Prevention, Early Detection and Management of Childhood Disabilities

Training Manual For Primary Health Care Physicians

This Publication Was Produced Within The Framework Of
Child-Focused Community-Based Rehabilitation Project
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Plan International - Egypt

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Association For Health and Environmental Development (AHED)

Disability Program

AHED is an Egyptian Non-Governmental Organization established and registered with the Ministry of Social Affairs in November 1987. It is a multi-disciplinary organization formed to assist in the development and implementation of alternative policies and systems in the areas of **H**ealth, **E**nvironment, and **D**isability, which are capable of responding to the needs and rights of Egyptian society as a whole**,** and the most disadvantaged and marginalized groups in particular. In all its work, **AHED** adopts the principles of equity, human rights, comprehensive integrated development and active community participation.

Foreword

This training manual was developed and published by the "Association For Health and Environmental Development (AHED)" within the framework of the "Child-Focused Community-Based Rehabilitation project". The project is being implemented in partnership among:

- International organizations; Plan International – Egypt and Save the Children - UK,
- National authorities represented by the Ministry of Health and Population, the Ministry of Education and the Ministry of Insurance and Social Affairs, and
- Egyptian Non-Governmental Organizations (NGOs) working on the national level which are: the Association for Health and Environmental Development (AHED) and Caritas-Egypt represented by SETI center for Training and Researches in Mental Disability.

The project aims at establishing copmprehensive Community-Based Rehabilitation (CBR) demostrative models in different four Egyptian Governorates, that are being implemented and will be sustained through community based organizations.

The training manual aims at enhancing the role of facilities of health care services provision on the primary level in the areas of prevention, early detection and management of childhood disabilities as a part of the needed multi-sectoral network to address the question of childhood disability in Egypt.

The role of facilities of health care services provision in addressing the childhood disability has been internationally recognized. In addition, the experience of establishing demonstrative CBR models indicates that the Primary Health Care (PHC) facilities can be the main player in the prevention, early detection and initial management including the referral. This is believed to be true because of two facts. First, Egypt has a relatively huge PHC infrastructure with well geographically distributed facilities. Second, the roles of these facilities, especially in compulsory immunizations, make them a good first contact for infants and children.
The manual has been made to enhance this role through providing the needed conceptual framework, knowledge and skills related to prevention, early detection and initial management of childhood disabilities.

The manual was developed with the contribution of sound experts in medical and rehabilitation fields.

Further development of the current manual in next editions will depend, in principle, on the feed-back of experts as well as the primary health care physicians who can enrich the manual with their practical experiences.

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Training Syllabus

Course Description: A hands-on, competency-based comprehensive training course for the primary health care physicians. It is designed to provide knowledge, skills and attitudes necessary for better understanding of the childhood disability question including its underlying causes and risk factors and process. It also provides the participants with effective schemes for prevention, early detection and initial management including proper referrals and follow up.

Course Duration: Eight training days (6 hours a day).

Language: English. Arabic language is used along the course in discussions, working groups as well as in clinical sessions.

Participants: 16-20 participants from the physicians working in the primary health care facilities.

Overall Objective: To assist in building the capacity of the primary health care facilities and health facilities attached to Non-Governmental Organizations (NGOs) in the area of prevention, early detection and initial management of childhood disabilities within the framework of a comprehensive community based approach.

Expected Outcome: At the end of the training course, it is expected that the health team that will undergo the training will be more capable of identifying and managing impairments and disabilities in children examined at the Primary Health Care (PHC) centres and the NGOs attached health facilities and to advice on proper referrals for the different types of disabilities. Moreover, they will be more capable of giving proper advice to mothers and parents on prevention from risk factors leading to disabilities.
It is also expected that relationships between the health team, that will undertake the course, will be strengthened with other sectors in the area of disability particularly with the educational and social services. In addition, the model will be monitored and evaluated by project team.

Specific Training Objectives: By the end of this course, participants should be able to:

1. Display a positive understanding and attitudes towards the needs of the disabled children and their families and the disability question in general in Egypt, discriminate between different models of rehabilitation and recognise the role of community based rehabilitation (CBR) model.
    1.1. Explain the process of disablement.
    1.2. Identify proper interventions at different levels of the process and identify the goals of the World Programme of Action in relation to the disability question.

1.3. Describe the role of social and environmental barriers in the disability process.
1.4. Recognise and describe the role of parents and care takers as primary actors and communities in the rehabilitation process.
1.5. Recognise the different models of rehabilitation with special focus on the role of PHC and its relationship to CBR.
1.6. Identify the size and nature of the problem of disability in Egypt and the situation of existing services.
1.7. Recognise the concept of rights of disabled people.

2. Provide appropriate advice to families on causes and risk factors of disabilities and means of prevention.
   2.1. Recognise and describe causes and risk factors of the major disabling conditions of children.
   2.2. Recognise important preventive measures of important disabling conditions.

3. Make early detection of impairments and disabilities in children from 0-18 years through conducting screening programmes.
   3.1. Explain the concept of screening in general and developmental screening in particular, criteria for their application and their role in early detection of childhood disabilities.
   3.2. Recognise and describe normal milestones of development.
   3.3. Identify watch signs of developmental delays and abnormalities in child development.
   3.4. Apply simple procedures for testing certain aspects and functions of development and make early detection of disability conditions (neurological & motor, visual, hearing, speech and mental).

4. Provide health care on the primary level to children with disabilities, including follow-up of their particular health problems and normal health needs (e.g. nutrition, growth etc.)
   4.1. Identify common health problems relevant to children with disabilities.
   4.2. Describe essential management procedures and care for the most important health problems.
      4.2.1. Identify proper medical, surgical, and therapeutic interventions required in the different types of disabilities at the primary care level.
      4.2.2. Display ability at conducting examination of the children to assess their disability, its type and degree.
      4.2.3. Identify and describe the required interventions in each case, including, medical, surgical, therapeutic.
      4.2.4. Assist parents to follow-up and successfully utilise equipment, aids, or different regimen and management procedures at their homes.
      4.2.5. Describe and design monitoring schemes and follow - up interventions with parents.
   4.3. Conduct psychological assessment and management.
      4.3.1. Conduct developmental assessment to all referred children.

4.3.2. Identify psychological problems of the child and family.
4.3.3. Identify and describe proper counseling and guidance to families.

4.4. Conduct social assessment and management.
4.4.1. Identify basic attitudes of family towards their disabled child and their coping mechanisms.
4.4.2. Identify available community resources and services and advice on appropriate ones for the child's and family needs and ways of procuring them.

5. Make proper referrals to other levels of health care as well as to other sectors.
5.1. Identify appropriate referral needs not fulfilled at the primary care level including medical, social and psychological.
5.2. Fill out standardised referral forms and follow up their procedures.
5.3. Identify available referral routs and institutions.
5.4. Recognise the roles and limitations of the different programmes, institutions providing rehabilitation in the community or on the secondary and tertiary level services and specialised rehabilitation institutions e.g. educational, vocational etc.
5.5. Assist families of disabled children to obtain proper services from the referrals.

6. Provide parents of children discovered as disabled with proper advice and assurance and ways of dealing with a disabled child.
6.1. Identify the most common problems facing families with disabled children.
6.2. Describe important management procedures and advice to parents.
6.3. Recognise stages of grief that parents go through upon receiving news that their child is disabled.
6.4. Provide support and advice to parents with disabled child and help them positively.

Course Methods: The course utilizes a variety of interactive training techniques that encourage group discussions, brainstorming and clinical practice demonstrations. Various types of audiovisual aids will be utilized by all the trainers; data show, overhead projector, slide projector and flip charts. Clinical training on screening techniques will be conducted.

Evaluation Techniques: A daily pre- and post-test will be used to assess participants' progress. The following scale will be used in evaluating participants' performance:

Multiple choice tests	40%
Clinical training	40%
Attendance and participation	20%

Participants who successfully complete this course will be awarded a certificate of completion.

Training Schedule and Timing

Day	*Session (1)*	*Break*	*Session (2)*	*Lunch*	*Session (3)*
	9.30 – 11.30	**11.30- 11.45**	**11.45 – 1.45**	**1.45 – 2.30**	**2.30 – 4.30**
1st	Introduction to Disability: concepts, types, and status of disability in Egypt		Child Rights and Disabled Rights		Rehabilitation and Community Based Rehabilitation (CBR)
2nd	Risk Factors and Causes of Disability		Effects of Disability		Prevention of Disability
3rd	Early Detection and Identification of Disability		Early Detection and Identification of Disability		Early Detection and Identification of Disability
4th	Visual Disability		Visual Disability		Visual Disability
5th	Mental Retardation		Mental Health Problems		Child Abuse
6th	Hearing Disability		Hearing Disability		Speech & Language Disabilities
7th	Neurological & Motor Disabilities		Neurological & Motor Disabilities		Neurological & Motor Disabilities
8th	Available Disability Services		Registration and Referral		Registration and Referral

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Chapter 1

Introduction to Disability

By the end of this chapter the PHC physician should be able to:

- Explain the process of disablement.
- Identify proper interventions at different levels of the process and identify their relationship to the goals of the world programme of action in relation to the disability question.
- Identify the size and nature of the problem of disability in Egypt and the situation of existing services.
- Recognize the effects of disabilities on disables and their families.
- Identify available facilities addressing the disability question in Egypt and recognize their gaps.
- Recognize the different models of rehabilitation with special focus on the role of PHC and its relationship to CBR.
- Describe the concept of rights of disabled people and particularly disabled children.

Section 1

Defining Disability and Required Interventions

Disability: What are we talking about?

> *"No leper shall come within the gates of the borough and, if one gets in by chance, the sergeant shall put him out at once. If one willfully forces his way in, his clothes shall be taken off him and burned, and he shall be turned out naked. For we have already taken care that a proper place for lepers shall be kept up outside the town, and that alms shall be there given to them"*
>
> **A law enacted in 14th century in Britain (Cartwright 1977)**

Analyzing the situation of children with disabilities requires foremost agreeing on the nature of the question at hand. This requires identifying and clarifying the multiple facets of the question and its consequences on the child, their family and community at large.

This step is essential if appropriate interventions are to be discerned. In turn, such an understanding becomes a frame of reference for assessing existing interventions whether on the policy, legislative levels or on the levels of existing services and social support systems.

Identifying a standardized definition for disability has been and continues to be both a national and an international problem.

The lack of consensus on defining disability is related to the complexity of the problem and its multi-facets.

What is disability? (Disability or the disabling process?)

In human development in general, as in childhood development, the health status of the human being is the result of the continuous interaction between the biological being on the one hand and the surrounding environment on the other. Disability is one of the possible outcomes of this interaction.

However, the effects of this interaction on the human being are not only governed by the biological outcome, it is modified and restructured through its interaction, once again, with the surrounding society and environment.

Hence, the way disability is defined by any society, at a certain time, reflects the way the society perceives the condition. This perception reflects, in its own term, the level of the prevalent understanding, attitudes to the question as well as the way this society is

economically and socially organized to deal with the needs of the different people and groups within it.
Old traditions of confusion and fear related to the explanation of causes of ill-health, particularly chronic ill-health, inherited from the past, played a role in passing on many misperceptions and negative attitudes towards disabilities across different generations.

For many centuries in Europe Leprosy was considered a crime and the disfigurement it produced was considered as a divine punishment for sin (Cartwright 1977). Accordingly, Lepers were treated as criminals who were to be locked away from society.

Among other causes, the attitude of isolation and segregation of the disabled can be traced back to such historical attitudes which were augmented by the fear of the disabled and the possibility of transmission of diseases.

The above cited law for people suffering from leprosy in fourteenth century Britain conveys such attitudes and fears from a specific type of disability which must have been transmitted to other types.

Until almost the end of the nineteenth century mental illness was considered 'akin to crime' and in the very few hospitals which catered for the mentally ill, the patients were confined to cells and chains.

Mental retardation, till recent times was confused with mental illness and mentally retarded persons were locked in mental hospitals and asylums.

The above examples from past and recent history are cited to show many of the causes behind inherited confusions in dealing with the subject of disability. This confusion reflected itself in the lack of clear consensus between different people, different societies and different professionals within the same society on the question to be addressed.

In addition, different "schools" or "professions" gave their own definitions and categories according to their particular interest, contribution and hence their approach to the question.
Consequently, different models or institutions of care, such as: Health, Social or Educational, tend to give their own interpretation to the phenomenon.

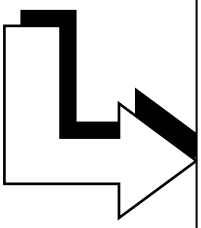
The "Biomedical Model" tends to describe it from the point of view of the etiology, or from the point of view of describing the physical or physiological characteristics of a "condition" or a "disorder" which is deviant from the "norm."

On the other hand, institutions such as insurance or social services tended to define it from the view of the degree of incapacitation produced by the disability; i.e. the inability to get a job and hence, the entitlement to allowances and/or financial support.

However, these various definitions gave rise to many problems. Discrepancy in available statistical data and information related to the size of the problem and its types reflected, more than anything else, the discrepancy in defining the problem at hand. In addition, erroneous definitions and narrow understanding lead to the propagation of sticky labels which compounded the problem of the disabled children, on the one hand, and lead to narrowing of interventions on the other.

International development in defining disability (A move towards a universally standardized definition of the phenomenon)

Within the above context a new international classification was developed by the WHO in 1980; namely the classification of Impairments, Disability and Handicap (WHO, 1980).



WHO Definition of Disability (1980)

First Phase: Impairment

In the context of health experience, an impairment is any loss or abnormality of psychological, physiological, or anatomical structure or function. i.e., it refers to an underlying molecular, cellular, psychological or structural disorder within an individual.

Second Phase: Disability

In the context of health experience, a disability is any restriction or lack (resulting from an impairment) of ability to perform an activity in the manner or within the range considered normal for a human being. i.e., it refers to a stable and persistent deficit in function, often the consequence of an impairment, and also confined to the individual.

Third Phase: Handicap

In the context of health experience, a handicap is a disadvantage for a given individual, resulting from an impairment or a disability, that limits or prevents the fulfillment of a role that is normal (depending on age, sex, and social and cultural factors) for that individual. i.e., it refers to the limitation on carrying out social roles defined as appropriate for an individual of certain age, gender or class.

In its definition the WHO described what can be called the disablement process which results in different outcomes through the interaction between the biological being and the environment and society.

The WHO manual provided three different planes of the experience where impairment described the condition on the level of the organ or the function, i.e. the pathology; while the disability described the condition on the level of the activity expected of the person and the handicap described the condition on the level of the social roles, i.e. when the person is hindered from taking on social roles appropriate to his/her age and sex.

New definition

Despite the important and historical development that took place in understanding the question of disability as a consequence of the 1980 WHO definition, the WHO continued in its effort to further develop a better understanding of the question particularly inspired by the rights of the disabled movement.

Accordingly, in the year 2000, the WHO produced final draft for a new definition (The International Classification of Functioning, Disability and Health "ICIDH-2").

The new definition and the classification of disability shed more light on the phenomenon of disablement through focusing on its relativity. This was shown through placing disability and ability on a continuum whereby all people lie at one point between two factious ends of complete disability and complete ability (functioning). The classification identified three determinants of the outcome, namely, the body and its organs, the human being as a whole and the social environment they live in. In doing so, the new definition emphasized that the human functioning state as a result of the interaction between these three dimensions i.e., the biological status with the psychological status and the coping mechanisms of the human being and the environment and the society surrounding.

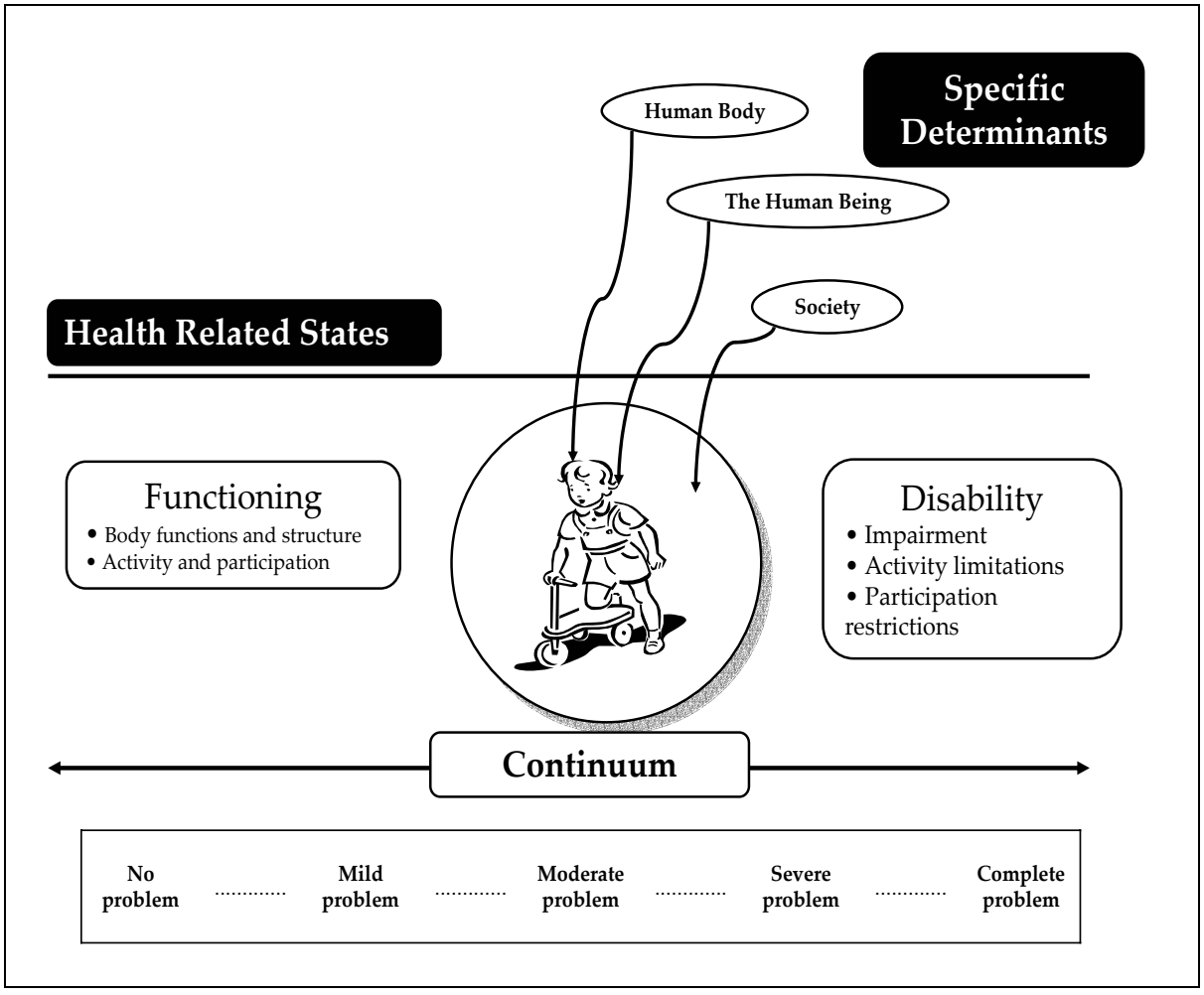


Figure (1-1): New WHO definition of disability (2000)

Classification/categorization of disability types

An important outcome of the WHO's definitions has been the attempts to classify types and degrees of disabilities in harmony with the definitions and internationally standardize them. However, in both the ICIDH (1980) and the ICIDH-2 (2000), the classifications were lengthy and complicated. In the International Epidemiological Study of Childhood Disability (IESCD), the following simplified list of categories was adapted from the (ICIDH). For more thorough categorization look (ICIDH – 2).

Table (1-1): Classification/categories of disability types

Categories	Definition
1. Motor disabilities including;	Decrease or difficulty in the ability to:
• **Fine Motor**	- Manipulate and the use of hands and fingers
• **Gross Motor**	- Sit and balance - Move
2. Hearing	Decrease in ability of hearing compared to peers
3. Speech	Difficulty in ability to speak compared to peers
4. Vision	Difficulty or decrease in ability to see compared to peers
5. Cognitive	Difficulty or decrease in ability to understand compared to peers
6. Fits or Seizures	Momentary loss of consciousness which is accompanied by jerking movements of muscles
7. Multiple Disabilities	Difficulty or decrease in two or more abilities excluding speech as secondary to MR or Hearing

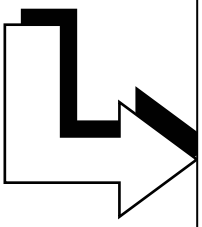
Table (1-2): Criteria for measuring the above disability categories according to the three stages of severity; namely , mild, moderate and severe.

Types and Categories	Degrees					
	Mild		**Moderate**		**Severe**	
Motor						
Fine motor	Some difficulty. Grasp may be weak or crude but can hold most instruments (pen, knife, glass,...etc.) Can dress oneself May not be able to fully raise arms above head		Difficulty in holding implements and using arms for dressing Unable to pick up small pill. In hemiplegia one hand completely useless		Does not use either hand for anything for more than reaching or pointing.	
Gross motor	• Some difficulty in prolonged sitting • Does not need support • Walks unaided - may have a limp or walk with an artificial limb. • May have difficulty in climbing steep steps or drags foot		• Needs support to sit upright • Needs maximum aid to walk, but still can move about		• Unable to maintain balance in upright position without full support • Cannot walk at all	
Hearing	**by dB**	**by Behavioral parameters**	**dB**	**Behavior**	**dB**	**Behavior**
	20 - 50 dB loss in best ear	Some difficulty in hearing. Needs to be placed in front of class. Understands normal speech, speaks normally. May wear a hearing aid.	50 - 70 dB loss in best ear	Difficulty in hearing speech even with hearing aid. Affects expression of speech	No response	No useful hearing evident
Speech						
	Speaks in sentences (depends on age). Can be understood but speech is not sophisticated enough to get more than simple ideas across.		Speaks in single words only which can get across basic needs		No speech at all	
Vision	**Measurements**	**Behavioral features**	**Measurements**	**Behavioral features**	**Measurements**	**Behavioral features**
	Can see Chart with pinhole	Corrected by glasses Can get around without difficulty	6/18 or 20/60	Not correctable by glasses	6/60 or 20/200	No vision except light Needs cane.
Cognitive	IQ 50 – 69		IQ 49 - 35		IQ 34 - 20 in addition to profound <20	
Fits or Seizures	2-4 Fits per year		1fit /week to 1/3 fits months		More than 1 fit /week	
Multiple disability	A mild decrease in 2 or more of the above mentioned functions		A moderate decrease in 2 or more functions		Severe decrease in 2 or more functions	

Required interventions

The WHO initiative not only provided for the first time internationally standardised definitions which help in cases of comparability, but probably what is more important is that it provided better understanding of the required interventions and the levels of interventions..

In the World Program of Action concerning Disabled persons in 1982, the U.N in its 37th session adopted for the first time three major areas of required interventions; namely prevention, rehabilitation and equalization of opportunities.



Prevention

All the measures aimed at preventing the onset of mental, physical and sensory impairments (primary prevention) or at preventing impairment, when it has occurred, from having negative physical, psychological and social consequence.

Rehabilitation

A goal-oriented and time-limited process aimed at enabling an impaired person to reach an optimum mental, physical and/or social functional level, thus providing her or him with the tools to change her or his own life. It can involve measures intended to compensate for a loss of function or a functional limitation (for example by technical aids) and other measures intended to facilitate social adjustment or readjustment.

Equalization of Opportunities

The process through which the general system of society, such as the physical and cultural environment, housing and transportation, social and health services, educational and work opportunities, cultural and social life, including sports and recreational facilities, are made accessible to all.

The diagram below is an attempt to show the relationship between the different dimensions of the disablement process and the required interventions.

If the disablement process is looked at holistically, the following important levels could be added.

1. The risk factors in the society and environment to which exposure may lead to a disease or a trauma or other types of ill-health conditions. Increase in the prevalence of these risk factors increases in turn the possibility of exposure and harm. Such risk factors vary from lack of safety measures in the environment including, roads, homes etc. to consanguinity, pollution…etc.

2. The second important level of the process in some cases is the stage of occult disease produced by these risk factors. The importance of this stage is that in many cases early detection could be life saving or disease and disability preventive. This ranges from

inherited diseases such as Phenyl Ketonurea and congenital hypothyroidism, to the early detection of hearing loss whose correction could prevent speech and language impairments.

3. The third level is that of apparent or clinical disease. At this level, prompt and appropriate treatment may in many cases prevent an occurring impairment and disability, such as the case of bacterial meningitis and recurrent otitis media…etc.

4. The fourth level is that of the occurring impairments. Again, interventions can prevent or decreases the degree of disablement produced. This includes proper diagnosis of impairments and providing proper management and rehabilitation efforts.

5. The fifth level is the level of disablement in which addressing the gaps in society and environment and equalization of opportunities can prevent or reduces the level of handicapping conditions produced through the interaction of the disability with the environment. A facilitative environment will produce a positive outcome while a hindering non facilitative environment and society will produce a negative outcome.

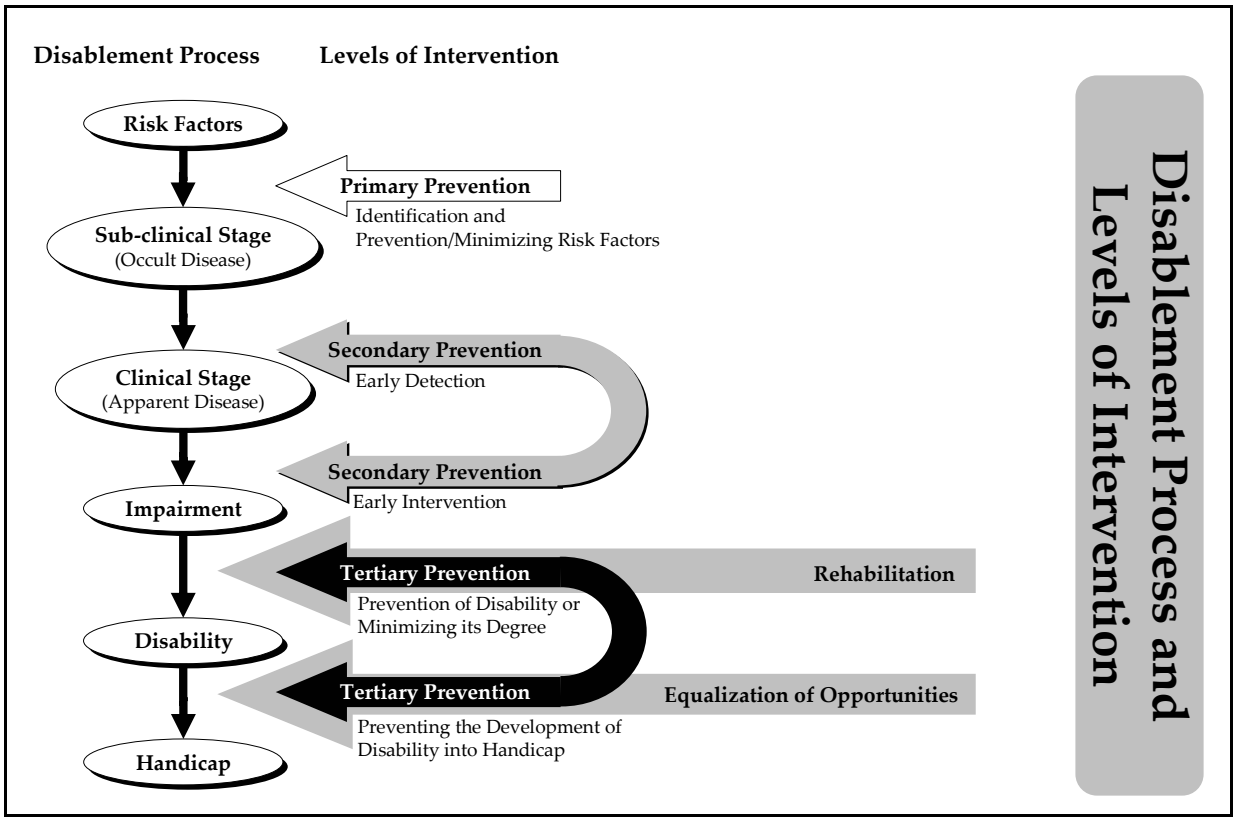
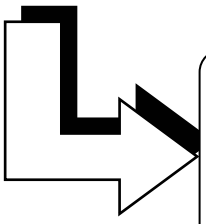


Figure (1-2): Disablement process and levels of interventions

Benefits of early intervention



Benefits of Early Intervention

- Helps children with disabilities
  - – Gains in cognitive, physical, language, and social skills
- Benefits for families
  - – Helps families manage the child
  - – Reduces stress
- Benefits for society
  - – Reduces need for institutional placement
  - – Reduces need for special education
  - – Saves money

Case study

Needs and challenges: *The story of "Warda"*

- I was called to see a girl "Warda", an eight years old child with pneumonia, about nine months after taking over a small health unit in Fayoum.
- I had not noticed the child before.
- Discovered that she is an old case of polio.
- General health condition was very bad.
- She had severe complications with severe muscle wasting, contractures and deformity of the joints, inability to use one leg completely.
- When inquired about her story and how these complications were not prevented was told:
  - Child got fever when she was 1.5 years old.
  - She took a shot at the local health unit
  - Later she had an acute case of paralysis which she nearly died from, after which the paralysis set in.
  - The family was told to go the polio institute in Cairo.
  - She received a caliper, however the caliper was too heavy and hurt her leg which made her stop using it.
  - When she became of 6 years of age she went to the school and was very happy at first even though the school was far from home, about three kilometers.
  - However, many times she arrived late and the schoolteacher used to punish her and the students used to laugh at her.
  - She stopped going to school and her family preferred to make her stay home, particularly that she is a girl and they feared that she could be taken advantage of.
  - Her psychological condition deteriorated rapidly and she stopped even wanting to play with other children and even her siblings, and her general health condition followed.
  - Her grand mother when upset used to call her crippled and she got upset, she would tell her siblings I hope god punishes you by making you crippled too.

I was at a loss, What is really the problem with Warda?

- Is it the pneumonia
- Or the general physical health condition,
- Is it her psychological condition, the underlying disability, or the lack of knowledge of the doctor
- Or is it the poverty and lack of awareness of the family
- Or is it the educational system including the attitudes of the teachers and the students?
- And hence what was my role?, the only thing I could offer was to send her again to the polio institute.
- However, I realized how much our existing medical system and we as doctors in general know very little about disability and offer even less.
- If we want to address the problem impairments and their sequel we have to deal with the underlying causes of disablement in the community and society.
- We cannot work alone, we have to work with others. Disability is not a disease it's a condition which requires the interventions of other sectors in society.

There is a need for a different system

Conclusion

- Any comprehensive health care system must include disabled population.
- Bringing disability to health care is a challenge to its development.
- It is a challenge that requires Primary Health Care to be truly a part of a comprehensive and integrated system of dealing with the causes and sequel of ill-health.

Questions

- What are the problems of Warda, or what are the different dimensions of Warda's problem?
- What is the impairment and the disability?
- What are the required interventions?
- What should be the role of the PHC and the health care system?

Section 2

Effects of Disability

For those of us who have not experienced severe disability, it is difficult to understand the effects that such conditions can have on one's life. It is often difficult to know what to do when we see someone with a disability. Will they be offended if I look at them? Can I ask what happened? Do they need help? Should I help? How?

The effects of being disabled

Emotional adjustment

Peoples' reactions to their disabilities vary with the individual, the severity of the handicap, and the time of life when the condition began. A person's emotional adjustment can be made more difficult by social discrimination and the limited concessions made by the community.

For those individuals who are born with an impairment, the emotional adjustment is on-going; it changes with the different phases of life. These people have never experienced what we consider a normal life, and the awareness that they are different may develop slowly as their social contacts increase. People with congenital handicaps may have more difficulty developing some skills. For example, developing "mainstream" language is harder for someone born deaf than it is for a person who becomes deaf after the age of three. People with congenital conditions must deal all their lives with the reality and permanence of their situations, but they do not experience the intense loss and adjustment difficulties of those who are suddenly disabled.

People who acquire an impairment through injury or sudden disease have a more dramatic and immediate adjustment to make. They must cope with their medical problems. They have to adjust to a new social situation and accept new attitudes and expectations from others. They may have to live with new limitations on their access to community facilities and services and suffer new limitations of opportunities. In addition, they may be coping with job loss, family crisis, and marital stress. Is it any wonder that their emotional turmoil often includes anger, guilt, grief, jealousy, resentment, helplessness, and frustration? Sudden disability is a terrible situation to cope with, but it is one that may happen to any of us at any time.

A slow degenerative condition puts somewhat different emotional stresses on affected individuals and those around them. The shock of adjustment is cushioned by time before impairment and functional limitation occur. The person may have time to put in place structural features required for continued mobility. Time may offer a chance for the person to get training to lessen the eventual effects of the condition, and thus avoid experiencing a disability which affects the quality of life. However, there is still the emotional turmoil of anger, resentment, guilt, and jealousy to work through before attaining acceptance.

Parents who discover that their baby is handicapped suffer tremendous emotional upheaval. It has been compared to the death of the imagined "ideal" child that every parent expects to have. The grieving process is compounded by resentment that an imperfect substitute has been left in the place of the anticipated infant. The reactions of friends and family members can be devastating at this time. However, the emotional reactions of parents vary, and professionals must not assume that a specific pattern of reaction is inevitable. Cultural values have a large influence on the degree of trauma the parent's experience.

Though common emotional reactions can be identified among large numbers of people who must cope with disabilities, they cannot be assumed to exist in every person or every family. Interventions should be individualized to meet the needs, wants, and culture of the person and the family.

Social issues

Disabilities have many effects on the social lives of those afflicted. These include the attitudes of others, the effect of personal appearance and behavior, and leisure activities.

Attitudes of others

The attitudes of other people are the major problems faced by people who have disabilities. Even when non-disabled people have good intentions, there is often friction between them and the disabled people they meet. On the one hand, there is a lack of understanding on the part of the non-disabled and, on the other hand, there is limited patience on the part of the individuals with handicaps. Unhappy situations occur because the non-disabled people assume that the disabled individual needs help, and the natural scenario which would occur between equals is upset. People who are disabled may react with anger and frustration when they are patronized or when inappropriate assistance is forced upon them.

Makas (1988) studied "positive attitudes" by comparing what disabled and non-disabled people meant by the term. Factor analysis identified two attitude clusters among the non-disabled which were rejected by those with disabilities. He described them as follows:

> *... the "Give the Disabled Person a Break" cluster and the "Disabled Saint" cluster. Items that comprise the Give the Disabled Person a Break cluster involve special concessions made for disabled individuals. Disabled respondents rejected this notion. They indicated that they neither want nor expect special treatment because of their disabilities. Items in the Disabled Saint cluster represent positive characteristics frequently attributed to persons with disabilities. Disabled respondents .... rejected these attributes as defining persons with disabilities.*

In Makas's study (1988), the attitudes that people with disabilities regarded as positive differed from those regarded as positive by the non-disabled respondents. The attitudes appreciated by those with disabilities either did away with the special category of "disability" altogether, or supported their civil and social rights.

People without disabilities thought that positive attitudes were those which were "nice" and "helpful". They did not realize that this sort of response actually placed the recipient in a needy position. When asked to respond in an especially positive manner, these behaviors, which the people with disabilities found offensive, increased. Consequently, the more positive the non-disabled people were trying to be, the more likely they were to offend.

The attitude conflict described above can be resolved by educating the non-disabled public. Relationships between people with disabilities and those without can be improved through open communication and personal interaction. Another way to change public attitudes toward people with disabilities is through positive media presentation.

Media portrayal and public information

Traditional images of people with handicaps include such figures as the Hunchback of Notre Dame; Mr. Hyde; Captain Hook; and Lenny from Steinbeck's Mice and Men. Dangerous people are often portrayed as "scarred, maimed, ugly, deformed, physically and mentally handicapped, monstrous.". Children are exposed to negative images about people with disabilities from books, movies, plays, television, and comics. Such stereotypes influence the reactions of people toward individuals who are obviously different. We can see these stereotypes at work when people react with hostility to the idea of a group home in their neighborhood.

Movies often show "people with disabilities in a negative and unrealistic way, preferring the sensational or pitiful to the everyday and human side of disability.". In spite of this, there have been many positive movie representations of disabled individuals.
The Special Olympics creates an active and appealing image of people with disabilities. Public news reports of these events are carried by many forms of media: radio, newspaper, and television.

Personal appearance related to social attitudes

Personal appearance can influence the way individuals are treated socially and affect their professional and economic opportunities. For example, consider the importance of a good first impression at a job interview.

For people with physical disabilities personal appearance is a particularly sensitive issue. Physical differences may be seen as unattractive or even revolting. People's initial reactions tend to focus on an obvious disability rather than on the person. Kaiser, Freeman and Wingate (1985) questioned people with disabilities. Sixty percent of those questioned believed that other people noticed their disabilities first and their individual appearance second.

Many of the respondents agreed that this trend reversed if they were attractively and fashionably dressed.

Individuals who have physical handicaps may find it difficult to get attractive clothing that is suitable, comfortable, and easy to put on. Clothes specifically designed for people with

disabilities often look odd because of big zippers, or velcro closures in unusual places. Regular clothing is preferred because it reduces the wearer's visible difference and helps the person to fit in with the general population. Some individuals choose clothes which will conceal their disabilities. Others divert attention by the use of eye catching clothing such as T-shirts with controversial slogans. A few people with physical disabilities enjoy the extra attention they get and dress flamboyantly to encourage it (Kaiser et al, 1985/1990). Clothes give people with disabilities the choice of normalizing their appearance or emphasizing their uniqueness, according to their mood of the moment.

Many people with physical disabilities manage their personal appearance in ways that reduce the stigma associated with their differences. This increases their social acceptance by non-disabled people.

Behavior and social acceptance

Enthusiastic and outgoing behavior can overcome the stigma of disability and increase a person's social acceptability. Conversely, socially inappropriate behavior can cause people to ostracize anyone displaying it. Such behavior as repetitive aimless motions, drooling, and messy eating habits stigmatize a person who exhibits them. According to Knoblock, *"particular behaviors such as lack of eye contact or difficulty moving ... may make it difficult for some ... to attend to children or to sustain their own level of involvement".*

The education of individuals with impairments includes training to help them behave in ways which fit the general perception of normal. For example, individuals with visual impairments are taught to face someone who speaks to them. This approximation of the eye contact common in most conversations helps a sighted person to feel at ease in the situation.

People who are mentally retarded find the mastery of normal social behavior patterns difficult. They have trouble learning to interpret social interaction, and judge appropriate responses, because continual analysis of complex, constantly changing, situations is required.

Social behavior skills are the most involved and convoluted skills for anyone to master, yet they are needed in order for a person to experience social acceptance and participation in community life. For many people this participation includes pray activities, parties, involvement in sports, clubs, and organizations. These and other leisure activities are highly valued parts of a normal social life.

Leisure activities

Because of their low rate of employment, people with handicaps tend to have more leisure time than the general population. According to a 1985 study by Day, disabled people usually choose leisure activities which are inexpensive, passive, or solitary. Watching television, reading, walking, relaxing, and listening to records were the five most common activities. A large number of the people queried said that they would rather travel, work, or socialize, but that they could not because of lack of money, motivation, or health (Day, 1985/1990). People with disabilities have the same social needs as everyone else. These needs include friendship, love, and intimacy.

Practical considerations

As evidenced by the personal remarks quoted earlier, people with disabilities have the same concerns that plague everyone else in our society: finances, social acceptance, and finding a washroom in a time of need. Handicapped people, however, have personal and practical problems specific to their particular requirements and limitations. The major concerns of disabled people fit into the following categories:
1. affordability and availability of assistive devices.
2. employment and financial concerns.
3. accessibility problems.

1. Assistive devices

These services include "mobility aids, respiratory equipment, and environmental aids such as wheelchairs, commodes, walkers, and other self-help devices which assist in daily living in the home. People who are paralyzed due to certain diseases or injury may be eligible for medical supplies, medication, and assistive devices through the Paraplegia Program.

Despite government programs, some people still have problems obtaining the assistive devices they need. Equipment is available in major centers, and rural residents may find it difficult to go there to get the devices they need. Long waiting periods sometimes occur before equipment is available.

The affordability of essentials is an issue for many people with disabilities because, as a group, their incomes are lower than average.

2. Employment and finances

The annual incomes of people with disabilities are lower than those for the general population. The discrepancy in income is related to the high unemployment rate among people with disabilities. Jobs held by people with disabilities tend to fall into the unskilled and semi-skilled categories rather than the managerial or professional categories.

According to the 1988 study done by de Balcazar, Bradford and Fawcett, people with disabilities are concerned about the following aspects of their employment situation:

1. lack of reasonable accommodations in the workplace,
2. work disincentives within the social welfare system,
3. lack of job-seeking skills and lack of information about job training and job search assistance,
4. job discrimination and lack of equality of opportunity.
5. limited job opportunities.

Another factor that limits job opportunities for many people with disabilities is their level of education.

Education

Educational requirements for jobs are often a barrier to people with disabilities because they tend to have less formal education than their peers. Two main reasons for this discrepancy: that disabilities caused interruptions in people's education, and the need to attend special classes or special schools. Some people are so severely disabled that they cannot go to school.

Medical costs and concerns

There are, however, health-related costs which are not paid for by public funds. These include the cost of some drugs, costs of dietary supplements, and costs associated with travel to service facilities. Prompt access to services is a problem for some people. The availability of specialized services is a concern and can be an extra expense.

3. Problems associated with accessibility

Disabled people's problems do not end when they finally reach a building. Exterior doors may be difficult to open. Doors and hallways may not be wide enough for a wheelchair or walker to be maneuvered easily. Washrooms can also pose many problems for someone who is in a wheelchair: they may be too small; they may not have grab-bars; and sink, towel dispensers, and mirrors may be inconveniently placed. All of these factors make people's lives more difficult and limit the number of places they can go.

Housing options for people with handicaps are also limited due to lack of accessibility. With proper adaptations for daily living, many people with disabilities can live independently. Unfortunately, many either cannot find, or cannot afford, such housing.

Accessibility is a major issue for people with disabilities. It limits the places they can go, what they can do, the stores they can shop in, the jobs they can hold, and the places they can live. Lack of access makes a joke of the entire concept of equality. Access, or lack of it, determines whether a functional limitation becomes a disability.

Section 3

Disability Services and Available Interventions in Egypt

Services requirements

The quality of life for people with disabilities depends on the availability of services. These services can include residential, therapeutic, medical, vocational, educational and developmental provisions. Not all people with disabilities require the same services, or the same intensity of services. For example, two people may require custodial care, but one may need diapering, while the other does not. Service needs vary with the severity of the disability.

Services needed by disabled children and their families

Families with disabled children become involved with a wide variety of services in order to meet their child's needs. These needs change as the child grows. For example, the services needed to maximize an infant's development are quite different from the services required by an adolescent who will soon need a job and an independent living arrangement.

Needs associated with a child's health may include services such as specialized medical procedures, assistive devices, personal care, and drug therapy. Assessment and programming services such as physiotherapy or occupational therapy may also be needed. Provisions may have to be made for health-related services in the school when the child becomes a student.

Educational and developmental needs also change as the child matures. Infants and toddlers can receive developmental services. Individualized education programs must be provided for all children with special needs. The programs may provide communication services such as speech therapy, sign language, and Braille instruction; teach mobility instruction and personal orientation; and develop plans to address the child's academic, social, and functional skills. As the child reaches high school age, vocational training and independent living skills may take precedence over academic studies and the adolescent could spend a large portion of the day in the community rather than in the school.

The daily care of a child who is severely handicapped can be a taxing job. For example, a child who may have a seizure at any time of the day or night requires constant monitoring, and a child whose behavior is self-injurious requires 24 hour a day protection. Services are available to help a family cope with this task. Social Services may provide respite care, to give families a temporary break from this burden. There is also a temporary custody alternative available through which parents can place their child in a special foster home for a limited period of time. These services can be immensely important to allow a family to recuperate from the constant strain of caring for a family member with a disability.

All of the services described above are provided by government either through social services, public health, or the education system. The drain on the public purse is immense, but money and effort put into the developing child may reduce later requirements and costs.

Availability of services

Availability of services is affected by lack of funds, and the need to serve a small population distributed over a large area.

Services for the disabled are concentrated in larger centers, and may not be available in rural areas. Less esoteric services such as speech and language services, physiotherapy, occupational therapy, and psychological assessments should be widely available in smaller centers. This is not always the case. Shortages of such services impose an extra burden for families who must travel long distances. Some people may simply not be able to take advantage of these services.

Group homes and supported living programs are scarce in rural areas. Even in the cities there are long waiting periods before residential spaces become available. Services which fill the needs of people with disabilities are extremely important to their quality of life.

Cost of services

Medical services
Most medical services are paid for by government funds, and Egyptians often are not aware of these costs. According to Birenbaum and Cohen (1993), the cost of medical care for a youngster with severe disabilities is almost 10 times as much as that spent on a non-disabled child. Birenbaum and Cohen's estimate did not include the costs of therapy, visual or dental care, drugs, special clothing, dietary supplements, aids and equipment, personal care services, or costs of assessment at a facility.

Residential services
Though it is difficult to get itemized costs for medical services, educational programs, residential services and vocational assistance, one general tendency is apparent: "high-need clients cost more to serve than either moderate- or low-need clients" (Schalock & Keith, 1986). Consideration of the cost of services for any individual who is disabled can only be done accurately on a case-by-case basis because each person's needs are unique. However, to give an idea of the life-time cost for people with one type of affliction, Dr. Brian Habbick stated that it would require an estimated $1,000,000 to provide the special services needed by one child who has Fetal Alcohol Syndrome (FAS).

Categorization of services

Categorization of services catering for the disabled is a very compound problem. Disability by nature requires the inputs of different sectors. Health, educational and social services are the three major relevant sectors, in addition to other inputs such as labour, youth and sports…etc.
Finally, categorization according to the status of provider; i.e. governmental, non-governmental and private will be used as a frame of reference for comparison.

1. Governmental services

To date, governmental services constitute the largest and most widely distributed infrastructure of services. Major inputs are provided through three ministries; namely, the Ministry of Health and Population (MOH&P), Ministry of Education (MOE) and the Ministry of Social Affairs (MOSA). Other ministries such as Ministry of Labour (MOL) and Ministry of Youth (MOY) have some input even though to a lesser degree.

a. Governmental/state health services

Governmental or state health services are numerous and constitute the widest infrastructure of facilities within these sectors. Several major institutions belong to this category, such as the MOH&P, the Health Insurance Organization (HIO), the Curative Care Organization (CCO), University Hospital, Teaching hospitals and institutes (THI), in addition to a large number of hospitals and clinics belonging to other ministerial bodies such as Army hospitals, Police hospitals and other public sector companies.

The Ministry of Health and Population is the body entrusted with the task of the overall planning, organization and supervision of the health care system in Egypt, as well as ensuring the provision of health care services for all people. It is still the largest provider of health service facilities in Egypt particularly on the primary level and to a certain extent the secondary levels of health care. It is the biggest employer of physicians in Egypt around 50% of all registered physicians are employed by it although 80% of them work in the private sector.

However one of the most distinctive features of the MOH&P which is particularly relevant to childhood health care in general and childhood disability in particular is its possession of a huge network of facilities on the primary health care level. A network represented by more than 4000 Rural and Urban health units, Maternal and Child health units and centers in addition to more than 400 Health Offices.

This primary health care network is the most widely spread and relatively the most equitably geographically distributed infrastructure of services in Egypt. It covers all the population with an average of 5 kilometers distance.

Moreover, this primary network is linked to a higher chain of rural village and urban district hospitals, potentially forming a comprehensive vertical network of referrals within the health care system belonging to the MOH&P.

However, there is still a clear bias in favor of urban vs. rural as well as Delta (North Egypt) vs. Upper (South Egypt) as witnessed by the ratio of beds to population as well as the ratio of physicians to population. There are about 2.3 beds per 1000 population in urban areas as compared to 0.3 to 1000 population in rural and 1 doctor to about 11,000 population.

On the secondary and particularly specialized tertiary health care levels stands a large number hospitals which are mainly urban based provided by the University hospitals (MOE) and those belonging to the (THI).

The role of several departments within the university hospitals as well as that of some specialized institutes have increasingly provided important diagnostic, medical, therapeutic and surgical interventions in the area of childhood disability.

Another major actor amongst the state health services and an increasingly important player in the area of child health in general as well as childhood disability in particular has been the Health Insurance Organization possessing around 37 hospitals and 151 centers and clinics (CAPMAS 1997), and covering around all Governorates. Originally providing mainly curative services and catering mostly for the working/governmentally employed population, the HIO's umbrella is increasingly covering new sectors of society and particularly children. In 1995 all children enrolled in schools became covered with the HIO, currently around 16 million and since September 1997 all children born since are to be covered by the HIO (around 6 million new children). In all the HIO covers around 21 million children to date and takes on around 1.2 million born each new year.

The HIO has become one of the major health care providers in the area of child health. It has also taken on an important task related to the prevention of disease and disabilities amongst children through its responsibility for school health services and for children born after September 1997. Moreover, the HIO being the major body responsible for child health it has taken on the duty of becoming the major body responsible for providing technical aids for disabled children.

Summary

The MOH&P has the widest infrastructure of services as compared to other sectors. Its infrastructure is the most widely geographically distributed, particularly in the countryside.

Its structure potentially provides a favorable opportunity for addressing many aspects of the question of childhood disability in a more comprehensive and integrated manner, particularly its prevention, early detection as well as provision of medical and therapeutic interventions.
Moreover, it is the most widely utilized by families seeking help or cure for their disabled children.

Previously the ministry did not show particular interest in the area of childhood disability. However, during the past five years the ministry showed rising interest in addressing the question comprehensively and many actions to develop programmes and structures in the area were undertaken.

However, some important challenges continue to face its interventions among which stand the following:

- The curative orientation of the system.
- The lack of integration with other sectors.
- The lack of orientation and appropriate training of doctors and health staff to the nature and needs of the disabled.
- The high turn over of doctors working in the area of primary health care which does not allow for establishing a continuous system for prevention and early detection on the local and district levels.

b. Governmental educational services for the disabled

The MOE is the governmental body entrusted with the overall responsibility of supervising, providing and certifying training and education in Egypt.

Aside from the role played by the MOE in health through its control over university hospitals, the MOE provides the following inputs in the area of childhood disability:

- Direct input in providing special education for different groups of disabled children.
- Its role in the training and certification of special education teachers.
- Developing and designing special education curricula.
- A supervisory role which includes setting the standards and supervising curricula given by other bodies such as NGOs who provide special education.

Special Education and the MOE: The Ministry of Education is the biggest provider of educational services for several groups of disabled children.

Up till the 1952 revolution the Ministry provisions were limited to the blind and to a lesser extent the deaf. These activities were under the jurisdiction of the "Section for the Abnormal" within the ministry, which followed the 'Department of Primary Education'. In 1964 this section became an independent department and was renamed the "Special Education Department." In 1969 the presidential decree number 156 re-organised the state of education for the disabled children, creating three departments each caring for one type of disability. However the three were under the auspices of the "Special Education Department".

- **The department of education and training for the visually impaired.** Training is provided in special schools for two categories: the Blind, and the partially sighted.

- **The department of education for the hearing impaired:** This overviews the education and training of two categories of the hearing impaired: the deaf and the partially deaf, in two types of special schools. The deaf has both day care as well as residential facilities. It includes three stages of education namely: primary, preparatory, as well as vocational training for three years and technical secondary schools for three years. The partially deaf: follows the normal curricula of the ministry, while providing special facilities for the students.

- **The department of the educable mentally retarded children with IQ between (50-70):** This received children with mild mental retardation only, who are mobile and have no other psychological or physical handicaps. Education is conducted on two

stages (Primary for 8 years including two preparatory years and a vocational stage for three years.

• **The department of the physically and visceral handicaps:** This group includes the motor disabilities as well as the visceral such as Rheumatic heart children. The department overviews education of these children and their examination in hospitals or a sanatorium.

