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An orienting diagnostic system in neonatal and infantile neurology

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A fast orienting diagnostic system was derived from a computerized diagnostic system in order to provide paediatricians and practitioners with a guide how to act in order to detect or exclude symptoms of pre- and perinatal brain damage endangering the normal development of the CNS. Other diseases of the nervous system are also included in the diagnostic system. The system is based on the neurological investigation and follow up of 2000 infants with suspicion of ante and perinatal CNS damage.

Complex diagnostic methods were applied in 2000 neonates and young infants in order to detect pre and perinatal brain lesions. On the basis of this clinical experience 10 diagnostic operative plans were devised. These together form a system containing the most important diagnostic methods and procedures to analyse and substantiate the existence of defects in brain function. A computerized system was developed for data acquisition and analysis to facilitate the diagnosis therapy and follow-up of neonates and young infants suffering from pre or perinatal brain defects. Another important programme of the system was the evaluation of applied therapies and rehabilitative procedures.

During the planning process of the computerized programme a more simple diagnostic system was devised for use without a computer. It serves as a guide to the physician who is not accustomed to deal with special problems in neonatal and infantile neurology. The aim of the system is to help the paediatrician or general practitioner in clinical orientation and investigation. In the presence of suspicious symptoms one should consult the diagnostic and therapeutic plans (ODPs) of the system for evaluation of the symptoms and for orientation concerning their possible correlations.

The system, called Fast Orienting Diagnostic System (FODS). consists of 9 subsystems containing guidelines, informations, suggestions and instructions how to act to obtain an early diagnosis of perinatal brain injury and how to initiate appropriate therapy and habilitation. The FODS informs the paediatrician concerning the most important questions to pose and the activities to fulfil and offers guidelines how to reach the ultimate diagnosis. Each subsystem represents in practice an operative plan in the form of a map containing questions and instructions. By following the guidelines one can proceed from a main question to

subquestions. The subsystems are called Operative Diagnostic Plans (ODP). The nine ODPs reflect the most important structural and functional trends of the maturing human nervous system, and possible pathological alterations produced by various defects such as fetal distress, neonatal hypoxia, etc. In this respect each ODP provides information about the development of all important functions of the central nervous system, and lists various possible defects in them. The ODPs also indicate the necessary steps to be taken to collect further data to the final diagnosis, and give suggestions for therapeutic and neurorehabilitative procedures. Thus, each ODP contains a plan of normal and abnormal developmental gradients in the maturing human nervous system, the symptomatology of possible abnormalities and the methods to detect and treat them if necessary.

The nine ODPs are as follows (Charts of ODPs can be found attached to the back cover).

I. Guide to the aetiology of pre- and perinatal brain injuries.

II. Guide to defects of the developing skull and brain.

III. Guide to defects of the developing spine and vertebral column.

IV. Guide to defects of the cranial nerves V, VII, IX, X, XII, and their central organization with special regard to feeding behaviour.

V. Guide to defects of the cranial nerves II, III, IV, and VI, and their central organization with special regard to early visual behaviour.

VI. Guide to defects of the cochlear portion of cranial nerve VIII and its central organization controlling early auditive behaviour.

VII. Guide to the development of sensorimotor functions and their defects in muscle tonus proportion, motor dynamics, and elementary posture.

VIII. Guide to defects in the development of sleep, alertness, orientation and general behaviour.

IX. Guide to evaluate defects in early mental activity and development of mental faculties as well as communication and social adaptation.

These 9 ODPs together represent a general outline of the structural and functional organization of the maturing nervous system with special regard to its defects, aberrations and pathological development.

Materials and Methods

General structure of ODPs

Each ODP is a map containing a number of guidelines. The map follows a definite pattern. On the left side we find a summary of the basic structures and functions to be investigated. For example in ODP No. V, from the sentence "Guide to defects of the cranial nerves II, III, IV, and their central organization with special regard to early visual behaviour" a guideline leads to the basic question whether all these functions are normal or abnormal. If any abnormality is found or suspected, the square following the words normal and abnormal must be used. In this case the square following the word abnormal should be marked.

