2009 Annual Report

of

The South Australian Cerebral Palsy Register

(part of The South Australian Birth Defects Register)

Children born 1993 to 2004 with cerebral palsy notified to the Register by 31st December 2009

July 2010

The South Australian Cerebral Palsy Register

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A/Prof Annabelle Chan	*Public Health Physician*
Mrs Phillipa van Essen	*Manager, SA Birth Defects Register (part-time)*
Dr Catherine Gibson	*Manager, SA Birth Defects Register (part-time)*
Mrs Rosie Rice	*Register Officer, SA Cerebral Palsy Register*
Ms Heather Scott	*Register Officer, SA Birth Defects Register*

Specialist Advisors to the Register

Dr Andrew Tidemann	*Paediatric Rehabilitation Specialist*
Dr Deirdre White	*Developmental Paediatrician*
Ms Terry Lyons	*Acting Director, Client Programs, Novita Children's Services*
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Register Management Group

A/Prof Peter Baghurst	*Representing the SA Birth Defects Register*
A/Prof Annabelle Chan	*Representing SA Department of Health, Epidemiology Branch*
Dr Ray Russo	*Paediatric Rehabilitation Specialist*
Mrs Phillipa van Essen	*Manager, SA Birth Defects Register (part-time)*
Dr Catherine Gibson	*Manager, SA Birth Defects Register (part-time)*
Mrs Rosie Rice	*Register Officer, SA Cerebral Palsy Register*
Ms Heather Scott	*Register Officer, SA Birth Defects Register*

Collaborating Organisations

Novita Children's Services
Flinders Medical Centre
Children, Youth and Women's Health Service

Funding

The Register is funded by the Children, Youth and Women's Health Service, with additional support provided by Novita Children's Services of South Australia

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Foreword

As mentioned elsewhere in this Annual Report of the South Australian Cerebral Palsy Register, there is no known cure for cerebral palsy. At a research level, however, there is growing evidence that a substantial proportion of cases of cerebral palsy may now be preventable through the very simple means of giving magnesium sulphate to women who may be about to give birth to a very preterm baby.

Around 42% of all cases of cerebral palsy are associated with preterm births with cerebral palsy being 30 times more frequent among babies who survive a birth at 28 weeks or less, compared with babies born at 37 weeks or more.

Administration of magnesium sulphate does not prevent a baby from being born prematurely, but it does appear to provide protection from the brain injury to which preterm babies are so vulnerable. During 2009 a Cochrane Review looked at the results of four clinical trials of magnesium sulphate versus a placebo treatment, and found that the risk of cerebral palsy in babies whose mothers were administered magnesium sulphate was nearly 30% lower than in babies whose mothers received the placebo. One of those four trials was conducted in hospitals across Australia and New Zealand under the leadership of South Australia's Professor Caroline Crowther. To ensure her results are translated into clinical practice, Professor Crowther has recently chaired a panel which has just released draft national clinical practice guidelines for administering magnesium sulphate prior to a preterm birth.

While magnesium sulphate has not been used consistently throughout the years in which Professor Crowther and colleagues undertook their research work, there is a fascinating suggestion in this Report that the prevalence of cerebral palsy may actually be starting to wane in South Australia. Before we get too excited, however, we must first decide whether there are other explanations for the decreases evident in Table 2.

One of the most difficult and time-consuming activities of any Register is to make sure that every case of interest has been identified and the diagnosis confirmed. At a time when privacy considerations are making it progressively more difficult for health-care organisations to share information, - and easier for individuals to deny access to that information, a disappointing interpretation of Table 2 would be that it simply reflects the increasing difficulty of identifying every case. If this explanation were correct, however, then one might expect ascertainment problems to affect the number of children with mild disabilities, (and who typically have less contact with healthcare providers) to be much more strongly affected than the numbers of children with moderate and severe disabilities. But Table 7 in this Report suggests that the prevalence of children with very severe limitations in motor function has decreased in similar proportion to those who can walk without restriction! Only continued monitoring by the Register, with close attention paid to finding every single case, can answer this question – but it is both exciting and refreshing to know that recent advances in antenatal medical treatment have the potential to prevent a condition for which there is no known cure.

The South Australian Cerebral Palsy Register is deeply indebted to the Paediatric rehabilitation specialists (and other specialists) who notified 53% of the cases identified in this Report, and to Novita Children's Services who identified 21%. The South Australian Birth Defects Register also identified 21% of the new cases.

I would like to conclude with special thanks to the Register Staff who possess that special eye for detail, and that hunger to track down every potential case, which are the essential ingredients of a successful Register.

A/Prof Peter Baghurst,
Head, Public Health Research Unit,
Women's and Children's Hospital,
Children Youth and Women's Health Service

South Australian Cerebral Palsy Register Staff

South Australian Cerebral Palsy Register Staff

Left to Right: Associate Professor Peter Baghurst, Associate Professor Annabelle Chan, Mrs Phillipa van Essen, Ms Heather Scott, Mrs Rosie Rice and Dr Catherine Gibson

Executive Summary

The SA Cerebral Palsy Register is a population-based collection of information on children with cerebral palsy born in South Australia in a population with an average of 18,468 live births per year (1993 – 2004). This report presents information for children with cerebral palsy born in the years 1993 – 2004.

The term cerebral palsy describes a group of permanent and non-progressive disorders of movement and posture that manifest early in life and result from a defect or lesion of the immature brain. Individuals with cerebral palsy have lifelong motor disabilities, frequently associated with intellectual disability, epilepsy and visual and hearing impairment. It is the most common chronic motor disability of childhood and it places a large emotional and financial burden on those affected and their families.

The types of cerebral palsy are based on clinical features. The types are spastic quadriplegia, spastic triplegia, spastic diplegia, spastic hemiplegia, monoplegia, ataxia and dyskinesia.

Children with cerebral palsy are ascertained through notifications to the SA Birth Defects Register from rehabilitation specialists, Novita Children's Services, other paediatricians and the Neonatal Long Term Follow-up Programs of Flinders Medical Centre and Children, Youth and Women's Health Service.

At around five years of age, a comprehensive medical history is obtained and the children are offered a formal clinical assessment to ensure that the diagnosis of cerebral palsy is correct; to document the type and severity of the disorder; and to define the disabilities experienced by the child. Parental consent is obtained for children to participate in the clinical assessment at five years of age.

For the years 1993 – 2004, 428 children with cerebral palsy were ascertained. At 31st December 2009, 91% (301/329) of children who were eligible to be assessed had completed their comprehensive clinical assessment, at around five years of age. For various reasons, there were 99 cases that were not able to complete a clinical assessment (14 had died before the age of five years, 26 did not want to participate in the register, 58 were not able to be contacted and one child was too old for the assessment).

The maximum and minimum prevalence of cerebral palsy between 1993 and 2004 is presented, per 1,000 live births. The minimum prevalence represents the cases confirmed by examination at five years, and the maximum prevalence reflects the total ascertained cases. Between 1993 and 2004 the minimum prevalence of cerebral palsy was 1.36 per 1,000 live births, and the maximum was 1.93 per 1,000 live births.

Paediatric rehabilitation specialists notified 48% of cases, Novita Children's Services 21%, the South Australian Birth Defects Register 21%, other sources 5% and other specialists 4%.

Hemiplegia (34%) and diplegia (33%) were the most common forms of cerebral palsy, followed by quadriplegia (22%), triplegia (3%) and ataxia (3%). Dyskinetic athetoid, monoplegia, hypotonia and dyskinetic dystonic were less common forms of the disorder. In 1% of cases, the type of cerebral palsy was unknown.

50.5% of affected children were born at term (≥ 37 weeks gestation) and 49.3% were born prematurely (< 37 weeks gestation).

The association of premature birth with cerebral palsy is demonstrated by a prevalence of 59.2 cases per 1,000 live births at 23-27 weeks gestation, compared with 1.1 cases per 1,000 live births at term (≥ 37 weeks gestation).