Table (1-3): Development in the number of schools, special education classes, staff working in special education including; teachers, psychologists, and social workers and numbers of students enrolled in Special Education Schools from 1991 to 2001

Year	Schools and classes		Number of Students			Number of Teachers	Number of Social workers	Number of Psychologists
	Schools with attached special classes	*Number of classes*	*Boys*	*Girls*	*Total*			
1991/1992	**136**	**827**	**9616**	**4919**	**14535**	**3169**	**81**	**106**
1995/1996	**202**	**2080**	**13609**	**7481**	**21090**	**4648**	**386**	**257**
2000/2001	**448**	**3076**	**19921**	**10849**	**30770**	**7303**	**565**	**328**

Training of the trainers or Personnel in special Education: One of the major problems facing efforts spent in the filed of special education is the shortage of specialized and well trained personnel. MOE data indicates that trained teachers in the field do not exceed 1/3 of the total number working. The rest 2/3 get their training on the job. The same applies for other personnel such as psychologists, social workers, vocational trainers as well as the administrative staff.

During the past five years an important step has been taken by the universities in the direction of developing the quality of special education for disabled children. A new undergraduate course was established first in Ain-Shams faculty of Education and followed by Helwan University. The new Cadre is a holder of a B.A. certificate.

However, despite the existing vast need, there is a problem in employment amongst the new Cadre. Once, again the question of the lack of coordination between efforts whether within between the different sectors or even within the same sector stand out.

Summary and conclusion

The MOE is the biggest provider of special education in Egypt. However, this is relative to other types of providers such as NGOs or the private sector rather than to coverage of the target population; i.e. disabled children at school age. As mentioned before, coverage of disabled at school age is estimated to be around 3%.

The reasons for this low coverage vary from lack of geographical accessibility - especially for the countryside – a low availability as shown by long waiting lists, for certain educational institutions, lack of knowledge especially for disadvantaged people, attitudes of parents of

disabled children, bad quality and bad reputation of these schools, in addition to the limitations in the types of disabilities served. Moreover, several groups of disabled such as the multiply handicapped as well as the moderate to severe mental retardation are not provided for by the Ministry of Education.

In addition, the state of special education seems to be going through a transitional stage. The old degree for primary school teachers has been stopped and new degrees for the preparation of the Cadres by the Ministry of Education has opened ranging from undergraduate training for four years up to post graduate diplomas

c. Sate and social services (Ministry of Social Affairs - MOSA)

The MOSA is the ministry entrusted with the overall responsibility for rehabilitation of children in Egypt. The ministry's input in the area of disability is carried out through the "Department of Social Rehabilitation" can be probably divided into the following categories:

- Development of general policies and strategies and the follow-up of their implementation
- The participation in the conduction of research in the area of disability
- The preparation of legislation to be presented to the parliament concerning the rights of disabled people
- Financial, technical and managerial support or assistance upon request to NGOs working in the field
- Supervision and legal control over all NGOs in Egypt
- Provision of services through its direct involvement, i.e. financing, managing and staffing as well as providing premises and equipment, and later on to incorporate these facilities under or into existing NGOs.
- Developing and collecting information on disability and the disabled people for planning purposes.

2. Non-governmental non-profit civil initiatives

By the year 2001, about 336 organizations were registered with the MOSA and worked in the disability field (Union of Organizations for the Care of Disabled, 2001).

However, the greatest majority of outlets from the different organizations provide services in the areas of registration, guidance of disabled and vocational rehabilitation. These activities are provided mainly through the Rehabilitation Bureau's. In reality the real function of these 'Bureau's' is the registration of the disabled and providing them with certificates entitling them to rights to special provisions, for medical and therapeutic rehabilitation, aids and appliances, vocational rehabilitation and jobs. Their direct role in providing rehabilitation services is minimal. These bureaus belong to the quasi-governmental group of organizations.

Types and numbers of services offered by NGOs (2001):

- Rehabilitation bureaus (144)
- Rehabilitation centers (37)
- Special factories for disabled (6)
- Factories for appliance (15)
- Intellectual institutions (18)
- Physiotherapy centers (17)
- Nurseries for disabled children (63)
- Vocational assessment centers (2)

Discussion and Conclusion

The role of the NGO sector in the field of disabilities is of great importance. Aside from the few educational facilities provided by the MOE for certain particular groups of disabled children, the bulk of other services, particularly in the areas of care, social support, training, vocational, advocacy and rights of disabled, provision of aids and appliances, certificates of disability...etc. are in the hands of the "NGO" sector.

Geographically, the majority of NGOs work in the metropolis and large cities, with a bias to the north of Egypt (the Delta) as compared to the south of Egypt. The rural areas hardly have any services at all.

The role of organizations working in the field of disability is directed mainly at service provision. It is mainly institutionally and professionally oriented.

However, during the past ten years, two important developments have taken place. The first is the emergence of organizations from parents of disabled children.

The second is the development of a number of initiatives that propagate a new approach to rehabilitation, namely the Community Based Approach (CBR).

3. Private services (profit-oriented services)

The input of the private sector in Egypt in the field of disability covers mainly two types of service provisions: the contribution of private health facilities and the contribution of educational ones.

a. The contribution of the private medical and health services

As in the governmental services, in the majority of cases, the contribution of the health and medical services has to be identified through the different medical outlets.
These outlets include private hospitals and polyclinics, accounting for 9% of total hospital beds in 1992 and private practitioners (60,000-80,000) running their private clinics (DDM, 1995).

The private hospitals range from large 'five star investment hospitals' to smaller and more moderate ones. With some exceptions that will be mentioned later, these hospitals do not

cater for disability per se but rather as part of management of disease through the different specialized departments, such as Neurology, ENT, and Pediatric departments...etc. In addition, some of these hospitals, have come to include other therapeutic interventions such as Physical therapy sessions which cater mainly for adult stroke patients and to a lesser extent children, while fewer have come to include speech therapy.

Alongside these hospitals, stands a huge army of polyclinics and individual practitioners. These mainly provide outpatient services. They provide the first point of contact for the majority of ill people in general and, in this case also families of disabled children. They range from General Practitioners (GP) to different specialists. Private General Practitioners are usually the first sought, while the most commonly utilized after the first point of contact is made, are the specialists, particularly university professors of such specializations as Neurology, Psychiatry and Pediatricians. These are followed by individual physical therapists and speech therapists.

In addition to the general private health care facilities, several more disability-oriented private centers and hospitals have opened, particularly during the past ten years - a fact which was stimulated by the increasing awareness to the problem and the gap in the quality of available services.

Among the first to function were hospitals for the mentally ill which opened units for mentally retarded children. In addition, a small number of specialized centers were opened during the past few years catering for both mentally retarded as well as physically disabled children. These centers were opened through the initiative of practitioners who gained experience in the governmental and non-governmental sector in the area of disability during that decade. Both the specialized hospitals and centers are geographically concentrated in the metropolis and mostly in Cairo.

b. The contribution of the private sector in education

The second main contribution of the private sector following health is in the field of education and training of disabled children.

This role is very new, and again has been stimulated by the growing need and awareness to the question and the demand for filling the gap in coverage and mainly in the quality of services provided by the governmental sector. However, this was in response mainly to the needs of rich Egyptian families or families from the rich Arab oil countries, who sought services for their children in the Arab world after experience in the Western countries.

Educational services provided by private institutions range from special nursery classes in ordinary schools as well as special classes in ordinary primary schools to special education schools. They all cater for intellectual disabilities (Mental Retardation) (Shukrallah, Personal interview, 1996).

The majority - about six schools - provide integrated classes on the level of kindergarten and primary education. Some have special units within their schools and some provide integrated classes.

In addition, three new special education schools were opened during the past few years.
In general, all these schools serve only mentally retarded children. Nearly all are English speaking or French speaking schools.

They are concentrated mainly in Cairo and to a lesser degree in Alexandria.
At the moment there are approximately four private schools that provide special education. All cater for intellectual disabilities (Shukrallah, Personal interview, 1996).

c. The contribution of the private sector in the field of "technical aids and prostheses"

Aside from the provision of eye-glasses which the private sector plays the most important role, the private sector has had an important contribution in the production of technical aids and prostheses for the physically disabled. In fact the role played by small manufacturers goes a long way back. These shops belong to old artisans who pass their professions over to their sons and are not produced in mass production. In many cases they are more appropriate to the community and cheaper than imported ones.

Recently, other medical companies and specialized import companies have opened to cater for importing Technical Aids for the disabled from different western countries. These are very few in number and also very recent in origin.

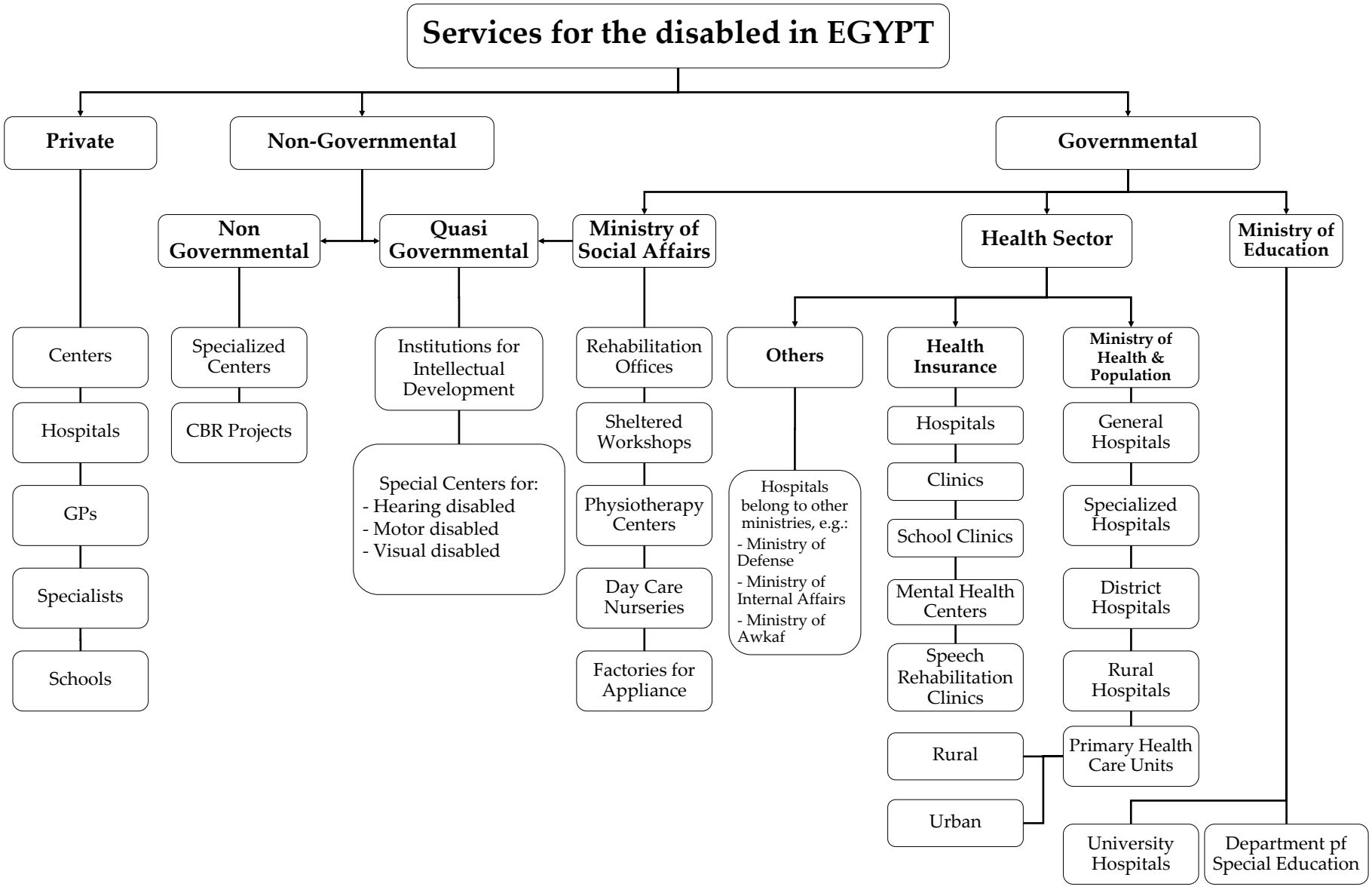


Figure (1-3): Figure 3: Services for the disabled in Egypt

Section 4

Community Based Rehabilitation

An integrated model to confront the disability question in Egypt from a developmental and rights approach

Definition of Community-Based Rehabilitation (CBR)

The definition below presents the essential elements of community-based rehabilitation (CBR):

- Community-based rehabilitation is a strategy within community development for the rehabilitation, equalization of opportunities and social integration of all people with disabilities.
- CBR is implemented through the combined efforts of disabled people themselves, their families and communities, and the appropriate health, education, vocational and social services.

Objectives of CBR

The major objective of CBR is to ensure that people with disabilities are able to maximize their physical and mental abilities, have access to regular services and opportunities and achieve full social integration within their communities and their societies.

This objective uses the broader concept of rehabilitation, that is, one which includes equalization of opportunities and community integration. As a broad concept, CBR is recognized as a comprehensive approach which encompasses disability prevention and rehabilitation in primary health care activities, integration of disabled children in ordinary schools, and provision of opportunities for gainful economic activities for disabled adults.

As a component of social policy, CBR promotes the right of people with disabilities to live within their communities; to enjoy health and well-being; and to fully participate in educational, social, cultural, religious, economic and political activities. CBR requires upon governments to transfer responsibility and necessary resources to communities so that they can provide the base for rehabilitation.

CBR is appropriate for both industrialized and developing countries. The broad methods used to implement it are applicable in either setting. However, the detailed methods of implementing CBR, and the resources available for it, will certainly vary among countries.

Methods for implementing CBR

CBR should be seen first and foremost as a programme which belongs to the community, as represented by the local government or authority. It should be considered an element of the social, educational and health policy at all levels, but particularly at the most decentralized level of the public sector. In national policy, CBR is part of a country's action in favour of

people of all ages who have any type of disability. In provincial and district level policy, it is supported with referral services and by transfer of knowledge to communities. At community level, the policy of integration is implemented under the control of the community, which "owns" the CBR programme and which gives a major role to people with disabilities and their families.

Hence, the broad methods for developing CBR include the formulation and implementation of policies to support CBR; the encouragement and support of communities to assume responsibility for the rehabilitation of their members who have disabilities; the strengthening of rehabilitation referral services for health, education and labor at district, provincial and national levels; and the establishment of a system for programme management and evaluation. The participation of disabled people in all of these activities is essential.

Community action for CBR is often initiated by a stimulus from outside the community, most likely from the ministry, committee or organization responsible for the programme. Following initial discussions with the representatives from outside the community, it is the community that decides whether CBR will become part of its ongoing community development activities. Various partners in the community, such as the community development committee, organizations of disabled people, and other nongovernmental organizations should be empowered to take responsibility for the programme. Once the community chooses to initiate a CBR programme, the CBR programme management should provide the necessary support and referral services, sufficient awareness, preparation and mobilization. It is important to underline the necessity of thorough preparation at all levels.

CBR may provide the means for all disabled people in a community to be socially integrated, or it may be more limited in its achievements. This depends on the commitment of the community, its resources, and the support services that it receives from both government and nongovernmental organizations. A municipal government in an industrialized nation has different resources than the local government of a rural town has in a developing country, although both may be committed to assisting their members with disabilities. Both will also be dependent - albeit to a varying degree - on some support from other levels. This consists of support to community members to enable them to actively participate in the CBR programme, as well as support directly to disabled people for treatment, education or equipment which they cannot obtain at the community level.

The definitions, objectives and implementation methods are taken from the "Joint Position Paper on CBR for and with People with Disabilities", (ILO/UNESCO/WHO, 1994).

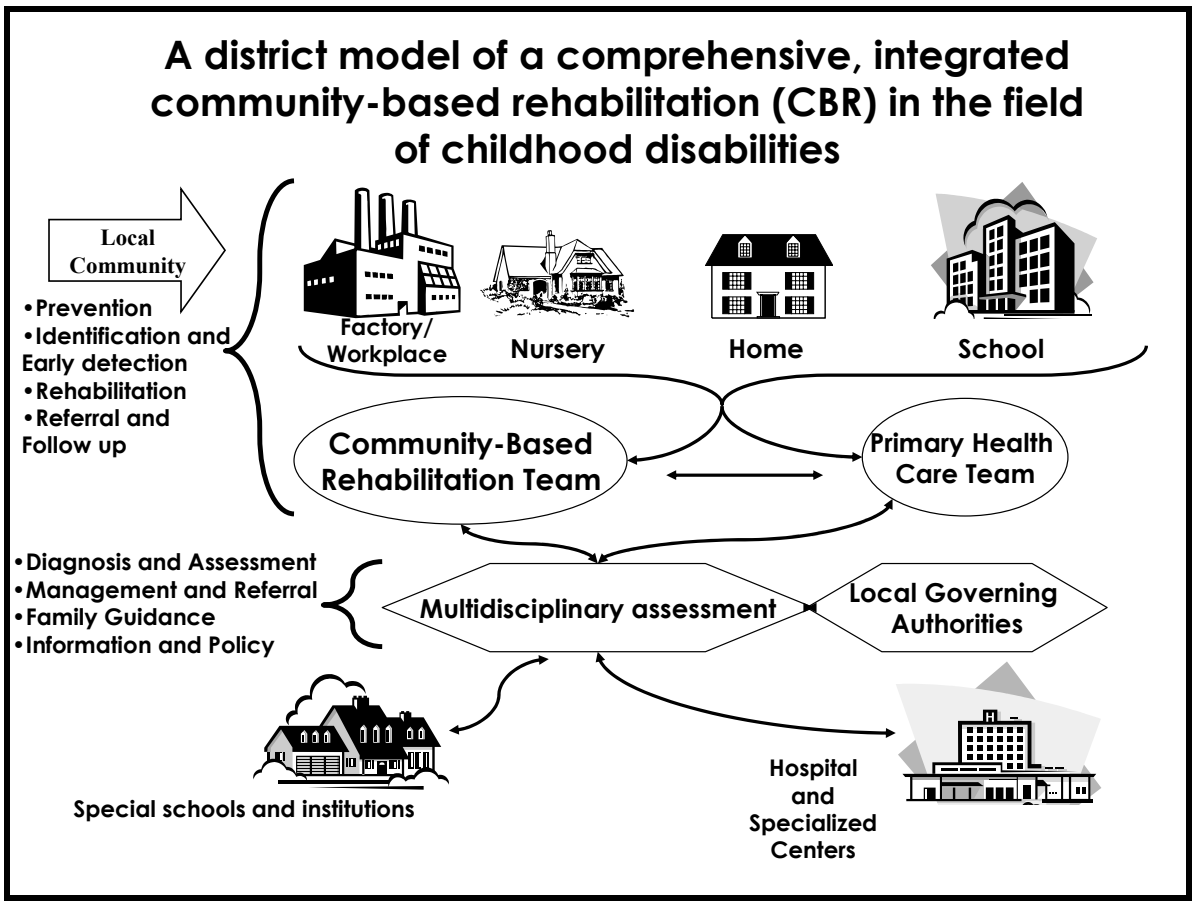


Figure (1-4): Community-based rehabilitation (CBR) model

Basic components of the CBR model

• **The community infra-structure of services, facilities and resources**, constitutes the base as well as the goal of the project. It includes normal main stream schools, nurseries, recreational and cultural facilities, transportation etc. and foremost the home of the child and his/her family.

• **The primary health care team as well as the community based rehabilitation workers**. These are the first line for professional and semi professional confrontation on the community level. These teams should work very closely with each other and with the communities they serve.

• **The inter-disciplinary assessment team/center.** A central link between the secondary and tertiary lines of confrontation and the primary and community level. The role of this team/center is to provide in-depth assessment of the needs of the child and its family, to devise a holistic inter-disciplinary plan, and to provide guidance to families as well as organize referrals.

• **The specialized centers and institutions** constitute the second and third lines for confrontation. They include services which provide backup support to the community level and the primary care level, such as hospitals, special schools, specialized centers,... etc.

Section 5

Childhood Disability as a Component of Child Rights and Child Protection

Historical evolution and growing international interest

In the area of childhood disability, the concept of rights surpasses the focus on the right to service provisions to embrace the right for inclusion in society.
This means that focus should not be on making the disabled fit within the norms of the society but on changing the norms of the society to include all the different groups within it, and among them the disabled.

In addition, placing the question of childhood disability on the sphere of rights emphasizes the indivisibility of these rights. We cannot separate the right to services such as education, health or social services from the right of access to the different facilities of society such as transport, recreational, cultural and religious ones, as well as the right to be protected from abuse and discrimination, whether due to disability, gender, race, religion and/or ethnicity…etc.

Finally, from within the conceptual framework of child rights, the concept of child protection emerged. The concept of protection embraces combating all causes which hinders the child's right for life, survival and development. In the area of childhood disability, this meant that all children have the right to be protected from the social and environmental risk factors which lead to disease and impairment in the first place as well as those which hinder the child from realizing his/her full potential development if and when the disability occurs.

The concept of rights hence, embraced, for the first time, the right of all children to be protected from all aspects of the disabling process.

The Convention on the Rights of the Child (CRC)

• The Convention on the Rights of the Child (CRC) is the first legally binding international instrument to incorporate the full range of:
  - Human rights
  - Civil and political rights as well as
  - Economic, social and cultural rights.

• Built on varied legal systems and cultural traditions, the Convention on the Rights of the Child is a universally agreed set of non-negotiable standards and obligations

• The Convention on the Rights of the Child was adopted and opened for signature, ratification and accession by General Assembly resolution 44/25 of 20 November 1989. It entered into force 2 September 1990, in accordance with article 49.

Who ratified it?

Convention on the Rights of the Child
• Ratified by 192 countries
• Only two countries have not ratified: *the United States and Somalia*, which have signaled their intention to ratify by formally signing the convention

The convention on the rights of the child: An overview

General principles	Clusters of rights
a. Non-discrimination (article 2); b. Best interests of the child (article. 3); c. The right to life, survival and development (article 6); d. Respect for the views of the child (article 12).	• Civil Rights • Survival and health • Development of Children's abilities (education, media, etc.) • Participation • Special Protection

Article related to rights of the disabled child in the CRC

Article 23	
1	States Parties recognize that a mentally or physically disabled child should enjoy a full and decent life, in conditions which ensure dignity, promote self-reliance and facilitate the child's active participation in the community
2	States Parties recognize the right of the disabled child to special care and shall encourage and ensure the extension …of assistance, …to the eligible child and those responsible for his or her care
3	Recognizing the special needs of a disabled child, assistance …shall be provided free of charge, whenever possible, taking into account the financial resources of the parents or others caring for the child and shall be designed to ensure that the disabled child has effective access to and receives education, training, health care services, rehabilitation services, preparation for employment and recreation opportunities in a manner conducive to the child's achieving the fullest possible social integration and individual development, including his or her cultural and spiritual development
4	States Parties shall promote, in the spirit of international cooperation, the exchange of appropriate information in the field of preventive health care and of medical, psychological and functional treatment of disabled children, including dissemination of and access to information concerning methods of rehabilitation, education and vocational services, with the aim of enabling States Parties to improve their capabilities and skills and to widen their experience in these areas. In this regard, particular account shall be taken of the needs of developing countries.

Disabled child rights in the Egyptian child law (law no. 12 for the year 1996)

In 1996 and within the context of the development of the past decade in the area of childhood in general, and in childhood disability in particular, the childhood law was passed. The law tackled the legal rights of disabled children in its 6th chapter "Care and Rehabilitation for the Disabled Child". The major items of the law came as follows:

The Egyptian Child Law Number 12, 1996	
Article 76	Disabled children have the right to enjoy special social, health and psychological care, which increases their ability to depend on themselves and to facilitate their integration and participation in their society.
Article 77	The disabled child has the right to rehabilitation, which includes all social, psychological, medical, educational and vocational services, required to help the disabled child and their families to overcome the consequences of their handicap. The state provides the rehabilitation services, the technical aids and appliances free of charge and according to the budget allocated for this purpose.
Article 78	The MOSA establishes the institutions and bodies required to provide rehabilitation services to children with disabilities. In addition, the MOSA can provide certificates giving the rights to other bodies to open such institutions according to the regulations identified by the internal charter. The MOE is entrusted with establishing schools and classes to educate the disabled children according to their abilities and potential, and the entry requirements, curricula, and examinations are to be decided by the internal charter.
Article 79	The bodies mentioned in the above two articles would provide, free of charge, a certificate to each disabled child who has been rehabilitated. The certificate would include the vocation that the child was trained for.
Article 80	The rehabilitation institutions should notify the Labour offices with the name of children who have been rehabilitated .... and the Labour offices have the duty to help those registered accordingly to acquire appropriate jobs to their ages, abilities and residence. In addition, the Labour Offices should inform the MOSA department with a monthly report on those children who have been allocated jobs, within their district.
Article 81	Both the Minister of Labour and the Minister of Social Affairs identify the suitable jobs in the governmental apparatus and public offices for the disabled children with certificates.
Article 82	Any employer using 50 workers or more, whether working in different sites or in one site, is to use disabled children who have been nominated by the Labour Office with a minimum of 2% of the 5 % allocated for the disabled population in the law 39 for the year 1975.

What is child protection?

Child protection embraces two basic principles namely,

- All measures to ensure that children enjoy their due rights.
- All measures to assist those at Risk of being denied more than others these rights. The concept of children at Risk includes those children:
  - At risk of being exposed to conditions which are hazardous to their survival and development and those
  - Already exposed to such conditions

Accordingly, it includes the preventive aspects of child protection, i.e. to identify those at risk of being exposed and provide them and their families with needed support as well as identify those exposed to risks in order to alleviate those risks and provide management and rehabilitation.

Concept of "Risk factors" from the point of view of "Child protection"

- **From the medical perspective:** It includes all factors whose increase places children at higher risk of being exposed to trauma and mental and physical and psychological illness

- **From the social perspective:** It includes all factors whose increase places children at higher risk of being exposed to delinquency and social marginalization

- **From the rights perspective:** It includes all factors whose increase places children at higher risk of being denied their child rights.

Who is at risk?

- Those exposed to conflict of war
- Those exposed to physical and sexual abuse
- Those exposed to economic exploitation (child labor)
- Those exposed to living in the streets
- Juvenile delinquents
- Children with special needs (Disabled) etc...

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Chapter 2

Causes, Risk Factors and Prevention of Disability

By the end of this chapter the PHC physician should be able to:

- Recognize and describe causes and risk factors of the major disabling conditions of children.
- Demonstrate the most common risk factors and causes of disabilities during the early childhood period.
- Describe the role of social and environmental barriers in the disability process.
- Recognize important preventive measures of important disabling conditions.
- Early recognition and intervention for prevention of disability and its complications.
- Recognize and describe the role of parents and care takers as primary actors and communities in the rehabilitation process.

Causes, Risk Factors and Prevention of Disability

Causes of disabilities

There are two main reasons that professionals strive to find the causes of disabilities: first, the identification of a specific cause can help in treating the condition, and second identification of the cause of a disability may help prevent the occurrence of such disabilities in future generations.

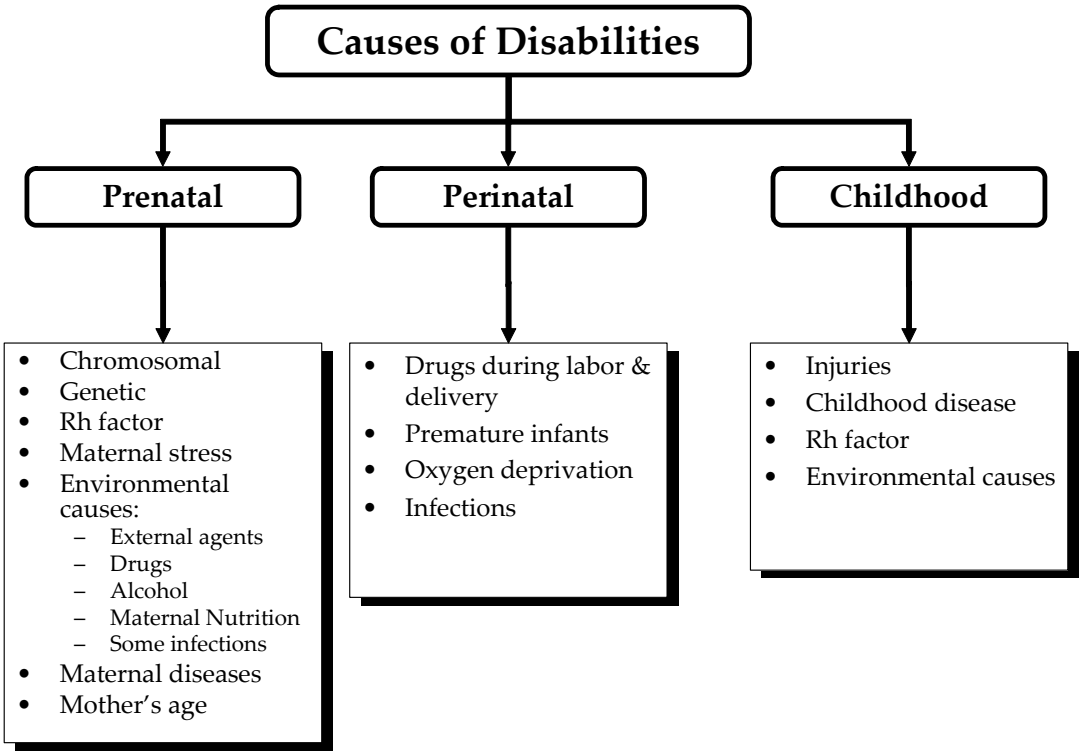


Figure (2-1): Causes of disability

There are thousands of known causes of disabilities, but in a great number of cases the exact cause of the impairment is never known. Several factors may combine to create a disability, but for the purpose of this discussion two major categories to classify the known causes of disabilities: biomedical or constitutional, and socio-cultural/environmental.

Biomedical or constitutional causes have a basis in the body of the individual; there is a biological aspect to the condition. Most severe and multiple handicaps include conditions which fall into this category. Congenital disabilities, those present at birth, are constitutional conditions.

Socio-cultural and environmental causes of disabilities are those which originate outside the individual's body. This includes not only those causes which stem from the social, cultural,

and physical environments, but also those causes which result from the individual's life-style and behavior. These factors are hard to isolate for two reasons. First, they are very complex, and second, they do not inevitably result in impairment. For example, risks among people of low socio-economic class can run through generations because the cycle of poverty creates conditions which contribute to the incidence of disabilities. In spite of these conditions, the majority of poor children do not develop disabilities. Since there is no direct cause and effect correlation between the associated socio-cultural and environmental conditions and disabilities, the conditions are generally called "risk factors". Those exposed to them are considered "at risk" for developing a disability.

Disability may be developmental or acquired and may arise from prenatal damage, perinatal factors, acquired neonatal factors and early childhood factors. These may include genetic factors, infections, traumatic or toxic exposure or nutritional factors, which result in perinatal or postnatal damage.

1. Prenatal causes of disabilities

The prenatal period extends from conception to the time of birth. Disabling conditions can occur at any point in the developmental process between those two events. Prenatal development can be divided into several periods. First, there is conception. Then, the time from conception to birth is usually divided into three phases. The first phase, the germinal stage, lasts from conception until implantation, when the developing organism becomes firmly attached to the wall of the uterus. This period is about 10 to 14 days long. The second phase, which extends from the second to the eighth week, is the embryonic stage. It is characterized by cell differentiation as the major organs begin to develop. The last phase, from 8 weeks until delivery, is the fetal stage. It is characterized mainly by growth. During this time the various body systems, which were led down in rudimentary form earlier, become quite well developed and begin to function.

Some prenatal biomedical causes of disability involve the basic building blocks of life: the genes and chromosomes which the person inherits. Other handicaps result from the prenatal environment within the womb. These causes can be considered separately, but it should be realized that heredity and prenatal environment work together to produce the infant.

Chromosomal causes of disability

Chromosomal abnormalities can involve the loss, gain, or exchange of genetic material from a chromosome pair. Such abnormalities often cause miscarriages, but may occasionally result in a baby with some kind of disability.

Down Syndrome, a congenital condition which usually includes health problems and mental retardation, is caused by an abnormality of the chromosomes. The twenty-first chromosome set is a triplet instead of a pair, hence the other name of this syndrome, Trisomy 21. Down Syndrome is associated with the mother's age. The incidence rate is high when mothers are extremely young, low for mothers in early adulthood, and increases with the mother's age after 35.

Genetic causes of disability

Chromosomes are made up of genes which, alone or in combination, govern all our inherited characteristics. Some disabilities are caused by specific genes that create damaging biomedical conditions. There are over 3,000 different genetic causes of disability.
There are definite patterns of inheritance which govern whether or not various traits affect us. An example of a genetic defect is Sickle Cell Disease, a blood disorder caused by recessive genes. Children only have the disorder if they receive the gene from both parents. If the gene is paired with a normal one, the individual does not have the condition, but can pass it on to his or her descendants. These individuals are called "carriers".

Rh factor

When an Rh-positive man and an Rh-negative woman have children together, there can sometimes be adverse consequences for their offspring. If their baby has Rh-positive blood, the mother's blood may begin to form antibodies against the "foreign" positive Rh factor. During the next pregnancy the antibodies in the mother's blood may attach the Rh-positive blood of the unborn infant. The resulting destruction may be limited, causing only mild anemia, or excessive, causing cerebral palsy, deafness, mental retardation, or even death. Fortunately, a way of preventing these consequences has been developed. The blood of the newborn infant is tested immediately after birth, using a blood sample from the umbilical cord. If an Rh-positive child has been born to an Rh-negative mother, the mother is given a vaccine that will seek out and destroy the baby's Rh-positive blood cells before the mother's body begins producing many antibodies. The red cells of later children will not be attached because the blood of the mother was never allowed to develop the antibodies.

Maternal stress

Even though there are no direct connections between the maternal and fetal nervous systems, the mother's emotional state can influence the fetus's reactions and development. This is true because emotions like rage, fear, and anxiety bring the mother's autonomic nervous system into action, liberating certain chemicals (e.g., acetylcholine and epinephrine) into the bloodstream. In addition, under certain conditions the endocrine glands, particularly the adrenals, secrete different kinds and amounts of hormones. As the composition of the blood changes, new substances are transmitted through the placenta, producing changes in the fetus's circulatory system. These changes may be irritating to the fetus. Bodily movements of the fetuses increase by several hundreds while their mothers are undergoing emotional stress. If the mother's emotional upset last several weeks, fetal activity continue at an exaggerated level throughout the entire period. When the upset is brief, heightened irritability usually last several hours. Prolonged emotional stress during pregnancy may have lasting consequences for the child. Infants born to upset, unhappy mothers are more likely to be premature or have low birth weights; to be hyperactive and irritable; and to manifest difficulties such as irregular eating, excessive bowel movements, gas pains, sleep disturbances, excessive crying, and excessive need to be held.

Prenatal environmental causes of disability

The prenatal environment is almost always a safe and nourishing one for a developing baby, but there are some environmental influences, which can damage a fetus. These influences include external agents, infections, toxins, and maternal health.

• External agents

External agents which can cause prenatal damage include injury and radiation. Seatbelts done up across the mother's abdomen can injure the baby in an accident. Any violent blow to the mother's abdomen can also hurt her child. Radiation such as X-rays can affect the fetus. This can result from treatment of pelvic cancer, from diagnostic testing, or from exposure to atomic energy sources, occupational hazards, or fallout. Although the hazards of radiation are not fully understood, it is clear that radiation can have a wide range of effects on unborn children, including death, malformation, brain damage, increased susceptibility to certain forms of cancer, shortened life span, and various mutations. Radiation that occurs between the time of fertilization and the time when the ovum becomes implanted in the uterus is thought to destroy the fertilized ovum in almost every case. The greatest danger of malformations comes between the second and sixth weeks after conception. Although the effects of X rays may be less dramatic later in pregnancy, there is still some risk of damage, particularly to the brain and other body systems.

• Many prescription and non-prescription drugs

These drugs can cross the placenta and adversely affect the developing child. Physicians and parents have become increasingly concerned about the potentially harmful effects of drugs on the developing embryo and fetus. One of the most dramatic reasons for this concern was the discovery around 1960 of the gross anatomical defects caused by a drug, thalidomide, that many women had taken during pregnancy. Thalidomide was introduced as a medication to control nausea in pregnant women, but it turned out to cause severe malformations in the legs and/or arms of the developing child. Many other drugs are suspected of producing birth defects when taken during pregnancy; substances that produce such effects are called teratogens. There is a long list of substances known or suspected to be teratogens, including legal drugs (alcohol, nicotine, caffeine), prescription drugs (some antibiotics, hormones, steroids, anticoagulants, anticonvulsants, tranquilizers, methadone), illegal drugs (cocaine, heroin, marijuana), and environmental pollutants (including lead, methylmercury, and polychlorinated biphenyls, or PCBs).

• Alcohol

Alcohol crosses the placenta easily and stays in the baby's system longer than it does in the mother's (FAS Symposium, 1993). Alcohol can create a spectrum of effects on the baby. These may include any or all of the following: various degrees of mental retardation, physical abnormalities, hyperactivity, autistic tendencies, and failure to thrive. There are many identified variables that influence the degree to which a mother's alcohol consumption affects the fetus. Some of these variables are: the amount of alcohol consumed, the stage of development of the fetus, whether the mother had been "binge drinking" or drinking on a regular basis, and the mother's general health, nutrition level, and metabolism.

• Maternal nutrition

Maternal nutrition affects the developing child. Deficiencies in iron, vitamins, and calorie intake can place the baby at risk. Some aspects of diet are especially important in the first few weeks of pregnancy, before many women know they are pregnant at all. For instance, not having adequate levels of folic acid is a risk factor for having a baby with spina bifida. Adequate levels are vital in the first weeks after conception and are difficult to get from diet alone. Babies born to mothers with nutritionally deficient diets are more likely to have low birth weights, to suffer from impaired brain development, to be less resistant to illnesses such as pneumonia, and bronchitis, and to have a higher risk of mortality in the first year of life. Severe maternal malnutrition may impair the child's intellectual development in addition to having adverse effects on physical development.

• Infections

Some infections that the mother suffers can damage the infant when the disease organisms cross the placental barrier. Viral diseases such as cytomegalovirus (which affects 5 to 6 percent of pregnant women), rubella (German measles), chicken pox, and hepatitis are particularly dangerous during the embryonic and early fetal periods. One of the most serious viral diseases during the first three months of pregnancy is rubella, which may produce heart malformations, deafness, blindness, or mental retardation. About 50 percent of babies whose mothers had German measles in the first month of pregnancy suffer birth defects; this figure falls to 22 percent in the second month, 6 percent in the third month, and only a small number thereafter. A pregnant woman can be tested to see whether she has already had rubella, but if she has not, she cannot be given vaccine for rubella because it contains the live virus. Thus, it is best for a woman who is considering pregnancy to ascertain whether she has had rubella before she becomes pregnant, and to receive vaccine at that time if she has not had it.

The rapid spread of the genital herpes virus among young adults poses another danger. Infection of the fetus with this virus usually occurs late in pregnancy, probably during delivery, and can result in severe neurological damage. When infection occurs several weeks prior to birth, a variety of congenital abnormalities can result. Prompt medical intervention is necessary if the presence of herpes during pregnancy is suspected.

Acquired Immune Deficiency Syndrome (AIDS) currently threatens the lives of a growing number of unborn and newborn babies. The percentage of women, who have developed AIDS or ARC (AIDS-related complex), principally through intravenous drug use or from a bisexual or drug-using partner, is rising rapidly. Mothers with AIDS can pass the virus to their babies, either across the placental barrier during pregnancy, during birth, or by breast feeding. Not all HIV-infected women infect their children, however. Estimates of transmission vary from 10 to 40 percent. Ongoing work is being done to determine if there are risk factors that influence the likelihood of transmission. It was found that mothers who transmitted HIV to their children had lower levels of vitamin A (low enough to constitute a deficiency) than mothers who did not. If this findings holds up, or other risk factors are identified, over which mothers have some control, it might be possible to reduce the number of babies born infected.

Infection of the fetus with syphilis is not infrequent. Fortunately, however, the placental barrier does not permit passage of the spirochetes that cause syphilis until after the fourth or fifth month of pregnancy. Consequently, transmission of the spirochetes (which otherwise

would take place in about 24 % of cases) may be prevented if treatment of a mother with syphilis begins early in pregnancy. When infection does occur, the spirochetes may produce miscarriage or a weak, deformed, or mentally deficient newborn. In some cases the child may not manifest symptoms of syphilis for several years.

• **Maternal diseases and disorders during pregnancy**

Some general disturbances of the mother during pregnancy may also affect the fetus. One of the most common of these is toxemia. Children whose mothers had severe toxemia during pregnancy run a risk of lowered intelligence. Illness of the mother, especially long-term illness, can also affect the child. Good prenatal medical care reduces these risks and increases the likelihood of having a healthy infant.

• **The age of the mother**

It is another factor associated with an increased risk of impairment. Teen-age mothers, especially those under 15 years of age, have a greater risk of having babies with low birth weight as well as neurological defects and childhood illnesses. Babies who are full term, but unusually small, are more likely to have a disability than are larger, more robust infants. Women with over 30 have a lower fertility rate than those in their twenties, and fertility continues to decline with age. They are also more likely than younger women to experience illnesses during pregnancy and to have longer and more difficult labor. Mothers over 40 run a sharply increased risk of having a child with a chromosomal abnormality, particularly Down syndrome. The average incidence of this disorder increases from less than 1 per 1000 through age 29 to 1.5 at ages 30 to 34, 6 at ages 35 to 39, 20 at ages 40 to 44, and 30 at ages over 45. Women over 35 are also more likely to have miscarriages and to give birth to underweight or stillborn babies. The older the woman, the greater the likelihood that these problems will arise; nevertheless, the absolute incidence of serious complications remains relatively small, especially for women who engage in good health practices and receive appropriate medical care. In cases in which there is reason to suspect the presence of a chromosomal or other abnormalities, procedures such as chronic villus sampling, ultrasound scans, and amniocentesis may be recommended.

• **Parental effects**

The risk of genetic disorders appears to increase with parental as well as maternal age. For instance, it has been found that older fathers have an increased risk of having children who suffer from genetic disorders caused by dominant genes, but where there is no history of the disease in the family. Such deviant genes may arise in the process of cell division leading to spermatogenesis. The overall genetic mutation rate in sperm cells may be six times higher than it is in eggs. As man's age increases, the number of times his sperm cells have gone through cell division also rises.

Fathers can cause problems in infants in non-genetic ways as well. They may transmit viruses to mothers along with semen, including sexually transmitted diseases such herpes, gonorrhea, syphilis, chlamydia, and AIDS. Furthermore, fathers' exposures to alcohol, nicotine, radiation, and pollutants such as lead have all been associated with risks to offspring.

2. Perinatal causes of disability

The perinatal period is the time immediately before and after birth. Disabilities originating from this time period are primarily biomedical ones. They may result from drugs taken during labor and delivery, prematurity, injury, oxygen deprivation, or infections acquired during the trip through the birth canal.

Drugs taken during labor and delivery

Because birth can be a painful process for women, various approaches have been devised to help deal with the pain. Perhaps the safest methods for both mothers and babies are methods for controlling pain through breathing, meditation, showers, appropriate movement, and support from a partner. Giving birth in this way is sometimes termed natural childbirth. However, other women feel they benefit from the use of medical intervention for pain control during delivery. Drugs such as pentobarbital or meperidine (Demerol) are one method of pain control. If taken just prior to delivery of a baby, however, they may make the infant less attentive, at least temporarily. Another medical method of pain control is anesthesia through epidural injection of painkillers into the mother's spinal cord. If administrated too close to delivery, however, such anesthesia can make it difficult for birthing mothers to push effectively in the second stage of labor.

One study of the effects of anesthetic drugs on sensorimotor functions in newborns found lags in muscular, visual, and neural functioning. While most such effects were greatest in the first few days of life, longer-term effects on cognitive functioning and gross motor abilities, particularly with heavy drug dosages, have been found at 1 year of age.

Premature infants

They are babies who were born too soon. Infants born earlier than the 38th week of gestation and weighing less than 2 Kg. Are referred to as premature. Prematurity is more frequent among economically disadvantaged mothers than among the affluent. In addition, smoking, alcohol,, and various drugs increase the likelihood that a baby will be born prematurely. Genetic factors play a role in determining birth weight.
The long-term effects of prematurity on development depend on how early the infant is born (gestational age), its birth weight, the type of postnatal care it receives, and the quality of its environment during early and middle childhood. Infants with gestation periods of less than 28 weeks (extreme prematurity) or weights of less than 1.5 Kg. Have a reduced chance of survival. In contrast, those who are only slightly premature (34 to 38 weeks) and whose weight is appropriate for their gestational age resemble full-term babies in many ways. They are generally healthy, though they are less mature, more vulnerable to illness, and slower to gain weight, and they must be monitored carefully. Recently, considerable progress has been made in caring for extremely premature infants and for "intermediate-term" infants, those that fall in the middle range of prematurity. These babies' gestational ages between 30 and 33 weeks, and their birth weights are at least average for their age, around 1500 grams or more at 30 weeks and 2000 grams or more at 33 weeks. Premature babies that have received intensive, highly specialized care have not only survived but gone on to develop normally.

A special problem for premature babies is respiratory distress syndrome, or breathing problems. This syndrome is typically caused by a lack of a substance called surfactant, a foamy lining the inside of the lungs that helps the lungs to expand and draw in air. Some progress is currently being reported in manufacturing substitute surfactant. Another difficult problem for premature babies is the occurrence of intracranial hemorrhage, which can cause permanent damage.

Premature infants need more than just medical care. Psychologists have worked on various kinds of programs to provide sensory and tactile stimulation and encourage parents to participate in the child's care while the child is hospitalized. Some investigators have thought that children would benefit from rocking (as on water beds) and gentile tactile stimulation (as from lying on sheep skins), interventions aimed at stimulating the conditions in the uterus. Others have provided visual, tactile, and auditory stimuli (such as mobiles, message, and music) that are thought to facilitate short-term benefits for premature children, including greater weight gain, less irritability and more quiet sleep, and better sensory responsiveness. The differences that can be attributed to the interventions typically decrease with age, as time since the interventions increases.

Like children who experience anoxia or complications during birth, premature children are particularly vulnerable to the effects of their environment. Premature children who are born into loving, nurturing homes where they receive competent physical and psychological care are usually show little long-range impairment unless they were very premature and did not receive appropriate neonatal or postnatal care. In homes with poor parental care and living conditions, premature babies are far more likely than full-term children to have both physical and psychological difficulties. Because prematurity and perinatal complications are more frequent among economically disadvantaged families than among middle-class families, poor children are more likely than richer ones to have to deal both with impairment at birth and a less favorable environment.

Oxygen deprivation

It may occur during a prolonged or difficult birth, and, because the brain suffers damage very quickly without a fresh and adequate supply of oxygen, brain damage can result. One major danger associated with birth is hemorrhaging, which is caused when very strong pressure on the head of the fetus breaks blood vessels in the brain. Another danger is failure of the infant to begin breathing soon after being separated from the maternal source of oxygen. Both hemorrhaging and failure to breathe affect the supply of oxygen to the nerve cells of the brain and produce a state called anoxia. The neurons of the central nervous system require oxygen; if they are deprived of it, some cells may die, and this can cause physical and psychological defects. If too many neurons die, the infant may suffer serious brain damage or, in extreme cases, may die.

Anoxia in a newborn is more likely to damage the cells of the brain stem than those of the cortex, and to result in motor defects. The child may experience paralysis of the legs or arms, a tremor of the face or fingers, or inability to use the vocal muscles. In this last case, the child may have difficulty learning to speak. The term cerebral palsy describes a variety of motor defects associated with damage to the brain cells, possibly as a result of lack of oxygen

during birth process. It is estimated that about 30% of cerebral palsy cases involve problems that occurred during birth or immediately afterward.

Anoxic infants are more irritable and show more muscular tension and rigidity than normal infants do during the first week. Infants with mild anoxia score lower on tests of motor development and attention during the first year and are more distractible. At age 3, they perform less well on tests of conceptualization. By age 7 or 8, behavioral differences between normal and mildly anoxic children are generally small, and their IQ scores are equal. In brief, the differences between mildly anoxic and normal children become smaller with age, and there is at present no firm evidence of serious and permanent intellectual damage.

Infections

Several sexually transmitted diseases can be contracted by a baby during the trip through the vagina. These infections include syphilis, AIDS, gonorrhea and herpes. Gonorrhea only affects the eyes of the infant, but herpes can result in severe disabilities due to nervous system damage. AIDS can also infect a baby through breast feeding.

3. Childhood causes of disability

Disabilities originating during childhood may be caused by biomedical and environmental factors.

Injury as a cause of disability

The types of injuries children are most likely to experience change with the age of the child. Tiny babies are rather floppy and slippery, and they have a tendency to put things in their mouths. These characteristics place them at risk for two types of accidents. First, they can fall when they slip out of restraints intended to care for and protect them, such as people's hands, cribs, high chairs, and playpens. The resultant injuries can produce motor or intellectual impairments as well as temporary damage such as bruises and broken limbs. Second, they can choke on small objects such as tiny toys, or pieces of toys. This type of accident can result in suffocation, anoxia, and brain damage.

Curiosity, lack of experience, and improved mobility combine to increase the toddler's chance of injury. Toddlers often fall when trying to get something they want, or see something that interests them. They may be injured when they investigate tools, pots on the stove, or electrical outlets. Cleaning products seem to hold a fascination for toddlers; according to the Saskatchewan Institute on the Prevention of Handicaps, thousands of poisonings involving household chemicals and children under four years old are reported each year (1991). Many of these accidents result in disabilities.

Spinal cord and brain injuries are of special concern due to the serious consequences of damage to the central nervous system. Such injuries often occur as a result of inadequate protection when children are riding in motor vehicles, or as passengers on bicycles.

Childhood disease as a cause of disability

Childhood diseases can retard a victim's future development. One of the most severe cases of disability is the result of meningitis suffered by the child in early infancy. It will develop sensory and motor limitations, and may have intellectual ones as well. It is impossible to assess the child's intelligence at present, because there is no appropriate method of evaluation for a youngster so severely handicapped.

Disabling conditions can sometimes result from common infectious diseases. Encephalitis, an inflammation of the brain which can cause mental retardation, is a possible complication of such childhood illnesses as mumps, chicken pox, and measles. Measles sometimes cause visual impairment. Ear infections which often accompany children's colds, can result in conductive hearing loss. Because of the tragic results of possible complications, even mild childhood diseases should be carefully monitored.

Environmental causes of disability during childhood

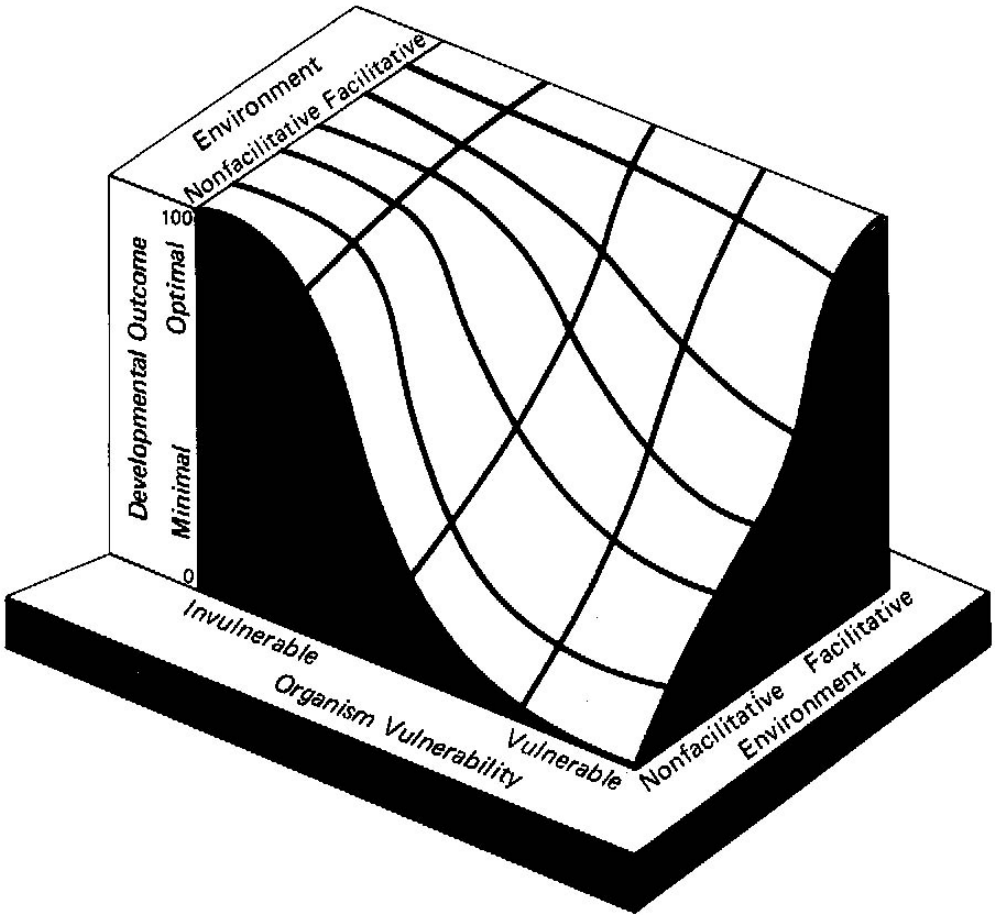


Figure (2-2): Horowitz's model of the interaction between the vulnerability of the child and the quality of the environment. The height of the surface is the "goodness" of the developmental outcome (like later IQ scores or language skills or social skills). The higher the surface, the better the outcome. As you can see, Horowitz proposes that a vulnerable infant in a non-facilitative environment will have by far the worst outcome – worse than the simple summing of the effects would predict. (Source: Adapted from Horowitz, 1982, Figure 2.1, p. 28.)

