DOCUMENT RESUME

ED 231 117 EC 151 992

AUTHOR McCrae, Marcia Q.
TITLE Medical Perspectives on Brain Damage and Development. Revised.
INSTITUTION Berks County Intermediate Unit 14, Leesport, Pa.; Pennsylvania State Dept. of Education, Harrisburg.
SPONS AGENCY Office of Special Education and Rehabilitative Services (ED), Washington, DC.
PUB DATE 82
GRANT G007802653
NOTE 31p.
PUB TYPE Viewpoints (120) -- Reports - Descriptive (141)

EDRS PRICE MF01/PC02 Plus Postage.
DESCRIPTORS Etiology; Incidence; *Intervention; *Medical Evaluation; *Minimal Brain Dysfunction; *Neurological Impairments; *Physical Development

ABSTRACT

The author describes damage and normal development of the brain, as well as assessment and intervention with brain-damaged children. After a brief introduction on the complex and delicate process of brain development and a review of incidence, aspects of etiology such as genetic and postnatal causes are discussed. Brain development is examined relative to such topics as cell migration, dendrite elaboration, and myelination. Mechanisms of brain damage covered include hemorrhage, hypoxia, and delayed response. Among suggestions given for assessment, observation of how the child functions is emphasized, a list of laboratory tests is presented, and seizures are addressed. Intervention is discussed in relation to aspects such as goals, maturation, skill development, and support for parents. (MC)

Medical Perspectives on Brain Damage and Development

MARCIA Q. McCRAE, M.D.
Developmental Pediatrician

Family-Centered Resource Project
FCRP

Medical Perspectives

on Brain Damage and Development

Marcia Q. McCrae, M.D.
Developmental Pediatrician

This work was developed under a grant from the United States Department of Education No. G007802653 (Office of Special Education). The content does not reflect necessarily the position or policy of that agency, and no official endorsement of these materials should be inferred.

Published by

The Family Centered Resource Project
Gilbert M. Foley, Ed.D., Director

of

The Pennsylvania Department of Education

and

The Berks County Intermediate Unit

June 1981

Revised July 1982

CONTENTS

I. INTRODUCTION--Complexity and vulnerability

The Japanese language has two words for intelligence: one for the mind of the older child or adult and one for the mind of the infant (Kagan, 1978). Thus, the Japanese have known for eons what we are just discovering through a burst of infant research: that the mind of the young child is very different from our own. In many respects the infant brain is much more remarkable than we ever suspected. For example, at birth the infant can distinguish "Ba" from "Pa," C from C-sharp. (Kagan, 1978), and can synchronize his body and tongue movement to the rhythm of any language he hears (Condon & Sander, 1974). At nine minutes of age he can follow an object and shows a definite preference for a human face pattern (Werner et al., 1971). By three days he can recognize his mother's voice saying his name (Bower, 1977), as well as the odor of her milk. By three years he has derived all the basic rules of grammar and has multiplied his vocabulary from two words at one year to 500 at three years.

It is wise to resist mechanistic views of human life. Nonetheless, one is tempted to compare the brain to a computer, if only to discover its superior design. Our "computer" is so efficient that it contains one billion connections or synapses in one cubic centimeter. There are 10^{15} synapses altogether. It is completely portable. It can run for three hours on the energy from one peanut. It operates unceasingly for over 60 years. It can be mass produced by unskilled labor. Best of all, each model is unique.

Brain development is a very complex and delicate process which is often imperiled. The infant brain is not only more competent than we

had realized, it is in some respects more fragile. It is put together during the most mysterious time of life: pregnancy. It is immature and growing rapidly during the most dangerous time of life: birth. It takes 18 months to two years to achieve a level of brain maturity typical of most mammalian species at birth.

II. INCIDENCE

Something goes wrong in one-fourth to one-fifth of pregnancies. Of 1,311 babies conceived, 1,000 will be born alive. Of these, 12 will die in the first year (4 will have a nervous system malformation), 30 to 40 will have a significant birth defect, 40 to 60 will have a handicapping chronic medical condition, another 40 to 60 will have a perplexing neurologic syndrome which used to be called minimal brain dysfunction and is now called attention deficit disorder, and 844 will enjoy good health and normal development at 2 and only 660 will be functioning adequately at age 10 (Bergsma, et al., 1976)!

To be "exceptional" is not unusual. Roughly, one child in 35 eventually will be diagnosed as neurologically impaired (Jones, Note 3). There is a child born with cerebral palsy every hour in America (Brant & Harris, Note 1). One family in ten includes a child with a developmental disability. Major disabilities are usually multiple. Every medical advance, such as rubella vaccine, eradication of Rh disease, and improved care of the premature baby, has been counterbalanced by an increased number of handicaps resulting from trauma and environmental causes and by increased ascertainment of low-severity handicaps. The total number of handicapped children has not

diminished significantly. Contrary to popular rhetoric, we are very far from the day when prevention obviates the need for sophisticated, expensive, comprehensive and painstaking services to impaired children.

III. ETIOLOGY

When a problem in development is identified, the most logical and appropriate first question is, "What caused it?" That is the question we are usually least able to answer. The possible causes are myriad.

