Clinical Practice Guidelines for Assessment and Management of intellectual disability

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INTRODUCTION

Mental retardation is a developmental disorder and is associated with significant limitations in intellectual functioning and adaptive behaviors. Currently, it is widely referred to as "intellectual disability (ID)" and "intellectual developmental disorders (IDDs)." In India, the Rights of Persons with Disabilities Act (2016) has introduced the term "intellectual disability" in the place of "mental retardation." However, India being a signatory country to the World Health Organization (WHO), where the International Classification of Diseases, 10th revision (ICD-10) guidelines are adopted in the clinical practice, the term "mental retardation" is still in clinical use (The WHO Working Group on the Classification of Intellectual Disabilities has recommended replacing the term "mental retardation" with "IDD" in ICD-11 [Salvador-Carulla *et al*., 2011]. Thus, both the terms, intellectual disability and mental retardation, are in use in India. Despite variation in the terminology and the differences in the criteria for diagnosis (e.g., ICD-10; Diagnostic and Statistical Manual of Mental Disorders, 5th Edition [DSM-5]) and assessment of disability (as notified in the guidelines in January 2018, which are based on the RPD Act), it is commonly agreed that significant impairments in intellectual functioning and adaptive behavior during the developmental period is the hallmark of the condition [Table 1].

It is estimated that nearly 2.5% of the global population will have low levels of intellectual functioning commensurate with ID. However, a wide variation in point prevalence of ID has been reported in India, from around 1/1000 to 32/1000, depending on the case definition, methodology, and population selected. An important point that can be noted in the literature is that prevalence rates vary depending on whether deficits in either intellectual functioning or adaptive behavior or both are considered. Although ID is recognizable in infancy or early childhood, it is often difficult to accurately diagnose it before 5 years of age. Hence, global developmental delay (GDD), which often predicts future development of ID, is used as a surrogate marker in children between the age group of 3 months and 5 years. Shevell *et al*. (2008) defined GDD as evidence of significant delay in two or more of the following developmental domains: gross/fine motor, speech/language, social/personal, cognition, and activities of daily living. However, not all cases of GDD may have cognitive deficits or end up as ID. Males are diagnosed with ID 30% more than females, especially in the milder ID range. However, this difference seems to disappear when the ID is more severe. ID is also associated with high morbidity and extreme costs of care. ID can cause significant impact on the individual, families, health-care system, and state.

Nature and needs of the condition

ID is a permanent condition therefore it creates special needs for both the individual and the family across the life span. The needs could be related to independent mobility, physical care, communication needs, modified curricula, aids and appliances, occupational and vocational opportunities, and medication if there are treatable, comorbid medical conditions. The special needs may necessitate support in varying degrees throughout the life span. Therefore, holistic programs should address the lifelong needs in a step-by-step fashion. For instance, when a child with ID is in preschool years,

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Table 1: Terminology and conceptual issues related to intellectual disability among different diagnostic systems

System	Term	Definition	Intellectual functioning	Adaptive behavior	Developmental period
ICD-10[a,1]	Mental retardation	It is a condition of arrested or incomplete development of the mind, which is especially characterized by impairment of skills manifested during the developmental period, which contribute to the overall level of intelligence, i.e., cognitive, language, motor, and social abilities	Components are cognition, language, and motor and social skills An intelligence quotient of 70 is the cutoff It categorizes ID into four severity levels that are based on IQ	Not clearly defined It is implied that assessment of adaptive behavior is part of assessment of intellectual functioning	Not explicitly defined, but understood to consider it as 18 years
DSM-5[b]	Intellectual disability (intellectual developmental disorder)	Intellectual disability (intellectual developmental disorder) is a disorder with onset during the developmental period that includes intellectual and adaptive functioning deficits in conceptual, social, and practical domains	Components are reasoning, problem-solving, planning, abstract thinking, judgment, academic learning, and learning from experience On standardized tests of intelligence, score of 65-75 is considered to indicate intellectual disability; where the test quotients have standard deviation of 15 and mean of 100 and standard error of 5	Deficits on adaptive functioning result in failure to meet developmental and sociocultural standards for personal independence and social responsibility Without ongoing support, the deficits will affect one or more activities of daily life, such as communication, social participation, and independent living across multiple environments Nomenclature of severity levels is same as it is in ICD-10, but the levels are decided based on the deficits only in adaptive functioning DSM-5 has explained the adaptive behaviors in each of the three domains of intellectual functioning such as conceptual, social, and practical domains in reference to the severity level and age	Defined as 18 years
Rights of Persons with Disabilities Act, 2016[c]	Intellectual disability	Intellectual disability, a condition characterized by significant limitation both in intellectual functioning (reasoning, learning, and problem-solving) and in adaptive behavior which covers a range of day-to-day, social, and practical skills	Like ICD-10, it has adopted the IQ cutoff of 70 for ID, and the same terminology to denote severity levels, but with different cutoffs. The severity levels are based on the scores of the Vineland Social Maturity Scale (a standardized, normative measure adaptive behavior scale) Profound disability=0-20 (100%) Severe=21-35 (90%) Moderate=36-54 (75%) Mild=55-69 (50%) Borderline=70-84 (25%) Note: Borderline disability is not a benchmark disability	Adaptive behavior is not defined but is understood to cover a range of day-to-day, social, and practical skills Scores on Vineland Social Maturity scale (a standardized, normative measure adaptive behavior scale) are considered to define the severity of ID	Not explicitly defined, but understood to consider it as 18 years
1CD-11 Working Group on Intellectual Disability (2011)[d; 1]	Intellectual developmental disorders	A group of developmental conditions characterized by significant impairment of cognitive functions, which are associated with limitations of learning, adaptive behavior, and skills	The working group advocated continuing clinical severity levels mentioned in ICD-10 due to their diagnostic and clinical utility. Therefore, IQ score should be considered as a clinical descriptor among others that are considered important in determining the severity levels of ID	Adaptive behavior is not defined but implied that difficulties in adaptive behavior will manifest in meeting the demands of daily life expected for one's age peers, cultural, and community environment. These difficulties include limitations in relevant conceptual, social, and practical skills	Not explicitly defined, but understood to consider it as 18 years

[a]World Health Organization (1992). ICD-10 Classification of Mental and Behavioural Disorders. Geneva: World Health Organization, [b]American Psychiatric Association (2013). Diagnostic and Statistical Manual of Mental Disorders, 5th ed.. Arlington, VA: American Psychiatric Association, [c]Government of India. Rights of Persons with Disabilities Act. New Delhi: Government of India, 2016, [d]Salvador-Carulla L, Reed GM, Vaez-Azizi LM, Cooper S-A, Martinez-Leal R, Bertell M, *et al*. Intellectual developmental disorders: Towards a new name, definition and framework for "mental retardation/intellectual disability" in ICD-11. World Psychiatry 2011; 10:175-180, [1]Need to refer to ICD-11 final guidelines as and when they become operational. ICD-10 – International Classification of Diseases, 10th revision; DSM-5 – Diagnostic and Statistical Manual of Mental Disorders, 5th Edition; ID – Intellectual disability; IQ – Intelligence quotient

the needs may center around self-care, sociocommunication skills, and school readiness skills but not so much about independent living or literacy. Similarly, for an adolescent with ID, the needs could be about education, prevocational training, and future independent living. As these examples indicate, ID will imply long-term, multidisciplinary approach to intervention for optimum outcome.

Etiological workup of intellectual disability

The etiology of ID/GDD is heterogeneous. The cause for ID and GDD can be nongenetic/environmental or genetic. Nongenetic causes such as prenatal infections, substance use like alcohol intake during pregnancy, and postnatal meningoencephalitis account for only one-third of cases and the rest are of genetic origin. The flowchart in Figure 1 is a modified version of the "Finnish approach." This provides a means for systematic etiological evaluation of ID. The common causes are also listed in the flowchart [Figure 1].

