CEREBRAL PALSY

Introduction

The term cerebral palsy refers to a group of disorders of movement or posture that arise from a non-progressive insult to the central nervous system during early development. The insult may occur prior to, during or shortly after birth and, while the insult itself is non-progressive, its physical consequences can evolve over time [1].

Cerebral palsy is the commonest cause of physical disability in early childhood. It occurs in 2–3 per 1,000 live births, although rates may be as high as 40–100 per 1,000 in very low birth weight or preterm babies [2]. Despite this, around 55% of children with cerebral palsy are born at term, while 20% are born at 32–36 weeks and 25% at <32 weeks [2].

As the typical neurological signs of cerebral palsy take time to develop, it is generally accepted that a child should be four years of age before a diagnosis is established, although earlier diagnoses are not precluded in individual cases. Around 80% of children with cerebral palsy have spastic cerebral palsy (characterised by weakness, increased muscle tone, overactive reflexes and a tendency to contractures), while 7% have dyskinetic cerebral palsy (characterised by involuntary movements that disappear during sleep) and 4% have ataxic cerebral palsy (characterised by problems with coordination, gait and rapid movements of the distal extremities) [2,3]. While the term 'cerebral palsy' refers to the motor impairment, additional features such as seizures, intellectual impairment and learning disabilities are common [3].

Children and young people with cerebral palsy require a variety of personal health care and disability support services to achieve their highest possible functioning within their family and community environments. Physical and occupational therapy are beneficial for the management of motor impairments, as proper positioning and handling are necessary to minimise difficulties with posture, trunk control, and feeding. Passive and active exercises to stretch tight tendons may be used to maintain normal alignment of bone, joint and soft tissue and to prevent contractures. Medical and surgical procedures may be necessary to correct contractures that do not respond to physiotherapy, and to re-establish motor balance between opposing muscle groups. In addition, a variety of equipment (e.g. walkers and standing frames, motorised wheel chairs, feeding tubes, computers to augment communication) and other supports (e.g. speech therapy, medications, ophthalmology, tailored educational programmes, respite care) may be required [4].

While plans for a New Zealand Cerebral Palsy Register are under way, at present there is no reliable information on the prevalence of cerebral palsy in New Zealand children and young people. In the absence of this information, the following section reviews hospital admissions for children and young people with any mention of cerebral palsy in any of their first 15 diagnoses, as well as mortality for children and young people with cerebral palsy listed as the main underlying cause of death, or as a contributory cause.

Data Source and Methods

Definition

1. *Hospital admissions for children and young people aged 0–24 years with cerebral palsy listed in any of the first 15 diagnoses*
2. *Mortality in children and young people aged 0–24 years with cerebral palsy listed as the main underlying cause of death or as a contributory cause*

Data Source

1. National Minimum Dataset

Numerator: Hospital admissions for children and young people aged 0–24 years with cerebral palsy (ICD-10-AM G80) listed in any of the first 15 diagnoses.

2. National Mortality Collection

Numerator: Mortality in children and young people aged 0–24 years with cerebral palsy (ICD-10-AM G80) listed as the main underlying cause of death, or as a contributory cause.

Denominator: Statistics New Zealand Estimated Resident Population (projected from 2007).

Notes on Interpretation

Note 1: This analysis focuses on hospital admissions and mortality for children and young people with cerebral palsy listed in any of the first 15 diagnoses, or as the main underlying or a contributory cause of death. The rationale for looking beyond the primary diagnosis was the need to highlight the full spectrum of health issues experienced by those with cerebral palsy, and their consequent requirement for health services.

For example, during 2008–2012, focusing on the primary diagnosis would have identified only 10% of acute and arranged hospitalisations in those with cerebral palsy, with the majority being admitted for other reasons (e.g. epilepsy/convulsions, pneumonitis). Similarly 51% of admissions were from the waiting list, with a large proportion being for injections into ligaments, tendons or soft tissue, or for other orthopaedic procedures. The presence of a small number of admissions which were unrelated to cerebral palsy (e.g. acute upper respiratory infections) however, may slightly overestimate the impact cerebral palsy has on acute service demand.

Note 2: As the majority of those with cerebral palsy are managed in the outpatient or primary care setting, it is likely that this analysis significantly underestimates the number of children and young people with cerebral palsy, particularly those at the milder end of the spectrum. The rationale for the methodology used however, was the absence of other more reliable sources of information on the number of children and young people with cerebral palsy in the community.

Note 3: If no mention of cerebral palsy was made in any of the first 15 diagnoses, these cases were not included, even if the patient had been assigned a cerebral palsy related code on a previous admission.

New Zealand Distribution and Trends

Distribution by Primary Diagnosis and Procedure

Acute and Arranged Admissions by Primary Diagnosis: In New Zealand during 2008–2012, only 10.4% of acute and arranged hospitalisations in children and young people with cerebral palsy listed in their first 15 diagnoses, had cerebral palsy listed as their primary reason for admission. Instead 17.5% of acute and arranged admissions were for epilepsy or convulsions and 22.3% for respiratory infections and diseases. Acute and arranged admissions collectively made up 48.6% of all admissions for children and young people with cerebral palsy during this period (**Table 1**).

Waiting List Admissions by Procedure: During the same period, 51.4% of admissions in children and young people with cerebral palsy were from the waiting list, with injections into ligaments, tendons, or soft tissue accounting for 42.2% of all waiting list admissions. Orthopaedic procedures collectively were the leading reasons for waiting list admissions in children and young people with cerebral palsy, followed by dental procedures (**Table 2**).

Table 1. Acute and Arranged Hospital Admissions for Children and Young People Aged 0–24 Years with Cerebral Palsy by Primary Diagnosis, New Zealand 2008–2012

Primary Diagnosis	Number of Admissions: Total 2008–2012	Number of Admissions: Annual Average	Admission Rate per 100,000 Population	% of Acute and Arranged Admissions	% of All Admissions in those with Cerebral Palsy
Acute and Arranged Admissions by Primary Diagnosis					
Epilepsy, Status Epilepticus, Convulsions	434	86.8	5.68	17.5	8.5
Cerebral Palsy	257	51.4	3.37	10.4	5.0
Influenza and Pneumonia	178	35.6	2.33	7.2	3.5
Unspecified Acute Lower Respiratory Infection	159	31.8	2.08	6.4	3.1
Pneumonitis due to Food and Vomit	128	25.6	1.68	5.2	2.5
Acute Upper Respiratory Infections	89	17.8	1.17	3.6	1.7
Other Respiratory Infections and Diseases	76	15.2	1.00	3.1	1.5
Constipation	56	11.2	0.73	2.3	1.1
Other Diseases Digestive System	152	30.4	1.99	6.1	3.0
Complications of Surgical and Medical Care	107	21.4	1.40	4.3	2.1
Infectious and Parasitic Diseases	103	20.6	1.35	4.1	2.0
Respite Care	32	6.4	0.42	1.3	0.6
Other Factors Influencing Health Service Contact	67	13.4	0.88	2.7	1.3
Other Diagnoses	645	129.0	8.45	26.0	12.6
Total Acute and Arranged Admissions	2,483	496.6	32.52	100.0	48.6
Total Waiting List Admissions	2,625	525.0	34.38		51.4
Total Admissions in those with Cerebral Palsy	5,108	1,021.6	66.89		100.0

