) or neurologically abnormal movements (e.g. observable hand asymmetry or suboptimal HINE* scores[3]) and/or motor activities that are substantially below that expected for chronological age. This may be seen by an abnormal score on standardised motor assessment, parental or clinical observation.

Clinical observations of motor dysfunction, delay and abnormal posture can be difficult in early infancy and may become more evident as the child gets older. As voluntary movement emerges and myelination occurs, the gap between normal and abnormal movement, motor dysfunction and activity limitations become more apparent.

The General Movements Assessment is the most predictive motor assessment tool for the likelihood risk of cerebral palsy and is considered the reference standard for early detection of cerebral palsy. With established validity[4] and inter-rater reliability[4,5,6] it has predictive validity superior to neuroimaging with best sensitivity as high as 98% and specificity as high as 91% in the early months.[7]

Normal GMs, especially in concurrence with other smooth fluid movements are shown to have high correlation with normal outcome[2], whilst abnormal GMs, in particular 'cramped synchronised' GMs in the 'writhing' period (which may be transient, or present for several weeks) followed by 'absent fidgety' (F-) in the 'fidgety' period has consistently shown the highest predictive value for spastic motor type cerebral palsy.[7]

Repeated assessment throughout both the 'writhing' and 'fidgety' stages of GMs assists in prediction of later motor severity of cerebral palsy.[10] The relationship and time of appearance of 'cramped synchronised' GMs predicts the degree of later functional impairment with the earlier the appearance, the more severe the functional impairment as classified by the Gross Motor Function Classification System.[8,9,10]

* Hammersmith Infant Neurological Examinations (HINE)

1. Novak et al 2017. Early Accurate Diagnosis and Early Intervention in Cerebral Palsy. *JAMA Pediatr.* 2017; 171(9):897-907.
2. Einspieler C, Prechtl H. Prechtl's Assessment of General Movements: A diagnostic tool for the functional assessment of the young nervous system. *Mental Retardation and Developmental Disabilities Research Reviews* 2005; 11: 61 – 67.
3. Haataja L, Mercuri E, Regev R, Cowan F, Rutherford M, Dubowitz V, et al. Optimality score for the neurologic examination of the infant at 12 and 18 months of age. *J Pediatr* 1999; 135: 153-61.
4. Einspieler,C.,Prechtl,H.F.R.,Bos,A.F.,Ferrari,F.,andCioni,G.Prechtl's method on the qualitative assessment of general movements in preterm, term and young infants. *Clin. Dev. Med* 2004; 167, 1–91.
5. Spittle AJ, Doyle LW, Boyd RN. A systematic review of the clinimetric properties of neuromotor assessments for preterm infants during the first year of life. *Dev Med Child Neurol* 2008; 50: 254-66.
6. Valentin T, Uhl K, Einspieler C. The effectiveness of training in Prechtl's method on the qualitative assessment of general movements. *Early Human Development* 2005; 81:623-627.
7. Bosanquet M, Copeland L, Ware R, Boyd R. A systematic review of tests to predict cerebral palsy in young children. *Dev Med Child Neurol* 2013; 55: 418-26.
8. Bruggink JL, Cioni G, Einspieler C, Maathuis CG, Pascale R, Bos AF. Early motor repertoire is related to level of self-mobility in children with cerebral palsy at school age. *Dev Med Child Neurol* 2009; 5: 878-85.
9. Yang H, Einspieler C, Shi W, Marschik PB, Wang Y, Cao Y, et al. Cerebral palsy in children: Movements and postures during early infancy, dependent on preterm vs. full term birth. *Early Hum Dev* 2012; 88: 837-43.
10. Ferrari F, Cioni C, Einspieler C, et al. Cramped synchronised general movements in preterm infants as an early marker for cerebral palsy. *Arch Pediatr Adolesc Med* 2002;156:460–7.

General Movements Assessment

What is the General Movements Assessment?

The General Movements Assessment, originating with Professor Heinz Prechtl in the 1980s, provides an evaluation of neurological integrity of the young nervous system[1]. It is a quick, non-invasive, qualitative assessment of spontaneous general movement patterns of young infants.

Assessment is based on observation and visual gestalt perception, through the use of a video recording, of an infant in supine without environmental interference. It is a standardised motor-assessment tool for pre-term and term infants up to 5 months post-term.

Although the tool can be utilised as a single assessment in the 'fidgety' period, a developmental trajectory and serial observations of GMs are preferred and more accurate[2]. This involves documenting 2 periods of GMs: the 'writhing' period from pre-term until 6–9 week post-term age (2 or more recordings) and the 'fidgety' period from 9–20 week post-term age (2 recordings recommended between 12–16 weeks post-term age).

A score of normal or abnormal is obtained in both periods with abnormal GMs being further classified into 'poor repertoire', 'cramped synchronised' or 'chaotic' in the 'writhing' period; and 'absent fidgety' or 'abnormal fidgety' in the 'fidgety' period.

Abnormal GMs will accurately detect the likely risk of cerebral palsy, whilst a detailed developmental trajectory can indicate likely severity of cerebral palsy.[3,4]

Scoring is completed by assessors, certified through the General Movements Trust.

General Movements Basic and Advanced training and certification is obtained via attendance at the General Movements Trust approved 3.5 day course, and high performance in summative assessment concluding the course for Basic and Advanced raters.[5]

A manual and demonstration video are also available.

1. Einspieler,C.,Prechtl,H.F.R.,Bos,A.F.,Ferrari,F.,and Cioni,G.Prechtl's method on the qualitative assessment of general movements in preterm, term and young infants. 2004; *Clin. Dev. Med*; 167: 1–91.
2. Einspieler C, Prechtl H. Prechtl's Assessment of General Movements: A diagnostic tool for the functional assessment of the young nervous system. *Mental Retardation and Developmental Disabilities Research Reviews* 2005; 11: 61 – 67.
3. Bruggink JL, Cioni G, Einspieler C, Maathuis CG, Pascale R, Bos AF. Early motor repertoire is related to level of self mobility in children with cerebral palsy at school age. *Dev Med Child Neurol* 2009; 5: 878-85.
4. Yang H, Einspieler C, Shi W, Marschik PB, Wang Y, Cao Y, et al. Cerebral palsy in children: Movements and postures during early infancy, dependent on preterm vs. full term birth. *Early Hum Dev* 2012; 88: 837-43.
5. General Movements Trust website **www.general-movements-trust.info/52/video**

Prechtl's General Movements Assessment – key evidence

Systematic review evidence (of large cohort studies in high-risk mainly pre-term infants) indicates that an abnormal General Movements (GMs) assessment score of 'absent fidgety' movements (i.e. the infant does not show tiny spontaneous movements of the neck, trunk and limbs in all directions with small amplitude, moderate speed and variable acceleration, which is a biomarker for neurological integrity) at 12 weeks corrected age until the end of fidgety period is 95–98% predictive of cerebral palsy.

Furthermore any 'absent fidgety' abnormal GMs scores should trigger the need for further investigations, assessments and referral for early intervention based on 'high risk of cerebral palsy.'

Abnormal GMs accurately detects the likelihood of risk of cerebral palsy and detailed GMs assessment predicts later severity of cerebral palsy.

GMs can detect both mild and more severe forms of cerebral palsy.[1]

CITATION	#EVIDENCE	#STUDIES	#PATIENTS	ACCURACY FOR CEREBRAL PALSY	QUALITY
Bosanquet 2013	Sys. Review	6	1358	Sensitivity = 98% Specificity = 91%	14/14
Burger 2009	Sys. Review	17	1830	Sensitivity = 92% Specificity = 82%	14/14
Darsaklis	Sys. Review	39	?	Sensitivity = 100% Specificity = 100%	14/14
Heinemen 2008	Sys. Review	7	?	No data in review	14/14
Spittle 2008	Sys. Review	5	344	Sensitivity = 83-100% Specificity = 57.96%	14/14

Normal GMs are shown to have high correlation with normal outcome, whilst abnormal GMs, in particular 'cramped synchronised' in the 'writhing' period followed by 'absent fidgety' (F-), has consistently shown the highest predictive value for cerebral palsy.[2]

1. Novak et al 2017. Early Accurate Diagnosis and Early Intervention in Cerebral Palsy. *JAMA Pediatr.* 2017; 171(9):897-907.
2. Bosanquet M, Copeland L, Ware R, Boyd R. A systematic review of tests to predict cerebral palsy in young children. *Dev Med Child Neurol* 2013; 55: 418-26.

Prechtl's General Movements Assessment definitions

	NORMAL RESULT	ABNORMAL GENERAL MOVEMENTS
'WRITHING' PERIOD **Seen up to 6–9 weeks post menstrual age and gradually fade**	Characterised by small to moderate amplitude and by slow to moderate speed. Fast and large extensor movements may occasionally break through, particularly in the arms. They typically are elliptical in form and this component creates the impression of 'writhing' quality of movement.	'Poor-repertoire' (PR); sequences of successive movement are monotonous and movements of the different body parts don't occur in complex way. 'Cramped synchronised' (CS); these appear rigid and lack smooth and fluent character, all limb and trunk muscles contract and relax almost simultaneously. 'Chaotic' (Ch); movements of all limbs are in a chaotic order without any fluency or smoothness. The consistently appear to be abrupt.
'FIDGETY' PERIOD **Seen up to 9-20 weeks post menstrual age and gradually fade**	Are circular movements of small amplitude, moderate speed and variable acceleration of neck, trunk, and limbs in all directions. They are continual during the awake child except during focused attention or when fussing or crying. They may be seen with concurrent other gross movements such as kicking, wiggling/oscillating, swiping of the arms, or pleasure bursts. Initially they occur as intermittent events (scored as F+), they gradually increase in frequency (scored as F++) and then decrease once again (scored as F+).	'Fidgety' movements are judged as abnormal if they are: 'Absent fidgety' (F-); are never observed from 9–20 weeks post-term. Other movements can however be commonly observed. 'Abnormal fidgety' (FA); they look like normal 'fidgety' movements but their amplitude, speed and jerkiness are moderately or greatly exaggerated.

References:

- General Movements Trust Training course notes.
- Einspieler C, Prechtl H. Prechtl's Assessment of General Movements: A diagnostic tool for the functional assessment of the young nervous system. *Mental Retardation and Developmental Disabilities Research Reviews* 2005; 11: 61–67.
- Prechtl H. F. R. (1990). Qualitative changes of spontaneous movements in fetus and preterm infants are a marker of neurological dysfunction. *Early Human. Dev.* 1990; 23: 151–158.

What is the validity and reliability of the General Movements Assessment?

There is substantial evidence supporting strong validity and reliability of the General Movements (GMs) demonstrated in many studies.[1,2,3,4] The inter-observer reliability has been demonstrated by several groups at 90% agreement.[5]

Systematic review evidence has demonstrated of all the assessment tools available to predict high-risk of cerebral palsy, the General Movements Assessment is considered the most predictive, with the best sensitivity 98% (95% CI 74–100%) and specificity 91% (95% CI 83–93%) of predicting cerebral palsy in the 'fidgety' period, 3 months post-term age.[6,7,2] Higher predictive validity for cerebral palsy has been demonstrated with GMs than with cranial ultrasound[8] and MRI neuroimaging.[2,7]

The combination of abnormal GMs at 3 months post-term age and abnormal neuroimaging at term age (white matter injury on MRI) has shown to be 100% predictive of a later diagnosis of cerebral palsy in a cohort of very pre-term infants.[9] Studies of term infants with hypoxic ischaemic encephalopathy (HIE) also demonstrate the predictive value of GMs assessment in term infants and correlation with lesions of the basal ganglia and thalamus.[10]

In particular a general movement trajectory of 'cramped synchronised' in the 'writhing' period, followed by 'absent fidgety' in the 'fidgety' period has consistently shown the highest predictive value for cerebral palsy.[7,11]

1. Einspieler C, Prechtl H. Prechtl's Assessment of General Movements: A diagnostic tool for the functional assessment of the young nervous system. *Mental Retardation and Developmental Disabilities Research Reviews* 2005; 11: 61 – 67.
2. Spittle AJ, Doyle LW, Boyd RN. A systematic review of the clinimetric properties of neuromotor assessments for preterm infants during the first year of life. *Dev Med Child Neurol* 2008; 50: 254-66.
3. Einspieler C, Prechtl HFR, Ferrari F, Cioni G, Bos AF. The qualitative assessment of general movements in preterm, term and young infants and review of the methodology. *Early Hum Dev* 1997; 50: 47- 60
4. van Kranen-Mastenbroek V, van Oostenbrugge R, Palmans L,Stevens A, Kingma H, Blanco C, et al. Inter- and intraobserveragreement in the assessment of the quality of spontaneous movements in the newborn. *Brain Dev* 1992;14:289– 93.
5. Valentin T, Uhl K, Einspieler C. The effectiveness of training in Prechtl's method on the qualitative assessment of general movements. *Early Human Development* 2005; 81:623-627.
6. Burger M, Louw QA. The predictive validity of general movements – a systematic review. *Eur J Paediatr Neurol* 2009; 13: 408-20.
7. Bosanquet M, Copeland L, Ware R, Boyd R. A systematic review of tests to predict cerebral palsy in young children. *Dev Med Child Neurol* 2013; 55: 418-26.
8. Einspieler, C., Prechtl, H.F.R., Bos, A.F., Ferrari, F., and Cioni, G. Prechtl's method on the qualitative assessment of general movements in preterm, term and young infants. *Clin. Dev. Med* 2004; 167, 1–91.
9. Spittle A, Boyd R, Inder T, Doyle L. Predicting motor development in very preterm infants at 12 months' corrected age: The Role of Qualitative and Magnetic Resonance Imaging and General Movements Assessment. *Pediatrics* 2009; 123(2):512-7.
10. Ferrari F, Todeschini A, Guidotti I, Martinez-Biarge M, Roversi MF, Berardi A, et al.General movements in full-term infants with perinatal asphyxia are related to basal ganglia and thalamic lesions. *J Pediatr* 2011;158:904–11.
11. Darsaklis V, Snider LM, Majnemer A, Mazer B. Predictive validity of Prechtl's method on the qualitative assessment of general movements: a systematic review of the evidence. *Dev Med Child Neurol* 2011; 53: 896-906.

General Movements Assessment video instructions

1 Corrected age and consent

- The General Movement Assessment should always be done based on corrected age
- Discuss the assessment with the parents of the child, provide the parents with printed information about the production of the video, and obtain parents' written consent
- Video cannot be produced without parental/guardian consent.

Methodology for video recording

2 Position

- Position the video camera securely above the child, preferably using a tripod
- Position the child in a supine position, with the infant orientated vertically
- Start with a shot of the child's information sheet: including the date, child's name and date of birth, UR number, corrected age, and gestational age at birth. Capture this information each time a video is created
- Ensure the baby's face is in view to note the behavioral state of infant. If infant is crying or sleeping, stop and try another time. Infant should be awake and calm for the video.
- Ensure there are no toys or other distractions in the immediate environment
- In the event of a strong head preference, reposition the head to midline position during the assessment period
- If postural instability, video in and out of nested position for comparison.

When recording in a Neonatal Intensive Care Unit (NICU)/Special Care Nursery (SCN) setting:

- remove blankets and clear bed area as much as possible to expose hands and feet (arms and legs if possible)
- ensure developmentally supportive care in place
- limit handling and disturbing the child unnecessarily
- the child should only wear a nappy if possible

When in an outpatient clinic setting, use a white sheet as background to ensure no other objects are captured in the video.

General Movements Assessment video instructions

3 Behavioural

- Video in active wakefulness – States 4 or 5
- Don't video when crying or fussing or State 1 sleep (regular respirations)
- Provide support and calm as required
- Time video to coincide with wakeful times e.g. feeding or baths. Liaise with parents and nurses
- Read the child's cues to ensure optimal state regulation – be careful of yawning
- Avoid videoing when child has prolonged hiccuping.

4 Environment (optimally)

Avoid:

- Interference by observer (keep away from the child, don't interact with parents)
- Noisy surrounds – limit discussions
- Toys and distractions
- Parent in field of vision
- Colourful blankets
- Mirrors
- Distractions on clothing e.g. staff ID badge
- Dummy in mouth.

5 Timing

- In the 'writhing' period: 5–15 minutes (may need to record up to 15 minutes of video)
- In the 'fidgety' period: 3-5 minutes of optimal recording
- Avoid videoing in the first days after birth, especially if the child was born extreme premature.

STATE	DESCRIPTION	BEHAVIOUR
State 1	Deep sleep	Lies quietly without moving
State 2	Light sleep	Moves while sleeping; startles at noises
State 3	Drowsiness	Eyes start to close; may doze
State 4	Quiet alert	Eyes open wide, face is bright; body is quiet
State 5	Active alert	Face and body move actively
State 6	Crying	Cries, perhaps screams; body moves in very disorganised ways

CLICK TO VIEW VIDEO

The General Movements Assessment

What about false positives and false negatives?

The Australian Cerebral Palsy Register (ACPR) indicates less than 5% of cerebral palsy diagnosis are false positive results with standardised tools.[1]

Almost all false positives result in the infant being diagnosed with a different neurological disability (e.g. intellectual disability, autism spectrum disorder) not a typical outcome.[2] False negatives resulting in late diagnosis and late intervention are detrimental to parents and infants.[3]

The General Movements Assessment has also been discussed in the literature in relation to other high-risk populations for developmental disorders and childhood disabilities.

Further research is required to better understand the predictive power of the General Movement Assessment in relation to other developmental disorders and childhood disabilities.

It is known that infants with cerebral palsy require and benefit from different evidence-based early interventions for infants 'at risk of developmental delay', or 'at risk of autism', or 'at risk of harm', or with 'social risk'. When the clinical diagnosis is unclear but the infant is perceived to be at risk of cerebral palsy, from either their perinatal history or clinical assessment data, the infant should be referred to intervention and then regular monitoring should commence to assist with forming a diagnostic picture.[3]

To reduce the likelihood of a false negative or positive, it is recommended to use the combination of standardised tools rather than any single assessment in isolation, as outlined in the clinical diagnostic pathway algorithm in the International Guidelines.

This represents the coupling of best available evidence tools with the best psychometric properties for the aim of accurate and early detection of high-risk of cerebral palsy and exclusion of differential diagnosis.

Does the General Movements Assessment have Automated Scoring?

Research into the automated analysis of General Movements using sensor technologies and computer based tools is underway. Although promising, this technology has not yet advanced to clinical practice.[4]

1. Report of the Australian Cerebral Palsy Register, Birth Years 1993-2006, February 2013. Sydney; Cerebral Palsy Alliance.
2. Nelson KB. Causative factors in cerebral palsy. *J Clin Gynecol Obstet* 2008; 5: 749-62.
3. Novak et al 2017. Early Accurate Diagnosis and Early Intervention in Cerebral Palsy. *JAMA Pediatr.* 2017; 171(9):897-907.
4. Marcroft, C., Khan, A., Embleton, N. D., Trenell, M., and Plötz, T. (2015). Movement recognition technology as a method of assessing spontaneous general movements in high risk infants. Front. Neurol. 5:284. doi: 10.3389/fneur.2014.00284

The General Movements Assessment

How to order an assessment

ASSESSMENT	TIME TO ADMINISTER	MANUAL	EQUIPMENT	TRAINING REQUIRED
General Movements	10–30 minutes 5–15 minutes to film 5–15 minutes to score at another time point	YES: DVD from General Movements Trust	Video, BabyMoves app	YES: Rater certification required. Source: **http://general-movements-trust.info** Video data can be collected by non-certified raters and scored remotely by trained raters

The General Movements Assessment is simply a video of an infant lying on their back whilst they are calm and alert taken before 16 weeks post-term age. It is non-invasive, non-disruptive to infants and relatively inexpensive.

Parent or carer education, counselling and informed consent is required prior to taking a video.

Clinicians or parents are able to record the video.

In addition to standard video capture, the BabyMoves app has been developed by Murdoch Children's Research Institute (MCRI) to capture videos via smartphone.[1]

Videos are scored retrospectively by at least two certified raters.

General Movements Basic and Advanced training and certification is obtained via attendance at Basic and Advanced Courses delivered by the General Movements Trust.

1. Spittle A, Olsen J, Kwong A, Doyle LW, Marschik PB, Einspieler C, Cheong J. The Baby Moves prospective cohort study protocol: using a smartphone application with the General Movements Assessment to predict neurodevelopmental outcomes at age 2 years for extremely preterm or extremely low birthweight infants. *BMJ Open* 2016; 6:e013446.

The General Movements Assessment

How to refer an infant for Prechtl's General Movements Assessment or have a video scored by a certified trainer in Australia?

For information about how to refer an infant for Prechtl's General Movements Video or have a video scored by a certified trainer in your area please follow the instructions for your state.

Cathy Morgan
CMorgan@cerebralpalsy.org.au

Alicia Spittle
aspittle@unimelb.edu.au

Jo George
j.george2@uq.edu.au

Kirstie Morgan
Kirstie.Morgan@sa.gov.au

Katherine Langdon
Katherine.Langdon@health.wa.gov.au

Kelly Paterson
Kelly.Paterson@nt.gov.au

Eliza Maloney
eliza.maloney@ths.tas.gov.au
Christy Dorward
Christy.dorward@ths.tas.gov.au

IN ADDITION

Queensland Early Detection and Intervention Network – Cerebral Palsy (QEDIN-CP): For clinical screening of infants at risk of CP and potential referral to early intervention clinical trials. Referral form on QEDIN website.

QEDIN Website

The Cerebral Palsy Alliance Early Diagnosis Clinic is a bulk-billed diagnostic clinic for babies at high risk of cerebral palsy. The clinic aims to fast track diagnosis and enable quicker access to early interventions, family support and better outcomes for the future.

Referral form

The General Movements Assessment

Can Prechtl's General Movements Assessment be performed via Telehealth?

The General Movements Assessment can be performed remotely by parents using the Baby Moves Smartphone application. This has been designed for parents to video their infants' spontaneous movements to enable remote scoring of the General Movements Assessment. The Baby Moves application provides reminders and instructions to capture General Movements videos.

The Baby Moves smartphone application is available for parents who participate in research trials.

For information about how to refer parents to use the Baby Moves App in your area please contact:

Cathy Morgan
CMorgan@cerebralpalsy.org.au

Alicia Spittle
aspittle@unimelb.edu.au

Jo George
j.george2@uq.edu.au

The Baby Moves App is currently not available in South Australia but other approved data-sharing platforms are in clinical use.

Kirstie Morgan
Kirstie.Morgan@sa.gov.au

The Baby Moves App is currently not available in Western Australia but other approved data-sharing platforms are in clinical use.

Katherine Langdon
Katherine.Langdon@health.wa.gov.au

The Baby Moves App is currently not available in the Northern Territory but other approved data-sharing platforms are in clinical use.

Kelly Paterson
Kelly.Paterson@nt.gov.au

The Baby Moves App is currently not available in Tasmania but other approved data-sharing platforms are in clinical use.

Eliza Maloney
eliza.maloney@ths.tas.gov.au
Christy Dorward
Christy.dorward@ths.tas.gov.au

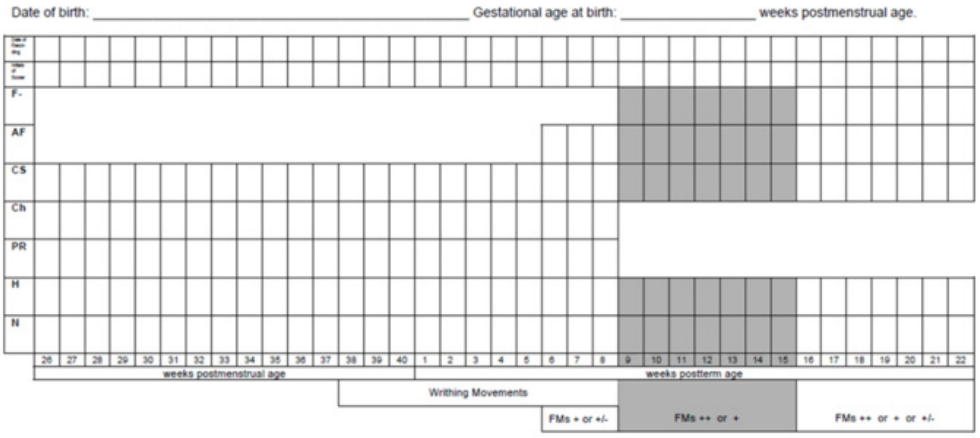
IN ADDITION

Queensland Early Detection and Intervention Network – Cerebral Palsy (QEDIN-CP): For clinical screening of infants at risk of CP and potential referral to early intervention clinical trials. Referral form on QEDIN website. QEDIN Website

The Cerebral Palsy Alliance Early Diagnosis Clinic is a bulk-billed diagnostic clinic for babies at high risk of cerebral palsy. The clinic aims to fast track diagnosis and enable quicker access to early interventions, family support and better outcomes for the future. Referral form

The General Movements Assessment: interpreting the results

What does a Prechtl's method 'General movement assessment individual developmental trajectory' look like?



N = normal age-specific GMs

FMS = 'fidgety' movements

H = hypokinesia (no GMs during the recording

PR = 'poor repertoire' of GMs

CH = 'chaotic' GMs

CS = 'cramped synchronised' GMs

AF = abnormal 'fidgety' movements

F- = absence of 'fidgety' movements

In the 'writhing' period abnormal GMs known as 'cramped synchronised' are highly predictive of spastic motor type cerebral palsy[1,2]. Negative predictive value for 'cramped synchronised' movements alone is shown to be high at 62-80% and positive predictive value ranging 87–100% for later spastic cerebral palsy.[3]

In high risk populations, 'cramped synchronised' GMs followed by 'absent fidgety' GMs in the 12–16 weeks post-term age has the highest predictive value for cerebral palsy with sensitivity 95–100%.[2,4]

An abnormal score of 'absent fidgety' GMs whether preceded by 'poor repertoire' or 'cramped synchronised' movements meets the essential criteria of motor dysfunction.

If there is additional criteria of abnormal neuroimaging and/or clinical history indicating risk of cerebral palsy the interim clinical diagnosis of 'high-risk of cerebral palsy' should be sensitively discussed with parents accompanied by referrals to cerebral palsy-specific early intervention services and parental emotional supports.

An abnormal GMs score of 'abnormal fidgety' in the 'fidgety' period (9–20 weeks post-term age) is more rare but may indicate a possible increased risk of neurological condition[5]. Referral for early intervention should be considered and ongoing developmental follow up including motor and cognitive development.

A GMs score of normal in the 'fidgety' period (9–20 weeks post-term age) can be considered low risk of cerebral palsy, ongoing developmental follow up may be required including motor and cognitive development.

CAVEAT

In rare cases, normal 'fidgety' movements do not preclude an adverse outcome; especially in mild unilateral cerebral[6].

In infants with milder cerebral palsy, especially unilateral cerebral palsy, it is possible for an infant to score within the normal range on a standardised assessment of motor performance whilst still displaying abnormal movements.

1. Spittle A, Boyd R, Inder T, Doyle L. Predicting motor development in very preterm infants at 12 months' corrected age: The Role of Qualitative and Magnetic Resonance Imaging and General Movements Assessment. *Pediatrics* 2009; 123(2):512-7.
2. Bosanquet M, Copeland L, Ware R, Boyd R. A systematic review of tests to predict cerebral palsy in young children. *Dev Med Child Neurol* 2013; 55: 418-26.
3. Ferrari F, Cioni G, Einspieler C, Roversi MF, Bos AF, Paolicelli PB, Ranzi A, Prechtl HFR. Cramped Synchronized General Movements in Preterm Infants as an Early Marker for Cerebral Palsy. *Arch Pediatr Adolesc Med.* 2002;156(5):460-467.
4. Synchronized General Movements in Preterm Infants as an Early Marker for Cerebral Palsy. *Arch Pediatr Adolesc Med.* 2002;156(5):460–467.
5. Darsaklis V, Snider LM, Majnemer A, Mazer B. Predictive validity of Prechtl's method on the qualitative assessment of general movements: a systematic review of the evidence. *Dev Med Child Neurol* 2011; 53: 896-906.
6. Einspieler C, Peharz R, Marschik P. Fidgety movements – tiny in appearance, but huge in impact. *J Pediatr* (Rio J). 2016;92(3 Suppl 1):S64---S70.
7. Einspieler C, Yang H, Bartl-Pokorny KD, Chi X, Zang FF, Marschik PB, et al. Are sporadic fidgety movements as clinically relevant as is their absence? *Early Hum Dev* 2015; 91: 247-52.