This modification of the Finnish approach at the preliminary level reliably distinguishes probable genetic and nongenetic etiologies in a majority of cases. Most of the nongenetic causes produce ID which is usually static in nature and potentially amenable for training. Further, the nongenetic causes in the subsequent pregnancies are either treatable or preventable, especially the maternal factors such as malnutrition, diabetes, teratogenic drugs, and substance use. The Finnish approach also paves way for further diagnostic and genetic testing among those cases initially suspected to be of genetic etiology. Genetic diagnosis is essential not only for accurate genetic counseling of recurrence risks and prenatal diagnosis, but also for appropriate management. This is in the light of newer strategies of treatment made available through thorough understanding of the pathophysiology of genetic disorders for several disorders. With the advances in the field of therapeutics, over eighty potentially treatable disorders have been identified. Majority of such inherited cases can be accurately diagnosed, provided that advances in the field of genetic diagnostics are utilized.

Comorbidities

Medical comorbidities

Various medical comorbidities are often associated with ID. Depending on the etiology, varying degrees of both neurological and nonneurological comorbidities are encountered. Some are a consequence of ID itself. Few of the common medical comorbidities are the following: epilepsy, spasticity, dystonia, ataxia, visual impairment, hearing impairment, congenital heart disease, cleft lip and cleft palate, limb anomalies such as congenital talipes equinovarus, congenital dislocation of hip joint, renal malformations, failure to thrive with vitamin and mineral deficiencies, recurrent infections, feeding disorder, and short stature.

Epilepsy is a common comorbidity with a prevalence of nearly 15%–30%. With increasing severity of ID, the prevalence increases to around 50%. Similarly, many electroclinical syndromes of epilepsies such as early infantile epileptic encephalopathies, West syndrome, and Ohtahara syndrome as well as other late-onset syndromes such as Lennox–Gastaut syndrome are invariably associated with ID. Ongoing seizures, especially if treatment refractory, often lead to developmental arrest. Such a condition remains a barrier against training and thereby against any hope of making developmental gains. Hence, it is essential that these disorders need particular attention and rigorous management.

Another domain of neurological disorder that impairs motor development and locomotion is the impairment in pyramidal, extrapyramidal, or cerebellar systems as well as combined. Spasticity, dystonia, tremors, and ataxia often lead to impairment in motor development and thereby successful locomotion. It is essential to differentiate the static or progressive forms of these disorders for appropriate counseling and management.

The medical comorbidities can be significant barriers for training and developmental learning and require attention in overall management. One can anticipate almost certainly the possibilities of physical disorders based on the etiology behind ID. With vast literature available on almost every one of the etiologies, we can predict and screen for the presence or absence of the comorbid medical disorders.

Behavioral and psychiatric problems

People with ID are 3–5 times at higher risk of any psychiatric disorder compared to the general population at all ages, with a cumulative prevalence of around 40%. It is conceivable that global cerebral functioning is affected by varying etiologies causing ID, which in turn can lead to a variety of neuropsychiatric manifestations. Besides these neurobiological underpinnings, social discrimination and deprivation can also influence the onset of psychiatric comorbidities in this group. Neuropsychiatric manifestations often commence insidiously with atypical presentation and are commonly written off as spectrum manifestations of ID. Hence, it is mostly underreported, misdiagnosed, and undertreated. This pattern of "diagnostic overshadowing and masking" is well documented. Limited choice in using structured diagnostic interviews is another barrier for accurate diagnosis of comorbid psychiatric condition. Eventually, only symptomatic treatment is resorted to, which may not solve the entire problems. Therefore, special efforts are required to identify behavioural and psychiatric disorders.

The etiology of ID can often provide clues to anticipate certain psychiatric comorbidities as certain behavioral phenotypes are frequently associated with some syndromes.

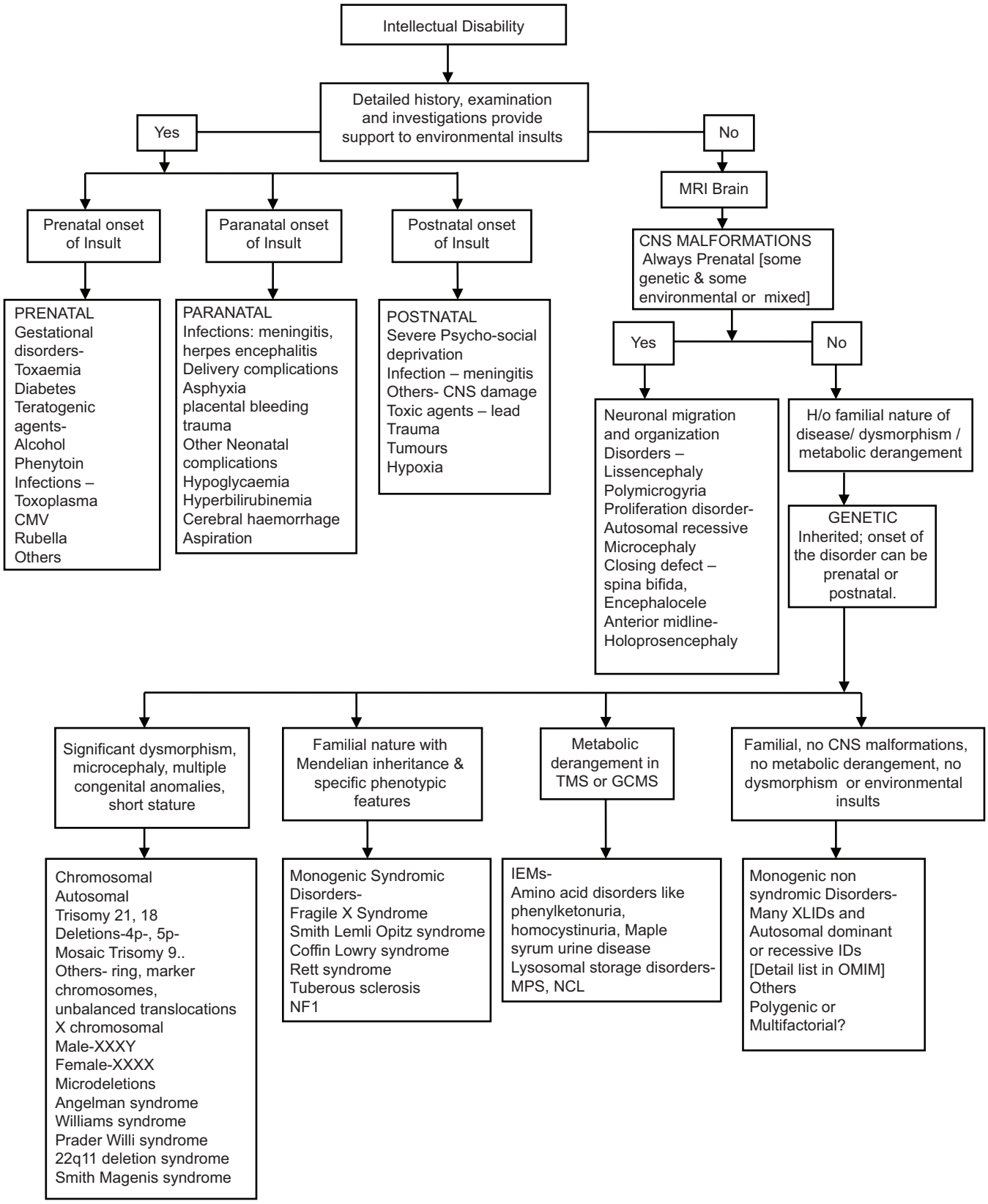

Figure 1: Finnish approach

Some examples are severe self-injurious behavior in Lesch–Nyhan syndrome; skin picking and obsessive-compulsive disorder in Prader–Willi syndrome; autistic traits and hyperactivity in Fragile X syndrome; self-hugging stereotypy and trichotillomania in Smith–Magenis syndrome; schizophrenia-like disorders in 22q11 deletion syndrome.

