Developmental Disorders

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ABSTRACT

Developmental disorders are a diverse group of physical, cognitive, psychological, sensory and speech impairments that appear anytime during development up to 18 years of age. Developmental disorders are heterogeneous group of disorders and have a prevalence rate ranging from 1.49% for severe developmental disorders to 16.8% for the entire spectrum of developmental disorders during childhood. In this chapter, the authors discuss three most commonly encountered developmental disorders — Mental Retardation, Pervasive Developmental Disorders, and Specific Developmental disorders. There is a detailed discussion of the clinical features, etiology, differential diagnosis along with management of these disorders.

INTRODUCTION

According to Centres of Disease Control, “Developmental disorders are a diverse group of physical, cognitive, psychological, sensory and speech impairments that appear anytime during development up to 18 years of age”.

Children with these disorders attend medical services not only because of the core signs and symptoms of these disorders but also because of the associated spectrum of behavioural and emotional complications. Thus, as found across studies and also from clinical experience, developmental disorders are often associated with social, cognitive, psychiatric concerns resulting in the challenges that persist throughout the lifespan. In this chapter, we will discuss these developmental disorders — their presenting symptoms, diagnostic criteria, etiology, prevalence, and management.

MENTAL RETARDATION

Mental retardation (MR) is a descriptive term for subaverage intelligence and impaired adaptive functioning arising in the developmental period (< 18 years). MR and other neurodevelopmental disabilities are seen often in a general paediatric practice.
Developmental delay is often used inappropriately as synonymous with MR. Developmental delay is an overly inclusive term and should generally be used for infants and young children (< 5 years of age) in which the diagnosis is unclear, such as those too young for formal testing.

Approximately 10% of children have some learning impairment; while as many as 3% manifest some degree of MR. MR originates during the developmental period (i.e., conception through age 18 years) and results in significantly sub-average general intellectual function with concurrent deficits in functional life skills. The diagnosis of MR requires an intelligence deficit of at least 2 standard deviations (SDs) below the mean IQ. This generally translates into an intelligence quotient (IQ) score of 70-75, given a population mean of 100. Along with low IQ, there should be deficits in at least two areas of functional life skills or adaptive skills to meet the diagnostic criteria for MR. Adaptive skills encompass functional life skills within the domains of communication, self-care, home living, social and interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health, and safety.

Table 1. Categorisation of MR by IQ

<table>
<thead>
<tr>
<th>Mental Retardation level</th>
<th>IQ Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>50-70</td>
</tr>
<tr>
<td>Moderate</td>
<td>35-49</td>
</tr>
<tr>
<td>Severe</td>
<td>20-34</td>
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<tr>
<td>Profound</td>
<td>&lt;20</td>
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</tbody>
</table>

Etiology of Mental Retardation

Some of the identified etiological factors are summarized in Table 2:

Table 2: Etiological Factors

| Genetic                  | i. Chromosomal Abnormalities e.g., Down’s Syndrome, Fragile X syndrome, Turner’s syndrome, Klinefelter’s syndrome.  
| Perinatal Causes         | ii. Inborn Errors of Metabolism involving amino acids (phenylketonuria), lipids (Gaucher’s Disease), carbohydrates etc.  
| Acquired Physical Disorders | iii. Single Gene disorders like tuberous sclerosis, neurofibromatosis.  
|                          | iv. Cranial abnormalities like microcephaly  
|                          | i. Infections like rubella, syphilis  
|                          | ii. Prematurity  
|                          | iii. Birth trauma  
|                          | iv. Hypoxia  
|                          | v. Intrauterine growth retardation  
|                          | vi. Placental abnormalities  
|                          | vii. Hypoxia  
|                          | i. Infections especially encephalitis  