54% of affected children had a birth weight ≥2500 grams, 19% weighed 1500-2499 grams, 13% weighed 1000-1499 grams, and 14% weighed <1000 grams.

Consistent with the association with premature birth, low birth weight is also strongly associated with cerebral palsy; the prevalence being 65.8 cases per 1,000 live births with birth weights in the range 500-999 g, but only 0.8 per 1,000 among live births in the range 3500-3999 g.

Of the 301 children assessed, 67% had relatively good levels of gross motor function (levels I and II), while 32% had more severely affected gross motor function (levels III-V) and in 1% of cases the level of motor function was unknown.

30% of the assessed children born between 1993 and 2004 had impaired intellectual ability (mild to severe disability), 39% had impaired vision and 12% had impaired hearing.

Thanks to Notifiers / Acknowledgements

Thanks to Notifiers

We wish to thank all notifiers to the Register, including paediatricians, staff of Novita Children's Services of South Australia and the families of children with cerebral palsy. In particular, we would like to thank the staff of the Paediatric Rehabilitation Service of the Children, Youth and Women's Health Service.

We would especially like to thank and acknowledge the Rehabilitation Specialists and paediatricians who complete the Data Collection Form: Dr Ray Russo, Dr James Rice, Dr Rosa Zarrinkalam, Dr Phil Egan, Dr Andrew Tidemann, Dr Kathy Lee, Dr Deirdre White and Dr Peter Flett.

We acknowledge the ongoing assistance of the staff of the Medical Records Departments at Flinders Medical Centre, Lyell McEwin Health Service and Children, Youth and Women's Health Service.

Special thanks must also go to staff of the Pregnancy Outcome Statistics Unit of the Epidemiology Branch, SA Health, for their invaluable assistance.

Acknowledgements

We would like to thank all those people and organisations who have contributed to the South Australian Cerebral Palsy Register during its planning stage and since its inception; their ongoing support and practical contribution is much appreciated.

Special thanks are due to:

- Novita Children's Services (formerly Crippled Children's Association of South Australia)
- The Community Accommodation and Respite Agency (CARA)
- Paediatricians from specialist centres and in private practice
- Staff of the Paediatric Rehabilitation Service of the Children, Youth and Women's Health Service
- Long term follow up program coordinators from the Children, Youth and Women's Health Service and Flinders Medical Centre
- Staff of the South Australian Clinical Genetics Service of the Children, Youth and Women's Health Service

Whilst there are many who have contributed to the South Australian Cerebral Palsy Register, there would be no Register without the kind cooperation of the families of affected children. These families live with the effects of cerebral palsy on a daily basis. Once again the Register owes them a debt of gratitude for allowing us to enter their lives and for their insightful comments over the year.

What is Cerebral Palsy?

Cerebral palsy is a term of convenience applied to a group of motor disorders of central origin defined by a clinical description [1]. It covers a range of cerebral disorders that result in childhood motor impairment. The impairment must stem from non-progressive malfunction of the brain (rather than the spinal cord or muscles).

Cerebral palsy affects an individual's ability to control movement and posture. Palsy is an expression used to describe paralysis. A more accurate description of the muscle symptoms might be weakness (paresis) and an inability to make voluntary movements and to suppress involuntary ones.

Unlike many other disorders that affect the motor system, cerebral palsy is not progressive – it does not get worse with time. In fact, family nurturing, therapy and education can result in improvements in functional outcomes and in quality of life.

Approximately 2 in every 1,000 children born in South Australia have cerebral palsy, which equates to approximately 37 newly diagnosed children each year in 1993 – 2004. Despite improvements in antenatal, intrapartum and postnatal care, there has been little change in the incidence of cerebral palsy.

Children affected by cerebral palsy will have lifelong disabilities and continue to represent a significant proportion of the children and young adults with disabilities in the community. Severity can vary greatly, from minor awkwardness to severe multiple disabilities.

Cerebral palsy can be associated with other disorders, such as epilepsy, learning difficulties, and problems with sight and hearing. There can be difficulties with communication, resulting from multiple disabilities, including problems controlling the speech muscles, intellectual disabilities, and poor hearing.

Orthopaedic complications are common, often requiring ongoing care. Most children with cerebral palsy survive to adulthood, although severely affected children may have a reduced life expectancy.

Whilst there is no single cause for cerebral palsy, we do know that there are a number of perinatal factors which are associated, both individually and in combination, with an increased risk of cerebral palsy. These include:

- Prematurity and very low birth weight
- Maternal infections, such as cytomegalovirus (CMV), group B streptococcus and rubella
- Feto-maternal haemorrhage
- Hypoxia during labour and delivery
- The use of certain drugs during pregnancy, such as cocaine
- Excessive alcohol intake during pregnancy
- Metabolic problems in the newborn period, such as severe jaundice and hypoglycaemia

Recent research using blood specimens from babies obtained soon after birth has demonstrated an association between evidence of exposure to infection before birth, markers of prenatal inflammation and certain inherited susceptibilities to thrombosis, and the subsequent development of cerebral palsy [2,3,4,5].

There are also events occurring in early childhood (before 2 years of age) which may result in "acquired" cerebral palsy. These include:

- Near drowning
- Brain hypoxia from any cause
- Some brain tumours
- Head injuries: accidental and non-accidental
- Severe infection involving the brain, such as meningitis or encephalitis
- Cerebrovascular accidents (strokes)
- Non-accidental injuries
- Complications of inborn errors of metabolism

Funding / Administration / Ethical Approval / Legislative Framework

Funding

The Register is funded by the Children, Youth and Women's Health Services, with additional funding from Novita Children's Services.

Administration

The Register is based at the Children, Youth and Women's Health Service and is located administratively within the Public Health Research Unit. Its location enables the Register to draw on the experience and expertise of the Paediatric Rehabilitation Service and Neurology Department of the Children, Youth and Women's Health Service. The Specialist Advisors to the Register are drawn from members of these departments and Novita Children's Services.

Ethics Approval

The establishment of the South Australian Cerebral Palsy Register was approved by the Human Research Ethics Committee of the Children, Youth and Women's Health Service, the North Western Adelaide Health Service and Flinders Medical Centre. Continuing oversight of the ethics of the activities of this state-based Register is carried out by the SA Health, Human Research Ethics Committee.

Legislative framework

The South Australian Cerebral Palsy Register is part of the South Australian Birth Defects Register and operates under the provisions of the South Australian Health Commission (Pregnancy Outcome Statistics) Regulations of 1999. These Regulations require the notification of all congenital abnormalities diagnosed before the child's fifth birthday. The historical data collected under the South Australian Health Commission Act continue to be subject to the privacy provisions of that Act. More recent data collected under the new *Health Care Act* 2008 are subject to slightly modified privacy provisions. This legislation does not alter the confidentiality guidelines under which the Register functions.

Part 7 of the *Health Care Act* 2008 allows the Register to carry out research into cerebral palsy while making provision for maintaining the confidentiality of participants.

Aims of the Register

The aims of the South Australian Cerebral Palsy Register are to:

- Determine and monitor the prevalence of cerebral palsy in South Australia
- Gather information about affected children that may provide clues to the causes of cerebral palsy
- Document the severity and range of disabilities experienced by children with cerebral palsy
- Provide information to help plan facilities for affected children
- Act as a source of information about cerebral palsy, for both families and the community
- Improve community and professional awareness of cerebral palsy, including its causes and outcomes
- Provide a resource for research into cerebral palsy
- Contribute to mortality and morbidity studies of cerebral palsy
- Contribute deidentified data to the Australian Cerebral Palsy Register

How the Register works

Ascertainment

Notifications of children with cerebral palsy come from a variety of sources, including Novita Children's Services, paediatricians, physiotherapists, neurologists and occupational therapists. The main sources of notification come from the Children, Youth and Women's Health Service, Novita Children's Services and Paediatric Rehabilitation Specialists. The Register encourages multiple notifications as a means of ensuring complete ascertainment of affected children.