From this square a guideline leads to the next question, "What kind of abnor-

mality?" At this point begins the instructive part of the map. From the main guideline, sub-guidelines lead to operative suggestions; these always indicate the necessity to investigate certain structures or functions, or both. In this case ODP No. V suggests to investigate the fundi, the lens and related structures, the eye movements, the reaction to light and visual orientation, visual behaviour. Along the guideline leading to these main directions we find the methods of the suggested investigations thus the investigation of light sensitivity with a lamp, investigation of the fundi by fundoscopy, investigation of the lens and related structures by fundoscopy, investigation of eye movements by activating the doll's eye symptom, fixation of human face, fixation and the following of objects, and various other procedures. The same and some other methods deal with the next problem, visual attention and behaviour. The guidelines starting from these five suggestions indicate the probability of more than 30 possible defects detectable during the diagnostic procedure. The presence of some of the defects or symptoms may indicate further diagnostic procedures using other methods. For example, abnormal visual orientation and behaviour must be analysed by application of the evoked visual potential method or by polygraphic analysis of the behavioural response to stimulation by light. As the physician follows the guidelines beginning at the left side of the map and proceeds along the arborizing sublines toward the right side, the probability of excluding symptoms and diseases arises, the differential effect increases. The circle around the possible diagnosis becomes narrower and it becomes possible to plan a strategy of the straightforward diagnostic approach by introducing the investigations suggested by the ODP. The arborizing guidelines represent the numerous contacts of the CNS correlating various functions. This mapping helps to connect one defective function to another and arrive at the full symptomatology of a disease. It must, however, be born in mind that the real correlative links between various functions as well as their central organization change in time in the maturing CNS. This is reflected for example in ODP No. V.

The real correlative links between the functions of the maturing CNS change according to the genetic programme of brain development. This is also reflected in ODP No. V.

In this map we find congenital prewired functions such as fixation of the human face, object following, turning towards light, etc. Various steps of the developing spontaneous visual behaviour, and visuomotor function will be encountered as we proceed on the map from left to right.

The maturing nervous system repeatedly reorganizes its vital control activity during development by temporally correlating certain cranial nerves to a distinct function. Such a system is represented by cranial nerves V, VII, IX and XII in order to coordinate sucking, swallowing and breathing. This functional combination offers an opportunity to analyse individual and coordinated performances of the nerves. Defects may readily be detected by the study of feeding. Later, however, new organizational systems arise to regroup the functions of various cranial nerves. For example, vocalization, speech development use the same cranial nerves, the nature of their organization and performance is, however, quite different. If the performance of the nerves is incorrect, dysarthria or other defects may develop, inhibiting normal vocalisation. This may be diagnosed well before this late period of development by analysing feeding behaviour in the first week of life together with crying, facial innervation, etc. In such cases early warning may initiate early therapeutical steps to prevent the integration of early defects in cranial nerve function into the next steps of neurological development.

Each ODP offers at least 3 choices how to begin and proceed with the diagnostical process on the map.

1. One may start with a systematic study of all functions included in the ODP. In this case one should begin to answer all questions one after the other from left to right. One must follow the guidelines proceeding step by step and perform the methods suggested to examine various problems. One is free to form any special individual strategy with the aid of the informations obtained by this procedure. One may go on slowly or may jump fast from the successful application of a method to some special conclusion. Sooner or later the diagnosis will be clear and then the therapeutic and/or neurorehabilitative process suggested by the ODP may begin. This is the analytical approach to the diagnosis.

2. A deductive way is also open. The physician may begin the diagnostic approach by hypothesising a certain diagnosis or opinion on the possible nature of the symptoms and then he may look at the right side of the map for a diagnosis which is nearest to his supposition. To find efficient proofs he has to backtrack all the ways by following the appropriate guiding lines. These are taking him provided his opinion was correct, to specific symptoms characteristic of the supposed diagnosis. The symptoms described in the ODP may or may not be present in the patient. If a similarity can be supposed between the symptoms described in the ODP and present in the patient, then further steps — suggested by the ODP — must be taken to find other symptoms in the patient. Each ODP may be considered a complex equation, which must equally be valid from the right side to the left and vice versa.