The table of etiological factors continues...
Comorbid psychiatric conditions are diagnosed more frequently in those with intellectual disabilities than in the general population. Even so, psychiatric disorders probably are underappreciated in this population. Attention deficit/hyperactivity disorder (ADHD) is diagnosed in 8-15% of children and 17-52% of adults with MR. Self-injurious behaviours require treatment in 3-15%, particularly in the severe range of MR. Major depression, bipolar disorders, anxiety disorders, and other mood disorders are the most common psychiatric diagnoses in adults with MR. Obsessive-compulsive disorder, conduct disorder, tic disorders, and other stereotypic behaviours are also diagnosed more commonly in those with MR. Schizophrenia may have a prevalence of 3% in individuals with MR, compared to 0.8% in the general population. In day-to-day clinical practice, as many as 30% of children with MR are observed to have an emotional or behavioural disorder. Occult visual and auditory deficits occur in 50% of those with MR, particularly when refractive errors are considered. One in five individuals with MR also has cerebral palsy (CP); however as many as 20% of individuals with MR have seizures. A profound social morbidity affects individuals with MR and their families. This morbidity can be measured in lost wages, dependence on social services, impaired long-term relationships, and emotional suffering.

**PERVASIVE DEVELOPMENTAL DISORDERS**

The term Autism Spectrum Disorders (ASD) also known as Pervasive Developmental Disorders [PDDs] refers to early onset, developmental disorders of varying clinical presentation characterised by pervasive qualitative impairment, delay and deviance in the development of reciprocal social, communicative and other skills. These ASDs include Autistic disorder, Rett’s Syndrome, Childhood Disintegrative Disorder, Asperger’s Syndrome and PDD, not otherwise specified. For the diagnosis of ASD, the child must have clinically significant, persistent deficits in social communication and interactions, as manifest by all of the following:(a) Marked deficits in nonverbal and verbal communication used for social interaction; (b) Lack of social reciprocity; and (c) Failure to develop and maintain peer relationships appropriate to developmental level. The restricted, repetitive patterns of behaviour, interests, and activities, are manifested as stereotyped motor or verbal behaviours, or unusual sensory behaviours; excessive adherence to routines and ritualized patterns of behaviour; and/ or restricted, fixated interests. Although all ASDs involve impairments in reciprocal social interaction skills, the degree of impairment in communication skills and cognitive abilities and the form disorder is associated with a slightly different set of diagnostic criteria as described below.
Autism

*Autistic Disorder* is also known as infantile autism, childhood autism and early infantile autism. Current diagnostic criteria for autism specify multiple impairments in social functioning and communication as well as restricted and repetitive behaviour present before the age of 3 years. The manifestations of this disorder vary greatly in terms of the degree of impairment, ranging from early estimates of 75% of individuals having comorbid mental retardation to recent reviews suggesting as few as 30% to 60% of individuals with this specific comorbidity (Fombonne, 2003). People with autism experience substantial social impairments that have an impact on almost every aspect of their interactions with others. Peer interactions are often avoided, and play behaviours often remain stereotypic and lacking in pretence. Even within the first 12 to 18 months of life, difficulties in joint attention, social responsiveness, and eye contact are noted and have been confirmed by retrospective videotaped studies (Tidmarsh et al. 2003). These social impairments contribute greatly to subsequent language delays by dramatically limiting the number of meaningful learning opportunities during a critical developmental period. Many children with autism fail to develop language at all unless dramatic intervention procedures are pursued. Restricted and repetitive behaviours occur more commonly in older preschool- and school-aged children than in young children or adolescents and adults and commonly include hand flapping, toe walking, and rocking.

Asperger’s Disorder

The criteria for Asperger’s disorder share some similarities with autism. To qualify for a diagnosis of Asperger’s disorder, an individual must demonstrate at least two characteristic criteria in the area of impaired social interaction and one characteristic criterion in the area of restricted, repetitive, and stereotyped patterns. Individuals with Asperger’s disorder do not have clinically significant delays in language skills, cognitive development, or age-appropriate self-help skills or adaptive behaviour. In fact, individuals with Asperger’s disorder are often verbally fluent and have above-average intelligence in many areas, whereas clear deficits and learning disabilities may be evident in other areas. Finally, developmental milestones are often within normal to advanced limits; thus, identification of these children typically occurs at a later age as difficulties develop on entry into preschool or day care or into general education environments.