All notifications are received by the SA Birth Defects Register. Once notified, the Cerebral Palsy Register approaches families through a clinician known to them, who will be one of the physicians working with the Register or the child's paediatrician. Information about the Register and its aims is sent to the managing clinician and an invitation to become involved with the Register is extended to the family. The managing clinician is also sent a package containing a family information leaflet and a "consent to contact" form. Once the clinician has communicated with the family, the package can be forwarded to them. The family can then respond directly to the Register.

The "consent to contact" form gives the Register permission to contact the family, allowing the Register to give the family detailed information about its aims and functions. Families are free to refuse to participate in the Register. Those who agree to participate may, at any time, ask to be withdrawn from the Register with the knowledge that this will not affect any aspect of the child's medical care. Children included on the Register are given a full clinical assessment at around school age, performed by a paediatrician working with the Register. By then, it is clear whether or not cerebral palsy is present, and the type and severity can be determined more accurately. Disabilities in addition to the motor disability can also be documented.

The purpose of the assessment is to collect data on the type of cerebral palsy, its severity and any associated disabilities. Information about learning problems, epilepsy, hearing, vision and the use of mobility aids is also collected. A brief family history is obtained to determine if cerebral palsy, intellectual disability or epilepsy has occurred within the immediate or extended family in the past. Information relating to the perinatal period is also collected.

Consent is obtained from each affected family to allow the Register Officer to access information from a variety of sources. The Register works closely with the Pregnancy Outcome Statistics Unit, medical records departments and clinicians in order to ensure accurate and complete data for each child.

Data Storage and Confidentiality

The collected data are stored on computer using unique identifier codes to maintain participant confidentiality. All information and participant files are backed up regularly to prevent data loss. The Register does not release identified information about a participant without the written consent of his or her parents or guardian.

Inclusion and Exclusion Criteria

The Register includes children who have a motor impairment, manifested early in life, which is the result of static cerebral pathology. The Register also collects cases of cerebral palsy acquired after the neonatal period (after the first month and before 2 years of age).

The following disorders are excluded: neurodegenerative conditions, neuromuscular disorders, neural tube defects, tumours, hypotonia occurring in isolation or with intellectual disability, many genetic syndromes and most inborn errors of metabolism. There are many conditions where a decision about inclusion or exclusion can be difficult, and guidance is provided by Badawi *et al* (1998) [1].

Participation in the Australian Cerebral Palsy Register

The Register has been involved in the establishment of the Australian Cerebral Palsy Register (ACPR). We have contributed to the development of the Australian Cerebral Palsy Register Agreement and Working Guidelines which have been developed to set out the methods and parameters required to facilitate the transfer of anonymous information about South Australian children with cerebral palsy to the ACPR as part of a national strategy to monitor the frequency of cerebral palsy and to undertake research into its causes.

Register Activities 2008 / 2009

During 2008 and 2009, Register staff have:

- Refined the data set, database and systems for analysis and interpretation of data.
- Participated in the South Australian Cerebral Palsy Research Group, which is investigating whether variations in genes (involved in blood clotting, inflammation and defence against infection) and perinatal infection are associated with cerebral palsy. This has been a highly productive collaboration, with many articles published in the international peer-reviewed literature in 2005-2009.
- Responded to general queries from representatives in other Australian States and Territories regarding the establishment of a cerebral palsy register, and requests for information on consent forms, information pamphlets and legislation.
- Responded to requests for information from the community and tertiary students who required statistics on the prevalence of cerebral palsy, types of cerebral palsy and associated disabilities in South Australia.
- Collaborated with staff from the Paediatric Rehabilitation Service of the Children, Youth and Women's Health Service on a study investigating notified cases of cerebral palsy who were subsequently found not to have cerebral palsy at or before the Register's five year clinical review.
- Collaborated with staff from the Paediatric Rehabilitation Service of the Children, Youth and Women's Health Service on a study examining motor profiles and associated health and developmental status of children with bilateral limb involvement on the SACPR.
- Worked on a project reviewing all the information collected from the 5 year clinical assessments between 1993 and 2003.
- Contributed to the development of the Australian Cerebral Palsy Register (ACPR) Agreement and Working Guidelines Document. This document has been developed to set out the parameters for the Australian CP Register.
- Represented South Australia on the Australian Cerebral Palsy Register Policy Group, participating in meetings and teleconferences to assist in the operation of the Australian Cerebral Palsy Register.
- Attended and participated in the 3rd International Cerebral Palsy Conference in Sydney, Australia (February 2009). This conference included a World Cerebral Palsy Register Congress, where Register staff from around the world met to discuss ongoing management and issues surrounding CP Registers.

Prevalence of cerebral palsy 1993 – 2004

Tables 1-6 and Figures 1-4 present data about children with cerebral palsy notified to the South Australian Birth Defects Register.

Table 1: SA children with cerebral palsy ascertained by the Register 1993-2004

Group	1993-1999	2000	2001	2002	2003	2004	1993-2004	
	No.	No.	No.	No.	No.	No.	No.	(%)
Assessment completed at 5 years	228	26	16	13	10	8	**301**	**(70.3)**
Awaiting assessment	0	1	2	0	5	6	**14**	**(3.3)**
Consents not complete	2	1	3	2	5	1	**14**	**(3.3)**
Medical reports only	55	11	8	5	3	3	**85**	**(19.9)**
Deceased prior to 5 years	10	1	1	1	1	0	**14**	**(3.3)**
Total*	**295**	**40**	**30**	**21**	**24**	**18**	**428**	**(100)**

*Does not include interstate or overseas births: three in 1993, five in 1994, six in 1995, six in 1996, five in 1997, six in 1998, four in 1999, two in 2000, five in 2001, four in 2002, one in 2003 and five in 2004 (total of 52)

Four hundred and twenty eight children, born in South Australia (SA) in 1993 – 2004, had been notified to the Register by 31st December 2009. Over this same period, the Register also received 52 notifications of cases born outside SA. A comprehensive clinical assessment at around school age has been completed for 301 (91%) of the 329 children eligible for assessment (see Table 1).

Experience has shown that some of the notified cases will not have cerebral palsy when assessed at five years of age; for this reason minimum and maximum prevalence estimates have been calculated for the years 1993 – 2004 (Table 2). The maximum reflects total ascertained cases and the minimum estimate is based on confirmed cases only

Table 2: Cerebral palsy ascertainment and prevalence 1993 - 2004

Year of birth	Live births	Ascertained cases	Confirmed cases*		Prevalence / 1,000# max / min	
1993-1999	133,522	295	228	(77.3%)	2.21	1.71
2000	17,765	40	26	(65.0%)	2.25	1.46
2001	17,584	30	16	(53.3%)	1.71	0.91
2002	17,623	21	13	(61.9%)	1.19	0.74
2003	17,710	24	10	(41.7%)	1.36	0.56
2004	17,409	18	8	(44.4%)	1.03	0.46
1993-2004	**221,613**	**428**	**301**	(70.4%)	**1.93**	**1.36**

*Have completed comprehensive clinical assessment at around 5 years of age
#Rate per 1,000 live births. Minimum rate represents confirmed cases and maximum rate reflects total ascertained cases