3. The third way to use an ODP is less systematic, but this may be the easiest approach. The physician who did find one or more symptoms in the newborn or young infant can look at the map and identify the same symptoms printed out. Now he can follow various guidelines which connect this symptom or symptoms with others. So he will be led to a syndrome, a complex defect which in the map points towards a full diagnosis. An example of the third possibility is that the physician finds a congenital paresis around the right corner of the mouth. He must identify this paresis as an important or an unimportant symptom. The symptom is found in ODP No. IV as a paresis of the cranial nerve VII on the right side. Now one has to follow all guidelines connecting this statement with the other statements reflecting in the functional integrity or defect of other cranial nerves.

Other symptoms mentioned in the ODP are also present, with some of them in the territory of other cranial nerves, the guidelines will indicate interrelations among the symptoms. Thus, a whole syndrome may appear suggesting the necessity of a thorough investigation. The ODP gives indications how to do it, what methods to apply and how to draw conclusions.

A short vocabulary of the ODPs

Each ODP contains notations, suggestions and instructions, questions, connecting guidelines and sub-guidelines. Each ODP is constructed from these items. The notations mark the existence of a certain structure or function and the necessity to investigate it. For example: turricephaly marks the possible existence of a characteristic shape of the skull. This may or may not occur in the given patient. A notation refers in each case to the necessity of looking for something else to observe and to describe it. The word skull in ODP No. I suggests to examine the skull. The words (notations) head and chest circumference contain a suggestion to measure them. Definite suggestions and instructions such as lumbar tap or EEG are usually instructions to undertake certain important investigations, to apply definite methods, or to perform the necessary

therapies and habilitative procedures. The guiding lines connect notations with notations, or notations with instructions. These connections mark the natural, biological links between various functions of the nervous system, mark the links between defective structures and functions forming syndromes, and mark the steps of various diagnostic procedures leading to a temporary or the final diagnosis. These are the baselines of the ODP structures. Each ODP intends to unify the main links in nervous functions and the main diagnostic steps to discover defects in them.

There are altogether 9 ODPs. In the following, summaries are given of each of them. These summarize very briefly the possible defects in a given structural entity such as the skull and the brain (morphology), or in a functional entity as the sensorimotor system of the neonate and the young infant.

ODP I. Guide to the aetiology of pre- and perinatal brain injuries

Brain development can seriously be affected during the embryological and fetal period by various endogenous and exogenous factors. All of them must be considered during consultations with young couples or during the control of pregnancy. According to these data should the labour be planned. Fetal development may be impaired by intrinsic factors such as various diseases of the mother and the father. The most important maternal risk factors are genetic anomalies, bacterial diseases, viral diseases, haemolytic diseases, hormonal disease, malnutrition, chronic heart and circulatory disturbances, renal deficiency, maternal toxicosis, placentar dysfunction, excessive bleeding, habitual abortion, and factors which lead to preterm or postterm birth. Many of these factors can be detected, prevented or treated in time depending on effective pregnant care. For example placentar circulation can be studied in the pregnant woman, biochemical and histological tests may reveal malformations of the fetal nervous system, sonography can detect the arrest of growth of the fetus.

Extrinsic factors such as chemical agents including drugs and physical agents such as irradiation may also act as risk factors. Both intrinsic and extrinsic factors may produce dysmaturity or prematurity, or damage the nervous system of the term neonate. Chronic fetal hypoxia is an extremely dangerous risk factor. Dysmature neonates are apt to develop brain injuries during their fetal life. It is sometimes more advisable to induce labour if there are definite signs of arrested fetal development than to wait and try to carry the pregnancy to term. In a well-equipped and experienced neonatal department the preterm baby can receive all possible help to assure its normal development.

The newborn can sustain brain injury also during labour. Protracted difficult labour, various impairments of the umbilical chord, aberrations in placentar adherence, hydramnios, forceps, vacuum extraction and other factors may play their part. Dysmaturity and prematurity by themselves may, however, be factors predisposing to specific complications which may lead to brain injury. IRDS in the premature may induce

peri- and intraventricular haemorrhage. Fetal distress can influence brain development. Incongruencies in maternal and newborn anatomy may produce head injuries. Neonatal asphyxia in the dysmature is more dangerous owing to lasting oxygen deficiency during fetal life. Hypoxia and intracranial haemorrhage are the most dangerous factors in the genesis of brain injury. A thorough neurologic investigation including EEG is necessary for judging the condition of the neonate, the possible prognosis, and the therapeutical and habilitative measures to be taken. In the case of convulsions, the serum Na^+, K^+, Ca^{++} and glucose levels should be estimated. All neonates who suffer perinatal injuries of any kind, or who have a history of considerable risk factors should be investigated carefully and controlled regularly later on. The next ODPs will suggest schemes of these examinations to discover all manifest and non-manifest defects of the nervous system [3, 5, 21]. A short summary is given how to investigate the neurologic condition of the neonate, including possible convulsions, and suggestions are given how to cope with them. At the right end of the map indications are found to long term follow-up if events reflected in the guide make it necessary.

