Child Disabilities Protocol of EHA

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Egyptian Clinical Practice Protocol

in

Child Disability

for

Egypt Healthcare Authority

First Edition

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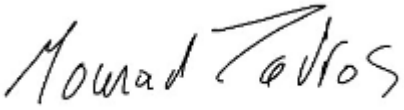

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PREFACE

Recently, there is an increasing need to provide programs with accurate competency-based assessments to ensure the delivery of high-quality healthcare. The aim of developing these Egyptian Clinical Practice Protocols in Child disability is to unify and standardize the delivery of healthcare to all newborns at all health facilities.

The current state of healthcare in which avoidable failures are abound. “We train longer, specialize more, use ever-advancing technologies, and still we fail.” Part of the problem, is that the ever-increasing complexity of medicine makes uniform care delivery impractical or impossible. That is, unless there are protocols, checklists, or care paths that are readily available to providers.

Standard textbooks, journals, and online resources currently available create excellent repositories of detailed information about the etiology, pathogenesis, clinical picture, diagnosis, and treatment of a condition. However, for a busy clinician looking for the best way to manage a sick patient, a standardized path for effective management of the patient may be impossible to discern. So, it would be a lot easier if we all managed simple things in a uniform way using the best available evidence and resources.

In child disability, busy clinicians have all felt the need for a concise, easy-to-use resource at the bedside for evidence-based protocols, or consensus-driven care paths where high-grade evidence is not available.

In this protocol, we offer comprehensive reviews of selected topics and comprehensive advice about management approaches and procedures based on the highest level of evidence available in each case. Our goal is to provide an authoritative practical medical resource for neonatologists, pediatricians, and other healthcare providers dealing with newborns after birth. This protocol is the product of contributions from numerous neonatologists from all over Egypt.

Members of the Working Group

For Development of the Egyptian Clinical Practice Protocol

In Child Disability

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Intellectual Developmental Disorder (IDD)

Definition:

- Disability is extremely diverse. While some health conditions associated with disability result in poor health and extensive health care needs, others do not. However, all people with disability have the same general health care needs as everyone else, and therefore need access to mainstream health care services.

- Levels of understanding and visibility have increased. And there have been profound changes in public attitudes towards disability, captured and catalyzed by national moments such as the Paralympic Games.

- **Intellectual Disability (ID)** (also known as **Intellectual Developmental Disorder (IDD),** and previously Mental Retardation) is a disorder with onset during the developmental period that includes both intellectual and adaptive functioning deficits in 3 domains:
    1. **Conceptual**
    2. **Social**
    3. **Practical domains**

- Intellectual disability is a heterogeneous condition with many different etiologies. Two other diagnoses exist under the intellectual disability diagnostic category in the DSM-5: Global Developmental Delay (GDD) and Unspecified Intellectual Disability (UID).

Diagnostic Criteria:

- Intellectual disability is a disorder with onset during the developmental period that includes both intellectual and adaptive functioning deficits in conceptual, social, and practical domains. The following 3 criteria must be met:

Criterion A

- Deficits in intellectual functions, such as reasoning, problem solving, planning, abstract thinking, judgment, academic learning, and learning from experience, confirmed by both clinical assessment and individualized standardized intelligence testing.

Criterion B

- Deficits in adaptive functioning that result in failure to meet developmental and sociocultural
- Standards for personal independence and social responsibility. Without ongoing support, the adaptive deficits limit functioning in one or more activities of daily life, such as communication, social participation, and independent living, across multiple environments, such as home, school, work, and community.

Criterion C

- Onset of intellectual and adaptive deficits during the developmental period.

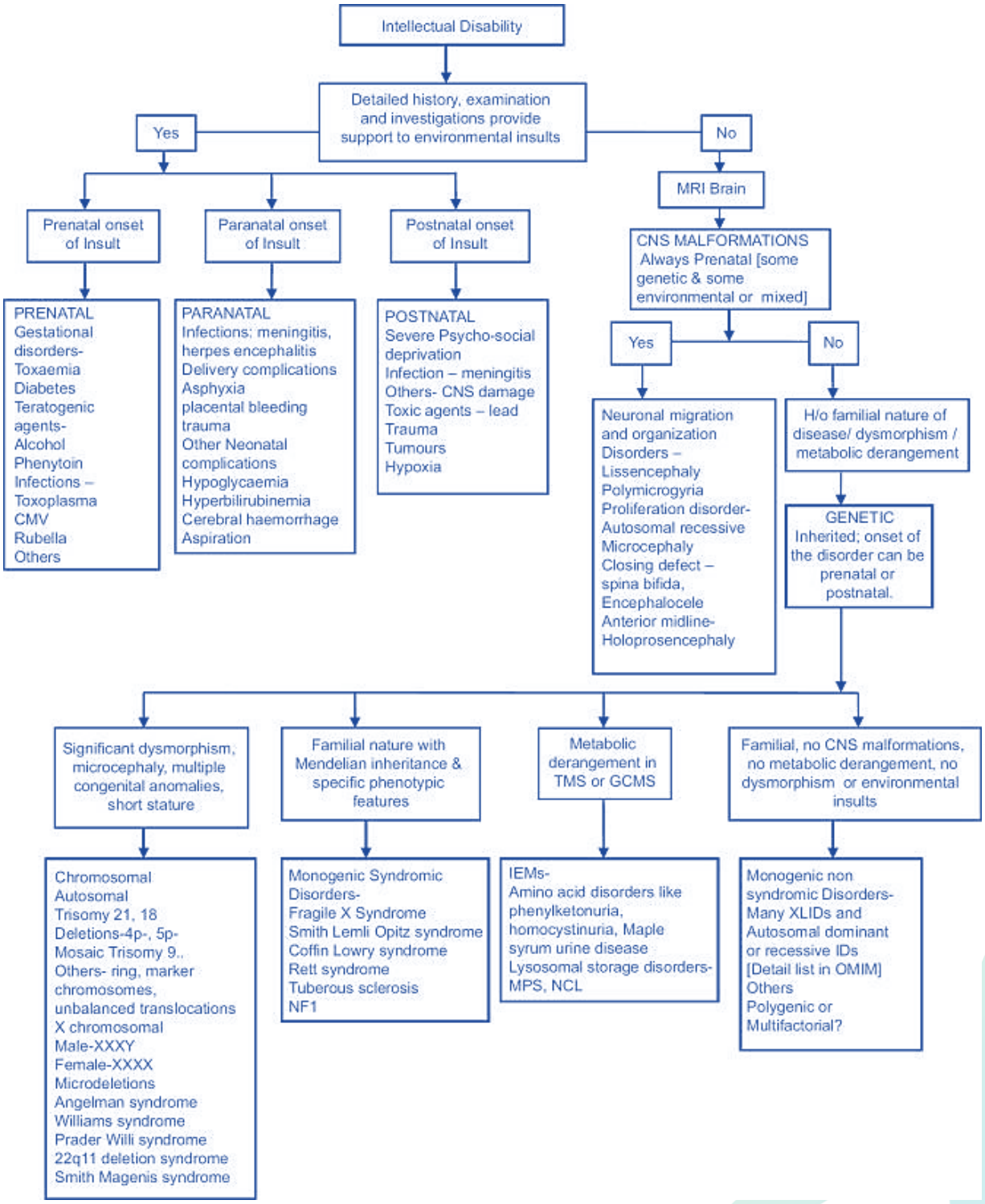
Pathophysiology:

- 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 common causes of ID are also listed in the flowchart [Figure 1].

Figure 1: Finnish Approach

Diagnosing Intellectual Disability 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.



I. History Taking:

- The purpose of eliciting the history is to establish that there is 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. A useful and comprehensive approach to assessment would include:

1. Noting **chief complaints** in chronological order with mode of onset, duration, and precipitating event followed by
2. **History of presenting illness** and a detailed prenatal and perinatal history.
3. **Developmental history** in greater detail, particularly related to motor, language, and communication; self-help skills; socioemotional skills; cognition; and occupational skills/leisure-time activities;
4. **Medical comorbidities** and its treatments;
5. **Psychiatric history** including the details of onset, evolution, and current status of behavioral and other psychopathological disturbances; and treatment history.
6. 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.