Rett’s Syndrome

This is a progressive condition that develops after some months of apparently normal development including normal perinatal head circumference. However, between five months to forty eight months (usually between six months to one year), head growth begins to decelerate followed by a loss of hand skills and appearance of midline stereotypic hand wringing movements or hand washing stereotypies. Once established, the disorder contains autistic features, such as impairments in social interactions, communication; and stereotyped behaviours. However, Rett’s disorder has enough distinguishing features to be considered an independent diagnostic entity. For instance, the course of the disorder is different from autism; with Rett’s disorder progressing to various forms of neurological impairments that are not seen in autism.
Childhood Disintegrative Disorder (CDD)

CDD is a rare condition characterised by: (1) onset of condition after a fairly prolonged period of normal development; and (2) marked deterioration in multiple developmental areas accompanied by development of various “autistic like features”. Children with CDD clearly resemble autistic children of the same intellectual level in terms of their behaviour, limited communication skills, pattern of long term outcome, and the need for various special services. Although available data are limited, the disorder has been reported to be quite rare.

Pervasive developmental Disorder not otherwise Specified

A diagnosis of PDD-NOS is used for milder problems on the spectrum when an individual displays a severe impairment in the development of reciprocal social interaction associated with verbal or nonverbal communication skills or with the presence of stereotyped behaviour, interests, and activities but without meeting criteria for another PDD.

Etiology

The cause of childhood autism is unknown. It is likely that the central abnormality is cognitive, affecting particularly symbolic thinking and language, and that the behavioural abnormalities are secondary to this cognitive defect.

Genetic influences have a major importance. The condition is 50 times more frequent in the siblings of affected persons than in the general population (Rutter et al. 1990). Several twin studies have shown a much higher concordance between monozygotic than between dizygotic twins. Autism has been linked to a region on chromosome 7q (International Molecular Genetic Study of Autism Consortium 1998), but further replication is required. Cognitive abnormalities are more frequent among the siblings of autistic probands than in the general-population, suggesting that the phenotype may be wider than-the- syndrome of autism-as currently defined (Bailey et al. 1998).

Organic brain disorder has been suspected as a cause of autism because there is an increased frequency of complications in pregnancy and child birth among these patients. Other evidence for biological causes Autism is associated with fragile X syndrome

Abnormal parenting — In a much-quoted paper, Kanner (1943) suggested that autism was response to abnormal parents who were characterised as cold, detached, and obsessive Kanner’s idea has not been substantiated. It is now thought that any psychological abnormalities in the parents are likely to be either a response to the problems of bringing up the autistic child, or a manifestation in-the parents, of the genes that have produced autism in the child.

SPECIFIC DEVELOPMENTAL DISORDERS

As already discussed mental retardation is a generalised impairment in nearly all areas of functioning. In contrast, the specific developmental disorders are characterised by an inadequate development in usually one specific area of functioning. The deficit in functioning may be in scholastic skills, speech and language, and motor skills. These may include reading, language, arithmetic, articulation or coordination. Sometimes more than one developmental disorder is present.
All the developmental disorders either cause impairment in academic functioning at school, especially when language is affected, or an impairment in the activities of daily living.