ODP II. Guide to the defects of the developing skull and brain

As pathological symptoms described in ODPs II–IX may or may not be consequences reflected in ODP I, the latter can be applied to collect the necessary information about the pre- and perinatal history of the infant and to connect the pathologic symptoms with the former events.

The first procedure is a close inspection of the head. In some congenital malformations the skin may be missing in a circumscribed area. Sometimes this is accompanied with a similarly circumscribed defect in the skull and occasionally of the dura. These defects need chronic or operative treatment according to their nature. The form and size of the head is important. Its circumference and that of the chest must be compared and registered for further comparisons. The size and form of the fontanelles, the cranial sutures must be observed and measured. The possibility of macro- or microcephaly must be investigated. One may find varieties in the form of the head such as brachycephaly, scaphocephaly, turricephaly, oxycephaly (acrocephaly) or plagiocephaly. These varieties as well as microcephaly may be part of well defined syndromes as the Apert, Carpenter, Crouson, Greig, Pfeiffer, Russel, Seckel or the Smith-Lemli-Opitz syndrome. Other malformations such as encephalocele may be detected usually in the suboccipital region or at the base of the skull, protruding through a hiatus in the palate. One must carefully observe the vascularization of the head. Occasionally teleangiectasis can be detected which may indicate malformations in the brain vessels. Enriched venous arborization, large head and a peculiar

shape of the frontal curvature may be identified as a sign of hydrocephalus or hydranencephaly.

For further observation of the interior of the cranium, transillumination of the head is necessary. The procedure may reveal the existence of fluid accumulation in the skull. This may be in the subdural space (early subdural effusion with increased protein content, occasionally high ICP) or subarachnoidally (porencephalia, hygroma). Fluid (CSF) may accumulate in the ventricular space and produce hydrocephalus. It often accompanies spina bifida (ODP No. III) or develops as a consequence of perinatal haemorrhage or encephalitis. Hydrocephalus may or may not be occlusive, hypertensive or normotensive. The circulation of CSF must be studied by stain tests. The existence of these malformations or defects must be clarified by neuroradiological procedures (CT, sonography, subdurography, /22/).

The presence of haemorrhage, softening of the cerebral tissue, oedema, porencephalia, watershed infarction, periventricular cavity formation and many other morphological defects can be detected early and followed up by CT and/or sonography. The early diagnosis of these alternations in the brain can be correlated to defects in various functions, for example in sensorimotor function (ODP VIII). The neuromorphologic investigation (neuroradiology) should be correlated to neurophysiologic studies (polygraphy, EEG, evoked potentials). ODP II reflects the applicability of these methods and the necessary steps in therapy if indicated by the investigations. Early chronic subdural effusion, detected by transillumination, neuroradiology, subdural pressure recording and high protein content of the subdural fluid accumulation has to be evacuated to prevent neomembrane formation. Hypertensive hydrocephalus calls for early implantation of an appropriate ventil system, to prevent atrophic changes in the brain. Convulsions necessitate drug treatment. The results of all therapies need careful longitudinal follow-up (indicated in ODPs IV–IX) and early application of appropriate neurohabilitative programmes if necessary.

ODP III. Guide to defects of the developing spine and vertebral column

Development of the vertebral column is determined by a specific genetic programme which includes the natural human specific movements. These motor functions exercise an important influence on the final shape of the vertebral column.

The spinal cord and the spinal column may sustain lesions during birth. Haemorrhages may occur in the epidural space, in the subdural space or in the spinal cord. Fractures of vertebrae may occur during difficult labour.