In majority, unspecified behavioral disorders are very common.

OVERVIEW OF ASSESSMENTS AND EVALUATION

Assessment is a process of collecting data for the purpose of making decisions. Assessment provides us with baseline information for intervention, whereas the evaluation is the assessment of outcome of an intervention. In clinical practice, therefore, we need both assessment and evaluation methods. The purpose of the assessment is as follows:

a. To identify the condition based on specific criteria and to establish that it is a clinical entity that requires appropriate mental health services and placement decisions
b. To identify and treat etiological factors and risk factors for ID
c. To identify the needs implicated by the condition and design a program plan to reduce the disability impact
d. To match the nature and needs of the conditions effectively with the best intervention methods available
e. To evaluate the effectiveness of intervention.

The following key questions could be asked to guide assessment, intervention, and outcome measure [Please see Appendix 1 for more details]:

- What is the nature of the delay – specific or global?
- Is the delay associated with significant limitations in intellectual functioning and adaptive behavior?
- Are there any comorbidities?
- Are there any treatable etiological conditions?
- What are the areas of intervention?
- Where and how the intervention should be carried out?
- Where the individual should be placed for maximum help (or, what are the existing agencies/service providers through which the interventions can be implemented)?
- How to evaluate the intervention outcome (or, what are the indices to stop intervening)?

DIAGNOSING INTELLECTUAL DIASBILITY AND ITS COMORBIDITIES

The diagnostic process of ID is similar to any other behavioral and mental disorders but with subtle differences. The diagnostic process involves history taking, observation including medical examination, intellectual and adaptive behavioral assessment, identification of comorbid psychiatric disorders, and need-based laboratory investigations for other medical conditions. Therefore, the diagnostic process encompasses several components that are described as follows:

History taking

The purpose of eliciting the history is to establish that there is an evidence for deficits in both intellectual functioning and adaptive behaviors that have an onset during the developmental period, to note possible etiology of ID, and to identify comorbidities and response to interventions, if any. Therefore, it requires interviewing of key people including the index patient and behavioral observation of the patient. Key people could be parents, caregivers, and service providers who know the birth and developmental history of the child.

A useful and comprehensive approach to assessment would include noting chief complaints in chronological order with mode of onset, duration, and precipitating event followed by a history of presenting illness and a detailed prenatal and perinatal history as a prelude. Developmental history in greater detail, particularly related to motor, language, and communication; self-help skills; socioemotional skills; cognition; and occupational skills/leisure-time activities; medical comorbidities and its treatments; psychiatric history including the details of onset, evolution, and current status of behavioral and other psychopathological disturbances; and treatment history. This should be followed by a comprehensive family history including the three-generation pedigree; consanguinity; family background; current living arrangements; and details of potential stressors, coping, and adaptation by the family.

Physical examination

It must involve routine systemic examination, anthropometric assessment, and observation of atypical morphological features suggestive of specific genetic disorders. Detailed physical examination helps to identify the etiology in a majority of cases, detect comorbid medical conditions, and also order appropriate investigations. Physical examination in cases with ID consists of three parts which are as follows:

Anthropometry

This provides indication toward nutritional status and underlying medical or genetic condition. The measures should include the following: height (length in case of neonates and infants), arm span, upper segment and lower segment lengths, sitting height, weight, head circumference, chest circumference, abdominal circumference, intercanthal and interpupillary distances, and palm and foot lengths.

Dysmorphology examination

Dysmorphology is the observation, documentation, and study of birth defects as well as syndromes. A thorough head-to-toe examination should be carried out to identify minor physical anomalies (MPAs), which provide clues toward etiological diagnosis, especially the genetic disorders [Table 2]. It requires keen observation and knowledge of normal versus abnormal morphology.

Examination of major organ systems

A systematic examination of all the organ systems to rule out multiorgan involvement and comorbid medical

conditions has to be performed for overall assessment and management. It is essential to be meticulous in observing and documenting the findings of physical examination as many of the MPAs can be easily missed. Hence, it may be important to take photographs or videos after informed consenting to document and revise the original findings at a later date. Some of the essential things to note are vision, hearing, locomotion (videos may help), and any major congenital anomalies. The presence of MPAs provides clues toward genetic versus nongenetic etiologies [Table 2]. Hence, branding every child universally with cerebral palsy which is often due to a nongenetic cause with a static course can be avoided. Presence of four or more MPAs should alert the physician toward probable genetic cause.

If MPAs are encountered in a child, such a case can be referred to a dysmorphologist/medical geneticist for further evaluation (the following are excellent sources for syndromes and standard terminology and definitions of MPAs – Smith's Recognizable Patterns of Human Malformations and "Elements of Morphology" in American Journal of Medical Genetics, 2009 [available from https://onlinelibrary.wiley.com/toc/15524833/149A/1]).

Further, progressive multiorgan dysfunction may be a clue toward a disorder of inborn error of metabolism which may be potentially treatable. Organ system examination is similar to any branch in medicine, and clinicians can refer to standard books like Hutchison's Clinical Methods.

Behavioral observation

The purpose of behavioral observation is to corroborate the clinical history with regard to intellectual functioning and behavioral repertoire. Therefore, it should start with observation of general appearance, any oddities in behavior, attention span, receptive and expressive speech abilities, and social and interpersonal abilities. Socioculturally appropriate stimuli could be presented to understand the level of general fund of knowledge, generic concepts, abstract thinking, reasoning, and problem-solving abilities that are not strictly dependent on academic learning. However, clinicians may use any standard format of general mental status examination for children to complement the behavioral observation.

Assessment of intellectual functioning and adaptive behavior

This step is to confirm the clinical diagnosis and identify the severity level of ID. Both ICD-10 and DSM-5 recognize the need for assessing the intellectual functioning with standardized tools that yield intelligence quotients (IQs). DSM-5 restricts the use of IQ to draw a cutoff of 65–75 (IQ 70± standard error of 5) for identifying ID. Conversely, ICD-10 advocates a IQ cutoff of 70 to identify ID and different IQ ranges for categorizing four severity levels such as, mild (IQ: 50–69), moderate (IQ: 35–49), severe (IQ: 20–34), and profound (IQ <20). The ICD-11 Working Group advocated that severity levels for IDD should rely on a clinical description of the characteristics of each subcategory, but the IQ score can be considered as one of the clinical descriptors that are important in determining the severity level. Therefore, till the time ICD-11 comes into force, the ICD-10 guidelines should be followed, which rely on IQ both for identifying the condition and ascertaining the severity levels of ID.

Clinicians may note that the choice of tests in the Indian context is limited notwithstanding the fact that the norms are in many cases are not revised [Appendix 2]. This is a major concern given the evidence for Flynn effect, which refers to observed rise in IQ scores over time and related norm obsolescence. Therefore, the IQ scores should not be rigidly interpreted.