Genetic disorders can involve a whole chromosome, as in Down's syndrome, or a single gene, as in PKU. More than 1,000 single gene disorders have been described, but altogether they account for less than one percent of developmental problems. More common are conditions of polygenic inheritance, which require a combination of genes and perhaps other factors as well. Meningomyelocele is an example of polygenic inheritance. Roughly, 25 percent of mental retardation (60 percent of severe retardation) and 5 percent of blindness and deafness are of genetic origin (Gilles, Note 2). Genetic determinants may operate more often in males and may account for the preponderance of males among the mentally retarded.

The devastating effects of prenatal infections, including rubella, toxoplasmosis, syphilis, cytomegalic inclusion disease and herpes are well known. Chicken pox and mumps also are suspect. The damage done depends on when the infection occurs during pregnancy, but frequently presents as microcephaly with mental retardation. Sometimes the effects are delayed, showing up as subtle learning problems or progressive hearing loss. If not diagnosed early in infancy, congenital infections

are difficult to substantiate. Preliminary data suggest that urinary infections, especially late in pregnancy, may be a common cause of mild brain damage (Gilles, Note 2). Maternal diabetes is another endangering prenatal condition.

Drugs (hormones, alcohol, tobacco, seizure medications and possibly aspirin) are the best known of the environmental factors interfering with development. Although drugs account for only a small percentage of handicapped children, present testing methods are inadequate to guarantee that any drug is safe in pregnancy. Experiences like Three-Mile Island and Love Canal are beginning to teach us about the effects of radiation and toxins in the environment. An epidemic of severe cerebral palsy occurred in Japan when mercury dumped into a bay contaminated local fish. Nutrition during pregnancy appears important even to newborn traits like irritability and recovery from stress.

Birth-connected causes of brain damage include prematurity and obstetric complications. This is a large group. At least 20 to 40 percent of cerebral palsy and 10 percent of severe mental retardation are due to either hemorrhage and subsequent scarring in the premature brain or ischemia in the term baby who is deprived of oxygen at birth. Among prematures, five percent will develop cerebral palsy. The otherwise healthy "premie" baby is at very low risk, except when medical and/or social complications arise.

Breech birth is an especially dangerous situation with a high risk of spinal cord injury in addition to brain damage. In fact, only 77 percent of breech babies survive the first month.

In the last ten years, there has been a marked increase in brain

damage due to infections (encephalitis, meningitis, Reye's syndrome) and dehydration or trauma (drownings, abuse, car accidents) in early childhood.

The list of rarer causes of brain damage easily could be expanded by several hundred. Despite this, in at least 40 percent of children with brain damage, no definitive cause can be identified. Many handicaps appear to result from an accumulation of small misfortunes rather than a single unitary cause. Important processes in the body have multiple regulatory systems, often four or five. This makes the developing human so resilient that perhaps permanent severe damage occurs only when adversity persists over a long period or comes from several directions and overlapping control systems break down. Certain tissues may have specific periods of vulnerability. In general, males are less resistant to any sort of developmental insult.

It may help to think of a spectrum of severity of handicap related to cumulative adversity (Figure 1).

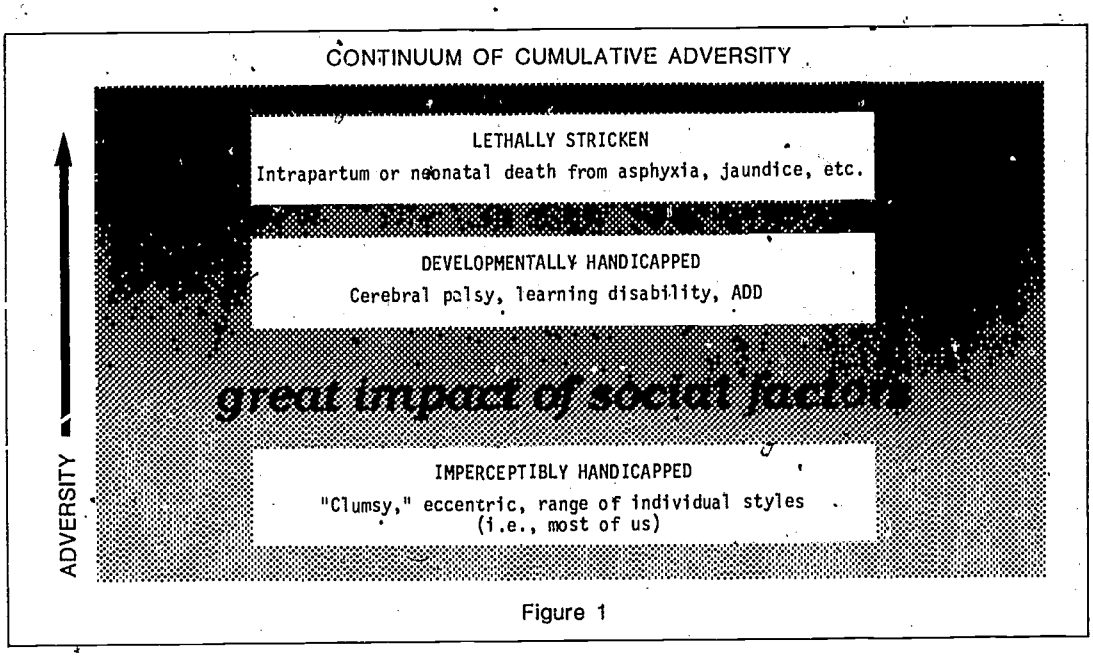

Figure 1

Very often social factors tip the child, especially the at-risk child, over the "line" from functioning to handicapped.