Source: Numerator: National Minimum Dataset, Acute and arranged admissions by primary diagnosis for children and young people with cerebral palsy listed in any of the first 15 diagnoses; Denominator: Statistics NZ Estimated Resident Population (projected from 2007)

Table 2. Waiting List Hospital Admissions in Children and Young People Aged 0–24 Years with Cerebral Palsy by Primary Procedure, New Zealand 2008–2012

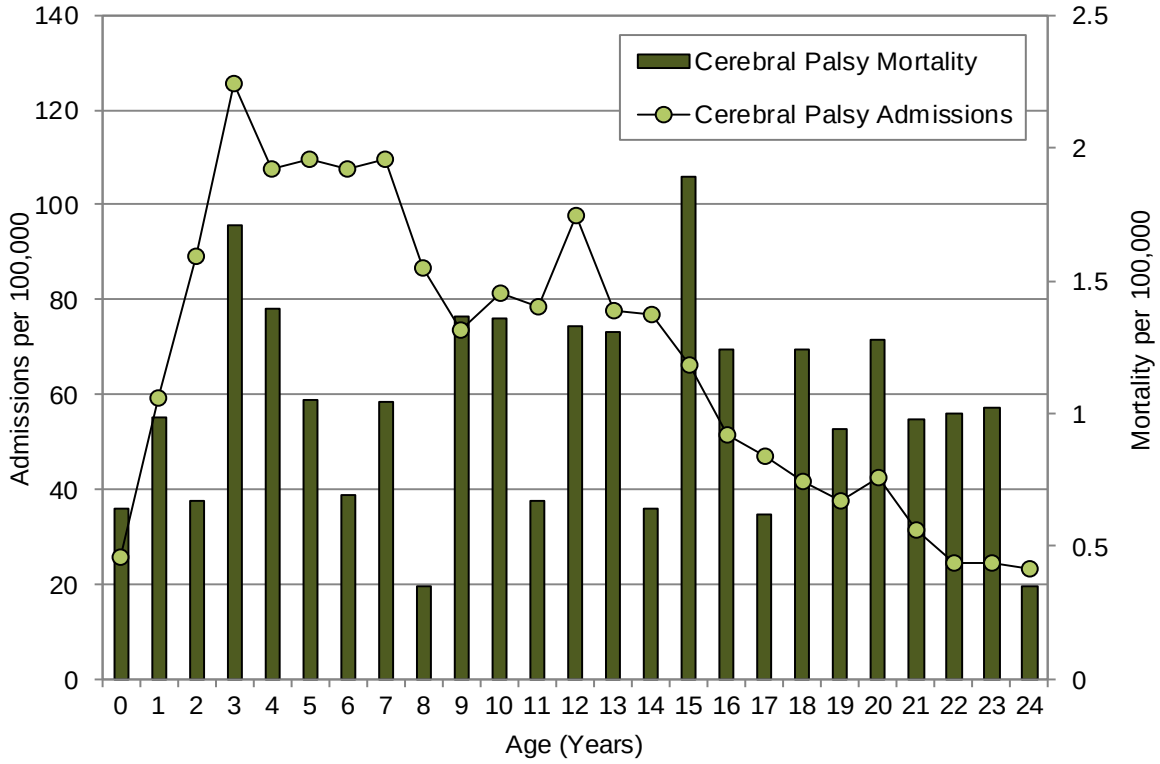
Primary Procedure	Number of Admissions: Total 2008–2012	Number of Admissions: Annual Average	Admission Rate per 100,000 Population	% of Waiting List Admissions	% of All Admissions in those with Cerebral Palsy
Waiting List Admissions by Primary Procedure					
Injection into Ligament, Tendon or Soft Tissue	1,107	221.4	14.50	42.2	21.7
Dental Procedures	233	46.6	3.05	8.9	4.6
Osteotomy of Proximal Femur	107	21.4	1.40	4.1	2.1
Release of Hip Contracture	85	17.0	1.11	3.2	1.7
Forage of Neck and/or Head of Femur	65	13.0	0.85	2.5	1.3
Lengthening/Repair of Achilles Tendon	60	12.0	0.79	2.3	1.2
Other Orthopaedic Procedures: Lower Limbs	147	29.4	1.93	5.6	2.9
Lengthening of Tendon, Unspecified	90	18.0	1.18	3.4	1.8
Spinal Fusion	48	9.6	0.63	1.8	0.9
Orthopaedic Procedures: Upper Limbs	31	6.2	0.41	1.2	0.6
Other Orthopaedic Procedures	160	32.0	2.10	6.1	3.1
Injection/Infusion of Other Therapeutic/Prophylactic Substance	57	11.4	0.75	2.2	1.1
Insertion of Percutaneous Endoscopic Gastrostomy (PEG) Tube	34	6.8	0.45	1.3	0.7
Insertion of Percutaneous Non-endoscopic Gastrostomy Button	19	3.8	0.25	0.7	0.4
Fundoplasty	34	6.8	0.45	1.3	0.7
Insertion/Replacement of Intrauterine Contraceptive Device	27	5.4	0.35	1.0	0.5
Tonsillectomy and/or Adenoidectomy	24	4.8	0.31	0.9	0.5
MRI of Brain	20	4.0	0.26	0.8	0.4
Other Procedures	208	41.6	2.72	7.9	4.1
No Procedure Listed	69	13.8	0.90	2.6	1.4
Total Waiting List Admissions	2,625	525.0	34.38	100.0	51.4
Total Acute and Arranged Admissions	2,483	496.6	32.52		48.6
Total Admissions in those with Cerebral Palsy	5,108	1,021.6	66.89		100.0

Source: Numerator: National Minimum Dataset, Waiting list admissions by primary procedure for children and young people with cerebral palsy listed in any of the first 15 diagnoses; Denominator: Statistics NZ Estimated Resident Population (projected from 2007)

Distribution by Age

In New Zealand during 2008–2012, hospital admissions for children and young people with cerebral palsy increased during infancy, reached a peak at three years of age, and then gradually declined. In contrast, mortality was more evenly distributed across the age range. During 2006–2010, 78 children and young people 0–24 years had cerebral palsy listed as their main underlying cause of death, or as a contributory cause (**Figure 1**).

Figure 1. Hospital Admissions (2008–2012) and Mortality (2006–2010) for New Zealand Children and Young People with Cerebral Palsy by Age



Source: Numerator Admissions: National Minimum Dataset, hospital admissions for children and young people with cerebral palsy listed in any of the first 15 diagnoses; Numerator Mortality: National Mortality Collection, Deaths with cerebral palsy listed as the main underlying or a contributory cause of death; Denominator: Statistics NZ Estimated Resident Population (projected from 2007)

Distribution by Ethnicity and Gender

In New Zealand during 2008–2012, hospital admissions for children and young people with cerebral palsy were *significantly* higher for males and for Pacific > European/Other > Māori > Asian/Indian children and young people (**Table 3**).