Table 2: Some common minor physical anomalies and other findings on physical examination

Anatomical Region	Features
Scalp hair	Sparse, light colored, double whorl on scalp, easily breakable
Shape of skull	Brachycephaly, scaphocephaly, trigonocephaly, oxycephaly, plagiocephaly
Facial appearance	Coarse facies, elongated, triangular, small
Eyes and periorbital structures	Deeply set, prominent eyes, microphthalmia, upslanting/downslanting palpebral fissures, hypertelorism, epicanthal folds, strabismus, ptosis, bushy eyebrows, synophrys, microcornea, corneal clouding, cataracts, coloboma of iris, blue sclera, telangiectasia, etc.
Ears	Low set, small, large, malformed, anteverted, posteriorly rotated, preauricular tags, pits, cup shaped, etc.
Nose	Depressed nasal bridge, short and stubby, beak shaped, bulbous tip, flaring or hypoplastic nostrils, anteverted nares, etc.
Palate	High arched, ridged palate, clefting, bifid uvula, etc.
Chin	Prominent, retrognathia, micrognathia, etc.
Hands	Broad hands, short hands, simian crease, Sidney line, spade shaped, etc.
Fingers	Clinodactyly, brachydactyly, syndactyly, camptodacyly, arachnodactyly, polydactyly, broad thumb, etc.
Chest	Pectus excavatum, pectus carinatum, nipple anomalies, gynecomastia
Abdomen	Protuberant, scaphoid, umbilical hernia, hepato-splenomegaly, inguinal hernia
Spine	Kyphosis, scoliosis, spina bifida
External genitalia	Micropenis, macro-orchidism, undescended testis, ambiguous genitalia, hypospadias, absent secondary sexual characteristics, shawl scrotum, etc.
Skin	Dry and coarse, café-au-lait spots, abnormal pigmentation, hemangioma, ichthyosis, absence of sweating
Feet	Pes planus, pes cavus, valgus/varus anomaly, broad hallux, increased distance between the 1st and 2nd toes
Skeletal	Exostoses, increase carrying angle, joint hypermobility

When IQ tests are not applicable because of young age (e.g., children below 3 years) or associated sensory-motor issues and gross understimulation, standardized developmental scales (e.g., Developmental Screening Test and Developmental Assessment Scales for Indian Infants) can be used as applicable. The developmental tests yield "developmental quotients" which are interpreted in the same way as IQ scores.

With regard to the assessment of adaptive behavior, Vineland Social Maturity Scale (VSMS) is the only standardized measure available in India at present. The VSMS yields social quotient (SQ) and a profile of eight important domains of adaptive behavior. If the administration of VSMS is not possible for any reason, clinicians can ask socioculturally relevant questions to understand the level of adaptive behavioral functioning. If needed, DSM-5 list of specifiers for severity levels of ID could be referred to assess the adaptive behaviors till the time ICD-11 guidelines come up.

Wherever IQ and SQ indicate different severity levels of ID, decisions are taken in favor of SQ scores because the latter denotes the degree to which the index patient is able to meet the standards of culture-appropriate demands of daily life. Thus, SQ reflects the severity of ID better than IQ under ordinary circumstances. However, when assessment of the severity of ID by means of the usual procedures is rendered particularly difficult or impossible by associated sensory or physical impairments and severe behavioral disturbances, the condition should be identified as "Other mental retardation." If there is evidence of mental retardation, but insufficient information is available to assign the patient to one of the four categories or other mental retardation, it can be identified as "unspecified mental retardation." In case of "Other mental retardation" and "unspecified mental retardation," more information on developmental skill repertoire and periodical assessments of intellectual and adaptive behavior is desirable to infer the current level of functioning and associated severity levels of ID. Test selection should be proper if the person has comorbid sensory-motor impairments [Appendix 3]. Lastly, it must be recognized that the use of IQ and adaptive measures for clinical diagnosis is different from disability assessment and the latter has specific guidelines that must be strictly adhered to.

Confirmation of intellectual disability diagnosis

Based on the information obtained through case history, observation, and testing, ID could be coded into any of the six categories such as mild, moderate, severe, profound, other, and unspecified mental retardation. The ICD-10 has provision for using a fourth character to specify the extent of the behavioral impairment if this is not due to an associated disorder (e.g., F7x. 0 to denote "no, or minimal, impairment of behavior") and an additional code from the ICD-10 should be used if the cause is known (e.g., F72 severe mental retardation plus E00. [congenital iodine-deficiency syndrome]). Evidence for additional coding of etiological causes may come from laboratory findings.

Diagnosis of comorbid psychiatric disorder

Any changes in behavior compared to previous period, dip in overall functioning, and changes in vegetative functioning should be carefully recorded in each visit. If it is pervasive and indicative of a comorbid psychiatric disorder, it has to be carefully considered. A timeline method would be helpful when in doubt. During clinical evaluation, a greater reliance on onset and chronological evolution of symptoms, intensity, frequency, context of occurrence of symptoms, and precipitating and relieving factors elicited through careful interviewing of parents and caregivers will help in uncovering the psychopathology. School report is a valuable additional source of information. A period of behavioral observation rather than just traditional psychiatric interview will often help the clinician to decide on the presence and type of psychiatric disorder.

The behavioral observation will start from the moment the child enters the consultation room. Equal attention needs to be paid to child's behaviors, parental reports, as well as to verbal interview in arriving at conclusions. If necessary, the child and its parents must be interviewed separately. Playroom observation and multiple baseline observations for a functional analysis (Antecedent-Behavior-Consequences analysis) of symptoms are sometimes required.

Clinicians may need to create child-friendly space with appropriate toys, picture books, and art and craft materials. The setting should be safe, well lit, and ventilated. It is preferable that in-depth interview is conducted only after developing rapport with the child. The rapport could be developed by allowing the child to sit where he/she prefers to sit or move; asking about their age, likes, and pet name; and offering toys. It is important to build partnership with parents from the outset, which could be achieved by listening and valuing their opinions, impressions, and efforts and appreciating the parents for the right things they have done.

Depending on the language development and conversational skills, verbal interview can be conducted with simple, structured, clear, and concrete questions. It is better to avoid leading questions. The examination may include the following:

- Basics: Behaviors suggesting sensory-motor impairments or physical health issues
- Response to interview situation: Excited, fearful, tense, shy, inhibited, guarded, uncooperative, or defiant
- Alertness: Overaroused, withdrawn
- Attachment to parents and response to separation: Clinging, wanting to be carried all the time, indifferent to separation
- Sociability: Social orientation, approachability,

social responsiveness, eye-to-eye contact, reciprocal interactions, and awareness of social boundaries
- Motor activity level: Fidgetiness, restlessness, hyperactivity, lethargy
- Course of motor behaviors during interview or response to firm instructions: Quiet initially, but restless later on; unresponsive to firm instructions
- Impulse control: Snatching, spilling, falling, bumping, climbing, interfering, temper tantrums; aggressive acts such as biting, throwing, beating, pulling hair, slapping
- Attention and concentration: Goal directedness, task completion, distractibility
- Speech, language, and communication: Verbal/nonverbal comprehension and expression; vocabulary, articulation, and flow
- Mood: Inhibited, excessively cheerful, whining and crying, irritable
- Play behavior: Type of activity, duration, themes, etc.
- Other inappropriate behaviors: Any excess behaviors that are inappropriate to the age and sociocultural context
- Impressions on current developmental attainment: Whether excess behaviors or skill deficits are typical of a known psychiatric or developmental disorder?
- Parent–child interactions: Quality of engagement with child; communication patterns; degree and quality of control over the child; response to good and bad behaviors.

Standardized instruments such as Psychiatric Assessment Schedule for Adults with Developmental Disability, Reiss Screen for Maladaptive Behavior, Psychopathology Inventory for Mentally Retarded Adults, Developmental Behavior Checklist, and Psychiatric Instrument for the Intellectually Disabled Adults can be utilized as per the need. However, rating scales should be used only to complement the clinical observations.

LABORATORY INVESTIGATIONS

Often, it is difficult to completely examine children with ID due to their inability to communicate or comprehend commands or due to their behavioral issues. Some of the malformations can be missed in spite of an exhaustive and careful examination. Malformations such as atrial septal defect in early infancy, single kidney, holoprosencephaly, and mild hearing/visual impairment can be missed during routine examination, which can be barriers for adequate management of ID. As highlighted in the earlier sections, an array of etiological factors can result in ID and at least some of them can be potentially treated. Hence, a bunch of investigations are essential not only to identify the cause of ID, but also to make sure the treatable causes have been investigated for [Table 3].