**Specific Reading Disorder**

Difficulties in reading accuracy (measured by tasks requiring decoding of single words) are distinct from difficulties with reading comprehension (measured by tasks tapping understanding of text). Whereas problems with decoding inevitably limit reading comprehension, there are a substantial minority of children who have difficulties with reading comprehension in the absence of problems with reading accuracy. Problems with word decoding are likely to predominate in the earliest years of schooling, whereas a later-emerging group of poor readers who cope with the initial stages of learning to read fare less well when the school curriculum places heavier emphasis on comprehension. Later, throughout adulthood, individuals may face persisting problems with spelling or written expression. The language in which children learn to read also affects the likelihood of reading difficulties, although data do not exist to allow fair comparison of prevalence rates, clinical experience suggests that children have more difficulty with English language than with Hindi.

**Spelling Difficulties**

Spelling problems may be seen as a direct consequence of difficulties in mastering the mappings between orthography (spelling patterns) and sound within the phonological pathway. The spelling errors made by people with dyslexia include both phonetic spelling errors, where the sound pattern of the word is represented accurately (e.g., biscuit ’biskit; chaos ’kaos), and phonetically unacceptable spelling errors (e.g., umbrella ’unbrl; adventure ’afveorl).

**Reading Comprehension Impairments**

In contrast to children with dyslexia, “poor comprehenders” decode well but have problems understanding what they read. Their difficulties often go unnoticed in the classroom because they can read aloud competently. Poor comprehenders typically have normal non-verbal abilities and good phonology; they do well on phoneme awareness tasks and on simple memory tests. However, they have neuropsychological impairments that encompass poor working memory, problems making inferences and deficits in metacognitive processes, such as comprehension monitoring. The reading comprehension impairments shown by these children are related to a range of oral language processing weaknesses.

**Specific Disorder of Arithmetic Skills**

Children experience different kinds of difficulty with numeracy skills. Some affect arithmetic (computation) whereas others affect more conceptual aspects of mathematics that require problem-solving. However, these are not well differentiated either in diagnostic manuals or in practice; tests of number skills differ in the extent to which they assess basic arithmetic as opposed to mathematical concepts (e.g., numerosity, magnitude, or geometry) and this leads to an unhelpful tendency to categorise all mathematical difficulties together. It is also important to note that mathematical learning is a cumulative process and therefore a basic deficit in computation will have downstream effects: addition, subtraction and multiplication processes are involved in higher-level mathematics, such as geometry, algebra and calculus. In addition, high levels of anxiety are often associated with
Developmental Disorders

Mathematics and these can pose obstacles to learning for children who find basic arithmetical tasks difficult. Children with arithmetic difficulties initially experience problems with the count sequence, as they grow older they may tend to rely on the “count-on” strategy in simple addition for longer than typically developing children, albeit error-prone, and they have difficulty in learning number facts. With more complex addition problems, they tend to guess more and less accurately than their peers.

**Developmental Co-ordination Disorder**

Developmental co-ordination disorder (sometimes referred to as “dyspraxia”) is a disorder of motor skills and is included here because problems with motor skills can adversely affect children’s educational achievements and self-esteem. Formally, the diagnosis of developmental co-ordination disorder (DCD) is used to describe problems of motor co-ordination that occur in otherwise normal children and significantly affect the activities of daily living. In practice, such problems often signify risk factors for other disorders, and DCD is commonly found in association with developmental disorders such as language impairment or autism, and the overlap with attention problems is high. The symptoms of DCD can vary considerably and may include gross motor difficulties, such as problems running, hopping, jumping, catching a ball and balancing, and fine motor difficulties including a lack of manual dexterity, difficulty in doing up buttons and laces, in dressing and in using eating utensils. Speech-motor skills can be affected and problems of pencil control are widespread. When DCD occurs in pure form, such children have been reported to have normal reading skills and only minor spelling problems even though their handwriting is usually very poor.