The child's constitution (genetic and biological attributes) and the environment are the two major influences on intellectual development. Most psychologists agree that both are important. Each contributes to a child's intellectual skills and academic accomplishments. A model for understanding one type of interaction between organism and environment is demonstrated by Horowitz (1987). In this model, organisms are placed on a continuum from constitutionally invulnerable to vulnerable and environment are classified on a continuum from facilitative to non-facilitative. Children with relatively invulnerable constitutions are expected to develop normally even in environments that are not highly stimulating or facilitating. Children in facilitative environments are expected to develop normally even when they have vulnerable constitutional attributes. The model is not intended to suggest that any child is completely invulnerable; a sufficiently bad environment can affect even a constitutionally strong child. Instead, it is intended to show that the greatest risk occurs for children who are biologically vulnerable and experience non-facilitative environments.

Though many children are resilient enough to grow up normally in spite of socio-environmental conditions such as neglect, poverty, famine, and even war, some do not. For some children environmental deprivation has a debilitating effect on the development of abilities such as language use, adaptive behavior, and cognition. Deprivation can include poor nutrition, poor housing, lack of social interaction and limited opportunity for varied experiences. These conditions are frequently associated with poverty, but can occur in any environment. They may sometimes be a symptom of child neglect or abuse.

Poor nutrition and starvation have been proven to have an effect on many areas of development. Hunger produces nervousness, irritability and a decreased ability to learn, thus it can have a negative effect on all aspects of a child's development. Negative effects increase with the degree of malnutrition. A severe vitamin A deficiency can cause blindness in children after they are weaned. A protein-calorie deficit during the first six months of life affects mental development and may damage the developing nervous system. Malnourishment is also associated with an increased susceptibility to infections.

Poor housing can be associated with a variety of disabling conditions. Chances of accidental injury increase when buildings are in poor condition. Cheap housing often consists of older buildings which may still have lead paint on walls and woodwork. Children often ingest this paint and get lead poisoning which affects the nervous system. Old buildings are often cold and drafty, and have substandard (or non-existent) plumbing and heating which increases the risk of disease and infection.

Limited social interaction and reduced opportunity for a variety of experiences are social-environmental factors which affect linguistic, emotional, and cognitive development. This type of environmental deprivation can occur in any home where language use is limited and life experiences lack variety.

Environmental deprivation can interfere with educational potential. Lack of exposure to reading and writing can adversely affect children's chances of academic success because this form of communication is unfamiliar. Reading and writing are less important when basic survival is in question; this can interfere with a child's motivation to learn academic skills.

Cultural differences can be risk factors for two reasons: discrimination and lack of understanding. Discrimination based on cultural differences may affect a family

economically and socially. Economic discrimination affects job opportunities, parental employment, and family income. It can result in hardship and poverty, which produce the risks described above. Social discrimination may add to the isolation of the family as a group and family members as individuals. Isolation can limit social interaction and experiences and have a harmful effect on the development of children in the culturally different family. Lack of understanding of cultural differences can result in a "mismatch" between what children experience at home and what is expected of them at school. This mismatch can lead to inappropriate schooling, which may result in intellectual impairments and functional limitations.

Deprivation during the developmental period is a contributing cause for many disabilities. Malnutrition, and increased risk of injury and infection, contribute to the number and severity of existing impairments. Limited opportunities for social interaction, lack of richly varied experiences, and low exposure to all forms of language, increase the risk for impairment of intellectual and linguistic development. Schools which do not consider the cultural and linguistic differences of students can deprive children of an adequate education.

Environmental risk factors, whether they are economic, social, cultural, or physical, intensify the effects of biological impairments and increase the likelihood of disability. Since deprivation is often associated with poverty, low socio-economic status itself becomes a risk factor.

Conclusion

There are many causes of disability, both biomedical and socio-environmental. Causal factors may operate independently, or combine to produce a disability. It is important to know the causes of a disability in order to prevent further occurrences. However, despite all that is known about the causes of various disabilities, there are many conditions for which no cause has been identified.

Prevention of disability

Every human society requires that its future generation be healthy. This depends on the birth and rearing of healthy children. To this end, preventive screening for genetic disorders, including developmental disabilities, is an essential component in uncovering possible disorders early, thus enabling timely medical intervention.

Such efforts are also required in order to reduce the expression and severity of disability. The ability of a physically or mentally disabled child to cope with and adapt to everyday life may be minimal compared to that of a normal child, and the disabled child may continuously suffer from trying to perform the functions so normal to others. This can have a major influence on the personality of the child which in turn can affect normal growth and development. Of particular distress is the fact that some disabled children never reach adulthood and some are at risk of developing other associated complications which may further disrupt their social and emotional development. These children and their families are under continuous mental and physical stress and require comprehensive services in order to help the children have a near normal life.

In addition, many disabled people require a continuous health care system, home help and other supportive services, which makes care programmes very costly. The management of childhood disabilities requires substantial medical, educational, social and rehabilitative care. The cost of preventive efforts is substantially lower and thus cost-effectiveness favors the prevention approach.

Stages of prevention

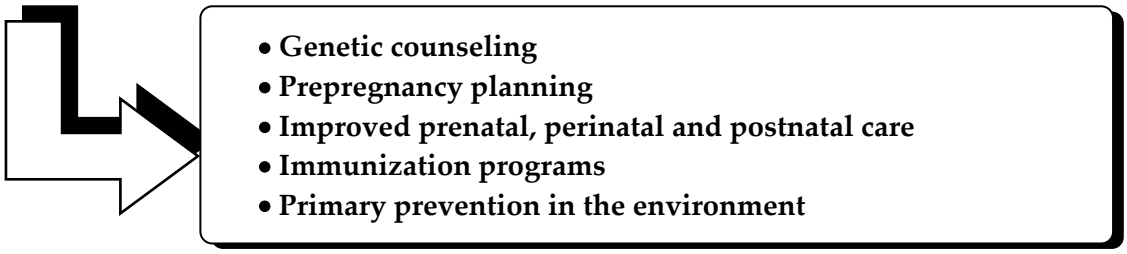
Endeavors for controlling disability can be categorized as primary, secondary and tertiary prevention.

1. Primary prevention

This involves the prevention of the manifestation of the disability. It may be universal (i.e. prevention desirable for everyone), or be restricted to a selected population (i.e. prevention recommended for high-risk groups) or to an indicated population (i.e. prevention in individuals with an identified risk).

Primary efforts are directed toward reducing the actual occurrence of disabilities and they employ measures that prevent the conception of a disabled individual or delay the disabling process.

Primary prevention efforts include:



Genetic counseling

Genetic counseling is an essential part of primary prevention strategies. It is the process of providing information on genetic (recurrence) risk, the nature and consequence of genetic disorders and the means available for the prevention of transmission of defective genes. Within this framework, there are three major aspects essential to effective counseling which are:

- Diagnostic aspects, where an accurate diagnosis is required for a secure foundation for advice
- Estimation of risk
- Preventive or ameliorative measures to ensure that those who are advised will benefit

One of the prime requirements of an effective genetic counseling program is to ascertain which individuals are at risk of having an affected child so that they can be offered advice. People who should consider genetic counseling include: those who are concerned that they might have, or "carry", an inherited disorder; couples who have a child with a disability that

may have been inherited; women who have had two or more miscarriages; and people who are concerned that their jobs or life-styles may pose a risk to pregnancy. Genetic screening and counseling prior to conception is important for the control of genetically determined disabilities. Many studies have shown that genetic counseling produces better understanding of the issues involved and hence has an impact on the subsequent reproductive decision. Several studies have shown that the number of children born with a serious genetic disorder leading to disability decreases significantly following genetic counseling.

Some inheritable genetic conditions can be identified through medical procedures such as blood analysis. Some "high risk" groups have been subjected to mass screening to try to reduce the incidence of genetic conditions common within those segments of the population.

"Pedigree analysis" is a form of genetic counseling which is done if there is a known abnormality in the family of either potential parent. This procedure takes the family histories, or pedigrees, of both partners, and analyzes them using the patterns of inheritance to determine the risk of this particular couple having a child with an inherited birth defect. Prenatal examinations check for another genetic cause of impairment, Rh blood factor incompatibility.

The best example of the successful application of this prevention strategy to prevent the birth of homozygotes for a certain disorder is the thalassaemia control program in Cyprus. Through health education, together with population screening and genetic counseling of carriers and prevention of carrier marriages, it was possible to reduce the homozygous affected births from 53 per 8594 births in 1974 to 0 per 10 752 in 1988 and 2 per 10 830 in 1990. The percentage of prevention achieved was 1.8% in 1974, 100% in 1988 and 97% in 1990. A similar approach when applied to other disorders could be equally successful.

Prepregnancy planning

Ideally, all babies are planned for and wanted by both parents; prepregnancy planning helps people achieve this goal.

People have many reasons for not wanting to have children at a particular time in their lives; impermanent relationships, educational or professional priorities, financial problems, or age and health concerns are only a few of the possibilities. Part of prepregnancy planning is to postpone pregnancy until both partners feel that they are ready to be parents. This requires the use of some form of birth control.

Once a couple decides to have a baby, they can increase their chances of having a healthy, normal child. Prepregnancy planning gives the parents-to-be time to establish good nutritional and exercise habits and to "kick" bad habits such as smoking or alcohol abuse. They should see a doctor at least three months before conception. At that time the doctor checks for medical conditions such as the: health problems in the mother, e.g., thyroid disease or diabetes; infections such as venereal disease; RH factor incompatibility; and the mother's immunization for rubella. All of these preconception efforts are primary prevention intended to reduce as much as possible the chances of having a child with an impairment.
Improved prenatal, perinatal and postnatal health care

This aspect of prevention concentrates on the management of maternal risk, factors at the time of delivery and support for the premature or compromised neonate.

Prenatal care helps insure that both mother and baby are as healthy as possible, and that any problems are addressed immediately. Prenatal clinics provide a combination of primary and secondary prevention. They not only try to prevent conditions that might put a fetus at risk, but also strive to reverse, reduce, or minimize risks that already exist.

Obstetricians attempt to ensure that all births are safe and that the infants are healthy. Medical procedures at the time of birth can prevent disabilities. Visual impairment is prevented by the use of medication to protect the eyes of the neonate from infections which may be picked up on the way through the birth canal. Careful monitoring of newborn babies and rapid treatment of anoxia can prevent mental disabilities.

After a baby is born, the public health nurses visit mother and child at home as soon as possible to give support and help. Later, at the Baby Clinics, nurses monitor babies' growth and screen them for sensory, physical, or developmental problems. Immunizations are started at the age of two months.

Immunization programs

Programs of general immunization during infancy have led to a remarkable decrease in, or in a few cases a complete absence of, several infectious diseases that used to be a major cause of disability. These include poliomyelitis, tuberculosis, meningitis and encephalitis.

Two good examples are rubella (German measles) and poliomyelitis. In the past, rubella epidemics occurred every three to ten years, leaving a legacy of babies with impairments such as blindness, hearing impairment, and mental retardation. Polio is leaving many people with physical impairments. The occurrence of such epidemics has been drastically reduced because immunization program reaches almost all of the population.

Primary prevention in the environment

The goal of primary prevention in the environment is to remove those causes of disability that reside in the social, cultural, and physical aspects of our lives. These risks include injuries associated with our daily activities, life-style and behavior patterns such as alcohol and drug abuse, lack of parenting skills, and the many characteristics of poverty which place people at risk. All of these socio-cultural/environmental areas should be addressed through primary prevention methods such as education, social and economic programs, and legislation.

2. Secondary prevention

Secondary prevention strategies aim at reducing the duration or severity of disability. These activities provide early identification of the disabling condition followed by prompt treatment and intervention to minimize the development of disability. These strategies can

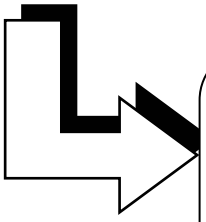
be applied either at the prenatal or neonatal level. Some of the conditions can be diagnosed during the prenatal and neonatal stages.

A history of certain risk factors indicates the need for careful evaluation. Prenatal insults such as maternal illness, infections, and teratogens (e.g., cigarettes, some anticonvulsants, and alcohol) increase the risk of DD in the child. Very low birth weight infants (under 1500 gm) have a 10% risk based on weight alone. This risk increases to 30-50% if there were associated problems such as hypoxia, infection, seizures, cerebral bleeding or being small for gestational age. Children in 1501-2500 gm birth weight group with a history of bronchopulmonary dysplasia, posthypoxic seizures, or meningitis have a 10% risk of a DD occurring. Larger babies who are appropriate weight for gestational age are at increased risk if there is a history of congenital malformation, birth trauma, perinatal hypoxia and seizures, meningitis, or adverse psychosocial factors. Low Apgar scores, especially at 10 or more minutes, may also suggest an increased risk for cerebral palsy.

Neonatal screening (organized examination of all neonates in order to diagnose specific disorders so that they can be treated) is a well established preventive approach and includes both clinical and biochemical screening. General developmental screening should include assessment of motor (gross and fine), social-personal, language, adaptive, and for older children, school-related abilities. Studies and experience have demonstrated that standardized, validated, and reliable screening systems are more successful in identifying developmental problems (especially mild ones) than the less accurate method of obtaining a developmental history from the parents and noting only major milestones on examination. In some countries, such information is available but in others, there is no information on detection frequency at birth of genetic disorders.

Follow-up of high-risk infants may use similar guidelines for developmental screening, with emphasis on testing at 6 and 12 months and 2 and 3 years of age. The preschool exam at 4-6 years should include vision and hearing screening. A few evaluation tools have been developed to measure school readiness, cognitive areas, speech and language skills, and emotional and behavioral problems that may interfere with school performance especially in children with very low birth weights.

Early intervention methods vary widely depending on the nature of the disability and its etiology but include the following:



- Genetic counseling to prevent further cases
- Specific treatment of underlying conditions, as in congenital hypothyroidism
- Treatment of specific contributory disabilities, e.g., hearing, vision
- Optimizing the functioning of the disabled individual and his or her family by:
  - Identifying and addressing the child's strengths and weaknesses, which will allow achievement of his or her full potential during the most crucial early years.
  - Preventing a single handicap from leading to secondary defects on other areas of development.
  - Preventing deterioration in development due to inappropriate physical and emotional handling by parents, peers, and society.
  - Improving interaction with siblings, between child and parents, and between parents, by lessening parental emotional reactions and promoting greater understanding the child's needs. This supportive relationship contributes to more successful developmental patterns later.
  - Helping families focus on the broad needs of the child.
  - Improving interaction and coordination among health and developmental providers.
  - Teaching parents how to utilize community resources more effectively and efficiently.
  - Providing direct supportive service to families in lower socioeconomic levels whose children are at increased risk for continuing developmental problems.

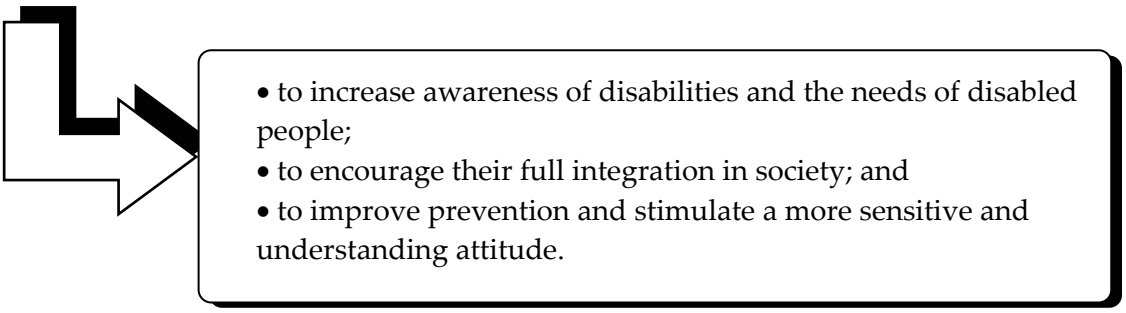
At the neonatal level, screening of neonates and proper intervention in those affected have been successful in reducing disability. The best known example of secondary intervention is that of neonatal screening for phenylketonuria (PKU), other aminoacidurias, hypothyroidism, the thalassaemias and other haemoglobinopathies. In PKU and other aminoacidurias, once the baby is diagnosed as having an abnormality, proper measures are taken by providing special diets.

Biochemical screening was first introduced for PKU in 1966 when it was shown that a low phenylalanine diet started in the first week of life prevents severe mental retardation. To screen for PKU, blood samples are usually taken by heel prick between 5 and 10 days after birth, when the body's metabolism has stabilized sufficiently for the results to be reliable. Screening for PKU is now established in several countries and screening for other abnormalities is also carried out, particularly since the advent of recombinant DNA technology. Hypothyroidism diagnosed during the neonatal period is treated by hormone replacement therapy, which encourages normal development and prevents complications such as mental retardation. In sickle-cell disease and thalassaemia patients, early detection enables better development and growth.

New forms of secondary prevention, such as genetic or surgical manipulation of an affected fetus to eradicate the biochemical or anatomical abnormality are being tried, some with a high degree of success. This is true for congenital heart disease, cleft lip and cleft palate, congenital dislocation of the hip and others.

3. Tertiary prevention

Tertiary prevention aims at limiting or reducing the effects of a disorder or disability that is already present. It involves long-term care and management of a chronic condition, e.g. rehabilitation or correction of the disability by surgical measures or by adopting strategies by which the disabled person can lead a normal or near normal life. **The main aims of rehabilitation of the disabled are**:



These measures also include special education programs. Only 50 years ago, the majority of disabled people were left illiterate. However, during the past three or four decades, considerable efforts have been made to develop special education programs to educate blind-deaf-mute, deaf-mute, blind and mentally retarded patients. Special schools with specially trained teachers have provided excellent education programs which have helped disabled people achieve goals that, in many ways, are similar to those of normal individuals.

Early recognition of disability

To apply any of the previously mentioned preventive measures successfully, the first step is an accurate and early recognition of the disability. Some impairment features are physical and obvious during clinical examination. These include skeletal abnormalities, blindness, hearing and speech disorders, some mental disorders and the chromosomal anomalies such as trisomy 18 or 21, Klinefelter syndrome and Turner syndrome. However, several other disorders do not become evident until later in life, although diagnosis may be made prior to the appearance of the disability or its complications. This has been possible using biochemical tests and, more recently, by applying recombinant DNA technology to the identification of the molecular basis of genetic disability.

The family in general and the mother in particular play a significant role in the early detection of disability. Abnormalities in development, both physical and others, and in learning ability may become obvious to the diligent eyes of the mother much earlier than a clinical diagnosis can be made. Early detection and early intervention can avoid the precipitation of several of the disabilities and can reduce the impact the disability may have on the family.

Once a diagnosis of genetic disorder is made in the carrier parents, proper counseling and premarital testing can prevent the pregnancy of a child with an abnormality. If conception has taken place, then prenatal diagnosis can be used; if the fetus is found to be abnormal, appropriate measures can be adopted and the parents can prepare themselves to look after a disabled child. If the child is diagnosed as having a disease that may lead to a disability, then proper intervention programs can be started at an early stage and can help ameliorate the effect of the disabling condition.

Medical and/or surgical approaches to preventive intervention provide whatever is necessary to overcome or correct disabilities and strengthen the family unit in order to enhance the abilities of disabled children and their families to cope.

Social and community efforts at tertiary prevention

Efforts to increase the accessibility of services in a community can be classified as tertiary prevention. As businesses and organizations provide better access, people with physical impairments are able to carry on their lives more normally. This reduces the effect of their limitations on their quality of life.

Social limitations caused by discrimination decrease as people become more accustomed to seeing and interacting with people with disabilities. Movies and television programs which emphasize the similarities between disabled and non-disabled individuals help encourage a positive attitude. School inclusion exposes students to individual differences and encourages social interaction between all types of people. Hopefully, the next generation of adults will accept people with disabilities as "just people" and grant them the equality they now lack.

Conclusion

Some disabilities can be prevented, others cannot. By the application of known techniques, a large number of disabilities can be prevented, or their severity reduced. Primary prevention is extremely effective because it targets the whole population, and, if it is successful, the disability addressed never occurs. Secondary and tertiary prevention efforts are also extremely valuable as they focus on specific groups with definite needs, and deal with their immediate situations. Some methods of disability prevention are controversial, but others involve the development of good health habits, good parenting skills, and adequate social supports.

Chapter Summary

Causes, Risk Factors and Prevention of Disability

Two major categories to classify the known causes of disabilities: biomedical or constitutional, and socio-cultural/environmental.

Prenatal causes of disabilities

Chromosomal abnormalities often cause miscarriages, but may occasionally result in a baby with some kind of disability; Down Syndrome. Some disabilities are caused by specific genes that create damaging biomedical conditions. The resulting destruction from Rh factor as a cause of disability may be limited, causing only mild anemia, or excessive, causing cerebral palsy, deafness, mental retardation, or even death. Moreover, the mother's emotional state can influence the fetus's reactions and development. The prenatal environment is almost always a safe and nourishing one for a developing baby, but there are some environmental influences, which can damage a fetus. These influences include external agents, infections, toxins, and maternal health.

Perinatal causes of disability

Drugs such as pentobarbital or meperidine (Demerol) are one method of pain control. If taken just prior to delivery of a baby, however, they may make the infant less attentive, at least temporarily. Infants born earlier than the 38th week of gestation and weighing less than 2 Kg. Are referred to as premature. The long-term effects of prematurity on development depend on how early the infant is born (gestational age), its birth weight, the type of postnatal care it receives, and the quality of its environment during early and middle childhood. Oxygen deprivation may occur during a prolonged or difficult birth, and, because the brain suffers damage very quickly without a fresh and adequate supply of oxygen, brain damage can result. Several sexually transmitted diseases can be contracted by a baby during the trip through the vagina.

Childhood causes of disability

The types of injuries children are most likely to experience change with the age of the child. Childhood diseases can retard a victim's future development; like Meningitis, Encephalitis, Mumps, Chicken pox, and Measles. Children are placed on a continuum from constitutionally invulnerable to vulnerable and environment are classified on a continuum from facilitative to non-facilitative; socio-environmental conditions, poor nutrition and starvation, poor housing, limited social interaction, lack of exposure to reading and writing, and cultural differences.

Prevention

Activities for controlling disability can be categorized as primary, secondary and tertiary prevention. Primary efforts are directed toward reducing the actual occurrence of disabilities and they employ measures that prevent the conception of a disabled individual or delay the disabling process. It includes genetic counseling, prepregnancy planning, improved prenatal, perinatal and postnatal care, immunization programs, and primary prevention in the environment. Secondary prevention strategies aim at reducing the duration or severity of disability. These activities provide early identification of the disabling condition followed by prompt treatment and intervention to minimize the development of disability. Tertiary prevention aims at limiting or reducing the effects of a disorder or disability that is already present. It involves long-term care and management of a chronic condition, e.g. rehabilitation or correction of the disability.

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Chapter 3

Early Detection & Screening

By the end of this chapter the PHC physician should be able to:

- Explain the concept of screening in general and developmental screening in particular, criteria for their application and their role in early detection of childhood disabilities.
- Recognize and describe normal milestones of development.
- Identify watch signs of developmental delays and abnormalities in child development.
- Apply simple procedures for testing certain aspects and functions of development and make early detection of disability conditions (neurological & motor, visual, hearing, speech and mental).

Section 1

Early Detection

What is early detection?

As mentioned in the first module, early detection was identified as a vital component of child health promotion. The situation analysis emphasized the fact that in Egypt the majority of children with impairments are discovered late when the condition has become less prone to respond to interventions and where complications have set in aggravating the original impairment and leading to the development of new ones thus hindering the child development and his/her relationship with family and society. Early detection belongs to the secondary and tertiary levels of prevention of impairments and disabilities in children. As part of a health promotion program, it can enlarge to embrace the early discovery of other conditions that threatens the healthy growth and development of children such as abuse, neglect and psychological disturbances.

Value of early detection

Concepts of critical and sensitive periods

The concepts of critical and sensitive periods could be applied both to the biological aspects of child development as well as the psychological and social aspects. For example, in the prenatal development, various body parts of the embryo develop at specific critical periods during which any external insult such as drugs, a disease of the mother is likely to produce a major structural deformity in that particular body part, e.g., the CNS and Heart from the third to the fifth month of the embryonic period.

Critical period is the period of development whereby exposure to insult or lack of proper timely intervention may cause permanent damage which may not be amenable to interventions at later stages. For example, inability to correct deafness at early stages of development leads to permanent dysfunction of speech even after correction of hearing at later stages.

Sensitive period has been described as the period during which a child may be particularly responsive to specific forms of experiences or particularly influenced by their absence. So, e.g., the period from 6 to 12 months of age may be a sensitive period for the formation of a core attachment to the parents. Other periods may be particularly significant for intellectual development or language. More generally still, some psychologists assume that the whole first five or six years of life form a kind of sensitive period; emotional patterns or skill levels established during these years are thought to be highly resistant to later changes.

Within the framework of the above two principles the value of early detection could be understood.

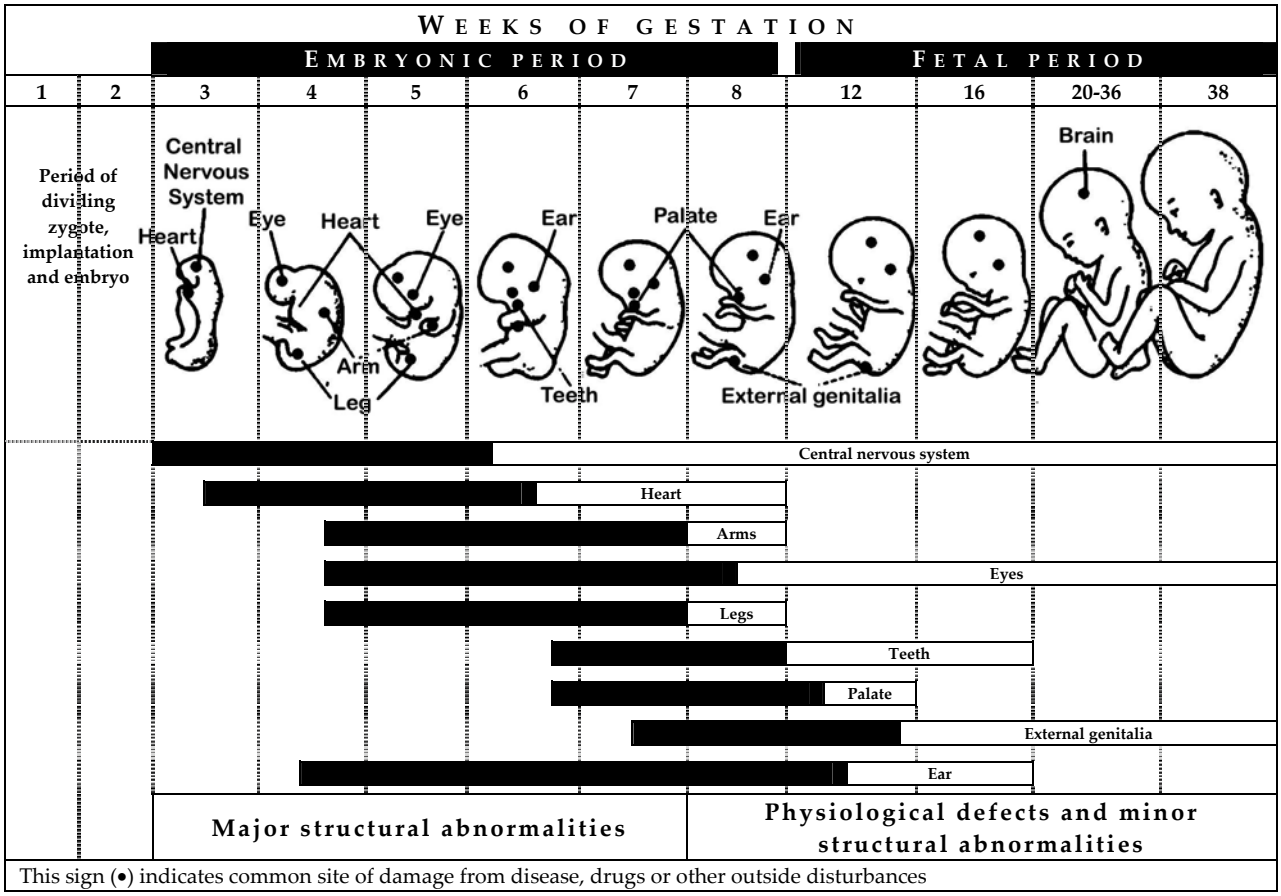


Figure (3-1): Critical periods in the prenatal development of various body parts. The darker portion of each line signifies the period during which any external insult – such as drugs, a disease in the other, or chemicals in the environment – is likely to produce a major structural deformity in that particular body part. The lighter part of the line shows the period in which more minor problems may result. You can see which body parts are most vulnerable to outside interference during a given week. The embryonic period is the time of greatest vulnerability for most body structures. (Source: Moore, 1982, Figure 8 – 14, p. 152).

Examples of the value of early detection

On the society level

In many incidences, efforts and expenses in addressing disabilities and their sequelae that discovered late markedly surpasses those of early interventions.

On the family level

Relationship of the family to the child and their acceptance of the child was shown to develop more appropriately when early detection is done and involvement of the parents in early intervention takes place. In addition, some families like to know if a problem exists early as to be better prepared to deal with it and plan ahead.

An early diagnosis may allow genetic counseling thus avoiding the birth of another handicapped child.

Some emphasized that the most important reason is that parents value early diagnosis. This is probably because it is easier to come to terms with a serious problem in a young baby than in an older child who has already acquired a personality and a shared life with the parents. It is not a kindness to keep parents in ignorance and leave them to find out for themselves that their child is abnormal.

On the child level

• *On the biological level*

Early treatment may reduce or even avoid permanent damage in some conditions such as congenital cataract, phenylketonuria, hypothyroidism, etc.

• *On the psychological and social level*

Lack of exposure to proper environmental stimuli or exposure to hazardous ones such as exposure to abuse or neglect may decrease the chances of the development of functions and behavioral maturity and the relationship to the family and society later in life.

Basic attachments between blind babies and their mothers

It was found that blind babies begin to smile at about the same age as sighted babies (about 4 weeks) but that they smile less often. And at about 2 months, when the sighted baby begins to smile regularly at the sight of the parent's face, the blind baby's smiles become less and less frequent. The blind infant's smile is also less intense, more fleeting.

The other thing blind babies don't do is enter into mutual gaze. They don't look right at their parent's, and everything we know about parent's responses to their babies underlines the importance of mutual gaze for the parent's feeling of attachment to the baby. When the blind baby does not look, the parents often report feeling "rejected".

Generally, the facial expressions of the blind infant are muted and sober. Many observers, including parents, conclude that the baby is depressed or indifferent.

It was found that most of the mothers of the blind babies gradually withdrew from their infants. They provided the needed physical care, but they stopped playing with their babies and gave up trying to elicit smiles or other social interactions. They often said they didn't "love" this baby.

Fortunately, it's possible to solve this particular problem. It was found that these mothers could be helped to form a strong bond with their infants if they could be shown how to "read" the babies' other signals. The blind child's face may be sober and relatively expressionless, but her/his hands and body move a lot and express a great deal. When the child stops moving when you come into the room, this means she/he is listening to you footsteps. Or she/he may move her/his hands when she/he hears your voice, rather than smiling as a sighted child would do.

When parents of blind children learn to respond to these alternative "attachment behaviors" in their babies, then the mutuality of the relationship can be reestablished. And when this happens, and the parents are able to provide more varied stimulation, blind children develop more normal behavior in other ways. In particular, they don't show the "blindisms" so often observed in blind youngsters, such as rocking, sucking, head banging, and other repetitive actions.

Section 2

Screening

What is screening?

In the area of early detection, screening is one of the major known methods. It refers to the examination of an apparently healthy population to distinguish those who probably have a condition from those who probably do not.

Other definitions describe it as,

- The process through which an apparently healthy population is investigated for an occult disease.
- The testing of asymptomatic individuals in the general population to identify those at high risk for selected disorders.

NB. Screening is not examining the child to see what is wrong with her/him, but the seeking of named conditions.

Criteria to be fulfilled by a screening procedure

(Wilson and Jungner's criteria for screening programmes)

- The condition to be sought should be an important public health problem as judged by the potential for health gain achievable by early diagnosis.

- There should be an accepted treatment or other beneficial intervention for patients with recognized or occult disease.

- Facilities for diagnosis and treatment should be available and shown to be working effectively for classic cases of the condition in question.

- There should be a latent or early symptomatic stage and the extent to which this can be recognized by parents and professionals should be known.

- There should be a suitable test or examination**:** it should be simple, valid for the condition in question, reasonably priced, repeatable in different trials or circumstances, sensitive and specific; the test should be acceptable to the majority of the population.

- The natural history of the condition and of conditions which may mimic it should be adequately understood.

• There should be an agreed definition of what is meant by a case of the target disorder, and also an agreement as to (i) which other conditions are likely to be detected by the screening programme and (ii) whether their detection will be an advantage or a disadvantage.

• Treatment at the early, latent, or presymptomatic phase should favourably influence prognosis, or improve outcome for the family as a whole.

• The cost of screening should be economically balanced in relation to expenditure on the care and treatment of persons with the disorder and to medical care as a whole.

• Case-finding may need be a continuing process and not a "once for all" project, but there should be explicit justification for repeated screening procedures or stages.

The characteristics of the ideal screening test *(Cochrane and Holland)*

• **Simple, quick and easy interpret:** capable of being performed by paramedics or other personnel.

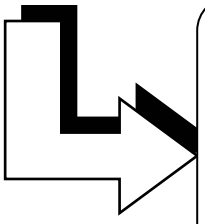
• **Acceptable to the public,** since participation in screening programmes is voluntary.

• **Accurate,** i.e. gives a true measurement of the attribute under investigation.

• **Sensitive:** this is the ability of a test to give a positive finding when the individual screened has the disease or abnormality under investigation.

• **Specific:** this is the ability of a test to give a negative finding when the individual screened does not have the disease or abnormality under investigation.

• **Repeatable:** this involves the components of observer variability, both within and between tests, subject variability and test variability.



Precision

This is the reliability or representability of the test. A test must give consistent results when performed more than once on the same individual. The main following factors should be considered.

• **Variation caused by the observer.** This may be due to differences between observers, or variation in the performance of the same observer, for example in recording positive and negative responses of infants to test sounds.

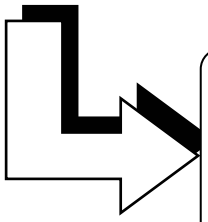
• **Variation within the subject.** The child's response to test sounds depends upon his level of interest and arousal. A hearing loss may fluctuate from day to day. Most physiological measurements – height, weight, blood sugar, blood pressure – vary in this way.

• **Variation in the method itself.** The equipment may be faulty or incorrectly calibrated. Test conditions such as levels of background noise during hearing tests may vary.

All these variations can be reduced or eliminated by standardization of equipment and procedures and intensive training and evaluation of observers.

Practical decisions to be made in implementing screening programme

In attempting to establish a successful programme of checks, a series of practical decisions need to be made.



Successful screening program should answer these questions:

- What is the population to be seen?
- Knowledge of the natural history of the disorder sought
- Who is responsible for carrying out the procedure on any individual?
- Where is the procedure going to be carried out?
- How do we contact the parents and ensure their cooperation by persuading them of the value of the service that we intend to provide?

1. What is the population to be seen? We need to be able to identify each individual to be seen and record the completion and result of the procedure. Some procedures may need to be applied to the total population, such as measurement of height and weight in order to identify growth disorders. Others may concentrate on the identification of high risk groups which will contain a much higher incidence of the problem being sought in the general population. Using this argument, selective screening of disadvantaged children will produce a high incidence of language problems, selective follow–up of certain neonates will yield a group of 6% which will contain 60% of all the sensorineural hearing losses.

2. Knowledge of the natural history of the disorder sought will determine the appropriate age for applying the screening procedure and the need for repetition. For example, screening for color blindness is undertaken on a once only basis whereas measurement of visual acuity needs to be repeated at intervals since changes occur with age.

A vital ethical consideration

The central ethical principal that should justify screening programs is no different from that which should guide all medical care: "Do no harm without compensating benefit and informed request"

3. Who is responsible for carrying out the procedure on any individual? Neglect of this essential aspect may cause a child not to be seen at all or result in a chaotic overlap and waste of effort between groups such as health visitors, school nurses, clinic doctors and family practitioners. Confusion can result from differing results or different decisions on future management.

4. Where is the procedure going to be carried out? The programme will fail if the center is inaccessible and distant from the patient's home or if appointments are offered at inconvenient times.

5. How do we contact the parents and ensure their cooperation by persuading them of the value of the service that we intend to provide? This raises issues of effective communication, appropriate health education and the delivery of services in such a way that

they are acceptable to the consumer. They need to be as convinced of the benefits as those providing the service are.

Information needed to assess a screening or a surveillance programme

- Clinical or laboratory expertise of those responsible for screening tests.
- Coverage achieved, i.e. what proportion of the total child population is examined.
- Number of children referred.
- Number of children referred who attend for specialist diagnosis.
- Number of children referred who are confirmed as true cases as a direct result of the screening test (yield).
- Number of false positives.
- Number of true cases missed.
- Number of true cases completing a course of treatment or other intervention.
- Number of cases effectively treated.
- The impact of the screening programme on other related services.
- The delays occurring between each step of the programme and the extent to which these cause or exacerbate parental anxiety.
- The quality, accuracy, and readability of the information provided for parents to explain the aims of the programme.
- The extent of the benefit accruing to those effectively treated.
- The cost of the programme, the cost per case detected, and the value of the benefits obtained.
- The training required to initiate the programme and maintain high standards.

Types of screening programmes for children

- **Periodic examination of all children**

  This entails all procedures and methods which examine children at their critical stages of development to identify predetermined conditions which interventions are necessary at these stages such as: laboratory investigations for conditions such as phenylketonuria, hypothyroidism, sickle cell anemia, etc and/or developmental screening tests.

- **Examination of only high risk groups**

  Limiting lab. investigations and follow up examinations and other investigations to high risk groups such as low birth weight babies, premature babies, incubated babies, etc.

- **Opportunistic screening**

  Screening children who come to clinic for other complaints.

- **In response to parents worries**

  Focusing on raising parent's awareness and knowledge of early signs of impairments and disabilities and limiting screening and examination to those children whose parents have doubts.

Section 3

Screening of Children's Growth & Development

Definition of developmental screening

It the process by which children are examined at serial key ages to detect conditions which are critically important at these developmental stages through recognizing decrease of functions, delayed development or deviation from normal development.

Definition of developmental assessment

The process through which children who have been screened positive i.e. suspected of developmental delay or behavior disorder, are assessed to establish whether there is a problem and if so the type and causes.

Its aim is to plan management, including treatment and genetic counseling, and to advise parents on how to cope and live with a disabled child

Some principles of development

- Development is a continuous process from conception to maturity.
- Development depends on the maturation and myelination of the nervous system.
- The sequence of development is the same for all children, but the rate varies from child to child.
- Certain primitive reflexes anticipate [develop before] corresponding voluntary movement and have to be lost before the voluntary movement develops.
- The direction of development is cephalocaudal [from head to toe].
- Generalized mass activity gives way to specific individual responses.

(Illingworth R.S., The Normal Child, p.149)

Development is a dynamic process in which both the child and its environment participate in which both select the influence of the other and through which both evolve in a particular direction.

The consequence or implication of this for assessment and intervention is that attention must not be focused exclusively on the child.

The interaction between the child and his parents or caregivers must constantly be born in mind, as mind factors of the family and the extended family.

Differences between growth and development

Growth: Increase in size
Progressions: Structural
Measured in terms of height, weight, head circumference, bone age etc.

Development: Increase in complexity
Progressions: Numerous, simultaneous and closely related but manifest many individual variations
It is a dynamic process in which the child and its environment participate

• **Normally,** growth and development of body, intellect and personality progress harmoniously and with relative predictability in rate and outcome. Development occurs in an integrated fashion.

• **Abnormally,** they are dissociated, producing widespread inconsistencies of and between somatic, cognitive and effective progressions, with unpredictable final results.

Biological characteristics of mankind and parallels of development for screening

Biological characteristics of mankind	Parallels of development for screening
• His upright posture, facilitating locomotion and enabling him to assume, adapt and maintain an enormous variety of effective attitudes while leaving his hands free for more precise activities	• Motor, involving body postures and large movements with combine high physical competence with economy of effort
• His finely adjustable visual equipment and uniquely flexible digits which bestow the ability to construct and use hand tools	• Vision and fine movements, involving visual competence in seeing and looking (far and near) and manipulative skills
• His possession of spoken language	• Hearing and speech, involving auditory competence in hearing and listening, and the use of speech and language codes
• His evolution of complex social cultures for the benefit of the groups and individuals comprising them, and for protection of the young during their relatively prolonged period of dependency	• Social behavior and spontaneous play, involving competence in organization of the self, i.e. self-care and progressively effectual self-occupation; together with increasing voluntary acceptance of satisfactory social standards with regard to personal relationships and cultural demands

Developmental disability

Developmental Disability means a significant deviation from the normally expected pattern of development, which is experienced by the person and/or by their environment as alarming. Such a disability may express itself in forms of retardation and/or dysfunction of a physical, cognitive and emotional-affective nature.

As a result of our starting from the transaction model of development, a developmental disability must also be regarded as a dynamic process in which a lot of factors can play a part. Among these are differences in norms between people, genders and cultures.
Retardation in development may thus occasionally be spontaneously removed or compensated for.

Developmental risk

Developmental risk implies that there is evidence of an increased probability that a disability may occur.
Three types of risk factors can be distinguished which are not mutually exclusive:

- Established risk in specific medically diagnosed conditions or syndromes, such as Duchenne's muscular dystrophy and Down's syndrome;
- Environmental risk, such as in children in deprived environments which limit their ability to profit from important learning experiences; and
- Biological risk on grounds of prenatal, perinatal and neonatal factors, such as in cases of prematurity and dysmaturity.

Developmental milestones

The categories for the milestones related to the "Denver Developmental Screening Test"

Emerging patterns of behavior during the first year of life	
Neonatal period (first 4 weeks)	
Prone:	Lies in flexed attitude; turns head from side to side; head sags on ventral suspension
Supine:	Generally flexed and a little stiff
Visual:	May fixate face or light in line of vision; "doll's-eye" movement of eyes on turning of the body
Reflex:	Moro response active; stepping and placing reflexes; grasp reflex active
Social:	Visual preference for human face
At 4 weeks	
Prone:	Legs more extended; holds chin up; turns head; head lifted momentarily to plane of body on ventral suspension
Supine:	Tonic neck posture predominates; supple and relaxed; head lags on pull to sitting position
Visual:	Watches person; follows moving object
Social:	Body movements in cadence with voice of other in social contact; beginning to smile
At 8 weeks	
Prone:	Raises head slightly farther; head sustained in plane of body on ventral suspension
Supine:	Tonic neck posture predominates; head lags on pull to sitting position
Visual:	Follows moving object 180 degrees
Social:	Smiles on social contact; listens to voice and coos
At 12 weeks	
Prone:	Lifts head and chest, arms extended; head above plane of body on ventral suspension
Supine:	Tonic neck posture predominates; reaches toward and misses objects; waves at toy
Sitting:	Head lag partially compensated on pull to sitting position; early head control with bobbing motion; back rounded
Reflex:	Typical Moro response has not persisted; makes defense movements or selective withdrawal reactions
Social:	Sustained social contact; listens to music; says "aah, ngah"

Emerging patterns of behavior during the first year of life	
At 16 weeks	
Prone:	Lifts head and chest, head in approximately vertical axis; legs extended
Supine:	Symmetrical posture predominates, hands in midline; reaches and grasps objects and brings them to mouth
Sitting:	No head lag on pull to sitting position; head steady, held forward; enjoys sitting with full truncal support
Standing:	When held erect, pushes wit feet
Adaptive:	Sees pellet, but makes no move to it
Social:	Laughs out loud; may show displeasure if social contact is broken; excited at sight of food
At 28 weeks	
Prone:	Rolls over; may pivot
Supine:	Lifts head; rolls over; squirming movements
Sitting:	Sits briefly, with support of pelvis; leans forward on hands; back rounded
Standing:	May support most of weight; bounces actively
Adaptive:	Reaches out for and grasps large object; transfers objects from hand to hand; grasp uses radial palm; rakes at pellet
Language:	Polysyllabic vowel sounds formed
Social:	Prefers mother; babbles; enjoys mirror; responds to changes in emotional content of social contact
At 40 weeks	
Sitting:	Sits up alone and indefinitely without support, back straight
Standing:	Pulls to standing position
Motor:	Creeps or crawls
Adaptive:	Grasps objects with thumb and forefinger; pokes at things with forefinger; picks up pellet with assisted pincer movement; uncovers hidden toy; attempts to retrieve dropped object; releases object grasped by other person
Language:	Repetitive consonant sounds (mama, dada)
Social:	Responds to sound of name; plays peek-a-boo or pat-a-cake; waves bye-bye
At 52 weeks (1 year)	
Motor:	Walks with one hand held; "cruises" or walks holding on to furniture
Adaptive:	Picks up pellet with unassisted pincer movement of forefinger and thumb; releases object to other person on request or gesture
Language:	A few words besides mama, dada
Social:	Plays simple ball game; makes postural adjustment to dressing

Emerging patterns of behavior from 1 to 5 years of age	
15 months	
Motor:	Walks alone; crawls up stairs
Adaptive:	Makes tower of 2 cubes; makes a line with crayon; inserts pellet in bottle
Language:	Jargon; follows simple commands; may name a familiar object (ball)
Social:	Indicates some desires or needs by pointing
18 months	
Motor:	Runs stiffly; sits on small chair; walks up stairs with one hand held; explores drawers and waste baskets
Adaptive:	Piles 3 cubes; imitates scribbling; imitates vertical stroke; dumps pellet from bottle
Language:	10 words (average); names pictures
Social:	Feeds self; seeks help when in trouble; may complain when wet or soiled
24 months	
Motor:	Runs well; walks up and down stairs, one step at a time; opens doors; climbs on furniture
Adaptive:	Tower of 6 cubes; circular scribbling; imitates horizontal stroke; folds paper once imitatively
Language:	Puts 3 words together (subject, verb, object)
Social:	Handles spoon well; often tells immediate experiences; helps to undress; listens to stories with pictures
30 months	
Motor:	Jumps
Adaptive:	Tower of 8 cubes; makes vertical and horizontal strokes, but generally will not join them to make a cross; imitates circular stroke, forming closed figure
Language:	Refers to self by pronoun "I"; knows full name
Social:	Helps put things away
36 months	
Motor:	Goes up stairs alternating feet; rides tricycle; stands momentarily on one foot
Adaptive:	Tower of 9 cubes; imitates construction of "bridge" of 3 cubes; copies a circle; imitates a cross
Language:	Knows age and sex; counts 3 objects correctly; repeats 3 numbers or a sentence of 6 syllables
Social:	Plays simple games (in "parallel" with other children); helps in dressing (unbuttons clothing and puts on shoes); washes hands

48 months	
Motor:	Hops on one foot; throws ball overhand; uses scissors to cut out pictures; climbs well
Adaptive:	Copies bridge from model; imitates construction of "gate" of 5 cubes; copies cross and square; draws a man with 2 to 4 parts besides head; names longer of 2 lines
Language:	Counts 4 pennies accurately; tells a story
Social:	Plays with several children with beginning of social interaction and role-playing; goes to toilet alone
60 months	
Motor:	Skips
Adaptive:	Draws triangle from copy; names heavier of 2 weights
Language:	Names 4 colors; repeats sentence of 10 syllables; counts 10 pennies correctly
Social:	Dresses and undresses; asks questions about meaning of words; domestic role-playing

After 5 years the Stanford-Binet, Wechsler-Bellevue and other scales offer the most precise estimates of developmental level. In order to have their greatest value, they should be administered only by an experienced and qualified person.

Section 4

Examples of Screening Programmes & Procedures

1. Examples of screening programmes

Antenatal screening – examples

Condition sought	Test to be applied	Agreed treatment	Justification for screening
Spina bifida Anencephaly	Serum alpha–fetoprotein	Termination of pregnancy if confirmed by raised amniotic fluid level	Prevention of severe handicap
Down's syndrome	Chromosomal analysis of amniotic fluid	Termination of pregnancy if agreed after counseling	Prevention of severe mental handicap
Duchenne muscular dystrophy	Chromosomal analysis of amniotic fluid	Counseling and termination offered	50% chance of an affected male infant

Neonatal Screening – examples

Condition sought	Test to be applied	Agreed treatment	Justification for screening
Phenylketonuria	Guthrie test – heel prick blood	Low phenylalanine diet	Prevention of mental handicap
Hypothyroidism	Blood T4/TSH – heel prick blood	Thyroxine	Prevention of mental handicap
Congenital dislocation of the hip	Barlow's and Ortolani's test. Hip must abduct fully and is non – dislocatable	Splinting and observation	Poor results and need for surgery in late diagnosed cases
Undescended testes	Clinical examination	Orchidopexy	Risk of infertility, malignancy, psychological problems
Hypospadias	Clinical examination	Surgery	Psychological effects of untreated condition
Cleft palate	Clinical examination	Referred to plastic surgeon	Development of normal speech
Talipes	Clinical examination	Orthopaedic referral	Prevention of handicap
Hydrocephalus	Measurement of head circumference	Insertion of ventriculo – atrial shunt	Prevention of handicap
Congenital heart disease	Clinical examination	Pediatric or pediatric cardiology clinic	Correction of defect. Prevention of SBE

Pre-school screening – examples

Condition sought	Test to be applied	Agreed treatment	Justification for screening
Squint	Cover test	Patching, orthoptic exercises or surgery	Prevention of amblyopia
Visual handicap	Detection of abnormal visual behaviour in infant - standardized test usually matching letters (Stycar) from age 2 ½ - 3 years	Correction of refractive error - educational and developmental guidance	Early intervention to reduce effect of possible handicap
Deafness	Distraction test Cooperation test Performance test or speech discrimination test Sweep test Tympanometry	To audiological center – depends on severity and cause	Minimize handicap and promote normal language development
Growth disorders	Accurate measurement of length	Referral to a specialist growth clinic	Discovery of conditions where treatment is available, e.g. Growth hormone deficiency, Coeliac disease, Hypothyroidism
Child abuse or potentially abuse situations	History (PMH, FH or SH). Clinical examination	Agreed procedures generally laid down at local level	Prevention of injury. Help for parents and child
Dental caries	Clinical examination	Dental treatment and health education	Prevention of pain and need for dental extraction

School age screening – examples

Condition sought	Test to be applied	Agreed treatment	Justification for screening
Scoliosis (11 – 14 years)	Clinical examination	Orthopaedic clinic	Prevention of severe deformity
Colour blindness	Ishihara colour test	Discussion of possible career limitations. No treatment	Minimal
Infestation	Clinical examination by school nurse	School treatment center or GP	Prevent spread
Depression	Interview. The quiet, isolated, withdrawn child is frequently missed	Discussion and referral	? Prevention of adult psychopathology

Examples of a schedule for periodic screening of children in United Kingdom (1990)

Age (approx.)	Aims and comments
At birth	Immediate evaluation to assess need for resuscitation and special care, and identify obvious disorders.
Between 6 – 10 days	Full post–natal examination: start of ongoing child health record (including relevant pre – and perinatal data). Arrangements for achieving this examination in association with policies of early discharge from hospital need to take into account the time factor and the later onset of cardiac and other important physical signs. *These examinations should ensure prompt recognition and explanation of apparent defects, and reassurance to mother of a normal infant. Mothers require positive advice, and mere absence of adverse comment is always insufficient.*
6 weeks	Introduction to the 'clinic' premises, facilities and staff: opportunity for professionals to listen and advise on infant management and family problems and to give sympathetic support (and, where necessary, to initiate treatment or referral) to mothers experiencing fatigue, isolation and possibly depression. *Unless for any reason the neonatal examination was nor carried out, there will be a relatively low emphasis at this stage upon detection of handicaps.*
7 – 8 months	Review of development, especially hearing and vision. The timing of this contact is important; 6 months is not a rewarding age to check motor development, while 9 – 10 months is often too late for the first routine hearing test.
18 months	Review of development, e.g. mobility, manipulative skills, hearing and early language, social relationships: opportunity to discuss the range of normal in growth and behavior.
2 ½ - 3 years	Review of development: opportunity to discuss behavior: language: appropriate vision testing and cover test for squint.
4 ½ - 5 years	Summing up of early health and development in relation to entry to school: early warning to teachers of potential or established difficulties (e.g. speech, behavior) which could affect child's response to school: appropriate testing of vision (including cover test for squint) and hearing. Exchange of information with teachers, school nurse and parents. *This examination, which should apply to all children should form the basis of a subsequent selective (i.e. non–routine) approach to ongoing oversight of the health and behavior of children in school.*
During school years	
Annually up to the age of 13 – 14 years	Health care interview with a school nurse, including a vision test and measurement of height and weight; dental examination by a dentist.
Twice during primary education	Hearing test by school nurse.
At approximately 13 years	Interview with the school doctor.

