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Introduction

Tang ML Kitty[1]

[1] Senior Medical & Health Officer

This issue of CASER is an update on one of the most commonly known problems in the field of Developmental Paediatrics - Intellectual Disability (ID).

It has been more than ten years since the last issue of CASER on this topic. During this period, many changes have taken place in this field. One of the most salient changes is on terminology. The term "intellectual disability" (ID) is now widely adopted in replacement of the term "mental retardation" that has been in use for decades. Clinically, both the American Association on Intellectual and Developmental Disabilities (AAIDD) and the Diagnostic and Statistical Manual of Mental Disorders (DSM-5) of the American Psychiatric Association (APA) now use ID in their diagnostic classifications. The International Classification of Diseases (ICD) from the World Health Organization (WHO) will be employing the term "Intellectual Developmental Disorder" (IDD) in their upcoming 11th edition.[1] The terms ID and IDD are considered as equivalent and interchangeable.[2] In the first part of this issue of CASER, there will be an update on changes in the concept and terminology of ID.

One important change for ID is the emphasis on adaptive functioning in the diagnostic criteria. DSM-5 relies more on adaptive functioning than the DSM-IV both for diagnosing ID and for determining its level of severity.[2] In the second part of this issue of CASER, we will discuss these changes of ID, together with a review in the situation in HK. Our clinical psychologist will talk about the application of The Adaptive Behavior Assessment System Assessment System of USA in our local setting. Our occupational therapists will share the local experience on measurement of participation and environment for children and youth with ID.

Finally, the use of genetic testing in Hong Kong (HK) for ID will be discussed. Current international guidelines for genetic testing recommend array comparative genomic hybridization (aCGH) or chromosomal microarray (CMA) as a first genetic test for individuals with unexplained global developmental delay or autism, together with a thorough history and physical examination. However, aCGH/ CMA is still not widely accepted as a first-tier test owing to the perception that this approach is more expensive.[3] What is the current practice in HK? In the last part of this issue of CASER, a local study on the genetic profile in children with unexplained ID and the clinical application of chromosomal microarray in these children in HK will be highlighted.

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Update on the Changes in Terminology and Concepts about Intellectual Disability

Ng TT Teresa[1], **Wong LW Polly**[1]

[1] Clinical Psychologist

The term "intellectual disability" (ID) is now widely adopted in replacement of the term "mental retardation" for policy, administrative and legislative purposes worldwide.[1] The American Association on Intellectual and Developmental Disabilities (AAIDD) first adopted the term ID in their definition and classification manual on the condition. In clinical practice, the World Health Organization (WHO) International Classification of Diseases, (currently ICD-10 (10th edition) with ICD-11 (11th edition) coming into effect in January, 2022), and the American Psychiatric Association (APA) Diagnostic and Statistical Manual of Mental Disorders, 5th edition (DSM-5) are the two most commonly used diagnostic classification systems. APA has also adopted the term ID in their diagnostic classification, while the ICD will employ the term "Intellectual Developmental Disorder" (IDD) in their upcoming version.[1,2] According to DSM-5, the terms of ID and IDD are clinically equivalent and could be used interchangeably.[3]

In clinical practice, both the ICD-10 and DSM-5 have comparable definitions on ID, which define the disorder as one that begins during the developmental period and is characterised by significant impairments in both intellectual and adaptive functioning.[3,4] Notably, one of the major changes introduced in the later versions of ICD and DSM is the re-integration of ID with other neurodevelopmental disorders.[1] ID/IDD is classified under the neurodevelopmental disorders in both DSM-5 and ICD-11 respectively, in contrast to the neurocognitive disorders. The revision reflected advanced and consensual understanding on the common aetiopathogenetic risks (such as genetic alteration, abnormal neural circuit development, etc.), clinical features (such as cognitive, behavioural, learning and communicative dysfunctions), as well as early emergence of deficits and the continuing course of neurodevelopmental disorders.[5] As such, a change is made in DSM-5 regarding the onset of ID from stating a specific age to a broader, generic developmental period.