Let us look at examples of the cumulative adversity concept based on rubella and prematurity. Rubella infection early in pregnancy can cause low muscle tone which will prevent the baby from turning in the uterus to be born head first. The resulting breech delivery exposes the baby to increased risk of birth trauma. The low tone creates a functional handicap in the face of gravity which will become a structural handicap without treatment. Impaired vision and/or hearing, caused by the infection, will result in cognitive and emotional handicaps unless the baby is blessed with exceptionally strong and intuitive caretakers.

Consider the young teenage mother. She might be less careful about prenatal care, nutrition or drugs, and is much more likely to have a premature baby. Even full-term infants of adolescent mothers are less alert, less socially responsive and have less motor control (Thompson, Cappelman & Zeitschel, 1979). Low birth-weight babies show decreased auditory and visual orienting at first. Expert mothering can "normalize" the developmental effects of moderate prematurity (Heber & Garber, 1975). But the young mother may not be equipped to give the very special care a premature baby needs in early infancy.

A final example shows how subtle and undramatic the causes of handicap can be. Turkewicz and Birch (1971) found that babies normally lie with heads turned to the right most of the time and they speculated that this motor "habit" contributes to selective language receptivity. Infants sedated with medication commonly given during labor do not

afterward show the normal asymmetry of head-turning and theoretically could be at risk of receptive language disorder.

Given the snowball or cascade effect of mild early insults, it should not be surprising that doctors are frequently unable to pinpoint a single devastating event causing most cases of brain damage. Fortunately, we can understand and help the child without needing to know the cause.

IV. BRAIN DEVELOPMENT

To understand brain damage, we need to look at how a baby's brain develops. All the basic divisions of the adult brain are in place by six weeks of pregnancy. They appear as rather undistinguished lumps of tissue, but their destinies have been assigned.

The basic geography of the brain is shown in Figure 2.

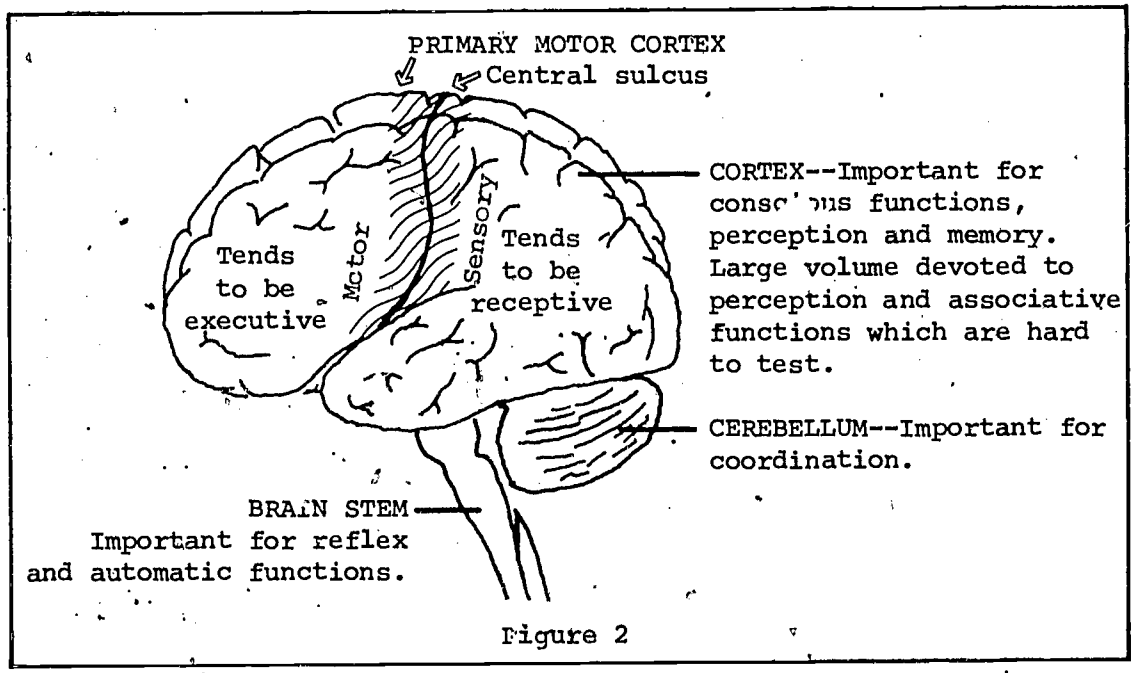

Figure 2

Each of the millions of cells in the cerebral cortex is spawned in the germinal layers lining the ventricular system as shown in Figure 3. While this germinal tissue is producing brain cells, it is a very active area containing many blood vessels. In its prime the germinal layer releases as many as 10^5 cells per day. Thousands of cells which begin to differentiate in the fetus degenerate and disappear before birth. The significance of this massive neuronal cell death is not clear. It may reduce the genetic burden in cortical development.

Each new brain cell migrates through the entire thickness of the cortex to a precise spot on its surface. These migrations occur in waves beginning at about 8 weeks and ending around 24 weeks of pregnancy. The last cells to leave end up on the outside of an orderly six layered cortex. Abnormal migration patterns have been seen in infants of mercury-poisoned mothers (Choi et al., 1979). Disruption of the layers has been discovered in a few cases of dyslexia.