Notifiers to the Register

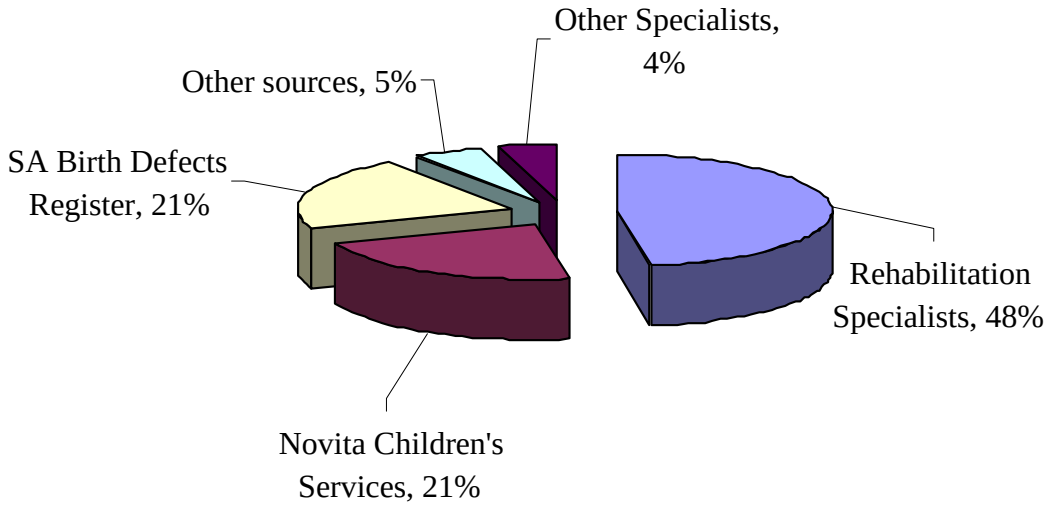
The sources of notification of children with cerebral palsy born in SA between 1993 and 2004 are shown in Table 3. Over this period, 90% of the notifications came from three main sources, Rehabilitation specialists, Novita Children's Services and the South Australian Birth Defects Register.

Table 3: Notifiers to the Register 1993 - 2004

Notifier*	**1993-1999**	**2000**	**2001**	**2002**	**2003**	**2004**	**1993-2004**	
	No.	**No.**	**No.**	**No.**	**No.**	**No.**	**No.**	**(%)**
Rehabilitation specialists	123	26	13	13	16	13	**204**	**(47.7)**
Novita Children's Services	88	0	2	0	1	0	**91**	**(21.3)**
SA Birth Defects Register	58	11	11	4	7	0	**91**	**(21.3)**
Other sources#	14	1	3	3	0	2	**23**	**(5.4)**
Other specialists	12	2	1	1	0	3	**19**	**(4.4)**
Total	**295**	**40**	**30**	**21**	**24**	**18**	**428**	**(100)**

* The Register encourages multiple notifications
# Other sources include allied health services such as physiotherapy and occupational therapy

Figure 1: Notifiers to the Register 1993 – 2004



Type of cerebral palsy

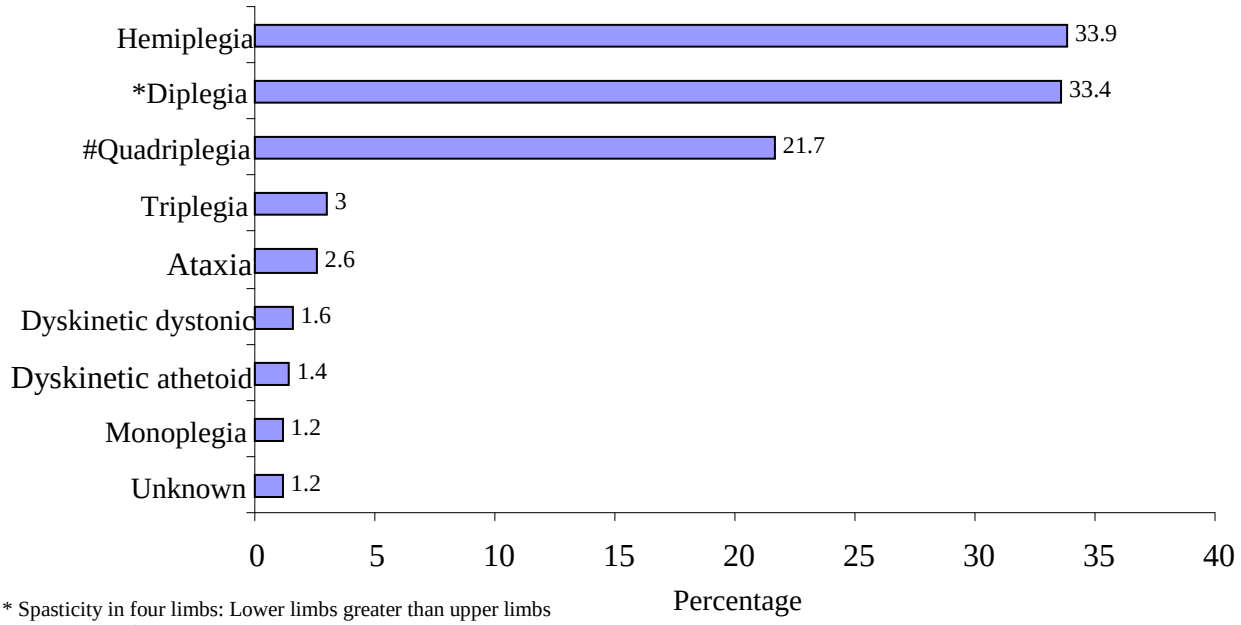
The different types of cerebral palsy for children born in SA between 1993 and 2004 are shown in Table 4. The most common forms of cerebral palsy were hemiplegia, diplegia and quadriplegia, accounting for 89% of all cases.

Table 4: Type of cerebral palsy 1993 - 2004

Type of cerebral palsy	1993-1999	2000	2001	2002	2003	2004	1993-2004	
	No.	No.	No.	No.	No.	No.	No.	(%)
Hemiplegia	98	16	10	8	6	7	**145**	**(33.9)**
Diplegia*	101	12	11	4	9	6	**143**	**(33.4)**
Quadriplegia#	61	9	8	6	5	4	**93**	**(21.7)**
Triplegia	12	0	0	1	0	0	**13**	**(3.0)**
Ataxia	9	2	0	0	0	0	**11**	**(2.6)**
Dyskinetic athetoid	6	0	0	0	0	1	**7**	**(1.6)**
Dyskinetic dystonic	1	1	1	1	2	0	**6**	**(1.4)**
Monoplegia	2	0	0	1	2	0	**5**	**(1.2)**
Unknown	5	0	0	0	0	0	**5**	**(1.2)**
Total	**295**	**40**	**30**	**21**	**24**	**18**	**428**	**(100)**

* Spasticity in four limbs: Lower limbs greater than upper limbs
# Spasticity in four limbs: Upper limbs greater than or equal to lower limbs

Figure 2: Type of cerebral palsy 1993 – 2004



* Spasticity in four limbs: Lower limbs greater than upper limbs
# Spasticity in four limbs: Upper limbs greater than or equal to lower limbs

Gestational age at birth

The gestational ages at birth for cases of cerebral palsy born in SA between 1993 and 2004 are shown in Table 5. There were 50.5% of cases born at term (≥ 37 weeks gestation).

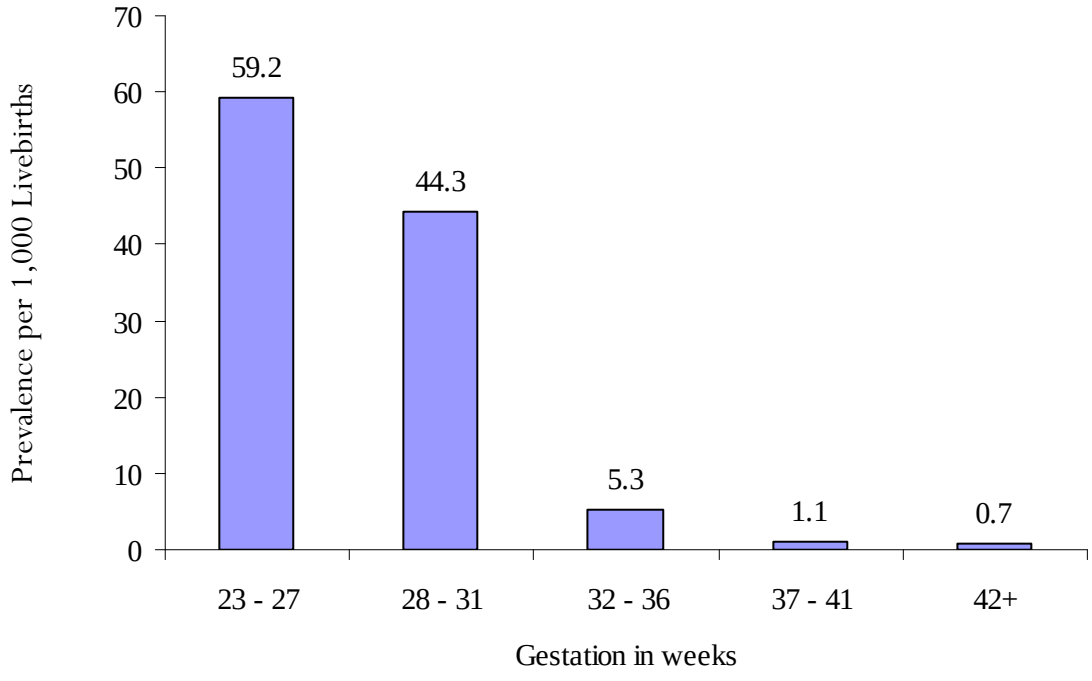
The prevalence of cerebral palsy by gestational age at birth 1993 – 2004 is also demonstrated in Table 5 and Figure 3 illustrating the association between cerebral palsy and prematurity (<37 weeks gestation), especially gestations <32 weeks.