Cerebral Palsy ALLIANCE

Centre of Research Excellence Cerebral Palsy

EARLY DIAGNOSIS – CEREBRAL PALSY

Cerebral palsy early diagnosis interpretation quick reference

GENERAL MOVEMENTS 11

General Movements Assessment

General Movements Assessment, to identify motor dysfunction

SCREENING TIME PERIOD	NORMAL RESULT	ABNORMAL RESULT
'Writhing' Up to 6–9 weeks post-term age	**Normal** Continue ongoing development follow up including motor and cognitive development	'Poor repertoire' (not predictive cerebral palsy)
		'Cramped synchronous' (predictive if persistent)
		'Chaotic' (rare and non predictive)
'Fidgety' Seen from about 9 weeks post-term age up to about 20 weeks Best assessed between 12-16 weeks post-term age Two recordings in the 'fidgety' period are recommended	**Low risk for cerebral palsy** Continue ongoing developmental follow up including motor and cognitive development	'Absent fidgety' (F-) High-risk cerebral palsy Referral for early intervention and parent supports
		'Abnormal fidgety' (AF) Less common. Possible increased risk of neurological condition Ongoing developmental follow up and consider referral for early intervention

Prediction of motor type and topography[1]

PRE-TERM GMs	'WRITHING' GMs (TERM-8 WEEKS)	'FIDGETY' GMs (3-5 MONTHS)	OUTCOME
'Poor repertoire' OR normal	'Poor repertoire' OR normal	Normal	Normal
'Poor repertoire' or 'cramped synchronised' GMs	'Cramped synchronise' GMs	'Absent fidgety' + abnormal neuro exam	Bilateral spastic cerebral palsy
'Poor repertoire' or 'cramped synchronised' GMs	'Poor repertoire' or 'cramped synchronised' GMs	'Absent fidgety' GMs + asymetrical segemental movements +/- abn neuro exam	Unilateral spastic cerebral palsy
'Poor repertoire' GMs	'Poor repertoire' GMs; circular arm movements with finger spreading	'Absent fidgety'; absence of foot-to-foot contact; circular arm movements and finger spreading	Dyskenetic cerebral palsy

1. Einspieker et al 2012

Cerebral Palsy ALLIANCE

Centre of Research Excellence Cerebral Palsy

General Movements Assessment – Interpreting the results

Severity

Repeated assessment throughout both the 'writhing' and 'fidgety' stages of General Movements (GMs) assists in prediction of later motor severity of cerebral palsy.[1]

The relationship and time of appearance of 'cramped synchronised' GMs predicts the degree of later functional impairment with the earlier the appearance, the more severe the functional impairment as classified by the Gross Motor Function Classification System.[2]

Unilateral cerebral palsy

Unilateral cerebral palsy may typically show abnormal GMs, usually 'poor repertoire' or 'cramped synchronised' in their first weeks of life followed by 'absent fidgety' at 12–14 weeks post-term age. At the age of 2–4 months the first asymmetries can be observed in the distal segmental movements, reduced or absent on the contralateral side of the lesion.[4,5]

Dyskinetic cerebral palsy

Typically 'poor repertoire' in the 'writhing' period, moving arms in circular movements and spreading their fingers. These movements can be present until 5 months, are unilateral or bilateral and monotonous, slow forward rotations starting at the shoulder. From 3–5 months 'fidgety' movements and movements towards the midline e.g. foot to foot were absent.[6]

1. Ferrari F, Cioni C, Einspieler C, et al. Cramped synchronised general movements in preterm infants as an early marker for cerebral palsy. *Arch Pediatr Adolesc Med* 2002;156:460–7.
2. Bruggink JL, Cioni G, Einspieler C, Maathuis CG, Pascale R, Bos AF. Early motor repertoire is related to level of self mobility in children with cerebral palsy at school age. *Dev Med Child Neurol* 2009; 5: 878-85.
3. Yang H, Einspieler C, Shi W, Marschik PB, Wang Y, Cao Y, et al. Cerebral palsy in children: Movements and postures during early infancy, dependent on preterm vs. full term birth. *Early Hum Dev* 2012; 88: 837-43.
4. Guzzetta A, Mercuri E, Rapisardi G, Ferrari F, Roversi MF, Cowan F, et al. General movements detect early signs of hemiplegia in term infants with neonatal cerebral infarction. *Neuropediatrics* 2003; 34: 61-6.
5. Cioni G, Bos A, Einspieler C, Ferrari F, Martijn A, Paolicelli P, Rapisardi G, Roversi M, Prechtl HFR. Early neurological signs in preterm infants with unilateral intraparenchymal echodensity. *Neuropediatrics* 2000; 31: 240-251.
6. Einspieler C, Prechtl H. Prechtl's Assessment of General Movements: A diagnostic tool for the functional assessment of the young nervous system. *Mental Retardation and Developmental Disabilities Research Reviews* 2005; 11: 61 – 67.

Cerebral Palsy ALLIANCE

Centre of Research Excellence Cerebral Palsy

Communicating results

Communicating results with parents or carers and the multidisciplinary team?

A General Movements (GMs) video cannot be taken without appropriate carer consent. Discussion regarding the assessment needs to take place with parents and a member or the treating multidisciplinary team to gain informed consent prior to videoing.

Newborn care 'writhing' period

Videos taken in the Neonatal Intensive Care Unit (NICU) or Special Care Nursery (SCN)

After review and scoring of videos by GM certified clinicians (at least 2) feedback should be given within 2 weeks to:

- Neonatologist, treating physician or multidisciplinary team
- Parents
- Medical and electronic medical records.

If an abnormal result of 'cramped synchronised' (predictive if persistent) or 'chaotic' (rare and not predictive) is found, a repeated video/s in the 'writhing' period should be performed in 1–2 weeks if abnormal movements persist.

If a normal result is found in the 'writhing' period, no further videos are required in the 'writhing' period and the baby can be filmed again in the 'fidgety' period (9–16 weeks corrected age) as an outpatient or via the BabyMoves app.

'Fidgety' period

When normal 'fidgety' GMs are scored by two GM certified clinicians (with one blinded to the clinical history), parents should be given reassurance of a low risk of cerebral palsy and advised to continue ongoing developmental follow up.

The result of 'absent fidgety' GMs should be communicated with the treating multidisciplinary team inclusive of neonatologists and paediatrician by the GM certified clinicians. Abnormal results of GMs should form part of the clinical reasoning with the combination of clinical history indicating risk for cerebral palsy and/or neuroimaging findings to inform the interim clinical diagnosis of 'high-risk of cerebral palsy'.

Informing parents on the interim clinical diagnosis of 'high-risk of cerebral palsy' should involve the multidisciplinary team in a sensitive, compassionate and well-planned way and always be accompanied by referrals to cerebral palsy-specific early intervention services, parental emotional supports and ongoing medical follow up. See fact sheet on 'Communicating diagnosis' and refer parents to 'Parent fact sheet'.

Data management of General Movements Assessment videos

Governance

Informed consent from parents/caregivers is required prior to capture of any General Movements (GMs) videos.

Clinical digital imaging forms part of the patient medical records.

Any cloud secure clinical data content share system and database needs to meet legislative and information management security classification requirements.

Consultation with Health Information Management, Clinical Information Management and Information Communication Technology stakeholders may need to be considered in the implementation of GMs programmes in local areas.

Hammersmith Infant Neurological Examination (HINE)

The Hammersmith Infant Neurological Examination (HINE) is recommended in the *International Clinical Practice Early Diagnosis of Cerebral Palsy Guidelines*, particularly in situations where the most predictive tools (General Movements and MRI) are not able to be used.

The HINE can assist in the early detection, diagnosis and prognosis of infants at risk of developing cerebral palsy. It can be used on infants aged between 2–24 months of age.

What is the HINE?

The HINE is a simple, scoreable, standardised clinical neurological examination for infants between 2 and 24 months of age. Specific cut-off scores for predicting cerebral palsy both in pre-term and full-term infants have been published.

- The HINE has good sensitivity and high predictive value for risk of cerebral palsy in high risk populations under 5 months.
- A HINE score < 57 at 3 months 96% predictive of cerebral palsy (sensitivity 96%; specificity 87%).[1]
- Over 5 months age corrected for prematurity it has 90% predictive accuracy for detecting the risk of cerebral palsy.[2,3]
- It provides objective information about likely motor severity and distribution of cerebral palsy.[1] Scores below 40 predict non-ambulant cerebral palsy.
- It provides information on other aspects of neurological function other than motor.
- It has good inter-observer reliability for all levels of clinical experience.[3,5,6]

Performing and scoring the HINE

There are three parts to the HINE: a neurological examination (which is scored), developmental milestones and behaviour (which are not scored).

The scoreable neurological examination is comprised of 26 items divided into 5 domains, assessing cranial nerve function, posture, quality and quantity of movements, muscle tone, and reflexes and reactions.

Each item is scored individually (0, 1, 2 or 3). The maximum score for any one item is a score of 3 and the minimum is a score of 0.

A subscore can be given for each section and the overall global score can be calculated by summing up all 26 items (range: 0–78), with higher scores indicating better neurological performance.

The maximum global score is 78.

1. Romeo DM, Cioni M, Scoto M, Mazzone L, Palermo F, Romeo MG. Neuromotor development in infants with cerebral palsy investigated by the Hammersmith Infant Neurological Examination during the first year of age. *Eur J Paediatr Neurol* 2008; 12: 24-31.
2. Pizzardi A, Romeo DM, Cioni M, Romeo MG, Guzzetta A. Infant neurological examination from 3 to 12 months: predictive value of the single items. *Neuropediatrics* 2008; 39: 344-6.
3. Romeo DM, Ricci D, Brogna C, Mecuri E. Use of the Hammersmith Infant Neurological Examination in infants with cerebral palsy: a critical review of the literature. *Dev Med Child Neurol* 2015. doi:10.1111/dmcn.12876.
4. Romeo DM, Cioni M, Palermo F, Cilauro S, Romeo MG. Neurological assessment in infants discharged from a neonatal intensive care unit. *Eur J Paediatr Neurol* 2013; 17: 192-8.
5. Haataja L, Mercuri E, Regev R, Cowan F, Rutherford M, Dubowitz V, et al. Optimality score for the neurologic examination of the infant at 12 and 18 months of age. *J Pediatr* 1999; 135: 153-61.
6. N.L. Maitre, O. Chorna, D.M. Romeo, A. GuzzettaImplementation of the Hammersmith Infant Neurological Examination in a high-risk infant follow-up program. *Pediatr Neurol* 2016: 65: 31-38.

EARLY DIAGNOSIS – CEREBRAL PALSY

Hammersmith Infant Neurological Examination (HINE)

HINE 01

How long does the HINE take?

The examination takes 10–15 minutes to perform.

Do I need certified training to use the HINE in clinical practice?

No you do not need certified training to use the HINE in clinical practice.

HINE is predictive of cerebral palsy

HINE scores at 3 months:

- <57 is 96% predictive of cerebral palsy
- <40 never occurs in children with normal outcomes[1,4]

HINE scores at (6, 9, 12 months):

- 90% predictive of cerebral palsy
- <73 predictive of cerebral palsy
- <40 almost always indicates cerebral palsy[3,4]

HINE is predictive of severity and topography of cerebral palsy

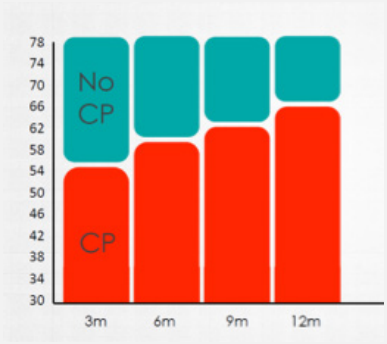
- Motor severity of cerebral palsy under two years of age is most accurately predicted using the HINE

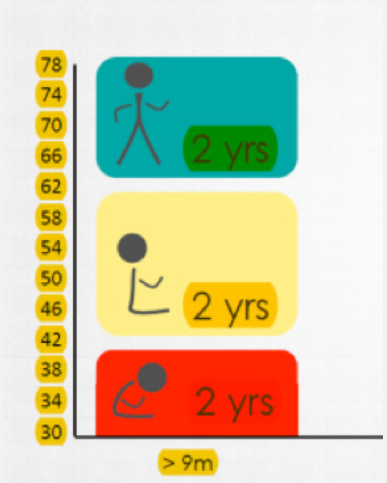
HINE scores at 3, 6, 9 or 12 months:

- 50-73 indicates likely unilateral cerebral palsy (i.e. 95-99% will walk)
- <50 indicates likely bilateral cerebral palsy

HINE scores at 3-6 months:

- 40-60 indicates likely GMFCS I-II
- <40 indicates likely GMFCS III-V





Click here to each the official Hammermith Website for resources, a video example and scoring forms.

Detailed face-to-face and live online trainings are avaialble through the BornTogether Project.

1. Romeo DM, Cioni M, Scoto M, Mazzone L, Palermo F, Romeo MG. Neuromotor development in infants with cerebral palsy investigated by the Hammersmith Infant Neurological Examination during the first year of age. *Eur J Paediatr Neurol* 2008; 12: 24-31.
2. Pizzardi A, Romeo DM, Cioni M, Romeo MG, Guzzetta A. Infant neurological examination from 3 to 12 months: predictive value of the single items. *Neuropediatrics* 2008; 39: 344-6.
3. Romeo DM, Ricci D, Brogna C, Mecuri E. Use of the Hammersmith Infant Neurological Examination in infants with cerebral palsy: a critical review of the literature. *Dev Med Child Neurol* 2015. doi:10.1111/dmcn.12876.
4. Romeo DM, Cioni M, Palermo F, Cilauro S, Romeo MG. Neurological assessment in infants discharged from a neonatal intensive care unit. *Eur J Paediatr Neurol* 2013; 17: 192-8.
5. Haataja L, Mercuri E, Regev R, Cowan F, Rutherford M, Dubowitz V, et al. Optimality score for the neurologic examination of the infant at 12 and 18 months of age. *J Pediatr* 1999; 135: 153-61.
6. N.L. Maitre, O. Chorna, D.M. Romeo, A. GuzzettaImplementation of the Hammersmith Infant Neurological Examination in a high-risk infant follow-up program. *Pediatr Neurol* 2016: 65: 31-38.

Cerebral Palsy ALLIANCE

Centre of Research Excellence Cerebral Palsy

Training

You DO NOT need certification to use or score the Hammersmith Infant Neurological Examination (HINE) but training is advised.

HINE scoring form

Online training to perform and score and interpret the HINE are available

Face to face HINE training workshops are available for workplaces.

Guidance notes to accompany the HINE face to face course training are available.

Where to find a HINE trainer in your area to perform a workshop in your workplace

Cathy Morgan
CMorgan@cerebralpalsy.org.au
Cathryn Crowle
cathryn.crowle@health.nsw.gov.au
Iona Novak
inovak@cerebralpalsy.org.au
Michelle Juarez
michelle.juarez@health.nsw.gov.au

Alicia Spittle
]aspittle@unimelb.edu.au
Joy Olsen
Joy.Olsen@thewomens.org.au
Catherine Hough
Catherine.Hough@monashhealth.org
Abbey Eeles
abbey.eeles@mcri.edu.au
Catherine Clancy
Catherine.Clancy@monashhealth.org

Jo George
j.george2@uq.edu.au
Rachel Thomas
Rachel.Thomas@health.qld.gov.au
Kym Morris
k.morris@uq.edu.au
Carly Dickinson
carly.dickinson@uq.edu.au
Bernadette Shannon
bernadette.shannon@uq.edu.au

Felicity Baker
Felicity.Baker@sa.gov.au
Kirstie Morgan
Kirstie.Morgan@sa.gov.au

Katherine Langdon
Katherine.Langdon@health.wa.gov.au
Jane Valentine
jane.valentine@health.wa.gov.au

Kelly Paterson
Kelly.Paterson@nt.gov.au

Eliza Maloney
eliza.maloney@ths.tas.gov.au
Christy Dorward
Christy.dorward@ths.tas.gov.au

EARLY DIAGNOSIS – CEREBRAL PALSY

Hammersmith Infant Neurological Examination: How to refer

HINE 01

How can I refer an infant for the Hammersmith Infant Neurological Examination?

For information about how to refer an infant for the Hammersmith Infant Neurological Examination (HINE) in your area please follow the instructions for your state or territory.

Cathy Morgan
CMorgan@cerebralpalsy.org.au

Alicia Spittle
aspittle@unimelb.edu.au

Jo George
j.george2@uq.edu.au

Kirstie Morgan
Kirstie.Morgan@sa.gov.au

Katherine Langdon
Katherine.Langdon@health.wa.gov.au

Kelly Paterson
Kelly.Paterson@nt.gov.au

Eliza Maloney
eliza.maloney@ths.tas.gov.au
Christy Dorward
Christy.dorward@ths.tas.gov.au

IN ADDITION

Queensland Early Detection and Intervention Network – Cerebral Palsy (QEDIN-CP): For clinical screening of infants at risk of CP and potential referral to early intervention clinical trials. Referral form on QEDIN website.

QEDIN Website

The Cerebral Palsy Alliance Early Diagnosis Clinic is a bulk-billed diagnostic clinic for babies at high risk of cerebral palsy. The clinic aims to fast track diagnosis and enable quicker access to early interventions, family support and better outcomes for the future.

Referral form

Cerebral Palsy ALLIANCE

THE UNIVERSITY OF SYDNEY

AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

HINE – Interpreting the results: scoring cut points

Hammersmith Infant Neurological Examination Scoring Cut Points

Scoring Cut Points Author: Dr Joanne George, The University of Queensland

Section 1: Median and range of HINE scores in different populations

Table 1: Data from cohorts of typically developing infants. Data are global score median and range

STUDY	n	Corrected age (months)				
		3	4.5 - 5.5	5.5 - 7	12	18
Haataja 2003[1]	74	67 (62.5-69)	70 (61.5-74)	73 (69-76.5)	73 (63-78)	
Haataja 1999	92 (12mo) 43 (18mo)				76 (63-78)	76 (63-78)

Table 2: Global scores median (range) in low-risk very preterm (≤32 weeks), late preterm (33-36 weeks) and term born infants with normal neurodevelopmental outcomes at 2 years and without cerebral palsy (Romeo et al[3])

STUDY	AGE	3 MONTHS	6 MONTHS	9 MONTHS	12 MONTHS
Romeo 2016[3]	Term infants (n=69)	65.5 (62-69)	69 (64-74)	72.5 (65-78)	74 (65-78)
Haataja 2003[1]	Term infants (n=74)	67 (62.5-69)	70 (61.5-74) [slightly younger than 6 months]		73 (63-78)
Romeo 2016[4]	Late preterm infants (n=71)	62 (57-69)	66 (60-72)	71 (63-75)	73 (64-77)
Romeo 2016[4]	Very preterm infants (n=48)	62 (51-67)	66 (52-71)	70 (57-76)	72 (60-77)

Hammersmith Infant Neurological Examination Scoring Cut Points

Scoring Cut Points Author: Dr Joanne George, The University of Queensland

Section 2:

Table 3: Cut points for lowest 10th percentile in typically developing children at 12 and 18 months of age

STUDY	n	OUTCOME	12	18
Haataja 1999[2]	92 (12mo); 43 (18mo)	Optimal (≥10th %) vs suboptimal (<10th %)	≥73	≥74

Table 4: Scoring cut points predicting cerebral palsy, motor function, ambulation

STUDY	POPULATION	n	OUTCOME	Corrected age (months)					
				3	6	9	12	15	18
Romeo 2013[5]	NICU (preterm & term) (149 very preterm, 754 late preterm, 638 term. Outcome: 1150 normal, 321 mild disability, 70 CP.)	1541	CP	<57 se 96%, sp 85%	<60 se 90%, sp 89%	<63 se 90%, sp 91%	<66 se 91%, sp 90%		
			Severe CP (quad/ diplegia/ diskinetic)	<40 se 100%, sp 99%	<42	<46	<47		
			Hemiplegia	<50 se 10% sp 99%	<56	<62	<64		
			General comments	<40 only seen in severe CP; Mild disability (no CP but low PDI &/or mild neuro signs) seen in infants with normal & abnormal HINE scores. Global suboptimal scores, when not associated with CP, were associated with minor neurological signs or low psychomotor development					
Romeo 2016[7]	Systematic review	3452 (831 Term, 2621 preterm)	CP	<57 (~90sens/ spec)			<66 (~90sens/ spec)		
				<40 = only found in association with severe CP					
Haataja 2001[8]	Term HIE	53	CP			<73 se 100%, sp 91%			
			Motor function at 2 years			≥67 97% indep walking; 40-66 100% sitting, 13% indep walking; <40 no sitting/walking			
			Motor function at 4 years			≥67 97% walking without restrictions; 40-66 25% walking without restrictions, 13% walking with assistive mobility, 62% no walking but some crawling/rolling; <40 no sitting/walking			

HINE – Interpreting the results: quick reference guide

STUDY	POPULATION	n	OUTCOME	Corrected age (months)					
				3	6	9	12	15	18
Frisone 2002[9]	Preterm <31wks	74	CP		<64 se 98%, sp 85%				
			Motor function at 2 years		>64 walk indep sens 98% spec 85% 52-64 sit indep <52 no indep sit or walking at 2 years				
Ricci 2006[10]	Term NE	15 (all CP)	CP						
Ricci 2006[11]	cPVL (23/24 preterm)	24 (18 CP, 6 no CP)	Mean (range)			<73 se 100%, sp 91%			
			Motor function at 2 years		>60 = walk indep; 41-60 = sitting but not walking; <40 = severe motor impairment (no indep sit)				
					increased neck & trunk extensor tone, & a posture of flexed arms & extended legs, abnormal arm protection & forward parachute reaction = no indep sit @ 2years; truncal hypotonia & extended arms & legs = indep sit but no walking				
Romeo 2008[12]	preterm	903	CP	<57 se 96%, sp 87					
				>50 & F- = hemiplegia; <50 & F- = mild to svere CP (diplegia or quadriplegia					
Pizzardi 2008[13]	Preterm & NE	658	CP						
Romeo 2008[6] (see table 4)	CP (all infants in cohort had CP	70	CP	>60 = GMFCS I; 40-60 = GMFCS I & II; <40 = GMFCS IV & V diplegia compared to quadriplegia = better scores in tone (scarf sign, pop. angle, adductors, pull to sit, ventral suspension) & posture (trunk & leg in sitting) Infants with hemiplegia scored much higher than those with diplegia & quadriplegia in all subsections & global scores, showed overlap of scores with infants with a normal outcome = HINE not sensible tool in identifying unilateral impairment.					
Gkoltsiou 2008[14]	Kernicterus	11	CP				<73 se 100%, sp 50%		
Romeo 2009[15]	preterm	103	Independent walking at 2 years	<51 se 93%, sp 100%	<53 se 93%, sp 100%	<60 se 93%, sp 95%	<61 se 93%, sp 95%		

Key: NE neonatal encephalopathy

Prepared by Dr Joanne George, The University of Queensland, Queensland Cerebral Palsy and Rehabilitation Research Centre (QCPRRC)

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

HINE – Interpreting the results: quick reference guide

1. Haataja L, Cowan F, Mercuri E, Bassi L, Guzzetta A, Dubowitz L. Application of a scorable neurologic examination in healthy term infants aged 3 to 8 months. J Pediatr 2003; 143: 546.
2. Haataja L, Mercuri E, Regev R, et al. Optimality score for the neurologic examination of the infant at 12 and 18 months of age. J Pediatr 1999; 135: 153-61.
3. Romeo DM, Brogna C, Sini F, Romeo MG, Cota F, Ricci D. Early psychomotor development of low-risk preterm infants: Influence of gestational age and gender. Eur J Paediatr Neurol 2016; 20: 518-23.
4. Maitre NL, Chorna O, Romeo DM, Guzzetta A. Implementation of the Hammersmith Infant Neurological Examination in a High-Risk Infant Follow-Up Program. Pediatr Neurol 2016; 65: 31-8.
5. Romeo DM, Cioni M, Palermo F, Cilauro S, Romeo MG. Neurological assessment in infants discharged from a neonatal intensive care unit. Eur J Paediatr Neurol 2013; 17: 192-8.
6. Romeo DM, Cioni M, Scoto M, Mazzone L, Palermo F, Romeo MG. Neuromotor development in infants with cerebral palsy investigated by the Hammersmith Infant Neurological Examination during the first year of age. Eur J Paediatr Neurol 2008; 12: 24-31.
7. Romeo DM, Ricci D, Brogna C, Mercuri E. Use of the Hammersmith Infant Neurological Examination in infants with cerebral palsy: a critical review of the literature. Dev Med Child Neurol 2016; 58: 240-5.
8. Haataja L, Mercuri E, Guzzetta A, et al. Neurologic examination in infants with hypoxic-ischemic encephalopathy at age 9 to 14 months: use of optimality scores and correlation with magnetic resonance imaging findings. J Pediatr 2001; 138: 332-7.
9. Frisone MF, Mercuri E, Laroche S, et al. Prognostic value of the neurologic optimality score at 9 and 18 months in preterm infants born before 31 weeks' gestation. J Pediatr 2002; 140: 57-60.
10. Ricci D, Guzzetta A, Cowan F, et al. Sequential neurological examinations in infants with neonatal encephalopathy and low apgar scores: relationship with brain MRI. Neuropediatrics 2006; 37: 148-53.
11. Ricci D, Cowan F, Pane M, et al. Neurological examination at 6 to 9 months in infants with cystic periventricular leukomalacia. Neuropediatrics 2006; 37: 247-52.
12. Romeo DM, Guzzetta A, Scoto M, et al. Early neurologic assessment in preterm-infants: integration of traditional neurologic examination and observation of general movements. Eur J Paediatr Neurol 2008; 12: 183-9.
13. Pizzardi A, Romeo DM, Cioni M, Romeo MG, Guzzetta A. Infant neurological examination from 3 to 12 months: predictive value of the single items. Neuropediatrics 2008; 39: 344-6.
14. Gkoltsiou K, Tzoufi M, Counsell S, Rutherford M, Cowan F. Serial brain MRI and ultrasound findings: relation to gestational age, bilirubin level, neonatal neurologic status and neurodevelopmental outcome in infants at risk of kernicterus. Early Hum Dev 2008; 84: 829-38.
15. Romeo DM, Cioni M, Scoto M, Pizzardi A, Romeo MG, Guzzetta A. Prognostic value of a scorable neurological examination from 3 to 12 months post-term age in very preterm infants: a longitudinal study. Early Hum Dev 2009; 85: 405-8.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

HINE – Interpreting the results: quick reference guide

Hammersmith Infant Neurological Examination (HINE)

A combination of neuroimaging, standardised motor assessments, standardised neurological examination and history taking about risk factors is recommended in the early diagnosis of cerebral palsy guidelines.

In infants who present as having high risk of cerebral palsy, cut-off scores predict both the likelihood of cerebral palsy and the probable motor severity of cerebral palsy.

HINE scores at 3, 6, 9 or 12 months:

- 50-73 indicates likely unilateral cerebral palsy (i.e. 95-99% will walk)
- <50 indicates likely bilateral cerebral palsy

HINE scores at 3-6 months:

- 40-60 indicates likely GMFCS I-II
- <40 indicates likely GMFCS III-V

In infants under 2 years of age, it is important to give parents accurate and clear information about the likelihood of cerebral palsy as a clinical diagnosis, while at the same time explaining that severity is difficult to predict accurately prior to two years of age. It helps parents to maintain hope by explaining that all infants can learn and that the condition has varying levels of severity, with mild being more common than severe in high income country contexts. See fact sheet on 'Communicating the diagnosis'.

Hemiplegia (Unilateral)

Quadriplegia (Bilateral)

Ambulant GMFCS I-II

Non-Ambulant GMFCS III-V

Practice points from international guidelines

Early detection in infants <5 months (corrected)

4

STRONG RECOMMENDATION based on MODERATE QUALITY evidence of test psychometrics in newborn-detectable risk populations

In contexts where the General Movements (GMs) assessment is not available and/or MRI is not safe or affordable (e.g. in low to middle income countries): early detection of cerebral palsy in infants with 'newborn detectable risks' and less than 5 months old (corrected age) is still possible and should be carried out to enable access to early intervention.

STANDARDISED NEURO EXAM + STANDARDISED MOTOR with history taking about risk factors

TEST: Hammersmith Infant Neurological Examination (HINE) [HINE<57 at 3 months is 96% predictive of cerebral palsy]. The HINE is a scored neurological examination, based on the Dubowitz.