Table 3. Hospital Admissions for Children and Young People Aged 0–24 Years with Cerebral Palsy by Ethnicity and Gender, New Zealand 2008–2012

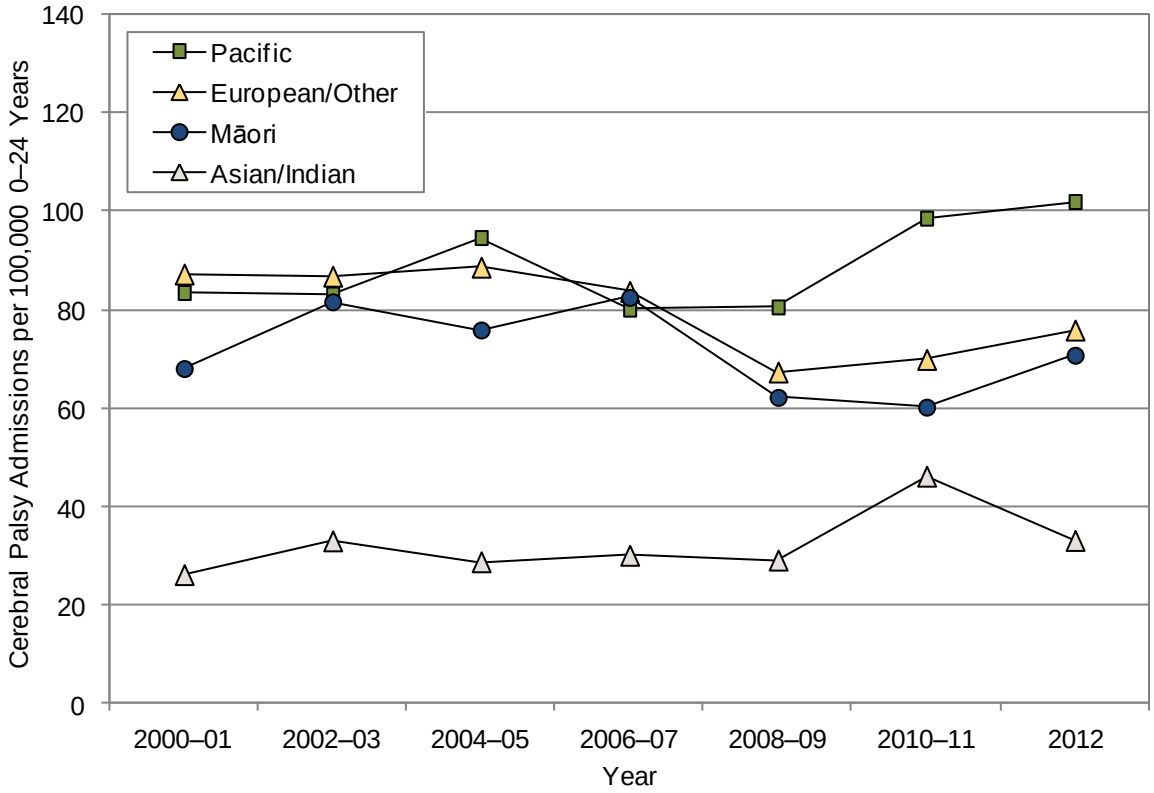
Variable	Rate	Rate Ratio	95% CI	Variable	Rate	Rate Ratio	95% CI
Cerebral Palsy							
Prioritised Ethnicity				Gender			
Asian/Indian	36.86	0.53	0.47–0.59	Female	57.56	1.00	
European/Other	70.01	1.00		Male	75.76	1.32	1.25–1.39
Māori	63.10	0.90	0.84–0.97				
Pacific	92.23	1.32	1.21–1.43				

Source: Numerator: National Minimum Dataset, hospital admissions for children and young people with cerebral palsy listed in any of the first 15 diagnoses; Denominator: Statistics NZ Estimated Resident Population (projected from 2007); Note: Ethnicity is Level 1 Prioritised; Rate is per 100,000 population

Trends by Ethnicity

While admission rates for Pacific, Māori and European/Other children and young people with cerebral palsy were similar during the early-mid 2000s, diverging trends saw rates for Pacific children and young people become higher than for European/Other and Māori children and young people from 2008–09 onwards. Admission rates for Asian/Indian young people however, were lower than for Pacific, Māori and European/Other children and young people throughout 2000–2012 (**Figure 2**).

Figure 2. Hospital Admissions for Children and Young People Aged 0–24 Years with Cerebral Palsy by Ethnicity, New Zealand 2000–2012



Source: Numerator: National Minimum Dataset, hospital admissions for children and young people with cerebral palsy listed in any of the first 15 diagnoses; Denominator: Statistics NZ Estimated Resident Population (projected from 2007); Note: Ethnicity is Level 1 Prioritised

South Island DHBs Distribution and Trends

South Island Distribution

In the South Island during 2008–2012, 52 Nelson Marlborough, 13 South Canterbury, 227 Canterbury, 11 West Coast, 72 Otago and 42 Southland children and young people were hospitalised with a diagnosis of cerebral palsy. Admission rates per 100,000 in Canterbury were *significantly* higher than the New Zealand rate, while in South Canterbury and Southland rates were *significantly* lower. Rates in the remaining DHBs were not *significantly* different from the New Zealand rate (**Table 4**).