While both the ICD-11 and DSM-5 continue to emphasise impairments of cognitive abilities in the diagnosis of IDD/ID, both move away from making the diagnosis and determining the severity of the condition by means of the general IQ test scores alone.[1,3] The move is informed by emerging research indicative of the limitations of IQ test scores in capturing the dynamic nature of cognitive impairment in persons with ID.[6] It also reflects the evolving conceptualisation of intelligence as an umbrella term that includes cognitive functioning, adaptive behaviour and learning that is appropriate to age, societal and cultural expectations and demands of everyday life.[1] There is also a call for assessment of other cognitive functions such as executive functioning to provide an individual profile and further inform diagnosis.[5,6]

In addition, adaptive behaviour, which is the collection of conceptual, social, and practical skills that have been learnt and are performed by people in daily living, were found to correlate with intellectual functioning and needs to be jointly considered in the diagnosis of ID.[7] Similar to the assessment of intellectual functioning, assessment of the level of adaptive functioning involves the use of standardised measures normed on the general population, and the test findings should be interpreted with consideration of respondent reliability as well as the broader context of community environments, expectations for same-age peers, as well as all other sources of measurement error.[7] While a number of tools have been developed for determining the severity and classifying support needed of persons with ID, more work is needed before such measures are available for worldwide use.[1]

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Application of The Adaptive Behavior Assessment System in Our Local Setting

Lau WY Kelly[1], Tse TY Stanley[2]

[1] Clinical Psychologist [2] Educational Psychologist

Assessment of Adaptive Behaviour

According to different classification systems, deficit in adaptive behaviour is one of the necessary criteria for confirming the diagnosis of intellectual disability. Educators may also collect information on a child's adaptive functioning to determine the presence and level of the individual's limitations, plans for educational goals, and to set the baseline performance for progress evaluation. The Adaptive Behavior Assessment System – Second Edition (ABAS-II) measures 10 specific skill areas of adaptive behaviour. These skill areas are divided into three groups, thereby producing three domain scores, namely Conceptual, Social and Practical. Its Chinese version (ABAS-II: C)[1] was normed in Taiwan with good psychometric properties on internal consistency, test-retest reliability, inter-rater reliability, factor structure, concurrent validity and discriminant validity.

In this study, we tried to examine the use of the ABAS-II: C as a measurement tool for assessing adaptive behaviour in Hong Kong. We hypothesised that:

1. High internal consistency will be found for the composite scores and the skill area subscales. Moderate to high inter-correlations will be found on skill area scaled scores with their respective domain scores and general adaptive composite (GAC). High inter-correlations will be found between adaptive domain scores and GAC.

2. Children with developmental disabilities will attain lower scores than children with typical development. Children with developmental delay and children with autism spectrum disorder and developmental delay will show significantly lower scores on GAC and three domain scores compared to the norm. Children with autism spectrum disorder will show specifically lower score on social and practical domain scores.

Methodology

Participants were children aged 4 to 5 and their parents. In the typical development (TD) group, 71 parents and children were recruited in 4 kindergartens. The cognitive subtest of the Preschool Developmental Assessment Scale for Hong Kong Chinese children which has been demonstrated to be valid and reliable,[2] was used to ensure that only children with typical development were included. Children suspected to have developmental delay (DD) (i.e. scaled score of two standard deviations below the mean) were excluded from further data collection procedures.

For the clinical group, 114 parents of children with developmental disabilities were recruited at Child Assessment Service (CAS), Department of Health. They were then classified into three sub-groups, namely DD (n=20), Autism Spectrum Disorder (ASD, n=43), and DD with ASD (n=51) (Table 1a, Table 1b).

Data Analysis and Result

Reliability was investigated using the Cronbach's alpha coefficients of GAC, three adaptive domain scores and the scaled scores of the ten skill areas to understand the relatedness of the items. The alpha coefficient for GAC (.987) and the three domain scores (between .961 and .969) were above the .90 threshold. The alpha coefficients for the ten skill areas were also high, falling between .841 and .945 (Table 2).

For the inter-correlations between different skill areas, the correlation coefficients fell within the moderate to high range, ranging from .50 to .82 with median of .66. For the inter-correlations between skill areas and adaptive domains, each skill area had the highest correlation with their respective domain and the correlation coefficients were moderate to high, ranging from .58 to .96 with median of .81. The correlations between the three domains were also high, with all of them above .85. For GAC, it correlated well with all ten skill areas (with correlation between .70 and .90), and all three adaptive domains (with correlation above .94) (Table 3).