The integrity of the spinal cord and the vertebral column is often impaired by various congenital malformations. Rachischisis may occur in the form of meningocele, myelo-

meningocele, myelodysplasia. These usually affect the lower part of the vertebral column, but occasionally they are observed at the cervical or upper thoracic level. Diminution in the number of the vertebrae can be detected in the sacrum. The deformity may produce sacral agenesis. The vertebrae may form abnormal contacts and develop continuities for shorter or longer extension as blocks, as in the case of the Klippel-Feil syndrome.

In addition to these local defects, deformities may develop due to other influences. This is the case if the activity of the neck muscles becomes abnormal. A pathologically increased tonus of the neck muscles on one side determines the position of the head. This in turn influences the form of the maturing spinal column. An exaggerated tonus of the neck muscles maintains either a lateralized position of the head or keeps the head in a medial position with upright chin and lower occiput. The former position is called increased asymmetric tonic reaction of the neck, the latter is known as an increased symmetric, tonic neck reaction. Both may be consequences of perinatal brain lesion. The first produces scoliotic changes in the vertebral column, the second gives rise to lordotic changes.

It is important to observe all symptoms which may lead to abnormalities in the development of the vertebral column which is the functional and structural basis of many important body postures such as of the sitting, standing and walking positions [12]. ODP III contains suggestions to the up-to-date diagnosis and treatment of meningomyelocele. Early preoperative diagnosis including early postnatal urodynamics and polygraphy may help the surgeon to consider the operative indication if selection is adopted, and in any case to compare pre- and postoperative states. Early postoperative neurorehabilitation is suggested to prevent further deformities in the structure and function of the lower extremities, the urinary bladder and the rectum. As meningomyelocele is often related to hydrocephalus, these conditions are briefly indicated in ODP III, though hydrocephalus is described in ODP II in detail.

ODP IV. Guide to defects of the cranial nerves V, VII, IX, X, XII and their central control with special regard to feeding behaviour

Reactions mediated by the Vth nerve can be studied by activization of the rooting reaction and the stimulation of the cardinal reflex points around the mouth. The function of nerve VII must carefully be studied if an eventual difference in the form of the right and left sides of the face is suspected. Laughing, crying and facial mimics and the technique of suction should be observed. While observing the activities of the baby during suction, the function of the IX and XII nerves should also be studied. Movements of the soft palate and successful swallowing reveal the integrity of nerves IX and X. Paresis

of the VIIth cranial nerve may be central or peripheral and due to some perinatal lesion. Defective sucking and swallowing may also indicate a perinatal lesion of the CNS.

Though the study of each individual cranial nerve is important, it is the investigation of feeding activity and nutritive behaviour which supplies information about the correlation of the nerves which take part in it. The trigeminal nerve acts as the trigger of the feeding reaction and coordinated response movements are mediated through the cranial nerves VII, IX, X and XII. The integrity of coordination between sucking, swallowing and breathing reveals any deficiencies in the nerves which execute these functions. This may be studied by observing breast or bottle feeding. If some defect in the feeding process is suspected, analysis of the feeding behaviour is necessary by polygraphy with simultaneous recording of the orofacial EMG, ECG, breathing, intrabuccal pressure (sucking), intrapharyngeal and intraoesophageal pressure (swallowing). The investigation occasionally detects defective sucking with good swallowing. This is usually due to an impaired coordination in consequence of a cerebral lesion at the supranuclear level over the brain stem. The absence of sucking activity can be a consequence of lesions in the brain stem. To analyse this problem, further investigations are needed. First a measurement of the threshold of the palatopharyngeal muscles to direct electric stimulation should be done. The stimulation of various trigger points in the soft palate and the upper constrictor muscle of the pharynx may reveal a high threshold or the absence of any reaction. In these cases mild or severe defects in the brain stem can be suspected. In the first case direct electrotherapy of the palatopharyngeal muscles followed by intensive feeding trainings is the method of choice. In the latter case no help is available [8, 9].

Sucking can be studied separately by polygraphy. In this case simultaneous EEG recordings should be taken together with orofacial EMG and intrabuccal electromanometry. Exteroceptive light and sound stimulation may reveal the state of consciousness of the infant as well as its orientation towards stimuli. This in turn informs about the plasticity of feeding behaviour.

Early diagnosis of neurological feeding defects is important because such defects may affect the condition of the infant and its future. For instance, the infant's condition can be affected by lung infections while his future by the danger of handicapping the development of speach. The therapies required to combat these factors will beneficially influence the development of vocalization.