There are two crucial periods in relation to education when every child would be seen by a doctor; on entering school and at about the period of puberty.

2. Denver Developmental Screening Test (DDST)

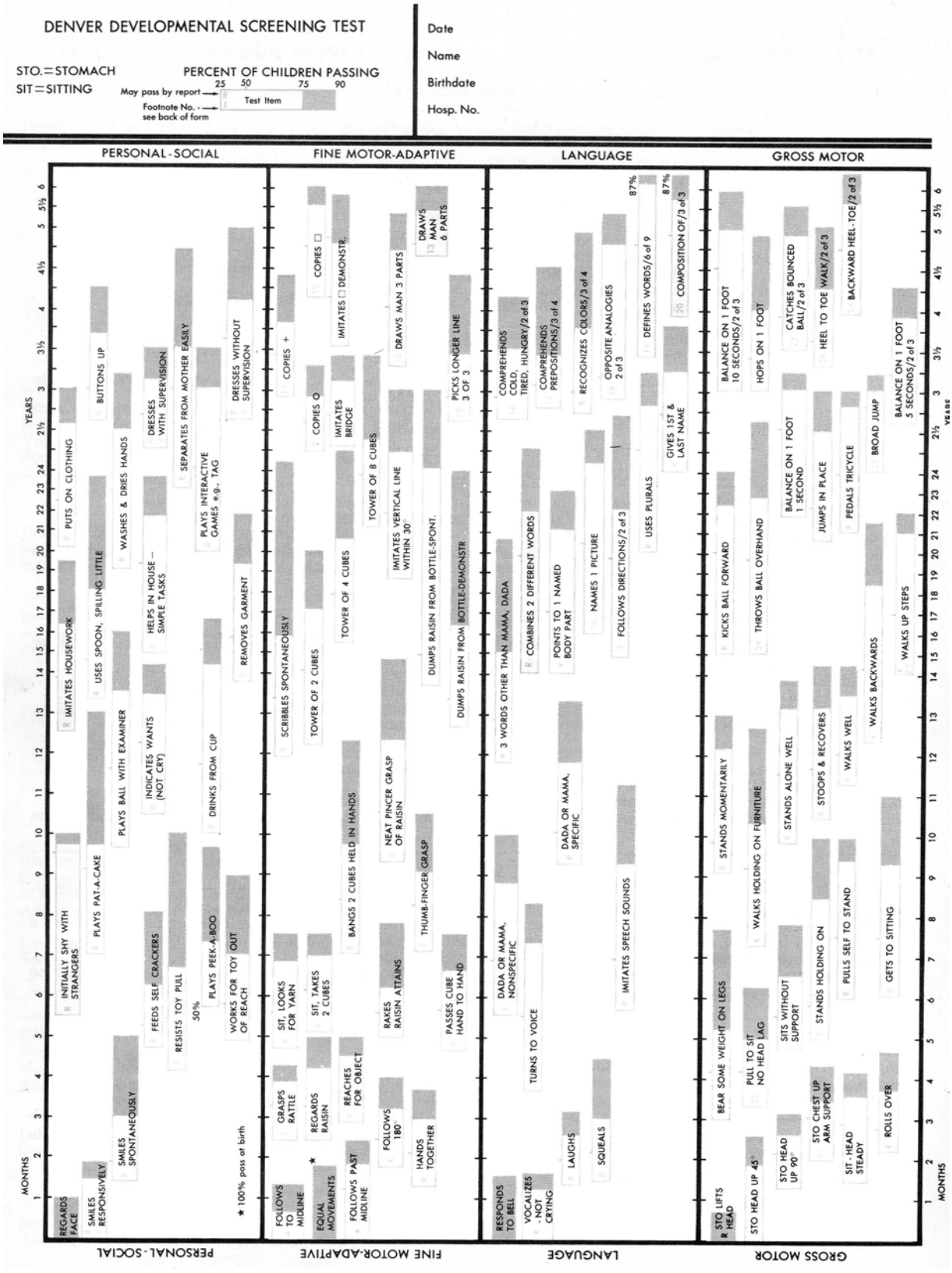


Figure (3-2): Denver developmental screening test

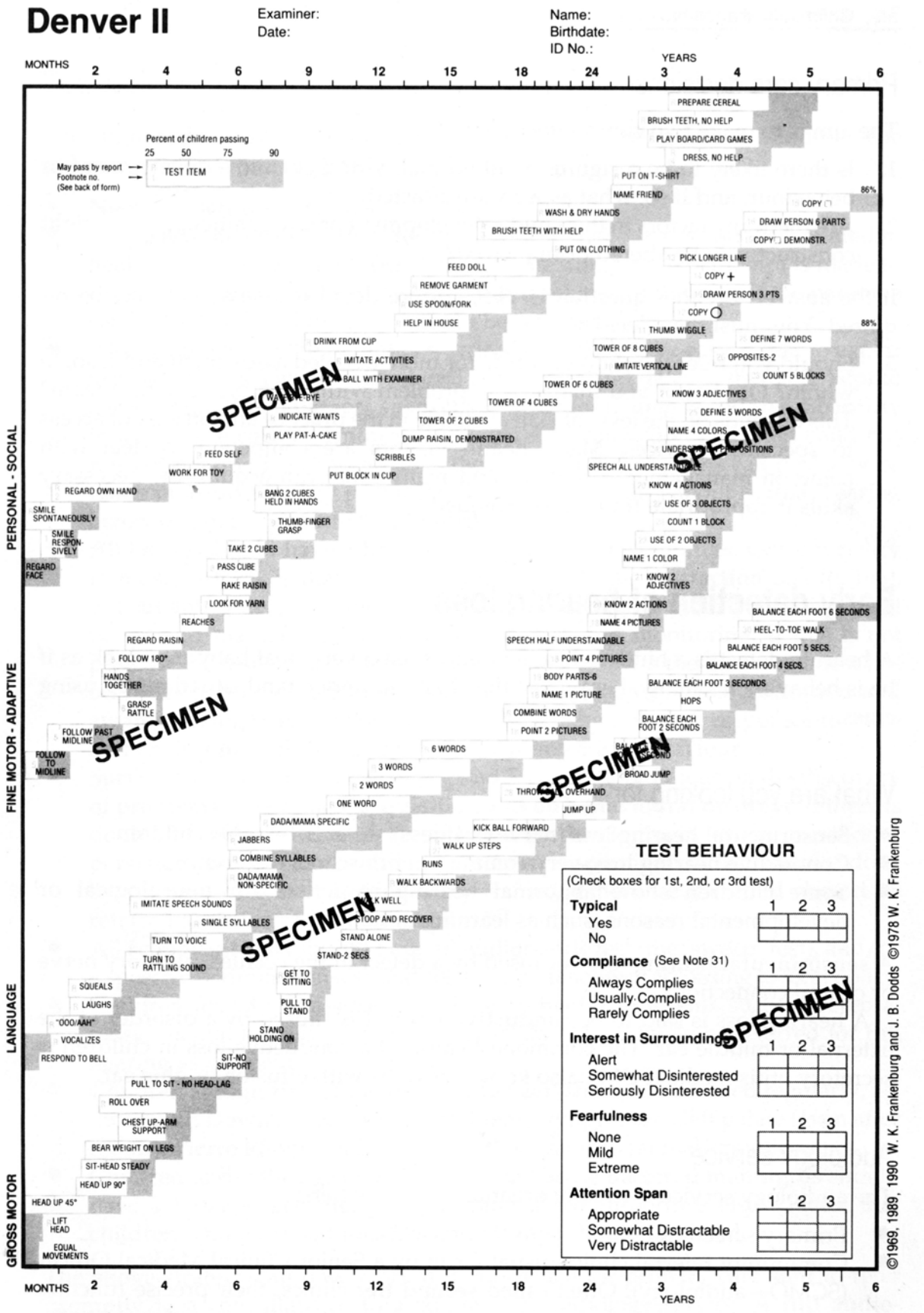


Figure (3-3): Denver II developmental screening test

Instructions for use of Denver Developmental Screening Test (DDST)

• **Date and behavioral observations** (how child feels at time of test, relation to tester, attention span, verbal behavior, self-confidence, etc,).

• **The Denver Developmental Screening Test**

The Denver Developmental Screening Test (DDST) is a device for developmental screening in infancy and the preschool years. It has been standardized on children of Denver. The test form is reproduced in Figure (3-2).

- Test materials

A piece of red wool; box of raisins; rattle with a narrow handle; small clear glass bottle with 5/8 inch opening; bell; tennis ball; test form; pencil; 8 1 inch cube blocks, colored red, blue, yellow, green.

- General instructions

The parent should be told that the purpose is to obtain an estimate of the child's level of development and that the child will not be able to perform all test items. The test relies on observations of the child and on report by a parent who knows the child. Direct observation should be used whenever possible. Every effort should be made to put the child at ease. The younger child may be tested while sitting on the parent's lap in such a way that he or she can comfortably reach the test materials on a table. One or two test materials may be placed in front of the child while the parent is queried regarding personal-social items. The first test items chosen should assure the child an initial successful experience. To avoid distractions it is best to remove all test materials from the table except those required for the test that is being administered.

- Steps in administering the test

1. Draw a vertical line on the examination sheet at the child's chronologic age. Place the date of the examination at the top of the age line. For children who were born prematurely, subtract the number of months of prematurity from the chronologic age. Adjust the age line appropriately and note the amount of adjustment at the top of the line.
2. The items to be administered are those in the Personal-Social, Fine Motor-Adaptive, Language, and Gross Motor sectors through which the child's chronologic age line passes. In each sector one should establish age levels at which the child passes all the items and at which all items are failed.
3. When a child refuses to do an item requested by the examiner, the parent may administer the item, provided this is done in the prescribed manner.
4. If a child passes an item, a large letter "P" is written on the bar. "F" designates a failure, and "R" designates a refusal.
5. Note is made of the child's adjustment to the examination (cooperation, attention span, self-confidence) and relationships to parent, to the examiner, and to the test materials.
6. The parent reports whether the child's performance was typical. This is recorded.

7. For retesting, use the same form, with different colors for each scoring line and age.
8. Instructions for administering footnoted items are on the back of the test form.

- Interpretations

Each test item is designated by a bar. The left end of the bar, the hatch mark at the top of the bar, the left end of the shaded area, and the right end of the bar designate respectively the ages at which 25 per cent, 50 per cent, 75 per cent, and 90 per cent of the reference population performed the item successfully.
Failure on an item achieved by 90 per cent of children of the same age should be considered a "delay." Performances are scored as abnormal if two or more sectors have two or more delays, or if one sector has two or more delays and one other sector has one delay and in the same sector the age line does not intersect one item that is passed; as questionable if anyone sector has two or more delays, or if one or more sectors have one delay and in the same sectors the age line does not intersect an item that is passed; as un-testable if refusals occur in numbers large enough to cause the test score to be questionable or abnormal if the refusals were to be scored as failures; and as normal if the performance is not abnormal, questionable, or un-testable.

Suspect performances should be evaluated. They may be due to temporary factors, such as fatigue, illness, hospitalization, separation from parent, fear, and so on; chronic unwillingness to do things requested; general retardation; pathologic factors, such as deafness or neurological impairment; or familial patterns of development.
If test results are abnormal, questionable, or un-testable, the child should be re-screened a month later. Without improvement, the child should be evaluated with more extensive and refined diagnostic studies.

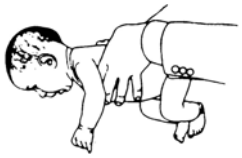
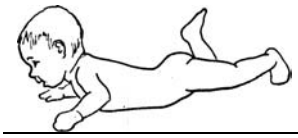
- Caution

The DDST is not an intelligence test and does not establish a DQ or an IQ. It is a screening instrument for use in clinical practice to identify children whose development may need critical study.

3. Suggested guide for developmental screening procedures

Developmental screening at 6 - 8 weeks

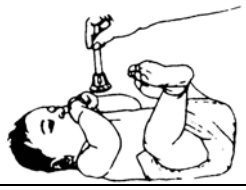
Gross Motor

Test or Sign Sought	Pass	*Achieved by*	*Possible Problems*
Head control			
Pull baby to sitting position 	Holds head upright	Usually 6 weeks	• Hypotonia • Cerebral disorder • Sedation • Intercurrent illness
Ventral Suspension			
Hold baby in ventral suspension 	Head in line or higher than body	4 – 6 weeks	• Floppiness • Spasticity • Retardation
Prone position			
Place baby face down on a flat surface 	Lifts head momentarily	All babies by 6 weeks	Same as head control
Moro's response			
Slight dropping of examiner's hand from head 	Extension & abduction of arms followed by adduction towards chest (symmetrical)	Neonates Disappears by 3 - 4 months	• Asymmetry: - *Dislocation* - *Infection* - *Fracture* • Weak reflex: - *Hypotonia* - *Severe hypertonia*

Fine Motor and Vision

Test or Sign Sought	*Pass*	*Achieved by*	*Possible Problems*
Stares			
Look and ask mother	Stares at a face 25 cm distance Mothers report	1st week - 1st month (one focal length camera)	• Visual dysfunction • Developmental delay
Follows Horizontally 90°			
Child on his back and face to one side Move a red woolen ball in front of face 	Eyes cross midline and follows at least 90° Make 3 trials, few minutes apart	4 weeks	Poor vision

Hearing and Speech

Test or Sign Sought	*Pass*	*Achieved by*	*Possible Problems*
Rattle or bell, 15 cm at ear level			
Hold rattle or bell so child can not see Shake rattle for few seconds at 15 cm at ear level with 5 seconds pauses Increase loudness if no response 	Baby becomes quiet Any other response, e.g. widens eyes etc.	Since birth	Poor hearing
Startle response			
Clap suddenly at ear level at 30 cm or ask parents	Blinking, stiffening, extending limbs or crying (Exam or history)	Since birth	Poor hearing

Social Behavior and Play

Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Smile			
Smile at child or ask parents to smile at 25 cm away	Child smiles without any stimulation in response to examiner or parents' face	90° by 6 weeks	• Visual dysfunction • Developmental delay

Developmental screening at 6 - 8 months

Gross Motor

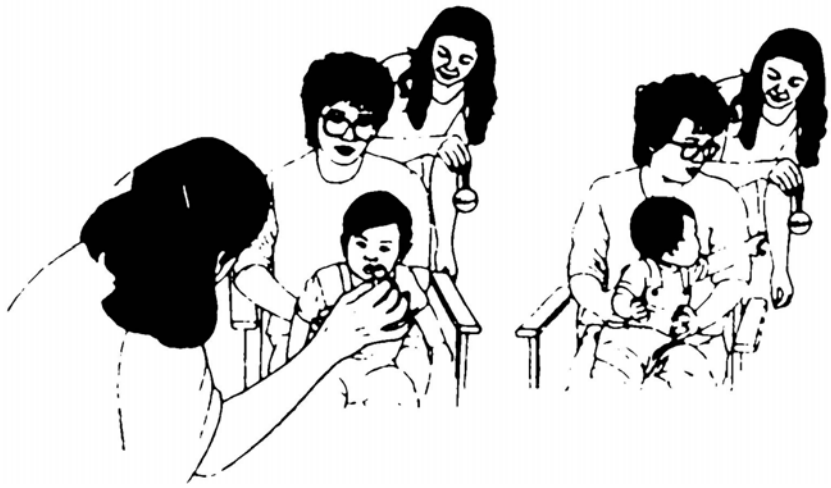
Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Sits			
Put child to sitting position, look at neck and back **Ask parents** 	Sits well with slight support	4 months with support 8 months (majority without support)	• Developmental delay • Weakness • Hypotonia or hypertonia • ?Normal floppy infants walk late
Bears weight on legs			
Hold child upright in vertical suspension and rest his feet on a table **Relax but not release**	Supports weight on legs	From 20 weeks bears most weight on legs	• Developmental delay • CP • Social deprivation • Floppy infant
Rolling			
Put child in prone and slightly help roll by edging leg	Rolls	From prone to supine by 24 weeks From supine to prone by 28 weeks	• Developmental delay • Spasticity • Floppiness

Fine Motor and Vision

Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Reaches out and grasp? Transfers and mouths			
Seat child on parents lap Place a brick, or other toy within easy reach and encourage 	Child picks up or tries to reach the toy Note the grasp	• Palmer grasp at 6 months • Scissoring at 7 months • Pincer at 9 months	• Developmental delay • Fine Motor problem
Watches rolling balls			
Seat child on mothers lap and roll a 6 cm white ball on colored ground at 3 meters	Child follows with eyes	Around 6 months	• Visual dysfunction • Developmental delay
Follows falling toys			
Seat child on mothers lap in front of table and let a bright toy fall to the floor 	Child looks down to see object	6 months	• Visual dysfunction • Developmental delay
Fixes on small objects			
Drop a smartie on a table or a ball 0.3 mm in front of child. Point to it if to get attention 	Looks at smartie or ball	4 - 6 months	• Visual dysfunction • Developmental delay

Hearing and Speech

Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Hearing distraction test			
Child on parents lap facing observer Attention held by observer moving an attractive object Suddenly observer hides object Examiner makes the sound at ear level	Child looks towards sound	6 - 8 months	• Hearing deficit • Developmental delay



Objects used:

- *Manchester rattle - 30 cm away*
- *Cup and spoon 1 meter away*
- *Low pitched sounds such as oo-oo or babies name in conversation*

N .B. All stimuli must be about 40 dB

Babbling			
Observe if child uses several babbles	Babbles and coos are heard	• Monosyllabic at 1 month • Polysyllabic at 6 - 9 months	• Hearing deficit • Developmental delay
Recognizes own name			
Seat child on mother's lap and whisper his/her name from 20 cm 	Turns to direction of voice Parents give History	4 - 6 months	• Hearing deficit • Developmental delay

Social Behavior and Play

Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Peak a boo game (Bekh) لعبة(بخ)			
Make a 1 cm hole in a A4 paper **Hide face and peep through** **Look out of paper twice on the same side and say (Bekh)** **Then look at where child look through hole**	Child looks in direction of where the face appeared	6 – 8 months	• Visual dysfunction • Developmental delay
Puts everything to mouth			
Observe whether child puts bricks into mouth	Child puts objects into his mouth Report by mother	• Starts by 4 months • Nearly all by 8 months • Disappears 9 - 12 months	• Developmental delay

Developmental screening at 18 months

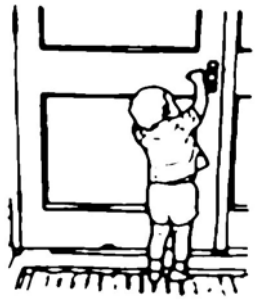
Gross Motor

Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Walks alone			
Observe or ask mother 	Child walks with wide gate	• Holding furniture at 7 – 11 months • Walks alone at 11 – 14 months	• Motor deficit • Developmental delay
Climbs into chair			
Observe or ask mother 	Mother says so	15 - 18 months	• Hypotonia or ataxia • Developmental delay
Climbs stairs			
Ask parents if child ascends stairs unaided or using rail for support 	Parents says he/she does or observed	15 - 18 months	• CP • Developmental delay • Hypotonia

Fine Motor and Vision

Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Delicate pincer grip			
Child sits on parent lap **• In front, on table, place a small object as raisin or smartie** 	Child picks it up using thumb and forefinger	9 - 12 months	• Vision • CP • Developmental delay
Builds tower of 3 – 4 bricks			
• Child on parent's lap **• Place 4 bricks on table and encourage him to build a tower** **• Demonstrate** 	Make a tower of at least 3 bricks 3 attempts	Average 18 months, goes up to 2 years	• Visual dysfunction • Developmental delay • CP

Hearing and Speech

Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Utters more than 3 words			
Ask parents to name number of recognizable words child speaks	Parents say so	From 11 months - 97 % by 18 months	• Hearing Developmental delay • Language problems • Developmental dysphasia
Points to nose and mouth			
Ask child or ask parents to ask child 	Child performs or parents say so	50 % by 18 months 10 % not by 2 years	• Normal • Developmental delay • Hearing
Obeys simple instructions			
Tell child to close door or parent to tell child 	Child performs or parents says so	Average 18 months 10% till 2.5 years	• Normal • Delayed language • Developmental delay

Social Behavior and Play

Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Indicates toilet needs			
Ask parents	Parents says so by pointing or showing potty etc.	15 -30 months	• Normal • Developmental delay
Drinks from cup			
Ask parents 	Parents report	10 – 18 months 10% don't achieve it by 2 years	• Environmental • Developmental delay • Psychosocial
Mimics mother's household activities			
Ask parents if child imitates mother's house work, e.g. brushing floor, etc.	Parents report	12 – 19 months	• Developmental delay • Deprivation

Developmental Screening at 3 years

Gross Motor

Test or Sign Sought	*Pass (History)*	*Achieved by*	*Possible Problems*
Walks upstairs alone one foot per step and two feet per step down stairs			
Ask parents or ask child to do it if possible 	Parents report or observation		• Developmental delay • Motor
Runs confidently			
Ask parents if child can run without falling	Parents say yes or observed		• Developmental delay • Motor
Paddles a tricycle			
Ask parents 	Parents reporting	Majority by 2.5 years	Most significant if child was able to and now not i.e. muscle is getting weaker

Fine Motor and Vision

Test or Sign Sought	*Pass*	*Achieved by*	*Possible Problems*
Copies circle			
Seat child on parents lap in front of a table **Give him a paper and a pencil and show him/her a circle and ask child to draw one** **N.B. don't draw in front of child**	Circle must be closed in any form, circular lines are not accepted	2.5 - 3.5 years	• 20% normal don't draw at 3 years, if fail repeat at 3.5 years • Deprivation • Developmental delay
Builds tower of 8 cubes			
Child must sit comfortably on a chair and a table **Put eight 2.5 cm cubes in front of child and asks him/her to make a tower** 	Child is able to build a tower of 8 bricks Give 3 trials	21 months - 3.5 years	• Poor coordination • Poor Vision • Developmental delay • Deprivation
Matches 2 colors			
Child must sit comfortably on a chair and a table **Put eight 2.5 cm cubes of four primary colors (red, green, yellow, and blue) in front of child and asks him/her to give you a red one** 	Child is able to identify 2 colors, usually red and yellow, but confuses blue and green	3 - 4.5 years	• Poor coordination • Poor vision • Developmental delay • Deprivation • Color blindness
If child can't (usual), show him the red and then asks him to give you one like it, then ask for another one, Repeat with yellow, then blue then green			

Hearing and Speech

Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Uses prepositions			
Ask parents **Ask child to put doll on chair, behind chair, under chair or in front of chair**	Parents report Child does it 3 out of 4	2.5 - 4.5 years 10% don't until 4.5 years	• Hearing Developmental delay • Deprivation
Sentences of 4 words			
Ask parents if child uses small sentences using at least 4 words **Check if possible by conversation**	Parents report or exam		
Gives full name, age and sex			
Ask child what is your name, how old are you, are you boy or girl	Child gives own name, sex and age	10% don't give their last name by 3.9 years	• General language delay • Hearing deficit • Developmental delay

Social Behavior and Play

Test or Sign Sought	*Pass (Ex. or history)*	*Achieved by*	*Possible Problems*
Eats without spilling			
Ask parent			
Dresses without supervision			
Dresses without supervision	Pass by history		• Developmental delay • Deprivation • Cultural

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Annex I: Growth Charts

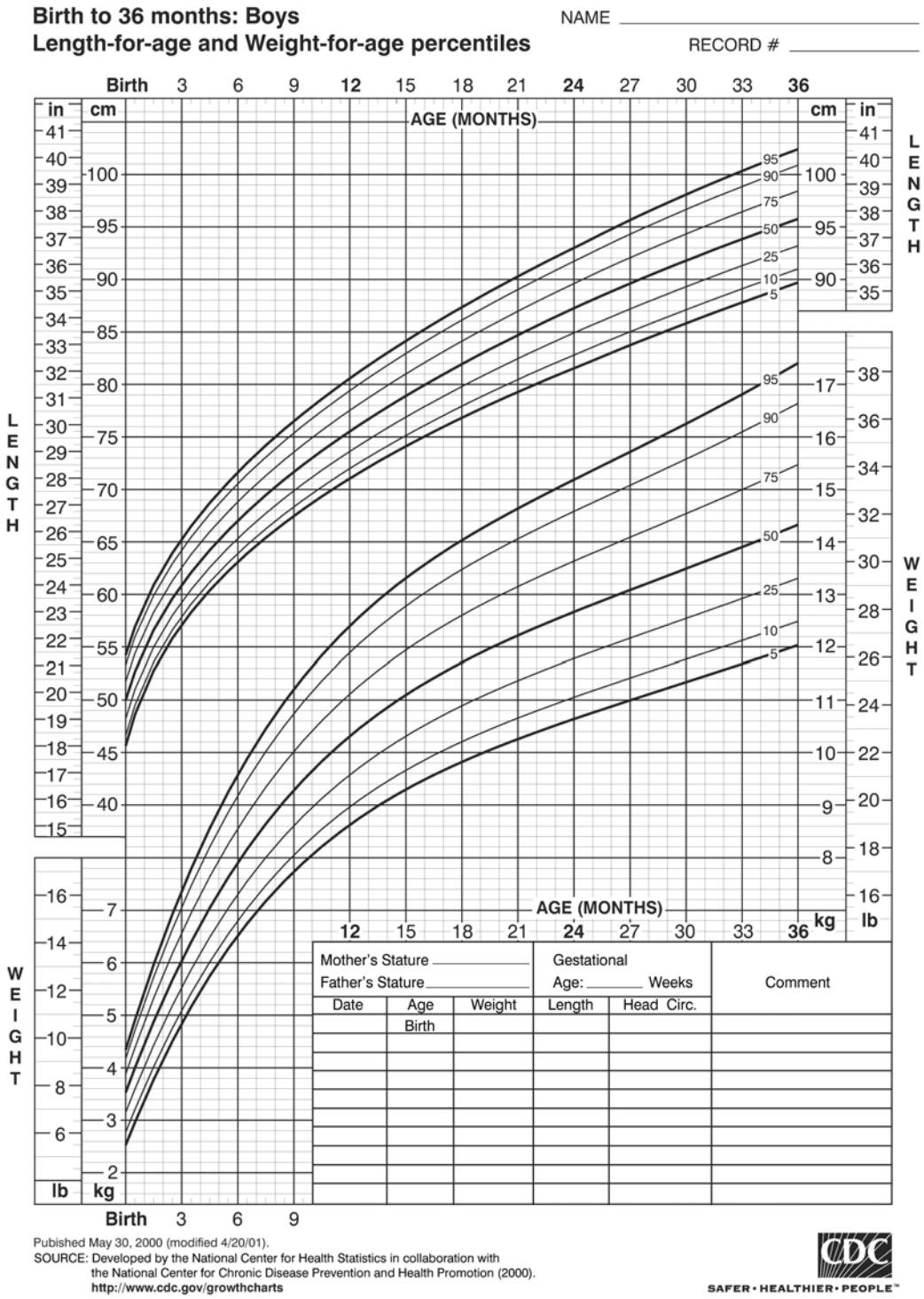


Figure (3-4): Length for age & weight for age percentiles (Boys; 0 to 36 months)

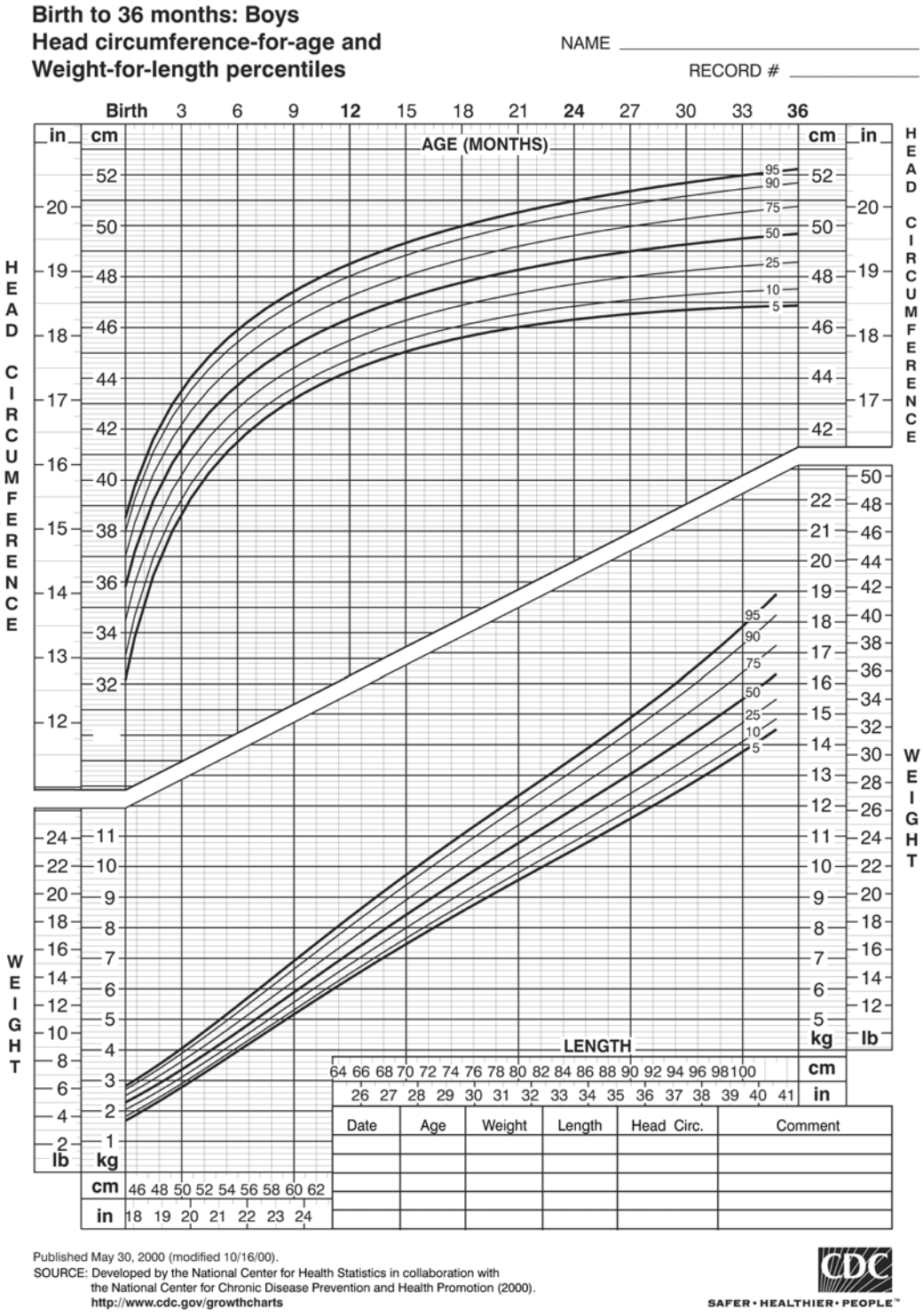


Figure (3-5): Head circumference for age & weight for length percentiles (Boys; 0 to 36 months)

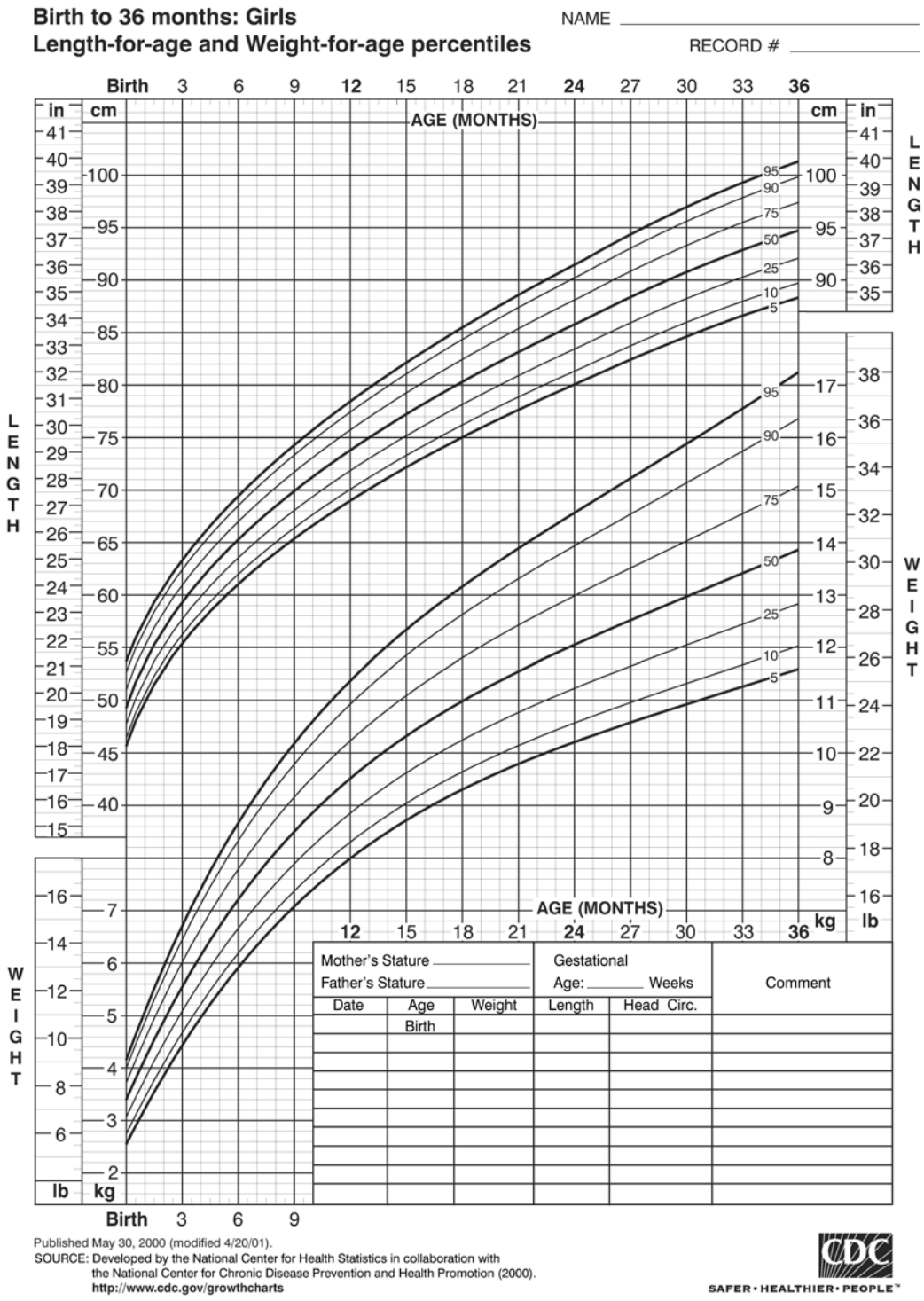


Figure (3-6): Length for age & weight for age percentiles (Girls; 0 to 36 months)

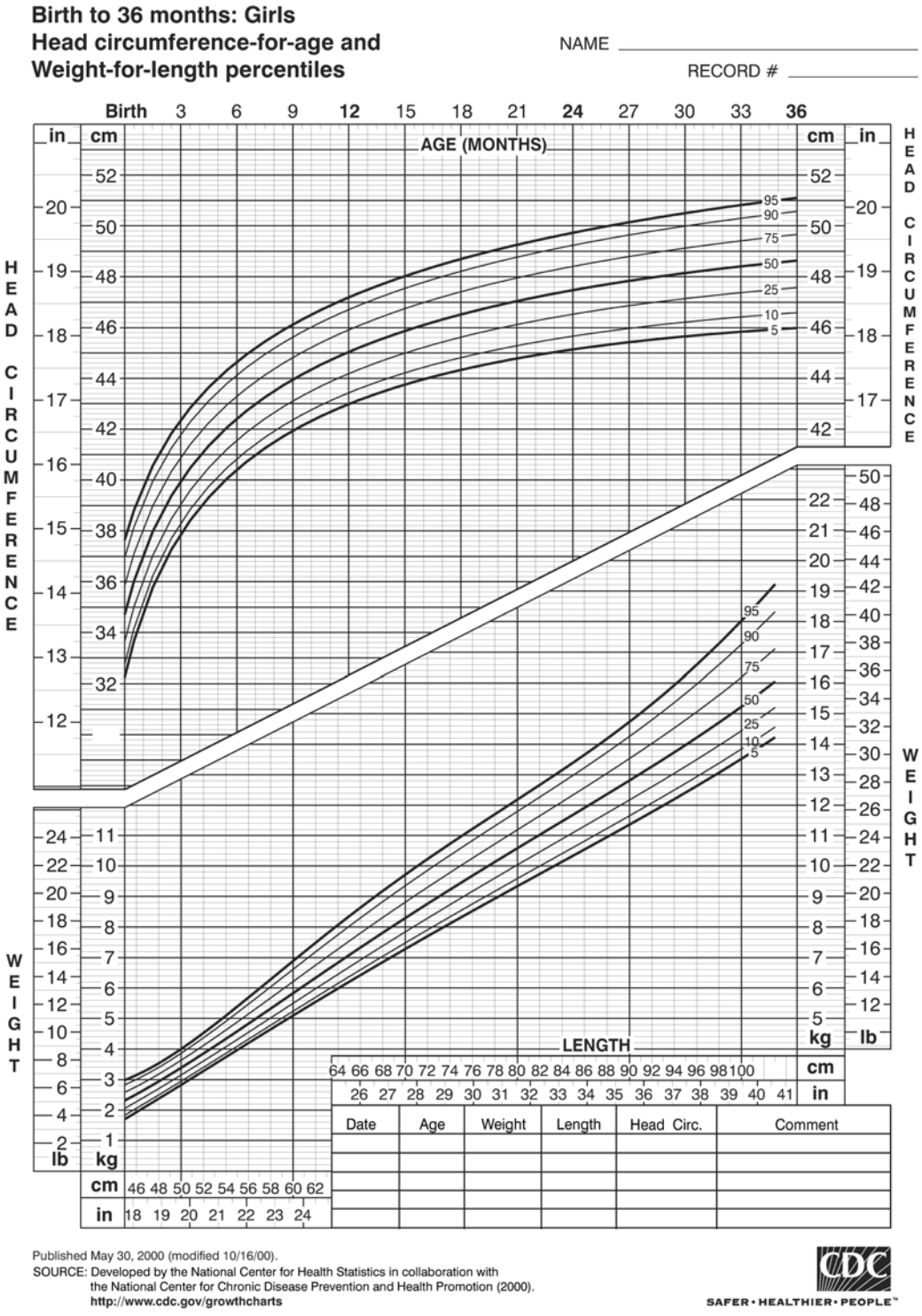


Figure (3-7): Head circumference for age & weight for length percentiles (Girls; 0 to 36 months)

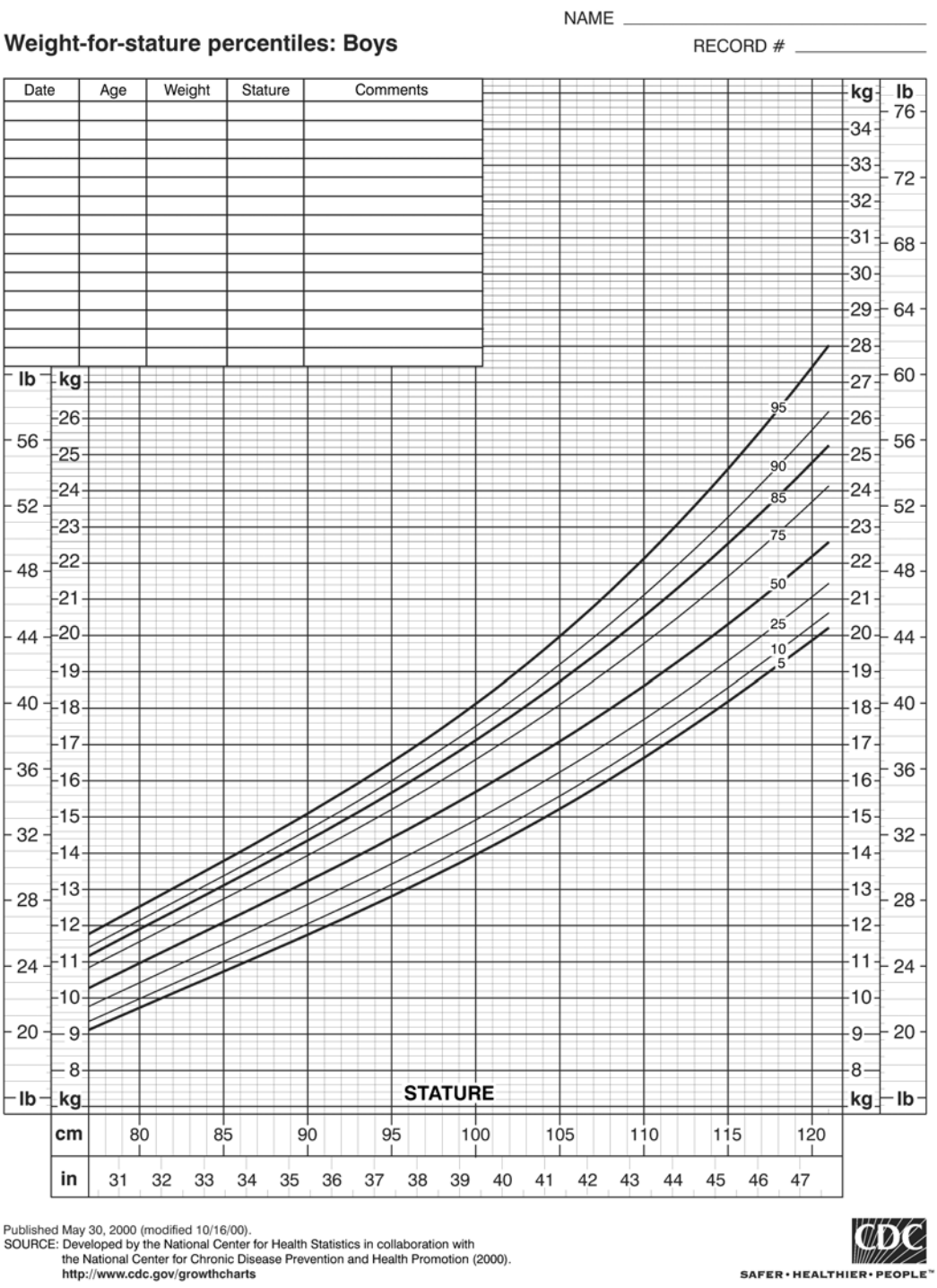


Figure (3-8): Weight for stature percentiles (Boys)

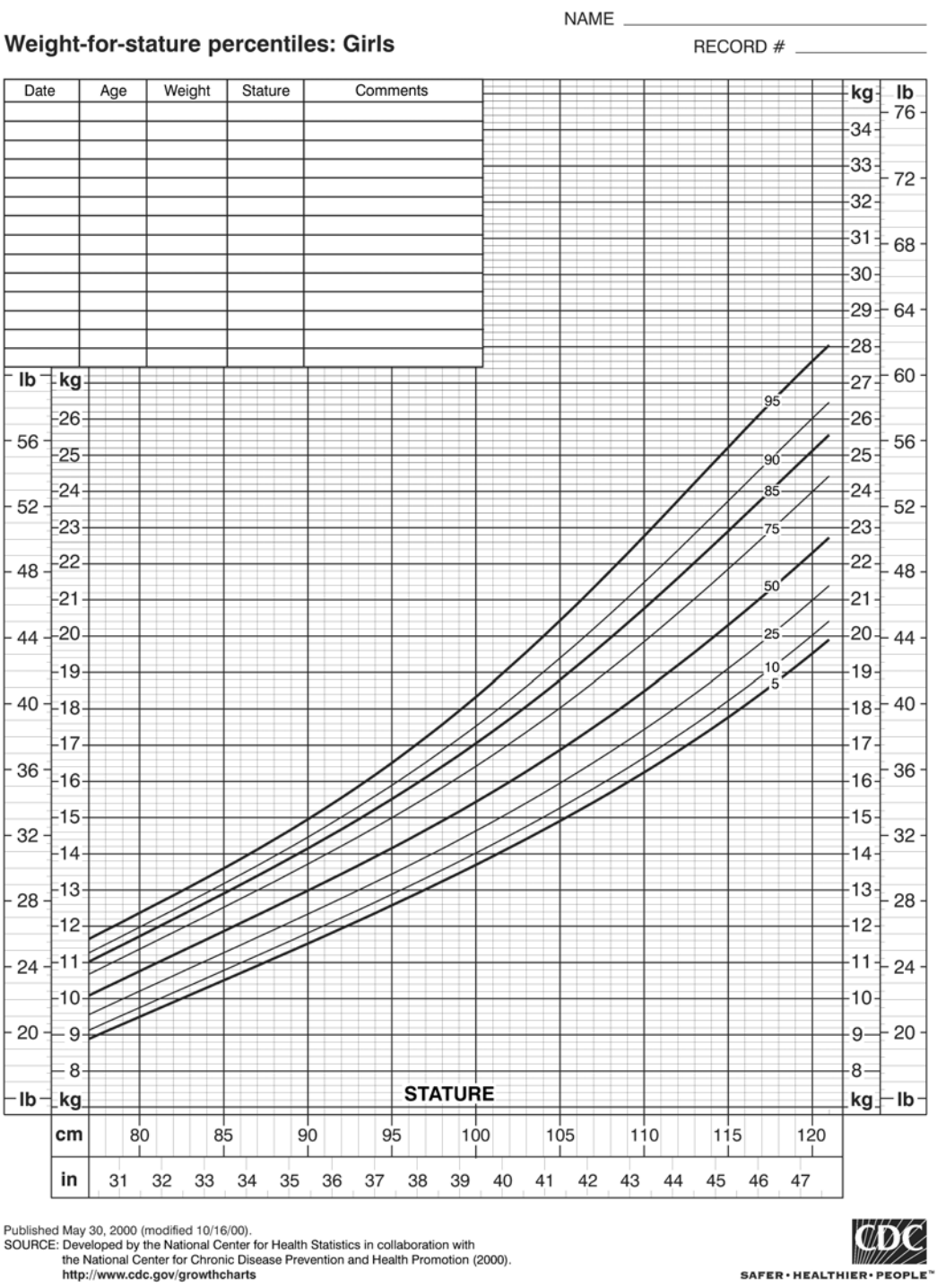


Figure (3-9): Weight for stature percentiles (Girls)

Chapter 4

Motor and Neurological Disabilities

By the end of this chapter the PHC physician should be able to:

- Recognize the definition of motor and neurological disabilities, their classification and their status in Egypt.
- Recognize and describe causes and risk factors of motor and neurological disabilities.
- Recognize important preventive measures of motor and neurological disabilities.
- Recognize and describe normal motor development.
- Make a provisional diagnosis of the motor and neurological disabilities according to the categories outlined.
- Describe essential management procedures and care for the most important health problems.
- To identify appropriate referral needs not fulfilled at the primary care level including medical, social and psychological.

Section 1

Motor Disability

Definition

Motor disability is defined as inability (reduced ability) to perform normal human motor functions, (as standing, walking, using hands, etc…).
It is caused by an interaction between an impairment affecting the structure and/ or function of the motor system & a physically or socially disabling environment.

A child with motor disability must have some structural or functional impairment affecting parts of his motor system and leading to;

- Bone deformities or defects
- Joint deformities, stiffness or hyper mobility
- Muscle weakness, spasticity, flaccidity uncontrolled spasms or contractions, involuntary or uncoordinated movements
- Interruption or disturbance of motor impulses passing through the spinal cord descending tracts, aches, spinal or cranial nerves or neuromuscular junctions.
- Absence or disturbance of motor commands and controls in the brain areas concerned with motor function regulation

Such impairments might cause motor disability manifesting as:

- Delayed or disturbed motor development.
- Inability or reduced ability to perform the main tasks of the motor system:
  - Maintaining postures (lying prone, supine & on either side, sitting on ground and seats, on hands and knees, kneeling and half kneeling, standing on both feet & on one foot…) .
  - Acquiring and changing postures (rolling, coming to sit, to stand and back…).
  - Moving from one place to another (rolling, creeping, crawling, walking, running, using stairs…).
  - Fine motor functions of the hands (supporting, reaching, grasping, releasing, manipulating…).

Types of motor disability

- Pure motor (myopathy)
- Motor with other disabilities (cerebral palsy)
- Part of a multiple disability (metabolic syndromes "cretinism")
- Progressive (myopathy, motor neuron D)
- Stationary (polio, CP)
- Remittent (D.S.)

Degrees of severity

Severity of motor disability constitutes a spectrum starting from no disability to very severe disability. It can be graded as follows:

Degree of severity	
Grade 0	No disability, motor functions performed without difficulty, assistance or aids
Grade 1	Motor functions performed with difficulty
Grade 2	Motor functions performed using aids
Grade 3	Motor functions performed with partial assistance
Grade 4	Motor functions performed with complete assistance
Grade 5	Motor functions performed with assistance and aids
Grade 6	Motor functions cannot be performed

Prevalence

Child disability prevalence in Egypt is justifiably considered to be in the range of 5% half of which (2- 3%) is the prevalence of significant disabilities requiring intensive rehabilitation services. It actually varies in different studies according to the criteria and methodologies used (0.03% - 11%).

Motor disability prevalence ranges between 0.56% and 1.2% (in different studies).

CP incidence is internationally recognized to be 1 in 200 living newborns.