STANDARDISED NEURO EXAM

Early detection in infants >5 months (corrected)

6

CONDITIONAL RECOMMENDATION based on MODERATE QUALITY evidence of test psychometrics in high risk populations

The most accurate method for early detection of cerebral palsy 'infant detectable risks', with children older than 5 months of age (corrected) but less than 2 years old, is to use a combination of:

STANDARDISED NEURO EXAM + ABNORMAL NEURO EXAM + STANDARDISED MOTOR with history taking about risk factors

TEST: HINE [90% predictive of cerebral palsy]. HINE scores <73 (at 6, 9 or 12 months) should be considered at high-risk of cerebral palsy. HINE scores <40 (at 6, 9 or 12 months) almost always indicate non-ambulant cerebral palsy.

STANDARDISED NEURO EXAM

EARLY DIAGNOSIS – CEREBRAL PALSY

Practice points from international guidelines

HINE 03

Early detection in infants >5 months (corrected)

7 **CONDITIONAL RECOMMENDATION based on MODERATE QUALITY evidence of test psychometrics in high risk populations**

In contexts where MRI is not safe or affordable (e.g. in low to middle income countries):
Early detection of cerebral palsy is still possible in those with 'infant detectable risks' between 5–24 months corrected age and should be carried out to enable access to early intervention.

STANDARDISED NEURO EXAM + STANDARDISED MOTOR with history taking about risk factors

TEST: Hammersmith Infant Neurological Examination (HINE) [90% predictive of cerebral palsy]. HINE scores <73 (at 6, 9 or 12 months) should be considered at high-risk of cerebral palsy. HINE scores <40 (at 6, 9 or 12 months) almost always indicate non-ambulant cerebral palsy.

STANDARDISED NEURO EXAM

Early detection of motor severity

8 **CONDITIONAL RECOMMENDATION based on LOW QUALITY evidence**

HINE 40–60

Ambulant more likely

Unilateral lesions (Grade IV haemorrhage, perinatal arterial ischemic stroke), periventricular leukomalacia (PVL non-cystic) moderate/severe white matter injury.

HINE <40

Non-ambulant more likely

Bilateral parenchymal haemorrhages (Grade IV), bilateral cystic periventricular leukomalacia (cPVL) (Grade III), brain maldevelopment, basal ganglia injury.

In infants less than 2 years old, prognosis of motor severity predictions should be made cautiously. **Always** use standardised tools, since incomplete development of voluntary motor skills and/or abnormal tone might confound clinical observations. Motor severity is most accurately predicted using standardised neurological exams and neurological imaging.

STANDARDISED NEURO EXAM + ABNORMAL NEURO EXAM

Cerebral Palsy ALLIANCE
Centre of Research Excellence Cerebral Palsy

Hammersmith infant neurological examination (HINE) clinical utility

Why do we need a standardised neurological examination?

- **To accurately identify children at risk of cerebral palsy early through the use of robust evidence-based standardised detection tools.**
- **To support the cerebral palsy diagnostic process in combination with other evidence-based tools.**
- **To help define the prognosis and provide information on the type and severity of impairment of neurological function.**
- **May assist in longitudinal follow up of high risk infants including effects of intervention.**

The Hammersmith Infant Neurological Examination (HINE) is recommended in the International Clinical Practice Guidelines. It can play a helpful role in early detection, diagnosis and prognosis of infants at risk of developing cerebral palsy.

1. Romeo DM, Ricci D, Brogna C, Mecuri E. Use of the Hammersmith Infant Neurological Examination in infants with cerebral palsy: a critical review of the literature. *Dev Med Child Neurol* 2015. doi:10.1111/dmcn.12876.
2. Haataja L, Mercuri E, Guzzetta A, Rutherford M, Counsell S, Frisone M, Cioni G, Cowan F, Dubowitz L. Neurologic examination in infants with hypoxic-ischemic encephalopathy at age 9 to 14 months: Use of optimality scores and correlation with magnetic resonance image findings. *J Pediatr* 2001; 138(3): 332-7.

Hammersmith infant neurological examination (HINE) clinical utility

Predictive/early detection children at risk of cerebral palsy

- Moderate quality evidence of test psychometrics in high-risk populations
- HINE<57 at 3 months is 96% predictive of cerebral palsy in infants older than 5 months of age (corrected for prematurity) but less than 2 years old
- 90% predictive of cerebral palsy at 2–24 months of age

HINE scores at 6, 9 or 12 months:

- <73 indicates high-risk of cerebral palsy
- <40 indicates abnormal outcome, usually cerebral palsy
- Meta analysis of predictive value of studies using HINE showed excellent sensitivity[1].

Early detection of motor severity and topography of cerebral palsy

- Moderate quality evidence of test psychometrics in high risk populations
- Cut-off scores predict the probable severity

HINE scores at 3, 6, 9 or 12 months:

- 50–73 indicates likely unilateral cerebral palsy (i.e. 95-99% will walk)
- <50 indicates likely bilateral cerebral palsy

HINE scores at 3-6 months:

- 40-60 indicates likely GMFCS I-II
- <40 indicates likely GMFCS III-V.

Provides information on other aspects of neurological function, not just motor

Easily performed

- Good inter-observer reliability even in inexperienced clinicians[2]

Accessible to all clinicians, no certified training required

1. Romeo DM, Ricci D, Brogna C, Mecuri E. Use of the Hammersmith Infant Neurological Examination in infants with cerebral palsy: a critical review of the literature. *Dev Med Child Neurol* 2015. doi:10.1111/dmcn.12876.

2. Haataja L, Mercuri E, Guzzetta A, Rutherford M, Counsell S, Frisone M, Cioni G, Cowan F, Dubowitz L. Neurologic examination in infants with hypoxic-ischemic encephalopathy at age 9 to 14 months: Use of optimality scores and correlation with magnetic resonance image findings. *J Pediatr* 2001; 138(3): 332-7.

Cerebral Palsy ALLIANCE

Centre of Research Excellence Cerebral Palsy

Hammersmith infant neurological examination

Hammersmith Infant Neurological Examination Clinical Patterns Predictive of Outcome

Clinical patterns from the examination predictive of abnormal outcome have also been discussed. Those most often associated with cerebral palsy were:[1]

- Abnormal posture (flexed arms and extended legs)
- Persistent abnormal axial tone (increased neck and trunk extensor tone)
- Limb tone (reduction of pROM in the items popliteal angle and hip adductors)
- Abnormal arm protection and forward parachute reaction during the second 6 months.

These may also provide an estimate of severity and functional level, with early abnormalities predicting those children who will not reach independent sitting. Specific items in tone (scarf sign, popliteal angle, adductors, pull to sit, ventral suspension) and posture (trunk and legs in sitting) also help distinguish between diplegia and quadriplegia.

1. Romeo DM, Ricci D, Brogna C, Mecuri E. Use of the Hammersmith Infant Neurological Examination in infants with cerebral palsy: a critical review of the literature. *Dev Med Child Neurol* 2015. doi:10.1111/dmcn.12876.

Neuroimaging– Interpreting the results: Quick reference guide

Magnetic Resonance Imaging (MRI)

MRI is the instrumental test recommended for the early detection of Cerebral Palsy (CP) for newborns and children with detectable risks of CP.

In preterm infants, serial cranial ultrasound (CUS) is used to assess brain abnormalities at any degree of prematurity. CUS is helpful in the evaluation of the risk of CP according to brain abnormalities, and may assist in recommending an MRI term equivalent age (TEA).

Therefore, in infants with newborn detectable risks, MRI is recommended at TEA.

Normal MRI findings do not preclude the clinical diagnosis of cerebral palsy, as abnormal MRI findings do not automatically precede cerebral palsy. 4,9% of the children with CP show no structural abnormalities on MRI, nevertheless, this group is commonly associated with limited ambulatory capacities.[1]

Abbreviazioni

CUS	**Cranial ultrasound**
DWI	**Diffusion weighted imaging**
DTI	**Diffusion tensor imaging**
fRMI	**Functional magnetic resonance imaging**
HIE	**Hypoxic ischaemic encephalopathy**
IVH	**Intraventricular haemorrhage**
MRS	**Magnetic resonance spectroscopy**
MRI	**Magnetic resonance imaging**
PLIC	**Posterior limb of the internal capsule**
PVE	**Periventricular echogenicity**
PVL	**Periventricular leukomalacia**
cPVL	**Cystic periventricular leukomalacia**
SWI	**Susceptibility weighted imaging**
TEA	**Term equivalent age**
T1w	**T1 weighted images**
T2w	**T2 weighted images**

1. Mailleux, L., Franki, I., Emsell, L., Peedima, M. L., Fehrenbach, A., Feys, H., & Ortibus, E. (2020). The relationship between neuroimaging and motor outcome in children with cerebral palsy: A systematic review—Part B diffusion imaging and tractography. Research in Developmental Disabilities, 97.

Cranial ultrasound (CUS) Key evidence

CUS is the most commonly used neuroimaging method during the early neonatal period as long as the acoustic access windows are usable (anterior and posterior fontanel, temporal and mastoid synchondrosis).

Advantages:

- Fast examination
- Bedside and widely available
- Non invasive (no radiation)
- Easy handling
- Images in real-time; useful for repeated and frequent imaging

Limitations:

- Variable depending on expertise (execution and interpretation)
- Need for an acoustic window with a less detailed visualisation of whole brain
- Cortical and subtle white matter and small cerebellar haemorrages abnormalities may be difficult to detect

In term or preterm infants, serial CUS can be used from the first days of life to monitor outcomes of a hypoxic-ischemic injury, including complications of germinal matrix hemorrhages.[1]

The most common abnormal cranial ultrasound patterns in preterm that later develop CP are:

- Cystic PVL (periventricular cystic lesions and/or tissue loss)
- IVH Grade III-IV
- Persistent ventricular dilatation/abnormal shape of ventricle at term age.

Although CUS is quite useful, especially for obtaining serial images, its spatial resolution is significantly lower as compared to MRI, therefore requires that the findings are confirmed and / or better defined with MRI.

1. Fiori, S., Canapicchi, R. e Guzzetta, A. (2018). Neuroimaging anatomico e funzionale. In Persico, A.M. (a cura di). Manuale di neuropsichiatria infantile e dell'adolescenza (pag. 183-198). Società Editrice Universo

Magnetic Resonance Imaging (MRI) – Key evidence

Neonatal MRI/MRI at near term equivalent age with neuroanatomical abnormalities predictive of cerebral palsy:

- 86-89% Sensitivity
- 89% Specificity

NB: Normal MRI findings do not preclude the clinical diagnosis of cerebral palsy, as 10% infants with cerebral palsy have normal MRI findings.

CITATION	EVIDENCE	#STUDIES	#PATIENTS	ACCURACY FOR CEREBRAL PALSY	QUALITY
Ashwal 2009	Practice Parameter Article	10	644	Sensitivity = 89%	14/14
Bosanquet 2013	Systematic Review	3	702	Sensitivity = 86% Specificity = 89%	14/14
Ment 2002	Practice Parameter Article	13	410	Basal ganglia injury = predictive of CP (50-94%) MRI = predictive DWI = probably predictive MRS = possibly predictive	14/14

Advantages

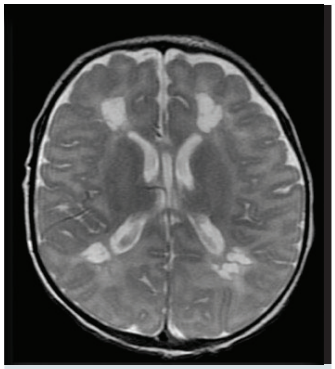
- No radiation
- High resolution
- Good contrast
- Detailed visualisation of whole brain
- Shows myelination
- Greater sensitivity white and grey matter injuries than cranial ultrasound
- SWI helps to distinguish ischemic and hemorrhagic punctate white matter lesions
- MRA allows visualisation of the arterial system
- MRV allows visualisation of the venous system

Limitations

- Care should be taken to use appropriate neonatal sequences with thin slices (2mm)
- Infants need to be stable and monitored for heart rate, respiration and saturation. Adequate hearing protection is also required
- High technical effort and costs may be a barrier in some centres
- Training with interpretation required
- Sedation or general anasthesia may be required for older infants (>6–10 weeks up to 2 years of age)
- Individual consideration given to risks benefits ratio

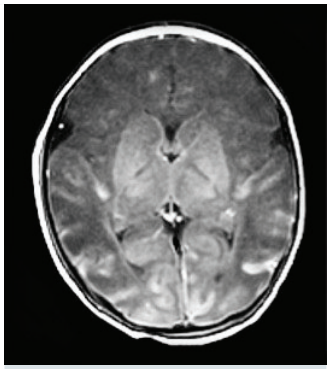
Magnetic Resonance Imaging (MRI) – Key evidence

The most predictive MRI patterns are:



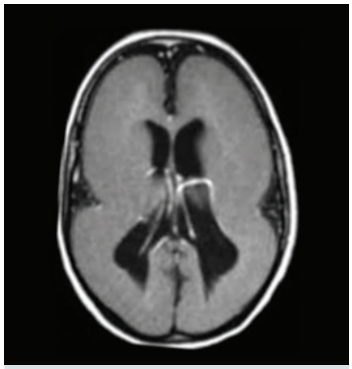
Predominant white matter injury (66,9%) [cystic periventricular leukomalacia (PVL) or periventricular haemorrhagic infarctions]

IMAGE 1



Cortical and deep grey matter lesions (18,6%) [basal ganglia/thalamus lesions, watershed injury (parasagittal injury), multicystic encephalomalacia, stroke]

IMAGE 2



Brain maldevelopments (4,3%) [lissencephaly, pachygyria, cortical dysplasia, polymicrogyria, and schizencephaly]

IMAGE 3

IMAGES 1 AND 2 PROVIDED BY ASSOCIATE PROFESSOR ANDREA GUZZETTA AND DR SIMONA FIORI; UNIVERSITY OF PISA

IMAGE 3 PROVIDED BY DR CATHY MORGAN, CEREBRAL PALSY ALLIANCE RESEARCH INSTITUTE.

In addition to the above mentioned patterns, it is possible to find in 1% of cases postnatal lesions (typical exclusively of unilateral CP) and in another 1% lesions classified as miscellaneous (cerebral and cerebellar atrophy, delayed myelination, ventriculomegaly, haemorrhages not classified as white or grey matter lesions, brainstem lesions or calcifications).[4]

Later MRI findings may also help estimate the presumed abnormality, timing and pathogenesis of the insult.

Studies including all types of CP show PVL to be the most common white matter lesion (75.2%). Grey matter lesions were the major cause of spastic CP (unilateral and bilateral) with an equal distribution across the GMFCS levels. Another finding was that grey matter lesions are found in 42.2% of children with dyskinetic CP.[4]

1. Ashwal, S., Michelson, D., Plawner, L., Dobyns, W.B.; Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Practice parameter: Evaluation of the child with microcephaly (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology. 2009 Sep 15;73(11):887-97.
2. Bosanquet, M., Copeland, L., Ware, R., Boyd, R. A systematic review of tests to predict cerebral palsy in young children. Dev Med Child Neurol. 2013 May;55(5):418-26.
3. Ment, L.R., Bada, H.S., Barnes, P., Grant, P.E., Hirtz, D., Papile, L.A., Pinto-Martin, J., Rivkin, M., Slovis, T.L. Practice parameter: neuroimaging of the neonate: report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology. 2002 Jun 25;58(12):1726-38.
4. Mailleux, L., Franki, I., Emsell, L., Peedima, M. L., Fehrenbach, A., Feys, H., & Ortibus, E. (2020). The relationship between neuroimaging and motor outcome in children with cerebral palsy: A systematic review—Part B diffusion imaging and tractography. Research in Developmental Disabilities, 97.

Neuroimaging basics

STANDARD MR TECHNIQUES	DESCRIPTION	CLINICAL USES
T1-weighted and T2-weighted images	Used for qualitative image assessment. T1-weighted images are MRI images that are designed to distinguish tissues with differing T1/T2 relaxation times and evaluate macroscopic changes in lesions and tissues, including sulci, ventricles, cysts.	Detects brain malformations, intracranial haemorrhage, ischemic-hypoxic injury, grey matter and white matter changes, ventriculomegaly, or atrophy. On T1 weighted images cerebrospinal fluid has low signal and appears dark, and myelinated white matter is brighter than grey matter. T1 is best for evaluation of signal abnormalities in the neonatal period and myelination. On T2 weighted images cerebrospinal fluid has a high signal and appears bright (except for proton weighted images), and myelinated white matter is darker than grey matter. T2 can be useful for additional information on signal abnormalities. Both T1 and T2 can also calculate tissue volumes. Both T1 and T2 can be used to perform 2D linear measurements to assess brain growth
SWI	Detects blood, iron, and calcifications within the brain.	Evaluates traumatic brain injury, coagulopathies, or other haemorrhagic disorders, vascular malformations, infarction, neoplasms, and neurodegenerative disorders. SWI can help to differentiate between hemorrhagic and ischemic PWML can diagnose punctate cerebellar haemorrhages better than with T2
DWI and ADC	Measures random motion of water in tissue and quantified as ADC.	Useful for early detection of hypoxic ischemic damage such as HIE or focal arterial infarctions or other toxic/neuro-metabolic disorders in acute and subacute phases.

RESEARCH MRI TECHNIQUES	DESCRIPTION	CLINICAL USES
Three-dimensional volumetric	Allows measurement of whole-brain volume as well as volumes of specific structures, ventricles and cerebellum.	Used for absolute quantification of brain structures and detection of deviations in normal volumes of tissues.
DTI	Measures water diffusion along axis that coincides with fibre tracts and quantified as Fractional anisotropy (FA). Used to identity and map WM tracts.	Used to generate tractography data to evaluate fibre tracts. Colour-coded FA map shows directionality of fibres. Can reveal premyelinated structures.
fMRI	Detects changes in blood oxygenation level dependent (BOLD) signals in spatially distinct regions that are correlated with task-related functional activity or resting state.	Resting state is more easily applied in the newborn. It can relate functional connectivity to neurodevelopmental outcomes.
MRS	Measures concentrations of metabolites in regions of the brain.	Used to study brain cellular metabolism, including metabolic disorders.

Neuroimaging - clinical utility

General points

At <6 weeks it is preferable to feed and wrap the baby and avoid sedation for MRI. Sedation may be required for an MRI examination >6 weeks (variable, depending on the individual patient and the resources of the center). MR team familiar with neonatal imaging is recommended.

Where possible use a 3 Tesla1 (3T)[1] scanner to improve the ability to detect subtle lesions.

Pre-term infants with unilateral cerebral palsy often have white matter injury due to periventricular haemorrhagic (presumed venous) infarction, while full term infants with hemiplegic cerebral palsy are more likely to show combined grey and white matter abnormalities following a perinatal arterial ischemic stroke.

When the posterior limb of the internal capsule (PLIC) and projections of the corticospinal tract are impacted, greater functional impairment is more likely to occur.[2,3]

Well defined lesions can be seen early, while subtle white matter lesions may not be seen with conventional MRI, they may result in poor brain growth, and/or delayed myelination.

Pre-term infants

Term equivalent age (TEA) (or as close as possible) MRI is most predictive of outcome.[4]

Sequential cranial ultrasound (CUS) can also predict non-ambulatory cerebral palsy but may fail to detect subtle lesions. Brain MRI may help in detecting subtle abnormalities such as punctate white matter lesions, which are difficult to detect on CUS.

Periventricular leukomalacia is the most common white matter finding in children with spastic forms of cerebral palsy born preterm.

Term-born infants

MRI in the first week of life (day 4-6 post delivery) is recommended for infants born at term with suspected brain insults. If the infant has had encephalopathy, conventional MR sequences may not show any signs of abnormality in the first 48 hours. Diffusion weighted imaging (DWI) and apparent diffusion coefficient maps (ADC) are likely to detect the injury early but waiting 3 – 5 days before imaging is recommended, to maximise identifying abnormal findings.

Conventional T1 beyond the first week and DWI before the end of the first week may also allow examination of the posterior limb of the internal capsule (PLIC) and the descending corticospinal tracts at the level of the cerebral peduncles, which is highly predictive of permanent motor dysfunction.[3,5]

Deep grey matter lesions are the most common finding in children with bilateral dyskinetic or spastic Cerebral Palsy[6,7].

Focal stroke is the most common finding with unilateral spastic cerebral palsy in term-born infants.[6]

Isolated grey matter damage is rare but can be seen in full-term infants following a sentinel event.

1. Plaisier, A., Govaert, P., Lequin, M.H., Dudink, J. Optimal timing of cerebral MRI in preterm infants to predict long-term neurodevelopmental outcome: a systematic review. AJNR Am J Neuroradiol 2014; 35: 841-7.
2. Korzeniewski, S.J., Birbeck, G., DeLano, M.C., Potchen, M.J., Paneth, N. A systematic review of neuroimaging for cerebral palsy. J Child Neurol 2008; 23: 216-27.
3. Cowan, F.M., de Vries, L.S. The internal capsule in neonatal imaging. In Semin Fetal Neonatal Med 2005; 10: 461-474.
4. Ashwal, S., Russman, B.S., Blasco, P.A., Miller, G., Sandler, A., Shevell, M., et al. Practice Parameter: Diagnostic assessment of the child with cerebral palsy. Report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurol 2004; 62: 851-63.
5. Martin, J.H., Chakrabarty, S., Friel, K.M. Harnessing activity-dependent plasticity to repair the damaged corticospinal tract in an animal model of cerebral palsy. Developmental Medicine & Child Neurology. 2011; 53:9-13.
6. Krägeloh-Mann, I., Horber, V. The role of magnetic resonance imaging in elucidating the pathogenesis of cerebral palsy: a systematic review. Dev Med Child Neurol 2007; 49: 144.
7. Mailleux, L., Franki, I., Emsell, L., Peedima, M. L., Fehrenbach, A., Feys, H., & Ortibus, E. (2020). The relationship between neuroimaging and motor outcome in children with cerebral palsy: A systematic review—Part B diffusion imaging and tractography. Research in Developmental Disabilities, 97.
8. Wisnowski, J. L., Wintermark, P., Bonifacio, S. L., Smyser, C. D., Barkovich, A. J., Edwards, A. D., ... & Publications Committee. (2021, October). Neuroimaging in the term newborn with neonatal encephalopathy. In Seminars in Fetal and Neonatal Medicine (Vol. 26, No. 5, p. 101304). WB Saunders

Topography

MRI signs indicating topography[1]

Topography is not easily determined from physical examination in early in life but MRI can provide valuable predictive information.

Unilateral abnormal MRI findings most commonly translate into unilateral cerebral palsy and milder physical impairments. Bilateral abnormal MRI findings almost always translate into bilateral cerebral palsy and higher likelihood of moderate-severe physical impairment. It should be noted however that 25% of children with an abnormal MRI such as PVL will not go on to develop cerebral palsy.

Other exceptions include some children with unilateral lesions who do not show signs of neurological asymmetric dysfunction.

SPASTIC FORMS			DYSKINETIC FORMS	ATAXIC FORMS
UNILATERAL	BILATERAL	QUADRIPLEGIC		
• Focal vascular insults (24%) • Malformations • Unilateral haemorrhage (grade IV) with porencephaly • Lesions in the parietal white matter involving the trigone • Middle cerebral artery stroke with asymmetry of myelination of the posterior limb internal capsule (PLIC)	• Bilateral white matter injury (31–60% of cases) • Cystic periventricular leukomalacia (PVL) (Grade II–III) with sparse or absent myelination of the posterior limb internal capsule (PLIC) • Periventricular echogenicity (PVE) • Extensive Punctate white matter lesions (PWML)	• Grey matter injury (34% of cases) • Malformations (16% cases) • Cystic periventricular leukomalacia (cPVL) (Grade III-IV) with absent myelination of the posterior limb internal capsule (PLIC) • Severe white matter injury +/- deep nuclear grey matter.	• Grey matter injury (21% of cases) with thalamic and lentiform nuclear injury.	• Malformations (18% cases) • Normal imaging (24–57% cases) • Cerebellar injury.

1. Reid SM, Dagia CD, Ditchfield MR, Carlin JB, Reddihough DS. Population-based studies of brain imaging patterns in cerebral palsy. *Dev Med Child Neurol* 2014; 56: 222-32.
2. Himmelmann, Kate, et al. "Neuroimaging Patterns and Function in Cerebral Palsy—Application of an MRI Classification. Frontiers in Neurology 11 (2021): 1889.

MRI findings and motor impairment severity

A correlation often exists between the location, size and extent of the injury and the severity of the child's motor impairment. Infants with brain malformations and cortical/subcortical lesions generally exhibit more severe motor impairments and have the highest risk for being non-ambulatory.

Non-ambulant cerebral palsy is more likely following:

- Bilateral parenchymal haemorrhages (Grade IV)
- Bilateral cystic periventricular leukomalacia (cPVL)
- Brain maldevelopment (although ambulation is possible in some cases)
- HIE with basal ganglia injury (although ambulation is possible in some cases)

Children with mild periventricular white matter lesions generally exhibit milder motor impairments and often less associated impairments. However, infants with more severe white matter injury may have more long-term motor and developmental concerns than those born at term age.

Ambulant cerebral palsy is more likely following:

- Unilateral lesions (Grade IV haemorrhage, perinatal arterial ischemic stroke)
- Periventricular leukomalacia (PVL) (non-cystic)
- HIE with moderate/severe white matter injury (and no or minimal basal ganglia injury)

Neuroimaging in clinical practice

MRI

Advantages

- No radiation
- High resolution
- Good contrast
- Detailed visualisation of whole brain
- Shows progress of myelination
- Greater sensitivity white and grey matter injuries than cranial ultrasound

Limitations

- High technical effort and costs may be a barrier in some centres
- Training with interpretation required
- Sedation or general anaesthetic may be required with infants in older range (>6–10 weeks up to 2 years of age)
- Individual consideration given to risks benefits ratio

Abnormal findings predictive of cerebral palsy

- White matter injury (both cystic periventricular leukomalacia (PVL) and non-cystic white matter injury or periventricular haemorrhagic infarctions
- Cortical and deep matter grey lesions (such as basal ganglia/ thalamus lesions, parasagittal injury, multicystic encephalomalacia, stroke
- Brain maldevelopments such as lissencephaly, pachygyria, cortical dysplasia, poloymircrogyria, schizencephaly
- Abnormal signal intensity within the posterior limb internal capsule (PLIC), abnormal myelination of PLIC

NB: Normal MRI findings do not preclude the clinical diagnosis of cerebral palsy, as some infants with cerebral palsy have normal MRI findings

ULTRASOUND

Advantages

- Bedside and widely available
- No radiation
- Images in real time and useful for repeated and frequent imaging

Limitations

- Less detailed visualisation of whole brain
- Cortical and subtle white matter abnormalities may be difficult to detect
- Interpretation may be variable dependent on expertise
- Cystic PVL (periventricular cystic lesions) and/or tissue loss
- IVH grade III-IV (PVHI)
- Persistent ventricular dilatation/abnormal shape of ventricle at term age

Neuroimaging in clinical practice

MRI

Recommendations international clinical practice guidelines early detection cerebral palsy

Pre-term infants

- Term equivalent age (TEA) (or as close as possible) MRI is most predictive of outcome[1].
- Where possible use a 3 Tesla (3T) scanner to improve the ability to detect subtle lesions.
- When an MRI is performed within a week after a presumed insult, diffusion weighted imaging (DWI) can be predictive of subsequent cystic evolution in the white matter.

Term-born infants

- MRI in the first week of life (day 5–7 post delivery is optimal) is recommended for infants born at term with suspected brain abnormalities.
- If the infant has had encephalopathy, conventional MR sequences may not show any signs of abnormality in the first 48 hours.
- Diffusion weighted imaging (DWI) and apparent diffusion coefficient maps (ADC) are likely to detect the injury early but waiting 3–5 days before imaging is recommended, to maximise identifying abnormal findings.
- Conventional T1 beyond the first week and DWI before the end of the first week may also allow examination of the posterior limb of the internal capsule (PLIC) and the descending corticospinal tracts at the level of the cerebral peduncles, which is highly predictive of permanent motor dysfunction[2].
- Parents may need to consider redirection of care or withdrawal of intensive therapy upon learning about the infant's pain and suffering levels[3].

ULTRASOUND

Recommendations international clinical practice guidelines early detection cerebral palsy

Pre-term infants

- Sequential cranial ultrasound (CUS) can also predict non-ambulatory cerebral palsy but may fail to detect subtle lesions, especially diplegia.