II. Physical Examination:

- Detailed physical examination helps to identify the etiology in most cases, detect comorbid medical conditions, and also order appropriate investigations. Physical examination in cases with ID consists of three parts which are as follows: It must involve:
  1. Routine systemic examination,
  2. Anthropometric assessment, and
  3. Observation of atypical morphological features suggestive of specific genetic disorders.
- **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 1). It requires keen observation and knowledge of normal versus abnormal morphology.

Table 1: 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	Deeply set, promment eyes, microphthalmia, upslanting/downslanting palpebral fissures, hypertelorism, epicanthal folds, strabismus,
structures	ptosis, bushy eyebrows, synophrys, microcomea, comeal 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	Chinodactyly, brachydactyly, syndactyly, camptodacyly, arachnodactyly, polydactyly, broad thumb, etc.
Chest	Pectus excavatum, pectus carinatum, nipple anomalies, gynecomastia
Abdomen	Protuberant, scaphoid, umbilical hernia, hepato-splenomegaly, ingamal 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 plans, pes cavus, valgus/varus anomaly, broad hallux, increased distance between the 1" and 2 toes
Skeletal	Exostoses, increase carrying angle, joint hypermobility

- **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.

III. Behavioral Observation:

- The purpose of behavioral observation is to corroborate the clinical history regarding intellectual functioning and behavioral repertoire. Therefore, it should start with:

1. Observation of general appearance,
2. Any oddities in behavior,
3. Attention span,
4. Receptive and expressive speech abilities, and
5. Social and interpersonal abilities.

- Any changes in behavior compared to previous period, should be carefully recorded in each visit. If it is pervasive and indicative of a comorbid psychiatric disorder, it has to be carefully considered. 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 will help in uncovering the psychopathology.
- 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.
- 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:

1. **Basics:** Behaviors suggesting sensory-motor impairments or physical health issues
2. **Response to interview situation:** Excited, fearful, tense, shy, inhibited, guarded, uncooperative, or defiant
3. **Alertness:** Over-aroused, withdrawn
4. **Attachment to parents and response to separation:** Clinging, wanting to be carried all the time, indifferent to separation
5. **Sociability:** Social orientation, approachability, social responsiveness, eye-to-eye contact, reciprocal interactions, and awareness of social boundaries
6. **Motor activity level:** Fidgetiness, restlessness, hyperactivity, lethargy
7. **Course of motor behaviors during interview or response to firm instructions:** Quiet initially, but restless later on; unresponsive to firm instructions
8. **Impulse control:** Snatching, spilling, falling, bumping, climbing, interfering, temper tantrums; aggressive acts such as biting, throwing, beating, pulling hair, slapping
9. **Attention and concentration:** Goal directedness, task completion, distractibility
10. **Speech, language, and communication:** Verbal/nonverbal comprehension and expression; vocabulary, articulation, and flow
11. **Mood:** Inhibited, excessively cheerful, whining and crying, irritable
12. **Play behavior:** Type of activity, duration, themes, etc .
13. **Other inappropriate behaviors:** Any excess behaviors that are inappropriate to the age and sociocultural context
14. **Impressions on current developmental attainment:** Whether excess behaviors or skill deficits are typical of a known psychiatric or developmental disorder?
15. **Parent–child interactions:** Quality of engagement with child; communication patterns; degree and quality of control over the child; response to good and bad behaviors.

Diagnosis of Comorbid Psychiatric Disorder:

- Some mental health, neurodevelopmental, medical and physical conditions frequently co-occur in individuals with intellectual disability, including Autism Spectrum Disorder, Cerebral Palsy, Epilepsy, Attention-Deficit Hyperactivity, impulse control disorder, and depression and anxiety disorders. Identifying and diagnosing co-occurring conditions can be challenging, for example recognizing depression in an individual with limited verbal ability an accurate diagnosis and treatment are important for a healthy and fulfilling life for any individual.

Differential Diagnosis:

- A diagnosis of intellectual disability should not be presumed simply because of a pre-existing genetic or medical condition. A differential diagnosis includes:

1.Major and Mild Neurocognitive Disorders:

- Intellectual disability is categorized as a neurodevelopmental disorder and is distinct from the neurocognitive disorders, which are characterized by a loss of cognitive functioning. Major Neurocognitive disorder may co-occur with intellectual disability (e.g. - an individual with Down syndrome who develops Alzheimer's disease, or an individual with intellectual disability who loses further cognitive capacity following a head injury). In such cases, the diagnoses of intellectual disability and neurocognitive disorder may both be given.

2.Communication Disorders and Specific Learning Disorder:

- These neurodevelopmental disorders are specific to the communication and learning domains and do not show deficits in intellectual and adaptive behaviour. They may co-occur with intellectual disability. Both diagnoses are made if full criteria are met for intellectual disability and a communication disorder or specific learning disorder.

3.Autism Spectrum Disorder:

- Intellectual disability is common among individuals with autism spectrum disorder. Assessment of intellectual ability may be complicated by social-communication and behaviour deficits inherent to autism spectrum disorder, which may interfere with understanding and complying with test procedures.

4.Genetic disorders: such as Fragile X Syndrome (FXS)

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.

Investigations:

- A comprehensive work up for intellectual disability includes assessing intellectual capacity and adaptive functioning, identifying genetic, non-genetic, and associated medical conditions (such as cerebral palsy and seizure disorders). A prenatal/perinatal history, family pedigree, physical examination, genetic evaluation (karyotype, chromosomal microarray, and/or genetic syndrome tests), metabolic screening, and neuroimaging assessment can also be important investigations.
- 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 2].

Table 2: 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

Psychosocial Assessments:

- Persons with ID will be at a high risk for neglect and abuse. Adaptive behavior is always impaired in people with ID, but the deficits are less evident in environments where support systems are in place. Therefore, psychosocial assessments are very important.

Assessment of Intellectual Functioning and Adaptive Behavior:

- Both ICD-10 and DSM-5 recognize the need for assessing the intellectual functioning with standardized tools that yield intelligence quotients (IQs). When IQ tests are not applicable because of young age (e.g., children below 3 years) or associated sensory-motor issues and gross under stimulation, standardized developmental scales (e.g., Developmental Screening Test and Bailey Scales for Infants) can be used as applicable.
- Regarding the assessment of adaptive behavior, Vineland Social Maturity Scale (VSMS) is the only standardized measure available at present. The VSMS yields social quotient (SQ) and a profile of eight important domains of adaptive behavior.

Treatment:

- Once a diagnosis is made, help for individuals with ID is focused on looking at the individual's strengths and needs, and the supports he or she needs to function. Hence formulating a treatment plan to address the following issues according to the severity of ID:

1.Etiology/syndrome
2.Associated medical problems
3.Associated psychiatric problems
4.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).

- Treatment needs **A Multidisciplinary Team I.e.** a collection of different professionals, who:

✓ Meet regularly
✓ Allocate time to the pursuit of team's objectives.
✓ Agree on explicit team's objectives.
✓ Have adequate administrative & clinical coordination to support the team's work.
✓ Respect skills & roles that are specific & unique to team individual members.

- **Case Manager is** any team member with:

✓ Appropriate interpersonal skills
✓ And broad background knowledge of the needs and requirements of the disabled person.

➢ **Role of the Case Manager:**

✓ The main source of information.
✓ Feedback
✓ Simple coordination within a single agency.
✓ Coordination across agency boundaries.

Medical Interventions:

- Medications, particularly antipsychotics, may be used to manage challenging behavior such as aggression in individuals with intellectual disability. When the behavior does not arise from an underlying mental illness, this is off-label use and evidence of efficacy is very poor. Furthermore, these patients face higher risks of drug-related side effects. 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.

- 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 3, and such cases have to be referred to specialists accordingly for further management.

Table 3: 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

Genetic Counseling:

- Genetic counseling not only provides accurate information on the prognosis of disorders and recurrence risks, but also helps in removing guilt and 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. 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.

- Psychiatric comorbidity not only presents itself more diffusely and atypically in these children, but also it is often difficult to treat. Management may need a multipronged approach usually involving pharmacological and psychosocial interventions.

- 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, the following is suggested:
  - 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 [Table4]. Prescribing Guidelines is a good source.