Causes / risk factors

The most common causes in Egypt include:	
• Poliomyelitis	• Polyneuropathy
• Cerebral palsy (secondary to causes occurring during pregnancy, labor and early after delivery)	• Myopathy
	• Fractures
	• Amputation
• Brachial plexus birth injury	• Metabolic bone and soft tissue diseases
• Stroke	• Rheumatoid arthritis. & other joint diseases
• Peripheral nerve injury	
• Spinal cord injuries and diseases	• Congenital deformities

Classification of causes

1. According to anatomical structures affected, causes can be classified as follows:

- **Brain**
  - Hypoxia
  - Hemorrhage
  - Infarct
  - Encephalitis/meningitis
  - Direct injury
  - Toxoplasmosis, German measles, cytomegalovirus

- Drugs (anti-epileptics, antibiotics, immunoregulatory, antidepressants…)
- Prematurity & low birth weight

- **Spinal cord**
  - Poliomyelitis
  - Transverse myelitis
  - Direct injury
  - Nerve roots and plexuses:
  - Brachial birth plexus injury
- **Nerves**
  - Nerve injury (e.g. sciatic)
  - Poly/mono neuropathy (acute infectious, diabetic, lead poisoning neuropathy…)
- **Muscles**
  - Myopathy
  - Myotonia
  - Myasthenia
- **Joints**
  - Rheumatoid arthritis
  - Arthrogryposis multiplex congenita
- **Bones**
  - Osteogenesis imperfecta
  - Congenital bone loss / deformities (ctev, cdh)
  - Fractures

2. According to the time of onset , the causes and risk factors can be classified as (with common examples):

- **Hereditary (genetically determined)**
  - Duchene muscular dystrophy
  - Werding Hoffman disease (AHC)
  - Metabolic diseases
- **Congenital**
  - Congenital dislocation of the hip joint
  - Congenital deformities or limb losses (arthrogryposis multiplex, talipes equino varus, amputations, osteogenesis imperfecta, leg length discrepancy ...etc.
  - Hydrocephalus - Meningeomyelocele
- **Prenatal**
  - German measles
  - Cytomegalovirus infection
  - Toxoplasmosis
  - Direct trauma.
  - The use of drugs (antibiotics, anti-epileptics, antidepressant…)
- **Natal**
  - Birth traumata leading to brachial plexus injury
  - Premature labor
  - Low birth weight
- **Postnatal**
  - Traumata leading to fractures, nerve injuries, spinal cord injury, muscle injury…
  - Encephalitis and meningitis
  - Poliomyelitis, polyneuritis

Early detection / identification

Screening guide for 0- 5 years (MOH)

Age	Gross motor milestones	Fine motor milestones
6- 8 weeks	- Ventral suspension: head in level with spine. - Prone: head momentarily raised. - Supine: symmetrical position & movements. - Pulled to sit: head rises with trunk.	- Hands grasped.
7- 9 months	- Sitting: unsupported for 10 minutes. - Rolling: prone to supine & vice versa. - Standing with support: puts weight on feet.	- Reaches and grasps. - Holds fine objects between index and thumb.
18- 24 months	- Walking unassisted. - Climbing chairs. - Climbing stairs with assistance.	- Picking fine objects between index & thumb. - Building a tower with 3- 6 cubes.
2.5- 3 years	- Climbing up and down chairs. - Jumping on both feet. - Running.	- Imitating vertical & circular lines. - Building a tower with 8 cubes.
4.5- 5 years	- Going up and down stairs alternating feet. - Walking backwards. - Holding a ball thrown to him.	- Imitating drawing a cube. - Building a bridge with 3 cubes. - Drawing a man with several features.

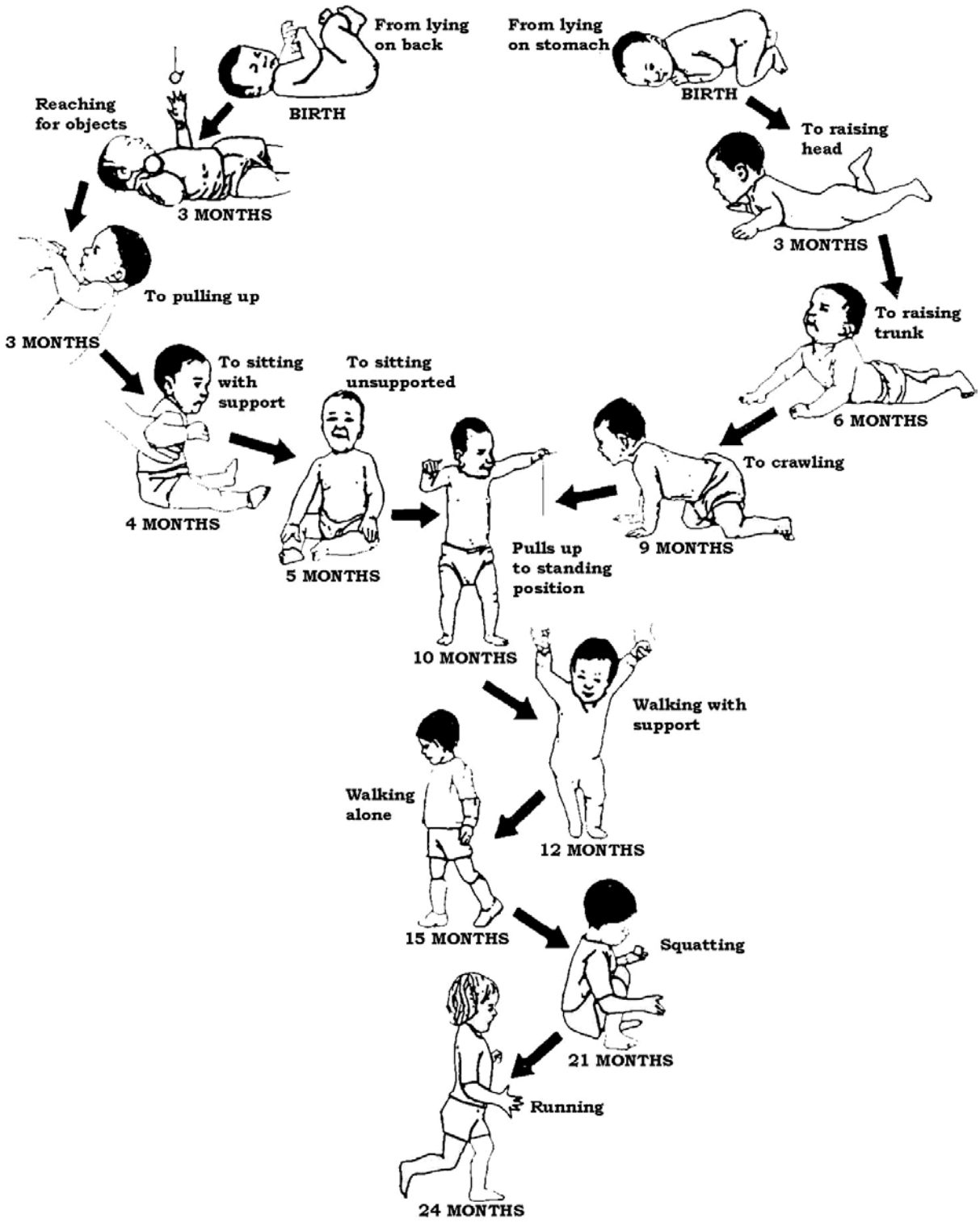


Figure (4-1): Development of strength and co-ordination in the movements of a child from birth to 2 years (known as motor development). *Notice that* **(i)** the process is one of continuous development, not steps and stages; **(ii)** the sequence is the same for all children *(for example, head control always comes before sitting, sitting before crawling, etc.)* but there may be many individual variations *(for example, some babies don't crawl but progress straight to walking)*; and **(iii)** the age at which a particular motor skill is achieved varies with certain limits. *(Illustration designed by* **Dr Pam Zinkin***, Institute of Child Health).*

Infant Neurological International Battery (INFANIB)

INFANIB Description of items

Child in supine position

1. Hand grasp: Observe hands, whether fisted, closed sometimes or open most of the time.
2. ATNR (Asymmetrical tonic neck reflex): Line up neck & body, turn head to side and observe for fencing position.
3. TLR (Tonic labyrinthine reflex): Place hand between scapulae, rub the back and observe for shoulder retraction, arms flexion or extension and leg extension.
4. Scarf sign: Grasp arm near shoulder, move it across chest and observe angle between arm and side of the body & place of elbow on chest.
5. Leg abduction: Grasp legs above knees, abduct them and observe angle between them.
6. Popliteal angle: Grasp legs at knees, keep buttocks on table, extend knees and observe angle of knee flexion.
7. Heel ear angle: Grasp legs near ankles, move feet towards face and observe angle between trunk and legs.
8. Dorsiflexion angle: Press foot up and observe angle between foot and leg.
9. Foot grasp: Place your thumb at the base of toes and watch for toes curling towards it.
10. Pulled to sit: Hold forearms and pull to sitting, observe head for lag , arms for flexion and shoulders for thrusting.

Child in sitting position

11. Observe back for straightening and rounding
12. Side parachute: Hold infant, tip him deliberately to each side and observe for arm extension
13. Back parachute: Hold infant, tip him deliberately backwards and observe for arms extension backwards or turning to one side.
14. Body rotative: Observe for spontaneous rolling from supine to prone (segmental / log) and for pulling to stand. (Can depend on mother's answer)
15. Body derotative: Grasp both feet, twist infant to initiate rolling, observe for (segmental /log rolling). (Can depend on mother' answer).

Child in prone position

16. On all fours: Observe upper & lower limbs.
17. TLR (prone): Flex head and observe for increase of limbs extension.
18. Forwards parachute: Hold infant from mid trunk with your hands at his sides, tip him forwards quickly and observe for arms extension.

Child in standing position

19. Positive support reaction: Hold infant from armpits, lower him down to table surface and observe feet whether flat or touching with toes only.
20. Weight bearing: Lower infant slowly and observe ability to support his weight and position of knees and hips.

Scoring INFANIB

Item	Normal (5)	Mildly/moderately abnormal (4,3)	Abnormal (2,1)
1. Hand grasp	Opened	Slight in 1 or 2 hands	Fisted always/almost
2. ATNR	Absent	In & can move out	Strong & persistent
3. TLR,supine	Absent	Change in tone/mild	Strong & persistent
4. Scarf sign	Same age level	One age level less	2 /3 age levels less
5. Leg abduction	Same age level	One age level less	2 /3 age levels less
6. Popliteal angle	Same age level	One age level less	2 /3 age levels less
7. Heel- ear angle	Same age level	One age level less	2/3 age levels less
8. Dorsiflexion angle	Same age level	One age level less	2 /3 age levels less
9. Foot grasp	Present (to 6 months)	Increased/asymmetric	Planter flexed, strong
10. Pulled to sit	Flexed head & arms	Partial head lag, arms not used	Complete head lag, thrusting
11. Sitting	Extended back, propping	Round back, no propping	Totally flexed, unable to sit alone
12. Side parachute	Present	Slow/asymmetric	Absent/support on elbows or fisted hands
13. Back parachute	Present	Slow/asymmetric	Absent/support on elbows or fisted hands
14. Body rotative	Present	Poor	Absent
15. Body derotative	Present	Poor	Absent
16. On all four	Head lifted 90/weight on hands	Head lifted < 90 / weight on forearms	No movement/no weight on forearms
17. TLR,prone	Absent	Change in tone/mild	Flexion of extremities
18. Forward parachute	Present	Slow/asymmetric	Absent
19. +ve support Rx.	Plantigrade	Partial	On toes
20. Weight bear stand.	Takes weight	Unequal weight bear	Absent/weak

French angles scoring

Months	1- 3	4- 6	7- 9	10- 12
Scarf sign				
Popliteal angle	90	105	120	165
Leg abduction	60	90	120	150
Heel- ear angle	90	105	120	165
Dorsi flexion angle	60	60	60	60

Cut points for INFANIB

Age	0 - 4 months	4 - 8 months	8 - 12 months
Abnormal	≤ 48	49- 65	>= 66
Transient	≤ 54	55- 71	>= 72
Normal	≤ 68	69- 82	>= 83

Assessment

Physical and neurological examination of a child with a possible motor disability for diagnosis and assessment:

- Examination should be as relaxing as possible to parents, as fun as possible for child and gaining confidence and cooperation.
- Examination should address the structure and functions of the motor system.
- The main structures are brain, spinal cord, nerve roots and plexuses, cranial, spinal and peripheral nerves, muscles, joints and bones.
- The main functions include maintaining postures, changing postures, moving from one place to another and hand functions.

Examination of locomotor system structure and functions include:

- **General intellectual brain functions;** attention, concentration, perception, orientation…etc.

- **General behavior**

- **Posture;** lying supine, prone, on sides, sitting on mother's lap, on chair, on ground (with legs extended, crossed and bent backwards, kneeling and half kneeling, standing…etc.

- **Changing position;** rolling, creeping, crawling, coming to sit, to stand and vice versa…etc.

- **Ambulation** (moving from one place to another; rolling, creeping, crawling, shuffling (in sitting), walking (gait description)…etc.

- **Manual dexterity;** in supporting body weight (while in prone, on hands and knees, kneeling, sitting, standing, crawling, walking…), in reaching for objects, grasping, releasing, manipulating objects and using both hands simultaneously.

- **Functional use of hands in ADL** (activities of daily living); eating, drinking, dressing & undressing, toilet activities, bathing, grooming, play activities…etc.

- **Muscle power;** when possible muscle power can be examined by asking child to move joints or putting joints in positions that call for muscles to contract (e.g., to prevent a limb from falling) and graded accordingly:
  - Grade (0) muscle showing no movement or flicker.
  - Grade (1) muscle showing a flicker but not moving the joint.
  - Grade (2) joint moved through full range with gravity nullified.
  - Grade (3) joint moved through full range against gravity.
  - Grade (4) joint moved through full range against gravity and some resistance.
  - Grade (5) joint moved through full range against gravity and maximal resistance.

• **Tone and reflexes.** To show hypotonia, normotonia or hypertonia, weak, normal or exaggerated reflexes in the four limbs.

• **Types of joint movements**

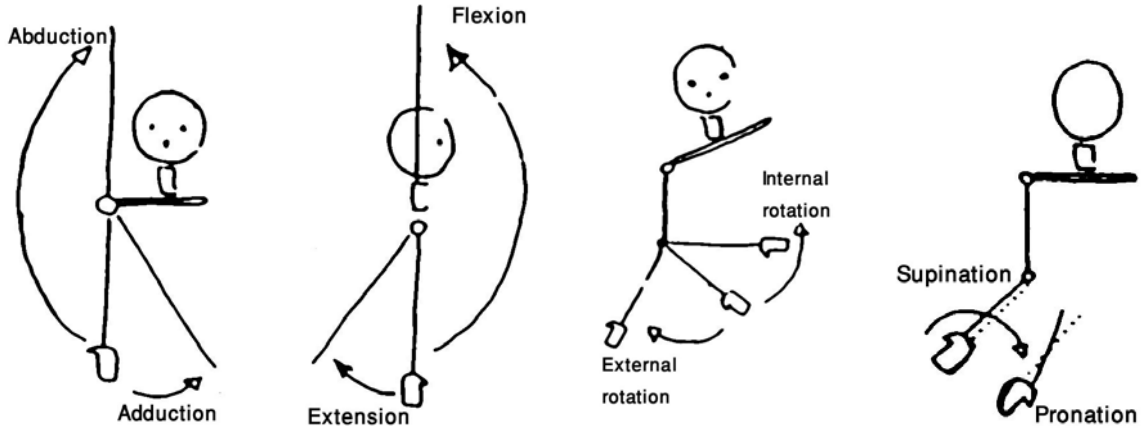


Figure (4-2): Types of joint movements

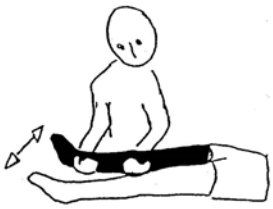
Simplified summary for the most important peripheral joints movements and range of mobility				
• **Shoulder**	Flexion	= 180°	Extension	= 45°
	Adduction	= 45°	Abduction	= 180°
	Internal rotation	= 90°	External rotation	= 90°
• **Elbow**	Flexion	= 135°	Extension	= 45°
• **Forearm**	Supination	= 90°	Pronation	= 90°
• **Wrist**	Flexion	= 75°	Extension	= 90°
	Adduction	= 45°	Abduction	= 15°
• **Hip**	Flexion	= 135°	Extension	= 15°
	Adduction	= 15°	Abduction	= 45°
	Internal rotation	= 30°	External rotation	= 45°
• **Knee**	Flexion	= 135°	Extension	= 0°
• **Ankle**	Plantar Flexion	= 30°	Dorsi flexion	= 15°
• **Foot**	Eversion	= 15°	Inversion	= 30°

• **Joint mobility, stiffness, deformities**… to assess passive and active joint range of motion (ROM) by comparing that to normal range of the examiner, of a child of same age or against a joint movement chart as well as the postures taken by joints while at rest or in use.

Flexion of the hip with fixation of the pelvis

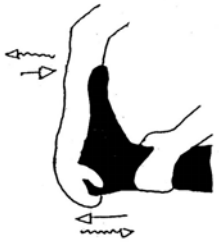
Extension of the hip

Internal and external rotation of hip



Abduction and adduction of the hip with fixation of the opposite limb

Knee flexion and extension



Ankle dorsi and plantar flexion

Toes flexion and extension

Foot inversion

Foot eversion

Figure (4-3): Examination of joint mobility of different joints

• **Bone deformities.** This might include bowing, shortening, lengthening, or swellings…etc.

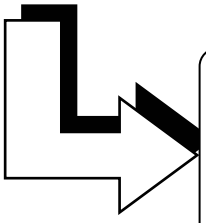
• **Ligamentous laxity.** Accompanying joint hyper mobility (hyper extension of knee, wrist, fingers…).
• **Ataxia.** Incoordination or imbalance observed in sitting, standing, walking or attempts for using hands.

• **Involuntary movements.** Observed at rest and more obviously while attempting to perform difficult tasks requiring fine movements.

• **Sensory impairments.** Particularly hypoesthesia in hands and feet.

• **Minor stigmata.** Particularly of head, face, eyes, ears, lips, nose and hair.

Important points to reach in diagnosis of neuromotor diseases and disabilities



• Is the condition progressive, regressive or stationary?
• Is there a probable diagnostic formulation that can be given?
• Is there any probable therapeutic measure that can be taken?
• Are there any probable associated diseases or impairments?
• Are there any probable environmental or genetic factors related?
• Are there any probable interventions that can ameliorate the condition?
• What can I do myself and what can be done elsewhere to complete the process of treatment and rehabilitation?

• **Is the condition progressive, regressive or stationary?**
If the condition is progressive thorough examination and investigations might be necessary to know the cause, the underlying progressive causes and accordingly the short term and long term interventions to control the causes and prevent progression of the disability and deterioration of the condition (hydrocephalus).

• **Is there a probable diagnostic formulation that can be given?**
Reaching a definite diagnosis is certainly helpful to prescribe treatment, make necessary interventions and prevent recurrences of genetically determined conditions in the families (hypothyroidism, muscle dystrophies).

• **Is there any probable therapeutic measure that can be taken?**
Medical or surgical therapeutic measures should be considered promptly to allow for obtaining best results (hypothyroidism).

• **Are there any probable associated diseases or impairments?**
With cerebral palsy (CP) visual, hearing and mental disabilities are rather common. They have to be searched for to be managed simultaneously with the motor problems.

• **Are there any probable environmental or genetic factors related?**

These might include dominantly, recessively or sex linked inherited diseases as well as environmentally determined conditions (lead poisoning…).

• **Are there any probable interventions that can ameliorate the condition?**

These interventions might include:

- *Medical treatment:*
  - Central and peripheral muscle relaxants to reduce spasticity.
  - Antiepileptic drugs to control fits.
- *Surgical operations:*
  - Tenotomy and tendon lengthening.
  - Bone corrections.
  - Muscle transfers.
- *Orthoses and prostheses:*
  - Lower limb braces (ankle foot, knee ankle foot, hip knee ankle foot orthoses, medical shoes…)
  - Upper limb splints (wrist, hand…)
  - Artificial limbs (above knee, below knee prostheses...)

• **What can I do myself and what can be done elsewhere to complete the process of treatment and reahabilitation?**

- Primary health care (PHC) doctor can give families some instructions and guidance regarding the care to be given to the child to help him moving, growing healthy and developing in all aspects and to prevent possible complications.
- Secondary services might be needed to provide specific services like simple operations, medical treatment, orthoses, physiotherapy.
- Tertiary level services might be needed to provide more sophisticated interventions like surgical operations needing special skills and orthoses/prostheses of very special specifications.

Early intervention & rehabilitation

Rehabilitation includes all processes performed to improve structure/function on the level of impairment, disability and handicap

It works through:

- Training to improve ability
- Assisting to enable to perform
- Reducing difficulties
- Finding alternatives

It utilizes:

- Early (or otherwise) intervention programs (portage program +)
- Developmental programs (programs)
- Physiotherapy (exercises, hydrotherapy, electrotherapy, functional training …)
- Occupational therapy (ADL, play therapy, psychomotor programs…)

- Assistive devices (orthoses, prostheses, walking aids, adl and learning aids …)
- Medical treatment (antiepileptic drugs, muscle relaxants…)
- Surgical interventions (neurectomy, tenotomy, rhizotomy, bone corrective surgeries, muscle transfers …)

Examples of very useful prevention, early detection & early intervention procedures

- Immunization of adolescent girls against german measles
- Immunization against poliomyelitis
- Early detection & early intervention programs with mild cases of mental retardation & learning disabilities
- Identification of Rh negative girls & taking the proper intervention measures
- Early detection of phenyl ketonuria and hypothyroidism
- Early detection & intervention with cases of congenital glaucoma & cataract
- Early detection of hearing defects and early intervention with hearing aids & speech therapy
- Good antenatal, natal & perinatal care of mother & child
- Good early management of epilepsy

Referral

- Early intervention centers
- Rehabilitation centers
- Physiotherapy centers
- Community Based Rehabilitation (CBR) projects
- District & general hospitals
- National institute for motor rehabilitation
- The book "My child, disability and the future"

Section 2

Cerebral Palsy (CP)

Definition

Cerebral palsy (CP) is essentially a motor disability that results from a brain impairment occurring during the brain development. It is characterized by an essentially non progressive impaired control of movement that appears in the first few years of life.

Prevalence

It is estimated that the prevalence of CP is about 2/1000 and that its incidence is about 1/200 new birth. Despite advances in preventing and treating certain causes of cerebral palsy, the number of children and adults it affects has risen over the past 30 years. This is partly because more critically premature and frail infants are surviving through improved intensive care. Unfortunately, many of these infants have developmental problems of the nervous system or suffer neurological damage.

Causes

Causes of cerebral palsy (CP) include different factors that may affect the brain development and cause an insult or damage to the motor areas, the cerebellum, the basal ganglia and/or the brain neuronal connections thus disrupting the brain's ability to control movement and posture. They can be classified as congenital and acquired. In the well developed countries only 20 % of cases of CP are acquired. The figure is much higher in underdeveloped countries.

1. Hereditary

- Microcephaly
- Metabolic diseases

2. Prenatal

- Kernicterus particularly due to Rh incompatibility (father Rh +ve and mother Rh –ve)
- Infections including toxoplasmosis, cytomegalovirus, german measles…
- Exposure to irradiation including X ray.
- Poisoning with lead, insecticides…
- Drug use including antidepressants, antibiotics, immunoregulatory drugs…
- Trauma including direct trauma, falls, accidents…
- Smoking, alcoholics, addiction…

3. Natal

- Pre maturity

- Post maturity
- Low birth weight
- Obstructed labor
- Brain hypoxia causing hypoxic- ischemic encephalopathy.
- Head injury by forceps or ventouse

4. Postnatal

- Trauma to the head
- Brain infection; encephalitis and meningitis
- Intoxication

Many of the causes of cerebral palsy are preventable or treatable; head injury, jaundice, Rh incompatibility, intoxication, complicated pregnancy & labor, and rubella.

Risk factors

Risk factors that increase the possibility of occurrence of CP include:

- Breech presentation
- Complicated labor and delivery leading to vascular or respiratory problems
- Low Apgar score. The Apgar score is determined by checking the baby's heart rate, breathing, muscle tone, reflexes, and skin color in the first minutes after birth. A low score at 10- 20 minutes after delivery is often considered an important sign of potential problems.
- Low birth weight and premature birth. The risk of cerebral palsy is higher among babies who weigh less than 2500 grams at birth and among babies who are born less than 37 weeks into pregnancy. This risk increases as birth weight falls.
- Multiple births (Twins, triplets, and other multiple births)
- Nervous system malformations. Some babies born with cerebral palsy have visible signs of nervous system malformation, such as an abnormally small head (microcephaly). This suggests that problems occurred in the early development of the nervous system.
- Maternal bleeding or severe proteinuria late in pregnancy. Vaginal bleeding during the sixth to ninth months of pregnancy and severe proteinuria are linked to a higher risk of having a baby with cerebral palsy.
- Maternal hyperthyroidism, mental retardation, or seizures.
- Seizures in the newborn. An infant who has seizures faces a higher risk of being diagnosed, later in childhood, with cerebral palsy.

Recognizing these warning signs helps to keep a close eye on children who face a higher risk for long- term problems in the nervous system. However, most such children do not have and do not develop cerebral palsy.

Manifestations

Manifestations of CP differ from one person to another and may even change over time in the same person. They can be classified into:

1. Primary (motor) manifestations including:

- Disturbance of muscle tone in the form of;
  - Hypertonia (spasticity)
  - Hypotonia (flaccidity)
  - Alternating hypotonia and hypertonia.
- Ataxia
  - Involuntary movements
  - Motor development delay and/or disturbance. The milestones of motor development as head control, rolling, creeping, crawling, sitting, standing, walking, running, hand use etc, might all be significantly delayed.

2. Accompanying manifestations including:

- **Mental retardation and learning disabilities.** About one- third of children who have cerebral palsy are mildly intellectually impaired, one- third are moderately or severely impaired, and the remaining third are intellectually normal. Mental impairment is even more common among children with spastic quadriplegia
- **Epilepsy.** As many as half of all children with cerebral palsy have seizures.
- **Delayed language development and speech problems,** mainly dysarthria.
- **Visual problems.** A large number of children with cerebral palsy have strabismus (squint), because of differences in the left and right eye muscles' power and control. In children, the brain often adapts to the condition by ignoring signals from one of the misaligned eyes. Untreated, this can lead to very poor vision in one eye and can interfere with certain visual skills, such as judging distance. Children with hemiplegia may have hemianopia, which is defective vision or blindness that impairs the normal field of vision of one eye. In homonymous hemianopia, the impairment affects the same part of the visual field of both eyes.
- **Hearing problems.** Impaired hearing is more frequent among those with cerebral palsy than in the general population.
- **Behavioral problems.** Due to the difficulties met by children with CP they are subject to develop problematic behaviors and sometimes neuroses or psychoses.
- **Sensory problems.** Some children with cerebral palsy have impaired ability to feel simple sensations like touch and pain. They may also have stereognosia, or difficulty perceiving and identifying objects using the sense of touch.
- **Growth problems.** Failure to thrive is common in children with moderate- to- severe cerebral palsy, especially those with spastic quadriparesis. Children seem to lag behind in growth and development despite having enough food. In babies, this lag usually takes the form of too little weight gain; in young children, it can appear as abnormal shortness; in teenagers, it may appear as a combination of shortness and lack of sexual development. Failure to thrive probably has several causes, including, in particular, poor nutrition and damage to the brain centers controlling growth and development. In addition, the muscles and limbs affected by cerebral palsy tend to be smaller than normal. This is especially noticeable in some patients with spastic hemiplegia, because

limbs on the affected side of the body may not grow as quickly or as large as those on the more normal side. This condition usually affects the hand and foot most severely.

Types

Cerebral palsy can be classified according to the nature or according to the distribution of the manifestations.

According to the nature of manifestations (the type of movement disturbance):

Ataxic CP **Athetoid CP**

Figure (4-4): Types of CP according to manifestations

• **Spastic CP** (70- 80% of cases) characterized by increased muscle tone that might lead to muscle contractures and shortening and accordingly joint stiffness. Spasticity increases usually with weight bearing and with stress.

• **Flaccid CP** characterized by decreased muscle tone and difficulty to start movements and to support & bear weight.

• **Ataxic CP** (the rarest type) affecting the sense of balance and depth perception. Affected persons often have poor coordination; walk unsteadily with a wide- based gait, placing their feet unusually far apart; and experience difficulty when attempting quick or precise movements, such as writing or buttoning a shirt. They may also have intention tremor. In this form of tremor, beginning a voluntary movement, such as reaching for a book, causes a trembling that affects the body part being used and that worsens as the individual gets nearer to the desired object. The ataxic form affects an estimated 5 to 10 percent of cerebral palsy patients.

• **Athetoid CP** with involuntary movements (10- 2- % of cases); characterized by uncontrolled, slow, writhing movements. These abnormal movements usually affect the hands, feet, arms, or legs and, in some cases, the muscles of the face and tongue, causing grimacing or drooling or the muscles needed for speech causing dysarthria. The movements often increase during periods of emotional stress and disappear during sleep.

• **Mixed CP** with manifestations of more than one of the three previous forms. The most common mixed form includes spasticity and athetoid movements

According to the distribution of manifestations:

- Hemiplegia affecting one side of the body
- Monoplegia affecting one upper or lower limb
- Diplegia affecting both lower limbs
- Quadriplegia affecting the four limbs (more significantly the upper limbs)

Hemiplegia **Diplegia** **Quadriplegia**

Figure (4-5): Types of CP according to distribution

Diagnosis

Diagnosis of CP is made through examining the neuromotor system and assessing the motor skills and functions, this includes detecting:

- Motor developmental delay
- Abnormal muscle tone in the form of hypotonia, hypertonia or changing tone
- Muscle shortening, fibrosis or contractures
- Early development of hand preference; before the first year of life
- Non disappearance of primitive reflexes like Moro's, Gallant, asymmetrical tonic neck reflexes
- Non occurrence or delay of protective and tilt reactions
- Excluding other conditions as genetic diseases, muscle diseases, disorders of metabolism, or tumors in the nervous system by assuring that the child's condition is not getting worse by continuous losing motor skills. The child's medical history, special diagnostic tests, and, in some cases, repeated check- ups can help confirm that other disorders are not at fault
- Use of CT, MRI and ultrasonography to identify underlying brain disorders.
- Searching for and detecting other conditions that are linked to cerebral palsy, including seizure disorders, mental impairment, and vision or hearing problems. EEG, IQ testing, hearing tests and visual assessment might be very helpful

Management

Cerebral palsy can not be cured, but treatment can often improve a child's capabilities. In fact, progress due to medical research now means that many patients can enjoy near- normal lives if their neurological problems are properly managed.

There is no standard therapy that works for all patients. Drugs can be used to control seizures and muscle spasms, special braces can compensate for muscle imbalance. Surgery, mechanical aids to help overcome impairments, counseling for emotional and psychological needs, and physical, occupational, speech, and behavioral therapy may be employed.

Physical, occupational, psychological, and behavioral therapy that assist with such skills as movement and speech and foster social and emotional development can help children who have cerebral palsy to achieve and succeed. Medications, surgery, and braces can often improve nerve and muscle coordination, help treat associated medical problems, and either prevent or correct deformities.

In general, the earlier treatment begins, the better chance a child has of overcoming developmental disabilities or learning new ways to accomplish difficult tasks.

The members of the treatment team for a child with cerebral palsy should be knowledgeable professionals with a wide range of specialties. A typical treatment team might include:

- a physician, such as a pediatrician, a pediatric neurologist, or a pediatric physiatrist, trained to help developmentally disabled children. This physician, often the leader of the treatment team, works to synthesize the professional advice of all team members into a comprehensive treatment plan, implements treatments, and follows the patient's progress over a number of years.
- an orthopedist, a surgeon who specializes in treating the bones, muscles, tendons, and other parts of the body's skeletal system. An orthopedist might be called on to predict, diagnose, or treat muscle problems associated with cerebral palsy.
- a physical therapist, who designs and implements special exercise programs to improve movement and strength.

an occupational therapist, who can help patients learn skills for day- to- day living, school, and work.

- a speech and language pathologist, who specializes in diagnosing and treating communication problems.
- a social worker, who can help patients and their families locate community assistance and education programs.
- a psychologist, who helps patients and their families cope with the special stresses and demands of cerebral palsy. In some cases, psychologists may also oversee therapy to modify unhelpful or destructive behaviors or habits.
- an educator, who may play an especially important role when mental impairment or learning disabilities present a challenge to education.

Individuals who have cerebral palsy and their family or caregivers are also key members of the treatment team, and they should be intimately involved in all steps of planning, making decisions, and applying treatments. Studies have shown that family support and personal

determination are two of the most important predictors of which individuals who have cerebral palsy will achieve long- term goals.

Too often, however, physicians and parents may focus primarily on an individual symptom -- especially the inability to walk. While mastering specific skills is an important focus of treatment on a day- to- day basis, the ultimate goal is to help individuals grow to adulthood and have maximum independence in society.

What specific treatments are available?

Physical, behavioral, and other therapies

Therapy - whether for movement, speech, or practical tasks - is a cornerstone of cerebral palsy treatment. The skills a 2- year- old needs to explore the world are very different from those that a child needs in the classroom or a young adult needs to become independent. Cerebral palsy therapy should be tailored to reflect these changing demands.

Physical therapy usually begins in the first few years of life, soon after the diagnosis is made. Physical therapy programs use specific sets of exercises to work toward two important goals: preventing the weakening or deterioration of muscles that can follow lack of use (called disuse atrophy) and avoiding contracture, in which muscles become fixed in a rigid, abnormal position.

Contracture is one of the most common and serious complications of cerebral palsy. Normally, a child whose bones are growing stretches the body's muscles and tendons through running and walking and other daily activities. This ensures that muscles will grow at the same rate. But in children with cerebral palsy, spasticity prevents this stretching and, as a result, muscles do not grow fast enough to keep up with lengthening bones. The resulting contracture can disrupt balance and trigger loss of previous abilities. Physical therapy alone, or in combination with special braces (sometimes called orthotic devices), works to prevent this complication by stretching spastic muscles. For example, if a child has spastic hamstrings (tendons located behind the knee), the therapist and parents should encourage the child to sit with the legs extended to stretch them.

A third goal of some physical therapy programs is to improve the child's motor development. A widespread program of physical therapy that works toward this goal is the Bobath technique, named for a husband and wife team who pioneered this approach in England. This program is based on the idea that the primitive reflexes retained by many children with cerebral palsy present major roadblocks to learning voluntary control. A therapist using the Bobath technique tries to counteract these reflexes by positioning the child in an opposing movement. So, for example, if a child with cerebral palsy normally keeps his arm flexed, the therapist would repeatedly extend it.

A second such approach to physical therapy is "patterning," which is based on the principle that motor skills should be taught in more or less the same sequence that they develop normally. In this controversial approach, the therapist guides the child with

movement problems along the path of normal motor development. For example, the child is first taught elementary movements like pulling himself to a standing position and crawling before he is taught to walk - - regardless of his age. Some experts and organizations, including the American Academy of Pediatrics, have expressed strong reservations about the patterning approach, because studies have not documented its value.

Physical therapy is usually just one element of an infant development program that also includes efforts to provide a varied and stimulating environment. Like all children, the child with cerebral palsy needs new experiences and interactions with the world around him in order to learn. Stimulation programs can bring this valuable experience to the child who is physically unable to explore.

As the child with cerebral palsy approaches school age, the emphasis of therapy shifts away from early motor development. Efforts now focus on preparing the child for the classroom, helping the child master activities of daily living, and maximizing the child's ability to communicate.

Physical therapy can now help the child with cerebral palsy prepare for the classroom by improving his or her ability to sit, move independently or in a wheelchair, or perform precise tasks, such as writing. In occupational therapy, the therapist works with the child to develop such skills as feeding, dressing, or using the bathroom. This can help reduce demands on caregivers and boost self- reliance and self- esteem. For the many children who have difficulty communicating, speech therapy works to identify specific difficulties and overcome them through a program of exercises. For example, if a child has difficulty saying words that begin with "b," the therapist may suggest daily practice with a list of "b" words, increasing their difficulty as each list is mastered. Speech therapy can also work to help the child learn to use special communication devices, such as a computer with voice synthesizers.

Behavioral therapy provides yet another avenue to increase a child's abilities. This therapy, which uses psychological theory and techniques, can complement physical, speech, or occupational therapy. For example, behavioral therapy might include hiding a toy inside a box to reward a child for learning to reach into the box with his weaker hand. Likewise, a child learning to say his "b" words might be given a balloon for mastering the word. In other cases, therapists may try to discourage unhelpful or destructive behaviors, such as hair- pulling or biting, by selectively presenting a child with rewards and praise during other, more positive activities.

As a child with cerebral palsy grows older, the need for and types of therapy and other support services will continue to change. Continuing physical therapy addresses movement problems and is supplemented by vocational training, recreation and leisure programs, and special education when necessary. Counseling for emotional and psychological challenges may be needed at any age, but is often most critical during adolescence. Depending on their physical and intellectual abilities, adults may need attendant care, living accommodations, transportation, or employment opportunities.

Regardless of the patient's age and which forms of therapy are used, treatment does not end when the patient leaves the office or treatment center. In fact, most of the work is

often done at home. The therapist functions as a coach, providing parents and patients with the strategy and drills that can help improve performance at home, at school, and in the world. As research continues, doctors and parents can expect new forms of therapy and better information about which forms of therapy are most effective for individuals with cerebral palsy.

Drug therapy

Physicians usually prescribe drugs for those who have seizures associated with cerebral palsy, and these medications are very effective in preventing seizures in many patients. In general, the drugs given to individual patients are chosen based on the type of seizures, since no one drug controls all types. However, different people with the same type of seizure may do better on different drugs, and some individuals may need a combination of two or more drugs to achieve good seizure control.

Drugs are also sometimes used to control spasticity, particularly following surgery. The three medications that are used most often are diazepam, which acts as a general relaxant of the brain and body; baclofen, which blocks signals sent from the spinal cord to contract the muscles; and dantrolene, which interferes with the process of muscle contraction. Given by mouth, these drugs can reduce spasticity for short periods, but their value for long- term control of spasticity has not been clearly demonstrated. They may also trigger significant side effects, such as drowsiness, and their long- term effects on the developing nervous system are largely unknown. One possible solution to avoid such side effects may lie in current research to explore new routes for delivering these drugs.

Patients with athetoid cerebral palsy may sometimes be given drugs that help reduce abnormal movements. Most often, the prescribed drug belongs to a group of chemicals called anticholinergics that work by reducing the activity of acetylcholine. Acetylcholine is a chemical messenger that helps some brain cells communicate and that triggers muscle contraction. Anticholinergic drugs include trihexyphenidyl, benztropine, and procyclidine hydrochloride.

Occasionally, physicians may use alcohol "washes" - - or injections of alcohol into a muscle - - to reduce spasticity for a short period. This technique is most often used when physicians want to correct a developing contracture. Injecting alcohol into a muscle that is too short weakens the muscle for several weeks and gives physicians time to work on lengthening the muscle through bracing, therapy, or casts. In some cases, if the contracture is detected early enough, this technique may avert the need for surgery.

Surgery

Surgery is often recommended when contractures are severe enough to cause movement problems. In the operating room, surgeons can lengthen muscles and tendons that are proportionately too short. First, however, they must determine the exact muscles at fault, since lengthening the wrong muscle could make the problem worse.

Finding problem muscles that need correction can be a difficult task. To walk two strides with a normal gait, it takes more than 30 major muscles working at exactly the right time

and exactly the right force. A problem in any one muscle can cause abnormal gait. Furthermore, the natural adjustments the body makes to compensate for muscle problems can be misleading. A new tool that enables doctors to spot gait abnormalities, pinpoint problem muscles, and separate real problems from compensation is called gait analysis. Gait analysis combines cameras that record the patient while walking, computers that analyze each portion of the patient's gait, force plates that detect when feet touch the ground, and a special recording technique that detects muscle activity (known as electromyography). Using these data, doctors are better equipped to intervene and correct significant problems. They can also use gait analysis to check surgical results.

Because lengthening a muscle makes it weaker, surgery for contractures is usually followed by months of recovery. For this reason, doctors try to fix all of the affected muscles at once when it is possible or, if more than one surgical procedure is unavoidable, they may try to schedule operations close together.

A second surgical technique, known as selective dorsal root rhizotomy, aims to reduce spasticity in the legs by reducing the amount of stimulation that reaches leg muscles via nerves. In the procedure, doctors try to locate and selectively sever overactivated nerves controlling leg muscles. Although there is scientific controversy over how selective this technique actually is, recent research results suggest it can reduce spasticity in some patients, particularly those who have spastic diplegia. Ongoing research is evaluating this surgery's effectiveness.

Experimental surgical techniques include chronic cerebellar stimulation and stereotaxic thalamotomy. In chronic cerebellar stimulation, electrodes are implanted on the surface of the cerebellum - - the part of the brain responsible for coordinating movement - - and are used to stimulate certain cerebellar nerves. While it was hoped that this technique would decrease spasticity and improve motor function, results of this invasive procedure have been mixed. Some studies have reported improvements in spasticity and function, others have not.

Stereotaxic thalamotomy involves precise cutting of parts of the thalamus, which serves as the brain's relay station for messages from the muscles and sensory organs. This has been shown effective only for reducing hemiparetic tremors (see glossary).

Mechanical aids

Whether they are as humble as velcro shoes or as advanced as computerized communication devices, special machines and gadgets in the home, school, and workplace can help the child or adult with cerebral palsy overcome limitations.

The computer is probably the most dramatic example of a new device that can make a difference in the lives of those with cerebral palsy. For example, a child who is unable to speak or write but can make head movements may be able to learn to control a computer using a special light pointer that attaches to a headband. Equipped with a computer and voice synthesizer, this child could communicate with others. In other cases, technology has led to new versions of old devices, such as the traditional wheelchair and its modern offspring that runs on electricity.

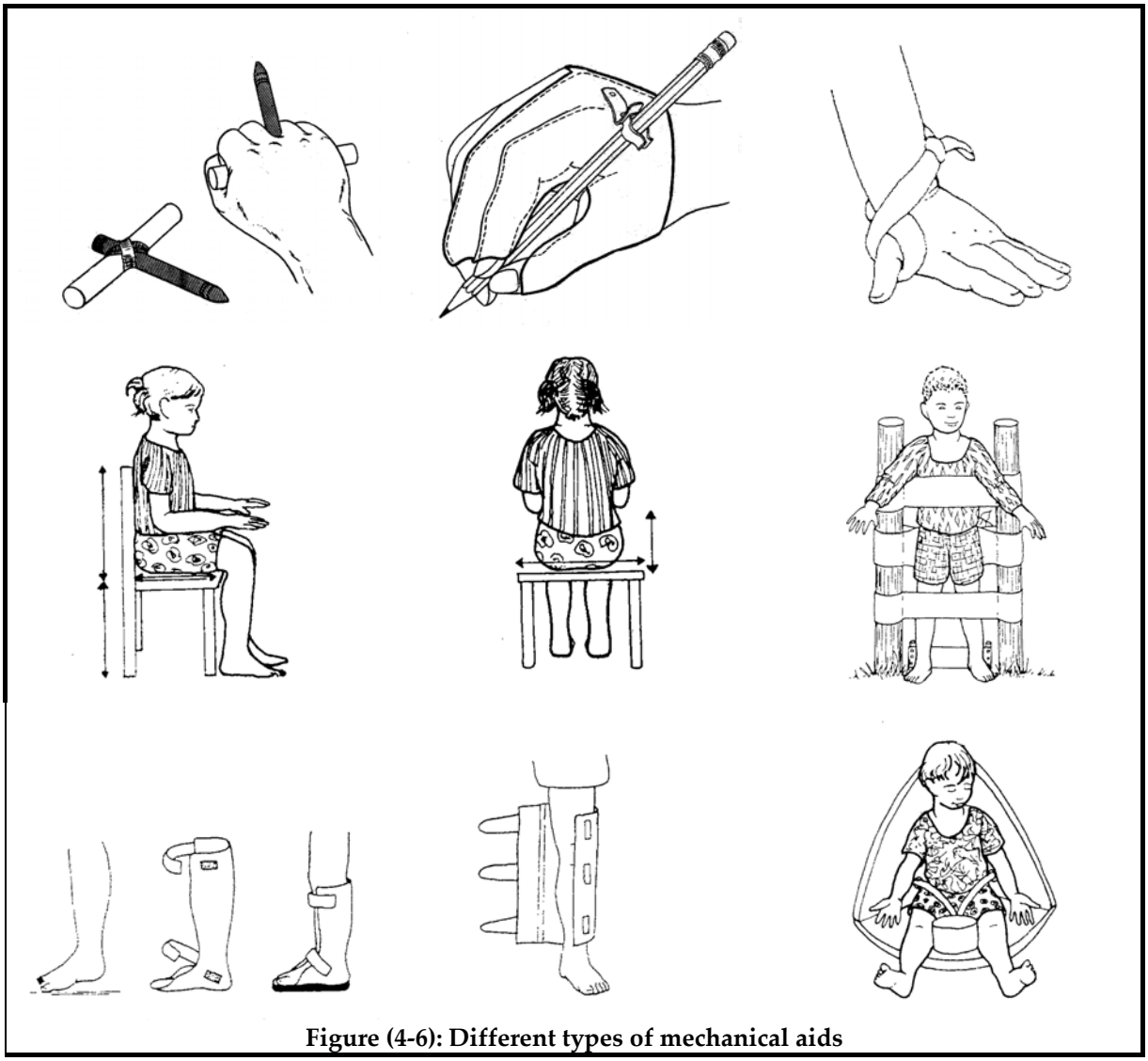


Figure (4-6): Different types of mechanical aids

Many such devices are products of engineering research supported by private foundations and other groups.

Other major problems associated with cerebral palsy

Drooling

Poor control of the muscles of the throat, mouth and tongue sometimes leads to drooling. Drooling can cause severe skin irritation and, because it is socially unacceptable, can lead to further isolation of affected children from their peers. Although numerous treatments for drooling have been tested over the years, there is no one treatment that always helps. Drugs called anticholinergics can reduce the flow of saliva but may cause significant side effects, such as mouth dryness and poor digestion. Surgery, while sometimes effective, carries the risk of complications, including worsening of swallowing problems. Some patients benefit from a technique called biofeedback that can tell them when they are drooling or having difficulty controlling muscles that close the mouth. This kind of therapy is most likely to work if the patient has a mental age of more than 2 or 3 years, is motivated to control drooling, and understands that drooling is not socially acceptable.

Poor nutrition

Difficulty with eating and swallowing - also triggered by motor problems in the mouth - can cause poor nutrition. Poor nutrition, in turn, may make the individual more vulnerable to infections and cause or aggravate "failure to thrive" - - a lag in growth and development that is common among those with cerebral palsy. To make swallowing easier, the caregiver may want to prepare semisolid food, such as strained vegetables and fruits. Proper position, such as sitting up while eating or drinking and extending the individual's neck away from the body to reduce the risk of choking, is also helpful. In severe cases of swallowing problems and malnutrition, physicians may recommend tube feeding, in which a tube delivers food and nutrients down the throat and into the stomach, or gastrostomy, in which a surgical opening allows a tube to be placed directly into the stomach.

Incontinence

Incontinence is a common complication is caused by faulty control over the muscles that keep the bladder closed. Incontinence can take the form of bed- wetting (enuresis), uncontrolled urination during physical activities (stress incontinence), or slow leaking of urine from the bladder. Possible medical treatments for incontinence include special exercises, biofeedback, prescription drugs, surgery, or surgically implanted devices to replace or aid muscles. Specially designed undergarments are also available.

Doctors may improve the effectiveness of the anti-spasticity drug called baclofen by giving the drug through spinal injections, rather than by mouth. In addition, scientists are also exploring the use of tiny implanted pumps that deliver a constant supply of anti-spasticity drugs into the fluid around the spinal cord, in the hope of improving these drugs' effectiveness and reducing side effects, such as drowsiness.

Other experimental drug development efforts are exploring the use of minute amounts of the familiar toxin called botulinum. Ingested in large amounts, this toxin is responsible for botulism poisoning, in which the body's muscles become paralyzed. Injected in tiny amounts, however, this toxin has shown early promise in reducing spasticity in specific muscles.

Scientists speculate that magnesium may play a role in brain development and possibly prevent bleeding inside the brains of preterm infants. Previous research has shown that magnesium may protect against brain bleeding in very premature infants. Animal studies have demonstrated that magnesium given after a traumatic brain injury can reduce the severity of brain damage.

Despite these encouraging research findings, pregnant women should not change their magnesium intake because the effects of high doses have not yet been studied and the possible risks and benefits are not known.

Prevention

Several of the causes of cerebral palsy that have been identified through research are preventable or treatable:

• Head injury can be prevented by regular use of child safety seats in cars & helmets during bicycle rides, and elimination of child abuse. In addition, common sense measures around the household - - like close supervision during bathing and keeping poisons out of reach - - can reduce the risk of accidental injury.

Jaundice of newborn infants can be treated with phototherapy or blood transfusion.

• Rh incompatibility is easily identified by a simple blood test routinely performed on expectant mothers and, if indicated, expectant fathers. In most cases, serum given after each childbirth can prevent the unwanted production of antibodies.

• Rubella, or German measles, can be prevented if women are vaccinated against this disease before becoming pregnant.

• Regular prenatal care and good nutrition and eliminating smoking, alcohol consumption, and drug abuse.

Section 3

Epilepsy

Introduction

Few experiences match the drama of a convulsive seizure. A person having a severe seizure may cry out, fall to the floor unconscious, twitch or move uncontrollably, drool, or even lose bladder control. Within minutes, the attack is over, and the person regains consciousness but is exhausted and dazed. This is the image most people have when they hear the word epilepsy. However, this type of seizure - a generalized tonic- clonic seizure is only one kind of epilepsy. There are many other kinds, each with a different set of symptoms.

Epilepsy was one of the first brain disorders to be described. It was mentioned in ancient Babylon more than 3,000 years ago. The strange behavior caused by some seizures has contributed through the ages to many superstitions and prejudices. The word epilepsy is derived from the Greek word for "attack." People once thought that those with epilepsy were being visited by demons or gods. However, in 400 B.C., the early physician Hippocrates suggested that epilepsy was a disorder of the brain - and we now know that he was right.

Epilepsy is not contagious and is not caused by mental illness or mental retardation. Some people with mental retardation may experience seizures, but seizures do not necessarily mean the person has or will develop mental impairment. Many people with epilepsy have normal or above- average intelligence. Famous people who are known or rumored to have had epilepsy include the Russian writer Dostoyevsky, the philosopher Socrates, the military general Napoleon, and the inventor of dynamite, Alfred Nobel, who established the Nobel prize. Several Olympic medalists and other athletes also have had epilepsy. Seizures sometimes do cause brain damage, particularly if they are severe. However, most seizures do not seem to have a detrimental effect on the brain. Any changes that do occur are usually subtle, and it is often unclear whether these changes are caused by the seizures themselves or by the underlying problem that caused the seizures.

Definition

Epilepsy is a neurological brain disorder in which clusters of nerve cells, or neurons, in the brain sometimes signal abnormally.

In epilepsy, the normal pattern of neuronal activity becomes disturbed, leading to a tendency to have recurrent seizures in the form of strange sensations, emotions, and behavior, or sometimes convulsions, muscle spasms, and loss of consciousness.

During a seizure, neurons may fire as many as 500 times a second, much faster than the normal rate of about 80 times a second. In some people, this happens only occasionally; for others, it may happen up to hundreds of times a day.

Prevalence

- Epilepsy can affect anyone, at any age and at any conditions of life.
- 1 in 20 people will have a single seizure at some time in their life.
- Having a seizure *does not* necessarily mean that a person has epilepsy. Only when a person has had two or more seizures is he or she considered to have epilepsy.

The prevalence of epilepsy is about 1 in 100 – 133.

Prognosis

- Majority of people with epilepsy can have their seizures well controlled by medication and the vast majority of them can take part in the same activities as everyone else, with the help of simple safety measures

- 80% of those diagnosed with epilepsy, seizures can be controlled with modern medicines and surgical techniques.

- 20 % of people with epilepsy will continue to experience seizures even with the best available treatment; called intractable epilepsy

- Many people who develop epilepsy below the age of 20 will grow out of it in adult life particularly if the epilepsy has been well controlled by medication.

- Children with idiopathic epilepsy, or epilepsy with an unknown cause, had a 68 to 92% chance of becoming seizure- free by 20 years after their diagnosis.

- Chances of becoming seizure-free are not as good for adults, or for children with severe epilepsy syndromes.

Causes & risk factors

What causes epilepsy?

Mechanism

- Epilepsy is a disorder with many possible causes.
- Anything that disturbs the normal pattern of neuron activity can lead to seizures.

Examples

- Abnormality of brain neurotransmitters *(abnormally high level of excitatory neurotransmitters or abnormally low level of inhibitory neurotransmitters).*
- Abnormal nerve connections – *may occur during brain development or following head injury, stroke, or other problem may cause epilepsy.*

- Disruption of neuron membrane structure, function, nourishment and repair may lead to epilepsy.
- Changes in glial cells that regulate concentrations of chemicals in the brain may lead to epilepsy.

About half of all seizures have no known cause. However, in other cases, the seizures are clearly linked to infection, trauma, or other identifiable problems.

Genetic factors

Genetic abnormalities are important factors contributing to epilepsy in that:

- May cause some types of epilepsy
- May increase susceptibility to seizures triggered by environmental factor.
- May increase resistance to drugs *(that is why anticonvulsant drugs do not work for some people).*
- May cause brain development abnormalities consequently lead to epilepsy.

