Definitions provide the mechanism to limit and demarcate the outlines of ‘something’ clearly, by explicitly providing its distinctive core features and immutable ‘essence’ or ‘quality.’ Definitions are declarative statements of meaning which are a dynamic product of usage and consensus which may change over time as our understanding of a concept changes. The objective of a definition is to provide for clarity and agreement in both verbal and written communication (Webster’s Encyclopedic Dictionary 1988).

Broadly speaking, ‘neurodevelopmental disability’ as a concept brings together under one rubric a group of related, but clinically distinct, chronic disorders whose essential and unifying feature is a documented disturbance in developmental progress, either quantitative or qualitative, or both, compared with established norms in one or more recognized developmental domains (American Psychiatric Association 1994). These domains traditionally include: motor (gross or fine), speech/language, cognition, personal/social, and activities of daily living. These domains are in themselves not mutually exclusive or independent. Collectively, these disorders are a common problem in child health which challenges practitioners at varying levels, including, but not limited to, accurate timely recognition, diagnosis, causation, intervention, resource allocation, and outcomes (First and Palfrey 1994).

Individual subtypes of neurodevelopmental disability defined below are essentially ‘symptom complexes,’ rather than specific disorders or diseases (Sherr and Shevell 2006). Etiologically heterogeneous, neurodevelopmental disability subtypes are diagnosed not by objective laboratory testing, but rather by observed clinical features. These subtypes can be conceptualized as ‘terms of convenience’ that encapsulate a number of commonly encountered children sharing impairments, which mandates a common diagnostic evaluation and approach, and broadly share medical requirements and therapeutic needs and individual/family challenges to participation and integration. In effect, they provide a shorthand way of communication between health professionals. Their conceptualization and recognition, in both a timely and accurate manner, are challenged by the awareness that the development of the child is an ongoing dynamic process involving the complex interplay of intrinsic (i.e., child) and extrinsic (i.e., families, environmental) factors, whose individual trajectories need not be smooth or consistent across time (Darrah et al.
There is also a wide variation of ‘normal’ to be accepted, and drawing a clear boundary line between normal and abnormal is often problematic when going from the general concept to the specific child. Ideally, the diagnosis of the subtypes outlined below should occur over time, rather than at a single point of clinical convenience (i.e. snapshot).

GLOBAL DEVELOPMENTAL DELAY
For the young child (i.e. less than 5 years of age), objective measurement of intelligence and cognition requires a consistent, standardized, and reliable measure of what is in essence an inferred concept. It also requires a universally agreed upon and widely applicable definition of intelligence itself. Both achieving this definition and an agreed upon measure of cognition have proved problematic, indeed elusive, to date in this age group (Gould 1981). Thus, the neurodevelopmental disability subtype of ‘global developmental delay’ has emerged to describe the child with limitations and delay in the widespread acquisition of skills that are directly observable and measurable in the context of the natural progression of all children (Simeonsson and Sharp 1992, Simeonsson and Simeonsson 2001). These skills are both developmental and functional in conceptualization and can be tracked systematically across time. Global developmental delay refers to a disturbance across a variety of developmental domains, which is defined operationally as a significant delay in two or more developmental domains (Shevell et al. 2003). From a practical standpoint, delay in two domains usually, but not invariably or necessarily, implies delay that is observable in all domains.

MENTAL RETARDATION/INTELLECTUAL DISABILITY
Global developmental delay may be an early marker of what is termed mental retardation, which is typically diagnosable in the child older than 5 years of age. The present consensus definition of ‘mental retardation,’ put forward by the American Association on Mental Retardation (now the American Association on Intellectual and Developmental Disabilities) in 2002, describes this entity as ‘a disability characterized by significant limitation both in intellectual functioning and adaptive behaviour as expressed in conceptual, social and practical adaptive skills’ (American Association on Mental Retardation 2002). Thus, the definition of mental retardation goes beyond a significant limitation in intelligence or cognition conceptualized as general mental capabilities, which is best objectively captured through an ‘intelligence quotient’ (IQ) score (subject to its own potential intrinsic errors in measurement) to include limitations in adaptive behavior (Spreat 1999). Adaptive

1 UK usage: learning disability.
behavior is conceptualized as skills that an individual learns in order to function in the context of his/her everyday life. These skills include those that are conceptual, practical, and social in orientation. Limitations in these skills impair performance and substantially dampen the ability to anticipate correctly and respond to the changes and demands encountered in daily life. Standards with respect to personal independence and social responsibility are not met. Intellectual disability is the currently preferred term for mental retardation (Schalock et al. 2007).

Clearly, global developmental delay and mental retardation are complementary, but non-synonymous, terms that share common features yet have distinctive characteristics (Sherr and Shevell 2006). At their core, they both represent a defect or disorder in learning. Though standardized testing solidifies their accurate diagnosis, the reality is that in practice experienced clinical judgment is typically substituted for such detailed intensive standardized testing (Sherr and Shevell 2006). Still to be determined through longitudinal studies is the precise relationship between these two related entities (Peterson et al. 1998).

