THE PEDIATRICIAN AND THE DEVELOPMENTALLY DELAYED CHILD

A Clinical Textbook on Mental Retardation

Monographs in Developmental Pediatrics, Volume 2

by
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and
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THE PEDIATRICIAN AND THE DEVELOPMENTALLY DELAYED CHILD is the second volume in the new **Monographs in Developmental Pediatrics** Series. Other volumes in this series include:

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The Embryology of Behavior
Infant Behavior Scales
Developmental Assessment of the Preschool Child
An Epidemiological Study of Developmental Disabilities

Marion Katz

People, even more than things, have to be restored, renewed, revived, reclaimed, and redeemed, and redeemed, and redeemed . . . Never throw out anybody.

Sam Levenson

Preface

Thus they are often obliged to do things which they have imperfectly learned, to say things which they imperfectly understand, and to devote themselves to work for which they are unprepared by long apprenticeship.
de Tocqueville

Medicine in general and the traditional pediatric training program in particular have demonstrated a preponderant interest in the diagnosis and treatment of acute organic illness. Although the importance of developmental and behavioral phenomena to the understanding of the whole patient has always received more than adequate lip service, the tremendous personal and social impact of chronic disease has failed to produce a truly valetudinarian medicine. At the beginning of the Christian era, the Methodist School did make brief inroads against the descendants of Hippocrates, the Dogmatic theorists and the Empirical skeptics. Adopting a peculiar middle position, the Methodists attempted to reform the attitude of physicians toward the treatment of the patient with chronic disease. In the long history of medicine, this movement was an abortive effort with no lasting effects.

Technologic and public health advances have today dramatically reduced the burden of organic childhood disease. Early diagnosis, parent education, more specific therapies, and regionalization of services have all allowed for striking reductions in the mortality and morbidity rates associated with classic pediatric disorders. The concern of society has now begun to focus more on the problems of chronic handicapping conditions, but scientific medicine has remained isolated and aloof from all but a minimal response to this trend.

For this and other reasons, it has become increasingly fashionable to decry the "medical model" in the evaluation and treatment of developmental disabilities. Usually the model that is under attack is a parody of the organ-localization pathology of late nineteenth-century medicine—an arrogant, quantitative mentality. It is unfortunate that traces of this orientation have survived in medical education and practice up to the present time, but it is an egregious error of historical interpretation for patients, physicians, or other professionals to identify this anomalous distortion with medicine proper. Medicine is an art whose proper goal is the use (and, if necessary, abuse) of science to help those suffer-

ing from bodily or mental disorders. Its proper study is, in Osler's phrase, the patient who happens to have a disease, and not the disease the patient happens to have. Numbers (such as IQ scores) and organ localization (such as brain cell heterotopia) are not the end, but rather some of the starting points of a comprehensive understanding of the handicapped child. The reduction of a human being to a number or a disease label cannot be justified as a shorthand description in the absence of a working knowledge of the processes of abnormal development.

A clarification of the reference to an "abuse of science" would appear to be in order. Scientific theories frequently fall victim to a vacillating faddishness (witness the history of the corpuscular and wave theories of light, the emotional and organic theories of autism, the genetic and environmental theories of intelligence, etc.); ivory tower researchers demonstrate amazing about-faces in their public pronouncements. As the pendulum swings to and fro, irrefutable (that is, statistically significant, but usually quite refutable) data are mustered to support first one hypothesis and then its antithesis. It is one of the more honorable tasks of humanistic medicine to steer a middle course between such extremes—keeping up to date on the latest clinical and laboratory advances, not subjecting patients to risky or difficult innovations based on tenuous hypotheses, and never being the cause of false hope or false despair.

The present text focuses on the clinical signs and symptoms of developmental delay in young children and attempts to aid the practicing pediatrician in the early diagnosis and habilitation of mental retardation. Serviceable guidelines to the understanding of developmental deviations seem to be a practical goal despite the absence of a unified theory of normal child development. In other words, a completely uniform and generally accepted "developmental physiology" is not a prerequisite to the understanding of developmental pathology. Stress upon early language milestones and such developmental phenomena as dissociation and coupling should make the book of practical value to all professionals working with young children. The concluding chapter analyzes sterilization as a debated issue which may signal clashes between social and ethical goals.

The material presented is part of a core curriculum in Developmental Disabilities in the Department of Pediatrics of The Johns Hopkins University School of Medicine. Medical

students, pediatric housestaff, and postdoctoral fellows in Developmental Pediatrics receive formal instruction at their respective levels of training along with intensive experience in the workings of the interdisciplinary team. Interest in historical and ethical issues is a pervasive characteristic of the program.