People with no family history of epilepsy may develop it due to newly developed abnormality, or mutation, in an epilepsy- related gene.

Other disorders

In many cases, epilepsy develops as a result of brain damage from other disorders including:

- Brain tumors
- Alcoholism
- Alzheimer's disease
- Brain hypoxia
- Meningitis, AIDS, viral encephalitis, and other infectious diseases
- Hydrocephalus
- Intolerance to wheat gluten
- Neuro-cysticercosis; a parasitic infection of the brain

Seizures may stop once these disorders are treated successfully, this however depends on the type of disorder, the brain region that is affected, and how much brain damage occurred prior to treatment.

- Developmental and metabolic disorders including;

- cerebral palsy	- neurofibromatosis
- pyruvate deficiency	- tuberous sclerosis
- Landau- Kleffner syndrome	- autism

Epilepsy in these cases is just one of a set of symptoms commonly found in people with these disorders.

Head injury

Head injury can lead to seizures or epilepsy. Safety measures such as wearing seat belts in cars and using helmets when riding a motorcycle or playing competitive sports can protect people from epilepsy and other problems that result from head injury.

Prenatal injury and developmental problems

The developing brain is susceptible to many kinds of injury such as
- Maternal infections
- Poor nutrition
- Hypoxia

These conditions may lead to CP, which often is associated with epilepsy, or they may cause epilepsy that is unrelated to any other disorders.

About 20 percent of seizures in children are due to cerebral palsy, dysplasia or other neurological abnormalities.

Poisoning

Seizures can result from exposure to lead, carbon monoxide, and many other poisons. They also can result from drugs and from overdoses of antidepressants and other medications.

Seizure triggers

Seizures are often triggered by several factors, these seizure triggers do not cause epilepsy but can provoke first seizures or cause breakthrough seizures in people who otherwise experience good seizure control with their medication. These include:

- Lack of sleep
  - Sleep deprivation is a universal and powerful trigger of seizures.
  - People with epilepsy should try to stay on regular sleep schedule as much as possible.
- Alcohol consumption
- Stress
- Hormonal changes associated with the menstrual cycle.
- Light flashing (e.g. flicker of computer monitor; this is called photosensitive epilepsy)
- Smoking; nicotine stimulate receptors of acetylcholine in the brain (excitatory)
- Sexual activity; very rarely

What are the different kinds of seizures?

More than 30- 40 different types of seizures have been described; a person may have more than one type. Seizures are divided into two major categories - partial seizures and generalized seizures. However, there are many different types of seizures in each of these categories.

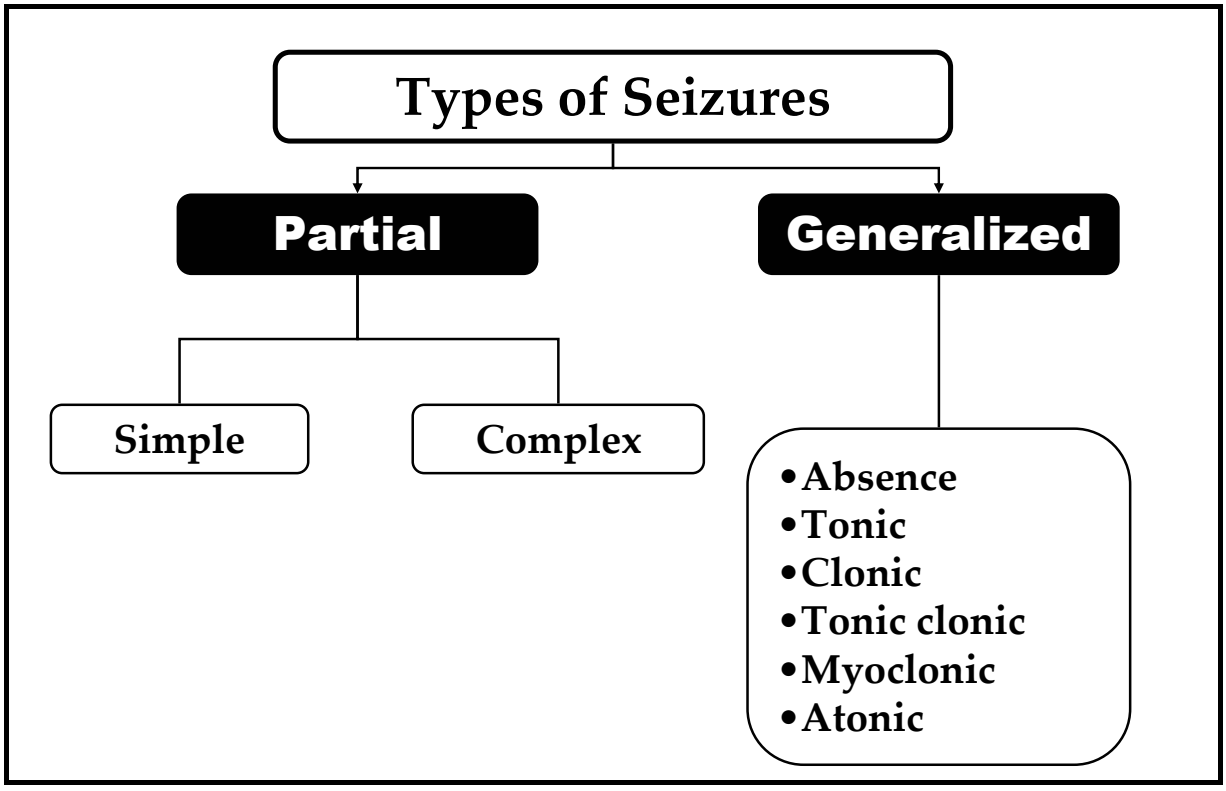


Figure (4-7): Types of seizures

• Partial seizures

- Occur in one part of the brain.
- About 60 percent of people with epilepsy have partial seizures.
- Partial seizures are described by the involved area of the brain from which they originate. (e.g. partial frontal lobe seizures).

Simple partial seizure

- The person will remain conscious
- The person may experience unusual feelings or sensations that can take many forms.
  - sudden and unexplainable feelings of joy, anger, sadness, or nausea
  - hear, smell, taste, see, or feel things that are not real.

Complex partial seizure

- The person has a loss of or change in consciousness (e.g. producing a dreamlike experience)
- The person may display strange, repetitious behaviors (called automatisms) such as
  - blinks
  - twitches
  - mouth movements
  - walking in a circle
  - fling objects across the room

- strike out at walls or furniture as though they are angry or afraid
• These seizures usually last just a few seconds.

Aura
• Some people with partial seizures, especially complex, may experience auras
• Auras are unusual sensations that warn of an impending seizure.
• These auras are simple partial seizures in which the person maintains consciousness.
• Symptoms and progression of aura tends to be similar every time in an individual.

Differential diagnosis
• Dreamlike perceptions associated with complex partial seizure may be misdiagnosed as migraine headaches
• Strange behavior and sensations caused by partial seizures also can be mistaken for symptoms of narcolepsy, fainting, or even mental illness

It may take many tests and careful monitoring by a knowledgeable physician to tell the difference between epilepsy and other disorders.

Generalized seizures

Generalized seizures are a result of abnormal neuronal activity in many parts of the brain. These seizures may cause loss of consciousness, falls, or massive muscle spasms.

There are many kinds of generalized seizures.

• *Absence seizures*
- Sometimes referred to as petit mal seizures *(Old term)*.
- The person appears as if staring into space and/or have jerking or twitching muscles.

• *Tonic seizures*
- Stiffening of muscles of the body, generally those in the back, legs, and arms.

• *Clonic seizures*
- Repeated jerking movements of muscles on both sides of the body.

• *Myoclonic seizures*
- Jerks or twitches of the upper body, arms, or legs.

• *Atonic seizures*
- Loss of normal muscle tone.
- Affected person will fall down or may nod his or her head involuntarily.

• *Tonic- clonic seizures*
- Mixture of symptoms including;
  ▪ stiffening of the body and
  ▪ repeated jerks of the arms and/or legs
  ▪ loss of consciousness.
- Tonic- clonic seizures are sometimes referred to by an older term *grand mal seizures.*

Not all seizures can be easily defined as either partial or generalized. Some people have seizures that begin as partial seizures but then spread to the entire brain. Other people may have both types of seizures but with no clear pattern.

Society's lack of understanding about the many different types of seizures is one of the biggest problems for people with epilepsy. People who witness a non- convulsive seizure often find it difficult to understand that behavior which looks deliberate is not under the person's control. In some cases, this has led to the affected person being arrested, sued, or placed in a mental institution. To combat these problems, people everywhere need to understand the many different types of seizures and how they may appear.

What are the different kinds of epilepsy?

- As there are many different kinds of seizures, there are many different kinds of epilepsy
- Doctors have identified hundreds of different epilepsy syndromes
- Epilepsy syndromes are frequently described

- by their symptoms or
- by where in the brain they originate.

Absence epilepsy

- Repeated absence seizures that cause momentary lapses of consciousness
- Seizures almost always begin in childhood or adolescence
- Tend to run in families

Associated symptoms

- Some people may have purposeless movements during seizures such as jerking arm or rapidly blinking eyes.
- Others have no noticeable symptoms except for brief times when they are "out of it"
- Immediately after a seizure, the person can resume whatever he or she was doing.
- Seizures may occur so frequently that the person cannot concentrate in school or other situations

Prognosis and effects

- Childhood absence epilepsy usually stops when the child reaches puberty
- Absence seizures usually have no lasting effect on intelligence or other brain functions.

Psychomotor epilepsy

- Recurrent partial seizures, especially seizures of the temporal lobe
- Psychomotor refers to strange sensations, emotions, and behavior seen with these seizures

Temporal lobe epilepsy

- Temporal lobe epilepsy is the most common epilepsy syndrome with partial seizures
- Often associated with auras
- Often begins in childhood
- Repeated temporal lobe seizures can cause the hippocampus to shrink over time
- Hippocampus is important for memory and learning

Frontal lobe epilepsy

- Cluster of short seizures with a sudden onset and termination
- There are many subtypes of frontal lobe seizures
- The symptoms depend on where in the frontal lobe the seizures occur.

Occipital lobe epilepsy

- Usually begins
  - visual hallucinations
  - rapid eye blinking or
  - other eye-related symptoms
- Otherwise, it resembles temporal or frontal lobe epilepsy

Parietal lobe epilepsy

- symptoms closely resemble those of other types of epilepsy.
- This may reflect the fact that parietal lobe seizures tend to spread to other areas of brain

Other rare types of epilepsy

Lennox- Gastaut syndrome

- begin in childhood. Children
- severe epilepsy with several different types of seizures, including atonic seizures
- This severe form of epilepsy can be very difficult to treat effectively

Rasmussen's encephalitis

- progressive type of epilepsy
- half of the brain shows continual inflammation
- It sometimes is treated with hemispherectomy

Classification of epilepsy syndromes according to effect cognitive functions or development

- ***Epilepsy syndromes that don't impair cognitive functions or development***

- are often described as benign epilepsy syndromes
- include
  - benign infantile encephalopathy
  - benign neonatal convulsions

- ***Epilepsy syndromes that impair cognitive functions or development***

- include early myoclonic encephalopathy
- Neurological and developmental problems are associated
- However, these problems may be caused by underlying neurodegenerative processes rather than by the seizures

- ***Epilepsy syndromes in which the seizures and/or the person's cognitive or motor abilities get worse over time***

- Called progressive epilepsy

When are seizures not epilepsy?

While any seizure is cause for concern, *having a seizure does not by itself mean a person has epilepsy.* First seizures, febrile seizures, no epileptic events, and eclampsia are examples of seizures that may not be associated with epilepsy.

First seizures

- Many people have a single seizure at some point in their lives.
- These seizures may occur in reaction to anesthesia or drug, but also may be unprovoked.
- These single seizures usually are not followed by additional seizures *unless*
  - the person has suffered brain damage
  - positive family history of epilepsy or other neurological abnormalities
- One recent study followed patients for an average of 8 years found that
  - Only 33% of people had a second seizure within 4 years after an initial seizure
  - People who didn't have second seizure within 4 years remained seizure- free for the rest of the study
  - People who had a second seizure, the risk of a third seizure was about 73% by the end of 4 years.

When someone has experienced a first seizure, the doctor will usually orders

1. EEG
- to determine what type of seizure
- to determine if there is any detectable abnormalities in brain waves

2. Brain scans
- to identify abnormalities that may be visible in the brain.

These tests may help the doctor decide whether or not to treat the person with antiepileptic drugs. In some cases, drug treatment after the first seizure may help prevent future seizures and epilepsy. However, the drugs also can cause detrimental side effects, so doctors prescribe them only when they feel the benefits outweigh the risks. Evidence suggests that it may be beneficial to begin anticonvulsant medication once a person has had a second seizure, as the chance of future seizures increases significantly after this occurs.

Febrile seizures

- Usually affects child during the course of an illness with a high fever.
- It is very alarming to the parents and other caregivers.
- *In the past;*
  - doctors used to prescribe a course of anticonvulsant drugs following febrile seizure for preventing epilepsy.
- *More recently, it was proved that;*
  - most children who have a febrile seizure do not develop epilepsy
  - long- term use of anticonvulsant drugs in children may damage the developing brain or cause other detrimental side effects.

- preventive treatment after a febrile seizure is generally not warranted unless
  - ▪ a family history of epilepsy
  - ▪ signs of nervous system impairment prior to the seizure
  - ▪ prolonged or complicated seizure.
- The risk of subsequent non- febrile seizures is only 2 to 3% unless one of these factors is present.

Researchers have now identified several different genes that influence the risk of febrile seizures in certain families. Studying these genes may lead to new understanding of how febrile seizures occur and perhaps point to ways of preventing them.

Non-epileptic events

Sometimes people appear to have seizures, even though their brains show no seizure activity. This type of phenomenon has various names, including nonepileptic events and pseudoseizures.

- It might be;

1. psychological in origin (psychogenic seizures).

- ▪ may indicate
- dependence - need for attention
- avoidance of stressful situations - specific psychiatric conditions
- ▪ Some people with epilepsy have psychogenic seizures in addition to epileptic seizures.
- ▪ Other people who have psychogenic seizures do not have epilepsy at all.
- ▪ Psychogenic seizures are often treated by mental health specialists.

2. Other nonepileptic events may be caused by

- narcolepsy
- Tourette syndrome
- cardiac arrhythmia
- other medical conditions with symptoms that resemble seizures

Distinguishing between true epileptic seizures and nonepileptic events can be very difficult and requires a thorough medical assessment, careful monitoring, and knowledgeable health professionals. Improvements in brain scanning and monitoring technology may improve diagnosis of nonepileptic events in the future.

Diagnosis

How is epilepsy diagnosed?

Different tests have been developed to determine whether a person has epilepsy and, if so, what kind of seizures the person has.
In some cases, people may have symptoms that look very much like a seizure but in fact are nonepileptic events caused by other disorders. Sometimes it is difficult to tell the difference between these disorders and epilepsy without close observation and intensive testing.

EEG monitoring

• EEG records brain waves detected by electrodes placed on the scalp.
• It is the most common diagnostic test for epilepsy
• Epileptic persons usually show changes in their EEG, even when they are not experiencing a seizure.
• It is very useful in diagnosing epilepsy.

Disadvantages
• Some people show normal EEG even after a seizure (as the unusual brain waves are generated deep in the brain where the EEG is unable to detect them).
• Many people who do not have epilepsy also show some unusual brain activity on an EEG.

Ideal EEG performance instructions;
• EEG should be performed within 24 hours of a patient's first seizure.
• EEGs should be performed while the patient is sleeping as well as awake.
• EEG should be performed with video monitoring
  - to determine the nature of a person's seizures.
  - to rule out other disorders such as arrhythmia or narcolepsy

Magneto Encephalogram (MEG)
• Experimental diagnostic technique used by some doctors
• Detects magnetic signals generated by neurons
• Allow doctors to monitor brain activity at different points in the brain over time
• MEG is similar in concept to EEG
• Advantages over EEG
  - does not require electrodes
  - can detect signals from deeper structures in the brain than an EEG can do

Brain scans

• One of the most important ways of diagnosing epilepsy
• Most commonly used brain scans include;
  - CT (computed tomography)
  - PET (positron emission tomography)
  - MRI (magnetic resonance imaging).

• *CT and MRI scans*
  - reveal the structure of the brain
  - useful for identifying tumors, cysts, and other structural abnormalities

• *PET and an adapted kind of MRI called functional MRI (fMRI)*
  - monitor the brain's activity
  - detect abnormalities in how brain works

• *SPECT (single photon emission computed tomography)*
  - new kind of brain scan

- used to locate seizure foci in the brain

- *Magnetic resonance spectroscopy (MRS)*
  - Experimental
  - Can detect abnormalities in brain's biochemical processes

- *Near- Infrared spectroscopy*
  - Experimental
  - Can detect oxygen levels in brain tissue.

Medical history

Taking a detailed medical history, including symptoms and duration of the seizures, is still one of the best methods available to determine if a person has epilepsy and what kind of seizures they have.

The doctor will ask questions about;
- the seizures
- any past illnesses or other symptoms a person may have had.

Since people who have suffered a seizure often do not remember what happened, caregivers' accounts of the seizure are vital to this evaluation.

Blood tests

Blood tests might be used for screening of metabolic or genetic disorders that may be associated with the seizures. They might also be used to check for underlying problems such as infections, lead poisoning, anemia, and diabetes that may be causing or triggering the seizures.

Developmental, neurological, and behavioral tests

Doctors often use tests devised to measure motor abilities, behavior, and intellectual capacity as a way to determine how the epilepsy is affecting that person. These tests also can provide clues about what kind of epilepsy the person has.

Prevention

Many cases of epilepsy can be prevented by;

1. Preventing head injuries and trauma by
- wearing seatbelts & bicycle helmets
- putting children in car seats

2. Preventing recurrent seizure activity by;
- prescribing medication after first or second seizures or febrile seizures

3. Preventing perinatal causes of seizure activity by;
- good prenatal care

- treatment of high blood pressure and infections during pregnancy

4. Prevent factors that may cause epilepsy in adulthood by;
- treating high blood pressure, infections

5. Identifying genes for neurological disorders
- provide opportunities for genetic screening and prenatal diagnosis .
- may ultimately prevent many cases of epilepsy.

Treatment

• Accurate diagnosis of the type of epilepsy is crucial for finding an effective treatment.
• Currently available treatments can control seizures in about 80% of people with epilepsy
• The remaining 20 % either have intractable seizures or get inadequate relief from available treatments.
• Once epilepsy is diagnosed, begin treatment as soon as possible before seizures and their consequences become established.

Team needed for evaluating and treating epilepsy should include
• Neurologists
• Pediatricians
• Pediatric neurologists
• Internists
• Family physicians
• Neurosurgeons

Some patients may need specialized or intensive care at large medical centers and neurology clinics at hospitals.

1. Medications

- By far the most common approach to treating epilepsy.
- There are more than 20 different antiepileptic drugs with different benefits & side effects.
- All medications aim to prevent seizures rather than treat them.
- Accordingly medications should be taken daily to reduce the risk of having seizures.

The choice of drug to prescribe, and dosage, depends on many factors, including:
- Type of seizures.
- Person's lifestyle and age.
- Frequency of the seizures.
- For a woman, the likelihood of becoming pregnant.

Medication choice; examples
- *Newly developed epilepsy;* carbamazapine, valproate, or phenytoin
- *Absence seizures;* ethosuximide is often the primary treatment.

Other medications
- Other commonly prescribed drugs include *clonazepam, phenobarbital, and primidone.*

• In recent years, a number of new drugs have become available, include;

- tiagabine - lamotrigine - gabapentin
- topiramate - levetiracetam - felbamate
- zonisamide - oxcarbazapine *(similar to carbamazapine but fewer side effects)*

• These new drugs may have advantages for many patients.

Monotherapy vs combination

• For most cases, seizures can be controlled with just one drug at the optimal dosage.
• Doctors usually prescribe monotherapy whenever possible.
• Combining medications usually amplifies side effects.
• Combinations of drugs are prescribed if monotherapy fails to control seizures.

Dosing intervals

• Determined by the drug's half- life
• Some drugs, such as phenytoin and phenobarbital, only need to be taken once a day
• Other drugs such as valproate must be taken more frequently.

Side effects

Introduction

• Antiepileptic drugs are largely safe
• Antiepileptic drugs can cause side effects but not to everyone.
• Most side effects are related to the mechanism of action of the drugs
• Side effects usually occur if the doses are increased too much or too rapidly
• Side effects are usually minimized once the dose is adjusted

Examples of side-effects

• Most side effects relatively minor; such as fatigue, dizziness, or weight gain.
• Severe and life- threatening side effects such as allergic reactions may occur rarely.
• Epilepsy medication may predispose to depression or psychoses.
• Teratogenic effects (congenital fetal malformations) in 5% of pregnant epileptic females

How to minimize possibility of side-effects

• **Patient should consult his/her doctor immediately if**

- suffered from any kind of rash while on medication
- find themselves depressed or otherwise unable to think in a rational manner.
- experience extreme fatigue, staggering, movement problems, or slurring of words.
- female patients are planning for pregnancy
- is going to undergoes any medical or surgical procedure

• People on anti-epileptic medications should consult their doctor administering other medications as epilepsy medications can interact with many other drugs in harmful ways.

Drug interactions

• Antiepileptic drugs when used in combinations may interfere with their beneficial effects and may worsen the side effects including sedation and depression.

• Drugs used for treatment of other conditions may increase or decrease the blood levels of antiepileptic drugs.
• Antiepileptic drugs increase the breakdown and accordingly the effectiveness of contraceptive pills.

Starting treatment

• *Treatment is not usually started after a first seizure EXCEPT IF*
- clearly unprovoked
- witnessed
- strong family history
- clear cut abnormality on EEG or brain scan.

• Once treatment has started the same dose should be taken daily preferably at the same time using the same brand.

Tailoring the dosage of antiepileptic drugs

People's bodies react to medications in very different and sometimes unpredictable ways, so it may take some time to find
- the **right drug**
- at the **right dose**
- to provide **optimal control of seizures**
- with **minimal or no side effects**.

• *When a person starts a new epilepsy drug, it is important to*
- tailor the dosage to achieve the best results.
- begin with low doses of the drug initially
- monitor the response and the blood levels of the drug carefully
- determine the best possible dose with best effects and minimal adverse effects

• *When a person switches between two equivalent anti-epileptic drug, it is important to consider that;*
- chemicals in generic drugs are exactly the same as in the brand- name drugs
- BUT they may be absorbed or processed differently in the body because of the way they are prepared.
- Therefore, switching to a generic version of the medication should be always considered with care.

Discontinuing medication

When to stop taking anti-epileptic medications
• **ONLY** upon doctor's instructions to do
• After long period of seizure free period (2-5 years).

Why shouldn't the patient stop the medication without Doctor's advice
• It is one of the major reasons of seizure activity recurrence
• Seizures that result from suddenly stopping medication can be very serious and can lead to status epilepticus.
• Uncontrolled seizures make it more difficult to treat the seizures in the future.

After stopping medications

- The chance that a person will eventually be able to discontinue medication varies depending on the person's age and type of epilepsy.
- More than 50% of children who go into remission with medication can eventually stop their medication without having new seizures.
- More than 60 % of adults who had been seizure- free for 2 years before stopping medication and 75 % of those who had been seizure- free for 3 years will be able to remain so.
- However, the chances of successfully stopping medication are not as good for;
  - people with a family history of epilepsy
  - people who needed multiple medications
  - people with partial seizures
  - people who continue to have abnormal EEG results while on medication.

2. Surgery

When seizures cannot be adequately controlled by medications, it might be recommended that the person be evaluated for surgery.

Considerations before surgical interference

To decide the benefit from surgery the physician should consider

1. Type of seizures

There are three categories of epilepsy that can be treated successfully with surgery.

- partial seizures
- seizures that begin as partial seizures before spreading to the rest of the brain
- unilateral multifocal epilepsy with infantile hemiplegia

2. The brain region involved & its importance for everyday behavior.

- Operating on areas of the brain that are necessary for speech, language, hearing, or other important abilities is usually avoided.
- Advanced tests might be performed to find areas of the brain that control speech and memory.

3. Patient circumstances

- Surgery should always be performed with support from rehabilitation specialists and counselors who can help the person deal with the many psychological, social, and employment issues he or she may face.

4. Possible outcomes of surgery

- While surgery can significantly reduce or even halt seizures for some people, it is important to remember that any kind of surgery carries some amount of risk.
- Surgery for epilepsy does not always successfully reduce seizures and it can result in cognitive or personality changes, even in people who are excellent candidates for surgery.

5. Medications after surgery

▪ Even when surgery completely ends a person's seizures, it is important to continue taking seizure medication for some time to give the brain time to re- adapt. Doctors generally recommend medication for 2 years after a successful operation to avoid new seizures.

Types of surgery

1. Surgery to treat underlying conditions

- In cases where seizures are caused by an organic brain disease
- e.g. brain tumor, hydrocephalus
- once the underlying condition is treated surgically, seizures will stop as well.

2. Surgery to remove a seizure focus

- The most common type of surgery for epilepsy
- It is appropriate only for partial seizures that originate in just one area of the brain.
- Have 55- 70% success rate, temporal lobe resection 70-90% success rate.

3. Multiple subpial transection

- When seizures originate in part of the brain that cannot be removed
- Surgeons perform series of cuts to prevent seizures from spreading into other parts of the brain
- 70 % of patients have satisfactory improvement in seizure control.

4. Corpus callosotomy

- Severing the network of neural connections between the right and left halves
- Done primarily in children with severe seizures that start in one half of the brain and spread to the other side.
- The procedure does not stop seizures in the side of the brain where they originate, and these partial seizures may even increase after surgery.

5. Hemispherectomy

- Removal of half of the brain's cortex
- Used only for children who have severe damage to one brain hemisphere and who also have seizures that do not respond well to medication.
- Very radical type of surgery and is performed only as a last resort
- Chance of a full recovery is best in young children
- Hemispherectomy should be performed as early in a child's life as possible.
- Should never be performed in children older than 13.

3. Devices

Vagus nerve stimulator (approved by the FDA in 1997)

- for use in people with seizures that are not well- controlled by medication.
- Battery- powered device that is surgically implanted under the skin of the chest
- Attached to the vagus nerve in the lower neck.
- This device delivers short bursts of electrical energy to the brain via the vagus nerve.
- Batteries need to be replaced every 5 years

- Battery replacement is a minor operation performed as an outpatient procedure.

Advantages
- Reduces seizures by about 20- 40 %
- Patients usually cannot stop taking epilepsy medication, but the device use
- reduces doses and subsequently reduce side effects
- reduces seizures.

4. Diet

Studies have shown that, in some cases, children may experience fewer seizures if maintain strict diet rich in fats and low in carbohydrates (ketogenic diet).
- About 25% of the children showed 90 % decrease in seizures activity
- About 50% the children have a 50% decrease in their seizures activity.
- Some children can discontinue ketogenic diet after several years and remain seizure-free.

The ketogenic diet is not easy to maintain, as it requires strict adherence to an unusual and limited range of foods. Possible side effects include retarded growth due to nutritional deficiency and a buildup of uric acid in the blood, which can lead to kidney stones.

How does epilepsy affect daily life?

- Most people with epilepsy lead normal lives.
- Approximately 80 % can be significantly helped by modern therapies
- Some may go months or years between seizures.

However, epilepsy does affect daily life for people with epilepsy and their families. People with severe seizures that resist treatment have
- shorter life expectancy
- increased risk of cognitive impairment *(particularly if seizures developed in early childhood)*

Behavior and emotions

It is not uncommon for people with epilepsy, especially children, to develop behavioral and emotional problems.
- Sometimes caused by embarrassment or frustration associated with epilepsy
- Other problems may result from bullying, teasing, or avoidance in school and other social settings.

In children, these problems can be minimized if parents encourage a positive outlook and independence, do not reward negative behavior with unusual amounts of attention, and try to stay attuned to their child's needs and feelings. Families must learn to accept and live with the seizures without blaming or resenting the affected person.

People with epilepsy have an increased risk of poor self- esteem, depression, and suicide. These problems may be a reaction to a lack of understanding or discomfort about epilepsy that may result in cruelty or avoidance by other people. Many people with epilepsy also live with an ever- present fear that they will have another seizure.

Driving and recreation

For many people with epilepsy, the risk of seizures restricts their independence, in particular the ability to drive. The risk of having a seizure- related accident decreases as the length of time since the last seizure increases.

The risk of seizures also restricts people's recreational choices. For instance, people with epilepsy should participate in sports such as swimming only with precautions and/or supervision. However, jogging, football, and many other sports are reasonably safe for a person with epilepsy.

Appropriate safety precautions should be considered. It is important to take steps to avoid potential sports- related problems such as dehydration, overexertion, and hypoglycemia, as these problems can increase the risk of seizures.

Education and employment

Significant barriers still exist for people with epilepsy in school and work. Many persons with epilepsy find it difficult to face the misunderstandings and social pressures they encounter in public situations. Antiepileptic drugs also may cause side effects that interfere with concentration and memory. Children with epilepsy may need extra time to complete schoolwork, and they sometimes may need to have instructions or other information repeated for them.

Pregnancy and motherhood

Possibility of pregnancy

• Most women with epilepsy can become pregnant despite that some seizure medications and some types of epilepsy may reduce a person's interest in sexual activity.

Chances of healthy baby

• Epileptic women have a 90 % chance of having a normal, healthy baby.
• The risk of birth defects is about5%.
• The risk of having epileptic children is about 5 % *unless the parent has a clearly hereditary form of the disorder.*

Precautions that women can take before and during pregnancy to reduce the risks

Before pregnancy

Women who are thinking about becoming pregnant should talk with their doctors prior to being pregnant to

1. Learn any special risks associated with their epilepsy
2. Know the medications they shouldn't administer during pregnancy
   valproate, trimethadione, and phenytoin, are known to increase the risk of having a child with birth defects such as cleft palate, heart problems, or finger and toe defects
3. Allow her doctor enough time to properly change medications

4. Begin vitamin supplements (especially folic acid), to reduce the risk of birth defects

Women who discover that they are pregnant but have not already spoken with their doctor should continue taking seizure medication as prescribed until that time to avoid preventable seizures.

During pregnancy

Seizures during pregnancy may lead to increased risk of
1. Miscarriage
2. Hemorrhage
3. Eclampsia
4. Premature labor
5. Cesarean section

Pregnancy may affect seizure activity through
• Decreased level of anti-epileptic drugs by
- increased blood volume leading to decreased blood level
- nausea and vomiting during pregnancy
- delayed gastric emptying affecting drug absorption

Pregnant epileptic lady show follow the following instructions
- administer prenatal vitamins
- get plenty of sleep to avoid seizures caused by sleep deprivation
- administer vitamin K supplements after 34 weeks of pregnancy
- follow good prenatal care
- avoid tobacco, caffeine, alcohol, and illegal drugs
- try to avoid stress
- consult her physician in case of change in seizure activity
- adjust dose of anti-epileptic drug guided by blood level and doctor's instructions

Precautions that should doctor follow in treating pregnant women with epilepsy
1. Close monitoring of drug blood level
2. Strict dose adjustment according to blood level
3. Frequent follow up visits

After labor
• Dose re-adjustment is needed as pre-pregnancy state is restored
• Most epilepsy medications don't influence decision about breast- feeding
• Minor amounts of epilepsy medications are secreted in breast milk; usually not enough to harm the baby.
• Rarely, baby may become excessively drowsy or feed poorly, and these problems should be closely monitored. However, experts believe the benefits of breast- feeding outweigh the risks except in these rare circumstances.
• Some epilepsy medications interfere with effectiveness of oral contraceptives. Different kind of antiepileptic medication might be used or other ways of avoiding an unplanned pregnancy might be suggested.

Status epilepticus

Severe, life- threatening condition in which a person either has
- prolonged seizures or
- does not fully regain consciousness between seizures.

Time of a prolonged seizure that must pass before a person should be diagnosed with status epilepticus is a subject of debate
- Many doctors diagnose it if a person has been in a seizure for 5 minutes
- Other doctors may diagnose status if the person had been in a seizure for 10 or 30 minutes.

While people with epilepsy are at an increased risk for status epilepticus, about 60 percent of people who develop this condition have no previous seizure history. These cases often result from tumors, trauma, or other problems that affect the brain and may threaten life themselves.

While most seizures do not require emergency medical treatment, someone with a prolonged seizure lasting more than 5 minutes may be in status epilepticus and should be taken to an emergency room immediately and be treated as soon as possible. It has been shown that 80% of people in status epilepticus receiving medication within 30 minutes of seizure onset can eventually stop having seizures, whereas only 40 % recover if treatment start 2 hours onset. Doctors in a hospital setting can treat status epilepticus with several different drugs and can undertake emergency life- saving measures, such as administering oxygen, if necessary.

People in status epilepticus do not always have severe convulsive seizures. Instead, they may have repeated or prolonged non-convulsive seizures. This type of status epilepticus may appear as a sustained episode of confusion or agitation in someone who does not ordinarily have that kind of mental impairment. The condition should still be treated as an emergency.

What to do if you see someone having a seizure?

If you see someone having a seizure with convulsions and/or loss of consciousness:

1. Roll the person on his or her side to prevent choking on any fluids or vomit.
2. Cushion the person's head.
3. Loosen any tight clothing around the neck.
4. Keep the person's airway open. If necessary, grip the person's jaw gently and tilt his or her head back.
5. Do NOT restrict the person from moving unless he or she is in danger.
6. Do NOT put anything into the person's mouth, not even medicine or liquid. These can cause choking or damage to the person's jaw, tongue, or teeth. Contrary to widespread belief, people cannot swallow their tongues during a seizure or any other time.

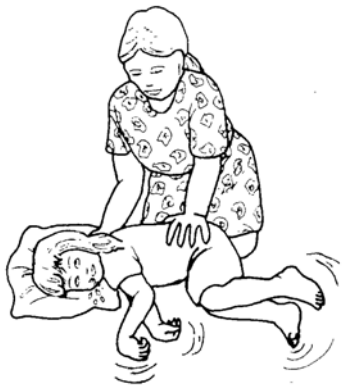


Figure (4-8): Management of seizure attack

7. Remove any sharp or solid objects that the person might hit during the seizure.
8. Note how long the seizure lasts and what symptoms occurred so you can tell a doctor or emergency personnel if necessary.
9. Stay with the person until the seizure ends.

Call ambulance if:

- The person is pregnant or has diabetes.
- The seizure happened in water.
- The seizure lasts longer than 5 minutes.
- The person does not begin breathing again and return to consciousness after the seizure stops.
- Another seizure starts before the person regains consciousness.
- The person injures himself or herself during the seizure.
- This is a first seizure or you think it might be. If in doubt, check to see if the person has a medical identification card or jewelry stating that he or she has epilepsy or a seizure disorder.

After the seizure ends, the person will probably be sleepy and tired. He or she also may have a headache and be confused or embarrassed. Be patient with the person and try to help him or her find a place to rest if he or she is tired or doesn't feel well. If necessary, offer to call a taxi, a friend, or a relative to help the person get home safely.

If you see someone having a non- convulsive seizure, remember that the person's behavior is not intentional. The person may wander aimlessly or make alarming or unusual gestures. You can help by following these guidelines:

1. Remove any dangerous objects from the area around the person or in his or her path.
2. Don't try to stop the person from wandering unless he or she is in danger.
3. Don't shake the person or shout.
4. Stay with the person until he or she is completely alert.

Chapter Summary

Motor and Neurological Disabilities

Motor disability & cerebral palsy

Definition:

Motor disability is an inability or reduced ability to perform normal human motor functions. (as standing, walking, using hands, etc…).
CP is essentially a motor disability caused by a damage of brain motor areas and frequently combined with other disabilities giving a good example of a multiple disability.

Risk factors

- Prenatal exposure to infections (toxoplasmosis, cytomegalovirus, german measles), poisoning (medications, lead), trauma, radiation,
- Premature birth, low birth weight and difficult labor leading to birth injury or hypoxia.
- Kernicterus and metabolic diseases.
- Exposure to trauma, infection or poisoning particularly in the early years after birth.

Warning signs

- Abnormal postures, non occurrence and/ or persistence of primitive reflexes..
- Deformities and/ or losses in trunk or extremities.
- Hypertonia or hypotonia.
- Significant delay in motor milestones.

Critical time for identification and screening

Identification &/ or early detection by screening should be done as early as possible to allow for starting early intervention programs to enhance motor development and prevent complications.

Suitable times for screening and identification are at birth (1st week of life), 2- 3 months, 9- 12 months, 1.5 – 2 years, 2.5 – 3 years and 5- 6 years as well as whenever a child is seen for examination or follow up. (as adopted by the screening tool developed by MOH).

Screening procedures

At the PHC Level
- Screening tools of MOH; (one previously prepared and one is currently being finalized and in field testing).

• Denver Developmental Screening Test.

Specialized testing

• INFANIB (A 20 items, 100 score tool for early detection of motor disability in the first year of life).
• Complete comprehensive neuromotor examination.

Management and rehabilitation guidelines

• Reach a diagnostic formulation describing the main manifestations, how they affect the child development, the possible causes & how persistent they are, the accurate diagnosis if possible and more importantly weather the condition is progressive or not.
• Use investigations (EEG, X ray, CT, MRI, genetic and metabolic tests) only when justified (not empirically).
• Use medicines only to produce a specific wanted effect that will probably improve the condition within a framework of an integrated rehabilitation programme (not empirically).
• Refer to more specialized centres for diagnosis and/ or intervention rehabilitation programmes.
• Follow up your children with disabilities as frequent as possible/ as needed understanding what programmes are given to them, giving feed back on their progress and caring for their general health, growth and development.
• Children with motor disabilities might need physiotherapy, occupational therapy, medical treatment, surgical interventions and assistive devices. They may need other services as speech, skill development, sensory stimulation and behavior management programmes if they suffer from accompanying difficulties.

Suggested referral places

• National Institute for Neuromotor Rehabilitation (Institute of Polio).
• Neurology and Rehabilitation departments of University hospitals.
• Faculty of Physical Therapy.
• Integrated Care Centre for Children with Special Needs; Zaitoun.
• Early Intervention & Rehabilitation Centre, Helwan.
• See also the guide in " My Child, Disability and the Future"

Epilepsy

Definition

Epilepsy is a neurological brain disorder in which clusters of neurons sometimes signal abnormally, leading to a tendency to have recurrent seizures in the form of strange sensations, emotions, and behavior, or sometimes convulsions, muscle spasms, and loss of consciousness.

Risk factors

• Familial tendency (genetic factors)

• Brain insults due to prenatal and/or postnatal factors, (ischaemia, hypoxia, intoxication, metabolic errors, trauma and developmental problems).

Warning signs

• A first seizure.
• Febrile seizures.

Critical time for identification and screening

Recurrence of febrile seizures, a first definite seizure or a second seizure.

Screening procedures

No screening is currently recommended.

Management and rehabilitation guidelines

• Diagnose accurately epilepsy and its type.
• Use EEG for diagnosis and monitoring.
• Start medical treatment as early as possible.
• Start treatment with carbamazapine, valproate, or phenytoin first, unless the epilepsy is a type that is known to require a different kind of treatment.
• Adjust dosage according to age, weight and response guided with serum drug level and EEG monitoring.
• Use ethosuximide for absence seizures.
• Use clonazepam, phenobarbital, and primidone as a second raw drugs.
• Use a combination therapy only for intractable seizures that do not respond to monotherapy.
• Inform patients (or families) about side effects including; skin rash, fatigue, dizziness, minor depression, attention and thinking difficulties and ask them to report to you if they complain of any of them.
• Discontinue drug therapy gradually after two or better more years passing without seizures.
• Refer to neurosurgeons patients who have tried two or three different medications without success, or those who have identifiable brain lesions believed to cause the seizures.
• For pregnant women switch treatment from valproate, trimethadione, and phenytoin that are known to increase the risk of having a child with birth defects to other drugs. Inform your women patients that they better allow for enough time to properly change medications, including phasing in the new medications and checking to determine when blood levels are stabilized before trying to become pregnant and that they should continue antenatal care and probably get supplementary vitamins especially folic acid throughout pregnancy.
• Refer cases of status epilepticus continuing for over 5 minutes to emergency units.
Attend to patients having seizures as an emergency state as described in the text.

Suggested referral places

• Cairo, Ain Shams & Al- Azhar University Hospitals (Pediatrics or Neurology departments)
• National Institute for Neuromotor Rehabilitation (Institute of Polio).

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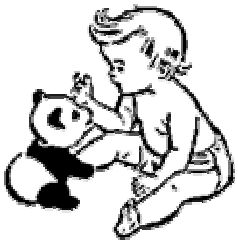
Chapter 5



Visual Disability

By the end of this chapter the PHC physician should be able to:

- Recognize the definition of visual disability, its classification and its status in Egypt.
- Recognize and describe causes and risk factors of the visual disability.
- Recognize important preventive measures of visual disability.
- Recognize and describe normal visual development.
- Make a provisional diagnosis of the visual disability according to the categories outlined.
- Describe essential management procedures and care for the most important health problems.
- To identify appropriate referral needs not fulfilled at the primary care level including medical, social and psychological.

Visual Disability

Definition

Based on visual acuity in the better-corrected eye, visual disability (impairment) is classified into the following categories:

Category	Visual acuity in the better-corrected eye
Low vision	Less than 6/18 but better than 6/60
Severe low vision	Less than 6/60 but better than 3/60
Blind	Less than 3/60

The above definition is based solely on visual acuity. However, visual impairment is a functional state rather than a numerical expression of visual acuity. The WHO set a more comprehensive and working definition that is:

> *"A person with visual impairment is one who has impairment of visual functioning even after treatment and/or standard refractive correction, and has a visual acuity in the better eye of less than (6/18 for low vision and 3/60 for blind), or a visual field of less than 10 degrees from the point of fixation, but who uses or is potentially able to use vision for the planning and/or execution of a task."*

Prevalence and burden of visual disability (globally and in Egypt)

Global burden

- There are 45 million blind people and 135 million with low vision, comprising of a total of 180 million people with significantly poor vision in the world today. These figures are likely to double over the next 20 years unless decisive action is taken now.
- 80% of blindness is avoidable (either treatable or preventable)
- 90% of the world's blind live in developing countries.
- Eye care is one of the most cost effective interventions of health care.

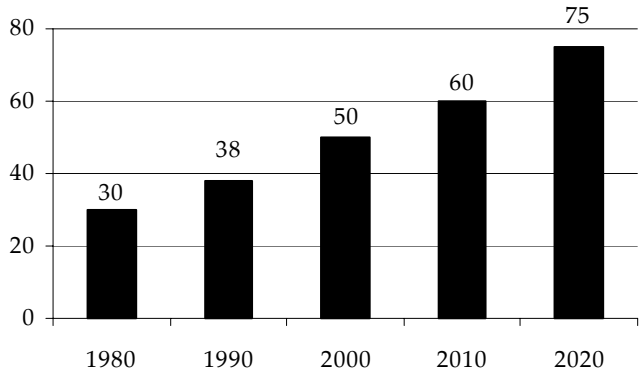


Figure (5-1): Global projections of visual disability

Burden in Egypt

- The prevalence of low vision for all ages is 3.1%, that is nearly equal to 2.2 million persons.
- The prevalence of blindness is 1.1%, that is nearly equal to 737,000 blind persons.
- Every year, an additional 15,000 Egyptians go blind.
- More than two-thirds of this blindness is treatable and preventable.
- Without proper interventions the number of blind in Egypt will increase to over one million by 2020.
- Annual GDP loss from blindness and low vision in Egypt is over $170

Causes / risk factors

Global causes

The leading causes for blindness for all age groups are as follows:

Cause	% From all causes
Cataract	50
Trachoma	16
Childhood blindness	3.1
Onchocerciasis	0.9
Others*	30

** Others include Glaucoma, Diabetic retinopathy, Trauma, Macular degeneration*

Major causes in Egypt

Major causes for blindness among age group 50+ are:

Cause	% From all causes
Cataract	54.8
Corneal opacity other than trachoma	18.8
Corneal opacity due to trachoma	7.6
Refractive error	7.0
Glaucoma	4.6
Others	7.2

List of causes by age group

In preschool children:

1. Prenatal phase

- Congenital disorders as cataract of genetic origin, glaucoma and retinoplastoma.
- Congenital cataract of infective origin (rubella)

2. Neonatal phase

- Retinopathy of premature
- Birth injuries and asphyxia
- Ophthalmia neonatorum
- Strabismus and/or amblyopia

3. Childhood phase

- Refractive errors.
- Vitamin A deficiency
- Measles
- Harmful eye practices

4. In adolescents

- Ocular trauma
- Corneal opacities either due to trachoma, other infective or traumatic causes.

Details of causes and burden by age group

Prenatal phase

- Congenital disorders as cataract of genetic origin, congenital glaucoma and retinoblastoma are not common. Consanguinity is associated with a greater risk of the congenital disorders.
- Congenital cataract of infective origin (rubella) has a relatively higher incidence in developing countries.

Neonatal phase

- Retinopathy of premature:
  - Recent studies have indicated an increasing incidence despite careful monitoring of oxygen use.
  - This is attributed to the increased survival rate of the immature infants.
  - The lower the weight and gestational age at birth, the higher the incidence.
- Birth injuries and asphyxia: Recent evidence suggests that this is no longer a common cause of childhood blindness with improving the labor and neonatal care.
- Ophthalmia neonatorum:
  - The incidence is low when good antenatal care is available.
  - However, it's an ocular emergency due to its aggressive ocular sequelae.

Childhood phase

- Vitamin A deficiency:
  - The major global cause of childhood blindness.
  - It causes corneal ulceration and opacity that ends with permanent blindness.
  - Worldwide, some 350 000 new cases occur annually among preschool children, and an estimated 60% of them die within a year of becoming blind.
  - No available data on the burden in Egypt.
- Trachoma:
  - The active stage prevails among preschool children and with repeated infections could lead later to corneal opacity and blindness in adults.
  - Data from recent surveys in Egypt have these following estimates for the prevalence of active trachoma among children of age group 1-6:

Survey site and date	Prevalence %
Menofiya, 1999	36.5
Menia, 2001	42.0
Fayoum, 2003	47.0

- Measles:
  - It's a severe infection in most developing countries with a fatality rate of more than 1%.
  - Blindness in children is due to corneal scarring.
- Harmful eye practices:
  - They appear to be of greater significance in developing countries.
  - Studies in East Africa attributed 20-25% of childhood blindness to the use of harmful eye medicines.

Preschool children

- Amblyopia and strabismus: many children suffer from amblyopia ("lazy eye"; loss of vision due to disuse) and strabismus (ocular misalignment), which, aside from congenital conditions, usually develop between infancy and ages 5-7.
- In the newborn, risk factors for developing strabismus or amblyopia include:
  - A family history of ocular malformations,
  - Anisometropia (a large difference in refractive power between the two eyes, more than 4 diopters in sphere and/or 2 diopters in astigmatism),
  - Congenital cataracts,
  - Ocular tumors,
  - Premature birth, or
  - Birth to a mother who suffered from infection such as rubella, genital herpes, or toxoplasmosis during pregnancy.
- Since normal vision from birth is necessary for normal binocular development, failure to detect and treat amblyopia, marked anisometropia, or strabismus at an early age may result in irreversible visual deficits.
- Resulting permanent amblyopia and cosmetic defects may lead to later restrictions in educational and occupational opportunities.
- Patients with amblyopia are at increased risk of blindness from loss of vision in their good eye.

School-aged children

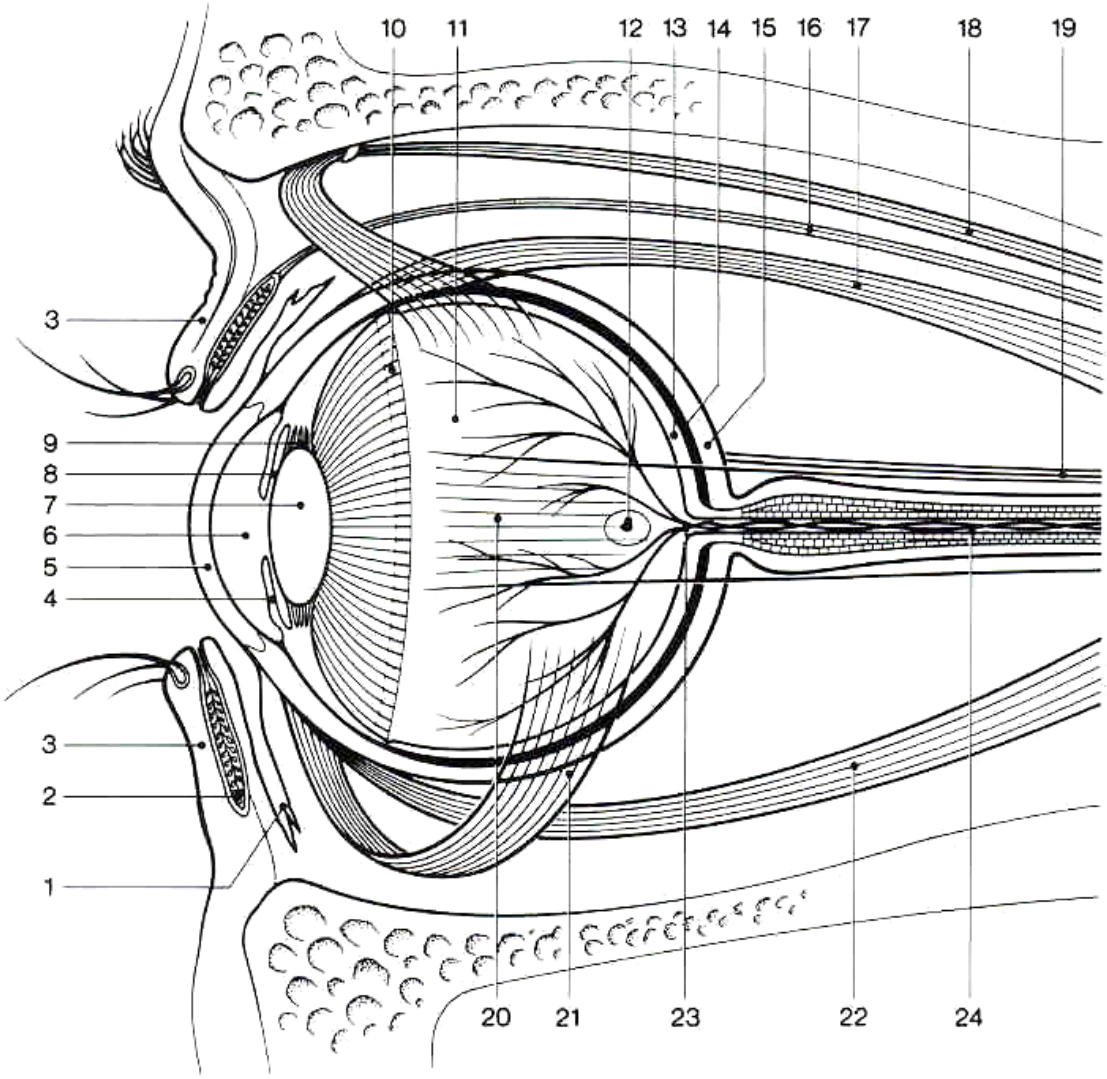
- Refractive errors, which often become manifest during school age, would carry prognostic implications. Myopia, hypermetropia are the main refreactive errors. (mentioned before in the section of eye functions)

Adolescents

- Refractive errors are the most common visual disorder in the adolescent and adult population.
- Corneal opacities either due to infective or traumatic causes constitute the third leading cause (16.4 % from all causes) for low vision among adults of age group 50+.
- For the same age group corneal opacities constitute the second leading cause for blindness (26.4 % from all causes).

Basic eye anatomy and function

Our eye



1. Conjunctiva
2. Sebaceous gland In the eyelid
3. Eyelids
4. Iris
5. Cornea
6. Anterior chamber of the eye
7. Lens
8. Posterior chamber of the eye
9. Zonule, the suspension apparatus of lens
10. Anterior edge of retina
11. Vitreous body
12. Yellow spot of the retina
13. Retina
14. Choroid
15. Sclera
16. Levator muscle of the upper eyelid
17. Superior rectus eye muscle
18. Superior oblique eye muscle
19. Medial rectus eye muscle
20. Lateral rectus eye muscle
21. Inferior oblique eye muscle
22. Inferior rectus eye muscle
23. Blind spot (entry of the optic nerve)
24. Optic nerve

Figure (5-2): Anatomy of the eye

How do we see?