Neuroimaging in clinical practice

MRI	ULTRASOUND
Recommendations international clinical practice guidelines early detection cerebral palsy (cont.) **Infants older than 5 months** Sedation will be required for neuroimaging infants in this older range (>6–10 weeks up to 2 years of age) and individual consideration will need to be given to the risk-benefits ratio. MRI has a reduced predictive value in this time window, because of the rapid growth, myelination, rapid change in T2 and T1 and activity-dependent plasticity that is occurring, all of which will confound interpretation of the scan results. Gliosis in the white matter first becomes visible during the second half of the second year. Furthermore, at 2 years of age when myelination is progressing, the final definition of the lesion boundaries and deep structure lesions are more evident on MRI. This is especially the case for subtle white matter lesions. The recommendations are for repeat MRI scans at 2 years of age for infants with initially normal MRI (at 12–18 months) but persistent motor dysfunction and/or neurological abnormality.	

1. Plaisier A, Govaert P, Lequin MH, Dudink J. Optimal timing of cerebral MRI in preterm infants to predict long-term neurodevelopmental outcome: a systematic review. *AJNR Am J Neuroradiol* 2014; 35: 841-7.
2. Cowan FM, de Vries LS. The internal capsule in neonatal imaging. In *Semin Fetal Neonatal Med* 2005; 10: 461-474.
3. Martin JH, Chakrabarty S, Friel KM. Harnessing activity-dependent plasticity to repair the damaged corticospinal tract in an animal model of cerebral palsy. *Developmental Medicine & Child Neurology.* 2011;53:9-13.
4. Thayyil S, Chandrasekaran M, Taylor A, Bainbridge A, Cady EB, Chong WK, et al. Cerebral magnetic resonance biomarkers in neonatal encephalopathy: a meta-analysis. *Peds* 2010.

Communication of diagnosis

Informing parents and carers of the diagnosis of cerebral palsy or high-risk of cerebral palsy is a difficult process. It is imperative that it is completed in the most sensitive, compassionate and well planned way to optimise outcomes for both parents and children.

The process of parental acceptance of a cerebral palsy diagnosis, grieving, coping and resiliency is ongoing, cyclical and requires a continuum of supports from diagnosticians.[1,2]

Parents experience grief and loss at the time of diagnosis, or when they are told their infant is at 'high-risk of cerebral palsy'. Emotional experiences at the time of diagnosis can be intense and contradictory, and include anger, fear, relief, confusion, guilt, despair and denial.[1,2]

For parents, the learning processes inherent in receiving information on the diagnosis can be impacted by various factors including experiential avoidance strategies, actively blocking the recall of bad news in order to cope and processing large quantities of new and complex information.[1,4]

The communication of high-risk cerebral palsy to a family should be conveyed through a series of well-planned and compassionate conversations, rather than a one-time event.

Qualitative evidence indicates many parents are dissatisfied with the diagnostic process.[3] Common criticisms include the amount of information received at diagnosis; lack of discussion about the likely impact on their child and family; information was unclear and conveyed a pessimistic outlook for the future.[3,5]

1. Whittingham, Koa, Wee, Diana, Sanders, Matthew R. and Boyd, Roslyn (2013) Sorrow, coping and resiliency: parents of children with cerebral palsy share their experiences. *Disability and Rehabilitation*, 35 17: 1447-1452.
2. Ahmann E. Review and commentary: Two studies regarding giving "bad news". *Pediatric Nursing.* 1998;24(6):554.
3. Baird G, McConachie H, Scrutton D. Parents' perceptions of disclosure of the diagnosis of cerebral palsy. *Arch Dis Child* 2000; 83: 475-80.
4. Jedlicka-Kohler I, Gotz M, Eichler I. Parent's recollection of the initial communicaiton of the diagnosis of cystic fibrosis. *Pediatrics* 1996;97:204.
5. Hummenlinck, A., & Pollock, K. (2006). Parents' information needs about the treatment of theirchronically ill child: A qualitative study. *Patient Education and Counseling*, 62(2), 228–234.

Communicating with parents

Best evidence-based communication strategies are recommended when communicating the diagnosis of cerebral palsy or the news about high-risk of cerebral palsy. Data from qualitative interviews suggests the following:

- Provide at least two face-to-face diagnostic information sharing sessions to facilitate comprehension, recall and acceptance.
- Ensure both parents and the infant are present to promote acceptance of the infant diagnosis.
- Use a quiet, private office.
- Provide the most honest, transparent and specific information about the diagnosis and prognosis as possible and explain the likely impact on the family.
- Use simple, direct and jargon free language.
- Use a hopeful, empathic and supportive tone.
- Tailor the information to the individual infant and the family communication style.
- Provide written information to allow absorption at a later stage.
- Provide information about the child's strengths as well as limitations, to promote development of an optimistic outlook.
- Invite questions.
- Invite discussion about feelings, as this promotes confidence in the parent's ability to cope and increases satisfaction.
- Recommend parent-to-parent and family support; parents indicate this facilitates long-term coping.
- Arrange a debriefing to help parents gather information and navigate service entry.
- Arrange early intervention, preferably initially at higher intensity to help parents come to terms with what is required of them.

Communication of diagnosis

Best practice recommendations for giving the diagnosis of cerebral palsy

1 Conveying the diagnosis of childhood disability to a parent is complex, but it is important that it is done well. When bad news is given well, hope can be conveyed, parent-child bonding can be facilitated, and satisfaction with the healthcare system is fostered.

Strong Recommendation For
based on high quality evidence for infant and parent outcomes

2 A diagnosis should be given as early as possible to:

- Maximise the child's potential from early intervention.
- To reduce parental stress and anger from diagnostic uncertainty.

Strong Recommendation For
based on high quality evidence for infant and parent outcomes

3 Diagnosis should not be delayed or withheld to protect parent's feelings.

Strong Recommendation AGAINST
based on high quality evidence for infant and parent outcomes

4 Mental preparation, factual preparation and environmental preparation is necessary before giving a diagnosis. A private, quiet room is recommended where both parents (where relevant) and the infant are invited to be present. [SPIKES STEP 1]

Strong Recommendation FOR
based on high quality evidence for parent outcomes and
low quality evidence for reducing stress in clinicians

5 Gain an understanding of the parent's current knowledge before giving a diagnosis. [SPIKES STEP 2][1]

Strong Recommendation For
based on high quality evidence for parent outcomes

6 Ask and invite questions. [SPIKES STEP 3][1]

Strong Recommendation For
based on high quality evidence for parent outcomes

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

7 Provide evidence-based facts. Answer questions honestly and using jargon-free language. Provide written information to allow later processing and information sharing with other family members (refer to Supplementary Table 2). [SPIKES STEP 4] [1]

Over a series of conversations, plan to discuss:

- Definition, prevalence, types, prognosis
- Cure
- Causes
- Prevention of complications
- Early intervention
- Medications
- Expected outcomes of treatment
- Caregiving stress
- Behaviour Management
- Adaptive Equipment
- Parent Support and Impact on the Family
- Future planning and life expectancy
- Reputable Sources of Information

Strong Recommendation For
based on high quality evidence for infant and parent outcomes

8 Respond emphatically to emotions. [SPIKES STEP 5][1]

Strong Recommendation For
based on high quality evidence for parent outcomes

9 Make a follow-up appointment to continue the diagnostic discussions. Arrange a treatment plan including early intervention and parent support [SPIKES STEP 6][1]

Strong Recommendation For
based on high quality evidence for infant and parent outcomes

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE

THE UNIVERSITY OF SYDNEY

AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

The Six Steps of SPIKES to Communicate a Diagnosis[1]

1 S - SETTING UP THE INTERVIEW

Take time to prepare

- use a quiet, private office[2]
- allow adequate time[3]
- schedule at least two information sharing sessions [4]
- Invite both parents and infants to be present[5]
- Prepare for difficult questions and different responses from each parent[6]
- use mental rehearsal to prepare for difficult questions[1]
- Prepare positive information about child strengths and hope for their future[5]

2 P - ASSESSING THE FAMILY'S PERCEPTION

- Use open-ended questions to gain a picture of what the parent already understands[1]
- Tailor information on parent's answers and individual child.[1]
- Reframe misunderstandings[1]
- Provide honest, transparent and specific information about future prognosis[7]

3 I – OBTAINING THE FAMILY'S INVITATION

Take time to respond to questions

- Invite questions[1]
- Communicate your willingness to listen the parents' questions both now and in the future.[1]
- Answer questions openly and honestly[7]

4 K – PROVIDE KNOWLEDGE AND INFORMATION

Warn that there is bad news ahead

- Use simple, direct, jargon-free language[7]
- Use a hopeful, empathic and supportive tone[5]
- Be clear and certain.[6]
- Provide written information to allow later absorption and communication of the news to other family members and friends.[8]

5 E— ADDRESSING THE FAMILY'S EMOTIONS

Respond empathically to emotions.

- Observe and name the emotions[1]
- Encourage and validate emotions[9]
- Invite discussion about their feelings[5]
- Offer assistance to tell others[4]

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE

THE UNIVERSITY OF SYDNEY

AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

6 S – STRATEGY and SUMMARY

- Check with the family to see if they are ready to discuss treatment planning[1]
- Involve the family in treatment planning[1]
- End sessions with something practical and helpful that parents can do[6]
- Arrange next review and debriefing
- Recommend parent-to-parent and family support[7]
- Appoint a key worker for service navigation[2]
- Arrange early intervention[3]

1. Baile WF, Buckman R, Lenzi R, Glober G, Beale EA, Kudelka AP. SPIKES – A sixstep protocol for delivering bad news: application to the patient with cancer. Oncologist 2000; 5: 302–11.
2. Rahi JS, Manaras I, Tuomainen H, Hundt GL (2004) Meeting the needs of parents around the time of diagnosis of disability among their children: evaluation of a novel program for information, support, and liaison by key workers. Pediatrics, 114(4), e477-82. doi:10.1542/peds.2004-0240
3. Girgis A, Sanson-Fisher RW, Schofield MJ. Is there consensus between breast cancer patients and providers on guidelines for breaking bad news? Behav Med 1999; 25: 69–77.
4. Hallberg U, Oskarsdottir S, Klingberg G (2010) 22q11 deletion syndrome – the meaning of a diagnosis. A qualitative study on parental perspectives. Child Care Health Dev, 36(5), 719-25. doi:10.1111/j.1365-2214.2010.01108
5. Ahmann E (1998) Review and commentary: two studies regarding giving "bad news". Pediatr Nurs, 24(6), 554-6. PMID: 10085998
6. Graungaard, A.H. & Skov, L. (2006). Why do we need a diagnosis? A qualitative study of parents' experiences, coping and needs, when the newborn child is severely disabled. Child: Care, Health and Development, 33(3): 296–307.
7. Reid A, Imrie H, Brouwer E, Clutton S, Evans J, Russel D, Bartlett D (2011) 'If I knew then what I know now: parents' reflection on raising a child with cerebral palsy. Phys Occup Ther Pediatr, 31(2), 169-183. doi:10.3109/01942638.2010.540311
8. Klein S, Wynn K, Ray L, Demeriez L, LaBerge P, Pei J, Pierre CS (2011) Information sharing during diagnostic assessments: what is relevant for parents? Phys Occup Ther Pediatr, 31(2), 120-132. doi:10.3109/01942638.2010.523450
9. Rabow MW, McPhee SJ. Beyond breaking bad news: how to help patients who suffer. West J Med 1999; 171: 260–63.

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE

THE UNIVERSITY OF SYDNEY

AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

Understanding the parent's perspectives of early diagnosis of cerebral palsy

The Bad News Response Model

Parents can respond to bad news by:
(a) Watchful Waiting,
(b) Active Change,
(c) Acceptance, and
(d) Nonresponding i.e. protective denial.

Clinicians should support parents to shift from "watchful waiting" to "active change" and "acceptance".

Parents require detailed information about the diagnosis, treatments, prognosis and supports.

The model redirects the focus from the clinician's emotions to the goal of fostering long-term adaptive parental responses to the bad news. The model also provides real-time feedback about whether the mode of news delivery was effective or not, and thus allowing re-communication.[1]

Active Change Response

- Seeks to bring about engaged parental responses towards addressing their bad news
- Clinicians need to teach and show parents how to help.

Active Change includes three types of behaviour on the part of parents:

(a) information seeking
(b) taking steps to prevent deterioration from the condition; and
(c) instigating treatment that brings about improvements in their child's development.

- Provide ongoing information, to lower anxiety.
- Provide information about a range of evidence-based treatments and where to access these treatments.
- Foster parental problem-solving and raising awareness about their child's needs, so that they can coordinate and plan their child's care.
- Parent education and coaching in how to parent their child is often required.[1]

1. Baile WF, Buckman R, Lenzi R, Glober G, Beale EA, Kudelka AP. SPIKES – A sixstep protocol for delivering bad news: application to the patient with cancer. Oncologist 2000; 5: 302–11.

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE

THE UNIVERSITY OF SYDNEY

AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

Acceptance response

The “Acceptance” response to bad news is where parents come to accept their circumstances and are able to create meaning in their loss, “reduce their dread over what lies ahead”, and seek support in order to cope .

Acceptance involves two types of behaviour on the part of parents:

(i) information sharing about their story with others, and
(ii) accommodation, which actively involves incorporating their child’s diagnosis into their family life, by reordering priorities and adjusting to a new future.

As we build relationships with parents over time, it is important to acknowledge and listen to their expertise.

Parents want equal and cooperative relationships with clinicians.

Hear and understand what is important to families, empowering them by discussing openly their hopes and goals, always referring to the child by name and avoiding labelling them as abnormal or by their diagnosis.

Clinicians also need to stay up to date regarding prevalence of disability and common comorbidities so that they can provide accurate and balanced information that is personalised to each family[1].

1. Sweeny K. & Shepperd JA. Being the Best Bearer of Bad Tidings. Review of General Psychology 2007, Vol. 11, No. 3, 235–257.

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Communication of diagnosis

Types of Information and Knowledge Needed by Parents

1 DIAGNOSIS

DEFINITION

Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitations, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain.[1]

PREVALENCE

Cerebral palsy is the most common cause of physical disability in childhood. Link: What is cerebral palsy?

TYPES

Motor Types, Topography and Classifications:

Four motor types exist but may emerge and change during the first two years of life:

(1) Spasticity. Spasticity is categorised topographically as (i) unilateral (hemiplegia) 38% and (ii) bilateral (including diplegia lower limbs affected more than upper limbs) 37% and quadriplegia (all 4 limbs and trunk affected) 24%[2]

(2) Dyskinesia including dystonia and athetosis;

(3) Ataxia; and

(4) Hypotonia.

There may be more than one motor disorder. A combination of spasticity and dystonia is common[2]. In childhood, several objective classification tools exist to classify the child's function, including the Gross Motor Function Classification System, (GMFCS- E&R)[3] and MACS Manual Ability Classification Systems[4].

EARLY DIAGNOSIS

Cerebral palsy or high risk of cerebral palsy can be accurately diagnosed early, in many cases under 6 months corrected age. High quality evidence indicates the combination of medical history and standardised tools should be used to predict risk. Before 5 months corrected age, neuroimaging (MRI), the Hammersmith Infant Neurological Examination (HINE) and Prechtl's General Movements Assessment (GMs) are the most predictive tools. After 5 months corrected age, MRI and the HINE are most predictive of risk for cerebral palsy. [5] When standardised assessments indicating cerebral palsy is suspected, the interim clinical diagnosis of "high risk" of cerebral palsy should be given. Essential criterion of motor dysfunction and at least one of the additional criteria of abnormal neuroimaging or clinical history indicating risk for cerebral palsy are required. This should be followed by referral for cerebral palsy specific early intervention and parent or carer support. Ongoing monitoring to assist in forming the diagnostic picture is recommended.[5]

Neurological Test: A Hammersmith Infant Neurological Evaluation (HINE) score below 57 at 3 months is 96% predictive of cerebral palsy. A HINE score below 40 at 3 months never occurs in children with normal outcomes.[6,7]

Motor Test: An abnormal General Movements Assessment score of "absent fidgety movements" at 12–20 weeks corrected age is 95-98% predictive of cerebral palsy.[8]

Link: https://www.cerebralpalsy.org.au/services/for-children/newly-diagnosed/

CURE

The complete causal path to cerebral palsy is unclear in 80% cases, but clinical risk factors are often identifiable and include risks prior to conception, during pregnancy, around the time of birth and postneonatally.[10]

PROGNOSIS

Diagnosticians should answer questions about prognosis as accurately and clearly as possibly, whilst maintaining a positive outlook. Where appropriate, the use of accurate prognostic facts such as "most children with cerebral palsy will walk" [11] can create a positive picture of a child with cerebral palsy for families.

0-2 years old: In children under 2 years of age motor severity is most accurately predicted using the HINE[12,13],and MRI.[14] Caution should be taken when giving prognostic information about motor prognosis under the age of 2 years, as voluntary movement, myelination and brain growth is still developing.

Over 2 years old: In children over 2 years of age, the severity of gross motor function is most reliably classified using the GMFCS.[3]

Link: Gross Motor Function Classification System (GMFCS – E& R)

PREVENTION OF COMPLICATIONS

The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems.[1]

- 3 in 4 experience chronic pain;
- 1 in 2 have an intellectual disability;
- 1 in 3 cannot walk;
- 1 in 3 have hip displacement;
- 1 in 4 cannot talk;
- 1 in 4 have epilepsy;
- 1 in 4 have a behaviour disorder;
- 1 in 4 have bladder control problems;
- 1 in 5 have a sleep disorder;
- 1 in 5 dribble;
- 1 in 10 are blind;
- 1 in 15 are tube fed;
- and 1 in 25 are deaf.[15]

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE

THE UNIVERSITY OF SYDNEY

AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

Co-occurring impairments are strongly linked to the severity of motor impairment. Medical investigations for associated impairments are always indicated in cerebral palsy. There are evidenced based recommendations and expert opinion care pathways for the medical and surgical management of children with cerebral palsy.[16] Early management of associated impairments and timely evidenced based interventions can improve outcomes.[17]

HIPS: Hip displacement occurs in 1 in 3 children with cerebral palsy. Children with non-ambulant bilateral cerebral palsy are most at risk. Hip dislocation can be prevented through early surveillance and management.[17]
Link: Australian Hip Surveillance Guidelines

PAIN: 3 in 4 children with cerebral palsy have chronic pain. Detailed pain assessment and management is important but often overlooked.[18]

SLEEP: 1 in 5 children with cerebral palsy have sleep problems, specialist assessments and early treatment are recommended.[19] Pain, breathing problems, vision impairment and epilepsy can affect sleep.[20]

Link: American Academy for Cerebral Palsy and Developmental Medicine Care Pathways

2 TREATMENT

TREATMENT PLAN

Goal based approach: Best practice rehabilitation and psychological evidence supports treatment planning based on child and family goals. Active involvement of the parents in all decision making and treatment goal setting, considering family values, expectations and preferences achieves better outcomes for children and parents.[21]

EARLY INTERVENTIONS

Cerebral palsy specific interventions exist, and are increasingly tailored to a specific type of cerebral palsy. Categorisation by typography of unilateral or bilateral is important to guide intervention. For example, early bimanual and constraint-induced movement therapy (CIMT) are recommended for unilateral cerebral palsy.[22] Emerging evidence is supporting early task-specific, child-initiated, and enriched environmental interventions for motor and cognitive gains[21]. Aims of optimising motor, cognitive, and communication outcomes for children, prevention of secondary impairments and promoting caregiver coping and mental health should all be considered in treatment plans.[22]

MEDICATIONS

Pharmacology can play a role for people with cerebral palsy in promoting health and secondary preventions. Effective pharmacologic interventions include the management of symptoms such as epilepsy, pain, tone management (baclofen, intrathecal baclofen (diazepam, botulinum toxin type A) and bone density (bisphosphonates).[23]

LEISURE

Children with cerebral palsy are able to be actively involved in a wide range of leisure activities, and experience a high level of enjoyment. Data suggests however, that they participate less in physically active leisure compared with peers, and that participation reduces over time. Parents of children with cerebral palsy rank participation as their second most important research priority.[26]

EXPECTED OUTCOMES

Communication regarding interventions and expected outcomes needs to be honest, holistic, family-centred and mindful of the ethical complexities in supporting and responding to families' hopes, goals and requests for treatments.[27] Parents are challenged from choosing a wide variety of therapy options, many of which have uncertain effects with some proven to be ineffective.[23] To enable discussion on expected outcomes of interventions, a defined goal, including its relative level of the International Classification of Functioning, Disability and Health (ICF) needs to be established.[28]

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

3 EQUIPMENT

ASSISTIVE DEVICES AND EQUIPMENT

Most children with cerebral will have some difficulty with functional independence. Assistive devices (such as for walking, communication) and equipment (such as wheeled mobility, seating and pressure mattresses) can help with independence, conserving energy and time, improving safety, and reducing your caregiving burden (such as shower chairs, toilet supports and lifting devices).

FINANCIAL SUPPORT

Understanding the complexities of funding systems for children with a disability can be challenging for health professionals and stressful and time consuming for parents.

4 DAILY CAREGIVING

BURDEN OF CARE GIVING

Parents and carers may experience an increased parenting burden, including complex care responsibilities, social isolation or financial hardship as a result of their child's disability.[29] Parent and carer wellbeing can impact on child outcomes.[29]

BEHAVIOUR MANAGEMENT

1 in 4 children with cerebral palsy have a behaviour disorder. Rates of behaviour disorders are higher in children with an intellectual impairment, in children with severe pain and in children with lower severity levels of cerebral palsy. Parent education is recommended in behaviour management. The positive parenting programme, (Triple P) can be effective in reducing disruptive behaviours of children with developmental disabilities.[30]

Link: Triple P Positive Parenting Program

5 FUTURE

DEVELOPMENTAL TRAJECTORY OF GROSS MOTOR SKILLS

Gross Motor curves exist that describe motor potential and the point at which motor development plateaus, which can inform the development of a realistic treatment plan. Children with ambulant cerebral palsy achieve 90% of their gross motor development potential by age 5 years, and children with non-ambulant reach 90% of their gross motor development by age 3.5 years. Before a child plateaus, focusing on active skill development is important, after the child plateaus it is important to prescribe compensatory equipment (such as a wheelchair) to ensure the child is fully included.

ADOLESCENCE

Children with cerebral palsy can show signs of ageing and physical decline in their adolescent years.[32]

ADULTHOOD

Compared with able bodied peers, adults with cerebral palsy are less likely live independently, have intimate relationships or maintain gainful employment.[33] Adults with cerebral palsy may require assistance with employment initiatives, advocacy needs to continue to counter discrimination, appropriate accommodation, transport, equipment and home modifications.[34]

EDUCATION

Children with disability have the same education rights as all other children. Educational rights are protected by law. Parents are encouraged to explore options available for their child. Access to appropriate support and adaptations to allow for inclusion in education should be provided.

Link: Raising Children Network Education Rights in Australia

LIFE EXPECTANCY

Cerebral palsy is permanent and lifelong. Life expectancy is almost always normal. Life expectancy decreases with increasing intellectual disability, epilepsy and increasing physical disability. There have been significant improvements in survival of children with severe cerebral palsy in recent decades.[36]

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE

THE UNIVERSITY OF SYDNEY

AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

6 HEALTHCARE PLANS

RECOMMENDED REVIEWS

Surveillance: Health Surveillance programs aimed at identifying onset of associated impairments and referring to timely interventions to prevent complications exist. Early management and evidenced based interventions can improve outcomes. Examples are CPUP in Sweden, CP CheckUp in Australia and hip surveillance programs.

7 SUPPORT

CEREBRAL PALSY STRATEGY

The Australian and New Zealand Cerebral Palsy Strategy reflects a united voice informed by people with cerebral palsy, their families, professionals and researchers across Australia and New Zealand.
www.cerebralpalsystrategy.com.au

PARENT TO PARENT SUPPORT

Parents indicate parent-to-parent and family support facilitates long term coping[37].
Link: Parent Tip Sheet Parent2Parent CanChild

COMMUNITY SUPPORT

Informal and formal community supports[37] can play important roles in active change and acceptance responses.

LEGISLATION, ADVOCACY & FINANCIAL

Advocacy is of utmost significance in a child's support system.[37] Anti-discrimination legislation exists to ensure people with a disability are not discriminated against.
Link: Raising Children Network

8 FAMILY SUPPORT

SIBLINGS

The impact of cerebral palsy on the whole family is complex and challenging. Siblings may require their own individual support.
Links and Books:
Raising Children Network Siblings
CP NOW Toolkit - Impact on CP Diagnosis on Family and Siblings
Views from our Shoes: Growing up with a Brother or Sister with Special Needs, Donald J. Meyer

COPING

The process of parental acceptance of a cerebral palsy diagnosis, grieving, coping and resiliency is ongoing, cyclical and requires a continuum of supports from diagnosticians. Parent child attachment and caregiver mental health interventions such as acceptance and commitment therapy are helpful interventions to assist with caregiver coping.
Books:
Uncommon Fathers: Reflections on Raising a child with a disability. Donald J Meyer.
Married with Special needs children: A couples guide to Keeping connected, Laura Marshak and Fran Prezant.

PARENT MENTAL HEALTH AND WELLBEING

Mothers of a child with a disability report high rates of distress, anxiety, depressions and suicidality.[38] 1 in 4 parents of children with CP have very high stress. Mothers report the perceived need for professional mental health support, and support is most wanted around the time of diagnosis.[38]
Link: Parent Wellbeing Resource

PARENT INFORMATION

Australian and New Zealand Cerebral Palsy Strategy Collaboration
www.cerebralpalsystrategy.com.au
Cerebralpalsy.org.au
Canchild.ca
Cpnowfoundation.org
Cpdailyliving.com
Cdc.gov
Neurodevnet.ca
Ucp.org
Scope.org.uk
Cpsn.org.au
Cpfamilynetwork.org
Reachingforthestars
yourcpf.org/
Aacpdm.org.
Cerebralpalsy.org.uk
Ausacpdm.org.au.

9 EXPLANATIONS TO OTHERS

EXPLAINING THE DIAGNOSIS

"Talking to family and friends, or showing them resources about your child's disability can help them understand and support you. But what you talk about, how much you say and who you talk to is up to you."
Links: Talking to Others About Your Child and CP.
Raising Children Network - Talking About Disability
CP NOW Toolkit: Impact on CP Diagnosis on Family and Siblings
Children's Books:
What Are Your Superpowers? By Marget Wincent ISBN 9781540897817;
Jessica's Box By Peter Carnavas ISBN 9781921928574;
My Friend Suhana. By Shaila Abdullah and Aanyah Abdullah ISBN 9781615992119.

