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Child Assessment Service Epidemiology and Research Gulletin

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introduction

Cerebral palsy (CP) is a major cause of childhood physical disability. The prevalence rate of children with CP in Hong Kong was estimated to be 1.3 per 1000 children.¹

In addition to physical disability, children with CP may also suffer from epilepsy, behavioural problems, learning difficulties, language and communication problems, visual and hearing impairment. These children often need a wide range of service in rehabilitation.

In this issue, we plan to explore specific areas of developmental needs and service for children with CP available in Hong Kong. Developmental profile of children with CP attending the Child Assessment Service, along with the development of Selective Dorsal Rhizotomy in Hong Kong, will be discussed.

Profile of Children with Cerebral Palsy at the Child Assessment Service

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"Cerebral palsy (CP) describes a group of disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behaviour, and/or a seizure disorder".²

In this issue, we present the profile of children with CP seen at the Child Assessment Service (CAS). We include all clients recorded by the CAS Information System (CASIS) from 2002 to 2008 and we try to differentiate developmental problems faced by children with different types of CP.

Type of cerebral palsy

The total number of children with CP from CASIS was 533. Majority (69%) were of the spastic type. The number of children with different types of CP were shown in table:

Table 1. Number of children by type of CP

Type of CP	Spastic quadriplegia	Spastic diplegia	Spastic hemiplegia	Dyskinetic CP	Ataxic CP	Other infantile CP
No. of children	73	195	102	66	15	72

Age at referral

The majority of clients (322) were referred to CAS at a very young age i.e. <18 months (Table 2). This is likely because physical impairment like CP presents early in life. Also, with the close collaboration with various hospital paediatric units and Family Health Service, babies are referred early. Another reason may be that babies born prematurely with low birth weight are followed up early at CAS through the high risk follow up program. Children at preschool and school-aged years are also referred for the comprehensive developmental assessment at CAS for recommendation of rehabilitation and placement needs.

Age (Years)	0 - 0.5	0.5 - 1	1 - 1.5	1.5 - 2	2 - 4	4 - 6	>6
No. of children	103	134	85	63	82	44	44

Physical functioning

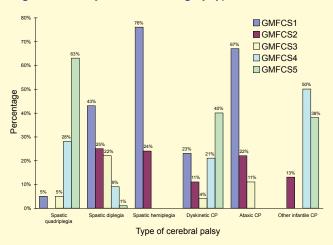
The physical functioning of the children is reflected by the Gross Motor Function Classification System (GMFCS), a standardized system to classify gross motor function of children with CP aged 12 months to 12 years. The classification is based on observation of a child's self-initiated movement and need for assistive technology and/or wheeled mobility.³

There are five levels of functioning:

- Children in level 1 can expect to walk and run, but has difficulty with more advanced skills
- Children in level 2 can expect to have some limitations in walking outdoors as they mature

- Children in level 3 can expect to have the ability to walk indoors on a level surface with a mobility device, and to use a wheelchair for community mobility
- Children in level 4 can expect reliance on wheeled mobility in home, school, and community settings, although they might walk with a walker for exercise
- Children in level 5 can expect limited selfmobility, even with assistive technology

Figure 1. Physical functioning by type of CP



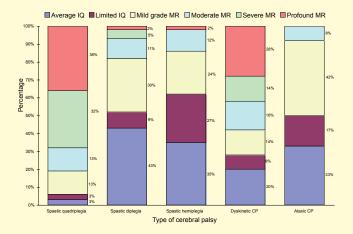
Among all types of CP, children with spastic hemiplegia had the best physical functioning as they were all independent walkers, 76% at level 1 and there is no child at level 3/4/5. On the other hand, children with spastic diplegia had a much wider variation of physical functioning. Majority of the spastic diplegic CP (68%) were independent walkers (level 1 & 2), 22% walked with aids, while the remaining 10% used wheelchair. For children with spastic quadriplegia and dyskinesia, they tended to have lower physical functioning, most of them being in Level 4 or 5 (91% & 61% respectively). The motor ability of children with dyskinetic CP skewed towards the extremes with 34% of them being able to walk alone. It is important to define the different levels of physical functioning as this entails different rehabilitation needs (Figure 1).