Table 4: Summary of Pharmacological Treatment Options

Symptom/disorder	Medication found to be effective in children with ID	Dose[a]	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[b] 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

[a]Doses are as used in non-ID children, but there is uncertainty regarding optimal dose in ID population. [b]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

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:** 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. 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.
- **Making provisions for additional disabilities:** the child may need aids and appliances and appropriate therapeutic interventions. For example, adapted furniture in cerebral palsy and hearing aids for hearing impairment
- **Special focus on early intervention:** Early identification, intervention is a top priority.
- **Referral and linkage:** Appropriate services can be obtained from programs.

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. 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.
- 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.
- 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. 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.

Working with Children with Neuro-Disability

- 0-18 years
- Wide range of developmental difficulties / disabilities
- Holistic approach
- Partnership with parents
- Multidisciplinary working
- Interagency working
- Communication

Settings:

- **Child Development Centres 0-5 years**
- **Mainstream School Health**
- **MLD Schools**
- **Complex Needs Schools**

Referrals:

- **Referrals from GP, nursery, educational psychology**
- **Referral centers for specialist developmental assessment**
- **Links with other agencies – education, social work**

Children with Additional Needs:

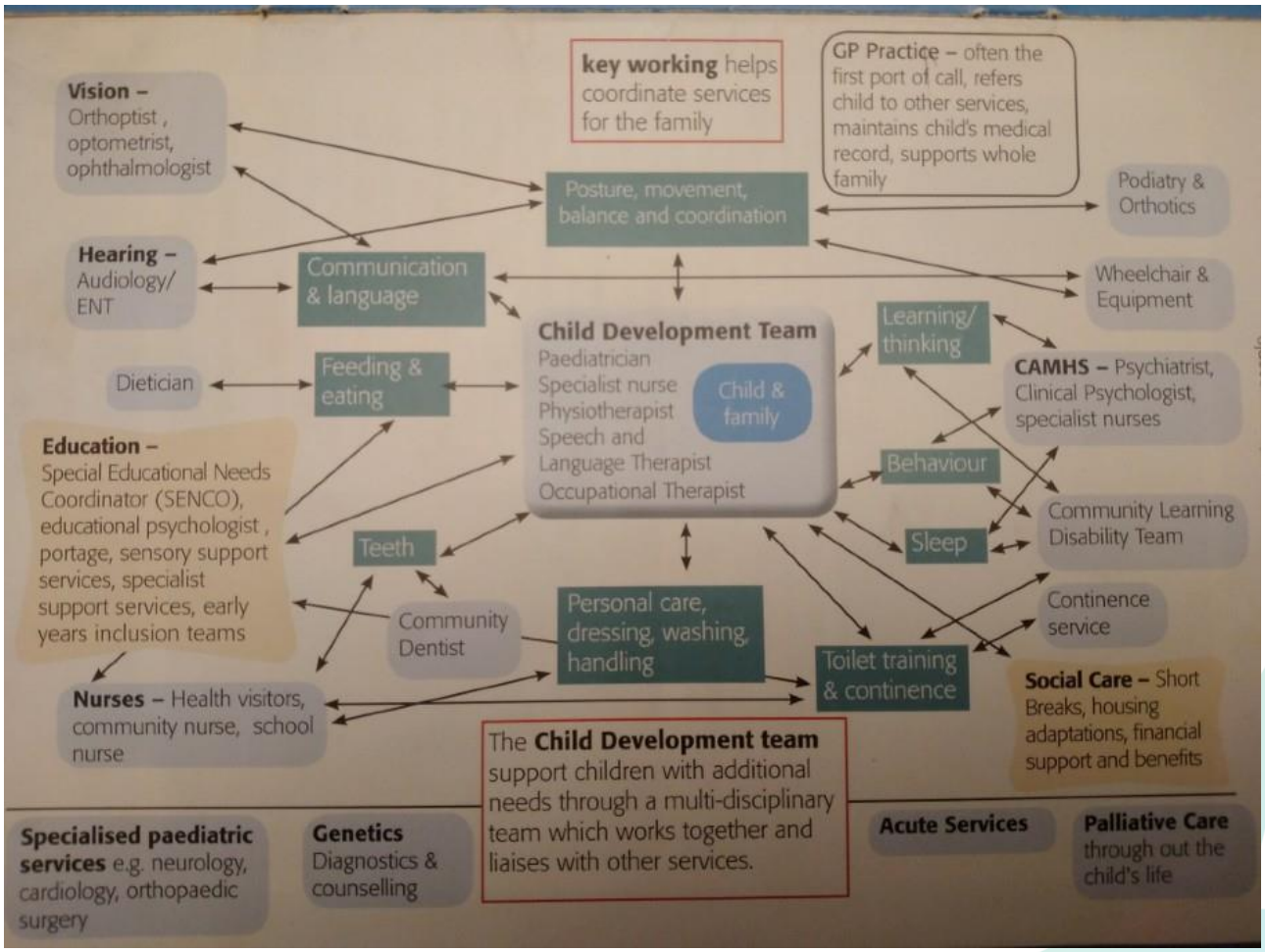
- **Ex – preterm**
- **Autism**
- **Down's syndrome**
- **Early developmental impairment ("global developmental delay")**

Child Development Centers

- It is composed mainly with a core team which is **the child development team**. It is mainly composed of:

  - Pediatrician,
  - Occupational therapist,
  - Physiotherapist,
  - Clinical psychologist,
  - Speech therapist,
  - Social worker,
  - And nurse

- Also, the team is joined or accessible to other services as mental health, audiology, dietitian, education.
- The following diagram summarizes the main multidisciplinary team and all other services that may be needed for the child with disability.



Autism Spectrum Disorder

- Autism is a brain disorder that limits a person's ability to communicate and relate to other people.
- It first appears in young children, who fall along a spectrum from mild to severe. Some people can navigate their world, some have exceptional abilities, while others struggle to speak.
- It is diagnosed according to DSM 5 or ICD 11
  - The 3 defining areas of impairment (social deficits; communication deficits; and restricted, repetitive behaviors and interest) were reduced to 2 domains by combining social and communication to "social/communication deficits" and retaining the behavioral impairment domain (RRB's).
  - Too difficult to separate social deficits from communication deficits combine into one unit.
  - Delays in language should be considered factors that influence symptoms rather than define the disorder.

Signs of Autism:

- Before a child turns three, careful observers can see signs of autism. Some children develop normally until 18-24 months old and then stop or lose skills. Signs of an ASD can include:
  - Repeated motions (rocking or spinning)
  - Avoiding eye contact or physical touch
  - Delays in learning to talk
  - Repeating words or phrases (echolalia)
  - Getting upset by minor changes

"It's important to note that these signs can occur in children without ASDs, too"

Early Warning Signs: First Year:

- Even young infants are very social, so it's possible to detect signs of autism in how babies interact with their world. At this age, a child with an ASD may:
  - ➢ Not turn to a mother's voice
  - ➢ Not respond to his own name
  - ➢ Not look people in the eye
  - ➢ Have no babbling or pointing by age one
  - ➢ Not smile or respond to social cues from others

Early Warning Signs: Year Two:

- The signs of autism are more noticeable in a child's second year. While other children are forming their first words and pointing to things they want, a child with autism remains detached. Signs of autism include:
  - ➢ No single words by 16 months
  - ➢ No pretend games by 18 months
  - ➢ No two-word phrases by age 2
  - ➢ Loss of language skills
  - ➢ No interest when adults point out objects, such as a plane flying

Other Signs and Symptoms:

- People with autism sometimes may have physical symptoms, including digestive problems such as constipation and sleep problems.

- Children may have poor coordination of the large muscles used for running and climbing, or the smaller muscles of the hand.

- About a third of people with autism also often have seizures.

Diagnosis of Autism (ASD)

- ASD is diagnosed according to DSM 5 or ICD 11 criteria.
- Diagnosing ASD can be difficult since there is no medical test, like a blood test, to diagnose the disorders.
- Doctors look at the child's behavior and development to make a diagnosis.
- ASD can sometimes be detected at 18 months or younger. By age 2, a diagnosis by an experienced professional can be considered very reliable.
- However, many children do not receive a final diagnosis until much older.
- This delay means that children with ASD might not get the early help they need.