Table 5: Cerebral palsy by gestational age at birth 1993 - 2004

Gestation Weeks*	1993-1999 No.	2000 No.	2001 No.	2002 No.	2003 No.	2004 No.	1993-2004 No.	1993-2004 (%)	1993-2004 Prevalence#
23^ – 27	51	3	3	0	2	0	59	(13.8)	59.2
28 – 31	57	9	3	3	4	3	79	(18.5)	44.3
32 – 36	49	9	7	3	1	4	73	(17.1)	5.3
37 – 41	136	19	17	15	17	10	214	(50.0)	1.1
42+	2	0	0	0	0	0	2	(0.5)	0.7
Unknown	0	0	0	0	0	1	1	(0.2)	-
Total	**295**	**40**	**30**	**21**	**24**	**18**	**428**	**(100)**	**1.9**

* Gestation at birth (best clinical estimate) in weeks, ^ There were no cases below 23 weeks, # Prevalence per 1,000 live births

Figure 3: Cerebral palsy by gestational age at birth 1993 – 2004



Birth weight

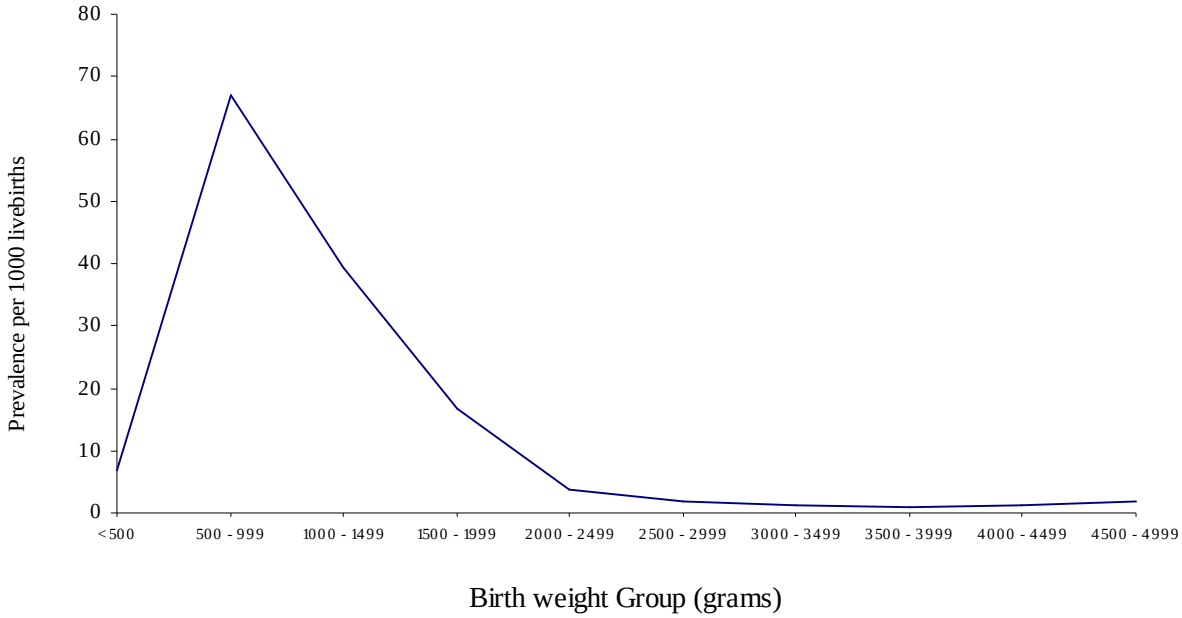
The prevalences of cerebral palsy per 1,000 live births, by birth weight, are provided in Table 6 and Figure 4 for the period 1993 – 2004. They demonstrate a strong relationship between cerebral palsy and low birth weight. As birth weight increases, the prevalence of cerebral palsy decreases, from 65.8 per 1,000 live births at 500 – 999g to 0.8 per 1,000 live births at 3,500 – 3,999g. Due to high mortality in the <500g birth weight group the prevalence of cerebral palsy appears relatively low at 6.7 per 1,000 live births.

Table 6: Cerebral palsy by birth weight 1993 - 2004

Birth weight range (grams)	1993-1999 No.	2000 No.	2001 No.	2002 No.	2003 No.	2004 No.	1993-2004 No.	1993-2004 (%)	1993-2004 Prevalence[#]
< 500	1	0	0	0	0	0	1	(0.2)	6.7
500 – 999	49	4	3	0	3	0	59	(13.8)	65.8
1,000 – 1,499	43	5	2	2	2	1	55	(12.9)	39.3
1,500 – 1,999	31	6	3	1	3	4	48	(11.2)	16.7
2,000 – 2,499	23	3	2	3	2	1	34	(7.9)	3.8
2,500 – 2,999	44	5	5	1	1	5	61	(14.3)	1.8
3,000 – 3,499	53	9	8	2	5	5	82	(19.2)	1.0
3,500 – 3,999	32	6	4	6	4	1	53	(12.4)	0.8
4,000 – 4,499	15	1	2	5	3	0	26	(6.1)	1.2
4,500 – 4,999	4	1	1	1	1	0	8	(1.9)	2.0
Unknown	0	0	0	0	0	1	1	(0.2)	-
Total	**295**	**40**	**30**	**21**	**24**	**18**	**428**	**(100)**	**1.9**

[#] Prevalence per 1,000 live births

Figure 4: Cerebral palsy by birth weight 1993 - 2004



Level of motor function

Tables 7-11 and Figures 5-6 present additional data collected at the 5 year assessment. At 31st December 2009, 301 (91%) of children who were eligible (329) to be assessed had completed their comprehensive clinical assessment, at around five years of age. For various reasons, there were 99 cases that were not able to complete a clinical assessment (14 had died before the age of five years, 26 did not want to participate in the register, 58 were not able to be contacted and one child was too old for the assessment).