South Island Trends

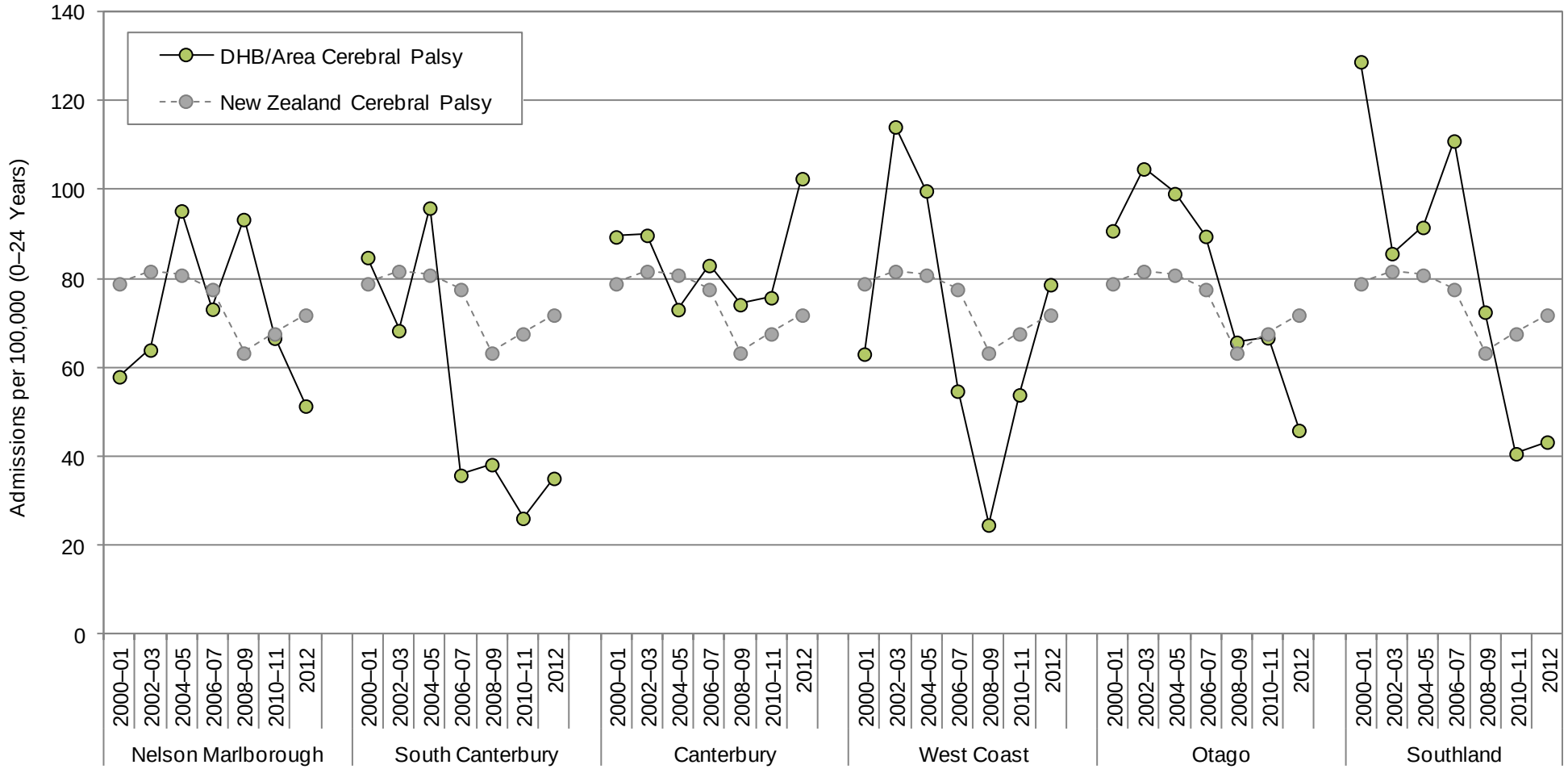
In the South Island DHBs during 2000–2012, large year to year variations (likely as a result of small numbers) made individual DHB's trends in admission rates difficult to interpret (**Figure 3**).

Table 4. Hospital Admissions for Children and Young People Aged 0–24 Years with Cerebral Palsy, South Island DHBs vs. New Zealand 2008–2012

DHB/Area	Total Number of Individuals 2008−2012		Total Number of Admissions 2008−2012	Average Number of Admissions per Individual per Year	Admission Rate per 100,000 Population	Rate Ratio	95% CI
	A*	B*					
Cerebral Palsy							
Nelson Marlborough	48	52	158	0.61	74.33	1.11	0.95–1.30
South Canterbury	12	13	28	0.43	32.88	0.49	0.34–0.71
Canterbury	224	227	670	0.59	80.57	1.20	1.11–1.31
West Coast	11	11	24	0.44	47.35	0.71	0.47–1.06
Otago	71	72	209	0.58	62.29	0.93	0.81–1.07
Southland	39	42	99	0.47	54.01	0.81	0.66–0.98
New Zealand	1,657		5,108	0.62	66.89	1.00	

Source: Numerator: National Minimum Dataset, hospital admissions for children and young people with cerebral palsy listed in any of the first 15 diagnoses; Denominator: Statistics NZ Estimated Resident Population (projected from 2007); Note: A*: Each individual only counted once in DHB in which they first reside (i.e. DHB total = NZ total); B*: Each individual counted once in each DHB in which they reside (i.e. the sum of DHB totals exceeds New Zealand total); Rate Ratios are compared to New Zealand rates and have not been adjusted for population demographics

Figure 3. Hospital Admissions for Children and Young People Aged 0–24 Years with Cerebral Palsy, South Island DHBs vs. New Zealand 2000–2012



Source: Numerator: National Minimum Dataset, hospital admissions for children and young people with cerebral palsy listed in any of the first 15 diagnoses; Denominator: Statistics NZ Estimated Resident Population (projected from 2007)

Policy Documents and Evidence-Based Reviews Relevant to the Diagnosis or Management of Cerebral Palsy

In New Zealand there is a paucity of policy documents relevant to children and young people with cerebral palsy. **Table 5** however summarises a range of overseas publications which may be relevant in this context.

Table 5. Policy Documents and Evidence-Based Reviews Relevant to Cerebral Palsy

International Guidelines and Useful Websites
Ferluga E D, Archer K R, Sathe N A, et al. 2013. **Interventions for Feeding and Nutrition in Cerebral Palsy. Comparative Effectiveness Review No. 94. (Prepared by the Vanderbilt Evidence-based Practice Center under Contract No. 290-2007-10065-I.) AHRQ Publication No. 13-EHC015-EF**. Rockville, MD: Agency for Healthcare Research and Quality. http://www.effectivehealthcare.ahrq.gov/ehc/products/436/1426/Cerebral-Palsy-Feeding-Nutrition-130318.pdf Children with cerebral palsy (CP) often have feeding and swallowing problems which may lead to growth failure and repeated aspiration of food resulting in chronic pulmonary disease. If a child cannot take sufficient food orally to maintain growth, there is frequent aspiration, or the level of work required by the child or their caregiver to maintain an adequate oral intake is excessive then feeding via a surgically implanted gastrostomy or jejunostomy tube may be recommended. This review assessed the effects of interventions for feeding and nutrition problems in individuals with CP using evidence from research studies. The authors explored the evidence relating to specified "key questions". They identified one systematic review on behavioural interventions and 12 unique primary studies, most of which were surgical case series. They stated that, overall, there was little data to guide care, that the study populations were almost entirely children with severe CP, and that most studies were short term and did not consistently assess harms of interventions. They concluded that the evidence for behavioural interventions ranged from insufficient to moderate and noted that some studies suggested that sensorimotor interventions such as oral appliances (moderate strength of evidence) and positioning (low strength of evidence) may be beneficial. They also concluded that the evidence for surgical interventions ranged from insufficient to low but that all studies to date had demonstrated significant weight gain with gastrostomy. They noted that there is considerable uncertainty over the harms of feeding interventions both in the short and the long term. They stated that harms associated with gastrostomy can be common and include overfeeding, site infection, stomach ulcer, and reflux.
Childhood Integrated Neuroscience Discovery Network (CP-NET) http://cpnet.canchild.ca/en/ This site is part of the website of the CanChild Centre for Childhood Disability Research located at McMaster University in Hamilton, Ontario, Canada. A collection of research articles on CP can be found here: http://cpnet.canchild.ca/en/Research_Articles_on_CP.asp
National Institute for Health and Care Excellence. 2012. **Spasticity in children and young people with non-progressive brain disorders: management of spasticity**. London: National Institute for Health and Clinical Excellence. http://www.nice.org.uk/nicemedia/live/13803/60029/60029.pdf This clinical guideline covers the management of spasticity and co-existing motor disorders and their early musculoskeletal complications in children and young people aged 0–18 years with non-progressive brain disorders, the most common of which is cerebral palsy. It does not cover the management of other aspects of cerebral palsy. This guideline is an abbreviated version of the full guideline: National Collaborating Centre for Women's and Children's Health (Commissioned by the National Institute for Health and Care Excellence). 2012. **Spasticity in children and young people with non-progressive brain disorders: management of spasticity and co-existing motor disorders and their early musculoskeletal complications** London: National Collaborating Centre for Women's and Children's Health. http://www.nice.org.uk/nicemedia/live/13803/60023/60023.pdf These comprehensive evidence-based guidelines are intended for healthcare professionals, service commissioners and planners, social service and education professionals, and families, carers and children and young people. They cover physical therapy (physiotherapy and/or occupational therapy), orthoses, oral drugs, botulinum toxin, intrathecal baclofen, orthopaedic surgery, and selective dorsal rhizotomy. There is also a chapter dealing with the health economics of these therapies. For each review question addressed in the guidelines there is an "evidence profile", a table listing the relevant studies, and, for each study, numbers of participants, size of effect (with 95% confidence interval) and study quality.