ODP V. Guide to defects of the cranial nerves II, III, IV, VI, and their central organisation, with special regard to early visual behaviour

Visual behaviour in the neonate and in the young infant can be studied

by both simple and sophisticated methods. The detection of visual impairment is of paramount importance because the young infant develops contact with the mother, her face, figure and movements partly through the visual system. Defective vision impairs this contact and may lead later to a prolonged defective development of the visuomotor process.

This indicates the necessity of an intensive examination of the visual system. The neonate and the young infant usually turn towards the light. The pupillary reflex may be slow and prolonged in the newborn. Sudden stimulus with bright light activates the startling reaction. In problematic cases this may be analysed by EEG and the activated brain stem potentials.

Vision can be studied effectively by activating rotatory nystagmus, by a rotating striped disk. Fundoscopy may reveal defects in the lens and related structures as well as in the fundi where haemorrhages, retinal defects, malformations can be seen.

The function of the eye muscles is equally important. Brain injuries frequently impair the activity of one or more nerves innervating the eye muscles. Investigations of the doll's eye phenomenon, placement of the infant in upright position and other methods serve to detect defects in the innervation of eye muscles. Detailed analysis of visual behaviour can be performed by the aid of polygraphy. Visual attention and behaviour of the neonate and the young infant can be analysed during an active process such as feeding.

Visual stimulation during feeding usually arrest the feeding process because the stimulus activates orientation and the young infant cannot handle two problems simultaneously. Thus, visual attention can be studied appropriately. Prolonged stimulation produces habituation. The infant first follows the flickering light with closing and opening the eyelids, later closes the eyes and remains quiet during the stimulation. When the stimulation terminates the infant continues to suck. A detailed polygraphic study together with other neuroophthalmologic examinations offers a sound basis to establish an opinion concerning the integrity of the visual system.

In the case of visual impairment due to optic defects or improper eye muscle coordination, visual attention will fail and therapeutic and rehabilitative procedures are needed [4].

ODP VI. Guide to defects of the cochlear portion of cranial nerve VIII and its central organization controlling early auditive behaviour

The auditive system of the fetus is responsive to external noises from the 25th—26th gestational week. Hence, the neonate responds well to auditive stimuli. It is imperative to investigate auditive functions in each neonate as early as possible. Impaired auditory function endangers the contact with the environment through this important sensory channel and handicaps the development of speech.

This is the reason why a thorough audiological investigation is necessary in all neonates and young infants.

The auditive behaviour of the young infant is characteristic. The cochleopalpebral reflex must be activated to observe primitive reactions to auditive stimuli. Auditory stimulus according to its intensity produces arousal reaction in the infant; this can be verified by EEG or activated potential analysis. Stronger stimuli activate the startling reaction in its various forms. Active orientation towards auditory stimuli is reflected by intensive movements or their arrest. The effect of the auditory stimulus depends partly on its nature and partly on the condition of the infant before stimulation. If the neonate was in a quiet state of behaviour the reaction is a startling reflex. On the contrary, if the infant was restless or crying, the most probable reaction will be a sudden quietness and reduction of movements. Both responses reflect the integrity of the auditory system. The responses reveal the existence of orientative behaviour toward auditive stimuli.

It is important to distinguish between peripheral and central auditory impairment. The latter is the consequence of brain injury. Its most important representation is the failure of auditive attention. While subjective audiometric studies can define in many cases the nature of the defect, special investigations will be necessary to detect an impaired attention to auditive stimuli. Polygraphic examination of the infant in a state of activity, for example during feeding, can give information in this direction. EEG, EMG of the extremities, ECG, and sucking must be recorded simultaneously. During the feeding process programmed stimulations with various sounds can be undertaken. The nature of the changing behaviour of the infant must be analysed. The normal reaction to the first sound is orientation. If the same initial sound is repeated regularly for a longer time, inhibition occurs and the attentive orientation diminishes. This reaction may supply information on the attentive processes of the brain. Inhibited attention, habituation, is a predisposing function to a novel orientation. This can be observed if some characteristic component of the stimulus is changed. A new frequency of sound serves again as a stimulus to activate orientation.

This and other examinations such as electroencephalography evoked brain stem potential audiometry etc. are necessary to detect an impairment of the auditory system and to introduce early therapy and rehabilitative training [10, 11].