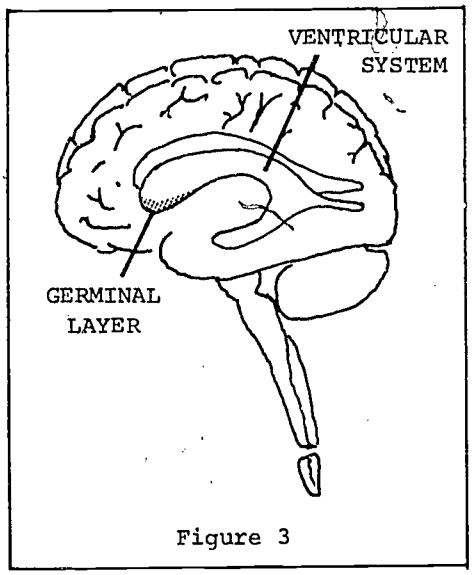

Figure 3

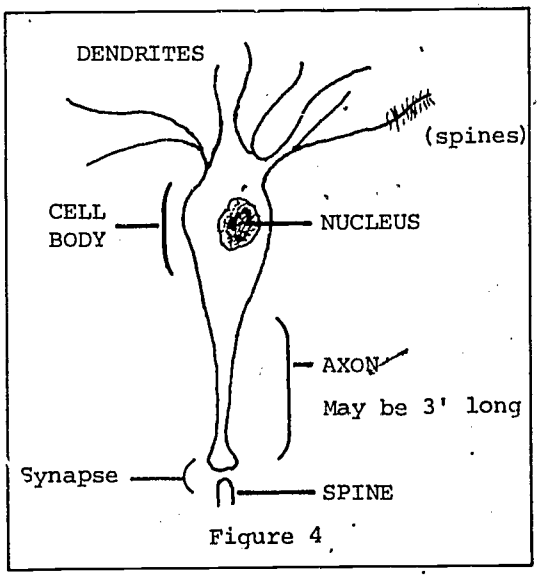

Figure 4

Once in place, each cell puts out hundreds of tiny branched processes called dendrites which hook up with other cells to bring impulses into the cell (Figure 4).

The dendrites are covered with tiny spines which eventually allow each cell to "connect" to an average of 1,000 others. Although for practical purposes no new brain cells can be made after birth, synapses continue to be established richly until age 5 and more slowly until age 18. The spines mature from stubby bumps with relatively high electrical resistance into sharp spindles which have greater ability to conduct impulses. In some conditions these spines may remain immature (Down's syndrome) or be reduced in number (PKU) (Purpura, 1974). The process by which cells become interconnected is not random. Axons find the proper target cells even in mutant mice with most of their cortical neurons in abnormal positions. Nicotine interferes with synaptogenesis, and malnutrition delays the timetable of synaptogenesis.

After the layers of the cortex are in place and as the cells are becoming larger and more elaborately connected, folds or sulci appear. The surface of the brain changes from smooth to convoluted as different areas "organize" for different functions. The right side organizes about two weeks before the left, and some asymmetry between hemispheres is normal. In general, receptive areas are located in the posterior cortex; executive areas are anterior. Evidence from cortical-evoked potentials suggests that even within areas devoted to a specific function, separate parallel neural "channels" develop handling components of sensation or movement (Hubel & Wiesel, 1979). Brain organization is influenced by sex and hormones. For example, language

tends to be more diffusely distributed in females. There is tremendous individual variation in brain organization. The pattern of folds on the brain surface is as unique as a fingerprint. Sulcation (formation of folds or sulci) occurs most actively from mid-pregnancy to two years. The brain achieves 45 percent of its adult surface area by one year, and 70 percent by age two. Some correlate this growth spurt with the appearance of capacity for internalized memory and symbolism. Although structure may not change much after infancy, there can be major shifts in functional organization during development. Animal research shows that monkeys younger than three years use different cortical areas on delayed-response learning tasks than do monkeys over three (Goldman, 1981). The possibility that different foci dominate the same function as the child ages may explain the puzzling and often dramatic improvements and also the late-emerging deficits seen in some childen.

Another important process is myelination. A fatty sheath somewhat like insulation is deposited around the axons and allows faster and more repetitive conduction of impulses. Although a marker of maturation, myelin may serve mainly to accommodate the increased length of neuronal tracks with growth (Springate, 1981). Myelin gives "white matter" its chalky appearance. Myelination also proceeds in an orderly hierarchic fashion. Peak activity for myelination is around birth but it continues significantly until age 9 and perceptibly into the 40s. An important feature of this process is that if myelination is suspended (by a life-threatening stress like meningitis or severe dehydration) it resumes with the proper step in the "timetable" rather than at the point of interruption, leaving some fibers permanently unmyelinated. Because

language and visual motor association tracts are so long, they are vulnerable over a considerable time span. Toxins may damage myelin. Vacuoles have been seen in the myelin of premature babies exposed to hexachlorophene.