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

References

1. Rosenbaum P, Paneth N, Leviton A, Goldstein M, Bax M. A report: the definition and classification of cerebral palsy April 2006. Dev Med Child Neurol. 2007;49:8-14.
2. Report of the Australian Cerebral Palsy Register, Birth Years 1993-2006, February 2013. Sydney;Cerebral Palsy Alliance.
3. Palisano R, Rosenbaum P, Walters SD, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Developmental Medicine & Child Neurology 1997; 39: 214-23.
4. Eliasson AC, Krumlinde-Sundholm L, Rosblad B, Beckung E, Arner M, Ohrvall AM, Rosenbaum P. The Manual Ability Classification System (MACS) for children with cerebral palsy: scale development and evidence of validity and reliability. Developmental Medicine & Child Neurology 2006; 48: 549-54.
5. Novak I, Morgan C, Adde L, Blackman J, Boyd RN, Brunstrom-Hernandez J, Cioni G, Damiano D, Darrah J, Eliasson AC, de Vries LS, Einspieler C, Fahey M, Fehlings D, Ferriero DM, Fetters L, Fiori S, Forssberg H, Gordon AM, Greaves S, Guzzetta A, Hadders-Algra M, Harbourne R, Kakooza-Mwesige A, Karlsson P, Krumlinde-Sundholm L, Latal B, Loughran-Fowlds A, Maitre N, McIntyre S, Noritz G, Pennington L, Romeo DM, Shepherd R, Spittle AJ, Thornton M, Valentine J, Walker K, White R, Badawi N. Early, accurate diagnosis and early intervention in cerebral palsy: Advances in diagnosis and treatment. JAMA Pediatr. 2017.
6. Romeo DM, Cioni M, Scoto M, Mazzone L, Palermo F, Romeo MG. Neuromotor development in infants with cerebral palsy investigated by the Hammersmith Infant Neurological Examination during the first year of age. Eur J Paediatr Neurol 2008; 12: 24-31.
7. Romeo DM, Cioni M, Palermo F, Cilauro S, Romeo MG. Neurological assessment in infants discharged from a neonatal intensive care unit. Eur J Paediatr Neurol 2013; 17: 192-8.
8. Bosanquet M, Copeland L, Ware R, Boyd R. A systematic review of tests to predict cerebral palsy in young children. Dev Med Child Neurol 2013; 55: 418-26.
9. ANZCTR.org.au. Preventing Adverse Outcomes of Neonatal Hypoxic Ischaemic EncephalopathyWith Erythropoietin: a Phase III randomised placebo controlled multicentre clinical trial. ACTRN12614000669695
10. McIntyre S, Morgan C, Walker K, Novak I. Cerebral palsy-don't delay. Dev Disabilities Res Rev. 2011;17(2):114-291
11. ACPR Group. Australian cerebral palsy register report 2018, birth years 1995-2012. Sydney, Australia: Cerebral Palsy Alliance; 2018
12. Romeo DM, Cioni M, Scoto M, Mazzone L, Palermo F, Romeo MG. Neuromotor development in infants with cerebral palsy investigated by the Hammersmith Infant Neurological Examination during the first year of age. Eur J Paediatr Neurol 2008; 12: 24-31.
13. Romeo DM, Cioni M, Palermo F, Cilauro S, Romeo MG. Neurological assessment in infants discharged from a neonatal intensive care unit. Eur J Paediatr Neurol 2013; 17: 192-8.
14. Towsley K, Shevell MI, Dagenais L, REPACQ Consortium. Population-based study of neuroimaging findings in children with cerebral palsy. Eur J Paediatr Neurol 2011; 15: 29-35.
15. Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy. Pediatrics. 2012;130(5):e1285-312
16. American Academy for Cerebral Palsy and Developmental Medicine Care Pathways https://www.aacpdm.org/publications/care-pathways
17. Hägglund, G., Andersson, S., Düppe, H., Lauge-Pedersen, H., Nordmark, E., & Westbom, L.Prevention of dislocation of the hip in children with cerebral palsy. The journal of bone and joint surgery (Br) 2005; 87:1: 95-101.
18. Westbom L, Rimstedt A, Nordmark E. Assessments of pain in children and adolescents with cerebral palsy: a retrospective population-based registry study. Dev Med Child Neurol 2017; 59: 858–863. doi:10.1111/dmcn.13459
19. Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy. Pediatrics. 2012;130(5):e1285-312.
20. Simard-Tremblay, E., Constantin, E., Gruber, R., Brouillette, R. T., & Shevell, M. Sleep in children with cerebral palsy: A review. Journal of Child Neurology 2011; 26:10: 1303-1310.
21. Morgan C, Novak I, Dale RC, Badawi N. Optimising motor learning in infants at high risk of cerebral palsy: a pilot study. BMC Pediatr 2015; 15: 30.
22. Eliasson AC, Holmefur M. The influence of early modified constraint induced movement therapy training on the longitudinal development of hand function in children with unilateral cerebral palsy. Dev Med Child Neurol 2015; 57: 89-94.
23. Novak I, McIntyre S, Morgan C, et al. State of the evidence: systematic review of interventions for children with cerebral palsy. Dev Med Child Neurol. 2013;55:885-910.
24. Majnemer A, Shevell M, Law M, Birnbaum R, Chilingaryan G, Rosenbaum P, Poulin C. Participation and enjoyment of leisure activities in school-aged children with cerebral palsy. Dev Med Child Neurol 2008; 50: 751–758. doi:10.1111/j.1469-8749.2008.03068.x
25. Majnemer A, Shikako-Thomas K, Schmitz N, Shevell M, Lach L. Stability of leisure participation from school-age to adolescence in individuals with cerebral palsy. Res Dev Disabil. 2015;47:73-9.
26. McIntyre S, Novak I, Cusick A. Consensus research priorities for cerebral palsy: A delphi survey of consumers, researchers, and clinicians. Dev Med Child Neurol. 2010;52 :3:270-5.
27. McIntyre S, Novak I, Cusick A. Consensus research priorities for cerebral palsy: A delphi survey of consumers, researchers, and clinicians. Dev Med Child Neurol. 2010;52 :3:270-5.
28. Novak I. Evidence-Based Diagnosis, Health Care, and Rehabilitation for Children With Cerebral Palsy Journal of Child Neurology 2014; 29:8: 1141-1156.
29. McLennan JD, Kotelchuck M. Parental Prevention Practices for Young Children in the Context of Maternal Depression. Pediatrics. 2000; 105:5:1090-5.
30. Skotarczak L, Lee G. Effects of parent management training programmes on disruptive behavior for children with a developmental disability: A meta-analysis. Science Direct 2015; 38:272-287.
31. Rosenbaum PL, Walter SD, Hanna SE, Palisano RJ, Russell DJ, Raina P, Wood E, Bartlett DJ, Galuppi BE. Prognosis for gross motor function in cerebral palsy: Creation of motor development curves. JAMA. 2002; 288: 11: 1357-1363. [PubMed: 12234229]
32. Novak I. & Msall, M. Cerebral palsy. In W.F. Malcom (ed). Beyond the NICU: Comprehensive Care of the High-Risk Infant. McGraw Hill Professional; 2014a; Chapter 24.
33. Michelsen SI, Uldall P, Kejs AMT, Madsen M.'Education and employment prospects in cerebral palsy' Developmental Medicine and Child Neurology 2005; 47:511-517
34. Economic Impact of CP in Australia 2007 http://cpaustralia.com.au/media/20379/access_economics_report.pdf Last accessed March 12 2018.
35. Donkervoort M, Wiegerink DJHG,Van Meeteren J, Stam H, Roebroeck ME, Transition to adulthood:vadlidation of the Rotterdam Transition Profile for young adults with cerebral palsy and normal intelligence. Dev Med Child Neurol. 2009;51910:53-62.
36. Brooks JC, Strauss DJ, Shavelle RM, Tran LM, Rosenbloom L, Wu YW. Recent trends in cerebral palsy survival. part II: individual survival prognosis. Dev Med Child Neurol 2014; 56:1065-1071
37. Reid A, Imrie H, Brouwer E, Clutton S, Evans J, Russel D, Bartlett D (2011) 'If I knew then what I know now: parents' reflection on raising a child with cerebral palsy. Phys Occup Ther Pediatr 2011; 31:2: 169-183.
38. Whittingham, Koa, Wee, Diana, Sanders, Matthew R. and Boyd, Roslyn (2013) Sorrow, coping and resiliency: parents of children with cerebral palsy share their experiences. Disability and Rehabilitation, 35 17: 1447-1452.
39. Raising Children Network -Talking About Disability http://raisingchildren.net.au/articles/talking_about_disability.html

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Communication of diagnosis

Common questions asked by parents of children with cerebral palsy

PARENT QUESTIONS MOTOR TYPE/ PROGNOSIS	ANSWERS: WHAT CAN WE SAY/ WHAT CAN WE DO NOW ?
Case 1: **12-months old, Spastic hemiplegia, GMFCS I, HINE at 6-months 68.5, HINE at 12-months 67.5**	
Does she have a problem with high tone in her right leg?	*Today she has a "catch", or resistance to movement, in her calf muscle, which means she does have mild spasticity in this muscle.*
Can spasticity and contractures get worse when she is older?	*As she grows, spasticity might develop in other muscle groups. For children who have mild spastic hemiplegia, it is not unusual for contractures to develop over childhood, but we do not know when they will develop. Most often it is during growth spurts, when bones are growing faster than the muscles.*
Do children who have a similar type of CP also have a limp?	*Children with mild hemiplegia can have changes to their muscles and joints over time, which can go on to cause a limp. We do not know if and when this is likely to develop for your daughter. We recommend regular checks of her muscles and developmental skills, to understand her development and the best course of treatment.*
Will she be able to run when she grows up?	*Right now, she is developing her gross motor skills on par with children her age. This is good news. We also know that motor tests often don't show difficulty with more intricate skills until later on. We do predict that she is going to be able to run, but she might have difficulty with some higher-level skill such as hopping and skipping. There are therapy interventions which can help to target developing these higher-level skills.*
Case 2: **4-months old , Type and Typography Unknown, GMFCS too young to determine, HINE at 3-months 47**	
Can the results of the HINE change over time and move into the optimal range?	*Usually the HINE improves a little bit with time as children get older. For each age, there is an optimal score range. Multiple tests of the HINE over time give us a better understanding of his brain function.*
His MRI is clear, does this mean he will be OK?	*You are right, his MRI didn't show any clear abnormalities. In some cases, children with a clear MRI can still go on to have developmental problems, for example 10% of children with CP have a normal MRI, which means we need to keep monitoring him. This is especially important because of his history of Encephalopathy and his HINE score. What we recommend is doing a repeat HINE in 1 month and see what that shows us. Is there anyone else you would like to be at that appointment with you?*
Some days his hands are fisting, on other days they aren't. Last week you felt tone in his legs, and this week it isn't there. What does that mean?	*Your observations are right. He does have changing tone in his arms and legs – we call this variable movement "dystonia". The fluctuations are involuntary. We will monitor his dystonia closely over time to see if we need to add any new treatments or change treatment plans.*

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE

THE UNIVERSITY OF SYDNEY

AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

PARENT QUESTIONS MOTOR TYPE/ PROGNOSIS	ANSWERS: WHAT CAN WE SAY/ WHAT CAN WE DO NOW ?
Case 3: 9-months old, (Emerging) , Spastic diplegia, GMFCS Too young to determine, HINE at 3-months 59	
I have noticed he stands on his toes and holds his knees stiffly. Is the stiffness in his legs of concern?	*Stiffness in his legs is something we are concerned about. Today, he wasn't showing what we call a spastic catch. However, often muscle spasticity (or over-activity) doesn't present fully until around 12 months of age. This is because the insulation of the nerves, called myelination, is still emerging. We recommend checking his muscles over time. He is starting to show some early control of his legs, which can help counterbalance involuntary muscle stiffness. For now, keep up the therapy focussed on learning skilled movement and control of movement.*
I think his hands are OK, they don't seem to get stiff– do you?	*I agree, it seems like he has more stiffness in his legs than his arms when he is moving and playing. In my assessments, today I haven't felt any spastic catches in any muscles. We will keep monitoring him over time, and keep a close eye on how his movements and skills are developing. For now, keep giving him lots of opportunities to practice developing his hand skills.*
Some days his hands are fisting, on other days they aren't. Last week you felt tone in his legs, and this week it isn't there. What does that mean?	*Your observations are right. He does have changing tone in his arms and legs – we call this variable movement "dystonia". The fluctuations are involuntary. We will monitor his dystonia closely over time to see if we need to add any new treatments or change treatments.*

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Communication of diagnosis

Communicating difficult news via telehealth

Communicating a cerebral palsy diagnosis to parents should always be delivered with sensitivity and be supported by facts and practical resources, as outlined in Communication Fact Sheets 1-5. These considerations are even more important if the diagnosis has to be delivered via telehealth.

Telehealth creates a virtual barrier between practitioner and patients/parents and this needs to be taken into consideration when delivering sensitive news.

To assist the parents in receiving and processing the news, consider the following:

- ensure that you have allowed adequate time for the telehealth appointment so that you do not risk being cut off or interrupted;
- ensure that disruption, distractions and external noise are kept to a minimum;
- be aware that the parents are likely to be on the call from their own home with the possible distractions of the patient and/ or other children demanding attention;
- ensure that you have prepared for the appointment and have all records and reference material immediately to hand to allow you to best respond to parents' queries;
- pre-preparing follow up communication and links that can either be sent immediately after the telehealth appointment so that the parents experience continued and reinforced support; and
- setting a follow up tele-health appointment either by yourself or a support worker to assist parents - noting that they will have a lot more questions in the days following diagnosis than they had during the initial appointment.

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Communication of diagnosis

Parents experience grief and loss at the time of diagnosis or 'high-risk' notification, and therefore communication with a family should be a series of well-planned and compassionate conversations.

Communication should:

- Be face-to-face. Provide at least two face to face diagnostic information sharing sessions to facilitate comprehension, recall and acceptance.
- Have both parents or caregivers present (where appropriate).
- Be private.
- Be honest and jargon free. Provide honest, transparent and specific information about the diagnosis and prognosis as possible and explain the likely impact on the family.
- Be tailored to the family.
- Be followed by written information.
- Include recommendations to use parent-to-parent support and arrangement of early intervention.
- Include identification of strengths as well as limitations, to promote development of an optimistic outcome.
- Include invitation to ask questions.
- Allow for discussion of feelings and arrangement for a debriefing to help parents gather information and navigate service entry.

Adapted with permission from: Novak I., Morgan C., McNamara L., te Velde A. Best practice guidelines for communicating to parents the diagnosis of disability. 2019 Early Human Development. 2019: 139.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Associated impairments

Early intervention for cerebral palsy prevents complications

1 in 5 have sleep disorders[1]

Early sleep management improves academic performance and behaviour. Investigate whether untreated pain is a cause of sleep disruption. Arrange sleep investigations.

1 in 10 have vision impairments[1]

Cortical vision impairment responds to treatment. Confirm whether a child can track an object in good light. If not, refer to specialist vision services early.

1 in 3 will not walk[1]

Children with severe cerebral palsy reach 90% of their motor potential by age 3.5 years. Refer early to physiotherapy and occupational therapy for intensive early motor training.

1 in 3 have hip displacement[1]

Hip dislocation is preventable with regular hip surveillance. Arrange hip x-rays according to hip surveillance guidelines.

3 in 4 have chronic pain[1]

Reflux is a common source of infant pain. Long-term chronic neuropathic pain is more likely without early pain management.

1 in 2 have dysphagia[1]

Aspiration pneumonia is the leading cause of premature death. Refer to a speech pathologist if feeding difficulties exist.

Screen for associated impairments

Clinical diagnosis of cerebral palsy or the interim diagnosis ‘high-risk of cerebral palsy’ should always include standard medical investigations for associated impairments and functional limitations (e.g. vision impairment, hearing impairment, epilepsy).

1. Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy. *Pediatrics*. 2012; 130: 5: e1285-1312.

Multi-Disciplinary Management

Medical management for children with cerebral palsy

Cerebral palsy is a diagnosis characterised by motor impairment, however, the clinical scenario is always part of a more complex, and multisystem condition. Brain injury itself, in addition to the impact on muscle and sensory function, affects multiple domains:

Pain

Neurobehavioural, cognition, mood and learning

Musculoskeletal: bone health, hips, spine, pain

Growth and nutrition

Feeding, drooling and oral health

Sensory: vision, hearing, taste, smell, tactile, vestibular

Seizures

Sleep

Family function, carer stress, siblings health

Emotional health

Although the brain injury is non-progressive, the co-occurring impairments change over time, impacting further on function and quality of life.

In Australia, there is variability in surveillance systems for comorbid disease, which happens over a vast array of clinical care models, and speciality services. The evolution of the National Disability Insurance Scheme is also influencing how surveillance is done, appreciating that there can be many care providers across different locations for one patient.

Communication to the whole multi-disciplinary team is essential in providing holistic care, as well as information gathering for children with cerebral palsy.

It is our role to consider the clinical presentation at each review carefully.

Long term survival and care of children with cerebral palsy has improved over the last decades.[1] There are increasing proportions of patients with significant impairment surviving to adulthood.[1]

The burden of chronic health conditions, as well as multi-comorbidity, is significant. This includes the metabolic, cardiovascular and social and emotional sequelae of having differences in motor function and activity participation.

Severe motor subtypes of cerebral palsy have a demonstrable risk for such multi-morbidity.[2]

Preventative holistic health care needs to start in childhood and encompass nutrition, activity and participation, and mental health.

In the childhood years, a diagnosis of cerebral palsy itself is a risk factor for hospitalisation.

More severe motor types are associated with more frequent, longer admissions as well as considerable risk of readmission within a 12 month period.[3]

Pneumonia is the leading cause of death in cerebral palsy.[1]

1. Blair, E., Langdon, K., McIntyre, S. et al. Survival and mortality in cerebral palsy: observations to the sixth decade from a data linkage study of a total population register and National Death Index. BMC Neurol 19, 111 (2019).
2. Cremer N, Hurvitz EA, Peterson MD. Multimorbidity in middle-aged adults with cerebral palsy. Am J Med 2017; 130: 744. e9–15.
3. Meehan E, Reid SM, Williams K, Freed GL, Sewell JR, Vidmar S, Donath S, Reddihough DS. 2016. Hospital admissions in children with cerebral palsy: a data linkage study. Dev Med Child Neurol. 2017;59:512–9.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Pain

Pain

Pain is common in children and adults with physical disability. In people with cerebral palsy, rates of pain vary from 40-75%.[3]

Pain is often missed by treating clinicians,[5] but can also be missed by parents and carers. Possible contributing factors to this include lack of awareness of signs, impaired communication and normalisation.

The causes of pain in children and adults can be multifactorial – including spasticity and dystonia, musculoskeletal subluxations, contractures and dislocations (joints, hips, spine, jaw), gastrointestinal (gastroesophageal reflux, constipation), bone (osteopenia, fractures), neuropathic and dental.

Un-treated pain in infancy increases the risk for long-term neuropathic pain.[1,2]

Chronic pain impacts on activity, participation, and quality of life.

Follow-on effects are seen in child and parental mental health (in the context of concern for a child's ability to enjoy and participate in life activities; as well as a child's independence).[4]

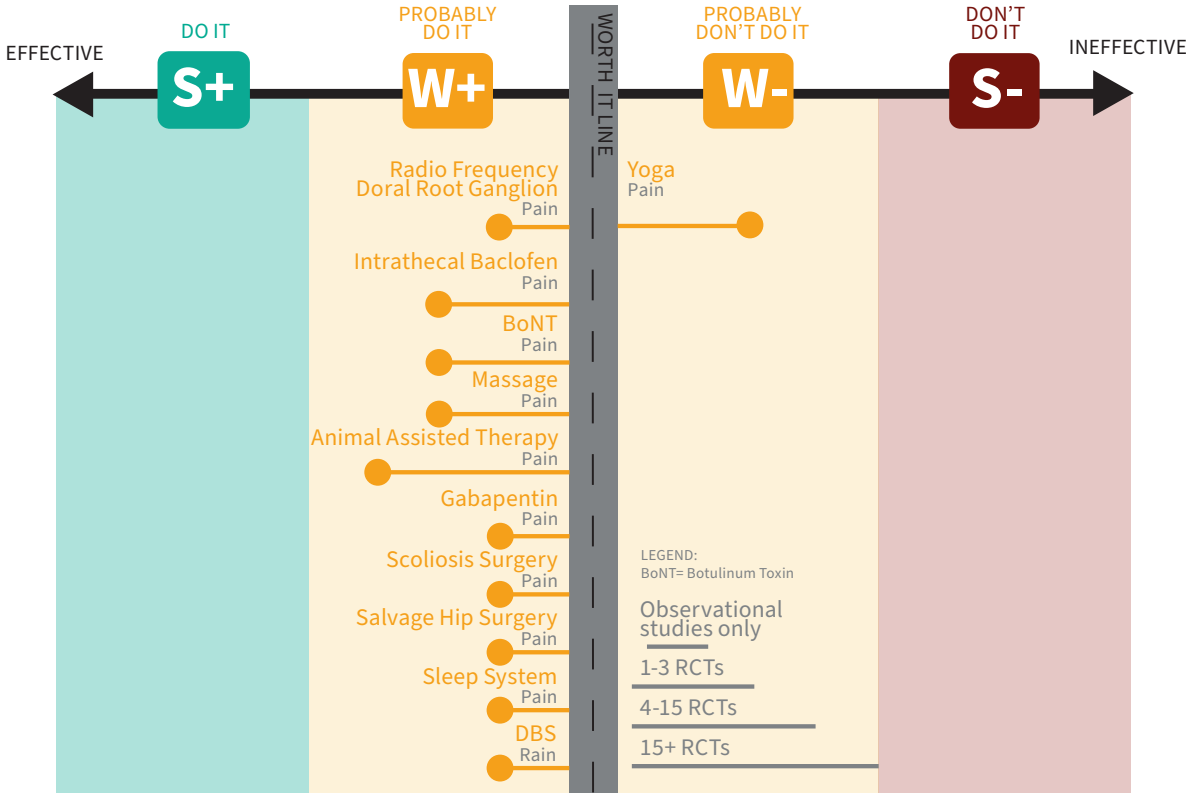
Consider pharmacological therapy and environmental interventions including analgesia for procedural pain in infants.[1,2]

Suggested pain management approaches[5] are to :

- Include screening in clinical practice.
- Educate patient and carer on pain, signs, and to "initiate a conversation with clinician.[5]
- Consider referral to a rehabilitation and/or pain specialist for management and monitoring.
- Refer for hip surveillance, and if indicated scoliosis surveillance.[6]

1. Anand KJ; International Evidence-Based Groupfor Neonatal Pain. Consensus statement for theprevention and management of pain in the newborn. Arch Pediatr Adolesc Med. 2001;155(2):173-180.
2. Novak I, Morgan C, Fahey M, et al. State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep. 2020;20(2):3. Published 2020 Feb 21.
3. Pain in adults with cerebral palsy: a systematic review and meta-analysis of individual participant data,Annals of Physical and Rehabilitation Medicine,2020
4. K. Ramstad, R. Jahnsen, O.H. Skjeldal, T.H. DisethParent-reported participation in children with cerebral palsy: The contribution of recurrent musculoskeletal pain and child mental health problems Developmental Medicine and Child Neurology, 54 (2012), pp. 829-835.
5. Fehlings D. Pain in cerebral palsy: a neglected comorbidity. Dev Med Child Neurol 2017; 59: 782–3.
6. Wynter M, Gibson N, Willoughby KL, et al;National Hip SurveillanceWorking Group.
7. Australian hip surveillance guidelines for children with cerebral palsy: 5-year review. Dev Med Child Neurol. 2015;57(9):808-820

Pain



Systematic overview of the best-available evidence (2012–2019) interventions in managing pain in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

Cognition

Cognition

Almost half of all children with cerebral palsy have co-occurring intellectual disability (46%) of varying severities.[1]

Cognitive impairment can include deficits in memory, reasoning, learning new skills, visuospatial awareness, attending and organising information. Impairment can coexist with differences in self-regulation.

These symptoms of brain dysfunction can be augmented by the physical condition and need to be considered in that context e.g. pain, postural fatigue, malnutrition and poor sleep.

EFFECTIVE COGNITIVE INTERVENTIONS

Assessments need to be multi-disciplinary and ideally include observation in the child's usual learning environment.

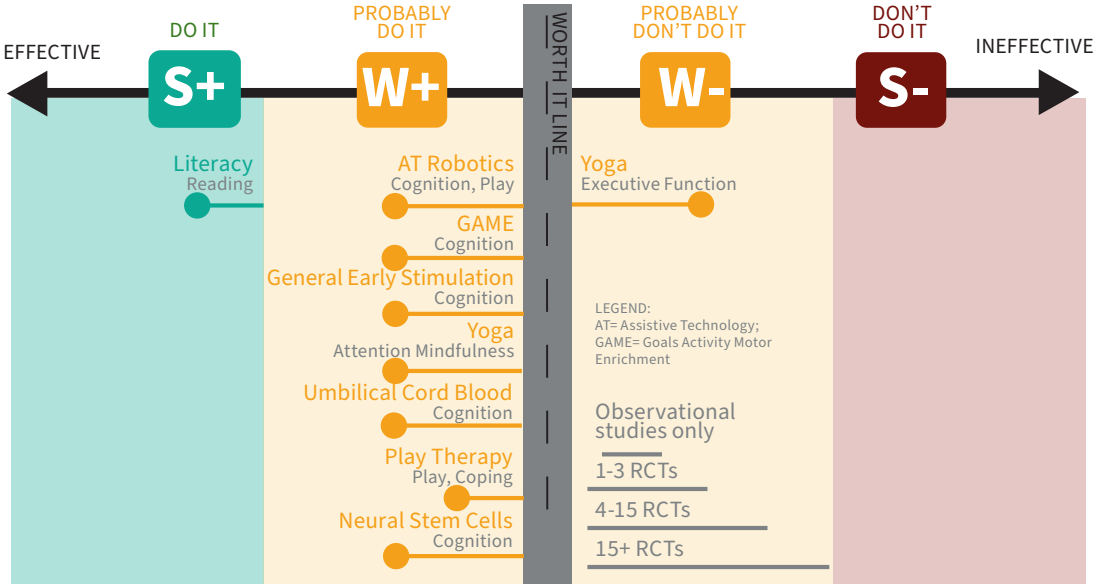
Intervention should be targeted to those gaps in function from such assessment, and relevant to each individual child.

Literacy interventions tailored for children with cerebral palsy using communication devices are effective.[2]

Physical and occupational therapy interventions should use child-initiated movement, task-specific practice, and environmental adaptations that stimulate independent task performance to target motor and cognitive outcomes.[3]

An example of an intervention that is suitable for all cerebral palsy subtypes is the GAME intervention (involving motor training, environmental enrichment, parent coaching and goal setting).[4] Infants who have received the GAME intervention have had better cognition at 1 year of age than age-matched peers on a norm-referenced test.[4]

The evidence for Cognitive orientation to occupational performance (CO-OP) intervention is also promising.[2,5]



Systematic overview of the best available cognitive interventions in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

1. ACPR Group. Australian cerebral palsy register report 2018, birth years 1995-2012. Sydney, Australia: Cerebral Palsy Alliance; 2018
2. Novak I, Morgan C, Fahey M, et al. State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep. 2020;20(2):3. Published 2020 Feb 21. doi:10.1007/s11910-020-1022-z
3. Morgan C, Darrah J, Gordon AM, et al. Effectiveness of motor interventions in infants with cerebral palsy: A systematic review. Dev Med Child Neurol. 2016; 58:9: 900-909.
4. Morgan C, Novak I, Dale RC, Badawi N. Optimising motor learning in infants at high risk of cerebral palsy: a pilot study. BMC Pediatr 2015; 15: 30.
5. Jackman M, Novak I, Lannin N, Froude E, Miller L, Galea C. Effectiveness of cognitive orientation to daily occupational performance over and above functional hand splints for children with cerebral palsy or brain injury: a randomized controlled trial. BMC Pediatr. 2018;18(1):248.

Behaviour

Behaviour

One in four children with cerebral palsy have a behaviour disorder.[1]

Essentially all of the medical comorbidities of cerebral palsy can influence behaviour. There are also a number of sociodemographic factors to consider, including family function and medical history.

Involve social work and psychology early.

Also consider issues relating to self-perception, cognition, emotional function of child and family and issues relating to grief and blame. Consider the lived experience of the child's condition and interventions.

BEHAVIOUR INTERVENTIONS

Parental education in behaviour management is recommended. An example is the Positive Parenting Program (Triple P)[2]. Parent-child attachment interventions are also helpful.

Referral to family centric services for support are recommended, such as behavioural therapists who can provide family-based intervention and supports.

Parent or caregiver mental health interventions are suggested.

One such intervention is Acceptance and Commitment Therapy (ACT).[3]

1. Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy. Pediatrics. 2012;130(5):e1285-312.
2. Skotarczak L, Lee G. Effects of parent management training programmes on disruptive behavior for children with a developmental disability: A meta-analysis. Science Direct 2015; 38:272-287.
3. Whittingham K, Sanders MR, McKinlay L, Boyd RN. Parenting intervention combined with acceptance and commitment therapy: a trial with families of children with cerebral palsy. J Pediatr Psychol. 2016;41(5):531–42.

Orthopaedics

Orthopaedics

HIP SURVEILLANCE

One in three children with cerebral palsy has hip dislocation, except in Nordic countries in which there are lower rates.[1,2] Children with severe cerebral palsy are most at risk.

Regular hip monitoring with x-ray can reduce hip dislocation and need for orthopaedic surgery.

There is moderate-quality evidence and a strong recommendation to use comprehensive hip surveillance practices to facilitate early detection and management of hip displacement.[3]

The frequency of ongoing hip surveillance is determined by Gross Motor Function Classification level, radiological measures and clinical assessment. [4]

https://www.ausacpdm. org.au/resources/australian-hip-surveillance-guidelines/

OSTEOPOROSIS

Low bone density is common in children with cerebral palsy. It can be asymptomatic and cause bone pain and also risk of traumatic fracture.

Once a child has had two long bone fractures, current standard practice is to refer to paediatric endocrinology for consideration of bisphosphonate.