**MANAGEMENT**

Early identification of children with developmental disorders is necessary to begin receiving early intervention services for children from birth to 3 years of age and early childhood education services for children aged 3-5 years, which are known to improve outcomes. The mainstay of treatment of MR is developing a comprehensive management plan for the condition. The complex habilitation plan for the individual requires input from care providers from multiple disciplines, including special educators, language therapists, behavioural therapists, occupational therapists, and community services that provide social support and respite care for families affected by MR. Adaptive equipment (e.g., for nonambulatory patients) and extra time (e.g., double time slots) may be required to accommodate such patients. In addition, family members or other support persons may be helpful. Written plans are helpful for interdisciplinary team communication. Physical activity and obesity are major contributors to various physical diseases in people with MR. Very few programs exist that target healthy lifestyles (nutrition/diet, exercise, self-care, stress reduction) in those with MR. Written, verbal and pictorial forms of communication as well as gestures and demonstrations are helpful for those with MR to ensure mutual understanding and improve treatment adherence. No treatments are available specifically for cognitive deficiency. Some specific treatments are discussed below:

**Behaviour Therapy**

Behavioural modification is effective for treatment of self injurious behaviour, pica and stereotypies in individuals with MR and PDD. Behaviour modification should be generalisable, consistent, and
focus on replacing maladaptive behaviours with adaptive prosocial behaviours/ skills. Change is
effected via training parents and adults to provide structure and reinforce behaviours. Once target
behaviours have been identified, they should be specifically described. A detailed behavioural analysis
must be carried out before starting modification. Evaluation should consist of assessing triggers of
the behaviour (antecedent), response of others (consequence) and responses to the consequences.
Continuity and consistency are important to maintain across caregivers and settings. If responses
are inconsistent, behaviours would be difficult to change. Positive reinforcement, negative
reinforcement and redirection should be considered. Studies with children with PDD have found that
behaviour therapy is most successful when principles of applied behaviour analysis are incorporated.
Functional behavioural assessment and analysis seek to examine the function of a behaviour, thereby
elucidating the cause of or motivation behind the behaviour. Functional analysis incorporates
manipulating or changing environment to effect behavioural change.

Social Interaction Interventions

Facilitating social skills has been a component of treatment for children with MR/PDD. This
area of intervention has been especially researched for individuals with PDD, given the hallmark core
delays in social development. Of the various facets of social interaction, interventions have been
found to improve initiation of social interaction, greeting others, responding to peers, perpetuating
interaction, taking turns, and sharing. Individuals with autism can be taught social interactions, and
generalise social interactions.

Many different approaches are currently used and being studied. These interventions include
changing the environment, developing collateral skills, developing child specific skills, and training
peers. “Collateral skills” training focuses on the development of activities that subsequently result in
improvement in social interactions. Studies find that individuals with PDD who are taught how to
play or how to ask questions can increase social interaction in other situations. “Child specific”
interventions which include social skills training, focus on developing social skills, teaching how to
initiate and sustain social interactions, and reinforcing social behaviour. This form of intervention has
been more effective in increasing the social initiation than sustaining the interaction. Peer mediated
training interventions focus on training peers to initiate interaction, engage and interact and increase
communication, with children with PDD.

Picture exchange communication system (PECS) (Bondy et al. 2001), a form of augmentative
and alternative communication (AAC) is a relatively newer intervention specially designed for children
with autism based on principles of applied behaviour analysis (ABA) and uses pictures instead of
words to help children communicate. PECS leads to improvement in communication of children
with autism who have difficulty in approaching another person (Malhotra et al., 2010) and in PECS
the child is made the incharge of the communication and since he is not expected to speak the initial
approach becomes less intimidating.

Cognitive Retraining

Cognitive retraining (CR) seeks to directly improve and/or restore cognitive functions utilizing a
variety of pen and paper or computerized tests or games requiring cognitive skills such as attention,
planning, problem-solving, and/or memory. Recent studies have reported the positive effects of CR interventions to ameliorate the known neurocognitive deficits in children with learning disability (Malhotra et al. 2009).

**Remedial/ Special Education**

Individualised education programmes based on each individual child’s strengths and weaknesses is to be planned for children with developmental disorder.

**REFERENCES**