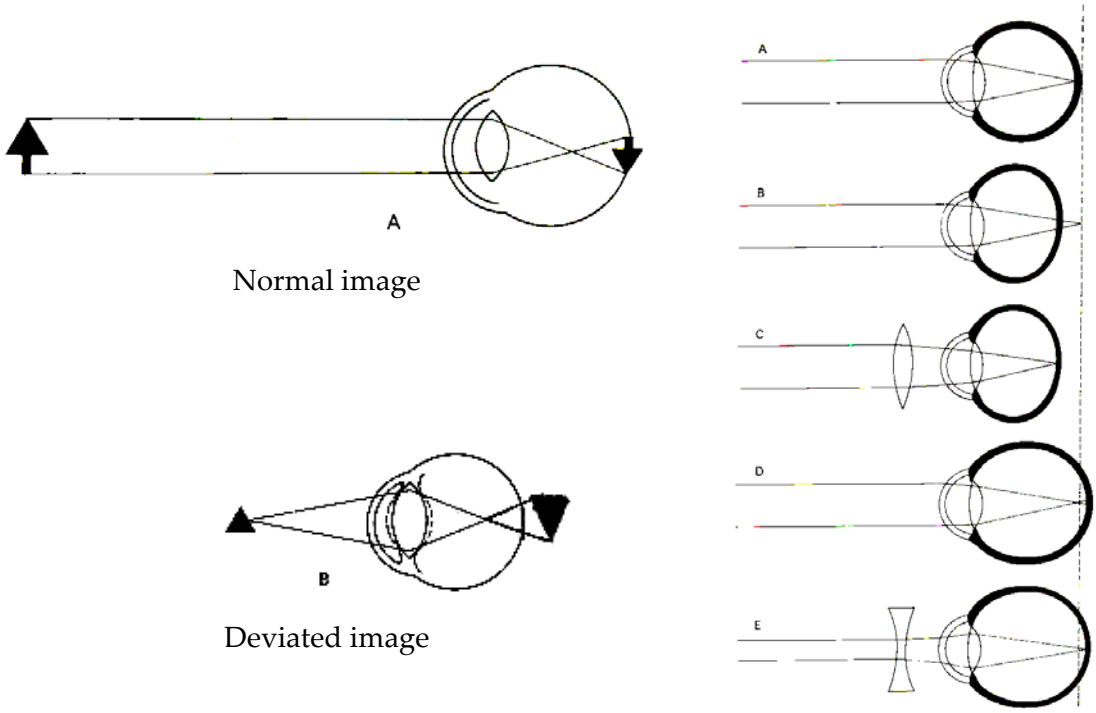


Figure (5-3): Normal and deviated images

For the normal sight, the image should be focused on the retina. The main power for focusing is performed by the cornea and lens.

Inability to bring the focused image exactly on the retina will lead to blurred image and a state of error in refraction. Myopia or short sight when the image is formed in front of the retina, and hyermetropia or long sight if it is formed behind the retina. These errors should be corrected by adding glasses that matches with the type of the error. Positive convex glasses for hypermetropia and negative concave glasses for myopia.

Landmarks for the development of eye and vision

The eye

- The pediatric eye is a growing eye, which differentiates it in many ways from the adult eye.
- The corneas of the infants appear large relative to the rest of the face. The infant sclera is about half of its adult thickness and strength.
- By birth, the diameter of the eye is about 66% of that in adulthood. The eye grows rapidly during the first 2 years of life; growth then slows until puberty.
- By birth, pupils are constricted (meiotic) because the dilator pupillae muscle is not well developed.
- Newborn infants cry without tears as the lacrimal gland (reflex) tear production does not begin until 20 or more days after birth.
- By birth, 75% of the growth of the optic nerve and disc occurred.
- Before 1 year of age, 90% of it occurred.
- Premature babies of body weight less than 1500 gm or 32 weeks gestation have peripheral retina still underdevelopment. They are at a higher risk of developing retinopathy.
- Within the first year of life, the darkening of the color of the eye increases. That is because additional pigment continues to be deposited in the anterior iris for approximately a year after birth.

The vision

- By birth, visual pathway is complete and images for different objects could be formed and perceived at visual center.
- Immediately after birth, infants see and look around. However, they still couldn't recognize or understand the images.
- Also development of eye muscles is complete and eye movements should be intact.
- During the first few months, shifting sight gaze towards light is elicited and babies start to recognize the mother and / or familiar persons by seeing as revealed by the facial expression.
- At the age of 3 months the infant starts to play with his/her hands.
- By 6 months, fixation reflex is intact and infant could fix sight gaze upon definite objects.
- During the first few months, infants become skilled users of their vision and develop eye-hand coordination. Both eyes normally look at the same object. The infant could identify certain objects by seeing, fixate on them and move the hand to catch them.
- During the first few years as the axial length for the eye increases the lens flatten. Myopic errors increase while hypermetropic errors decrease with age.
- It is important to diagnose a lazy eye early and to teach the infant to look with it. Otherwise that eye will be amblyopic and never see well.

Assessment of visual disability

A. Preschool children

Children under 3 years

• Early identification of children with visual impairment has important implications as it can prevent or minimize the developmental delay that may follow the visual impairment and give the child the chance to learn how to compensate or adapt.

• Patterns of presentation:
- An ocular abnormality is noticed during routine postnatal screening.
- Abnormal visual behavior.
- Child complains of symptoms.

• Patterns of detection:
- Children with obvious structural ocular abnormalities or nystagmus are likely to be identified early, although delay in referral to the appropriate specialist may jeopardize the outcome of visual rehabilitation.
- Children with severe visual loss, but without obvious external structural abnormalities are likely to present early because of abnormal visual behavior.

Children 3-5-year-old age

Screening to assess the distance visual acuity, near visual acuity and visual function should be done

• **Assessment of distance visual acuity**
Although the following tests do not reflect the resolving power of the eye, i.e. visual acuity, they can indicate whether an abnormality is present or not:
- Pupillary reactions to light.
- Head turn towards a light source.
- Blink response to a bright light.

• **Assessment of near vision**
- The Illiterate E or the Landolt C is used, as they require naming or matching of letters with independency to reading skills.

• **Assessment of functional vision**
- When children are not able to be assessed using distance and near acuity tests, then an assessment of functional vision should be carried out to obtain qualitative data on the level and use of vision.

- The aim is to assess:
▪ The ability of fixation (the ability of the eyes to fixate a target and maintain steady fixation),

▪ Eye movements (tracking; the ability to follow moving objects),
▪ Shifting gaze from one object to another (scanning to find objects).

- It should be performed with both eyes together, and with spectacles if these are normally worn.
- Observation and comment should be done for these following capacities;

▪ *Child can see to walk around:*
1. Place two chairs 1 meter apart in a well-lit room.
2. Ask the child to walk in and out of, or between the obstacles without assistance.

▪ *Child can recognize faces:*
Is the child able to identify a person known to him e.g. a teacher or a relative by visual recognition of the face alone at a distance of 3 meters?

▪ *Child can see print:*
1. Clearly draw a cross, square or circle approximately 2 cm in size.
2. Ask the child to describe the shape or draw it.
This test can be performed at any distance within ½ meter.

Repeated testing over time with the previous tools may be necessary to assess the level of vision.

Screening tests for detecting strabismus and amblyopia include:

- Simple inspection,
- Cover test,
- Visual acuity tests, and
- Stereovision assessment.

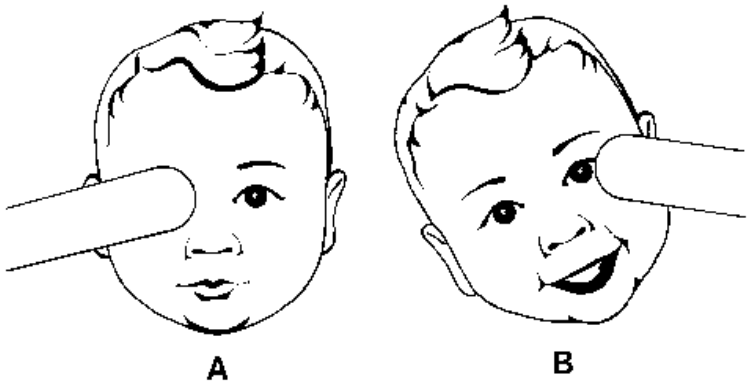


Figure (5-4) Cover test

• Testing for visual acuity should include testing visual acuity with pinhole to detect the correctable refractive errors.
• Screening for active trachoma by everting of the upper lid and examining for follicles is very simple and effective in identifying active cases. Although it's not a blinding disease in children, however it's a potentially blinding.
• The same previous test is valuable in detecting active cases with spring catarrh and allergic conjunctivitis that prevailed in hot dry climates as in Egypt. It could interfere with proper vision due to induced allergy and astigmatism.

- External eye examination using a light source and /or simple magnifier can verify the congenital cataract as a lenticular opacity that interferes with red reflex and pupilary reflex.
- The same examination could verify congenital glaucoma with abnormal increased corneal diameters, epiphora and photophobia.

B. School-aged children

- Screening of visual acuity is generally accomplished with standard Snellen vision charts.

- Near and distance vision are not always affected to the same degree in all eye conditions. In children, near vision is often not as severely affected as distance vision.

- Tests of visual acuity and visual field do not show how a person can use his/her vision. It gives a starting point in the assessment of visual impairment.

- Visual acuity can be used to identify children who have low vision or blindness and who need a further examination to see if their eye conditions are curable. If the condition is not curable, the effect of low vision should be assessed.

Techniques for visual acuity tests

Distance visual acuity test

- Visual loss is categorized according to the International Classification of Disease (ICD) as listed before.

Category	*Visual acuity*
Visual impairment	Less than 6/18 but better or equal to 6/60
Severe visual impairment	Less than 6/60 but better or equal to 3/60
Blind	Less than 3/60

- Check that you have the best lighting available during testing. The normal test distance is 6 meters.
- The vision should be tested separately for each eye and then with both eyes.
- The criteria for vision at a certain level are: 4 correct consecutive showings or 5 correct out of 6 showings or 6 correct out of 8 showings.

Pinhole test

- A special test using the pinhole mask should be considered to identify those with poor vision who may need spectacles to improve their vision.
- It should be used when distance visual acuity is less than 6/18.
- It is performed using a simple mask that has multiple tiny holes to allow examined person to look through at the test chart.

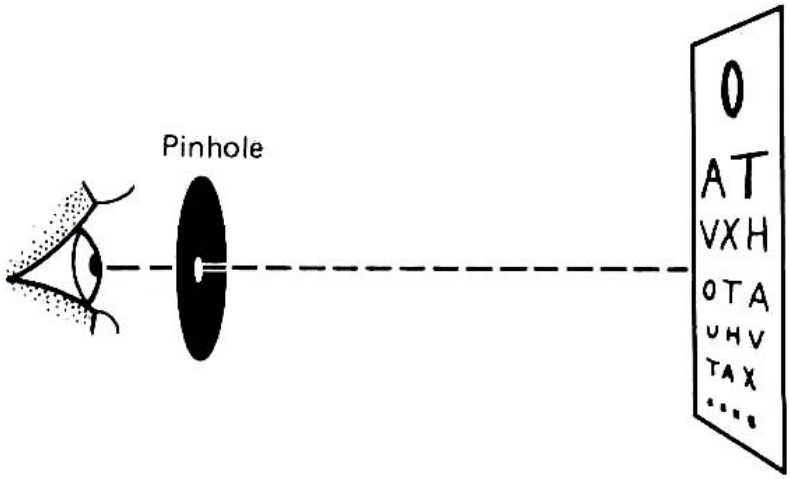


Figure (5-5): Pin hole test: If visual acuity improves to ≥ 6/18 = Glasses

• People who have their visual acuity improved with the pinhole should be referred for prescription of glasses.

Test for near vision

• Near vision tests show a person's ability to discriminate the details of near object (within arm's distance from the body)

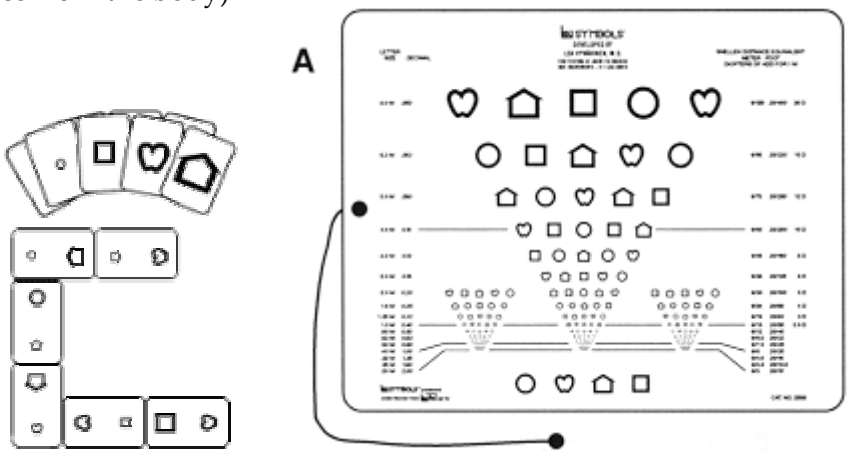


Figure (5-6): Tests for near vision

• Children who cannot recognize the smallest symbols should be referred to check if spectacles or low vision devices might be helpful.

Testing instructions

• No standard distance is required.
• The test card is held at the distance preferred by the person.
• Young children may see better when the card is held close to the eyes.
• Start with the largest Es. If a person cannot see these, tell him or her to hold the card closer to his or her eyes.

Landmarks for screening

Age group	Sign	Test
First week	• Premature baby with - body weight < 1500 gm - or gestation < 32 weeks.	Frequent fundus examination.
	• The normal shape and structure of the eye.	External Examination
	• Light (Pupillary) reflex	Test for pupillary reflex
	• Red reflex	Test for red reflex
Up to 8 to 10 weeks	• Head and eye turn to light source.	Test by light source
	• Normal eye movements	Test for eye movements
6 months	• Fixation reflex	Test for fixation reflex
	• Conjugate eye movements with the two eyes.	Test for eye movements
	• Patent lacrimal orifices	External examination
18 month	• Corneal luster and transparency	Corneal stain particularly for children with measles, recurrent respiratory tract infections, severe diarrhea or malnutrition.
Up to 3 years	• Shifting gaze from near to far objects. • Discrimination of objects. • Imitation of movements.	Tests for near and far vision
	• Vitamin A deficiency • Night blindness • Corneal luster and transparency	External examination
	• Frequent eye infections particularly trachoma	External examination with inspection for upper lid.
	• Frequent lacrimation / itching and redness	External examination
Up to 6 years	• Visual acuity for far and near vision (For each eye separately)	Test for visual acuity for far and near
	• Conjugate eye movements	Cover test
Primary school	• Errors of refraction • Eye Trauma	Tests for visual acuity
Preparatory and secondary school	• Lid deformity and trichiasis	Inspection of upper lid

How to perform basic eye tests for screening?

Tool	Procedure	Output
Body weight for baby		
Baby weighing		All babies of B.W. > 2000 gm.
External Examination		
• **Naked eye or simple magnifier (X 2.5)** • **Torch**	• Approach the child's eye either by your naked eye or wearing a simple magnifier. • Approach the torch to the child's eye from laterally and shed the light on the corneal surface.	• Clear and transparent cornea. The shadow of the torch on the corneal surface is pointed and clear. • Clear conjunctiva. • Normal eye lid.
Pupillary reflex		
• **Pointed light source (torch)** • **Naked eye or simple magnifier (X 2.5).**	• In dim light. Approach the torch to the pupil of one eye slightly from peripheral and observe the movement of the pupil. • Remove the torch away from the eye rapidly and observe the pupilary reaction. • Repeat the same while observing the reaction of the other pupil (indirect reaction). *As the test is simple, fast and not much annoying for the baby, it could be repeated to assure the output.*	• Pupil is round regular and reacts both directly and indirectly to light. • Pupil constricts and dilates on approaching and removing light to and from the eye respectively.
Red reflex		
• **Lamp preferably with beam light.** • **Concave mirror with a central hole or pocket retinoscope or direct ophthalmoscope**	• In a dark room. • No need to dilate the child's pupil. • Place the child in a way that light comes laterally from behind the child's head. • Sit one meter away facing the child. • Hold the mirror in your hand facing the light and reflecting it onto the child's right or left eye. • Look from the central hole with one of your eyes on the child's eye that faces the mirror. • Observe the color of the reflected light and any structures might be lying in the pass of the light. • Repeat the same test for the other eye after adjusting the site of the light source. • In case of availability of retinoscope or direct ophthalmoscope, use the light of the retinoscope or ophthalmoscope instead of the ordinary light source and the mirror.	• The light is reflected in red color. (as it's reflected normally from the retinal blood vessels). • The pupil is round regular and there are no structures obscuring the pass of light.
Corneal stain		
• **Flurocein strip papers** • **Torch** • **Liquifilm eye drops or saline solution 0.9 %**	• Place the tip of the fluorecein paper in the lower conjunctivaal fornix and leave it there for 5 seconds. • Wash the eye gently by the eye	• Normal corneal surface is smooth, transparent, intact and continuously washed by tears. It'd not be stained by any stain.

Tool	Procedure	Output
• **Magnifier (X 2.5)**	drops. • Observe the corneal surface using magnifier (X 2.5)	• Abrasions, wounds or ulcers will be stained by the green color of fluorecein dye and will be easily recognized on examination.
Cover test		
• **Hard cover to be placed ulternatively in front of each eye.** • **Torch**	• Face the child one meter away with your eyes in the same level of the child's eye. • Hold your index finger and ask the child to fix at it. • Place the cover in front of the child's right eye. • Remove the cover and observe any change in the direction of the eye that was covered.	• The eye keeps fixed on the fixation point and does not change its direction on removing the cover. • In latent squint, after covering the eye it could not keep fixation and deviates to one side. • On removing the cover the eye starts to fix again and re deviate to the opposite side.
Test for eye movements		
Pointer or index	• Test each eye separately first then the two eyes together. • Face the child one meter away with your eyes lying in the same level of the child's eye. • Move the pointer in the main eight directions in front of each eye and ask the child to follow the pointer by his or her corresponding eye without moving the head. • Observe the movement of the eye in all the defined directions. • Bring the pointer from far to near (up to 25 cm) from the child' eyes and observe its movement.	• Each eye moves smoothly and freely in the identified directions. • Both eyes move in conjugate movement in the identified directions. • Any deviation in movements of the eye will be viewed in manifest squint.

Tests for the basic assessment of the eye and vision

• Those tests are selected out of a wider group mentioned within the text in section (7). Landmarks for screening) considering those tests that could be performed within the age group (from birth up to six years old).

• The aim of this schedule is to allow for early detection and referral of cases suspected to have any sort of visual impairment within that age group.

• Details for how to perform each test are mentioned within the text in section (8. How to perform basic eye tests for screening).

• For any negative finding in any test, children should be referred to the eye doctor (ophthalmologist) for further assessment.

Table (5-1): Tests for the basic assessment of the eye and vision

Test	Possible finding
First two months	
External examination for the eye	• Abnormal shape, size or structure of the eye
Pupillary (light) reflex	• Abnormal retina, optic nerve or visual pathway
Third month	
Eye moves to direction of the light.	• Abnormal visual pathway • Delayed development of vision
Red reflex is clear (light reflected from inside the eye is red in color and round in size)	• Congenital cataract • Abnormal vitreous and / or retina • Retinoplastoma
Sixth month	
Eye could fixate on certain near object	• Delayed development of vision • Squint • Nystagmus
First year	
Recognizing small objects at a near distance (30 cm)	• Low vision • Long sighted and /or astigmatism
Eye movements are intact and synchronize.	• Squint • Nystagmus
Up to the second year	
Follow moving objects (e.g. rolling ball)	• Delayed development of vision
Up to the third year	
Cornea is clear and transparent	• Trauma • Infections • Vitamin A deficiency
Identifying tiny objects at a near distance (30 cm)	• Delayed development of vision • Low vision
Up to the sixth year	
Matching objects similar in size, color and shape	• Delayed development of vision • Low vision
Far vision. Identifying the types of the visual acuity chart	• Abnormal long or short sight and / or astigmatism

Prevention of visual disability

The main preventable causes for visual impairment are:

- Trachoma
- Ocular infections
- Vitamin A deficiency
- Ocular trauma
- Harmful eye medications

In Egypt, Trachoma and corneal opacities due to infections and trauma have higher prevalence.

As preventive measures are so effective in eliminating of the preventable causes of visual impairment, a full data is listed here in the following table to highlight the basic preventive and primary curative measures. Causes are listed in the table according to age of onset to simplify the reading. However the priority for the above mentioned group of causes should not be overlooked.

Primary	Secondary	Tertiary
Prevention of prenatal causes		
Congenital cataract & congenital glaucoma of genetic origin		
• Genetic counseling. • Avoidance of consanguineous marriage.	• Early diagnosis. • Early surgical treatment.	• Low vision care for those with poor visual outcome.
Congenital cataract of infective origin (rubella)		
• Rubella immunization.	• Early diagnosis. • Early surgical treatment.	• Low vision care for those with poor visual outcome.
Retinoblastoma		
• Genetic counseling.	• Early detection.	• Low vision care.
Other prenatal determined conditions		
• Education regarding use of drugs, smoking, and alcohol in pregnancy.	• Early detection	• Low vision care.
Prevention of neonatal causes		
Ophthalmia neonatorum		
• Recognition and treatment of maternal infection • Cleansing of lid margins and application of prophylactic tetracycline 1% eye ointment.	• Early diagnosis • Ceftriaxone, 50 mg/kg of body weight intramuscularly as a single dose.	• Surgical treatment for corneal opacities.
Retinopathy of premature		
• Measure to prevent low birth weight / premature birth. • Monitoring of oxygen therapy.	• If oxygen treatment given without monitoring: • Screen all babies < 2000 g, < 36 weeks.	• Treatment of complications (glaucoma, retinal detachment) • Low vision services
Birth asphyxia		
• Improve obstetric care.	• Early detection of visual disturbance in anoxic infants.	• Low vision care.
Prevention of childhood causes of blindness		
Vitamin A deficiency		
• Increase maternal vitamin A supply, encourage breast-feeding and proper weaning. • Prevent and control measles, diarrheal diseases, acute respiratory infections, and protein-energy malnutrition. • Vitamin A supplements.	• Identify and treat signs and symptoms of vitamin A deficiency. • For children over one year: Immediately on diagnosis 200 000 IU of vit.A orally. Repeat the same dose the	• Low vision care.

Primary	Secondary	Tertiary
	next day and 4 weeks later. • For children under one year or less than 8 Kg: Treat with half the above dose.	
Measles		
• Promote and provide measles immunization.	• Vitamin A (200 000 IU) and topical antibiotic particularly when cornea is affected.	
Harmful eye practices		
• Health education • Community awareness • Primary eye care facilities	• Appropriate treatment of ocular complications.	
Trachoma		
• Improved community and individual hygiene, including regular face washing	• Mass treatment with topical tetracycline 1% twice daily for 6 weeks in endemic areas. • Treatment with a single dose of Zithromax based on body weight. 25 mg/kg. • Surgical correction of trichiasis (uncommon in childhood)	
Ocular trauma		
• Legislative measures e.g. seat belts, fireworks. • Public health education	• Early detection and primary treatment • Speedy referral for definite treatment.	

Key points for doctors and parents

• Vision screening to detect amblyopia and strabismus is recommended once for all children prior to entering school, preferably between ages 3 and 4.

• Clinicians should be alert for signs of ocular misalignment when examining infants and children. If an infant before the age of 6 months seems to have crossed eyes (squint) all the time, he/she should be seen by an ophthalmologist soon not after several weeks. An intermittent crossing of eyes in children older than 6 months should be examined by ophthalmologist.

• Signs for congenital causes of visual impairment as cataract and glaucoma should not be neglected as early detection is crucial in visual prognosis.

• Signs for corneal opacities that interfere with visual axis should be considered when examining infants, children and adults.

• Regulations should be made for combating the spread of active stages of trachoma and its blinding sequences. Particularly clinicians at the rural health units in Egypt should be alert with its signs and management.

• Screening for diminished visual acuity with Snellen visual acuity chart and pinhole test is recommended for elderly persons.

• Screening for diabetic retinopathy should be considered for all diabetics.

• Screening for glaucoma should be considered for those with age 40+ and have positive family history for glaucoma.

Chapter Summary

Visual Disability

Definition

A person with visual impairment is one who has impairment of visual functioning even after treatment and/or standard refractive correction, and has a visual acuity in the better eye of less than (6/18 for low vision and 3/60 for blind), or a visual field of less than 10 degrees from the point of fixation, but who uses or is potentially able to use vision for the planning and/or execution of a task.

Major causes / risk Factors

Globally the leading causes for blindness for all age groups are as follows:
- Cataract
- Trachoma
- Childhood blindness
- Onchocerciasis
- Others: Glaucoma, Diabetic Retinopathy, Trauma, Macular degeneration

Warning signs and needed screening procedures

Age group	Sign	Test
First week	• Premature baby with - body weight < 1500 gm - or gestation < 32 weeks.	Frequent fundus examination.
	• The normal shape and structure of the eye.	External Examination
	• Light (Pupillary) reflex	Test for pupillary reflex
	• Red reflex	Test for red reflex
Up to 8 to 10 weeks	• Head and eye turn to light source.	Test by light source
	• Normal eye movements	Test for eye movements
6 months	• Fixation reflex	Test for fixation reflex
	• Conjugate eye movements with the two eyes.	Test for eye movements
	• Patent lacrimal orifices	External examination
18 month	• Corneal luster and transparency	Corneal stain particularly for children with measles, recurrent respiratory tract infections, severe diarrhea or malnutrition.
Up to 3 years	• Shifting gaze from near to far objects. • Discrimination of objects. • Imitation of movements.	Tests for near and far vision

Age group	Sign	Test
	• Vitamin A deficiency • Night blindness • Corneal luster and transparency	External examination
	• Frequent eye infections particularly trachoma	External examination with inspection for upper lid.
	• Frequent lacrimation / itching and redness	External examination
Up to 6 years	• Visual acuity for far and near vision (For each eye separately)	Test for visual acuity for far and near
	• Conjugate eye movements	Cover test
Primary school	• Errors of refraction • Eye Trauma	Tests for visual acuity
Preparatory and secondary school	• Lid deformity and trichiasis	Inspection of upper lid

Management guidelines

• Vision screening to detect amblyopia and strabismus is recommended once for all children prior to entering school, preferably between ages 3 and 4.
• Clinicians should be alert for signs of ocular misalignment when examining infants and children. If an infant before the age of 6 months seems to have crossed eyes (squint) all the time, he/she should be seen by an ophthalmologist soon not after several weeks. An intermittent crossing of eyes in children older than 6 months should be examined by ophthalmologist.
• Signs for congenital causes of visual impairment as cataract and glaucoma should not be neglected, as early detection is crucial in visual prognosis.
• Signs for corneal opacities that interfere with visual axis should be considered when examining infants, children and adults.
• Regulations should be made for combating the spread of active stages of trachoma and its blinding sequences. Particularly clinicians at the rural health units in Egypt should be alert with its signs and management.
• Screening for diminished visual acuity with Snellen visual acuity chart and pinhole test is recommended for elderly persons.
• Screening for diabetic retinopathy should be considered for all diabetics.
• Screening for glaucoma should be considered for those with age 40+ and have positive family history for glaucoma.

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Chapter 6

Mental Disability

By the end of this chapter the PHC physician should be able to:

- Recognize the definition of mental disability and its classification.
- Recognize and describe causes and risk factors of mental disability.
- Recognize important preventive measures of mental disability.
- Recognize and describe normal psychological development.
- Make a provisional diagnosis of the mental disability according to the categories outlined.
- Describe essential management procedures and care for the most important health problems.
- To identify appropriate referral needs not fulfilled at the primary care level including medical, social and psychological.
- Provide parents with proper guidance and support in dealing with the condition.

Section 1

Introduction to Mental Disability

All psychological disorders in children have both a physical and psychological component. For many disorders-such as asthma, eczema, and ulcerative colitis, the emotional factor plays important role in the course of the disease.

Stress precipitates symptoms in people of all ages. Headaches, leg aches, stomachaches, and vomiting are common symptoms of stress in children. The sort of stress that causes such symptoms may be problems at home such as parental discord, inconsistent parental behavior, rivalry with siblings, or unrealistic expectations by parents—or problems at school or with friends. The loss of a parent or a move to a new home can be an acute stress.

Minor behavior disturbance involving enuresis (urinating), soiling (defecating), or sleep disorders are common. Similarly, habit spasms (tics) involving repetitive involuntary movements, usually of the head and neck, are extremely common.

Most children who exhibit such behaviors should not be considered psychologically abnormal if they receive the appropriate management in the right time

Serious childhood psychological and mental disorders are relatively uncommon, early detection and intervention make huge difference in improving the quality of life of these children.

Although widely prevalent, child abuse often is overlooked by family, friends, and health professionals. Prejudice, anxiety, and shame—not lack of information—seem to be the major reasons for the failure to recognize these private acts of violence—a form of tacit denial that leads to their perpetuation.

Child abuse can have serious future consequences for its victims. Early detection of child abuse including identification of the risk factors and ending the silence around its prevalence can save the children from the permanent damage that may result from the exposure to the insult or lack of proper intervention in the proper time.

Normal psychological development of the child

Age	Cognitive /social	Behavioral	Motor	Speech
1st week	Normal cry		Normal Sucking	
8-12 weeks	Smile, recognize breast and bottle, and anticipate being lifted.	Communicate hungers fears and discomfort	Lift head and chest when lying on his stomach	Cooing and gurgle sounds
9-12 months	Follow hidden objects	Shy with strangers, imitates gestures	Sitting without assistance, crawls on belly	Says dad and mama, imitates words
18-24 months	Recognize, names of familiar people, follow simple instructions	Enthusiastic about company with others, Say No	Walk alone	Single words (18 months) simple phrase (2 years)
2- 3 years	Matches objects with pictures, sort objects by colors shapes, complete puzzle of 3-4pieces	Express affection openly, express wide range of emotions, shows affection for play mates	Pedal tricycle, holds a pencil in writing position	Identifies all common objects, says names and ages, pronouns, I, You, --
4-5 years	Names and colors, concept of time, knows every day things (Money, food)	Wants to please friends, aware of sexuality	Hops, swings, Uses folk, brush his teeth.	Speaks sentences more than 5 words, uses future tense

Normal psychological development of school age child (6-12)

Cognitive / social

- Metalinguastic understood
- Concrete – Children begin to think logically, understand conservation of matter (frozen milk same amount as melted)
- Hierarchical classification – arrange cars by type
- Reversibility- play games backwards
- Decent ration – worry about details, obsessive
- Spatial operation – likes models for direction
- Child focus on social cues to recognize gender
- Gender remains constant regardless of clothing or behavior
- Child accepts respect for rules; negotiate with peers Concept of reciprocity
- Child argues about punishment.
- Boys develop more motor skills

Landmarks to be observed & common problems during the first five years

Landmarks to be observed during the first year

- Attachment (Eye contact, smiling, response to familiar faces
- Special interest in special figures
- The onset of the language development (the first meaningful word)
- Fears (related to unexpected change in the surroundings)
- Review the motor development)

Landmarks during the second year

- More attachment skills
- More reactivity because of more motor development
- First NO, which means the start of perceiving him/herself as an independent ego
- Anger tantrum
- Imitation of gestures
- Toilet training
- Fears of sudden changes and strangers start to decrease by the end of the 2nd year but the beginning of developing specific phobias start by this year (like phobias from animals)
- The cognitive development during this stage is mainly sensory motor which means the child learn to use the sense (like taste) and the patter of motions become more purposeful.

Common problems during the first two years
• Delay in development (Motor, emotional, behavioral, and cognitive)
• Involuntary movement & seizures
• Feeding problems (failure to thrive)

Landmarks during the third – fifth years

- Development of the fine purposeful movement (review motor development)
- Development of the fine discrimination (by the age 5 the child is able to fully recognize pictures and draw it.
- Sex differences in attitude
- Gender identity
- Identification with other figures (parents, teachers, etc.
- Imaginary thinking & fears
- Attachment to an object like a toy or similar thing

Common problems
• Tics, thumb sucking
• Sleeping problems
• Involuntary movement
• Absence & seizures
• Delayed development (manifested by behavioral problems)
• Eating problems
• Autistic behavior

Situations dangerous for children mental health

• **Malnutrition:** Malnourished child is particularly prone to chronic infection which itself may result in long term neurological and mental health impairment

• **Poverty:** Poverty deprives children from the essential elements of development, poor families are disadvantaged because of the lack of supportive environment and the lack of accessible services or space required for normal and health development.

• **Migration:** Refugee children are among the most vulnerable groups to emotional problems.

• **Environmental hazards:** Lead poisoning result from lead water pipes and lead base paint, insecticide in the rural areas also proved to affect the mental state and ability of the children

• **School factors:** Bullying and physical punishment at school inhibit healthy psychological development

General psychiatric assessment of children

Set routine for psychiatric examination of children is quite difficult or impossible. It depends on the age, language, social and motor skills

General appearance:

Size, level of nourishment, general hygiene, any abnormalities of facial, head, body build or limbs, bruises, cut, burns, mode of dress and its appropriateness to the weather.

Self – regulation:

• **Digestive system:**
Eating habits, vomiting, abdominal pain, chronic diarrhea, faecal soiling, and pica.

• **Urinary system:**
Bedwetting, wetting by day, over-frequent or painful micturation.

• **Sleep:**
Problems of going to bed, nightmares, night terrors, excessive sleepiness, interrupted sleep.

• **Motor System:**
Restlessness, overactive, under activity, tics or other involuntary movements, motor weakness, abnormal gait.

• **Neurotic habits:**
Nail biting, thumb sucking, nose picking, head banging,

• **Speech:**
Over talkative, mutism, articulation, faulty speech, stuttering, vocabulary.

- **Temperamental traits:**

Each child is born with specific temperamental pattern of behavior response. These traits can be discerned throughout the child's life span. There are two main Categories:

Easy children

- Regular in biological functions
- Positive approach to new stimuli
- High adaptability to change

Difficult children

- Irregularities in biological functions
- Negative withdrawal responses to new stimuli
- Non or slow adaptability

- **Behavior:** Fearfulness, reactive, irritable, tearfulness temper tantrum.

- **School:** attitude to school, behavior at school, progress and social adjustment at school is quit significant from the mental health point of view

- **Episodic disorders:** Convulsions, fainting, breath holding attacks.

- **Signs of physical abuse.**

Formulation:

- **Formulation grid**

	Constitutional	Temperamental	Physical	Environmental
Predisposing				
Precipitating				
Perpetuating				
Protective				

* The grid above may be used to help in the process of developing formulation of the case, it is not just to list the causative or contributing factors but the interaction and interplay between each of them is crucial to have a successful treatment plan It may be used also to update the information during the treatment process.

Child psychological disorders are usually the results of combination of different factors in the child and in his environment including the family. Strengthens as well as weakness should be included in our formulation of the case

- Constitutional Factors Like: genetic factors, chromosomal abnormalities, intrauterine disease
- Temperamental Factors: As mentioned above the pattern of reaction and behavior of the child
- Physical: Physical damage of the brain, Physical signs of abuse
- Environmental: family status and environment

Section 2

Mental Retardation and Associated Disorders

Mental retardation is characterized by incomplete or insufficient general development of mental capacity, causing a delay in the normal development of motor, language, and social skills. Behavioral abnormalities and impaired emotional control are also common. Before birth, chromosomal and developmental diseases, genetic and other metabolic disorders, and intrauterine infections or toxicity are possible causes. In the postnatal period, asphyxia, hemolytic disease, nutritional deficits, infections such as meningitis and encephalitis, trauma, and toxins are the most frequent causes of mental retardation.

Mentally retarded people are those develop at low average rate and experience difficulties in learning and social adjustment (it means sub average general intellectual functioning concurrent with maladaptive behavior).

Mentally retardation is not a disease and should not be confused with mental illness, children with mental retardation become adults (develop in the same way but at a slower) and they do learn but slowly and with difficulties rate)

Missing the early detection of mental retardation is common because of the (Myth) misbelieve that Normal appearance and ambulation are less likely to happen with retardation than the testing intellectual abilities in children is not possible.

Warning signs

- Delayed speech
- Dysmorphic features
- Hypotonia
- General inability to do things
- Extremes in infant temperament are often the first clue to an atypical course in child development.

Categories of mental retardation

- Borderline
- Mild
- Moderate
- Severe
- Profound

Table (6-1): Categories of mental retardation

	Borderline	Mild	Moderate	Severe	Profound
I.Q.	70 – 80 Educable	50 –70 Educable	35 –50 Trainable	25 – 35 Ineducable Little training	Below 25 Inducable Untrainable
Preschool	Develop normal milestone	Social & communicable	Can Talk	Poor motor Minimal speech	Semi motor functioning
School	Achieve Sec. School	Achieve 6 grade	Can achieve occupational skills	Elementary hygiene skills	Minimal Self care
Adult life	Normal	Social skills	Self support by unskilled work	Work under close supervision	Minimal self care
Stress needs		Guidance	Supervision	Close supervision	Constant Supervision

Common clinical syndromes associated with mental retardation

1. Down syndrome

Results from extra copy of chromosome 21, 1/600 births.

Risk factors

- Inherited in 2% from a carrier parent
- Early or late pregnancy

Manifestations

- Hypotonia
- Flat facial profile, flat bridge of the nose
- Broad feet with short toes
- Up slanting papeperal fissures
- Small ears, (Small ear canal)
- In curving 5th finger
- Short neck, Small head, Small oral cavity
- Single transverse palmer creases
- Short high pitched cries in infancy

Specific health problems common in Down syndrome

- Congenital heart defect (in one third of the of the babies with Down syndrome)
- Thyroid dysfunction?
- Visual problems (congenital cataract, nystagmus, strabismus far or near visions are more common)
- Developing Hearing loss or difficulties are common in children with DS.
- Atlanto Axial Instability (AAI)

Effectiveness of early detection

- The Paradigm of DS (They are retarded and no treatment) is proved to be wrong with early detection and intervention. They are not born retarded, rather they become retarded.
- Individuals with DS need loving home, early intervention and appropriate continuous medical care.

2. Phenyl ketonuria

- Defect in the phenylalanine hydroxylase with the accumulation of phenylalanine.
- Clinically: absent neonatally, later development of seizures (25%) fair skin, blue eye, blond hair, rash, if untreated mild to profound mental retardation, speech delay, and hyperactivities.

3. Fetal alcohol syndrome

Maternal consumption of alcohol, short stature, midface, microgentalia, mild to moderate retardation, irritability, inattention

4. Social retardation

Mildly retarded children who normally do not show any medical or neurological abnormality. Their retarded development is mainly due social and care deprivation and the lack of the appropriate programs and also lack of cognitive stimulation that help them to achieve their potentials. This may result in a lower level of functioning and more deterioration in their performance

Clinical management of mentally retarded children

There two main categories of mentally retarded population; the first and the larger group are those with mild retardation and do not have any specific or associated clinical disorders, the other group consists of those who are seriously handicapped mentally either because of specific clinical syndrome or due to massive brain damage.

For the mildly retarded children, the main goal of intervention is to integrate them into the society and manage them as equal citizens. Educational programs in special classes or special schools are provided to those who cannot benefit from the regular education, such programs include continuous skill assessment and potentials development.

Supportive programs (Financial, moral, educational) for the families to enable them to retain their handicapped children are recommended.

For behavioral and emotional problems of the mentally retarded children, we follow with them same treatment approaches in other psychological disorders like behavioral therapy, family therapy and psychopharmacological treatment.

Treatable cases like Phenylketonurea should start treatment as early as possible. The treatment is mainly a diet low in phenylalanine during the child's early years; this usually leads to normal child development.

Last few years have witnessed the development special treatment programs for Down syndrome that create more hope for those children and shaken the myth of the Down syndrome as an untreatable condition. It includes medications (antioxidants) and special diets.

Special supportive and educational programs for the families and communities that aim changing the attitude towards the Downs and promote more integration in the society.

Other treatment approaches for mentally retarded children are applicable in Down syndrome depending on the severity of the case, the age and the degree of the mental retardation.

Section 3

Specific Mental Health Problems

Mind-body relationships in children

1. Eating disorder:

The common eating disorders in childhood are:

- Lack of appetite
- Excessive appetite
- Perverted appetite; Pica, which means eating a non nutritive (non-food) substances, it may be a symptom of: MR, Neglect and abuse.

2. Bed-wetting-day time wetting (enuresis)

Enuresis is defined as involuntary or intentional voiding of urine, severity is determined by the frequency of urination not the quantity .The definition precludes a physical cause for the disorder and the child's chronological and mental age is at least 5years.

• **Primary:** the child has never become dry. In most of the cases no specific cause can be found for 1ry enuresis. It is often ascribe to delay in neurological maturation or severely disorganized family and the child receives inconsistent toilet training.

"Delay in achieving bladder control is a feature of mental retardation; it is marked in severe retardation"

• **Secondary:** enuresis starting after the child has achieved continence after a certain period of time (3 months).

It may be a symptom of regressive behavior appearing in situations of emotional stress; it may be also symptoms of conduct disorder.

Diurnal wetting is probably more often associated with psychiatric disorder.

Other specific causes like UTI, nocturnal epilepsy, anatomical abnormalities were founded.

Enuresis usually complicated by feelings of anxiety or guilt especially if the child is blamed or punished for enuresis.

3. Sleeping problems

Children's need for sleep varies from child to another. At the age of 2 months, infants undergo a shift from endogenous control to more exogenous control as his waking hours become longer. Children use the waking hours for more exploratory behavior

Normal sleep pattern

- Newborn infant sleep an average of 17 hours/ day
- 6 months sleep 14 hours /day
- 2 years sleep 12 / day
- 6 years sleep 11 hours / day
- 6 – 11 years, sleep about 10 hours
- After 11, the average sleep time is about 8 hours.
- Regular wake up time in the morning

Sleep disorders

Insomnia

Primary insomnia characterized by excessive worry during the day time about not being able to sleep or getting nightmares

Secondary insomnia: insomnia related to another mental disorder like anxiety or depression, or related to a known organic factor (psychoactive substance, arthritis)

Parasomnia

Night terrors, Nightmares, sleep walk, sleep apnea

Night terrors: the child wake up in the morning in a terrified state, inaccessible, not responding when spoken to, twilight state, looking at people and objects not actually present (hallucinating)

- **Tics**

A sudden movement of a muscle or group of muscles (may be facial, vocal, trunk or limbs), serving no purpose and not under voluntary control.

Usually associated with other emotional disorders
It is handicapping socially especially when it involve large movements

4. Anxiety disorders

Normal anxiety is the major human motivator, when exceed a certain limit it interfere with the performance level and needs intervention.

It can be general anxiety disorder or anxiety related to specific situations (separation anxiety, phobia and post traumatic stress disorder)

A. General (over) anxiety disorder

- The child tends to be extremely self-conscious.
- Worry about the future events, such as exam, meeting expectations
- Feeling of tension, sense of insecurity, fears of danger from within
- Lack of concentration
- Insomnia and nightmares
- Dry mouth, heartburn, nausea, headache, blurring of vision, fatigue

B. Separation anxiety disorder

Continuous anxiety concerning separation from those whom the child is attached in the form of:

- Unrealistic and persistent worry about possible harm befalling attachment figures
- Reluctance to go to school in order to stay with the attachment
- Difficulties to go to sleep in order to stay with the major attachment figure
- Repeated nightmares
- Physical complaint

C. Phobias

Fears are natural and instinctual feeling like fear from death, heights, dark, and monsters. However when fears persist for long time or irrationally exist, then it is not normal (phobias)

Phobias in children may exist in the form of:

- *Simple phobias:* fears of a specific objector situation
- *Social phobias:* fear of being embarrassed or humiliated in social situation.

D. School refusal

- At the age of 5 – 7, associated with separation anxiety disorder
- At the age of 11, probably associated with school change
- At the age of 14, frequently associated with depression

E. Posttraumatic stress disorder

- **Stressor:** The child must have been exposed to a stressful event of exceptionally threatening nature, which would be likely to cause pervasive distress to almost any one. The symptoms composed of 3 categories:

  1. Re experiencing: Flashbacks, reliving the trauma events in the form of vivid memories, recurrent dreams

  2. Avoidance and Numbing: Avoiding behavior to the circumstances that resemble or associated with the stress.

  3. Hyper arousal: Persistent anxiety symptoms in the form of, difficulties in falling asleep, difficulties in concentration and exaggerated startle response.

Management of anxiety & anxiety related disorders

• **Drug therapy:** It may be helpful in some cases that show severe symptoms of anxiety or other emotional disturbances. New generation of antidepressants that characterized by low profile of side effects and rapid onset of action are preferable, short courses of minor tranquilizers may be also helpful in some cases.

• **Psychotherapy:** the essence of the psychotherapy intervention and regardless the specific approach is the building of a relationship between the therapist and the patient. Children are specifically sensitive to the therapeutic context, every contact with the child is emotionally significant and may be either therapeutic or anti therapeutic.

• **General principles to be considered in dealing with children:**

DO NOT
• Criticize or disapprove the child. • Allow the child to feel at fault. • Press for an answer the child is unwilling to give. • Go straight in discussing symptoms unless brought up by the child.

DO
• Listen and feel the child point of view. • Help and facilitate the expression of anger and other negative feelings. • Appraise the ideas expressed by the child. • Gain the child's confidence, remember that children do not come for treatment by them selves, they brought by the caregivers who are worried and concerned and may be angry with the child.

• **Family therapy:** Many children's psychiatric problems can be well understood in the context of family situation. The focus in family therapy is the family system, when this system is dysfunctional, it manifest directly as psychological symptoms in children.
The types of family intervention differ widely depending on the theoretical models of the therapist. Direct intervention consists of offering the family a different way of functioning.

• **Behavioral Therapy:** Commonly used in some child psychiatry like phobias and enuresis. It is based on the learning theory; its goals are either elimination of undesirable way of response or behavior or development of desirable behavior. The treatment is often located in child's environment like home or at school rather than in clinic or therapist's office and in collaboration with the family or the caregivers.

Conditioning process is the basis of behavioral therapy, it may be:

• Classical conditioning where a neutral stimulant (UCS) comes to evoke a new response by pairing it with another stimulant that reflexively (CS) evokes that response

• Operant conditioning based on the fact that responses followed by reinforcement tend to increase in rate; those not followed by enforcement tend to decrease in rate.

5. Attention deficit with hyperkinesias (ADHD)

- The child appears as if does not listen
- Does not stick with any task to the end, frequently shifts from one unfinished activity to another, noisy
- Fails to follow the role of structured game
- Poor peer relation
- Poor social problem solving skills
- Interrupt the others
- Can't wait his role
- Can't remain seated
- Low self esteem
- Impulsive, unpredictable mood
- Poor perspective on the future consequences
- Immature physical size
- Poor motor coordination
- High pain tolerance
- Under reactive CNS
- High frequencies of minor physical anomalies

The course of the disease may be:

- *Self-limited:* In case of early detection and good management, all the symptoms disappear at puberty.
- *All symptoms persist:* this common if the cause is minor brain damage
- *Improvement of the hyperactivity symptoms and persistence of the attention deficit*

Management

The treatment plan should be based on careful assessment of the predominant symptoms (hyperactivity/attention deficit); any associated learning difficulties, perceptual or behavioral abnormalities.

The medications used in ADHD are mainly two categories:

- Cerebral Stimulants like Methylphenidate, Amphetamines and pemoline have been found to improve the on – task behavior, decrease the impulsivity and the purposeless activities, improve the goal directed activities, concentration and the information processing.

- Tranquillizers: like Haloperidol, chlorpromazine and thioridazines, may be helpful to reduce the hyperactivities symptoms but at the cost of sedating the child.

Specific psychotherapeutic programs for behavioral modifications are proved to be helpful in improving the impulsive behavior and the lack of attention. Family and teachers education about the disease and the needs of the child is crucial.

6. Autism

Autism may develop early before the 2nd year (infantile autism). Or later after the third year (childhood autism).

Infantile autism

Usually associated with mental retardation (75%). the mental retardation is not related to the autistic process itself as it tended to remain at about the same level even after the recovery from the condition itself.

Childhood autism

Characterized by:

- Qualitative abnormalities in communication skills; language skills as well as facial expressions and gestures
- Qualitative abnormalities in reciprocal social abnormalities; lack of eye – eye contact, lack of spontaneous seeking to share enjoyment
- Stereotype restricted activities; like moving an object endless repetitive movement
- Attachment to an odd object

Asperger syndrome
Same criteria of autism accept no significant delay in the spoken or receptive language skills.

Clinical management

Treatment of autistic disorder should be a venture of planners of the program, the behavioral therapist, speech therapist and special teacher. The best location for treatment is at home or within the community itself.

Treatment should aim to promote healthier and more normal developmental progress with the various development delays seen in autistic child.

Focusing on the communication and speech skills is quite important, however it is a long and difficult process.

Involvement of the family is crucial for the progress in treatment although the family itself need support and guidance.

Section 4

Child Abuse & Maltreatment

Definition

Child abuse is not only beating child,

- ***Physical abuse;*** take the form of physical violence that includes beating, burning, suspension and other possible forms of physical punishment and violations.

- ***Emotional abuse;*** results from Rejections, deprivations of affections or stimulations, exposure to domestic violence or severe domestic disharmony, inappropriate criticism, threats, humiliation sand accusations.

Signs

Physical signs

- Skin injuries like cigarette burns or hematoma
- Head and eye injuries
- Long bone or rib fracture or multiple fractures.
- Bite marks
- Signs of lack of care like malnutrition, inappropriate clothing (lack of warm clothing in cold weather, chronic infections)
- Recurrent dysuria or vaginitis, sexually transmitted disease, bruising or lacerations to the vulva or the anus

Behavioral signs

- Aggressiveness, phobias, fears, sexually inappropriate behavior
- Reluctance to be alone with a particular adult
- Repeated running away from home
- Withdrawn, avoidance behavior
- Any form of drug abuse

Emotional signs

- Sadness, detachment or blunted (emotional death)

Family signs

- Young parents under employment of the care givers, poor marital relation, social isolation
- Alcoholic, Drug abusers or psychotic parents
- Positive family history; Parents blame the child, see the child as bad or an evil and not attractive

Female genital mutilation

Is performed on an estimated two million girls world wide every year, it is practiced across diverse socioeconomic classes and different cultural, ethnic and religious groups, commonly girls are circumcised between the age of 4 and 12. This practice has been criticized world wide including the WHO and other major health groups and considered child abuse in many countries

Child abuse: How to manage?

When signs of physical abuse are founded or suspected, full physical exam should be done including careful exam of the genetalia and X ray and any other investigations as indicated. Proper, careful and early physical treatment for the physical injuries is essential in preventing any possible disabilities.

For psychological injuries, we apply the same general principles used in managing anxiety related disorders.

If the child has been abused by a family member or within the family environment, try to adopt an empathetic, non-punitive attitude. Explain to the family member your professional obligations to inform the legal authorities and your willingness to continue offering them the needed help and support for the benefit of the child. It is not helpful to show anger or outrage or to forcefully extract a confession from the family about the abuse.

The case is different when the abusive is some one outside the family; in such cases the family members usually show active cooperation in the treatment and investigation of the case. Make sure that they arte not judging the child or blaming him/her and try to emphasize the importance of being supportive and understanding for the treatment of their child and reducing the sequel of the abusive events.

Treatment of the abused child require a multidisciplinary well trained team that involved the therapist, family member, social authority, legal authority and a school member. Continuous cooperation between the team members is crucial for the success of the treatment plan.