Concurrent with these processes, chemical maturation is occurring. Passage of messages (impulses) from cell to cell is accomplished by chemical neurotransmitters (Figure 4), which are released only if four, sometimes five, different regulatory systems are in the proper configuration. More than two dozen neurotransmitters have been identified. The chemical briefly changes the structure of the membrane of the next cell so that energy can be used to start a new impulse, contract a muscle or release a hormone. The sensitivity of receptors to chemical transmitters varies widely from region to region on the brain and from person to person. These differences in sensitivity are probably genetic in part but many believe they are also influenced by early childhood experience. They speculate that over- or understimulation of auditory, optic or other sensory receptors may set them to be super- or subsensitive for life.

The brain and sensory organs are mutually dependent on each other for normal structural and functional development. For example, the inner ear will not develop without the brain. The brain also needs the ear. If the embryonic ear is damaged, one sees decreased number and size of cells in the auditory nuclei and cortex. Even when both brain and ear are structurally normal and hearing is acute, temporary sound deprivation causes permanently decreased auditory response threshold. Hormones also influence brain development. For example, high levels of

progesterone during pregnancy seem to lead to improved performance in elementary school.

There is a tremendous range of normal in the time schedules of the various categories of maturation. At birth there is a two-fold range of normal for brain weight. The range for development of sulci is two months, for myelination it is 14 weeks (all within a nine-month life span!). Within individuals, development proceeds at uneven rates. The vast range of normal in rate of maturation and in cortical organization complicates the task of distinguishing the limits of normal development from pathology. Variance-versus-deviance controversies continue to rage about dyslexia, extremes of temperament, attention deficits and dyspraxia. We must at least conclude that children of any age, stage or condition are entitled to look quite different from each other (and from the textbook).

V. MECHANISMS OF BRAIN DAMAGE

As a rule, pervasive or chronic insults like PKU, hypothyroidism, malnutrition, and rubella cause more profound and long-lasting deficits in the immature brain. Hypoxia and mechanical injury are two exceptions to this rule. In these cases the younger brain is relatively more resilient than the mature brain. Generally, however, the younger child responds more globally to any adversity. Several areas of development, as well as integration of functions are usually affected. Let us return to our rough timetable to see why it is so risky to be born between 28 and 34 weeks. At 28 weeks the germinal matrix is full of the tiny blood vessels needed to supply oxygen to produce all those cortical cells. It

is a watershed area for blood vessels and has continued high metabolic activity. But many cells are gone, leaving the tissue lax, unsupported, and very susceptible to hemorrhage. Any event which increases or sharply decreases the blood pressure inside the head is apt to cause rupture of these fragile vessels. Getting born is a good way to raise the blood pressure inside the head. A small hemorrhage will be contained within the germinal layer and cause few symptoms. A larger hemorrhage may rupture into the ventricular system and cause hydrocephalus, or it can erupt into the surrounding white matter and cause spasticity. Since the motor fibers to the legs pass closest to the germinal matrix, a moderate hemorrhage yields the spastic diplegia so typical of prematurity. Massive hemorrhage can rupture far enough into the white matter to cause quadriplegia and severe retardation. Half of babies born before 32 weeks have some degree of hemorrhage. It is often detectable only by X-ray, and severe problems occur only in the tiniest and sickest prematures.

After 32 to 34 weeks the germinal layer involutes and richest blood supply shifts to the midzone cortex, and changes in blood pressure tend to result in multiple small hemorrhages in the cortical surface watershed areas, resulting in diffuse damage. The same mechanisms (changes in blood pressure due to lack of oxygen or problems with acid-base balance, etc.) which hit the germinal matrix now hit the cortex because of the shift with age in vulnerable blood supply.

There are other special vulnerabilities in the young brain. Certain deeper structures are especially vulnerable to specific changes in body chemistry such as jaundice or low blood sugar. Certain

populations of cells throughout the nervous system are especially sensitive to mild lack of oxygen. Because the brain uses one-fifth of the body's oxygen and is ill-equipped to use alternative pathways, it is an organ likely to be damaged by oxygen deprivation.

Baby brains are very susceptible to external compression which obstructs blood vessels and causes hemorrhage. The compression tends to be applied over the cerebellum where hemorrhage will impair coordination. In the very tiny premature, just the tape holding an oxygen mask, or the back-pressure from prolonged bottle sucking in a supine position can cause cerebellar hemorrhage.

Baby brains respond differently to infection. They do not swell easily. They do not have a well-developed barrier between brain and blood which keeps toxins, drugs, etc. out of brain tissue. They can't mobilize the usual mature defenses to kill germs or neutralize toxins. A common cause of bladder and urinary infections in pregnant women is a germ (E. coli) which secretes a toxin (endotoxin) which enters the mother's blood stream in minute amounts. It is easily removed and does not make her sick. In the process, however, tiny amounts reach the defenseless baby brain where it can kill neurons and result in tiny islets of damage scattered throughout the cortex.

The brain often shows a delayed response to injury. Scar formation may take years. Because a certain level of maturity is needed for full expression of a generalized seizure, birth injury may not produce seizures until 4, 7 or even 50 years later. Lead ingested at 2 may affect association functions that are not measurable until 7 or 8.

Because of the complex interdependencies between parts of the

brain, remote damage can occur. Destruction of the visual cortex is followed by atrophy of visual nuclei in the midbrain (lateral geniculate bodies). An untreated seizure focus in one temporal lobe can produce a mirror image focus in the other temporal lobe.