Cognitive abilities of different types of cerebral palsy

Assessment of intellectual functioning of children with CP poses a challenge to clinicians as some children are unable to complete full testing due to physical impairment. If these children's motor impairment also coupled with language impairment, such as those with dystonic and dyskinetic CP, this could further mask some of their cognitive abilities. Augmentative and Alternative Communication may be needed to achieve a valid assessment.

In CAS, among all CP cases, children with spastic quadriplegia had the lowest intellectual functioning. Majority (68%) were either severely or profoundly retarded. Only 3 % achieved average IQ or above. The remaining (13 % each) had mild or moderate mental retardation.

Figure 2. Cognitive abilities by type of CP



In contrast, nearly half of the children with spastic diplegic or spastic hemiplegic CP had average IQ or above. For those with mental retardation, most were in the mild grade range.

Children with dyskinetic CP showed a more scattered profile. 20 % had average IQ or above; 8% had limited IQ; 14 % had mild grade mental retardation; 16 % had moderate grade mental retardation; 14% had severe grade mental retardation and 28% had profound grade mental retardation (Figure 2).

For children with ataxic CP, 33% had average IQ or above; 17% had limited IQ; 42% had mild grade mental retardation; 8% had moderate grade mental retardation and none had IQ below moderate grade mental retardation.⁴

Associated/Co-morbid developmental disabilities in children with different types of cerebral palsy

Due to an insult to the developing brain, associated developmental disabilities are expected in children with various forms of cerebral palsy. It can be related to different sites of injury in the brain. In our locality, children with CP had increased rate of visual-perceptual problems (9% in spastic diplegia; 7% in hemiplegia; 20% in ataxic CP) and handwriting problems (14% in spastic diplegia; 11% in hemiplegia; 20% in ataxic CP).^{5,6} Moreover, children with CP often have higher incidence of language and phonological problems (25% in hemiplegia; 12% in dyskinesia; 20% in ataxia). For those children with CP born prematurely, studies showed that there was an increased risk for attention problem.⁷ However, in our cases, only 2% of the spastic diplegia and 7% of the hemiplegia were noted to have attention deficit (Table 3).

Visual problems, such as squint and ambylopia, are other important comorbidities found in overseas studies and in our locality. Among our cases with CP,. squint (quadriplegia 28%; diplegia 36%; hemiplegia 16%; dyskinetic 24%; ataxia 33%) and amblyopia (ataxia 13%; spastic diplegia 6%; hemiplegia 5%; dyskinesia 3%) were common, especially among ataxic CP.. Blindness or low vision was more evident in spastic quadriplegia and dyskinetic CP. Occurrence of significant hearing impairment, worse than moderate grade loss, increased in quadriplegia (7 %) and dyskinetic CP (14%).

Table 3.	Associated/co-morbid	developmental
disabilities by	/ type of CP	

	Spastic quadriplegia	Spastic diplegia	Spastic hemiplegia	Dyskinetic CP	Ataxic CP
Squint	28%	36%	16%	24%	33%
Amblyopia	0%	6%	5%	3%	13%
Low vision	17%	1%	1%	8%	0%
Nystagmus	7%	4%	1%	2%	0%
Significant hearing loss	7%	0%	1%	14%	0%
Language problem	3%	7%	25%	12%	20%
Visual perception	0%	9%	7%	0%	20%
Handwriting problem	1%	14%	11%	6%	20%
At risk of dyslexia	0%	4%	1%	0%	0%
ASD	0%	2%	1%	2%	0%
AD/HD	0%	2%	7%	2%	0%
Emotional problem	0%	1%	1%	0%	0%

Some of the above problems seemed to be less obvious in the severely physically-impaired children as seen from the figures above. Children with spastic quadriplegia appeared to have fewer comorbidites. We feel that this is in fact due to their globally impaired intellectual functioning, which disqualified them in the diagnostic criteria of some developmental disabilities. But these children actually have the greatest functional needs among all the CP children.

It is important not to overlook the other developmental needs beyond physical impairment and intellectual functioning in children with CP. These developmental disabilities can pose a further challenge in providing rehabilitation for children with physical impairment in both mainstream and Physical Handicap schools. Further study would be worthwhile to document their needs.