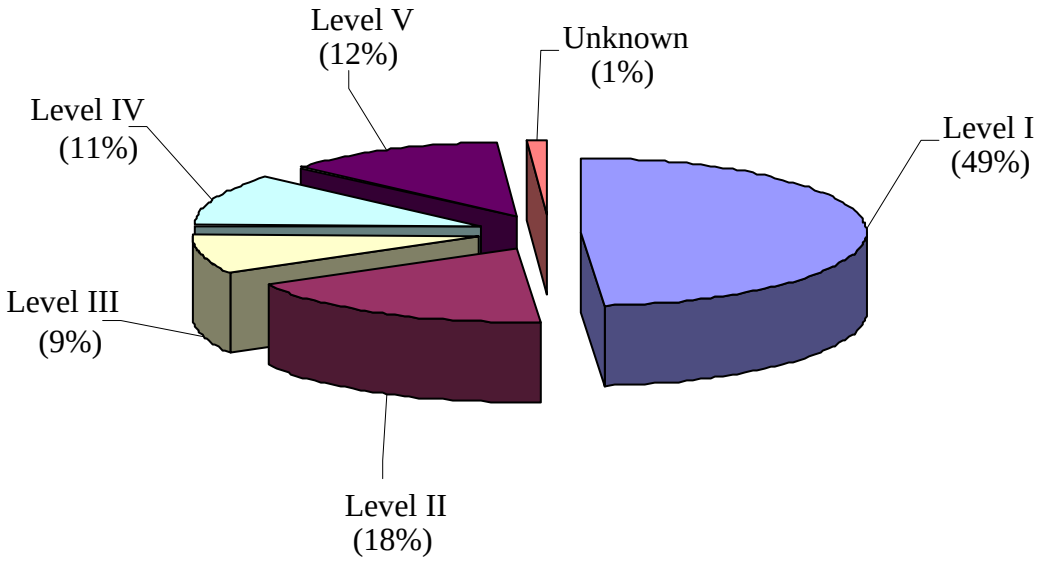
Table 7 and Figure 5 show the level of motor function, using the Gross Motor Function Classification System (GMFCS) for children with cerebral palsy born 1993 – 2004. The majority of children had less restricted motor function Levels I and II i.e. walking without restriction (49%) and walking without assistive devices (18%) respectively, while 32% were more severely affected in their motor function Levels III – V, with limitations or severe limitations to self-mobility. A definition of motor function appears in Appendix 1.

Table 7: Gross Motor Function Classification 1993 - 2004

Level of function*	**1993-1999**	**2000**	**2001**	**2002**	**2003**	**2004**	**1993-2004**	
	No.	**No.**	**No.**	**No.**	**No.**	**No.**	**No.**	**(%)**
Level I	110	18	7	6	3	3	147	(48.8)
Level II	45	1	2	2	2	3	55	(18.3)
Level III	22	1	1	0	1	1	26	(8.6)
Level IV	21	5	2	3	2	1	34	(11.3)
Level V	29	0	4	2	2	0	37	(12.3)
Unknown	1	1	0	0	0	0	2	(0.7)
Total	**228**	**26**	**16**	**13**	**10**	**8**	**301**	**(100)**

* Definition of Gross Motor Function Classification System (GMFCS) appears in Appendix 1

Figure 5: Gross Motor Function Classification 1993 - 2004



Co-morbidities

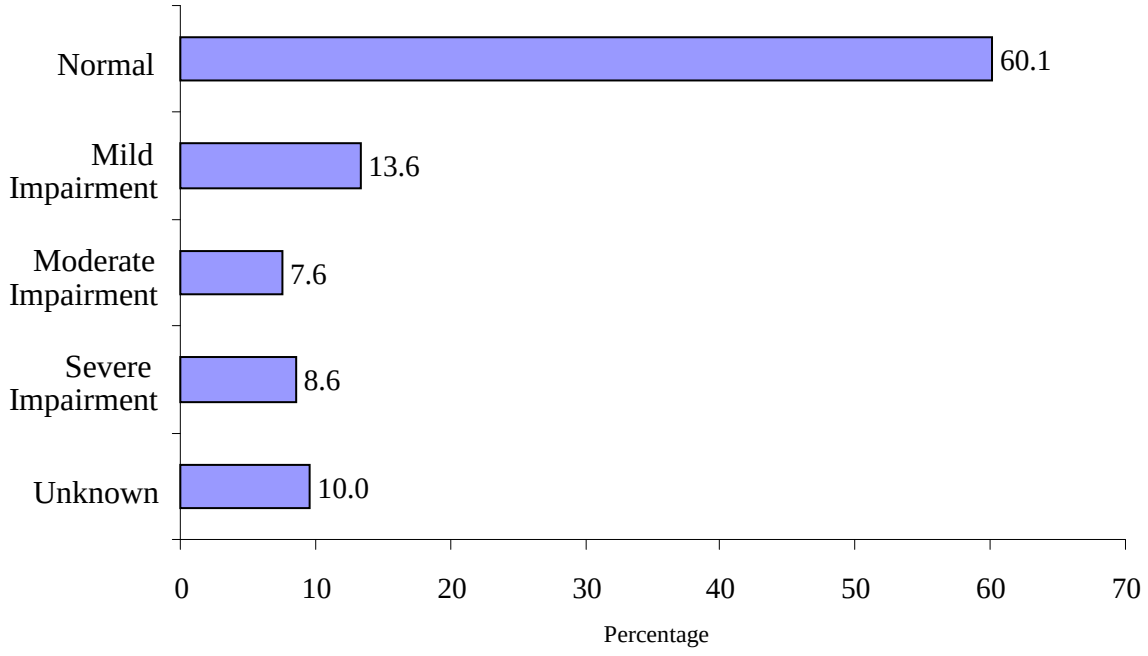
Table 8 and Figure 6 illustrate the level of intellectual ability for children with cerebral palsy born 1993 – 2004. In 2009, SA adopted the categories used by the Australian Cerebral Palsy Register to illustrate intellectual ability. They show that 60% of children with cerebral palsy had normal intellectual ability (i.e. an intelligent quotient score of ≥ 70), 14% had mild intellectual disability, and 16% had moderate to severe intellectual disability.

Table 8: Intellectual ability 1993 - 2004

Intellectual ability #	**1993-1999**	**2000**	**2001**	**2002**	**2003**	**2004**	**1993-2004**	
	No.	**No.**	**No.**	**No.**	**No.**	**No.**	**No.**	**(%)**
Normal (IQ ≥ 70)	140	19	8	7	2	5	181	(60.1)
Mild Impairment (IQ 50-69)	37	2	1	1	0	0	41	(13.6)
Moderate Impairment (IQ 35-49)	20	0	1	1	1	0	23	(7.6)
Severe Impairment (IQ < 35)	19	1	4	1	1	0	26	(8.6)
Probably greater than borderline: severity uncertain	0	0	0	0	0	0	0	(0.0)
Unknown	12	4	2	3	6	3	30	(10.0)
Total	**228**	**26**	**16**	**13**	**10**	**8**	**301**	**(100)**

Various scales, appropriate to the child's age and physical abilities, were used to assess intellectual ability

Figure 6: Intellectual ability# 1993 – 2004



Various scales, appropriate to the child's age and physical abilities, were used to assess intellectual ability

Co-morbidities

The frequency of visual problems in children with cerebral palsy born 1993 – 2004 is shown in Table 9. 52% of children had normal vision and 39% had impaired vision. Impaired vision includes: strabismus +/- amblyopia, refractive error, cortical blindness, nystagmus and optic nerve abnormalities.

Table 9: Visual problems 1993 – 2004

Visual problems*	**1993-1999**	**2000**	**2001**	**2002**	**2003**	**2004**	**1993-2004**	
	No.	**No.**	**No.**	**No.**	**No.**	**No.**	**No.**	**(%)**
Normal	115	18	9	5	6	4	157	(52.2)
Impaired	90	7	6	7	4	4	118	(39.2)
Unknown	23	1	1	1	0	0	26	(8.6)
Total	**228**	**26**	**16**	**13**	**10**	**8**	**301**	**(100)**

* Impaired vision includes: strabismus +/- amblyopia, refractive error, cortical blindness, nystagmus and optic nerve abnormalities

The frequency of hearing problems in children with cerebral palsy born 1993 – 2004 is shown in Table 10. 84% of children had normal hearing and 12% had impaired hearing. In 4% of cases, the level of hearing was unknown. Impaired hearing includes: sensorineural and/or conductive hearing loss determined by audiology.