Analysis was conducted to examine the ability of the scale to differentiate children with developmental disability from children with typical development. Analysis of Covariance (ANCOVA) was conducted to compare the scores of the TD group and the clinical group on GAC, while Multivariate Analysis of Covariance (MANCOVA) was conducted to compare the scores of the TD group and the clinical group on the three domain scores and ten skill area scaled scores. Age of the child and child's gender were used as covariates in both of the analyses due to a significant difference found between the two groups on these two variables. For the ANCOVA, a significant difference was found between the two groups on GAC. The GAC of the TD group (M=102.93, SD=11.95) was significantly higher than that of the clinical group (M=79.88, SD=12.32), $F(3,181)=58.23$, $P < .001$. For the MANCOVA, separate analyses were conducted to compare the two groups on their 3 domain scores and 10 skill area scaled scores. The multivariate tests using Wilks' Lambda were significant for the 3 domain scores ($F(3,179)= 25.31$, $P < .001$) and 10 skill area scaled scores ($F(10,172)= 10.41$, $P < .001$). Result of the follow-up univariate test indicated significant differences in all 3 domain scores and 10 skill area scaled scores between the two groups at $P < .001$, with the TD group having a higher score on all domain scores and skill area scaled scores than the clinical group. Thus, GAC, 3 domain scores and 10 skill area scaled scores significantly discriminated between the TD group and clinical group (Table 4).

Analysis was further conducted to examine the ability of the scale to differentiate groups with different developmental disabilities on GAC using ANCOVA and 3 domain scores using MANCOVA with Bonferroni test for post hoc comparisons. For the analysis on GAC using ANCOVA with age of the child and child's gender as covariates, result was significant with $F(5,177)=45.92$, $P< .001$), indicating that the four groups, namely TD, ASD, DD, and DD with ASD, were significantly different on GAC. Follow-up pair-wise post hoc tests comparing the means of the four groups showed that GAC of the TD group (M=103.24, SD=11.27) was the highest and significantly differed from the other three groups. GAC of the ASD group (M=86.47, SD=11.68) was significantly lower than the TD group, but significantly higher than the DD group and the DD with ASD group. No significant difference between GAC of the DD group (M=78, SD=15.45) and that of DD with ASD group (M=73.63, SD=8.33) was noted (Table 5).

For the analysis on the 3 domain scores using MANCOVA with age of the child and child's gender as covariates, results was significant using Wilks' Lambda, ($F(9,430.92)= 15.78$, $P< .001$). The follow-up univariate analyses indicated that the four groups differed on all 3 domain composite scores, Conceptual Domain ($F(5,179)=55.07$, $P < .001$), Social Domain ($F(5,179)=39.18$, $P < .001$) and Practical Domain ($F(5,179)=28.05$, $P < .001$).

Post hoc comparisons were conducted by Bonferroni. On all three domains, the TD group had the highest domain scores and the scores were significantly different from the other three groups. Each clinical group had their unique patterns in the different domains. In the Conceptual Domain, the score of the ASD group (M= 89.6, SD=11.97) was significantly higher than the DD group (M=74.8, SD=16.55) and the DD with ASD group (M=72.84, SD=11.07), while no significant differences were found between the score of DD group and the DD with ASD group. In the Social Domain, no significant differences were found between the scores of the ASD group (M=84.42, SD=13.68) and the DD group (M=82.35, SD=17.79), but scores of both groups were significantly higher than that of the DD with ASD group (M=72.06, SD=10.52). In the Practical Domain, the score of the ASD group (M=88.51, SD=13.8) was not significantly different from the DD group (M=79.45, SD=17.92), but was significantly higher than that of the DD with ASD group (M=75.53, SD=12.82). Also, no significant differences were found between the score of the DD group and the DD with ASD group.

For the TD group, the mean scaled scores of the ten skill areas were between 9.44 and 11.21, except functional academics with a mean scaled score of 13.15, and the standard deviations were between 2.25 and 3.01. For the three adaptive domain scores, the mean scores were between 103.24 and 106.66 and the standard deviations were between 10.97 and 12.18. For GAC, the mean score was 103.24 and the standard deviation was 11.27. The mean of the skill area scaled scores and domain scores, except for the functional academics scaled score, were comparable to the mean of 10 for scaled scores and 100 for domain scores of the Taiwan normative sample, but the standard deviation of the skill area scaled scores and domain scores were slightly smaller than the standard deviation of 3 for scaled scores and 15 for composite scores of the Taiwan norm. The mean scaled score of functional academics was about one standard deviation higher than that of the Taiwan norm.