National Institute for Health and Care Excellence. 2010. **IPG373 Selective dorsal rhizotomy for spasticity in cerebral palsy: guidance**. London: National Institute for health and Care Excellence.
http://www.nice.org.uk/nicemedia/live/11220/52083/52083.pdf

This brief document provides guidance on selective dorsal rhizotomy for spasticity in cerebral palsy. The procedure involves surgery to cut some of the sensory nerve rootlets leaving the lumbar region of the spinal cord with the aim of reducing sensory input to the sensory–motor reflex arcs responsible for increased muscle tone. Three non-randomised comparative studies (142, 108 and 142 patients) have reported positive results from the procedure. This publication states that there is a risk of serious but well-recognised complications and that the evidence for efficacy is adequate. More details of the studies on which the guidance document is based are contained in the following publication:

National Institute for Health and Care Excellence. 2010. **Selective dorsal rhizotomy for spasticity in cerebral palsy - Overview**. London: National Institute for Health and Care Excellence.
http://www.nice.org.uk/nicemedia/live/11220/49551/49551.pdf

Guidelines from the James M. Anderson Center for Health Systems Excellence at Cincinnati Children's Hospital
http://www.cincinnatichildrens.org/service/j/anderson-center/default/

The Anderson Center at Cincinnati Children's Hospital has published a number of "Best Evidence Statements" related to care issues for children with cerebral palsy. They contain details of the studies relevant to the clinical questions addressed and care recommendations. Guidance on the following CP-related topics (with year of publication) can be found at these links:

- Lower Extremity Orthoses for Children with Hemiplegic Cerebral Palsy (2010)
- Aquatic Therapy for Children with Hemiplegic Cerebral Palsy (2010)
- Strengthening (progressive resistive exercise) for individuals with Cerebral Palsy age 4-20 years who demonstrate muscle weakness (2010)
- Biofeedback Intervention for Children with Hemiplegic Cerebral Palsy (2010)
- Pediatric Constraint Induced Movement Therapy (CIMT) (2009)

American Academy for Cerebral Palsy and Developmental Medicine http://www.aacpdm.org

This organisation describes itself as "A global leader in the multidisciplinary scientific education of health professionals and researchers dedicated to the well-being of people with childhood-onset disabilities." It provides scientific information for health professionals and promotes excellence in research and services. It publishes the journal **Developmental Medicine & Child Neurology.** The following evidence reports can be found on this website: http://www.aacpdm.org/publications/outcome. Titles are followed by publication date. Where there is no publication date on a report, the date given is the latest year of the literature search on which each report is based.

- A systematic review of the effectiveness of aerobic exercise interventions for children with cerebral palsy (2006)
- A systematic review of the effects of casting on equinus in children with cerebral palsy (2006)
- Effects of therapy for children with CP following botulinum toxin-A injections (2003)
- Effects of Conductive Education for Cerebral Palsy (2003)
- Effects of Intrathecal Baclofen for Spastic and Dystonic Cerebral Palsy (2000)
- Effects of Neurodevelopmental Treatment (NDT) for Cerebral Palsy (2001)
- Effects of Gastrostomy Feeding in Children with Cerebral Palsy (2002)
- Effect of Surgical Adductor Releases for Hip Subluxation in Cerebral Palsy (2003)

Systematic and Other Reviews from the International Literature

Novak I, McIntyre S, Morgan C, et al. 2013**. A systematic review of interventions for children with cerebral palsy: state of the evidence.** Dev Med Child Neurol, 55(10), 885-910
http://onlinelibrary.wiley.com/doi/10.1111/dmcn.12246/pdf

This thorough and concise review, describing itself as a systematic review of systematic reviews, aimed to describe the best available evidence on interventions for children with cerebral palsy (CP) using the GRADE system and to assist clinicians with translating evidence to practice and deciding what to do. This evidence is complemented with the Evidence Alert Traffic Light System. The authors found 166 articles meeting the review's inclusion criteria, 74 of which were systematic reviews. These articles addressed 64 discrete interventions and reported on 131 outcomes. The 'green' interventions (those with the highest quality favourable evidence and therefore strong recommendations) included anticonvulsants, bimanual training, botulinum toxin, bisphosphonates, casting, constraint-induced movement therapy, context-focused therapy, diazepam, fitness training, goal-directed training, hip surveillance, home programmes, occupational therapy after botulinum toxin, pressure care, and selective dorsal rhizotomy. Most interventions (70%) had lower level evidence and the authors noted that many of these interventions are in common use in standard care which is a problem for people with CP, healthcare providers, funders and purchasers.

Vernon-Roberts A, Sullivan PB. 2013. **Fundoplication versus postoperative medication for gastro-oesophageal reflux in children with neurological impairment undergoing gastrostomy**. Cochrane Database of Systematic Reviews(8).

The development or worsening of gastro-oesophageal reflux (GOR) is a widely reported complication of gastrostomy tube placement. To reduce the possibility of this occurring surgical anti-reflux treatment in the form of fundoplication or another anti-reflux procedure is frequently performed at the same time as gastrostomy surgery. Fundoplication involves wrapping the fundus of the stomach around the oesophagus at the gastro-oesophageal junction to strengthen the barrier to acid reflux. The authors did not identify any RCTs involving children aged up to 18 years with neurological impairments and GOR who were undergoing gastrostomy tube insertion and therefore they concluded that there is considerable uncertainty about the optimal treatment when faced with the choice of fundoplication vs. anti-reflux medications for children with GOR and neurological impairment who are undergoing gastrostomy insertion.