The immature brain shows remarkable plasticity of function. Although it cannot repair damage with new cells, the brain has some capacity to reorganize. This versatility probably diminishes with age. However, the concept of differential recovery of young and old brain has recently been challenged. Both within and across cortical areas, recovery of function is inversely related to the degree of myelination. Many skills are stored bilaterally, and sometimes the other hemisphere can take over a lost function. After brain damage, it may be that surviving related neurons from either hemisphere "compete" for developing synaptic sites. Some sensory areas are surrounded by a fringe of dormant cells with the potential to serve any of several adjacent functions if the core cells are lost. (Geshwind, 1979). Researchers have ablated large portions of animal cortex attempting to produce an animal model for cerebral palsy. To their surprise, removal of almost half the cortex in fetal monkeys produced not spasticity but impulsive, very active baby monkeys with many features of what used to be called minimal cerebral dysfunction, now called attention deficit disorder. Even these symptoms disappeared when the babies were reared by experienced, exceptionally nurturing mother monkeys. (Similarly, good human mothering can reverse many of the effects of prematurity.) An identical syndrome can be produced by depleting the brain of dopamine. In either case, the symptoms abate with maturity. Some

animals and children have shown dramatic recoveries from isolation and deprivation. Critical periods for sensory stimulation impose constraints on this functional plasticity.

VI. ASSESSMENT

This marvelous potential for recovery of function makes early diagnosis and prognosis very difficult. Only 23 percent of children with cerebral palsy show definite neurologic abnormalities in the nursery (Nelson & Ellenberg, 1979). Conversely, of children who appear neurologically abnormal in the nursery only 16 percent (Volpe, 1979) will develop cerebral palsy. Of children who appear to have cerebral palsy at six months, 50 percent will have normal tone at one year. The latter are important nonetheless because many will emerge with mild retardation or speech disorders. There is currently no really dependable early screening test for developmental problems. Neonatal hearing screening combined with a social assessment like the Home Observation for Measurement of the Environment (Caldwell & Bradley, 1978) and a careful examination will probably pick up most babies who need service. By four months abnormal tone is the best indicator of disturbed development. It is important to recall that even in the face of serious risk factors, most babies escape unscathed. For example, neonatal seizures signify a 55 to 75 percent increased risk of cerebral palsy, but 70 percent of neonates with seizures develop normally.

Development is such a complicated process that it is astonishing that it comes out "right" as often as it does. It reminds us that we cannot explain brain damage by any unitary theory such as hypoglycemia.

It means that we must abandon any preconceived notions and take each child as he comes. This is the essence of developmental assessment. Formal screening tests excell at identifying abnormal children but detailed application of expert clinical judgement is best for defining an abnormality once it has been detected. Examiners and techniques vary but the common denominator is meticulous observation of how the child functions. The history is usually the most valuable part of the medical examination. The examiner can be of little use unless the parents and past records are available to him. He should have the opportunity to see the child play, eat, interact with people and objects, experience frustration, etc. A sign such as an asymmetric tonic neck reflex seen in free play rather than on a test can be ascribed functional importance rather than mere presence or absence. The physician will be interested in collating minor anomalies of skin, hair, ears, eyes, palate, etc. as indicators of risk for major problems and as clues to the timing and breadth of past insults. The traditional adult-based neurologic exam will have a low yield. Many handicaps are clarified only by developmental observation. In all areas, the elements of style and process should be evaluated in addition to skill levels. The assessment should produce a functional description of the child's temperament, strengths, neuromaturational status, learning and relating styles, and sensory competence, as well as whatever factors might interfere with optimal development. It is not so much a search for pathology as a search for ways around the pathology. Disentanglement of genetic from constitutional from environmental factors is very difficult--often impossible. A specific diagnosis and etiology are dividends of the

descriptive process. Findings should be couched in language that will elicit a helping response from all who encounter the child.

Laboratory tests commonly employed include:

1. Audiogram--cheap, painless, accurate; should be done whenever anyone thinks of it. Of all handicaps, deafness suffers the longest delay in diagnosis and greatest underestimation of its impact. Many forms of congenital deafness are now thought to be progressive and warrant repeat audiograms. Children with fluctuating hearing loss due to serous otitis may need to be tested weekly.

2. Electroencephalogram (EEG)--measures the electrical activity on he surface of the brain. It is a test of function and says nothing about structure. Since most EEGs measure only from the outer 3 mm of cortex, it is far from a complete evaluation. Currently it is useful mainly for evaluation of seizures.

3. Computerized Axial Tomography (CAT Scan)--is a serial X-ray technique from which a three-dimensional image of the brain can be reconstructed. It is a test of structure and says nothing about function. It is excellent for the detection of hydrocephalus, cysts, tumors, atrophy, etc., and has eliminated the need for many more dangerous or painful tests.

4. Electromyogram (EMG) and muscle biopsy--these are painful tests which should be restricted largely to cases of progressive weakness. They are used to diagnose muscle and lower motor neuron disease.

All information derived from tests and assessment should be interpreted

constructively in great detail to the parents. The assessment process cannot be neutral, so should be therapeutic. The assessor should have the supportive skills to help the parents with their need to know "what happened," "why me," and to help them start asserting control over "what next." The examiner's most important job is to give the parents the best possible understanding of the problem so that they can be effective advocates-allies to the child. Among his many services, he should enable the family to see what is normal in the child. He also provides a data base for understanding the child in relation to similarly disabled children. From the outset, efforts should be made to prevent secondary problems ranging from scoliosis to emotional problems to genetic recurrence. Reevaluation should be part of the initial plan. A longitudinal perspective will answer many more questions than the most elaborate single work-up.