Discussion and Recommendation

The present results indicated that the ABAS-II: C demonstrated good reliability when used in the local context. It has a high internal consistency (>.96) over GAC and the three domain scores, i.e., Conceptual, Social, and Practical. It also has good internal consistency (>.84) over the ten skill area scaled scores. Compared to the results of the validation study of the manual of the Chinese ABAS II, results of the current study demonstrated comparable level of reliability. Among all skill areas, the lowest internal consistency was the self-care subscale with alpha coefficients of .84, which is still much higher than the acceptable level of .70.

The inter-correlations between different skill areas were found to be ranging from moderate to high (between .50 and .82). This finding confirmed with the definition of adaptive behaviour from the literature and theoretical structure of the ABAS-II that these skill areas are independent but related with one another. At the same time, the skill areas have strong correlations with GAC ($r >.74$) and their own respective domains ($r >.80$), and the strongest correlations were found between the three adaptive domain composite scores and GAC ($r >.85$). The results on inter-correlations between different scores replicated the results of validation study from the manual of Chinese version, with correlation of .51 to .77 between skills areas, .61 to .79 between skill areas and their respective domains, .63 to .84 between skill areas and GAC, and .91 to .95 between adaptive domains and GAC. In conclusion, the data of the current study provided supporting evidence on the theoretical constructs of the ABAS-II.

Present results showed that children with disabilities performed significantly lower than those with typical development. The clinical group has lower mean score on GAC, three adaptive domain scores than children from TD group and the effect sizes were large for the GAC (Partial Eta Squared=.49) and for the three adaptive domains (Partial Eta Squared from .24 to .29). The mean of GAC, Conceptual domain, Social domain and Practical domain scores of the clinical group were 79.24, 79.60, 78.58 and 81.31 respectively, which fell within the classification of borderline to low average and are more than one standard deviation below the mean. For the ten skill areas, the clinical group also has significantly lower scores in all ten skill areas than the TD group, with largest difference found in the communication skills (Partial Eta Squared=0.48) and social skills (Partial Eta Squared=0.44) subscale. The result also replicated the findings in the ABAS-II: C validation studies that compared clinical groups with matched control group using the Taiwan sample. In conclusion, the result showed that ABAS-II: C demonstrated good ability in differentiating children with disabilities from children with typical development.

Data also supported that ABAS-II: C demonstrated good ability in differentiating different levels of adaptive functioning for different clinical conditions. While the TD group showed significantly higher scores on all adaptive domain composites and GAC than the other three clinical groups, it is interesting to find that different patterns emerged in different domains among the three clinical groups. The ASD group was found to have significantly higher scores only in GAC and the Conceptual domain score than the DD group, but showed no significant difference in the Social domain and Practical domain score with the DD group. Yet, the DD with ASD group had similar scores with the DD group on GAC, Conceptual domain and Practical domain scores, but showed significantly lower score in the Social domain score. Such profile of adaptive skills for the ASD group and DD group matched well with the previous research studies on the adaptive skills of children with Autism Spectrum Disorder and Developmental Delay. For instance, several research studies on individuals with high functioning ASD showed that they exhibit deficit in adaptive skills, especially on the Social and Practical domain of the adaptive behaviour.[3,4] For pre-schoolers, Milne et al[5] concluded that preschoolers' adaptive function was highly correlated with their developmental ability, with preschoolers with developmental delay showing poorer performance on their adaptive behaviour.

Examining the distribution of the scores of the TD group provided evidence that the scoring system of the ABAS-II: C functions quite well for the TD group. Almost all subscale scaled scores and domain scores of the TD group had their means comparable to the mean score of the Taiwan norm. The standard deviation of subscale scaled scores and domain scores of the TD group were found to be slightly smaller than the standard deviation of the standardised score. A possible reason is due to the limited age range and size of the sample in the present study which limited the range of deviation of the scales. Another point to note is that the mean score of the Functional Academics skill area of the TD group was more than one standard deviation above the mean score. A possible explanation is that the academic skills of preschoolers in general had been enhanced since the establishment of the Taiwan norm in 2008. Another possible reason may be due to the difference in the level of functional academic skills between children of Hong Kong and Taiwan. A study that explored the early childhood education in Hong Kong also revealed that the early childhood education in Hong Kong had great academic pressure on local preschool children under the market-driven context to put emphasis on academic learning and prepare the children to enter primary school due to the highly competitive environment in the primary education level.[6] With the demand to display academic skills in early childhood from parents and preschools in Hong Kong, the functional pre-academic adaptive skills may have been fostered in children in Hong Kong to meet the expectation from environment, thus leading to a significant higher level of functional academic skills than children in Taiwan.