DEVELOPMENTAL LANGUAGE IMPAIRMENT

‘Developmental language impairment’ is characterized by a predominant, almost exclusive, delay in the speech/language domain (Webster and Shevell 2004). Developmental language impairment features either an expressive or receptive deficit in the absence of any observed cognitive limitations, significant hearing loss or co-existing autistic features (Nass and Trauner 2006). A multitude of synonymous terms have been utilized including (but not limited to) specific language impairment, developmental language disorders, and developmental dysphasia. Various elements of language may be affected including articulation, phonologic decoding and programming, oral motor planning (dyspraxia), semantics, syntax, lexicon, and pragmatics. Though various classification schemes for developmental language impairment have been put forward, consensus agreement on the precise subcategorization for this neurodevelopmental disability subtype remains elusive thus far.

Longitudinal studies have emphasized that the delay in language skills initially observed in the child is not merely one of maturational delay (Shevell et al. 2005). Indeed, the documented delay may over time not be solely restricted to the language domain, with early language delay perhaps functioning as a harbinger of later observed and more early recognized cognitive difficulties. Careful studies at the time of diagnosis have suggested that these children may often have overlooked, indeed subtle, neurologic and motor difficulties that may impact on overall developmental trajectory and thus have therapeutic implications (Webster et al. 2008). Thus, labeling a child originally with a developmental language impairment may be a matter of the language domain’s degree and predominance of impairment, rather than the apparent exclusivity of this domain’s deficit.
AUTISTIC SPECTRUM DISORDERS

‘Autistic spectrum disorders’ have also been known by various terms including autism syndrome and pervasive developmental disorders. First described in the 1940s, diagnostic criteria for this neurobehavioral syndrome have been extended over time, resulting in more children with milder and varying degrees of deficits meriting the ‘autistic’ diagnostic label (Hirtz et al. 2006). This broadening clinical phenotype accounts for much of the recent observed ‘autism epidemic.’

Present criteria encapsulated in the Diagnostic and Statistical Manual of Mental Disorders, 4th edition revised (DSM-IVR) define a range of deficits that reflects a quantitative and qualitative impairment in both reciprocal social interactions and language skills, combined with a restrictive repertoire and stereotyped patterns of behavior, interest, and activities (American Psychiatric Association 1994). The qualitative impairment in social interaction is signaled by abnormalities in non-verbal behavior, peer relationships, sharing with others, and demonstrated reciprocity. Language impairment is signaled by actual delay, repetitive idiosyncratic or non-varied language use, and conversational limitations with respect to initiating and sustaining a conversation. Motor mannerisms, adherence to routine, and observed restricted patterns of interests or bizarre preoccupations frequently characterize behavior in the autistic child. Among children with autistic spectrum disorders, there is marked variability in the precise distribution and actual severity of symptoms. Cognitive function also can be highly variable between affected individuals.

Diagnosis of an autistic spectrum disorder requires an application of a DSM-IVR diagnostic checklist or standardized interview (e.g. Autism Diagnostic Interview (ADI), Autism Diagnostic Observaton Schedule (ADOS)) or the application of a widely accepted autism rating scale (e.g. Childhood Autism Rating Scale (CARS)) (Hirtz et al. 2006). Some children may not reach the threshold for formal diagnosis of an autistic spectrum disorder; thus the label of a ‘pervasive developmental delay – not otherwise specified’ or a ‘global developmental delay with autistic features’ may be warranted (Shevell et al. 2001). The distinction between these two entities is not clear at this time and they may be synonymous with widely overlapping terms or diagnostic formulations.

CEREBRAL PALSY

The concept of ‘cerebral palsy’ dates from the latter part of the 19th century (Ingram 1994) and the penultimate consensus definition states that cerebral palsy is ‘an umbrella term covering a group of non-progressive, but often changing, motor impairments syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development’ (Mutch et al. 1992). Cerebral palsy is clearly conceptualized as a symptom complex featuring heterogeneous etiology, pathologies, and clinical
manifestations (Shevell and Bodensteiner 2004) and the most recent consensus definition highlights a disorder of movement and posture with activity limitations and a varying range of associated sensory, behavioral, cognitive, perceptual, language, medical, and musculo-skeletal difficulties (Rosenbaum et al. 2007).

The non-progressive nature of cerebral palsy refers to the underlying pathologic lesions that neither progress nor resolve once they occur. Neurodegenerative, neurometabolic, and neoplastic processes do not underlie cerebral palsy; however, some controversy exists regarding the inclusion of potentially progressive vascular (i.e. moyamoya) or traumatic (i.e. shaken baby syndrome) etiologies under this term. While the pathologic lesion may not change, clearly the clinical manifestations of a child’s cerebral palsy may evolve over time, reflecting the interplay of the lesion with a dynamic maturing nervous system (Badawi et al. 1998).