Gantasala S, Sullivan PB, Thomas AG. 2013. **Gastrostomy feeding versus oral feeding alone for children with cerebral palsy.** Cochrane Database of Systematic Reviews(7).

The authors of this review aimed to evaluate the effects of nutritional supplementation given via gastrostomy or jejunostomy to children with feeding difficulties due to CP. They did not identify any RCTs relating to this issue and so they concluded that there remains considerable uncertainty about the effects of gastrostomy for children with CP.

Nguyen TN, Crowther CA, Wilkinson D, et al. 2013. **Magnesium sulphate for women at term for neuroprotection of the fetus.** Cochrane Database of Systematic Reviews(2).

Magnesium sulphate is widely used in obstetrics for the prevention and treatment of eclampsia and it has been shown to be effective for neuroprotection of the fetus when given to women at risk of very preterm birth. Since more than half of all cases of cerebral palsy occur in children born at term it is important to determine whether antenatal administration of magnesium sulphate to women at term would also protect the fetus from brain injury and associated disabilities, including CP. This was the aim of this review. The authors identified one RCT involving 135 women with mild pre-eclampsia at term. This trial did not report on any of the review's pre-specified primary outcomes and so the authors concluded that there is currently insufficient evidence to assess the efficacy and safety of magnesium sulphate when given to women at term for neuroprotection of the fetus.

Scholefield B, Duncan H, Davies P, et al. 2013. **Hypothermia for neuroprotection in children after cardiopulmonary arrest.** Cochrane Database of Systematic Reviews(2).

Severe brain damage as the result of hypoxic ischaemia may follow a cardiopulmonary arrest and resuscitation and lead to the development of cerebral palsy, blindness, seizures and hypothalamic and pituitary insufficiency. Very severe brain damage can be fatal or lead to a persistent vegetative state. Therapeutic hypothermia (lowering the core body temperature to between 32°C and 34°C) may reduce brain injury in the period after circulation has been restored. This therapy has been effective for neonates with hypoxic ischaemic encephalopathy and adults after witnessed cardiopulmonary arrest with ventricular fibrillation. This review considered the clinical effectiveness of therapeutic hypothermia after paediatric cardiopulmonary arrest. The authors were unable to find any RCTs or quasi-RCTs meeting their criteria, but they found four on-going RCTs which may provide data for analysis in the future. They stated that, based on their review findings, they were unable to make any recommendations for clinical practice.

Jacobs SE, Berg M, Hunt R, et al. 2013. **Cooling for newborns with hypoxic ischaemic encephalopathy.** Cochrane Database of Systematic Reviews(3).

Pilot studies in humans, as well as animal studies, suggest that inducing mild hypothermia (cooling) after peripartum hypoxia-ischaemia in newborn infants may reduce neurological sequelae without adverse effects. This review considered the effect of therapeutic hypothermia in encephalopathic asphyxiated newborn infants on mortality and long-term neurodevelopmental disability and assessed whether there are clinically important side effects. The review included 11 RCTs involving 1505 term and late preterm infants with moderate/severe encephalopathy and evidence of intrapartum asphyxia. The results of these trials indicated that therapeutic hypothermia led to a statistically significant and clinically important reduction in the combined outcome of death or major developmental disability at age 18 months (typical relative risk 0.75 (95% CI 0.68 to 0.83); typical risk difference −0.15, 95% CI −0.20 to −0.10); number needed to treat for an additional beneficial outcome (NNTB) 7 (95% CI 5 to 10) (8 studies, 1344 infants). Cooling also produced statistically significant reductions in mortality (typical RR 0.75 (95% CI 0.64 to 0.88), typical RD −0.09 (95% CI −0.13 to −0.04); NNTB 11 (95% CI 8 to 25) (11 studies, 1468 infants) and in neurodevelopmental disability in survivors (typical RR 0.77 (95% CI 0.63 to 0.94), typical RD −0.13 (95% CI −0.19 to −0.07); NNTB 8 (95% CI 5 to 14) (8 studies, 917 infants). Adverse effects of cooling included an increase sinus bradycardia and a significant increase in thrombocytopenia. The authors concluded that therapeutic hypothermia is beneficial for term and late preterm infants with hypoxic ischaemic encephalopathy. It reduces mortality without increasing long term disability and the benefits outweigh the short term adverse effects. They stated that hypothermia should be instituted in term and late preterm infants with moderate-to-severe hypoxic ischaemic encephalopathy if this is identified before six hours of age.

Walshe M, Smith M, Pennington L. 2012. **Interventions for drooling in children with cerebral palsy.** Cochrane Database of Systematic Reviews(11).

Drooling is a common problem for children with CP and their parents and caregivers. It may lead to social rejection, damp and smelly clothing, chapped skin, mouth infections, dehydration, speech difficulties and damage to learning materials like books and computers. This review had three aims: firstly to evaluate the effectiveness and safety of interventions aimed at reducing or eliminating drooling in children with CP, secondly to provide the best available evidence to inform clinical practice and, thirdly, to assist with future research planning. The review included studies only if they were RCTs or controlled clinical trials (CCTs). The authors found six relevant studies, five RCTs and one CCT, with approximately 198 participants altogether. Four were of botulinum toxin A (BoNT-A) and there was one study on each of the pharmacological treatments benztropine and glycopyrrolate. There was considerable heterogeneity between the BoNT-A studies and so a meta-analysis was not possible. All six studies showed some statistically significant effects of the intervention for up to one month following the intervention but the authors considered that all studies had methodological flaws. They concluded that it was not possible to reach a conclusion on the effectiveness and safety of either BoNT-A or the pharmaceutical interventions, benztropine and glycopyrrolate, and that there is insufficient evidence to inform clinical practice on interventions for drooling in children with CP.

Morgan AT, Dodrill P, Ward EC. 2012. **Interventions for oropharyngeal dysphagia in children with neurological impairment.** Cochrane Database of Systematic Reviews(10).

Oropharyngeal dysphagia (OD) encompasses problems with chewing and preparing the food for swallowing, moving the food or fluid posteriorly through the oral cavity with the tongue into the back of the throat, and swallowing the food or fluid and moving it through the pharynx to the oesophagus. It is commonly experienced by children with neurological impairment, including those with CP. The authors of this review considered the effectiveness of interventions for OD in children with neurological impairment. They found three small studies meeting their inclusion criteria, two of which they considered to have high risk of bias. Two studies compared oral sensorimotor interventions to standard care in children with CP and the other compared lip strengthening exercises to no treatment in children with myotonic dystrophy type 1. The two trials of oral sensorimotor interventions were too different for meta-analysis to be possible. The authors concluded that there is currently insufficient high-quality evidence from RCTs or quasi-RCTs to provide conclusive results about the effectiveness of any particular type of oral-motor therapy for children with neurological impairment and that more research with larger-scale RCTs is urgently needed.

Spittle A, Orton J, Anderson P, et al. 2012. **Early developmental intervention programmes post-hospital discharge to prevent motor and cognitive impairments in preterm infants.** Cochrane Database of Systematic Reviews(12).