Table 10: Hearing problems 1993 – 2004

Hearing problems*	**1993-1999**	**2000**	**2001**	**2002**	**2003**	**2004**	**1993-2004**	
	No.	**No.**	**No.**	**No.**	**No.**	**No.**	**No.**	**(%)**
Normal	189	23	14	9	9	8	252	(83.7)
Impaired	30	1	2	3	1	0	37	(12.3)
Unknown	9	2	0	1	0	0	12	(4.0)
Total	**228**	**26**	**16**	**13**	**10**	**8**	**301**	**(100)**

* Impaired hearing includes: sensorineural and/or conductive hearing loss determined by audiology

Co-morbidities

Table 11 presents the types of cerebral palsy with associated co-morbidities of hearing, vision, epilepsy and intellectual disability. Some differences can be seen, for example between quadriplegic and diplegic types of cerebral palsy, especially with respect to vision, epilepsy and intellectual disability. The proportions of these co-morbidities were found to be significantly higher in those with quadriplegia. Some of the children have multiple co-morbidities, therefore the totals do not add to 100%. Intellectual disability is defined as mild to severe disability (IQ <70).

Table 11: Type of cerebral palsy and associated disabilities 1993 – 2004

Type of cerebral palsy	Total in category	Hearing impairment		Vision impairment		Epilepsy		Intellectual disability*	
		No.	(%)	No.	(%)	No.	(%)	No.	(%)
Hemiplegia	106	15	(14.2)	33	(31.1)	28	(26.4)	23	(21.6)
Diplegia	114	12	(10.5)	38	(33.3)	17	(14.9)	26	(22.8)
Quadriplegia	45	7	(15.6)	31	(68.9)	27	(60.0)	28	(62.2)
Triplegia	12	1	(8.3)	3	(25.0)	3	(25.0)	4	(33.3)
Ataxia	8	1	(12.5)	5	(62.5)	2	(25.0)	5	(62.5)
Dyskinetic athetoid	6	0	0	2	(33.3)	1	(16.7)	2	(33.3)
Dyskinetic dystonic	7	1	(14.3)	5	(71.4)	2	(28.6)	2	(28.6)
Monoplegia	2	0	0	0	0	0	0	0	0
Unspecified type	1	0	0	1	(100)	1	(100)	0	0
Total (% of total)[#]	**301**	**37**	**(12.3)**	**118**	**(39.2)**	**81**	**(26.9)**	**90**	**(29.9)**

* Known to have mild to severe intellectual disability, IQ <70
[#] Some of the children have multiple co-morbidities, therefore the totals do not add up to 100%

Presentations / Conferences / Publications

Conference presentations utilising data from the Register 2008/2009

1. McMichael, GL, CS Gibson, AH MacLennan, PN Goldwater, EA Haan, K Priest, GA Dekker. Apolipoprotein E genotype is not associated with cerebral palsy. International Cerebral Palsy Conference, Sydney, Australia 2009 (oral presentation).
2. R Zarrinkalam, R Russo, CS Gibson, P van Essen, A Peek, EA Haan. Inaccuracy rate of the diagnosis of cerebral palsy in South Australian Cerebral Palsy Register. International Cerebral Palsy Conference, Sydney, Australia 2009 (oral presentation).
3. P Flett, P van Essen, CS Gibson, R Russo. Clinical examinations at five years of age provide a "gold standard" for cerebral palsy registers. International Cerebral Palsy Conference, Sydney, Australia 2009 (oral presentation).
4. R Russo, R Atkins, EA Haan, M Crotty. Use of upper limb orthoses, adaptive equipment and therapy in a community based sample of children with hemiplegic cerebral palsy. International Cerebral Palsy Conference, Sydney, Australia 2009 (oral presentation).
5. M Djukic, CS Gibson, GL McMichael, AH MacLennan, PN Goldwater, EA Haan, K Priest, GA Dekker. Is there a genomic basis to cerebral palsy? Seeking candidate genes. Perinatal Society of Australia and New Zealand Conference, Gold Coast, Australia, 2008 (oral presentation).
6. GL McMichael, CS Gibson, AH MacLennan, PN Goldwater, EA Haan, K Priest, GA Dekker. Stored cord serum: is it an additional resource for research into fetal viral exposure? Perinatal Society of Australia and New Zealand Conference, Gold Coast, Australia, 2008 (poster presentation).
7. CS Gibson, AH MacLennan, PN Goldwater, EA Haan, K Priest, GA Dekker. Mannose binding lectin haplotypes are associated with cerebral palsy. Society for Maternal Fetal Medicine Conference, Dallas, USA, 2008 (oral presentation).
8. CS Gibson, AH MacLennan, PN Goldwater, EA Haan, K Priest, GA Dekker. Mannose binding lectin haplotypes are associated with cerebral palsy. Perinatal Society of Australia and New Zealand Conference, Melbourne, Australia, 2007 (oral presentation).

Conferences attended by Register staff 2008/2009

1. International Cerebral Palsy Conference, Sydney, February 2009 (CS Gibson, H Scott).
2. Australian Cerebral Palsy Register meeting, Sydney, February 2008 (CS Gibson, H Scott, A Peek).

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Annual Reports

1999 – 2009 Annual Reports of the South Australian Cerebral Palsy Register.

Appendix 1

GROSS MOTOR FUNCTION CLASSIFICATION SYSTEM IN CEREBRAL PALSY [6]

LEVEL I

Walks without restrictions: limitations in more advanced gross motor skills.

Before 2nd birthday: Infants move in and out of sitting and floor sit with both hands free to manipulate objects. Infants crawl on hands and knees, pull to stand and take steps holding onto furniture. Infants walk between 18 months and 2 years of age without the need for any assistive mobility device.

From age 2 to 4th birthday: Children floor sit with both hands free to manipulate objects. Movements in and out of floor sitting and standing are performed with adult assistance. Children walk as the preferred method of mobility without the need for any assistive mobility device.

From age 4 to 6th birthday: Children get into and out of and sit in a chair without the need for hand support. Children move from the floor and from chair sitting to standing without the need for objects for support. Children walk indoors and outdoors, and climb stairs. Emerging ability to run and jump.

From age 6 to 12: Children walk indoors and outdoors and climb stairs without limitations. Children perform gross motor skills including running and jumping but speed, balance and coordination are reduced.

LEVEL II

Walks without assistive devices: limitations walking outdoors and in the community.

Before 2nd birthday: Infants maintain floor sitting but may need to use their hands for support to maintain balance. Infants creep on their stomach or crawl on hands and knees. Infants may pull to stand and take steps holding onto furniture.

From age 2 to 4th birthday: Children floor sit but may have difficulty with balance when both hands are free to manipulate objects. Movements in and out of sitting are performed without adult assistance. Children pull to stand on a stable surface. Children crawl on hands and knees with a reciprocal pattern, cruise holding onto furniture and walk using an assistive mobility device as preferred methods of mobility.

From age 4 to 6th birthday: Children sit in a chair with both hands free to manipulate objects. Children move from the floor to standing and from chair sitting to standing but often require a stable surface to push or pull up on with their arms. Children walk without the need for any assistive mobility device indoors and for short distances on level surfaces outdoors. Children climb stairs holding onto a railing but are unable to run or jump.

From age 6 to 12: Children walk indoors and outdoors and climb stairs holding onto a railing but experience limitations walking on uneven surfaces and inclines, and walking in crowds or confined spaces. Children have at best only minimal ability to perform gross motor skills such as running and jumping.

Distinction between Levels I and II: *Compared with children in Level I, children in Level II have limitations in the ease of performing movement transitions; walking outdoors and in the community; the need for assistive mobility devices when beginning to walk; quality of movement; and the ability to perform gross motor skills such as running and jumping.*

LEVEL III

Walks with assistive mobility devices: limitations walking outdoors and in the community.

Before 2nd birthday: Infants maintain floor sitting when the low back is supported. Infants roll and creep forward on their stomachs.

From age 2 to 4th birthday: Children maintain floor sitting often by "W-sitting" (sitting between flexed and internally rotated hips and knees) and may require adult assistance to assume sitting. Children creep on their stomach or crawl on hands and knees (often without reciprocal leg movements) as their primary methods of self-mobility. Children may pull to stand on a stable surface and cruise short distances. Children may walk short distances indoors using an assistive mobility device and adult assistance for steering and turning.

