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Foreword

Just as we had begun to think that researchers were getting a hold on the causes of mental retardation and other developmental disabilities, two more threats to the fetus have burst forth on the scene. They are crack and pediatric AIDS. These two problems are, of course, related. Over 25% of AIDS cases in the United States are drug addicts, their partners and their babies.
The New York State Office of Mental Retardation and Developmental
Disabilities, or OMRDD, as we are known in the shorthand of New York State bureaucracy, has an active interest in research into the causes and prevention and treatment of mental retardation and developmental disabilities. We believe that research, training and education undergird a strong policy commitment to people with developmental disabilities. We also hold that ongoing contact and interaction between service and research strengthens this policy commitment. There is a dialectic and we must all understand that there is this dialectic. That science informed service and service informs research and education and together, they bring about a more coherent and reasonable public policy. At the OMRDD we have worked to this end by supporting the Institute for Basic Research and other activities such as our Medical and Dental Fellows Program.

The subject of this conference, Brain and Behavior in Pediatric HIV Infection, is of great import to New York State and to the OMRDD because if infants and children present themselves to us with a developmental disability, regardless of their disease, we must know what services they need and how best to provide them.

New York is struggling with the policy issue of who should care for children with HIV infection. This is a complicated problem because there are good reasons for children being served by the child welfare system, but sometimes in hospitals and sometimes by the OMRDD or the voluntary agencies it supervises. No matter how this question gets resolved, however, the Office of Mental Retardation and Developmental Disabilities is going to promote research and try to facilitate information exchange in this area.

Policy is affected and shaped by research. We need to know, for instance, how many children will present with HIV infection over the coming years? How many will be developmentally disabled? What kinds of services will they require? Is there the potential to treat the infection, to curtail the developmental disability and/or the infection itself?

This conference is the first time information on these aspects of this problem has been shared. We are going to focus on what we know and also what we don't know. We are waiting for science and public policy to catch one another, and as the poet, Ferlinghetti, said: 'To catch up with one another and share a common destiny'. I thank you and wish you well in your endeavors.

Howard B. Gold
Director,
Policy and Planning,
Preface

At the beginning of the epidemic, the AIDS virus was perceived as the agent selectively affecting the immune system. At present, we know that the human immunodeficiency virus (HIV), the causative agent of AIDS, is not only lymphotropic, but also neurotropic. Devastating nervous system involvement, although insidious and in the early stage of the disease, not always obvious clinically, occurs in almost all children infected with HIV. Most of the funding now supporting the research on pediatric AIDS comes from federal sources. The burden of providing health care for these children, on the other hand, is faced by local state and city governments. New York State is one of the epicenters of the AIDS epidemic in the United States. The vast majority of cases are concentrated in New York City. The New York State Institute for Basic Research in Developmental Disabilities (IBR) decided to organize this meeting because pediatric AIDS is a serious emerging problem and the aspects of the problem related to brain and behavior fall clearly within our mission. The Institute, or IBR, has a fourfold mission: research into the causes, diagnosis, prevention and treatment of developmental disabilities; clinical service; education and the technology transfer.

Pediatric AIDS is certain to have a major impact on the field of developmental disabilities, although it is not yet clear how various health and human service delivery systems will divide the responsibilities. A new group of infants and children with HIV infection will enter the health care delivery system and they will require long-term care and commitments. This group will differ from the persons now served. They carry a virus that produces a fatal disease and all infected are not expected to live longer than a few years from the time of diagnosis. They present special medical challenges that are not present in persons who are developmentally disabled, but not HIV-infected. The disease is progressive and there is, at this time, no effective treatment. There is also a declining neurological status frequently associated with mental retardation. Also, there is unfounded public fear of contagious spread of HIV from these infected children. The staff has to be specially trained with new levels of competence and responsibility. Most of these children are from minority groups and many of them are or will be orphaned. There has been a rather bleak perspective of finding adoptive
homes for these children, although this may be changing. Improved medical
care and well-designed social interventions will extend their lives; however, it
will not necessarily improve or even preserve the existing quality of life.
HIV infection is not only a challenge to the health care system, it also
represents a major challenge to basic research investigators in the field of
neuroscience. It bridges neuroscience with immunology. The damage
inflicted by HIV to the nervous system is astounding and represents a new
phenomenon in human neurovirology.