Compared to infants born at term, infants born pre-term are at higher risk of developing cognitive and motor impairments. This review considered the effectiveness of early developmental intervention post-discharge from hospital for improving motor or cognitive development in preterm (< 37 weeks) infants with no major congenital abnormalities. Twenty-one studies met the review's inclusion criteria (3133 randomised patients) but only ten were RCTs with appropriate allocation concealment. Meta-analysis indicated that intervention improved cognitive outcomes at infant age (developmental quotient (DQ): standardised mean difference (SMD) 0.31 standard deviations (SD); 95% confidence interval (CI) 0.13 to 0.50; P < 0.001; 13 studies; 2147 patients), and pre-school age (intelligence quotient (IQ); SMD 0.45 SD; 95% CI 0.34 to 0.57; P < 0.001; six studies; 1276 patients). However, this effect was not sustained at school age (IQ: SMD 0.25 SD; 95% CI -0.10 to 0.61; p=0.16; five studies; 1242 patients). There was significant heterogeneity between studies for cognitive outcomes at infant and school ages. In respect of motor outcomes, meta-analysis of 10 studies showed a significant effect in favour of early developmental interventions but the effect was small (motor scale developmental quotient (DQ): SMD 0.10 SD; 95% CI 0.00 to 0.19; p=0.04; 10 studies; 1745 patients). There was no effect on the rate of cerebral palsy in survivors; risk ratio (RR) 0.89; 95% CI 0.55 to 1.44; five studies; 737 patients). There was little evidence for a positive effect on motor outcomes in the long term, and only five studies reported outcomes at pre-school or school age. The authors concluded that early intervention programmes for preterm infants have a positive influence on cognitive and motor outcomes during infancy and the cognitive benefits persist into pre-school age. Substantial differences between studies make comparing intervention programmes hard.

Valentin-Gudiol M, Mattern-Baxter K, Girabent-Farres M, et al. 2011. **Treadmill interventions with partial body weight support in children under six years of age at risk of neuromotor delay.** Cochrane Database of Systematic Reviews(12).

A number of childhood disorders, including Down syndrome, cerebral palsy (CP), spina bifida and a broad range of other neuromuscular disorders are associated with delays in motor development in infancy. Treadmill training, in which the child is supported by a harness, can give children the opportunity to walk with support for long enough periods of time to acquire the motor skills necessary for independent walking. This review assessed the effectiveness of treadmill interventions on locomotor motor development in pre-ambulatory infants and children under six years of age who are at risk for neuromotor delay. The review included five studies (RCTs or controlled clinical trials) reporting on treadmill interventions. The studies had methodological limitations as outcome assessors were not blinded. In total 139 children were involved, 73 in the intervention groups and the rest in the control groups. Three studies involved children with Down syndrome (90 children in total), one involved children with CP (8 in total) and one involved children at risk of neuromotor delay. The studies were quite diverse with regard to the types of comparisons made, the time of evaluation and the parameters assessed, so meta-analysis was only possible for the results of three studies. The authors stated that, given the limited number of studies, and their heterogeneity, they could not provide any firm evidence for the clinical application of treadmill training but that, for children with Down syndrome, treadmill training may facilitate earlier walking and children with Down syndrome who received more intensive treadmill intervention may be more accomplished in their gait parameters that the children who received less intensive treadmill intervention.

Pennington L, Goldbart J, Marshall J. 2011**. Speech and language therapy to improve the communication skills of children with cerebral palsy.** Cochrane Database of Systematic Reviews(9).

Children with CP often have difficulties with speech, language and gesture due to motor, sensory and intellectual impairment. Speech and language therapy (SLT) is used to help children maximise their communication skills. It may involve augmentative and alternative communication systems, such as symbol charts or communication aids with synthetic speech, as well as enhancing children's natural forms of communication. This review considered the effectiveness of SLT focusing on the child or their familiar communication partners (as measured by changes in patterns of interaction) and whether some forms of SLT are better than others. The review criteria included any experimental study with an element of control and sixteen such studies were identified by the authors. Nine studies evaluated treatment given to the children and seven training for communication partners. The authors concluded that their review did not indicate firm evidence for the positive effect of SLT for children with CP but there were some positive trends in communication. They recommended further more methodologically rigorous research.

Ade-Hall R, Moore P. 2009. **Botulinum toxin type A in the treatment of lower limb spasticity in cerebral palsy.** Cochrane Database of Systematic Reviews(1).

Children with CP often have spasticity (stiffness) of the legs due to involuntary muscle over-activity caused by brain or spinal cord damage. Spasticity is a major component of the disability and deformities associated with CP and it causes poor coordination, spasms, abnormal posture and pain. Botulinum toxin blocks the release of acetylcholine from the neuromuscular junction and weakens the muscle. This review considered whether botulinum toxin A(BoNT-A) is safe and effective treatment for lower limb spasticity in children with CP. The authors identified three small, short term, randomised studies with follow-up periods of between four and 26 weeks. They concluded that there was no strong evidence to either support of refute the use of BoNT-A for the treatment of leg spasticity in CP.

Hoare BJ, Wallen MA, Imms C, et al. 2010. **Botulinum toxin A as an adjunct to treatment in the management of the upper limb in children with spastic cerebral palsy (UPDATE).** Cochrane Database of Systematic Reviews(1).

This review assessed the effectiveness of injections of Botulinum toxin A (BoNT-A)**,** alone or in combination with occupational therapy, for the treatment of the upper limb in spasticity and hypertonia children with CP. The authors identified ten RCTs meeting their inclusion criteria, nine of which were considered to be of high quality. They concluded that there was high level evidence to support the use of BoNT-A as an adjunct to occupational therapy in managing the upper limbs in children with spastic CP and moderated evidence that BoTN-A alone is not effective. They recommended that practitioners follow the Fehlings guidelines:

Fehlings D, Novak I, Berweck S, et al. 2010. **Botulinum toxin assessment, intervention and follow-up for paediatric upper limb hypertonicity: international consensus statement**. Eur J Neurol*,* 17 Suppl 2, 38-56.

Katalinic OM, Harvey LA, Herbert RD, et al. 2010. **Stretch for the treatment and prevention of contractures**. Cochrane Database of Systematic Reviews(9).

Stretch is widely used for the prevention of contractures and the preservation of joint mobility in a range of neurological and musculoskeletal conditions, including CP. This review assessed the effectiveness of this practice. The authors identified 35 RCTs or controlled clinical trials meeting their criteria with 1391 participants in total. No study lasted longer than seven months. The authors found that in people with neurological conditions, there was moderate to high quality evidence to indicate that stretch does not have clinically important effects on joint mobility either immediately (mean difference 3°; 95% CI 0 to 7), in the short-term (mean difference 1°; 95% CI 0 to 3) or the long-term (mean difference 0°; 95% CI −2 to 2). The results were similar for people with non-neurological conditions. For all conditions, they found little or no effect of stretch on pain, spasticity, activity limitation, participation restriction or quality of life. They concluded that stretch does not have clinically important effects on joint mobility in people with, or at risk of, contractures if performed for less than seven months and they stated that the effects of stretch performed for periods longer than seven months have not been investigated.