Many handicapped children have seizures. A seizure is a spontaneous, excessive discharge from a group of abnormal neurons which triggers synchronous firing in normal neurons resulting in involuntary function. The symptoms reflect the brain sites involved and range from laughter, inattention, sensory hallucinations or complex automatisms, to jerking of the whole body in the grand mal seizure. The kinds of seizures a child has may change over time. Petit mal seizures do not occur under 3 and are usually outgrown in adolescence. Infantile spasms generally cease by 5 years but are often replaced by akinetic or generalized seizures.

It is quite common for seizures to be recognized first by the teacher or therapist. Lapses of attention are most apparent in an

instructional setting. It is easy for parents to misinterpret seizures as colic or as deliberate movements. Seizures may be aggravated by fatigue, fever, illness, emotional stress, and sometimes by specific stimuli such as light and sound.

It is important to treat seizures vigorously because:

1. They are usually unpleasant.
2. They consume energy, sometimes interfering with growth.
3. They disrupt attention, often interfering with learning.
4. Uncontrolled seizures appear to cause further brain damage and/or can lower the threshold for subsequent seizures. Rarely, a permanent aphasia may result.

Treatment of seizures is difficult and tedious; it requires optimal communication among parent, teacher and doctor. Every seizure should be described to the doctor. Most of the drugs commonly used are slow-acting, quite toxic and have a narrow therapeutic range (i.e., the therapeutic dose is close to the toxic dose). Because of the long delay between administration and full effect (14 to 21 days for phenobarbitol, 7 to 8 days for phenytoin), changes must be small and deliberate, and adjusted by feedback. A single drug should be used whenever possible to avoid drug interactions. For example, valproic acid potentiates phenobarbital and dosages should be changed in concert. The availability of blood-level monitoring is a major advance in seizure therapy. Anyone on anticonvulsants who is sedentary should take a vitamin preparation. Common side effects of anticonvulsants include sedation, nystagmus and impaired coordination. Recently, promising reports have appeared on the effects of psychotherapy for intractable seizures.

VII. INTERVENTION

Although early developmental intervention techniques are diverse, they share common goals and developmental premises. Therapy for handicapped children should strive to simulate normal developmental experience, promote a nurturing environment, prevent secondary effects, define and address associated deficits, and strengthen assets in all areas. These goals are important to the extent that they foster happiness and comfort.

Development is the product of both maturation and experience. Harlow's monkey studies began a long series appreciating the effects of environment on behavior. It appears that brain structure is affected by the complexity of early experience (Bennett et al., 1964). Tactile stimulation is an important channel in many species. Kittens, for example, who are stroked twice a day open their eyes and emerge from the nest sooner, develop deeper coat color and accelerated maturation of EEG pattern (Meier, 1961). It is becoming clear that many structural handicaps originate as functional deprivation of normal early experience. The contractures and deformities of untreated cerebral palsy begin as the effects of low flexor tone on infant posture.

Development is a hierarchic and sequential process. Complex skills are synthesized from previously mastered components. Herein lies a rationale for early intervention. Compensations must be prevented from evolving into abnormal motor or perceptual habits from which distorted complex skills are built. Certain progressions are evident in every domain of development. These sequences proceed from head to toe (cephalocaudal), from proximal to distal (decentering), from mass

responses to discrete responses (dissociation), from lateralized to midline responses, from reflexive to voluntary control. The development of selective or focused inhibition is a useful behavioral marker of maturation. Axial motor development, beginning with acquisition of extension against gravity, balanced by flexion, augmented by righting reactions, allowing rotation, and, finally, dissociation of movement, can be followed visibly and palpably down the trunk. Difficulty at any level can be understood only in the context of the total motor status, including the reflex repertoire. Speech requires the highest degree of dissociation and can be refined only when head and trunk control are well established. Perceptual hierarchies exist also. For example, the infant can discriminate face from nonface patterns by 2 weeks, orthodox face from scrambled face by 4 to 5 months, old from young faces and male from female faces by 5 to 6 months. By 7 months he can recognize individual faces but by criteria different from those used by adults.

Skills are learned through practice of age-specific operations. It follows that they can be taught. Skills like walking, talking and feeding evolve so smoothly in the normal child that they appear to be automatic but are actually self-taught largely by trial and error. One can observe the progression of age-appropriate strategies in the elaboration of almost any concept. For example, the 5-month infant's sensorimotor knowledge of size is evident as he opens his hand wider to reach for larger objects. This occurs at least a year before he can sort objects by size and several years before he can verbally mark objects by size. Over and over we see important concepts expressed as motor "ideas" before they can be used symbolically. Fully actuated

development depends on collaboration of specific forms of environmental stimulation at various levels of maturity. Obviously, it is essential that the infant have some opportunity to use his body as a spatial laboratory.

The various areas of development are intimately and irretrievably interrelated. Problems in one area have repercussions across the spectrum. For example, excessive extensor tone, which causes the baby to thrust away from the mother when picked up, will be interpreted as an aversive reaction and will undermine attachment. Normal exclusive attachment facilitates development of object permanence. Attachment depends on a degree of motor competency, and attachment failure imperils language and cognition as well as adult emotional capacity.