The roles of parents and teachers are very important in the management of CP cases. A comprehensive team approach to assessment and reviewing concerns as raised by either parents or teachers across the years will address the developmental needs of children with CP effectively.

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Selective Dorsal Rhizotomy: Revisit

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Introduction

Cerebral palsy (CP) was first described by Little in 1861 as "cerebral paresis". In 1964 Bax defined CP as a disorder of movements and posture due to a non-progressive lesion of the immature brain. It excluded disorder of posture and movement that are of short duration, due to progressive disease or due solely to mental deficiency.

The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication, behaviour, epilepsy and secondary musculoskeletal problems.

Yam et al found that the point prevalence for children aged 6 to 12 years with CP was 1.3 per 1000.¹ Spastic CP remains the most common type. Spasticity is defined as a motor disorder characterized by velocity-dependent increase in tonic stretch reflexes that exaggerate tendon jerks, resulting in hyperexcitability of the stretch reflex. This results in increase muscle tone, weakness and abnormal movement, thus further interfere with function & contributes to discomfort, pain and ultimately affects quality of life. Without intervention, evidence suggested progressive degeneration and detrimental changes in gait and function would occur within 1 to 4 years time.

Selective Dorsal Rhizotomy: the theory behind

Children with CP require treatment for their spasticity and their orthopaedic deformities acquired over time. There are different treatment targets on spasticity reduction aiming at various sites in the reflex arc. Selective dorsal rhizotomy (SDR) removes the excitatory influences from the dorsal root and is one of the treatment choices to reduced lower limb spasticity. It aims to improve existing gross motor function and gait.

History of SDR

In 1913, Forester first described 159 cases of complete posterior rhizotomy performed in patients with spasticity. Due to persistent loss of sensation and proprioception, the operation was not widely accepted. In 1960, Gros divided part of the posterior root but he found residual spasticity. Fasano divided rootlets with abnormal evoke response and report excellent reduction in spasticity in 1979. In 1987, Peacock performed the operation with more extensive laminectomy and with more accurate identification of levels. A more limited laminectomy approach had been adopted in certain centres recently.

Outcome of SDR

Meta-analysis of three randomized controlled trials published in 2002 revealed that SDR with physiotherapy is efficacious in reducing spasticity in children with spastic diplegia and has a small positive effect on gross motor function.² Chan et al reported short term outcome on SDR in Hong Kong.³

SDR Clinic in CAS

In Child Assessment Service (CAS), we have established "Physical Impairment Clinic" (PI Clinic) and "Selective Dorsal Rhizotomy Clinic" (SDR Clinic) to serve clients with CP (and other neuromuscular diseases) for assessment, management and rehabilitation plan. The following are the data of SDR clinic in the year 2001-2008.

SDR clinic is held at Tuen Mun Child Assessment Centre once every one to two months. The first SDR clinic jointly organized by CAS and the Tuen Mun Hospital (TMH) Neurosurgical Unit was established in March 1997 with the aim of providing a forum for clinicians to share experience in the treatment of CP, and to serve appropriate clients who might benefit from the surgery. Core members include Paediatricians from CAS and TMH Paediatric Unit, Neurosurgeons from TMH Neurosurgical Unit, Physiotherapists from both Departments, as well as Orthopaedic surgeons from TMH and United Christian Hospital and Urologist from TMH. School therapists and referrers are the clinic's other members.

Indication for SDR

Table 1. Ir	ndications and	contraindications	for SDR
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Indication (6S)	Contraindications
Slim	Weakness
Smart	Presence of dystonia or ataxia
Straight	Weak truncal control
Strong	Severe contractures
Spastic	Severe scoliosis
Support: good family support	Hip subluxation
+/- good Selectivity	Hereditary spastic paraparesis

Pre- SDR case assessment

Candidates for SDR are scheduled for pre-SDR assessment. Developmental paediatricians and physiotherapists will assess them in CAS. Gait analysis, urodynamic studies, X-rays of hips and other relevant radiological investigations are also be arranged in TMH.