Henry M. Wisniewski, MD, PhD
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Introduction

The purpose of this volume is to share with other scientists the papers
that were presented at the Conference on 'Brain and Behavior in Pediatric
HIV Infection'. The meeting was organized by the New York State Institute
for Basic Research in Developmental Disabilities and held July 24-25, 1989,
at the Halloran House in New York City. A total of 115 scientists participated.
We would like to express our gratitude to them for taking time from
their busy schedules and scientific work to participate in the conference.
We would also like to thank the agencies that supported the conference.
These include the New York State Office of Mental Retardation and Developmental
Disabilities, the United States Bureau of Maternal and Child
Health, and the National Institute of Child Health and Human Development.
At the beginning of the AIDS epidemic, AIDS was perceived as a disease
of homosexual men and, to lesser extent, individuals who received transfusions
of HIV-contaminated blood or blood products.
At present, the disease is spreading most rapidly among intravenous
drug abusers, including women. A small population of women become
infected through heterosexual transmission, from HIV-infected sexual
partners. The increased number of HIV-infected women has been followed
by increasing numbers of congenitally infected infants and children. The
number of infected children is growing so rapidly that it is projected that in
the next 4 years, HIV will become the leading infectious cause of mental
retardation in children.
Central nervous system involvement in pediatric HIV infection is early
and severe. Neurologic manifestations are often the first signs of AIDS, and
immunodeficiency in an affected child or infant may not yet be evident. HIV
infection in infants and children produces not only a fatal immune deficiency,
but also chronic and devastating central nervous system damage. This damage may occur even in the absence of opportunistic central nervous system infections. In many cases, the brain is so affected by the virus that the outcome is a profoundly damaged, blind, and incontinent child.

The elucidation of clinicopathological correlations in pediatric AIDS has been difficult. First, the complex 'natural history' of the disease has to be projected on a background of a rapidly developing brain. The second reason that these clinicopathological correlations have been difficult is that the number of pediatric AIDS cases in each of the AIDS centers is relatively small, so that only small groups of cases have been analyzed thus far. Improved communication among researchers is crucial. Only by combining findings from various centers, can a large and significant population be assessed and future trends be defined. We will then be able to prepare ourselves for the explosion of pediatric HIV cases that is certain to come in the near future.

Nervous system complications of pediatric AIDS were the major focus of this meeting. For the purpose of these proceedings, the presentations are organized as follows. First, two introductory papers present the epidemiology of pediatric HIV infection and show the extent and scope of the problem. These are followed by four papers presenting the clinical aspects of this infection. The neurological and neurodevelopmental aspects are then presented, followed by reviews of treatment and case management. In the last part, general pathology and neuropathology is presented. Development of new diagnostic tools and assessment of brain involvement is crucial for the development of effective therapy. The necessity of this work is put in a capsular form by Dr. Richard T. Johnson (1988), an eminent neurovirologist.

'Rational therapy may depend on our clarification of the pathogenesis of the neurological diseases. If immune enhancers clear opportunistic infections or antiviral drugs inhibit systemic replication of HIV without altering neural infection, we may simply prolong the lives of a growing population of mute, blind, demented, paralyzed and incontinent young patients.'

R.T. Johnson, 1988

This perspective is grim, but quite real as the number of pediatric AIDS patients is growing dramatically. Though the disease is ultimately fatal, with improved therapy, the patient may live longer. We hope that the papers presented in this volume will be found thought
provoking and stimulating. That they will improve the exchange of information, help to clarify many issues, and, finally, benefit the care and treatment of the 'innocent' victims of this dreaded disease.

Piotr B. Kozlowski, MD, PhD

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