Bisphosphonates improve bone mineral density, and can reduce bone pain. [5]

There is low-quality evidence for routine supplementation with Vitamin D.[5]

1. Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy. Pediatrics. 2012;130(5):e1285-312.
2. Hagglund G, Alriksson-Schmidt A, Lauge-Pedersen H, RodbyBousquet E, Wagner P, Westbom L. Prevention of dislocation of the hip in children with cerebral palsy: 20-year results of a population-based prevention programme. The Bone & Joint Journal. 2014;96-b(11):1546–52.
3. Novak I, Morgan C, Fahey M, et al. State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep. 2020;20(2):3. Published 2020 Feb 21
4. Wynter M, Gibson N, Willoughby KL, et al; National Hip SurveillanceWorking Group. Australian hip surveillance guidelines for children with cerebral palsy: 5-year review. Dev Med Child Neurol. 2015;57(9):808-820.
5. Novak I, McIntyre S, Morgan C, et al. State of the evidence: systematic review of interventions for children with cerebral palsy. Dev Med Child Neurol. 2013;55:885-910

Tone Management

Tone Management

SPASTICITY

85% of children with cerebral palsy have muscle spasticity as their primary motor type,and 7% have dyskinesia (including either dystonia or athetosis).[1]

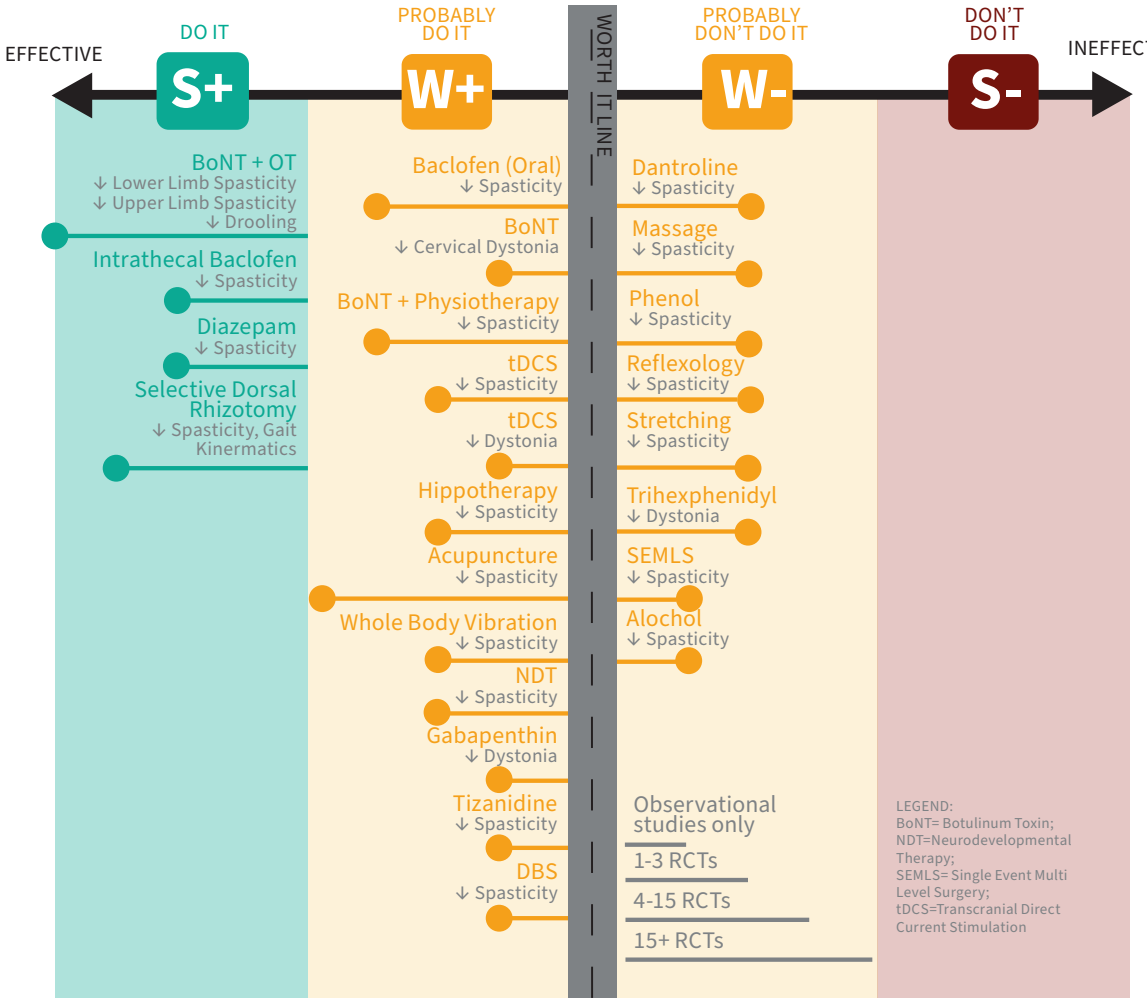
There are non-pharmacological and pharmacological treatments available. Management should be goal directed, and planned with involvement of the multidisciplinary team.

The following pharmacological agents and neurosurgical procedures effectively reduce spasticity: Botulinum toxin A, intrathecal baclofen, diazepam, and selective dorsal rhizotomy.[2]

Weak positive pharmacological evidence for treating dystonia include local injections of Botulinum toxin A, oral gabapentin, intrathecal baclofen via a pump, and oral trihexyphenidyl, but side effects may outweigh benefits for some children. [2]

Weak positive evidence supports Botulinum toxin A, intrathecal baclofen and gabapentin to reduce pain. [2]

Further research is indicated for deep brain stimulation for children with dystonia that causes pain.[2]



Systematic overview of best-available evidence (2012-2019) tone management interventions in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

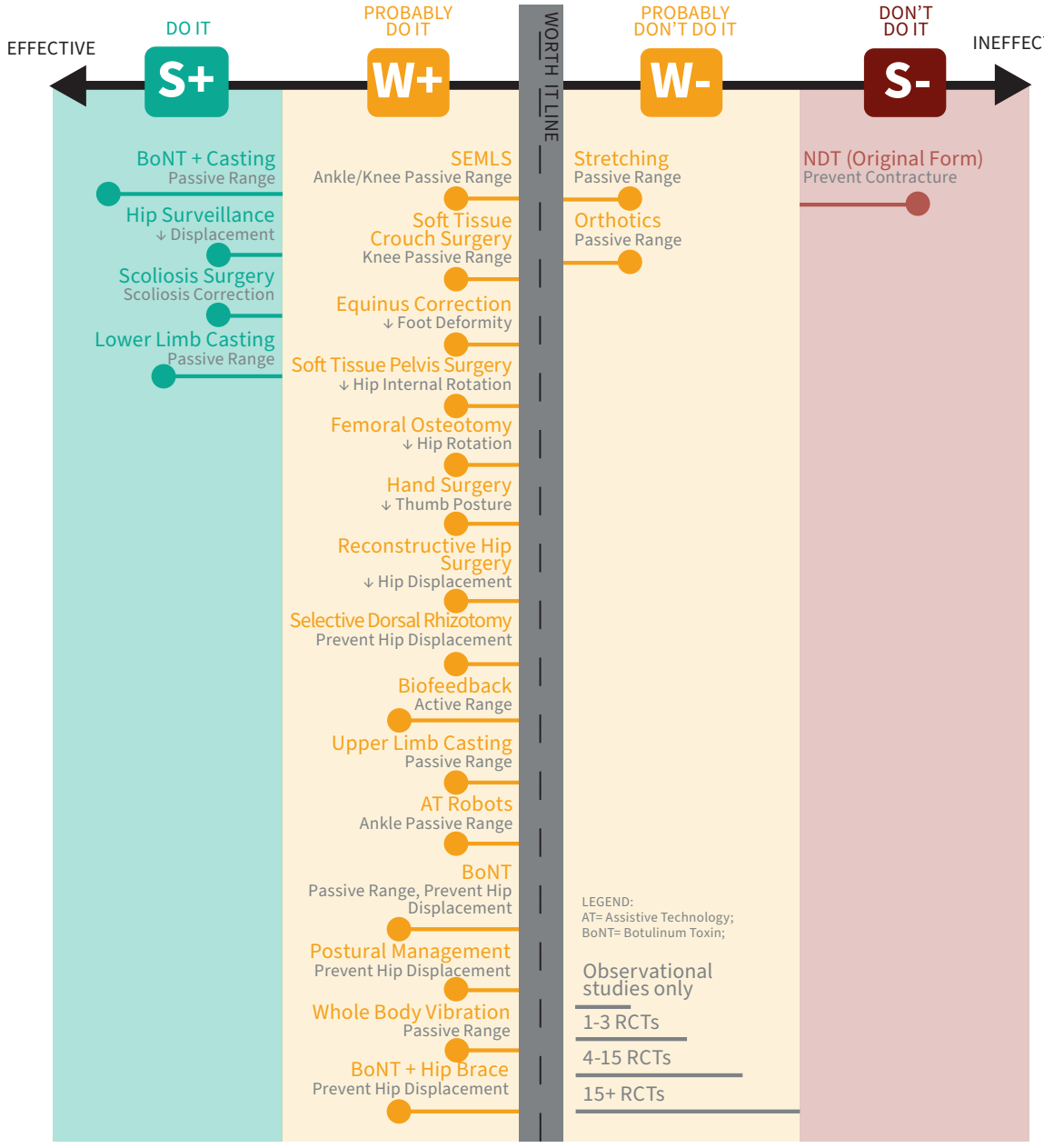
1. ACPR Group. Australian cerebral palsy register report 2018, birth years 1995-2012. Sydney, Australia: Cerebral Palsy Alliance; 2018
2. Novak I, Morgan C, Fahey M, et al. State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep. 2020;20(2):3. Published 2020
3. Hagglund G, Alriksson-Schmidt A, Lauge-Pedersen H, RodbyBousquet E, Wagner P, Westbom L. Prevention of dislocation of the hip in children with cerebral palsy: 20-year results of a population-based prevention programme. The Bone & Joint Journal. 2014;96-b(11):1546–52.

Contracture and Alignment

Contracture Prevention and Management

Early high intensity self-generated active movement to prevent the onset of weakness, disuse and muscle contracture is recommended. [1,2] Once a contracture has developed, serial casting is effective in the short term, followed by active movement, strength and goal-directed training.[1]

For severe contractures, orthopaedic surgery should be considered with the treating surgeon.[1]



Systematic overview of best-available evidence (2012-2019) contracture management interventions in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

1. Novak I, Morgan C, Fahey M, et al. State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep. 2020;20(2):3. Published 2020
2. Hagglund G, Alriksson-Schmidt A, Lauge-Pedersen H, RodbyBousquet E, Wagner P, Westbom L. Prevention of dislocation of the hip in children with cerebral palsy: 20-year results of a population-based prevention programme. The Bone & Joint Journal. 2014;96-b(11):1546–52.

Growth and Nutrition

Growth and Nutrition

Regular review of nutritional state, including weight, should be undertaken, as severe disability elevates the risk for malnutrition.[1]

One in 10 children with cerebral palsy requires neonatal tube feeding as a result of malnutrition.[2] Percutaneous endoscopic gastrostomy (PEG) and jejunostomy (J-tube) can help improve weight and growth.[3]

FEEDING

Feeding can be a highly emotional and stressful activity for caregivers and children. [4] Consideration needs to be given to co-ordination of movement patterns for a safe swallow, fatigability, efficiency, volume tolerance, sensory processing, impact of posture, impact of concurrent medical conditions and medications. Access to appropriate textures, nutrition and feeding equipment needs to be considered within a multi-disciplinary framework.

A multi-disciplinary assessment is required, led by a speech pathologist and/or dietitian.

For non-oral feeding, swallowing safety should be comprehensively assessed if concerns or clinical history of aspiration exists. Aspiration resulting in respiratory complications is the leading cause of death in individuals with cerebral palsy (45%)[5] and is mitigated by tube feeding.[6] Consideration needs to be given to the psychosocial support needs of parents and caregivers considering tube feeding.[4]

DYSPHAGIA MANAGEMENT

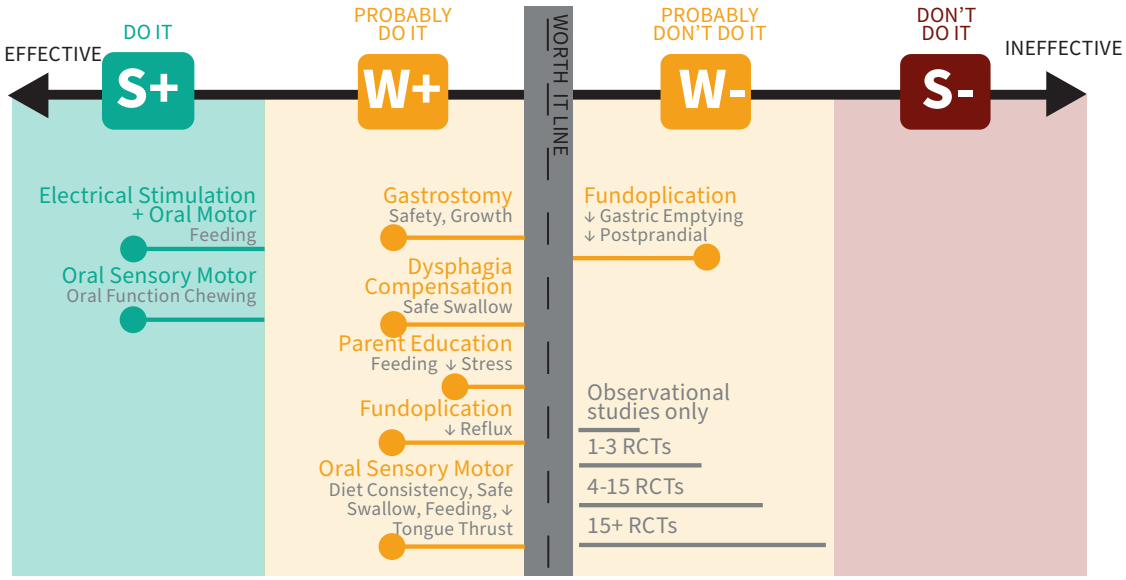
Half of all children with cerebral palsy have dysphagia[2] with the prevalence higher in infants.

Any child with cerebral palsy is at increased risk of swallow safety compromise, and as such, complications of aspiration, and longer term respiratory health.

New effective dysphagia management approaches to potentially lower the risk of aspiration include: electrical stimulation plus oral sensorimotor therapy[8]; and motor-learnings based oral sensorimotor intervention functional chewing training (FuCT).[9]

1. Kuperminc MN, Stevenson RD. Growth and nutrition disorders in children with cerebral palsy. Dev Disabil Res Rev. 2008;14(2):137-146.
2. Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy. Pediatrics. 2012;130(5):e1285-312.
3. Novak, I., McIntyre, S., Morgan, C., et al. . A systematic review of interventions for children with cerebral palsy: state of the evidence. Dev Med Child Neurol. 2013; 55(10), 885-910.
4. Craig, G. Psychosocial aspects of feeding children with neurodisability. Eur J Clin Nutr 67, S17–S20 (2013). https://doi.org/10.1038/ejcn.2013.226
5. Blair E,Watson L, Badawi N, Stanley FJ. Life expectancy among people with cerebral palsy in Western Australia. Dev Med Child Neurol. 2001;43
6. Sullivan PB, Juszczak E, Bachlet AM, et al.Gastrostomy tube feeding in children with cerebralpalsy: a prospective, longitudinal study. Dev MedChild Neurol. 2005;47(2):77-85.
7. Benfer K, Weir KA, Bell KL, Ware RS, Davies P, Boyd RN, Pediatrics. 2013;131 (5) e1553-e1562
8. Umay E, Gurcay E, Ozturk EA, Unlu AE. Is sensory-level electrical stimulation effective in cerebral palsy children with dysphagia? A randomized controlled clinical trial. Acta Neurol Belg. 2019.
9. Inal Ö, Serel Arslan S, Demir N, Tunca Yilmaz Ö, Karaduman AA. Effect of functional chewing training on tongue thrust and drooling in children with cerebral palsy: a randomised controlled trial. J Oral Rehabil. 2017;44(11):843–849.

Growth and Nutrition



Systematic overview of best-available evidence (2012-2019) feeding interventions in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

Respiratory

Respiratory

Attention is required for respiratory health as pneumonia is the leading cause of death in cerebral palsy.[1,2]

Careful assessment of swallow, secretion management and severity of reflux need to be assessed.

One in ten children require non-oral tube feeding as a result of aspiration.

There is a lifelong risk of increased vulnerability to respiratory complications. Children with truncal weakness and weak bulbar function may benefit from chest physio, and cough assist interventions.

Where possible, all children with cerebral palsy (and those sharing their care environment) should have immunisations for communicable disease effecting respiratory tract, including influenza. Children with severe impairment should also be considered for the pneumococcal vaccine.

GASTROESOPHAGEAL REFLUX

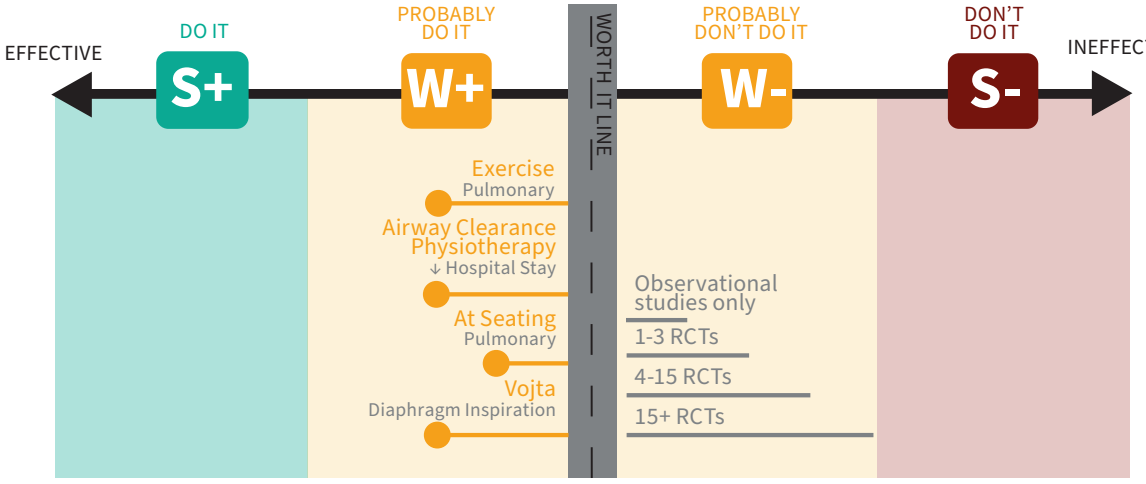
Three in four children with cerebral palsy have gastrooesophageal reflux, but prevalence has not been studied well in this population.

Early detection of typical signs and symptoms of gastroesophageal reflux disease (GERD) is important.

Signs and symptoms of GERD are often non-specific, including excessive crying or distress, back-arching posture, regurgitation, episodes of apnoea, feeding difficulties and / or poor growth, recurrent aspiration pneumonia, frequent otitis media.[3]

Management of GERD should follow a step-wise approach that initiates with non-pharmacological approaches (changes in feed volume and / or consistency, feed frequency, postural adjustments) when possible, and follows to pharmacological interventions, when necessary3.
Alginates and antacids (proton pump inhibitors and histamine H2 receptor antagonists) can be used in the management of gastroesophageal reflux with no cerebral palsy specific recommendations.[4]

Enteral nutrition (gastric or transpyloric) could be considered when GERD negatively affects growth. Finally, antireflux surgery (fundoplication) should be reserved for children with GERD in whom pharmacological approaches, as well as enteral feeding, have proved unsuccessful or impractical.[3]



Systematic overview of best-available evidence (2012-2019) respiratory interventions in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

1. Blair E,Watson L, Badawi N, Stanley FJ. Life expectancy among people with cerebral palsy in Western Australia. Dev Med Child Neurol. 2001;43 (8):508-515.
2. Novak I, Morgan C, Fahey M, et al. State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep. 2020;20(2):3.
3. Lopez RN, Lemberg DA. Gastro-oesophageal reflux disease in infancy: a review based on international guidelines. Med J Aust. 2020 Jan;212(1):40-44. doi: 10.5694/mja2.50447. Epub 2019 Dec 13. PMID: 31834639.
4. Vernon-Roberts A, Sullivan PB. Fundoplication versus post-operative medication for gastro-oesophageal reflux in children with neurological impairment undergoing gastrostomy. Cochrane Database Syst Rev 2007; 1: CD006151.

Drooling and Oral Health

Sialorrhea

Rates of drooling are highest with children with a more severe disability.[2] Drooling is a significant issue for both children and families. The negative impact of drooling goes beyond the physical function of saliva and can have a significant impact on self-esteem, confidence and image; as well as hygiene.

Saliva has an important role in dental health, lubrication and digestion of food. Drooling can vary in volume, can spill anteriorly from the mouth or posteriorly into the pharynx.

Abnormal loss of saliva in most children with spastic cerebral palsy is related to a disturbance in oral motor control rather than excessive production of saliva.[3] In children with dyskinetic cerebral palsy increased saliva may be the result of hyperkinetic oral movements.[3]

Assessment of cause requires a multi-disciplinary approach to consider factors such as reflux, nausea, food intolerance, oral health; posture and head position; swallow, oral and sensory-motor functions, medications (including clobazam and also oversedation) and seizure status.

Botulinum toxin A, benztropine mesylate, or glycopyrrolate management should be considered.[1]

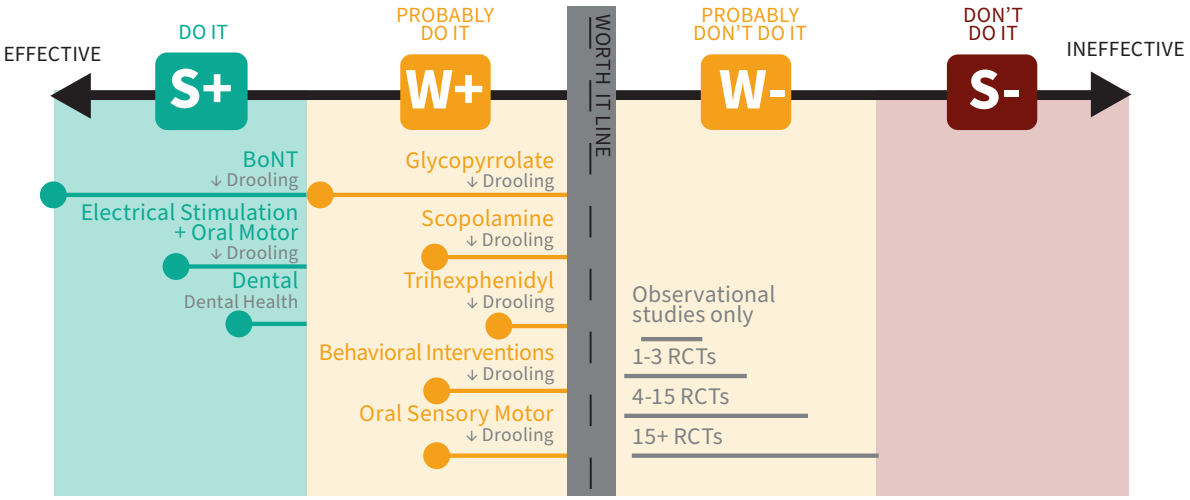
A comprehensive resource book "Saliva Control in Children" outlining interventions and management has been developed by the Royal Children's Hospital Melbourne. http://ww2.rch.org.au/emplibrary/plastic/salivabook.pdf\

Oral Health

Children with cerebral palsy are at higher risk of dental problems, which can significantly impact on the quality of life and well-being – including sources of infection and pain.

Screening for dental disease should be part of an initial assessment, and referral for management should be provided by specialised services with experience in treating children with neurological and developmental impairments.

Families should be provided with skills and guidance on appropriate equipment, such as modified brushes, to provide adaptive oral health care.



Systematic overview of best-available evidence (2012-2019) drooling and oral health interventions in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

1. Walshe M, Smith M, Pennington L.Interventions for drooling in children with cerebral palsy. Cochrane Database Syst Rev. 2012;11: CD008624.
2. Parkes J, Hill N, Platt MJ, Donnelly C. Oromotor dysfunction and communication impairments in children with cerebral palsy: a register study. Dev Med Child Neurol 2010; 52: 1113–9.
3. Erasmus CE, Van Hulst K, Rotteveel LJC, Jongerius P, Van den Hoogen, FJA, Roeleveld N, et al. Drooling in cerebral palsy: Hypersalivation or dysfunctional oral motor control? Developmental Medicine and Child Neurology 2009;51:454–459.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Management of Associated Impairments

Urinary Tract

Children with cerebral palsy are at higher risk of urinary tract complications. Abnormal anatomical findings are common in cerebral palsy and may require investigation.[1]

Any child with a central lesion can manifest signs of neurogenic bladder. There are a number of medications commonly used in people with cerebral palsy, including Baclofen, Botulinum toxin A, and anticholinergics that can result in detrusor relaxation and urinary retention.

A careful history of voiding ability and urinary tract infections should be collected.

Consider renal tract investigations, and referral to a paediatric urologist.

Constipation

One in three children with cerebral palsy will have constipation.[1]

Constipation is likely multi-factorial with nutrition, trunk movement and hydration status being involved.

Management principles are the same as for non-cerebral palsy populations.

In addition to this, supported standing in the upright position (e.g. in a standing frame)[2] and massage may help alleviate symptoms.[3]

Vision Assessments

Vision impairments can range from mild, requiring glasses, to functionally blind.

The visual pathway is complex, traversing and interconnecting with many levels of brain to result in visual function. Children can test to have normal acuity, but can have deficits in co-ordination of the pathway at any level, which results in cortical visual impairment. Carefully consider and assess not just acuity, fields and occulomotor function, but also how the child navigates in their environment, how they respond to visual information, and visual crowding.

Consider referral to optometry, and paediatric optometry with training in cortio-visual assessment.

Recommendations to use standard clinical visual functional examinations, MRI, visual evoked potential and a battery of behavioural assessments have been made.[4]

Early assessment of vision is possible in the first 48 hours of life in full-term infants.[5]

Any infant with abnormal vision at term-equivalent age should receive vision aware intervention and be reassessed at 3-6 months.[6] If vision awareness is uncertain at 6 months, assessment for cerebral vision impairment should occur.

Vision Australia is a good resource for families with visual impairment.