Clinical protocol for SDR case follow up

Candidates for SDR are followed up in CAS under a clinical protocol set up in 2003. Table 2 provides a summary of their follow up protocol from 2006 January, where patients being considered for SDR are assessed by Developmental Paediatricians and Physiotherapists in CAS as shown.

Table 2.Clinical protocol for SDR cases follow up

Time Assessment (Discipline)	Pre op	Post 6 mth	Post 12 mth	Post 36 mth	Remarks (might need OT assessment if indicated)
Developmental Paediatrician	+	0	+	+	FU 3 yearly until 12/16 years old
Physiotherapist	+	+	+	+	FU 3 yearly until 12/16 years old

Note:

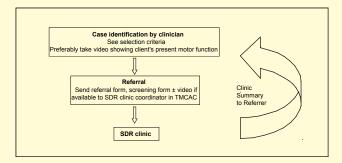
+: provide assessment; O: nil assessment; number of SDR performed after January 2006

Gait analysis review are scheduled for patients at post-SDR 6 month and 12 months in TMH. Family support and commitment are needed for intensive post-SDR physiotherapy program (at least daily PT for at the first 2 months). Close collaboration between therapists of different settings to continue the exercises upon discharge from hospital is also crucial for success.

Referral to SDR clinic

Potential patients could be referred to SDR clinic by clinicians or therapists. The clinic coordinator will schedule the patient to be seen in the clinic. Referrers are most welcomed to sit in the clinic as well. The clinic summary will be sent back to the referrer as a reply for their reference (see figure 1 for the flow chart).

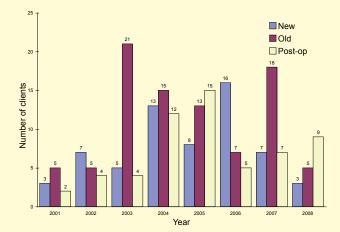
Figure 1. Flow chart of SDR clinic



Statistics of SDR clinic from 2001 to 2008

The number of clients attending the SDR clinic is shown in figures 2 and 3. In figure 2, new cases represent those who attended the clinic for the first time. If the same patient was seen in the next clinic, they would be regarded as old cases. Figure 3 showed the number of attending patients with different types of CP. Patients with diplegic CP were further categorised by using the Gross Motor Function Classification System (GMFCS).⁴ In GMFCS, there are 5 levels of gross motor functions. GMFCS I/II would include those who could walk independently, but with different level of ability for stairs walking and jumping. Patient with GMFCS III/IV could walk or practice walking with walking aids. Patients with diplegic CP were the majority of the cases attending the SDR clinic (Figure 3). More female attendants were noted in our clinic (M:F ratio around 1: 1.4). Table 3 showed the number of SDR performed in TMH as recorded from 2003 to 2008.

Figure 2. Types of attendance





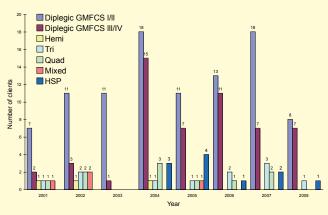


Table 3. Number of SDR performed in TMH*

Year	2003	2004	2005	2006	2007	2008
No. of SDR	6	10	11	8	8	5

* Data was recorded from 2003 when the cohort study

Of the 16 patients who had undergone SDR from 2003 onwards, all of them have definite improvement in spasticity. However, up to date, only one showed improvement in GMFCS level. Therefore, functional gains of these children still need to be followed up.



The CAS PI team (2008)

Future plans for SDR clinic

Our team plans to regularly review post-SDR cases at the clinic for functional outcome and further intervention needs. Follow up study for children who have undergone SDR is being planned, to understand their long term functional outcome.

Conclusion

SDR is one of the treatment modalities for spastic lower limbs. Evidence suggests definite reduction in spasticity. Combination with other modalities of treatment such as orthopaedic surgery might be needed for correction of bony alignment problem and for contractures. Postoperative complications such as weakness and initial deterioration in gait should be looked after carefully.

The care of children with cerebral palsy involves management by multiple disciplines. It is now time for us to review the care for these children. Treatments that benefit their motor control will in turn help to improve their activities of daily living, and ultimately lead to better qualities of life.