In summary, the present results provided preliminary supporting evidence for the use of the ABAS-II: C in the Hong Kong context to assess adaptive behaviour of preschool children. The data in the present study supported the hypotheses that the scale has good reliability and validity, and that it is useful in discriminating children with various levels of disabilities in adaptive functioning. The findings in the present study provided some ground to demonstrate the scale as an evidence-based Chinese scale to assess adaptive behaviour based on Hong Kong' s local sample.

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Table 1a Demographic characteristics of participants by normal group and SEN group

Characteristic	Normal Group (n=71)	SEN Group (n=114)	Significance
Sex of the target child: boy	36 (50.7%)	95 (83.3%)	$\chi^2(1) = 22.54$ P < .001
Mean age of children	4.71 (SD=0.50)	5.60 (SD=0.23)	t(183) = 14.24 P < .001
Mean weeks of pregnancy at child birth	38.43 (SD=2.58)	38.03 (SD=2.58)	t(165) = -.96 P = .34
Children's mean length of residence in HK	3.77 (SD=1.09)	4.9 (SD=.52)	t(181) = 8.12 P< .001
Relationship with target child: mother	54 (76.1%)	93 (81.6%)	$\chi^2(2) = 1.68$ P = .43
Father's education: upper secondary or above	59 (84.3%)	84 (76.4%)	$\chi^2(1) = 1.64$ P = .20
Father working	64 (94.1%)	103 (94.5%)	$\chi^2(1) = .01$ P = .92
Mother's education: upper secondary or above	52 (74.3%)	91 (80.5%)	$\chi^2(1) = .99$ P = .32
Mother working	36 (52.2%)	51 (45.5%)	$\chi^2(1) = .75$ P = .39
Family income above median domestic household income (HK$20,000 or above)	35 (50.7%)	64 (57.7%)	$\chi^2(1) = .83$ P = .36

Table 1b Demographic characteristics of participants by normal group and type of developmental disability

Characteristic	Normal (n=71)	ASD (n=43)	DD (n=20)	DD with ASD (n=57)	Significance
Sex of the target child: boy	36 (50.7%)	38 (88.4%)	13 (65.0%)	44 (86.3%)	$\chi^2(3) = 26.53$ P < .001
Mean age of children	4.71 (0.50)	5.58 (0.23)	5.59 (0.25)	5.63 (0.23)	F(3,181) = 91.15 P < .001
Mean weeks of pregnancy at child birth	38.43 (2.58)	38.3 (2.21)	37.47 (3.66)	38.02 (2.36)	F(3,163) = 0.75 P = .526
Children's mean length of residence in HK	3.77 (1.09)	4.98 (0.15)	4.85 (0.49)	4.86 (.70)	F(3,179) = 29.70 P < .001
Relationship with target child: mother	54 (76.1%)	36 (83.7%)	14 (70.0%)	43 (84.3%)	$\chi^2(6) = 10.40$ P = .109
Father's education: upper secondary or above	59 (84.3%)	34 (81.0%)	17 (85.0%)	33 (68.8%)	$\chi^2(3) = 4.80$ P = .187
Father working	64 (94.1%)	39 (90.7%)	17 (94.4%)	47 (97.9%)	$\chi^2(3) = 2.23$ P = .526
Mother's education: upper secondary or above	52 (74.3%)	37 (86.0%)	13 (68.4%)	41 (80.4%)	$\chi^2(3) = 3.39$ P = .336
Mother working	36 (52.2%)	16 (38.1%)	6 (31.6%)	29 (56.9%)	$\chi^2(3) = 5.79$ P = .122
Family income above median domestic household income (HK$20,000 or above)	35 (50.7%)	28 (66.7%)	9 (47.4%)	27 (54.0%)	$\chi^2(3) = 3.29$ P = .35

Table 2 Internal consistency

Scale	Cronbach's alpha coefficient
Skill area	
Communication	.933
Community Use	.941
Functional Academics	.945
Home Living	.912
Health and Safety	.921
Leisure	.925
Self-Care	.841
Self-Direction	.921
Social	.933
Motor Skills	.878
Adaptive Domain Composite	
Conceptual Composite	.968
Social Composite	.961
Practical Composite	.969
General Adaptive Composite	.987