Early Screening for ASD:

- Many children aren't diagnosed with an autism disorder until preschool or even kindergarten, and may miss getting the help they need in the early years. That's why guidelines call for screening all children at nine months old for delays in basic skills. Special ASD checkups are needed at:
  - 18 months
  - 24 months
- As needed for children with worrisome behaviors or a family history of autism

Screening and Diagnostic tools for ASD:

Screening tools for ASD

- The Checklist for Autism in Toddlers (CHAT) for children 18-24 months of age.
- The modified Checklist (M-CHAT) for children 16-30 month of age.
- The Social communication questionnaire (SCQ) for children 4 years and older.

Diagnostic tools for ASD

- Autism Diagnostic interview- Revised (ADI_R).
- Autism Diagnostic Observation Schedule (ADOS-G).
- Childhood Autism Rating Scale (CARS).
- Diagnostic Interview for Social and Communication Disorders (DISCD).

Diagnosis: Speech Problems:

- At regular checkups, the physician will check how the child responds to the mother's voice, smile, or other expressions. Is he cooing or babbling? Problems or delays in speech call for a visit to a speech therapist.
- A hearing test may be needed, too.
- Most children with autism will eventually speak, but they do so later than others.
- Making conversation may be especially tough.
- Children with ASDs also may speak in a sing-song or robotic way.

Musculoskeletal Disorders & Examination

Basic Categories:

1. Hyperkinesia or dyskinesia, characterized by excessive movement.
2. Hypokinesia, bradykinesia or akinesia, characterized by slowness or a lack of movement.

Ataxia:

- Ataxia describes a lack of coordination while performing voluntary movements. It may appear as clumsiness, inaccuracy or instability:
  - **A.** Movements are not smooth, and may appear disjointed or jerky.
  - **B.** Associated tremor due to over-correction of inaccurate movements.
  - **C.** Past-pointing when an attempted reach overshoots the target.
  - **D.** Poor performance of regular, repeated movements, such as handclapping.
  - **E.** When it affects mechanisms of walking, there will be instability with a tendency to fall.

Examination:

- The first feature to observe about ataxia is where it occurs in the child's body. It may affect only walking or arm and eye movements may be involved. It is important to recognize if:
  1. There are variations in the severity of the symptoms during the day.
  2. There are variations in the severity of symptoms at mealtime.
  3. Symptoms become worse when the child is tired, hungry or ill.
  4. Whether particular types of foods that affect the symptoms.

Treatment:

1. Physical therapy to train and strengthen muscles to compensate.
2. Gait and balance training are essential.
3. Cane, crutches or walker is often beneficial.
4. Ddapted utensils and other tools may be helpful.

Bradykinesia (slow movement):

1. Bradykinesia essentially refers to a component of parkinsonism.
2. A child with bradykinesia has slow and painstaking movements of the affected limbs.
3. If the whole body is affected, there may be an unnatural stillness or frozen quality.
4. In some cases, there are reduced movements of the face leading to an expressionless look referred as a “mask face”.
5. It may affect one limb, one side of the body, or the entire body.
6. Rigidity, refers to resistance to passive movement that may make the limb feels like a “lead pipe”.
7. Rigidity, also affects the response to gravity, leading the child maintaining his or her arm in a fixed posture while walking rather than swinging it loosely at the side.
8. Cog wheeling, when rigidity and tremor are present at the same time.

Chorea:

1. Irregular, rapid, uncontrolled, involuntary, excessive movement that seems to move randomly from one part of the body to another.
2. The affected child often appears restless and unable to sit still.
3. The jerky movements of the feet or hands are often similar to dancing or piano playing.
4. When chorea is severe, the movements may cause motion of the arms or legs that results in throwing whatever is in the hand or falling to the ground.
5. Walking may become bizarre, with inserted excessive postures and leg movements.
6. Unlike parkinsonism, the movements of chorea, athetosis and choreoathetosis occur by themselves, without conscious attempts at movement. In some cases, attempts to move may make the symptoms worse.
7. Athetosis is a slow twisting movement.
8. Choreoathetosis is a movement of intermediate speed, between the quick, flitting movements of chorea and the slow twisting movements of athetosis.
9. Ballism is a violent flinging of one or more limbs out from the body.
10. Choreoathetosis is the most common form in children.
11. These disorders may affect the hands, feet, trunk, neck and face. In the face, they often lead to nose wrinkling, continual flitting eye movements and mouth or tongue movements.
12. These disorders may be distinguished from tics, as tics tend to repeat the same set of movements. In addition, the child often describes a “build-up” in the need to make the tic, with a sense of release afterwards.

Dyskinesia:

Clinical Features and Classification:

1. Paroxysmal Kinesigenic Dyskinesia (PKD):

- The episodes of hyperkinetic movements are provoked by sudden voluntary movement or unexpected stimuli (startle).

2. Paroxysmal Non-Kinesigenic Dyskinesia (PNKD):

- The attacks may occur spontaneously while at rest or out of a background of normal motor activity, but may be exacerbated by alcohol or caffeine consumption, stress, fatigue or other factors.

3. Paroxysmal Exertion-induced Dyskinesia (PED):

- A relatively rare form of paroxysmal dyskinesia has been described in which episodes are induced by prolonged exertion.

4. Paroxysmal Hypnogenic Dyskinesia (PHD):

- A rare disease variant, characterized by transient attacks of involuntary movements occurring during non-REM (NREM or non-rapid eye movement) Sleep.
- In rare instances, episodes may be preceded by potentially painful sensations. Paroxysmal dyskinesias may also be further categorized according to the duration of the attacks. They may be described as “short-lasting” if episodes are less than or equal to 5 minutes or “long-lasting” if attacks are longer than 5 minutes.

Dystonia:

1. A syndrome of sustained muscle contractions, frequently causing twisting and repetitive movements, or abnormal postures”.
2. The muscle contractions related to dystonia may be quite rapid and not sustained.
3. The movements may not be repetitive and not lead to fixed postures.
4. Usually occurs only during voluntary movement or with voluntary maintenance of a posture of the limbs or body.
5. Characteristic postures that are frequently seen in dystonia include:
   a. Spooning, during which the fingers of the hand are bent backward with the wrist flexed.
   b. Elbow and wrist flexion with the hand held near the body.
   c. Foot in-turning or inversion at the ankle, which is frequently pronounced with walking.
   d. Extension of the great toe.
   e. Turning of the neck or torticollis.

6. Focal dystonia: (only one body part is involved, such as a hand, foot or the neck).
7. Segmental dystonia: (two contiguous parts are involved, such as the face and neck).
8. Multifocal dystonia: (two noncontiguous parts of the body are involved, such as the face and one leg).
9. Hemidystonia: (one half of the body is involved).
10. Generalized dystonia: (both legs, as well as one additional body part are involved).
11. A dystonic posture of the right hand may occur while the left hand is performing a rapid movement, or a dystonic posture of the foot may occur during walking. The triggering movements may be very specific; for example, walking forward may be a trigger, while walking backward may not be a trigger.

Examination:

1. The child being evaluated for dystonia must be observed at rest, with action of the parts of the body affected by dystonia, as well as actions unrelated to the dystonia.
2. A child with foot dystonia must be observed while sitting, standing, walking and performing tasks with the hands.
3. Mental distraction is also helpful; when possible, the evaluator may use language or mathematical tasks to distract the child. These types of distractions may help to determine the specific triggers for the dystonic movements.
4. They may also assist in evaluating if other body parts are subtly affected when provoked by attempted movement, stress or distraction.
5. It is important to test the child during the certain activities to observe dystonia-obstructing movement or excessive movements. These activities include:
   a. Reaching movements of the arms.
   b. Speaking.
   c. Tongue movement.

Spasticity:

1. A velocity-dependent increase in resistance to passive movement of a limb.
2. Spasticity does not necessarily interfere with the child's attempts at voluntary movement. It is essentially a property of passive movement. Therefore, spasticity is triggered by the interaction between the child and the environment.
3. In children is most commonly due to Cerebral Palsy (CP) and there are sets of spasticity syndromes or patterns that are well recognized. These includes:
   i. Spastic diplegia (both legs involved greater than arms).
   ii. Hemiplegia (involves an arm and a leg on the same side of the body).
   iii. Double hemiplegia (both arms involved more than legs).
   iv. iv. Tetraplegia (all four limbs involved usually severely).