ODP VII. Guide to the development of sensorimotor functions and the defects of muscle tonus proportion, motor dynamics and elementary posture

The motor function of the neonate and the young infant consists of seemingly haphazard movements.

These, however, are only fragments of complicated stereotype elementary motor patterns. The blueprints of

elementary motor patterns can be activated by stimulation of the vestibulo-cerebello-reticular system and various proprioceptors in the extremities. Positioning the infant according to various patterns activates stereotype series of movements of the extremities and the muscles of the spinal column. Chain-reactions develop, such as elementary crawling, sitting, walking and other motor activities.

The defects of motor function may produce a great variety of symptoms. Generalized hypotonicity may develop, giving place later to hypertonicity and extensive spasticity. Latent hemiparesis may occur, masked by a relative immobility of the infant and becoming manifest later when spontaneous movements begin. Neck muscle defects may give rise to forced positions of the head, for example in the lateral direction. This in turn inhibits the maturation of rotatory functions of the trunk, orientation of the head, and the free activity of the upper arm. Hypotonicity of the neck muscles inhibits elevation of the head and handicaps the development of erect posture. In many cases some important muscle groups are hypotonic (neck and spine) while others produce hypertonicity of the extremities. Neither the myotactic reflexes nor spontaneous movements give sufficient information on the nature and place of the defect. Passive movements of the extremities are of no help. Elementary motor patterns, however, contain ample information because of the manifold movements produced by the stimulus-activated nervous system itself. The activating function of the midbrain and the brain stem, the integrating function of the spinal cord can well be analysed by activating the most important elementary patterns.

Early detection of motor defects is important for the sake of early habilitation and rehabilitation [1, 2, 7, 14].

ODP VIII. Guide to defects in the development of sleep, alertness, orientation and general behaviour

ODP VIII follows the main gradients of behavioural maturation with special regard to the development of affections, emotions and attention. This ODP reflects important steps in the normal and abnormal development of sleeping and awake behaviour spontaneous and activated attention, contact readiness, infant-mother, mother-infant relation, tendencies to preference, longing, activity and withdrawal. Developing trends in the formation of personality are intimately connected with the maturation of learning, socialisation, and communication. These human faculties are treated in ODP IX. ODPs VIII and IX contain many items of various well-known tests as the Bayley Scales of Infant Development, the Cattell Infant Intelligence Scale, the Denver Developmental Screening Test, the Gesell Developmental Schedules and others. These ODPs, however, differ in several items from all known tests.

The study of sleeping behaviour must include an analysis of the quan-

tity and quality of sleep, the duration of diurnal and nocturnal sleep. The rhythm of sleep, the nature how the infant falls asleep are all important facts. So are the behavioural patterns during the waking state. The level of spontaneous activity must be observed, symptoms such as restlessness, hypermotility, deprivation symptoms including deprivational movements, the motility patterns during alertness are all basic informations.

Signalization towards the environment is beginning at birth. Vocal signalization and signalization with facial muscles and movements should be noted and studied. These are often significant behavioural patterns to activate contact with the environment, principally with the mother. In this respect the number of signals towards the mother, the nature of the contact, the number of daily interactions are all important. When the general behaviour or mental development of the infant is investigated, a study must be dedicated to the activity of the mother and the environment towards the infant. The nature and number of contacts between the environment and the infant should be regarded as bio-social feedbacks acting on the development of social patterns in the baby's behaviour.

The level of socialization is mirrored in the development of feeding behaviour, dressing, maturation of self-reliance, manifestations of will, tolerance of frustrations, emotional maturation. All these can be studied on the manifestation of various acts reflecting the baby's adaptation to social conditions, for example urinary and rectal continence.

The behaviour of the young child among other children, in children's communities, etc., offers data to the physician and psychologist how to act when irregular behaviour is suspected and therapy is considered.

The physician must be aware of the family's relation to the maturing infant. This is especially important if the infant is sick and active aid is required from the family. The mother as the principal person in habilitation including the execution of various sensory and motor trainings, plays the main part in this aid. The physician has to know how the family reacts to the infant's disease, what aid is given to the infant and how the prescribed therapy and habilitative procedures are performed. The control of the baby's maturing behaviour with all its ever multiplying patterns needs a close and good contact between the physician or psychologist and the family.