Hearing

Impairments can range from mild impairment to bilateral deafness.[7]

For hearing, standard early hearing accommodations are recommended.[8]

1. Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy. Pediatrics. 2012;130(5):e1285-312.
2. Rivi E, Filippi M, Fornasari E, Mascia MT, Ferrari A, Costi S. Effectiveness of standing frame on constipation in children with cerebral palsy: a single-subject study. Occup Ther Int. 2014;21(3):115-123.
3. Orhan C, Kaya Kara O, Kaya S, Akbayrak T, Kerem Gunel M, Baltaci G. The effects of connective tissue manipulation and Kinesio taping on chronic constipation in children with cerebral palsy: a randomized controlled trial. Disabil Rehabil. 2018;40(1):10–20.
4. Chorna OD, Guzzetta A, Maitre NL. Vision Assessments and Interventions for Infants 0-2 Years at High Risk for Cerebral Palsy: A Systematic Review. Pediatr Neurol. 2017;76:3-13.
5. Ricci D, Cesarini L, Groppo M, et al. Earlyassessment of visual function in full term newborns. Early Hum Dev. 2008;84(2):107-113.
6. Ricci D, Romeo DM, Gallini F, et al. Early visual assessment in preterm infants with and without brain lesions: correlation with visual and neurodevelopmental outcome at 12 months. Early Hum Dev. 2011;87(3):177-182.
7. Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy. Pediatrics. 2012;130(5):e1285-312.
8. Novak I, McIntyre S, Morgan C, et al. State of the evidence: systematic review of interventions for children with cerebral palsy. Dev Med Child Neurol. 2013;55:885-910.
9. Novak I, Morgan C, Adde L, Blackman J, Boyd RN, Brunstrom-Hernandez J, Cioni G, Damiano D, Darrah J, Eliasson AC, de Vries LS, Einspieler C, Fahey M, Fehlings D, Ferriero DM, Fetters L, Fiori S, Forssberg H, Gordon AM, Greaves S, Guzzetta A, Hadders-Algra M, Harbourne R, Kakooza-Mwesige A, Karlsson P, Krumlinde-Sundholm L, Latal B, Loughran-Fowlds A, Maitre N, McIntyre S, Noritz G, Pennington L, Romeo DM, Shepherd R, Spittle AJ, Thornton M, Valentine J, Walker K, White R, Badawi N. Early, accurate diagnosis and early intervention in cerebral palsy: Advances in diagnosis and treatment. JAMA Pediatr. 2017

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

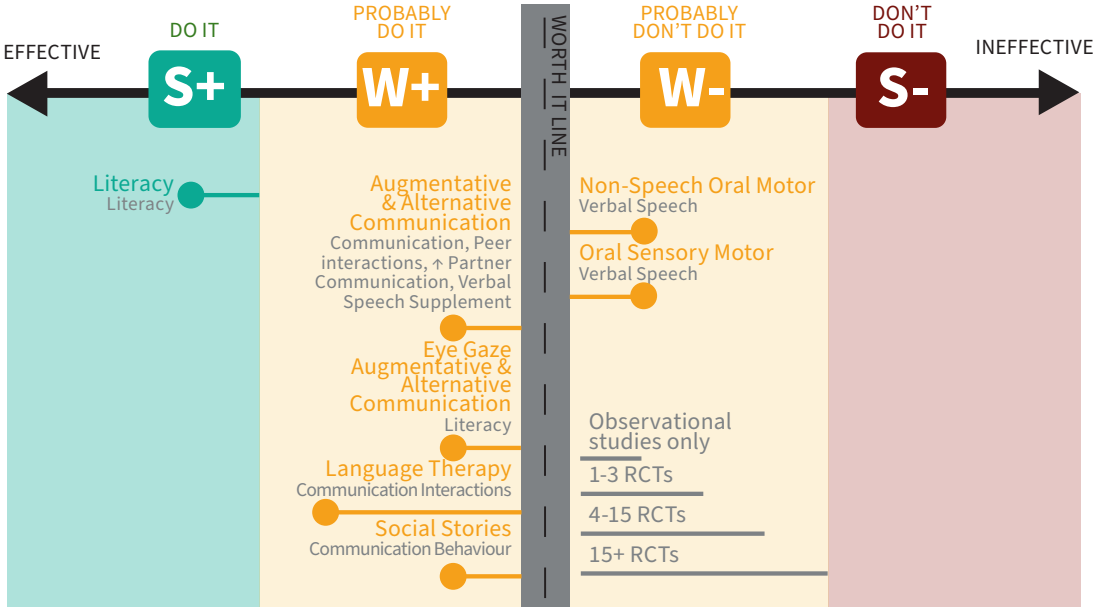
Communication

Communication Non Verbal (1 In 25)

Up to 60% of children with cerebral palsy have a speech impairment, and up to 40% are non-verbal, requiring alternative communication.[1] Children are more likely to be non-verbal if not walking, GMFCS IV and V.

For infants with cerebral palsy, interventions should promote parent-infant transactions.[2]

Examples include the “Hanen It Takes Two to Talk” and “More Than Words” programmes,[3] as well as alternative and augmentative communication.[2]



Systematic overview of best-available evidence (2012-2019) communication interventions in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

1. Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy. Pediatrics. 2012;130(5):e1285-312.
2. Novak I, Morgan C, Adde L, Blackman J, Boyd RN, Brunstrom-Hernandez J, Cioni G, Damiano D, Darrah J, Eliasson AC, de Vries LS, Einspieler C, Fahey M, Fehlings D, Ferriero DM, Fetters L, Fiori S, Forssberg H, Gordon AM, Greaves S, Guzzetta A, Hadders-Algra M, Harbourne R, Kakooza-Mwesige A, Karlsson P, Krumlinde-Sundholm L, Latal B, Loughran-Fowlds A, Maitre N, McIntyre S, Noritz G, Pennington L, Romeo DM, Shepherd R, Spittle AJ, Thornton M, Valentine J, Walker K, White R, Badawi N. Early, accurate diagnosis and early intervention in cerebral palsy: Advances in diagnosis and treatment. JAMA Pediatr. 2017
3. Chorna O, Hamm E, Cummings C, Fetters A,Maitre NL. Speech and language interventions forinfants aged 0 to 2 years at high risk for cerebral palsy: a systematic review. Dev Med Child Neurol. 2017;59(4):355-360.

Epilepsy and Sleep

Epilepsy

One in three children with cerebral palsy has epilepsy at some point. One in four has ongoing epilepsy. Rates are higher in children with a severe physical disability and intellectual disability.[1]

For epilepsy, standard antiepileptic pharmacological management is recommended.[2]

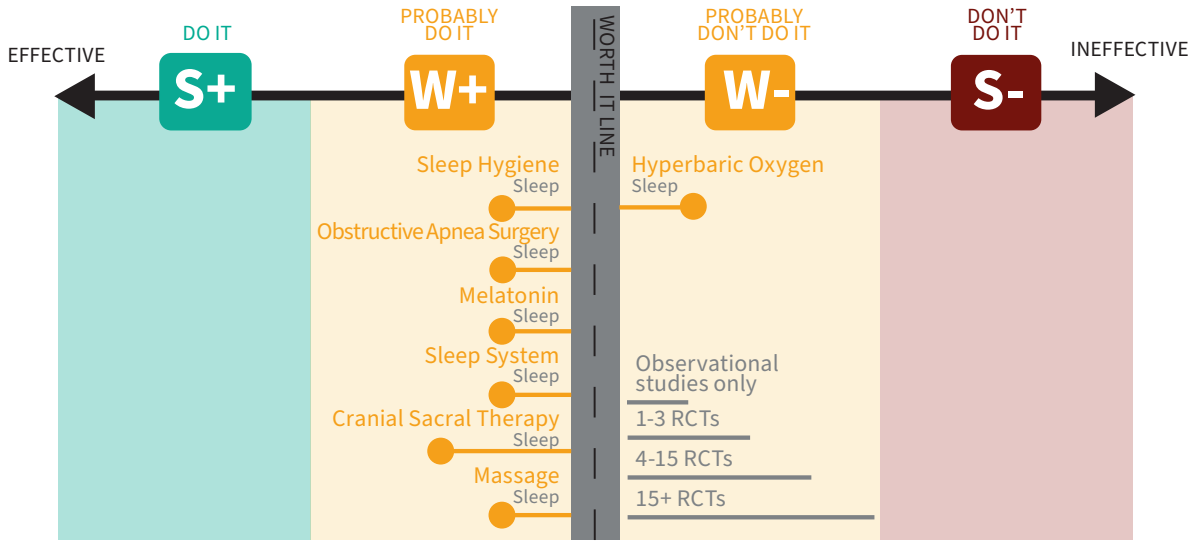
Sleep Disorders

1 in 5 children with cerebral palsy has a sleep disorder.[1]

Sleep should be systematically addressed in all children with cerebral palsy. High likelihood of chronic sleep disorders can be multi-factorial, including epilepsy, abnormal postures, more severe physical impairment and severe visual impairment, apnoea, pain, clinical sleep disorders, environmental factors and behavioural problems.

Optimal management involves ensuring pain is assessed and treated thoroughly, and anticonvulsants and tone management medications may also be used.

Examples include sleep hygiene, parental education, spasticity management, melatonin (2.5-10mg), and gabapentin (5mg/kg).[2,3]



Systematic overview of best-available evidence (2012-2019) sleep interventions in cerebral palsy. .

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

1. Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy. Pediatrics. 2012;130(5):e1285-312.
2. Novak I, McIntyre S, Morgan C, et al. State of the evidence: systematic review of interventions for children with cerebral palsy. Dev Med Child Neurol. 2013;55:885-910.
3. Novak I, Morgan C, Adde L, Blackman J, Boyd RN, Brunstrom-Hernandez J, Cioni G, Damiano D, Darrah J, Eliasson AC, de Vries LS, Einspieler C, Fahey M, Fehlings D, Ferriero DM, Fetters L, Fiori S, Forssberg H, Gordon AM, Greaves S, Guzzetta A, Hadders-Algra M, Harbourne R, Kakooza-Mwesige A, Karlsson P, Krumlinde-Sundholm L, Latal B, Loughran-Fowlds A, Maitre N, McIntyre S, Noritz G, Pennington L, Romeo DM, Shepherd R, Spittle AJ, Thornton M, Valentine J, Walker K, White R, Badawi N. Early, accurate diagnosis and early intervention in cerebral palsy: Advances in diagnosis and treatment. JAMA Pediatr. 2017.

Parent Outcomes

Family Functioning

The impact of cerebral palsy on the whole family is complex and challenging. Early interventional supports for parents are required.

Effective parenting interventions include stepping stones triple P[1] and acceptance and commitment therapy (ACT).[2]

PARENT WELL-BEING

1 in 4 parents of children with cerebral palsy have very high stress.[3]

Mothers of a child with a disability report high rates of distress, anxiety, depressions and suicidality.[4] Mothers report the perceived need for professional mental health support, and support is most wanted around the time of diagnosis.[3,4]

Link: Parent Wellbeing Resource
Books: (1) Uncommon Fathers: Reflections on Raising a child with a disability. Donald J Meyer. (2) Married with Special needs children: A couples guide to keeping connected, Laura Marshak and Fran Prezant.

SIBLING SUPPORT

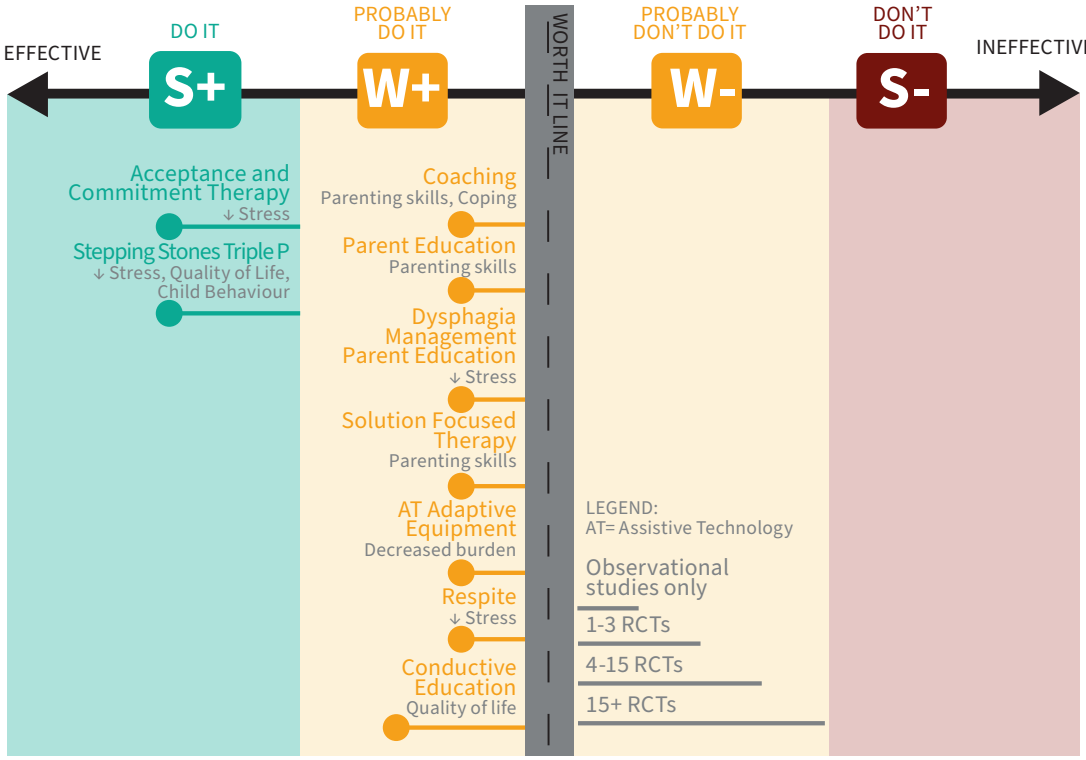
Siblings may require individual support.

Links and Books:

Raising Children Network Siblings

CP NOW Toolkit. Impact on CP Diagnosis on Family and Siblings

Views from our Shoes: Growing up with a Brother or Sister with Special Needs, Donald J. Meyer



Systematic overview of best-available evidence (2012-2019) parenting interventions in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

1. Roberts C, Mazzucchelli T, Studman L, Sanders MR. J Clin Child Adolesc Psychol. 2006 Jun; 35(2):180-93.
2. Whittingham K, Sanders MR, McKinlay L, Boyd RN. Parenting intervention combined with Acceptance and Commitment Therapy: a trial with families of children with cerebral palsy. J Pediatr Psychol. 2016;41(5):531-542.
3. Parkes J, Caravale B, Marcelli M, Franco F, Colver A. Parenting stress and children with cerebral palsy: A european cross-sectional survey. Dev Med Child Neurol. 2011; 53:9: 815-821.
4. Gilson K,M, Davis E, Johnson S, GainsJ, Reddihough D, Williams K. Mental health care needs and preferences for mothers of children with a disability. Child CareHealth Dev. 2018;1–8.https://doi.org/10.1111/cch.12556

Self-Care

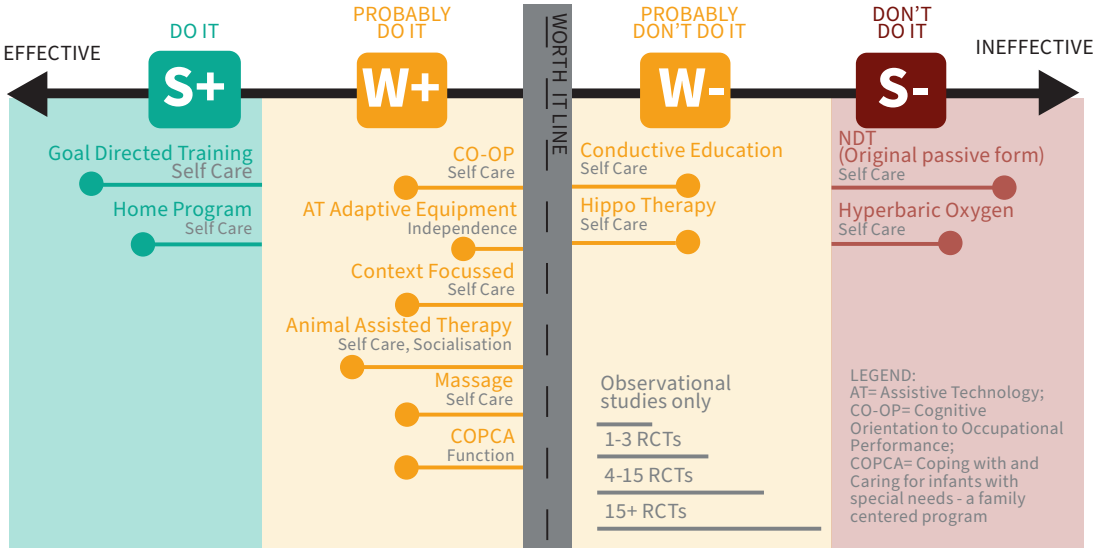
Participation

Children with cerebral palsy are able to be actively involved in a wide range of leisure activities, and experience a high level of enjoyment.[1]

Data suggests, however, that they participate less in physically active leisure compared with peers, and that participation reduces over time.[2]

Parents of children with cerebral palsy rank participation as their second most important research priority.[3]

Interventions are available that target participation, and address barriers that prohibit participation and their effects.[4,5]



Systematic overview of best-available evidence (2012-2019) self-care interventions in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

1. Majnemer A, Shevell M, Law M, Birnbaum R, Chilingaryan G, Rosenbaum P, Poulin C. Participation and enjoyment of leisure activities in school-aged children with cerebral palsy. Dev Med Child Neurol 2008; 50: 751–758. doi:10.1111/j.1469-8749.2008.03068.
2. Majnemer A, Shikako-Thomas K, Schmitz N, Shevell M, Lach L. Stability of leisure participation from school-age to adolescence in individuals with cerebral palsy. Res Dev Disabil. 2015;47:73-9.
3. McIntyre S, Novak I, Cusick A. Consensus research priorities for cerebral palsy: A delphi survey of consumers, researchers, and clinicians. Dev Med Child Neurol. 2010;52 :3:270-5.
4. Novak I, Morgan C, Fahey M, et al. State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep. 2020;20(2):3. Published 2020 Feb 21.
5. Reedman SE, Boyd RN, Trost SG, Elliott C, Sakzewski L. Efficacy of Participation-Focused Therapy on Performance of Physical Activity Participation Goals and Habitual Physical Activity in Children With Cerebral Palsy: A Randomized Controlled Trial. Arch Phys Med Rehabil. 2019;100(4):676-686.

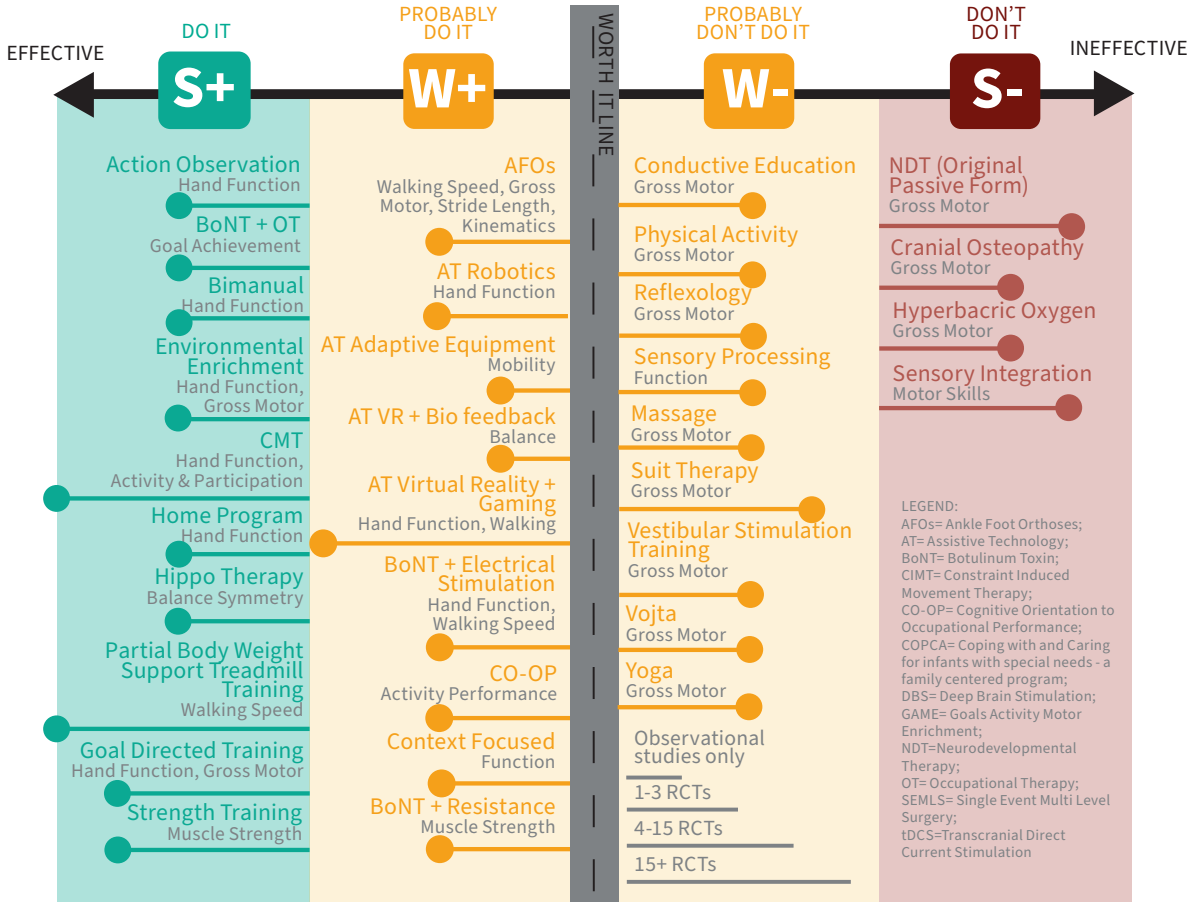
Effective motor interventions for children and adolescents with cerebral palsy

Effective motor interventions for children and adolescents with cerebral palsy

Effective motor interventions involve practice of real-life tasks and activities, using self-generated active movements, at a high intensity, where the practice directly targets the achievement of a goal set by the child (or a parent proxy if necessary).

Clinical trial data supports training-based interventions, including action observation training, bimanual training, constraint induced movement therapy, functional chewing training, goal-directed training, home programs using goal-directed training, mobility training, treadmill training, partial body weight support treadmill training, and occupational therapy post Botulinum toxin A.

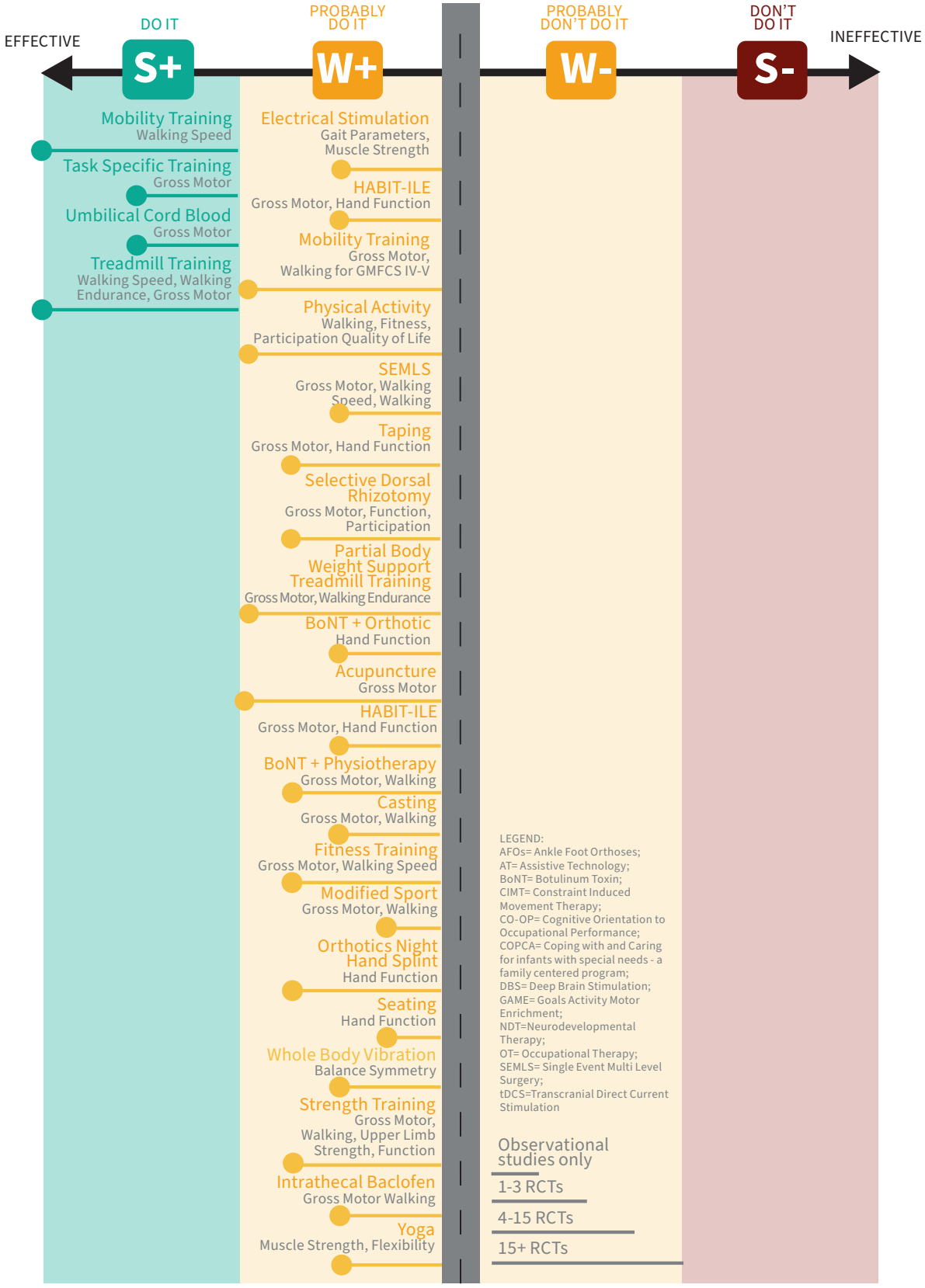
Weak positive evidence supports adjunctive interventions when combined with task-specific motor training: electrical stimulation, hydrotherapy, taping, transcranial direct current stimulation, and virtual reality serious gaming. Complementary and alternative medicine trials have demonstrated weak positive evidence efficacy with acupuncture, and animal-assisted therapy.



1. Morgan C, Darrah J, Gordon AM, Harbourne R, Spittle A, Johnson R, et al. Effectiveness of motor interventions in infants with cerebral palsy: a systematic review. Dev Med Child Neurol.2016;58(9):900–9.
2. Kolb B, Muhammad A. Harnessing the power of neuroplasticity for intervention.Frontiers in Human Neuroscience. 2014;8(6):377.
3. Shepherd RB, ed. Cerebral Palsy in Infancy:Targeted Activity to Optimize Early Growth and Development. Oxford, England: Elsevier Health Sciences;2014.
4. Morgan C, Novak I, Dale RC, Guzzetta A, Badawi N. Single blind randomised controlled trial of GAME (Goals Activity Motor Enrichment) in infants at high risk of cerebral palsy. Research in Developmental Disabilities. 2016;55:256-67.