Hereditary Spastic Paraplegia (HSP):

1. A group of genetic, degenerative disorders of the spinal cord.
2. Characterized by progressive weakness and stiffness of the legs. C- This group of disorders is also sometimes referred to as:
   a. Familial Spastic Paraparesis (FSP).
   b. Familial Spastic Paraplegia (FSP).
   c. Hereditary Spastic Paraparesis (HSP).
   d. Strumpell-Lorraine Syndrome.
   e. Strumpell's Disease
3. Primarily characterized by varying degrees of stiffness (rigidity) and weakness of leg muscles and hip abductors, with associated gait disturbances and increasing difficulties walking.
4. Symptoms may begin as early as infancy or early childhood to as late as the eighth or ninth decade of life. In some kindreds, symptoms appear to occur at a progressively younger age with successive generations.
5. (HSP) that is characterized by progressive spasticity as an isolated finding often described as uncomplicated or "pure" (HSP). In those patients' initial findings may include:
   a. Rigidity and increased tone of certain leg muscles, including those of the inner thigh, front and back of the thighs and of the calves.
   b. Weakness of certain leg muscles, dorsi and planter flexors, hip flexor, knee flexors.
   c. Delayed walking, abnormal shuffling gait.

Examination & Evaluation of Children with Neurological Disorders

1. Mental Status Examination:

a. Is the child awake, alert and responsive??
b. Does the child behave in an appropriate manner?
c. Are speech, language, reading and writing skills appropriate for the child's age?
d. Is judgment intact and memory for objects and numbers appropriate for the child's age?

2. Cranial Nerve Function:

a. Are eye, facial, mouth and tongue movements intact?
b. Is sensation on the face normal?
c. Are hearing and balance functions intact?
d. Are smell and taste normal?

3. Sensation:

a. Is the child able to feel light touch, temperature, pinprick and the position of joints throughout the body?
b. Is the sensation the same on both sides of the body and the same in the face, arms and legs?

4. Motor function:

a. Strength normal in all parts of the body?
b. Are there any areas of unusually reduced muscle bulk?
c. When relaxed, can the examiner move the limbs and neck easily?
d. Is there a change with the speed of movement or a "spastic catch"?
e. Is there a change when a distant part of the body is moved or when the child is lying down, seated or standing?
f. Are there any abnormal postures of the limbs, trunk or face?
g. Are there excess involuntary movements such as choreoathetosis, myoclonus, tremor or tics?

5. Coordination:

a. Can the child reach accurately to touch an object far from their body?
b. Are movements appropriately rapid?
c. Can the child manipulate objects with two hands and pick-up small objects between the fingers?
d. Can the child perform rapidly alternating movements such as opening and closing the hands or fingers?

6. Gait:

a. How does the child support his or her weight against gravity?
b. How does the child respond to a loss of balance?
c. Can the child stand and walk stably (depending on the child's age)?
d. Is it possible to walk on the toes or heels, to walk on a narrow line or to walk backwards?
e. Is the child unsteady while standing with eyes closed?
f. Is there any unusual posturing of the arms, legs or face while walking or running?

Neurological Examination:

1. All 12 cranial nerves must be reviewed.
2. Sensory examination should include superficial touch and pain, deep pain, position sense (large joints as well as small).
3. Vibration sense, two-point discrimination, hot and cold perception, and the presence or absence of extinction to bilateral confrontation.
4. Cerebellar and coordination functions need careful review, as do the deep tendon and pathological reflexes.
5. Communication skills are assessed during the neurological examination either as part of or following a broad assessment of the optic and auditory cranial nerves.
6. Speech assessment includes evaluating articulation as well as the content of expression.
7. The listening (i.e., receptive) skill can be assessed by presenting concrete instructions and abstract ideas of increasing complexity that require responses by the patient. Similar performance requirements will help assess reading and writing skills.

Musculoskeletal System:

1. A screening examination is useful in localizing abnormalities when the disability problems are minor.
2. For conditions that may result in major disability, individual joint examinations are necessary. Such examinations include:
   a. **Inspection:** (the two sides of the body should be observed for symmetry in contour and size and differences measured.
   Atrophy, masses, swellings, and skin color changes must be noted).
   b. **Palpation:** (the origin of a pain symptom may be localized by palpation of the various anatomical structures about the joint. Palpation of masses and swellings for consistency can allow to distinguish between bone masses, edema, and joint effusions. To determine the presence of muscle spasm).
   c. **Passive Range of Motion:** (tests are performed by the examiner while the patient is relaxed. When range of motion is limited, range limitation is due to joint surface abnormality; joint fluid excess or loose bodies; or capsule, ligament, or muscle contractures.
   d. **Stability:** (assess whether a pathological condition of the bone, capsule, or ligament is causing abnormal movement (subluxations or dislocations). The joint should be moved under stress in the direction it is not supposed to move by virtue of its contour, ligaments and capsule, with the patient at rest).

e. **Active Range of Motion:** (tests should be performed prior to strength tests in the event pain is a problem. Muscle tension and joint compressions induced by an active movement are less stressful than in a strength test. If pain is minimal in an active range of motion test, the examiner can more easily proceed with a strength test. When active range of motion is less than passive range of motion, the examiner must decide between true weakness, hysterical weakness, joint stability or pain as possible causes).

f. **Muscle Strength:**

i. **GRADE 5:** Normal strength, full range of motion against gravity and against "full" resistance applied by the examiner.

ii. **GRADE 4.** Good strength, the muscle can move the joint it crosses through a full range of motion against gravity with only "moderate" resistance applied by the examiner.

iii. **GRADE 3:** Fair strength, the muscle can move the joint it crosses through a full range of motion against gravity only.

iv. **GRADE 2:** Poor strength, the muscle can move the joint it crosses through a full range of motion only if the pant is positioned so that the force of gravity is not acting to resist the motion**.**

v. **GRADE 1:** Trace strength, muscle contraction can be seen or palpated but strength is insufficient to produce motion even with gravity eliminated.

vi. **GRADE 0:** Zero strength, complete paralysis. No visible or palpable contraction.

vii. The key muscle grade with regard to disability assessment is grade 3. Since any activity a patient may perform is done in a gravity field, if at least grade 3 function is present. then the involved body part can be used.

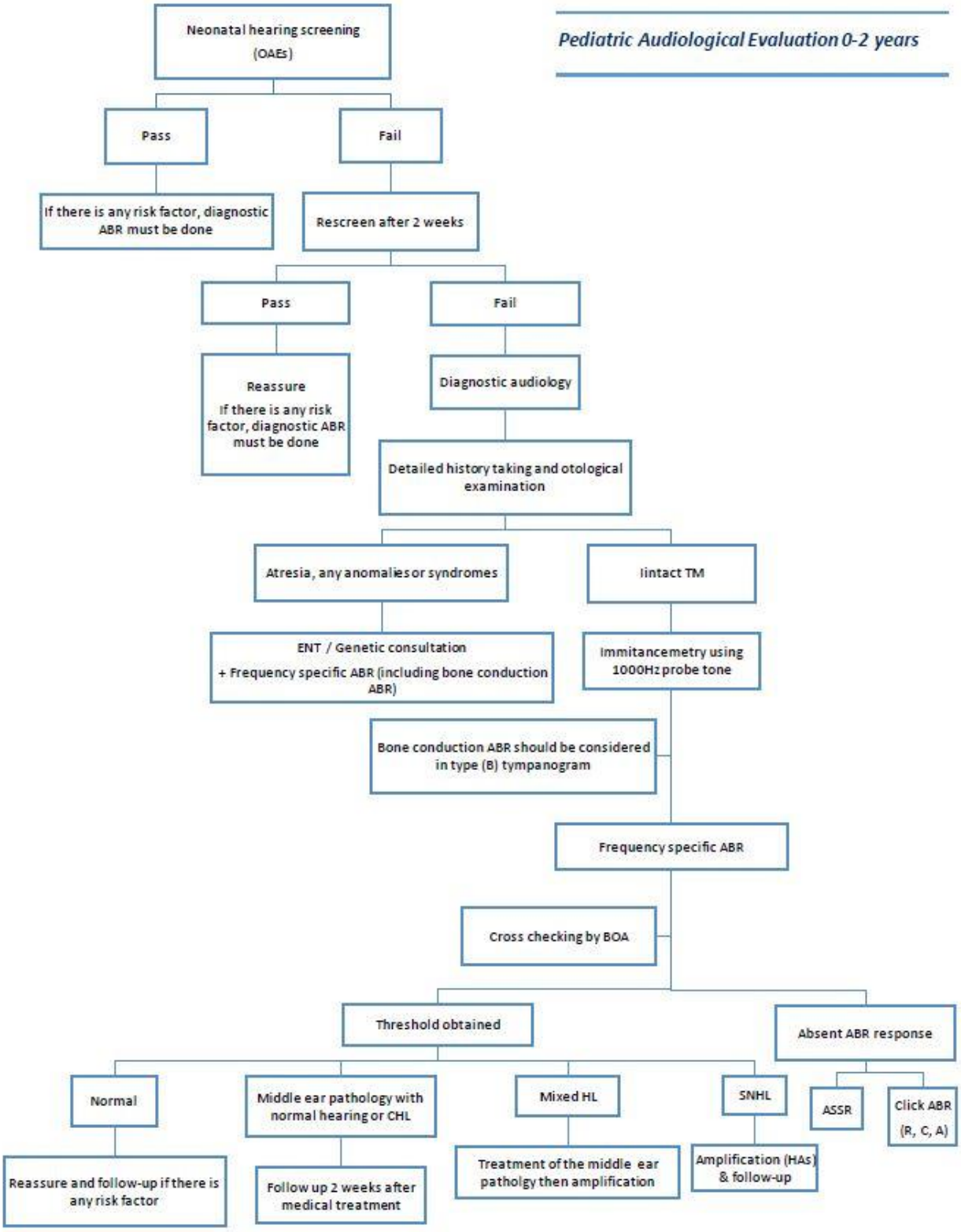
viii. For grades less than 3. external support may be necessary to make the involved part useful to the patient.