A necessary part of the education of a pediatrician is a thorough grounding in the roles various nonmedical disciplines play in the diagnosis and treatment of handicapped children. A refusal to enter into meaningful dialogue with such paramedical professionals can only contribute to further handicap the child. The boundaries of traditional medicine need to be considerably extended so that the following fictional (but all too true) account may evolve from the typical to the obsolete:

> I could tell stories. An outsider wouldn't believe the number of conflicting opinions the different doctors gave us and the backbiting judgments they made of each other, but we did. We believed them all, the good and the bad. And disbelieved as well (we had no choice) and had no choice but to search for others, like wandering supplicants.
>
> "It's organic."
>
> "It's functional."
>
> "It's largely organic with functional complications now."
>
> "He is not deaf but may not be able to hear."
>
> "At least he's alive."
>
> "The prognosis is good."
>
> "For what?"
>
> "The prognosis is bad."
>
> "It would not be possible to offer a prognosis at this time."
>
> Not one of them ever had the candor, the courage, the common sense, the character to say:
>
> "Jesus—I really don't know."
>
> It began with:
>
> "You're making too much of it."
>
> And moved to:
>
> "He will never speak."
>
> "He probably will not surpass a mental age of five, if he attains that. His coordination and muscular control will never be good.
>
> It will require tremendous patience."
>
> We hate them all, the ones who were wrong and the ones who were right. After awhile, that made no difference. The cause didn't matter. The prognosis was absolute.[1]

It is too easy a defense against such charges to reply that the experts involved were themselves misdiagnosed and mislabeled.

[1] Reprinted by permission from *Something Happened* by Joseph Heller, 1975, pp. 497-498. Ballantine Books, New York.

The more appropriate response is the establishment of high professional standards. The subspecialty area of developmental pediatrics has the major goal of fostering professional education and interdisciplinary communication in the field of developmental disabilities. It is hoped that the present text, although aimed predominantly at the practicing pediatrician, will fulfill this dual role by clarifying to the nonphysician the contribution that medicine can make to the multidisciplinary team approach to mental retardation.

This work was supported in part by Project 917, Maternal and Child Health Service. Figures and line drawings were prepared under the supervision of Mr. Leon Schlossberg. Instruments were photographed by Mr. William Diehl. The photograph of the young child with Down syndrome on the dedication page is taken from a photographic exhibit of handicapped children entitled "A Different Beauty" by Marion Katz. Zuhair Kareem, Chief of Biomedical Photography at The Johns Hopkins Medical Institutions, prepared all the prints for publication. Barbara Kelner, MLS, hunted up many of the references. The handwritten manuscript was reduced to a legible typescript by Karen A. Torbit. To all those who contributed their support (moral and otherwise) the authors wish to extend their gratitude.

Pasquale J. Accardo
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HISTORY

> You may alter the place to which you are going; but you cannot alter the place from which you have come.
>
> Chesterton

The recognition of childhood as a distinct phase of human development is a recent phenomenon—so recent that its social, legal, and educational implications have yet to be fully worked out. Some historians optimistically view the evolution of childhood as a rational progression toward more humane conditions (Figure 1). But, in the same way that the history of medicine can be said to be the history of humanity itself, the history of mental retardation is but a special aspect of the history of childhood. The major roles of deviant persons (Table 1) which Wolfensberger (1972) used to stereotype past attitudes toward retardation can easily be extended to serve as a descriptive typology of the place children occupy in different cultures. These deviancy roles, whether applied to normal children or to the retarded, evidence relatively little linear progress. Rather, they seem to be cyclic with different roles being dominant at different time periods in the same culture. Thus when the deviant individual was viewed as a social menace during the "genetic scare" (1890–1925), laws for the sterilization and euthanasia of the retarded were passed. More refined genetic theory discredited the pseudoscientific aspects of this movement, but utilitarian social planners are currently attempting to revive the menace role by employing more palatable euphemisms. In a similar vein, Haffter (1968) documents some of the modern survivals of the ancient concept of the "changeling"; attitudes toward children and the retarded seem to be grounded in the more primitive levels of the psyche. The real history of mental retardation would seem to lie in the most recent historical subspeciality—psychohistory, the history of childhood and family life. Attempts to understand the complex evolution of social attitudes toward developmental incompetence should have significant impact on the future of mental retardation.

Ancient and medieval medicine rarely attempted to distinguish between retardation (a permanent, hereditary condition) and lunacy (in which there was some hope of a cure or temporary remission) because there existed no effective therapies for either

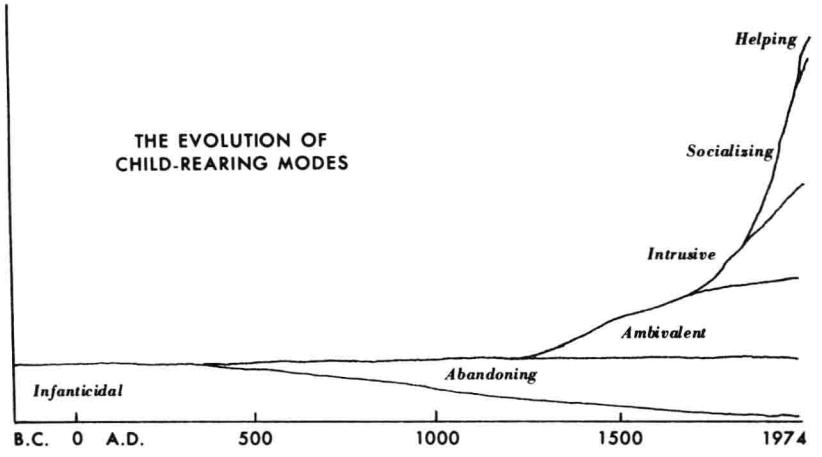

Figure 1. The progressive evolution of parent-child interactional modes. From DeMause (1974) by permission.