Majority of Indian families do not possess medical insurance scheme. Hence, clinicians have to carefully consider the financial circumstances of the family, clinical hints, and treatability to order appropriate investigations. One should always consider the possibility of recurrence of the same disorder in the next pregnancy before an investigation is deemed unnecessary. This has to be discussed with the family and appropriate genetic counseling should be provided. Family can then chose to proceed or not to proceed with further investigations. Magnetic resonance imaging of brain and screening of metabolic disorders are considered mandatory investigations in all cases of ID. The American Academy of Pediatrics and American Academy of Neurology provide useful guidelines in this regard (Please refer to Moeschler (2008) and Michelson *et al*. (2011) for more details).

PSYCHOSOCIAL ASSESSMENTS

Persons with ID will be at a high risk for neglect and abuse. Therefore, risk assessment should be an integral part of comprehensive assessment plan in ID. Adaptive behavior is always impaired in people with ID, but the deficits are less evident in environments where support systems are in place. Hence, support systems available to the family and child must be reviewed. Therefore, psychosocial assessments are very important.

Assessment of family needs and functioning

Parents and families are the main source to implement the intervention plan in any condition that requires extensive long-term support. Specifically in the context of ID, studies indicate that their perceptions of the condition, disability impact, perceived support, and stress and coping mechanisms are very important moderators of intervention. Therefore, clinicians must consider assessing these areas further. Need may be appropriate tools such as the following could be used for this purpose: Global Entrepreneurship Monitor Questionnaire, Disability Impact Scale, Family Support Scale, Family Efficacy Scale, Family Needs Schedule (note: these scales are available in public domain at www.nimhindia.org/punblications); and Family Interview for Stress and Coping in Mental Retardation for assessing stress and coping of the parents of children with ID.

Psychoeducational assessments

With universalization of elementary education and the Right to Education act, many children with ID are in the mainstream as compared to a decade ago. Children both in the mainstream and in special school settings may need appropriate psychoeducational assessment. Tools such as the Grade Level Assessment Device and Functional Assessment Checklists for Programming (available at www.nimhindia.org/punblications) can be used for this purpose. Another gray area is the assessment of "school readiness skills" because there are no standardized measures. Often, children diagnosed with GDD or young children with ID are referred to mental health professionals for the assessment of school readiness skills. In such a scenario, a clinical assessment could be carried

Table 3: Physical investigations in intellectual disability

Test	Examples of conditions detected
Brain imaging with MRI and MRS*	CNS malformations, cerebral creatine deficiency, hypomyelinating and dysmyelinating disorders
Thyroid function test	Hypothyroidism
Advanced metabolic tests such as GCMS, TMS*	Fatty acid oxidation disorders, amino acid disorders, urea cycle disorders and organic acidurias
Enzyme studies	Tay-Sachs disease, metachromatic, leukodystrophy, some NCLs, MPS
Urine screen for mucopolysaccharides and oligosaccharides	MPS and Oligosaccharidosis
Karyotyping	Down syndrome, large deletions, ring/marker chromosomes, translocations
FISH and MLPA	Prader-Willi syndrome, William syndrome, Subtelomeric deletions
Chromosomal microarray	CNVs (many microdeletion duplication syndromes)
Next-generation sequencing/Sanger sequencing	Monogenic disorders such as Rett syndrome (MECP2 mutation), XLID, tuberous sclerosis, NF1
Repeat primed PCR	Fragile X syndrome
EEG	Epileptic encephalopathies such as West syndrome
Hearing evaluation (BAER)	Sensorineural hearing impairment
Visual evaluation	Wilson disease, cataract, optic atrophy, cortical blindness, refractive error
Blood group of child and parents	Rh iso-immunization
Immunologic tests (IgM antibodies)	TORCH infections (to be performed preferably within 6-8 weeks of delivery)
Investigations for organ system functioning	Cardiac malformations
ECHO	Renal malformations, nephropathy, hepatosplenomegaly due to storage disorders
Renal and Liver function tests with ultrasound abdomen	

*Mandatory investigations if obvious etiologies (such as Down syndrome, NF1) are not found clinically. GCMS: Gas chromatographic mass spectroscopy, TMS: Tandem mass spectroscopy, MRS: Magnetic resonance spectroscopy, MRI: Magnetic resonance imaging; FISH – Fluorescence *in situ* hybridization; MLPA – Multiplex Ligation-Dependent Probe Amplification; BAER – Brainstem auditory-evoked response; ECHO – Echocardiography; NCLs – Neuronal ceroid lipofuscinoses; MPS – Mucopolysaccharidoses; XLID – X-linked Intellectual Disability; CNS – Central nervous system; ID – Intellectual disability; TORCH – Congenital taxoplasma infection, rubella, cytomegalovirus and herpes

out by focusing on the following: sensory-motor abilities; eye–hand coordination skills; activity-based attention span; receptive and expressive skills (but, not necessarily verbal communication); independent personal care, particularly toilet indication; drinking and eating; sitting tolerance; and basic social skills such as eye contact, waiting for turn, following the authority, staying without primary caregivers, and ability to engage in play. In addition to this, any significant medical history (e.g., seizures, attention-deficit/hyperactivity disorder [ADHD]) which needs supervision of medication in the classroom should also be counted. A special note should be made if any aids and appliances are required to enhance the functional abilities of the child (e.g., reading glasses, hearing aids, wheelchairs, and specially adapted furniture). Accordingly, the assessment report must include appropriate recommendation for placement and intervention.

DISABILITY ASSESSMENT

According to the guidelines based on the Rights of Persons with Disabilities Act 2016 (Government of India, 2018, p. 94), disability assessment is done through three stages such as screening, diagnosis, and disability calculation [Table 4]. The minimum age for certification is one completed year. Children above 1 year and up to the age of 5 years shall be given a certificate with a diagnosis of GDD. Children above the age of 5 years shall be given a diagnosis and certificate as ID. The medical superintendent or chief medical officer or civil surgeon or any other equivalent authority as notified by the state government shall be the head of the medical board. The authority shall comprise the following: (a) the medical superintendent or chief medical officer or civil surgeon or any other equivalent authority as notified by the state government; (b) pediatrician or pediatric neurologist (where available)/psychiatrist or physician (if age >18 years); (c) clinical or rehabilitation psychologist; and (d) psychiatrist. It is preferable that clinicians time to time refer to relevant source to be updated with the guidelines. Temporary certificate can be issued for children <5 years, which will be valid for a maximum of 3 or 5 years of age, whichever is earlier. For children aged >5 years, the certificate will mention when to renew. As per the act, the certificate will have to be renewed at the age of 5, 10, and 18 years. The certificate issued at 18 years of age will be valid lifelong.

FORMULATING A TREATMENT PLAN

The treatment plan needs to address the issues related to the following five dimensions as indicated in a given case (Note: The multiaxial system of comprehensive diagnosis of IDD is taken from NIMHANS evaluation pro forma for IDDs):

1. Level of intellectual functioning (i.e., severity of ID)
2. Etiology/syndrome
3. Associated medical problems
4. Associated psychiatric problems
5. Family and psychosocial factors (e.g., awareness, attitude-overprotective, negligent, hostile, favorable; expectations; consistency of parenting; quality of stimulation; stressors in the family, family discard; caregivers' burnout).