Table 3 Intercorrelations among subtests scaled scores and composite scores

	CU	FA	HL	HS	Le	SC	SD	Soc	MS	ConC	SocC	PraC	GAC
Com	.814**	.778**	.590**	.675**	.753**	.587**	.677**	.779**	.548**	.917**	.796**	.776**	.868**
CU		.749**	.669**	.786**	.758**	.569**	.687**	.765**	.525**	.850**	.799**	.886**	.875**
FA			.511**	.656**	.666**	.503**	.582**	.649**	.530**	.909**	.685**	.707**	.806**
HL				.701**	.648**	.633**	.705**	.699**	.540**	.659**	.693**	.845**	.782**
HS					.794**	.610**	.752**	.784**	.532**	.774**	.826**	.894**	.867**
Le						.595**	.811**	.823**	.555**	.827**	.956**	.809**	.896**
SC							.628**	.625**	.554**	.629**	.632**	.803**	.741**
SD								.822**	.557**	.818**	.852**	.794**	.855**
Soc									.570**	.826**	.943**	.830**	.899**
MS										.610**	.584**	.621**	.699**
ConC											.868**	.850**	.946**
SocC												.860**	.940**
PraC													.946**

** P < .01

Note. Com = Communication; CU = Community Use; FA = Functional Academics; HL= Home Living; HS = Health and Safety; Le = Leisure; SC = Self-Care; SD = Self-Direction; Soc = Social; MS = Motor Skills; ConC = Conceptual Composite; SocC = Social Composite; PraC = Practical Composite; GAC = General Adaptive Composite

Table 4 Mean scores and standard deviations of the normal group

	Mean	SD
Skill area		
Communication	9.97	2.55
Community Use	11.21	2.44
Functional Academics	13.15	2.25
Home Living	9.44	2.61
Health and Safety	10.30	2.38
Leisure	11.13	3.01
Self-Care	10.41	2.84
Self-Direction	10.35	2.59
Social	10.46	2.59
Motor Skills	9.94	2.53
Adaptive Domain Composite		
Conceptual Composite	106.66	11.07
Social Composite	104.13	12.18
Practical Composite	101.68	10.97
GAC	103.24	11.27

Table 5 Mean difference between the four groups on GAC and 3 domain composites

	Normal	ASD	DD
GAC			
ASD	19.76 **		
DD	28.33 **	8.56 *	
DD with ASD	32.79 **	13.03 **	4.47
Conceptual Composite			
ASD	19.80 **		
DD	34.25 **	14.45 **	
DD with ASD	36.66 **	16.86 **	2.41
Social Composite			
ASD	19.74 **		
DD	21.89 **	2.16	
DD with ASD	32.11 **	12.38 **	10.22 *
Practical Composite			
ASD	18.06 **		
DD	27.41 **	9.35	
DD with ASD	31.38 **	13.32 **	3.98

* P < .05 ** P < .01

Measurement of Participation and Environment for Children and Youth with Intellectual and Developmental Disabilities

Lin SY Lenzs[1], Yu WH Joanna[1]

[1] Occupational Therapist

Introduction

Young children with developmental delay and disabilities are at risk of experiencing problems when participating in activities as compared with typically developing peers.[1,2,3,4] During the early years of a child's life, supportive environments optimise outcomes of the child and family.[5] In recent years there has been expansion of the rehabilitation services for children with developmental delay and disabilities, such as the On-site Preschool Rehabilitation Services in kindergartens, and enhanced rehabilitation services in special schools for students with mild and moderate intellectual disabilities in Hong Kong. The collaboration among the carers, professionals and teaching staff to promote children's positive participation in home, school and community activities by providing practical measures and accommodations in a real-life situations is essential for their well-being.[6]

The International Classification of Functioning, Disability and Health (ICF)

With the introduction of the International Classification of Functioning, Disability and Health (ICF) framework,[7] the concept of participation becomes a central construct in health care, rehabilitation, and occupational therapy.[8] Participation in everyday activities is a key indicator of children' s health and well-being.[9] Participation has been broadly defined as "involvement in a life situation" in the International Classification of Functioning, Disability and Health-Children and Youth Version (ICF-CY).[10] The components of participation and environment have been identified as two important aspects of the framework that should be addressed and measured.[9]