From age 4 to 6th birthday: Children sit on a regular chair but may require pelvic or trunk support to maximise hand function. Children move in and out of chair sitting using a stable surface to push on or pull up with their arms. Children walk with an assistive mobility device on level surfaces and climb stairs with assistance from an adult. Children frequently are transported when travelling for long distances or outdoors on uneven terrain.

From age 6 to 12: Children walk indoors or outdoors on a level surface with an assistive mobility device. Children may climb stairs holding onto a railing. Depending on upper limb function, children propel a wheelchair manually or are transported when travelling for long distances or outdoors on uneven terrain.

Distinction between Levels II and III: *Differences are seen in the degree of achievement of functional mobility. Children in Level III need assistive mobility devices and frequently orthoses to walk, while children in Level II do not require assistive mobility devices after age 4.*

LEVEL IV

Self-mobility with limitations: children are transported or use power mobility outdoors and in the community.

Before 2nd birthday: Infants have head control but trunk support is required for floor sitting. Infants can roll to supine and may roll to prone.

From age 2 to 4th birthday: Children floor sit when placed, but are unable to maintain alignment and balance without the use of their hands for support. Children frequently require adaptive equipment for sitting and standing. Self-mobility for short distances (within a room) is achieved through rolling, creeping on stomach, or crawling on hands and knees without reciprocal leg movement.

From age 4 to 6th birthday: Children sit on a chair but need adaptive seating for trunk control and to maximise hand function. Children move in and out of chair sitting with assistance from an adult or a stable surface to push or pull up on with their arms. Children may at best walk short distances with a walker and adult supervision but have difficulty turning and maintaining balance on uneven surfaces. Children are transported in the community. Children may achieve self-mobility using a power wheelchair.

From age 6 to 12: Children may maintain levels of function achieved before age 6 or rely more on wheeled mobility at home, school and in the community. Children may achieve self-mobility using a power wheelchair.

Distinction between Levels III and IV: *Differences in sitting ability and mobility exist, even allowing for extensive use of assistive technology. Children in Level III sit independently, have independent floor mobility and walk with assistive mobility devices. Children in Level IV function in sitting (usually supported) but independent mobility is very limited. Children in Level IV are more likely to be transported or use power mobility.*

LEVEL V

Self-mobility is severely limited even with the use of assistive technology.

Before 2nd birthday: Physical impairment limits voluntary control of movement. Infants are unable to maintain antigravity head and trunk postures in prone and sitting. Infants require adult assistance to roll.

From age 2 to 12: Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At level V, children have no means of independent mobility and are transported. Some children achieve self-mobility using a power wheelchair with extensive adaptations.

Distinction between Levels IV and V: *Children in level V lack independence even in basic antigravity postural control. Self-mobility is achieved only if the child can learn how to operate an electrically powered wheelchair.* (Palisano et al, 1997)[6].

Appendix 2

South Australian Cerebral Palsy Register Family Information Sheet

About the South Australian Cerebral Palsy Register

Approximately one in every 500 children born in South Australia has cerebral palsy. It is now recognised that most cerebral palsy is due to factors present before labour begins and only a small fraction of cases stem from events occurring during labour and delivery. However, what actually causes cerebral palsy is not clear. In order to determine these factors and to improve services, it is important to collect information about children with cerebral palsy.

The South Australian Cerebral Palsy Register was established in 1998 with support from the Crippled Children's Association (now Novita Children's Services) and the Women's and Children's Hospital. It is located at the Women's and Children's Hospital in the Public Health Research Unit and is funded by Children, Youth and Women's Health Service, with support from Novita Children's Services, It is a collaboration between the Register, the Paediatric Rehabilitation Service (Children Youth and Women's Health Service) and Novita Children's Services.

The Register contributes de-identified data to the Australian Cerebral Palsy Register as part of a national strategy to monitor the frequency of cerebral palsy and to undertake research into its causes.

What is the purpose of the Register?

The purpose of the Cerebral Palsy Register is to collect information about South Australian children with cerebral palsy. This information will enable us to:

- Find out how many children in South Australia have cerebral palsy;
- Detect changes in the number of children with cerebral palsy;
- Carry out research into the causes of cerebral palsy;
- Identify the full range of disabilities experienced by children with cerebral palsy;
- Help in the planning of services for children with cerebral palsy and;
- Increase knowledge in the community about cerebral palsy.

What benefits are there for my child?

When your child is around school age he/she will receive a free, comprehensive medical assessment by a paediatric rehabilitation specialist. This assessment can take place at the Women's and Children's Hospital or Novita Children's Services during a scheduled appointment with one of the Paediatric Rehabilitation Doctors. Information gained by the specialist may be used to make recommendations to your child's doctor about services or treatment. You may choose to receive newsletters, a copy of the Annual Report and other information which may be of interest to you, including information about current research.

What other benefits may come from participating?

By participating you will also benefit other children with cerebral palsy by contributing to research. Such research may:

- Find ways of preventing cerebral palsy;
- Find possible causes;
- Lead to new treatments or ways of caring for children with cerebral palsy;
- Increase community awareness and recognition and;
- Lead to the development of new and/or improved services.

Who are the Register staff we will have contact with?

The person with whom you will have the most contact will be the Register Officer of the South Australian Cerebral Palsy Register. The paediatric rehabilitation specialists who work with the Register are Dr Ray Russo, Dr James Rice, Dr Andrew Tidemann, Dr Phil Egan, Dr Deepa Jeyaseelan, Dr Nick Ricci and Dr Rosa Zarrinkalam. One of these specialists will conduct your child's medical assessment.

Will information about my child be kept confidential?

All information contained on the South Australian Cerebral Palsy Register is strictly confidential. Only Register staff and those carrying out research in collaboration with the Register will use this information, and any such use must first be approved by a Human Research Ethics Committee. The Register provides anonymous information about South Australian children with cerebral palsy to the Australian Cerebral Palsy Register for research purposes only. *No information, which identifies your child, will be released to other people unless you give written permission.*

Are there any risks for my child?

There are no risks for your child associated with the medical assessment at five years of age. ***It will not involve*** the taking of blood samples or any other invasive tests.

Has the Register received approval from an Ethics Committee?

The South Australian Cerebral Palsy Register has received approval from the Human Research Ethics Committees of the Children, Youth and Women's Health Service, Flinders Medical Centre, and the North Western Adelaide Health Service.

How do I give my consent?

You will be asked to give written consent to include your child on the Register. If, in the future, you change your mind, you can withdraw your consent. Refusing to include your child will not disadvantage your child in any way or change your relationship with health professionals or hospitals.

What happens now?

By now you will have received some information from your child's doctor about the South Australian Cerebral Palsy Register, and the Register Officer will have contacted you either by phone or letter.
The next step is to decide whether you would like your child to be included on the Register. Feel free to contact the Register Officer and ask any questions you may have at that time.

If you decide that you would like your child to be included then please sign and return the Consent Form in the prepaid envelope provided, (*if you decide not to participate please let the Register Officer know this by return mail so we know not to contact you in future*).

When your child is around five years of age, a paediatric rehabilitation specialist will conduct a one-off medical assessment. This will usually be conducted at your local Regional Office of Novita Children's Services. (We also have rural clinics in key central areas. It may be possible to make other arrangements for country families and those who would find it difficult to get to Adelaide).

The medical assessment will be conducted free of charge. You will receive no payment for your participation.

Although paediatric rehabilitation specialists working with the Cerebral Palsy Register will conduct the medical assessment, your child will remain in the care of his/her usual doctor. If, at any time, you have any questions about your child's treatment or development, you should ask your child's usual doctor. To collect complete information about your child's health it may be necessary to consult hospital records of your child's birth and current health.

If you have any further questions please feel free to contact the Register Officer, South Australian Cerebral Palsy Register, on (08) 8161 7242 during office hours.

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