Each of these five dimensions will have implications for biological, psychological, and social intervention. Consider, for example, a person with mild ID and ASD and seizures, with limited access to services and health-care facilities

Table 4: Disability certification process as per the guidelines based on Rights of Persons with Disabilities Act

Screening	Diagnosis	Disability calculation
Many of the children with ID/GD are on follow-up with pediatricians as developmental delay. Hence, they can be assessed by pediatricians and screened for associated comorbidities, namely, hearing/vision/locomotor impairments/ epilepsy. Then, these children are referred for detailed assessment	The screened children will be referred to child/ clinical psychologists for adaptive functioning and IQ testing. The tools that can be used for the same include VSMS for the adaptive functioning and BKT/MISIC for IQ testing. Based on these, the diagnosis of ID will be confirmed. Based on adaptive functioning assessment, severity scoring will be done and disability for ID will be charted	The disability calculation will be done based on VSMS score. The following will be used for disability calculation VSMS score 0-20: Profound disability - 100% VSMS score 21-35: Severe disability - 90% VSMS score 36-54: Moderate disability - 75% VSMS score 55-69: Mild disability - 50% VSMS score 70-84: borderline disability - 25%

VSMS – Vineland Social Maturity Scale; BKT – Binet–Kamat Test; ID – Intellectual disability; MISIC – Malin's Intelligence Scale for Indian Children; IQ – Intelligence quotient

in his/her community will have significant impairment in functioning as compared to a person with ID alone. In this example, the former will need appropriate medical, behavioral, and psychosocial interventions to address all these issues.

Setting for intervention is an important factor. Unless there is an indication for careful monitoring of medication on daily basis or poor therapeutic outcome if the patient is anywhere other than in the institutional setting, individuals with ID must be offered services in the community or on day-care basis. The idea is that the persons with ID should be in a least intrusive environment so that they can have maximum opportunities for learning and development in natural environment.

Medical interventions

Every attempt should be made to identify treatable causes of ID or at least potentially treatable symptoms such as hearing impairment and spasticity. Some of the conditions which present with ID are nearly completely preventable or to some extent reversible with appropriate management, provided that it is treated early in the course. Examples of treatable disorders are listed in Table 5, and such cases have to be referred to specialists accordingly for further management.

It is also important to treat associated medical problems along with therapies aimed at altering the pathophysiology among children with ID. Specialists need to be consulted for appropriate management to obtain maximal benefits. Few examples are treatment of epilepsy with antiepileptic drugs, spasticity with antispasticity medications, hearing impairment with hearing aids and cochlear implantation, sleep problems with sedatives as well as sleep hygiene techniques, and so on.

Genetic counseling

Genetic counseling is often deemed as a specialty in the current medical practice though it can be practiced by all clinicians to varying degrees depending on their expertise. As two-thirds of cases of ID have genetic etiologies, genetic counseling becomes mandatory. The most common situation is when genetic counseling is required in ID is when parents have one child with ID and would like to know the risk of recurrence and possibility of prenatal diagnosis. Genetic counseling not only provides accurate information on the prognosis of disorders and recurrence risks, but also helps in removing guilt and allaying ongoing recrimination in families.

Management of comorbid behavioral and psychiatric disorders

Nearly 20%–80% of the ID population can have problematic behaviors ranging from hyperactivity, temper tantrums, odd behaviors, to aggression. Behavioral problems are potential reasons for stigma, segregation, and caregiver's burden. Lack of occupation and limited developmental opportunities and communication deficits are major factors of problematic behaviors. While problematic behaviors can be a source or trigger for psychiatric problem and/or part of psychiatric disorder, they can also exist independently. In either case, a thorough behavioral plan is required. Identification of the problematic behaviors is the first step in management. Behaviors that lead to social exclusion, stigma, and those that interfere with learning should be given priority. Based on the hierarchy, target behaviors can be selected and functional analysis can be conducted to understand the antecedents and maintaining factors. Basic premise of the behavioral management is that opportunities are created to facilitate positive behaviors that would otherwise serve the same function as the problematic behaviors do (e.g., reinforcing any form of socially appropriate communication as a substitute for temper tantrums secondary to verbal communication deficits). In principle, the techniques should be least intrusive and culturally appropriate; therefore, the behavioral management plan can be implemented through the following three levels:

i. Restructuring the environment to control the antecedents and provide ample opportunities for positive learning
ii. Differential reinforcement to strengthen the adaptive behaviors by providing opportunities for reinforcement of adaptive behaviors
iii. Controlling inappropriate reinforcement of problematic behaviors.

It is also important to recognize that all problematic behaviors are not due to environmentally mediated, inappropriate reinforcement practices. Problematic

Table 5: Summary of medical interventions

Therapeutic modality	Examples of disorders
Replacement of deficient molecules	Thyroxine supplementation for hypothyroidism
	Enzyme replacement therapy for MPS
	Copper histidine for Menkes disease
Small molecule therapy	Usually provided at high doses (beyond daily recommended doses). Tetrahydrobiopterin along with low phenylalanine diet for PKU
	Creatine monohydrate for CCDS
	Pyridoxine, Vitamin B12, and folate for homocystinuria
Bone marrow transplantation	For alpha-mannosidosis and MPS 1
Pharmacotherapy	Vigabatrin for succinic semialdehyde dehydrogenase deficiency and tuberous sclerosis
Special/modified diet	For many organic acidurias and aminoacidopathies such as PKU, glutaric aciduria type 1, and MSUD.
Chelation of excess metals	Wilson disease and manganese transporter deficiency

MPS – Mucopolysaccharidoses; MSUD – Maple syrup urine disease; CCDS – Cerebral creatine deficiency syndromes; PKU – Phenylketonuria

behaviors may be an atypical presentation of psychiatric comorbidity or an indicator of the onset of a psychiatric episode. In some cases, problematic behaviors may be a manifestation of ineffective coping strategies to manage the psychiatric distress.

Psychiatric comorbidity not only presents itself more diffusely and atypically in these children, but also it is often difficult to treat. Carefully studying behavioral profile may point to a particular psychiatric disorder. Management may need a multipronged approach usually involving pharmacological and psychosocial interventions. If there is inadequate information to establish a psychiatric diagnosis, psychosocial interventions should be attempted first.

As only a handful of medications have been licensed for use in children, often, it is difficult to manage these disorders. This has to be discussed with parents in detail and their expectations should be handled regarding the outcome of such a treatment. Very few large systematic controlled trials are available in ID group; however, open drug trials, case reports, and expert reviews suggest the following:

- Begin with low dosage and increase it slowly
- Adequate trial time should be allowed before deeming failure of a medication
- Outcome to be monitored at multiple settings (home, school)
- Rationalize medications when multiple medications are being used and change one drug at a time
- Pediatric dosing schedules and guideline should be followed.

There are few studies on medications in comorbid disorders in ID, namely, methylphenidate in ADHD, or antipsychotics for schizophrenia. Risperidone also is widely studied as symptomatic treatment for problematic behaviors such as stereotypes and aggression [Table 6]. For further details in dosing and indications, the latest edition of the Maudsley Prescribing Guidelines are a good source. Specific details on pharmacological management could also found in condition-specific clinical practice guidelines of the Indian Psychiatric Society.

Nonpharmacological management

Child-centric interventions

Nonpharmacological interventions should be guided by life span and functional approaches. Accordingly, the following general framework can be adapted in regular clinical practice:

- Life span approach: Life span approach is that it regards the developmental needs and the tasks that the individual must achieve at each developmental stage to adapt to the environment. Accordingly, the skill training focuses on all important domains of adaptive behaviors such as conceptual, social, and practical skills that are considered important at a given developmental stage. In the initial 3 years, the focus should be on acquiring sensory-motor skills, socio-communication skills, basic self-help skills, and concepts. During 3–6 years of age, the focus can be on school readiness skills and mastery of culturally appropriate adaptive behaviors. During 6–18 years of age, the focus should be on the consolidation of academic and independent personal skills that can lead to future vocational training, employment, and adult independent living
- Functional approach: It is preferable that the tasks taught to the individual enable him or her to function well in day-to-day tasks. For example, there is no functional utility of mastering the spelling of five exotic animals as compared to mastering the sight words essential for daily functioning and community use (e.g., danger, exit, stop, price, and own name). Irrespective of the age and sociocultural context, each individual first needs training in self-care (toilet control, bathing, eating, dressing, and grooming), motor skills (especially, eye–hand coordination skills), receptive and expressive language abilities, social skills, and concepts in one set. Later, the children can be recommended for academics or functional academics, finally leading to vocational training, gainful occupation, and independent living skills. Throughout the program, health and safety skills should be strengthened.
- Making provisions for additional disabilities: Depending on additional disabilities, the child may need aids and appliances and appropriate therapeutic interventions.