ix. For conditions in which weakness is associated with spasticity, the grading system is not as useful in predicting how much the patient may get out of the muscle for the performance of his basic skill needs.

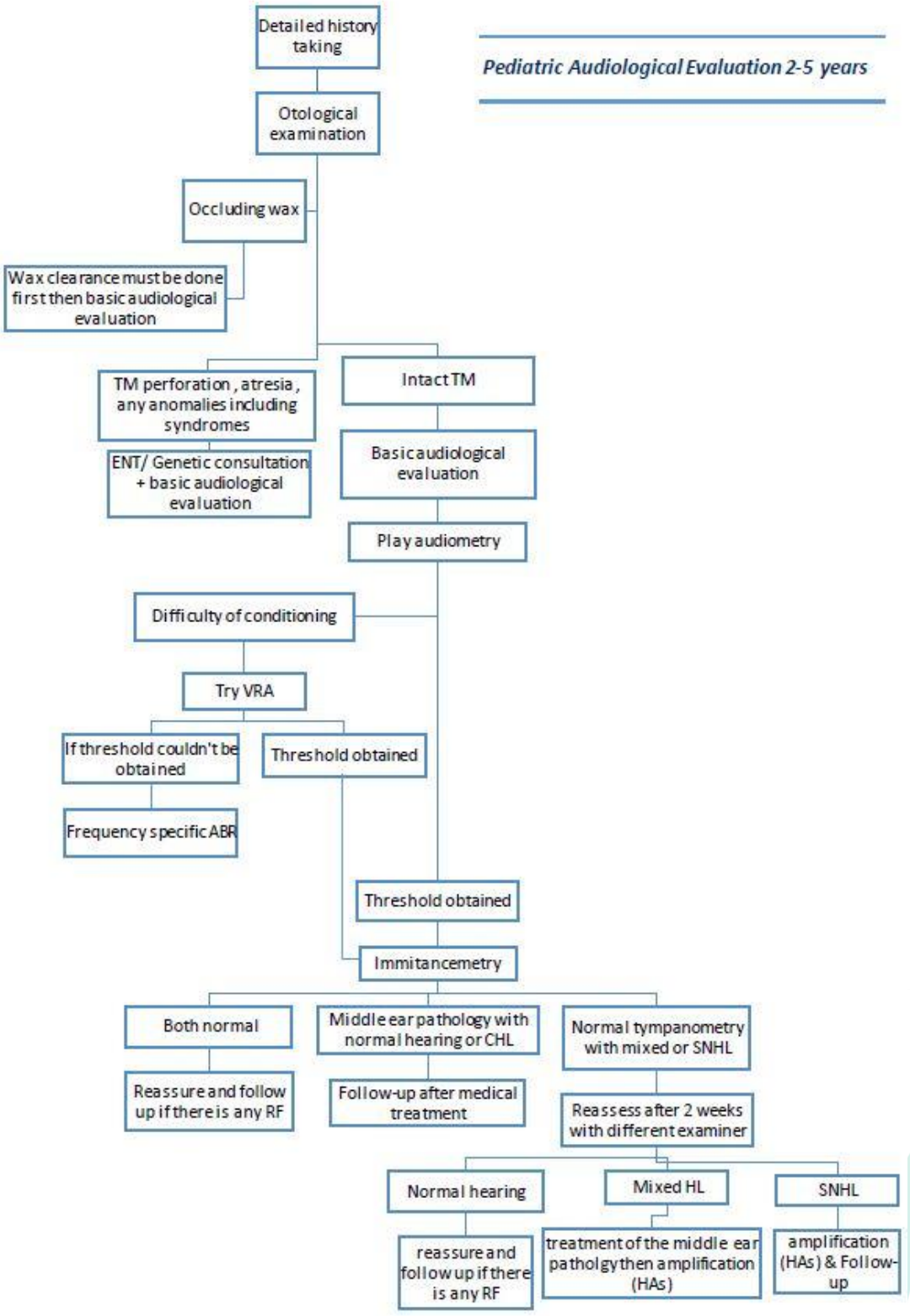
Functional Neuromuscular Examination:

1. The functional examination is the actual translation of the objective neurological and musculoskeletal examinations into performance.
2. It defines at a given point in time the skill of the patient in the execution of the activities of daily living.
3. It is the starting point from which improvement can occur through treatment even if the objective neurological and musculoskeletal signs may not tie alterable owing to the nature of the disease.
4. The functional examination confirms the skill status reported by the patient in the history under Present Illness with regard to ambulation. transfers, eating, dressing, and personal hygiene. The functions to be tested are as follows:
    - **a.** Sitting Balance.
    - **b.** Transfers (abilities for turning from supine to prone and back, rising to a sitting position, rising from sitting to standing, and moving from a bed or low examining table to a chair).
    - **c.** Standing Balance (a necessary prerequisite for safe ambulation. Assessed without support and, if balance is present, nudging from side to side should then be done to assess the patient's ability to recover.
    - **d.** Eating Skills (assessed by demonstration of hand-to- mouth abilities utilizing various examining room objects or by means of actual observation at mealtime).
    - **e.** Dressing Skills (assessed in the examining room if the examiner is present at the time the patient removes the clothes prior to the examination and puts them on at the conclusion).
    - **f.** Personal Hygiene Skills (motions necessary for face, perineal, and back care can usually be mimicked in the examining room. Direct observation of the specific task when actually performed may be necessary if personal hygiene functions are significant disability problems.
    - **g.** Ambulation (walking should be observed if the patient has standing balance. The patient should be essentially unclothed. Walking should be inspected with and without street shoes, and from the front and back as well as from the side. Abnormalities should be described in relation to the phase of the gait at which they occur):
        - **i.** Cadence: Symmetrical? Asymmetrical? Consistent?
        - **ii.** Trunk. Fixed abnormal posture? Abnormal anterior, posterior, or lateral movements?
        - **iii.** Arm Swing. Symmetrical?
        - **iv.** Pelvis.' Fixed abnormal posture? Abnormal pelvic tilt or drop?
        - **v.** Base: Narrow? Broad?
        - **vi.** Stride Length: Short? Asymmetrical?
        - **vii.** Heel Strike and Push Off Present?
        - **viii.** Swing Phase: Knee flexion? Circumduction?