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- Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Developmental Medicine & Child Neurology 1997;39, 214-23.

Recent Publications and Scientific Presentations

Publications

<u>Chan BMY</u>. Why and how is mathematics so difficult for some children: studies and experience sharing on mathematics disorder. Journal of Basic Education 2009;(18)2:137-56.

Lau WY, Chan CK, Li JC, Au TK. Effectiveness of group cognitivebehavioral treatment for childhood anxiety in community clinics. Behav Res Ther 2010;48(11):1067-77. 10.1016/ j.brat.2010.07.007.

Leung C, Lau J, Chan G, Lau B, Chui M. Development and validation of a questionnaire to measure the service needs of families with children with developmental disabilities. Res Dev Disabil 2010;31(3):664-71. doi:10.1016/j.ridd.2010.01.005.

Leung C, Mak R, Lau V, Cheung J, Lam C. Development of a preschool developmental assessment scale for assessment of developmental disabilities. Res Dev Disabil 2010;31(6):1358-65. doi:10.1016/j.ridd.2010.07.004.

Liu KPY, <u>Fong AKH</u>. Overview of occupational therapy practice with children in Hong Kong. Journal of Occupational Therapy, Schools, & Early Intervention 2010;3(3):282-89. doi: 10.1080/19411243.2010.515183.

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<u>Tsang L</u>, Chak WK. Neurocognitive assessment for a 7-year-old girl with left mesial temporal sclerosis. Brainchild 2010;11(1):30-4.

Wong AMY, Leung C, <u>Siu EKL</u>, <u>Lam CCC</u>, <u>Chan GPS</u>. Development of the language subtest in a developmental assessment scale to identify Chinese preschool children with special needs. Res Dev Disabil 2011;32(1):297-305. 10.1016/ j.ridd.2010.10.004.

Scientific Presentations

The following presentations were conducted between January and December 2010:

有特殊學習需要兒童的評估、診斷及治療 教育局屯門區學校發展組,社會福利署屯門區家庭及兒童福利服務 協調委員會,屯門區小學校長會合辦 2010/11學年屯門區小學聯校教育發展日 二零一零年十二月十五日 葉佩華

Mathematics disability on 18 November 2010 at course of Master of Educational and Child Psychology, Hong Kong Polytechnic University by CHAN Mee-yin, Becky.

認識視覺學習策略支援自閉症/亞氏保加症學童 (TEACCH) on 3 November 2011 at Special Learning Needs Education Course in Autism/Asperger's Syndrome, HKU SPACE by LAM Ling.

Nature and nurture: parenting for parents with children aged 3-6 on 30 October 2010 at Parent seminar: effective home-school cooperation makes a difference, Quality Assurance Division, Education Bureau by Dr LEE Mun-yau, Florence. 自閉症/亞氏保加症學童的診斷方法 on 20 October 2010 at Special Learning Needs Education Course in Autism/Asperger's Syndrome, HKU SPACE by LAM Ling.

Developmental disorders: nature and nurture on 16 October 2010 at 12th Beijing/Hong Kong Medical Exchange - Children, development, environment by Dr LAM Chi-chin, Catherine.

Developmental screening and services for children with DD in HK and Learning and development - children with specific learning difficulties; screening diagnosis and management on 30 September 2010 at MSc course "Issues in children with Developmental Disabilities (DD)", Department of Rehabilitation Sciences, Hong Kong Polytechnic University by Dr LAM Chi-chin, Catherine.

Attention deficit hyperactivity disorder and autism on 22 September 2010 at Post-registration Certificate Course in the Child and Adolescent Nursing, Hospital Authority by Dr LIU Kayee, Stephenie.

Dyslexia and specific learning disabilities on 22 September 2010 at The Institute of Advanced Nurse Studies by Dr LAM Chichin, Catherine.

Social and communication development: selected population: children with ASD - screening, diagnosis and general management on 20 September 2010 at MSc course "Developmental issues in children with or without disabilities", Department of Rehabilitation Sciences, Hong Kong Polytechnic University by Dr WOO Kai-fan, Estella.

Autism spectrum disorder - management on 7 September 2010 at "精靈一點", Radio Television Hong Kong by Dr LIU Kayee, Stephenie.