Table 6: Summary of pharmacological treatment options

Symptom/disorder	Medication found to be effective in children with ID	Dose[#]	Caution/side effects
Hyperactivity, ADHD	Methylphenidate (IR) Clonidine Risperidone (especially in the presence of aggression and irritability)	Start with 5-10 mg, increments of 5-10 mg/week, maximum up to 2.1 mg/kg 0.1 to 0.5 mg/kg in 2-3 divided doses 0.5-2 mg	Tics, insomnia, anorexia (height and weight monitoring) Excess somnolence, hypotension (monitoring of BP required) extrapyramidal symptoms, and somnolence
Aggression, self-injurious behaviors, and irritability	Risperidone Clonidine	In general, dose in pediatric population is 0.5-2 mg Start with 0.25 mg/day for children <20 kg weight and 0.5 mg/day for children >20 kg weight* 0.1-0.5 mg/kg in 2-3 divided doses	Postural hypotension and excess somnolence
Stereotypy and RRBI	Risperidone SSRIs especially fluoxetine for other RRBI (Cochrane review 2013 showed no evidence of effectiveness and emerging evidence of harm)	Dosage as above Start with 2.5 mg/day up to 10 mg/day May be lower than usual doses used to treat depression in neurotypical children	As above Agitation, insomnia, anorexia, suicidal ideation
Depression, obsessionality, and anxiety	SSRI	Fluoxetine 5-10 mg/day is the starting dose Sertraline 25-50 mg daily. Effective dose is 50-100 mg	Higher risk for hypomania in ID children
Sleep disturbance	Melatonin If insomnia is associated with hyperarousal, then clonidine or clonazepam	1-10 mg doses have been tried; usual starting dose in children is a 2 mg single late evening dose Wide range of benzodiazepines such as clonazepam (0.25-0.5 mg) and lorazepam (0.5-1 mg) have been tried. Best titrated based on symptoms starting from lowest dose. However, less preferred due to paradoxical reactions	Epilepsy (no conclusive evidence) Paradoxical heightened agitation, impulsivity, and disinhibition Excess somnolence

[#]Doses are as used in non-ID children, but there is uncertainty regarding optimal dose in ID population. *Doses are as also used in autism, but the literature is limited regarding dosage in ID. RRBI – Restricted repetitive behaviors and interests; IR – Immediate release; BP – Blood pressure; ID – Intellectual disability; ADHD – Attention-deficit/hyperactivity disorder

For example, adapted furniture in cerebral palsy and hearing aids for hearing impairment

- Special focus on early intervention: Early identification and intervention with children at risk for GDD or ID should be a top priority. It is also important to recognize that early intervention can start from prenatal period in terms of identifying high-risk pregnancies, providing appropriate health care, and dealing with psychosocial adversities. Nonetheless, the postnatal early intervention plan should include accurate diagnosis of ID and comorbid conditions; identification of underlying etiological processes and methods of treatment as applicable; and activities to facilitate sensory-motor integration, speech and language development, and socioemotional development. The basis of early intervention is healthy bonding and attachment between mother and the child. Therefore, any stable caregiver can also be involved in early stimulation. In principle, early intervention programs should aim at stabilizing the current developmental milestones and create opportunities for the development of future tasks. Play-based methods and culturally rooted good practices of early child care should be strengthened. Materials recommended for intervention should be easily available and culturally appropriate; otherwise, parents will be overwhelmed if they are not easily available. For more formal intervention, referrals can be to the District Early Intervention Centers of the *Rashtriya Bal Swasthya Karyakram* (RBSK) and the *Anganwadi Centers* of the Integrated Child Development Services (ICDS)
- Referral and linkage: Appropriate services can be obtained from programs under the Sarva Shiksha Abhiyan, National Institute of Open Schooling, District Disability Rehabilitation Centers, Composite Rehabilitation Centers, national institutes, and local nongovernment agencies (more details of the government schemes can be found at www.socialjustice.nic.in). Wherever possible, it is better to refer the individual to the agencies in their own locality to cut down the costs of rehabilitation. Therefore, a registry of local, regional, and national agencies working in the area of developmental disabilities can be maintained for this purpose.

Family-centered interventions

- Parents and families should be given proper information regarding the nature, needs, and management of ID and its comorbidities in simple language devoid of any technical terms. Need may be appropriate literature, and specific web-based sources can be recommended for further reading. Siblings and other key family members can also be involved in the program plan.
- Parents and families should be supported in finding right resources for health care, therapy, education, and vocational and occupational needs.
- Ensure that parents and families are aware of the social provisions and importance of disability certificate for the child to avail the same.

- Emphasize on self-advocacy by creating awareness about various policies and provisions related to ID. If the person with ID is an adult, information regarding the guardianship and National Trust Act must be compulsorily provided
- Making meaning of the condition and developing a sense of control are crucial for optimum functioning of families. Various methods such as individual counseling, group counseling, parent training programs, and self-help groups can be used to achieve this.
- Each family is unique therefore the family support programmes must be individualized to meet the needs of the family in the context of care-giving and disability impact.
- Parents and primary caregivers must be routinely screened for stress-related disorders because there is ample evidence to suggest that syndromal depression and anxiety are high among parents of children with ID.

In summary, ID is a developmental disorder that affects general intellectual functioning and adaptive behaviors. It has no definite cause, but has multiple risk factors including genetic, biological, and environmental factors. Depending on the severity of the condition and the underlying etiological processes, ID can also present with comorbid conditions. It is important to identify the treatable conditions and treat the same. Special attention should be paid to psychiatric and behavioral disorders, which are common in ID and cause stigma, caregiver burden, and need for medication and segregation. Since ID causes disability, appropriate measures should be taken to certify disability and guide the families for appropriate support systems including the social benefits.

Nonpharmacological intervention should focus on skill development leading to educational and vocational competencies so that the individual will acquire necessary capacities for future adult independent living. Depending on the stage of development, referral can be made to different service agencies starting from early intervention services under the RBSK and ICDS to educational provisions under the Sarva Shiksha Abhiyan to vocational training and guardianship under the National Trust Act. Parents and families should be involved along with the individual at all levels of decision-making in order to promote self-advocacy.