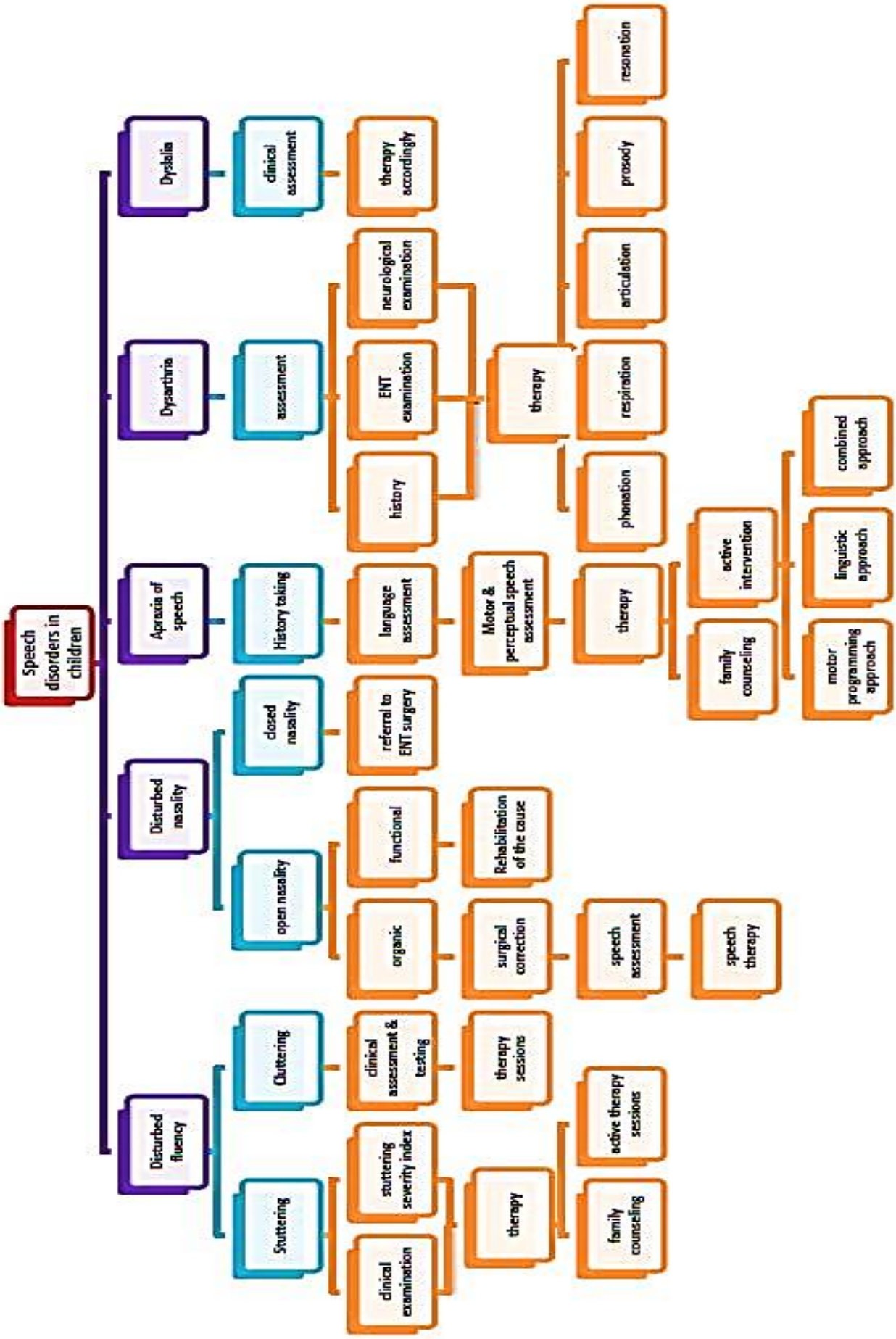
Hearing Assessment Flowchart (0-2yrs)



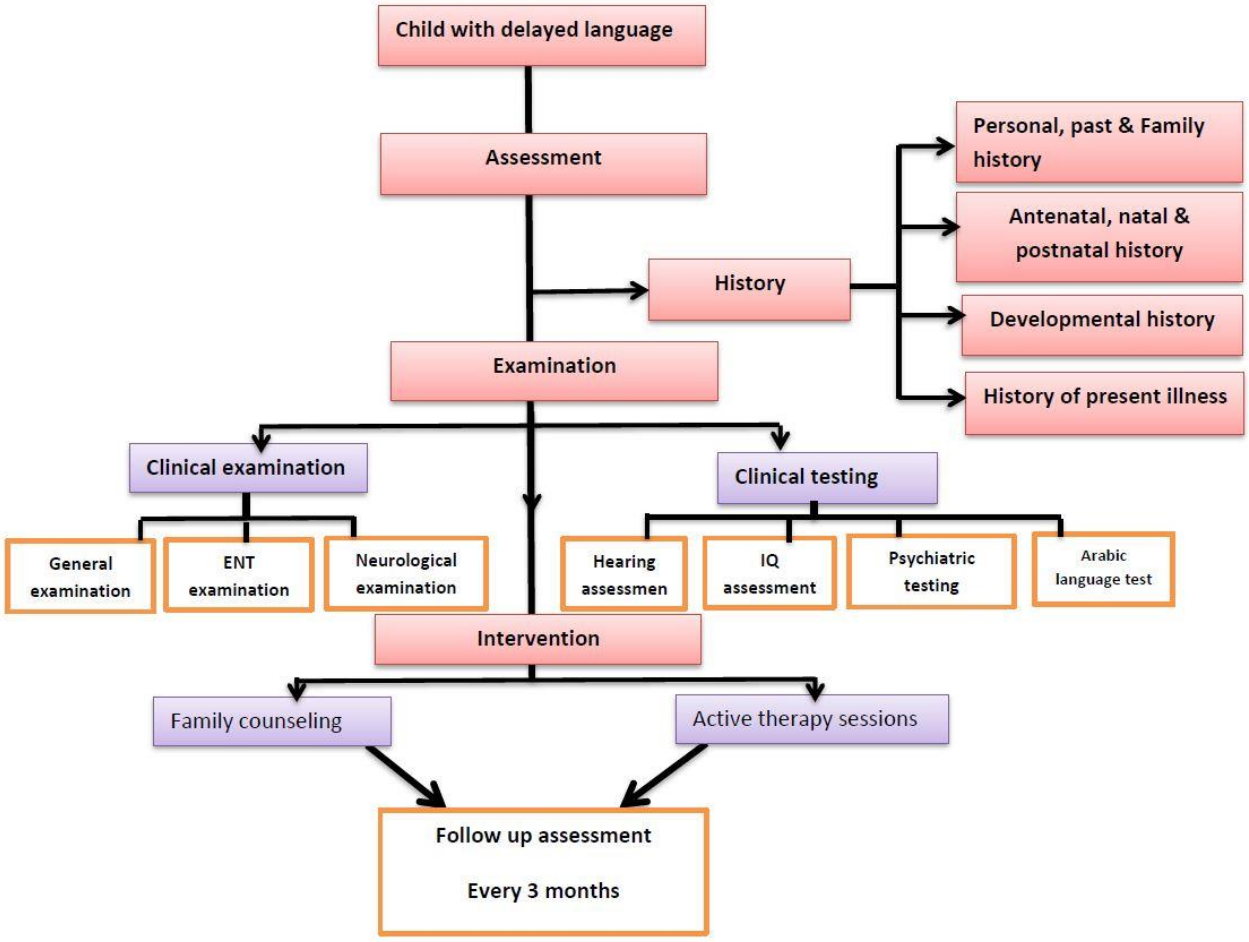
Hearing Assessment Flowchart (2-5yrs)



Speech Disorders



Delayed Language



Amblyopia

Diagnosis:

1. History:
   - Eye problem in childhood such as misaligned eyes, patching or muscle surgery?
2. Ocular examination to rule out an organic cause for the reduced vision.
3. Cover – uncover test to evaluate eye alignment.
4. Refraction: Cycloplegic in children too young to cooperate.

Treatment:

1. Patients younger than 10 years:
   a. Appropriate spectacle correction
   b. Patching: patch the eye with better corrected vision 2 to 6 hours /day for a week per year of age with at least 1 hour of near activity.
   c. Continue patching until the vision is equalized or show no improvement after three cycles of patching.
2. Patient more than 10 years of age: atrial of Spectlacle correction, patching and or may be atropine considered if not attempt previously.
3. Treatment of media opacity.
4. Treatment of anisometropic amblyopia: give the appropriate spectacle correction at the youngest age possible (best if given before age 5).