Measuring Participation and Environment for Children and Youth

Individuals with intellectual disabilities (ID) have limited cognitive capacities and adaptive behaviours for participating in activities of daily living.[11] It is important to comprehensively examine the interaction between the individual and the environment in order to create the best person-activity-environment match, through providing appropriate adaptations (e.g., activity grading and environmental modification) that promote his/her activity participation and thereby develop new skills, make friends, have fun and gain quality of life.[12]

The Participation and Environment Measure for Children and Youth (PEM-CY) is a parent-report questionnaire that is developed based on the ICF-CY conceptual framework. It measures participation and environment simultaneously across three settings: home, school and community.[9] It is designed to help parents, service providers and researchers better understand activity participation of children and youth aged 5 to 17 years. Items for each life situation consist of a set of activities that are typically performed by the child in that environment. Examples are "personal care management" at home;[13] "classroom activities" at school and "organised physical activities" in the community.[14] Participation in each type of activity is measured in terms of frequency (from daily to never), level of involvement (from very involved to minimally involved), and parent' s desire for changes in that activity (yes or no and, if yes, five options can be chosen). Environmental impact on participation in each setting is measured by asking whether certain features of the environment (e.g. physical layout) will help or make it harder for the child to participate.

Following the development of the PEM-CY, the Young Children' s Participation and Environment Measure (YC-PEM) was designed for the younger age group from 0 to 5 years old. The YC-PEM is modelled after the PEM-CY, but underwent extensive revision so that the questions and response options are better applicable to caregivers of younger children. It assesses caregivers' perceptions of their young child' s participation in various types of activities that take place at home, daycare/ preschool and community settings. The YC-PEM has undergone initial validation of its online version with families in North America.[15] A number of cultural adaptation and research projects involving the YC-PEM and PEM-CY had already been done, e.g. in Sweden,[16] Korean[17] and Singapore,[18] with others in progress.

Validation of the Participation and Environment Measures in Hong Kong

Recently, members from the Occupational Therapy Programme of Department of Rehabilitation Sciences, Hong Kong Polytechnic University conducted a validation study on a culturally adapted version of the YC-PEM for the children aged 0-5 years in Hong Kong. The original English version was translated into Chinese. The Occupational Therapy team of the Child Assessment Service was invited to provide expert review on the linguistic clarity and cultural relevance of the translated version. Earlier to this, the children and youth version, PEM-CY, has also been adapted and validated in Hong Kong for the children aged 5-12 years, where evidence from psychometric properties supported its use in assessing Chinese children' s participation and environmental supports/barriers.[19] Both validated Chinese versions of the YC-PEM and PEM-CY are now available for purchase online in CanChild Centre for Childhood Disability Research.

Conclusion

The use of these validated Chinese versions of the YC-PEM and PEM-CY for evaluating children with developmental delay or intellectual disabilities in Hong Kong can be considered. The instruments will help identify children' s participation restrictions in different settings, and the environmental factors that hinder their participation. The assessment results will enable the making of evidence based recommendations for the schools and agencies to provide environmental modifications and resources to clients. The information will be useful in setting practical goals for training, and to come up with strategies to improve children' s participation in daily activities. The findings will also serve as a basis for research studies to examine the similarities and differences in activity participation for children with different disabilities across home, school, and community settings.

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Local Study Highlight & Current Practice:

Genetic Profile and Clinical Application of Chromosomal Microarray in Children with Intellectual Disabilities in Hong Kong

Tang ML Kitty[1], Chan YT Purdy[2]

[1] Senior Medical & Health Officer, [2] Acting Senior Medical & Health Officer

According to a cross-sectional study performed by the Child Assessment Service (CAS) and the Clinical Genetic Service (CGS) in 2016/17, 11.6% children with unexplained Intellectual disability (ID) demonstrated genetic abnormalities, as detected through chromosomal microarray (CMA) testing. This was comparable with finding from other international cohorts. The diagnostic yield of CMA increased with severity of ID (moderate, severe, and profound ID were 8.7%, 17.6%, and 23.5%, P<0.05) and was much higher in children with dysmorphism (45.8% vs 4.4%, P<0.05).[1]