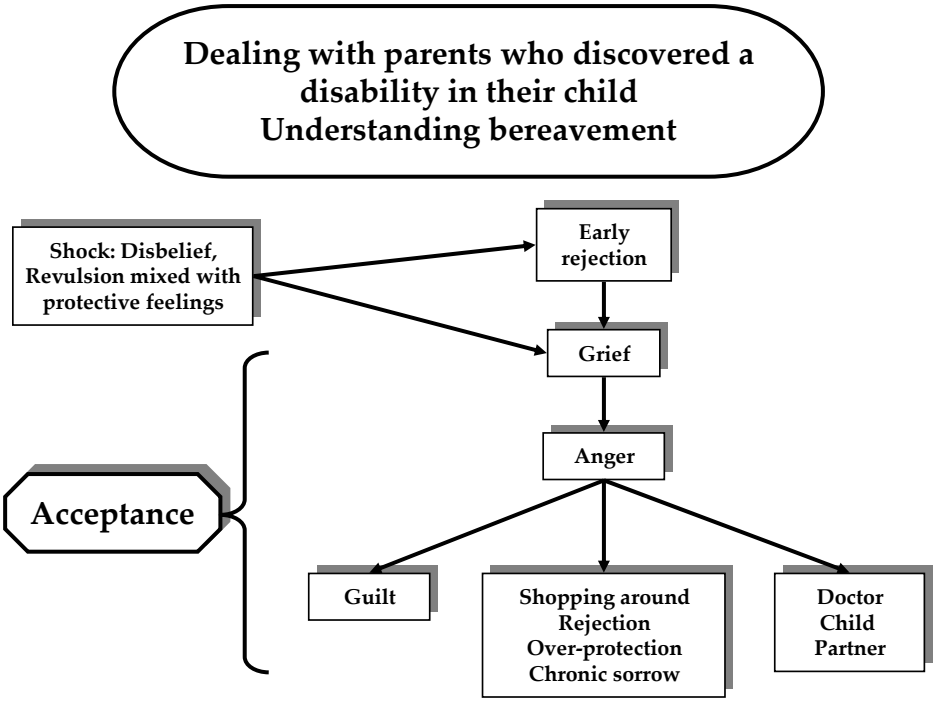
Because treatment of child abuse is quite long and difficult process, it is believed now that the best treatment is the preventive one that involved increase the awareness of the health and social workers in the primary care facilities, especial educative programs in schools and with the families as an essential part of the anti and post natal care. Hotlines for helping abused children and special units at the police and court facilities for the same purpose was established in many of the developed countries and were proved to be valuable in preventing child abuse and reducing its psychological sequels.

Section 5

Guidelines in Dealing with the Families

• Current mental literature describes families of the children as the most important resource. The role of the professionals should be to assist in meeting their goals and to empower them to achieve control over their life circumstances.

• Families and mental health professionals know that the stresses of caring for a child with mental or psychological disorders are real. It includes changes to daily family life, anger, worry; feelings of loss (grief), social isolation, feeling blamed, and increased financial expenses.



• In addition to the stress of having a child with mental disability, some families have the added stress of social and economical- cultural origin.

Questions that frequently asked by the parents

1. Why is this happening to us?
2. What have I done wrong?
3. When does it end?

• The best model for empowering the families to care for their children with mental disorders is a partner ship between families and the professionals involved in their child's care (therapeutic alliance). This partnership relationship is based on sharing the skills, knowledge and experiences of each partner. This empowers the parents; they begin to take

an action to get what they need and they begin to contribute and offer valuable information to help in developing the best and most appropriate treatment decisions.

Some ideas that will help to foster a professional parents partnership

• Families with children who have special needs are more like other families than they are different, though they are handling more intense stresses and they are handling them more often, they are trying to achieve the same quality of life and are using the same methods for coping stress as are all families in our culture.

• Experiences and research have shown that parents of children with special needs are generally welcome parent to parent support and often seek it in times of need. Parents who have been there can be effective resources for information and guidance and can act as role models and advocate for other parents.

• Take long range perspectives rather than focus on accomplishing short term objectives.
Family members can provide valid and well in formed assessment and information including identifying and describing their child in regard to their strengths and needs.

• When parents are not involved in decision making and service delivery, they can develop feelings of helplessness and dependency on t he one hand, and resentment on the other.
Listen carefully to what parents tell you and be open to new perspectives.
Support system such as extended family, neighbors mosques, churches, schools and volunteers organizations are critical for family success.

• Parents are very aware of their responsibilities to provide care for their children but they are seldom aware of their rights as care givers. Remind the parents their rights as human beings:

1. You have the right to accept that you are doing the best you can, and that is good enough.
2. You have needs that are as important as the needs of your child.
3. You have your own hobbies and interests.
4. Have a vacation away from your child every year.
5. Have celebrations, weekends away, and time together with your partner to enhance your relationship.

Chapter Summary

Mental Disability

Risk factors

- Constitutional: hereditary and congenital anomalies
- Temperamental: difficulties in coping
- Physical: Malnutrition,
- Environmental: Broken family, Loss, migration, severe poverty

Warning signs

- Congenital anomalies
- Failure to thrive
- Involuntary movements
- Epileptic activities
- Disturbance in bowl control
- Sleep disorders
- Eating disorders
- Lack of basic social and linguistic communication skills

Screening procedures

At the PHC level

- General medical exam
- General Psychological exam
- Assessment of the risk factors

Management and rehabilitation guidelines

- Multi disciplinary assessment (Bio- Psycho- Social)
- Multi axial treatment (Bio Psycho social}
- Family or care givers are involved from the beginning
- Supportive, educational measures for the local community, family and teachers are crucial

Suggested referral places

- Ain Shams Child Psychiatry unit
- Kasr El Aeiny child psychiatry unit
- Child Psychiatry clinics, school health insurance
- General Psychiatric hospital

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UNICEF website

Chapter 7

Hearing Disability

By the end of this chapter, the PHC physician should be able to:

- Recognize the status of hearing disability in Egypt and its classification.
- Recognize and describe causes and risk factors of hearing disability.
- Recognize important preventive measures of important hearing disability.
- Make a provisional diagnosis of hearing disability according to the categories outlined.
- Describe essential management procedures and care for the most important health problems.
- Identify appropriate referral needs not fulfilled at the primary care level including medical, social and psychological.

Abbreviations

• dB	Decibel
• STORCH	Syphilis, Toxoplasmosis, Rubella, Chlamydeous, Herpes
• KHz	Kilohertz
• HL	Hearing loss
• SNHL	Sensorineural hearing loss
• ABR	Auditory brainstem response
• OAEs	Otoacoustic emissions

Hearing Disability

Introduction

Hearing disability has gained great interest in the last decades. This growing interest stemmed from its devastating effects on communication, person's life and above all the economics of countries. All these aspects are, however common to all types and forms of other disabilities, yet hearing loss is proved to be the number one birth defect among newborns. Current international research studies have also shown that if hearing loss is discovered very early in life with early implementation of proper intervention procedures, these children can lead a normal life. This chapter will provide the reader with the basic knowledge to identify the size of the problem, its different types and causes, together with different lines for diagnosis and management of hearing disability.

Definition

Hearing disability is best defined as a lack or reduction in the ability to hear clearly due to a problem somewhere in the hearing mechanism. A hearing impairment can occur in the outer, middle, or inner ear, or along the pathway to the brain.

The impact of this disability is reflected on the function of the individuals in the community and this lead to limitations on the part of the individual and obligations on the part of the community.

Prevalence of hearing disability

Many factors shape and influence the impact of the disability. Hearing loss represents a common birth defect however, it is difficult to detect, due to its 'invisible' nature. Statistics for children with hearing impairment are reported to be 2-6 per 1000 live birth (Parving, A., 1999). Mehl and Thompson (1998) stated that hearing loss represents a significant public health problem especially when compared to other commonly known childhood disease (2%). In Egypt, profound hearing loss is shown to be 6-7 per 1000 child.

Types of hearing disability

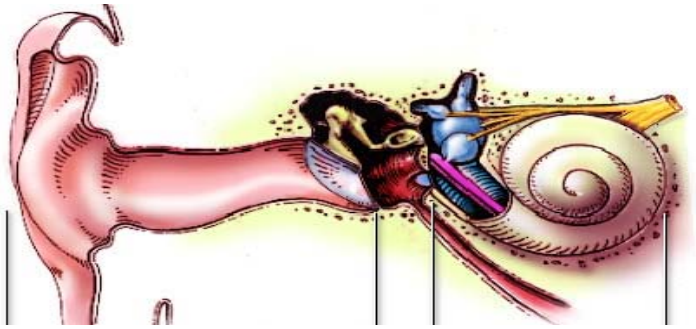
Peripheral hearing loss

There are three types of peripheral hearing loss: conductive, sensorineural, and mixed.

- **A conductive hearing loss** is a result of damage to outer or middle ear. Conductive losses are not severe and often times can be surgically corrected. A person with a conductive loss may reap great benefits from hearing aids.

• **A sensorineural hearing loss** is a result of damage to the hair cells of the inner ear or nerves. This type of loss ranges from mild to profound and is permanent. In other words, surgery cannot be performed to correct a sensorineural hearing loss. While the aids may amplify sounds, speech discrimination may be still affected.

• **A mixed hearing loss** simply means that the hearing problem occurs in the outer or middle ear and inner ear.



External ear **Middle ear** **Inner ear**

Figure (7-1): The Auditory System Peripheral pathway

Levels of hearing disability

There are different degrees of hearing loss. Below is a table that lists and classifies the degrees of hearing loss according the dB range in which sound is heard.

Degree of hearing loss	dB range
Normal Hearing	0-20 dB
Mild Hearing Loss (HL)	> 20-40 dB
Moderate HL	> 40-55 dB
Moderately severe HL	> 55-70 dB
Severe HL	> 70-90 dB
Profound HL	> 90 and up dB

Mild hearing loss

A mild hearing loss causes loss of 25-40% of the speech signal. Usually this results in problems with clarity since the brain is receiving some sounds but not all of the information. Symptoms of mild hearing loss include problems understanding someone farther away than a normal distance for conversation, or even up close if the background environment is noisy. Weak voices are also difficult to understand for people with mild hearing losses.

Moderate hearing loss

A moderate hearing loss leads the patients to miss 50-75% of the speech signal. This means they would not have problems hearing at short distances and understanding people face-to-face, but they do have problems if distance or visual cues changed. Symptoms of moderate

hearing loss include problems of hearing even normal conversations and problems hearing consonants in words

Severe hearing loss

People with severe hearing loss have difficulty hearing in all situations. Speech may be heard only if the speaker is talking loudly or at close range. A severe hearing loss may sometimes attenuate up to 100% of the speech signal. Symptoms of severe hearing loss include inability to have conversations except under the most ideal circumstances (i.e., face-to-face, in quiet, and accompanied with speech reading).

Profound hearing loss

Profound hearing loss is the most extreme hearing loss. A profound hearing loss means that the patients do not hear loud speech or any speech at all. They are forced to rely on visual cues instead of hearing as the main method of communication. This may include sign language and/or speech reading (also commonly referred to as "lip-reading").

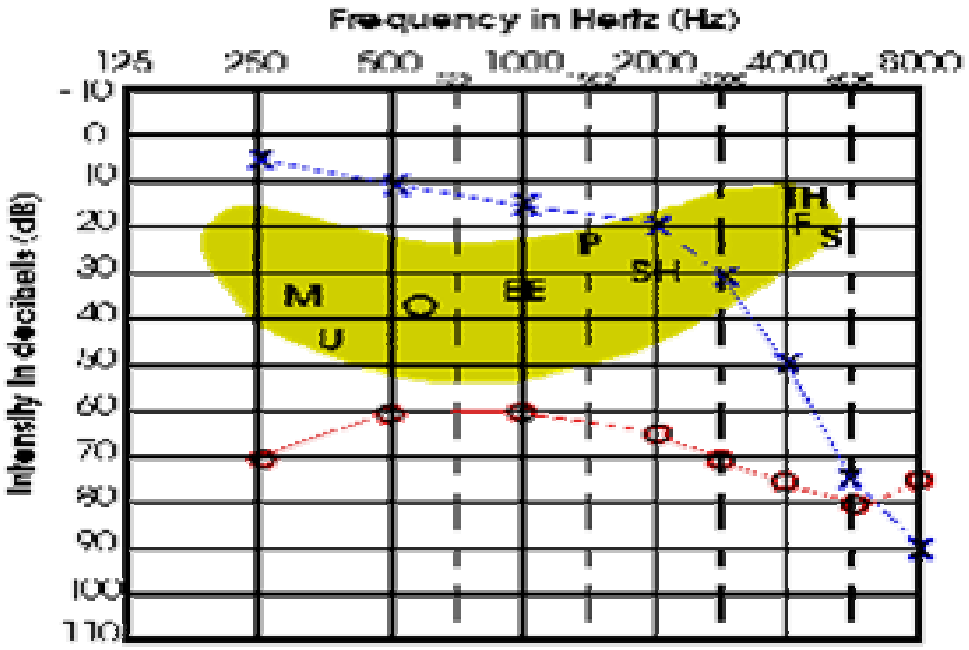


Figure (7-2): Hearing thresholds of Right ear (o....o) and Left ear (x….x) represented on an audiogram. The yellow banana shaped figure represents all the sounds that make up the human voice when speaking at normal conversational levels.

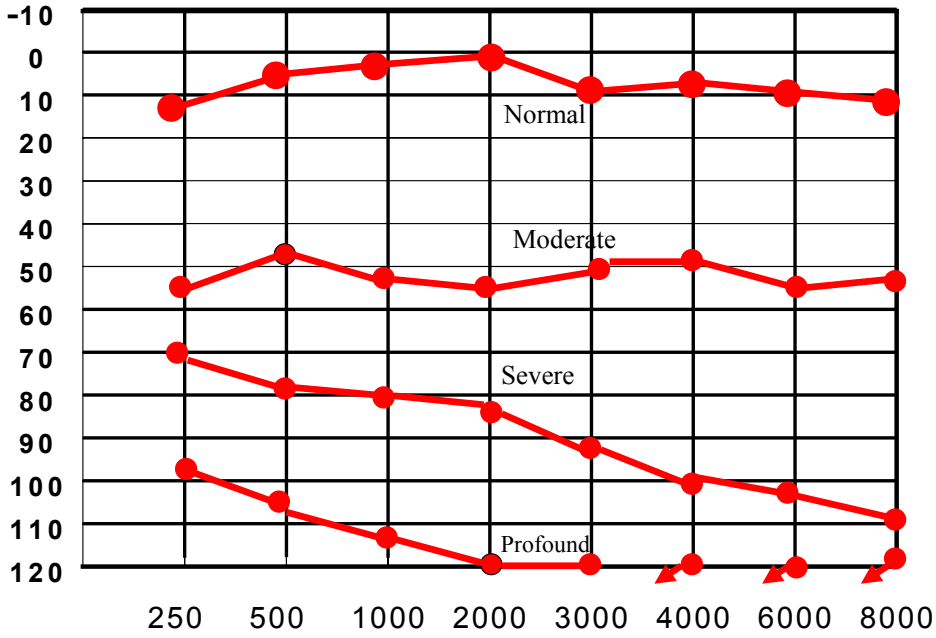


Figure (7-3): Normal hearing thresholds & the different degrees of hearing loss

Causes & risk factors of hearing disability

Conductive hearing loss

Commonest causes

- A buildup of fluid in the middle ear (middle ear effusion)
- Wax in the ear canal
- Puncturing of the eardrum (traumatic)
- Problems or injury to the bones or membrane — which carry sound from the external ear through the middle ear to the inner ear (e.g. chronic suppurative otitis media).
- Congenital malformation of the external / middle ear.

Sensorineural hearing loss

Commonest causes

1. Prenatal

- Intrauterine infections (STORCH)
- Prematurity
- Toxemia
- Maternal intake of ototoxic medication especially in first trimester Or maternal exposure to radiation
- Genetic Disorders

2. Perinatal

- Obstructed labor
- Cyanosis
- Birth trauma
- Depressed Apgar score - Prolonged mechanical ventilation

3. Postnatal

- Hyperbilirubinmea
- Infections: Mumps, measles, meningitis
- Trauma
- Ototoxic medications: Aminoglycosides (e.g., gentamicin, streptomycin, tobramycin, neomycin etc.), Antineoplastics, and Loop diuretics
- Syndromes known to include HL

Mixed hearing loss

Mixed hearing loss is a combination of both conductive and sensorineural hearing loss at the same time. Both the middle and inner ear are involved. This hearing disorder can also occur when a child first just has a permanent sensorineural hearing loss and then also develops a conductive hearing loss e.g. middle ear infection. The two types of loss combine to create a greater hearing loss. Some other instances of mixed hearing loss are the result of the outer and inner ear being malformed, which causes both types of hearing loss.

With mixed hearing loss, the conductive part may be treated, but the sensorineural part is usually permanent.

Assessment of hearing disability (formal testing)

1. History taking

Proper history taking is mandatory in order to identify the possible risk factors and to reach an etiological diagnosis.

2. Otological examination: to visualize eardrums

A. Audiological evaluation:

It aims at measuring hearing thresholds at frequency range of 250 – 8000 Hz by air conduction and possibly bone conduction routes. Also it measures the speech understanding scores. This is done according to the age of the child.

1. Subjective methods

a. Free field examination

- *Behavioral / observational audiometry (4months-2.5 years)*

The child sits on his mother lap in **sound treated room** facing the examiner. The audiologist presents sound stimulus (warble tones from 250-8000Hz from loudspeaker) and observes the child's behavior manifested by change in suckling pattern, eye widening, cessation of movement, crying or a head turn. This test represents the thresholds of the better ear, yet its sensitivity & specificity are very low.

• *Visual reinforcement audiometry (children between 6 months and 2.5 years)*

The child sits on his mother lap in **sound treated room** facing the examiner. The child is conditioned and trained to look to the light (toy) when sound is presented. The stimuli are warble tones from 250-8000Hz delivered from loudspeaker with variable intensities. This test represents the threshold of the better ear only. Moreover, it needs exceptional child and examiner.

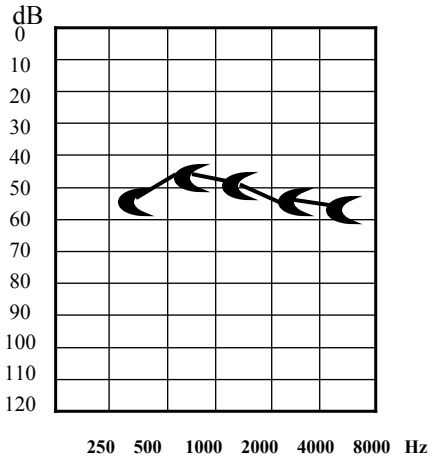


Figure (7-4): Sound field results
Moderate hearing loss (response of better ear)

b. Conditioned play audiometry

Used with children 2.5 to 5 years old. The child is seated in **a sound treated room** wearing **headphones** i.e. examine each ear separately. The child conditioned to drop a block in the jar when sound is presented (250-8000Hz) at variable intensities.

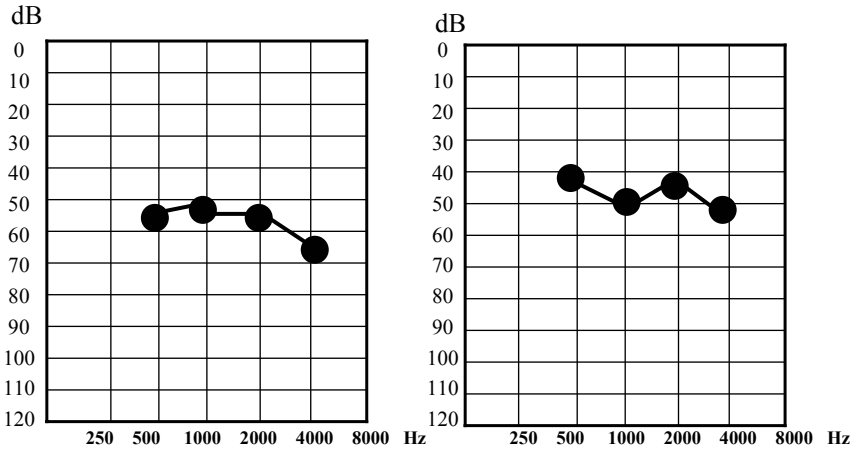


Figure (7-5): Play audiometry (Bilateral moderate hearing loss)

c. Conventional audiometry (Adult type)

For children 5years and older.The child is seated in **a sound treated room** wearing **headphones**. The child asked to raise his hand in response to sounds that are presented at variable intensities at frequency range of 250 – 8000Hz. Then **a bone vibrator** is placed over the mastoid and bone conduction threshold is also tested in a

similar pattern. In this way air-conduction and bone conduction thresholds are measured together with speech audiometry.

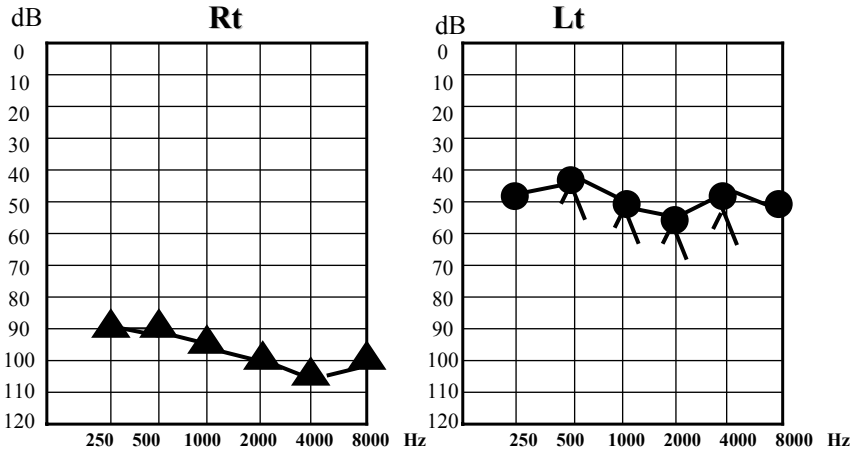


Figure (7-6): Conventional Audiometry
Speech Discrimination Scores = Rt. Ear: 20% Lt. Ear: 76 %
RT profound, Left moderate SNHL hearing loss

B. Objective methods: (for all age groups)

Tympanomerty

It measures the middle ear status. However it is not a measure of hearing thresholds. It can quickly differentiate middle ear effusion from Eustachian tube dysfunction versus normal middle ear. It needs passive cooperation i.e. the child sits quietly or asleep.

A probe is placed in the ear canal of the child and acoustic admittance of the ear with various amounts of air pressure in the ear canal is measured.

Types of tympanograms

- *Type A:* peak around atmospheric pressure (Normal, SNHL)
- *Type B:* flat tympanogram (effusion, cholesteatoma)
- *Type C:* negative peak (Eustachian tube dysfunction)

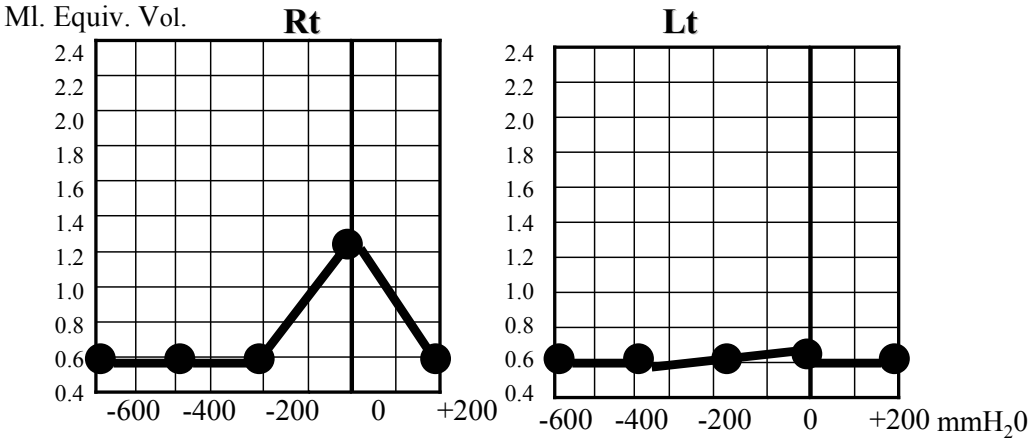


Figure (7-7): Tympanometry results
Right type A tympanogram, Left type B tympanogram

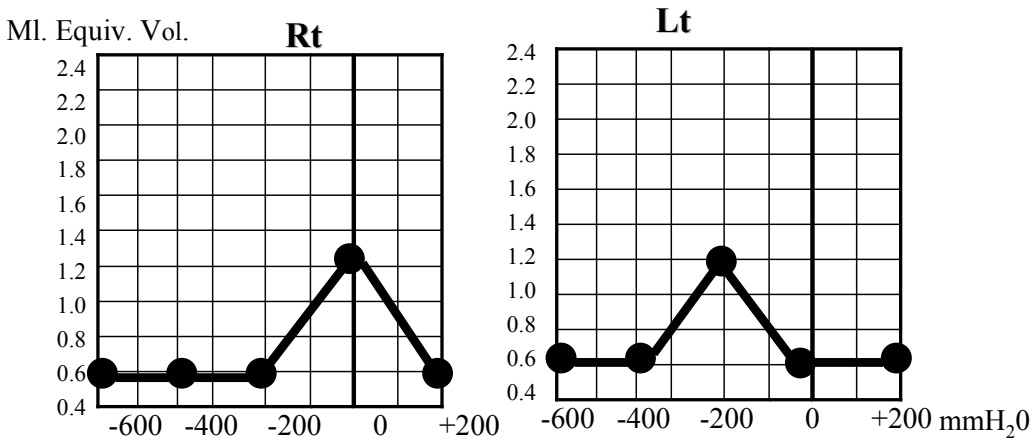


Figure (7-8): Tympanometry results
Rt (type A) tympanogram, Left (type C) tympanogram

Auditory Brainstem Response (ABR)

ABR are very small electrical voltage potential originating from the brain recorded from the scalp in response to auditory stimuli**.**

The child is usually sedated, electrodes are placed over the forehead and mastoids and click stimuli are presented at various intensities through **headphones**.

The Auditory Brainstem Response consists of five to seven waves. Originating from the VIIIth cranial nerve (waves I and II) and brainstem auditory structures (wave V: region of lateral lemniscus and inferior colliculus). It determines hearing sensitivity in high frequency range (2-4 KHz).

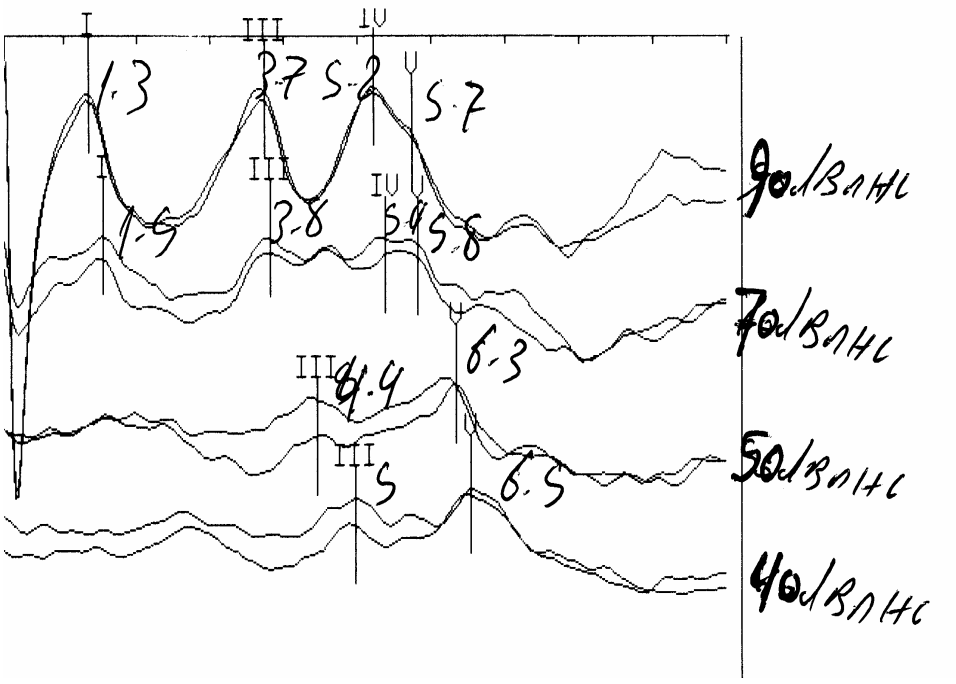


Figure (7-9): Normal ABR

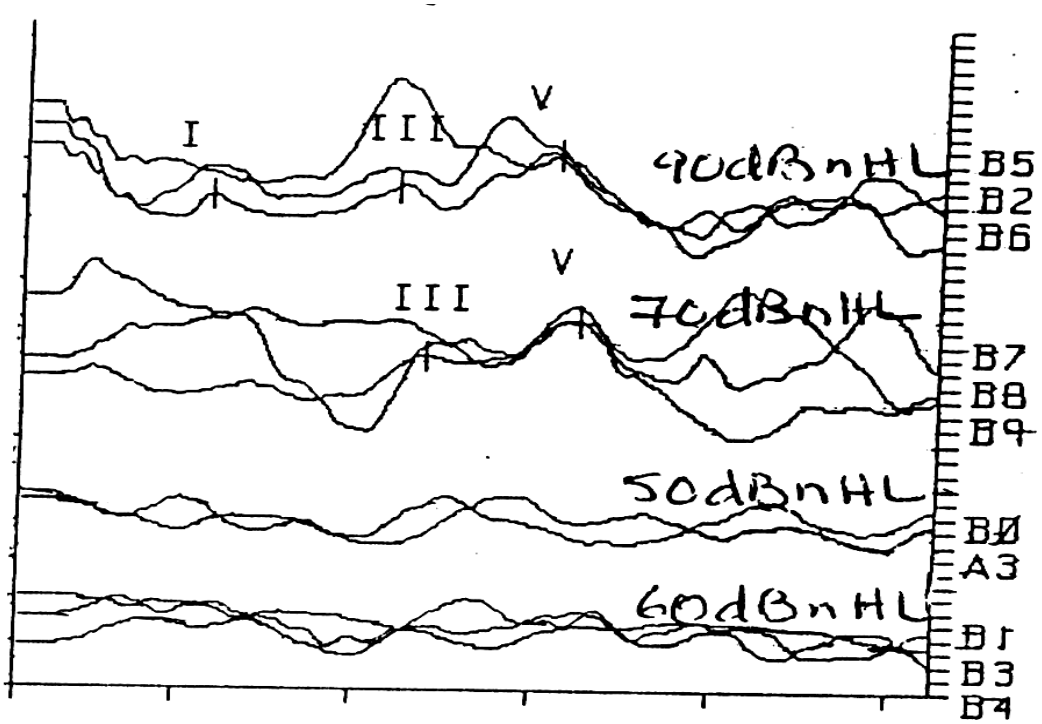


Figure (7-10): ABR showing Hearing loss (moderate)

Otoacoustic Emissions (OAEs)

They are sounds generated by the cochlear outer hair cells in response to an external sound stimulus. When this mechanism loses the peak of its performance, OAEs diminish and hearing threshold is raised. As OAEs can be recorded in the outer ear. It provides an objective, non-invasive and quantitative measure of hair-cell function.

The child is sedated or sits quietly in a sound treated room and a probe is placed in the ear canal

Types:

- *Transient-evoked OAEs (TEOAEs),* a response to acoustic clicks delivered to the outer ear, are currently thought to be the most clinically useful OAEs, as they are detectable in normal hearing, regardless of age or sex, and the two ears of any individual produce similar TEOAEs. TEOAEs are considered as the mvost important tool in newborn hearing screening.

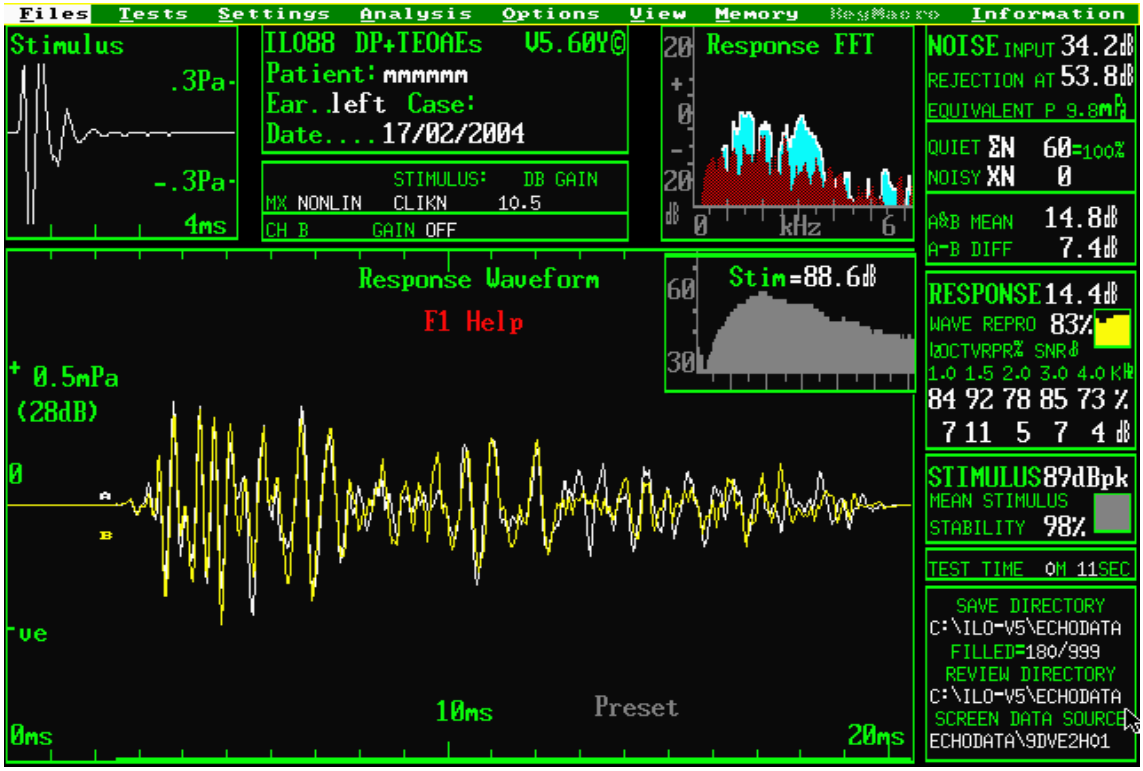


Figure (7-11): Transient OAE (Pass response)

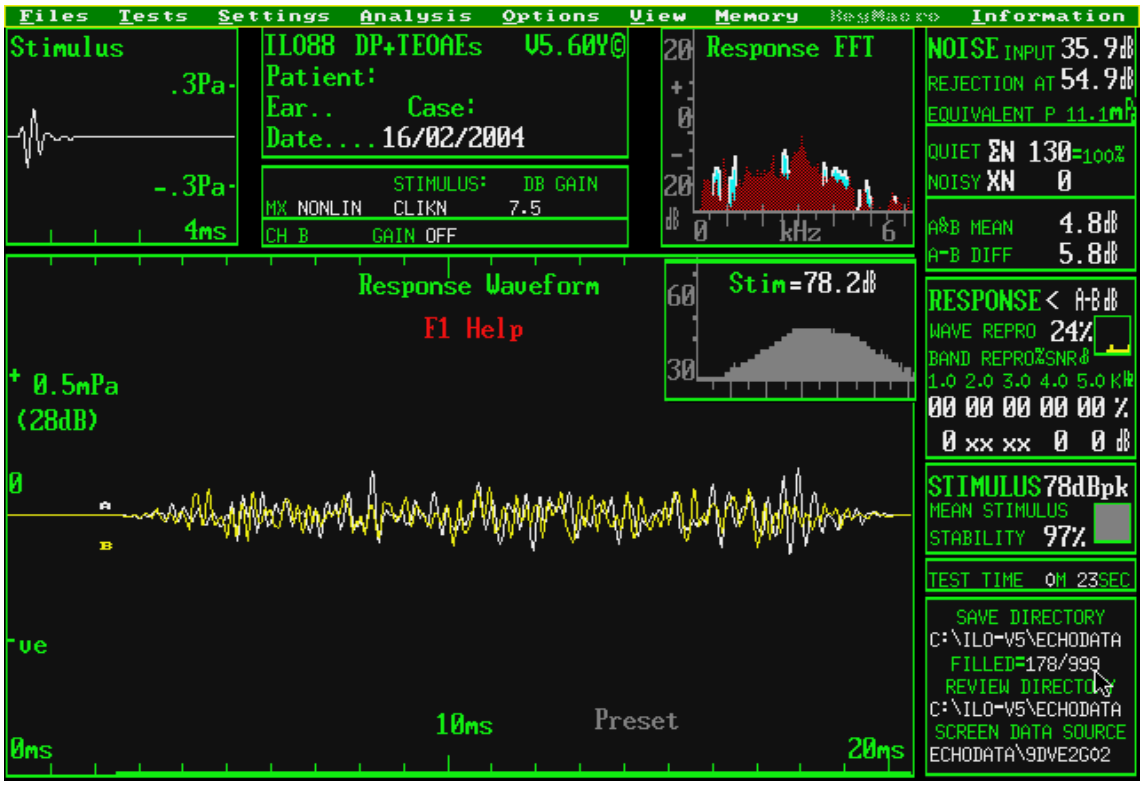


Figure (7-12): Transient OAE (Absent response)

• *Distortion-product OAEs (DPOAEs)* also occur in 100% of people with normal hearing, while small in amplitude in SNHL up to moderate degree. It can be used to intentionally test a specific frequency region of the cochlea for a more depth of knowledge.

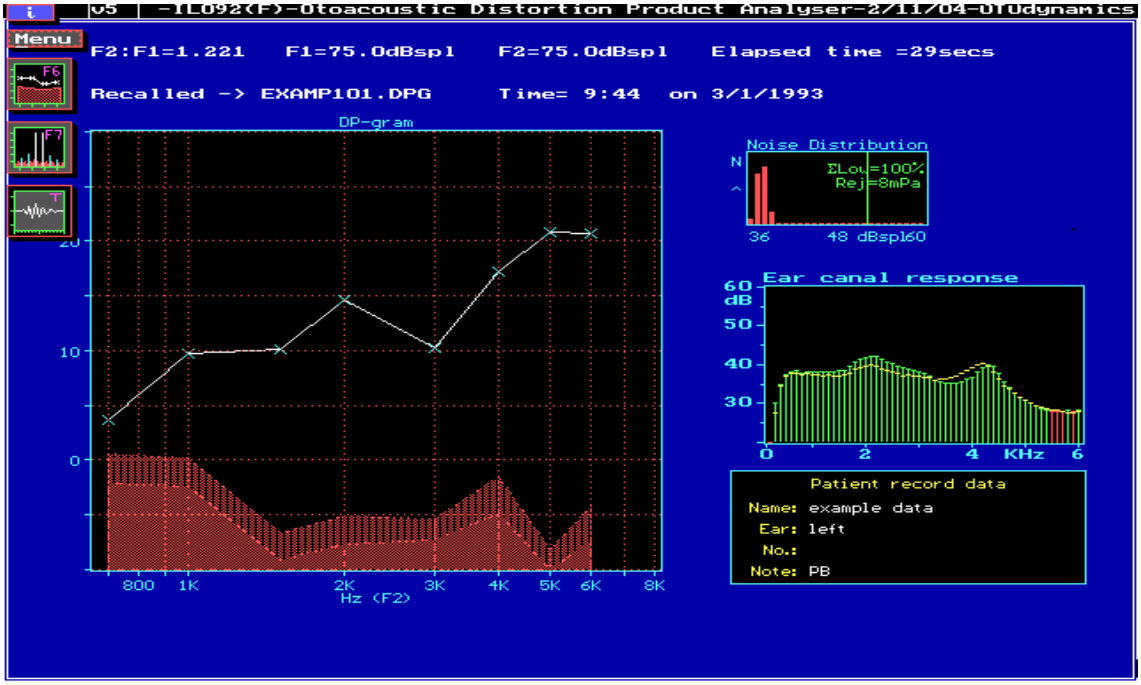


Figure (7-13): Distortion OAE (Pass response)

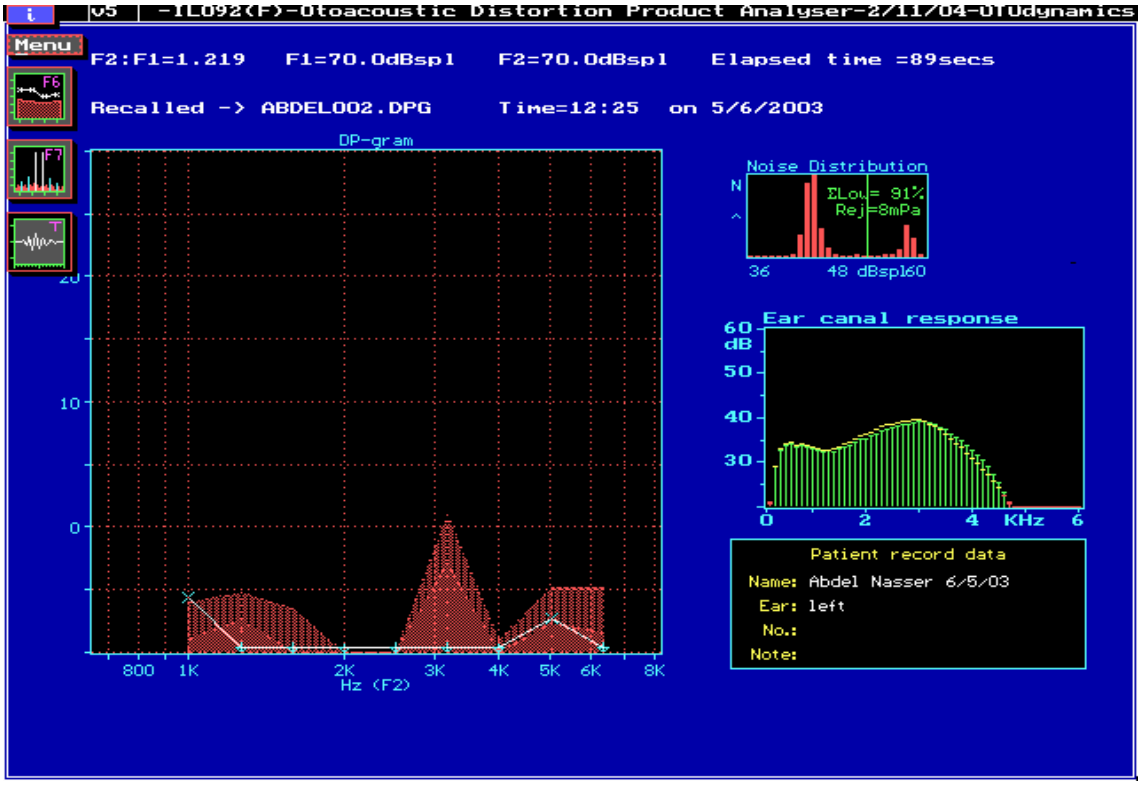


Figure (7-14): Distortion OAE (Absent response)

Management of hearing disability

Prevention (reduction of factors leading to disability)

- Genetic counseling to the parents to avoid heredofamilial hearing loss
- Improved vaccination against Rubella and CMV
- Adequate antenatal care, together with good care during labor
- Vaccination of children during the 1st year of life against measles, mumps and meningitis
- Better hygienic conditions to avoid systemic infections, e.g. Typhoid and local ear infections
- Prevention of the intake of ototoxic drugs
- Media education of the causes and impact of hearing loss

Early detection & intervention

A. Neonatal hearing screening (formal instrumental testing)

It is the process of applying to a large number of individuals certain rapid, simple measurement that will identify those individuals with high probability of hearing loss.

Importance of screening

- Hearing loss is number one birth defect
- Early & appropriate intervention (< 6 months of age) helps children develop speech, language & social skills as normal peers.
- Later identification causes 40 - 50% developmental language delay.
- Availability of acceptable technologies for effective screening.

Types of screening (candidates)

- Universal: all live births
- Targeted: high-risk population i.e. with history of one or more of the known causes of hearing loss.

Methods:

The child should receive an objective hearing-screening test before 1 month old. The test commonly used is transient OAE, ABR or both.

B. Informal testing for screening (6 months – 5.5 years)

This can be carried out by anyone who has the necessary skills. It uses simple equipment and straightforward techniques, and a quiet place free of distractions. The results provided by an informal hearing test neither accurate nor detailed, but they will tell us if a child has a profound hearing loss. Every child who is suspected of having a hearing loss should be formally tested.

Child age	Name of test
6 months – 2 years	Distraction test
2 years – 5.5 years	Co-operative test

1. Distraction test (6 months – 2 years)

The child is held on the parent's knee facing forward, head erect. One tester sits in front of the parent and child. His role is to get the child's attention by showing him a small toy, which dose not make a noise. The other tester sits behind the parent, being sure to keep out of sight of the child. His role is to present the sound stimulus.

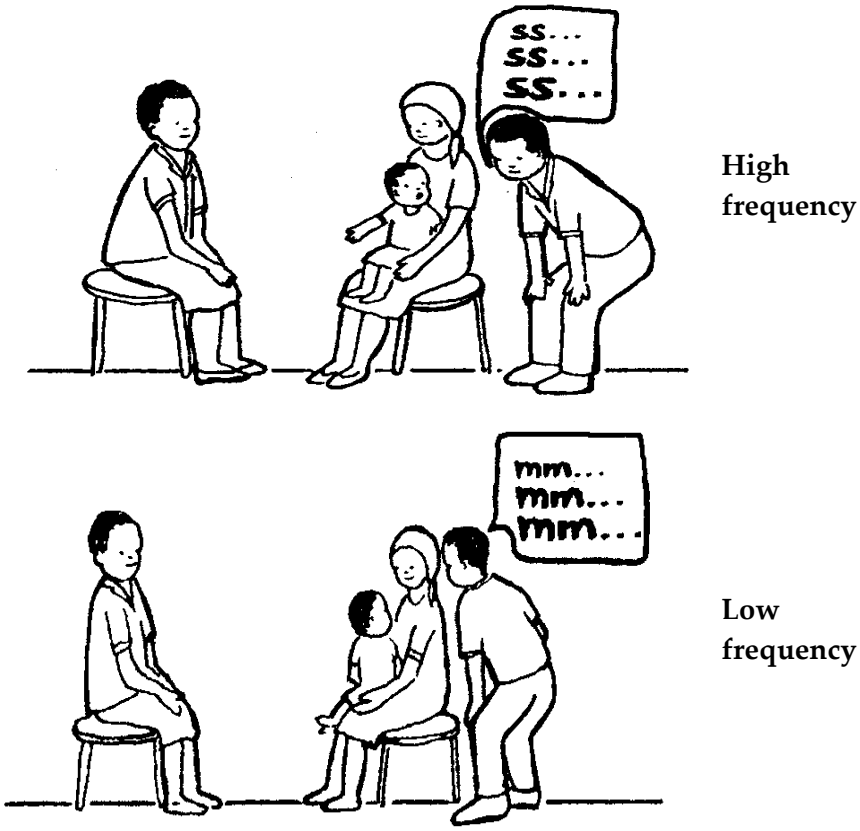


Figure (7-15): Distraction test

Both high and low frequency sounds should be used. To make a **"high frequency"** sound, the tester makes a clear **"S"** sound and repeats it rhythmically. To make a **"low frequency"** sound, the tester makes a **humming noise**. The sound should be presented as quietly as possible, if the child does not respond, refer him for formal testing.

2. Co-operative test (2 years – 5.5 years)

The aim of this test is to check that the child can hear simple instructions given very quietly, and can hear the difference between similar sounding words.

Procedure:
Four objects which are familiar to the child. If possible, use pairs of objects which have similar sounding names in the child's language, for example, cup/duck, spoon/shoe in English. Ask the child in quiet voice to point to the objects. If the child failed to point, refer for formal testing.

Figure (7-16): Co-operation test

C. Screening of school children

Candidates

- New Arrivals to school system
- Have speech & Language problems
- Suspected of having hearing loss
- Suspected of have medical condition causing HL
- Back to school after serious illness

Methods

1. Pure Tone Audiometry: Pass / Fail

Failure to respond at 20dB at 1,2,4, KHz in one or both ears

2. Immitancemetry: Pass / Fail

Negative pressure > 150-mmH_2O or f lat tympanogram

Treatment of hearing loss

- **Medical:** e.g. for otitis media.
- **Surgical:** e.g. ventilation tube for otitis media.

These would apply almost for some types of conductive hearing loss while in SNHL there is no place.

- **Aural rehabilitation**

  It is the restoration as much as possible of the function, which had been lost (hearing) i.e. to minimize consequences of hearing loss. It is considered the primary line for managing SNHL.

Steps

1. Guidance & Support to parents

Good counseling to parents about the nature of the problem and the proper way of management.

2. Amplification

- Hearing aids.
- Cochlear implantation.
- Assistive listening devices

3. Speech & Language therapy

4. Follow-up

Regular follow up once / year to re-evaluate hearing thresholds and hearing aid performance.

1. Guidance & support to parents (parents counseling)

a. Personal adjustment counseling

Deals with emotional issues related to having a child with hearing loss to accept the limitation and to assist families in dealing with such disability.

Parents & families usually go through different stages of emotional disturbances.

- Grief reaction: the parents realize that their ideal child has been lost.
- Shock and denial: are ways of protecting oneself from crises
- Anger and guilt: As the parents become convinced that something they have done in the past may be responsible for the hearing loss
- Acceptance: Finally the parents become convinced that their child's hearing loss is reality.

b. Informational counseling

To provide technical information about:

- Hearing loss: general information on child development and specific information on hearing loss and language development.
- Amplification: choice of hearing aids style, earmolds and maintenance…
- Communication mode: oral or sign language…

c. Decision-making

A family guide to achieve informed decision-making and to provide an access to professional, educational, and consumer organizations. Families should have opportunities to interact with adults and children who are hard of hearing and deaf.

2. Amplification

a. Hearing aids

Over the past few years, tremendous advancements have been made in hearing instrument technology. Hearing aids offer a superb level of sound quality (digital hearing aids) packaged in tiny instruments, practically invisible to most observers.

Types of hearing aids

• *Behind-the-Ear (BTE)* hearing aids are worn behind the ear and are connected to a plastic earmold that fits inside the outer ear. The components are held in a case behind the ear. Sound travels through the earmold into the ear. BTE aids are used by children of all ages for mild to profound hearing loss.

• *In-the-Ear (ITE)* hearing aids fit completely in the outer ear and are used for mild to severe hearing (for older children).

• *Canal aids fit into the ear canal.* A Completely-in-Canal (CIC) hearing aid is largely concealed in the ear canal and is used for mild to moderately severe hearing loss. Because of their small size, canal aids may be difficult for the user to adjust and remove. They are not typically recommended for children.

Figure (7-17): Different Styles Of hearing aids

• People with profound hearing loss now seldom use body aids. The aid is attached to a belt or a pocket and connected to the ear by a wire.

b. Cochlear implants

A cochlear implant is a device that restores hearing to people with very severe or profound deafness. Cochlear implants have been used since the late 1980s. It is a safe electronic device that is implanted beneath the skin and into the inner ear. Once the outer skin has healed, an external device is placed on the skin over the implanted device and turned on. It generates electrical stimulation of the auditory nerve directly.

Indications of cochlear implantation in children

- Severe to profound hearing loss with no benefit from hearing aids.
- Normal mentality.
- No surgical, medical and radiological contraindications.
- Good family support.

Cochlear implants allow totally deaf children to hear common sounds, spoken voice and to develop normal speech & language if they are implanted at 2 to 5 years old. However, they need extensive speech and language therapy.

Figure (7-18): Cochlear implant

c. Assistive listening devices

Great advances have been made in the evolution of "assistive listening devices". These devices help the deaf and hard-of-hearing lead a more independent life. They are used together with other amplifications. They include lights that flash when the telephone, doorbell or alarm rings; pillow or bed vibrating alarm clocks: as well as infra red or FM listening systems that help children better hearing in classrooms, live performances, or even watch television at home.

Chapter Summary

Hearing Disability

Definition

Hearing disability is best defined as a lack or reduction in the ability to hear clearly due to a problem somewhere in the hearing mechanism.

Risk factors

- Prenatal: Maternal infection (STORCH)
- Perinatal: Birth trauma , prematurity, incubation and neonatal jaundice
- Postnatal: Family history, infections and ototoxicity

Warning signs

Inconsistent response to sounds + delayed speech & language development.

Critical time for identification and screening

Early & appropriate intervention of hearing loss (< 6 months of age) helps children develop speech, language & social skills as normal peers.

Screening procedures

- At the PHC level: screening using noise makers.
- Specialized test:
  - Otoacoustic emission (OAE);
  - Auditory Brainstem Response (ABR); or
  - Both (using pass / fail criteria)

Management and rehabilitation guidelines

- Good counseling
- Amplification:
  - Hearing aids
  - Cochlear implantation
- Speech & language therapy
- Follow-up

Suggested referral places

Hearing clinics

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جمعية التنمية الصحية والبيئية

١٧ ش بيروت – مصر الجديدة – القاهرة

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