The child is more active in initiating developmental change than was previously realized. Many studies have shown that the child learns better when he has active control of the stimuli. He is equally active in emotional development. Maternal behaviors are in place but are elicited by the child. Separation is initiated by the child and either tolerated or resisted by the mother. The child sets and changes the tempo of parent-infant play, and the parent follows. A major advantage of breast feeding is that the child starts and stops sucking sequences and often terminates the feeding, enjoying a reciprocity which seems to be important for language development with differences seen in sentence completion at age 15.

When intervening in development, we must remember how little we know about the process and how crudely we approximate it. Modern child-rearing itself is a radical social experiment. We know little about

making stimulation sufficiently contingent and active for the handicapped child. The average toddler at play changes position 50 times in two minutes! The most devoted physical therapist could not provide an equivalent experience to a spastic or low-tone child. Very young children teaching themselves a skill practice it just to the point of earliest mastery. They then appear to lose interest, but in several months are apt to incorporate it into a more complex achievement. In therapy how do we tell when a skill has been practiced to that exquisite point where it is just mastered but is not so automatized that it cannot be an ingredient for a new, more complex skill? Children do not engage in true repetition as they practice. Rather they are constantly revising internal reference criteria as they "problem-solve" toward a motor goal. Observing them reminds us of the magnitude of the task we undertake in trying to simulate the normal developmental sequence. Furthermore, we must know when and how some interventions can interfere with attachment.

Intervention should never displace the parent. If we do our jobs very, very well we can be important allies to the family, but the burden for management ultimately devolves to the parents. Any program is only as good as its ability to support parents in their job of helping the child to develop a healthy self-concept and to perceive the world as a place of pleasure and a place upon which he can act. It is the parents' success at these two tasks, more than any kind or amount of therapy or education, that determines the outcome for a handicapped child.

Reference Notes

1. Brant, R. L. & Harris, M. I. Prevention of embryonic fetal and perinatal disease (DHEW Publication No. NIH 76-853). Bethesda, MD: U.S. Government Printing Office, 1975.

2. Gilles, R. Developmental neuropathology. Lecture given at Children's Hospital Medical Center, Boston, 1978.

3. Jones, M. Diagnosis and general assessment in cerebral palsy. Paper presented at 27th annual meeting, American Academy for Cerebral Palsy, Washington, D.C., December, 1973.

References

Bennett, E. L. et al. Chemical and anatomical plasticity of the brain. Science, 1964, 146, 610-619.

Bergsma, D. et al. Developmental disabilities: psychological and social implications. New York: Alan Arliss, Inc., 1976.

Bower, T. G. R. A primer of infant development. San Francisco: W. H. Freeman & Company, 1977.

Caldwell, B. P. & Bradley, R. Home observation for measurement of the environment. Little Rock, Arkansas: University of Arkansas at Little Rock, 1978.

Choi, G. H. et al. Abnormal neuronal migration, deranged cerebral cortical organization, and diffuse white matter astrocytosis of human fetal brain: A major effect of methol mercury poisoning in utero. Journal of Neural Pathology and Experimental Neurology, 1979, 37, 719.

Condon, W. S., & Sander, L. Neonate movement is synchronized with adult speech. Science, 1974, 183, 99-101.

Geshwind, M. Specializations of the human brain. Scientific American, 1979, 241, 180-199.

Goldman, P. S. An alternative to developmental plasticity: heterology of CNS structures in infants and adults. In D. G. Stein, J. F. Rosen & N. Butters (Eds.), Plasticity and the recovery of function in the central nervous system. London: Academic Press, 1974.

Heber, R., & Garber, H. The Milwaukee project: a study of the use of family intervention to prevent cultural-familial retardation. In Friedlander, B. Z., Sterritt, G. M., and Kirk, G. (Eds.), Exceptional Infant (Vol. 3). New York: Brunner/Mazel, Inc., 1975.

Hubel, D. H., & Wiesel, T. N. Brain mechanisms of vision. Scientific American, 1979, 241, 150-163.

Kagan, J. The growth of the child. New York: W. Norton and Company, 1978.

Meier, G. W. Infantile handling and development in siamese kittens. Journal of Comparative and Physiological Psychology, 1961, 54, 284.

Nelson, K. B., & Ellenberg, J. H. Neonatal signs as predictors of cerebral palsy. Pediatrics, 1979, 64, 225-232.

Purpura, D. P. (Ed.). Methodological approaches to the study of brain maturation and its abnormalities. Baltimore: University Park Press, 1974.

Springate, J. E. The neuroanatomic basis of early motor development: a review. Developmental and Behavioral Pediatrics, 1981, 2, 146-150.

Thompson, R. J., Cappelman, M. W., & Zeitschel, K. A. Neonatal behavior of infants of adolescent mothers. Developmental Medicine and Child Neurology, August 1979, 21, 474-482.

Turkewitz, G., & Birch, H. G. Neurobehavioral organization of the human newborn. In J. Hellmuth (Ed.), Exceptional infant: studies in abnormalities (Vol. 2). New York: Brunner Mazel, 1971.

Volpe, J. J. Neonatal neurologic examination. Pediatrics, 1979, 64, 541-548.

Werner, E. D. et al. The Children of Kaui. Honolulu: University of Hawaii Press, 1971.