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Conflicts of interest

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Appendix 1: Key questions to aid assessment, intervention, and treatment plan

Decision area	Key questions to be answered
Diagnosis	What precipitated the consultation? Is there a delay in important areas of development such as motor; speech, language, and communication; personal care/self-help skills, cognition/learning, and emotion? If yes, is it a global delay (i.e., delay in more than one important area of development)? Or, specific delay (i.e., deficit in only one area, e.g., speech and communication deficits in case of hearing impairment)? Does the global delay suggest significant impairments in intellectual functioning as is reflected in the adaptive functioning that is considered appropriate for the age and sociocultural standards for personal independence and social responsibility? (Note: Asking key questions related to adaptive behavior that reflect practical, conceptual, and social skills is important; presenting questions based on the behavioral indicators given in DMS-5 [American Psychiatric Association, 2013; p. 35-36] will be useful in this regard) Are the deficits in intellectual functioning and adaptive behavior appeared during the developmental period (i.e., before the age of 18 years)? Special circumstances: Is there an evidence for significant impairments in intellectual functioning and adaptive behaviors but no reliable early developmental history as in case of children reported with sheltered homes, orphanages, and adopted or those under foster care; or who do not have valid birth records or the caregiver does not have adequate information?
Comorbidities/co-occurrences	Are there any identifiable comorbid conditions or co-occurrences? If yes, do they have specific implications for health care and other forms of interventions? Do the comorbid conditions increase the severity of ID because of additional disability? And, do they denote "multiple disabilities"? If yes, do they have specific implications for health care and other forms of interventions?
Etiology and risk factors	Are there any treatable etiological conditions of ID? Or, are there any risk factors associated with the present condition or have the potential to aggravate the disability in future? Does it need further medical examination and laboratory investigations to confirm the screening results? If yes, specify them (for specific details, see the subsection on medical comorbidities for possible etiological conditions, risk factors, and essential laboratory investigations)
Nature and needs of the condition	Given the developmental stage and the sociocultural background of the individual with ID, what are the immediate needs of the child and family and other service providers? What would be the impact on the child, family, and the immediate environment if the needs are not met? How does the current needs impact the future independent living of the individual with ID? Does the person with ID require any special assistance and adaptations to meet the identified needs? What are the resources available at various levels (e.g., family, neighborhood, and community) to meet the identified needs?
Intervention plan	Is there a need for medication to treat associated medical conditions including psychiatric comorbidities? If yes, identify the condition and medical intervention Does the child require referrals for any therapies (e.g., speech, audiological, physiotherapy, and occupational therapy, educational [supported/special/integrative]) to restore or enhance the functional abilities Does the child need specific behavioral management plan for managing challenging behaviors? If the evidence-based intervention plan including all or any of the above strategies is implemented, in what the quality of life of the person with ID will be better? Is the intervention plan cost-effective? Are there any significant side effects or offshoot troubles of the intervention? Will the intervention plan facilitate inclusion of the person with ID in the mainstream? Knowledge, attitudes, and perceptions of the caregivers with regard to the condition? What are the needs of the caregivers? Will the intervention plan help reduce the caregiver's burden?
Placement decisions	Which is the best setting to deliver the targeted interventions (e.g., home, hospitals, vocational/rehabilitation centers; day-care or residential schools, *Anganwadi* centers, and District Early Intervention Centers)? If medical and therapeutic interventions are required, which is the best setting to obtain the maximum positive outcome for the individual with ID - outpatient or inpatient? Who are the people or agencies through which the intervention can be delivered? Is the placement least intrusive that the persons with ID will continue to have normal developmental and learning opportunities and appropriate sociocultural experiences? Are all options for community integration are exhausted before considering segregation from the mainstream for any reason including safety, dignity, optimizing the potential, quality of life, and well-being?
Evaluating the outcome of intervention	Are there any indicators other than the direct measures to suggest that the intervention is effective? Is the positive therapeutic outcome observed in one setting is maintained or generalized to other settings?

ID – Intellectual disability

Appendix 2: Scales of intellectual functioning and adaptive behavior adapted or normed for Indian population

Test	Indian Adaptation	Age	Content	Merits	Challenges
Seguin Form Board	Bharatraj (1971) Goel and Sen (1984); Revalidated by Venkatesan (1998)*	Reliable for 3 to 11 years old, but valid for all age groups of people with ID	Performance test	It serves as a quick measure of general intelligence	It is not much valid for children aged above 11 years of age, when it becomes more a measure of visuo-motor speed than global intelligence
Binet-Kamat Test of intelligence	Kamat (1967) adapted 1916 revision of Binet-Simon Test; Reappraised by Venkatesan (2002a)*	3 years - adulthood	Age scale; Predominantly verbal	Balances verbal and performance items	Specific test items depend on formal education. Verbal items not available for vernacular languages other than Kannada and Marathi. some items are completely redundant
Stanford-Binet Intelligence Scale	Kulshreshta (1971)	3 years - adulthood	Age scale; Predominantly verbal	Balances verbal and performance items and also offers a short scale. More suitable for Hindi-speaking population	Did not include people with low intelligence in the sample
Malin's Intelligence Scale for Indian Children	Malin (1973) adapted the original scale of Wechsler's Intelligence Scale for Children	6-16 years	Has verbal and performance tests	It measures both verbal and performance intelligence	Some of the verbal scales depend on formal education
Developmental Screening Test	Bharat Raj (1977)	0-15	Developmental tasks	It assesses global development	DST is highly loaded with speech and language items; hence, it must be interpreted cautiously in case of conditions such as cerebral palsy, autism, and speech and hearing impairment
Vineland Social Maturity Scale	Malin (1968); expanded by Bharatraj (1992)	0-15	Culturally appropriate adaptive behavioral skills	It gives a comprehensive profile of adaptive behavior	It may need revision in tune with the changing concepts of adaptive behavior
Progressive matrices a. Standard	Raven (2003); Indian norms are available (Deshpande *et al*., 2002)	11 years to adults	Nonverbal	It assesses the general intellectual abilities trough form comparisons and analytical reasoning. This test is culture-fair to a large extent	Not suitable for illiterates and persons at the lower end of ID spectrum. It does not yield IQ scores.
b. Colored	Raven (2003);	5-11 years		It gives percentiles	Same as above
Gessel's Drawing Test	Verma *et al*. (1972); Revalidated by Venkatesan (2002b)*	15 months to 8 years	Performance test	It is a reliable screening test of mental developmental	It is not a valid test for the children who have not attended school or have no experience with a pencil or children with specific finger dexterity problem
Bhatais's Battery of Performance Test of Intelligence	Bhatia (1955)	11 years and above	Performance test	Many subscales are indigenous	It measures IQ above 70; hence, not suitable for use with suspected cases of ID
Wechsler Intelligence Scale for Children - fourth edition (India)	Wechsler (2003)	6-16 years and 11 months	Contains both verbal and performance scales	It has updated areas of assessment in accordance with the development of children in India	Time consuming and costly

The table is adapted from Arya, S., Kishore, M.T., Ranga, S., Bisht, J. Current Status of Intelligence testing in India: Perspectives on disabilities. NIMH News Letter, 2005; 18 (2 and 3), 19-23. ©NIEPID (formerly, NIMH), Secunderabad. *Revalidation/reappraisal details could be found in Madhavaram, T.K. Intelligence testing and its implications for disability evaluation in individuals with mental retardation. Psychol Stud 2011;56(3):289-294. DOI 10.1007/s12646-011-0093-y.
DST – Developmental Screening Test; IQ – Intelligence quotient; ID – Intellectual disability

Appendix 3: Tests indicated in case of intellectual disability and comorbid conditions

Type of disability	Screening	Adaptive behavior	Global Intelligence Scale (in the order as below)
ID alone	DST	VSMS	BKT
ID and VI	DST	VSMS	Prorate the IQ based on MISIC verbal scales BKT
ID and HI	GDT and SFB	VSMS	Prorate the IQ based on MISIC performance scales BKT is not suitable because of high loading of verbal and language items
ID and CP (or, locomotor disability)	DST and GDT	VSMS	BKT; Profile analysis will help identify specific effect of motor deficits on test performance Prorate the IQ based on MISIC verbal scales

Source: Kishore MT. Trends in intelligence testing of persons with mental retardation and its implication for certification of disability and service provisions. An unpublished study funded by the Indian Council of Social Sciences Research, 2011. DST – Developmental Screening Test; VSMS – Vineland Social Maturity Scale; GDT – Gessel's Drawing Test; SFB – Seguin Form Board; BKT – Binet–Kamat Test of Intelligence; MISIC – Malin's Intelligence Scales for Indian Children; ID – Intellectual disability; VI – Visual impairment; HI – Hearing impairment; CP – Cerebral palsy