Smeulders JCM, Coester A, Kreulen M. 2009. **Surgical treatment for the thumb-in-palm deformity in patients with cerebral palsy**. Cochrane Database of Systematic Reviews(4).

Thumb-in-palm deformity impairs the ability of a person with CP to use their thumb and severely limits hand function. A variety of surgical procedures to correct this problem have been described. This review considered: the efficacy of surgical interventions for the thumb-in-palm deformity in patients with spastic CP; the selection criteria for surgical treatment of thumb-in-palm deformity in these patients; and the outcome assessment used in studies of these treatments. The authors did not find any relevant RCTs or controlled clinical trials. They did find 14 prospective studies and included the best nine of these, which compared preoperative and postoperative assessment, in their review. They concluded that because of the poor methodological quality of the studies it was not possible to provide a reliable judgement of the role of surgery for thumb-in-palm deformity in patients with CP but surgery appeared to improve hand function, to facilitate hygiene, and to improve appearance and quality of life.

Hoare BJ, Wasiak J, Imms C, et al. 2009. **Constraint-induced movement therapy in the treatment of the upper limb in children with hemiplegic cerebral palsy.** Cochrane Database of Systematic Reviews(4).

Children with hemiplegic CP often neglect or fail to use their affected limb. Constraint-induced movement therapy (CIMT) is an emerging treatment for hemiplegic CP. It aims to increase spontaneous use of the affected upper limb and thereby limit the effects of developmental disregard. CIMT is based on two fundamental principles: constraint of the non-affected limb and massed practice of therapeutic tasks with the affected limb. This review examined the effectiveness of CMIT for children with upper limb hemiplegic CP. The authors identified three studies (RCTs or controlled clinical trials) meeting their inclusion criteria with 18, 41 and 31 children. The studies used a variety of constraint methods and outcome measures. One trial found a significant treatment effect using modified CIMT and another found a positive trend favouring modified CMIT and Forced Use. The authors concluded that, given the limited evidence, the use of CIMT, modified CIMT and Forced Use should be considered experimental.

Delgado MR, Hirtz D, Aisen M, et al. 2010. **Practice parameter: pharmacologic treatment of spasticity in children and adolescents with cerebral palsy (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, 74(4), 336-43 http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3122302/

This review evaluated the efficacy and safety of pharmacologic treatments for childhood spasticity due to cerebral palsy. The authors did not find any peer reviewed studies on phenol, alcohol, or botulinum toxin type B that involved more than nine patients with CP aged 19 years or younger and had English language abstracts. They stated that there was insufficient evidence to either support or refute the use of BoNT-B, phenol, and alcohol injections as a treatment for spasticity in children with spastic CP.

Botulinum type A: They found 148 studies of botulinum toxin A (BoNT-A) fulfilling the same criteria. Fifteen of these were Class I and five were Class II according to the American Academy of Neurology criteria. (A Class I trial is a randomized, controlled clinical trial with masked or objective outcome assessment in a representative population and a Class II trial is either a RCT that lacks one of the criteria for a class I trial or a high-quality prospective matched cohort study). A total of 573 children received BoNT-A in the Class I and II studies. All but three of these studies reported spasticity reduction but there was conflicting evidence regarding functional improvement. Seventeen studies reported transient adverse events, most commonly localised pain, excessive weakness, unsteadiness and increased falls, and fatigue. Five patients were reported to have developed urinary incontinence and two, dysphagia. The authors recommended that for children with CP who have localised or segmental spasticity in the upper and lower extremities, BoNT-A should be offered as an effective and generally safe treatment. They stated that there was insufficient evidence to either support or discourage the use of BoTN-A to improve motor function in these children.

Diazepam: Based on the results of one Class I study (n= 180) and one Class II study, the authors concluded that diazepam is probably effective for the short term treatment of spasticity in children with CP. They noted that no studies formally considered whether it improved motor function and that therefore there was insufficient evidence to support or refute its use for this purpose. Most studies reported ataxia and drowsiness as side effects. They recommended that diazepam be considered a short term anti-spasticity treatment and noted that adverse effects such as sedation, drowsiness, hypersalivation and weakness limit is usefulness for long term use and that prolonged use can produce physical dependence.

Dantrolene: The authors identified one Class I, two Class II and two Class IV studies. The Class I study and one of the Class II studies had conflicting results. Weakness, drowsiness and irritability were frequent side effects. The authors stated that there was insufficient evidence to support or refute the use of dantrolene and noted that, in their experience, this agent is rarely used in clinical practice.

Baclofen (oral): There were two Class II studies (one with 20 children and one with 15) and one class IV study. The results of the Class II studies were conflicting and some participants in the larger study suffered somnolence or sedation (20%) and hypotonia (15%). The authors concluded that there was insufficient evidence to support or refute the use of this medication but they noted that it is widely used in clinical practice.

Tizanidine: There was one small Class II placebo-controlled parallel study (10 children in the treatment group and 30 in the placebo group). There was a significant reduction in spasticity in the treatment group and no side effects were reported. The authors concluded that this agent is possibly effective and recommended that its use may be considered in children with CP and spasticity. They noted that this agent is used in adults with multiple sclerosis and spinal cord injury with reported adverse effects including hypotension, sedation, asthenia, dry mouth, dizziness, hallucinations, and hepatotoxicity.

Intrathecal baclofen pump: This device is a surgically implanted pump that delivers medication via a catheter into the spinal cord. There was one Class III and five Class IV studies and all reported reduced spasticity. Common adverse effects were: catheter malfunction (43%), wound infection (39%), seromas (29%) and CSF leaks (17%). Less common were headache, vomiting, lethargy, disorientation, agitation, irritability, and meningitis. The authors concluded that the data regarding the use of continuous intrathecal baclofen in children with CP and spasticity are inadequate and that there is insufficient evidence to support or refute its use. They noted the impracticality of RCTs of this technology.

Other Relevant Publications

Reddihough D, Ong K, Bajraszewski E, et al. 2008. **Cerebral Palsy: An information guide for parents**. Melbourne: The Royal Children's Hospital. http://ww2.rch.org.au/emplibrary/cdr/CPBooklet.pdf

This concise guideline intended for parents provides an overview of cerebral palsy and its treatment.

Note: The publications listed were identified using the search methodology outlined in Appendix 1 (Search Methods for Policy Documents and Evidence-Based Reviews)

References

1. Blair E, Stanley F. 1997. Issues in the Classification and Epidemiology of Cerebral Palsy. *Mental Retardation and Developmental Disabilities Research Reviews* 3 184-93.
2. Kragenloh-Mann I, Cans C. 2009. Cerebral palsy update. *Brain and Development* 31 537-44.
3. Rudolph A. 1996. Rudolph's Pediatrics. London: Prentice Hall International.
4. Krigger KW, Krigger KW. 2006. Cerebral palsy: an overview. *American Family Physician* 73(1) 91-100.