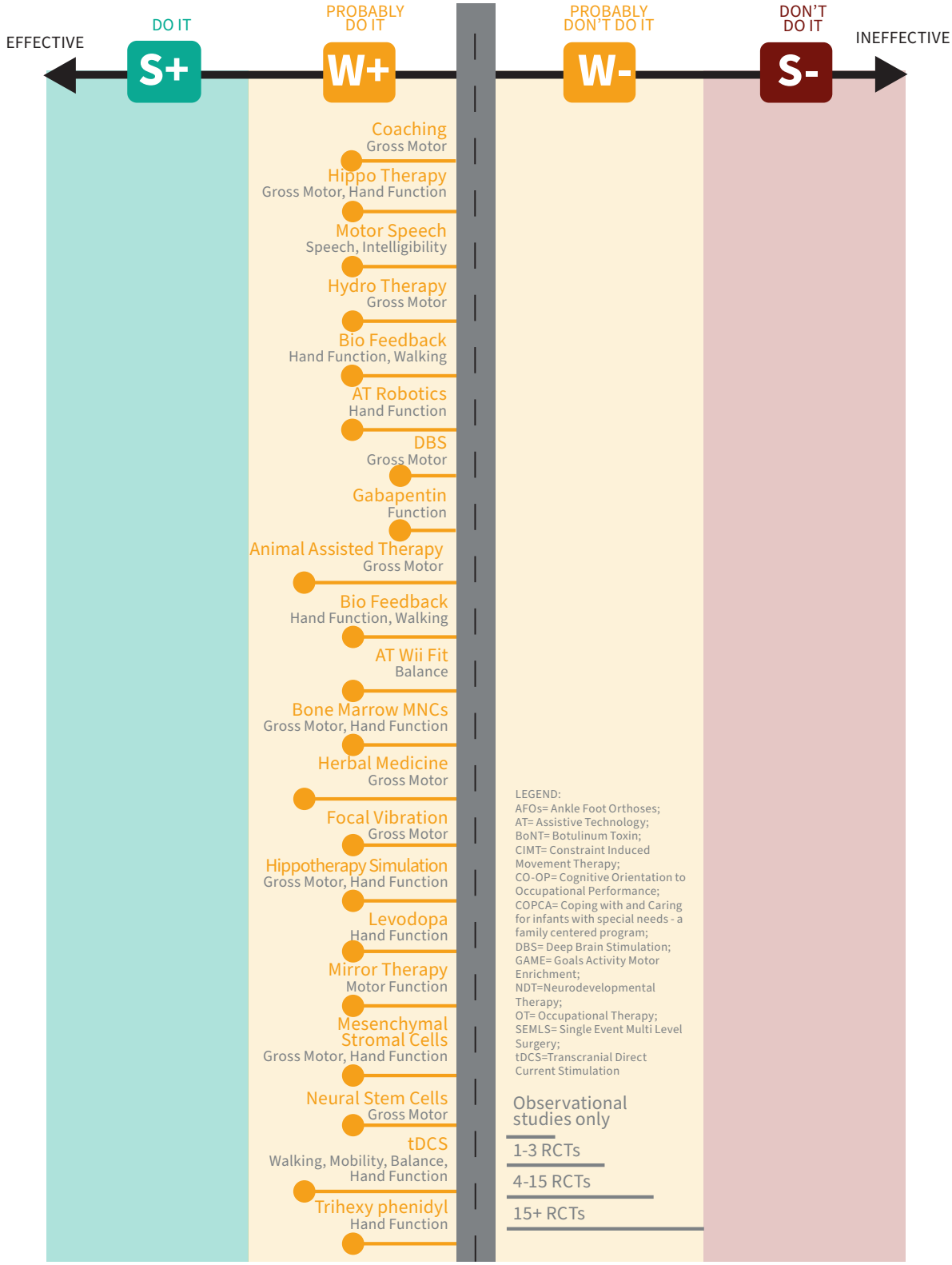
Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Effective motor interventions for children and adolescents with cerebral palsy



Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Effective motor interventions for children and adolescents with cerebral palsy



Systematic overview of the best-available evidence (2012–2019) motor interventions in managing motor in cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network
CENTRE FOR RESEARCH EXCELLENCE

Prevention of Cerebral Palsy

Prevention of Cerebral Palsy and Emergent Prophylactic, Reparative and Restorative Brain Interventions

- Significant improvement in cerebral palsy prevention has been seen over the last decade.[1]
- Antenatal magnesium sulfate before delivery of an infant less than 30 weeks' gestation prevents 30% of cerebral palsy.[2]
- Antenatal corticosteroids decrease intracranial haemorrhage and act as an effective neuroprotectant.[2]
- Prophylactic caffeine (methylxanthines) prior to extubation in mechanically ventilated premature infants effectively prevents cerebral palsy.[3]
- In term babies with neonatal encephalopathy or asphyxia, therapeutic hypothermia commenced within 6-h of delivery is neuroprotective and prevents 15% of cerebral palsy associated with intrapartum hypoxia.[3]
- A genetic contribution is likely in one-third of all children with cerebral palsy, especially in those without traditional risk factors such as prematurity and hypoxia.[4]
- New prevention and treatment is predicted as the understanding of neurobiology and genomics expands.[4]
- Regenerative medical treatments are being explored.[1]
- Erythropoietin is emerging as a promising intervention for preterm populations with trials underway in hypoxic ischemic encephalopathy populations.[3]
- Umbilical cord blood as a cell therapy, coupled with rehabilitation, is slightly more effective than rehabilitation alone for improving motor skills in children with cerebral palsy. [1,5,6]

1. Novak I, Morgan C, Fahey M, et al. State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep. 2020;20(2):3. Published 2020 Feb 21.
2. Shepherd E, Salam RA, Middleton P, Makrides M, Mcintyre S, Badawi N, et al. Antenatal and intrapartum interventions for preventing cerebral palsy: an overview of Cochrane systematic reviews. J Paediatr Child Health. 2017;53(Supplement 2):90.
3. Shepherd E, Salam RA, Middleton P, Han S, Makrides M, McIntyre S, et al. Neonatal interventions for preventing cerebral palsy: an overview of Cochrane systematic reviews. Cochrane Database Syst Rev. 2018;6:Cd012409.
4. Fahey MC, Maclennan AH, Kretzschmar D, Gecz J, Kruer MC. The genetic basis of cerebral palsy. Dev Med Child Neurol. 2017;59(5):462–469.
5. Novak I, Walker K, Hunt RW, Wallace EM, Fahey M, Badawi N. Concise review: stem cell interventions for people with cerebral palsy: systematic review with meta-analysis. Stem Cells Transl Med. 2016;5(8):1014–25.
6. Kulak-Bejda A, Kulak P, Bejda G, Krajewska-Kulak E, Kulak W. Stem cells therapy in cerebral palsy: a systematic review. Brain Dev. 2016;38(8):699–7

Early intervention

Clinicians should understand the importance of prompt referral to diagnostic-specific early intervention to optimise infant motor and cognitive plasticity, prevent secondary complications, and to optimise caregiver well-being[1].

Neuroscientific evidence indicates that brain development and refinement of the motor system continues postnatally, driven by motor cortex activity. Early active movement and intervention is essential because infants who do not actively use their motor cortex risk losing cortical connections and dedicated function.[2,3] Practice of real-life tasks and activities, using self-generated active movements, at a high intensity, where the practice directly targets the achievement of a goal set by the child (or a parent proxy if necessary) is recommended. [4]

There is increasing evidence that the infant's motor behavior, through discovery and interaction with the environment, controls and generates the growth and development of muscle, ligament, and bone, as well as driving ongoing development of the neuromotor system. Evidence is emerging that commencement of cerebral palsy-specific early intervention before 6 months (corrected age) and the completion of the corticospinal tract, improves children's motor and cognitive outcomes.[5,6]

Early intervention improves child outcomes

Commencement of cerebral palsy-specific early intervention before 6 months (corrected age) and the completion of the corticospinal tract development improves motor and cognitive outcomes.

Early intervention is about taking action as soon as possible to tackle problems for children and families before they become more difficult to reverse.

1. Novak et al 2017. Early Accurate Diagnosis and Early Intervention in Cerebral Palsy. *JAMA Pediatr.* 2017; 171(9):897-907.
2. Eyre J. Corticospinal tract development and activity dependent plasticity. In: Shepherd R, ed. Cerebral Palsy in Infancy. Oxford, England: Elsevier; 2014:53-66.
3. Martin JH, Chakrabarty S, Friel KM. Harnessing activity-dependent plasticity to repair the damaged corticospinal tract in an animal model of cerebralpalsy. *Dev Med Child Neurol.* 2011;53(suppl 4):9-13.
4. Novak, I., Morgan, C., Fahey, M., Finch-Edmondson, M., Galea, C., Hines, A., ... & Shore, B. (2020). State of the evidence traffic lights 2019: systematic review of interventions for preventing and treating children with cerebral palsy. Current neurology and neuroscience reports, 20(2), 1-21
5. Morgan C, Novak I, Dale RC, Guzzetta A,Badawi N. Single blind randomised controlled trial of GAME (Goals–Activity–Motor Enrichment) in infants at high risk of cerebral palsy. *Res Dev Disabil.* 2016;55:256-267.
6. Eliasson AC, Holmefur M. The influence of early modified constraint-induced movement therapy training on the longitudinal development of hand function in children with unilateral cerebral palsy. *Dev Med Child Neurol.* 2015;57(1):89-94

Effective Early Intervention Recommendations

Effective early interventions for cerebral palsy

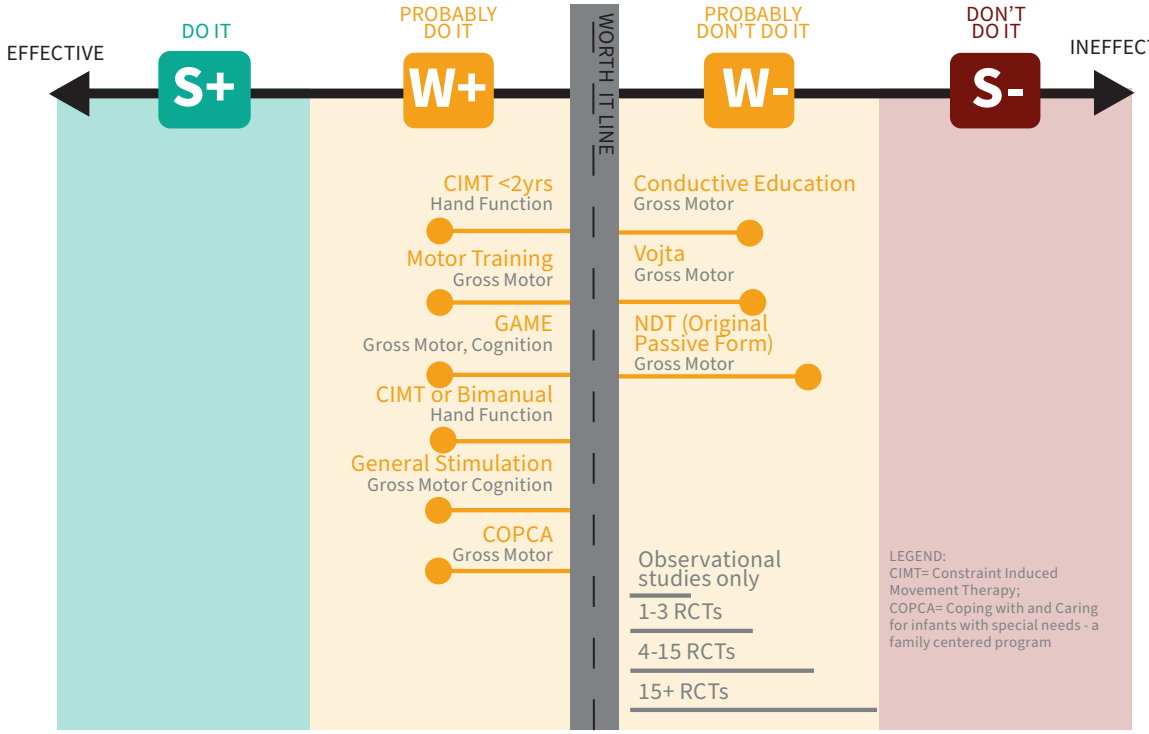
Systematic review evidence has shown that historical treatments for cerebral palsy using passive movements (such as positioning, facilitated normal movement patterns and stretching) are ineffective for improving motor skills.[1]

Effective early motor interventions specific to cerebral palsy should harness neuroplasticity[2], encourage infants to learn that their purposeful actions have meaningful consequence[3], and involve key ingredients of:

(1) active child-initiated movements;
(2) task specificity;
(3) high intensity;
(4) parent goal setting;
(5) repetition;
(6) variability and
(7) enrichment of the home environment.

Novel promising motor learning interventions are emerging, demonstrating weak positive evidence, such as baby-CIMT (constraint-induced movement therapy), baby-bimanual, GAME (a combination of motor training, environmental enrichment) and small steps. They have reported positive gains in movement skills with larger replication trials currently underway (ACTRN12617000006347).

Trials into early interventions targeting other developmental domains such as cognition, feeding and communication will emerge in the near future.[5]



Systematic overview of best-available evidence (2012-2019) in early intervention for cerebral palsy.

Adapted with permission from Novak, I., Morgan, C., Fahey, M., et al. (2020). State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy. Curr Neurol Neurosci Rep, 20(2), 3. doi:10.1007/s11910-020-1022-z

1. Morgan C, Darrah J, Gordon AM, Harbourne R, Spittle A, Johnson R, et al. Effectiveness of motor interventions in infants with cerebral palsy: a systematic review. Dev Med Child Neurol.2016;58(9):900–9.W
2. Kolb B, Muhammad A. Harnessing the power of neuroplasticity for intervention.Frontiers in Human Neuroscience. 2014;8(6):377.
3. Shepherd RB, ed. Cerebral Palsy in Infancy:Targeted Activity to Optimize Early Growth and Development. Oxford, England: Elsevier Health Sciences;2014.
4. Morgan C, Novak I, Dale RC, Guzzetta A, Badawi N. Single blind randomised controlled trial of GAME (Goals Activity Motor Enrichment) in infants at high risk of cerebral palsy. Research in Developmental Disabilities. 2016;55:256-67.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

How to Refer an Infant for Early Intervention

How to refer an infant with a diagnosis of 'high-risk' of cerebral palsy for early intervention services.

The diagnosis of 'high-risk' of cerebral palsy has been acknowledged in National Disability Insurance Scheme (NDIS) access request practice guidelines.[1]

Medical reports submitted to the NDIS with a 'high-risk' of cerebral palsy diagnosis should include early diagnosis of cerebral palsy guideline recommendations[2] of:

- Prechtl's General Movements Assessment results
- Magnetic Resonance Imaging results
- Hammersmith Infant Neurological Examination results.

1. Practice Guide – ECEI. Supporting an Access Request. (2020). National Disability Insurance Scheme. Accessed June 2020.
2. Novak, I., Morgan, C., Adde, et al.(2017). Early, Accurate Diagnosis and Early Intervention in Cerebral Palsy: Advances in Diagnosis and Treatment. JAMA Pediatr, 171(9), 897-907. doi:10.1001/jamapediatrics.2017.1689

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

Early diagnosis – cerebral palsy

Early detection and diagnosis recommendations from best available evidence

STRONG RECOMMENDATION

CONDITIONAL RECOMMENDATION

1 The clinical diagnosis of cerebral palsy can and should be made as early as possible. When the clinical diagnosis is suspected but cannot be made with certainty, the interim clinical diagnosis of 'high-risk' of cerebral palsy should be given.

MOTOR DYSFUNCTION GMs +/- HINE + ABNORMAL NEURO IMAGING MRI +/- HINE + CLINICAL HISTORY

Based on MODERATE QUALITY evidence for infant and parent outcomes.

2 Early standardised assessments and investigations for early detection of 'high-risk' of cerebral palsy should always be conducted in 'high-risk' of cerebral palsy populations, i.e. infants born pre-term, infants with neonatal encephalopathy, infants with birth defects or infants admitted to Neonatal Intensive Care Unit (NICU).

Based on HIGH QUALITY evidence of test psychometrics.

Early detection of cerebral palsy before 5 months corrected age

Option A: The most accurate method for early detection of cerebral palsy in infants with newborn-detectable risks and younger than 5 months corrected age (CA) is to use a combination of a standardised motor assessment, neuroimaging and history taking about risk factors.

MOTOR DYSFUNCTION + ABNORMAL NEURO IMAGING

3 **TEST:** General Movements Assessment (GMs), to identify motor dysfunction [95–98% predictive of cerebral palsy]; combined with neuroimaging.

STANDARDISED MOTOR

TEST: MRI (before sedation is required for neuroimaging) to detect abnormal neuroanatomy in the motor area/s of the brain [80–90% predictive of cerebral palsy].
Note: Normal neuroimaging does not automatically preclude the diagnosis of risk of cerebral palsy.

ABNORMAL NEURO IMAGING

Based on HIGH QUALITY evidence of test psychometrics in newborn-detectable risk populations.

Option B: In contexts where the General Movements Assessment is not available or MRI is not safe or affordable (e.g. in countries of low to middle income), early detection of cerebral palsy in infants with newborn-detectable risks and younger than 5 months (CA) is still possible and should be carried out to enable access to early intervention.

STANDARDISED NEURO EXAM + STANDARDISED MOTOR

4 **TEST:** Hammersmith Infant Neurological Examination (HINE) [HINE<57 at 3 months is 96% predictive of cerebral palsy].

STANDARDISED NEURO EXAM

Based on MODERATE QUALITY evidence of test psychometrics in newborn-detectable risk populations.

TEST: Test of Infant Motor Performance (TIMP).

STANDARDISED MOTOR

Based on LOW QUALITY evidence of test psychometrics in newborn-detectable risk populations.

Early detection of cerebral palsy after 5 months corrected age

Accurate early detection of 'high-risk' of cerebral palsy in those with infant-detectable risks and age 5-24 months can and should still occur as soon as possible, but different diagnostic tools are required.

5 Any infant with:
(a) inability to sit independently by 9 months; or

(b) hand function asymmetry: strong early preference for one side; or

(c) inability to take weight with feet flat on the floor should receive standardised investigations for cerebral palsy.

Based on HIGH QUALITY evidence of motor norms.

Option A: The most accurate method for early detection of cerebral palsy with infant-detectable risks older than 5 months (corrected age) but younger than 2 years old is to use a combination of a standardised neurological assessment, neuroimaging, and a standardised motor assessment with a history taking about risk factors.

STANDARDISED NEURO EXAM + STANDARDISED MOTOR + ABNORMAL NEURO IMAGING

6 **TEST:** HINE (90% predictive of cerebral palsy). HINE scores lower than 73 (at 6, 9 or 12 months) should be considered at 'high-risk' of cerebral palsy. HINE scores lower than 40 (at 6, 9 or 12 months) almost always indicate cerebral palsy; combined with neuroimaging and standardised motor assessments.

TEST: MRI to detect abnormal neuroanatomy in the motor area/s of the brain (sedation required >6 weeks up to 2 years of age).

STANDARDISED NEURO EXAM

ABNORMAL NEURO IMAGING

Specific tests of movement and development called the Developmental Assessment of Young Children (DAYC) and the Alberta Infant Motor Scale (AIMS) are also recommended and can be performed and scored by experienced clinicians.

MOTOR DYSFUNCTION

Based on MODERATE QUALITY evidence of test psychometrics in newborn-detectable risk populations.

LOW-MODERATE QUALITY evidence of test psychometrics in newborn-detectable risk populations.

180139 April 2018

Early diagnosis – cerebral palsy

Early detection of cerebral palsy after 5 months corrected age (continued)

Option B: In contexts where MRI is not safe or affordable, early detection of cerebral palsy is still possible with infant-detectable risks between 5-24 months (corrected age) and should be carried out to enable access to early intervention.

STANDARDISED NEURO EXAM + STANDARDISED MOTOR

7

TEST: HINE [90% predictive of cerebral palsy at 2–24 months of age] HINE scores at 6, 9 or 12 months: <73 indicates 'high-risk' of cerebral palsy. A score of <40 indicates abnormal outcome, usually cerebral palsy.

STANDARDISED NEURO EXAM

Based on MODERATE QUALITY evidence of test psychometrics.

TEST: Developmental Assessment of Young Children (DAYC) to quantify motor delay [83% predictive of cerebral palsy].

STANDARDISED MOTOR

TEST: Motor Assessment of Infants (MAI) to quantify motor delay [73% predictive of cerebral palsy].

STANDARDISED MOTOR

LOW-MODERATE QUALITY evidence

Early detection of motor severity of cerebral palsy

Prognosis of long-term motor severity is most accurate in children over 2 years using the Gross Motor Function Classification System (GMFCS).

8

In infants under 2 years old, prognosis of motor severity predictions should be made cautiously and always involve the use of standardised tools, because incomplete development of voluntary motor skills or abnormal tone might confound clinical observations. Motor severity of cerebral palsy under 2 years of age is most accurately predicted using the Standardised Neurological Assessment.

STANDARDISED NEURO EXAM + ABNORMAL NEURO IMAGING

Based on LOW QUALITY evidence.

TEST: HINE. Cut-off scores predict the probable severity.

HINE 40–60

HINE <40

STANDARDISED NEURO EXAM

TEST: MRI Normal imaging does not preclude cerebral palsy, and abnormal imaging does not automatically lead to cerebral palsy.

ABNORMAL NEURO IMAGING

Based on MODERATE QUALITY evidence in newborn-detectable risk populations.

Early detection of motor sub-type and topography of cerebral palsy

9

Early detection of motor sub-type and topography can be difficult in infants under 2 years old, but wherever possible it is very important to identify unilateral versus bilateral cerebral palsy early, as the early interventions (e.g. constraint induced movement therapy) and long-term musculoskeletal outcomes and surveillance needs differ (e.g. hip surveillance).

Based on LOW QUALITY evidence.

Early intervention

10

Clinical diagnosis of cerebral palsy or the interim diagnosis of 'high-risk' of cerebral palsy should always be followed by a referral to cerebral palsy-specific early intervention (e.g. constraint induced movement therapy and hip surveillance). Parent concern is a valid reason to trigger formal diagnostic investigations and referral to early intervention.

Based on HIGH QUALITY evidence.

Early detection of associated impairments

11

Clinical diagnosis of cerebral palsy or interim diagnosis of 'high-risk' of cerebral palsy should always include standard medical investigations for associated impairments and functional limitations (e.g. vision impairment, hearing impairment and epilepsy).

Based on HIGH QUALITY evidence.

Communicating the diagnosis to parents compassionately

12

Parents experience grief and loss at the time of diagnosis or 'high-risk' notification; therefore communication with a family should be a series of well-planned and compassionate conversations. Communication should be empathetic and involve the family, face-to-face with both parents or caregivers present (where appropriate), private, honest and jargon-free. This should be followed by written information, identification of strengths, invitation to ask questions, discussion of feelings, recommendations to use parent-to-parent support and arrangement of early intervention.

Based on HIGH QUALITY qualitative parent interviews.

Adapted with permission from: Novak et al 2017. Early, Accurate Diagnosis and Early Intervention in Cerebral Palsy. *JAMA Pediatr.* 2017;171(9):897-907. doi:10. 1001/jamapediatrics.2017.1689 Available from: http://jamanetwork.com/journals/jamapediatrics/article-abstract/2636588

Cerebral Palsy ALLIANCE

CENTRE OF RESEARCH EXCELLENCE Cerebral Palsy

Algorithm for early diagnosis of cerebral palsy or high-risk cerebral palsy

A Best available evidence pathway

B Next best available evidence pathway when some pathway A tools are not available

Newborn-detectable risks

Infant-detectable risks

Pre-term

Encephalopathy

History or neurological risk factors (e.g. birth defect, IUGR)

Parent identified concern

Unable to sit at 9 months or hand asymmetry

Risks or concerns warrant an investigation for cerebral palsy

Conduct a medical history and clinical examination with or without investigations for etiology and differential diagnosis (as indicated)

<5 months CA

>5 months CA

STANDARDISED NEURO EXAM

ABNORMAL NEURO IMAGING

STANDARDISED MOTOR

	A	B
<5 months CA	3 MRI + 3 GMs	4 HINE + 4 TIMP
>5 months CA	6 HINE + 6 MRI + 6 DAYC + 6 AIMS + 6 NSMDA	7 HINE + 7 DAYC + 7 MAI

Combined assessment data indicates:

1 High risk of CP

1 Definitely CP

1 Unclear

1 Definitely *not* CP

8 Determine preliminary severity of cerebral palsy

HINE	MRI WMI	HINE	MRI GMI
STANDARDISED NEURO EXAM	ABNORMAL NEURO IMAGING	STANDARDISED NEURO EXAM	ABNORMAL NEURO IMAGING
HINE 40–60		HINE <40	

As indicated, continuous testing for differential diagnosis and relevant associated impairments

9 Determine preliminary topography

11 Assess for associated impairments

12 Communicate findings to parents compassionately

10 Arrange early intervention and parent support

Monitor → Confirm diagnosis

Acronyms

CP	Cerebral palsy
IUGR	Intrauterine growth restriction
CA	Corrected age
HINE	Hammersmith Infant Neurological Examination
MRI	Magnetic Resonance Imaging
WMI	White matter injury
GMI	Grey matter injury
GMs	Prechtl's General Movements Assessment
TIMP	Test of infant motor performance
DAYC	Developmental Assessment of Young Children
AIMS	Alberta Infant Motor Scale
NSMDA	Neuro Sensory Motor Developmental Assessment
MAI	Motor assessment of infants

Adapted with permission from: Novak et al 2017. Early, Accurate Diagnosis and Early Intervention in Cerebral Palsy. *JAMA Pediatr.* 2017;171(9): 897-907. doi:10. 1001/jamapediatrics.2017.1689 Available from: http://jamanetwork.com/journals/jamapediatrics/article-abstract/2636588

Cerebral Palsy ALLIANCE

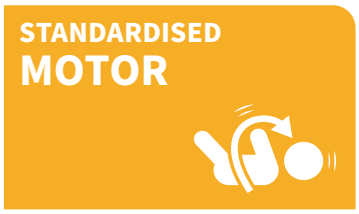
CENTRE OF RESEARCH EXCELLENCE Cerebral Palsy

1801139 April 2018

EARLY DIAGNOSIS – CEREBRAL PALSY

Prechtl's General Movements Assessment

Prechtl's General Movements Assessment is simply a video of your baby lying on their back whilst they are awake, calm and alert.



- The assessment can be completed from birth up to 20 weeks of age (corrected age).
- It is non-invasive and non-disruptive.
- The video can be taken by parents or clinicians with appropriate consent.
- The video may be recorded by medical professionals whilst your baby is an inpatient, an outpatient, or by you in your home via the BabyMoves app.

What's involved?

It is a standardised test of movement that can be scored, based on observation of your baby's movements, by certified assessors trained by the General Movements Trust. The length of the video depends on how old your baby is.

AGE	PHASE	VIDEO DURATION
Up to 6-9 weeks, corrected age	'Writhing' phase	Up to 15 minutes
From about 9 weeks corrected age through to about 20 weeks corrected age	'Fidgety' phase	3-5 minutes

Prechtl's General Movements (GMs) are predictive of how the young central nervous system is developing. They can identify neurological issues predictive of cerebral palsy and other developmental disabilities. The results of your GMs videos will be discussed with you thoroughly by your medical team.

If your baby's GMs indicate an abnormal result, your multidisciplinary team may guide you through further investigations, assessments and early intervention supports.

Specific early intervention and supports for babies and their families who are identified 'at risk of cerebral palsy' is proving to demonstrate better outcomes for children and families.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

EARLY DIAGNOSIS – CEREBRAL PALSY

Prechtl's General Movements Assessment

STANDARDISED
MOTOR

What are General Movements?

General Movements (GMs) are distinct spontaneous movement patterns that are evident in babies before birth and after birth up to 20 weeks of age (corrected age).

They are seen spontaneously when the baby is awake, calm and alert and not externally stimulated (such as a parent playing or talking with them).

You may be familiar with other spontaneous motor patterns seen in young babies such as startles, twitches, yawning and breathing movements.

GMs involve the whole body and are variable, complex, fluent and elegant.

GMs mature and change in a specific order:

AGE	PHASE
Up to 6-9 weeks, corrected age	'Writhing'
From about 9 weeks through to about 20 weeks, corrected age	'Fidgety'

What are the benefits of GM assessment?

- General assessments are a cost-effective way of assessing a baby's young nervous system.
- The standardised Prechtl's GMs assessment provides an assessment of these General Movement patterns of young infants.
- If GMs are identified as 'absent' or 'abnormal' it may indicate risk of neurological conditions, in particular cerebral palsy.
- GMs videoed around 3 months of age (12–16 weeks corrected age) provide the most predictive information about the likelihood risk of cerebral palsy.
- Identifying infants at 'high risk of cerebral palsy' early using the GMs assessments means that parent supports and specific treatments can start very early with potentially better outcomes for infants and families.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE

EARLY DIAGNOSIS – CEREBRAL PALSY

Prechtl's General Movements Assessment

How is the assessment done?

STANDARDISED
MOTOR

Consent to the video must be given prior to videoing, and clinicians will discuss the assessment with you if the video is being taken by a clinician.

The assessment can be done by observing the spontaneous movements of your baby, lying on their back on a mat on the floor in quiet surrounds while they are awake, calm and alert.

Your baby should not have any toys or pacifiers and be lightly dressed (no socks).

Try not to play or talk with your baby while the video is taken as this can change the movements that are seen.

Comfort your child as required, however babies that are upset or crying change the movements and make the video difficult to score.

If your baby has a strong head preference, try to reposition their head towards the middle during the video assessment.

The clinician taking the video may position your baby nested in a pillow if your baby has severe reflux or is more settled in a nested pillow position.

Video quietly over the top of your baby, with your baby orientated vertically and make sure that you can see all of your baby including their hands and feet.

The clinician taking the video may set up a tripod and will need to document your baby's date of birth, date of video and corrected age.

Regardless of who takes your baby's video, the General Movements Assessment will be scored by certified assessors trained by the General Movements Trust.

There are a growing number of certified assessors throughout Australia qualified to score the GMs assessment.

Results of your baby's GMs assessment will be communicated with you by your multidisciplinary team.

Your baby's GMs video will be stored securely following all state and federal policies and standards.

Cerebral Palsy ALLIANCE
THE UNIVERSITY OF SYDNEY
AusCP-CTN
Australasian Cerebral Palsy Clinical Trials Network
CENTRE FOR RESEARCH EXCELLENCE

EARLY DIAGNOSIS – CEREBRAL PALSY

Prechtl's General Movements Assessment

Should my child have the General Movements Assessment?

STANDARDISED MOTOR

The General Movements Assessment may give extra information of how your baby's neurological system is developing if your baby is under 20 weeks age (corrected age) and:

- there were medical concerns at birth (spent time in a Neonatal Intensive Care Unit, prematurity, lack of oxygen, stroke or congenital heart disease); or
- your baby is not developing typically like babies of similar corrected age.

The General Movements Assessment is not currently used as a screening tool for all healthy babies without any developmental concerns.

Please speak to your doctor or multidisciplinary team if you have concerns about your baby or you would like to know more about the General Movements Assessment.

Who can do this assessment?

In some hospitals and community centres across Australia, videos are taken and scored by General Movements Trust trained medical professionals.

Videos can also be taken by parents including via the BabyMoves app.

These videos are then scored by professionals who are trained by the General Movements Trust.

Please follow the instructions on how the video assessment is done, and read the BabyMoves handout if taking a video at home.

If you have any questions, please don't hesitate to discuss these with your doctor or therapist.

Cerebral Palsy ALLIANCE

THE UNIVERSITY OF SYDNEY

AusCP-CTN Australasian Cerebral Palsy Clinical Trials Network CENTRE FOR RESEARCH EXCELLENCE