ODP IX. Guide to defects in early mental activity and development of mental faculties, communication and social adaptation

ODP IX reflects outlines of the maturation of memory, cognition, learning and communication. The development of metacommunication, verbal communication, with the possible main abnormalities during their evolution are stressed. Emerging defects in reasoning, understanding,

interpretation of objects, situations are described in the guide. As mentioned before, the last two ODPs interpret the close correlation in the development of human behaviour, intelligence, learning and adaptive social activities.

The diagnostic approaches in this plan are concentrated on the development of attention, exploration, the maturity of interpretation and studying, the development of conceptual faculties and the utilization of comprehension in daily life. This diagnostic approach begins in early infancy. The readiness of attention and attentive reactions such as orientation, habituation and dyshabituation can be studied as well as in the later months the stability of attention in various situations. Evoked potential and polygraphic studies may inform the physician how the infant proceeds in handling more and more external stimuli simultaneously. Preference behaviour the development of attentive behaviour can be analysed and used to consider possible anomalies.

In the later stages of development, exploration activity of the infant is very informative. The exploration of objects, situations, of the environment and space are important steps in development. All these signs of growing intelligence are deeply connected to the maturation of character. Comprehension and behaviour are inseparable factors mutually influencing each other. Affective behaviour is reflected in utilization of comprehension, for example in problem-solving during play or activity transfer.

The development of conceptual behaviour can be studied by conditioning by non-verbal stimuli and by estimation of the grade of transferability. The comprehension of suggestion or instructions and the capability to express affects, needs, requirements are mile-stones in the development of mental faculties.

The maturation of attentive behaviour is deeply connected to the ability of studying. The comprehension of a task, problem-solving is based on vigility, the stability of attentive behaviour and an inborn need to face and solve situations. Short and long memory can be studied by objective methods as well as the need to stabilize the above mentioned processes with other stimuli. The maturity of verbalization is naturally important in the development of study and comprehension.

The development of special patterns (gradients) influencing comprehension and studying are significant factors of the maturing human conceptual behaviour. The development of visual-motor patterns is especially useful to the physician for discovering early aberrations. One of the first signs of comprehension, problem-solving and attention are object handling and related patterns.

In order to be able to detect anomalies of the development of the mental faculties, the physician must often rely on the family. Data on the developing comprehensiveness and intelligence of the young child must partly be based on family assumptions. The physician must carefully

compair his own observations with the informations received from the family to estimate an eventual abnormality and to indicate further investigations and treatment.

Discussion

The basic approach to the problem of early diagnosis, prognosis and early therapy as well as habilitation in cases of pre- and perinatal brain injury must be based on thorough neurological studies and close longitudinal control. Early treatment and early habilitation should only be indicated on the basis of a series of detailed neurological examinations. The treatment must concentrate on the source of the discovered neurological defect [17, 18, 20].

Adequate ante- and perinatal care has decreased the danger of brain damage during fetal or infantile age. Neonatal departments feel that nowadays the danger of brain damage is minimal. Rehabilitation centres, paedagogic institutions do not, however, share readily this kind of optimistic view.

The maturing CNS fulfils two basic functions, 1) the direction and control of daily life; 2) the direction and control of its own maturation processes. The first function may be controlled well by modern neonatology even in acute situations. This, however, does not automatically mean that it can always normalize the second function, too.

The 9 ODPs together produce a complete guide: the comprehensive Fast Orienting Diagnostic System. The aim was the illustration of the main interdependent, correlated ontogenetic gradients of the human nervous system, together with normal variations and pathologic alterations. The close interdependence of the 9 ODPs reflects the necessity to investigate the whole patient instead of suspicious symptoms. In all ODPs time was represented as an essential factor in normal and abnormal brain maturation. The initial brain injury (illustrated mainly in ODP I) produces subsequent brain lesions (illustrated in the other ODPs) which create, during time correlated brain maturation various pathological symptoms. There is much to learn about the natural history of cerebral palsy, the development of its partly uniform, partly individual symptomatology. The Fast Orienting Diagnostic System may help to accumulate more data on the correlation of the developing pathological symptoms for early diagnosis, and to objectivate the control of therapeutic and rehabilitative effects.

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