Motor impairment is at the core of cerebral palsy (Minear 1956) and is manifested by motor delay, gait disturbance, and objective neurologic findings involving changes in tone (typically increased), passive resistance to stretch, the quality of possible limb movements (i.e. dyskinesias), the presence of primitive reflexes, and the exaggeration of stretch reflexes. The combination and severity of observed features and the resulting impairments are highly variable. Furthermore, the observed motor difficulties may co-exist with developmental impairments in other domains (e.g. global developmental), cognitive and/or adaptive limitations (e.g. mental retardation), primary sensory impairments (e.g. hearing or visual loss), learning disorders (e.g. learning disability or attention-deficit–hyperactivity disorder), or a seizure disorder (e.g. epilepsy) (von Wendt et al. 1985).

By definition, only lesions affecting the brain can result in cerebral palsy and only those occurring in the early stages of brain development prior to its complete maturation (Mutch et al. 1992). The precise timeframe corresponding to this early stage has not been consistently and universally agreed upon (Stanley et al. 2000). This has led to local geographic variation in etiologic patterns for cerebral palsy reported depending on the precise local ‘early’ timeframe employed.

GROSS MOTOR DELAY
Like developmental language impairment, ‘gross motor delay’ is delay affecting exclusively or predominantly a single developmental domain, in this instance motor skills (either gross or fine in character). It is a single domain developmental delay (Shevell et al. 2000). Children with isolated cerebral palsy and no other impairments have gross motor delay, but not all children with gross motor delay have cerebral palsy. This is so because some children with gross motor delay will either not have objective neurologic findings, aside from the motor delay itself, or they may indeed manifest an etiology arising not from the brain but rather from the spinal cord or peripheral nervous system (i.e. lower motor unit).
OTHER NEURODEVELOPMENTAL DISABILITIES
While primary sensory impairments occur at increased frequency in the setting of the neurodevelopmental disabilities, occasionally they occur in isolation, manifesting as either hearing loss (i.e. ‘deafness’) or visual impairment (i.e. ‘blindness’), which brings forth unique and particular challenges to the affected child, family, and those involved in primary care, especially from a rehabilitative perspective. Previously intact school age children may manifest, in the context of school attendance, either a ‘specific learning disability’ or an ‘attention-deficit–hyperactivity disorder,’ both of which can be considered as a neurodevelopmental disability subtype. Developmental co-ordination disorder is a recently parsed and defined neurodevelopmental disability that provides a diagnostic label to children previously considered as ‘clumsy’ or ‘maladroit.’ Diagnosis of this particular neurodevelopmental disability is based on currently formulated DSM-IVR criteria.

INTERNATIONAL CLASSIFICATION OF FUNCTIONING, DISABILITY AND HEALTH
The above definitions reflect a pronounced and somewhat narrowly configured biomedical approach which may be overtly limiting or narrow when considering chronic health conditions such as the neurodevelopmental disabilities. While not altering the basic unifying definitions in any way, a complementary framework is offered by consideration of the International Classification of Functioning, Disability and Health (ICF) (World Health Organization 2001). The product of a decade of effort by the World Health Organization (WHO), the ICF offers a different model for disability and functioning which is more holistic and robustly biopsychosocial in orientation (Fig. 1.1).

This model emphasizes the continual bi-directional dynamic interaction between contextual factors and health conditions (Rosenbaum and Stewart 2004). Contextual factors may be personal (i.e. age, gender, education, lifestyle) or environmental (i.e. social, cultural, institutional, financial). Disability is envisioned as a social construct, reflecting the unique product of the interactions between the individual and the broader family, community, and society in which the individual resides. In this schema, activity and participation are important potentially modifiable determinants of health that could – and should – be addressed through intervention (i.e. therapy). By being a social construct, the range of modifications that may affect disability is broadened considerably. Indeed, desired modifications are individualized to fit the particular needs of the individual family unit. Furthermore, by expanding outcomes beyond that of the modification of body structures or function alone, the potential for therapeutic change and benefit is also considerably expanded.

The ICF model can be applied in a unique and particular way to each child with a neurodevelopmental disability. Within this framework, properly speaking
the disability does not rest with the child. This has led to the emergence of a ‘family centered care’ approach to intervention (Rosenbaum et al. 1998). Rather than fixing the deficit, however physiologically defined, therapeutic efforts are directed at resolving difficulties and barriers to participation and activity that are identified by the child and family as subjectively desired goals. Together with our emerging scientific understanding of childhood neurodevelopmental disability, this offers the hope of improving overall outcomes for these children.

ACKNOWLEDGMENTS
MS is grateful for the support of the MCH Foundation and YCC during the writing of this manuscript. Alba Rinaldi provided the necessary secretarial assistance.

REFERENCES


CONCEPTS AND DEFINITIONS