Congenital Cataract

- Examination of the red reflex is an essential part of healthy baby/child visits in nonverbal children.
- Infantile cataracts that are not extracted in the first 6-8 weeks of life may be associated with irreversible visual loss and nystagmus.

Diagnosis:

1. History :
   - Maternal illness or drud ingestion during pregency?
   - Radiation exposure or trauma?
   - Family history of congenital cataract.
2. Visual assessment of each eye alone.
3. Ocular examinations
4. Cycloplegic refraction.
5. B-scan US may be helpful when the fundus view is obscured.
6. Medical examination by pediatrician looking for associated abnormalities.
7. Red blood cell galactokinase (galactokinase level) with or without RBC galactose -1- phosphate uridyl transferaseactivity to rule out galactosemia.

Treatment:

1. Referral to a pediatrician to treat any underlying disorder.
2. Treat associated ocular diseases.
3. Cataract extraction: usually within days to weeks of discovery to prevent irreversible amblyopia …. Cataract extraction is performed in the following condions:
   a. Vision is obscured.
   b. Cataract progression threatens the health of the eye .
4. After cataract extraction. treat amblyopia in children younger than 9 to 11 years.

Congenital Glaucoma

- When excess tearing is associated with photophobia (light aversion), corneal enlargement and clouding, an immediate referral should be made for possible congenital glaucoma

Diagnosis:

1. History:
   - Other systemic abnormalities?
   - Rubella infection during pregency?
   - Birth trauma?
   - family history of congenital glaucoma.
2. Ocular examination, including a visual acuity assessment of each eye separately, apen light or portable slit lamp examination to detect corneal enlargement and haziness, a dilated fundus examination is performed to evaluate optic disc and retina.
3. IOP and pachymetry are performed, axial length is measured with us to monitor the progression of the disease.

Treatment:

- Definitive treatment is usually surgical.
- Medical therapy is to temporizing measure before surgery.

Medical:

- Oral carbonic anhydrase inhibitor (e.g. acetazolamide, 10 to 15 mg / kg / day) most effective often to clear cornea prior goniotomy.
- Topical carbonic anhydrase inhibitor (e.g brinzolamide) less effective.

Surgical:

- Goniotomy is procedure of choice.
- If the cornea is not clear, trabeculotomy is usually the preferred procedure.

Note:

- **Amblyopia may be superimposed on glaucoma and should be treated by patching.**

Follow Up:

1. Repeated examinations, under anaesthesia when needed, are necessary to monitor corneal diameter, iop , cup / disc ratio , and axial length .
2. These patients must be followed throughout life to monitor for progression.

Congenital Ptosis

- Mechanical obstruction of vision can produce severe visual loss (derivational amblyopia).
- Droopiness of Eyelid (ptosis) or Eyelid hemangioma can also cause visually significant Astigmatism that can result in Refractive amblyopia

Diagnosis:

1. History:
   - Age of onset?
   - Duration? Family history?
   - History of trauma or prior surgery?
   - Any crossing of eyes?
2. Visual acuity for each eye separately, with correction to evaluate for amblyopia.
3. Manifest and cycloplegic refraction checking for anisometropia.
4. Pupillary examination.
5. Ocular motility examination.
6. Measure interpalbral fissure distance, distance between corneal light reflexand upper eyelid margin, levator function, postion and depth of upper eyelid crease . check for bell phenomenon.
7. Slit-lamp examination looking for signs of corneal exposure.
8. Dilated fundus examination.

Treatment:

1. Observation if degree of ptosis mild, no evidence of amblyopia, and no abnormal head positioning.
2. Simple congenital ptosis; if levator function is poor, consider frontalis suspention, if levator function is moderate or normal, consider a levator resection.
3. Macus gunn jaw winking: No treatment if mild, in general, the jaw winking gets better around school age.

Follow Up:

1. If observing, patient should be reexamined every 3 to 12 months.
2. After surgery, patients should be monitored for undercorrection or overcorrection and recurrence.

Retinopathy of Prematurity

- Very premature infants, <1500g or <32wks, are at risk for development of retinopathy of prematurity.

Diagnosis:

1. Screening recommendations
    a. Birth weight <1500g.
    b. Gestional age <32wks.
2. Dilated retinal examination 4 weeks after birth.

Treatment:

1. **Photocoagulation (laser therapy)**
    - Photocoagulation is the first line of defense against ROP. The setup is much like a retinal exam, except your child will be given local or general anesthesia. The ophthalmologist uses a diode laser mounted on the indirect ophthalmoscope to make tiny “burns” in the periphery of the retina, to prevent further growth of abnormal blood vessels.
    - Your child's doctor will set follow-up exams — usually every one to two weeks — to see how the eyes are responding to the laser treatment. If the ROP continues to worsen, your child may need additional laser treatments or possibly eye surgery.
2. **Cryopexy (cryotherapy)**
    - Formerly the procedure of choice for treating ROP, cryopexy uses a penlike instrument called a cryoprobe to freeze parts of the retina's periphery through the outer wall of the eye. Though it's largely been replaced by laser therapy, cryopexy is useful when the retina can't be fully seen (because of a hemorrhage, for example).
    - Because both photocoagulation and cryopexy destroy part of the retina's periphery, your child may lose some of his side vision with these treatments. However, the procedure aims to save his “central vision”—the most important part of sight — which is necessary for things like reading and driving.
3. **Eye surgery**
    - If your child's retinal becomes partly or completely detached — Stage 4 or 5 — your doctor may refer him to a retinal surgeon for treatment, usually scleral buckling or vitrectomy.
        a. Scleral buckling involves placing a silicone band around the eye and tightening it until the retina is close enough to the wall to reattach itself. The band, called a scleral buckle, can be left in place to protect the eye for months, or sometimes years.
        b. Vitrectomy involves removing the vitreous (the gel-like substance that fills the back of the eye) and replacing it with saline solution or oil. The scar tissue on the retina can then be peeled back or cut away, allowing the retina to flatten back down against the wall of the eye.

The Bilaterally Blind Infant

Diagnosis:

1. History:
   - premature?
   - Normal development and growth?
   - Maternal infection, diabetes or drug use during pregensy ?
   - Family history of eye disease?
2. Evaluate the infant ability to fixate on an object and follow it.
3. Pupillary examination.
4. Look carefully for nystagmus.
5. Fundus examination for optic nerve and retinal evaluation.
6. Cycloplegic refraction.
7. ERG.
8. Consider a CT or MRI of the brain.

Treatment:

1. Correct refractive errors and treat known or suspected amblyopia.
2. Parental counseling is necessary in all conditions.
3. Referral to educational services for the visually handicapted may be helpful.
4. Provide genetic counseling.
5. If neurological or endocrine abnormalities are found or suspected, the child should be referred to a pediatrician for appropriate work up or management.

Environmental Conditions for prevention of child disability

There's increasing numbers of disabled newborn children that can be referred to many factors that can be classified into two categories:

Genetic and/or Environmental

A) genetic factors which can be caused by abnormal gene that caused due to parents inherited gene or exposure of the mother before, during or after labor such as

- Infection; German measels; Toxoplasmosis
- Premature baby
- Neonatal hypoxia
- Exposure to irradiation either to Ionizing such as x Ray or gamma Ray or one neglected indoor source called Radon, non-ionizing radiation such as exposure to electromagnetic I field that our life for a lot of electrical and electronics Utensils.
- Food additive such as artificial sweeteners color flavor smells etc
- Junk food
- agricultural pollutants such as bisticids, fertilizer, herbicide, hormones, Vermont..etc
- A lot of medications

  Don't give medications to pregnant woman except if severely indicated

The best way is the prevented and genetic study and by genotherapy otherwise we have to follow the rules of palliative regime of rehabilitation training reeducation exercise.

Child Disabilities

"We offer comprehensive reviews of selected topics and comprehensive advice about management approaches based on the highest level of evidence available in each case. Our goal is to provide an authoritative practical medical resource for pediatricians.

We hope that such an approach will encourage clinicians to apply available evidence to their practice and also track compliance with desired practices."

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