This is the first local genetic study on CMA testing in children diagnosed with unexplained ID. Unexplained ID was defined as children with no identifiable causes for ID. These children were non-syndromic and non-dysmorphic. From July 2016 to June 2017, 339 children with history of developmental delay (DD) were assessed at pre-primary one and diagnosed with more severe forms of ID: 241 (71%) children had moderate ID, 49 (14.5%) had severe ID, and 49 (14.5%) had profound ID. 114 children were excluded or opted out for different reasons. Among the remaining 225 children, 138 underwent CMA testing. The diagnostic yield of CMA increased with severity of ID: it was 8.7% in moderate ID, 17.6% in severe ID, and 23.5% in profound ID (P<0.05) and the yield was much higher in children with dysmorphism (45.8% vs 4.4%, P<0.05). The abnormalities demonstrated included copy number loss (deletion), copy number gain (duplication), unbalanced translocation, Angelman syndrome and Cri du chat syndrome, etc. In total, 69% of pathogenic or likely pathogenic copy number variants (CNVs) were de novo.[1]

ID is estimated to affect 1% to 3% of the population in Western societies. It is almost two-fold greater in prevalence in low and middle-income countries, compared with high-income countries.[2] Importantly, the General Household Survey in 2014 showed the prevalence rate of ID to be approximately 1.0% to 1.4% in Hong Kong (HK).[3] The aetiology of ID is complex. While milder forms of ID are suspected to typically result from the interplay of genetic and environmental factors; biological causes, particularly genetic causes, are often identified in children with significant intellectual delay (IQ <50).[4] However, 50% to 60% of these children did not exhibit a known aetiology (unexplained ID).[5]

About Chromosomal Microarray

In recent years, chromosomal microarray (CMA) or array comparative genomic hybridisation, is recommended by many international professional organisations as a first-tier genetic investigation for children with unexplained DD, ID, or autism spectrum disorder (ASD).[4,6] Compared with conventional karyotyping, CMA is able to detect CNVs with much finer resolution and is not reliant on staining and visual resolution limits. In the past decade, CMA has allowed more comprehensive unbiased discovery of microdeletion and microduplication syndromes throughout the human genome. According to the American Academy of Neurology evidence report in 2011, CMA testing was abnormal in approximately 7.8% of patients with global DD or ID.[7] The yield was higher (10.6%) in those with syndromic features. Children with ASD who had co-morbid ID were more likely to yield molecular diagnoses.[7]

CMA has high clinical utility. First, it shortens the diagnostic odyssey and may avoid unnecessary investigations, which reduce both individual and societal costs associated with testing and medical care. Secondly, it may lead to a clinically actionable recommendation. Information from CMA may alert the clinician to other potential co-morbid conditions that could not have been predicted on the basis of physical examination alone. In HK, a detection rate of 8.6% was reported for clinically actionable CNVs,[8] which was comparable to the reported rates of 3.6% to 7% in Western studies.[9] Thirdly, it allows estimation of recurrence risk and informed decisions regarding reproductive options for future pregnancies. Together, these considerations support the use of CMA as a first-tier investigation for children with significant unexplained ID in HK.

Current Practice in HK

Currently, children with significant global delay, syndromic or dysmorphic features, multiple congenital anomalies, or significant family history, are referred to CGS for a formal genetic consultation before any genomic investigations would be offered. The waiting time for the first appointment is long. In order to alleviate the situation, there has been a modification of the referral mechanism since 2018 between CAS and CGS, after the published results of this study. An expedited pathway is now offered for children with unexplained ID (moderate ID or below diagnosed after intellectual assessment at 5 years old or above). Pre-genetic counselling is provided by a paediatrician at CAS, followed by direct blood examination for CMA and Fragile X syndrome (FGX) testing at CGS. Consultation with a geneticist will be arranged if either CMA or Fragile X testing yielded abnormal outcomes. Otherwise, clients will receive a report issued by CGS stating no abnormalities detected in CMA and are not required to see a geneticist. This modification will significantly reduce the waiting time for both pre-testing genetic counselling and investigation turnover time in cases with unexplained ID.

In the coming future, the ideal approach may work towards the expedited mechanism for children with early-onset significant DD. It can avoid unnecessary investigations, thus lowering stress for both child and parent; importantly, it may reduce societal costs.

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Contact

Email: ro_cas@dh.gov.hk
Website: http://www.dhcas.gov.hk

Address: 2/F., 147L Argyle Street, Kowloon, Hong Kong