condition. However, ancient and medieval law evolved careful distinctions between natural fools (idiots) and lunatics because the disposition of such persons' property by the crown turned on the existence of lucid intervals (Neaman, 1975). Thus in 1534, the jurist Sir Anthony Fitz-Herbert declared:

> And he who shall be said to be a sot [i.e., simpleton] and idiot from his birth, is such a person who cannot account or number twenty pence, nor can tell who was his father or mother, nor how old he is, etc., so as it may appear that he hath no understanding of reason what shall be for his profit nor what for his loss. But if he hath such understanding, that he know and understand his letters, and do read by teaching or information of another man, then it seemeth he is not a sot nor a natural idiot (Doll, 1972, pp. 49–50).

Table 1. Major historic roles of deviant persons

Subhuman Organism
Menace
Unspeakable Object of Dread
Object of Pity
Holy Innocent
Diseased Organism
Object of Ridicule
Eternal Child

It is important to note that the legal focus has until recently been exclusively centered on the protection of property rights with little or no consideration for the person of the mentally handicapped. The literacy criterion in the passage cited above would not have much relevance for the nonpropertied commoner class. Nevertheless, even this early, one can observe a consideration of social and adaptive behaviors in addition to purely cognitive performance. The legal distinction was clear to philosophers such as John Locke (1632–1704), who wrote in his book titled *An Essay Concerning Human Understanding* (1690):

> Herein seems to lie the difference between idiots and madmen, that madmen put wrong ideas together, and so make wrong propositions, but argue and reason right from them, but idiots make very few or no propositions, and reasons scarce at all (1959, Vol. 1, p. 210).

The above examples notwithstanding, throughout most of recorded history disorders of the mind have been confused—St. Dymphna of Gheel (feastday May 15) became the patroness of both the emotionally disturbed and the mentally retarded.

Psychiatry was one of the first of the modern branches of medicine to enter its scientific phase late in the eighteenth century; it recognized the retarded as a distinct type of mental disorder and promptly discarded them as incurable. Within less than a century neurology would establish itself as a specialty and follow psychiatry in abandoning the retarded except for that minority demonstrating interesting neuropathology.

But there was another stream of medical interest that went its way quietly and succeeded in revolutionizing the area of behavioral science. Jean Marc Gaspard Itard (1775–1838) received a grant from the French government to educate Victor, a "wild child" found roaming in the woods of Aveyron. Itard spent 5 years applying a program of graded stimulus discrimination in an attempt to civilize his pupil. This "physiologic method" drew its theoretical inspiration from the erratic speculations of Rousseau's *Emile*, but it was much more firmly grounded in Itard's practical experience with deaf children and his creative responses to his daily interactions with Victor. At the end of the experiment, Itard considered that he had failed; by norm-referenced standards Victor could neither talk nor function independently in society—he was still not normal, still retarded. The French government felt otherwise; compared to his functional level before his sojourn with Itard, Victor had made dramatic progress. The government

implicitly used a criterion-referenced standard in honoring the physician's achievement. Dr. Itard went on to become one of the founders of otolaryngology; in his youthful idealism he had fathered special education. One of Itard's physician pupils, Edouard Seguin (1812–1880), further refined the physiologic method, elaborating the sensorimotor drills and exercises and inventing some of the hardware still in use today (Figure 2). After his emigration to the United States in 1848, Seguin became one of the architects of the institutional movement in this country. His classic text on the principles of educating the retarded, *Idiocy and Its Treatment by the Physiological Method* (1866; reprinted in 1971), contains some of the earliest descriptions of Down syndrome and the fetal alcohol syndrome. Maria Montessori (1870–1952) was the first woman to receive a medical degree in Italy. The Montessori method extended the techniques Itard and Seguin had devised for the education of the retarded to the acceleration of the development of the normal child.

In 1877, William Ireland (1832–1909) published *On Idiocy and Imbecility,* the first medical textbook of mental retardation. His classification of idiocy was the first attempt at a comprehensive medical nosology (Table 2). Genetous did not refer to genetics because that branch of science did not yet exist in 1877; rather it was what today would be called idiopathic. It was Ireland's largest category; his observations supported a study of Langdon Down's which described the frequent occurrence of high-arched palate in this group. Absolute microcephaly was an adult head circumference below 17 inches; it was noted to be much more common in males. Ireland recognized that there was no constant relationship between intelligence and head size; he compared the head

Table 2. Classification of idiocy

Genetous
Microcephalic
Eclampsic
Epileptic
Hydrocephalic
Paralytic
Cretinism
Traumatic
Inflammatory
Idiocy by Deprivation