Early childhood development on 28 August 2010 at Hong Kong College of Paediatrician by Dr LAM Chi-chin, Catherine.

Autism spectrum disorder: Complementary and Alternative Management (CAM) on 3 July 2010 at Hong Kong College of Paediatricians by Dr LIU Ka-yee, Stephenie.

Word reading in Chinese kindergarteners with and without specific language impairment (SLI) on 2 July 2010 at Research in Reading Chinese and Related Asian Languages Conference (Toronto, Canada) by Dr LAM Chi-chin, Catherine.

介紹傷殘運動員評級方法及傷殘人士運動 on 17 June 2010 at Thematic Course on Education of Students with Physical Disability, Hong Kong Institute of Education by LEUNG Yim-fan, Cannie.

Knowledge management in Child Assessment Service, Department of Health on 11 June 2010 at Knowledge management in government: building organizational intelligence for service excellence, Hong Kong Polytechnic University by Dr LIU Ka-yee, Stephenie.

讀寫困難學生的校內及公開考試調適需知 on 3 June 2010 at Certificate in Special Education, HKU SPACE by CHAN Mee-yin, Becky.

Neurocognitive outcome of paediatric traumatic brain injury: a cohort study in Hong Kong on 19 April 2010 at 2nd UK Paediatric Neuropsychology Symposium (London, UK) by TSANG Yee-ha, Lucia.

讀寫困難學生的校內及公開考試調適需知 on 29 March 2010 at Certificate in Special Education, HKU SPACE by CHAN Mee-yin, Becky.

Assessment of children with visual impairment and hearing impairment on 18 March 2010 at Hong Kong Polytechnic University by Dr LEE Mun-yau, Florence and TSANG Fung-king, Janice.

How to handle special needs children's temper and misbehaviour on 13 March 2010 at Benji's Centre by TSANG Yee-ha, Lucia.

解讀評估工具及結果分析 on 4 March 2010 at Certificate in Special Education, HKU SPACE by CHUNG Wai-hung, Angela.

Workshop on the Hong Kong Cantonese Oral Language Assessment Scale (HKCOLAS) at The University of Hong Kong on 26 February 2010:

- · Opening Speech by Dr YIP Pui-wah, Lesley
- Administering HKCOLAS & Test of Hong Kong Cantonese Grammar by NG Kwok-hang, Ashley
- Textual Comprehension Test by CHAN Yvonne Binva
- Word Definition Test by MAN Yuk-han, Yonnie
- Lexical-Semantic Relations Test & Expressive Nominal Vocabulary Test by CHAN Wai-ki, Amy
- Nonword Repetition Test & Hong Kong Cantonese Articulation Test by CHEUNG Sau-ping, Pamela

Visual assessment in children with developmental problems on 25 February 2010 at Hong Kong Polytechnic University by CHEUNG Pui-yi, Josephine and LIU Sau-kuen, Dilys.

及早識別及評估方法 on 25 February 2010 at Certificate in Special Education, HKU SPACE by CHUNG Wai-hung, Angela.

讀寫困難學生的校內及公開考試調適需知 on 11 February 2010 at Certificate in Special Education, HKU SPACE by CHAN Mee-yin, Becky.

Specific learning difficulties in reading and writing on 11 February 2010 at Certificate in Special Education, HKU SPACE by Dr LAM Wai-fan, Fanny.

Specific learning difficulties in reading and writing on 4 February 2010 at Certificate in Special Education, HKU SPACE by Dr LAM Wai-fan, Fanny.

Developmental coordination disorder in young children on 1 February 2010 at The Chinese University of Hong Kong by CHUI Mun-yee.

輔導專注力不足幼兒的方法與技巧 on 23 January 2010 at Hong Kong Society for the Protection of Children by CHEUNG Manching, Jasmine.

Cognitive, language and psychological development and related disorders on 19 January 2010 at Post-registration Certificate Course in Child and Adolescent Psychiatric Nursing, Castle Peak Hospital by CHUNG Wai-hung, Angela.

Next Issue

The next issue of CASER will be released in June 2011. The featured topic is on hearing impairment.

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